LIMB LENGTHENING and RECONSTRUCTION SURGERY

edited by
S. Robert Rozbruch, M.D.
Svetlana Ilizarov, M.D.

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informa
healthcare

New York London

Informa Healthcare USA, Inc.
270 Madison Avenue
New York, NY 10016

Printed in the United States of America on acid-free paper
10 9 8 7 6 5 4 3 2 1

International Standard Book Number-10: 0-8493-4051-9 (Hardcover)
International Standard Book Number-13: 978-0-8493-4051-2 (Hardcover)

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This book is dedicated to . . . .
My wife, Yonina, and my children, Jason and Libby, who have given to me so much love, joy, encouragement, and support.
The memory of my father, Max Rozbruch (1918–2001), who was a humble, courageous, generous, industrious, and great man.
My mother, Frieda Rozbruch, whose pride and encouragement have been a continual source of strength and inspiration.

S. Robert Rozbruch, MD

The memory of my father, Gavriil A. Ilizarov, MD (1921–1992) whose work I have admired all my life and whose life is a continual inspiration.
My mother, whose love, intellect, patience, advice, and continuous support became a cornerstone of everyone's achievement in our family. Her "behind the scenes" help allowed my father to use his extraordinary abilities to overcome obstacles and to earn worldwide recognition and success.
My dear son, Gabe—my strength and my heart.

Svetlana Ilizarov, MD

Foreword

It is my privilege to be invited to write a foreword for this book. The greatest pride for a teacher comes when a student accomplishes great achievements with what the teacher taught or inspired. Dr. Robert Rozbruch was my student and fellow in 1999, and it has given me tremendous satisfaction and pride to witness and participate in this achievement. This book is the first comprehensive text on limb lengthening and reconstruction surgery since the publication of Professor Gavriil Abramovich Ilizarov's textbook in 1992. It represents the next generation of Ilizarov's teachings, not only because one of its coeditors is Gavriil Abramovich's daughter, Svetlana, but also because it is coauthored by so many first- and second-generation students of the late professor. I know that Professor Ilizarov would be proud to witness the revolution he launched when he first traveled to the West in 1981. I am sure he would be impressed to read through this book.

Founded on the influence of the Russian and Italian schools, this book, *Limb Lengthening and Reconstruction Surgery*, presents the orthopedic surgeon with the first English language didactic reference text that teaches the basic science, clinical science, and surgical methodology of the broad applications of compression–distraction histogenesis. Unlike related texts in this field, this book is not based on any single manufacturer's device. Dr. Rozbruch has recruited many internationally renowned authors to write 49 chapters on the state of the art of this relatively new, rapidly evolving field of surgical practice and research study. Although the international contribution in this field is enormous, this to me is a testimony to the central role that the introduction of the Ilizarov method to the United States in 1987 has played in the proliferation and development of compression–distraction surgery. Finally, this book provides a glimpse into the exciting future of limb lengthening and reconstruction surgery.

Dror Paley, MD, FRCSC
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Preface

I hope that this book will complement the great classic works in limb lengthening and reconstruction surgery. I have been influenced and educated by Ilizarov's *Transosseous Osteosynthesis* (Springer Verlag, 1992) and Paley's *Principles of Deformity Correction* (Springer Verlag, 2002). The former is predominantly an atlas of cases demonstrating the author's pioneering work and the creation of a new discipline. The latter focuses on principles and planning for lengthening and deformity correction using the center of rotation and angulation method.

Interestingly, these classics were not compiled as traditional medical textbooks. My goal was to organize and edit a traditional textbook with many expert contributors that would cover the gamut of limb lengthening and reconstruction surgery. Authors were asked to share their practical approaches to a clinical problem. There are also chapters on basic science and technology. This book is a comprehensive approach to the many state-of-the-art applications of limb lengthening and reconstruction surgery. The book is divided into sections on principles, acute trauma, post-traumatic reconstruction, foot and ankle, knee, hip, pediatric, tumor, upper extremity, stature lengthening, technology, and postoperative care. There are 49 well-defined chapters presented by many leaders in the field from around the globe. In addition, the introduction, The Ilizarov Method: History and Scope, was written by my coeditor and partner, Dr. Svetlana Ilizarov.

This book is a necessity for the practitioner of limb-lengthening reconstruction and deformity correction. The chapters are organized to include an explanation of the clinical problem, a practical approach to each subject, and a concise review of the literature. Tables outlining the authors' classification of each problem subtype and their decision-making with regard to treatment are included. A review of the authors' surgical approach including technical tips, complication management, and clinical case examples are covered in each chapter. Although the authors worked within guidelines, their individuality will show with some heterogeneity of chapter format. Common abbreviations such as leg length discrepancy, anteroposterior, and intramedullary are used liberally throughout the book.

Many people have asked me, ''How did you get into limb lengthening?'' I would like to acknowledge the mentorship, help, and support of many people by briefly telling the story of my orthopedic limb lengthening journey. My mother's dream to become a physician was cut short by the Nazis and her deportation from Romania in 1941. She and my older brother, Jacob Rozbruch, MD, who is a successful New York orthopedic surgeon, were major early role models.

During my residency at the Hospital for Special Surgery, I was turned on to trauma and the AO/Association for the Study of Internal Fixation school. I did an Arbeitsgemeinschaft für Osteosynthesefragen (AO) fellowship with Professor Reinhold Ganz in Bern, Switzerland, and this was my first real exposure to osteotomy for deformity correction. After returning to New York, I practiced general orthopedics, with a special interest in trauma, for three years. The most complex post-traumatic problems were bone loss and large deformity, and we did not have a good solution for these. Dr. David Helfet, chief of the Orthopaedic Trauma Service and Dr. Russell Warren, then surgeon-in-chief at the Hospital for Special Surgery encouraged me to take a sabbatical and do a second fellowship to learn the Ilizarov method and to develop a niche at the Hospital for Special Surgery. The Baltimore fellowship with Dr. Dror Paley and Dr. John Herzenberg was life changing. I was amazed by their clinical work. They are great teachers and great men, and I am indebted to them for teaching me the tools of this amazing trade.

When I returned to New York, Dr. Warren gave me the task and opportunity to develop the limb lengthening program at the Hospital for Special Surgery. Although many of the faculty saw this as an exciting new discipline and supported this, others felt that it was only

a technique to be used very sparingly and were resistant to this strange ''new'' field. I met Arkady Blyakher who had come to the United States from Russia and was trained by and had worked with G. A. Ilizarov in Kurgan. He and I worked together at the Hospital for Special Surgery for five years on clinical and academic pursuits, and he exposed me to many aspects of the classic Ilizarov method. At one time, he was a great friend and colleague. He introduced me to Svetlana Ilizarov, MD, at one of our holiday parties. Despite her great experience with and knowledge of her father's method, she was surprisingly not involved in this discipline. With the support of John Reynolds, the chief executive officer and president of the Hospital for Special Surgery, and Greg Lutz, MD, the chief of physiatry at Hospital for Special Surgery, I recruited her and we formed the Institute for Limb Lengthening and Reconstruction at the Hospital for Special Surgery.

I attended a Taylor Spatial Frame course soon after my fellowship and was exposed to the genius of J. Charles Taylor, MD and his frame/computer program. Immediately, I was seduced by this lengthening and deformity correction machine and began using it feverishly. I met an orthopedic resident named Austin Fragomen, MD, while I was lecturing at a Taylor Spatial Frame meeting. His desire to do a fellowship with me led to the creation of the Limb Lengthening Fellowship at the Hospital for Special Surgery. Austin was my first fellow and student. He taught me how to teach, and his excellence confirmed to me that we had great program. I recruited him to join the faculty and with the leadership, support, and vision of Dr. Thomas Sculco, the current surgeon-in-chief at the Hospital for Special Surgery, we formed the Limb Lengthening and Deformity Service at the Hospital for Special Surgery. We have developed a successful collaborative relationship with the other Hospital for Special Surgery services including foot and ankle, trauma, sports, arthroplasty, hand, and tumor. Joint conferences have been very productive for the sharing of ideas and patients. With more experience, we have come to appreciate the many indications for the Ilizarov method. I thank these people and my many colleagues at the Hospital for Special Surgery who support our work.

A man named Geoff Greenwood from Informa Healthcare publishing house contacted me two years ago and proposed this book project. He helped us organize the job at hand. I also want to thank all the contributors to this book who are giants in this field. Thanks to my coeditor and partner, Dr. Svetlana Ilizarov, the editorial team at Informa Healthcare led by Dana Bigelow, and Nancy Collins, the project editor at The Egerton Group.

While I never met Professor Gavriil A. Ilizarov, I owe a great deal of gratitude to him. His pioneering genius paved the way for our clinical and academic work. Many of the contributors to this book were taught by him first hand. The rest of us learned about the method second hand. It is my hope that this book will stimulate young doctors to explore this subspecialty of orthopedic surgery. Ilizarov's work inspired the formation of the Association for the Study and Advancement of the Method of Ilizarov, which is an international academic society with national branches. The North American branch of the Association for the Study and Advancement of the Method of Ilizarov is one of the subspecialty societies of the American Academy of Orthopaedic Surgeons and is called the Limb Lengthening and Reconstruction Society (www.LLRS.org). I encourage people who are interested in this field to attend and join this society. It is a great place to learn, share ideas, and meet fellow ''Ilizarovians.''

S. Robert Rozbruch, MD

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Part I INTRODUCTION

1 The Ilizarov Method: History and Scope

Svetlana Ilizarov
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Imagination is more important than knowledge.

Albert Einstein (1879–1955)

INTRODUCTION

Daring, hard work, struggle, and perseverance—these are the words that come first to mind when I sit down to describe my father, Professor Gavriil A. Ilizarov, and his life's work. I find the task daunting. How does a daughter adequately describe the life of a man she knew so well personally, yet whose life was so full of complicated and extraordinary events that it lay before her like a novel of fiction?

I witnessed some of these events and heard about others from my parents and their friends. Growing up in a family of physicians, I was introduced very early to their work and my father's method in particular. I remember attending conferences, listening to my father's presentations, and observing him in the operating room (OR) since I was in high school. Later, I was fortunate to work with my father (1985–1991), assisting in surgeries, discussing treatment plans for patients, traveling with him abroad, and helping in preparation of lectures and its technical support.

My father had realized early in his life that new medical concepts sometimes emerge very slowly, and only when a critical mass of evidence has gathered with enough strength to push back the doubters so that a new paradigm arises. He once wrote:

> It is a matter of fact that medicine has a conservative nature. And it is not surprising that its main principle is to first do no harm. But unfortunately, it often leads to the testing of soundness and endurance of not just an invention but the inventor, himself, even in the presence of convincing evidence confirming its expediency and efficiency. However, facts do not lie. I continued to accumulate them patiently. I had to prove my case by convincing and persistent facts. And I stood my ground.

Work is a necessary and vital part of life. In the life of Gavriil A. Ilizarov, work was synonymous with life.

HOW IT ALL STARTED

His interest in medicine began in his childhood after a local doctor treated him for a bad case of food poisoning. He was better in no time after taking the medications and was impressed by the doctor and his ''miraculous cure.'' He then became full of youthful confidence in the power of medicine and decided he would pursue a career in medicine himself.

Due to family circumstances, my father entered elementary school at the age of eleven; however, he quickly caught up with his peer group. Amazingly, his spelling and grammar skills were superb and later in life he used it frequently while editing articles and PhD theses of his colleagues and apprentices.

Figure 1 Gavriil Ilizarov—first years in medical school in Crimea.

At the age of 18, in 1939, he entered medical school in Simferopol, Crimea (Fig. 1). However, when the Soviet Union was invaded by Germany in June 1941, the government wanted to protect its medical resources; so the entire school, professors and students, had to be evacuated to Kazakhstan. My father remembered these war years as extremely difficult, with constant hunger and fatigue from lack of sleep. To earn his living, he worked night shifts at the rail station unloading freight trains; this to not only live but also to continue with his studies.

After graduating in 1944, the new physician, Dr. Gavriil Ilizarov, was posted as a family practitioner to Dolgovka in the Kurgan province of western Siberia. During Tsarist Russia, this remote area was used as a place of exile, apparently from the time of Peter the Great.

My father was the only physician covering this large remote region, the size of a small European country; he had to perform a wide variety of surgeries that addressed many different medical conditions. There were no older colleagues from whom to seek advice and no telephones. He had only his medical books to consult and his courage and reliance on a resourceful gift with which he was abundantly blessed: an ability to analyze a life/medical problem from a multifaceted perspective, arrive at a practical solution, and act on that solution. During his first medical practice in Dolgovka, Ilizarov understood the faith with which people came to him for medical care, including treatment of conditions that were beyond his formal training. There was no other physician to whom they could turn for help. Medicine and surgery in Siberia, during that era, sometimes had to rely on the courage of both the physician and the patient. Ilizarov had taken a six-month course after medical school in military field surgery. This, combined with confidence in his ability to educate and train himself further, enabled a self-taught Ilizarov to master various surgical techniques that were needed by his patients of this remote region. I recall him recounting a case of a young woman in whom he surgically repaired a longstanding congenital upper lip defect and another case in which he restored a boy's nose, which had been bitten and torn from his face by a dog. He would study and practice these plastic surgery procedures by cutting out a pattern on paper and on fabrics until he had the requisite understanding and skill. Ilizarov also, in his practice on occasion, used hypnosis techniques self-taught during his youth. He applied them to help patients with some conditions such as stuttering. This remote Siberian region, so

unsympathetic to survival with its harsh winters and bereft of most of life's conveniences and civilized comforts, was blessed with the best of physicians to care for its population.

Some of the patients whom my father treated were returning soldiers from the frontlines of battles of World War II who had sustained fractures from various causes of war. From these and the bone injuries of the local inhabitants, my father became impressed over time that healing and treatment of bone fractures was very prolonged, and caused great suffering and delays in returning to work. Treatment modalities of the time were limited to casts and skeletal traction. Internal fixation was not widely used due to a lack of antibiotics. He believed and persevered in his thoughts that there must be a way to improve bone healing. He came to see this problem as so important that he made a determination to devote his career to orthopedics. At first, he studied all the available Russian books on orthopedics. Later, he took courses, learning from the best surgeons in the country. He acquired all available training in classic orthopedics of that time, which emphasized physical examination and an understanding of biomechanics and function. However, he noticed that there was not much progress in this area of medicine especially in regard to duration of treatment, improvements of patient mobility, and function.

By nature, my father was a keen observer and always paid attention to details. He was a dreamer as well, but not an idle dreamer. ''I like to play with (my) imagination,'' he often said. One day, while traveling in a horse carriage to see a patient, he became curious about the shaft-bow ''Duga'' harness construction connecting the horse to the carriage through shafts and how it provided a fixed and stable distance between them as well as maneuverability. The Duga reminded him of the metal bows from skeletal traction. He thought of an idea to immobilize bone, using construction of bows connected by parallel rods. My father first applied this idea as a ''treatment'' on a broken broomstick; however, he discovered that the broom fragments were not adequately secured but remained mobile. Fortunately, this was simply an idea that did not prove successful and he did not abandon his belief or his search for an improved treatment for slow healing fractures and nonunions.

THE SEARCH CONTINUES

In 1950, Ilizarov moved to the city of Kurgan to accept a position within a general surgery department, which also included trauma patients, at the Kurgan Regional Hospital. Part of his assignment was to travel to rural areas in single-engine biplane constructed of plywood. Because my mother was also a physician, a radiologist, each traveled by this precarious mode to provide medical care where and when it was needed.

In Kurgan, Ilizarov continued to explore ways to achieve improved results in bone healing and immobilization of fractures. Toward this end, while he was studying mechanics, he had an insight into the stability that an external ring with crossed wires would bring to a fracture setting. It was an insight born of his interdisciplinary knowledge and the principles of basic mechanics he was learning. He then asked a local metal worker to fashion these specially designed parts for a new orthopedic device, and, as before, did his preliminary testing on a broken broomstick. This time, Ilizarov became convinced that his invention would provide secure immobilization. He also continued to test different types of materials for strength and elasticity in order to improve the original design.

He sent an application for a certificate of invention and was shortly invited to Moscow to demonstrate his external fixator (Figs. 2 and 3). A story told by my father years later was that upon arrival in Moscow, he was offered an ''arrangement'' by a highly-placed medical official who suggested that his name might be added after they worked together on improving the device and he could help him in the patenting process as well. My father refused this offer of assistance. His application for authorship of the device was accepted in June 1952 and was finally approved in 1954 (Fig. 4) (1).

The first patient to be treated with the new external fixation device was a worker with a nonunion from the factory where metal parts for the device were made. After successful treatment, he started to apply it for fractures and for immobilization after knee arthrodesis for arthritis secondary to tuberculosis. The results surpassed his expectations—successful healing in a significantly reduced time compared to traditional methods.

When he presented his data at the conferences, other physicians were skeptical because his results of treatment were dramatically shorter. That being said, devices similar to the Ilizarov

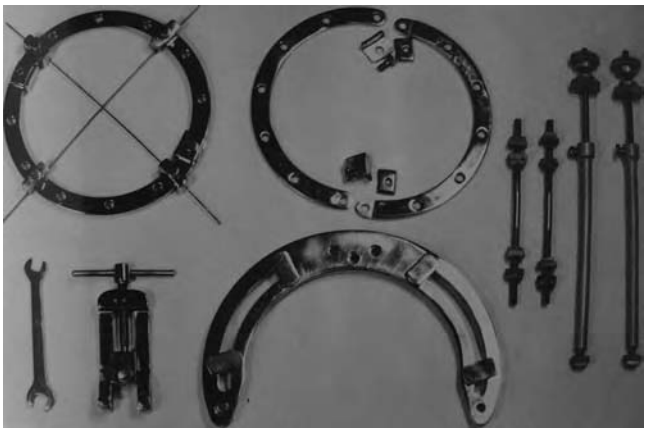

Figure 2 Parts of the Ilizarov apparatus designed in 1951.

apparatus started to emerge, in spite of the skepticism, using bow, circular, and rectangular shaped fixators. One of them was a half-ring (bow) external fixator by a physician (O.N. Gudushauri) from the Central Institute of Traumatology and Orthopaedics (CITO) in 1955. Later, the Gudushauri device was given a green light and became the "official" external device used in Moscow for many years. At that time, due to politics, Moscow "coryphaei" did not want to give a priority to a "nonyielding" province doctor from Siberia.

Dr. Mstislav V. Volkov, who became head of the CITO in Moscow in 1961, was one of the prominent figures who actively worked against official acceptance of the Ilizarov device and method. In 1968, Dr. Volkov, together with Dr. Oganesyan, had patented a similar device to the one presented by Ilizarov at the conference on Tuberculosis of Bones and Joints in Tomsk in 1963 (2,3). Volkov used his prestige and position to promote the application of his device in the Soviet Union and at international conferences. However, the fortunes of Dr. Volkov would dramatically shift during perestroika years later, when he was removed in 1985 from his position

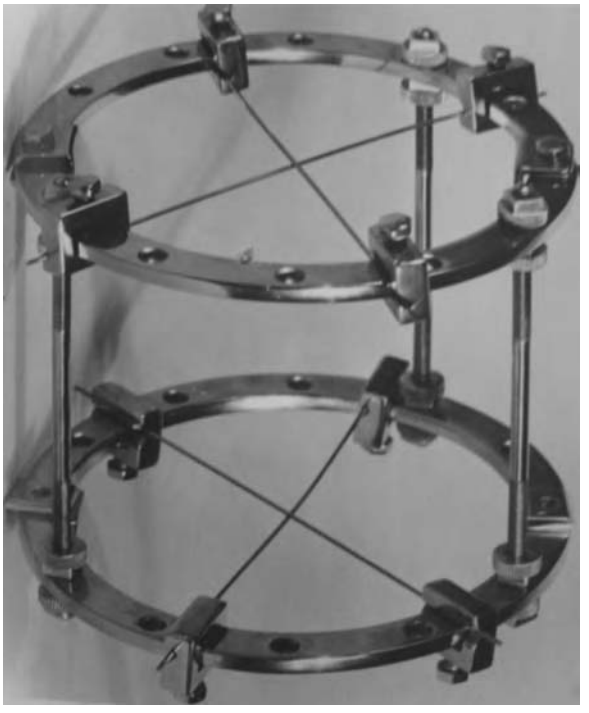

Figure 3 One of the first designs of the Ilizarov apparatus.

Figure 4 Doctor Gavriil Ilizarov after receiving authorship certificate for his apparatus.

as director of CITO. Among the reasons for his dismissal was oppression toward the acceptance and distribution of Dr. Gavriil A. Ilizarov's external fixator device and his method (4).

TREATING A CELEBRITY CAN MAKE ALL THE DIFFERENCE

Despite the negative experiences in Moscow and his first attempts to introduce his new device to the medical society of the Soviet Union, Dr. Ilizarov and his innovative treatment started to gain recognition locally. In 1955, Ilizarov became Chief of the Department of Trauma and Orthopaedics in the veterans' hospital in Kurgan.

In 1965, he was awarded the title of ''Honored Physician of Russian Federation'' for his achievements in medicine. He also became known among patients by a less official, but nonetheless gratifying, title as the ''magician from Kurgan'' and had a long waiting list of patients. This ''title'' had little to do with Ilizarov's lifelong love of learning and showing off magic tricks, which became his hobby throughout his life and provided him great joy and relaxation.

In 1966, owing to his growing local and regional reputation, with support from the local government, he was given a clinical and research laboratory affiliated with Sverdlovsk Research Institute of Trauma and Orthopaedics in a newly built city hospital.

In 1965, the Ministry of Health decided to send a group of physicians to Kurgan to observe more closely the surgery and progress of the patient according to the Ilizarov method. The response was very positive but may have threatened the officially sanctioned devices by CITO.

In 1968, my father operated on Valery Brumel (1942–2003), a famous Russian athlete renowned in international sports, a high jumper who had set six world records during the 1960s and was a silver (1960) and gold (1964) Olympic medalist. His highest jump of 2.28 m (7 ft. 5.75 in.), made in 1963, was unbeaten for seven years. Tragically, for Brumel, in 1965, at 23 years of age, he suffered a compound fracture of the distal tibia in a motorcycle accident. The accident and his injuries and treatment received wide notice in the Soviet press. Brumel spent approximately three years in various clinics and underwent about 20 unsuccessful surgeries. Ultimately, he developed an infected nonunion as well as a significant leg length discrepancy. During his last admission to the CITO, when he started to lose hope of recovery, his girlfriend heard about a doctor in Kurgan from a physician who preferred to remain anonymous. He was later revealed to be Dr. Vladimir Golyakhovski, then a young successful surgeon in Moscow. Golyakhovski was among the first group of specialists sent to Kurgan, in 1965, to observe and evaluate Ilizarov's work. CITO had assigned Dr. Golyakhovski the task of looking out for aspects of the Ilizarov work that might not be sound. After one month in Kurgan, Golyakhovski had returned to Moscow, instead wholly enthusiastic about my father's advances in treatment. Volkov, however, was apathetic about supporting the device within CITO.

Brumel immediately called Kurgan to seek a consultation. On being reassured that his infected nonunion could be healed and his leg length discrepancy of 3.5 cm could be corrected,

Figure 5 Doctor Gavriil Ilizarov with Valery Brumel during his treatment in Kurgan.

he left the Moscow clinic and flew to Kurgan. The surgery was successful (Fig. 5). Brumel resumed his athletic training sessions in 1968. These events brought substantial recognition and attention of high officials as well as fame to Dr. Gavriil Ilizarov within the Soviet Union. Brumel's recovery and his surgeon's name were also published in international articles. One of these articles appeared in the U.S. medical press, the Journal of Podiatry, in 1973 and was titled ''Kurgan: Revolution in Orthopaedics.''

However, the news impact in the Soviet Union of Brumel's recovery was fast and very positive for Ilizarov. In 1969, the Ilizarov laboratory had become affiliated with the Leningrad Institute of Traumatology and Orthopaedics. The media exposure of a famed athlete recovering against such overwhelming odds and prior treatment failures helped to gather support for financing a new orthopedic institute in Kurgan in 1971. The Research Institute of Experimental and Clinical Orthopaedics and Traumatology (KNIIEKOT) would be committed to research and applications of the Ilizarov method. Later, in 1982, an additional building in the shape of a snowflake was added to expand the clinical, research, and diagnostic services of the institute. This snowflake design was an original concept by my father to prevent the spread of infection, by placing patient wards furthest away from the administrative center of the building and providing them with independent entrances. In case of a serious infection breaking out in one ward, that block could be effectively isolated without interruption of the work of the hospital.

Valery Brumel and my father developed a close friendship over the years and he became the best man at Brumel's wedding. Brumel later wrote an autobiographically based novel about the physician who saved his leg and athletic career entitled ''Do not Betray Yourself.'' A movie was later produced in 1973, in Russia, based on the novel, and was retitled ''Every day of Dr. Kalinnikova.'' The character, Dr. Kalinnikov, was meant to be the role of Dr. Gavriil Ilizarov; however, for political reasons, at the last moment, in order to save the production of

the movie, this role was changed to be a female physician and was filled by an actress. She did an excellent job playing her character. However, for me and for the people who knew my father, it was very strange to watch this movie. Just imagine that in the story of Dr. Zhivago, the gender changes and the title role is given to Barbara Streisand instead.

DEVICE IS NOT A METHOD

There has been a long history, beginning in the 19th century, of various physicians and engineers inventing different external fixation devices. Ilizarov's distinction was that he came up, independently, with an idea of rings and crossed wires for improved fixation of bone fragments. He also developed the most universal design of the external fixator, allowing its application to any bone, providing and maintaining stable fixation during treatment as well as applying forces to the bone fragments in different directions and planes, preserving functions of muscles and joints and allowing weight bearing from the first days of treatment. He took his invention to the next level and succeeded in developing a well-established method supported by biomechanics and basic science.

For those interested in history, I recommend a most complete and accurate description of the historical development of the devices, concepts, and research in the area of bone healing in ''The Historical Background of Transosseous Osteosynthesis'' chapter written by my father in his book in 1992 (5).

Despite all the successful results, my father had to struggle for his work to be recognized in the medical community. In December of 1956, at the scientific orthopedic conference in Sverdlovsk (renamed Yekaterinsburg), when he reported his results showing that he achieved bone healing after knee arthrodesis in 16 to 18 days while allowing patients to ambulate after 10 days during treatment without crutches, he expected other specialists to be interested and supportive. Instead, his colleagues accused him of recklessness, applying a carpenter's approach to surgery, and even fabrication of his results (6). However, he said he was ready to prove his data if the Sverdlovsk Scientific Research Institute for Restorative Surgery, Traumatology and Orthopaedics would give him an opportunity and help with a series of experimental canine studies.

Curiosity of scientists won over their skepticism and during 1957 to 1960, Ilizarov conducted a series of experiments in Sverdlovsk. The results of those studies surprised Ilizarov himself. Bone healing during compression arthrodesis of the canine knee occurred on days 8 to 10 after surgery (Figs. 6 and 7), with complete remodeling of the regenerated tissue on days 30 to 45.

The data of those experiments on 73 dogs and his early clinical work was reported in his dissertation ''Transosseous Compression Osteosynthesis by the Author's Apparatus,'' in 1968 (7). The main topic of his research was the importance of stable fixation by eliminating movements of bone fragments using compression osteosynthesis to achieve primary bone healing in the shortest possible time.

He presented the clinical results of 444 patients after arthrodesis of major joints (Fig. 8), correction osteotomies (Fig. 9), and treatment of nonunions (Fig. 10) and fractures.

In his clinical data, the average time of the bone healing after resection arthrodesis of the knee using compression osteosynthesis was 17.59 days, and 31.3 days for healing of diaphyseal fractures.

Ilizarov wrote in his dissertation:

''Conditions for developing primary bone healing are provided by creating complete immobility of bone fragments and maximum contact area. In explaining the reasons of accelerated healing during compression osteosynthesis one should proceed not from the idea of accelerating the regeneration but by admitting that any movement at the bone end junction slows formation of healing.''

He advocated, from this work and his prior experience, minimal resection, and stated that he was against resection to correct deformities. Instead, he recommended gradual lengthening of soft tissue before proceeding with fusion.

In the dissertation conclusion he wrote:

''It is difficult to find any other biological problem, to studying of which were devoted so many research investigations, as the problem of regeneration of bone tissue. At the same time, it is difficult to find any problem in orthopaedics and trauma, the study results of which had so many contradicting conclusions, deposits of subjectivism, and as a consequence (resulting

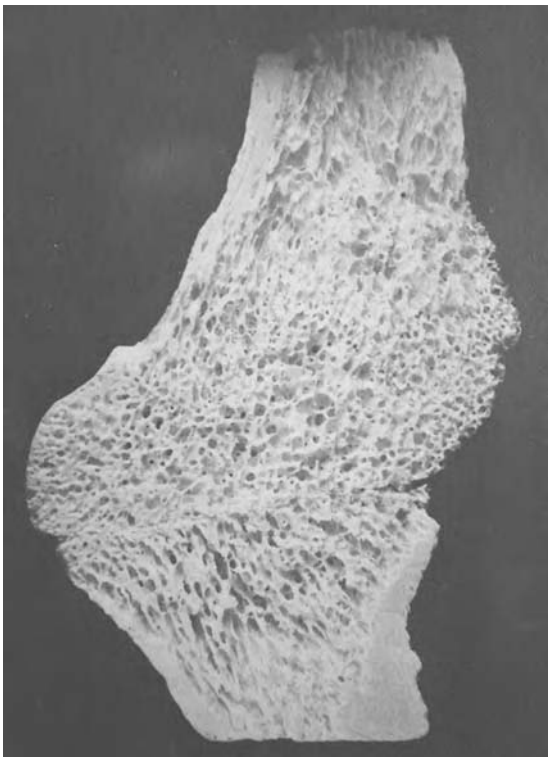

Figure 6 Canine knee specimen shows bony healing eight days after knee arthrodesis surgery (saggital cut).

in a) multitude of unjustified recommendations.... A slow process of regeneration was considered to be a specific quality of bone tissue and led scientists down the road of looking for biologic stimulators of osteogenesis. At the same time, the research of some other scientists (Kapsammer, 1897; Vasiliev, 1889; Matzuoka, 1903; Syngaevski, 1911) regarding ability of bone tissue to heal as soft tissue with primary intension in utmost short time was not given much attention. The results of our research and years of clinical experience convinced us that healing by primary intention may and ought to serve as the only standard for bone tissue regeneration. And if until recently majority of authors observed bone healing with secondary intension, than in the first place the cause of that was imperfect fixation of bone fragments.''

My father defended his dissertation in Perm, Ural, in 1968. He was awarded the title of Doctor of Medical Science instead of Candidate of Medical Science (equal to a Ph.D.) for which

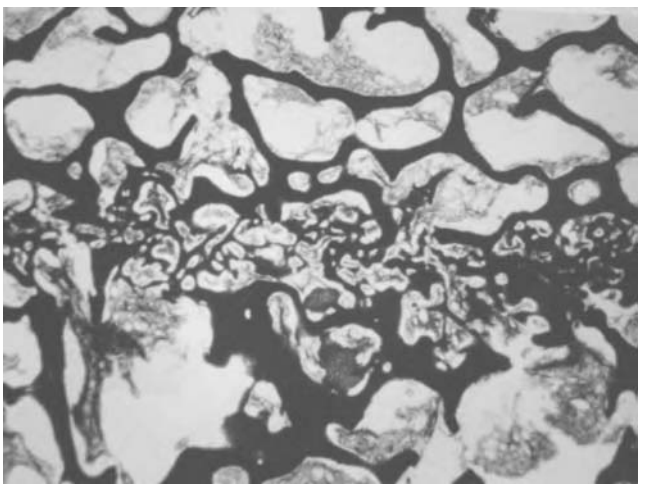

Figure 7 Histology slide of canine knee specimen shows bony healing eight days after knee arthrodesis surgery.

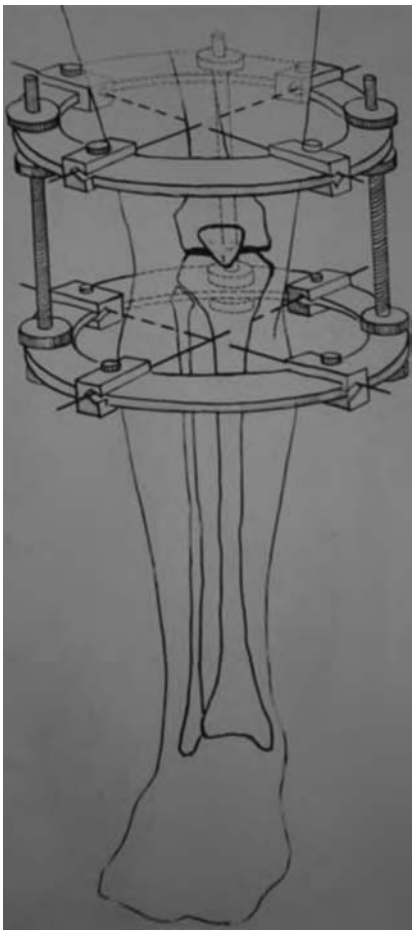

Figure 8 Schematic drawing of the apparatus for compression knee arthrodesis from Ilizarov's dissertation (1968).

he applied (Fig. 11). This uncommon event was even reported in the Soviet media (TASS). Ilizarov formulated the following principles of fracture treatment in his dissertation, which revolutionized the approach and treatment of fractures.

- Preservation of the blood supply
- Preservation of the osteogenic tissue
- Complete anatomic reduction
- Stable fixation
- Functional activity of the muscles and joints
- Early patient mobilization

By providing bone and other tissues with ideal conditions for healing and without compromising an already injured limb further, we should see faster healing time and speedier recovery as was shown in my father's work.

The development of new bone enhancement products is exciting, especially for conditions with compromised bone healing. However, we should be careful not to forget or underestimate the principles stated above and rely on those strong bone enhancement products alone.

Ilizarov started to perform different bone lengthening procedures in early 1950s. At that time, he was using classic long oblique or Z-shaped sliding osteotomies and his frame for distraction and immobilization. The ability of bone to regenerate by distraction (or distraction osteogenesis) was discovered a few years later by serendipity. A case of hypertrophic nonunion was supposed to be treated by compression osteosynthesis; however, the nuts on the rods were turned in the wrong direction creating distraction forces instead of compression. When Dr. Ilizarov observed some cloudy density on a radiograph of that patient, he believed

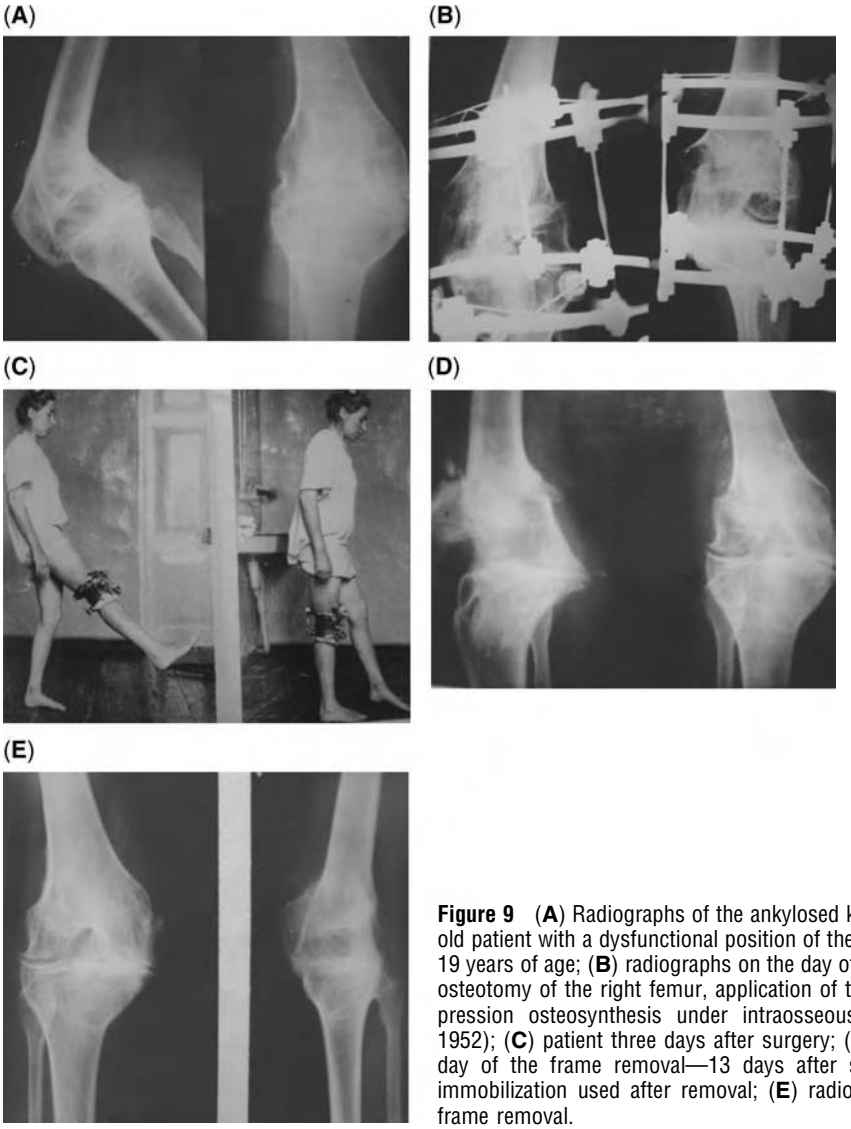

Figure 9 (**A**) Radiographs of the ankylosed knee joint of a 41-year-old patient with a dysfunctional position of the knee after infection at 19 years of age; (**B**) radiographs on the day of hinged supracondylar osteotomy of the right femur, application of the apparatus for compression osteosynthesis under intraosseous anesthesia (July 7, 1952); (**C**) patient three days after surgery; (**D**) radiographs on the day of the frame removal—13 days after surgery, no additional immobilization used after removal; (**E**) radiographs one year after frame removal.

it to be new bone formation. A few years after that, in the early 1960s, similar density had been noted by him in a case of accidental epiphyseolysis in a child with premature consolidation at the osteotomy site. He confirmed his conjecture by experiments and brought both of these techniques into his practice. He called this method ''bloodless'' surgery for lengthening (without osteotomy).

In 1963, he reported on a use of distraction hinges mounted in the apparatus at the knee joint so that, ''the diastasis [of the knee joint] remained unchanged at any position of flexion. Hinges could be fixed at any angle of flexion.... Apparatus... eliminates side-to-side movements and at the same time allows to do range of motion in one (sagittal) plane keeping a constant set distance between articular surfaces'' (2,3).

Ilizarov worked to improve both instrumentation and technique. He emphasized preservation and gentle handling of the soft tissue avoiding use of Kocher's type of instruments. He adjusted the traditional osteotomy to a so-called corticotomy, based on his own approach to the idea of minimally invasive surgery. This resulted in less damage to the soft tissues and bone marrow during surgery and even faster healing and recovery. He perfected his technique and routinely used an osteotome that was 5 mm wide for children and a 10 mm for adults, performing osteotomies through even smaller skin incisions. He did not use a drill or a Gigli saw.

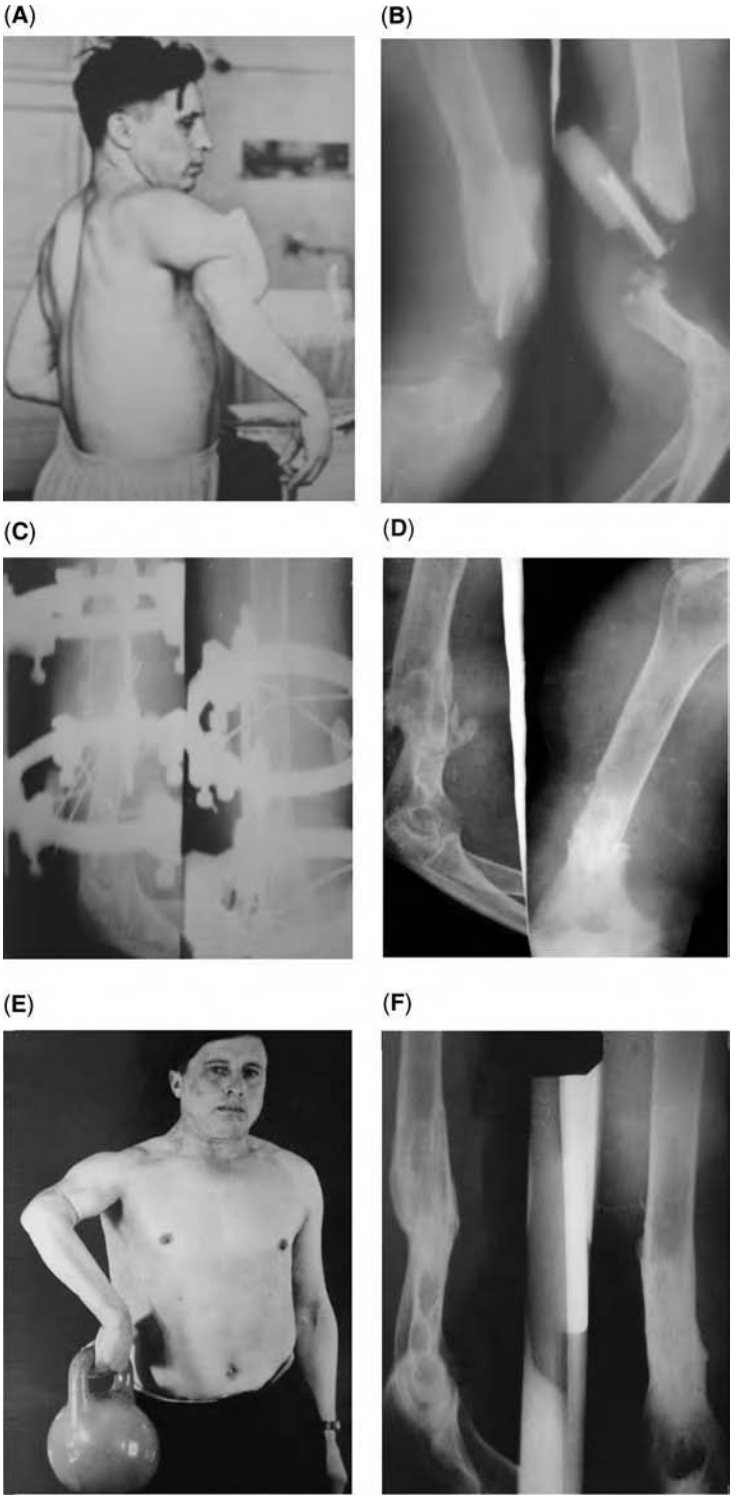

Figure 10 (**A**) Patient with nonunion of distal third of the right humerus (six years after injury and four unsuccessful surgeries including bilateral tibial bone autografts and plastic endoprosthesis); (**B**) radiographs before surgery (note a free floating plastic prosthesis from previous surgery); (**C**) radiographs on the day of the application of the frame under regional anesthesia (April 3, 1964); (**D**) radiographs on the day of frame removal (total time in the frame: 33 days); (**E**) patient four years after surgery; (**F**) radiographs of the right humerus four years after surgery.

Figure 11 Professor Gavriil A. Ilizarov and his national and international awards.

He also developed a new approach in limb reconstruction surgery such as treatment of bone defects without use of bone grafts but using bone's ability to heal (bone transport techniques).

Ilizarov also thought of and performed experiments to evaluate the influence of the stability of fixation, as well as the degree of preservation of bone marrow, nutrient vessels, periosteal, and soft tissues on osteogenesis during distraction.

As his investigations continued, he discovered ability of other tissues—blood vessels, nerves, skin, etc.—to regenerate during incremental distraction as well. He called it the Law of Tension–Stress.

The stimulating effect of tension–stress on regeneration was officially registered by the State Committee of the USSR on inventions and discoveries as a discovery under the number 355 on September 15, 1988 entitled ''General Biologic Property of Tissues to Respond to Incremental Tension with Growth and Regeneration.''

However, even in those days, there were opponents to this registration. Interestingly, in 1991, Professor A.G. Babayeva, in her review of Ilizarov's Law of Tension–Stress, argued with his opponents: ''Indeed many researches described this phenomenon; however, no one found a new law. All saw that any object thrown in any direction falls to the ground .... However, the Law of Gravity was discovered by one man—Newton.'' (Fig. 12).

He used this discovery for developing new directions in research and clinical practice such as transverse bone distraction for the treatment of circulatory limb disorders (Buerger's disease and atherosclerosis) by neovascularization, for eliminating defects in one of the paired bones, or for remodeling the shape of a limb (Fig. 13). He performed experiments on bone formation under the influence of tension–stress for the filling of skull defects. Those techniques are currently used in clinical practice in maxilla- and craniofacial surgery. He also conducted experiments for elongation of vertebral bodies, which were done on puppies by distraction epiphyseolysis, and created an animal model of scoliosis by changing blood supply to one side of the spine in young growing dogs.

The Ilizarov method of gradual deformity correction, use of hinges in the frame, and calculation of their location, revolutionized the process of deformity correction.

Figure 12 Prof. Golyakhovsky in New York also noted a resemblance and drew well-meant caricature of Ilizarov as Newton in 1988 in an attempt to answer the curious orthopedic residents' question at the Hospital for Joint Diseases, "How could Ilizarov come up with such an idea?"

He also studied the influence of rate and rhythm of distraction on regeneration of bone, comparing bone formation in a series of experiments with complete osteotomy and closed osteoclasia with different rates and rhythm of distraction. These studies demonstrated that both the rate and the frequency of distraction are important to osteogenesis. When the limb was lengthened 0.5 mm daily, at a frequency 0.125 mm $\times$ 4 a day, osteogenesis overtook the speed of distraction and caused premature consolidation, especially when closed osteoclasis technique was used. A rapid distraction rate of 2 mm a day at a frequency of 0.5 mm every six hours, in contrast, not only retarded osteogenesis but also caused detrimental changes in the soft tissues surrounding the site of distraction. Lengthening by 1 mm a day (0.25 mm every six hours) led to more favorable results. Using a greater frequency with autodistraction (1 mm in 60 steps instead of 4) brought even more superior results (8). It was also observed that when autodistraction was used, the proliferative, metabolic, and biosynthetic changes in cellular activity in many tissue elements took on features characteristic of histogenesis during embryonic, fetal, and postnatal limb growth. These discoveries changed the way limb lengthening and deformity correction is done today.

Later, Ilizarov studied the interrelation of osteogenesis and hemopoiesis and expanded his research into different fields using the newest available technology in biology (including stem cell research), physiology, nuclear medicine, hematology, chemistry, biochemistry, immunology, and others. He published a significant part of his experimental work in the book "Transosseous Osteosynthesis," in 1992.

It was not just the device that made the difference, but years of experimental and clinical work discovering the potential of bone tissue and its growth, the development of the tension–stress theory on regeneration of all tissues, and incorporating this into a new biological law. "It was learning from nature," Professor Ilizarov often said.

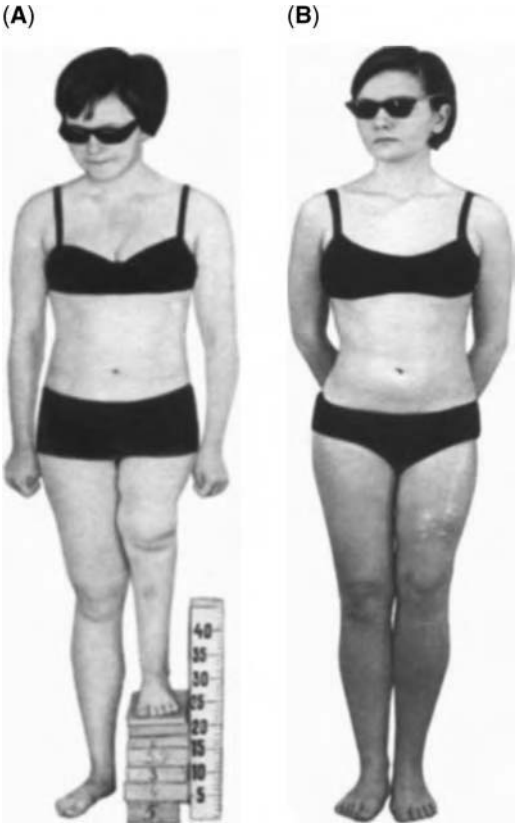

Figure 13 (**A**) Patient before treatment; (**B**) Patient after lengthening and remodeling the shape of her leg.

AMBULATORY SURGERY EDUCATION SOCIOECONOMIC IMPACT

The ambulatory care department was created in 1971 in order to reduce the time spent as an inpatient, the cost of treatment, and to accommodate a greater volume of patients coming for treatment from other parts of the country. This department took care of patients mostly with fractures and nonunions as well as some patients for lengthening with achondroplasia. It was a novel approach to treatment and was not widely accepted at that time.

Ilizarov paid a lot of attention to education and training of specialists. He realized that the only way to help huge numbers of patients coming to him for help and to reduce decade-long waiting lists in his hospital was to train other physicians interested in mastering this technique, in order to provide care in other regions of the country. In 1979, continuous instructional courses on the method of transosseous osteosynthesis for surgeons were organized at the Kurgan Institute. Ilizarov also trained researchers and guided over 50 medical dissertations for candidate and doctor of science degrees.

Russian studies of the Ilizarov system demonstrated significant reduction in treatment time, cost of treatment, and disability payments. In the management of fractures and post-traumatic nonunions, the use of this system decreased primary disability three to five times and in cases of open fractures, eightfold, in comparison to traditional treatment. An important factor in successful treatment was the time from the original injury to the application of the frame, which had a direct impact on the percentage of disability; the earlier treatment was started, the better the outcome for the patient. Even more impressive was the percentage of patients who returned to work after long-term disability secondary to the sequelae of trauma. Capacity for work was restored in 96% of the cases (9). The socioeconomic impact was calculated in Russia and could be difficult to assess in other countries. However, it showed significant economic advantage, which should be important in any country, especially with continuous growth of medical and social cost.

THE MICHELANGELO OF ORTHOPEDICS

Another patient, Carlo Mauri (1930–1982), who was a well-known Italian journalist, alpinist, and explorer, helped to introduce the Ilizarov system into Western Europe. Mauri was part of an international expedition crossing the Atlantic on a papyrus boat. The boat was reconstructed entirely of reed and designed from ancient Egyptian drawings. The expedition was organized by Thor Heyerdahl, (1914–2002), a famous Norwegian anthropologist and explorer, who was best known for his theories about migration patterns of various ancient peoples. By his successful crossing of the Atlantic from North Africa (Morocco) and a safe landing at Bridgetown, Barbados, in 57 days, he proved that ancient Egyptians could have reached South America and founded the Aztec and Inca cultures 4000 years ago. Carlo Mauri had suffered a distal tibia fracture during an accident in the mountains 10 years earlier. The team physician of the Atlantic crossing, Dr. Yuri Alexandrovich Senkevich (1937–2003), was from the Soviet Union. On treating Mauri during the trip when his old leg wound reopened, he advised him to consult with Dr. Gavriil Ilizarov in Kurgan, Siberia.

After crossing an ocean in an ''ancient paper boat,'' the remoteness and Siberia's reputation would not deter a man of Mauri's courage and adventure. His surgery in 1980 was a success, and Mauri's infected tibial nonunion completely healed. On his return to Italy, Mauri wrote an article in an Italian newspaper, naming the Russian physician Gavriil Ilizarov as the ''Michelangelo of Orthopaedics.'' This would prove to be the break in the dam that would release the news of the Ilizarov method and external fixator to the world medical community and set in motion events that have led to worldwide application and study of the Ilizarov method.

Carlo Mauri's physicians in Italy were amazed by the healing that had occurred of his longstanding nonunion condition. Mauri subsequently invited Ilizarov to Italy, in June of 1981, and arranged for his participation in XXII Italian AO conference in Bellagio, Italy, through his physician friends. It may be difficult today to imagine the effort to bring news of such medical accomplishment out of the Soviet Union, until it is realized that the meeting in Bellagio was Ilizarov's first clinical presentation on the other side of the ''iron curtain,'' Dr. Roberto Cattaneo recalled later (10):

''Some of the cases which he presented were so dramatic that more than one surgeon at the Congress became suspicious and insinuated that he was displaying elegant photomontages for promotional purposes. His friends in Lecco, however, believed in him, trusted his scientific work, and honoured his humanity . . . . Procedures such as arthroscopy and progress in joint replacement surgery and microsurgical operations reflect technological advances, but Ilizarov's contribution has been to detect a natural law of regeneration of biological tissues such as bone, musculotendinous units, and neurovascular structure, during slow and progressive distraction, and to turn these findings into a practical expression. Distraction osteogenesis mimics the physiological process of the bone growth, which occurs at the growth plate.''

During this trip to Italy, my father performed surgery on the lower leg of a little girl with a leg length discrepancy (Fig. 14). I remember it was quite an event in the OR complicated by the fact that a medical translator was not available. Though we had Carlo Mauri's young niece who studied some Russian, it was her first time in the OR and she was very nervous. She was trying to translate initially but soon stopped her attempts especially when my father requested ''three holes male . . . post.'' The poor OR nurse looked completely lost (she had never seen this equipment before—a heavy suitcase filled with a complete set of the apparatus was lost by an airline and was found and delivered on the day of surgery). I got closer to her table where all parts of the set were spread out. I myself was two years out of high school, but fortunately, by that time, I had some knowledge of the device. I tried to help the nurse by pointing to the area of the table where the asked part of the frame was located, but she would always pick the wrong one. In desperation, the nurse gave me a sterile glove and we managed to get my father necessary equipment on time. I remember he completed an osteotomy in record time and was very pleased with the surgery.

Italian physicians immediately realized the significance of this ''Siberian technique'' and were enthusiastic to learn the procedure. The following year, in 1982, Professors Roberto Cattaneo, Antonio Bianchi-Maiocchi, G.B. Benedetti, and Dr. Angelo Villa formed an Italian society dedicated to learning and dispersing knowledge about the Ilizarov system: the Association for the Study and Application of the Methods of Ilizarov. In 1983, the first course

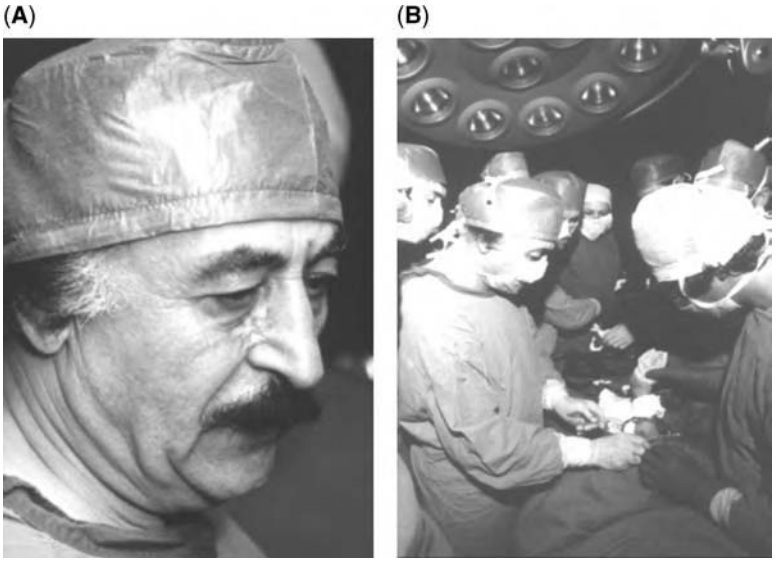

Figure 14 (**A**) Professor Ilizarov in Italy (June 1981); (**B**) Professor Ilizarov performing surgery in Italy (June 1981).

was held in Lecco, Italy. The organization was successful in stimulating an interest in the Ilizarov method in Europe and other countries.

American surgeons first learned of this technique from their European colleagues abroad. Dr. Victor H. Frankel, President of the Hospital for Joint Diseases (HJD) in New York, saw the external fixator device, which caught his attention on display in the scientific exhibit area at the meeting in Spain. After making inquiries, Frankel together with Dr. Stuart Green, a colleague from California, visited the Ilizarov center in Kurgan in 1987. Drs. Frankel and Green became newly converted ''Ilizarovians.'' Green returned to Kurgan several more times, and later provided editorial assistance in preparation of the English translation of my father's book ''Transosseous Osteosynthesis,'' published in 1992.

Dr. Frankel, after his trip to Kurgan, was decisive in bringing the Ilizarov method to the United States and invited Dr. Ilizarov to give the Sir Robert Jones lecture of 1987 at the HJD. Dr. Golyakhovski recalled this event: ''Ilizarov requested three carousel projectors for his lecture, which surprised the inviting party. The auditorium was packed. People were sitting and standing in the aisles. (Ilizarov) showed all 700 slides in one and a half hours. When he finished, the audience jumped from their seats and applauded, standing, for about 10 minutes'' (11). Dr. Golyakhovski by this time had immigrated to the United States, and was recruited by Frankel to work on the Ilizarov method with Dr. Frankel and Dr. Alfred Grant, a pediatric surgeon and Director of the Center for Neuromuscular and Developmental Disorders at HJD.

Interest in the Ilizarov external fixator in United States was contagious and many started to use the apparatus without proper training, making mistakes that led to complications as well as discouragement and misplaced blame on the external frame. My father sometimes enjoyed reminding others of a Russian expression that, ''Boldness should not exceed one's skills.'' Fortunately, many others carefully studied his method, attended courses and conferences, and became world-known experts.

DON QUIXOTE AND HIS LAST BATTLE

During the course of Gavriil Ilizarov's life, there were many battles he had to fight, not only the ones to prove the validity and value of his work. Because of the poor economic conditions in the country, including those for his own employees of the hospital, other doctors, and healthcare workers, he took on the burden of improving the basic quality of their lives. At times, that meant going to Moscow to arrange for funds for food and housing. He knew that whatever he could do would improve, directly or indirectly, the care of his patients. These added stresses contributed to weakening his health. In 1988, he traveled to New York and underwent several vascular surgeries at Hospitals. At that time, he was strongly advised to

have further tests for heart disease. His electrocardiography had showed some signs of previous myocardial infarctions of which he was not aware. However, physician Gavriil Ilizarov won out over patient Ilizarov and was in a rush to return to his work and his waiting patients. Regrettably, though my father completed several trips between New York and Kurgan between 1988 and 1991, he never seemed to take as good care of himself as he did of his patients, and the tests were never done.

Plans for Dr. Ilizarov to become a visiting professor at HJD were set for September 1992. Not surprisingly, it took some time for Dr. Frankel to convince a Soviet orthopedist obsessed with work and his hospital, his ''Siberian creation,'' to come to New York. Tragically, Dr. Gavriil Abramovich Ilizarov died in Kurgan, Siberia, on July 24, 1992, from transmural myocardial infarction following one of his numerous litigations to save residential apartments for his staff that were being taken away by the city. That particular litigation concerned a newly constructed building for which he had worked to obtain finances from the Ministry of Health. This was to be his last fight. People sometimes gave my father small figures of Don Quixote, perhaps for the blind faith that Quixote maintained in the goodness of mankind. While Quixote's life pursuit was to bring order to a tumultuous world, my father fully understood and engaged the tumultuous realities of his world to bring forth the best in himself and to do the best by his patients and those for whom he felt responsible.

FUTURE DIRECTIONS: RESEARCH, TECHNOLOGY, AND EDUCATION

Future directions of tissue regeneration research begun by Ilizarov lie in the collaboration of multidisciplinary experts in the other fields such of spinal cord and nerve injury research, vascular disease, muscle disease, and hematological conditions.

Progress in technology will improve all aspects of treatment using the Ilizarov method, to include preoperative planning with computer-assisted software calculation and prediction of all possible strategies of treatment, based on (i) patient symptoms; (ii) radiographic images using auto measurements of deformities, length discrepancies, and bone quality; (iii) appearance of soft tissue from digital imaging; and (iv) gait analysis. Use of navigation and robotic technology will help during surgery, as well as during the postoperative period, utilizing satellite technologies to monitor auto-adjustments with motorized remote control systems. All of this is feasible in the future.

Today, however, the most attention should be paid and every effort made on education. Teaching residents with an emphasis on mechanical and biological principles is essential. The Ilizarov technique should become part of every training program relative to the care of acute complicated trauma patients, especially in Category I trauma centers. Residents should be as comfortable applying circular frames as they are placing spanning external fixators. Though the latter can be more quickly applied, temporary spanning frames do not provide the same degree of immobilization, correction ability, nor allow any joint movement or weight bearing during recovery and require additional surgery. Residents should become comfortable with the device and its modularity together with understanding mechanical and biological principles of the Ilizarov method. Malunions should not be viewed as the most common complication of external fixation because it is still stated on orthopedic written board exams. Many malunions with any external device are caused by a poor primary reduction or, in some cases, by loss of reduction due to instability of these external devices or due to inability to maintain stable fixation. Failures of any method and transosseous osteosynthesis, in particular, could be explained by insufficient training and experience. Dr. Solomin listed the top three main contraindications for using external fixation, which preceded health-related contraindications to surgery: (i) lack of necessary qualification by a surgeon to perform transosseous osteosynthesis of estimated complexity; (ii) inadequate organizational/technical conditions for performing the surgery and absence of trained personnel; and (iii) absence of an adequate follow-up system postoperatively (12). I believe the perception of external fixation will be changed by centers developing an effective system of training that enters the curriculum and creating necessary conditions for surgeons and patients to use this method safely, effectively, and successfully.

> *All truth passes through three stages. First, it is ridiculed. Second, it is violently opposed. Third, it is accepted as being self-evident.*
>
> Arthur Schopenhauer (1788–1860)

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Part II PRINCIPLES

2 Basic Science and Biological Principles of Distraction Osteogenesis

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EDITOR'S INTRODUCTION

This chapter is the first complete publication of a manuscript titled ''Distraction Osteogenesis, Experimental Studies of Strain-induced Bone Growth in the Canine Tibial Lengthening Model.'' The paper authored by Dr. Aronson was selected for the Arthur H. Huene Award by the pediatric society of North America. It describes a series of experiments performed over a 10-year period to study the biology of distraction osteogenesis. This research was supported by the orthopedic research and education foundation, Arkansas Children's Hospital and the University of Arkansas for medical sciences.

An understanding of the biology and the basic science of the Ilizarov method is essential for the practicing limb lengthening surgeon. These principles guide clinical treatment. This chapter can be studied on two different levels. Most of the biological principles and clinical significance are outlined in the sections Summary, Clinical Significance, Introduction, and Discussion. The sections Materials and Methods and Results contain great detail about the experimental method and may be beyond the interest of the average clinician. This detail is included because it will be invaluable for the next generation of basic scientists. These experimental models may be used to test the benefit of new modalities such as systemic drugs, local bone healing enhancements, and new external fixator and intramedullary nail designs.

SUMMARY

Distraction osteogenesis is a unique clinical method for regenerating local bone deficiencies in length, width, or alignment or in bones with intercalary gaps, nonunions, or osteomyelitis. As introduced by Ilizarov, gradual mechanical distraction of a low-energy osteotomy spontaneously produces potentially unlimited new bone from the local host bone that rapidly remodels to normal structure, even in skeletally mature bone.

Over a 10-year period, a series of basic research projects was completed to answer three questions: (i) What growth process, if any, occurs within the biological interface that can spontaneously regenerate bone? (ii) Which clinically available variables are important to modulate this local biology and to affect outcome? and (iii) Do mechanical factors play an intrinsic role in this process of bone formation that might relate to our understanding of Wolff's law and help to predict outcome?

Sixty-five adult mongrel dogs, divided into subgroups, underwent left tibial lengthening in order to test the effects of variations in technical factors including rate and rhythm of distraction, latency period, osteotomy site, fixator type, and stability. Distraction osteogenesis was also characterized at different points in time by biopsy and whole bone specimens using decalcified and nondecalcified histology, India ink injection with Spalteholz clearing, back-scattered scanning electron microscopy, gravimetric and chemical analysis, and biomechanical testing. Relatively noninvasive methods including plain radiography, arteriography, technetium scintigraphy, computerized axial tomography, and strain gauge measurements were used in order to correlate findings with the invasive techniques.

Under ideal conditions—stable fixation, a low-energy osteotomy located in the proximal metaphysis, following a five to seven day latency, distracted at a rate of 1 mm/day and a rhythm of two to four times per day—distraction osteogenesis occurred reliably as pure intramembranous ossification. This bone regenerate arose between the entire cross-section of each distracted bone surface and a central radiolucent "fibrous interzone" (FIZ), histologically comprised of parallel collagen bundles. New bone trabeculae formed directly from this central collagen bridge extending toward both host bone surfaces (HBS) as extremely uniform microcolumns with average diameters of 150 microns, oriented parallel to the distraction force and surrounded by slightly larger diameter blood vessels oriented in the same fashion. Following distraction, the microcolumns bridged across the central zone and rapidly remodeled to a macro- and microstructure similar to that of the host bone region (consolidation). This experimental model added about 26% new bone mass and volume to an adult canine tibia, that tested at 43% to 47% as stiff as the unoperated control side, four months following the initial operation. The bone bridge, though mechanically not normal, allowed full painless weight bearing by the dogs.

Unstable fixation, distraction at a rate exceeding 2 mm/day, or a sporadic rhythm led to poor bone formation and nonunions. Prolonged latencies (14–21 days), especially in the metaphyseal region, carried a high risk of premature consolidation, which prevented distraction osteogenesis from occurring.

Quantitative technetium scintigraphy (QTS) reflected a major increase in regional blood flow that temporally paralleled that reported in the injury-repair model of the canine tibia. Quantitative computer tomography (QCT) was more sensitive than plain radiography in measuring the initial mineralization at two to three weeks of distraction; a QCT-based finite element model also predicted the actual mechanical strain of the regenerate bone.

In-line strain gauges measured the loads during distraction, which reflected the changing mechanical properties of the developing regenerate bone. A differential dissection of the soft tissues during in vivo load measurement revealed that a mean of 72% of the resistance to distraction (after 30% lengthening) resided in the osteogenic area. The measured loads increased with time during distraction. Loads generated at metaphyseal sites exceeded the loads generated at diaphyseal sites. These loads were inversely proportional to the length of unmineralized collagen bridge FIZ remaining, which radiographically decreased during distraction. The loads also increased directly proportional to the cross-sectional area of the osteogenic zone, as measured by computed tomography (CT) at the level of the collagen interface FIZ bridging the new bone columns. The loads from both metaphyseal and diaphyseal lengthenings were normalized to osteogenic zone "stress" by dividing each measured load by the CT calculated cross-sectional area of the FIZ in each animal. The increase in osteogenic zone stress was uniform over time, independent of the bone site. A low osteogenic zone stress predicted a future nonunion as early as three to four weeks of distraction, presumably due to disruption of the normal biological bridge.

CLINICAL SIGNIFICANCE

The technical conditions for successful distraction osteogenesis as stipulated by Ilizarov were supported by our experimental work. Further insight into the process revealed that the bone formed by direct intramembranous ossification of a primary collagen bridge that progressively mineralized. Several other clinical correlations became apparent from this work.

New bone growth at the zone of Ranvier (the circumferential periosteal bridge surrounding the growth plate), which is histologically identical to that seen in distraction osteogenesis, probably forms as a result of distraction loads imparted to the periosteum from the growth plate pushing the epiphysis away from the metaphysis (just as the external fixator stretches the FIZ).

The sudden appearance of new radiodense bone within the distraction gap during the third week of distraction, despite histological, chemical, and QCT evidence that the microcolumns were already mineralizing one week earlier, confirmed the visual limitations of standard radiography, as suspected but not previously proven, that a 40% change in radiodensity is necessary to see a change on a radiograph.

The massive increases in local and regional blood flow did not differ significantly from those measured in normal fracture healing, indicating that the distraction process did not

prolong or enhance this regional reparative response. Increasing the rate of distraction seemed to approach the limits of local blood flow, which may be a rate-limiting factor in successful distraction osteogenesis.

QCT had the potential to measure the structural stiffness of a long (tubular) bone by conversion of CT density to apparent density and then to modulus of elasticity for a geometric finite element model.

Strain gauges attached in series to the ring external fixator could directly measure the loads within the collagenous interface, predicting either a normal biological interface, which would proceed to union, or an abnormal interface, which would proceed to nonunion as early as the third week of distraction. With these loads converted to osteogenic zone stress, normalized values could be applied to any bone or site within a bone.

INTRODUCTION

Ilizarov (1,2) first introduced this method both experimentally and clinically over his 40-year career in Siberia. Distraction osteogenesis involves the mechanical stretching of the reparative process invoked by a low-energy osteotomy using external fixation. Ilizarov's work implies that this process is regenerative rather than reparative; consequently he referred to the new bone as ''regenerate'' (3,4). His clinical successes saving the limbs of thousands of patients with conditions that traditionally resulted in amputation have revolutionized the current practice of modern orthopedic surgery (5,6). This procedure has been successfully used to regenerate bones deficient from congenital conditions such as hemimelias and other failures of formation, from acquired shortening secondary to fractures or infections during childhood that stopped normal bone growth, and from intercalary bone defects where large segments of a bone have been lost due to open fracture, osteomyelitis, or a variety of local tumors or dysplasias of bone (7–9). Bone deformities and discontinuities (e.g., nonunions) have also been corrected with this technique (7,8). The procedure has been successful in patients at nearly any age from early childhood to middle-aged adults (8). The actual length of new bone produced from a single procedure can be as much as 18 to 20 cm per limb segment, sometimes extending an individual bone by over 100% of its initial baseline length (8). Multiple lengthening sites are possible for simultaneously producing new bone segments, lengthening the limb even more rapidly. The new bone lengths are usually of equivalent cross-section and quality to the local site in the host bone. Bone production seems to be highly successful in these procedures but soft tissue growth and preservation of normal joint function may limit the clinical applications (10,11). Certain clinical applications of the distraction procedure are more controversial, including stature lengthening in dwarfs and arthrodiastasis (stretching of joint contractures).

Ilizarov and his scientific colleagues conducted a series of experiments using canine tibia to study distraction osteogenesis. By varying the stability of fixation, energy of the osteotomy (i.e., degree of vascular damage) and the ''rate and rhythm'' of distraction, he postulated that all four factors were critical to osteogenesis (3,4). He concluded that a distraction rate of 1 mm/day was necessary for the spontaneous regeneration of new bone that would successfully bridge the bony gap. Earlier attempts by others to lengthen limbs using similar mechanical external fixation devices failed to produce new bone in the gap. A historical review indicates that rate and rhythm were never considered to be important. Codivilla (12) attempted to stretch the entire gap under anesthesia all at once, while Wagner (13,14) had the patient turn the distraction knob as fast as pain or neurological problems would allow, routinely planning to fill the gap with bone graft, which could then take years to fully consolidate. Interestingly, Sunderland (15) discussed experimental evidence that spontaneous nerve regeneration occurred at a rate of 1 mm/day long before the importance of rate was correlated to distraction osteogenesis.

Over 10 years, a series of experiments were conducted using 65 dogs to better understand the process of distraction osteogenesis. Invasive analysis using decalcified and nondecalcified histology, India ink injection with Spalteholz clearing, back-scattered scanning electron microscopy, gravimetric and chemical analysis, and biomechanical testing have all been utilized in our laboratory to characterize this process. Relatively noninvasive methods including plain radiography, arteriography, technetium scintigraphy, computerized axial tomography, and strain gauge measurements have been used in order to correlate findings with the invasive techniques. By studying the animals in groups of four, the treatment

conditions could be altered to produce significant variations in outcome from the normal process. Using these different outcomes confirmed by invasive tests, the noninvasive tools were compared in order to develop reliable clinical methods to monitor patients in the clinical setting.

MATERIALS AND METHODS

Experimental Design

Ilizarov's original model of a 28 mm (15%) lengthening of the adult canine tibia was reproduced to study the histology, blood supply, and radiology of bone formation that occurs in the distraction gap, with four per group, compared by a Wilcoxon nonparametric statistical analysis (16).

Canine Tibial Lengthening Model

Adult mongrel dogs weighing 15 to 25 kg were used in all experiments. The operation was performed on day 0. In all animals, an external fixator was applied with four pins and a low-energy osteotomy was performed using the technique described by Ilizarov (17). This ''corticotomy'' was performed with a chisel and rotational osteoclasis to crack the peripheral cortex subperiosteally, attempting to preserve the endosteal blood supply (18). After a seven-day latency period following the index operation (the latency period was varied in later experiments), mechanical distraction was carried out from day 7 to day 35 for a total of 28 days (in one group distraction was prolonged to 56 days). The distraction rate of 1 mm/day at a rhythm of four increments per day (0.25 mm every six hours as recommended by Ilizarov) was compared to more sporadic rhythms and more rapid rates (19–22). From day 35 to day 77, the frame was left intact for a 42-day consolidation period. At day 77, the frame was removed under sedation and the dogs were finally euthanized on day 119, 42 days later.

Metaphyseal Versus Diaphyseal Corticotomy Sites

To test the effect of the osteotomy site on the osteogenic potential (22,23), 32 dogs underwent left tibial corticotomy for a 28 mm lengthening and were randomized into two groups by corticotomy location: 16 in the proximal metaphysis and 16 in the mid-diaphysis. Because different latency periods had been advocated for optimal osteogenesis (1,24,25), each major group was then divided into four subgroups of four dogs each on the basis of 0-, 7-, 14-, and 21-day latency periods (Fig. 1A–C).

Histology Specimen Preparation

In vivo biopsies were performed in several animals at different stages of distraction. Whole bone specimens were paired and measured fresh for gravimetrics, photographed, radiographed and then submitted for routine (decalcified) or special histology [nondecalcified, Spalteholtz, back-scattered electron microscopy (26,27)]. Qualitative X-ray microanalysis (Tracor Northern 5500, Noran Instruments, Middleton, WI) for chemical constituents including calcium and phosphorus was also performed (28).

Gravimetrics

Using a modified method of Robinson (29), the whole bone specimens were weighed wet and dry and the volume established by water displacement in a volumetric cylinder. Changes in mass, volume, and density were compared between lengthened and opposite side controls.

Quantitative Chemical Analysis of the Five Major Organic and Inorganic Constituents of Bone

Serial 2-mm thick transverse specimens taken from the fresh nondecalcified tibiae were used for chemical analysis, alternating with histological samples that correlated to each defined zone—FIZ, primary mineralization front (PMF), microcolumn formation (MCF), and HBS. Following desiccation, fat content (% of wet weight) was measured by subtracting the defatted weight from the dry weight following overnight treatment in serial cloroform/methanol solutions (30). The dried, defatted specimens were then hydrolyzed (wet ashed) in 6 N

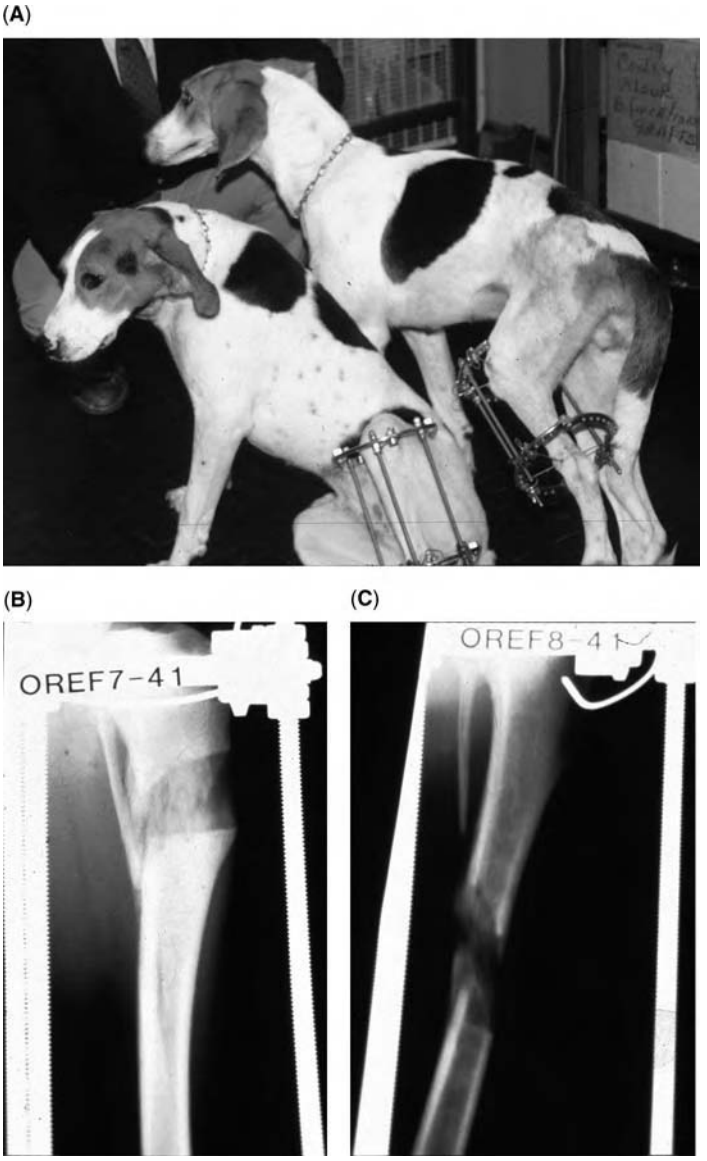

Figure 1 (**A**) Two dogs ambulating on the two-ring fixators during distraction phase. The rings are positioned identically on the tibia, but the distraction site was either in the proximal metaphysis or mid-diaphysis for comparison of bone formation and distraction loads. (**B**) Radiograph of metaphyseal bone formation at the end of distraction. (**C**) Radiograph of diaphyseal bone formation at the end of distraction.

hydrochloric acid. Because bone (hydroxyapatite) exhibits close to a 1.66 Ca/P molar ratio (31), a single dilution was sufficient for both Ca and P ions. Aliquoted portions from the diluted solutions were analyzed in our hospital's clinical laboratory for quantization of calcium and phosphorus. Sample aliquots from each wet ash solution were analyzed for collagen content by using the hydroxyproline assay (spectrophotometer) method of Bergman and Loxley (32). A correction factor of 13.5 g hydroxyproline/100 g collagen was used to calculate the collagen content (33). Results were expressed as milligrams per milligram of wet bone and as a percentage of the normal control bone from adjacent HBS and from the contralateral unoperated tibia at similar anatomical sites. This method was standardized, reproducible, and consistent with values in normal tibiae of about 15% water, 5% lipid, 25% calcium, 12% phosphorus, and 24% collagen; the Ca/P ratio held at about 1.60 and the Ca/collagen ratio was about 1.1.

India Ink/Spalteholz Technique

Selected animals were deeply sedated (ketamine) prior to sacrifice and the femoral artery and vein were surgically exposed, cannulated, and connected to a manometrically controlled pressure gauge for gentle injection of India ink (34). When the effluent channel demonstrated complete India ink replacement, the animal was sacrificed and the tibia isolated by sharp dissection for submersion in Spalteholtz solution. Both whole bone and cut slabs, longitudinal and transverse, were thus rendered translucent. Each specimen was placed in a Petri dish of the Spalteholz solution and photographed on the stage of a microscope for analysis.

Plain Radiology/Photodensitometry

Standardized radiographs were taken with an aluminum stepwedge, weekly from preoperation to end-distraction, and then at biweekly intervals until sacrifice at day 119. The criteria (22) used for evaluation of the radiographs was as follows: (i) *time of first new bone formation*: the initial appearance of faint radiodense new bone formation within the lengthening gap, (ii) *time of first bridging callus*: the first radiodense callus connecting the distracted bone ends, (iii) *time of first cortex*: the peripheral radiodensity increased such that the bone ends were connected by a cortex equal to the adjacent cortices in thickness, and (iv) *time of first canal*: the initial medullary canal appeared in lengthening zone surrounded by cortices on all surfaces (Fig. 2A–D). If a transverse radiolucency persisted within the osteogenic zone by day 77, a *nonunion* was presumed. If the wires became bowed toward the center of the fixator and the radiographs showed no gap of the bone ends after distraction, with early bridging callus, *a premature consolidation* was diagnosed. The final distraction gap between proximal and distal bone ends was measured on the radiograph taken at day 77 using a dial caliper (Mitutoyo #505-635-50, Tokyo, Japan). The *bone healing index* (treatment time per centimeter new bone) was used to compare bone production between the groups and subgroups.

Following calibration for attenuation coefficients using the graduated stepwedge by the modified method of Vose (35), a pinpoint photodensitometer was used to gather relative light transmission data along x–y coordinates tracing the entire osteogenic area from proximal to distal.

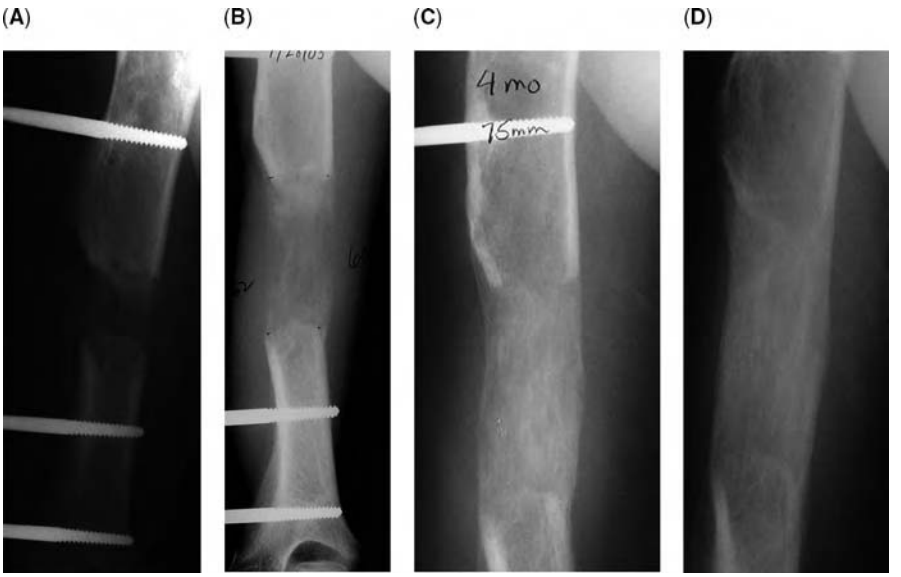

Figure 2 (**A**) Radiograph of humeral lengthening in a patient, demonstrating typical early bone formation at the third week after surgery and second week of distraction. (**B**) Radiograph of same patient at the end of distraction, demonstrating early bridging of the 75 mm distraction gap. (**C**) Radiograph of same patient at four months demonstrating early cortex formation. (**D**) Radiograph of same patient prior to fixator removal when three of four potential cortices had fully bridged the distraction gap as medullary canal remodeling was nearly complete.

Quantitative Technetium Scintigraphy

Ten dogs were randomized into three groups, based on time of distraction: 14, 28, and 56 days. The normal rate of distraction and twice the normal rate were selected to test the effect, if any, of rate of stretching on the blood flow. The third group was subjected to twice the distraction time and length in order to see if the peak flow period could be prolonged by prolonging the distraction. Eight dogs underwent a 28-mm distraction (15% lengthening), four at a rate of $1\,mm/day \times 28$ days and four at $2\,mm/day \times 14$ days. The two other dogs underwent a 56-mm distraction (30% lengthening) at a rate of $1\,mm/day \times 56$ days.

Eight dogs in the first two groups each underwent eight scans: preoperative baseline, weeks 1, 2, 3, 4, and 5 postoperatively (through the entire distraction period) and then during the consolidation period, at week 11 (fixator removal), and at week 17 (presacrifice). The two dogs in third group each underwent scans on week 3 and 7 postoperatively during the prolonged distraction period.

Under ketamine, standard dosages of 10 mCi of technetium-99m were administered intravenously via the forepaw. The gamma camera was placed directly over the tibial crests held parallel to the surface of the collimator. Dynamic data were accumulated at the rate of one image per three seconds for the first three minutes postinjection to calculate flow (60 seconds) and pool (three minutes) images. Two hours later, static bone images of 500,000 counts were accumulated.

Using the left leg (experimental side), a computer color-enhanced isocontour map was used to outline a region of interest (ROI) that corresponded to the 40% region of highest uptake, quantitated as the average number of counts per pixel in each ROI (5). The mirror image of this ROI was then transferred to the opposite (control) side in order to measure the average number of counts per pixel in the exact same anatomical position and area. A third ROI, a fixed rectangular area, was placed over the lateral soft tissues adjacent to the tibial diaphysis on the control right side for the average counts per pixel in the soft tissues. After correcting for decay and actual body weight and subtracting the soft tissue uptake, the experimental side was divided by the opposite to determine the ratio or increase over normal. The control side was compared to baseline (preoperative) measurements and between dogs to confirm its use as a common denominator for the experimental side (36). The three groups were compared over time and to previously published data on injury models (37).

Quantitative Computer Tomography

The serial in vivo density measurements of the new bone mineralization using QCT were standardized with graduated dibasic potassium phosphate phantoms. Under ketamine, each dog underwent a total of five CT scans: preoperation, start distraction, end-distraction, day 77 (fixator removal), and day 119 (presacrifice). Scanning commenced at the proximal HBS, continued contiguously through the entire osteogenic area to the distal HBS, and concluded with an isolated cut through the distal diaphyseal bone. Both legs were carefully positioned within the scanner to align both knees so that the proximal lengthened area correlated anatomically to the opposite side. Each transverse cut was imaged to include both tibiae and the underlying phantom. The outermost perimeter of each bone and the graduated phantoms were then individually outlined using a manual cursor. The mean density per pixel in Hounsfield units represented the average radiodensity of each outlined region of interest from contiguous 4 mm CT slices of the new bone. The average density per pixel of the phantom provided a standard for comparison between scans and different points in time, controlling for scanner drift and more importantly, for any density changes in the contralateral, opposite side ''control.'' In this way, density changes were monitored from proximal to distal across the osteogenic area and compared with the equivalent levels on the control side. The three minimum QCT readings taken from contiguous cuts (the ''weak link in the chain'') were averaged for each dog at each time point. The density ratio of the experimental to the control side was calculated to compare average bone density between groups and subgroups. The cross-sectional area of each bony cut was also obtained automatically by the CT scanner's computer to monitor remodeling to cortex and medullary canal (Fig. 3A–D). Objective information using the geometric distribution of each pixel (relative density) was eventually reduced to hard numbers for conversion to a finite element model (see below).

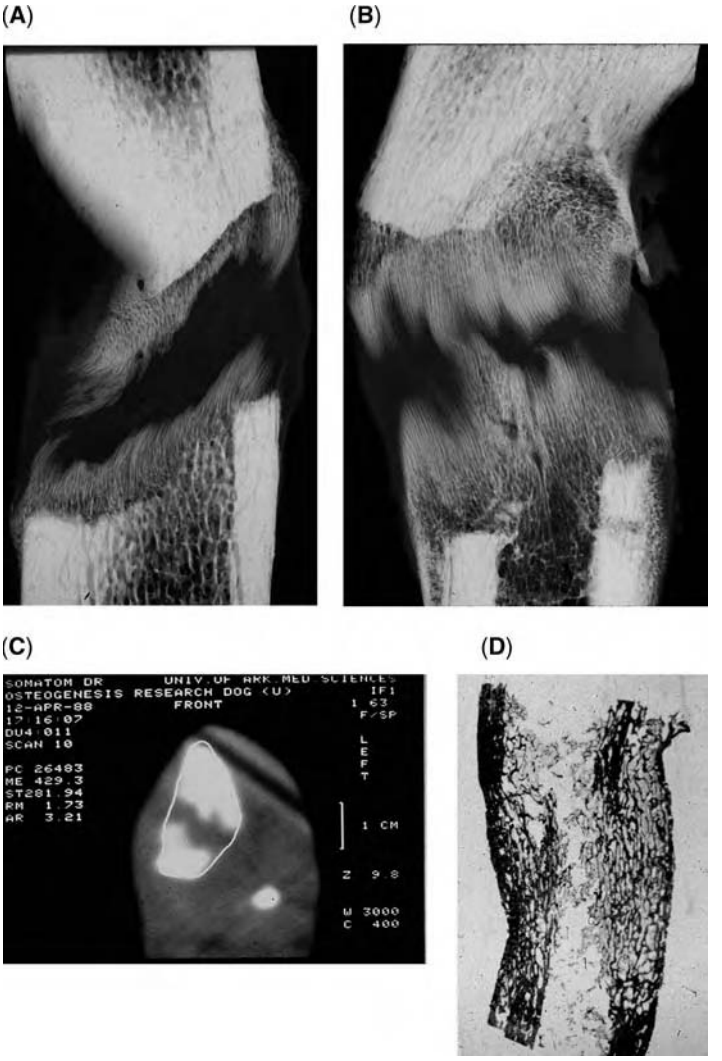

Figure 3 (**A**) Microradiograph of dog specimen at early bone formation from each distracted surface, projecting toward the central radiolucent, collagenous interzone FIZ. (**B**) Microradiograph of dog specimen at first bone bridge across the FIZ shortly after distraction had been discontinued. (**C**) Computed tomographic transverse scan across the distraction gap. The radiolucent zone is coursing obliquely through dense new bone formation during distraction and prior to consolidation or corticalization. The free hand region of interest drawn around the entire distraction gap provides the mean number of Hounsfield units per pixel within the gap. This measurement can provide objective data regarding distribution and density of bone formation. (**D**) This histological macrosection across the distraction gap in a dog specimen demonstrates early consolidation of the individual microcolumns of new bone during corticalization. *Abbreviation*: FIZ, fibrous interzone.

QCT-Based Finite Element Analysis Model

Because this tibial lengthening model produced an isolated segment of new bone with a wide range of apparent densities by prior gravimetric, calcium, and QCT analyses, it was felt to provide an ideal test for the accuracy of a QCT-based finite element analysis (FEA) model (20,26,38). Following the standard protocol for a 15% (28 mm) diaphyseal lengthening, one dog with radiographic union was selected for analysis. Forty-two days after fixator removal (day 119 of the experimental model), the dog was sedated and in vivo QCT scans were made of the newly formed bone zone and the corresponding zone on the contralateral leg using a dipotassium hydrogen phosphate solution phantom. The dog was then euthanized and both tibiae were harvested and macerated for mechanical testing (MTS Bionix^{TM} Test system, MTS Systems Corporation, Eden Prairie, Minnesota, U.S.A., info@mts.com). A clip-on strain gauge

that had been modified for a 25 mm gauge length was attached to each tibia and the assembly was loaded axially to two times body weight (400 N) in both tension and compression. A total of six strain gauge readings were obtained from each tibia along the anterior, medial, and lateral aspects of the tibiae spanning the zones 65 to 91 mm and 91 to 117 mm from the tibial plateau.

Numerical matrices of the QCT images were obtained and processed to determine the cortical geometry and material properties. The endosteal and periosteal surfaces were delineated by using 500 HU as the threshold value for cortical bone. Each QCT slice was modeled as a ring of eight three-dimensional plate elements using a FEA program.

The four highest QCT numbers in each segment were averaged; this was felt to decrease volume-averaging effects at the edge of the dense cortical areas without discounting the high stiffness of denser regions in the cortex. The QCT values were adjusted for differences in phantom readings. The distracted tibia mean QCT number was 1174 HU; the control tibia mean was 1476 HU. The average QCT numbers were then used to calculate the apparent density of each segment (39).

A finite element model (40,41) was constructed for both distracted and control tibiae by calculating the modulus of elasticity from the segment apparent density. Separate FE models of each tibia were constructed; one model (Model I) used the Carter and Hayes equation (42) and the other (Model II) used the Schaffler and Burr relationship (43).

An axial loading of 400 N in compression was applied to the actual bone and simulated in the FEA. Displacements were matched to the strain gauge locations during mechanical testing.

Biomechanical Testing In Vitro

As each dog for mechanical testing was euthanized, the tibiae were harvested for similar testing in an MTS Bionix^{TM} system. Each tibia was loaded axially at 8 N/sec to two times body weight (400 N), in six cycles of both tension and compression. A total of three strain gauge readings were obtained from each tibia along the anterior, medial, and lateral aspects of the tibiae spanning the osteogenic zone on the experimental side and at the same level measured from the tibial plateau on the control side.

Torsion testing of each tibia followed the axial test. Using the same steel mounts and a linear variable differential transformer, torsional stiffness was measured in Newton-meters per radian. External torque was applied in 2.5 N-m increments, up to a 10 N-m maximum. The experimental side was normalized to the control opposite side as a percentage for cross-group comparison.

In Vivo Load Measurements During Distraction

A subgroup of these 32 dogs was used to measure load readings during distraction in order to predict outcome (44). This group consisted of 21 randomly selected dogs with either metaphyseal or diaphyseal corticotomies and varying latency periods (0, 7, 14, or 21 days) between surgery and the start of distraction. At one-week intervals during the four-week distraction phase, the dogs were sedated and positioned supine on a holding frame. The external fixator was suspended so that the tibia was horizontal to eliminate gravity loads. The dial caliper was used to measure the baseline ring separation at the three distraction rod locations. Load cell assemblies were installed and the load transferred from the distraction rods to the load cell assembly by loosening the nuts on the distraction rods, which were left loosely in place to provide translational control between the rings. Load measurements were recorded (23,38,44) for one minute before distracting each rod by 0.5 mm. Loads were then measured for an additional 9 minutes. After the monitoring period, the load cell assemblies were removed and the rings reset to the proper position.

Differential Stress During Distraction Osteogenesis

Four dogs underwent a 30% (56 mm) left tibial lengthening (twice the standard experimental model used in all the other experiments) in order to measure loads during distraction of larger magnitude and to differentiate the effects of soft tissue resistance from that in the osteogenic zone (45).

Weekly radiographs and load measurements were made as described previously (Fig. 4A,B). Following a seven-day latency, distraction at 0.5 mm B.I.D. was carried out for

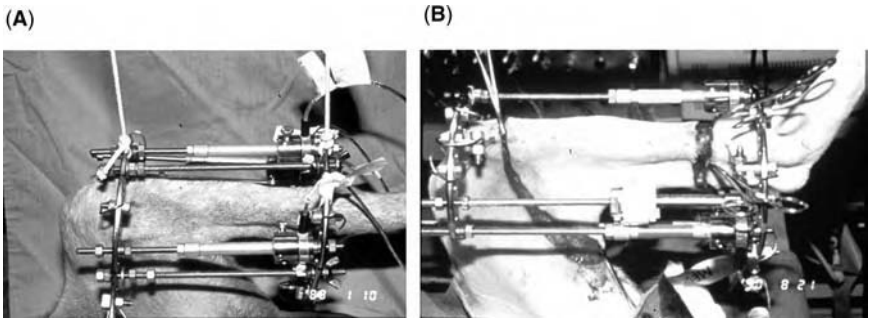

Figure 4 (**A**) This dog is under sedation while the frame is suspended horizontally to gravity for load measurement. Three load cells are inserted adjacent to the distraction rods, which are then loosened to test in vivo distraction loads. (**B**) This dog is undergoing sequential dissection under anesthesia with load cells in place to measure the relaxation of distraction loads as different soft tissue layers are surgically released: skin, fascia, muscle, and periosteum. The animal was sacrificed immediately following surgery for radiographic and histological analysis.

twice the normal period, 56 days rather than 28 for a 30% lengthening. On the last day of distraction, each dog was placed under general anesthesia with the external fixator suspended to hold the tibia parallel to the ground. Using sterile and hemostatic surgical technique, with strain gauges in place for ongoing load measurements, the lengthened leg was differentially dissected in order to observe the changes in resistance to the distraction load as each layer of soft tissue was released. In sequence, the skin, fascia, individual muscle groups, the fibula, and, finally, the periosteum, were circumferentially released until only the tibia itself remained with the external fixator and in-line strain gauges.

RESULTS

Rate and Rhythm Experiments

The initial experiments were based on the hypothesis that rate and rhythm are critical to distraction osteogenesis. The null hypothesis was tested first. Seven days following external fixation of a corticotomy in the proximal tibia, the entire distraction gap was created under sedation to see if new bone would bridge an acute 2-cm gap between the cut surfaces. All of these animals went on to an atrophic nonunion. This experiment reproduced the original method of limb lengthening reported by Codivilla at the turn of the century (12), which routinely resulted in an empty bone gap in his patients.

In order to further examine rate and rhythm, the next group again underwent the Ilizarov corticotomy but with sporadic daily distractions of 1.0 to 1.5 mm similar to the standard and currently accepted technique of limb lengthening developed by Wagner (13,14). More bone was produced in the gap, but again all trials resulted in a nonunion. Accordingly, the Wagner technique is routinely planned in three stages: to distract the osteotomy, to graft the defect with autograft and internal plate fixation, and then years later, to remove the plate when all of the graft is incorporated and remodeled.

When the corticotomized fragments were distracted at the rate and rhythm recommended by Ilizarov, 0.25 mm every six hours, all bones bridged and went on to remodel to normal lengthened bone. Using whole bone gravimetrics to confirm that new bone was actually added, the average 12% (26–28 mm) lengthening produced an average 27% increase in mass and an average 26% increase in volume for a minimal change in whole bone density (29). Chemical analysis of the newly consolidated bone demonstrated that averages of water 15%, lipid 5%, calcium 25%, phosphorus 12%, and collagen 24% with calcium:phosphorus ratios of 2.1 and calcium:collagen ratios of 1.1 were similar to those measured in normal bone (20,28,46–48).

Rate was further tested by comparing two groups of dogs distracted at 1 versus 2 mm/day. All animals in both groups bridged the gap and remodeled to normal appearing bone; however, noninvasive monitoring techniques QTS detected a significant decrease in blood flow at week 3 and QCT detected a significant decrease in mineralization at weeks 4 and 5 in the more rapid, 2 mm/day group (36).

Distraction osteogenesis produced by the Ilizarov corticotomy, latency, and rate and rhythm was then tested for fixator specificity by comparing the tensioned-wire ring external fixator modeled after the Ilizarov device, used in all previous experiments, to the standard half-pin fixator used by Wagner (21,49–51). The radiographic and histological appearance of new bone formation was identical, except that the medially placed half-pin device tended toward uncontrolled angulation (valgus) during the distraction process.

When the external fixator was unintentionally destabilized (e.g., untensioned wires, loose pins, or flexible frame), the gradual distraction produced bone that either angulated (malunion) or failed to bridge (nonunion).

Histology

Histological preparations were made by biopsy and by whole bone sectioning in the coronal and transverse planes (Fig. 5A–D).

The initial latency period appeared to be no different than routine fracture healing, as might be expected. Hematoma and inflammatory cell infiltrates filled the gap at the corticotomy site.

After the start of distraction, mesenchymal-like cells began to organize a bridge of collagen and immature vascular sinusoids. As distraction proceeded, the fibrovascular bridge seemed to organize itself parallel to the direction of distraction (21). The collagen network became denser and less vascular, almost resembling tendon, while the vascular channels

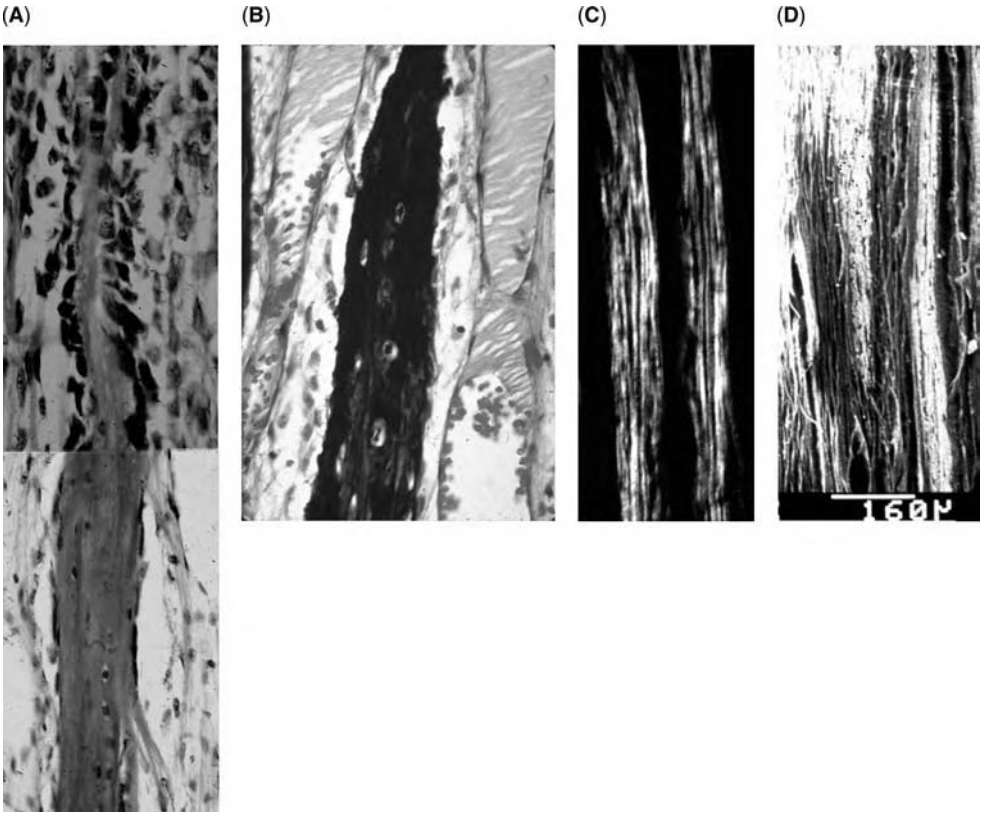

Figure 5 (**A**) This composite histological preparation demonstrates how collagen is incorporated into each individual bony trabeculum during the transition from the fibrous interzone to primary mineralization front to microcolumn formation (top to bottom). Note how collagen is added along the surface as osteoblasts are incorporated within the bone matrix become osteocytes. Similar to naturally occurring trabecular bone, each microcolumn reaches a uniform diameter of approximately 150 microns because it does not contain haversian canals. (**B**) Each microcolumn is surrounded by large vascular sinusoids for nutrition by diffusion. In this case, osteogenesis is closely linked to angiogenesis. (**C**) Polarized light microscopy demonstrates uniform incorporation of collagen bundles within each of the two microcolumns pictured, all parallel to the distraction force. (**D**) Back-scattered scanning electron microscopy demonstrates mineralized columns of new bone surrounded by loose collagen and vascular spaces.

remained at the proximal and distal edges closely approximated to the cut surfaces of the corticotomy segments. During the first week of distraction, this central zone of hypovascular fibrous tissue bridged the entire 6 to 7 mm gap (26). This region has been named the FIZ to signify that the two bone surfaces are actually bridged by collagen. Spindle-shaped cells resembling fibroblasts were loosely interspersed between collagen bundles; neither osteoid nor osteoblasts were present. Bone mineral was distinctly absent by von Kossa staining and back-scattered scanning electron microscopy of nondecalcified specimens. Cartilage islands were rarely seen, contrary to the large collections of subperiosteal cartilage routinely found during normal fracture healing.

During the second week of distraction, osteoblast-like cells appeared in clusters adjacent to the vascular sinuses on either side of the FIZ. Collagen bundles became fused with pink staining matrix-resembling osteoid by routine staining of decalcified specimens. The osteoblastic cells initially rested on the surface of these primary bone spicules, and eventually became enveloped within, because the spicule gradually enlarged by circumferential apposition of collagen and osteoid (5,21). By the end of the second week, the osteoid began to mineralize. These early bone spicules could be described as the PMF. The mineralization within the columns was confirmed by von Kossa staining of the nondecalcified specimens and by X-ray microanalysis (5,27). This osteogenic process was seen uniformly covering the entire cross-section of the cut bone, including periosteum, cortex, and medullary spongiosa (5) by microradiography. The microcolumns extended from each corticotomy surface toward the central FIZ-like stalactites and stalagmites (26), as visualized by back-scattered scanning electron microscopy.

From the third week on, this process continued with the FIZ undulating across the center at average thicknesses from 4 to 8 mm. As the distraction gap increased, the bridge was perpetuated by the elongation of the new bone spicules. These bone spicules were actually longitudinally connected by the collagen fibers as demonstrated by routine and polarized light microscopy (5). The tips of the spicules began at a diameter of approximately 7 to 10 microns and rapidly expanded to diameters of up to 150 microns toward each corticotomy surface (5,21,26). Each microcolumn of new bone was surrounded by large thin-walled sinusoids (5). The columns were devoid of Haversian canals. No cartilage or osteoclasts were seen. These regions on either side of the FIZ can be described as the zones of MCF.

At the conclusion of distraction, the FIZ ossified, creating one zone of MCF, completely bridging the gap. Some cartilage islands formed centrally in areas more than 300 microns from local vessels (5). This cartilage formation was rarely seen if distraction was carried out at the proper rate and rhythm and the bone fixation remained stable. Cartilage interposition was found in sites with delay of the osseous bridge, usually seen when the local biology had been traumatized by instability of fixation. This phenomenon was noted in specimens with recognized pin loosening at the bone or fixator.

In this experimental model, the frame was removed six weeks after the four-week distraction period. During this six-week ''consolidation period,'' the dogs usually resumed weight bearing.

During the six weeks following frame removal, the osteogenic area remodeled into cortex and medullary canal. The bony columns took on the staining characteristics of mature lamellar bone with cement lines and smaller osteocytes resting in lacunae. The fibrovascular tissue that filled the spaces around bone columns was replaced by normal appearing marrow elements. Normal osteoclastic remodeling was present.

Blood Supply

Histologically, each column of new bone was completely surrounded by large vascular sinusoids. The appearance of clusters of osteoblasts at the tip of each column was in close proximity to these sinusoids (5). India ink injection studies with Spalteholz clearing technique demonstrated that these vessels paralleled the bone columns and the distraction force, but very few vessels actually crossed the FIZ, which remained relatively avascular (5). These vessels were fed from both endosteal and periosteal sources from each corticotomy surface.

Quantitative Technetium Scintigraphy

The preoperative baselines demonstrated left–right equality within a relatively large range of 25% variation while the postoperative increases always exceeded 100% of baseline. The opposite

leg was noted to decrease its uptake from the preoperative baseline during all future scans during the experiment, a so-called "steal effect" where the massive increase in flow to the experimental limb sequestered enough of the isotope so as to decrease the scan uptake on the normal unoperated limb. This steal effect was quite consistent in all animals throughout the experiment, so the data was felt to be consistent and normalized by a left/right ratio.

In the first group (1 mm/day × 28 days), the average flow phase within the distraction area increased by eight times the opposite, peaking at week 2 of distraction and then decreasing to six times the opposite (week 3), five times the opposite (week 4), and reaching a plateau at three to four times the opposite until sacrifice at week 17. In the second group (2 mm/day × 14 days), the average flow phase within the distraction area increased by nine times the opposite, peaking at week 2 and then decreasing to four times opposite (week 4), and finally falling to two times opposite by week 17. In the third group (1 mm/day × 56 days), the data for prolonged distraction measured the blood flow by this technique well above the opposite at 11 times (week 3) and at 5.5 times opposite (week 7), but the temporal decrease paralleled the first two groups (Fig. 6). By a Wilcoxon-two sample test, all values during distraction were significantly greater than the control ($p = 0.05$). None of the increases measured between these three groups were significantly different at any one point in time.

When both the distracted left tibiae (metaphyseal sites) and control right tibiae were divided into proximal and distal halves for comparison of relative blood flow, the proximal halves were always greater than the distal halves at each time point from preoperative baseline until week 17. The proximal to distal ratios ranged from 1.6 to 5 times, averaging three times greater, even in the unoperated leg (36).

The blood flow measured in the distal half of the left tibiae (a regionally distant site in the same bone from the distraction osteogenesis zone in the proximal metaphysis) was always increased compared to the right opposite control. The mean ratios of the distal tibiae were calculated for all dogs from the first two groups. The peak flow of seven to eight times control, even at this distant site, occurred at weeks two and three reaching a plateau at two to three times control from weeks 4 through 17 (36).

One dog from each of the first two groups went on to nonunion. The one nonunion from the first group demonstrated the same pattern of increased blood flow measured in the other dogs that went on to union; the other nonunion from the second group had only a moderate increase in blood flow that was consistently below the average for the group and for all dogs tested; the difference was not significant, however.

Standard Radiography

Standard radiography provides a good weekly or biweekly check on the progress of the distraction gap (length and alignment) and usually by the third week of distraction, new bone

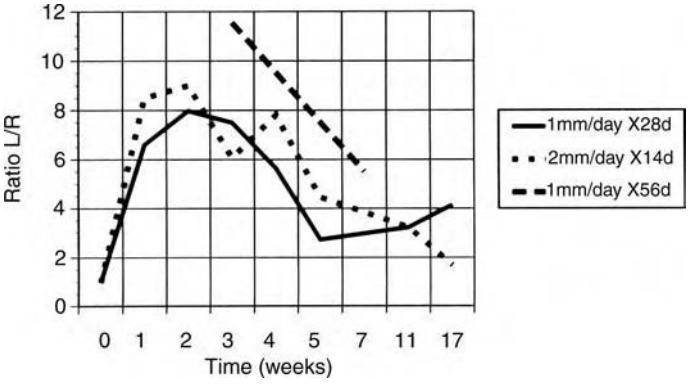

Figure 6 Quantitative technetium scintigraphy was used to measure regional blood flow over time during different distraction conditions (rate and duration). A massive increase in blood flow (about 8 × normal baseline) persists for the first several weeks, which seems necessary for coupled osteogenesis and angiogenesis during distraction osteogenesis and may account for the tendency to edema of the limb when not actively exercised. Prolonging the distraction duration did not prolong this vascular effect, which may be related more to the operative intervention (similar to fracture healing) than to distraction.

mineral appears as fuzzy, radiodense columns extending from both cut surfaces toward the center (20,22,52). As distraction proceeds, the central FIZ remains as an undulating radiolucent zone, between 4 and 8 mm wide, while more and more new bone is added from each end (26). The new bone spans the entire cross-sectional area of the HBS on both orthogonal views.

Metaphyseal Versus Diaphyseal Corticotomy Sites

All 32 dogs survived the experiment (22). Of these, 23 had successful lengthening to the predetermined length (72%) and nine fused prematurely (28%). Of the 32 lengthenings, four resulted in nonunion (12.5%). Among 16 metaphyseal lengthenings, there were six premature consolidations (37.5%), all four with a 21-day latency and two with a 14-day latency, and one nonunion (6.2%). In the 16 diaphyseal lengthenings, three fused prematurely (18.7%), two with 21-day latency and one incomplete corticotomy with seven-day latency, and three resulted in nonunion (18.7%). All animals, both metaphyseal and diaphyseal, successfully bridged the distraction gap after a zero-day latency. The average length gained (s.d.), excluding premature consolidation, was 23.9 mm (3.7) in metaphyseal and 23.8 mm (2.0) in diaphyseal lengthening.

Evaluation of roentgenograms showed that in metaphyseal sites, the first new bone was seen seven days earlier, the first bone bridge, 10 days earlier, the first cortex, five days earlier, and the first canal formation, six days earlier than in the diaphyseal sites.

Although the bone healing index averaged six days earlier in metaphyseal than in diaphyseal sites, no significant difference was found by *t*-test. Comparison of the latency groups showed that new bone consolidation occurred soonest at a zero-day latency in both metaphyseal and diaphyseal lengthenings, with a significant difference between zero-day and seven-day latency groups ($p = 0.01$).

The minimum QCT density ratio of the experimental to the contralateral side, excluding the nine premature consolidations, decreased during distraction, reaching a minimum at the end of distraction, and then increased gradually during the consolidation period (22). Among the subgroups, no significant difference was found at preoperation or at the start of distraction. When the metaphyseal ($n = 10$) and diaphyseal ($n = 13$) lengthenings were compared, significant differences in the minimum QCT were found at the end of distraction ($p = 0.001$), at fixator removal ($p = 0.001$), and at sacrifice ($p = 0.04$), with the diaphyseal sites always lower.

Biomechanical Testing In Vitro

Fourteen pairs of tibiae ($n = 28$) were tested in cycles of axial tension and compression, and in torque. All the experimental tibiae underwent a corticotomy followed by external fixation for 11 weeks and full weight bearing (out of external fixation) for an additional six weeks, totaling 17 weeks from the index operation. These groups of tibiae were randomly chosen from the metaphyseal–diaphyseal latency experiment. The mechanical testing group had seven unions (six diaphyseal and one metaphyseal), six premature consolidations (five metaphyseal and one diaphyseal), and one nonunion (diaphyseal). All the premature consolidations underwent attempted distraction for several days, but once corticotomy healing was diagnosed by failure of the radiographic gap to develop, distraction was discontinued; this group consequently represented healing of a low-energy fracture for comparison to the distracted tibiae.

Axial stiffness was represented by the slope of a linear regression line and expressed in kilo-Newtons per millimeter. The control tibiae ($n = 14$) averaged 51.3 KN/mm. The six premature consolidations (fracture controls) averaged 35.5 KN/mm (69% of control) while the seven distracted tibiae averaged 24 KN/mm (47%) with the highest values averaging 32 KN/mm (62%) in two diaphyseal lengthenings with 7- and 14-day latencies. The one nonunion was tested only axially in tension because the interface was soft, measuring 0.7 KN/mm (1.4% of control).

Torsional stiffness was similarly calculated at an average of 395 N-m/rad in the controls, at an average of 379 N-m/rad (96%) in the premature consolidations (fracture controls), and at an average of 169 N-m/rad (43%) in the distracted tibiae. The two best diaphyseal lengthenings at 7- and 14-day latencies averaged 314 N-m/rad (83%).

QCT-Based FEA

The predicted axial displacements in compression from the finite element models were compared with the measured displacements in compression using a simple regression (38).

Both models (I and II) of the control and distracted tibiae had significant correlations when the displacements from both tibiae were pooled together ($n = 12$). The Carter and Hayes formulation had a higher correlation coefficient ($r^2 = 0.671$) than the models based on Schaffler and Burr ($r^2 = 0.423$). When the displacements were correlated by tibia ($n = 6$), the Carter and Hayes formula (Model I) predicted the displacements better in the distracted tibia ($r^2 = 0.914$) versus Model II ($r^2 = 0.654$); the Schaffler and Burr formula (Model II) predicted displacements better in the control tibia ($r^2 = 0.820$) compared to Model I ($r^2 = 0.652$).

Load Measurements During Distraction

The clinical outcome of the lengthening procedure was determined from examination of the last radiograph prior to sacrifice (44). Observer bias was prevented by using an investigator to analyze the radiographs, who was unaware of the load measurement results. An outcome was judged to be a nonunion if there was a continuous radiolucency in the osteogenic area. An outcome of malunion was assigned if angular deformity ($>5^\circ$) existed without signs of a nonunion.

The variables for each subject were CT measurements of baseline tibia length, osteogenic zone area, soft tissue area at the osteogenic zone, latency from surgery to start of distraction, and distraction loads at weekly intervals. This data was grouped by both osteotomy site and outcome, and then analyzed using a biomedical statistics package (BMDP Statistical Software, Inc., Los Angeles, California, 90025). The BMDP programs used were 3-D (*t*-tests), 1R (Linear Regression by Groups), and 5V (Unbalanced Repeated Measures Models with Structured Covariance Matrices).

Six dogs had premature fusions early during the distraction phase. These were easily discerned on the plain radiographs. Loads were measured in these subjects within the safety limits of the load cells so as not to overload the gauges and endanger the instrumentation system.

Fifteen dogs completed the distraction load measurement phase of the study. Diaphyseal ($n = 40$) and metaphyseal ($n = 20$) observations from 15 subjects were divided evenly over the four observation periods. One animal died of lymphosarcoma (unrelated to the experimental protocol) prior to the scheduled sacrifice. Fourteen dogs completed the entire protocol to final bone healing for outcome assessment. The clinical outcomes of these 14 dogs were assessed radiographically prior to sacrifice at day 119 after surgery. There were 11 unions and 3 nonunions at the completion of the study. Two of the unions developed angular deformity that was felt to be secondary to premature removal of the fixator.

No significant difference was found by *t*-test in the baseline tibia length when grouped by corticotomy site, metaphyseal versus diaphyseal ($p = 0.6424$). There was a significant difference between osteogenic cross-sectional areas ($p = 0.0138$) as calculated by CT scans; the mean (and standard deviation) of the metaphyseal sites were $3.31\,cm^2$ (0.57) and of the diaphyseal sites were $2.34\,cm^2$ (0.43).

The mean (and standard deviation) of the weekly load measurements were grouped by corticotomy site. Diaphyseal ($n = 40$) and metaphyseal ($n = 20$) observations from 15 subjects were divided evenly over the four observation periods. The mean load at the end of distraction was 155 N for metaphyseal ($n = 5$) and 111 N for the diaphyseal ($n = 10$) sites, respectively.

That the load increased steadily over time from surgery was highly significant ($p < 0.0001$). The effect of corticotomy site on load was also significant ($p = 0.0207$).

A linear regression for all dogs tested based on the distraction loads versus time was significant, $R^2 = 0.55$ ($p < 0.0001$). An analysis of variance of regression coefficients over groups was highly significant ($p = 0.00161$) indicating that the slopes of the regression lines between metaphyseal and diaphyseal sites are statistically different for measured loads over time.

The individual distraction loads were then divided by the respective cross-sectional area of the osteogenic zone in each subject. The resulting quantity was called the ''osteogenic zone stress.'' The regression analysis on this data increased significantly with time, $R^2 = 0.50$ ($p < 0.0001$). An analysis of variance of regression coefficients between metaphyseal and diaphyseal groups was not significant ($p = 0.87$), indicating that there was no statistical difference in the slopes of the regression lines for calculated stress.

When loads were compared to latency periods, a trend to higher loads over time developed with longer latencies in both the metaphyseal and diaphyseal groups. When the

serial in vivo radiographs were used to measure the thickness (linear length) of the central radiolucent zone (FIZ), a significant inverse relationship was found such that higher loads were measured with the narrower FIZ. Over time, the FIZ narrows, even during distraction. By plain radiography, the corresponding radiolucent zone appears to increase until week 3 (week 2 of distraction) at which time the FIZ narrows even during further distraction. Widening of the FIZ is accentuated at the faster 2 mm/day rate. The loads increased despite the early increase in the radiolucent zone because radiographic evidence of mineralization lags behind that in histology and QCT.

There was a significant difference in the ultimate tibial length ($p = 0.0292$) when grouped by outcome. The mean lengths and standard deviations were 175 mm (5.24) and 185 mm (9.70) for the unions and nonunions, respectively. The three nonunions had osteotomies at the diaphyseal site. There were no significant differences in latency, osteogenic zone area, or soft tissue area. A linear regression of the load readings demonstrated a significant difference in the slopes ($p < 0.0023$) between unions and nonunions. When compared by t-test, there was a significant difference in loads between unions and nonunions at the end of the third week ($p = 0.044$) with the unions averaging 47 newtons per cm^2 and the nonunions averaging 27 newtons per cm^2. There was even a suggestive change as early as the end of the second week ($p = 0.056$) (Fig. 7A–C).

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The loads continued to rise over time throughout the 56-day experiment (45). The major resistance to distraction was determined to be in the distraction zone itself. Each of the soft tissue

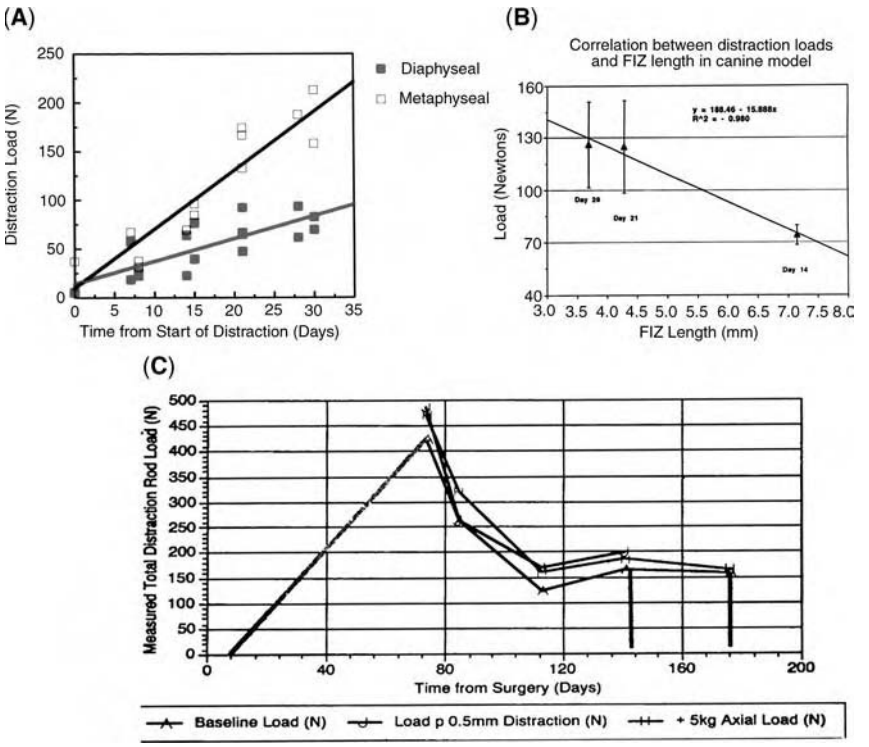

Figure 7 (**A**) In vivo load testing is compared over time during distraction between metaphyseal and diaphyseal distraction sites. The metaphyseal sites required higher distraction loads, which correlated directly to the cross-sectional areas of new bone formation using computed tomography scans to measure the area. (**B**) In vivo loads were inversely related to the length of the radiolucent zone FIZ measured on radiographs, reflecting the different modulus of elasticity between collagen and mineralized bone. As distraction proceeded over time, the radiolucent zone decreased until a homeostatic range of 2 to 4 mm was reached prior to bridging at the end of distraction. (**C**) When a patient was subjected to weekly load measurements, the loads rose during distraction, fell off to a plateau during neutralization (consolidation), but did not fall to zero until the frame was completely dynamized. *Abbreviation*: FIZ, fibrous interzone.

layers (skin, fascia, muscles, and periosteum) accounted for 4% to 7% of the load; the bone (osteogenic area) itself still contained 72% of the baseline load.

DISCUSSION

Histology

The histological sequence of events during distraction osteogenesis, though not unique, is essential to understand before correlating the mechanical influences, imaging studies, and clinical management. Under ideal conditions, a low-energy osteotomy preserving blood flow to each apposed surface, distracted at a regular, incremental rate of 1 mm/day by a stable external fixation system, will reliably regenerate a new bone segment that fills the physical gap with normal bone of similar macro- and microstructure. The majority of published experimental work (25,53–56) including that of Ilizarov (1–4) supports the findings in this paper that under these conditions, bone forms by intramembranous ossification. Some researchers have found more cartilage-like cells and matrix comprising the microcolumns (57), consistent with chondroid bone formation (58). These experimental models varied techniques from those described here, using an oscillating saw osteotomy and different latency, rate, and rhythm. The oscillating saw probably creates thermal necrosis, altering the local biological process (54).

Prior to distraction, the histology reflects expected patterns of fracture healing. During distraction, the local histology transforms into five zones: a central FIZ with immature collagen bundles and fibroblast-like cells arranged parallel to the distraction force spanning the entire cross-section of the bone gap and ranging from 4 to 8 mm in length; two adjacent zones where clusters of osteoblast-like cells produce an osteoid-like matrix, which consolidates the collagen into longitudinal microcolumns with nearby capillary buds, termed the PMF; and adjacent to the PMF bridging to each HBS are the zones of MCF where the primary bone units begin to mineralize, expand to maximal diameters of 150 to 200 microns, and cross-link to each other, each surrounded by vascular sinusoids, all spanning the entire cross-section of the gap. Following distraction, the bridge consolidates and remodels. The PMF traverses the FIZ followed by the MCF. Osteoclastic remodeling of a normal medullary canal, condensation of the peripheral MCF into cortex and replacement of fibrovascular channels with bone marrow or fatty elements occurs late in the consolidation process.

Some clinicians using the Ilizarov method refer to this area of bone regeneration as a ''growth plate,'' and in the sense of new bone formation, it is; however, histologically, distraction osteogenesis is intramembranous ossification in its purest form. This direct appositional bone growth, with osteoblasts laying collagen and osteoid, which is then mineralized, is seen in normal bony remodeling; it is also seen with periosteal new bone. In fact, the only part of a growth plate that resembles distraction osteogenesis is at the zone of Ranvier where periosteum is stretched across the physis (59). The relationship between periosteal new bone and distraction osteogenesis is histologically identical, which lends indirect evidence to the historical hypothesis that periosteum is stretched from the epiphysis to the metaphysis by the expanding endochondral sequence of the physis, similar to the external fixator stretching the FIZ to stimulate new bone formation during distraction osteogenesis.

Cartilage and endochondral sequence has been noted in some specimens when the local blood supply has been disrupted or when bone-fixator instability exists. The endochondral sequence of ossification seems to occur much more slowly and residual islands may persist indefinitely. In all of these experimental canine tibial lengthenings as well as those reported by others (32), and in two human biopsies on my patients, intramembranous ossification has been the predominant finding.

The linear rate of distraction osteogenesis (length of new bone segment per day of external fixation time) averages about 300 microns per day. The adolescent distal femoral physis adds new bone at a linear rate of 50 microns per day and the fetal femur has been estimated to grow at a linear rate of 400 microns per day. Distraction osteogenesis approaches the growth rate of the fetal femur.

Rate and Rhythm

A specific distraction rate of 1 mm/day at a regular rhythm of 2, 3, 4, or more increments per day similar to the conditions postulated by Ilizarov has been confirmed by this work to be

critical for successful bone regeneration in the distraction gap. Excessive rates (greater than 2 mm/day) seemed to outstrip the local angiogenesis and blood flow necessary for the biological process of osteogenesis. Lesser rates allowed premature bony consolidation of the (collagenous) FIZ, thus bridging the bone fragments with more rigid material that prevented further distraction. At rates faster than 2 mm/day, experiments did not produce new bone in the distraction gap, reconfirming the findings of both historical and traditional methods of limb lengthening utilized by Codivilla (12) and Wagner (13,14). Sunderland (15) has reported on several experiments indicating that peripheral nerves regenerate axons at a rate of 1 mm/day. Linear growth of bone tissue by this process is dependent upon several histological components: a central bridge of moderately loose collagen bundles parallel to the distraction force that remains at a minimum length of 4 to 8 mm during active distraction; peripheral zones of uniform microcolumns of new bone each of which is circumferentially surrounded by budding sinusoidal vessels, all of which parallel the distraction force; and regions of cell proliferation that allow for such rapid tissue growth. If the distraction rate is too rapid, inadequate tissue regeneration due to finite limits of local cell growth, insufficient delivery of nutrients by local vessels, and actual disruption of the biological bridge most likely inhibits the osteogenic process. The measured increase of the distraction gap in nonunions over unions lends some evidence to an actual disruption of the collagenous bridge leading to an eventual nonunion. This effect may also be seen in the more rapid 2 mm/day rate where the radiolucent zone significantly widened.

Latency

Because metaphyseal blood flow far exceeds that measured in the diaphysis of the normal canine tibia at all ages (60), it was hypothesized that bone formation in the diaphyseal region would be enhanced by a longer latency than that in the metaphyseal region. The results of the latency experiment surprisingly indicate that the latency period did not enhance bone formation in the diaphyseal site; in fact, diaphyseal bone formation equaled that measured in the metaphysis at both zero- and seven-day latency. Longer latencies of 14 and 21 days carried a high risk of premature consolidation preventing distraction osteogenesis, greater in the metaphysis than the diaphysis. These findings differ from a similar study by Kenwright (25) using a half-pin monolateral frame. The difference between this monolateral fixator and the tensioned-wire ring fixator involves thin wires, which increase their rigidity as they deflect (49,50). This wire deflection effectively delays the actual bone separation for three to five days so that the latency at the bone level was actually longer in each group as bone separation lagged behind the ring separation, which is more consistent with Kenwright's work.

Blood Supply

Physiologically, the regional and local blood supply is probably the most consistent requirement for new bone formation (61). Research using a combination of angiography, histology, India ink injections, and QTS substantiates this assertion. Technetium scintigraphy of the osteogenic area is intensely hot with a central cool area corresponding to the FIZ. Using the method of QTS to automatically outline the hottest 40% of the distracted tibia and measure the average number of counts per pixel in this region of interest (36), the initial or flow phase of the technetium scan was correlated to blood flow (62). During the four weeks of distraction, the experimental side peaked at eight times the normal side and then decreased to three times normal for at least the next three months. The temporal pattern of blood flow increased as measured by QTS paralleling that of a similar study for simple fracture healing, although the actual values during distraction osteogenesis were twice those of the fracture model (37). Although the absolute numbers during distraction osteogenesis were higher than during fracture healing, probably related to experimental technique, the temporal patterns were nearly parallel. These studies would imply that blood flow increases related to distraction osteogenesis are more closely related to the injury-repair response than to a regenerative response because the effect is not temporally extended by prolonged distraction.

Pathophysiology

Certain conditions documented from experimental animals and clinical patients that reliably lead to poor osteogenesis are excessive rate, sporadic rhythm, initial diastasis, frame or

bone-fixator instability, inadequate consolidation period, poor regional or local blood supply, and a traumatic corticotomy (5,8). The massive increase in regional blood flow measured experimentally must still circulate evenly on the microscopic level as described in the histology section. The orderly zones of bone formation seen in normal distraction osteogenesis involve collagen deposition, osteoid formation, and mineralization. It is easy to postulate that an initial diastasis between the cut bone surfaces would inhibit the formation of a primary fibrovascular bridge, which seems essential to transmit distraction force to the tissue level. External fixation frame instability results in macromotion, especially shear forces that could disrupt the delicate bone and vascular channels. The regulatory importance of rate and rhythm may well involve the biosynthetic pathways on the cellular level by rate-limited steps such as protein synthesis and mitosis, especially those involved in angiogenesis. If the host bone suffers from inadequate vascularity initially, then the regional hyperemic response to the corticotomy may be insufficient to drive the local biology. A systemic process such as diabetes, smoking, or peripheral vascular disease may limit vascularity or a traumatic corticotomy could severely disturb the local flow. In clinical practice, patients with these conditions have been more likely to result in nonunion (63). Biopsies from sites of failed osteogenesis reveal ischemic, atrophic fibrous tissue when the cut bone surface is devoid of osteocytes in lacunae and red cells in vascular spaces (7). If the bone ends are initially separated more than a centimeter or distracted too quickly, then islands of cartilage proliferate in the gap (5). Gaps and motion (64) have traditionally been implicated in the formation of nonunions and pseudoarthroses. Even after successful osteogenesis, remodeling to lamellar bone and normal macrostructure is necessary for normal weight bearing. Experimental destabilization of the frame prematurely has also led to late breakdown of the microcolumns and subsequent replacement with fibrocartilaginous nonunion (22).

Noninvasive Monitoring

During the process of distraction osteogenesis, it is clinically helpful to assess the progress of bone formation. Earlier on, the surgeon might adjust the length of latency to enhance the osteogenic potential. During distraction, rate or rhythm adjustments may be necessary to optimize osteogenesis. During consolidation, it is important to know when the osteogenic area is strong enough to remove the fixator.

The absence of new radiodensity by the third week of distraction is cause for concern. The findings of histology and chemical analysis correlate to the QCT graduated increase in mineralization from each HBS toward the central radiolucent zone. The ''sudden'' appearance of radiodense projections by week 3 (week 2 of distraction) is an artifact of plain radiography, where an estimated increase of 40% in radiodensity is necessary to visualize a change on a routine radiograph. Ultrasonic examination is sensitive to mineral depositions within cartilage prior to appearance on plain radiographs confirmed by the large, clinical experience in the neonatal hip. Unfortunately our experience with ultrasound during distraction osteogenesis was not as reliable. Vascular channels are also echogenic and may mimic the new microcolumns of bone. The high-resolution probe may not fit easily between rings even with silicone spacers. Circumferential exams are usually obscured by the rings, rods, and anatomy. Occasionally, the distraction gap appears empty by ultrasound, indicating a cystic cavity. In this rare instance, distraction should cease and the gap should be gradually closed until the corticotomy surfaces engage for a repeat latency prior to redistraction.

The osteogenic gap should demonstrate radiodense projections of new bone extending parallel from each osteotomy surface toward the central radiolucent zone FIZ. If the new bone segment appears to be bulging on its surface and the thickness of the radiolucent zone FIZ is narrowing, then osteogenesis is proceeding too rapidly, risking premature consolidation and the distraction rate should be accelerated. If the new bone forms an hourglass appearance, the FIZ is widening, then osteogenesis is proceeding too slowly, risking nonunion; the distraction rate should be decelerated. These structural alterations in the radiographic shape are indirect indications that strain within the osteogenic tissues is related to the biological outcome, as if a rubber band were stretched, becoming thinner (Poisson effect).

QCT is a developmental technique (65–68) that was used extensively in these experiments. The histological zones of distraction osteogenesis created a predictable pattern of mineralization, which was actually measured with QCT (26). The average number of

Hounsfield units per pixel in a freehand region of interest drawn around the perimeter of the osteogenic area was computer generated at each level in a series of transverse cuts spanning the distraction gap. These values were reproducible with minimal interference by the connecting rods or aluminum telescopic rods (69). Heavy metal such as rings, wire fixation bolts, or the steel head of the clicker units will cause significant interference, so measurements in these areas should be avoided (69). When compared to a similar anatomical region on the contralateral, normal side, the average QCT density was converted to a percentage of normal. The FIZ averaged about 25% to 35% of normal, the PMF usually rises to about 40% to 55% of normal, and the MCF remains at about 60% to 75% of normal during distraction (26). These values are roughly supported by a similar increase in the calcium to collagen ratios measured from the different zones experimentally. The chemical analysis of specimens taken from the different radiologic–histologic zones reflected the QCT density in the corresponding zones. Calcium was measured from dried, defatted bone (micrograms of calcium per milligram of bone) and expressed as a percentage of the normal contralateral bone: FIZ 30%, PMF 40% to 50%, and the MCF 60% to 70% (26). If this uniform sequence was present by QCT, then distraction osteogenesis proceeded normally, despite concurrent radiolucency on plain films.

When new bone mineral cannot be demonstrated by plain radiography or QCT, then the triphase technetium bone scan may be useful. Based upon experimental evidence, the distraction gap should be very hot in all three phases (5,36). It is important to discern that technetium uptake is increased on both sides of the osteogenic gap. In cases of poor vascularity or traumatic corticotomy, a triphase bone scan can also be performed after the predicted latency to confirm that both sides of the corticotomy have adequate flow. If the scan is cold, then distraction must be discontinued and the local problem carefully assessed. Arteriography, which is rarely indicated in these circumstances, can be helpful preoperatively if congenital or posttraumatic variation in the major arteries is suspected. In these cases, special care can be taken to avoid injury to the critical vessels.

During consolidation, plain radiography can be obtained on a monthly basis until the osteogenic area has cortex and medullary canal on orthogonal views. Despite the appearance of these radiographic findings, the overall bone density may be significantly reduced. QCT is helpful to demonstrate quantitatively that the new bone is strong enough by mineral density and cross-sectional distribution (38).

Biomechanical Testing In Vitro

The addition of a segment of new bone, averaging 26% mass and volume, to the adult canine tibia took 11 weeks treatment time (in external fixation) and a total of 17 weeks from the index operation (corticotomy). This new segment of bone was capable of normal weightbearing. The mechanical stiffness measured 47% axially (compared to 69% for the fracture controls) and 43% torsionally (compared to 96% for the fracture controls). Because all of these animals ambulated without limp, pain, or radiographic collapse of the tibia, it is postulated that the measured stiffness, though significantly lower than normal, is sufficient for normal weightbearing activities. The final time period until remodeling to normal bone stiffness is yet to be established.

Strain Gauge Testing In Vivo

The in vivo load measurements statistically support earlier case reports that suggest distraction loads increase with time (70,71) and vary with corticotomy site (25,56). We hypothesized that this load is directly related to distraction of the collagenous bridge central to the osteogenic interzone. The measured loads were converted to stress by dividing the osteogenic zone area into the load. When metaphyseal stresses were compared to diaphyseal stresses by linear regression, there was no longer a statistically significant difference between groups and the regression lines for the two groups were essentially parallel and collinear.

These findings are consistent with our previous assumption that the FIZ carries the primary distraction load and therefore the cross-sectional area of this zone is directly proportional to the load. It is postulated that the width or thickness of this zone (length of unmineralized collagen) is indirectly proportional to the load. As distraction proceeds, the collagen matrix becomes progressively more mineralized and therefore stiffer. More of the resistance is thus concentrated on shorter lengths of collagen, giving rise to higher loads over time. The load increment seems to decrease with time as the FIZ reaches a stable length. A proper range of

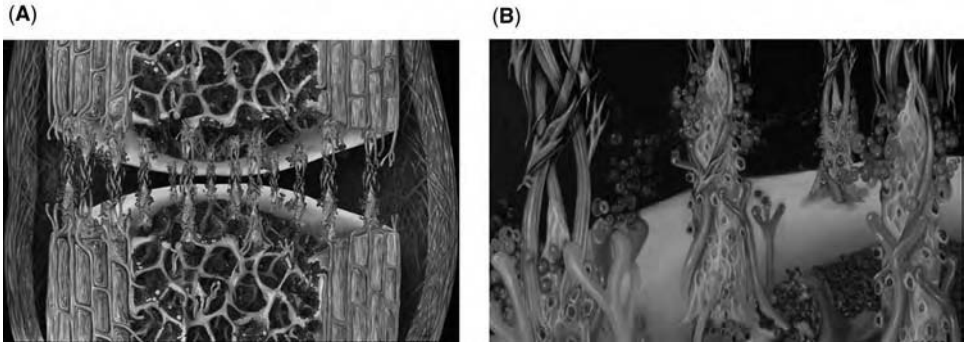

Figure 8 (*See color insert.*) (**A**) Artist rendition of histological findings demonstrates multiple bone forming units (BFUs) bridging the entire cross-section of the two distraction surfaces. Note the central collagenous zone of each BFU, where strain energy is absorbed following each distraction. This drawing demonstrates why the overall distraction force (load) is proportional to the number of BFUs (cross-section of bone formation) and indirectly related to the length of unmineralized collagen. (**B**) Closer examination of the BFUs reveals massive zones of proliferating precursor cells, which migrate from local bone marrow and periosteum, concentrating on the tips of each new microcolumn at the zone of primary matrix formation. *Source*: Courtesy of Ron Tribbel.

osteogenic zone stress is probably critical for perpetuation of the distraction osteogenesis process. Our data suggest that by week 3 of distraction, an osteogenic zone stress in the range of $47\,N/cm^2$ is consistent with healthy distraction osteogenesis and an osteogenic stress below $27\,N/cm^2$ will lead to nonunion, secondary to an abnormal biological bridge (Fig. 8A,B).

Arguments for Strain-Induced Bone Growth (Wolff's Law)

1. Histologically, the distracted bone surfaces are joined by a collagenous bridge that progressively mineralizes.
2. Differential dissection in vivo proves that the majority of load during distraction is carried by this bridge.
3. The actual loads measured during distraction correlate directly to the cross-sectional area of this bridge and inversely to the length of the unmineralized portion.
4. Prolonged latency allows for advanced mineralization of the bridge, increasing loads, while more rapid rates of distraction inhibit the mineralization of this bridge, decreasing loads.
5. FEA of the osteogenic tissue based on converted QCT data correlates highly with measured strains.
6. Critical levels of stress are found relative to future outcome such that lower stresses lead to nonunion, presumably due to disruption of the biological bridge, and higher stresses mean premature bony consolidation, while an intermediate stress level leads to healthy bone formation.

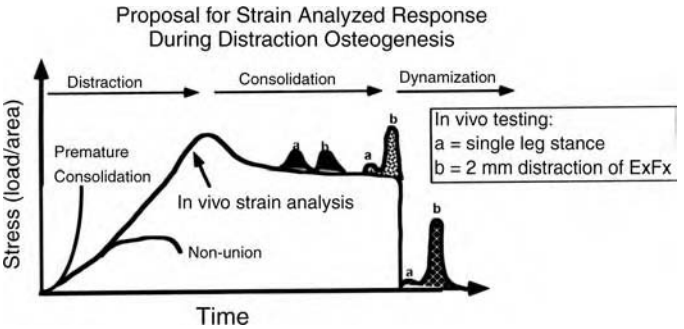

Figure 9 Proposed graphic for in vivo strain analysis during distraction osteogenesis based on these studies. Different conditions (premature consolidation, nonunion, and normal load generation) are depicted over time, as well as during consolidation and dynamization using either distraction or single leg stance to test the load, which allow for early prediction of normal bone formation and solid healing.

Proposal for Strain Analyzed Response for Guiding Treatment

In-line strain gauges used with ring external fixation during distraction osteogenesis can guide the clinician to make critical decisions during treatment that will maximize the chances for successful bone formation. Early disproportionate increases in measured loads can identify premature consolidation before plain radiography, allowing for either increase in rate or repeat corticotomy, saving time. Early disproportionate decreases in load can predict a disruption in the biological bridge, allowing either decrease in rate or reversal of distraction (compression) to reactivate the local osteogenesis, saving treatment time and maximizing the result. During consolidation, strain analyzed response to loading can predict the mechanical stiffness of the osteogenic tissue bridge, allowing for appropriate timing of frame dynamization and removal while minimizing the risk of collapse (Fig. 9).

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3 Mechanical Principles of the Ilizarov Method

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INTRODUCTION

The success of the Ilizarov method relies on the presence and collective function of several biologic and mechanical factors. Ilizarov observed that for bone to realize its full osteogenic potential, a versatile fixation system was needed that would provide stability, soft tissue preservation, adjustability, and functionality. Unique features of the Ilizarov frame and method are the use of a stable yet dynamic frame that allows the surgeon the options of acute and gradual correction, frame modularity to build as complex a frame as needed, and the ability of the patient to bear weight and move adjacent joints as tolerated.

There are a lot of different external fixation devices that have been developed and used. However, the term ''external fixator'' in traditional orthopedics is usually synonymous with a spanning temporary device that is associated with marginal bony reduction and fixation that leads to malunions and nonunions. We are not going to discuss such devices here. The purpose of this chapter is to review the mechanical principles of the Ilizarov method. Observed through biomechanical research and clinical experience, these principles provide a foundation for the proper application of fixators, which play an increasingly vital role in modern orthopedics.

THE MECHANICS OF CIRCULAR EXTERNAL FIXATION

Rings and Connecting Rods

Popularized by Ilizarov, circular fixators are comprised of several components, the most fundamental of which are rings and connecting rods. Varieties of Ilizarov rings include full (closed) rings, partial (open) rings, and arches. Although full rings provide the most rigidity, partial rings and arches are particularly helpful when working near joints and in areas where a closed ring would prevent normal extremity motion or would be poorly tolerated, such as the proximal tibia and femur. Open rings are also helpful when wound access is needed after trauma.

Bony stability is of paramount importance for osteogenesis and is dependent on the stability of the external frame. Frame stability is greatly impacted by ring properties. Rings of larger diameter are less stable than smaller rings (1–3). Gasser et al. found that decreasing the ring diameter by 2 cm increased axial frame stiffness by 70% (4). Ideally, the smallest diameter ring that will fit the extremity should be used. A guideline is that one should allow for 2 cm of space between the skin and the ring circumferentially for possible soft tissue swelling. In general, the bone should occupy a central position in the ring, but an eccentric bone position in the ring as occurs around the proximal tibia has no adverse effect on ring stability (5,6). The distance between the rings and the type of ring connections used will affect stability. Rings that are far apart and connected with long rods will be relatively unstable. In order to minimize the unsupported length between rings, additional connecting rods or an intermediate free ring secured in the mid-portion of the long rods should be used.

The concept of a *ring block* is important to understand with respect to frame stability. Simply defined, a ring block is the portion of the frame that is attached to a bone segment. Bones are divided into segments by fractures, defects, or osteotomies. The stability of a ring block will increase by using two rings instead of one, controlling both near and far ends of each bone segment, having a minimum of four connections between the rings, and using at least four points of fixation to the bone with two to three points per ring (Fig. 1). With short metaphyseal segments, ring blocks commonly consist of one ring with three to four points of fixation maximally spread across the bone segment. Lengthening frames usually gain and sustain additional stability from distraction forces that are generated for overcoming resistance of the soft tissue envelope. In this case, the ring block can be shorter, consisting of one ring for each segment of bone with multiple wires in different planes. Atrophic, mobile nonunions have little inherent stability and require double ring blocks if allowed by the segment length. Although hypertrophic, stiff nonunions have inherent stability and one ring block could be sufficient for each segment; the frame would not be efficient enough to correct deformity through a stiff nonunion. Increased frame stability is needed with addition of rings, olive wires, or multiplaner half-pins. Bone transport requires more stability than lengthening, in part because the docking site is essentially a mobile nonunion.

Wires

The ringed scaffolding of the Ilizarov frame supports the limb through the use of transfixion wires and half-pins. Ilizarov used *all wire* frames and this is still the case today in Kurgan, Russia. Transfixion wires have many advantages when used with circular frames. Stable fixation of small bone segments is made possible by using multiple wires. This is especially useful in pediatric patients where fractures commonly have small metaphyseal segments and where epiphyseal fixation can be obtained without crossing the growth plate. Moreover, it is simple to remove an infected or painful wire in the office.

The mechanics of transfixion wires including proper insertion and tensioning techniques have been elucidated. Frame stability increases with increasing wire diameter and tension, with the use of more wires per ring, with the use of a drop wire, by placing wires on opposite

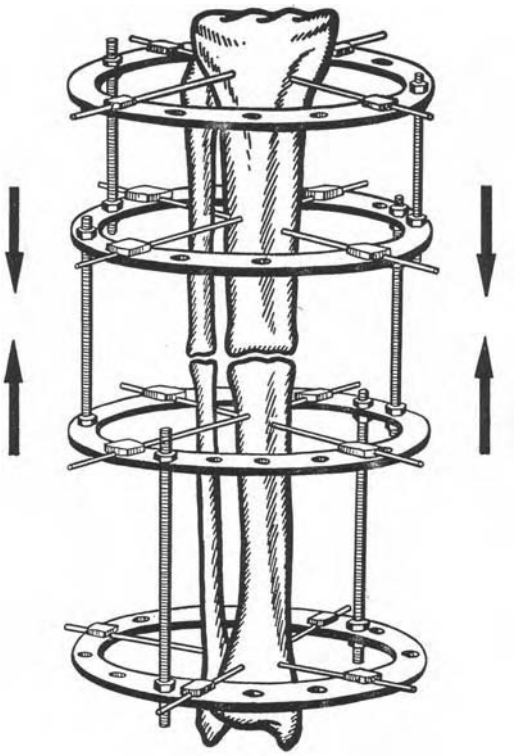

Figure 1 Depicted are two distinct ring blocks being compressed together. Note that each block consists of two rings with two points of fixation (wires) per ring. Threaded rods are parallel to each bone segment, preserving alignment of the entire long bone. (Although this illustration shows two connecting rods between each ring, four rods are needed for adequate stability.)

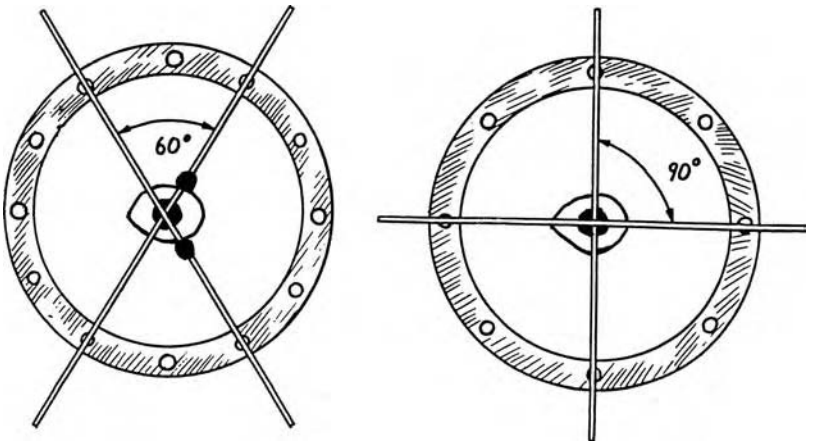

Figure 2 The picture on the right shows a 90° crossing angle. The left-sided diagram demonstrates how opposing olive wires can be used to achieve stability for wire crossing angles less than 60°.

sides of the ring, and by inserting wires in different planes including crossing from the top to the bottom of the ring (2,5–9). Increasing crossing angles of wires approaching 90° provides maximal stability (3). Crossing angles of less than 60° may allow the bone to slide along the wires requiring the use of opposing olive wires or the addition of a half-pin (Fig. 2) (Table 1).

Wire Insertion Techniques

A thorough knowledge of the cross-sectional anatomy of the extremity is necessary to avoid neurovascular injury. If under general anesthesia, the patient should not receive paralytic

Table 1 Methods to Increase Frame Stability

Ringed fixators	
Rings	Decreasing ring diameter
	Decreasing ring-to-skin distance
	Securing near and far ends of bone segment
	Increasing number of rings (use "dummy ring" to span long distances)
Ring connections	Increasing number of connections
	Increasing rigidity of connections
	(Telescopic rods to span long distances)
Wires	Increasing diameter of wires
	Increasing number of wires
	Increasing tension across wire
	Maximizing crossing angle wires
	Opposing olive wires
	Drop wires
Half-pins	Increasing diameter of pins
	Increasing number of pins
	hydroxyapatite coated pins
	Maximizing crossing angle half pins
	(Out-of-plane/multiplanar pins)
Bone considerations	Maximize bone end contact
	Apply compression/distraction
	With distraction: short, wide regenerate
Unilateral fixators	Decreasing bar-to-bone distance
	Multiple connecting rods (pin-to-bar)
	Increasing diameter of connecting rod
	Securing near and far ends of bone segment
	Multiplanar pins

Source: From Refs. 1–14.

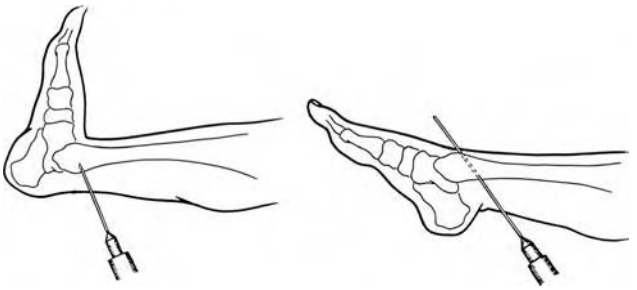

Figure 3 Positioning of the foot while inserting a wire to allow ankle movements after application of the frame.

agents because this will mask the important signs of flickering of the distal extremities when a motor nerve is irritated by a wire. Spinal or epidural anesthesia will not mask the irritation of a motor nerve. Proper wire insertion demands the use of a low heat technique. It is important to minimize the heat generated during drilling of the wire through the bone to prevent bony and soft tissue necrosis, which is directly related to infection and loosening of the wire. To prevent thermal necrosis, the tourniquet should never be inflated prior to drilling because normal blood circulation will help cool passing wires. Alcohol- or saline-soaked sponges can be used to cool and direct the wire during drilling. If the bone is particularly hard as is the case of diaphyseal wires, then frequent pauses will prevent heat buildup. The all-cortical wires should be avoided because this will generate heat rapidly. When the tip of the wire is through the soft tissue on the opposite side of the extremity, the wire should be pushed the rest of the way using a mallet.

When inserting a wire near a joint, the joint should be placed in the end range of motion (e.g., dorsiflexion—before inserting a wire on posterolateral or posteromedial side; plantar flexion—before the wire comes out on anteromedial or anterolateral site) to allow joint mobility (Fig. 3). The same concept is used if passing through fascial compartments, although transcompartmental wires should be avoided because they are uncomfortable and may become infected from movement of the soft tissue at the wire site when activating the muscles (3).

Olive Wires

Olive wires provide an important buttress effect in the correction of angular deformity using the Ilizarov frame. The ''rule-of-thumb'' is a guide to olive wire placement about a deformity. Proper bead positioning will increase the efficiency of the frame (Fig. 4). Olive wires are also used to fine-tune reduction of the bony fragments and/or to provide oblique or side-to-side compression at the fracture or nonunion site.

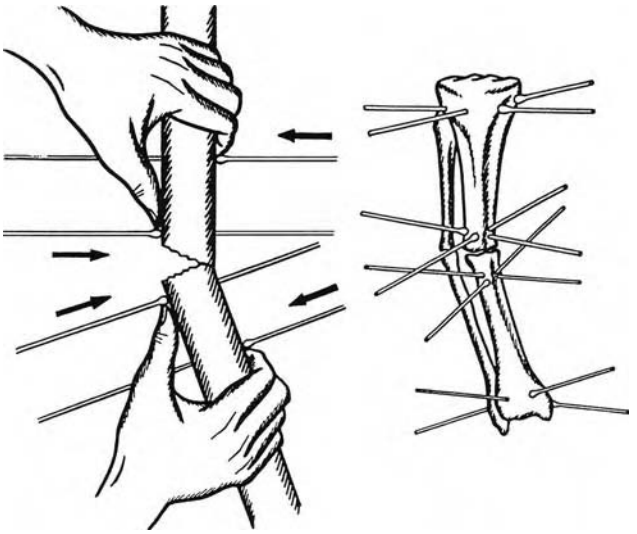

Figure 4 The deformity is ''grabbed'' with both hands so that pushing with both thumbs would straighten the bone. Olive wires are placed where the thumbs would be and on the opposite side where the other fingers would be. The olive wires provide improved pushing power, allowing the frame to work more efficiently in correcting the deformity.

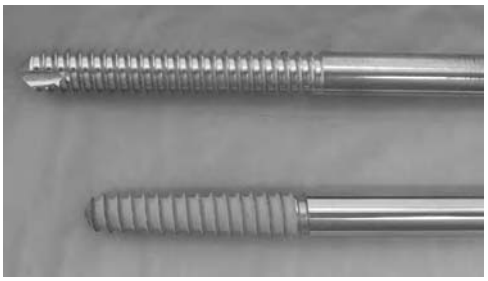

Figure 5 Bicylindrical uncoated 6 mm half-pin (*top*) is shown adjacent to a 5/6 mm tapered hydroxyapatite coated pin (*bottom*).

Wire Tensioning

Wire tensioning greatly enhances the rigidity of the wire and the stability of the frame (6). Smooth wires are typically tensioned to 130 kg. Tension beyond 155 kg will cause stretching and plastic deformation of the wire (10). Wires that cross at an angle of less than 60° should ideally be tensioned simultaneously to ensure equal tension across both wires. This is especially important if using on an open ring and when using olive wires.

Half-Pins

The use of half-pins in lieu of some wires gained popularity in the West where surgeons are more accustomed to inserting half-pins. Almost all external fixators used in the United States rely on half-pins to provide bony fixation. Advantages of half-pin fixation include familiarity in application, patient comfort, rigid fixation, and a low infection rate especially in the tibia, where half-pins are not inserted through the muscles or in close proximity to the tendons. The mechanics of these pins follow the same trends as those of transfixion wires. As the diameter of the pins increases, so does the rigidity. A 90° crossing angle of half-pins is desirable for improved control in multiple planes. Calhoun et al. (11) studied wire-only and combination with half-pin frames and showed that the use of half-pins increased the bending and torsional stiffness of the frame. Knowledge of the anatomic safe zones and respect for the soft tissues apply. The principle of low heat generation during half-pins' insertion is again of paramount importance. All half-pins should be bicortical and predrilled with a tissue protection sleeve. In hard bone, frequent pauses are prudent, and the drill flutes may need to be wiped clean before passing through the far cortex. Pins are inserted by hand.

More recently, hydroxyapatite (HA) coated pins have become popular, especially in limb lengthening and deformity surgery where frames stay on for several months. HA coated pins have been shown to have significantly increased extraction torque, lower rates of loosening, decreased infection rates, and even a lower incidence of secondary deformity during lengthening (8,12–14). Although both tapered and bicylindrical designs exhibit excellent pin extraction torque and microscopic evidence of direct bone-pin contact (15), we prefer 6 mm tapered HA coated pins (Fig. 5). Pins are available in 4, 5, and 6 mm diameter sizes. When selecting a pin, keep in mind that the diameter of the half-pin should be less than one-third the bone diameter to minimize the risk of fracture at the pins site (Table 2).

WIRES OR HALF-PINS?

This has become a subject of debate between classic and modern Ilizarov surgeons. Decisions will be made based on surgeon preference, anatomic constraints, and mechanical principles. While *all wire* or *all half-pin* frames can be correct if applied with sound mechanical principles, we prefer hybrid frames in which wires and half-pins are used optimally. Transverse wires are useful in the metaphysis where they avoid muscle compartments and help establish proper ring orientation (*reference wire*). The *reference wire* helps with the application of a *reference ring* to a given bony segment. Wires are useful for tibia and fibula fixation in both the proximal and distal leg. Because wires can be easily removed in the clinic, they can be used when only temporary fixation is felt to be needed.

Half-pins are particularly useful in the diaphysis where large crossing angles can be achieved without invading the muscle compartments of the leg. In the metaphysis of the tibia,

Table 2 Pearls of Frame Mounting

Rule of twos:
2 cm between skin and frame
2 rings/bone segment
2 points fixation/ring
2×2 (4) connecting rods b/w rings
Fixation both (2) ends of the bone segment (near-near and far-far
Pin and wire fixation in 2 planes
Other:
1.8 mm smooth or olive wires (adult)
6.0 mm tapered hydroxyapatite coated half pins (adult)
Wires in metaphysis
Pins in diaphysis

Source: From Refs. 1–14.

half-pins can be inserted in a greater anterior-to-posterior orientation than could be achieved with wires. Half-pins are very useful in the femur and humerus for anatomic reasons.

When deciding whether to use half-pins or wires for fixation, the clinical scenario may call for one over the other. We have concerns about the use of half-pins in patients with neuropathy because the pins have a tendency to become loose and infected, or fatigue and break at the level of the cortex. On radiographs, we have observed large areas of bone resorption around the half-pins, which is likely the result of uncontrolled weight bearing in patients who lack the protection of pain feedback (minimal weight bearing is recommended to these patients). In children, we tend to use more wires close to the growth plate. In children with congenital pseudoarthrosis of the tibia, we avoid half-pins because of their poor bone remodeling potential around larger holes from half-pins.

HINGE CONCEPTS AND ANGULAR DEFORMITY CORRECTION

Ilizarov hinges are used to perform gradual corrections of bony angular deformities, joint contractures, and to span and stabilize joints during limb lengthening procedures. Hinge types include simple ones that allow movement only in a single plane, and universal ones that have greater degrees of freedom (Fig. 6).

The simplest method to correct an angular deformity is to place a hinge at the apex of the deformity. Placing the hinge along the bisector of the deformity ensures that no translational deformity will occur during the correction. If the hinge is placed at the cortex over the convexity of the deformity, then subsequent correction will result in an opening wedge type osteotomy (Fig. 7). If the hinge is placed off the bone but still in the convexity, the result will be a simultaneous correction and lengthening (Fig. 8). If the hinge is placed within the confines of the bone, then a neutral wedge correction will result with opening along the concavity and compression of the convex cortex. This compression could result in fracturing of

Figure 6 Simple hinges allow motion in one plane and can be assembled by connecting two plates (*left*) or two posts (*middle*). The universal hinge allows greater freedom (*right*).

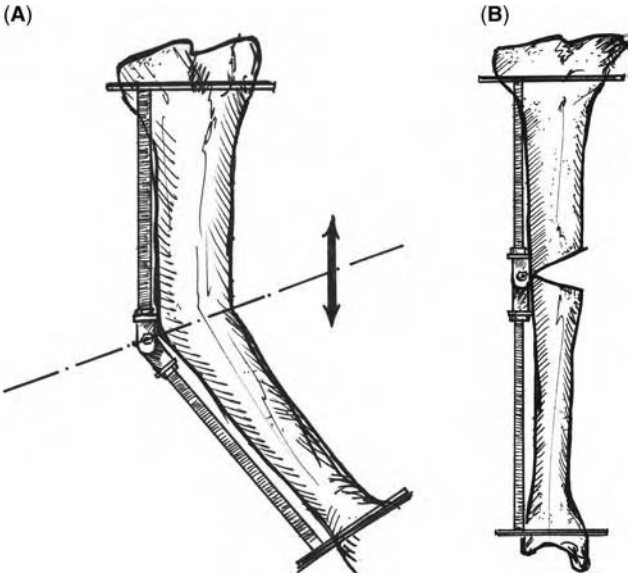

Figure 7 (**A** and **B**) This simple hinge is placed on the bisector line at the apex of the deformity on the convex cortex (**A**). Straightening the bone will result in a true opening wedge correction (**B**).

the cortex, the inability to achieve full correction, or undue stress on the bony fixation points with displacement of the less stable fragment. Hinge placement off the bisector line will result in translation during angular correction (Fig. 9).

When gradually straightening a deformity, the ideal rate of correction is 1 mm/day. The distance of 1 mm is measured at the location that the surgeon chooses, typically the concave cortex because this cortex will travel the furthest, or the concave border of soft tissues in a case of joint contracture.

The hexapod type frames have gained popularity in the gradual correction of deformities. These frames use a virtual hinge in lieu of a stainless steel one and are capable of correcting a deformity in multiple planes simultaneously. With the use of a computer program to generate schedules for corrections, modifications of the frame are minimized. The placement of the virtual hinge could be done by using the same Ilizarov principles or utilizing the *origin-corresponding point* method described by Charles Taylor. We usually choose the center of the bone for virtual hinges especially in cases of rotational correction. Central placement of the *origin* simplifies rotational correction; however, about 5 to 8 mm of distraction is

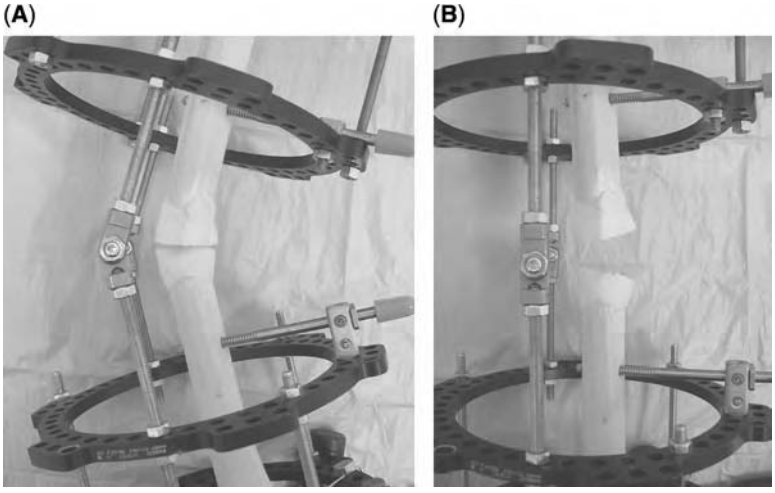

Figure 8 The hinges lie on the bisector line of the deformity in the convexity of the curve but outside the bony confines (**A**). By placing the hinge on the bisector line and off the cortex, straightening of the bone will achieve distraction of the osteotomy concurrently with the deformity correction (**B**).

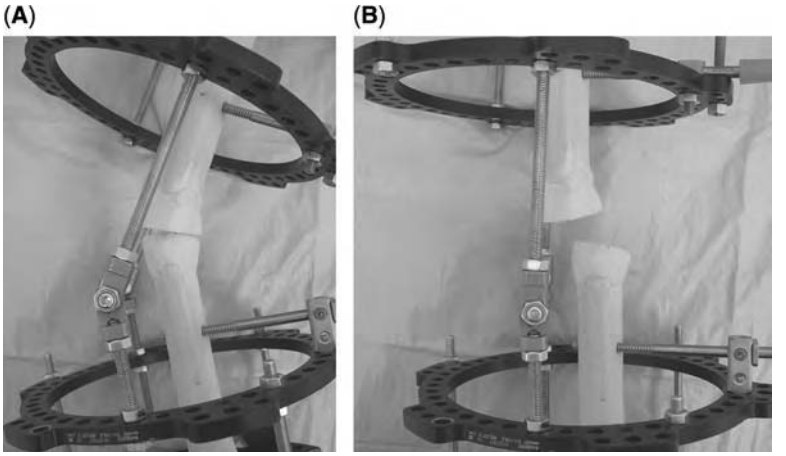

Figure 9 A hinge that is positioned not on the bisector (**A**) will result in a translational deformity (**B**).

required prior to correction of the rotational deformity. In cases of angular correction in one of the vertical planes, the osteotomy site should also be gradually distracted by several millimeters to protect the convex cortex from compressive forces and allowing the bone ends to clear one another. When using the Taylor Spatial Frame (TSF) (Smith and Nephew Inc., Memphis, Tennessee, U.S.A.) program, the rate of correction is determined by selecting 1 mm of movement per day at the *structure at risk* (SAR). The SAR is typically the concavity of the deformity, but it can be in any chosen location. If the soft tissue (not the bone) in the concavity is chosen to be the SAR, then it will move at the rate of 1 mm while the bony cortex will move more slowly because it lies closer to the hinge point. Similarly, if a valgus knee is corrected through a proximal tibial osteotomy at a rate of 1 mm/day, then the peroneal nerve is selected as the SAR ensuring that the nerve will be stretched at 1 mm/day. This mandates that the osteotomy move at a slower rate, increasing the risk of premature consolidation. One has to recognize these features of the TSF program and use clinical judgment to decide the optimal rate of correction.

COMPRESSION AND DISTRACTION FORCES

The effect of compression or distraction of the frame is an increase in frame stability. The mechanics of this improved stability are attributable to an increase in wire tension and rigidity, which enhances the stiffness of the entire frame. Good bony contact in combination with compression at the fracture site dramatically augments frame stability, load sharing between frame and bone, and the ability to bear weight (3,6). During a consolidation phase, the stability could be also improved with additional compression across a nonunion site. Distraction of the frame tightens the surrounding soft tissue envelope providing increased stability. A lengthening frame could consist of a simple single ring per segment construct.

The geometry of the fracture or osteotomy also affects stability and direction of compression forces. In case of a comminuted fracture, stable fixation of the bone in a neutral mode (no compression or excessive distraction) should be achieved. Excessive distraction will decrease bony contact and impair bone healing. Excessive compression will lead to shortening and deformity.

In the case of a fracture or nonunion with incongruent or oblique ends, axial compression should also be avoided because it will lead to shortening and malalignment. With oblique bony edges, side-to-side compression can be achieved by using directional wires (olive or arched) and/or by compression of the ring blocks perpendicular to the oblique end of the bone.

MONOLATERAL EXTERNAL FIXATORS

Monolateral fixators have been found to afford limb function and provide good bony stability (16). These frames are particularly useful in the femur and humerus where rings are

uncomfortable. The stability of these frames is enhanced by the use of a heavy and rigid frame design and by decreasing the distance between the frame and the bone. Other techniques to improve stability include using larger diameter half-pins and more points of fixation, maximizing the spread of pins over a bone, and placing pins in multiple planes (16).

FUTURE DIRECTIONS

The Ilizarov method has greatly contributed to the fields of trauma, limb lengthening, limb reconstruction, and deformity correction. Central to the success of this method is the Ilizarov circular frame both in its traditional *all wire* and in combination with half-pin forms, whose mechanical properties have been demonstrated. Over the last decade, the TSF has been introduced and embraced by the trauma and limb lengthening communities. This frame has been used for multiplanar deformity corrections, lengthening, nonunion management, and fracture care. When under tension, TSF clinically provides a reasonable stability; however, while in its neutral mode it allows for increased motion (due to the universal joint struts-to-ring connections). This frame can be further enhanced with mechanisms to stabilize a neutral frame and to axially dynamize the frame.

The TSF struts and program can be further improved to allow more gradual frame adjustment in increments of a millimeter. Further biomechanical testing needs to be done as well as experimental and clinical studies looking at frame stability and healing time compared to the Ilizarov system. Limb lengthening and reconstruction is a rapidly growing and exciting field of orthopedics with ample opportunities for further biomechanical studies.

REVIEW OF LITERATURE

Authors	Journal	Title	Model	Results	Conclusions
Orbay et al.	Clin Orthop, 1992	The effect of wire configuration on the stability of the Ilizarov external fixator	Comparison varying wire number and orientations	Increasing number of wires—greatest effect on axial and torsional stiffness Drop wire greatly improved bending stiffness Opposing olive wires improved shear stiffness	Bending and axial stiffness is directly proportional to the number of wires
Calhoun et al.	Bull Hosp Jt Dis, 1992	Rigidity of half-pins for the Ilizarov external fixator	Comparison Ilizarov vs. Hybrid Ilizarov	Ilizarov high axial stiffness hybrid higher stiffness bending and torsion	Three-6 mm half-pin-only system provide more stiffness than two-wire (W) construct in all loading modes; hybrid system (1 W plus 1 or 2 half-pins) exhibits stiffness comparable to 2 W system
Podolsky and Chao	Clin Orthop, 1993	Mechanical performance of Ilizarov circular external fixators in comparison with other external fixators	Four ring Ilizarov fixators were tested for parametric variations in wire tension, wire diameter, crossing wire angle, and bone position in a	Nonlinear stiffness behavior under axial compression loads. Wire diameter 1.8 mm increased the stiffness. Fixators with wires crossed at 45° had significantly less stiffness in axial compression	Stiffness curve increases under higher axial compression loads. Eccentric position of bone has no adverse effects. Tortional stiffness

(*Continued*)

REVIEW OF LITERATURE (*Continued*)

Authors	Journal	Title	Model	Results	Conclusions
			ring in axial compression, torsion, bending		increases under coupled axial compression
Antoci et al.	J Orthop Trauma, 2005	The effect of transfixion wire number and spacing between two levels of fixation on the stiffness of proximal tibial external fixation	Comparison of differing wire and ring number on frame stiffness	The use of 5 wires at a single level of fixation provided comparable bending stiffness to that of a double level ring block of 2 rings with 2 wires off each ring	Short metaphyseal bone segment can be stabilized effectively with 5 wires

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4 Enhancements of Regenerate Bone Healing

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INTRODUCTION

Distraction osteogenesis is arguably the most dramatic expression of the anabolic potential for new bone formation and repair. Its enthusiastic adoption into established orthopedic practice stems from the capacity for simultaneous lengthening and complex deformity correction. No other process is currently reliably capable of producing entirely new spans of bone up to and beyond 6 to 8 cm in length. In fact, the length of new bone gained may be only limited by the soft tissues' capacity for distraction.

Various biological and mechanical strategies have developed, aimed at improving our ability to enhance the rate and volume of regenerate formation. These include: allowance of a latency period, the performance of low-energy metaphyseal osteotomy, careful soft-tissue preservation, and load-sharing fixator designs. We assume, for the purposes of this chapter, that the influences of these variables are well understood from the preceding chapters on biological and mechanical principles.

In some cases, the individual biological condition of the host and/or the mechanical environment conspires against optimal regenerate formation or consolidation in the desired timeframe. All surgeons who utilize distraction osteogenesis for limb reconstruction have experienced cases of regenerate insufficiency. We characterize these as:

1. Failure of adequate regenerate formation in an expected time frame
2. Fracture through the regenerate or adjacent bone (Fig. 1)
3. Bending of the regenerate postframe removal

Solutions that have been devised include biological and mechanical alteration of the limb environment. Mechanical solutions, which have been introduced to attempt to overcome some of these problems, include lengthening over a nail and lengthening nails (2–4). Slow healing is not necessarily a problem if the defect is spanned with a nail and functionality has been returned to the limb. However, these techniques are not universally applicable and have their own intrinsic limitations. The development of adjunctive therapies aimed at enhancing the biology of regenerate formation and managing cases of regenerate insufficiency remains a worthwhile endeavor.

CLINICAL RELEVANCE

The clinical need for enhancements of regenerate bone healing is well accepted. Although powerful, distraction osteogenesis takes time. Therapeutically, increasing the relative anabolic response to distraction has a potential for both reducing the healing time in more basic cases and increasing success rates in more difficult reconstructive circumstances. Avoiding negative influences that inhibit the anabolic response, such as smoking and drugs, are also real and important issues in clinical practice.

On the other side of the healing equation, the catabolic element of bone repair has a potential contribution in distraction osteogenesis. Controlled bone resorption coupled with new bone formation is intrinsic to the remodeling process. However, when uncontrolled, catabolic bone resorption can produce complications such as stress-shielding responses, local and regional disuse osteopenia, regenerate deformation, and regenerate fracture. These problems

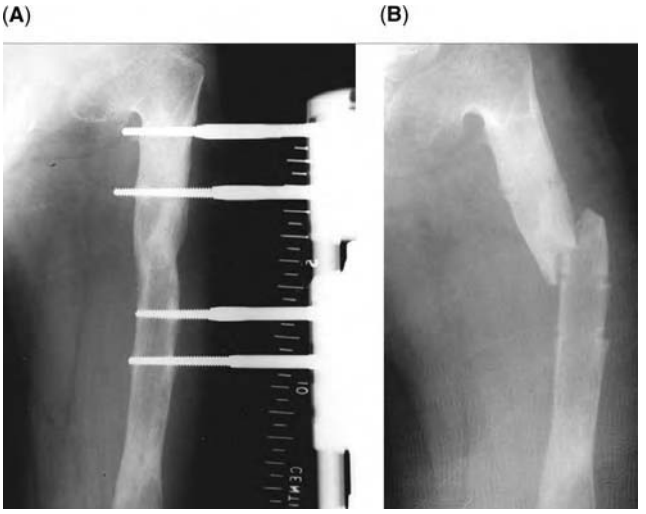

Figure 1 Radiographs of a child with a congenitally short femur after bifocal lengthening showing (**A**) good consolidation and (**B**) a fracture through the regenerate. *Source*: From Ref. 1.

continue to complicate our surgery and have not been fully eliminated by enhancements in mechanics; they could potentially be addressed by therapeutic means.

Many of the methods of enhancement of regenerate discussed in this chapter are already utilized in clinical practice. However, none has been developed to the point, where practitioners of distraction osteogenesis techniques can fully agree on their utility. As with any prescribed surgical technique, many practitioners favor a particular method of enhancement, usually based on their personal experience. Where possible, clinical trial results are referred to; however, much of this chapter focuses on the preclinical evidence relating to the potential of various approaches to enhance regenerate bone healing. In preclinical testing, more weight is given to studies that have shown improvement in mechanical outcomes in terms of strength of the regenerate. Observational histology without mechanical testing does not necessarily provide definitive evidence of improvement.

MAIN CONTENT

The processes of distraction osteogenesis have been detailed in previous chapters. However, in order to understand how interventions may help or hinder the progress of regenerate healing, we will separate the anabolic (bone forming) and catabolic (bone resorbing) components of bone repair.

Briefly, the osteotomy and subsequent distraction process triggers and maintains a cascade of events that begins as an inflammatory response, followed by cellular recruitment and proliferation at the osteotomy site. Cytokine and growth factor-derived stimuli and mechanical stimuli are important mediators of these events. These stimuli influence subsequent cellular differentiation, and thus the type, quality, and quantity of matrix are produced. These processes are coordinated with the revascularization of the distracted region, essential to the anabolic response. Simultaneously, the activated catabolic response acts to both remove unwanted tissue and initiate the remodeling process. Remodeling incorporates a coupled system of woven bone resorption, followed by mature lamellar bone formation.

Each of these processes may be a focal point for amplification or diminution of the net regenerate ''product.'' We have summarized the potential effects of various interventions on distraction osteogenesis utilizing this concept (Table 1). The net effect on the regenerate ''product'' in terms of quantity and quality is determined by the magnitude and interaction of the anabolic and catabolic responses. For example, weight bearing increases the anabolic response, but as this is coupled to resorption, catabolism increases secondarily. We might consider that this, in turn, produces increased remodeling, which in the setting of a healthy anabolic response is advantageous. The product in these circumstances is large amounts of remodeled, lamellar bone.

Table 1 Interventions

Therapeutic Agents	Anabolic Response	Catabolic Response
Weight bearing/mechanical stimulation	↑↑↑	↑
Ultrasound	↑	—
Electrical stimulation	↑	↓
Motorized distraction	↑↑	↑
BMP	↑↑↑	↑↑
Bisphosphonates	↓	↓↓↓
PTH	↑↑↑	↑↑
Hyperbaric oxygen	↑	—
Potentially harmful agents		
Rigid fixation	↓↓	↑↑↑
Smoking	↓↓↓	↑↑
NSAID	↓↓	—
Chemotherapy	↓↓↓	↓

Legend: ↑ mild increase; ↑↑ moderate increase; ↑↑↑ large increase; – no change; ↓ mild decrease; ↓↓ moderate decrease; ↓↓↓ large decrease.

Table 2 Summary of Literature

Reference	Animal/ Human	Results	Conclusion
5	Rat	Weight bearing in presence of fixator induced morphologic changes and remodeling but no change in strength	Changes in morphology cannot be presumed to result in increases in strength
1	Human	Refracture occurred in 10% of 173 patients after frame removal	Fractures occur in regenerate, at the regenerate junction and at distant sites. Associated disuse osteoporosis
6	Human	Increases in bone mineral content (BMC) on the limb receiving low intensity pulsed ultrasound (LIPU)	LIPU can influence regenerate formation in hemicallotasis
7	Human	Randomized trial showed no effect of pulsed electromagnetic fields (PEMF) on regenerate. Osteopenia in surrounding segments was reduced	PEMF does not enhance regenerate formation
8	Rabbit	Doses of OP-1 did not change histologic, densitometric, or biomechanical parameters	Anabolic response in distraction osteogenesis at 1 mm/day may not respond to bone morphogenetic protein (BMP)
9	Rat	BMP-7 treatment prior to distraction resulted in significant increases in bone mineral density (BMD) at day 14. By day 48, strength was increased fourfold over controls. Control healing was relatively poor at this rate of distraction	BMP may assist in regenerate formation when given at time of osteotomy, when distraction rate is high
10	Rabbit	BMC and bone volume were increased with administration of bisphosphonate. Significant increases in strength and stiffness	Bisphosphonates may prevent osteopenia and lead to increased strength of regenerate, when stress-shielding is present
11	Rabbit	Smoking decreases BMD and torsional strength	Smoking can create adverse outcomes in distraction osteogenesis
12	Human	Cultured MSCs and autologous PRP were injected into the distracted callus. These patients with dysplastic bone conditions rapidly consolidated with mean healing index 23 days/cm	Injection of MSC-derived proanabolic cells may form a basis for enhancing regenerate formation, particularly in cases with underlying bone abnormalities

In contrast, for example, a rigidly fixed segment combined with lack of weight bearing may manifest not only a diminished anabolic response, but the associated stress shielding may also potently drive premature and excessive bone catabolism. As the increase in catabolism is the primary driver, the further formation of new bone through coupling is promptly resorbed as the bone adapts to a mechanically inferior environment. This rapid bone turnover results in osteopenia in and around the regenerate. The osteopenic bone, although remodeled, has a decreased ability to take load and is at risk of failure upon fixation removal.

Therapeutic interventions and potentially harmful agents will be considered in the light of this anabolic versus catabolic paradigm. In this way, some (but not all) of the paradoxes that are present in the literature, and in clinical practice, maybe better explained and understood (Table 2).

ANABOLIC THERAPY

Mechanical Stimulation

The intrinsic structure and biology of bone relates to its primary load-bearing function. It is, therefore, logical that mechanical factors have such a profound effect on outcome in distraction osteogenesis, and adjunct mechanical stimulation is a logical possible solution to preventing regenerate insufficiency. The underlying theoretical basis for most of these techniques utilized is that increased loading of the regenerate in compression and/or distraction (but not shear) is anabolic and may remove the catabolic effects of ''hardware-induced'' stress shielding.

It is puzzling then, that while the animal study of Pacicca et al. demonstrated that weight bearing increased callus area and other morphological changes that would seem favorable although no effect was evident in improving regenerate stiffness, strength, or mineral content (5). It is possible that uncontrolled load bearing can damage primitive regenerate by introducing strains outside the optimal range. This study also further demonstrates that it is dangerous to interpret incomplete analyzes based on morphology alone and assume these have a mechanically beneficial effect. Had mechanical testing not been performed, a positive result may have been erroneously concluded.

Motorized Distraction

Ilizarov showed when distracting at 1 mm/day, 60 increments was better than 4 or 1 increments (13). It is likely that smaller, more evenly distributed incremental distraction lessens the injury to the forming regenerate. In support of this concept, Mizuta randomized limbs to eight-step distraction (0.125-mm increments), compared to four-step (0.25-mm increments) daily distraction. At all intervals until six weeks after the completion of distraction, bone mineral density (BMD) in the distraction gap was significantly higher in the eight-step group than in the four-step group (14). In another review, motorized distraction in 1440 increments was found to produce equivalent regenerate to mechanical distraction four times per day (15).

Although improving initial callus formation during distraction is desirable, the quality and amount of bone at frame removal is heavily modulated by the events during the much longer consolidation period than those of the distraction phase. This may explain why early gains are not translated to grossly improved outcomes. There is certainly no evidence that motorized distraction is harmful, and it may increase patient compliance and comfort.

Dynamization

Dynamization is another mechanical method of regenerate stimulation. Late dynamization, by allowing regenerate compression, may increase the axial and torsional load absorbed by the bone; this may stimulate it to strengthen by allowing remodeling in a less stress-shielded environment.

Despite the obvious merits of mechanical approaches, the complexity of frame design and variability of cases has meant that no particular system is fully accepted. There is currently no feedback mechanism whereby we can be certain that the loads and strains we apply are the optimal ones. This is a valuable area of ongoing research.

Mechanical Callus Stimulation (''Pumping'' the Regenerate)

This procedure involving sequential alternate small increment distraction and compression of the regenerate in cases of regenerate insufficiency is practiced quite by often some clinical practitioners. It is interesting that there is no hard evidence studying the practice, other than anecdotal reports.

A study by Mofid et al. in an animal model of rabbit mandibular distraction osteogenesis, applied alternate compression and distraction to one group, in 1-mm increments for a three-week period followed by a five-week period of further consolidation. While cortical: cancellous bone ratio (remodeling), cortical thickness, and mineral apposition were found to be greater in the experimental group, there was no difference in bending strength or stiffness on mechanical testing (16). Another experimental study, by Greenwald et al., of pumping the regenerate at the commencement of consolidation, demonstrated no significant difference in radiological or histological parameters measured (17).

In a variation on this theme, Kassis et al. showed that programmed micromovements of the fixator at the end of distraction significantly improved regenerate area, BMD, and bone mineral content (BMC) (18). This study also displayed a mean 11% increased capacity for axial loading to failure. There is no reported use of programmed micromovements of the fixator in clinical practice.

In summary, although morphologic changes are apparent in many studies changing the applied load to the regenerate, few of these studies show that this translates into significant changes in strength or stiffness at an appropriate outcome time point. Although the most basic tool at our disposal, our current understanding of the transduction of mechanical stimulation remains insufficient to make definitive recommendations from the literature.

LOW-INTENSITY PULSED ULTRASOUND

Low-intensity pulsed ultrasound (LIPU), a noninvasive form of mechanical energy transmitted transcutaneously as high-frequency acoustical pressure waves, is probably the best-studied method of regenerate enhancement included in this review.

Experimental Evidence

LIPU may work by enhancing endochondral ossification. LIPU can affect aggrecan gene expression (19), and is characterized by increased calcium incorporation in both cartilage and bone cell cultures. LIPU is demonstrated to modulate transforming growth factor-B synthesis in osteoblastic cells, and accelerate soft callus formation and enchondral ossification of the callus at the fracture site in animal models (20–22). In these models, the agent acts principally on the anabolic response, and perhaps advances the rate of endochondral ossification (19). Animal data on distraction osteogenesis is less impressive. While Shimazaki et al. showed early increases in percentage BMD in rabbit distraction osteogenesis, the effect was lost by the time of frame removal (23). Two further rabbit distraction studies show no effect of LIPU on outcome in terms of strength of the regenerate (24,25). A rat study showed trends toward increased strength and stiffness, but these did not reach significance (26). In bone transport, LIPU has been shown to increase axial and indentation stiffness in sheep (27). Destructive strength testing was not performed.

Clinical Evidence

The evidence for use of LIPU in fracture repair is considerably more robust than in distraction osteogenesis. Clinical studies have shown that LIPU has proven efficacy in randomized studies of tibial (28) and scaphoid (29) fractures. So, it is a logical choice to possibly enhance regenerate formation. Some clinical experience favors the use of LIPU in distraction osteogenesis. In a randomized trial during bilateral tibial hemicallotasis, Tsumaki et al. showed increases in BMD when LIPU was applied during the consolidation phase (6). However, this study involved a very small amount of distraction in a defined area and cannot be readily compared to limb lengthening.

An uncontrolled study by El-Mowafi et al. reports significantly improved healing indices in 10 patients treated LIPU during the consolidation phase, when compared with a

similar-sized group with no LIPU treatment (30). However, this paper describes the technique being employed mostly in bone transport, and no analysis is made allowing for the severity of initial limb injury, which is a major prognostic factor. The paper raises the need for further randomized prospective studies. These studies are the first to suggest the efficacy of LIPU in situations involving stable mechanical fixation. The majority of successful randomized studies of LIPU were in cast-treated fractures and a randomized study in nailed fractures produced no effect (31). LIPU may, therefore, not be effective when lengthening over a nail.

An overall view can be supported that the anabolic effects of LIPU in bone repair are proven. However, in distraction osteogenesis and other situations involving stable fixation, this stimulus may not always be enough to overcome the negative effects of stress shielding, which drive catabolism. Further high-level studies are required.

ELECTRICAL STIMULATION

Electrical stimulation has been pursued for some time as a method of bone-repair enhancement. The initial basis for this was that electrical potentials are created by the mechanical stresses and fluid flows in bone, and are likely to be one of the ways in which mechanical signals are transduced to the bone. Many different types of stimulation have been recommended, including direct current, capacitive coupling, and pulsed electromagnetic fields (PEMF). A discussion of the relative merits of these approaches is beyond the scope of this review.

Most of the available data in bone repair relates to the effect of electrical stimulation in fracture repair. These studies have shown efficacy in some cases equivalent to bone grafting (32). A randomized trial showed a positive effect of pulsed electric current in nonunions, even in the presence of smoking (33). A similar trial in delayed union also showed a positive effect (34).

The evidence in distraction osteogenesis is again less clear-cut. PEMF were found to have no effect on regenerate mineralization in a randomized study; however, catabolic effects on surrounding bone were reduced (7). A study in rabbits revealed no effect (35). Direct electrical stimulation in experimental mandibular distraction osteogenesis showed a transiently positive effect (36). However, in tibial distraction in beagle dogs, capacitive coupling electrical stimulation has been reported to have a negative effect (37).

The current data cannot be used to recommend electrical stimulation in distraction osteogenesis without further high-level trials being performed.

BONE MORPHOGENETIC PROTEINS AND OTHER ANABOLIC AGENTS

Bone morphogenetic proteins (BMPs) potently stimulate many phases of the anabolic cascade, including cellular recruitment, proliferation, differentiation, and bone production. Both recombinant BMP-2 and BMP-7 osteogenic protein-1 (OP-1) are clinically available. Such anabolic molecules hold potential for the improvement of regenerate formation, or the reinitiation of the anabolic response after distraction has ceased. However, despite their enormous potential, BMPs have not yet been optimized for clinical use in distraction osteogenesis.

Experimental Evidence

Two preclinical studies attest to the anabolic potential of BMPs (Fig. 2), but both of these utilized models where the distraction rate may have muted the normally profound anabolic response (9,38). In one study, the distraction rate was 2.0 mm/day in rabbits, and the untreated controls formed only attenuated, periosteally derived bone (38). The other study in rats was 0.5 mm/day, and the controls did heal (9). In that study, the BMP-7 was placed at the site of the osteotomy before the latency period. In contrast, Hamdy et al. attempted anabolic augmentation in a rabbit distraction model by injecting OP-1 at the end of a slow 2.0-cm distraction. This failed to change the outcome in terms of the histologic, densitometric, and biomechanical parameters assessed over vehicle treatment, even in doses of up to 2.0 mg per rabbit (human dose 3.5 mg) (8). These authors considered the lack of effect might be due to decreased BMP receptors being present after the end of distraction. It is possible that in properly carried out distraction, the anabolic response is already maximally stimulated. Another explanation may be catabolic stimulation by the OP-1, which in the presence of a rigid fixator may have actually

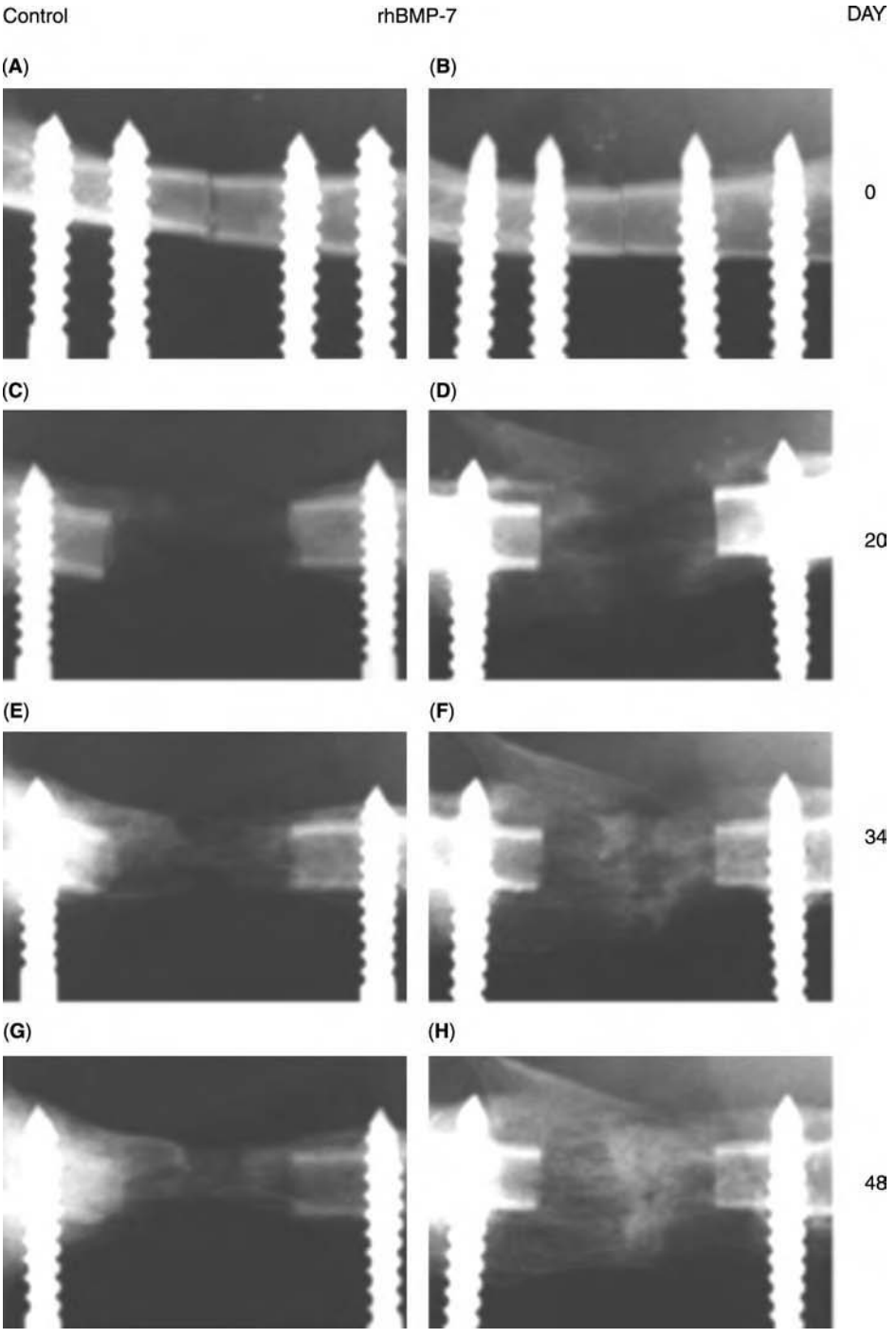

Figure 2 Radiographs of specimens from the control group (**A**, **C**, **E**, and **G**) and the bone morphogenetic protein (BMP)-7 group (**B**, **D**, **F**, and **H**). At day 20, the regenerate area in the BMP-7 group (**D**) is larger than that in the control group (**C**). At day 34, the regenerate area in the BMP-7 group (**F**) is larger than that in the control group (**E**). At day 48, the mature regenerate area in the BMP-7 group (**H**) is larger than that in the control group (**G**). *Source*: From Ref. 9.

had a detrimental effect. The paradox may relate to the fact that BMP-2 and BMP-7 are known to stimulate osteoclastogenesis through osteoblast-mediated RANK/RANKL signaling (39) and directly stimulate osteoclastic bone resorption (40). It is known that osteoclasts are already upregulated in distraction osteogenesis (41). Further stimulation could indeed be detrimental.

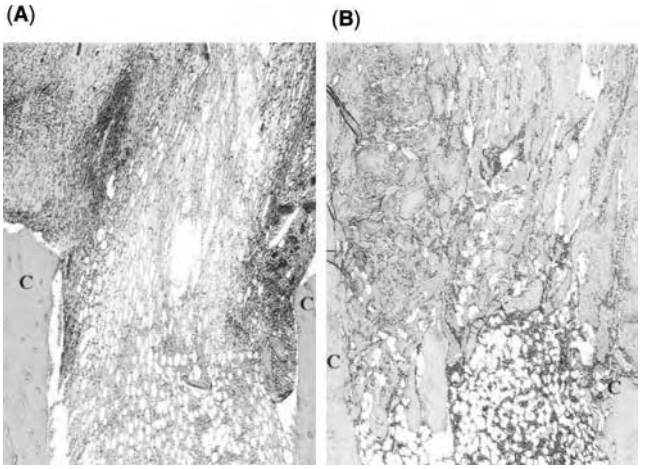

Figure 3 Histologic comparison was done of new bone formation from the distal end of the distraction gap in old rats with local delivery of either control substance or FGF-2 by an osmotic pump. (**A**) This old rat received a control substance and did not produce endosteal new bone. (**B**) This old rat received FGF-2, which restored endosteal new bone formation to levels typical of young rats. *Abbreviation*: C, cortex. *Source*: From Ref. 43.

In gap healing studies, it has been shown that bone formation and strength can be doubled by administration of OP-1 and a bisphosphonate over OP-1 alone, lending further support to this anabolic versus catabolic paradigm (42). Further study and optimization of BMPs in distraction osteogenesis is required before they can be recommended.

Other Anabolic Agents

Other growth factors have been applied successfully in animal models of distraction osteogenesis (Fig. 3). These include local FGF (43) and vitamin D analogs (44), and several studies into the anabolic effects of systemic applied human growth hormone (hGH) (45) and locally applied hGH (46). Daily parathyroid hormone (PTH) administration has an anabolic effect on fracture repair (47) and has been shown to be effective in distraction osteogenesis (48). Various blood-related derivatives and concentrates have biological effects in tissue healing. A thrombin-related peptide, TP508, has been shown to promote soft-tissue healing and fracture repair. In a rabbit distraction study, animals in the TP508-treated groups had complete bony union of the distraction gaps when compared to the saline-treated group at two weeks (49). Examination of the regenerates demonstrated an improvement in BMD and the histological parameters examined. Mechanical testing was not performed.

Platelet-rich plasma and platelet concentrates also have potential anabolic effects in bone healing; however, this simple potential therapy, used alone, or in combination with anabolic cell lines or other agents (typically bone marrow aspirate, or cultured mesenchymal cells), remains largely unresearched (12,50).

HYPERBARIC OXYGEN THERAPY

The effect of hyperbaric oxygen (HBO) therapy on bone healing has been a controversy. Coulson et al. (51) and Yablon and Cruess (52) noted that fracture healing in rats was enhanced by HBO in a mechanical strength study and a histologic study, respectively. Penttinen et al. (53) noted that exposure of fractured rats to HBO for two hours twice daily at 2.5 atm resulted in hypertrophy of cartilage and then in increased bone formation. In 1998, Ueng et al. reported that bone healing of tibial lengthening in rabbits was enhanced by HBO therapy at 2.5 atm for two hours per day (54).

No clinical trials have been reported on HBO therapy. Mobilization of patients undergoing distraction osteogenesis to attend therapy on a daily basis is likely to be impractical; however, such therapy may be viable for patients with identified regenerate insufficiency.

ANTICATABOLIC THERAPY

Bisphosphonates

Bisphosphonates are bone avid molecules that have antiresorptive effects. Modern nitrogen-containing bisphosphonates (N-BPs) prevent fractures from occurring in osteoporotic

conditions, control pain and deformity in Paget's disease, and decrease rates of skeletal-related events in many cancers affecting bone. The rationale for the use of N-BPs in distraction osteogenesis is to prevent catabolic effects, which may decrease net regenerate production and lead to osteopenic bone, which is prone to deformation or fracture. Simpson and Kenwright (1) and O'Carrigan et al. (55) have reported refracture in 8% to 10% of distraction cases. The problems of refracture are likely to be related to osteopenia, with reported decrease in local BMD during the healing period as great as 60% (56,57). Many authors report correlate regenerate torsional stiffness, resistance to bending and axial compressive loading, and overall regenerate strength to the BMC of the regenerate and in the affected limb. Although bone resorption is a natural and necessary component of bone repair, in some situations the amount of catabolic resorption is clearly detrimental, and arguably pathological, in its nature and effect.

N-BPs are administered orally or parenterally. Oral administration may result in gastrointestinal side effects, and some time is required before an effect is realized. Intravenous N-BPs give a faster onset of action, but must be used with care to avoid hypocalcemia or renal problems. Bisphosphonates have a very long half-life in bone, and lack of data on whether this may have unwanted effects has limited their use in younger patients, where more studies are needed.

In rabbit distraction studies, the N-BPs pamidronate and zoledronic acid (ZA) have been shown to increase BMC, BMD, and bone volume, both in and around the regenerate area (Fig. 4) (10,58,59). These increases have led to significant increases in strength, up to 89% in one ZA study (10). Stiffness was also significantly increased. Careful study through histomorphometry has revealed that the mechanism of ZA in distraction osteogenesis is to transiently decrease bone catabolism—there is no in vivo evidence anabolic effect (60). There is in fact a slight decrease in early bone formation, but this is more than offset by decreases in resorption leading to significant increased net regenerate size and strength.

This delay in catabolism is clearly also associated with decreased remodeling. However, it can be argued that regenerate is sometimes overremodeled, such that its mineral content and size are inappropriate to deal with the forces placed on the bone once the fixator is removed. Eventual remodeling after frame removal is an absolute necessity, and this had been observed to occur by 44 weeks in rabbits (60) and anecdotally approximately 1 to 2 years in adolescent patients (Fig. 5) (61).

Clinical Evidence

A case series of patients, N-BPs in distraction osteogenesis demonstrates potential for bisphosphonate use as adjunct therapy in cases of regenerate insufficiency (61). However, large controlled studies of the effects of N-BPs in distraction osteogenesis are lacking to date.

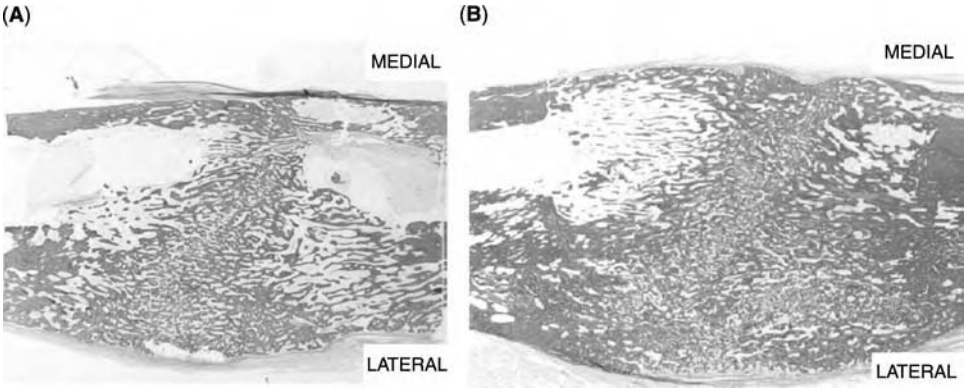

Figure 4 Regenerate bone at six weeks showing original cortical bone, trabecular bone organized in the line of tension from distraction, and predominantly occupying the lateral callus region. (**A**) Saline group. (**B**) Redosed zoledronic acid group. Saline group shows significantly less bone volume fraction in the distraction gap (Van Gieson's stain; magnification X1.25). *Source*: From Ref. 10.

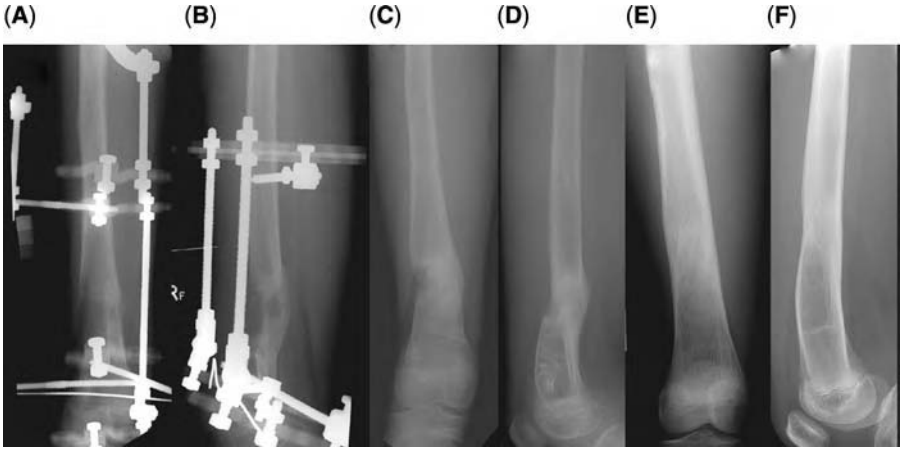

Figure 5 Case of regenerate failure and osteopenia (**A** and **B**). Short-term bisphosphonate therapy resulted in dense trabecular callus allowing removal of frame (**C** and **D**). Bone mineral density measured by dexascan (DXA) increased from 50% of the nonoperative limb before treatment to close to 100% by the time of frame removal. Remodeling was evident by two years post frame removal (**E** and **F**), with reformation of cortical/medullary definition.

OTHER ANTICATABOLIC THERAPIES

Other anticatabolic strategies have been attempted; however, none currently show the utility of bisphosphonates. A study of calcitonin in distraction osteogenesis has shown no effect and this study in rabbits demonstrated no change in the limb in terms of absolute BMD and BMD relative to the baseline BMD; however only one dose level was explored (62). Until further anticatabolic molecules are tested, N-BPs remain the mainstay, when the objective is to reduce bone resorption. Reduction of catabolism can only be a successful strategy in the presence of an adequate anabolic response.

OTHER EXPERIMENTAL THERAPIES

Newer techniques being explored for bone repair in general include gene therapy and stem cell technology.

In gene therapy approaches, cells are harvested and modified to overexpress a particular gene or combinations of genes are capable of rapidly forming bone tissue. This is particularly applicable in areas of rapid cell proliferation and turnover, as in the bone healing process. For safety, these overexpressing cells are terminally differentiated to give a limited lifespan or may even be programmed with a suicide gene such that ongoing pathological or malignant bone formation does not occur.

A recent paper by Ashinoff et al. in rodent distraction osteogenesis studied injection of the regenerate with an adenovirus carrying the BMP-2 gene. The gene therapy, administered after a 10 days latency and distraction period, demonstrated improved osteogenesis over a four-week period of consolidation (63). Mechanical results are lacking.

At the time of writing, the field of gene therapy would probably be limited to those patients with specific gene defects, or with problems refractory to other modalities of therapy, due to the high cost and possibility of unwanted side effects. These issues may well be overcome in the near future and allow the realization of this potentially powerful technique.

Stem cell technology takes multipotent cells, usually not genetically modified, and places them in the defect in a number many thousands of times higher than normally available. For this technique to be effective, the correct environmental stimuli to accelerate differentiation and bone production are needed, or the cells may remain quiescent or differentiate into an unwanted lineage. Many of these stimuli are indeed present in the distraction phase (64). Li et al. have shown a myriad of BMP genes expressed during the temporal phases of distraction osteogenesis, and programmed cellular events and apoptosis, which indicate the intricacy of this anabolic evolution.

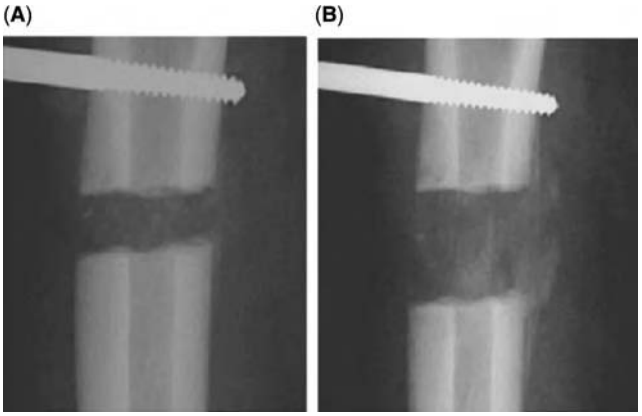

Figure 6 Radiographs of the right femur of a case before the first transplantation of MSCs (**A**) and two weeks later after transplantation (**B**). Callus formation was enhanced within the distracted area. *Source*: From Ref. 12.

Some clinical data and case studies have been reported using cultured mesenchymal stem cells, stimulated to develop along an osteoblastic differentiated pathway, and then implanted usually by injection into the distraction gap (Fig. 6). Although these results are still preliminary, transplantation of osteoblast-like cells and platelet-rich plasma, which seemed to be a safe and minimally invasive cell therapy, could shorten the treatment period by acceleration of bone regeneration during distraction osteogenesis (12).

DETRIMENTAL FACTORS

Smoking

Multiple studies highlight the negative effects of smoking on bone-forming cells and in bone metabolism. Clinically, smoking has been linked to adverse effects (65). Reports document a higher rate of nonunion in smokers after spinal fusion (66,67). Similarly delayed healing and nonunion of tibial shaft and mandibular fractures occur more frequently in patients who smoke (68). Cobb et al. examined cigarette smoking and nonunion after ankle arthrodesis and reported results showing the relative risk of nonunion increased 3.75 times for smokers versus nonsmokers (69).

However, it is very difficult to stop patients from smoking. In elective situations, smoking habits may need to be taken into account before embarking on complex time-consuming reconstructions because of this associated risk of anabolic failure. In trauma situations, this is not so easily done.

In 1997, the work of Ueng et al. demonstrated that cigarette smoke inhalation delays the bone healing of the tibial regenerate in an experimental rabbit model (11). Subsequent studies from Ueng et al. demonstrated that HBO therapy reversed some of the negative effects of smoking on the formed regenerate in the rabbit (70).

Nonsteroidal Anti-inflammatory Drugs

Much controversy exists around these commonly prescribed analgesics. Nonsteroidal anti-inflammatory drugs (NSAIDs) can be very helpful agents in managing postoperative pain, and as night pain can also be a feature of limb lengthening, NSAIDs may also be prescribed for pain relief later in the process. These agents inhibit the enzyme cyclooxygenase (COX). COX-1 and COX-2 are differentially induced in different tissues, but both may play a role in fracture and bone repair. It has been known for many years that NSAIDs in high doses have a negative effect. Part of the initial inflammatory cascade involves the conversion of arachidonic acid to prostaglandins, interleukins, and leukotrienes, which interact in a complex manner to stimulate the initial anabolic response. It has been shown that fracture healing does not proceed in COX-2-deficient mice, indicating the definite importance of that enzyme (71). Specific COX-2 inhibitors were very popular until the withdrawal of rofecoxib (Vioxx®) due to unwanted cardiac effects. Evidence was accumulating that this class of inhibitor had negative effects on bone healing, although this has been difficult to prove in clinical studies. Controversy exists over the potential negative effects in short term NSAID therapy postoperatively,

while early NSAIDs, most commonly indomethacin, have been characterized for their potent effect in reducing potential fracture healing even with short-term therapy (72,73). Long-term therapy with COX-2 inhibitors at high doses had minimal effects in a recent study (74), and the short-term effects have shown little or no significant effect (75,76).

As with the negative cardiac effects, negative effects on bone repair are likely to be dose related. Clinical reports regarding NSAIDs are inconclusive. Overall, the advice of Gerstenfeld and Einhorn in a recent editorial sensibly suggests that in situations where a robust anabolic response is absolutely a prerequisite for success, it is suggested that physicians consider short-term administration or use of other drugs in the management of these patients (77). No definitive study regarding the role of NSAIDs in distraction osteogenesis is available.

SUMMARY

Not all cases of distraction osteogenesis require augmentation or adjunctive therapy. Further refinement is required in an effort to identify those at risk of a poor outcome, either preoperatively or in the early distraction phase. Diagnostic tools such as DXA and 99-Tc bone scanning or other dynamic tests may assist us in future in better assessing these patients. Simple radiographic analysis will not necessarily give us the information we require. To properly apply an adjunctive therapy, we need to identify the biological or biomechanical problem. Is there an anabolic, catabolic, or mixed etiology for the regenerate insufficiency? Can it be predicted and prevented?

Patients with anabolic deficiency could be treated with anabolic interventions, whether these be one or a combination of mechanical, pharmacological, growth factor, or cell-based therapies. Those cases manifesting good bone formation but overt catabolism would be best treated by either increasing the stresses on the bone or directly blocking catabolism with bisphosphonates or other drugs. Patients with severe deficiencies or those likely to require prolonged treatment may benefit from combinations of treatments aimed at boosting the anabolic response and simultaneously controlling catabolism to an appropriate level that prevents osteopenia but allows remodeling.

Efforts at improving regenerate bone formation are continuing. The optimal combination of mechanical and biological management and the most cost-effective strategy has not yet been clearly demonstrated. We have described some of the underlying mechanisms involved in pathogenesis of regenerate insufficiency. Focusing attention to the actual deficit a patient has may allow a more targeted individual therapeutic strategy. We suggest that such an analysis can allow the clinician to potentially manipulate the anabolic and catabolic responses to optimize the outcome of regenerate bone healing in distraction osteogenesis.

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Part III ACUTE TRAUMA

5 Acute Trauma Applications

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INTRODUCTION

In the 21st century, because the number and severity of road accidents have risen dramatically, and military and terrorist activities have led to an increase in serious injuries, acute trauma in orthopedic surgery is more genuine today than ever.

Trauma patients can be found in all age groups. Today, lifespan is longer and we meet more and more elderly patients suffering from trauma rather than traditionally age-related accidents. The patient's general condition, especially in the event of chronic illness, may influence treatment and outcome after trauma. Or, conversely, trauma may aggravate an existing illness or even cause new chronic conditions in a previously healthy individual (1).

Limb trauma is often accompanied by open injuries. The percentage of open fractures of the tibia is about 24% (2). Soft tissue injury accompanying fracture may vary from an insignificantly small scratch or abrasion to damage that may lead to loss of the limb.

Multifracture and/or polytrauma condition, which are frequently life-threatening situations, require a special approach for treatment (3,4). The preservation of life is of primary importance, yet should be combined, if at all possible, with the preservation of functioning extremities and with a minimum of complications. Our goal should be a stable limb without deformation, of equal length, with functioning muscles, good range of joint motion, and free of pain. It is important to preserve normal innervation. Besides the obvious fact that all fractures must heal, additional key points of the treatment process are: decreasing time of disability, minimizing the number of surgical procedures, and lowering expenses. During the course of treatment, the life of the trauma patient should be as comfortable and dynamic as possible.

The Ilizarov external fixation technique is a valuable tool that enables achievement of these goals. In this chapter, we will discuss our approach to acute trauma with an emphasis on applications of the Ilizarov method. Tibial plateau and pilon fractures are among the most common indications for the use of a frame and separate chapters will follow to discuss these situations. Our focus in this chapter will be on other acute fractures and traumatic conditions.

The Ilizarov method is particularly advantageous for the following acute trauma situations:

1. Tibial plateau and pilon fractures
2. Open fractures with contamination
3. Poor soft tissue
4. Bone loss (acute shortening vs. bone transport)
5. Segmental fractures (multiple level stabilization)
6. Adjacent joint instability (fixation across joints)
7. Polytrauma patients with multiple limb injuries to enable early weight bearing

CLINICAL EVALUATION

Clinical evaluation of the trauma patient should follow a steady algorithm to avoid overlooking hidden pitfalls and to allow for correct planning in patient care.

An injured patient may present a wide range of complex problems. Therefore, initial evaluation must follow the well-established principles of Advanced Trauma Life Support and those of Damage Control of trauma (3,4).

Orthopedic assessment of an injured limb begins with the first view, as the surgeon surveys the skin color, the presence of wounds, visible deformities, swelling, and presence of contamination. Peripheral pulse and capillary refill should be checked. It is important to inspect skin sensitivity and examine movements of the hand or foot in order to evaluate the neurological status of the extremity. In the case of a possible compartment syndrome, immediate measurement of intracompartmental pressure should be taken (5). Palpation of the involved limb may help to exclude joint effusion caused by hemarthrosis.

If the patient's condition allows performing X rays in the emergency room, the surgeon should be present to ensure that the technician takes appropriate pictures. Temporary immobilization by splints should be applied before transfer to radiological investigation. Roentgenograms of limb segments should include proximal and distal adjoining joints. It is very important to keep in mind the mechanism of injury and to order additional radiograms of potentially involved areas. For example, in a case of a fractured calcaneus as the result of a fall from height, imaging of the thoracic and lumbar spine should be performed routinely (6). Careful analysis of radiographs is essential. The type of fracture and pattern of fracture lines should be noted. The fracture may be simple, severely comminuted, or segmented. The possibility of joint involvement in the fracture must be considered, and if it is found, the surgeon should recognize possible impaction of metaphyseal bone and a possible bone defect after fracture reduction.

We perform computed tomography (CT) with three-dimensional reconstruction for all intra-articular fractures. CT provides more detailed recognition of the fracture and allows proper planning of the treatment and adequate fixation for specific types of injury. This way, we are usually able to decide before surgery if there is a need for bone graft or bone substitute during the reconstruction.

If, during the initial evaluation of the patient, suspicion of vascular injury arises, the vascular surgeon is summoned, and angiography or CT angiography is performed preoperatively. At times, surgical intervention is performed by the orthopedic surgeon together with the vascular surgeon. It may be necessary to provide fixation of bone fragments prior to vascular repair (7).

In cases of open fractures, systemic antibiotic treatment begins in the emergency room; the decision regarding types of antibiotics or their combination is made according to the severity of the injury.

In patients with involvement of one limb only, it is very important to also investigate the uninjured side. There may be bilateral knee varus or rotational malalignment, and anatomical correction of this peculiarity may cause limb asymmetry in the future. If possible, these aspects should be discussed with the patient before treatment.

In cases of severe bone defect due to severe comminution or loss of bone fragment at the time of injury, it is crucial to measure the length of the normal limb in order to restore original length during treatment. Usually, we perform CT measurement of the length during the postoperative period in order to acquire precise parameters. However, at the time of surgery we use the simple method of measurement between two anatomical landmarks in order to predict the size of the frame and possible lengthening in the future, without having to later remodel the external fixator.

After fracture reduction and its primary stabilization into the frame, we perform final evaluation of the injured limb under general or regional anesthesia. It is of primary importance to check the stability of adjacent joints and to decide about extending the fixator over the unstable joint or performing arthroscopy. Then, careful additional investigation of the wound should be carried out and a decision made about soft tissue management.

CLASSIFICATION OF OPEN FRACTURES

Many systems of classification of open fractures have been introduced in the last 30 years. Gustilo and Anderson's classification is considered the gold standard today (8). They published their classification in 1976 and modified the system in 1984. However, in the past 20 years, surgical techniques and approaches to treatment of open fractures have changed considerably. New internal and external fixation devices have been developed and many old concepts modified. Gustilo and Anderson's classification, built upon wound size, periosteal soft tissue damage, periosteal stripping, and vascular injury, no longer reflects modern guidelines for treatment (9).

Another important classification, described by Tscherne and Gotzen and revised by the Association for the Study of Internal Fixation (AO-ASIF) group, includes both closed and open injuries, muscle-tendon injury, and neurovascular injury (10). Despite the emphasis on the importance of soft tissue injury in this classification, no consideration is given to contamination. Furthermore, in practice, it is very difficult to quantify the degree of soft tissue injury, which this system requires.

We have devised our own system of open fracture classification and would like to introduce it here. Our scheme is based upon practical estimation of wound, bone, and vascular conditions as well as extent of contamination.

We have removed wound size from our classification because that particular detail does not influence the modern treatment approach. This is evident in fractures considered to be type Gustilo IIIA, in which internal fixation is often recommended (11). In our opinion, the crucial factor is the status of the soft tissue at injury and following debridement and how its condition may compromise the limb in specific types of fixation.

Shtarker-Volpin Classification (WBVC System)

W: Wound Condition (soft tissue)

1. Simple wound—punctiforme or linear wound without crush of adjoining soft tissue
2. Soft tissue crush without insufficiency after debridement
3. Partial loss of soft tissue—wound closure is technically impossible
4. Massive loss of soft tissue

B: Bone condition

1. Simple fracture—two or three part (butterfly) fracture
2. Comminuted fracture (more than three fragments)
3. Segmented fracture
4. Bone loss or high velocity fracture (bone burst)

V: Vascular and neurological condition

1. *No* vascular injury
2. Severe swelling of limb without compartment syndrome, possible blisters
3. Compartment syndrome
4. Vascular injury requiring repair

C: Contamination

1. No visual contamination
2. Visual contamination of soft tissue only
3. Visual contamination of soft tissue and bone
4. Severe contamination, farm injuries, or delayed arrival of untreated infected open fractures

Therefore, types W1 and W2 may be suitable for internal fixation, but type W3, even in uncomplicated degrees of BVC, should be considered for external fixation only. Type W4 presupposes acute shortening of the segment or complicated soft tissue reconstruction in order to achieve appropriate bone covering (Table 1).

Type B1, even with a butterfly fragment, theoretically may be treated by any method of fixation. Even type B2 fractures with severe comminution may be treated with an intramedullary nail or, for example, locked compression plate fixation (12). However, the combination of this type of fracture with a problematic wound condition such as W3, and especially W4, is an absolute indication for use of external fixation. Fracture types B3 and B4 are better treated by circular external fixation, without taking into account other parameters.

The Ilizarov external fixator, or other comparative circular device, allows three-dimensional restoration of bone alignment, with stable fixation and desired length of the segment. Its versatility makes it possible to perform additional rotational or axis corrections and, if necessary, to stabilize adjacent joints. Fixation of small bone fragments and application

Table 1 Shtarker-Volpin Classification (WBVC System)

Parameters and Their Degree	W Wound Condition (Soft Tissue)	B Bone Condition	V Vascular and Neurological Condition	C Contamination
1	Simple wound—punctiforme or linear wound without crush of adjoining soft tissue	Simple fracture—two or three part (butterfly) fracture	No neurovascular injury	No visual contamination
2	Soft tissue crush without insufficiency after debridement	Comminuted fracture (more than three fragments)	Severe swelling of limb without compartment syndrome, possible blisters	Visual contamination of soft tissue only
3	Partial loss of soft tissue—primary wound closure is technically impossible	Segmental fracture	Compartment syndrome	Visual contamination of soft tissue and bone
4	Massive loss of soft tissue	Bone loss or high velocity fracture (bone burst)	Vascular injury requiring repair	Severe contamination, farm injuries, or delayed arrival of untreated, infected open fractures

of compression on them are possible by use of thin wires with stoppers (olives). In many B4 cases, there is a need for bone transport either after initial loss of bone at trauma, or after debridement and removal of unviable bone fragments. Ilizarov external fixator is the device of choice for completion of these procedures (13). Although a unilateral external fixator may be successfully applied as temporary fixation for 2 to 14 days, it should later be replaced by a circular device as part of the definitive treatment.

The vascular condition of the limb may cause serious complications. Even uncomplicated V2 injuries may require delay in internal fixation due to difficulties in wound closure. Development of a compartment syndrome (V3) necessitates wide fasciotomy, making internal fixation inappropriate (Fig. 1C) (14). If arterial damage (V4) was diagnosed and repaired, then prophylactic fasciotomy is routinely performed because serious swelling is to be expected after surgery (15). Stable bone fixation is imperative in order to allow normal healing of an injured artery. External fixation in these cases eliminates the need for cast fixation and allows good access to the limb segment for monitoring and wound treatment. On a temporary basis, unilateral fixation may be more convenient in this situation; even transbridging fixation over the joints may provide enough stable fixations in this case (16).

All open fractures should be considered as contaminated. If the wound looks clean initially, the fracture may be considered C1 and may even tolerate a loose wound closure for approximation (17). Contamination of soft tissue (C2) requires debridement of the tissues and the use of pulsatile lavage. In these circumstances many surgeons would use, following debridement, internal fixation if the wound is W1 or W2 (9,12). In patients with C3 injuries, resection of the grossly contaminated bone may be the best choice. In such conditions, initial wound closure should not be performed and external fixation is obligatory.

The C4 group demands a very intensive and meticulous approach to treatment: bone resection followed by external fixation and aggressive systemic antibiotic treatment seems to be the only choice in many cases. One of the following combinations—C3 to C4 with W3 to W4, B3 to B4, or V3 to V4—converts an open fracture into a serious problem that demands a complex multidisciplinary approach and the use of external fixation.

LONG BONE FRACTURES

In the case of tibial fracture that cannot be treated with a cast or internal fixation (for example, our classes W3–W4, B3–B4, V2–V4, and C3–C4), we use circular Ilizarov external fixation as a definitive mode of treatment. In some other cases, the frame may be the treatment of choice following discussion with the patient.

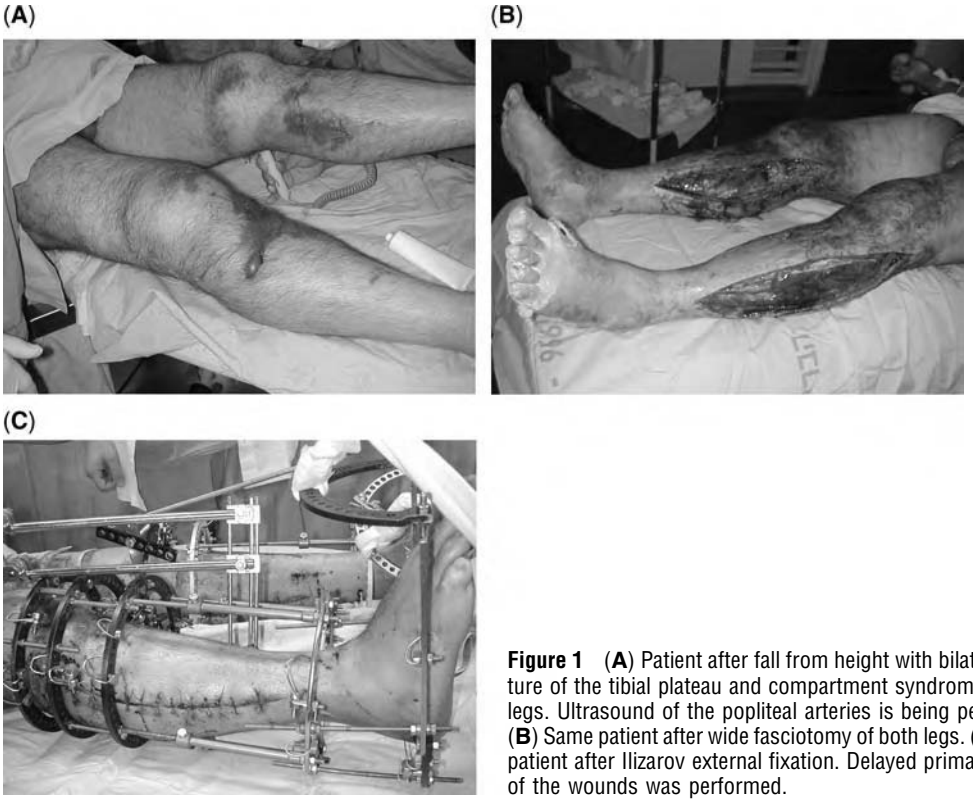

Figure 1 (**A**) Patient after fall from height with bilateral fracture of the tibial plateau and compartment syndrome of both legs. Ultrasound of the popliteal arteries is being performed. (**B**) Same patient after wide fasciotomy of both legs. (**C**) Same patient after Ilizarov external fixation. Delayed primary suture of the wounds was performed.

In closed fractures without vascular compromise, when a decision to use a circular frame has been made, it is possible to begin treatment in the emergency room by applying skeletal traction through the calcaneus. A definitive procedure must/should be performed in the next 24 to 48 hours. In open fractures or in cases with vascular compromise (V3–V4), surgery should be performed in the first hours after admission to hospital.

Careful planning is essential for success in each individual case. The surgeon should choose the appropriate ring size and construct a stable but not too bulky frame. The inner border of ring-to-skin distance should be at least 3 cm in all perimeters of the widest part of the limb segment, to allow for swelling in the early postoperative period. A four-ring frame composition is usually stable enough for fracture fixation and even permits weight bearing. In B3 to B4 injuries, the device may include five or even six rings (Fig. 2C). In order to allow a better range of motion of the knee joint, the proximal ring should be a 5/8-ring. At the level of comminution, carbon rings are more suitable than metal because they allow for better visualization of the fracture on X ray. Two rings above the fracture and two below is, mechanically, the most stable composition. It is also possible to use cubes with half pins as a substitute for additional rings.

In all cases of tibial fractures, the use of a fracture table makes the procedure easier to perform. After insertion of temporary skeletal traction through the calcaneus, the limb is positioned on the fracture table. The skeletal traction clamp is fixed in the clamp adaptor of the fracture table and then an initial traction is applied. In this way, exposure is maximized for both surgery and C-arm imaging. When initial reduction is performed, it is very important to control the plane and the rotational alignment of the limb segment.

Many different combinations of wires, pins, and frames have been introduced and are in use today. We have had success with the following technique. Fixation of the frame to bone begins with insertion of a half pin and its fixation to the most proximal ring—the ''reference pin.'' This is done by an initial insertion of a 1.8 mm Ilizarov wire through the fibular head, from the lateral to the anteromedial side of the tibia. Then, with a 4.5 mm cannulated drill, a hole is made from the tibial side through the proximal tibia and through the fibular head using this pin as a guide. A 6 mm half pin with a long cortical thread is inserted from medial

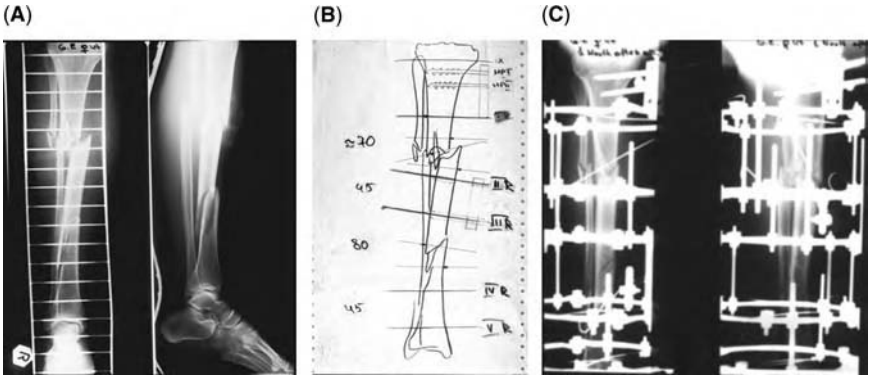

Figure 2 (**A**) X rays of severe comminuted segmented fracture of the tibia and fibula. (**B**) Preoperative planning of fracture fixation shown in Figure 2A. (**C**) Postoperative X Rays after fixation performed according to preoperative planning.

to lateral. Connection of this pin to the ring is achieved by its fixation to a one-hole post, allowing for three-dimensional reorientation of the prebuilt frame relative to the limb segment. After proper positioning of the frame, the half pin is fixed in the cube. Next, a distal reference olive wire is inserted through the distal tibia and fibula and attached and tensioned on the distal leg ring. The next step in the surgery is completion of reduction; olive wires allow very precise restoration of normal bone anatomy. When the frame is applied and the fracture is in an anatomic position, additional stabilization of the device is performed (Fig. 2). Overstabilization of the fracture may cause a delay in callus formation; however, the system should be stable enough to maintain the planned position of bone fragments and should tolerate forces applied to the limb during motion and weight bearing (18).

If the fixation is performed after a wide fasciotomy due to compartment syndrome or a vascular procedure, the surgeon should consider skin retraction during insertion of the wires. Before skin penetration by the wire and its further insertion into the bone, the skin should be stretched in order to approximate wound edges as much as possible. After pin insertion the skin pulls back, but later it allows delayed wound suture or decreases the necessary skin graft size.

In severely comminuted fractures or conditions with bone loss, wounds after debridement should be orientated transversally in order to facilitate closure during acute shortening and to avoid the additional need for soft tissue reconstruction. Loose wound closure by staples and rubber slings is then possible. It is vital to check the vascular condition of the limb after acute shortening, due to possible kinking of the blood vessels. Restoration of length of the injured segment begins gradually after the wound has filled with granulation tissue, and in the absence of purulent discharge. It is preferable to delay osteotomies for bone transport until complete epithelization or closure of the wound.

If the bone defect is too large for acute shortening, a wound with longitudinal orientation will heal more easily and more quickly. Bone ends should be covered by soft tissue. Also, in this situation bone transport may begin after filling of the wound by granulation, and the frame should be stabilized maximally. A stiffer frame decreases the potential for development of osteomyelitis (19). If it appears the least bit necessary, additional debridement of wound and bone should be done without further thought.

In some cases, the fracture may extend into the joints; however, if fracture fixation is stable enough and no joint instability is observed, the joints should be left free in order to allow early motion and a better functional outcome. Reduction of intra-articular fractures may be completed with arthroscopic assistance. Arthroscopy enables accurate reduction, removal of free cartilage or bone fragments, and treatment of other intra-articular injuries. During the arthroscopy, a large cannula should be inserted in the joint for drainage, in order to avoid compartment syndrome. If some concern about joint stability of the knee is present, the frame should be extended to the distal femur, and these rings connected by hinges in order to allow motion (Fig. 3A–D).

Distal femoral intra-articular fractures may be treated by the same approach. Frame composition for femur may be different. Attachment of a unilateral fixator to two or three

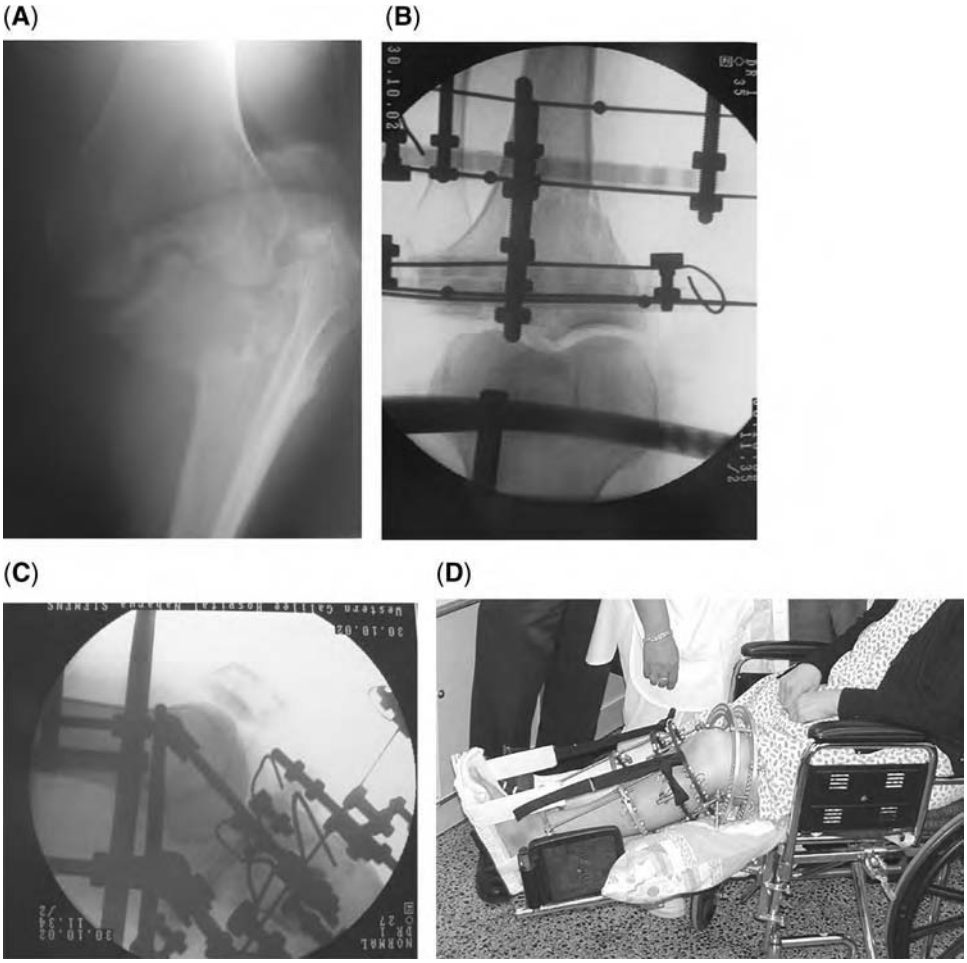

Figure 3 (**A**) X-ray of severe comminuted fracture dislocation of knee. (**B**) and (**C**) Postoperative A-P and lateral X-rays after reduction and Ilizarov external fixation of previously shown fracture. (**D**) Clinical picture of the above patient. The Ilizarov frame is extended to the proximal femur with use of knee hinges.

distal rings makes the frame more convenient for the patient. The use of carbon rings allows better visualization of the fracture on X rays. This assembling of a hybrid combined fixator may be simply performed by connecting the unilateral fixator to cubes with cut half pins. Even in cases of femoral bone transport, a unilateral frame can be used for bone lengthening. If the wound and bone conditions allow for it (W1–W2, B4, V1–V4, and C1–C2), an unreamed intramedullary distal femoral nail may be used as a guide for future bone transport.

CALCANEAL FRACTURES

Another useful application of the Ilizarov frame is in calcaneal fractures. External fixation allows for restoration of the shape of the calcaneus and provides stable fixation. Final results of treatment using this method are comparable with those of internal fixation. The reduction of the fracture does not require extensive exploration or internal fixation of bone, thus decreasing the possibility of complications (20). The rate of subtalar arthrosis is less or equal to other methods of treatment (21). Weight bearing on the injured limb may already be allowed several days after surgery.

Surgical treatment of calcaneal fractures begins with closed reduction of the fracture. This is done by initial application of skeletal traction through the most distal part of the

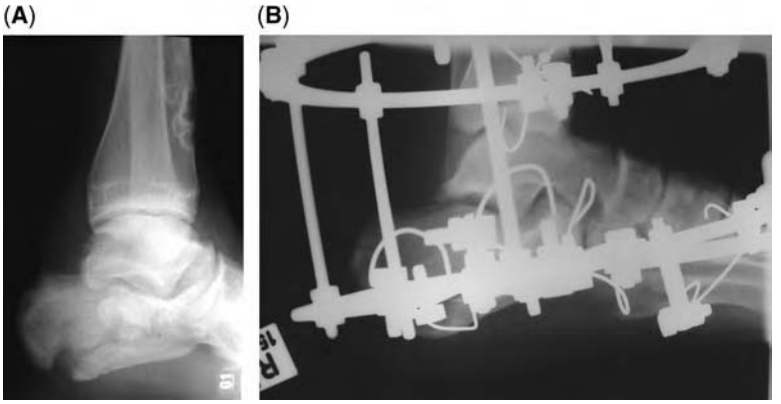

Figure 4 (**A**) X-ray of displaced intra-articular fracture of the calcaneus. (**B**) Postoperative X-ray following Ilizarov fixation of this calcaneal fracture.

tuber of the calcaneus. In cases of severe comminution, even subperiosteal insertion of wire may be suitable. The leg is then elevated 10 to 15 cm above the table (we usually use a folded drape), so that the ankle and foot are free. Then a sterile clamp is attached and the wire is tensed. A sterile cord is attached to the clamp and passed off the table for connection to 4 to 6 kg weight. The direction of the cord should be approximately 45° to the leg axis distally and downward. Manual reduction of fracture is performed by squeezing the heel in both hands and applying pressure from medial and lateral sides distally and backward. It is important to keep the ankle joint in mild dorsal flexion during the manipulation of the fracture. This entire procedure is performed under X ray control with C-arm. After restoration of the Böller angle, reduction of depressions in subtalar joint is performed through a minimal incision or percutaneously with bone tamps. Upon completion of reduction, the Ilizarov frame is applied (Fig. 4A and B). The external fixator is assembled with a foot ring attached to a ring block across the ankle joint. Previously induced traction forces, as well as their direction, should be maintained with the Ilizarov frame by passing the wires through the distal tibia, metatarsals, and finally through the calcaneus. Then skeletal traction may be removed and additional olive wires inserted into the calcaneus. These wires apply side compression on broken bone in order to decrease its widening and restore shape. Moreover, fixation of bone fragments and articular parts is achieved by their multidirectional insertion. After further stabilization of the frame by the addition of half pins and wires to proximal rings, bone substitutes may be injected into cavities that appear on fluoroscopy. At the completion of surgery, the subtalar joint should be distracted by 6 to 8 mm.

POLYTRAUMA PATIENTS

Polytrauma extends the indications for use of external fixation and especially the Ilizarov frame. Even relatively simple types of fractures of both lower limbs are better treated by circular frames because they allow early ambulation and weight bearing by the patient. Because these patients need to bear weight on their upper extremities when using crutches or a walker, it is preferable to provide stable fixation to fractures of the upper limb by circular external fixation in presence of lower limb fractures (Fig. 5A–C).

COMPLICATIONS

Pin tract infection (PTI) is the most common complication of external fixation. Its rate varies from 10% to 50% according to different authors (22). Usually, superficial infection appears in areas of mobile skin near the joints or due to excessive tension of the skin during bone transport and lengthening. Careful local treatment and prevention of overtension of the skin helps avoid this complication. If superficial PTI has appeared, a small incision around the pin under local anesthesia and oral antibiotic treatment can quickly eliminate infection (23). Sometimes,

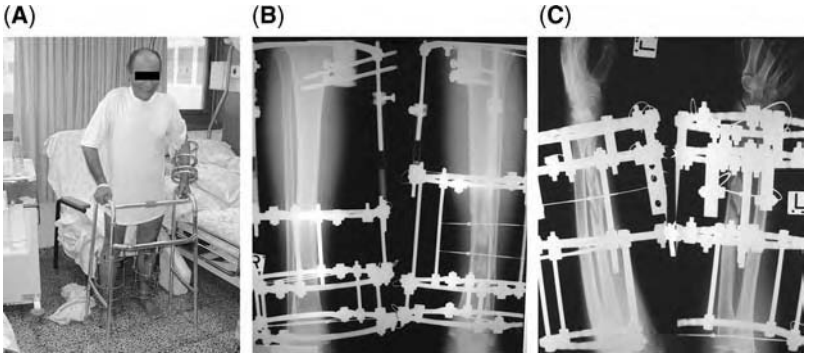

Figure 5 (**A**) Multiple trauma patient with fractures of both legs and left forearm. Of note, the patient is able to ambulate independently. (**B**) Postoperative X-ray of both legs after anatomic reduction. (**C**) Postoperative X-ray of left forearm after anatomic reduction.

in cases of prolonged or recurrent infection, exchange of the wire or half pin is necessary. Untreated cases of PTI may cause bone involvement and development of pin tract osteomyelitis. Such cases require aggressive surgical treatment with curettage of the pin canal and intravenous antibiotic treatment.

Because the lateral aspect of the proximal leg above the fibular head is the most common place for PTI, we avoid insertion of wires in this region and use a medial half pin connecting the tibia and fibula, as described earlier.

Another possible complication is iatrogenic nerve injury, although the chance of this complication can be decreased with proper anatomic position of wires and pins and correct technique of insertion. Use of a nerve stimulator during surgery may be very helpful. We do not use long acting muscular relaxants during the surgery in order to visualize nerve irritation, if present. Iatrogenic vascular injury is a very rare complication and should not occur if the surgeon follows accepted, anatomically based rules of fixator application.

Malposition of the fracture may require revision of external fixation and additional acute or gradual correction. Delay in callus appearance may require treatment by either gradual compression of the fracture or just the opposite dynamization of device. In cases of nonunion bone, grafting may be the procedure of choice (24,25).

In order to avoid joint contractures, early and aggressive physical therapy with restoration of range of motion should be performed. Early weight bearing is the best prophylaxis against disuse osteoporosis.

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6 Wire Ring Fixation of Complex Tibial Plateau Fractures

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INTRODUCTION

Complex tibial plateau fractures are typically the result of high-energy blunt-trauma. The mechanism of this trauma transfers force to bone, cartilage, and soft tissue structures creating a combination injury, requiring a well-planned treatment strategy. The majority of these plateau fractures are bicondylar and display significant displacement. The results of nonoperative treatment are poor, yet even in the most experienced hands, operative treatment using standard techniques can produce further injury to an already compromised soft tissue envelope. The keys to successful outcome are: (i) restoration of articular cartilage, (ii) preservation of the biology, (iii) alignment of the mechanical axis, (iv) restoration of joint stability, and (v) preservation of functional motion.

The technique of wire ring external fixation offers many advantages to achieve these goals. This chapter will concentrate on the protocols involved in patient evaluation, practical classification, treatment strategy, and detailed surgical techniques of articular reduction with minimal internal fixation and wire ring fixation of complex tibial plateau fractures. Techniques to minimize complications and improve functional outcomes will be stressed.

CLINICAL EVALUATION

Patients who sustain high-energy tibial plateau fractures require a comprehensive trauma evaluation. A mechanism of injury should be assessed. Was the fracture produced by a fall from standing (low energy) or a fall from a roof (high energy)? Polytrauma patients are appropriately stabilized and urgent needs are addressed.

Clinical examination of the extremity must include inspection for an open fracture or open knee joint. Realign the leg if gross deformity exists and cover all open wounds with a sterile dressing. Try to obtain X rays at this time to avoid plaster artifact. Does the patient have palpable pulses with the leg straight? If not, Doppler pulses need to be obtained. An ankle/brachial index should be checked if a vascular injury is suspected.

Evaluate the neurological function. Common peroneal nerve injury at the fibula neck with resultant foot weakness is not uncommon. Palpate the compartments of the leg. Do they feel very tight? Are they severely tender? Is there pain with passive toe movement? These may be early signs of compartment syndrome and need aggressive repeat clinical evaluation and surgical release. Comatose patients require more aggressive compartment pressure monitoring. Treatment for compartment syndrome and a tibial plateau fracture is four-compartment fasciotomy and a bridging four-pin fixator.

Other limb-threatening injuries that require urgent treatment include open plateau fractures, plateau fractures associated with multiple long bone fractures, and plateau fractures associated with vascular injuries.

Closed fractures may not initially demonstrate the full degree of soft tissue trauma involvement. Evolving swelling may produce fracture blisters that form after several days. A surgical incision made in that area, even before the blister forms, can produce poor wound healing, dehiscence, and eventual infection. External fixation wires or half-pins placed through fracture blisters are poorly tolerated and can result in soft tissue failure and infection. Definitive wire ring external fixation with reduction of the articular surface should most often be delayed.

Bicondylar plateau fractures are placed in a well-padded splint with side slabs and several layer of webril to add gentle, evenly distributed compression. Most high-energy

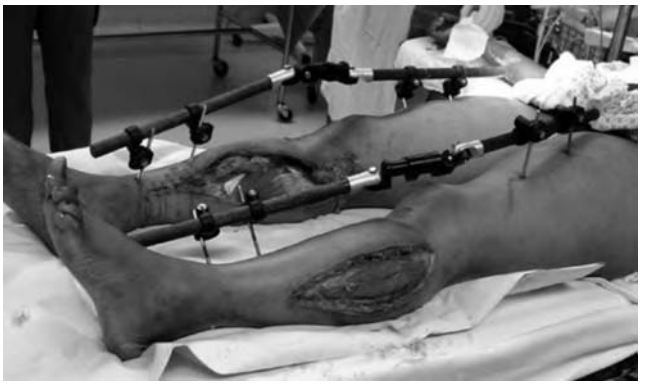

Figure 1 Temporary bridging knee external fixation. Half-pins are placed in the femur and tibia, distant from the planned operative site.

significantly displaced bicondylar tibial plateau fractures require temporizing bridging knee external fixation (1–3). Closed fractures are placed on the elective schedule for closed reduction and application of a bridging temporary fixator. This should be done in the subsequent 24 hours, but is not urgent unless associated with an open fracture, compartment syndrome, vascular injury, or multiple long bone fractures.

Temporary bridging fixation can take less than half-an-hour to apply and should not interfere with, but aid eventual definitive treatment. Tibial half-pins are placed anterior-medial and femoral half-pins are placed anterior-lateral to avoid vital anatomy (Fig. 1). Radiodense connection clamps are avoided over the fracture, to permit unobstructed imaging in the fixator. The external fixator half-pins should not be near future incisions or hardware. Gentle traction is applied with the fixator under anesthesia and fluoroscopy is used to confirm a provisional fracture reduction using ligamentotaxis. The fracture blisters should be un-roofed under sterile technique and covered with a dry dressing. Oral antibiotics should be started. The ankle is placed in a neutral position with a removable splint to support the leg compartments.

The patient is admitted for observation. Elevation of the leg is encouraged. Anteroposterior (AP) and lateral radiographs and a computed tomography (CT) scan with coronal and sagittal reconstructions are performed with the external fixator providing provisional distraction across the fracture. The studies are used to classify the injury and a plan is made for definitive treatment. Most patients are discharged and followed in the outpatient setting until the soft tissue is healed adequately to permit reduction of the articular surface and stabilization of the metaphyseal-diaphyseal junction. The soft tissue may take from 7 to 21 days to accommodate definitive treatment. In most cases, it is the soft tissue condition, fracture pattern, and comorbities that direct the definitive treatment plan.

CLASSIFICATION

The classification of tibial plateau fractures should help the orthopaedic surgeon in choosing a treatment strategy that best addresses the reduction. The Schatzker classification, which is still widely used, includes types I, II, and III, which are unicondylar with only lateral plateau involvement (4). Type IV patterns are unicondylar medial plateau fractures often associated with ligament injuries. High-energy unicondylar fractures occasionally benefit from ring fixation to prevent soft tissue complications (5). However, most unicondylar fractures are best treated with standard plating if the soft tissue allows.

Bicondylar fracture patterns and nonarticular patterns with complete metaphyseal-diaphyseal disruption are generally those patterns most amenable to using wire ring fixation. In the Schatzker classification, this would include the type V and VI patterns. Type V is bicondylar fracture with a central vertical split, which does not extend into the articular surface of either the medial or lateral condyle. The split usually extends into the intercondylar eminence, avoiding the articular surface (Fig. 2A). These fractures tend to have better outcomes than more complex patterns that directly involve the articular surface.

Most bicondylar fractures are Schatzker Type VI patterns with involvement of the articular surface. The pattern can vary widely and detailed analysis is required for each fracture (Fig. 2B). In the severely osteoporotic patient, the fracture pattern is generally more impacted

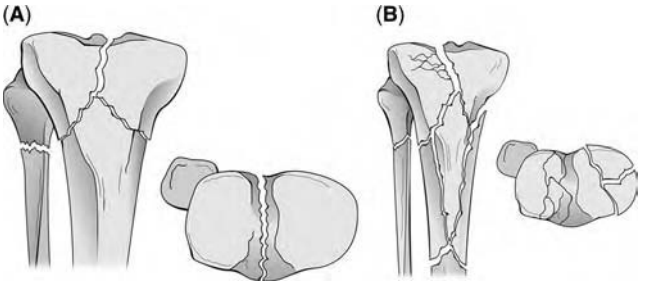

Figure 2 (**A**) Schatzker V tibial plateau fracture which is bicondylar with extension into the intercondylar eminence. (**B**) Schatzker VI tibial plateau fracture which is bicondylar with extension into the articular cartilage.

and can be limited to within 3 cm of the joint surface (no distal extension). This Schatzker VI pattern, with less than 3 cm of distal extension, is difficult to treat with wire ring fixation due to the close proximity to the joint surface. The fracture, which extends distally into the diaphysis, is more ideally suited for wire ring frames. Most Schatzker VI fractures have more comminution and impaction of the lateral plateau than the medial plateau. This can focus all the attention laterally, overlooking the medial articular displacement. A posterior-medial fracture of the medial plateau can be easily missed using standard radiographs (Fig. 3A). Often this occurs in the coronal plane and is best seen with a CT scan reconstruction (Fig. 3B). This medial plateau pattern requires a separate posterior-medial approach for reduction (see Treatment section). Although not specified by the classification, involvement of the tibial tubercle needs special attention. Separate fixation is usually required for the tubercle.

The other major classification is the AO/Orthopaedic Trauma Association (OTA) type. Type 41 A, B, and C fracture patterns are further subdivided into 1, 2, and 3 depending on the location and degree of comminution. The type A fracture pattern does not exist in the Schatzker classification scheme but it does have relevance in the discussion of wire ring fixation of proximal tibia fractures. The type A is a nonarticular pattern fracture with variable amounts of metaphyseal comminution (Fig. 4). These are often high energy (bumper injury) and can have significantly compromised soft tissue. Wire ring fixation is a very useful tool in the treatment of type A fractures. Type B fractures are unicondylar (partial articular) and correspond to the Schatzker 1, 2, 3, and 4 types. These patterns are most amenable to plating. Type C fractures are bicondylar with variable degrees of comminution corresponding to Schatzker V and VI patterns. The AO/OTA classification further divides the fractures into groups and subgroups. C1 fractures are bicondylar but with no articular involvement. C1 fractures correspond to the Schatzker V pattern. C2 fractures have simple articular and comminuted metaphyseal patterns and C3 have comminuted articular and metaphyseal patterns (Fig. 5). The AO/OTA classification is more descriptive and thus more useful in comparing outcomes of distinct subgroups (31).

A discussion of classification would be incomplete without mentioning the soft tissue-grading scheme. Open fractures are still routinely graded using the Gustilo classification (6,7). Type I open fractures have less than 1-cm wounds, usually created by an in to out puncture. Type II open injuries have wounds larger than 1 cm, but do not have extensive periosteal stripping or crushing of soft tissue. Type III injuries represent severe soft tissue injury with

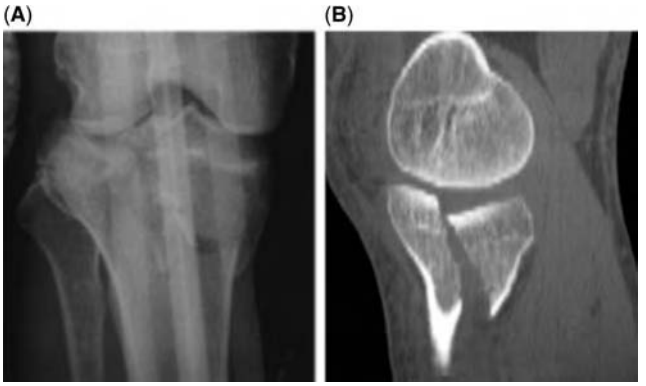

Figure 3 (**A**) Schatzker VI tibial plateau fracture distracted by temporizing fixator. (**B**) Schatzker VI tibial plateau fracture with posterior medial plateau fracture which is best seen by coronal computed tomography images.

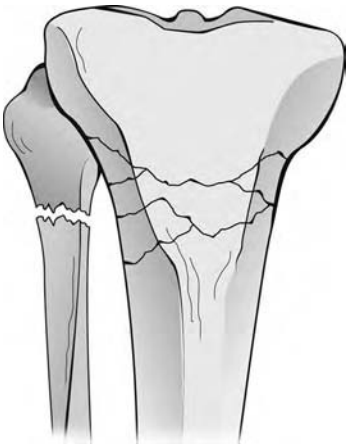

Figure 4 AO/OTA 41 A—nonarticular tibial plateau fracture.

extensive contamination and periosteal stripping. Grade IIIA does not require a muscle flap, grade IIIB requires a muscle flap, and grade IIIC requires artery repair for limb salvage.

The Tscherne classification grades the condition of the soft tissue in closed fractures (8). It is useful in directing the timing and method of fracture treatment but can take several hours to days to fully evaluate the extent of damage at the plateau level. Grade 0 injuries occur from indirect trauma and have negligible soft tissue damage. Grade 1 displays superficial contusions or abrasions of the soft tissue over the fracture. Grade 2 injuries have significant muscle contusion and full thickened abrasions with a risk for compartment syndrome. Grade 3 injuries have extensive crushing with subcutaneous avulsion or degloving and established compartment syndrome.

Finally, a host classification needs to address the health of the patient and clarify the increased risks associated with those comorbities. We have modified the Cierny/Mater classification used to treat elective osteomyelitis (9). In the modified system, the type A host is healthy with no preexisting medical problems locally or systemically compromising the injury. The type B host has moderately compromised health that affects healing, such as early vascular disease, smoking, controlled diabetes, obesity, or osteoporosis. The treatment strategy needs to address the compromised B host to minimize surgical injury but the goals of treatment remain the same. Type C hosts are those patients with multiple comorbities, such as severe diabetes on dialysis, in which the treatment strategy and the goals of treatment are altered. The type C patient has preexisting conditions that may make open articular reduction or wire placement an unwise choice, and limit the goals of articular reduction or perfect mechanical axis alignment. The treatment should not make the condition worse.

TREATMENT OPTIONS

The treatment options for tibial plateau fractures depend on the fracture pattern, energy of the injury, the soft tissue condition, and the type of host. No one treatment option is appropriate for all types of tibial plateau fractures.

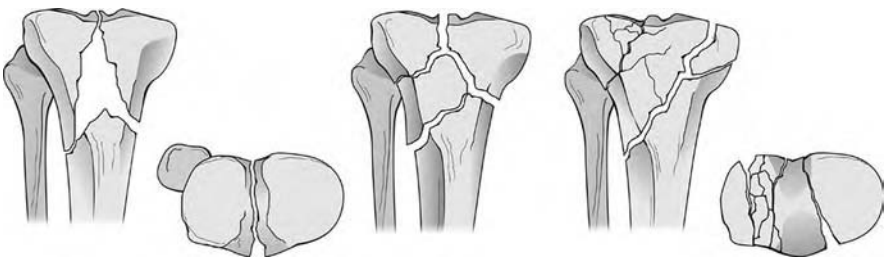

Figure 5 AO/OTA 41 C1 fracture—simple articular and simple metaphysis; AO/OTA 41 C2 fracture—simple articular and complex metaphysis; AO/OTA 41 C3 fracture—complex articular and metaphysis.

The treatment should start with preservation of the biology. No surgical trauma should be created through a compromised soft tissue envelope. Bridging the knee with a simple external fixation and waiting is an excellent means of initial stabilization, regardless of the choice for definitive treatment. It is unwise to initiate definitive treatment until the extent of the soft-tissue injury is fully declared. The patient can be sent home and followed-up as an outpatient until the appropriate time for definitive treatment.

Nonoperative treatment, including cast-bracing and traction, has poor results in the treatment of bicondylar tibial plateau fractures (10,11). It does not achieve the goals of treatment but may have some justification in the C host, when open operative options are not likely to be successful. When appropriate for general anesthesia, the C host may undergo bridging external fixation to produce a closed reduction. This can become the definite treatment for the C host when left in place for six to eight weeks. This is followed by a hinged knee brace. In all other patients, the definitive treatment requires the delayed planned reconstruction of the articular surface and the mechanical axis.

Articular cartilage incongruity of greater than 3 mm elevates joint contact pressures, which leads to premature arthritis. Ali et al. (12) had higher failure rates with articular cartilage step-off of greater than 3 mm or malalignment greater than 5°. Kumar and Whittle (13) presented 55 patients with bicondylar fractures in which 45 patients had anatomic reduction with knee scores average of 83. Conversely, nine patients did not undergo open restoration of the articular cartilage and were left with nonanatomic reductions. The average knee score in this group of patients was 52. Mikulak et al. (14) showed that his patients with better articular reductions in combination with the use of wire ring fixation had less arthritis changes. Articular cartilage incongruity is not well tolerated and leaves the surgeons with few options for reconstruction in the young patient. Residual displacement of more than 3 mm should not be accepted unless other comorbidities prevent adequate articular reduction.

On the other hand, massive exposure with periosteal stripping of the tibia plateau and dual plating, particularly through a single midline approach, has resulted in unacceptable outcomes. Experienced traumatologists using this technique in several series have reported infection, wound breakdown, and subsequent osteomyelitis ranging from 25% to 87% (15–17). Infected nonunions of the tibial plateau can result in chronic osteomyelitis and bone loss and are challenging limb salvage problems. Unfortunately, this can result in an above-knee amputation. Preventing this problem should be a high priority when considering options for bicondylar plateau fractures.

Two incisions to reduce the articular surface appear to be much better tolerated than a single anterior approach to both condyles. Barie et al. (2) reports on 47 patients with good results utilizing two incisions (anterior-lateral and posterior-medial), less periosteal stripping, and dual plating. Their infection rates are reported at less than 11%. Two incisions, which minimize periosteal disruption anteriorly, preserve the local biology and result in a better healing environment. Two incisions can be utilized safely to approach the articular reduction but this does not require that dual plating be used to stabilize the bone. Subchondral screws can stabilize the articular surface while the metaphysis is linked to the diaphysis with wire ring fixation.

Wire ring fixation of bicondylar tibial plateau fractures compared to open reduction and internal fixation demonstrates similar knee range of motion, time to union, and surgery time. The number of wound complications, deep infections, and time to full weight bearing, however, is reduced with wire ring fixation (13,18) compared to standard dual plating techniques.

The advent of locked plating techniques has suggested that bicondylar fractures can be plated from the lateral side alone with equal stability compared to dual standard plates. However, Mueller in 2003, using cadaver models of bicondylar fractures with no medial comminution and anatomic reduction of the medial cortical fracture (seldom seen clinically), compared dual plating to single lateral locked plating. Axial loading produced more medial displacement with the locked plate alone than with dual standard plating (19). The paper still recommended locked plates for bicondylar fractures but the data suggested less stability with locked plating alone. Wilson in 2003 describes his experience with locked less invasive stabilization system plates with high rates of soft tissue problems and infections due to plate-prominence (20). This produced a 14% rate of infection due to metal prominence, despite being a percutaneous procedure. It has been our experience that grade IIIA open plateau fractures can be easily converted to Grade IIIB fracture, requiring a muscle flap, by the introduction of a plate in proximity to the open area (Limb Lengthening and Reconstruction Society

Specialty Day 2003). This has been termed ''the grade IIIA to IIIB conversion'' and is avoided by using wire ring fixation for most high-energy open plateau fractures. This also applies to plateau fractures associated with compartment syndrome when the plates may have direct contact to the fasciotomy sites.

The advantages to wire ring fixation includes better biology at the metaphyseal-diaphyseal junction. Even percutaneous plating requires metal plates across the fracture site and disruption of at least part of the periosteum. Metal itself at the fracture site has a higher rate of infection because the plate is avascular no matter the percutaneous method of introduction. The minimal infected dose of bacteria is 10,000 times lower in the presence of a metal implant (21,22). Wire ring fixation does not damage the periosteum, does not add volume to a compromised sort-tissue-envelope, does not promote bacteria at the fracture, and eliminates the need for future plate removal.

Angular deformity of the proximal tibia is not well tolerated. More than 5° of varus/valgus causes significant mechanical axis deviation producing shear forces on an already traumatized articular surface. The newer wire ring fixators, such as the Taylor Spatial Frame (TSF) (Smith-Nephew, Memphis, Tennessee, U.S.A.), permits computer-assisted adjustment of the mechanical axis during the postoperative period. The alignment can be fine tuned in the post-operative period when hip to ankle X rays are more easily attainable, without repeat surgery. This is not limited to angulation but can adjust rotation, translation, and length if needed. Plating techniques do not permit postoperative adjustments.

Another advantage to wire fixation is that early weight bearing is possible without fixation failure (12). This can be critical to recovery of pulmonary function in patients with multiple injuries. Early weight bearing prevents disuse changes in the bone and soft tissue. Weight bearing reduces peripheral vascular resistance, increases vascularity to the leg, and promotes fracture healing (23). Wire ring fixators are designed to prevent shear and allow axial compression with weight bearing. Applying a stable fixator is crucial to support early weight bearing. Ali et al. (24) has shown that a four-wire construct of ring fixation of the proximal tibia is as stable as dual plating in the bicondylar fracture. Wire ring fixation can be axially compressed or dynamized after the surgery to stimulate bone union during the healing process. Plates are static and cannot increase compression during healing.

The indications for wire ring fixation in tibial plateau fractures are in high-energy bicondylar (C1, C2, and C2) or nonarticular (A2 and A3) patterns, particularly those patients who have soft tissue problems. This includes patients with significant fracture blisters, abrasions, grade II and IIIA open fractures, and patients with compartment syndrome. Grade IIIB open fractures that require flaps for limb salvage may be treated with plating or wire ring fixation. Temporary bridging fixation and early debridement are a crucial first step to plan bone stabilization and prompt soft tissue coverage. Generally, fractures that extend distally through the metaphysis into the shaft are prime candidates for wire ring fixation. Patients that have comorbidities (including smokers) have less ability to heal and are in poor general health and may be better served with ring fixation. This includes Type B and C hosts when possible.

SURGICAL TECHNIQUES

The surgical technique of fixation of the complex tibial plateau includes: (i) elevation and reduction of the articular surface and/or tibial tubercle, (ii) stabilization of the articular surface and tubercle, (iii) application of the ring–ring fixator, (iv) provisional reduction of the metaphyseal/diaphyseal junction, and (v) fine tune the reduction of the articular surface to the shaft.

The process of reduction of the articular surface is usually delayed for 7 to 21 days depending on the condition of the soft tissue. Distraction X rays and CT scans should be scrutinized to determine the best approach to reduce the articular surface. The surgery is performed on a flat radiolucent table. The leg is elevated on a nonsterile bump in order to obtain unobstructed lateral images. It is preferable to remove and sterilize the temporary external fixator leaving the half-pins in the femur and tibia. The half-pins are prepped into the field and the fixator is reapplied sterilely as a femoral distracter for the articular reduction. A sterile tourniquet may be placed between the femoral half-pins while the articular reduction is addressed (Fig. 6).

The articular reduction depends on the fracture pattern. AO type A type fractures do not require articular reduction. Schatzker V or AO type C1 fractures do not usually require open

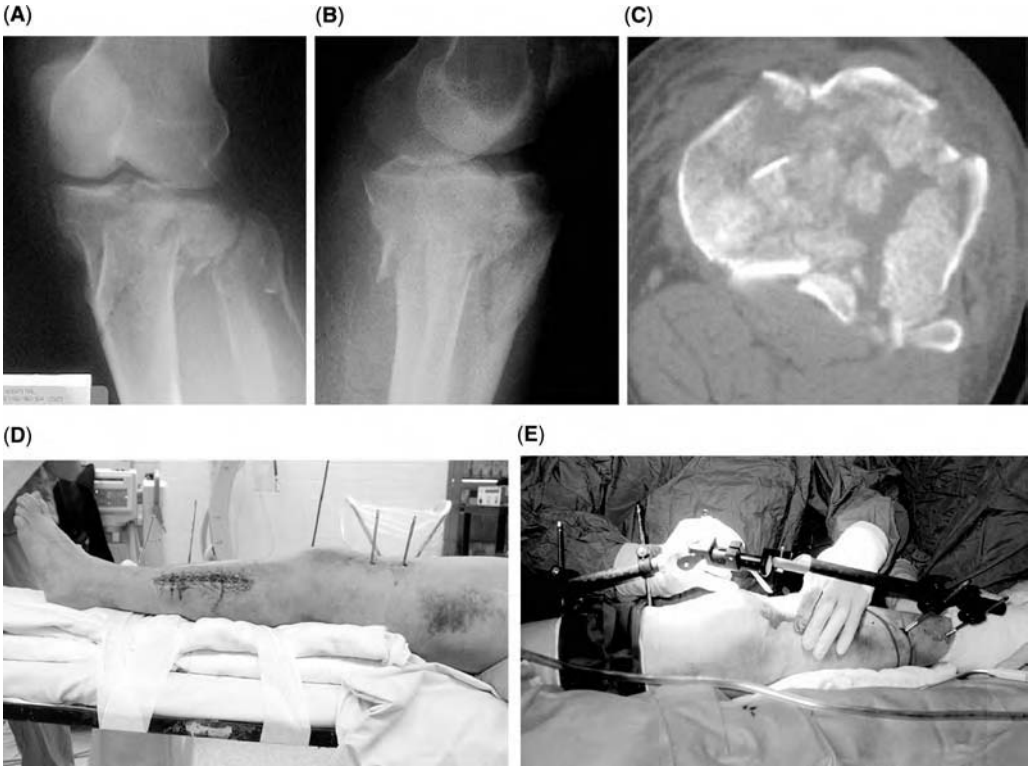

Figure 6 (**A** and **B**) High-energy bicondylar Schatzker VI (41 C3), tibial plateau fracture with compartment syndrome. (**C**) Computed tomography scan of complex plateau fracture. (**D**) Stage II—definitive treatment two weeks after compartment release and temporizing external fixation. Half-pins are prepped into the field and the fixator is removed and sterilized to be placed back on for intraoperative distraction. (**E**) Definitive articular reduction using the bridging fixator as a distraction device.

reduction of the articular surface but do require reduction the nonarticular portion of the plateau, and the intercondylar eminence. This can usually be accomplished with distraction from the bridging frame, placing temporary half-pins in each condyle, reducing the medial to the lateral side independently, and holding the reduction with a periarticular clamp. This is done under fluoroscopic guidance, followed by two lag screws from lateral to medial, just below the subchondral level. Nondisplaced Schatzker VI, AO types C2 and C3 patterns can be treated with similar subchondral lag screw, generally placed from the lateral to medial condyles under fluoroscopic guidance. Unfortunately, most of the bicondylar fractures do have displaced intra-articular fracture patterns, which require an open reduction of the joint surface.

The posterior-medial and anterior-lateral approaches are the most useful and direct methods to obtain the reduction of the articular surface. If only the lateral articular surface is displaced, then only an anterior-lateral approach is needed. *A midline approach is not recommended*.

If both plateaus have articular displacement, address the least comminuted side first. This is usually the medial plateau. The posterior/medial approach is the best way to visualize the problematic posterior-medial split. The Pes tendons are elevated proximally and the medial gastrocnemius is retracted posteriorly. The medial articular surface is best reduced by indirect reduction of the medial cortical fracture line. This is most easily accomplished with a small fragment antiglide plate placed posterior medially (Fig. 7A and B). The plate is under contoured and only one or two screws are required distal to the fracture to push and hold the medial plateau. Screws are *not* placed across to the lateral plateau that has not yet been reduced. The reduction is directed by fluoroscopic image and aided by reduction clamps, tamps, ball-spike-pusher, and distraction by the temporary frame. The plate lies under the pes tendons and seldom is longer than five holes. Another option is to reduce the fracture at the posterior medial cortex but to place a screw percutaneously from anterior to posterior to hold the reduction (Fig. 7C).

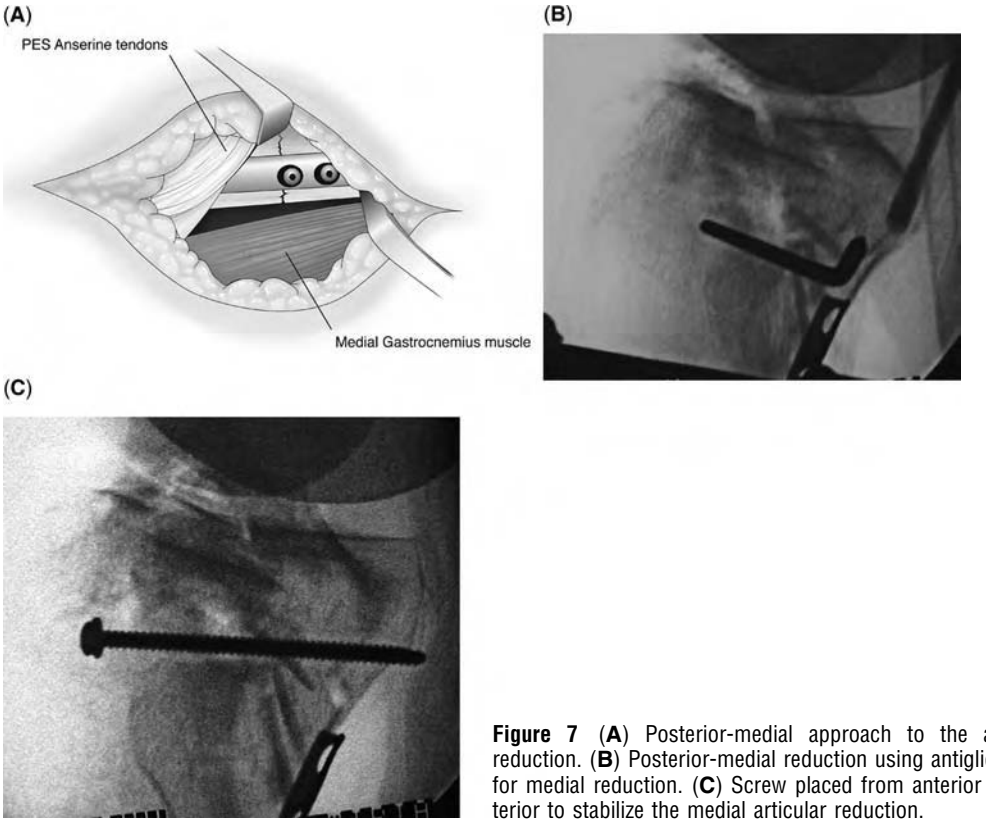

Figure 7 (**A**) Posterior-medial approach to the articular reduction. (**B**) Posterior-medial reduction using antiglide plate for medial reduction. (**C**) Screw placed from anterior to posterior to stabilize the medial articular reduction.

The posterior-medial approach has excellent soft tissue coverage in an area that is poorly tolerated by an olive wire. Attention is then turned toward the lateral plateau.

Many times the lateral approach is all that is needed because the medial articular surface is not displaced. We prefer a modified transverse incision, which permits a wide exposure of the articular surface but does expose the metaphysis (Fig. 8A). The incision is made just below the plateau but above the site for wire insertion. The tibial tubercle can be exposed with the anterior portion of the incision if needed. The anterior muscle compartment is elevated only just enough to reduce the articular surface of the lateral plateau. The lateral side of the joint is distracted with the temporary external fixator to prevent femoral crush down. The indirect reduction is preformed under fluoroscopy visualization (Fig. 8B and C). Curved elevates are very helpful. If reduction is not obtainable, a submeniscal approach can be done for direct visualization. Often a torn lateral meniscus is trapped in the articular fracture. Allograft bone graft or autograft is used to back fill the subchondral defect.

The elevated lateral plateau is then reduced to the medial plateau using large periarticular clamps. The lateral and medial plateau articular cartilage can be perfectly reduced, but if the condyles are malreduced with respect to each other, the alignment of the entire plateau will suffer. Malrotation of the condyles in the coronal plane can be avoided by ensuring equal joint spaces on AP fluoroscopic imaging tilted 10° caudally. Once the condyles have been reduced and held with periarticular clamps, the fragments are stabilized using lag or neutralization screws in the proximal 14 mm of subchondral bone (Fig. 8D). This author prefers 3.5-mm fully threaded pelvic screws as scaffolding rather than 7-mm cannulated screws. The 3.5-mm screws have better purchasing strength and less likely to split smaller fragments.

If the tibial tubercle is fractured, it should also be stabilized. One or two screws may be place through the tubercle from anterior to posterior. Alternatively, a 1/3 tubular plate can be use with a single screw in the tubercle and one distal to the tubercle for better purchase. The screws should *not* transfix the main metaphyseal-diaphyseal fracture, which may limit the reduction and dynamization that can be preformed later by the external fixator. Occasionally, a coronal split fracture into the diaphysis may be secured with a lag screw if

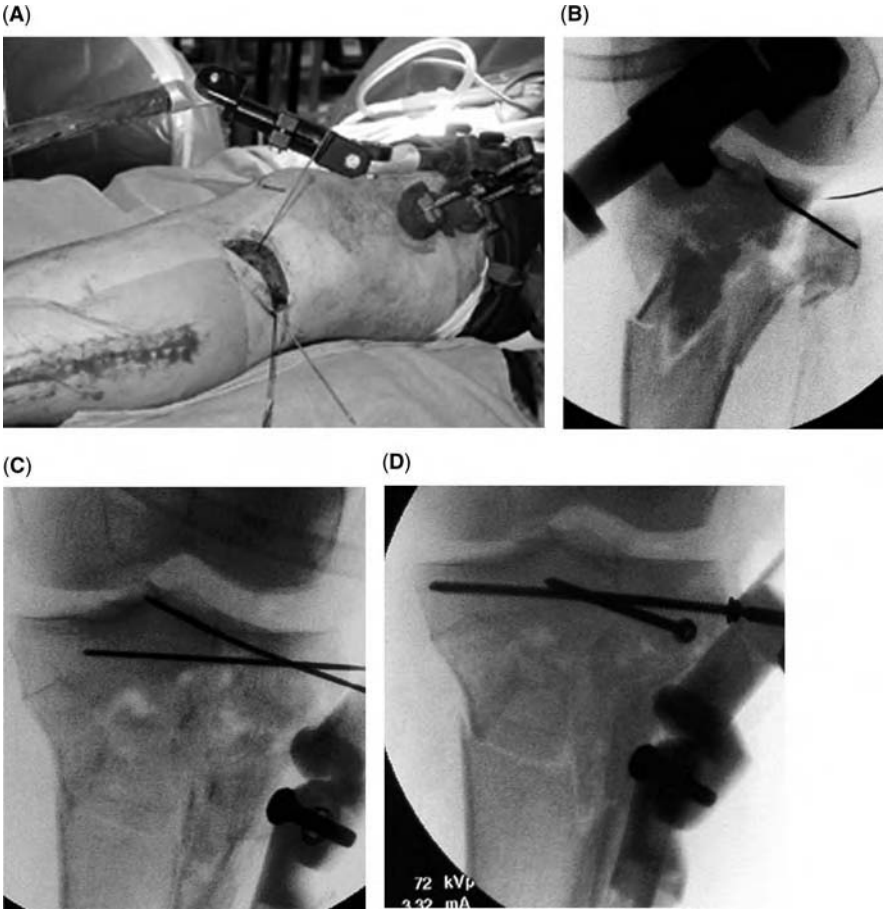

Figure 8 (**A**) Lateral exposure of the articular surface. (**B**) The lateral plateau articular comminution. (**C**) Reduction of the joint surface with temporary K-wires. (**D**) Articular cartilage stabilization with 3.5-mm pelvic screws.

it does not interfere with the reduction at the metaphyseal/diaphyseal junction later. When the articular surface and the tubercle have been restored, the tourniquet is released. All incisions are closed without tension using nylon sutures and the bridging external fixator is removed except for the two femoral half-pins. Attention is directed to the application of the wire ring external fixator.

Understanding the anatomy of the proximal tibia is important for safe and comfortable wire and half-pin placement. The recesses of the joint capsule of the knee extend 14 mm below the joint surface (25). It is important to place wires outside of the joint capsule to prevent future joint sepsis. Obviously, the popliteal fossa should be avoided. Other structures that should be avoided are the common peroneal nerve at the fibular neck, the patella tendon, and the pes anserine tendons. Transfixing the pes anserine tendons causes chronic pain and reduced knee motion. The anterior tibial artery pierces the interosseous membrane into the anterior compartment in the proximal 1/3 of the leg. It is at risk with wire placed too posterior in the anterior compartment. The center of the leg serves as the lower fixation block for the external fixator frame. Generally, half-pins are used at the second ring. The entire medial, anterior, and anterior-lateral surface of the tibia are safe zones used in half-pin fixation. Ninety-degree circumferential spread is possible from directly medial to anterior-laterally.

Preassemble the frame with appropriate soft tissue clearance, which also accommodates for postoperative swelling. A 180-mm diameter ring is the most common size used. For application of the TSF, I prefer that a single full ring be used at the plateau level and that a single full ring is used distally. A full ring is more stable at the plateau level but does sacrifice some

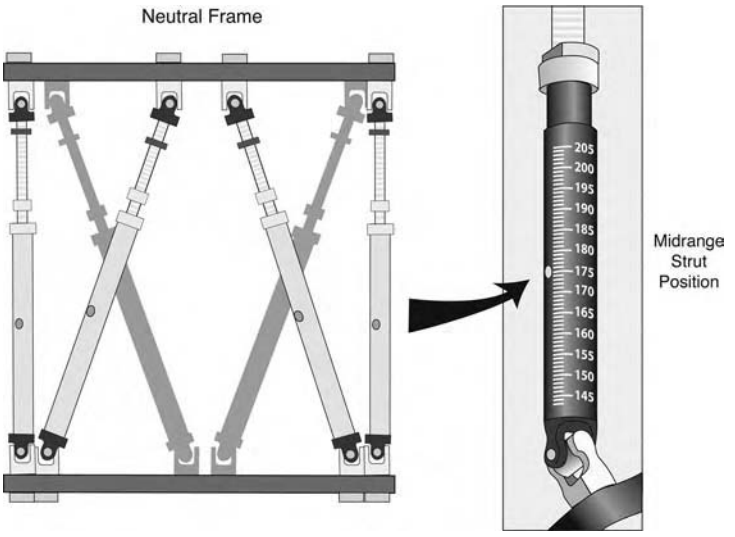

Figure 9 Neutral frame with struts adjusted to the midrange position.

postoperative knee flexion. A 2/3 ring at the plateau level is also an option, but should be used with more stable fracture patterns.

Start with a neutral frame in which both rings are parallel. Either all medium or all large fast-fix struts are used and locked at the midrange setting connecting the two rings (Fig. 9). Placing the struts at their midrange setting allows maximum adjustment for the reduction without having to exchange different size struts. Under fluoroscopic image, a reference olive wire is placed from lateral to medial, anterior to the fibula head, 15 mm below and parallel to the joint surface. The frame is placed over the foot and ankle, brought to the level of the reference wire and rotated until the master tab is positioned anteriorly. The ring is centered for appropriate soft tissue clearance and the wire is attached and tensioned to the proximal ring (Fig. 10A). This places the ring parallel to the joint surface and orthogonal to the proximal segment on the AP image.

A second olive wire is placed from posterior-medially (anterior to the pes anserine) to anterior-laterally to align and stabilize the ring on the lateral image. The proximal ring should now be orthogonal to the proximal tibia on the AP and lateral images and secured with two olive wires (Fig. 10B). Two wires are not stable enough to completely secure the tibial plateau but they are stable enough to provide a provisional stabilization of the proximal ring. This allows wires to be added later to the proximal ring in order to fine-tune the fracture alignment and stabilize the frame. Attention is turned to mounting the distal ring.

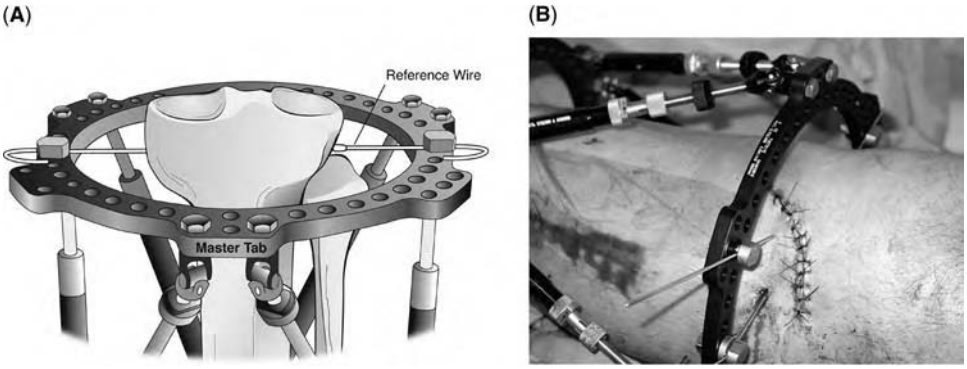

Figure 10 (**A**) Attach reference wire to the proximal ring with appropriate soft tissue clearance and master tab centered. (**B**) Second posterior/medial to anterior/lateral wire which secures the proximal ring in orthogonal position and avoids the incision.

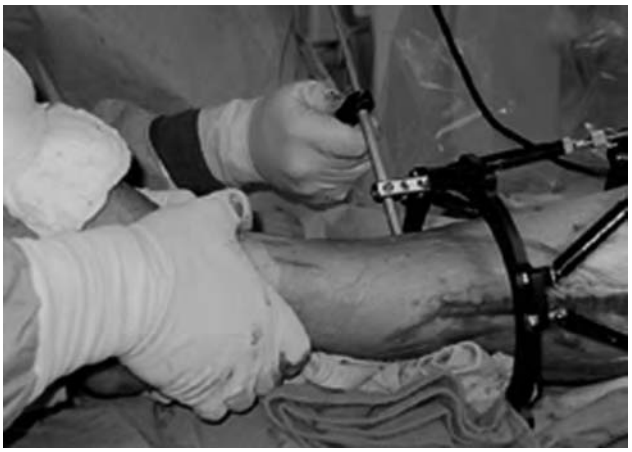

Figure 11 Manual distraction is held while distal half-pins are placed.

Manual traction is placed on the leg by an assistant in order to grossly realign the fracture as the distal ring is mounted. Two half-pins on the medial face of the tibia, one proximal to the ring and one distal, are inserted from the distal ring while manual traction is maintained (Fig. 11). With two wires proximally and two half-pins placed distally, the six frame fast struts are unlocked. An assistant holds the proximal ring and the surgeon pulls, angulates, and rotates the distal ring with the fracture reduction visualized under fluoroscopic image (Fig. 12). The fast struts are relocked. Translation may be difficult to achieve at this stage, but can be addressed in the proceeding steps.

Fine-tuning the reduction occurs while at the same time improving the stability of the fixation blocks above and below the metaphyseal-diaphyseal junction. At least two additional olive wires are placed proximally, creating a four-wire fixation block. One olive wire is usually placed through the fibular head into the tibia providing added lateral stability. The fourth proximal wire is a ''drop wire.'' It can also function as a fine-tuning reduction tool for translational displacement. This wire is secured from a post, distal to the proximal ring to increase stability of the proximal fixation block. Four half-pins are generally placed in the distal fixation block, including one anterior laterally, to create a 90° spread distally. The last half-pin is placed from the distal ring proximally and closest to the fracture. This half-pin may be placed parallel to the main fracture to serve as a steerage pin construct, which increases compression and reduces shear during weight bearing (Fig. 13). It is important *not* to cross the main fracture with a wire or half-pin, as this will obviate the ability to reduce and compress the main fracture line with the external fixator. Olive wires are not to be used as lag screws at the metaphyseal-diaphyseal fracture line. Adequate frame stability should now be obtained above and below the metaphyseal fracture.

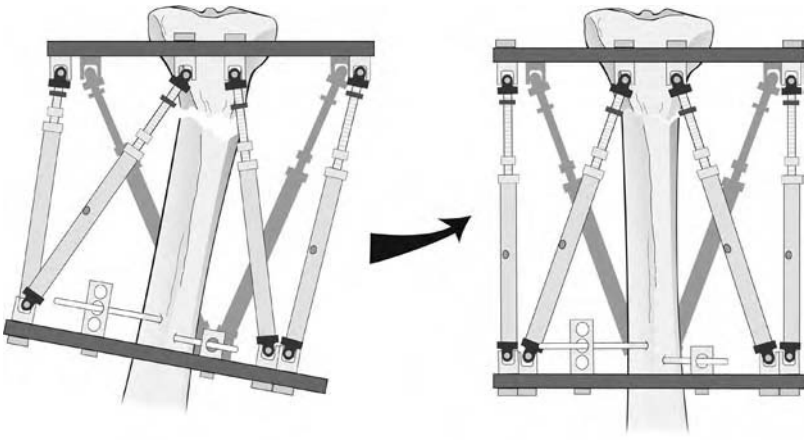

Figure 12 Unlock the six struts, and manually reduce the fracture under C-arm visualization, then relock the struts.

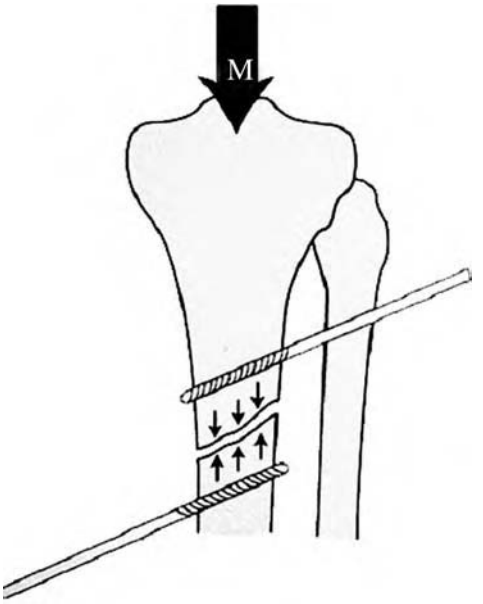

Figure 13 Steerage half-pins produces compression through the fracture with weight bearing.

Occasionally, it is advantageous to cross the knee with an external fixation bridge for up to six weeks (Fig. 14). For example, if the proximal fixation block is limited to one level or three wires because of the fracture pattern, early knee movement may cause fracture motion. Traumatic soft tissue damage, such as open fractures or compartment syndrome, may heal more quickly with delayed knee motion and a bridging knee fixator. Femoral articular fracture or ligament injuries at the knee with a plateau fracture benefit from bridging the knee for up to six weeks. This can be accomplished by attaching the two femoral half-pins from the temporizing frame to the proximal ring with a simple bar. Most ring fixators have a clamp, which attaches one or two straight bars to the proximal ring. This increases stability at the plateau thus improving early soft tissue and bone healing. Hutson (26) has demonstrated that removing the bridging frame by six weeks does not significantly decrease long-term knee motion in C-type plateau fractures.

Final reduction of the plateau can now be accomplished using the TSF computer-assisted technique and the ''Total Residual Correction'' (TRC) program. The details of the technique are beyond the scope of this chapter, but it does provide an excellent fracture reduction tool. The TRC can be initiated in the operating room if Internet assess is available using AP and lateral orthogonal images based on the proximal reference ring. The ability to translate and shorten the fracture during the TRC allows the oblique fracture at the metaphyseal-diaphyseal junction to be compressed perpendicularly to the major fragments, similar to compression

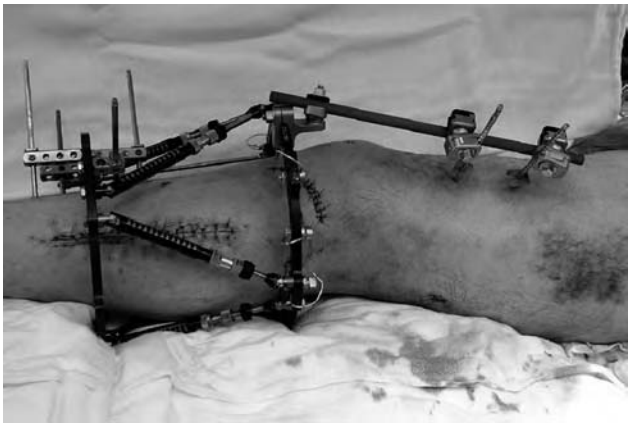

Figure 14 Final wire-fixator with temporary bridging knee frame due to soft tissue damage; the femoral extension is removed in four to six weeks.

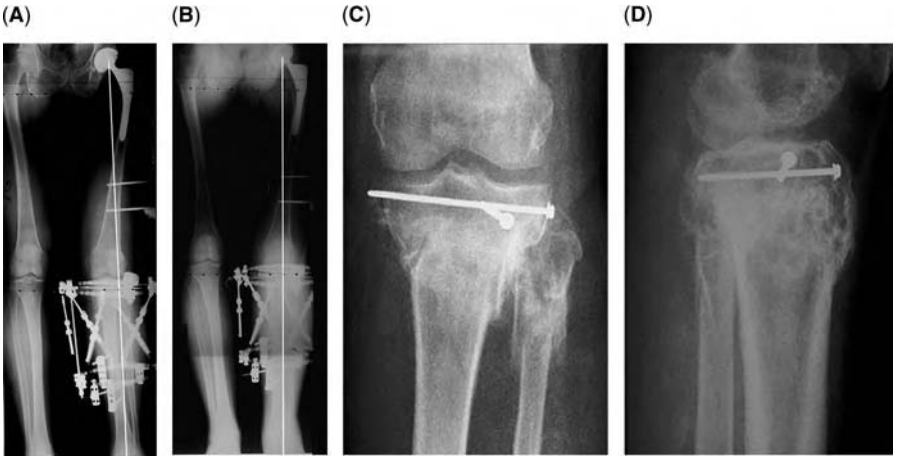

Figure 15 (**A**) Postoperative standing hip to ankle X ray displaying lateral axis deviation. (**B**) Mechanical axis alignment corrected after surgery using the total residual correction program. (**C** and **D**) Healed complex tibial plateau fracture.

afforded by a lag screw. More or less compression can be adjusted during the healing process. Also, the mechanical axis alignment can be adjusted to match the contralateral leg using standing 51-inch hip-to-ankle X rays. Small angular adjustments at the plateau have a profound affects on the mechanical axis due to the close proximity to the knee (Fig. 15A and B). The ability to adjust this alignment postoperatively can be very beneficial and prevent the development of posttraumatic arthritis (Fig. 15C and D).

Postoperative care is patient and injury dependent. Weight bearing is usually limited during the first six weeks to 25% to 50% depending on the amount of articular comminution. Early range of motion is started for those patients without a bridging frame. Half-pins and wires, particularly those that are adjacent to compromised soft tissue or close to the joint, are monitored closely for infection and treated aggressively with antibiotics if the need arises. The bridging frames are removed in the office by six weeks and range of motion of the knee encouraged. Outpatient physical therapy progresses with weight bearing as tolerated with crutches or a walker initiated by the sixth week. By three months all patients should be full weight bearing with an assisted device for balance. X rays are repeated monthly. The frame is dynamized after the third month to encourage strengthening of the callus. Most patients have the frame removed as an outpatient surgical procedure between the fourth and fifth months. Patients are placed in a hinged knee brace for weight bearing activities for one month after frame removal. Physical therapy is continued, improving range of motion and increasing strength and function.

COMPLICATIONS

Acute complications can occur preoperatively, intraoperatively, or in the immediate postoperative period. Failure to distract high-energy bicondylar fractures with temporary bridging fixation initially can produce poor CT scan imaging, poor soft tissue healing, and greater difficulty with the definitive articular reduction. Unrecognized compartment syndrome can lead to disastrous results. All patients should be monitored with around-the-clock physical examinations for 24 hours after the initial high-energy trauma. Questionable exams should undergo compartment pressure monitoring and surgical release if needed. Immediate definitive fixation of high-energy plateau fracture should be avoided to prevent early soft tissue failures and wound infections. A midline approach to access both condyles is discouraged.

Comprehensive understanding of the safe zones defined by the local anatomy can reduce intraoperative complications. When placing the proximal fibula wire, one must avoid the common peroneal nerve at the neck of the fibula. The patient should not be chemically paralyzed during wire placement so as to monitor nerve penetration. If a nerve is penetrated, noted by foot or toe movement, removal of the wire intraoperatively does not generally result

in long-term sequelae. Binding the pes anserine tendons is painful for the patient and can reduce knee motion. Wires should not be closer than 14 mm from the joint surface to avoid wire penetration into the knee joint. All half-pins and wires should be bicortical and placed with minimal heat generation to avoid bone necrosis and pin sepsis. Half-pins should be predrilled to prevent the generation of heat, which produces early pin failure and infection. Half-pins in the proximal fixation block should not extend into fracture lines, which extend into the joint. This can cause a joint infection from metaphyseal extension of half-pin sepsis. For this reason, it is prudent to avoid half-pins in the proximal fixation block on most complex bicondylar fractures. Finally, articular reduction is paramount to satisfactory functional outcomes and should be addressed prior to the placement of the ring fixator. This may be the most time consuming surgical step but poor reductions lead to poor results with very few options available for delayed reconstructions.

Deep venous thrombosis has been reported in the range of 12% (27). Generally, the longer period of immobilization prior to definitive treatment produces a greater risk of deep venous thrombosis. All patients at our institution receive sequential compression stockings and 5000 units of SQ heparin two times daily while in the hospital and a routine Doppler ultrasound screening of the lower extremities prior to discharge. The popliteal vein and proximal veins are easily seen with the scan. Venous thrombosis distal to the popliteal vein is not treated with aggressive anticoagulation. A positive scan proximal to the calf receives prolonged anticoagulation treatment. A negative scan receives no home going anticoagulation.

Deep infection and soft tissue failure can still occur with delayed surgery and wire ring fixation. If wires or half-pins are within the zone of soft tissue failure, they need to be removed. A new strategy for stabilizing the reduction, without compromising the access to the problem area, needs to be developed. This usually requires extending the frame across the knee to stabilize the bone and soft tissue envelope. Acutely infected fractures require marginal surgical debridement of necrotic soft tissue and bone (Fig. 16A and B). Acutely infected fractures generally do not require en bloc debridement, which is common in osteomyelitis cases. Infected fracture treatment includes marginal debridement with removal of dysvascular tissue, intravenous antibiotics, antibiotic spacer, and soft tissue coverage. Stripped cortical fragments are removed. Excessive cancellous bone debridement is not necessary. It is important to provide access for the plastic surgeon to address the soft tissue defect. The medial gastrocnemius muscle is the most likely for soft tissue coverage (Fig. 16F) but a free vascular flap is sometimes required due to trauma to the posterior compartment. A skin graft alone is a poor choice. It does not bring in new blood supply to the area nor is it useful as a window to bone graft if needed at a later date.

Soft tissue loss can also be addressed by angulating or shortening to close down a defect primarily, but should only be used in extreme situations. It does not bring fresh tissue to the area, it is painful, and can lead to malunions or nonunions. The technique is reserved for small full thickness defects in a narrow zone of injury or those patients who are not candidates for muscle flaps.

Finally, an early postoperative hip-to-ankle X ray standing should be taken to assess the mechanical axis alignment. Angular deformity of greater than 5° at the plateau level deviates the mechanical axis of the extremity to a greater extent than the same deformity would if it were farther from the knee. The wire ring fixator should be adjusted in the clinic to restore the mechanical axis deviation and reduce the risk of future arthritis.

Loss of reduction postoperatively with early weight bearing has not appeared to be a problem in healthy patient with normal bone density (12). However, early weight bearing in the elderly has been shown to have loss of reduction in up to 79% if started before six weeks (28). Weight bearing in patients with poor bone quality should be delayed to prevent loss of reduction.

THE FUTURE

The goals of treatment including the preservation of biology, reduction of the articular surface, restoration of the mechanical axis, maintenance of joint stability, and the preservation of motion will continue to drive the technological advancements to improve functional outcomes. The TSF allows minimal biological insult with computer-assisted reduction at the metaphyseal-diaphyseal junction even after the surgical procedure has been completed. This is a powerful tool and will set the standard for future technological advancements in the treatment of complex tibial

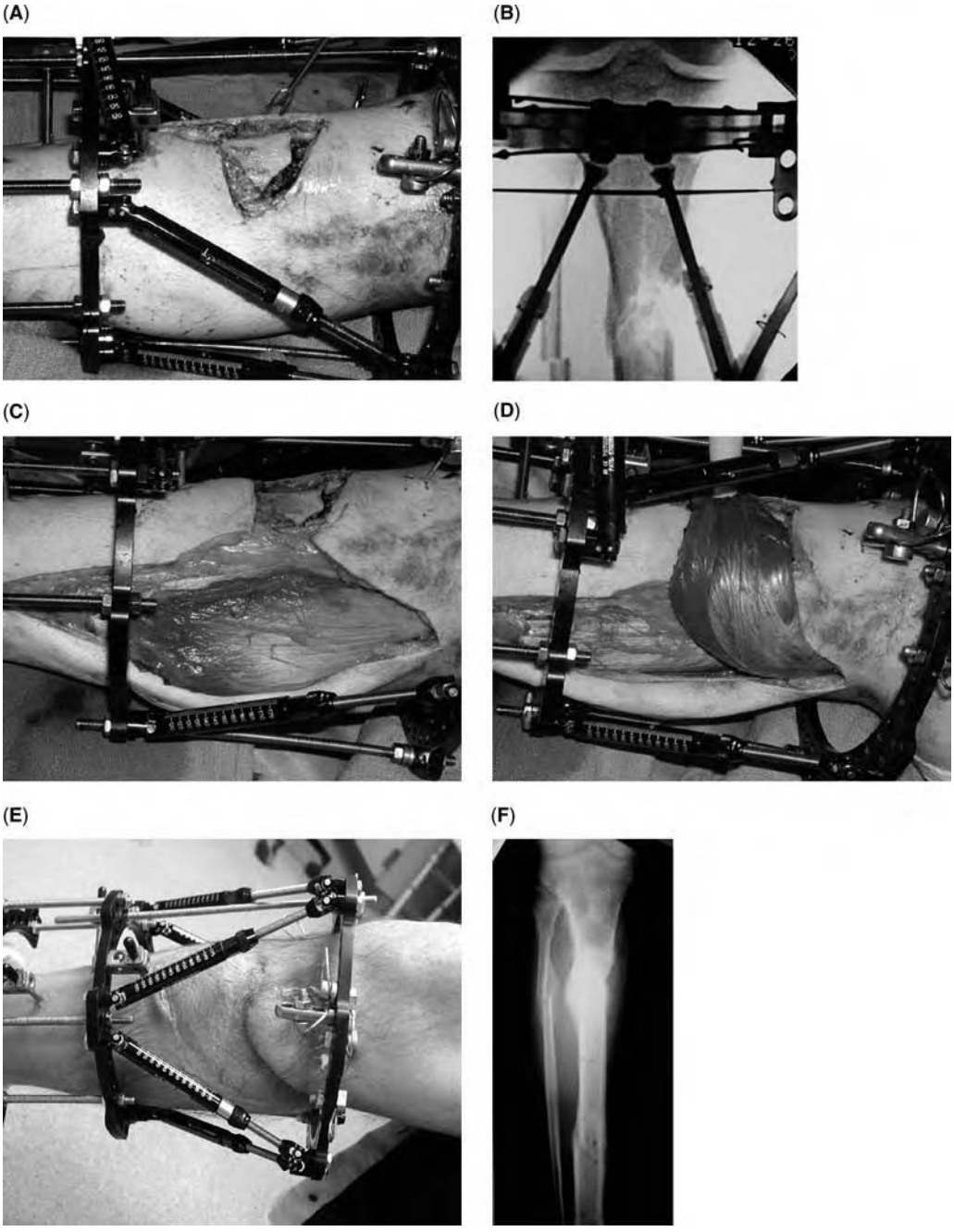

Figure 16 (**A**) Infected open tibia plateau with necrotic soft tissue and bone removed. (**B**) Stable fixation with bone defect. (**C**) Medial gastrocnemius exposure. (**D**) Transferred medial gastrocnemius muscle. (**E**) Healed soft tissue. (**F**) Result after staged iliac crest bone grafting, healed, excellent alignment, with no infection.

plateau fractures. External fixators will continue to be improved and redesigned to simplify the ease of application and adjustment. New advancement in external fixation will *not* be a substitute for good clinical judgment, appropriate surgical timing, and careful soft tissue handling.

In a society of limited recourses, the wire ring fixator compared to plates and screws, is still the more expensive device. However, this should be compared to the resources preserved by reducing the postoperative complications in high-energy injuries or compromised hosts. The wire ring technique will continue to be a powerful tool for this problematic fracture.

REVIEW OF LITERATURE

Authors	Journal, Year	Title	Number of Patients	Results	Conclusions
Ali AM, et al.	JOT 2003	Treatment of displaced bicondylar tibial plateau fractures (OTA-41C2&3) in patients older than 60 years of age	12 pts, average age 72, displaced bicondylar	F/U 8 months, 82% satisfactory, 3 malunions >10 degrees, 3 osteotomies, 1 TKR, 5 superficial infections	Safe and rellable technique
El Barbary, et al.	Int Orthop 2005	Complex tibial plateau fractures treated with illzarov external fixation with or without minimal internal fixation	30 Schatzker VI fractures, 18 combined with minimal internal fixation	Knee society score 10 excellent, 7 good, 1 fair, 2 poor	Success of minimal internal fixation and the illzarov
Katsenis D, et al.	JOT 2005	Minimal internal fixation augement by small wire transfixation frames for high energy tibial plateau fractures	Retrospective review 48 pts 40 (VI) 8 (V) 37.5% open, 63% bridged the knee	Fracture treated with provisional bridging frames tended to have superior clinical result compared to unbridged frames with same ROM, all united 13.8 weeks, ave. 38 mos., 81% excellent or good results	Complications included injury to peroneal nerve, 6 DVT's, 10 pin tract infections, one diaphyseal infection, 1 arthofribrosis, 4 varus deformities. Internal and external fixation with a bridging frame is a good treatment for complex plateau fractures
Ali AM, et al.	Clinical Biomech 2003	The stength of different fixation techniques for bicondylar tibial fractures, a biomechanical study	7 sawbones fixed with dual plating, a ring-ring external fixator (EF), right-bar hybrid EF, plate and medial EF, lateral plate and medial screw	All Failed in medial plateau, no difference between dual plating and ring-ring EF (4200n), other fixation methods significantly inferior in study	Choice between wire fixation and dual plates depends on the soft tissue and not biomechanics, both equally strong
Ali AM, et al.	JBJS B 2003	Outcome of complex fracture of the tibial plateau treated with beam-loading ring fixation systems	21 patients prospective consecutive standard protocal bicondylar, eatly weight bearing	Bony union 100%, good to excellent result 85%, full weight bearing at 6 wks in 60%, genral helath status correlated with knee score	Stable, may allow early weight bearing without loss of bony reduction
Kumar A, et al.	JOT 2000	Treatment of complex (Schatzker VI) fractures of the tibial plateau with circular EF: retrospective case review	55 pts, 35 closed, 21 open, limited open 7, avg follow-up 42 months	45 anatomic reductoin knee score was 83 (funtional 69); 9 with nonanatomic reduction knee score 52 (functional 19)	19/56 compartment syndrome; Results may have been improved by more frequent ORIF with elevation of depressed fragments and bone grafting

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7 Tibial Diaphyseal Fractures

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INTRODUCTION

The use of the Ilizarov method for treating acute closed tibial fractures is an attractive concept as it offers many advantages over existing techniques of fracture stabilization. It minimizes surgical trauma at the fracture site, with a percutaneous approach that does not further compromise the biological condition of the fracture site, therefore utilizing the full capacity of the bone and soft tissue to achieve bone healing.

The high incidence and variety of tibial fractures may explain the wide choice of fracture stabilization techniques available to treat this common fracture. The tibia has many unique features that make it vulnerable to many complications. It is a subcutaneous bone with poor soft tissue coverage on the medial border with a high incidence of open fractures (1). Its blood supply is also limited particularly, at the distal end. These factors results in a higher incidence of significant soft tissue damage and susceptibility to infection, wound break down, and nonunion.

The Ilizarov apparatus is distinguished from other external fixators. It is a universal system of fracture care that allows for individualized care for each type of fracture. It is a stable and yet dynamic construct that permits functional axial loading of the injured limb. This in turn stimulates bone angiogenesis and promotes osteogenesis, leading to quicker remodeling (2). The apparatus is also versatile and allows correction of any residual deformity. These characteristics allow the Ilizarov method to be used to treat tibial fractures with minimal interference at the fracture site, minimizing the rate of deep infection and nonunion.

The use of the frame in acute injuries did not pick up momentum outside certain geographical locations (3), despite its great advantages (Table 1). The use of Ilizarov apparatus requires expertise and can be time consuming. Pin site problems have been a discouraging factor for many surgeons. In addition to the well-established role in the treatment of complex fractures, nonunion, and malunion (4,5), we feel that it has a role to play in the treatment of closed tibial fractures especially in patients with narrow medullary canals and adolescents with open growth plates (4). With the advancement in technology and the introduction of the Taylor spatial frame (TSF), we anticipate that circular frames will be used more to treat this common but controversial fracture. The versatility of the Ilizarov method and frame enables the surgeon to treat both simple and complex tibia fractures, including open fractures, bone loss, and delayed healing (6–8).

EVALUATION

Assessment of the injury includes full clinical exam to exclude associated injuries. Examination of the soft tissue envelope is essential to ensure full clinical evaluation of the injury site. The soft tissue injury associated with closed tibial fractures is graded by the Oestern and Tscherne classification (Table 2) (9). Documentation of the neurovascular status is important and monitoring the patient for signs and symptoms of compartment syndrome is a fundamental part of the treatment. Radiological assessment should include anteroposterior (AP) and lateral views of the full length of the tibia.

Table 1 Advantages

Stability achieved without compromising the soft tissue envelope
Minimal blood loss
Minimally invasive
Ability to achieve anatomical reduction comparable to open reduction techniques
Ability to adjust the degree of rigidity of the fixation as required to suite the healing stage
The limb is suspended in the frame, avoiding pressure in vulnerable areas in the heel avoiding therefore pressure sores
Early mobilization
Reduced length of hospitalization

CLASSIFICATION

The tibial diaphysis is defined as the part of the tibia excluding the proximal and the distal 5 cm of the tibia. The morphology of the fracture is assessed (Table 3), taking into account the degree of displacement. Studies have confirmed the relation between the degree of displacement as a predictor of outcome and the potential for delay in achieving union (2,5,10). Many classification systems have been described but the Arbeitsgemeindschaft für Osteosynthesis fragen (AO) classification provides the most comprehensive classification of tibial fractures morphology and takes into account the degree of displacement as a predictor of success (11). It correlates well with Tscherne soft tissue classification (1).

INDICATIONS

The use of the circular frame to treat tibial fractures is not restricted to any particular group of patients and can be safely applied in the majority of cases (Table 4). Anatomically, the tibia is an optimal bone to treat with the Ilizarov apparatus. It is an easily accessible bone throughout its full length. Attention to safe corridors for placement of pins and fine wires is crucial to avoid neurovascular injuries (12). Managing the fracture can be accomplished using either the classic Ilizarov frame or the Ilizarov/TSF frame. The principles are identical but the use of TSF frame with computer assistance simplifies residual deformity correction. The aim of initial treatment is to obtain a satisfactory acute reduction. Residual deformity can be corrected gradually with the Ilizarov/TSF.

METHODS OF TREATMENT

The Ilizarov Apparatus

The fracture is reduced with the help of distraction. Positioning of the patient should be adjusted using bumps to ensure a patella forward position. With patella pointing forward, the second web space can be used as a guide to fracture reduction. To facilitate the application process, the leg is suspended on a frame that allows 360° accesses from above the knee to below the ankle. Direct reduction maneuvers of acute fractures using traction technique are usually sufficient to achieve good alignment. Further adjustments of the reduction can be achieved using different techniques,

Table 2 Oestern and Tscherne Classification

Grade C 0	Soft tissue damage is absent or negligible. Simple fracture caused by indirect violence
Grade C I	Superficial abrasion or contusion of skin from within. The fracture has a mild to moderately severe configuration
Grade C II	Deep, contaminated abrasion with localized skin or muscle contusion from direct trauma. Moderately severe to severe fracture configuration. Impending compartment syndrome is included in this category
Grade C III	The skin is extensively contused or crushed and muscle damage may be severe. Also, subcutaneous avulsion, compartment syndrome or rupture of major blood vessel associated with closed fracture

Table 3 Classification of Diaphyseal Fractures

Simple
With a third butterfly fragment
Segmental
With bone loss

which when used individually or in combination, can correct any residual deformity including angulations and translations in the coronal, sagittal, or axial planes.

FRAME DESIGN

The basic frame design is made of four rings (Fig. 1). Two ring blocks are applied to the proximal and the distal segments of the bone. An additional ring is added in cases of segmental fractures to stabilize the middle fracture segment or a butterfly segment, although stabilization of the transfixing wires can also be achieved by high profile post (Fig. 2). The preassembled frame is applied to the leg leaving sufficient room proximally for 90° knee flexion and room distally to allow ankle extension above neutral (Fig. 3). The two central rings should be several centimeters above and below the fracture. The frame design varies according to the configuration of the fracture.

OPERATIVE TECHNIQUE

Acute Reduction with a Neutral Frame

The reference wire insertion is crucial to ensure optimal frame alignment in the frontal and the sagittal planes. One smooth 1.8 mm reference wire is inserted transversely orthogonal to the proximal tibial axis. This is a very important step and must be performed meticulously. The frame is applied around the tibia and fixed to the reference wire. A distal wire (1.8 mm smooth wire) is inserted 1.5 cm proximal to the ankle joint parallel to the joint and the first proximal wire. The tibial fracture is reduced by traction and correction of angulation and rotation is performed simultaneously under image intensifier control. The proximal tibia is stabilized in the proximal ring block with two points of fixation. Care is taken to set this ring block orthogonal to the proximal segment. The distal wire in the tibia is positioned on the distal ring to achieve correct rotation and translation. Distraction is then placed across the frame and fracture to correct shortening (in acute fractures). The distal segment is further stabilized, ensuring that it is orthogonal to the distal tibia ring block. Leg Bumps and arched olive wire technique are used to fine tune the position at the fracture site. Once optimal reduction is obtained, the excess distraction is removed. The proximal and distal reference wires ensure that the frame is orthogonal to the tibia and simplifies any further adjustments. As a rule, the threaded rods connecting the rings must be parallel to the tibia in both planes. A reliable judgment can be made if the fracture has minimal remaining displacement. Image intensifier can be used to judge the degree of residual displacement. The frame should be applied to the leg with adequate space between the frame and the skin and, as a rule, two finger breadths between the frame and the skin (especially posteriorly) and a longer distance is recommended if swelling is anticipated.

METHODS OF REDUCTION

Correction of Longitudinal Displacements

The first step of fracture reduction is to correct the shortening and to distract the fracture site (Fig. 4). This maneuver will disengage the fracture fragment and allow for more complex reduction techniques to be performed.

Table 4 Indications

Narrow intramedullary canal
Adolescents with open physis
Compromised soft tissue envelope
Skin scarring or pathologies that impede open surgical approaches
Patient choice
Avoidance of anterior knee pain (associated with intramedullary nailing)

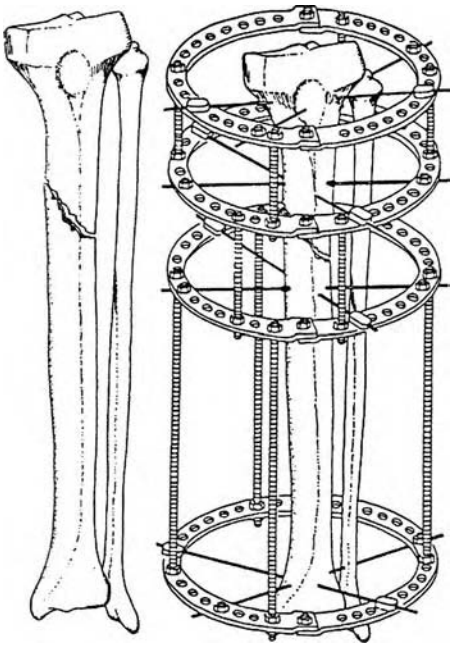

Figure 1 Simple diaphyseal fractures. The basic frame design consisting of four rings is used to treat this type of fracture. Olive wires are placed transversely on the intermediate ring allow reduction of the fracture in the frontal plane. *Source*: From Ref. 13.

Correction of Translation

Translation of the fracture in the frontal and the sagittal planes can be corrected using a variety of techniques. Translation of the fracture can occur in the frontal plane, sagittal plane or both. The techniques available can deal with the translation separately or simultaneously. Translation can be corrected with use of either the olive wire technique or the push-pin technique.

Olive Wire Technique

Olive wires can be used to correct translation in the frontal plane, by using the tensioner on the opposite end of the olive (Fig. 5). As tension is applied, the bone fragment is pulled in the direction of the tensioner, correcting the translation. The degree of tension is judged based upon the amount of translation that needs to be corrected. This could be done with help of fluoroscopy. The frame can be used to correct translation by attaching two olive wires one to the proximal and the other to the distal in opposing directions. Translation is achieved by pulling the displaced fragment with the olive wire to the stable fragment (Fig. 6).

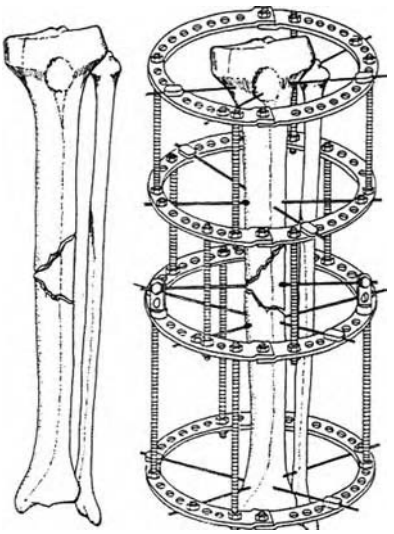

Figure 2 Diaphyseal fractures with a third (butterfly) fragments. The frame is built with four rings. Two olive wires are applied from the same side and the third is placed from the opposite side stabilizing the butterfly fragment. This wire is fixed to the ring with posts. *Source*: From Ref. 13.

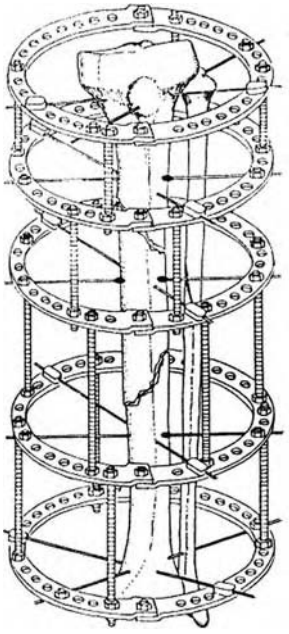

Figure 3 Diaphyseal fractures at two levels (segmental). A preassembled frame with five rings is required to stabilize this type of injury. Two sets of two rings at the level of the proximal and distal fragments and a ring in between. *Source*: From Ref. 13.

Push Pin Technique

Half pins can be used to correct translation (Fig. 7A–C). The pin is inserted into the bone close to the fracture using the standard pin insertion technique. With the pin in place, the fracture translation can be corrected by pushing the distal fragment. Once correction is achieved, the pin is fastened to the cube using a set-screw to hold the fracture reduction.

Arched Wire Technique

Fracture translation in the sagittal plane is corrected using the arched wire technique (Fig. 8). A fine wire is introduced on the middle rings in a frontal plane. The ends of the wire are fixed to the ring in a forward or backward direction depending on the direction of displaced fragment assuming an arched shape. Tensioning of the wire gradually reduces the displaced fragment in the direction of the bow. It is highly recommended to replace the wire once the task of reduction is complete because it usually has tented the skin.

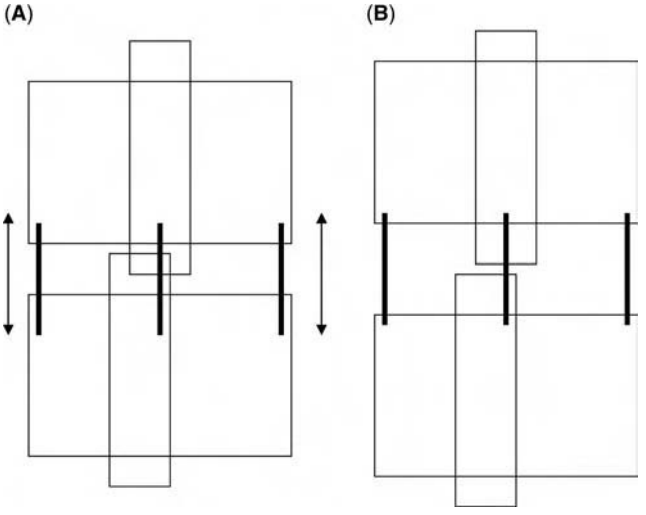

Figure 4 (**A**,**B**) Disengagement of the fracture fragments is achieved by lengthening the rods connecting the proximal and distal ring blocks.

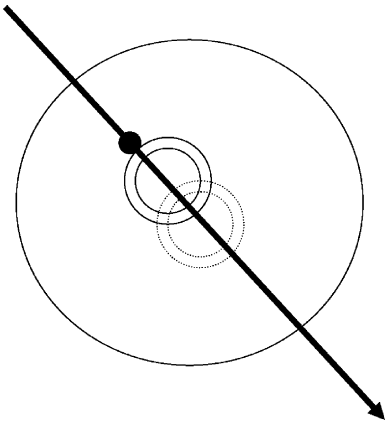

Figure 5 Correction of translation using olive wire technique.

Arched Olive Wire Technique

The olive wire is introduced into the bone and fixed in a forward or backward direction on the ring (Fig. 9). The tensioner opposite to the olive wire corrects the frontal translation. This part of two plane correction is accomplished first before correcting the sagittal plane translation, which is corrected by applying tension on the side of the olive.

Correction of Translation, Using the Frame

Movement of the distal part of the construct in respect to the proximal will allow the correction of the translation of the fracture (Fig. 10A–D). Three threaded rods are used to loosely join the two translated rings in an offset manner. The translation of the rings and bone is corrected by tightening the rods. Alternatively, translation boxes can be used to translate rings relative to each other. Additional compression may be achieved by then shortening the rods.

Reduction of Butterfly Fragment

Medial/Lateral Butterfly Fragment

An olive wire is used to reduce the butterfly fragment (Fig. 11). The wire should transfix the butterfly fragment to the either the proximal or the distal part of the bone—whichever has a greater contact area.

Anterior/Posterior Butterfly Fragment

A wire is introduced in the frontal plane through the butterfly fragment and arched with the concavity facing the desired direction of displacement (Fig. 12A,B). By applying tension, the fragment is reduced to the stable segment.

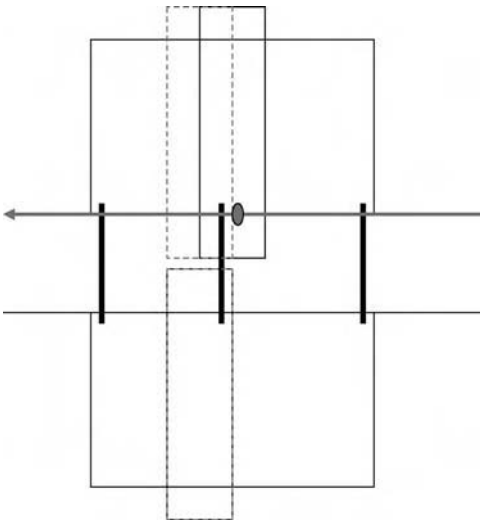

Figure 6 Correction of translation using two olive wires.

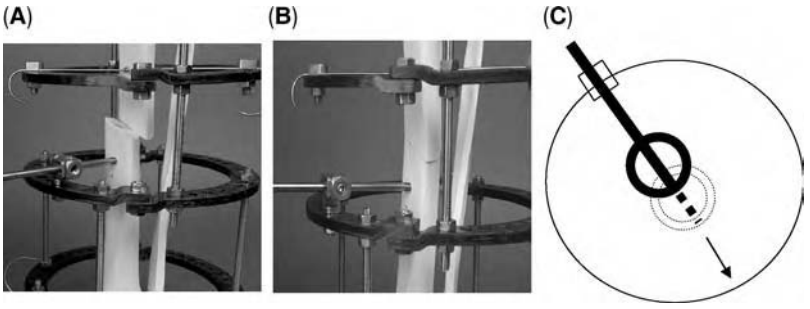

Figure 7 (**A**–**C**) Push pin technique.

Varus or Valgus Malalignment

Acute reduction techniques usually correct frontal plane malalignment. Residual displacement of less than 7° is correctable by shortening the rods at the convex side of the angular deviation, and lengthening the rods on the concave side of the deformity. Conical washers are helpful in this regard. If conical washers are not used, the rods will bend and less correction can be achieved.

Recurvatum or Procurvatum Malalignment

The major part of sagittal malalignment is corrected by applying axial traction and appropriate support of the leg holding frame or bumps to correct the sagging of the fracture site. The residual part of the correction can be accomplished by lengthening of the rods on the anterior part of the frame and shortening of the rods of posterior aspect of the frame.

Correction of Rotational Malalignment Using the Frame

Like translation, rotation may be corrected with rods placed loosely in an offset manner or with use of translation boxes between the two rings. It is vital to bear in mind that if the bone is not centered, as in the case of tibia, there will be a translational deformity from this maneuver.

Correction of the Rotation with Twisted Wires

The points of fixation of the wires are shifted symmetrically (Fig. 13). Tensioning the wires will rotate the bone fragments.

Taylor Spatial Frame

Acute Fracture Reduction—FastFix™ Struts (Rings First Method)

A TSF is usually composed of two ring blocks from four rings. The proximal ring block is made of an appropriately sized two-thirds ring and a full ring. Rings blocks are applied to the proximal and the distal fragments. Each ring block should be orthogonal to the mechanical axis of the fragment and secured to the bone at three to four points, using either wires, or pins, or a combination of these.

The leg is placed supported with bolsters to achieve a provisional reduction. Fast-fix struts are used to connect the proximal and distal ring blocks. With the struts in their sliding mode, the fracture is acutely reduced under fluoroscopic guidance. The fracture is reduced

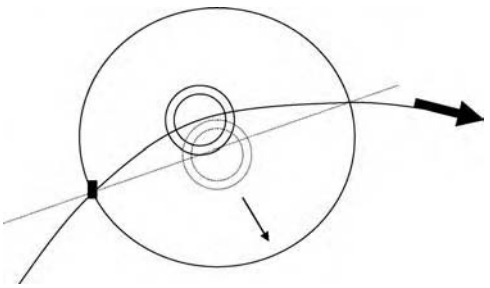

Figure 8 Arched wire technique.

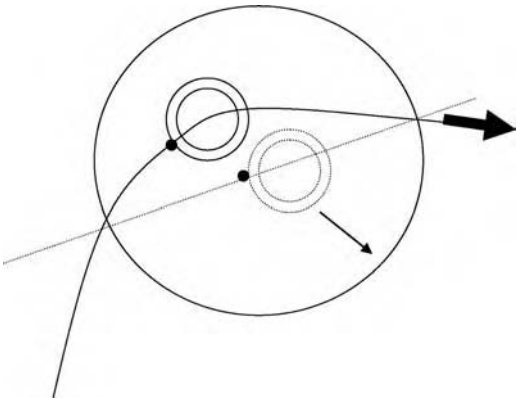

Figure 9 Arched olive wire technique.

with traction through the frame. The length, rotation, angulation, and translation are usually corrected with the initial reduction technique. The struts are locked once the desired fracture position is achieved. The fracture position is then assessed and further adjustment can be made. Olive wires technique can be used on the second and the third ring to achieve reduction of the butterfly fragment if present to correct residual malalignment. Additional fixation with 6 mm hydroxyapatite half pins off the second and the third rings is used (Fig. 14E and 15E). It is helpful if fixation of the proximal and distal ring is made using fine wires only, while using half pins off the second and the third ring. This will help in the later stages of treatment, especially if dynamization of the frame is desired, the proximal and the distal rings can be removed in the office. Frame and deformity parameters are obtained at the end of the procedure using the fracture site are used as the origin. Using these parameters, further improvements in the position can be gradually performed without the need for anesthesia utilizing the total residual deformity correction program (14).

POSTOPERATIVE CARE

Residual Correction

Correction of residual displacement can be done in the office, without the need for anesthesia. Angulation, translation, rotation and shortening can be corrected using the traditional Ilizarov adjustment techniques. In the case of TSF, strut lengths can be adjusted based on a schedule made with the help of the total residual deformity program.

Pin Care

Pin site infection is a fairly common complication with the circular frames (15). Pin site care begins on the second postoperative day. The patient is taken through the various steps of pins site care before being discharged home. The patient or a responsible relative should be conversant with the pin care protocol in order to ensure optimal pin site care management.

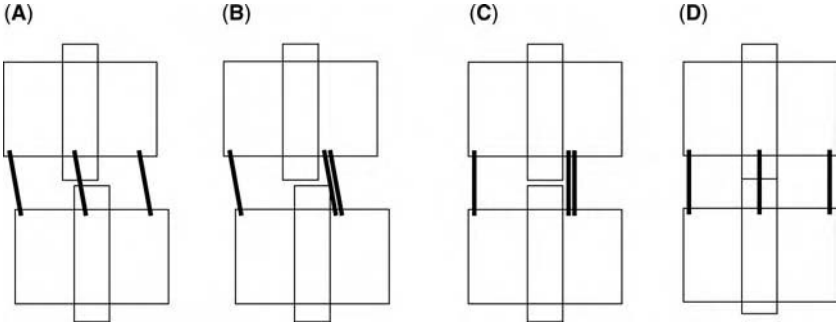

Figure 10 (**A**–**D**) Stages of correcting fracture translation using the frame.

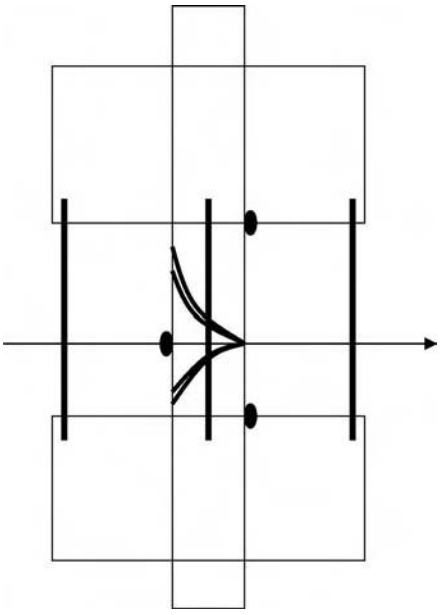

Figure 11 Reduction of butterfly fragment using an olive wire.

Our pin care protocol involves cleaning the pin sites with a half strength mixture of sterile normal saline and hydrogen peroxide. The pin sites are then covered with Xeroform gauze. This routine of pin care is repeated on daily basis to ensure healthy pin site. Patients are also encouraged to shower daily. It is also vital to ensure that patients have a supply of oral antibiotics, which can be started as soon as pin site is suspected. We also allow our patients to swim in a chlorinated pool.

Weight Bearing Status

The patient is allowed to bear weight from the first postoperative day. Weight bearing as tolerated is encouraged as a general principle in all patients. Patients typically progress to full weight bearing over a two to three week-period.

Fracture Union and Frame Removal

Clinical and radiological examination is the key to assessing fracture union. The average union time using the Ilizarov frame is between two to five months (2,16). Shorter periods in the frame were reported with consolidation achieved in between 21 and 72 days in children, and 30 and 90 days in the elderly (17). The ability of the patient to mobilize without a walking aid is indicative of increased fracture stiffness (18). At this stage, the rigidity of the frame can be decreased to allow dynamization of the fracture, before the frame is finally removed. The dynamization process is helpful in reducing the fracture gap and helps in achieving union (19). It is usually done at the later stages of the treatment and serves to increase the rate of bone healing and gives assurance that the consolidation of the fracture is underway. Cyclical loading in contrast to continuous compression has been shown to enhance fracture healing,

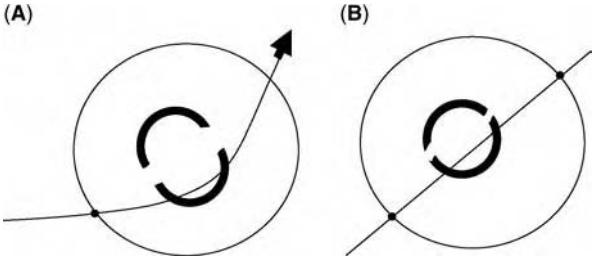

Figure 12 (**A**,**B**) Reduction of anterior/posterior butterfly fragment using arched wire technique.

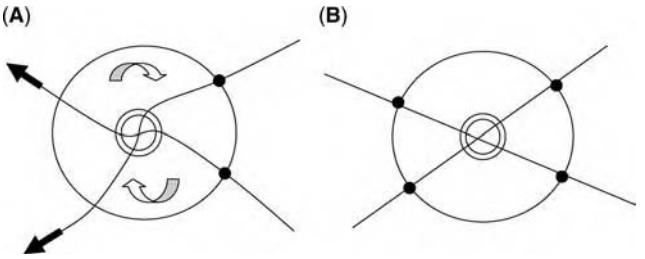

Figure 13 Correction of rotational deformity using twisted wire technique.

particularly at the later stages of the treatment (20). Dynamization of the Ilizarov frame is accomplished in a variety of ways, including removing wires, removing distraction, loosening bolts to allow free sliding of the vertical rods, and the removal of rings. Final check of the fracture union is done on the day of the removal. We remove frames in the operating room. At this time we check the stability of the fracture with manual testing after the connecting rods or struts are removed. We also analyze an AP and lateral X-ray. Based on this information, we

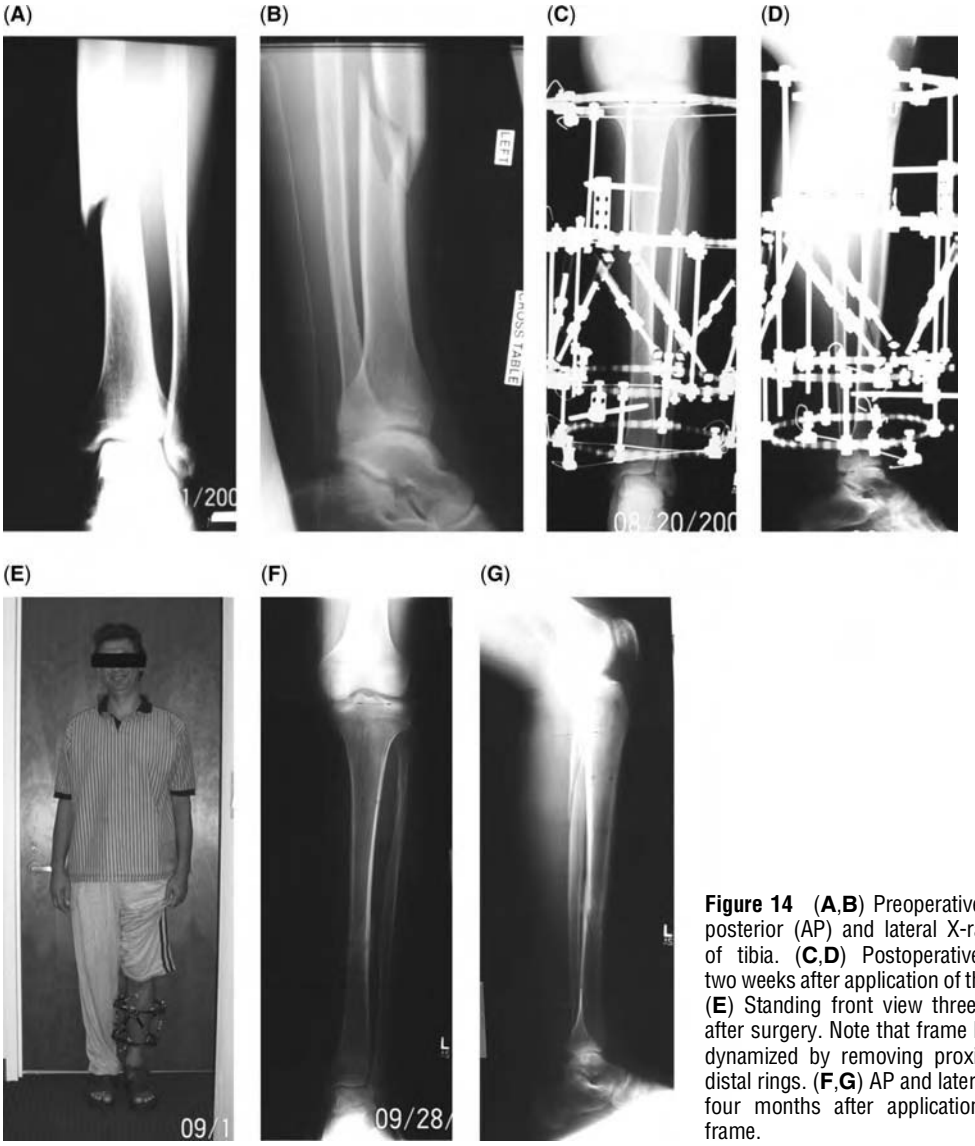

Figure 14 (**A**,**B**) Preoperative anteroposterior (AP) and lateral X-ray views of tibia. (**C**,**D**) Postoperative X-rays two weeks after application of the frame. (**E**) Standing front view three months after surgery. Note that frame has been dynamized by removing proximal and distal rings. (**F**,**G**) AP and lateral X-rays four months after application of the frame.

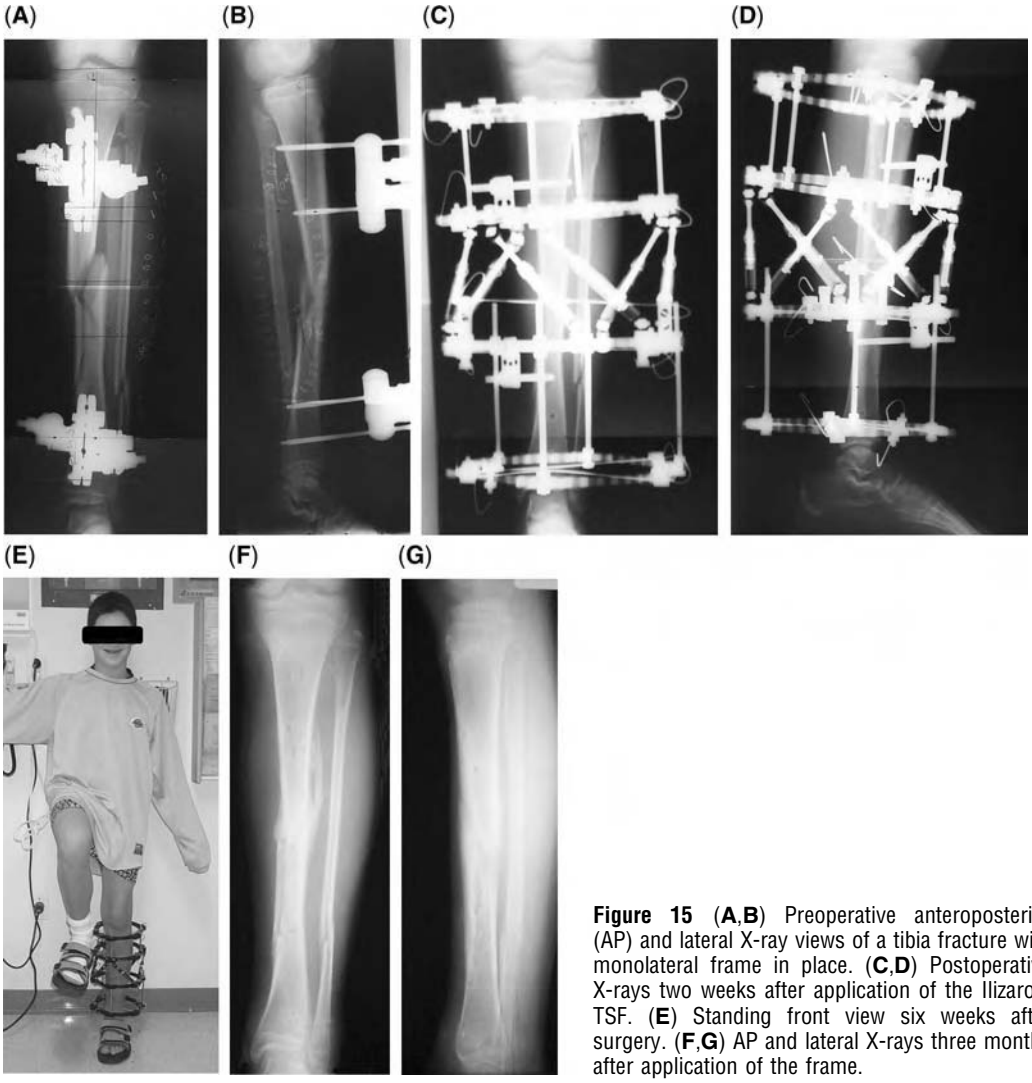

Figure 15 (**A**,**B**) Preoperative anteroposterior (AP) and lateral X-ray views of a tibia fracture with monolateral frame in place. (**C**,**D**) Postoperative X-rays two weeks after application of the Ilizarov/TSF. (**E**) Standing front view six weeks after surgery. (**F**,**G**) AP and lateral X-rays three months after application of the frame.

either remove the frame and apply a protective cast or leave the frame on the leg for some more time.

SUMMARY

The Ilizarov method and frame is very versatile and empowers the skilled surgeon to treat all types of tibia fractures. Using the methods outlined above, simple tibia fractures can be treated resulting in anatomic reduction, quick functional return, and frame times of three to four months.

These same methods can be used to treat the most complex tibia fractures including open fractures, fractures with bone loss, segmental fractures, tibia with open growth plates and with small intramedullary canals. Advanced techniques of bone and soft tissue transport or temporary intentional deformation to enable wound closure can be implemented early to optimize the clinical results.

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8 Tibial Pilon Fractures

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INTRODUCTION

Circular tensioned wire fixators (Ilizarov fixators) are powerful tools, that allow a trauma surgeon to construct a custom external fixator to treat tibial pilon fractures. The circular fixator may be a simple three-ring frame to treat extra-articular Type A (11) distal tibia fractures. More complex situations of intra-articular comminution, bone loss, and infection can be addressed with more complex frames for bone transport, acute shortening, arthrodesis, and supplemental fixation of hind foot injuries into the reconstruction. Pilon fractures present a challenge to the trauma surgeon because of a wide spectrum of skeletal and soft tissue injuries. Preexisting comorbidities, especially diabetes, tobacco abuse, and ischemic vascular disease may alter the approach to salvaging the patient's extremities. The accuracy of reconstruction of the plafond and mortise will have a direct effect on the stability of the joint and the ability of the cartilage to survive and heal after the injury (12–15). The reconstruction goal should be anatomic reconstruction of the joint surface and reconstruction of the normal width of the mortise (16). If anatomic alignment cannot be obtained, the minimal reduction of the joint should be a step off no greater than the thickness of the cartilage (2–3 mm) (12,15). The metaphyseal zone of comminution and the axial alignment of the limb require accurate reduction to promote functional outcome (17). This technical effort must be balanced against the morbidity of surgical approaches, introduction of metallic implants, and the use of autograft, allograft, and bone graft substitutes. Avoidance of surgical disasters is an important concept advocated by J. L. Marsh and considered in the preoperative planning (18). The treatment goals of the pilon fracture surgeon are to align the fracture axially, reduce the plafond and mortise, reduce and reconstruct bony comminution and bone loss, protection of the soft tissues from further damage, and rehabilitation of the extremity during healing. Increasing levels of bony comminution and soft tissue injury potentially compromise the functional result. It is the surgeon's task to balance the goal of anatomic bony reduction with complication avoidance. This chapter will focus on tensioned circular wires as the method of treatment, one of the three strategies of treatment. Other methods include open reduction and internal fixation and bridging half pin frames with limited open reduction and internal fixation. The latter methods have wide application and are beyond the scope of this chapter.

CLINICAL EVALUATION

The patient with a pilon fracture requires complete evaluation for multiple traumas. High-energy frontal impact car crashes and falls from height are the most common mechanisms of injury for pilon fractures (7). Once the patient is stabilized, a careful physical examination of the injured extremity is conducted. If there is gross deformity, the ankle and foot should be gently distracted to realign the fracture and stabilized in a splint. Specific sensory evaluation of the superficial and deep peroneal, calcaneal, medial/lateral plantar, and saphenous nerves is recorded. Overall capillary flow in the digits is evaluated. Vascular examination of the dorsalis pedis and posterior tibial pulse are observed. The pulse may initially be undetectable. Reduction and splinting of the fracture will help reduce the spasm of the vessels and the pulse will usually reappear. Profound ischemia indicates vessel injury. If there is an open wound, the local area is cleansed with an antibacterial solution and a sterile dressing applied. Intravenous antibiotics are administered. Crude manual examination of open fractures in the emergency room is unwarranted. The lower leg and foot are evaluated for compartment syndrome. Both foot and leg compartment syndromes are associated with pilon fractures (1).

The leg is splinted with bulky padded medial and lateral splints and X rays are obtained. Low-energy pilon fractures without significant displacement may be maintained in splints until surgery. High-energy pilons with shortening, comminution, and fractures with open wounds and compartment syndromes require emergency surgery. Open fractures are surgically debrided. The goal is removal of all devitalized tissue and foreign material. This is carried out with small forceps and tenotomy scissors. Foreign debris (dirt and grease) is removed with the contaminated bone to clean bleeding bone. Irrigation of the wound with pulsatile lavage without meticulous debridement is an inadequate technique. The potential space between the fibula and tibia is inspected if the tibia has protruded through the skin medially. Compartment syndromes are released. The fracture is distracted and bridged with a half pin frame (Fig. 1). The talar dome must be aligned axially in the anterior posterior (AP) and lateral view, the foot rotated to align the second toe and tibial tubercle, and the fracture slightly overdistracted to facilitate later reduction. The forefoot is controlled with a half pin in the first metatarsal to prevent equines and cavus of the forefoot. The patient is sent for computed tomography (CT) scans after the initial stabilization to evaluate the fracture pattern. Fractures with a lot of contamination will need to have second and even third surgical debridements. Amputation should be considered when there is mutilating soft tissue injury and crushing of the hindfoot associated with the pilon fracture. If there is a question of viability, the extremity is debrided and stabilized. Subsequent examination of the injury over the following 48 to 72 hours after injury will help clarify the decision of amputation versus limb salvage.

CLASSIFICATION

The fracture is evaluated for comminution, bone loss, and soft tissue condition. The pilon fracture will be sorted to one of two treatment strategies (2,3,5,6). Fractures with moderate comminution associated with moderate soft tissue injuries and in a reasonably healthy host, will be reconstructed to anatomic length. Fibula fixation is indicated and, the plafond and metaphysis will be augmented with bone graft. The goal is anatomic reconstruction of the tibia and fibula to normal axial length. Fractures with severe comminution, bone loss, and compromised soft tissue envelopes are approached with the concept of salvaging the extremity. This may require strategies of axial shortening, bone transport, and/or ankle arthrodesis.

SURGICAL TECHNIQUE

The injured extremity is elevated and the forefoot mobilized for 7 to 21 days until the soft tissue envelope has recovered from the initial trauma. The patient receives gait training and is encouraged to exercise. Anticoagulation medication is administered during the preoperative resuscitation before surgery. Type A (extra-articular) and C2 (partial articular) fractures may be approached with acute reduction without delay because a closed approach or only a limited

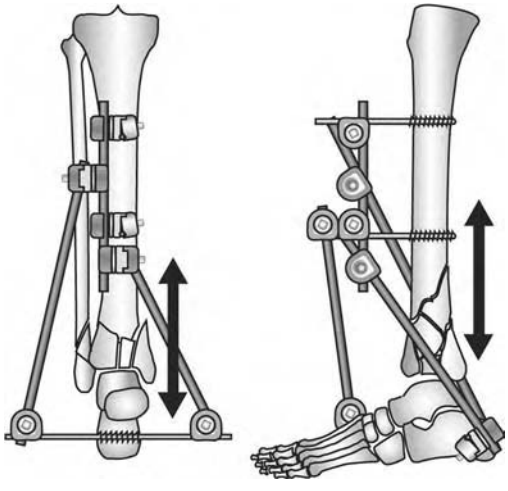

Figure 1 Half pin trauma resuscitation frame. The dome of the talus is axially aligned with the tibial shaft on anterior posterior and lateral fluoroscopy images. The foot is aligned rotationally with the tibial tubercle. The forefoot is controlled with a first metatarsal pin to maintain plantar neutral position. The fracture is distracted to length or mildly overdistracted.

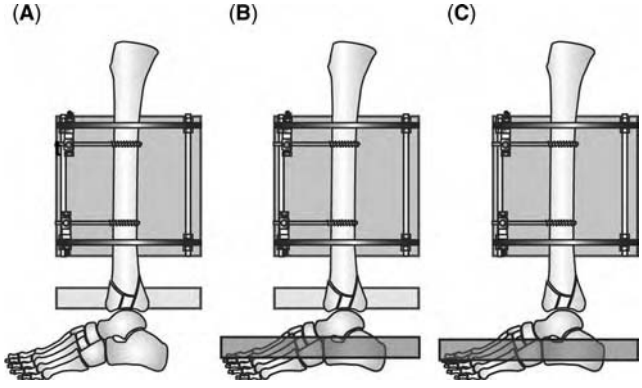

Figure 2 The reduction and fixation of distal tibia fractures is based on a fixation block placed orthogonally on the tibial shaft. Two AP half pins are mounted on universal cubes to allow precise alignment and subsequent adjustment. (**A**) The fixation of the fracture will be at the level of the plafond for Type A and C2 fractures. (**B**) C3 fractures will have initial distraction through the hindfoot followed by reduction and tensioned wire fixation of the joint. (**C**) Comminuted fractures or fractures with limited internal fixation are treated with spanning fixation without tensioned wires placed at the plafond.

open approach may be needed for an adequate reduction (11). This is less risky even for swollen soft tissues. Distraction across the fracture is the key to reduction. Because pilon fractures are located at the terminal end of the extremity, the reduction and fixation are based on a fixation block placed orthogonally on the midtibial shaft (Fig. 2). The stable base is constructed with a two-ring block separated with threaded rods 120 to 150 mm in length. The stable base is secured with two AP half pins mounted on universal Rancho cubes (Smith Nephew, Memphis, Tennessee, U.S.A.) and a medial face half pin (Fig. 3). This fixation block provides the proximal base for distraction across the fracture. Type A and Type C2 distal tibia fractures will have a horizontal reference wire placed one centimeter above the plafond and the metaphyseal fracture is distracted (Figs. 4 and 5) (19). Comminuted pilon fractures will be distracted by a horizontal reference wire placed in the calcaneus (20).

Distraction across the ankle joint is a powerful technique to gain initial axial alignment of the fracture. The distraction can result in near anatomic alignment and percutaneous fixation can be employed, or an open reduction and limited internal fixation will be necessary to align the plafond.

A subset of pilon fractures with comminution can have a level of fragmentation and crushing in which there are no fragments large enough to be fixated with tensioned wires at the level of the plafond. The tensioned wires would pass through ''bone mush'' with poor blood supply, produce little fixation, and provide a conduit for bacteria into the joint and fracture. These fractures are treated with a calcaneal distraction frame between the stable base and hind foot, with small screws and Steinman pins placed at the joint level to create a salvage joint surface. This treatment strategy requires more frame time, four to six months, for the fracture to heal, but the patients are able to walk with 50% weight. The hindfoot fixation block has opposed olive wires in the calcaneus and a medial to lateral talar neck olive wire.

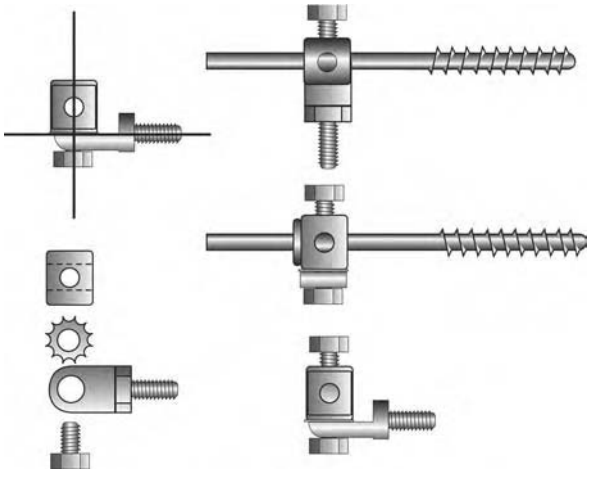

Figure 3 The universal pin mounting consists of an 8-mm bolt securing a one-hole Rancho cube to a long male hinge. This mounting allows the stable base to be precisely aligned on the tibia. Small secondary alignment modifications are also possible. If Rancho cubes are used without a universal mounting, the frame position is fixed by the alignment of the half pins drilled into the bone.

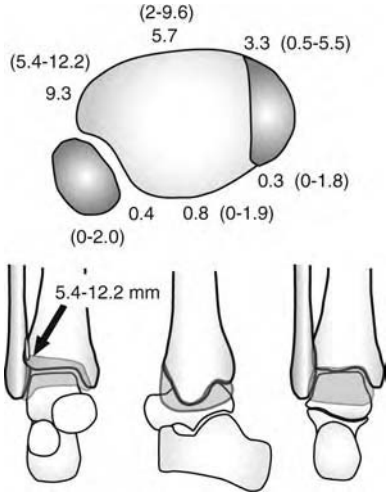

Figure 4 The capsule of the ankle extends over the anterior and anterior lateral plafond. The proximal reflection in the anterior lateral joint ranges from 5.4 to 12.2 mm. The most proximal extension is between the fibula and tibia. The posterior and medial plafond has extension of less than 2 mm. The figures in parenthesis are the range of capsular extension. *Source*: From Ref. 19.

The Ilizarov method provides powerful techniques for reconstruction of bone loss. The technique of acute shortening can be used to compress a comminuted metaphysis into the plafond to promote healing in patients with poor soft tissue envelopes and patients who are poor hosts. The lateral malleolus may require a small oblique segmental resection to compress without distorting the mortise. To compress a comminuted metaphysis or reconstruct bone loss, a proximal corticotomy and intercalary transport is used to reconstruct defects. Bone loss or gross contamination of the plafond may require excision. The extremity is salvaged with acute arthrodesis or intercalary transport to arthrodesis to obtain soft tissue closure, axial alignment of the tibia and hindfoot. Shortening of the metaphysis and lateral malleolus, bone transport, ankle arthrodesis, and limited fixation of the plafond are techniques used to reconstruct this group of fractures. Soft tissue injuries are treated with wet to dry dressings, skin grafts, hyperbaric oxygen, and rarely, free flaps. The soft tissues of the ankle have the ability to heal significant wounds if there is no metal implant or necrotic bone between the soft tissues and underlying viable bone.

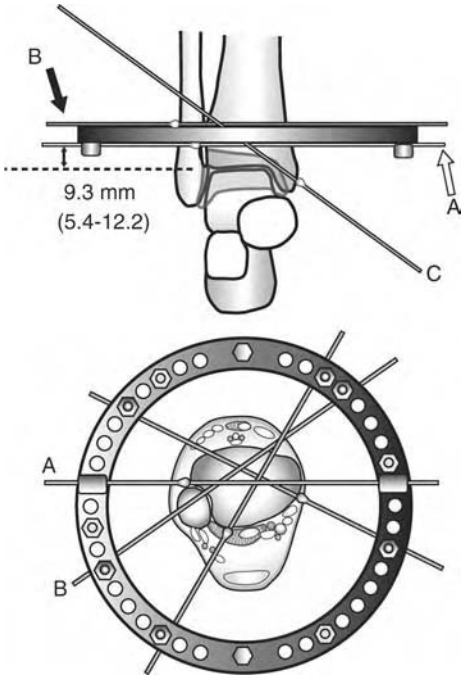

Figure 5 The anterior lateral capsular expansion needs to be cleared by the horizontal reference wire. (**A**) The wire should be placed 1 cm (or more proximal) above the subchondral bone of the dome of the plafond on the lateral centered view. (**B**) Wires fixating the fibula are located on the superior surface of the fracture reduction ring clearing the joint extension between the tibia and fibula. (**C**) Wires can be placed safely in the coronal plane to fixate medial malleolar fragments.

Surgical Techniques Type A and C2 Fractures

The sagittal plane C2 fracture is reduced and stabilized with 3.5 mm lag screw (9,10). The screw is placed just above the plafond. A stable orthogonal base is secured to the tibia with two 5 mm half pins on Rancho universal cubes (Smith and Nephew, Memphis, Tennessee, U.S.A.) (Fig. 6). A horizontal reference wire is placed at least 10 mm above the plafond. The olive is placed from medial to lateral or lateral to medial, based on the initial deformity of the fracture. If the fracture was initially displaced into the varus, the olive is placed medially. If the fracture was displaced into the valgus, the olive wire is placed lateral to medial. The distal tibia, metaphysis, and plafond are aligned on the distal fixation ring with the proximal shaft (Fig. 7). The dome of the talus must be centered under the axis of the tibia. The second toe is aligned with the tibial tubercle and patella to achieve correct rotational alignment. The knee is always included in the sterile field when treating pilon fractures to provide a guide for rotational alignment. The fracture is distracted to length. The distal tibia is rotated and manipulated into alignment with pin and wire reduction techniques (9). If the lateral malleolus is aligned by distraction across the fracture, then fixation of the lateral malleolus is optional. The fibula is fixated with an intramedullary pin or 1/3 tubular plate (Fig. 8). The plafond is fixated with three or four opposed divergent olive wires (Fig. 9). The wire pathways are chosen to stabilize the fracture fragments. Small adjustments of the tibial shaft alignment are possible by manipulating the two AP half pins mounted on the universal Rancho cubes. A medial face half pin is added to the stable base. Once aligned, the fracture is moderately compressed. Proximal fracture extension to the distal shaft requires a working length ring to fixate the fragments between the stable base and the fracture reduction ring (Fig. 10).

Surgical Technique C3 Pilon Fractures

An orthogonal stable base is applied. A horizontal reference wire is placed in the calcaneus in the posterior tubercle to avoid the tibial nerve branches (Fig. 11) (7,10,20). A carbon fiber fracture ring is placed on the threaded rods between the stable base and calcaneal footplate. The hind foot is manipulated on the footplate to align the dome of the talus with the AP and lateral axis of the tibia. The foot is rotated to align with the tibial tubercle. The fracture is acutely distracted out to length. If the goal is anatomic reduction of the plafond, then the lateral malleolus is reduced and fixated with a Steinman pin or 1/3 tubular plate. Distraction may produce near anatomic reduction of the plafond. Based on the CT scan, one can use 3.5 mm cannulated lag screws to reduce and fixate the joint fragments. If the joint does not reduce with distraction, a limited open reduction can be done (Fig. 12). The preoperative CT scan and soft tissue injury

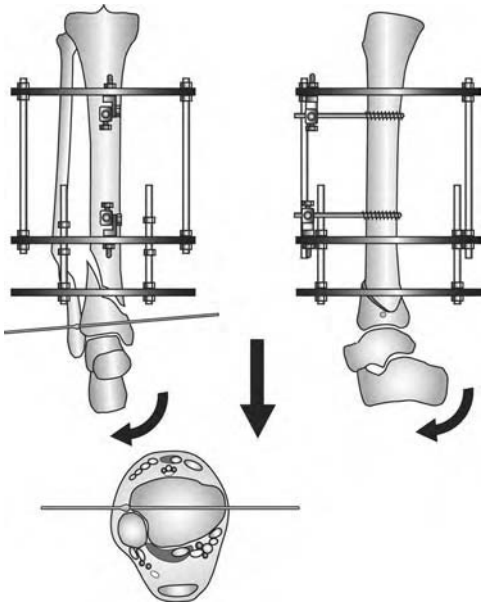

Figure 6 After applying the stable base, a horizontal reference wire is placed in the plafond. If the stable base is orthogonal, a reference wire aligned on the fracture reduction ring will produce near anatomic alignment in the AP plane. The fracture reduction ring is moved superior temporarily to provide clear fluoroscopic imaging of the horizontal reference wire.

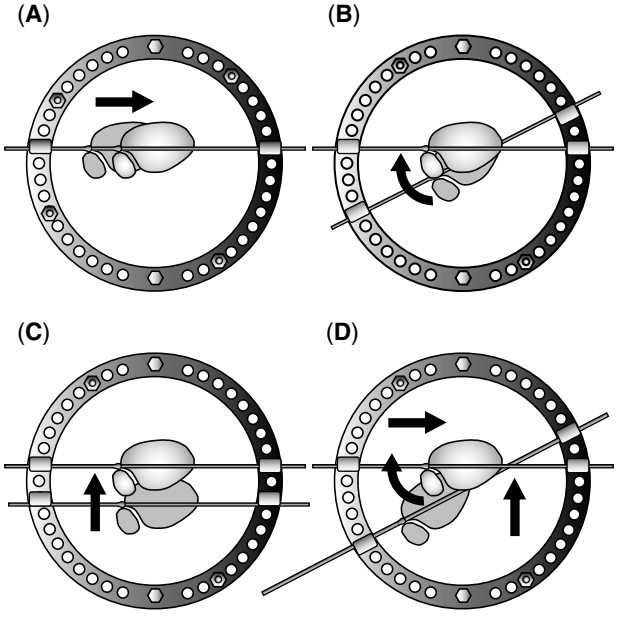

Figure 7 The plafond and horizontal reference wire is manipulated on the ring to align the plafond on the AP and lateral fluoroscopic views and to rotationally align the foot with the tibial tubercle. (**A**) Medial lateral alignment. (**B**) The reference is rotated to align the second toe with the tibial tubercle. (**C**) The wire is repositioned anterior-posterior by repositioning the slotted fixation bolts. (**D**) All three alignments are combined when positioning the distal metaphyseal block with the tibia shaft fixation stable base.

will be helpful for deciding on the optimal surgical approach as indicated—anterior medial, anterior lateral, posterior medial, or posterior lateral. The posterior fragment is often the key to reduction (21). It must be reduced to the posterior edge of the proximal tibial shaft and be pulled anterior over the plafond and pinned in place with proper relationship to the talus (Fig. 12). Both the medial and the anterior lateral fragments are reduced using the posterior malleolar fragment and the dome of the talus as a template for reduction of the joint surface. Autograft or bone graft substitutes are used to fill in the gap between the joint and metaphyseal fragments. Small interfragmentary screw and Steinman pins are placed to fixate the joint fragments in a reduced position. The approach is closed. The deep retinacular tissue should be closed over the reduction; skin closures without retinacular closure leads to wound dehiscence and infection.

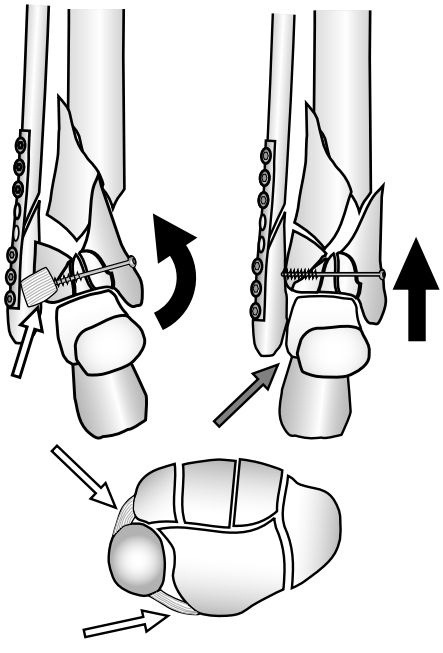

Figure 8 Fixation of the fibula demands reconstruction of the tibial column to anatomic length. If the tibia-fibula ligaments (*white arrows*) are intact and the tibial column not reconstructed, varus malalignment occurs. If the ligaments are disrupted, the plafond shortens, causing a fibula plus impingement (*gray arrow*).

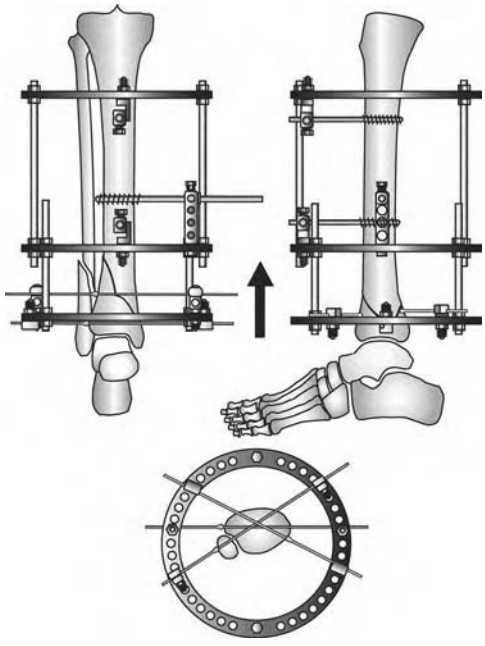

Figure 9 The distal plafond is aligned on the lateral view by rotating the hindfoot on the reference wire to achieve axial alignment. Two to three further olive wires are placed in the 60° arc of safe wire pathways to fixate the fracture. Draw wire and arc wire techniques are used to align and reduce metaphyseal fragments. The fracture is compressed.

The carbon fiber fracture ring is moved distally over the plafond. A horizontal wire is placed 1 cm above the joint and two or three opposed divergent olive wires are placed through the fragments (Fig. 13). The posterolateral wire (posterior to the fibula) is not routinely used. If the metaphysis is comminuted beyond potential for anatomic reduction, no tensioned wires are placed at the level of the fracture, and the frame is used as a distraction frame for four to six months (Fig. 14). The foot ring for this technique has additional fixation. Two opposed divergent olive wires are placed in the calcaneus and a medial to lateral olive wire is placed in the talar neck to stabilize the hindfoot for the four- to six-month healing time. Alternatively, fixation wires can be placed from off a plafond ring even without anatomic joint reconstruction. This limited-goals approach is to restore ankle joint congruity and metaphyseal axial alignment while avoiding an aggressive open approach and likelihood of deep infection. Fixating the fracture with tensioned wires at the plafond allows the hindfoot distraction foot frame to be removed six weeks after surgery to start ankle motion and increase gait to 50% weight bearing. The example shown in Figure 13 illustrates use of a spanning frame combined with open reduction and internal fixation after recovery of the soft tissues.

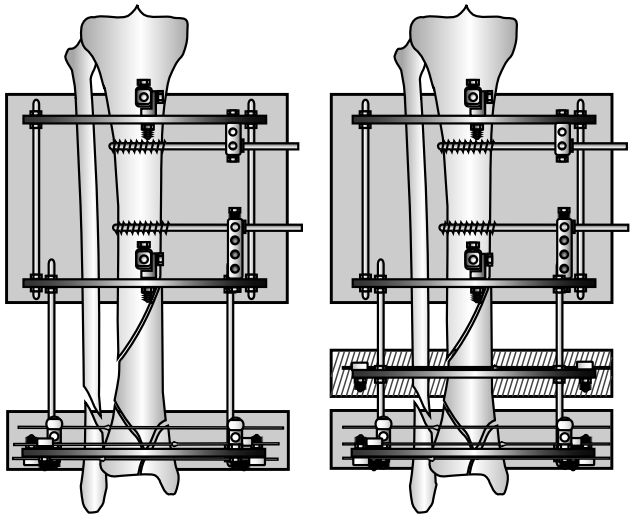

Figure 10 Pilon fractures may have proximal extension. A working length ring is added to the frame and wires are placed to fixate the proximal extension. Preoperative planning will determine when this ring is needed.

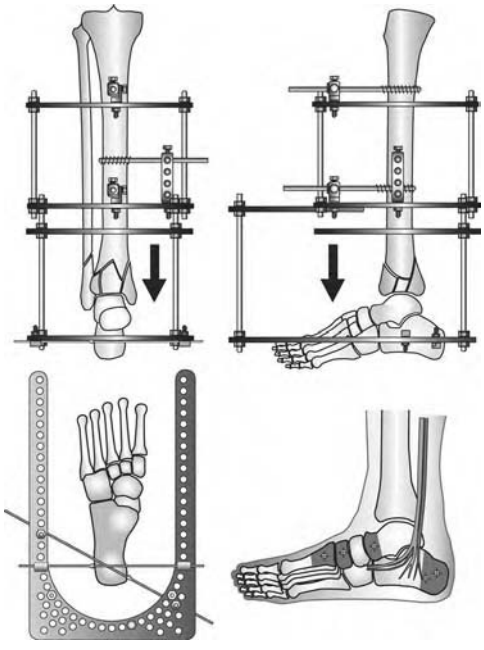

Figure 11 C3 pilon fractures are distracted initially between the stable base and a horizontal reference wire in the calcaneus. The calcaneal wires are placed in the posterior tubercle to avoid the calcaneal branch of the tibial nerve. The hindfoot is manipulated on the footplate to align the dome of the talus axially and rotationally. The fracture is distracted. Notice the fracture reduction ring, which is located adjacent to the base. The fracture reduction ring will be moved over the plafond once the fracture has been reduced by percutaneous or limited open technique.

Acute shortening is used when there is severe comminution or bone loss of the metaphysis and the soft tissue envelope is compromised (Fig. 15) (9). Diabetes, ischemic vascular disease, and tobacco abuse are factors that would lead to consideration of acute shortening as a reconstruction technique. The joint surface is reduced with limited internal fixation. Rather than distracting the metaphysis, the fracture is shortened until the fragments are compressed to promote healing. Acute shortening may assist with wound closure and will often reduce the open interval, which will close by secondary intention. If the fibula has a simple fracture, a segmental resection is done to allow the lateral side of the pilon to compress symmetrically (Fig. 16). A proximal lengthening is considered in younger patients with good health. Compromised patients

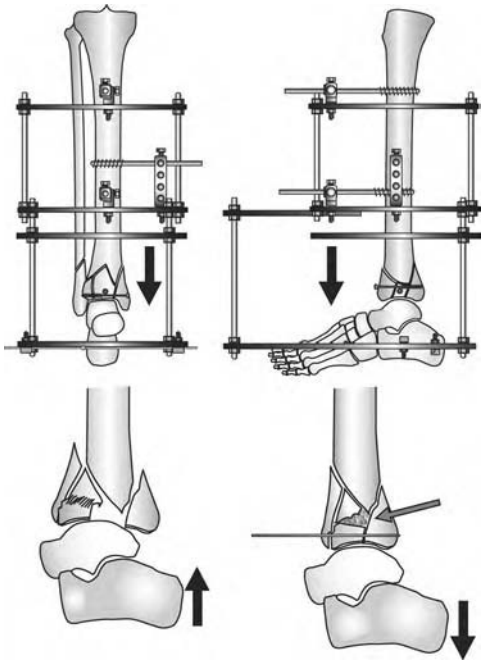

Figure 12 With distraction, the fracture may align. Percutaneous 3.5 mm screws are placed across the fragments based on preoperative computed tomography planning. If the fracture does not reduce, a limited open reduction and internal fixation is indicated. The anterior lateral or anterior medial approach is easily accomplished with the fixator in place. The key to reduction is reducing the posterior fragment anteriorly over the dome of the talus (*gray arrow*).

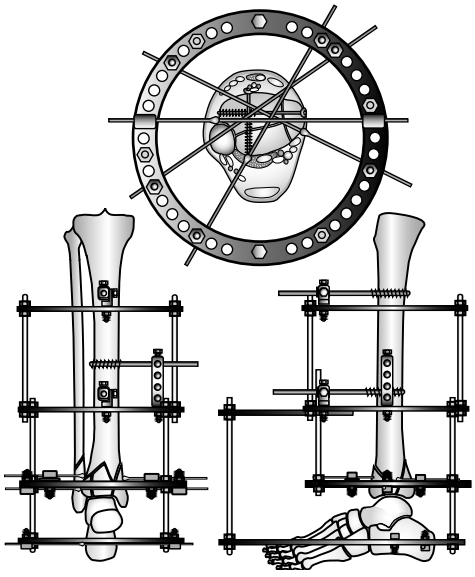

Figure 13 The fracture is fixated with at least three opposed olive wires in the safe wire pathways on the fracture reduction ring. The ring is moved inferiorly over the fracture and two anterior threaded rods are placed to stabilize the ring. The footplate is removed four to six weeks later in the clinic to start ankle motion.

use full sole lifts to equalize leg length. Large defects in the metaphysis are reconstructed with intercalary bone transport or with combined shortening and transport (Fig. 17).

COMPLICATIONS

Wire infection is the most common complication when treating pilon fractures with tensioned wire fixators (2,3,6,7). Meticulous pin and wire care is essential to reduce the rate of inflammation and wire infection. Rarely wires will develop deep infection requiring removal, debridement, and intravenous antibiotics. Septic arthritis and deep fracture infection require debridement, removal of necrotic bone, and revision with transport and arthrodesis. Methicillin resistant Staph Aureus is the most common infecting organism. Aggressive infections can develop, and this could lead to a need for amputation (4,8,22).

Equines contracture of the ankle and cavus contracture of the foot can be prevented with physical therapy, use of a foot ring, and elastic toe loops. The foot must always be maintained in a plantar neutral position until the patient is able to walk on the extremity.

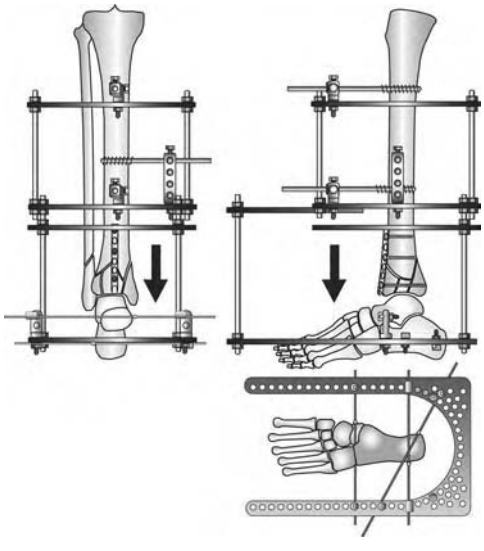

Figure 14 Pilon fractures with severe comminution or fractures, which have had limited plating are stabilized with a spanning frame. A medial to lateral talar neck wire is added to control the hindfoot until healing.

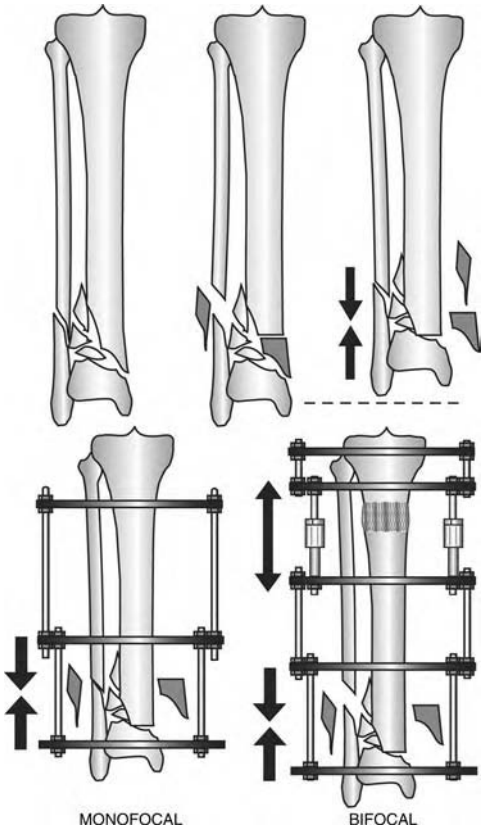

Figure 15 Acute shortening is used to salvage comminuted fractures in patients with physiologic and soft tissue compromise. Shortening may require fibula resection. Proximal lengthening is combined with shortening in appropriate patients.

Pilon fractures require four to six months of frame time to become stable. The fixator should not be removed until the fracture is united. Secondary bone grafting, fracture site compression, or bone transport is used to treat delayed union. Premature frame removal leads to nonunion, deformity, and malunion (1,22). Frame removal is done with monitored sedation or general anesthesia. A fracture orthosis is used until mature callus bridges the fracture.

Physical therapy should be continued for a year after surgery. The functional outcome 12 to 18 months after injury plateaus and reflects the long-term function (4,23,24). Alignment of the mortise, axial alignment of the extremity, soft tissue fibrosis, and cartilage damage ultimately determine the severity of post-traumatic arthritis. Socioeconomic factors affect

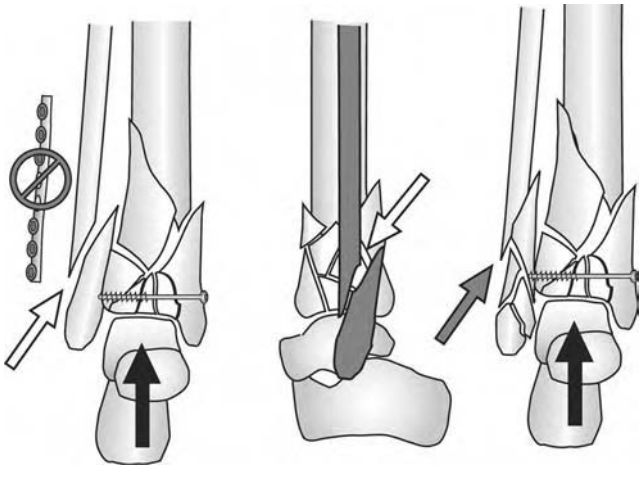

Figure 16 Shortening can cause significant malalignment of the mortise when the fibula has no fracture or a simple fracture pattern. A carefully planned osteotomy is indicated. Comminution of the fibula tolerates moderate shortening.

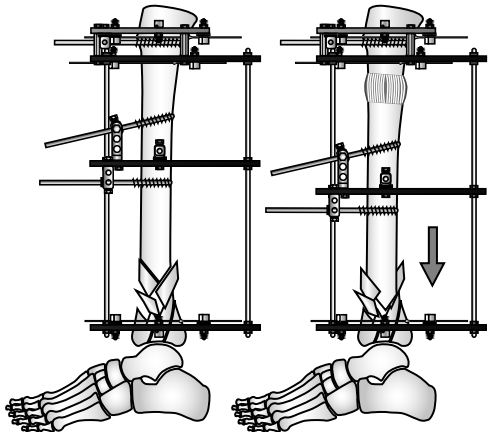

Figure 17 Intercalary bone transport to compress metaphyseal comminution or reconstruct bone loss is an excellent reconstruction technique.

the functional scores of the patients (4,24). Pilon fractures cause significant loss of function, especially to patients who were employed as physical laborers in the workplace (4,24).

There will be a subset of fractures, that will not heal with the initial treatment. These fractures will have poor callus formation on the medial aspect of the fracture, necrotic bone at the fracture site, or an infection that prevents healing. The frame is removed and the pin sites are allowed to heal. The patient is encouraged to walk on their cast or orthosis. The fracture will usually deform into varus because the fibula has healed. In fractures with viable bone, varus collapse will induce new hypertrophic callus and a Taylor Spatial frame (Smith Nephew, Memphis, Tennessee, U.S.A.) is used to distract through the callus to realign the fracture and heal the nonunion. Fractures with necrotic bone and infection require excision of the necrotic bone, leaving only viable bleeding bone. Reconstruction is done with squaring osteotomies and acute shortening combined with proximal lengthening for defects 3 cm or less. Larger defects are reconstructed with intercalary transport. If the plafond is necrotic or infected, the pilon will be reconstructed with ankle arthrodesis and proximal leg lengthening.

FUTURE TRENDS

Small titanium screws (2.0–2.7 mm) will eventually become available, which are long enough (45–55 mm) to fixate the plafond and allow fixation of smaller fragments. Bioengineered bone morphogenic proteins may eventually be developed that will be placed in the comminuted metaphysis and plafond, which will stimulate fracture healing, reducing the incidence of nonunion. Infection of the fracture continues to be the dreaded complication. Prevention of infection must be considered during the entire treatment course. Antibiotic coated pins may be helpful in this regard. Lastly, direct articular cartilage damage causes matrix breakdown and post-traumatic arthritis (25,26). Research into cartilage repair and amelioration of acute impact injury may develop biologic treatments, which will reduce the long-term functional degradation of cartilage after plafond fractures.

REVIEW OF LITERATURE

Authors	Population, Method	Results	Conclusions
Bacon et al. (1)	42 fractures, 28 ORIF, 14 circular fixator	Increased incidence nonunion, malunion, and infection in circular fixator group	Only study with poorer result Ilizarov technique compared to ORIF
Harris et al. (2)	63 ORIF, 16 circular fixator; selection bias for circular fixators to treat open and more severely comminuted fractures	Functional outcome was equivalent comparing ORIF to circular fixation	Recommended circular fixator for pilon fractures with open fractures and severe comminutions

(*Continued*)

REVIEW OF LITERATURE (*Continued*)

Authors	Population, Method	Results	Conclusions
Watson et al. (3)	105 fractures, 41 ORIF, 64 circular fixator; fractures comminution and soft tissue injury determined treatment group; Tscherne (2–3) and all open fractures treated circular fixator	Increased rate of malunion, nonunion, and wound complication ORIF group; poor outcome in C3 fractures; frame time 15 weeks	Ilizarov technique effective method to treat complicated tibial pilon fractures with severe soft tissue trauma
Pollack et al. (4)	80 fractures, 42 ORIF, 38 half-pin bridge; half pin group higher level comminution soft tissue injury	5/38 pilon fracture (12%) amputation rate; poorer ROM ankle bridging distraction	Midterm outcomes after pilon fractures not good; poor physical and psychosocial outcomes
Endres et al. (5)	50 fractures; all open fractures treated with Ilizarov; high incidence of C2/C3 fractures and soft tissue injuries in the group treated with Ilizarov technique	No incidence of pseudoarthrosis or osteitis in Ilizarov group; ORIF group, 5% osteitis, 2.5% delayed union, 8% arthrodesis; 18% wire infection Ilizarov group	Ilizarov technique effective method to treat complicated tibial pilon fractures with severe soft tissue trauma
Leung et al. (6)	31 fractures. 14 C2/C3	29% pintrack infection; frame time 14 weeks	Ilizarov comparable result to ORIF
Hutson and Zych (7)	100 fractures; 27 Type A, 73 Type C treated with Ilizarov fixator; 39% open fractures	Compartment syndrome foot (5), leg (4). Arthrodesis (4). Pintrack infection (10), deep infection (6), septic arthritis (1), nonunion (4), malunion (1), wound problem (0). 17/100 infection incidence; all infections in C3 fractures; frame time 23 weeks	Ilizarov technique effective method to treat complicated tibial pilon fractures with severe soft tissue trauma
Wyrsch et al. (8)	38 fractures. 18 ORIF, 20 bridging frame; only randomized prospective study	ORIF 3 open, 6 free flap, 6 deep infection, 3 amputation; EF, 7 open, 1 free flap; 1 septic arthritis, 5 malunion; bridging frame high incidence non/malunion; ORIF increased soft tissue problem	Study predated two-stage reconstruction, which has greatly improved soft tissue problems; testing of concept that circular wire fixators are indicated in pilon fractures with severe soft tissue will require randomized prospective study using 2-stage technique compared to circular wire technique
Hutson (9)	Technique article	An illustrated monograph with detailed technique chapters explaining biomechanics, reduction techniques, and fixation of tibia fractures	
Watson (10)	Technique article	Step-by-step approach for treating pilon fractures	

Abbreviation: ORIF, Open reduction internal fixation

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9 Acute Trauma: Soft-Tissue Reconstruction of the Leg

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INTRODUCTION

Obtaining adequate soft tissue coverage is one of the most important goals in the management of severe open injury of the lower limb. The primary goal of soft tissue coverage is satisfactory wound coverage with restoration of function. Aesthetic outcome often becomes a secondary concern. Many lower extremity trauma victims present with a paucity of soft tissue available for reconstruction. These conditions necessitate knowledge of the anatomy of the lower extremity as well as options for local soft tissue coverage, muscular rotational flaps, and free flap transfers.

The overall status of the traumatized patient must be thoroughly evaluated to ascertain the presence and extent of coexisting injuries. These injuries must then be addressed in accordance with optimal trauma management. Further analysis of the patient's baseline health is an integral part of the patient assessment. The patient must be in sufficiently sound health to undergo a potentially prolonged anesthetic course (1).

CLINICAL EVALUATION

The clinical examination of lower extremity injuries begins with evaluation of the size, location, and components of the wound. The examination proceeds with evaluation of the skin and surrounding soft tissue, including the quality of the fat, fascia, muscle, and bony defect or loss. The type of injury is divided into an avulsion, a crush, or an amputation subgroup. The status of the wound should also be determined as being clean, contaminated, or dirty. Contaminated and dirty wounds with obviously devitalized tissue are debrided, while tissues with marginal viability are observed in an effort to conserve as much tissue as possible.

Evaluation of the neurovasculature status of the leg begins with presence of pulses, active bleeding, exposed vasculature structures, and any obvious evidence of peripheral vascular disease. The clinical bedside evaluation including vascular exam of pulses in the context of injury may be followed by ankle brachial index, Duplex scan, magnetic resonance angiogram, or angiogram. In acute trauma, neurological assessment of nerve injury is paramount in the preoperative planning strategy. The neurological exam includes test of tactile sensation, vibration, and proprioception. Nerve injury repair is crucial to preserve protective sensation and more importantly provide postinjury ambulation. The presence of vascular or nerve injury may require special consideration necessitating possible vascular bypass and/or free flap reconstruction.

Special consideration is given to bony defects requiring hardware. These injuries often necessitate additional soft tissue coverage [i.e., microvascular free flap (MVFF)] to keep the hardware from becoming contaminated and to help prevent extrusion.

In our institution, we have found the vacuum-assisted device (VAC^{R}) to be an adjunct in the preparation of the wound for soft tissue reconstruction. The VAC device provides controlled, localized negative pressure wound therapy to help uniformly draw wounds close (2). It assists in granulation tissue, removes infectious material, and, additionally, can be applied to wounds with external hardware (3).

Table 1 Clinical Management

Classification	Clinical Evaluation	Surgical Approach	Pearls	Complications/Pitfalls
Thigh	I. Skin and subcutaneous loss	Pedicled rectus abdominus	Latissimus dorsi MVFF for large defects	Limited availability of local flaps
	II. Presence or absence of bony defects	Gracilis		Exposed femoral vessels with flap loss
	III. Muscle quality	Tensor fascia lata	Pedicled rectus or VRAM for exposed femoral vessels	
	IV. Vascular assessment			
	V. Available flaps	MVFF-Latissimus dorsi		
Upper third and knee	Same	Soleus, medial, or lateral gastrocnemius	Knee extensor mechanism has to be covered reconstructed flap	Lateral sural nerve injury with lateral gastrocnemius flap
Middle third	Same	Proximal soleus FDL EDL EHL Tibialis anterior	Flap length: width ratio can be extended to 3 : 1 or twice standard lengths	Functional loss with tibialis anterior harvest Great toe drop-extensor hallicus longus
Lower third and ankle	Same	MVFF Rectus abdominus Latissimus dorsi Serratus anterior Gracilis	Vascular structures coursing through zone of injury can be used	MVFF loss Infection
Foot	Same	MVFF Rectus abdominus Latissimus dorsi Serratus anterior Gracilis	Forefoot is an excellent recipient of STSG	MVFF loss Infection STSG loss

Abbreviations: VRAM, vertical rectus abdominus muscle; MVFF, microvascular free flap; STSG, split thickness skin graft; FDL, flexor digitorom longus; EDL, extensor digitorum longus; EHL, extensor digitorum longus.

CLASSIFICATION

Table 1 summarizes our approach to lower extremity soft reconstruction and review of the literature, respectively.

There are many classification systems available to organize patients in terms of a treatment plan in the acute trauma setting. Historically, the Gustilo-Anderson classification system (4,5) for lower extremity trauma has been used (Table 2). This system grades the severity of the soft tissue injury and correlates it with long-term limb function. We further subdivide the limb into thirds as described by Griffin and Thornton. The reconstructive ladder is then employed

Table 2 Gustilo-Anderson Classification of Open Fractures of the Tibia

Type	Description
I	Open fracture with a wound <1 cm
II	Open fracture with a wound >1 cm without extensive soft tissue damage
III	Open fracture with extensive soft tissue damage
IIIA	III with adequate soft tissue coverage
IIIB	III with soft tissue loss with periosteal stripping and bone exposure requiring soft tissue coverage
IIIC	III with arterial injury requiring repair

Source: From Ref. 6.

based on the soft tissue available, beginning with skin grafts (split-thickness or full-thickness), and proceeding to local advancement flaps, muscle and fasciocutaneous flaps, perforator flaps, and, ultimately, MVFF. Crush injuries and loss of tissue or large muscle prevent a local reconstruction leaving MVFF as the final common pathway. This will be impacted by zone of injury and determine the flap selected.

The tenets of lower extremity reconstruction include adequate debridement and preparation of the wound with full debridement and control of any wound infection prior to coverage as well as stabilization and management of associated orthopedic injuries. Additionally, the patient has to be prepared, with an overall assessment of patient suitability for reconstruction and rehabilitation potential.

Timing of reconstruction becomes a major consideration, especially in the postdebridement assessment. The factors influencing wound closure are bacterial status of the wound, type of fracture, different types of tissues involved in the injury, and the presence of exposed structures (7). In the case of an unstable patient, consideration is given to second tier reconstruction with a split-thickness skin graft (STSG) being used to initially close the wound. Microvascular free tissue transfer may then be employed at a later timeframe when the patient is otherwise optimized. The subsequent requirement for further osseous fixation, including the potential use of internal rigid fixation, mandates a close and continual relationship with the orthopedic service to determine the ultimate soft tissue coverage requirements.

Thigh

Defects of the thigh are well tolerated and will usually be covered with STSG or local advancement flaps. Thorough assessment of skin and muscle quality and availability is necessary for local advancement flaps. Exposed muscles without bony defects are typically covered with STSG from the contralateral leg or avulsed amputated tissue. Anterior thigh contour deformities and exposed femoral vessels are covered with pedicled Rectus abdominus or vertical Rectus abdominus myocutaneous flaps with or without skin grafts (6). Additional options include gracilis or tensor fascia lata muscle flaps. The microvascular free tissue transfer of the latissimus dorsi flap with or without skin is a favorite for very large defects of the thigh (6).

Upper Third and Knee

Rotational muscle flaps are the workhorse for defects of the upper third of the leg and the knee (8–10). Flap options include the medial head of the gastrocnemius muscle or the lateral head of the gastrocnemius muscle (with special care to avoid injury to the lateral sural nerve). Dibbell and Edstrom (11) recommend wide scoring of the fascia to facilitate long advancement. Moderate size defects, as large as 10×7 cm, can be covered with an extended gastrocnemius muscle adipofascial flap as an alternative method before considering free muscle transfer of the prepatellar region and the lower extremity (Figs. 1–4) (12). The proximally based soleus muscle or a bipedicled tibialis anterior muscle can be used for the lower portion of the upper third of the leg.

Rotational fasciocutaneous flaps are viable options for this portion of the leg as well. The blood supply for these flaps is based on the superficial perforating vessels of the deep arterial system and Doppler assessment is strongly recommended before undertaking these flaps (6). Although fasciocutaneous flaps are options for the proximal third of the leg, the standard

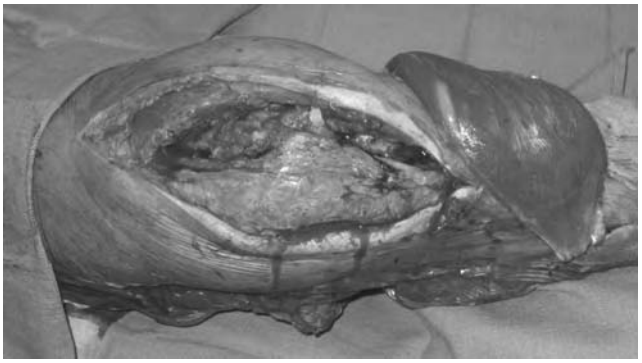

Figure 1 Proximal third defect with medial gastrocnemius muscle exposed.

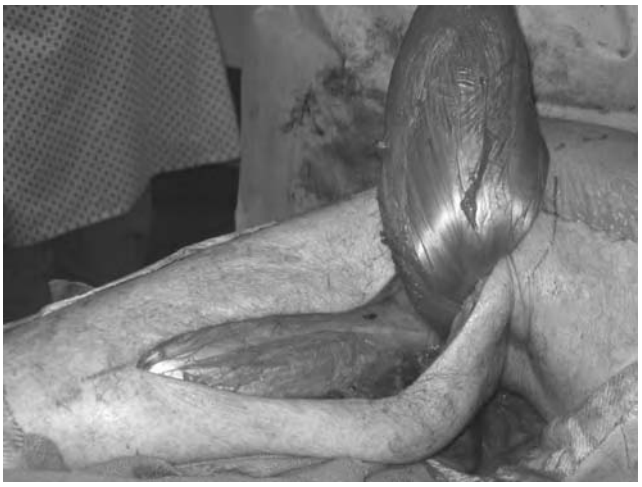

Figure 2 Medial gastrocnemius muscle elevated with scoring of the fascia to facilitate advancement.

rotational flap remains the medial gastrocnemius muscle flap. Keeping in mind that medial and lateral gastrocnemius and soleus flaps require STSG, special consideration for reconstruction of knee extensor mechanism has to be employed (6).

Middle Third

Multiple muscle rotational flaps options are available for middle third deficits; however, the medial gastrocnemius and soleus are the most reliable (13). Each flap option has special considerations (Table 3). Fasciocutaneous flaps for the middle third are typically based on medial or posterior lateral septocutaneous perforators and can be designed without identification of a perforating artery (3). The length to width ratio can be extended to 3:1 or twice that of a standard random cutaneous flap (13).

Lower Third and Ankle

MVFFs are primarily used in this area due to insufficient soft tissue available for transposition at this level (6). Several muscle and fasciocutaneous flaps can be designed to cover small defects in this region; however, MVFFs provide ideal coverage.

Microvascular free tissue transfer can deliver both soft tissue and skeletal support to large, complex wounds of the leg and are particularly useful in distal third of the leg and ankle (Figs. 5–7) (6). They provide large bulk to fill in dead space and can be expected to atrophy and

Figure 3 Medial gastrocnemius muscle advancement over defect.

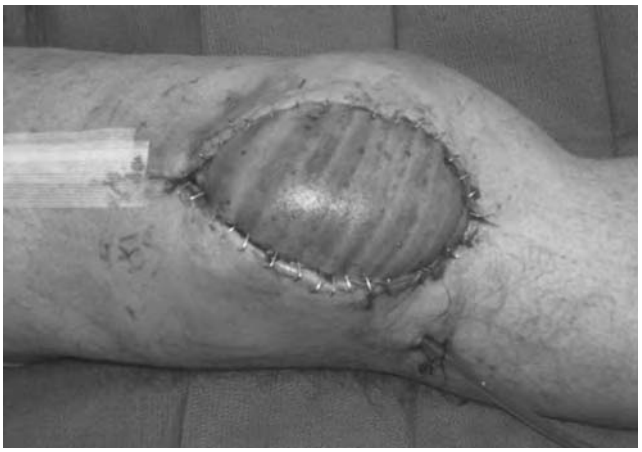

Figure 4 Medial gastrocnemius muscle and skin covering defect.

recontour if appropriately inset with the necessary tension and use of compression garments. Useful guidelines for free flap to lower extremity are (14):

1. Make end-end arterial and end-side or end-end venous anastomoses.
2. Reconstruct soft tissue first and then restore skeletal support.
3. Size of defect and the requirement necessary to fill any dead space.
4. Required pedicle length.

While the traditional teaching is using a pedicle out of the zone of injury (14), we prefer to rely on vasculature coursing through the zone of injury, which has been shown to have adequate flow, as proven by angiography (Figs. 8 and 9).

The preferred donor flaps are the rectus abdominus, latissimus dorsi, serratus anterior and, gracilis muscle flaps. The rectus abdominus provides a significant volume of muscle for moderate to large defects; however, an intraoperative positional change is required for leg defects. Using the single thoracodorsal pedicle, the latissimus can be combined with the Serratus for massive lower extremity defects. Both the latissimus and serratus have long vascular pedicles, which allow anastomoses to be performed well outside of the zone of injury. The gracilis muscle is an ideal flap for smaller volume defects, is relatively easy to harvest, has little or no donor morbidity, and adapts well to the leg contour. It has a short pedicle and is ideal for distal tibia, ankle, and foot defects (6).

Khouri and Shaw (15) report failure rates of MVFF reconstruction as high as 8% versus 3% for nonlower extremity cases. They found that the magnitude of the traumatic insult was the most significant factor associated with anastomotic failure.

Foot

Plantar surface defects are usually covered with STSG; however, they can only be used when a substantial portion of subcutaneous plantar pad is intact (6). Touam et al. (16) recommend the

Table 3 Middle Third Leg Flap Considerations

Donor Tissue	Special Considerations
Proximal soleus	Transferred without significant functional loss; can be carried to a point 5 cm above its tendinous insertion
Flexor digitorum longus	Transferred without significant functional loss because of supplemental action of the flexor digitorum brevis
Extensor digitorum longus	Used for small wounds < 5 cm. Superficial peroneal nerve must be preserved
Extensor hallicus longus	The distal tendon has to remain attached to the extensor digitorum communis to avert great toe drop
Tibialis anterior	Not expendable—necessary for dorsiflexion

Source: From Ref. 6.

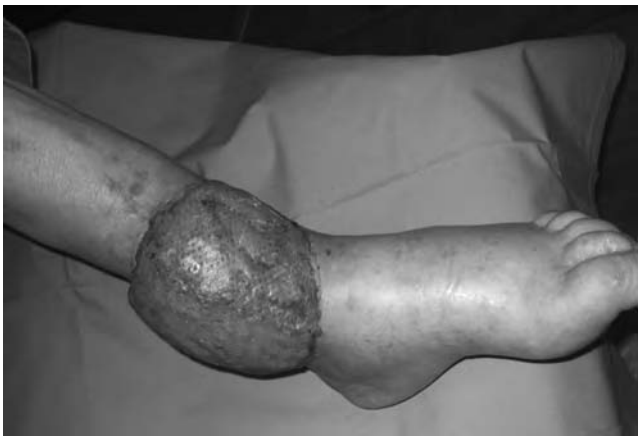

Figure 5 Coverage of lower third defect with microvascular free flap and split-thickness skin graft.

distally based sural neurocutaneous flap as the method of choice for covering skin defects over the foot, heel, ankle, and the lower one-fourth of the leg. Rotational flaps available for the heel include the plantar flap and the instep flap (17). MVFFs, such as the serratus anterior or gracilis, can also be used in this area given their small size.

The forefoot is an excellent recipient of STSG of full thickness skin graft (FTSG). They routinely provide adequate soft tissue for the non weight-bearing surfaces. When designing coverage for forefoot defects, the incision should not be placed on weight-bearing surfaces. Keeping in mind that the amount of tissue available after transfer is often less than expected, free flaps and fascial flaps containing skin or soft tissue are considered when bony surfaces have no overlying subcutaneous pad and local cutaneous coverage is not available (6).

While cross leg flaps are an option for lower extremity soft tissue reconstruction, they are seldom used instead of MVFF (18). It is only used when local pedicled flaps are unavailable, the patient is not a free flap candidate, or the patient has to remain immobilized for other reasons (19). Complications include a 40% rate of local flap necrosis and a 28% rate of infection.

SPECIAL CONSIDERATIONS

Tissue Expansion

Tissue expansion is primarily used to resurface areas of unstable soft tissue or unsightly scars. Their use is contraindicated around the ankle and foot, especially the plantar surface of the foot, and should be used with caution (20). The expansion begins one to two weeks after implantation. It often requires multiple operations and patients frequently voice their

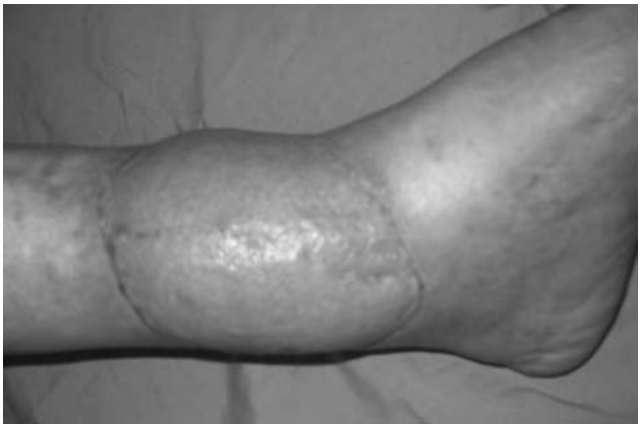

Figure 6 Lower third microvascular free flap two months postoperative (side view).

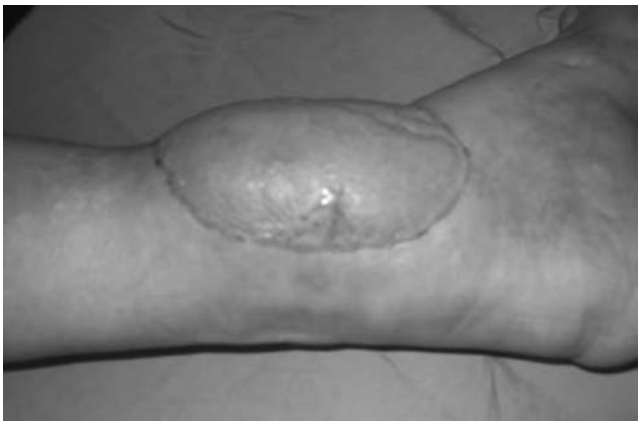

Figure 7 Lower third microvascular free flap two months postoperative (second view).

displeasure with the appearance. The infection rate ranges from 5% to 30% and overall complications are quoted to be as high as 75%.

Replantation

Replantation of the lower extremity is only considered when the patient is willing to accept multiple operations, transfusions, and the significant risk of complications (21). The goal of replantation must be directed not only at replanting the amputated part but also at providing a durable, sensate, and painless extremity capable of supporting the individual during the conduct of his or her normal daily activity. If the replanted part falls short of these goals, the alternative use of prosthetic device may be preferable (1).

Excessive bone loss is a factor for replantation but is no longer a contraindication. Bone loss can be reconstructed using conventional bone grafting for moderate loss and interpositional vascularized bone grafts for large loss. The Ilizarov technique has been successfully employed for defects of 15 cm or greater. Ischemia time is a key factor with a direct relationship between ischemia time and replant survival.

Elderly

The initial evaluation of elderly patients should proceed in the same manner as any other trauma patients. The presence of substantial comorbidities does not preclude free flap transfers (22). We advocate thorough medical workup, including cardiac and pulmonary risk stratification as well as evidence of peripheral vascular disease (PVD). In many instances, the presence of substantial comorbidities is better served with amputation given the difficult postoperative healing

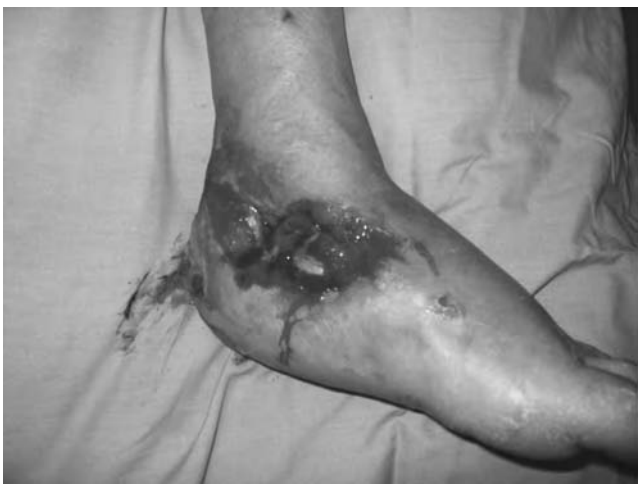

Figure 8 Complex wound with exposed bone after vacuum-assisted device dressing preparation.

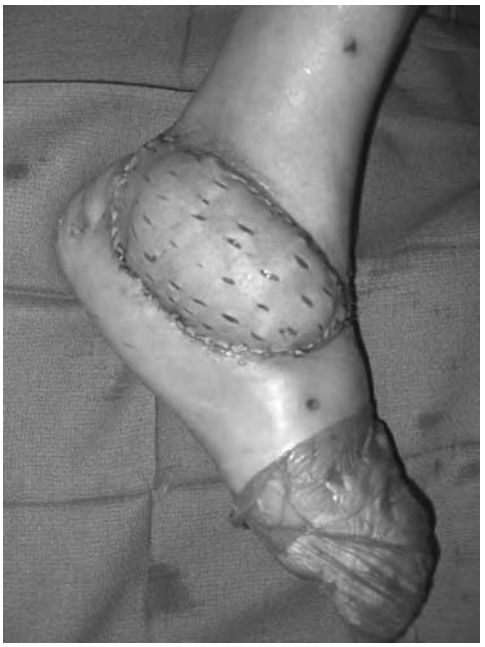

Figure 9 Coverage with microvascular free flap and split thickness skin grafts.

(i.e., diabetes, PVD, and chronic obstructive pulmonary disease). There is evidence that the results in elderly patients are worse than results in younger patients with microvascular free tissue transfers; however, flap failure and complications are more often related to patient comorbidities rather than purely patient's physiologic age. Complication rates include a 10% failure rate, 30% complication rate, and 8% to 10% mortality for patients older than 70 years (23,24).

FUTURE DIRECTIONS

The Ilizarov device is now routinely being used for soft tissue distraction in wound coverage of the lower extremity (6). The device has eliminated bone loss as a contraindication to replantation and facilitated treatment of those injuries once thought to be unsalvageable. The application of the Ilizarov technique is very encouraging; however the treatment time can be long and it is technically demanding (6). The complication primarily encountered is pin site sepsis, reported to occur from 0% to 30%. These infections are successfully treated with local care and antibiotics in most cases (6).

Another evolving tool in the armamentarium of treatment of traumatic soft tissue injuries is the VAC appliance. It effectively reduces the size of the wound, allowing more reconstructive options to be used.

REVIEW OF LITERATURE

Authors	Journal, Year	Title	Number of Patients	Results	Conclusion
C. Touam et al.	Plastic and Reconstructive Surgery February, 2001	Comparative study of two series of distally based fasciocutaneous flaps for coverage of the lower one-fourth of the leg, ankle and the foot	63	18.5% failure rate for lateral supramalleolar flap, 4.8% failure rate for sural neurocutaneous flaps	Distally based sural neurocutaneous flap is the method of choice for covering skin defects over the foot, heel, ankle, and lower one-fourth of the leg

(*Continued*)

REVIEW OF LITERATURE (*Continued*)

Authors	Journal, Year	Title	Number of Patients	Results	Conclusion
Yoon Jae Chung et al.	Annals of Plastic Surgery, 2002	Reconstruction of a lower extremity soft-tissue defect using the gastrocnemius musculoadipofascial flap	7	One partial skin necrosis	Extended gastrocnemius muscle adipofascial flap is an alternative to free flaps for moderate-size defects of the prepatellar region and lower extremity
Hiroshi et al.	Microsurgery, 2002	Vascularized composite tissue transfers for open fractures with massive soft-tissue defects in the lower extremity	39	9 revisions, 37 successful transfers	Free and island flaps can be used with satisfactory results in Type III fractures
Yazar et al.	Plastic and Reconstructive Surgery, 2004	One-stage reconstruction of composite bone and soft tissue defects in traumatic lower extremities	61	Flap survival 88.9%	One stage procedures for complex lower extremity defects equal to microsurgical staged procedures
DeFranzo	Plastic and Reconstructive Surgery, 2001	The use of vacuum-assisted closure therapy for the treatment of lower-extremity wounds with exposed bone	75	Successful coverage in 71 patients	VAC useful treatment of traumatic lower extremity wounds

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Part IV POST-TRAUMATIC RECONSTRUCTION

10 Aseptic Nonunions of the Tibia

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INTRODUCTION

Nonunions are a significant clinical problem in this country. It is estimated that 2% to 10% of all tibia fractures go on to nonunion. This results in a large number of revision procedures for nonhealing fractures, significant added morbidity for patients, and the expenditure of vast numbers of scarce healthcare dollars.

Bone is one of the few tissues in the body that heals by itself. Bone does not heal with scar, as does muscle and connective tissue. It heals with bone. Thus, fractures do not heal; they are repaired by a process of regeneration. A fracture union is one that is repaired to a degree that it is mechanically able to function like the de novo bone. The patient experiences no pain, and there is clinical stability at the fracture site. Fracture unions are accompanied by radiographic signs of healing. A delayed union is a fracture that although making progress toward union, has not healed in the usual amount of time for a similar fracture. A nonunion is a fracture that will not heal. It has sustained an arrest of the repair process, and has not shown radiographic or clinical progress toward healing for months. Nonunions may have some clinical stability, as they will have cartilage or fibrous interposition instead of bone. Others will be atrophic, with little healing tissue, and have no clinical stability. Though nonunions cannot be predicted, some fractures are destined to go on to nonunion from the beginning of treatment.

CLINICAL EVALUATION

The evaluation of a patient with a nonunion, just as with an acute injury, requires a thorough look at more than just the fracture pattern and the radiographs. One must determine the "personality of the fracture" as coined by Schatzker. This involves a complete history of the events of the injury, the fracture, the host, the treating physician, and the institution at which the treatment will occur. Only with this kind of analysis can one do proper preoperative planning and optimize the chance for success.

A complete history is essential. One must determine the mechanism of the fracture and other associated injuries that may have occurred such as those involving head, chest, abdomen, or other fractures. Was the initial injury open or closed? Was there a high-energy mechanism such as a motorcycle accident or a lower energy trip and fall? Were there any neurovascular issues at the time of initial injury or after treatment? A complete history of the initial and all other treatment provided is also necessary. A determination of the type and number of previous surgeries is essential, as is the presence and treatment of previous infection. If there is retained hardware at the fracture, old operative notes can be helpful in identifying the type and manufacturer for planned removal.

A complete picture of the host must also be developed. A thorough past medical and surgical history must be taken, as well as a list of current medications, allergies, and social habits. Have previous fractures healed in a timely fashion? Patients with recreational drug habits or other substance abuse may have compliance issues. Smokers are at risk because of the well-documented relationship between nicotine use and delayed healing. Patients using nicotine gum are not immune to this problem. The occupation of the patient is important, as treatment that requires a non-weight bearing gait will cause a longer period off work for a laborer than a patient with a more sedentary occupation. The knowledge of the avocations

and hobbies of your patient are also important, as it rounds out the level of activity to which the patient must return. Hospital discharge planning often begins before surgery. The patient's living situation, amount of support from family or friends, their financial resources, the location of their home, and the type of dwelling in which they reside are helpful in planning successful aftercare.

A complete musculoskeletal examination is important. Examination of the patient's other extremities will provide clues as to other disabilities that may play a role in mobility and later rehabilitation.

Examination of the nonunited segment includes an inspection for gross deformity and overall limb alignment. Gross limb length can be checked, and if the patient is ambulatory, the gait pattern should be examined. The skin should be inspected for the presence, location, and healing status of previous open wounds and incisions. The presence or absence of lymphedema or venous stasis should be noted. If previous external fixators have been in place, the condition of the old pin sites should be examined. A complete neurovascular examination should be carried out. The presence and character of the pulses should be noted. Patients with suspected dysvascular limbs should be sent for more thorough testing including transcutaneous oxygen tension and ankle-brachial indices. A complete neurological exam is mandatory. Existing nerve deficits can be tested by electromyography to determine the likelihood of recovery. The fracture site should be checked for pain to manual stress, as well as the presence of gross or subtle motion. The motion of adjacent joints should be examined. If there is joint contracture, it should be determined if it is due to soft tissue contracture, heterotopic ossification, or both.

Radiographic evaluation includes true anteroposterior and lateral films of the problem limb segment, orthogonal to the ''normal'' portion of the limb. If deformity or limb length issues are suspected, special films are required. Long leg alignment films and scanograms should be obtained. Comparison films of the contralateral leg are helpful in determining the normal alignment of the patient, and population normals can be used if the problem is bilateral. Computed tomography (CT) scans with reconstructions can be helpful in analyzing subtle nonunions, but can be hard to interpret with fracture fixation devices in place. Plain tomography can be very helpful in these instances. If infection is suspected, a combined bone scan and tagged white cell study can help differentiate bone turnover from active infection. Sinograms can be used to determine if chronic wounds communicate with the fracture site. Magnetic resonance imaging can be helpful in evaluation of a bone for infection, or looking at ligaments in adjacent joints, but are not commonly used in the evaluation of most nonunions.

Laboratory studies can round out the clinical picture of the patient. In addition to routine preoperative chemistries and blood counts, patients suspected of infection should have their erythrocyte sedimentation rate and a c-reactive protein checked. Patients suspected of malnutrition should have a complete nutritional panel drawn including liver enzymes, total protein, and albumin levels.

The last part of developing the personality of the fracture is an examination of the surgeon and the treating facility. Preoperative planning should include timely and appropriate consultation from plastic or microvascular surgeons if flaps or wound issues are anticipated, and vascular surgeons if poor vascularity is suspected. The patient's primary care physician can be helpful with chronic medical conditions. Surgeons should honestly examine whether they have the training, skill, patience, and experience necessary to treat a complex nonunion. The hospital is the final piece. Is the correct equipment in the house or available to be brought in? Is there experienced nursing and surgical assistance available? Can the anesthesia staff care for the needs of a sick patient?

At the end of the evaluation, the surgeon should create a problem list in anticipation of preoperative planning. This list should include pertinent positives about the patient's social condition and history, the physical examination, the bone, the skin, and the retained hardware. The consults required should also be listed as well as the equipment anticipated for the case. A preoperative plan should be prepared and drawn out in detail for all but the simplest of conditions.

CLASSIFICATION

Unlike acute fractures, there is no single definitive classification system for nonunions. Nonunions can be classified on the basis of their anatomy, the presence or absence of infection,

their biological potential, or their stiffness. Often more than one method of describing the nonunion will be helpful in determining a treatment plan.

The first issue is whether the fracture is a delayed union or a true nonunion. A delayed union may go on to a successful outcome if given more time, while a true nonunion will require intervention to achieve union. This is not a trivial question to answer for the patient. Though most nonunions will be diagnosed if the surgeon waits long enough, it is imperative to identify fractures that are falling behind as soon as possible in order to shorten overall treatment time and restore the patient back to full function. Delaying intervention for an arbitrary length of time before calling a fracture a nonunion results in more disability, more time off work, and greater psychological stress for the patient. As soon as slow healing is identified, a frank discussion of the possibility of nonunion should be had with the patient about the need for further future treatment. Many patients will opt for early intervention if it means an earlier return to work or recreational activities.

Nonunions are also classified by their anatomic location. Diaphyseal nonunions have relatively less biological potential as they involve cortical bone, but are amenable to a wide variety of treatment methods including nails, compression plating, and external fixation. The goal in this instance is to restore length and axial alignment while achieving fracture union. As the nonunion reaches the metaphyseal region, the goals remain the same, but the options for fixation are more

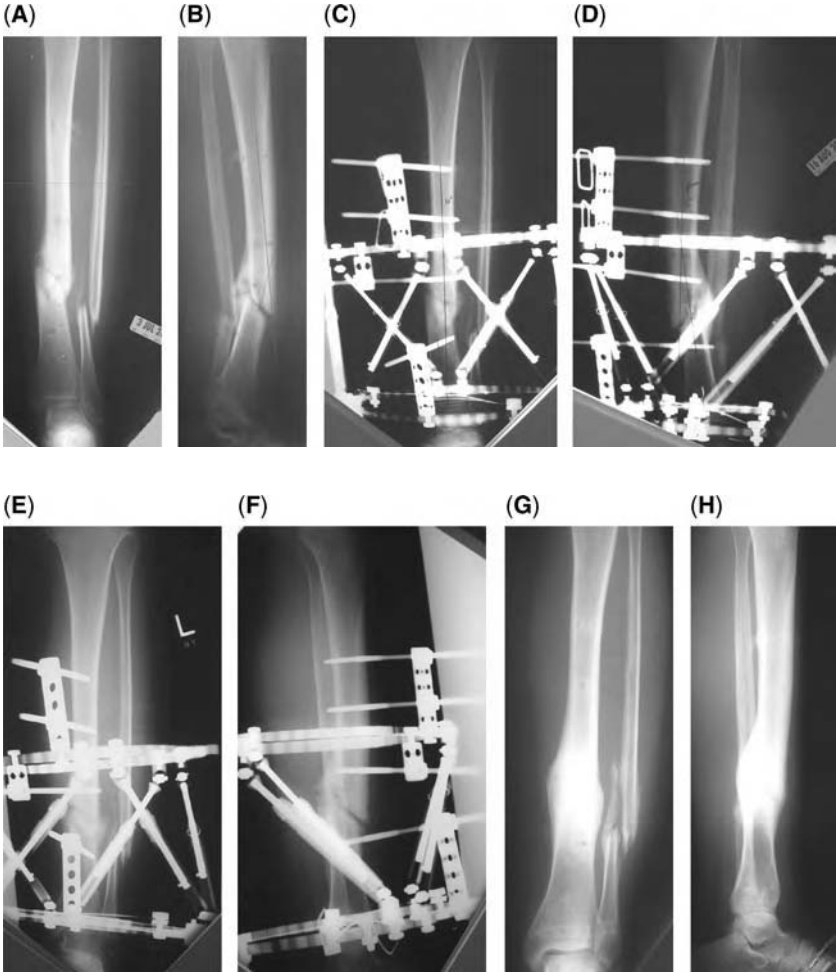

Figure 1 Anteroposterior and lateral (**A**, **B**) of a 15-year-old girl with an aseptic malpositioned nonunion after an open tibia fracture treated initially with a uniplanar external fixator. The nonunion was treated percutaneously with a Taylor Spatial Frame. The initial mounting (**C**, **D**) and post correction views (**E**, **F**) are illustrated. The patient went on to union (**G**, **H**) at 12 weeks.

limited. Periarticular nonunions may also be associated with stiff or contracted joints that must be accounted for in the preoperative plan. Nonunions of the articular surface are particularly challenging. Defining the extent of the nonunited segment may require multiple radiographs and CT scans. Step-offs, gaps, and injury to the joint surface may lead to local or global articular arthritis. Treatment may consist of open reduction and rigid fixation, arthrodesis, or arthroplasty.

Nonunions may be aseptic or infected. Though many authors have shown that bone constructs with adequate stability can heal in the face of infection, the general goal is to convert an infected nonunion into a noninfected nonunion, and then proceed with treatment of the fracture. Though many infected nonunions will have skin breakdown, open wounds, and drainage, the diagnosis is not always obvious. Laboratory studies can be helpful as can nuclear medicine studies. The patient should be counseled that treatment may take several staged procedures for hardware removal, debridement of dead bone, soft tissue coverage, and stabilization. A period of intravenous antibiotics-based thorough deep cultures is followed by definitive reconstruction. Depending on the extent of the infection and bone resected, this may require a period of months. Failed soft tissue coverage, failure to eradicate the infection, or obtain union may lead to eventual amputation.

Nonunions can be classified on the basis of their biological potential. Hypertrophic nonunions are characterized by abundant bone formation and are often referred to as having the appearance of an elephant foot. In general, they are stiff and relatively stable. Patients are often able to bear weight with pain on a hypertrophic nonunion. They have excellent blood supply and biological potential, and often require only the addition of stability for the fracture to unite. Atrophic nonunions, on the other hand, have little biological potential. Atrophic nonunions are often the result of open fractures or previous surgical procedures that have caused a disruption of the normal vascular supply to the bone. They have had a cessation of the regeneration process, resorption of the bone ends and sometimes capping off of the endosteal canal of the bone. These nonunions are mobile; patients usually are unable to bear weight and may require external immobilization for comfort. A special case of the atrophic nonunion is a true pseudarthrosis in which a false joint has been created between the two ends of the bone. These fractures need biological stimulation in addition to skeletal stability. Bone grafting and other adjuvants often play a role in their treatment. Oligotrophic nonunions are somewhere in between these two extremes. They have very little callous formation, but the bone ends are vital. They often require both biological and mechanical augmentation.

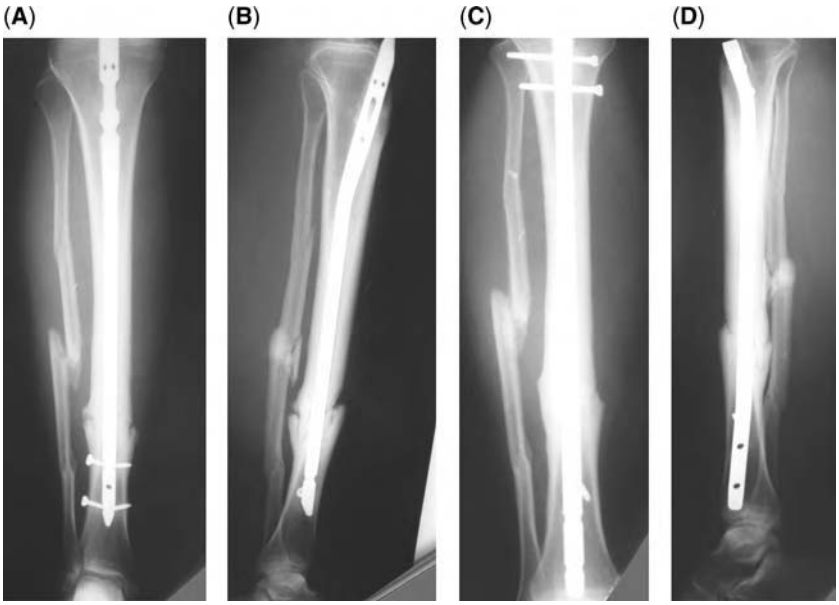

Figure 2 Anteroposterior and lateral (**A**, **B**) of a hypertrophic nonunion after nailing of a closed tibial fracture. The fracture went on to rapid union after a reamed exchange nailing (**C**, **D**).

Table 1 Clinical Management

Classification Group	Goal	Surgical Tactic	Pearls	Pitfalls
Hypertrophic nonunion	Provide stability	Plate, nail, external fixation	Does not require grafting, do not disturb biology	Failure to provide adequate fixation
Atrophic nonunion	Provide biological simulation and stability	Bone graft or substitute, provide stability	Thorough debridement of bone ends is a must	Failure to provide biological stimulation
Nonunion with deformity	Correct deformity and nonunion	Osteotomy or osteoplasty, provide biology and stability	Fully analyze deformity including length	Failure to correct deformity
Diaphyseal nonunion	Maintain axial alignment and length	Nail, external fixation, plate	Exchange nailing is primary technique	Maintain length, rotation, and axial alignment
Metaphyseal nonunion	Maintain axial alignment and length	Plate, external fixation	Carefully plan periarticular fixation	Maintain angular alignment
Articular nonunion	Preservation of the reconstructed joint	Rigid internal fixation, arthroplasty	Comminuted nonunions require arthroplasty	Prognosis of the joint is poor

TREATMENT OPTIONS

The options available include internal or external fixation and acute or gradual correction (Table 2). External fixation is especially helpful if there is large deformity that is most safely handled with gradual correction. The presence of limb length discrepancy (LLD) is an indication for external fixation and the Ilizarov method where simultaneous lengthening can be done either at the nonunion site or through an osteotomy at a different location. Internal fixation is most suitable when there is a healthy soft-tissue envelope and when an acute correction can be done.

Hypertrophic nonunions are biologically viable, and require mechanical stability and correct alignment to promote union. Stable constructs allow compression and have no shearing at the nonunion site. A stable construct made up of the patient's bone and the fixation device allows stable vascular in-growth and the progression of fracture healing. Multiple methods including plating, exchange nailing, or external fixation can provide this stability depending upon the personality of the injury. An excellent application of gradual correction

Table 2 Treatment Options (Editor's Addendum)

Treatment Approach	Best Indication
Acute correction	Minimal deformity
	Atrophic nonunion with open bone grafting
	No LLD
Gradual correction	Large deformity
	Stiff nonunion with deformity
	Associated LLD
	Bone defect
Plate and screw fixation	Metaphyseal/periarticular location
	Excellent soft-tissue envelope
	No infection
Intramedullary nailing	Intramedullary nail in place
	Need for exchange nailing
	Diaphyseal location
	No infection
Circular external fixation	Large deformity
	Stiff nonunion with deformity
	Associated LLD
	Poor soft-tissue envelope
	Concern about infection
	Bone defect
	Metaphyseal/periarticular location
	Diaphyseal location

with external fixation is the treatment of a hypertrophic stiff nonunion with deformity. Focal compression and distraction, with simultaneous correction of associated angular deformities according to the methods of Ilizarov is performed. The hypertrophic nonunion has fibrocartilage tissue in the nonunion site and has the biologic capacity for bony union. It lacks stability and axial alignment. Gradual distraction of this type to achieve normal alignment results in bone formation. The nonunion acts like regenerate and bony healing occurs. Modest lengthening of no more than 1.5 cm should be done through the nonunion. If additional lengthening is needed, a second osteotomy for lengthening is performed. If preexisting hardware is present, it is removed. Fractures without previous hardware can be treated percutaneously in a frame only. A stable external fixator is mounted on the tibia, and compression is instituted for 10 to 14 days. At the end of this period, the limb is distracted gently. Concurrent angular deformities are corrected at this time. Once any deformity is corrected, the patient is encouraged to bear weight as tolerated. This method may be used with a traditional Ilizarov external fixator with hinges and threaded rods or a Taylor Spatial Frame. Several studies have confirmed Ilizarov's success with this technique. The principal advantages are: not having to open the nonunion site in the face of poor skin and widened callus and gaining length through an opening wedge correction. This technique is not useful for mobile atrophic nonunions and less applicable to infected nonunions.

Hypertrophic nonunions can also be treated with internal fixation. Compression plating and intramedullary nailing can be used successfully, especially in the absence of large deformity and with a healthy soft-tissue envelope.

Atrophic nonunions require biological augmentation and mechanical stability. Patients with bone loss and infection fall into this category, but are discussed in more detail in other chapters. These nonunions require the most preoperative planning as the physiological environment is inadequate to promote healing. Medical problems must be treated, while vascular surgery and plastic surgery consults may be necessary to correct soft tissue problems. These nonunions must be opened and the bone ends debrided back to healthy viable tissue. All nonviable scar tissue but be removed and the endosteal canal of the bone must be opened, either with a curette or a drill bit. These fractures require bone grafting. Though the gold standard remains autogenous cancellous bone from the iliac crest, other methods such as allograft bone with or without bone marrow aspirate or commercially available bone graft substitutes are under active investigation. The addition of biologically active compounds such as platelet derived growth factor or bone morphogenic proteins is supported by good early data, and holds great promise for the stimulation of recalcitrant nonunions. Stability must then be provided either with an external fixator or other internal fixation device.

COMPLICATIONS

The complications incurred in the treatment of nonunions are those of each individual form of fixation. External fixators are prone to pin tract infections, which may be avoided by careful pin and wire insertion technique. Plating techniques require careful handling of the soft tissues and the avoidance of stripping. The biggest complication that the nonunion surgeon may avoid is the failure to adequately plan the case. Poor hosts with untreated medical, vascular, or soft tissue problems will not do well. Failure to adequately debride the nonunion and provide biological stimulation will doom an atrophic nonunion to failure. Finally, the surgeon must provide a stable construct that allows fracture healing to occur while the patient begins early functional rehabilitation.

FUTURE DIRECTIONS

With the advent of locked plating constructs and computer-assisted wire external fixation techniques, fracture fixation technology has made quantum leaps forward in the past five years. Larger series with long-term results will help clarify the indications and technical aspects used to optimize their success. The introduction of biologically active factors directly to the fracture construct promises to revolutionize fracture treatment in the years to come. Though some of these factors are commercially available now, it remains to be seen which bone morphogenetic protein (BMP) or growth factor is most active and which method of administration is most useful.

REVIEW OF THE LITERATURE

Authors	Source	Title	Results	Take Home Message
Wiss and Stetson	J Orthop Trauma, 1994	Nonunion of the tibia treated with reamed IM nail	47 pts, 89% union	Effective technique, care must be taken after external fixation for extended periods
Richmond et al.	J Orthop Trauma, 2004	Nonunions of the distal tibia treated by reamed IM nailing	32 patients, reamed IM nail, 29/32 healed in 3.5 mo, all eventually unite	Reamed IM nailing is an excellent technique in the distal tibial shaft
Schmitz et al.	Clin Orthop Relat Res, 1999	Effect of smoking on tibial shaft fracture healing	146 pts with tibia fracture, smokers healing significantly longer (269 to 136 days)	Smokers requiring treatment require more time to heal than nonsmokers in nailed or ex-fix treatment
Feldman et al.	J Orthop Trauma, 2003	Correction of tibial malunion and nonunion with six-axis analysis deformity correction using the Taylor Spatial Frame	18 pts, 17 unions with deformity correction; 15 returned to preinjury activity	TSF is a reliable technique for tibial malunions and nonunions
Paley et al.	Clin Orthop Relat Res, 1989	Ilizarov treatment of nonunions with bone loss	25 pts, 100 % union at mean 13.6 mo	Bone transport with deformity correction is a reliable technique for managing nonunions with bone loss and deformity

Abbreviation: IM, intramedullary

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11 Nonunions of the Femoral Shaft and Distal Femur

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INTRODUCTION

Nonunion following a femur fracture is relatively uncommon. The rate of nonunion following intramedullary nail fixation for a femur fracture is generally believed to be 2% or less (1–3). Although most femur fractures heal uneventfully, those that progress to nonunion tend to be stubborn. Healing the bone presents the primary clinical challenge (4). Soft tissue problems are infrequent in femoral nonunions due to the nature of the thigh musculature.

Various treatment options have been described to treat these challenging problems. Table 1 reviews the available literature on femoral nonunions by treatment, anatomic location, subgroup (uninfected, infected, and segmental defect), and most recent failed treatment.

CLINICAL EVALUATION

The clinical evaluation consists of the patient history and physical examination. The patient history should include the date and mechanism of the initial injury as well as any preinjury medical problems (diabetes, malnutrition, metabolic bone disease, etc.), disabilities, or associated injuries that might affect the treatment plan or outcome. All prior surgeries to treat the fracture and fracture nonunion should be reviewed. The history should also include details regarding infection. Finally, the patient should be questioned regarding other possible contributing factors for nonunion. Any current use of nonsteroidal anti-inflammatory drugs (NSAIDs) should be discontinued.

The physical examination is used to document the current status of the affected limb and the functional status of the patient. The nonunion site and the hip and knee joints should undergo manual testing to evaluate motion, pain, and stability. The presence of active drainage and sinus formation should be noted. The presence of deformity at the fracture site should be noted and described. A neurovascular examination should be performed to rule out or document vascular insufficiency and motor or sensory dysfunction.

RADIOLOGIC EVALUATION

The radiologic evaluation begins with a review of the original fracture films and subsequent radiographs of the salient aspects of previous treatments. This review allows evaluation of the character and severity of the initial injury and of the progress or lack of progress toward healing. The prior plain films should be carefully examined for the status of any orthopedic hardware (e.g., loose, broken, inadequate in size or number of implants) including its removal or insertion on subsequent films. The evolution of deformity at the nonunion site over time should be evaluated. The time course of missing or removed bony fragments, bone grafting, and implanted bone stimulators allows for an assessment of the associated fracture repair response.

Current radiographs should be taken and should include (i) a 36 in anteroposterior (AP) and lateral radiograph of the entire femur, including the hip and knee joint; (ii) AP, lateral,

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Table 1 Literature Review of Femoral Nonunions

Anatomic Location	Subgroup	Most Recent Failed Treatment	Authors	Treatment (Number of Cases)	Success Rate	Time to Bony Union	Adjunctive Treatments
Subtrochanteric	Uninfected	Intramedullary nail	Haidukewych and Berry (5)	Exchange nailing (11 cases)	100%	Not reported	Cancellous autograft or allograft or both (number of cases not reported)
		Plate and screw fixation	Haidukewych and Berry (5)	Repeat plate and screw fixation (8 cases)	88%	Not reported	Cancellous autograft or allograft or both (number of cases not reported)
			Charnley and Ward (6)	Intramedullary nail (2 cases)	100%	5 mo	None
			Kempf et al. (7)	Intramedullary nail (7 cases)	86%	4 mo	None
			Haidukewych and Berry (5)	Intramedullary nail (4 cases)	100%	Not reported	Cancellous autograft or allograft or both (number of cases not reported)
Diaphyseal	Uninfected	Intramedullary nail	Bungaro et al. (8)	Plate and screw fixation (7 cases)	100%	Not reported	Autologous cancellous bone grafting (all cases)
			Bai et al. (9)	Plate and screw fixation (6 cases)	83%	6 mo	Composite graft of bovine BMP and plaster (all cases)
			Wu and Shih (10)	Plate and screw fixation (14 cases)	79%	5 mo	Autologous cancellous bone grafting (13 cases)
			Bellabarba et al. (11)	Plate and screw fixation (23 cases)	91%	3 mo	Autologous cancellous bone grafting (13 cases)
			Cove et al. (12)	Plate and screw fixation (8 cases)	100%	6 mo	Autogenous bone grafting (all cases)
			Ueng et al. (13)	Augmentative plating (10 cases)	100%	8 mo	None
			Ueng and Shih (14)	Augmentative plating (5 cases)	100%	5 mo	Cancellous bone grafting (3 cases)
			Pihlajamaki et al. (15)	Exchange nailing (11 cases)	64%	10 mo	Autogenous bone graft (3 cases)
			Wu and Chen (16)	Exchange nailing (36 cases)	92%	4 mo	None
			Wu and Shih (10)	Exchange nailing (32 cases)	81%	4 mo	Autologous cancellous bone grafting (9 cases)
			Heiple et al. (17)	Exchange nailing (5 cases)	96%	4 mo	None
			Christensen (18)	Exchange nailing (8 cases)	100%	Not reported	None
			Cove et al. (12)	Exchange nailing (2 cases)	100%	8 mo	Autogenous bone grafting (1 case)
			Furlong et al. (19)	Exchange nailing (25 cases)	96%	7 mo	Autogenous bone grafting (12 cases)
			Kempf et al. (7)	Exchange nailing (6 cases)	83%	4 mo	None
			Hak et al. (20)	Exchange nailing (18 cases)	72%	Not reported	None
			Harper (21)	Exchange nailing (8 cases)	75%	7 mo	Autogenous bone grafting (5 cases)
			Oh et al. (22)	Exchange nailing (11 cases)	100%	Not reported	None

	Wu et al. (23)	Exchange nailing (45 cases)	96%	4 mo	Corticocancellous bone grafting and acute lengthening (all cases)
	Wu and Chen (24)	Exchange nailing (16 cases)	100%	4 mo	None
	Weresh et al. (25)	Exchange nailing (19 cases)	53%	8 mo	None
	Finkemeier and Chapman (26)	Exchange nailing (28 cases)	68%	11 mo	Autogenous bone grafting (11 cases)
	Pihlajamaki et al. (15)	Nail dynamization (19 cases)	74%	5 mo	Autogenous bone graft (2 cases)
	Menon et al. (27)	Slow compression over a nail using external fixation (SCONE)(2 cases)	100%	8 mo	None
	Brinker and O'Connor (28)	SCONE (3 cases)	100%	4 mo	None
	Wu and Chen (24)	Open autogenous bone grafting (19 cases)	100%	5 mo	None
	Pihlajamaki et al. (15)	Autogenous bone grafting (5 cases)	0%	Not applicable	None
Plate and screw fixation	Bai et al. (9)	Repeat plate and screw fixation (4 cases)	100%	6 mo	Composite graft of bovine BMP and plaster (all cases)
	Cove et al. (12)	Repeat plate and screw fixation (12 cases)	100%	8 mo	Autogenous bone grafting (10 cases), allograft (2 cases)
	Wu et al. (29)	Intramedullary nail (8 cases)	88%	4 mo	Autologous corticocancellous bone grafting (all cases)
	Wu et al. (30)	Intramedullary nail (21 cases)	100%	5 mo	Reaming bone grafting (all cases)
	Bai et al. (9)	Intramedullary nail (5 cases)	100%	6 mo	Composite graft of bovine BMP and plaster (all cases)
	Wu and Shih (10)	Intramedullary nail (29 cases)	86%	4 mo	Autologous cancellous bone grafting (14 cases)
	Heiple et al. (17)	Intramedullary nail (4 cases)	100%	4 mo	None
	Christensen (18)	Intramedullary nail (4 cases)	100%	Not reported	None
	Kempf et al. (7)	Intramedullary nail (8 cases)	100%	4 mo	Autogenous cancellous bone grafting (4 cases)
	Harper (21)	Intramedullary nail (3 cases)	100%	4 mo	Autogenous bone grafting (2 cases)
	Wu et al. (23)	Intramedullary nail (6 cases)	83%	3 mo	Corticocancellous bone grafting; acute lengthening (all cases)
Traction and casting	Heiple et al. (17)	Intramedullary nail (16 cases)	94%	4 mo	None
	Wu and Shih (10)	Intramedullary nail (4 cases)	75%	5 mo	Autologous cancellous bone grafting (1 case)
	Christensen (18)	Intramedullary nail (5 cases)	100%	Not reported	None
	Harper (21)	Intramedullary nail (5 cases)	80%	8 mo	Autogenous bone grafting (4 cases)

(*Continued*)

Table 1 Literature Review of Femoral Nonunions (*Continued*)

Anatomic Location	Subgroup	Most Recent Failed Treatment	Authors	Treatment (Number of Cases)	Success Rate	Time to Bony Union	Adjunctive Treatments
Diaphyseal	Infected	Intramedullary nail	Klemm (31)	Exchange nailing (16 cases)	100%	Not reported	Internal irrigation after nail placement; irrigation, and debridement after nail removal following consolidation (all cases)
			Christensen (18)	Exchange nailing (3 cases)	100%	Not reported	None
			Hak et al. (20)	Exchange nailing (5 cases)	100%	Not reported	None
			Oh et al. (22)	Exchange nailing (2 cases)	100%	Not reported	None
			Kostuik and Harrington (32)	Incision and drainage, retained nail (10 cases)	40%	18 mo	None
			Cove et al. (12)	Serial debridements followed by plate and screw fixation (3 cases)	100%	18 mo	Vascularized fibular transfer and autogenous bone grafting (all cases)
			Barquet et al. (33)	Serial debridements followed by external fixation (6 cases)	83%	9 mo	Cancellous bone grafting (all cases)
			Slätis and Paavolainen (34)	Debridement and external fixation (2 cases)	100%	5 mo	Cancellous bone grafting (all cases)
			Ueng et al. (35)	Debridement followed by external fixation (9 cases)	100%	8 mo	Antibiotic-eluting beads for 2 to 6 wk and cancellous bone grafting (6 cases), and vascularized fibular transfer (3 cases)
		Plate and screw fixation	Klemm (31)	Intramedullary nailing (21 cases)	86%	Not reported	Internal irrigation after nail placement; irrigation, and debridement after nail removal following consolidation (all cases)
			Kostuik and Harrington (32)	Incision and drainage, intramedullary nail (4 cases)	50%	36 mo	None
			Cove et al. (12)	Serial debridements followed by external fixation (2 cases)	100%	7 mo	Autogenous bone grafting (all cases)
			Barquet et al. (33)	Serial debridements followed by external fixation (4 cases)	100%	9 mo	Cancellous bone grafting (all cases)
			Slätis and Paavolainen (34)	Debridement and external fixation (3 cases)	67%	5 mo	Cancellous bone grafting (all cases)
		External fixation	Ueng et al. (35)	Debridement followed by external fixation (5 cases)	100%	10 mo	Antibiotic-eluting beads for 2 to 6 wk and cancellous bone grafting (all cases)
		Traction and casting	Cove et al. (12)	Serial debridements followed by external fixation (5 cases)	80%	11 mo	Vascularized fibular transfer and autogenous bone grafting (all cases)

	Segmental defect	Intramedullary nail	Ueng et al. (13)	Augmentative plating over a retained intramedullary nail (7 cases)	100%	7 mo	Cancellous bone grafting (all cases)
		Following debridement or traumatic bone loss	Jupiter et al. (36)	Vascularized fibular graft (7 cases)	71%	5 mo	None
			Muramatsu et al. (37)	Vascularized fibular graft (17 cases)	94%	8 mo	None
			Yajima et al. (38)	Vascularized fibular graft (20 cases)	75%	6 mo	Autogenous bone grafting (9 cases)
			Song et al. (39)	Vascularized fibular graft (17 cases)	60%	9 mo	Autogenous bone grafting at docking site (all cases)
			Wei et al. (40)	Vascularized fibular graft (10 cases)	50%	8 mo	None
			Wei et al. (40)	Vascularized fibular graft (7 cases)	100%	8 mo	None
			Hou and Liu (41)	Vascularized fibular strut graft (5 cases)	100%	7 mo	None
			Chapman (42)	Intramedullary nailing and closed intramedullary bone grafting (8 cases)	100%	Not reported	None
			Song et al. (39)	Bone transport (20 cases)	70%	10 mo	None
			Smrke and Arnez (43)	Bone transport (3 cases)	100%	34 mo	Free flap transfer (all cases)
			Jaffe et al. (44)	Bulk allograft (4 cases)	75%	6.7 mo	Fixation with dynamic compression plate (all cases)
Supracondylar	Uninfected	Intramedullary nail	Brinker and O'Connor (28)	SCONE (2 cases)	100%	6 mo	None
		Plate and screw fixation	Bellabarba et al. (45)	Repeat plate and screw fixation (20 cases)	100%	4 mo	Bone graft substitute (1 case)
			Chapman and Finkemeier (46)	Repeat plate and screw fixation (16 cases)	94%	8 mo	Autogenous bone graft (15 cases) and bone-graft substitute (1 case)
			Wang and Weng (47)	Repeat plate and screw fixation (10 cases)	100%	5 mo	Cortical allograft struct grafts and corticocancellous autograft (all cases)
			Koval et al. (48)	Retrograde intramedullary nailing (16 cases)	25%	17 mo	Autogenous bone graft (13 cases)
			Kempf et al. (7)	Antegrade intramedullary nail (5 cases)	100%	4 mo	None
			Ali and Saleh (49)	External fixation (10 cases)	100%	10 mo	None

(*Continued*)

Table 1 Literature Review of Femoral Nonunions (*Continued*)

Anatomic Location	Subgroup	Most Recent Failed Treatment	Authors	Treatment (Number of Cases)	Success Rate	Time to Bony Union	Adjunctive Treatments
			Haidukewych et al. (50)	Total knee arthroplasty (15 cases)	80%	Not applicable	None
			Freedman et al. (51)	Tumor replacement prosthesis (2 cases)	100%	Not applicable	None
	Infected	Plate and screw fixation	Ali and Saleh (49)	Debridement followed by external fixation (5 cases)	80%	11 mo	None
			Freedman et al. (51)	Tumor replacement prosthesis (1 case)	100%	Not applicable	None
		External fixator	Chapman and Finkemeier (46)	Plate and screw fixation (2 cases)	100%	14 mo	Autogenous bone graft (all cases)
	Periprosthetic		Anderson et al. (52)	Total knee arthroplasty, long femoral stem (6 cases)	83%	9 mo	None
Condylar (intra-articular)	No cases reported in the literature						

Abbreviation: BMP, bone morphogenetic protein

and two oblique views of the nonunion site itself on small cassette films for improved magnification and resolution; and (iii) standing AP, 51 in alignment radiographs of both limbs to assess leg length discrepancies and deformities.

The current plain films are used to assess the following characteristics: (i) anatomic location, (ii) healing effort, (iii) bone quality, (iv) surface characteristics [(a) surface area of adjacent fragments, (b) extent of current bony contact, (c) orientation of fracture lines, and (d) stability to axial compression], (v) status of previously implanted hardware, and (vi) deformities [that should be characterized by location, magnitude, and direction and should include a description of the deformity in terms of i. length, ii. angulation, iii. rotation, and iv. translation (53–55)].

For femoral nonunions, the anatomic location is classified as subtrochanteric, diaphyseal, supracondylar, or condylar (i.e., when intra-articular involvement is present). Diaphyseal nonunions involve primarily cortical bone, whereas distal femoral metaphyseal nonunions largely involve cancellous bone.

The radiographic assessment of healing effort includes evaluating radiolucent lines and gaps and callus formation. The assessment of bone quality includes observing (i): sclerosis; (ii) atrophy; (iii) osteopenia; and (iv) bony defects.

COMPUTED TOMOGRAPHIC SCANNING AND TOMOGRAPHY

Assessment of bony healing in femoral nonunions may be difficult because overlying hardware may obstruct plain radiographs. In such cases, computed tomographic (CT) scans are particularly helpful in estimating the percentage of the cross-sectional area that shows bridging bone. The cross-sectional area of bridging bone may be followed on serial CT scans to evaluate the progression of fracture consolidation.

CT scans are also useful for assessing articular step off, joint incongruency, and bony healing in cases of intra-articular nonunions. Rotational deformities of the femur may be accurately quantified using CT by comparing the relative orientations of the proximal and distal segments of the involved bone to the contralateral normal bone (56–60).

NUCLEAR IMAGING

A variety of studies, when used in concert, are useful for assessing: (i) bone vascularity at the nonunion site, (ii) the presence of a synovial pseudarthrosis, and (iii) infection.

Technetium-99m-pyrophosphate (bone scan) complexes will show increased uptake in viable nonunions but decreased tracer uptake in nonviable nonunions. The diagnosis of synovial pseudarthrosis can be made by technetium-99m-pyrophosphate bone scanning, which will show a ''cold cleft'' at the nearthrosis between the hot ends of the ununited bone (61–64).

Radiolabeled white blood cell scans [such as with indium-111 or technetium-99m hexamethylpropyleneamine oxime (HMPAO)] are useful tools for the evaluation of acute infections of bone. Gallium scans are useful for the evaluation of chronic infections of bone. The combination of a gallium-67 citrate scan and a technetium-99m sulfa colloid bone marrow scan can clarify the diagnosis of chronic infection.

OTHER RADIOLOGIC STUDIES

Ultrasonography is useful for assessing the status of the bony regenerate (distraction osteogenesis) during bony transport or lengthening. Ultrasonography is also useful for confirming the presence of a fluid-filled pseudocapsule in cases of suspected synovial pseudarthrosis by nuclear medicine study.

Magnetic resonance imaging may be used to evaluate the soft tissues at the nonunion site or the cartilaginous and ligamentous structures of the adjacent joints. Sinograms may be used to image the course of a sinus tract in cases of infected nonunions. Angiography provides anatomic detail regarding the status of vessels as they course through a scarred and deformed limb. This study is unnecessary for most patients who have a femoral nonunion, unless there is concern regarding the viability of the limb.

(Text continues on page 154.)

Table 2 Clinical Management of Subtrochanteric Femoral Nonunions

Subgroup	Most Recent Failed Treatment	Treatment Options	Pearls	Surgical Technique
Uninfected	Intramedullary nail	Plate and screw fixation with intramedullary and extramedullary autogenous iliac crest bone graft	An oblique osteotomy may be necessary in order to improve surface characteristics and to facilitate interfragmentary lag screw fixation Autologous iliac crest bone graft is delivered to the nonunion site via a chest tube placed into the medullary canal, as described by Chapman (42) Consider the use of BMPs (66,67) Proximal femoral locking plates increase construct rigidity	
	Plate and screw fixation	Repeat plate and screw fixation with intramedullary and extramedullary autogenous iliac crest bone graft; interfragmentary lag screw fixation	An oblique osteotomy may be necessary in order to improve surface characteristics and to facilitate interfragmentary lag screw fixation Consider the use of BMPs (66,67)	
Infected	Intramedullary nail	Nail removal and serial debridements followed by plate and screw fixation with intramedullary and extramedullary autogenous iliac crest bone graft	Autologous iliac crest bone graft is delivered to the nonunion site via a chest tube placed into the medullary canal, as described by Chapman (42) Place an intramedullary antibiotic-eluting nail at the time of each debridement	The antibiotic-eluting nail is constructed by using a chest tube as a mold and placing liquid PMMA with antibiotic powder inside the chest tube with a wire as a central core

	Plate and screw fixation	Hardware removal and serial debridements followed by repeat plate and screw fixation with intramedullary and extramedullary autogenous iliac crest bone graft	An oblique osteotomy may be necessary in order to improve surface characteristics and to facilitate interfragmentary lag screw fixation	
Segmental defect	Intramedullary nail	Defect <4 cm: plate and screw fixation with autogenous iliac crest bone graft	Consider acute shortening with immediate bone-to-bone contact with the plan of restoring length later in a staged procedure (trim the nonunion site in order to improve surface characteristics); an oblique osteotomy may be necessary in order to facilitate interfragmentary lag screw fixation Consider the use of BMPs (66,67)	
		Defect >4 cm: Ilizarov bone transport or intercalary bulk allograft over an intramedullary nail	Consider bone transport over an intramedullary nail	Proximal femoral half-pin fixation anterior or posterior to the nail is facilitated by a "miss-a-nail" targeting device
	Plate and screw fixation	Defect <4 cm: repeat plate and screw fixation with autogenous iliac crest bone graft	Consider acute shortening with immediate bone-to-bone contact with the plan of restoring length later in a staged procedure (trim the nonunion site in order to improve surface characteristics); an oblique osteotomy may be necessary in order to facilitate interfragmentary lag screw fixation Consider the use of BMPs (66,67)	
		Defect >4 cm: Ilizarov bone transport or intercalary bulk allograft over an intramedullary nail	Consider bone transport over an intramedullary nail	Proximal femoral half-pin fixation anterior or posterior to the nail is facilitated by a "miss-a-nail" targeting device

Abbreviation: PMMA, polymethylmethacrylate

Table 3 Clinical Management of Diaphyseal Femoral Nonunions

Subgroup	Most Recent Failed Treatment	Treatment Options	Pearls	Surgical Technique
Uninfected	Intramedullary nail	Exchange nailing	The new nail should be 1 mm to 4 mm larger than the nail being removed Custom nails may be needed for patients who have large nails in situ or large femoral medullary canals	
		Consider nail dynamization for axially stable nonunions that are 3 to 4 mo out from surgical treatment		
	Plate and screw fixation	Repeat plate and screw fixation with autogenous iliac crest bone graft	An oblique osteotomy may be necessary in order to facilitate interfragmentary lag screw fixation Consider dual plating Large fragment locking plates increase construct rigidity Consider the use of BMPs (66,67)	
	External fixator	Compression/distraction with the external fixator, if the fixator allows or exchange for an Ilizarov external fixator or external fixator removal with plate and screw fixation with autogenous iliac crest bone graft	An oblique osteotomy may be necessary in order to facilitate interfragmentary lag screw fixation if internal fixation is the method chosen Consider dual plating Consider the use of BMPs (66,67) Large fragment locking plates increase construct rigidity	
Infected	Intramedullary nail	Nail removal and serial debridements via intramedullary reaming followed by exchange nailing	Consider autologous iliac crest bone graft delivered to the nonunion site via a chest tube placed into the medullary canal, as described by Chapman (42) Place an intramedullary antibiotic-eluting nail at the time of each debridement	The antibiotic-eluting nail is constructed by using a chest tube as a mold and placing liquid PMMA with antibiotic powder inside the chest tube with a wire as a central core

	Plate and screw fixation	Hardware removal and serial debridements followed by repeat plate and screw fixation with autogenous iliac crest bone graft or Ilizarov compression–distraction	An oblique osteotomy may be necessary in order to facilitate interfragmentary lag screw fixation Consider dual plating Large fragment locking plates increase construct rigidity	
	External fixator	Serial debridement followed by Ilizarov compression–distraction or bone transport or external fixator removal and serial debridements followed by plate and screw fixation with autogenous iliac crest bone graft		
Segmental defect	Intramedullary nail	Defect <4 cm: Plate and screw fixation with autogenous iliac crest bone graft	Consider dual plating Consider the use of BMPs (66,67) Consider acute shortening with immediate bone-to-bone contact with the plan of restoring length later in a staged procedure (trim the nonunion site in order to improve surface characteristics); an oblique osteotomy may be necessary in order to facilitate interfragmentary lag screw fixation	
		Defect >4 cm: Ilizarov bone transport or intercalary bulk allograft over an intramedullary nail or vascularized fibular graft	Consider bone transport over an intramedullary nail	Nonunions proximal to the midshaft are most commonly treated with bone transport over an antegrade nail (femoral half-pin fixation anterior or posterior to the nail is facilitated by a ''miss-a-nail'' targeting device) Nonunions distal to the midshaft are most commonly treated with bone transport over a retrograde nail
	Plate and screw fixation	Defect <4 cm: Repeat plate and screw fixation with autogenous iliac crest bone graft	Consider dual plating Consider the use of BMPs (66,67) Consider acute shortening with immediate bone-to-bone contact with the plan of restoring length later in a staged procedure (trim the nonunion site in order to improve surface characteristics); an oblique osteotomy may be necessary in order to	

(*Continued*)

Table 3 Clinical Management of Diaphyseal Femoral Nonunions (*Continued*)

Subgroup	Most Recent Failed Treatment	Treatment Options	Pearls	Surgical Technique
			facilitate interfragmentary lag screw fixation	
		Defect >4 cm: Ilizarov bone transport or intercalary bulk allograft over an intramedullary nail or vascularized fibular graft	Consider bone transport over an intramedullary nail	Nonunions proximal to the midshaft are most commonly treated with bone transport over an antegrade nail (femoral half-pin fixation anterior or posterior to the nail is facilitated by a "miss-a-nail" targeting device) Nonunions distal to the midshaft are most commonly treated with bone transport over a retrograde nail
	External fixator	Defect <4 cm: Plate and screw fixation with autogenous iliac crest bone graft or Ilizarov compression–distraction or Ilizarov bone transport	Consider dual plating Consider the use of BMPs (66,67) Consider acute shortening with immediate bone-to-bone contact with the plan of restoring length later in a staged procedure (trim the nonunion site in order to improve surface characteristics); an oblique osteotomy may be necessary in order to	

		facilitate interfragmentary lag screw fixation	
	Defect >4 cm: Ilizarov bone transport or intercalary bulk allograft over an intramedullary nail or vascularized fibular graft	The technique of bone transport over an intramedullary nail is useful for cases with large segmental defects; however, the risk of deep infection is increased in patients who have had previous external fixation (4,68). In general, the risk is highest in patients whose prior external fixation was recently removed and was in situ for an extended period. The risks and benefits of conventional transport versus transport over a nail in patients with prior external fixation must be weighed by the treating surgeon on a case-by-case basis	Nonunions proximal to the midshaft are most commonly treated with bone transport over an antegrade nail (femoral half-pin fixation anterior or posterior to the nail is facilitated by a "miss-a-nail" targeting device) Nonunions distal to the midshaft are most commonly treated with bone transport over a retrograde nail
Periprosthetic	Revision arthroplasty with a long-stem femoral component or plate and screw fixation with autogenous iliac crest bone graft or both revision arthroplasty with a long-stem femoral component and plate and screw fixation with autogenous iliac crest bone graft	Revision arthroplasty is most appropriate for loose prostheses or those readily amenable to revision Consider dual plating Consider the use of BMPs (66,67) Large fragment locking plates increase construct rigidity and allow for unicortical screw placement in areas where the femoral prosthesis occupies the medullary canal Consider the use of strut cortical allograft with cable fixation to augment stability Specialized periprosthetic cable-plate systems may be advantageous in certain cases	

Venous Doppler studies should be performed preoperatively to rule out a deep venous thrombosis in patients with a lower extremity nonunion who have been confined to a wheelchair or bedridden for an extended period.

LABORATORY STUDIES

In addition to routine lab work, the sedimentation rate and C-reactive protein are useful for following the course of infection. In cases of suspected infection, the nonunion site may be aspirated or biopsied and the material sent for a cell count and Gram stain, and cultured for aerobic, anaerobic, fungal, and acid-fast bacillus organisms. In order to encourage the highest yield possible, all antibiotics should be discontinued at least one week prior to aspiration.

CLASSIFICATION

Nonunions of the femoral shaft and distal femur can be classified according to anatomic location, the presence or absence of infection or a segmental defect, the most recent failed surgical treatment method, and nonunion type (4).

Weber and Cech (65) have classified nonunions based on radiographic healing effort and bone quality into two categories:

1. Viable nonunions—those capable of biological activity, and
2. Nonviable nonunions—those incapable of biological activity.

Viable nonunions include hypertrophic nonunions and oligotrophic nonunions. Hypertrophic nonunions possess adequate vascularity and display callus formation. They arise because of inadequate mechanical stability with persistent motion at the fracture surfaces. Oligotrophic nonunions possess an adequate blood supply but little or no callus formation. Oligotrophic nonunions arise secondary to inadequate reduction with displacement at the fracture site.

An atrophic nonunion is the most advanced type of nonviable nonunion. Atrophic nonunions do not display callus formation and a radiolucent gap is observable on plain radiographs. This gap is bridged with fibrous tissue that has no osteogenic capacity. The ends of the bony surfaces are avascular and usually appear partially absorbed and osteopenic.

Anatomic location is divided into four regions: subtrochanteric, diaphyseal, supracondylar, and condylar (intra-articular).

TREATMENT OPTIONS AND SURGICAL TECHNIQUES

An overview of treatment options and surgical techniques is given in Tables 2 through 5. In cases of infected nonunion, the initial treatment is aimed at eliminating infection regardless of the anatomic location and most recent failed treatment. After elimination of infection, treatment can then proceed.

SUBTROCHANTERIC FEMORAL NONUNIONS (TABLE 2)

The incidence of subtrochanteric femoral nonunions due to failure of internal fixation (intramedullary nail or plate and screw fixation) has been reported to range from 0% to 12% (69,70). Regardless of the most recent failed treatment, plate and screw fixation (or repeat plate and screw fixation) with bone grafting is the most common technique that we employ (Fig. 1), unless there is a large segmental defect (>4 cm). Repeat plate and screw fixation with autogenous cancellous bone grafting or allografting has been reported to have high union rates in the treatment of subtrochanteric nonunions (5).

Reamed intramedullary nailing with or without bone grafting has also been reported to be successful in subtrochanteric nonunions that are amenable to nailing (5–7). A defect in this region larger than 4 cm may require Ilizarov bone transport or an intercalary bulk allograft over an intramedullary nail to restore bony continuity (71).

(Text continues on page 160.)

Table 4 Clinical Management of Supracondylar Femoral Nonunions

Subgroup	Most Recent Failed Treatment	Treatment Options	Pearls	Surgical Technique
Uninfected	Intramedullary nail	SCONE (28) or plate and screw fixation with autogenous iliac crest bone graft	An oblique osteotomy may be necessary in order to improve surface characteristics and to facilitate interfragmentary lag screw fixation if internal fixation is the method chosen Specialized supracondylar locking plates increase construct rigidity	A retrograde nail facilitates proximal femoral half-pin placement because the nail does not occupy the most proximal portion of the femur
	Plate and screw fixation	Repeat plate and screw fixation with autogenous iliac crest bone graft	Consider dual plating Specialized supracondylar locking plates increase construct rigidity	
	External fixator	Ilizarov compression–distraction or plate and screw fixation with autogenous iliac crest bone graft		
Infected	Intramedullary nail	Nail removal and serial debridements via intramedullary reaming followed by plate and screw fixation with autogenous iliac crest bone graft or Ilizarov compression–distraction or serial debridements followed by bony resection through or proximal to the nonunion and reconstruction with a tumor replacement prosthesis	Consider autologous iliac crest bone graft delivered to the nonunion site via a chest tube placed into the medullary canal, as described by Chapman (42) Place an intramedullary antibiotic-eluting nail at the time of each debridement	The antibiotic-eluting nail is constructed by using a chest tube as a mold and placing liquid PMMA with antibiotic powder inside the chest tube with a wire as a central core
	Plate and screw fixation	Hardware removal and serial debridements followed by repeat plate and screw fixation with autogenous iliac crest bone graft or Ilizarov compression–distraction Serial debridements followed by bony resection through or proximal to the nonunion and reconstruction with a tumor replacement prosthesis	An oblique osteotomy may be necessary in order to improve surface characteristics and to facilitate interfragmentary lag screw fixation if internal fixation is the method chosen Consider dual plating	
	External fixator	Serial debridements followed by Ilizarov compression/distraction or bone transport or external fixator removal and serial debridements followed by plate and screw fixation with autogenous iliac crest bone graft or external		

(*Continued*)

Table 4 Clinical Management of Supracondylar Femoral Nonunions (*Continued*)

Subgroup	Most Recent Failed Treatment	Treatment Options	Pearls	Surgical Technique
		fixator removal and serial debridements followed by bony resection through or proximal to the nonunion site and reconstruction with a tumor replacement prosthesis		
Segmental defect	Intramedullary nail	Defect <4 cm: plate and screw fixation with autogenous iliac crest bone graft or bony resection through or proximal to the nonunion site and reconstruction with a tumor replacement prosthesis	Consider acute shortening with immediate bone-to-bone contact with the plan of restoring length later in a staged procedure	
		Defect >4 cm: Ilizarov bone transport or bony resection through or proximal to the nonunion site and reconstruction with a tumor replacement prosthesis or intercalary bulk allograft over an intramedullary nail	Consider bone transport over a retrograde intramedullary nail	
	Plate and screw fixation	Defect <4 cm: repeat plate and screw fixation with autogenous iliac crest bone graft or bony resection through or proximal to the nonunion site and reconstruction with a tumor replacement prosthesis	Consider acute shortening with immediate bone-to-bone contact with the plan of restoring length later in a staged procedure	
		Defect >4 cm: Ilizarov bone transport or bony resection through or proximal to the nonunion site and reconstruction with a tumor replacement prosthesis or intercalary bulk allograft over an intramedullary nail	Consider bone transport over a retrograde intramedullary nail	A retrograde nail facilitates proximal femoral half-pin placement because the nail does not occupy the most proximal portion of the femur

	External fixator	Defect <4 cm: plate and screw fixation with autogenous iliac crest bone graft or Ilizarov compression–distraction or Ilizarov bone transport or bony resection through or proximal to the nonunion site and reconstruction with a tumor replacement prosthesis Defect >4 cm: Ilizarov bone transport or bony resection through or proximal to the nonunion site and reconstruction with a tumor replacement prosthesis or intercalary bulk allograft over an intramedullary nail	Consider acute shortening with immediate bone-to-bone contact with the plan of restoring length later in a staged procedure
Periprosthetic		Revision arthroplasty with a long-stem femoral component or plate and screw fixation with autogenous iliac crest bone graft or retrograde femoral nailing with autogenous iliac crest bone graft or both revision arthroplasty and fixation with plate and screw fixation or retrograde intramedullary nail fixation with autogenous iliac crest bone graft	Revision arthroplasty is most appropriate for loose prostheses or those readily amenable to revision Consider dual plating Consider the use of BMPs (66,67) Large fragment locking plates increase construct rigidity and allow for unicortical screw placement in areas where the femoral prosthesis occupies the medullary canal Consider the use of strut cortical allograft with cable fixation to augment stability Specialized periprosthetic cable-plate systems may be advantageous in certain cases

Table 5 Clinical Management of Condylar (Intra-articular) Femoral Nonunions

Subgroup	Most Recent Failed Treatment	Treatment Options	Technique Pearls
Uninfected	Intramedullary nail	Interfragmentary lag screw fixation or knee replacement arthroplasty	Knee replacement arthroplasty is most appropriate in older adults or those whose nonunions have failed to unite despite multiple surgical attempts or those with significant arthritis
	Plate and screw fixation	Repeat plate and screw fixation or knee replacement arthroplasty	Knee replacement arthroplasty is most appropriate in older adults or those whose nonunions have failed to unite despite multiple surgical attempts or those with significant arthritis
Infected	Intramedullary nail	Serial debridements followed by knee replacement arthroplasty, osteoarticular allograft reconstruction, or knee arthrodesis	An antibiotic-eluting spacer may be useful following debridement but prior to reconstruction
	Plate and screw fixation	Serial debridements followed by knee replacement arthroplasty, osteoarticular allograft reconstruction, or knee arthrodesis	An antibiotic-eluting spacer may be useful following debridement but prior to reconstruction

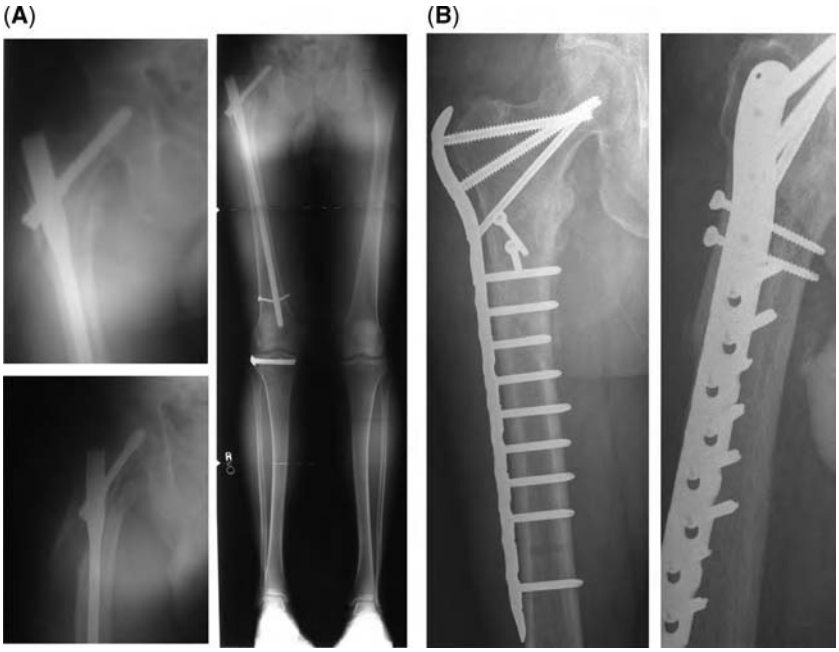

Figure 1 (**A**) Presenting anteroposterior (AP) and lateral radiographs of a 51-year-old man referred in four months following intramedullary nail fixation of a reverse obliquity intertrochanteric/subtrochanteric femur fracture. The lateral view shows poor bone-to-bone contact with no evidence of progression to healing whatsoever. This patient complained of increasing pain and a sensation of abnormal motion in the thigh. (**B**) Follow-up AP and lateral radiographs five months following reconstruction with open reduction, interfragmentary lag screw fixation, fixation with a proximal femoral locking plate (Synthese, Paoli, Pennsylvania), and intramedullary and extramedullary autogenous bone grafting. At follow-up, the nonunion site is solidly healed and the patient has returned to preinjury function without any symptoms.

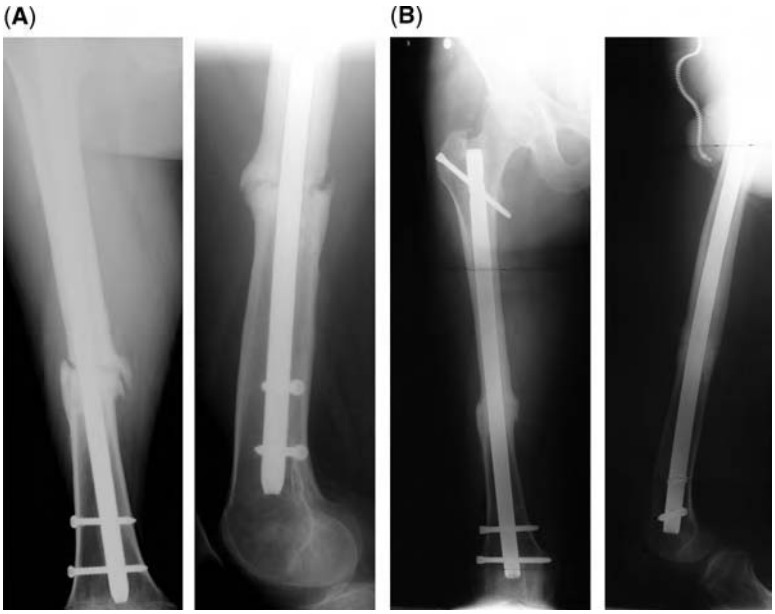

Figure 2 (**A**) Presenting anteroposterior (AP) and lateral radiographs of a 33-year-old man referred in for a diaphyseal nonunion 11 months following intramedullary nail fixation. (**B**) Follow-up AP and lateral radiographs seven months following exchange nailing show solid bony union.

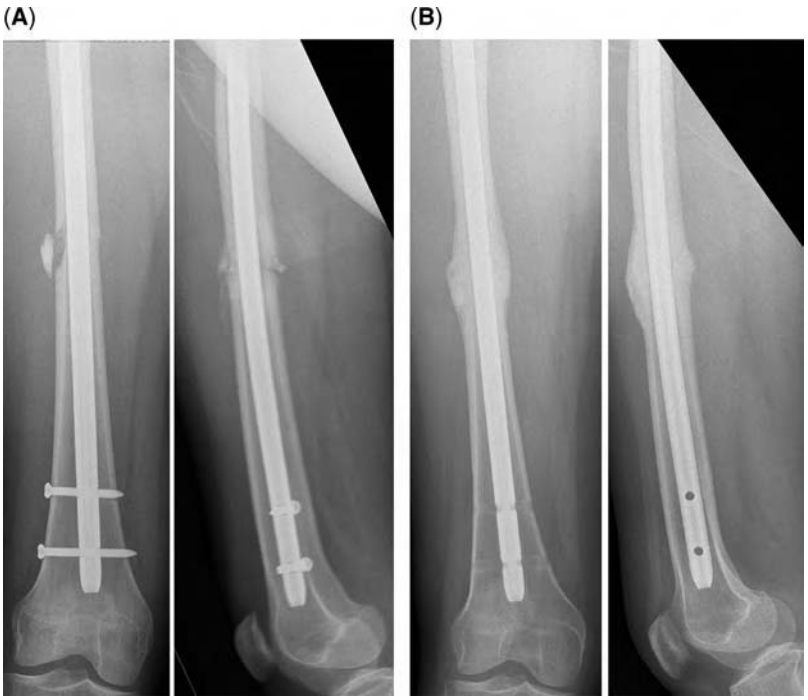

Figure 3 (**A**) Presenting anteroposterior (AP) and lateral radiographs of a 17-year-old girl with cerebral palsy referred in 3.5 months following intramedullary nail fixation of a femoral shaft fracture. The patient complained of progressively worsening pain in the right thigh. (**B**) Follow-up AP and lateral radiographs five months following nail dynamization show solid union. At follow-up, the patient is asymptomatic and has returned to preinjury functional status.

DIAPHYSEAL FEMORAL NONUNIONS (TABLE 3)

The incidence of diaphyseal femoral nonunions following intramedullary nailing of an acute fracture has been reported to range from 2% to 13%; the highest nonunion rates are associated with unreamed nailing (15,72).

Patients with diaphyseal femoral nonunions who have failed intramedullary nailing generally receive exchange nailing (Fig. 2) or nail dynamization when they are three to four months out from surgical treatment and the nonunion is axially stable (Fig. 3) (7,12,15–23,25,26,29,31). Patients who have failed one or more exchange nailings may require slow compression over a nail using external fixation (SCONE) (Fig. 4) (28). Augmentative plating over the nail has also been reported to be successful, although this technique requires exposure of the nonunion site and has a slightly longer healing period (13,14). Alternatively, the nail can be removed and the nonunion site can be stabilized using plate and screw fixation (8–12). Autogenous bone grafting around a previously placed nail without providing augmentative stabilization has had mixed results and we do not recommend this technique (10,15). Infected diaphyseal nonunions may require external fixation after debridement to provide stability or address segmental defects (12,33,34).

Failed plate and screw fixation usually responds well to repeat plate and screw fixation with bone grafting (9,12), but may require treatment with Ilizarov external fixation if infection or a segmental defect is present (73). Reamed intramedullary nailing with or without bone grafting following failed plate and screw fixation has been reported to have a very high success rate (7,9,10,17,18,21,23,29–32); this is not a strategy that we often employ.

Patients with failed external fixation may require plate and screw fixation and bone grafting after removal of the fixator, a change in the treatment mode of the fixator (for example, from static to compression–distraction), or conversion to Ilizarov external fixation (compression–distraction or bone transport).

In the event of a segmental defect larger than 4 cm, Ilizarov bone transport may be the method of choice regardless of the most recent failed treatment (39,43). Vascularized fibular grafts, either single or double, can also be successful in the treatment of large segmental defects, but are associated with vascular complications and are at significant risk for subsequent fracture (36–40). Intercalary allograft over an intramedullary nail is also a useful treatment option (4,44).

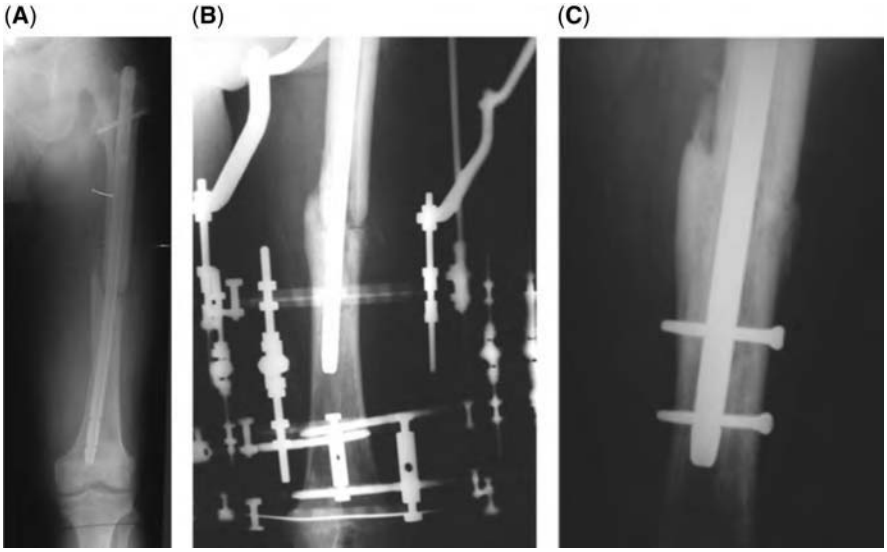

Figure 4 (**A**) Presenting anteroposterior (AP) radiograph of a 67-year-old man referred in for a nonunion 30 months following an open femoral shaft fracture and seven previous surgeries, including two previous exchange nailings and bone grafting. (**B**) AP radiograph on postoperative day 75 showing slow compression over a nail using external fixation (SCONE). (**C**) Follow-up AP radiograph four months following SCONE shows solid bony union.

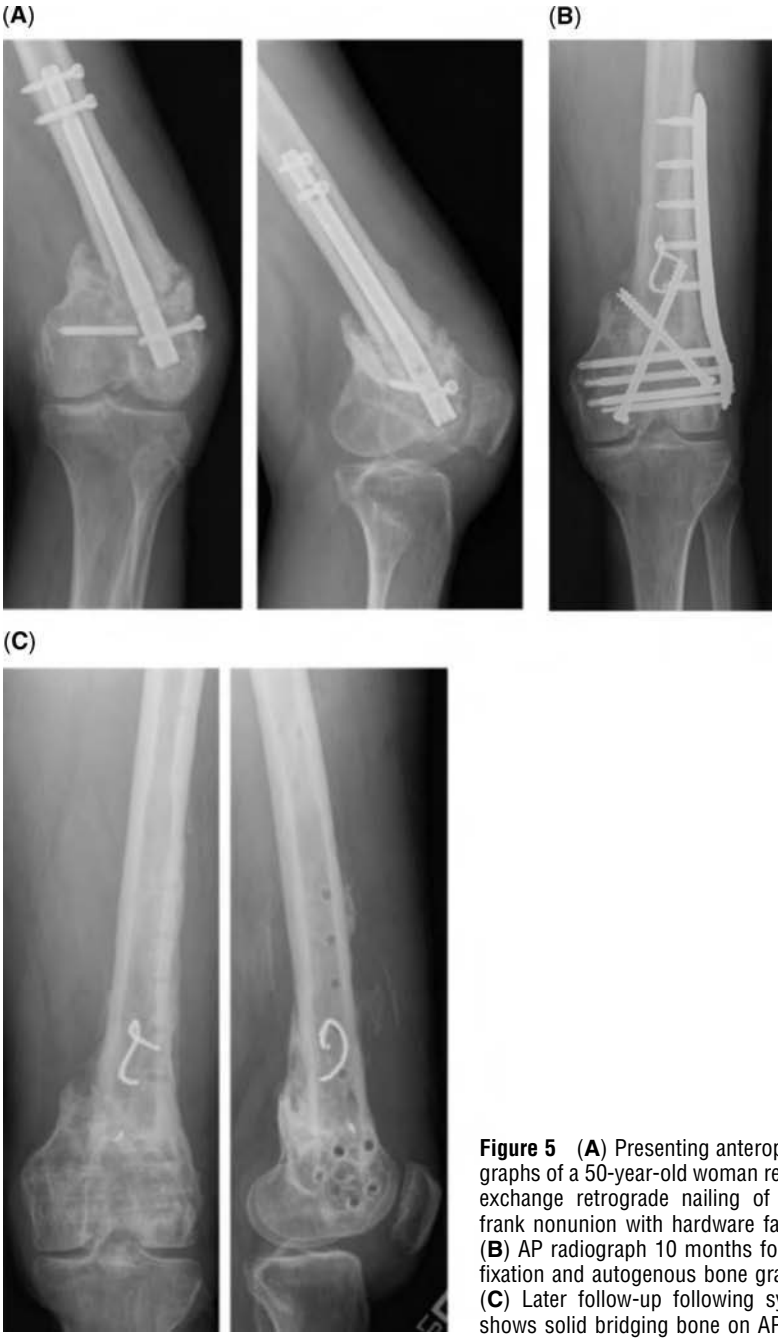

Figure 5 (**A**) Presenting anteroposterior (AP) and lateral radiographs of a 50-year-old woman referred in nine months following exchange retrograde nailing of a supracondylar nonunion. A frank nonunion with hardware failure and deformity is present. (**B**) AP radiograph 10 months following open reduction internal fixation and autogenous bone grafting reveals solid bony union. (**C**) Later follow-up following symptomatic hardware removal shows solid bridging bone on AP and lateral views.

Periprosthetic diaphyseal nonunions may require revision arthroplasty, plate and screw fixation with bone grafting, or both.

SUPRACONDYLAR FEMORAL NONUNIONS (TABLE 4)

The incidence of supracondylar femoral nonunions has been reported to range from 3% to 6% when treated with a supracondylar intramedullary nailing system (74–76), and from 0% to 13% with plate and screw fixation (77–80). Intramedullary nailing following failure of plate and screw fixation is a poor treatment option for supracondylar nonunions. Koval et al.

reported a 75% failure rate for distal femoral nonunions treated with retrograde intramedullary nailing (48). In contrast, Kempf et al. reported good success using a dynamically locked antegrade intramedullary nail in five cases (7).

Supracondylar nonunions that have most recently failed intramedullary nailing are most commonly treated at our institution by SCONE (28). Other options include Ilizarov compression–distraction, external fixation (49), and plate and screw fixation with bone graft (Fig. 5) (11,46,47). Older patients or patients with limited physical demands may benefit from total knee arthroplasty using a long femoral stem, a megaprosthesis, a tumor replacement prosthesis, or an allograft–prosthesis composite (50,51,81).

Patients who have most recently failed external fixation are treated by Ilizarov compression–distraction or bone transport, or plate and screw fixation with bone grafting. Ilizarov bone transport or intercalary bulk allograft over an intramedullary nail are used to treat segmental defects larger than 4 cm. Cases of infected supracondylar nonunion or nonunion associated with a large segmental defect may require bony resection proximal to the nonunion site and reconstruction using a tumor prosthesis.

Periprosthetic supracondylar nonunions may be treated by revision arthroplasty with a long-stem femoral component, plate and screw fixation or retrograde femoral nailing with autogenous iliac crest bone graft, or a combination of these techniques.

CONDYLAR (INTRA-ARTICULAR) FEMORAL NONUNIONS (TABLE 5)

The incidence of condylar femoral nonunions has been reported to range from 1% to 13% (82,83). These cases may be successfully treated to union with interfragmentary lag screw fixation (Fig. 6). In older adults or patients who have failed multiple surgical attempts, or those with significant arthritis, knee replacement arthroplasty may be appropriate. In cases with infection, an osteoarticular allograft or knee arthrodesis may be alternatives to knee replacement arthroplasty. Following serial debridement, an antibiotic spacer may be used to decrease the chance of reinfection following the later reconstruction.

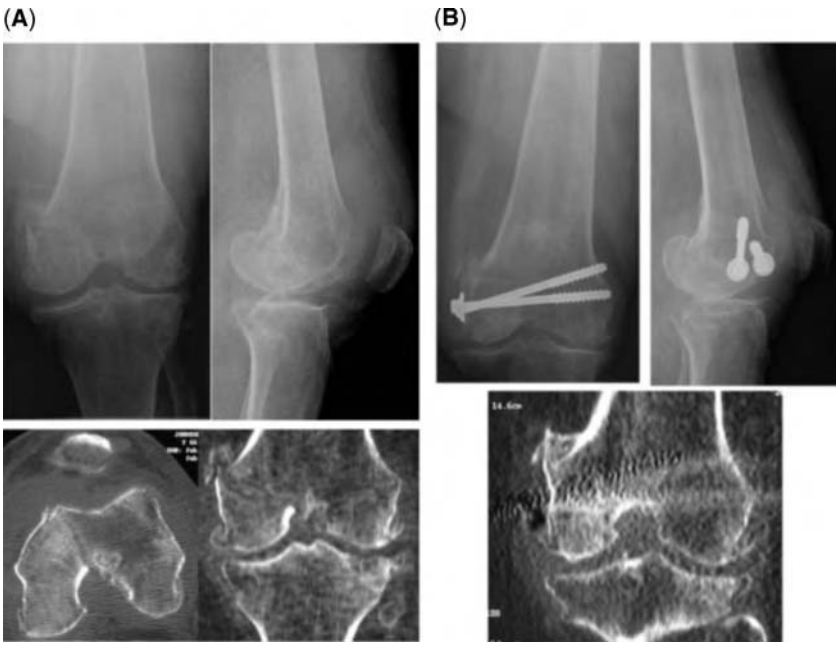

Figure 6 (**A**) Presenting anteroposterior (AP) and lateral radiographs and computed tomographic (CT) scan of a 81-year-old woman referred in 4.5 months following a fall at home. Radiographs and CT scan reveal a nonunion of the medial femoral condyle. (**B**) Follow-up AP and lateral radiographs and CT scan three months following interfragmentary lag screw fixation show solid bony union.

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12 Tibial Malunions

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TREATMENT OF TIBIAL MALUNIONS: CHARACTERIZATION BY A GRAPHICAL METHOD

INTRODUCTION

If ever an ounce of prevention was worth a pound of cure, it would be in achieving and maintaining accurate fracture reduction. Remembering the trauma of the original fracture, there is usually a significant problem with pain, function, or cosmesis before a patient would present for consideration of malunion correction.

Whereas the need to treat a tibial nonunion is usually clear, the decisions of whether to divide the tibia and how to stabilize the fragments in the name of improving the anatomic elements of alignment, length, and rotation are usually more difficult. Historical guidelines suggest 5° in the coronal plane and 10° in the sagittal plane as upper limits of malalignment. Some mild deformities seem to be better tolerated than others. External rotation of the tibia does not pose the same immediate problem with ambulation as internal rotation. The increased varus play in the subtalar joint can partially ameliorate a supramalleolar valgus deformity. Although as surgeons we strive to reduce and maintain fractures within these limits, there must be very careful consideration before surgically correcting an established malunion at these threshold values. The symptoms, age, activity level, general medical condition of the patient, and condition of the tibia must be considered. In some cases, protracted healing as a result of multiple comorbidities led to the original malunion. Serious initial problems with soft-tissue coverage and/or infection may have also contributed to the malunion by limiting the type and zone of fixation. What will be different the second time around after the fracture is recreated? The best healing time in adults is 3 to 4.5 months. Are the benefits of improved alignment worth this second period of healing following general anesthetic?

COMPENSATED DEFORMITIES

In the nondisplaced or anatomic position, the mechanical axis passes through the center of the ankle (Fig. 1A). In Figs. 1B and C, the distal fragment is in 20° varus. In Fig. 1B, the mechanical axis still passes through the center of the ankle and is compensated. Although the angular deformity in Fig. 1C is the same, it will be more cosmetically apparent and will result in mechanical axis deviation.

THREE-DIMENSIONAL DEFORMITY

The French geometer Charles Chasles was the first to show that one object can be moved to any ''deformed position'' with respect to a reference object by a rotation about a doubly oblique axis. As surgeons, we tend to split this true three-dimensional rotation into the three orthogonal components and talk about a threshold in each. Probably the patient's symptoms are proportional to the true three-dimensional rotation. The equation is expressed as follows where s equals the three-dimensional angle.

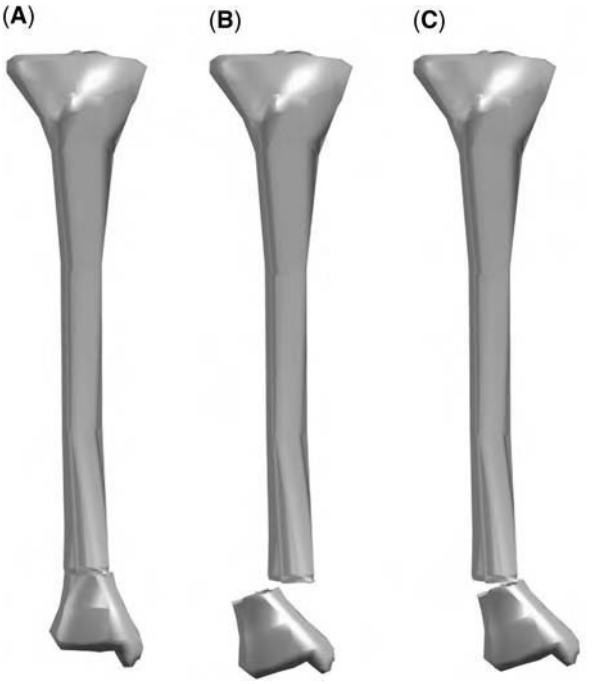

Figure 1 (**A**) Normal tibia in which the mechanical axis passes through the center of knee and ankle. (**B**) Compensated angular deformity in which the mechanical axis still passes through the center of knee and ankle. (**C**) Uncompensated deformity is more apparent clinically and will result in mechanical axis deviation at the knee.

HIDDEN CONTRACTURES

Long-standing malunions may have associated soft-tissue contracture or even bony block preventing normal range of motion of the ankle or knee. A full extension lateral X ray of the knee and a full dorsiflexion lateral of the ankle should be part of the preoperative evaluation. An equines contracture of the ankle is often associated with a recurvatum deformity of the distal tibia. It may be necessary to treat the equines contracture with a hinged frame from distal tibia to foot with or without tendoachilles lengthening. Similarly, a recurvatum deformity with a flexion contracture of the knee might be addressed by less than full correction of the usual 10° posterior slope of the plateau.

NAIL, PLATE, AND EXTERNAL FIXATION

Intramedullary (IM) nails tend to be self-proving for diaphyseal fractures and malunions, in that simply inserting a nail that practically fills the canal automatically realigns the fragments; however, it does not guarantee correct rotation and cannot address significant length discrepancy. In fact, malunions are seen with IM nailing of relatively proximal and distal fractures that are technically more demanding and not self-proving. Recent reports site significant numbers of rotational malunions with nailing of even diaphyseal fractures. The role of IM nailing in the treatment of malunions should be confined to diaphyseal deformity in which there is no significant length discrepancy, no history of osteomyelitis, and a sufficient or restorable canal. The surgeon must be able to accurately assess rotation intraoperatively. Some patients have postoperative and occasionally chronic anterior knee pain following IM nailing of the tibia.

Plates may be used for malunions at all levels of the tibia. Like IM nailing all correction is achieved in surgery. Sliding oblique osteotomies fixed with plates can correct some length in addition to correcting alignment, but there are limitations to how much length can be acutely and safely gained at the expense of fragment apposition. The soft tissues and bone must be able to tolerate the exposure necessary for precise osteotomy and plate fixation. The surgeon must be able to assess the mechanical axis as well as the rotation in surgery. A history of prior infection is a relative contraindication to plate synthesis.

External fixation has proven to be a powerful method for correcting all anatomic elements of a malunion and providing skeletal stability while the osteotomy heals. It is also used as a

reduction tool to achieve a more anatomic realignment, which is then fixed in situ by a plate or IM nail, and the fixator is removed. As thoroughly shown by Ilizarov, an osteotomy stabilized by external fixation and gradually distracted is capable of creating regenerate bone filling in an opening wedge correction of the angular deformity. Also, additional length to correct any inequality in the limbs is achieved by continuing to gradually lengthen through the osteotomy to correct the angular deformity or a second osteotomy in more metaphyseal bone for more extensive lengthening. Present day external fixation allows the surgeon to build an external fixator that exactly mimics the deformity. After the fixator is fixed to the tibia and the osteotomy performed, the fixator is gradually adjusted to a home position, thereby correcting the malunion. Since 2002, the total residual method has been possible with the spatial frame allowing the surgeon to fix the rings of the frame to the skeleton first and create an accurate adjustment schedule for the malunion even after surgery. The Spatial Frame is a six-axis manipulator and is able to address all components of a deformity including rotation with essentially the same frame. The goals in surgery for correction of malunions with external fixators is different from IM nail or plate fixation. The surgeon has only to provide stable fixation of each bone segment and perform an osteotomy. The gradual highly accurate reduction is achieved after surgery. Further adjustments can be made for weeks until the patient and surgeon are satisfied. External fixation is relatively indicated in cases of prior infection or with poor soft-tissue coverage.

Because of the simpler surgical goals, applicability to the entire shaft of the tibia, use in cases with prior infection, late adjustability, extreme accuracy, and high success rate, the external fixator is a powerful tool for the treatment of malunions.

The remainder of this chapter will address the topics of characterizing the skeletal deformity, determining the level of osteotomy, and locating the external fixator.

Overcorrection/Undercorrection

In the case of varus deformity of the proximal tibia in conjunction with arthritis of the medial compartment of the knee, it is advisable to slightly overcorrect the skeletal deformity to have the mechanical axis passing lateral to the center of the knee up to the Fujisawa point for bone on bone arthritis, or slightly short of the Fujisawa point for articular cartilage thinning.

DEFINITIONS AND MEASUREMENTS

The opposite normal side is a convenient standard when characterizing tibial malunion. Good standard measurements have been published by Paley and Herzenberg as well as others. The mechanical axis in the sagittal plane passes from the center of the femoral head, through the center of the knee joint, and on through the center of the ankle. In the coronal plane, the mechanical axis passes through the junction of the anterior one-fifth of the joint in the knee and the center of the ankle. The knee slopes 3° on anteroposterior (AP) view, the ankle is essentially 90° on AP view. The knee slopes 9° posterior on lateral view. The ankle opens 10° anteriorly on lateral view.

Compared to congenital deformities, more malunions have additional translation in a plane perpendicular to the oblique plane of angulation, and thus require more than just an angular correction to reestablish the mechanical axis.

The information needed to completely characterize a skeletal deformity is present in orthogonal radiographs, AP and lateral, and in an assessment of rotation, which may be clinical or based on computed tomographic landmarks. From a practical standpoint, the amount of additional length needed during skeletal correction will depend upon the cross-sectional shape of the bone at the level of osteotomy and conceivably extrinsic information such as scanograms or growth charts. Most external fixation systems have a means of lengthening along the axis of the fixator, although most do not have a convenient means of gradually derotating the deformed fragment about its mechanical or anatomic axis.

THE IMPORTANCE OF THE REFERENCE FRAGMENT

If both radiographs are taken orthogonal to the reference fragment and the magnification is taken into account, then the axis of the reference fragment is like a scale that can be used to measure the position of the deformed fragment. The only foreshortening is in the deformed fragment as expected. The mechanical axis of a short periarticular fragment is usually most accurately represented as a line with a given relation to the joint line, such as a 90° line with

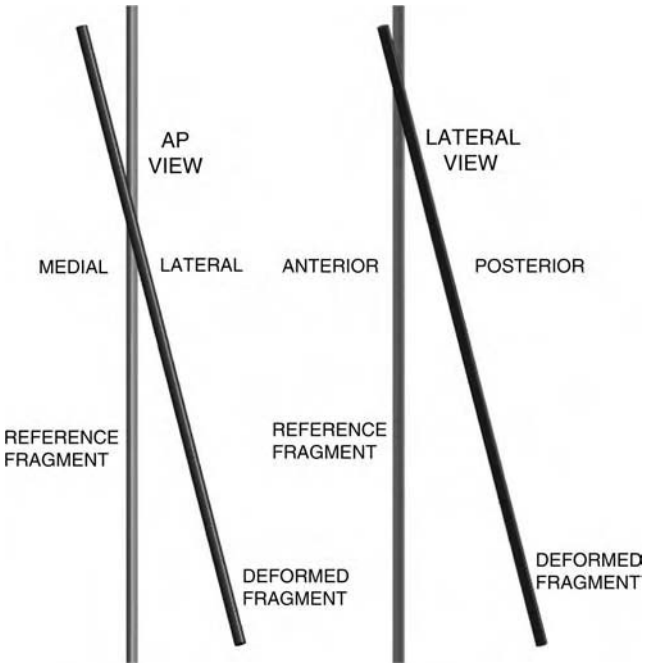

Figure 2 On anteroposterior and lateral radiographs or tracings made to the same scale, extend the centerlines or mechanical axis of each fragment and label the anterior, posterior, medial, and lateral directions. This is the first step to creating an axial view and locating the line of closest approach.

respect to the AP ankle line or an 87° line with respect to the AP knee line. Because it is difficult to draw the joint lines accurately in oblique views, the short periarticular fragment often is the best choice for reference fragment. Also, the X-ray technician can usually do a better job aligning to the patella for distal femoral or proximal tibial deformities and the foot for distal tibial deformities and the elbow or hand for upper extremity deformities.

SYNTHESIS OF THE AXIAL VIEW

Like a mechanical drawing where the information on the *front* and *side* view can be used to create a *top* view, the *AP* and *lateral* projections contain the information to create an *axial* view. On AP and lateral radiographs, draw extended mechanical axes (Fig. 2).

On a piece of graph paper begin the axial view by placing a small circle at the intersection of the sagittal and coronal planes. This is the reference fragment. Label anterior, posterior, medial, and lateral (Fig. 3).

The position of the deformed fragment mechanical axis relative to the mechanical axis of the reference fragment must be measured at two levels 15 to 20 cm apart. Level 1 might be the

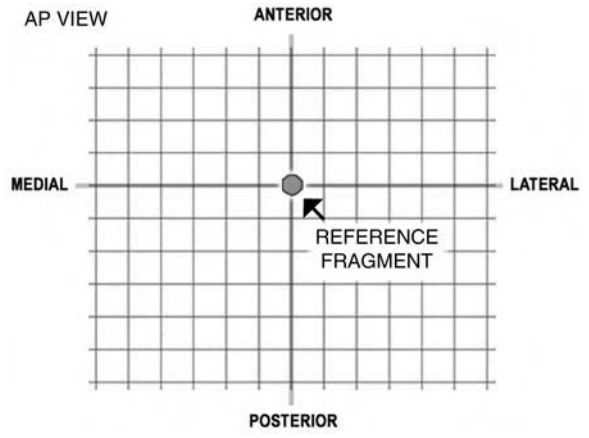

Figure 3 Creating an axial view of the reference fragment with directions labeled.

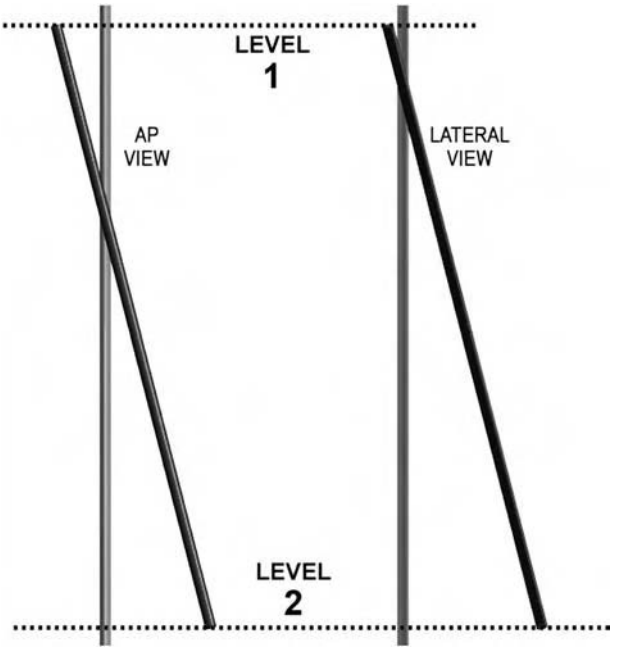

Figure 4 Establish two levels at least 20 cm apart at which to measure the relative position of the deformed fragment centerline to the reference fragment centerline.

level of the knee joint or other good landmark easily discerned on AP and lateral views. Level 2 is located on both AP and lateral views by measuring the same distance from Level 1 (Fig. 4).

At Level 1 on AP and lateral measure the position of the deformed axis with respect to the reference axis. Represent this as a small circle, point 1, on the axial view (Fig. 5).

At Level 2 (approximately 20 cm away) on AP and lateral measure the position of the deformed axis with respect to the reference axis and represent this as another small circle, point 2, on the axial view (Fig. 6).

Connect the two circles corresponding to the deformed axis at Level 1 and Level 2. This new line is the axial view of the deformed fragment mechanical axis between Level 1 and Level 2 (Fig. 7).

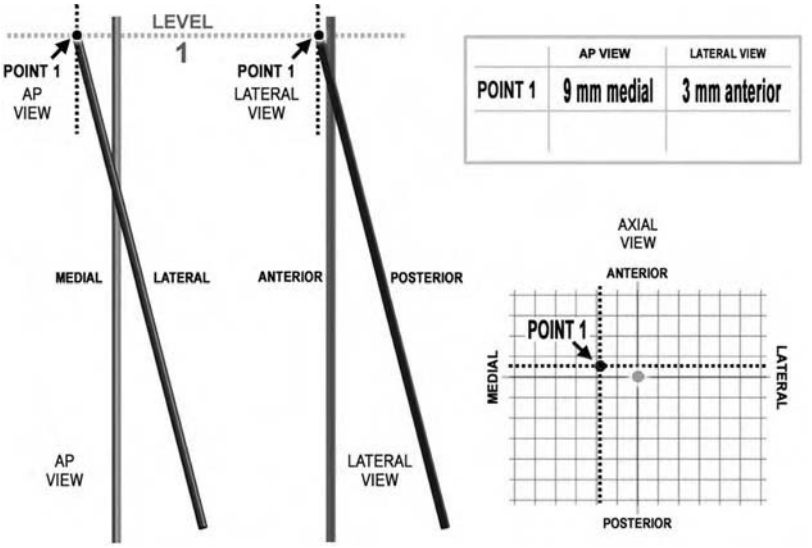

Figure 5 Measure the position of the deformed fragment centerline at Level 1 on anteroposterior and lateral views. Place this point 1 on the axial view.

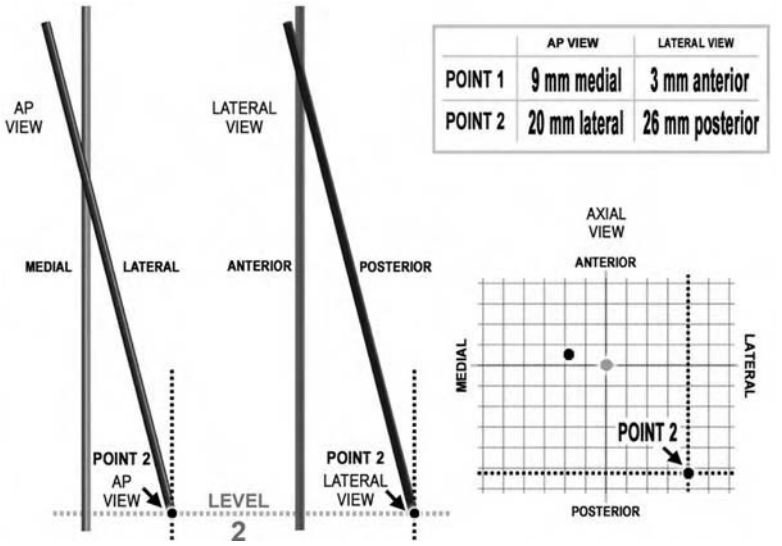

Figure 6 Measure the position of the deformed fragment centerline at Level 1 on anteroposterior and lateral views. Place this point 2 on the axial view.

LINE OF CLOSEST APPROACH—THE KEY TO CHARACTERIZING AND CORRECTING DEFORMITY

Given two lines, one arbitrarily oblique to a reference line, analytic geometry shows that there is only one line that is perpendicular to both. This line is also the shortest distance between the two lines and is the line of closest approach or line of closest approach (LOCA).

On the axial view, draw a line that is perpendicular to the deformed axis and passes through the reference axis. This is the LOCA (Fig. 8).

An external fixator hinge placed collinear with the LOCA will correct the angular deformity without creating any additional translation. The axial view gives the orientation of the LOCA in the transverse plane, but it must also be located at the correct level. To determine the correct level for the LOCA, look at the intersection of the LOCA with the mechanical axis of the deformed axis, point 3, as seen on the axial view. Measure the amount of medial/

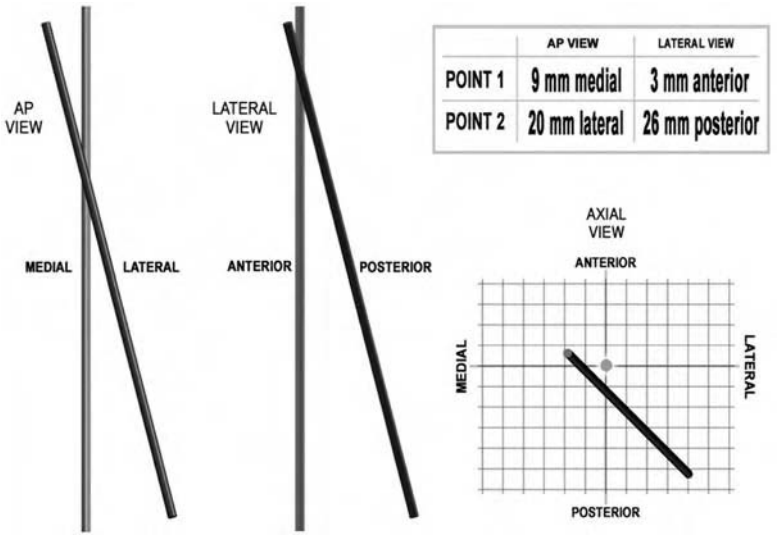

Figure 7 Connect point 1 and point 2 with a line. This line is the axial view of the deformed fragment between Level 1 and Level 2.

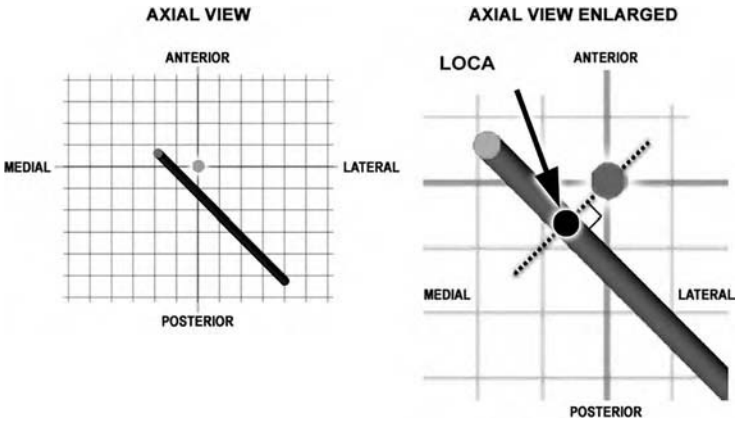

Figure 8 Draw the line perpendicular to the deformed axis and passing through the reference fragment. This line is the line of closest approach, line of closest approach, for the two fragment centerlines.

lateral translation of point 3 with respect to the reference fragment and find the level on the AP view where the deformed fragment has this same translational relation. This level is the level of the LOCA. Likewise, the anterior/posterior translation of this same point and the lateral view can be used to identify this same level of the LOCA (Fig. 9).

MEASURING TRUE ANGULAR DEFORMITY

If the length of the line between point 1 and point 2 is taken as the base of a right triangle and the distance between Level 1 and Level 2 is taken as the height of the right triangle, then the inscribed angle, delta, is the true magnitude of the oblique plane angular deformity (Fig. 10).

DETERMINING TRANSLATION PERPENDICULAR TO THE PLANE OF ANGULAR DEFORMITY

The LOCA by definition is the shortest distance between the reference fragment and deformed fragment. The length of the LOCA as seen on the axial view is the distance the deformed fragment

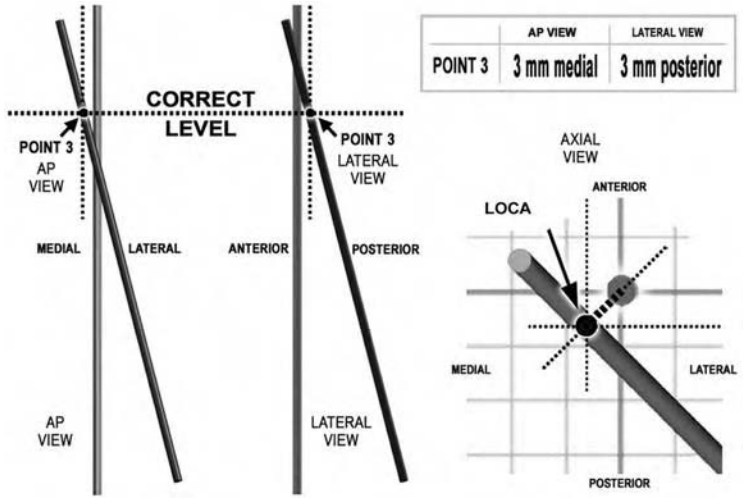

Figure 9 On the axial view measure the medial–lateral position of the point of intersection of the line of closest approach (LOCA) with the deformed fragment centerline. Find the level on the original AP drawing where the deformed fragment centerline has this same medial–lateral position relative to the reference fragment centerline. This Level 3 is the real level of the LOCA.

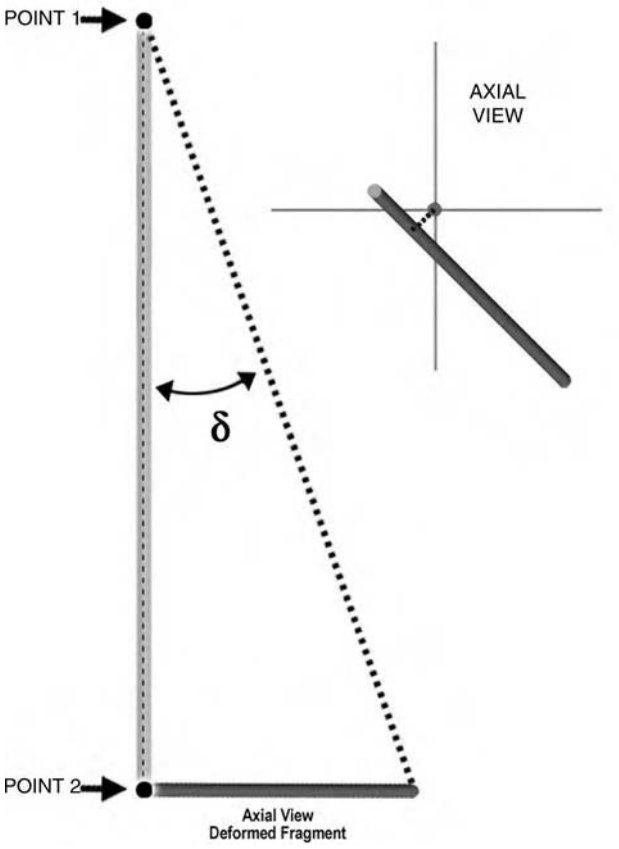

Figure 10 Draw a triangle with a base equal to the length of the line from point 1 to point 2 on the axial view. The height of the triangle is equal to the distance from Level 1 to Level 2. The inscribed angle equals the true magnitude of the oblique plane angular deformity.

needs to be translated in a plane perpendicular to the plane of angulation. In the Ilizarov system, this is the setting for the translational blocks or the offset in the translational hinge (Fig. 11).

Unless there are other considerations, such as skin and bone condition or fragment size for sufficient fixation, the LOCA is usually the best level for osteotomy. Osteotomy at the level of the LOCA will result in the least possible offset after correction of the translational deformity. Gigli saw osteotomy in the metaphysis and multiple drill hole osteotomy for the diaphysis have both been used.

PUTTING IT ALL TOGETHER

Discussion

The preceding graphical method generates an axial view and solves for the LOCA. The axial view yields the plane of angulation, the length of the LOCA, and can be used to plan fixator hinge orientation (Fig. 12).

A hinged fixator placed collinear to the LOCA can correct oblique plane angulation without introducing translation in that oblique plane. The length of LOCA is the amount of translation in a plane perpendicular to the plane of angulation that the deformed fragment must be moved to bring it collinear to the reference fragment. Also, the true magnitude of the angular deformity is determined. In many instances, the level of the LOCA represents the best choice for the level of osteotomy. This method is efficient, only an AP and lateral radiograph are required, and precise.

The six deformity parameters used to control the spatial frame may be measured at any level such as Level 1, 2, or 3 in the preceding example. It is unnecessary to determine the orientation of the hinge axis, the true magnitude of angulation, or the magnitude of translation perpendicular to the plane of angulation. The computer program basically establishes a virtual hinge axis and virtual translation device that completely corrects the deformity. It is still advisable to determine the level of the LOCA when using the spatial frame on malunions to plan

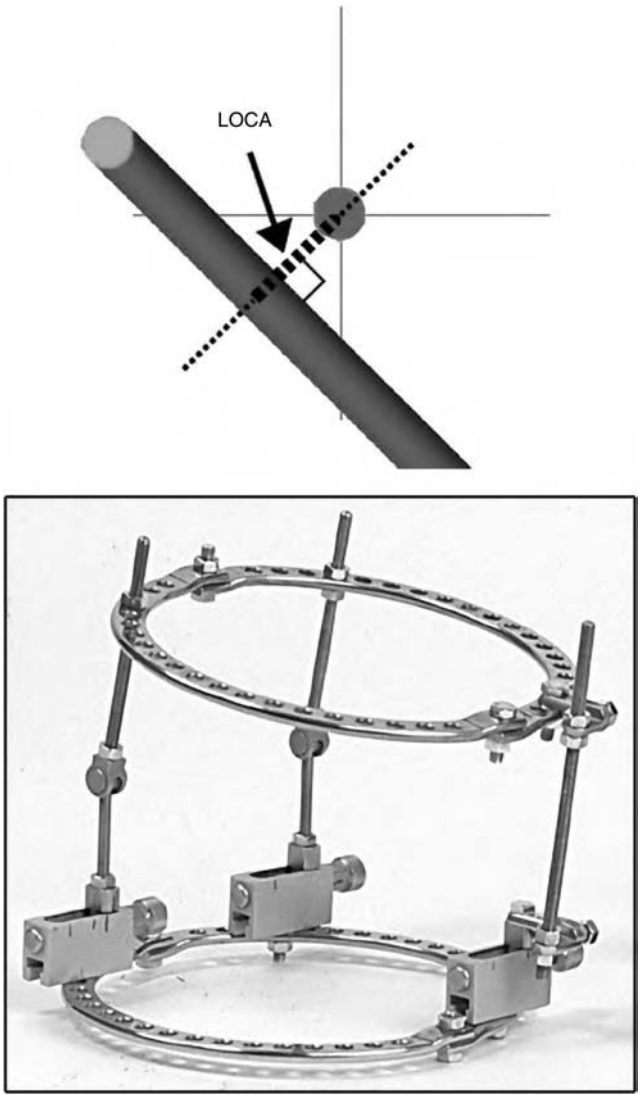

Figure 11 The length of line of closest approach (LOCA) is the distance the deformed fragment must be translated to bring the fragments coplanar. The LOCA is perpendicular to the plane of deformity. In the Ilizarov system, this may be accomplished with multiple translation blocks.

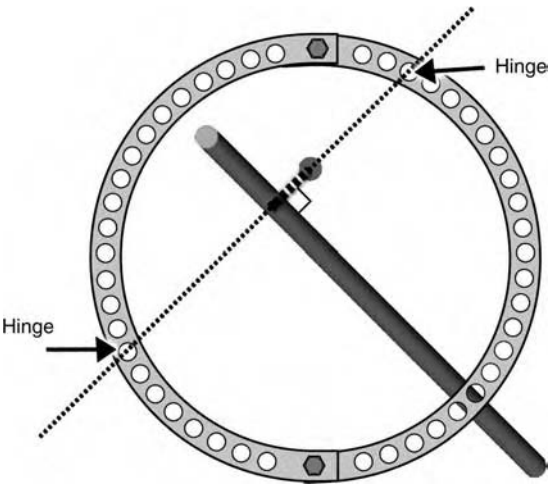

Figure 12 To correct a complex deformity, the hinge axis of the fixator is made collinear with the line of closest approach (LOCA) and an angular correction equal to the true magnitude of the angle between the fragments is performed. The deformed fragment is translated along the hinge axis by an amount equal to the length of LOCA.

the osteotomy. LOCA type analysis is also required for treatment of nonunions with conventional fixators, but is not necessary for Spatial Frame cases because no osteotomy is required.

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13 Post-Traumatic Reconstruction: Femoral Malunion

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INTRODUCTION

Femoral malunion may result in suboptimal function and altered joint loading through various combinations of angulation/translation, shortening, and rotational malalignment (1,2). The most important consideration is the long-term effect of malalignment and altered joint mechanics, resulting in abnormal joint loading and leading to premature osteoarthritis (1–4). Malunion may result in symptoms when angular deformities exceed 5° in the coronal plane or 10° in the sagittal plane, when rotational deformities exceed 10°, and when shortening exceeds 2 cm. It is critical to understand that absolute values of the angular magnitude alone cannot be considered the single most important consideration (1,2,5). Associated translation and the level of the deformity are other crucial aspects that must be considered. Those deformities with the apex near the knee have the greatest effect on the mechanical axis (1,2,5,6), whereas deformities more proximal in the diaphysis may have far less impact. Isolated rotational deformities are still prevalent even in the modern era of intramedullary (IM) fixation (7–10). Length discrepancy is still not an uncommon deformity, though certainly less frequent than when femoral fractures were managed nonoperatively.

There are many options available to manage femoral malunions, and various strategies are still evolving. Internal fixation with plates (3,11–13) or intramedullary rods (14) can be useful for acute corrections, particularly when length is not a significant element of the deformity. In the past decade, telescoping (lengthening) rods have been developed to address length using a totally implanted device. Alternatively, lengthening can be achieved gradually using a temporary external fixator combined with an intramedullary (IM) nail (15). However, if infection has been recently active it is generally considered prudent to avoid permanent implants. External fixation still has the broadest indications, but is the most inconvenient device for patients and surgeons (5,6,16,17). External fixation can be used for either acute or gradual correction, and is most useful when lengthening is indicated. Decisions regarding the most suitable device for a particular patient are based on features identified during the preoperative clinical evaluation.

CLINICAL EVALUATION

A thorough history and careful physical examination are more important considerations than arbitrary radiographic values when advising a patient as to the potential benefit of a corrective osteotomy. Within the history, it is important to identify if there would be an increased risk of infection if implants were introduced. The history should also identify prior nerve or vascular injuries, an indication for gradual rather than acute correction.

The physical examination should focus on these same issues, while assessing further aspects that are not visible radiographically. Rotation, joint contractures, and ligamentous instability are specifically assessed. Soft-tissue contractures are not uncommon, and both hip and knee range of motion should be examined carefully. When knee range of motion is less than 90°, consideration should be given to performing a Judet quadricepsplasty in conjunction with a corrective osteotomy. Hypersensitivity, paresthesias, or a positive Tinel's sign can help identify the patient at risk of developing neurologic compromise after acute correction. Vascular compromise is less common but should be considered, particularly in the patient with a history of a prior open injury. Internal rotation deformities with significant shortening would be considered at greatest risk.

Good-quality radiographs are mandatory when considering a possible corrective osteotomy. Long-standing radiographs of both lower extremities are extremely useful, and when properly exposed can be considered the single most reliable means of assessing the various characteristics of the malunion, including both limb length discrepancy and coronal plane alignment (5,6). Good-quality lateral radiographs of the knee in extension are also indispensable regarding sagittal plane deformities. Determining the level of deformity is a critical consideration, and again highlights the reason why the absolute value of the angular deformity can never be considered an adequate criterion to determine possible indications for surgery (1,2,5,6). Computed tomographic scans have been considered the most accurate means of determining rotational malalignment, although clinical examination should be able to reproducibly delineate the magnitude and direction of any rotational deformity (7–9).

Perhaps as important as assessing the mechanical alignment (5,6) and joint orientation is the need to carefully scrutinize the intramedullary contour of the involved bone. If the canal is occluded by internal callus, it may make it very difficult to stabilize the deformity using an intramedullary rod. It is therefore critical to assess the radiographs and determine if it is possible to pass an intramedullary rod down the canal of the involved bone. Note should also be made of remnants of prior internal fixation, and broken drill bits or screws may be a relative contraindication to use of an intramedullary rod. Prior or planned arthroplasty, particularly hip replacement, could be a further consideration that may preclude the use of intramedullary fixation. Preoperative assessment is conducted as a process of discovery, designed to identify the critical aspects of the patients' condition and clinical presentation. After thorough evaluation, a patient would be regarded as most suitable for intramedullary fixation, internal fixation, or external fixation. These treatment options are not mutually exclusive, and one or more options may be suitable for any given case. Regardless, when all aspects are considered it is most likely that a single option will possess specific advantages compared to the alternatives.

CLASSIFICATION

For femoral deformities, the main consideration focuses on the possibility of stabilization with an intramedullary rod. The preoperative assessment, both clinical and radiographic, should directly evaluate the suitability of the involved limb for intramedullary fixation. In the absence of any specific contraindication, intramedullary fixation is generally preferred. In those cases where any one of the multiple clinical factors indicates that the limb is unsuitable for intramedullary fixation, most often external fixation should be utilized. This includes both an active infection and a well-defined history of prior infection. Open physes or a medullary canal that is significantly occluded may also be considered contraindications to intramedullary fixation.

Most femoral malunions are amenable to intramedullary fixation. This includes limbs with no significant history of infection and a minor limb length discrepancy that is generally less than 2 cm, including both diaphyseal and metadiaphyseal deformities. Metaphyseal deformities can be corrected using the focal dome osteotomy technique. Both angular and rotational deformities are clearly amenable to acute fixation and stabilization with an intramedullary rod. Significant translational deformities often result in occlusion of the medullary canal, which may make passage of an IM rod more difficult, though still possible.

Classification should account for all significant factors identified in the preoperative evaluation, both clinical and radiographic. Length discrepancy and rotation, as well as the level and magnitude of the deformity, must all be considered. Any history of prior infection, the presence of open physes, and limb length discrepancy greater than 2.5 cm would all be considered more amenable to external fixation. Open physes are clearly a relative contraindication to intramedullary fixation. Soft tissues must be assessed critically but, in contradistinction to the tibia, with femoral malunion rarely does the degree of soft-tissue injury impact significantly on the decision whether or not it is suitable for acute correction. Vascular compromise and joint contractures, such as a stiff knee, are further considerations. Consideration must also be given to local factors, including the internal characteristics of the medullary canal of the involved bone. In many instances this is the most important consideration, and certain femoral malunions may be extremely difficult to stabilize with an IM rod. In the presence of any single aspect that would be considered a contraindication, external fixation would be preferred.

TREATMENT OPTIONS

Those patients who remain symptomatic despite conservative measures are clearly candidates, as are those patients whose deformity places them at great risk of developing premature osteoarthritis (1,4). Rotational deformities are suitable for correction when the gait disturbance is obvious or when attempts by the patient to compensate for the deformity result in secondary complaints, usually in the vicinity of the hip. Conservative management, including the use of appropriate lifts and braces is appropriate, but when a patient has failed to respond to nonoperative measures, corrective osteotomy is an attractive option. Further considerations include not only the elements of deformity but also the patient's age, activity level, and body mass, as well as patient's expectations and comorbid conditions.

The majority of post-traumatic femoral deformities can be corrected acutely, although limb length discrepancy is the most common contraindication to full acute correction. Corrective osteotomies can be used for acute correction and restoration of limb length when the discrepancy is less than 2.5 cm (13). With larger leg length discrepancies one may consider staged reconstruction. An initial acute correction of the angular and rotational deformities can be stabilized with an intramedullary rod. Secondary lengthening of the limb can be performed as a lengthening over the nail (15), or alternatively lengthening rods are now available and may be considered.

Options include internal fixation, using either open (3,12,13) or percutaneous (11) exposures. Fixed angle devices, including both blade plates and locking plates, have definite advantages. Intramedullary rods are preferred in many circumstances, and can be inserted either antegrade or retrograde. External fixation (16) has the broadest indications, and is preferred for gradual corrections when limb length discrepancy is significant. External fixation is generally preferred for those cases with either a history of infection or an evidence of active infection. Totally implanted gradual lengthening devices are now available and are also useful when limb length discrepancy is significant. These are currently limited to intramedullary devices, specifically telescoping rod designs. The choice of intramedullary rods or internal fixation is often influenced by the quality of the patient's bone, as well as the internal characteristics of the medullary canal.

The spectrum of treatment alternatives includes virtually every means of stabilization from external fixation alone through internal fixation. External fixation is preferably unilateral, although circular and multiplanar devices have advantages in limited circumstances. Certainly, in adults, the mobility and convenience of a unilateral fixator is an overwhelming consideration. Unilateral devices provide excellent stability in adult bone and for the majority of cases is preferred. Intramedullary fixation is suitable in isolation for the majority of diaphyseal deformities. However, for metaphyseal and metadiaphyseal lesions, it is best to consider a combination of external fixation and intramedullary stabilization, the fixator-assisted nailing technique.

Fixator-assisted nailing involves application of a temporary external fixator used intraoperative to control correction of the osteotomy. After the deformity is corrected acutely, an intramedullary rod is inserted as definitive stabilization and the fixator is then removed (Fig. 1). The fixator is used to effectively limit secondary deformity such as flexion through a distal femoral osteotomy. Correction of the deformity can be achieved to a very high degree of accuracy and stabilization with the intramedullary rod can be augmented with appropriate blocking screws to contain or limit motion of the metaphyseal fragment. If one fails to recognize the potential for additional motion of the metaphyseal fragment independent of the intramedullary rod, it allows for the introduction of residual deformity. Blocking screws in effect constrict the canal and confine motion after the deformity has been corrected. An intramedullary rod in isolation is generally suitable only for true diaphyseal lesions. One could consider use of the femoral distractor as the temporary external fixation device, and this certainly is a common application as a fixator-assisted nailing.

This combined approach can be further applied as a combination of external fixation with intramedullary fixation for the purpose of lengthening over a nail (15). In this instance, the external fixator is applied and the osteotomy is performed. The deformity correction is completed to the satisfaction of the treating surgeon, and then an intramedullary rod is passed for more definitive stabilization. The external fixation then remains in place, only to act as a device for applying a distraction force through the osteotomy to gradually achieve lengthening (15).

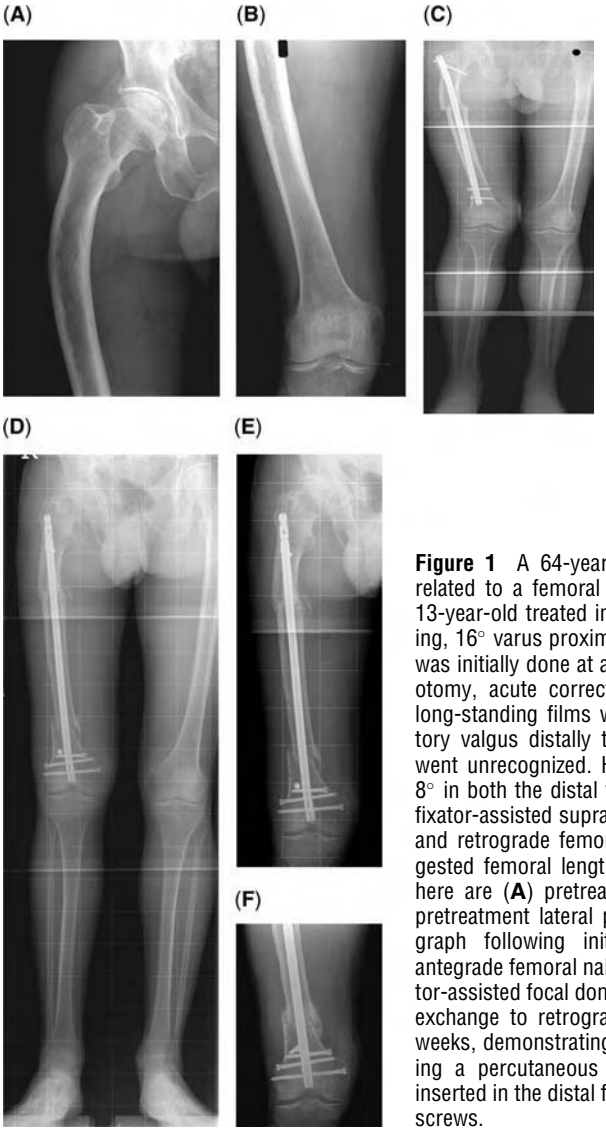

Figure 1 A 64-year-old man developed chronic hip and knee pain related to a femoral malunion resulting from a fractured femur as a 13-year-old treated in traction. The deformities included 3 cm shortening, 16° varus proximal femur, and 15° internal rotation. The treatment was initially done at another institution through a subtrochanteric osteotomy, acute correction, and antegrade femoral nail. Unfortunately, long-standing films were not obtained preoperatively, and compensatory valgus distally that had developed during his remaining growth went unrecognized. He presented with a residual valgus deformity of 8° in both the distal femur and proximal tibia. This was treated with a fixator-assisted supracondylar focal dome osteotomy, acute correction, and retrograde femoral nail. Given the age, the person declined suggested femoral lengthening to equalize the limb lengths. Reproduced here are (**A**) pretreatment anteroposterior (AP) proximal femur; (**B**) pretreatment lateral proximal femur radiographs; (**C**) erect leg radiograph following initial corrective subtrochanteric osteotomy and antegrade femoral nail; (**D**) erect leg; (**E**) AP radiographs following fixator-assisted focal dome supracondylar osteotomy, acute correction, and exchange to retrograde femoral nail; and (**F**) AP radiograph at nine weeks, demonstrating bridging callus and typical of rapid union following a percutaneous osteotomy. Note the use of a blocking screw inserted in the distal fragment immediately proximal to the three locking screws.

Diaphyseal deformities corrected using any internal fixation device are the most limited, because there are very few options when the intramedullary nail fills the canal both proximal and distal. The correction achieved is dictated by the characteristics of the bone, and it can be frustrating and very difficult to attempt to make any further modification or adjustment to the correction initially achieved. The same considerations hold true when using plates for correction of femoral malunion. With diaphyseal deformities, the local anatomy of the contour of the femoral cortex often dictates the accuracy of the correction. As with intramedullary nails, there is little margin for error and the correction achieved may be compromised by anomalies of the femoral cortex. Locking plate technology and minimally invasive techniques (10) have made internal fixation a more attractive option than conventional plates and open exposure of a femoral osteotomy. The femoral distractor (alternatively, a temporary external fixator) can again be used quite reliably to correct the deformity acutely. Certain metaphyseal deformities are very amenable to this technique, including both supracondylar and subtrochanteric femoral deformities.

Whichever form of stabilization is chosen, the same principles of deformity correction apply. Both alignment and joint orientation should be corrected according to well-defined standards and criteria (5,6). One may consider intramedullary rods as a means of internal

fixation through a truly percutaneous approach, and impressive corrections are possible with minimally invasive surgery. Use of locking plates through limited incisions (11) also preserves local vascularity and soft-tissue attachments. Locking plate technology has the added advantage of providing stability in osteoporotic bone, whereas intramedullary rods may be less stable when the diameter of the canal is significantly larger than the rod itself. Locking plates are an alternative device that may be regarded as potentially advantageous in experienced hands.

SURGICAL TECHNIQUES

The patient can be positioned on a radiolucent table, either lateral or (most often) supine, with a sandbag under the pelvis to provide circumferential access. The limb is prepped and draped free to facilitate clinical assessment of rotation. For true hip or subtrochanteric osteotomies, there are advantages to placing the patient supine on a fracture table. For distal femoral osteotomies, a retrograde IM nail has significant benefits. If an IM rod is planned for fixation, the initial portions of the procedure involve preparation for its eventual insertion. A limited incision and fluoroscopically guided dissection can allow the femoral canal to be opened and cannulated with a guide wire. Reaming is performed only after the osteotomy and acute deformity correction are complete. It is imperative to vent the femur prior to reaming, to limit the potential for clinically significant emboli including fat and bone fragments. A popular technique is to insert a cannulated drill bit into the distal femoral metaphysis in a unicortical manner. This is then simply left in situ while reaming is completed.

There can be little doubt that application of a unilateral external fixator is the safest and least complicated means of addressing femoral malunion (Fig. 2). There is little need for sophisticated technology and the lateral aspect of the femur provides a means of safe and convenient access. Multiple pins proximal and distal provide adequate stabilization in the vast majority of cases. Insertion of half-pins perpendicular to the mechanical axis of the femur both proximal and distal to the apex of deformity is a very reliable technique. The osteotomy itself can almost always be completed using a modification of the De Bastiani technique. Multiple drill holes are made at the chosen level, percutaneously through a small lateral incision. A small osteotome is then inserted through the same incision, and rotation used to complete the osteotomy. Using this technique is safe and reliable, provided an image intensifier is available to assess the level of the osteotomy and the position of the osteotome, as well as to monitor progress.

Correction of rotational deformities is easiest to achieve by initial insertion of one pin proximal and one pin distal, rotated axially in the direction matching the deformity identified preoperatively. After the osteotomy is completed, rotation is corrected acutely by simply realigning the two pins to a parallel orientation. This is in almost all circumstances the most expeditious and convenient way to address rotational deformity. This is true regardless of whether external fixation is used as definitive stabilization, or when it is only used on a temporary basis. If the correction is planned as a fixator-assisted nailing or lengthening over a nail, particular attention must be taken when inserting the pins to confirm that there is adequate room for the intramedullary rod.

For gradual correction of angular deformities, the placement of the hinge becomes critical because the hinge placement ultimately dictates the quality of the correction. As in other long bone deformities the hinge should correspond to the level of the deformity, not necessarily the osteotomy (5,6). The osteotomy is performed at a convenient level, where the chosen method of fixation will indeed provide stability, and is also performed through an area of bone that has a high probability of satisfactory union. Using a circular fixator, such as the Ilizarov device, hinge placement can be modified to correspond to the precise level, position, and orientation that will facilitate correction of the deformity. When using a unilateral fixator for gradual deformity correction, it can be difficult to position the hinge at the ideal position and orientation to avoid introducing secondary elements of deformity. When a unilateral device is applied for acute deformity correction any secondary elements of deformity that would have been introduced can be recognized and addressed intraoperatively. As a general rule, a unilateral device is best used to acutely correct the angular and rotational elements of deformity, and then a gradual lengthening performed to equalize length.

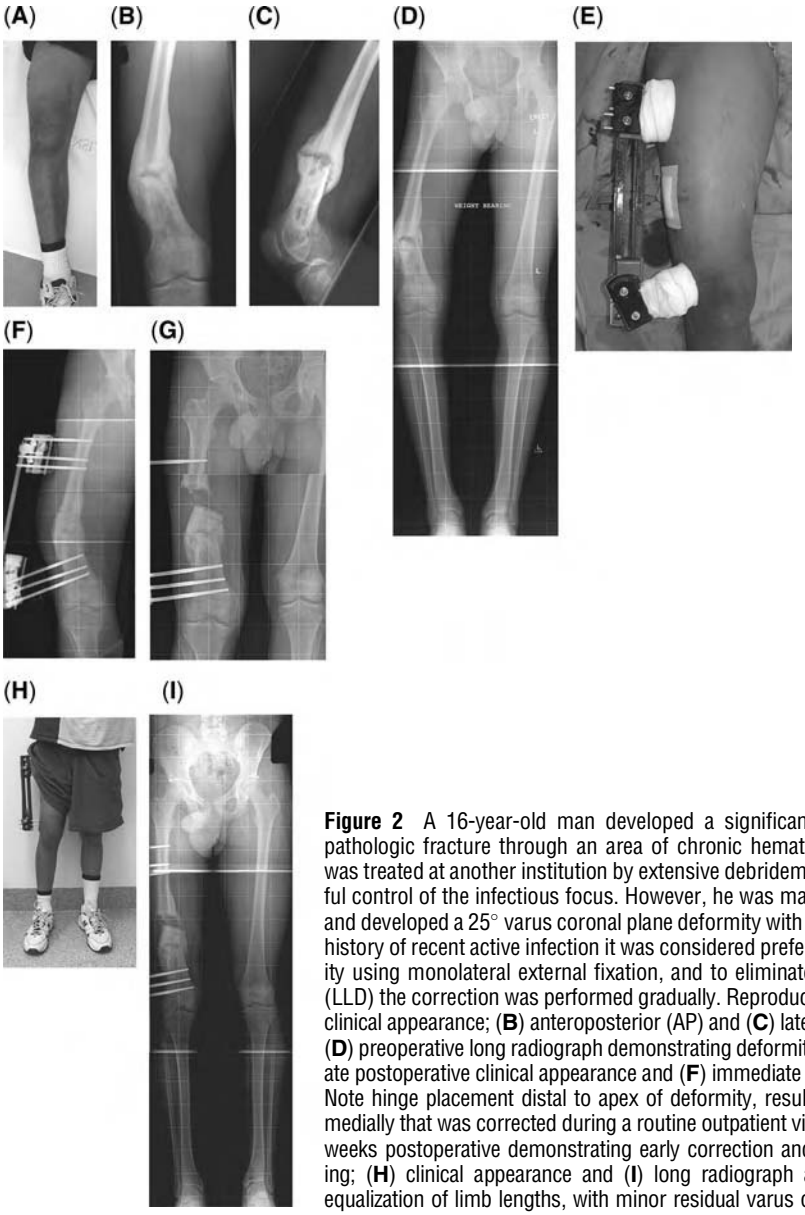

Figure 2 A 16-year-old man developed a significant femoral deformity after a pathologic fracture through an area of chronic hematogenous osteomyelitis. This was treated at another institution by extensive debridement, and resulted in successful control of the infectious focus. However, he was managed in traction and a cast, and developed a 25° varus coronal plane deformity with 3.5 cm shortening. Given the history of recent active infection it was considered preferable to address this deformity using monolateral external fixation, and to eliminate the leg length discrepancy (LLD) the correction was performed gradually. Reproduced here are: (**A**) preoperative clinical appearance; (**B**) anteroposterior (AP) and (**C**) lateral preoperative radiographs; (**D**) preoperative long radiograph demonstrating deformity including LLD; (**E**) immediate postoperative clinical appearance and (**F**) immediate postoperative AP radiograph. Note hinge placement distal to apex of deformity, resulting in secondary translation medially that was corrected during a routine outpatient visit; (**G**) AP radiograph at four weeks postoperative demonstrating early correction and medial translation developing; (**H**) clinical appearance and (**I**) long radiograph at 10 weeks, demonstrating equalization of limb lengths, with minor residual varus of less than 2°.

COMPLICATIONS

The potential complications associated with post-traumatic femoral deformity correction are those commonly associated with limb lengthening, deformity correction, external fixation, and intramedullary rodding of any long bone. Residual deformity, delayed union, and nonunion are the typical complications of bone, and are not uncommon problems. Joint contracture and subluxation would be much less commonly encountered, but are certainly possible when length is a significant element of the deformity. Nerve and vascular injuries can occur both acutely intraoperative and postoperatively with gradual distraction; neither would be commonly encountered. Pin site infection is a common issue, and is managed as indicated based on the severity of infection. It is uncommon to require pin removal to control infection. Persistence of infection after pin removal is distinctly uncommon. Deep infection complicating lengthening over a nail (15) has a reported incidence of 6%. This is typically managed by rod removal and reaming, and generally results in successful resolution of infection.

FUTURE DIRECTIONS

The most important recent development has been the introduction of totally implantable lengthening devices (Fig. 3). There are several telescoping rod designs currently available, using different sources to power the lengthening mechanism. Although potentially minimizing the complications often associated with limb lengthening, surgeons have found that they

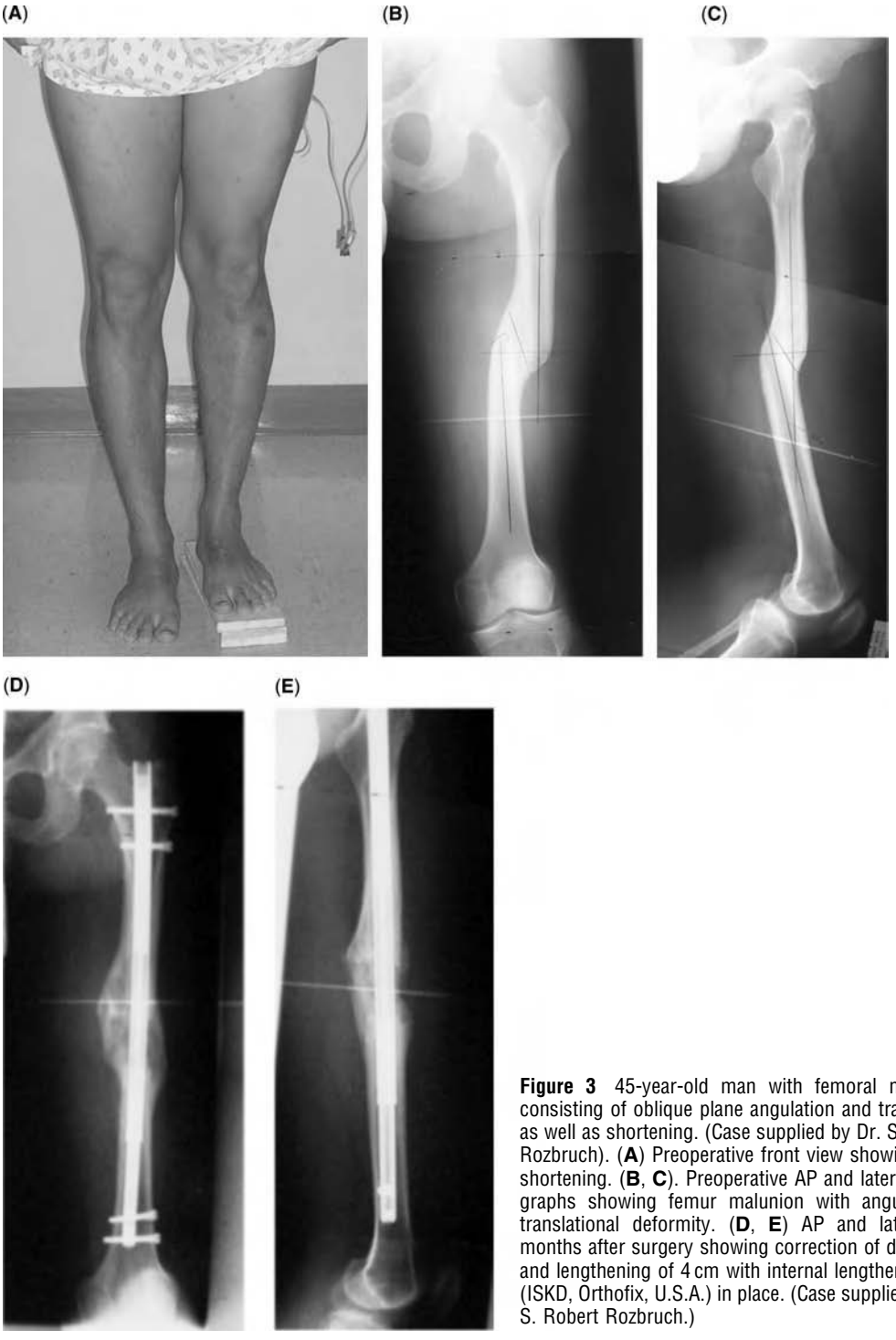

Figure 3 45-year-old man with femoral malunion consisting of oblique plane angulation and translation as well as shortening. (Case supplied by Dr. S. Robert Rozbruch). (**A**) Preoperative front view showing 4 cm shortening. (**B**, **C**). Preoperative AP and lateral radiographs showing femur malunion with angular and translational deformity. (**D**, **E**) AP and lateral six months after surgery showing correction of deformity and lengthening of 4 cm with internal lengthening nail (ISKD, Orthofix, U.S.A.) in place. (Case supplied by Dr. S. Robert Rozbruch.)

have their own set of unique complications and problems. However, these devices offer significant advantages, particularly with regard to femoral lengthening and deformity correction. The future should eventually see totally implantable devices also used for gradual deformity correction, in an analogous manner.

The other major development has been the rapid rise in popularity of locking plates for internal fixation (11). The next step should see the introduction of more sophisticated instrumentation for truly percutaneous applications. Fixator-assisted plating will then become an even more attractive option, particularly when integrated with computer navigation. Although innovation and technology will undoubtedly play an important role in determining future developments, the surgeons' skill and imagination will continue to dictate how, when, and where these new devices will be best utilized.

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14 Bone Defects

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INTRODUCTION

The treatment of bone loss, occurring as the result of acute trauma or segmental resection for reconstructive procedures in the skeleton, has traditionally been a complex surgical problem. Numerous procedures have been devised to reconstitute bone stock, obtain fracture union, and provide a stable functional limb.

In an attempt to avoid the problems associated with deficient graft materials and free tissue transfers, internal bone transport is a technique that has been a successful methodology for bony reconstruction for both acute and reconstructive bone loss (1).

CLINICAL EVALUATION

Most published reports utilizing bone transport have dealt primarily with nonunions or infected nonunions for which transport techniques are undertaken as a planned elective procedure. The situation is more complex when the surgeon has to deal with segmental defects following an acute traumatic situation. Fractures often will have extensive soft-tissue loss in concert with large skeletal defects. For these types of injuries, the principles of open fracture management must be adhered to prior to the institution of bone transport techniques.

When considering bone transport in an acute or chronic situation, it is paramount to determine if a biologically sound healing environment is present or can be achieved at both the site of the proposed corticotomy and/or the docking sites. The success of both corticotomy and solid docking involves well-vascularized segments of bone and soft tissue (2). If soft-tissue incompetence (dysvascularity) is present at the proposed corticotomy site, the production of healthy regenerate may be compromised (3–5). Severe open fractures with a wide zone of injury are often associated with very poor soft-tissue coverage at the site of injury (6). Associated soft-tissue compromise may be coexistent elsewhere in the limb, which may involve the site of the proposed corticotomy.

The periosteal blood supply is derived primarily from the surrounding soft-tissue envelope. If this is inadequate and unable to provide a vascularized, viable periosteal sleeve, the prospects for the development of an inadequate regenerate is very real. In these situations, an alternative corticotomy site, performed through healthy tissues, should be selected.

Solid healing of the docking site requires all the biologic components necessary to heal what is equivalent to an acute fracture. The ability to revascularize the ends of the docking segments and facilitate the migration of pluripotential cells is dependent on the revascularization process. If the docking fragments are excessively mobile, the moving bone ends will traumatize the local blood supply. Thus, the influence of a stable mechanical environment facilitates docking site union. The hallmark of these events is the inflammatory phase of fracture healing that promotes the revascularization process. This area must be manipulated to provide the appropriate vascular response either through aggressive debridement or through soft-tissue coverage techniques.

In cases of infected nonunions, draining sinuses with atrophic and scarred soft tissues are often present at the nonunion/proposed docking site. Consideration of these issues helps to determine the extent of nonviable tissue debridement necessary to obtain healthy vascular tissue. Magnetic resonance imaging can be helpful to determine the extent of marrow dysvascularity found in a proposed transport segment or proposed docking site (4,7–10). Additionally, arteriography may also be useful to determine distal vascularity (blush) with regard to docking segment viability as well as soft-tissue viability (Fig. 1).

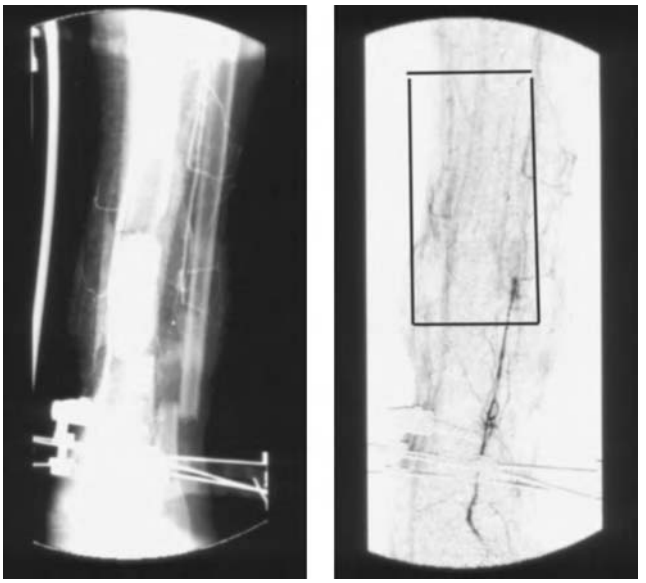

Figure 1 Angiogram of tibial defect prior to corticotomy and transport. Note well-vascularized transport tract and docking site blush (*black outline*).

If the soft-tissue defect is similar in size to the bone defect, then soft-tissue transport in conjunction with the bone transport is possible (11,12). Tissue loss that exposes bone is not amenable to combined soft-tissue/bone transport without first addressing the exposed bone. This is accomplished through rotational or free tissue transfer to cover the bone. Alternatively, the bone should be resected back until healthy soft tissue covers the bony segment (6,12,13).

TREATMENT OPTIONS

During the initial presentation of an acute traumatic defect, a simple monolateral, four-pin external fixator is applied to the limb (Fig. 2). Every effort should be made to remove any

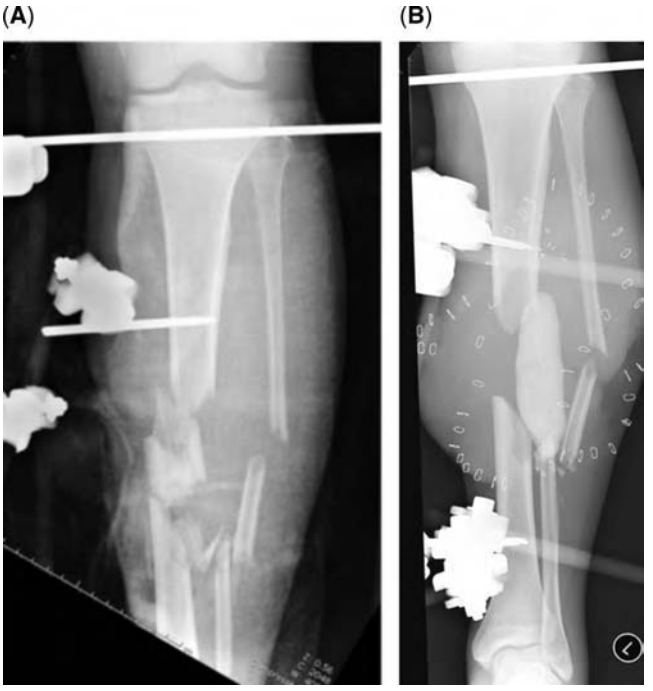

Figure 2 (**A**) Severe open tibia stabilized with a simple four-pin external fixator. (**B**) Antibiotic block placed beneath free flap coverage to maintain prospective transport pathway prior to frame application. In this case, the flap was placed prior to transport frame placement. The usual circumstance is to place frame before free flap application.

Table 1 Clinical Management

	Degree of Bone Loss	Soft-Tissue Integrity	Vascularity Vasculature/Small Vessel Disease	Pearls of Management	Potential Complications
Bone transport	> 3–4 cm	Competent at corticotomy, transport and docking sites	Vasculature intact no small disease (good)	Antibiotic beads in transport tract	Modulate distraction rate site to avoid deficient regenerate; Graft docking site
Bone transport with open wound	> 3–4 cm	Incompetent at nonunion or fracture site	Good	Flap coverage over transport/docking site	Transport tract cutting through flap pedicle
Bone and soft-tissue transport	No minimum defect size	Incompetent	Dysvascular limb; small vessel disease	Resect bone back to healthy bone until covered by granulation soft tissue	Poor regenerate. Invagination of soft tissue into transport tract
Acute shortening	<4 cm	Incompetent	Dysvascular limb; small vessel disease	May bridge larger defects with gradual shortening 0.5 cm per day	Kinking of vasculature; bunching necrosis of soft tissues
Combination transports	Massive >10–12 cm	Competent	Good	Bifocal, trifocal treatment; fibular transport	Number of potential complications rises exponentially with complexity of transport

devitalized bone and necrotic soft tissue. The patient is returned to surgery every 48 hours for additional irrigation and debridement procedures until the zone of injury has declared itself, and the wound has become culture negative. At this point, the decision to proceed with transport is made (6,13–15). This strategy should also be employed when initiating a staged reconstruction for an infected focus. Alternatively, acute or gradual shortening offers advantages over transport in the patient who presents with vascular insufficiency, i.e., a one vessel leg where free vascularized tissue transfer is contraindicated. Cases in which the patient has a systemic small vessel disease process, i.e., diabetes, severe peripheral vascular disease, connective tissue disorder, etc., are also candidates for shortening strategies (6,9,16–19) (Table 1).

Shortening acutely can be accomplished safely for defects up to 3 to 4 cm in the tibia. More shortening can be tolerated acutely in a femoral defect up to 5 to 7 cm. In some situations, it is advantageous to decrease the transport distance and thus time in the frame. Shortening aids in soft-tissue coverage by decreasing tension and gaps in the open wound; this approach may allow wounds to be closed by delayed primary closure, or healed by secondary intention or simple skin grafting. With this technique, one may avoid extensive free flap coverage (Table 1) (6,10–12,17).

Acute shortening of more than 4 cm can cause the development of tortuous vasculature and actually produce a low flow state with detrimental consequences. Open soft-tissue wounds when acutely compressed can become notably bunched and dysvascular, with the development of significant edema and the possibility of additional tissue necrosis and infection (11,12,14,16). More than 4 cm may be safely accomplished in the femur; however, similar problems with wound edema and bunching may occur (1,20,21).

If the defect is larger than can safely be closed acutely, a gradual shortening can accomplish the same goals. Shortening at the rate of 0.5 cm per day in divided doses will rapidly oppose the skeletal defect as well as avoid the detrimental soft-tissue consequences and vascular element kinking of acute defect compression (6).

Massive defects, greater than 8 to 10 cm, are candidates for combined treatment options. The success of massive transport is directly proportional to the number of complications associated with these rigorous reconstructions (4,8–10,14,15,17,20–23). It is recommended that combination methodologies be initiated with great caution, in cases where all transport parameters are optimized, i.e., intact vascularity, small vessel disease, intact soft-tissue sleeve, etc. (1,8,16). Acutely shortening the defect can reduce the transport time required to achieve

docking. Once docking is accomplished, straightforward lengthening can then be carried out. The ability to stop the lengthening process is available for patients who may experience ''frame fatigue'' during a prolonged bone transport. It is much easier to stop a lengthening procedure and allow the lengthened regenerate to consolidate with the only morbidity being a short functional limb. Alternatively, once a stable limb has been achieved, delayed lengthening can be accomplished by alternative measures such as lengthening over an intramedullary (IM) nail or achieving resolution using an internal lengthening nail. Alternatively, it is very difficult to salvage a limb if transport is stopped during mid-distraction prior to docking.

Bifocal and trifocal strategies can be employed such as double-level transport in combination with acute shortening (4,8,9,16,17,22). Free vascularized fibula, combined with acute shortening and bone transport, has also been reported as a methodology to reduce the substantial frame time required for massive bone defect reconstruction (1,13). Transport over nails has also been employed for larger defects in both tibial and femoral deficiencies (1,21,24). Lastly, transverse ipsilateral fibular transport has been reported for reconstruction of massive tibial defects.

For acute bone loss, it is advantageous to avoid local rotational flaps because the rotated muscle is often involved in the acute zone of injury, and thus performing a rotational myoplasty may further damage a compromised muscle. Ultimately, the additional vascularity supplied by this compromised muscle may be of minimal value. The use of free tissue transfer helps to provide a well-vascularized tissue bed through which bone transport, docking, and eventual healing of the docking site can occur.

Once a healthy wound is achieved, a specific bone transport frame is then applied. It is much easier to perform a radical debridement in the presence of a simple external fixator, rather than through and around a complex bone transport device. Although access is more difficult for the plastic surgeon, it is preferable to have the transport frame in place prior to the placement of the free flap. In this way, the vascularized pedicle can be planned and located away from any transport wires or pins that may eventually impinge upon the flap anastomosis.

SURGICAL TECHNIQUE

The majority of bone transport is performed for tibial defects. A basic tibial transport frame consists of a multiple ring construct. Depending on the size of the limb in question, anywhere from three to five rings may be used. For a proximal or midshaft defect, a single proximal ring will be attached, parallel to the knee joint, at the level of the fibular head. For more distal tibial defects, two proximal rings can be utilized (Fig. 3).

For distal one-third tibial bone loss, the distal fixation segment is not large enough to accommodate two rings, and therefore only one distal ring is used. A single, intermediate transport ring is utilized. The proximal and distal ring blocks are attached to each other by four long-threaded rods. Location of the rods on the rings can be varied to allow for large open corridors, to facilitate plastic surgeon's placement of free flaps with the frame in place. The transport ring is placed midway on the four long-threaded connecting rods. Initially, this ring is left to ''float'' up and down on the four connecting rods. Frames should be preassembled to facilitate mounting in surgery (Fig. 4).

Precise placement of the frame is crucial to ensure that the proposed docking side is aligned with sufficient cortical contact for union to occur (Fig. 5). The preassembled frame is placed on the limb, and a transverse 1.8 mm wire is inserted as a proximal reference wire. This wire is placed parallel to the knee joint at the level of the fibular head, attached and tensioned to the proximal ring.

A transverse olive wire or perpendicular schantz pin is next placed into the proximal ring fixation block. Once the overall alignment of the frame is confirmed, the wire is tensioned, or the half-pin is connected to the ring, locking the proximal limb segment in place and preventing the frame from shifting during subsequent distal fixation. If shortening has occurred or the limb is acutely shortened intentionally, a proximal fibular capture wire or half-pin construct should be utilized to avoid dislocation of the proximal tibia–fibula joint when limb lengthening occurs (Fig. 6).

The distal ring fixation proceeds with the insertion of a smooth 1.8-mm reference wire at the level of the distal ring, parallel to the ankle joint line. The wire should be placed just anterior to the fibular shaft in the lateral view. The single smooth wire in the distal segment will be used to ensure appropriate alignment of the docking site in both the coronal and

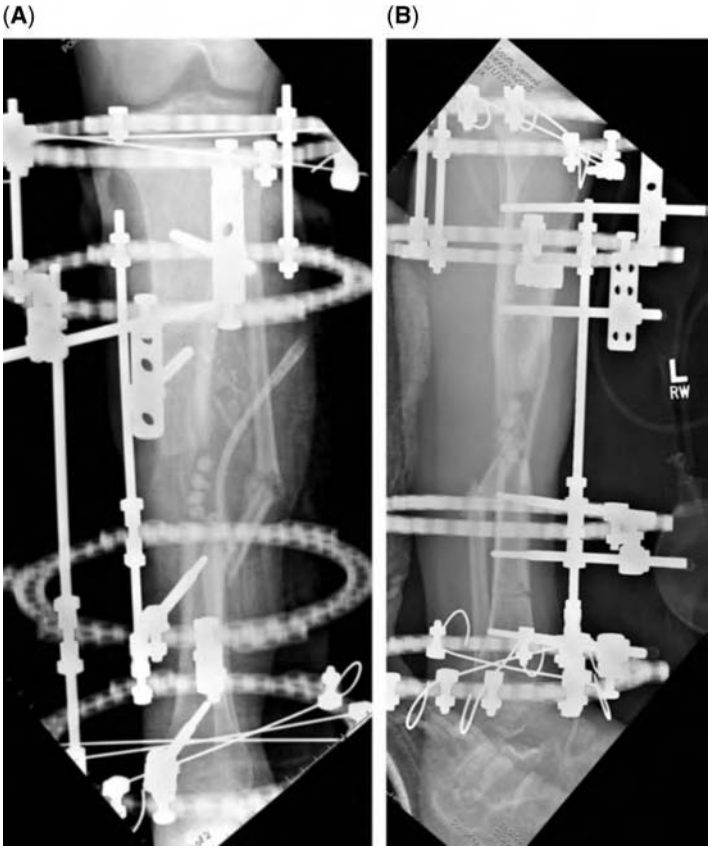

Figure 3 (**A**) and (**B**) Four-ring frame application following removal of antibiotic spacer. A distal corticotomy was selected. The limb was acutely shortened decreasing total transport time. A short chain of antibiotic beads is placed into transport tract.

the sagittal planes. Prior to attaching the solitary distal wire, the anteroposterior displacement of the proposed docking site is corrected. The image intensifier is utilized in the lateral position to view the segmental defect region. The distal fragment can be raised or lowered appropriately to align the posterior cortices across the defect. When the correct anteroposterior alignment is achieved, the smooth wire location is noted on the ring and the wire fixation bolts are attached to the ring at this location.

The image intensifier is now positioned to obtain an AP view of the segmental defect proposed docking site. Because only one smooth transverse wire is present distally, the entire distal fragment including the fibula can be translated on the smooth wire in order to correct any translational offset.

The distal segment is thus moved on the smooth wire in a medial or lateral direction until the lateral cortices above and below the defect are aligned. It may be helpful to position a threaded rod along the lateral cortex to gauge the adequacy of alignment in both the AP and the lateral views. After correct alignment is achieved, a second smooth wire is placed as a distal fibular capture wire. This wire locks in the position of the distal fragment and the proposed alignment of the docking site. This is followed by application of the remainder of distal fixation, using tensioned wires or half-pins as per surgeon's preference.

The intermediate transport ring is left unattached to the bone, especially if further plastic surgery is contemplated. The ring is ''floated'' in a proximal or distal location, out of the region of the open wound and proposed plastic surgery procedures. A reciprocating saw can be used to achieve congruent surfaces on the ends of the proposed transport and docking segment. An antibiotic cement spacer can also be placed across the defect. The block should be oversized to achieve a wedging effect into the defect, which confers additional frame stability by achieving temporary cortical contact (Fig. 3).

At the time of free flap or primary closure, the antibiotic spacer is removed, and a solitary chain of antibiotic cement beads is placed into the defect (Fig. 4). The beads provide and maintain a ''potential space'' or fibrous tunnel through which the transport segment will

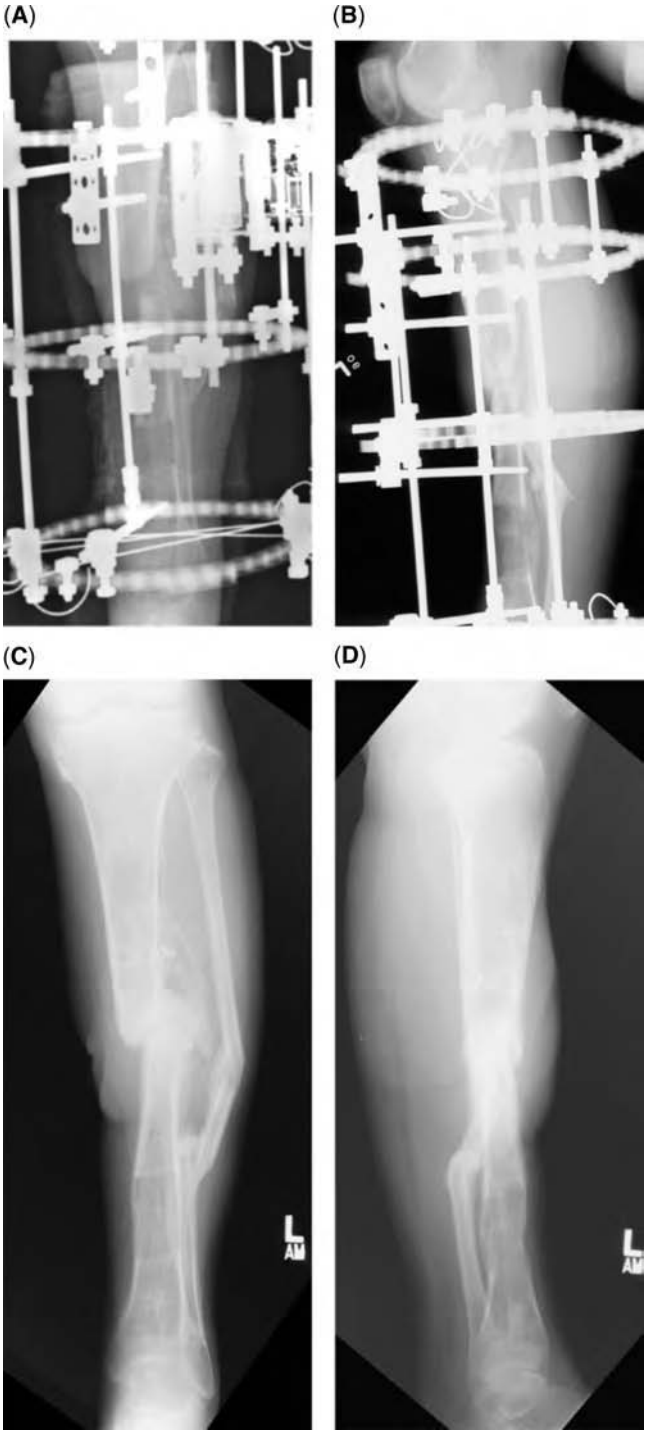

Figure 4 (**A**) Autodistractors used as transport ''motors'' sliding segment proximally onfour long-threaded rods (guide tracks) The transport ring attached to bone using 90° divergent 6 mm HA pins. Note the pins are attached proximally in the transport segment to ''pull'' the bone. The antibiotic beads have been exchanged for bone graft at the docking site. (**B**) Distractors removed and docking site compressed until healed and regenerate has consolidated. (**C** and **D**) Frame removal following healing of docking site and maturation of the regenerate with slight translation of docking site. Lateral view shows anatomic alignment with solid union at docking site.

travel. If the wound is closed primarily, antibiotic beads are still used to prevent invagination of the intact soft-tissue envelop into the transport pathway.

If combined soft tissue and bone transport is considered, then the bone must be resected back until covered by healthy remaining tissue (Fig. 7). The region of the proposed transport tract may be completely devoid of covering tissue, and this area is allowed to granulate. Once healthy tissue is achieved in this location, it can be covered with a split thickness skin graft

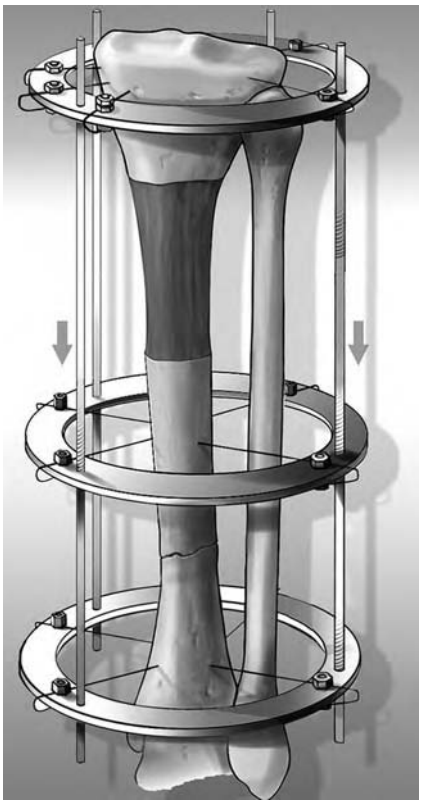

Figure 5 Three-ring transport frame with proximal and distal reference wires.

and transport of the segment undertaken. Other times, the bone and soft-tissue defect close simultaneously, with transport obviating the need for any plastic surgery coverage.

Transport should be delayed for at least three weeks following free flap coverage. The delay allows the free flap anastomotic site to become fully epithelialized and healed and is then able to withstand the inevitable tension forces that will be subjected to it during transport. If the wound is closed primarily or bone resected back to healthy tissue, corticotomy and transport can be undertaken immediately.

Two methods are commonly used to transport the bone segment. Traditional fixation of the transport segment is accomplished using obliquely placed olive wires across the transport segment. The resultant pull vector of these wires should be parallel to the bone axis. These wires exit inferiorly through the soft tissues. These free wires are attached to the distal ring using a slotted rod on a hinged assembly. The angle of these wires changes as the transport progresses, and this is accommodated by a hinge on the pulling mechanism (Fig. 8).

Once the transport segment approaches the distal docking site, the longitudinal wires are exchanged for transverse wires and attached to the intermediate transport ring. This is done to maintain constant compression at the docking site.

The second method of transport utilizes a transport ring throughout the entire treatment. The ring is positioned at the mid to distal third of the proposed transport segment. The eccentric location of the transport ring is chosen because the ring will "pull the bone" into docking position rather than "push the transport segment." A "pushing" construct occurs if the ring and bone attachment is located closer to the corticotomy, rather than the docking end of the transport segment. This "pushing" construct results in an unstable transport segment that will have a tendency to deviate during transport (Fig. 4). A useful analogy is that it is easier to pull a string of spaghetti (transport segment) into a straight line, rather than trying to push it into a straight line.

The transport ring is rigidly fixed to the bone by utilizing transverse tensioned wires or half-pins placed on either side of the transport ring. It is recommended that two 6-mm half-pins be utilized for segmental transport, and that the half-pins be placed at 90-degree angles to each other (Fig. 4). This pin orientation avoids excessive cutting through the

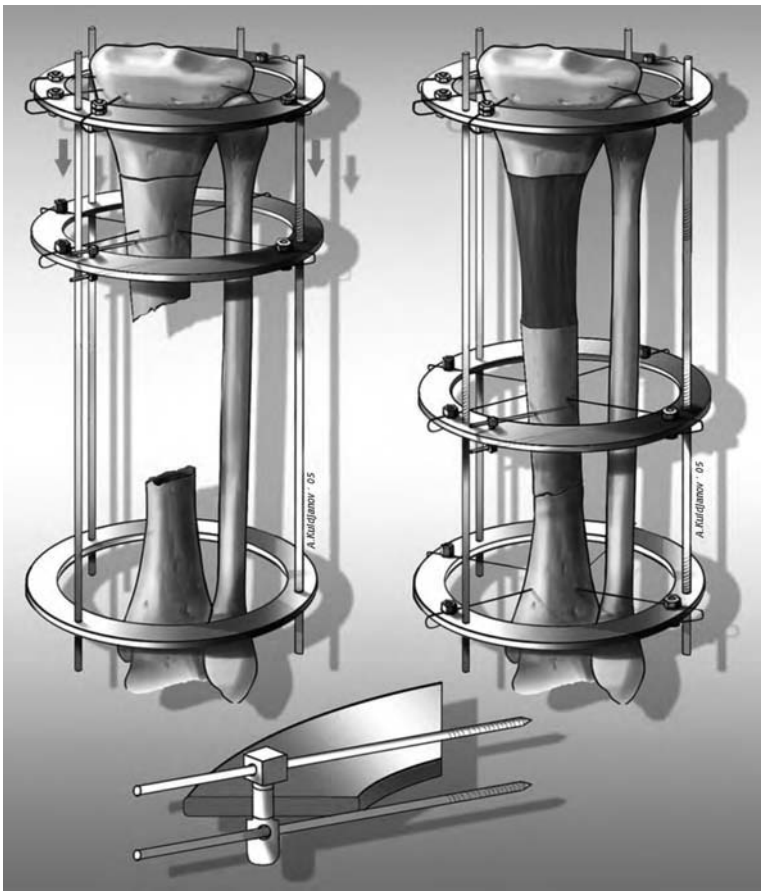

Figure 6 Ring transport using either transverse wires or two Schantz pins mounted above and below the transport ring.

transport tract. Parallel orientation of transport half-pins has been reported to cause a ''double-hit'' wound dehiscence. The first transport pin cuts through the soft tissues, and just as the transport tract begins to heal, the second pin reopens the same tract with subsequent wound breakdown (13).

Another way to assemble the transport frame is for simultaneous but independently controlled shortening and lengthening. This can be connected with rods or Taylor spatial frame struts. In this manner, the defect shortening can be performed faster than the regenerate lengthening, leading to earlier docking at the nonunion site. The fibula cannot be intact if differential shortening and lengthening is to occur.

Half-pins and transport wires will easily cut through a free flap; however, one must note the location of the flap anastomosis to ensure that transverse transport wires or half-pins will not impinge on this area.

Following fixation of the transport segment, a proximal or distal corticotomy is performed. In cases of acute bone transport following high-energy fractures, a wide zone of injury is often present. It is better to perform the corticotomy away from any region of previous soft-tissue compromise or zone of injury and, as such, double-level transport with two corticotomies is not easily achieved (Fig. 4).

Double-level transports have been performed in cases of large bone defects. These complicated reconstructions have been associated with a high rate of complications as well as nonunion of the corticotomy site. However, if double-level transport is to be carried out, additional proximal or distal transport rings will be required as described above (4,8,13,22,23).

Bifocal Approach

This is called bifocal because there are two segments with activity. One segment (the defect) is undergoing compression/shortening, and one segment (the bony regenerate) is undergoing

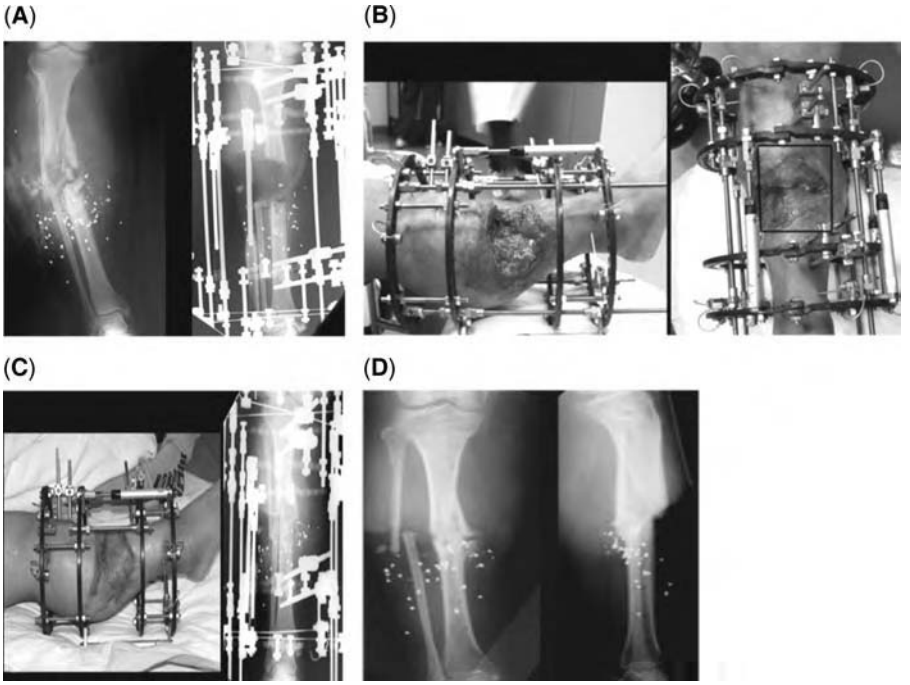

Figure 7 (**A**) Severe gunshot wound with segmental soft-tissue and bone loss. Patient had a one vessel limb as well as severe uncontrolled diabetes and heavy smoking history. (**B**) Gradual shortening at the rate of 0.5 cm/day was undertaken once healthy bone was resected back under soft tissues. As shortening progresses, bone is covered with healthy tissue (box). (**C**) After docking, the soft tissues were skin grafted. (**D**) Follow-up X rays at one-year post frame removal.

distraction/lengthening to maintain the length of the limb. A ring block is applied on either side of the bone defect. Another ring block is placed on the other side of the anticipated lengthening osteotomy site. Rods or struts are applied across this segment and are set up for lengthening or distraction. The rods are then disconnected in preparation for the osteotomy. The osteotomy is done in a percutaneous fashion using either the multiple drill hole and osteotome technique or the Gigli saw technique. Care is taken to perform this osteotomy outside the zone of injury in healthy bone. Ideally, this osteotomy is done in the metaphyseal bone. The proximal metaphyseal location is preferable to the distal metaphysis because of increased bone regeneration potential (Fig. 9).

Trifocal Approach

This is called trifocal because there are three segments with activity. One segment (the defect) is undergoing compression/shortening, and two segments of bony regenerate are undergoing distraction/lengthening. This can maintain the length of the limb. Rings are placed on either side of the defect. Additional rings are placed around what will be two lengthening sites. If the defect is in the middle of the tibia, two osteotomies are performed—one in the proximal and one in the distal tibia (Figs. 10 and 11). Two intercalary bone segments are transported toward each other (Fig. 10). If the defect is in the proximal or distal tibia, another trifocal option exists where two intercalary segments are transported in the same direction (not shown in Fig. 10).

Ilizarov Frame Considerations

The frame should be applied to the leg so that rings are perpendicular to the tibial axis, the rods are parallel to the bone axis, and there is adequate clearance between the soft tissues and the rings, especially at the posterior leg. The bone defect edges should be perfectly pointed toward each other to avoid deformity and to optimize contact at the anticipated

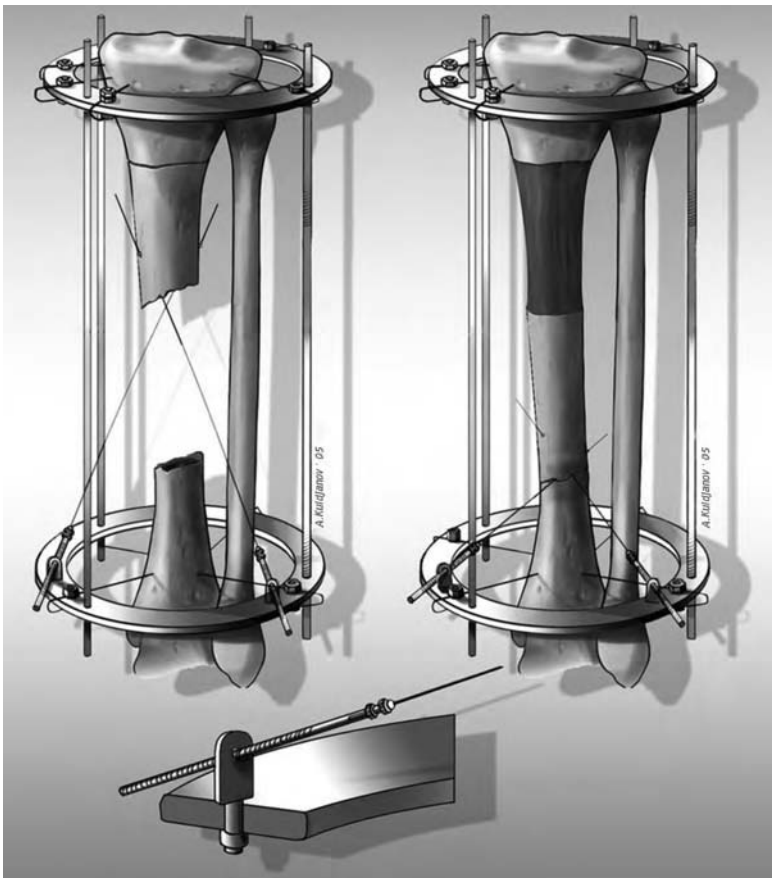

Figure 8 Traction wire transport using longitudinal wires. This methodology requires the threaded rod attachment to be connected to a hinged mechanism as transport proceeds.

docking site. If deformity should occur, this can be managed with frame modification and/or a surgical procedure to optimize contact at the docking site.

Taylor Spatial Frame Considerations

Rings are placed on either side of the defect site and the anticipated lengthening site(s). The rings can be placed independently to optimally fit the leg. This is called the rings first method. One ring is chosen as the reference ring for each level of movement, and it is important that this ring be placed orthogonal to the axis of the tibia. Mounting parameters are defined by the center of the reference ring, and this will define the point in space where the deformity correction will occur (Fig. 11). It is important to maintain enough distance between rings so that the struts can fit properly. In this frame, one is limited by the shortest length of strut. The advantages of this frame are that the application is easier, and the fit on the leg is better when using the rings first method. Also, residual deformity at the lengthening and docking sites can be addressed by using the same frame to correct angulation and translation simultaneously in the coronal, sagittal, and axial planes, without major frame modification. This allows precise docking with optimal bone contact and minimizes angular deformity at the docking and lengthening sites.

Fibular transport has been described for the treatment of massive tibial defects. Commonly, this involves the transport of the entire fibula transversely into the tibial defect. Precise frame orientation must be assured such that the fibula correctly "docks" at either end of the defect. A variation of fibular transfer is the split fibular transfer. The fibula is cut in the sagittal plane, accomplished through small lateral incisions. The medial half of the fibula is then transported into

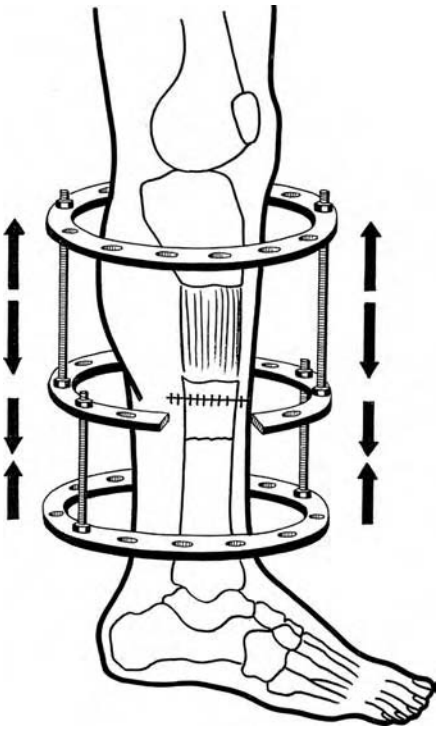

Figure 9 Schematic representation of bifocal treatment depicting gradual closure of bone and soft-tissue defect with bone transport. While there is shortening across the defect, there is simultaneous lengthening through a proximal tibial osteotomy. This maintains the length of the limb. *Source*: Courtesy of Arkady Blyakher.

the tibial defect via multiple olive wires. Olive wires are inserted into the fibula through 2-mm holes drilled in the lateral cortex of the fibula. The wires are then advanced out of the medial cortex of the fibula and attached to slotted, threaded rods linked to swivels on a medial tibial connecting plate. Because tension is placed on the wires, the olives will abut the medial fibular cortex and gradually transport that segment, leaving the lateral fibula intact (Fig. 12).

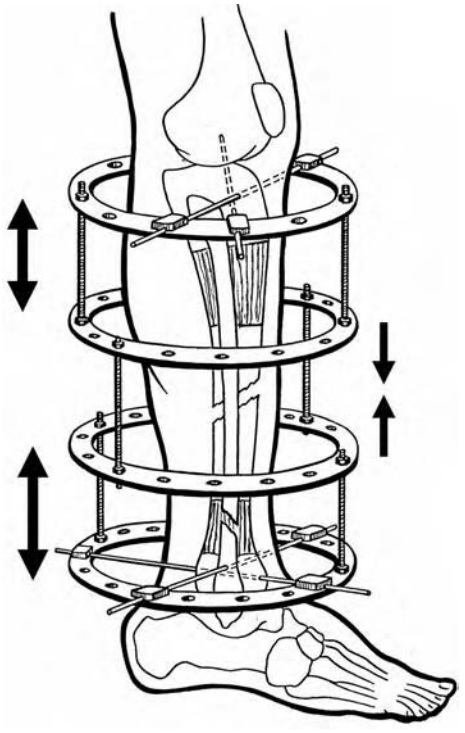

Figure 10 Schematic representation of trifocal treatment. Here, there are three foci of dynamic activity. There is gradual closure (shortening) across the defect and simultaneous lengthening (transport) through two osteotomies of the tibia. This maintains the length of the limb. *Source*: Courtesy of Arkady Blyakher.

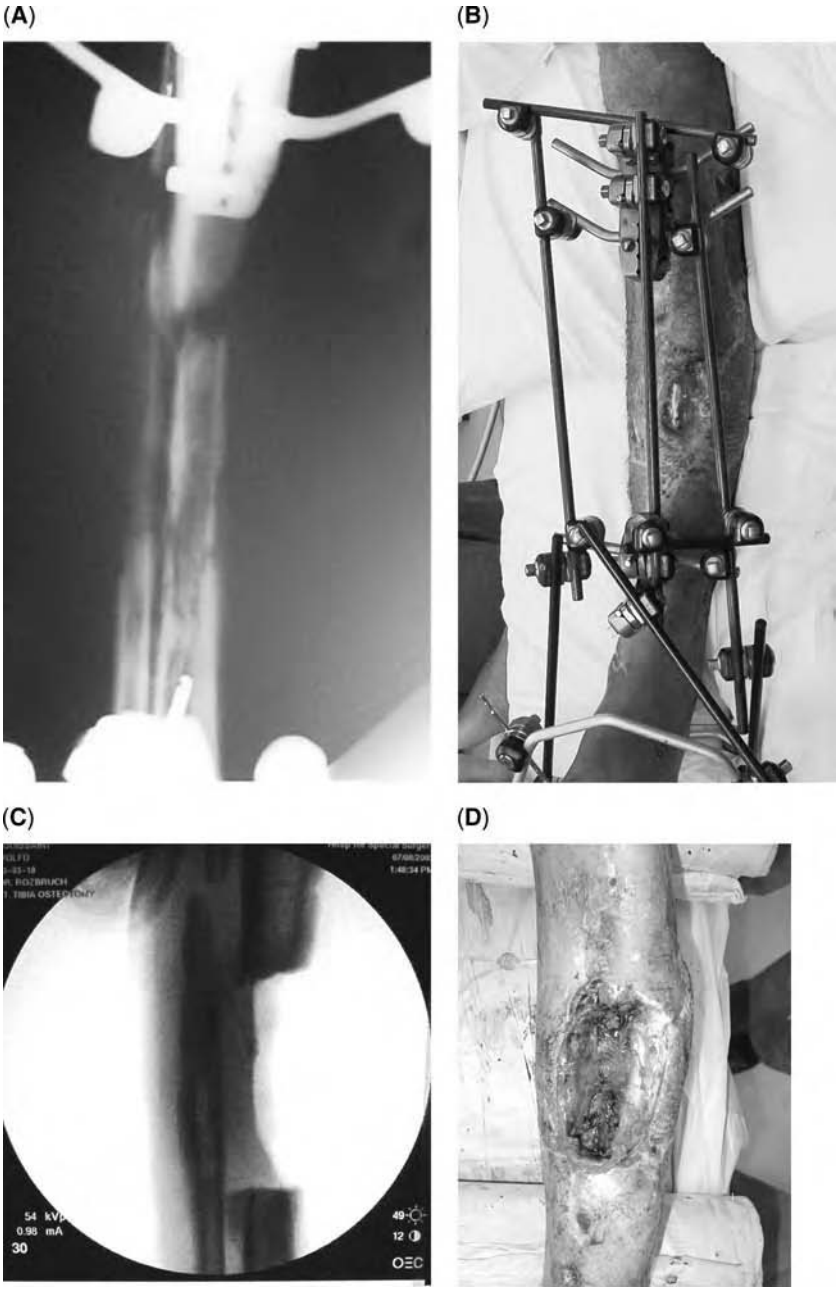

Figure 11 Clinical case example of trifocal bone transport. (**A**) Anteroposterior radiograph of an infected tibial nonunion one year following a pedestrian versus motor vehicle bumper crush injury, which was a Gustilo Anderson grade IIIC open fracture. (**B**) Clinical appearance of this infected tibial nonunion. Previous free flaps were performed and exposed desiccated bone was present. Subsequent operative cultures grew methicillin-resistant *Staphylococcus aureus*, *Pseudomonas aeruginosa*, and *Klebsiella pneumoniae*. (**C**) Lateral intraoperative radiograph following resection of dead, infected bone showing an 11 cm defect. (**D**) Intraoperative appearance of leg following wound debridement and bone resection showing a 13×8 cm soft-tissue defect. (**E**) Leg with Ilizarov/Taylor Spatial Frame in place. This frame has struts across the middle defect and rods across the proximal and distal tibial lengthening sites. There is a vacuum-assisted closure device covering the wound. There is extension of the frame across the ankle for treatment of an ankle equines contracture. (**F**) Interim appearance during gradual closure of the wound. Complete closure occurred after 23 weeks. (**G**) Standing side view at seven months postoperative. (**H** and **I**) Anteroposterior and lateral radiographs of the leg at seven months showing excellent alignment, closure of the defect, and partial bony healing of the docking site, and the proximal and distal tibial lengthening regenerate sites. (**J**) Standing front view three months following frame removal. Total time in frame was 53 weeks. (**K** and **L**) Anteroposterior and lateral radiographs of the leg at three months following frame removal. (Case supplied by S. Robert Rozbruch).

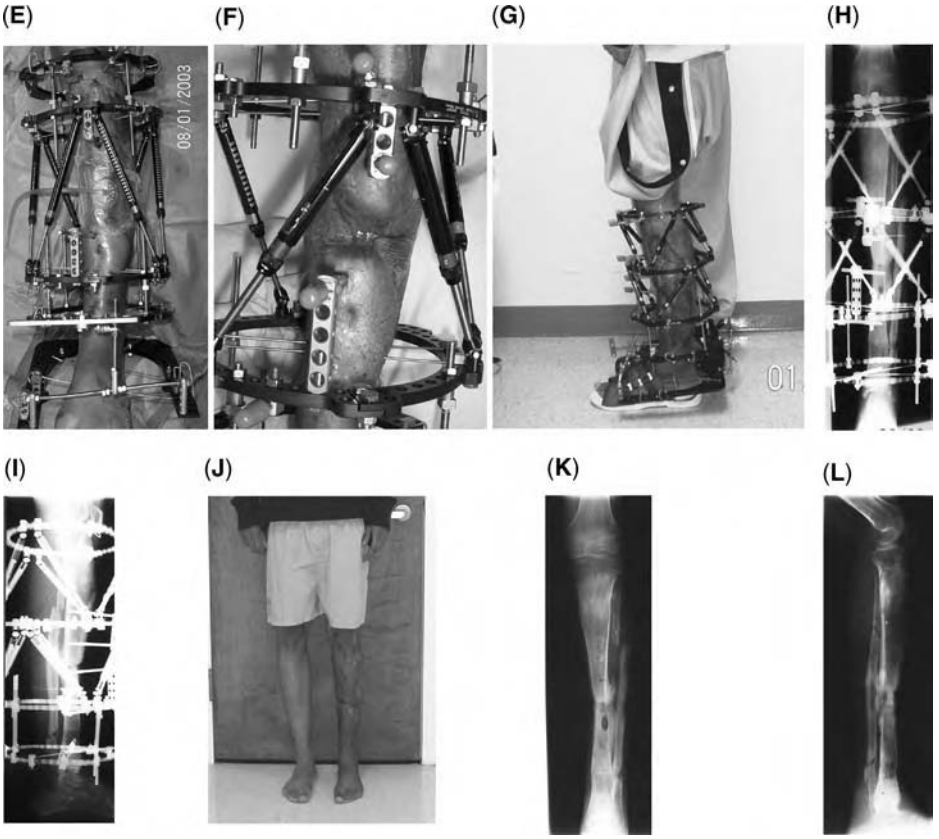

Figure 11 (*Continued*)

TRANSPORT MANAGEMENT

A latency period of 7 to 10 days following corticotomy is allowed prior to initiation of transport. Following corticotomy, auto distractors or threaded distractors are attached between the proximal fixation block and the transport ring. The distractors function as transport motors and the longitudinal threaded rods are utilized as a guide track (Fig. 4). The initial rate of distraction begins at approximately 0.25 to 0.5 mm per day. A slower distraction rate is initially undertaken because of the wide variability of the injury patterns and in the vascularity of the limb. In more extensive fractures or nonunions with a wide zone of injury, transport should be initiated very slowly. When regenerate bone is visualized at approximately two to three weeks postcorticotomy, the distraction rate can be modulated depending on the adequacy of the regenerate bone seen. In general, transport in the acute situation proceeds at a slower rate of 0.5 to 0.75 mm per day as opposed to the standard rate of 1 mm per day.

DOCKING SITE MANAGEMENT

Bone transport is continued until the antibiotic beads have been compressed to approximately the width of one bead. At this time, the patient is returned to surgery and the docking site exposed with removal of antibiotic beads. A high-speed burr is used to freshen the docking site and any irregular areas of bone-contoured flush to ensure maximal cortical contact and stability once docking occurs (Fig. 4). Autogenous iliac crest grafts, as well as numerous alloplastic and recombinant materials, have been used to augment and aid in the rapid consolidation of the docking site. Docking site augmentation has been shown to decrease the overall rate of nonunion and decrease frame time (2,4,7–9,13,15,17,22,23,25). Distal transport is continued within 24 hours of grafting, and the site is compressed once docking has occurred.

During the consolidation phase, compression is maintained at the docking site by compressing the transport ring 0.25 mm every other day until the docking site is radiographically

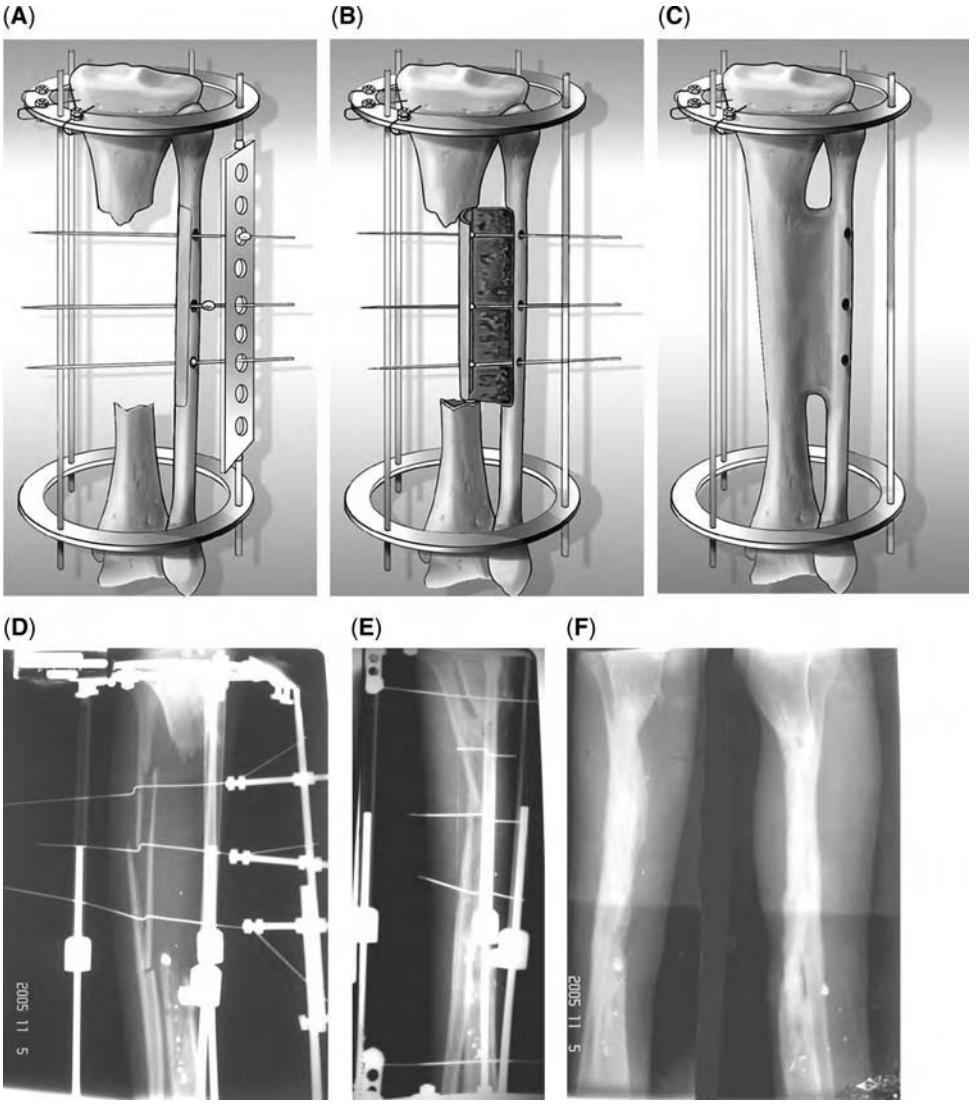

Figure 12 (**A**–**C**) Hemifibular transport using olive wires to transport the medial fibula. (**D**–**F**) Medial half of fibula transported with angled wires. Incomplete fibular corticotomy deformed intact fibula but still allowed complete transfer and docking.

healed. In cases of extremely long transports, the docking site usually heals long before consolidation of regenerate occurs. Frame removal should not occur until the regenerate has matured. Electrical stimulation, as well as ultrasound, has been used with encouraging results to help speed the consolidation of these very extensive regenerate segments (25). Frame removal requires the development of a neocortex, visualized on at least three of four cortices on the AP and lateral radiographs. Prior to frame removal, the frame is dynamized, allowing the transport rings to ''float'' on the longitudinal threaded guide rods.

COMPLICATIONS

The most common complication is nonunion of the docking site. Numerous authors have demonstrated many successful secondary procedures to manage docking site failure. Of prime importance is the period (time from frame removal to secondary procedure) prior to the undertaking of secondary procedures (6,13,22,23). It is crucial to ensure that contaminated pin tracts heal, and no additional pin site pathology is noted. Delay periods of at least one

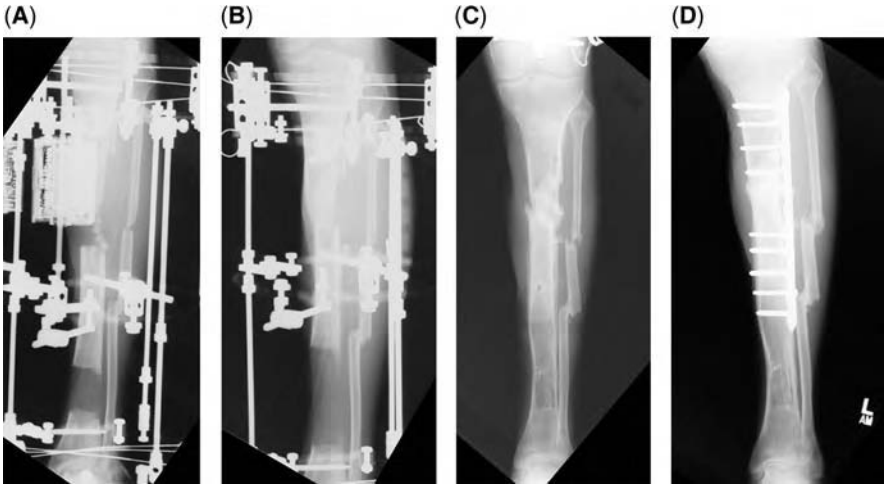

Figure 13 (**A**–**D**) Long retrograde transport under free flap. Despite grafting docking site, progressive nonunion of docking site resulted following frame removal. Nonunion was initially treated with an orthotic, and following a latency period of four months, open reduction internal fixation (ORIF) with bone morphogenic protein (BMP) augmentation resulted in complete healing.

month prior to revision IM nailing or plating should be strictly adhered to. Delayed procedures following frame removal have been shown to result in high rates of union and low rates of infection for the treatment of docking site nonunion (Fig. 13) (6,22,23).

Inadequate regenerate is a concern, especially in cases of extremely long transports. Late deformation and regenerate collapse can occur and the treatment of these complications is very problematic and thus it is best to avoid them at all costs. Modulation of the distraction rate aids in the development of a competent regenerate. Adjuvant modalities, such as ultrasound and electrical stimulation, also assist in the development of a viable regenerate (25). Thorough radiographic evaluation, including computed tomography scan, should be obtained if there is any doubt with regard to the adequacy of regenerate bone. In a few instances, autogenous grafting may assist in the consolidation of a marginal regenerate. One should adhere to the caveat that it is much easier to treat an inadequate docking site (nonunion) than it is to treat an inadequate regenerate (deformation, collapse, recurrent deformity, etc.).

FUTURE DIRECTIONS

Bone transport is a reliable and successful technique; however, it is laborious, requires extreme patient compliance, and has a relatively high rate of complications. With the advancement in external fixator technology, newer frame configurations have simplified the mechanics of frame mounting and transport. The Taylor Spatial Frame[â] (Smith & Nephew, Memphis, Tennessee, U.S.), other hexapod frames, as well as many monolateral transport constructs have devised less complex frame mountings that allow for simplistic application. These frames permit constant adjustment of the proposed docking site without the malalignment potential that

REVIEW OF LITERATURE

Authors	Journal; year	Title	Patient number	Results	Conclusions
Cattaneo R, Catagni M, Johnson EE (4)	Clin Orthop Relat Res 1992	The treatment of infected nonunions and segmental defects of the tibia by the methods of Ilizarov	28	Functional results were good to excellent in 21, fair in six, and poor in one	Hemicircumferential corticotomy and partial bone fragment internal transport useful for partial defects

(*Continued*)

REVIEW OF LITERATURE (*Continued*)

Authors	Journal; year	Title	Patient number	Results	Conclusions
Green SA, Jackson JM, Wall DM, Marinow H, Ishkanian J (23)	Clin Orthop Relat Res 1992	Management of segmental defects by the Ilizarov intercalary bone transport method	17	All but one patient eventually healed, five bone grafts at docking site, one at regenerate.	Complications: prolonged time in frame. Need to graft docking sites
Raschke MJ, Mann JW, Oedekoven G, Claudi BF (26)	Clin Orthop Relat Res 1992	Segmental transport after unreamed intramedullary (IM) nailing. Preliminary report of a ''Monorail'' system	20 patients; 13 tibia, 7 femur	Three patients. residual leg-length discrepancies, one hypertrophic nonunion, two deep infections,	Successful use of transport over IM rod decrease time in ex-fix frame
Marsh JL, Prokuski L, Biermann JS (10)	Clin Orthop Relat Res 1994	Chronic infected tibial nonunions with bone loss. Conventional techniques versus bone transport	25	Similar rates of healing; residual infection; treatment time; final angulation; complications and procedures	Unilateral transport device can be successful. Distraction techniques have lower rate of residual leg length discrepancy (LLD)
Lowenberg DW, Feibel RJ, Louie KW, Eshima I (15)	Clin Orthop Relat Res 1996	Combined muscle flap and Ilizarov reconstruction for bone and soft-tissue defects	36	Union and absence of infection were achieved in 35 of 36 patients	Combined approach provides competent biology for grafting of docking site and permits the accurate restoration of limb length
Polyzois D, Papachristou G, Kotsiopoulos K, Plessas S (21)	Acta Orthop Scand Suppl 1997	Treatment of tibial and femoral bone loss by distraction osteogenesis. Experience in 28 infected and 14 clean cases.	42	Infection eradicated in all patients with septic defects. four patients. required grafting at docking site	Radical debridement necessary; grafting of docking site reduces rates of nonunion
Paley D, Maar DC (17)	J Orthop Trauma 2000	Ilizarov bone transport treatment for tibial defects	19	12 excellent, six good, one poor	Extended treatment time, graft docking site
Song HR, Kale A, Park HB, et al.(1)	J Orthop Trauma 2003	Bone transport vs. vascularized fibula for femoral bone defects	37patients; 17 fibular graft, 20 transport pts	Improved outcome for transport patients	Complete debridement of infection required; bone grafting at the docking site
Mahaluxmivala J, Nadarajah R, Allen PW, Hill RA (16)	Injury 2005	Ilizarov external fixator: acute shortening and lengthening versus bone transport in the management of tibial nonunions	18	All healed. union at 12.1 mo shortening and lengthening, 17.2 mo Transport. 8 mo stabilization only	Transport group required grafting at docking sites; acute shortening of 4.6 cm
Beals RK, Bryant RE (7)	Clin Orthop Relat Res 2005	The treatment of chronic open osteomyelitis of the tibia in adults	30	Two patients residual drainage, one patient with aseptic nonunion	Advantages of circular frame, bone transport, bone graft, and long-term antibiotics

(*Continued*)

REVIEW OF LITERATURE (*Continued*)

Authors	Journal; year	Title	Patient number	Results	Conclusions
Rozbruch SR, Weitzman A, Watson JT, Freudigman P, Katz HV, Ilizarov S (27)	J Orthop Trauma. 2006	Simultaneous treatment of tibial bone and soft-tissue defects with the Ilizarov method	25	Bony union in 96%, time in frame 43 wk, lengthening 5.6 cm, LLD 1.2 cm, all wounds closed, no osteomyelitis, no amputations	This limb salvage method can be used without need for flap coverage; trifocal approach should be considered for large bone defects

can occur with traditional circular frame constructs. Development of HA (hydroxy-appetite) - coated pins and other substrate biomaterials have improved the pin/bone interface such that these frames may remain on patients for extended periods of time without pin loosening or infection, decreasing the need for rigorous pin care regimes. It is hoped that continued advancement in pin and frame design will overcome the current limitations of prolonged external fixation with respect to patient compliance managing complex frame adjustments. Advancements in IM designs may eventually decrease the need for external transport devices.

The emergence of orthobiologics holds great promise for large skeletal transports. The ability to augment large regenerate segments with percutaneously applied growth factor adjuvants to reduce prolonged consolidation times is an attractive alternative to spending 1.5 to 2 years in a transport device. Similarly, some of these same materials are currently being used to augment docking site union with early good results (8,13,15,17,23,25). Ideally, the ability to rapidly distract a transport segment with a simplistic fixator construct, followed by percutaneous application of docking and regenerate site enhancements to allow rapid consolidation and frame removal, is the ultimate goal.

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15 Osteomyelitis and Infected Nonunions

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INTRODUCTION

Osteomyelitis is a condition that has existed for thousands of years. Today, even with modern technology, the prevalence of osteomyelitis is increasing. Many patients are surviving life-threatening war injuries only to have a battle with a limb-threatening infection. The offending bacterial organisms, such as *Staphylococcus aureus* and *Enterococcus faecalis*, are becoming more difficult to treat as antibiotic-resistant bacterial strains evolve (1). The existence of antibiotic-resistant bacterial strains ensures that osteomyelitis will continue to be a challenging condition for limb reconstruction surgeons to treat. This chapter discusses the current classifications of osteomyelitis, the treatment options, and the surgical techniques that can be used to treat different stages of osteomyelitis and infected nonunions.

CLINICAL EVALUATION

During an initial clinical evaluation, the surgeon should obtain a thorough medical history of the patient and a history of the events that precipitated osteomyelitis. Medical problems, such as diabetes, rheumatoid arthritis, and immunocompromise, will affect wound healing and increase the patient's risk of infection (2). If the patient has a history of smoking, it should be noted in the initial evaluation report because it compromises the patient's ability to heal bone and soft tissue (3). Several other factors might compromise the host (4).

Systemic Factors	Local Factors
Malnutrition	Chronic lymphedema
Renal/liver failure	Venous stasis
Alcoholism	Major vessel compromise
Immunodeficiency	Arteritis
Chronic hypoxia	Extensive scarring
Malignancy	Radiation fibrosis
Extremes of age	
Steroid therapy	
Diabetes mellitus	
Tobacco abuse	

In 1985, Cierny and Mader developed a staging system for adult patients with osteomyelitis based on the anatomic type of osteomyelitis and physiologic class (4). It is called the University of Texas Medical Branch Staging System for Adult Osteomyelitis.

Anatomic Type

Type I. Medullary osteomyelitis
Type II. Superficial osteomyelitis
Type III. Localized osteomyelitis
Type IV. Diffuse osteomyelitis

Physiologic Class

A host. Good immune system and delivery
B host. Compromised locally (B^L) or systemically (B^S)
C host. Requires suppressive or no treatment; treatment worse than disease

Host factors often can be modified to improve the clinical stage, and this modification can positively influence the prognosis.

It is important to note previous incisions and to evaluate the blood supply during the clinical evaluation. If the pulse is not palpable or the blood flow to the limb is in question, an arterial duplex study should be conducted. Previous incisions often dictate the surgical approach, especially if the patient has undergone flap coverage of the bone or experienced extensive scarring of the limb (Fig. 1).

Draining sinuses are very important to note during clinical evaluation. The duration of time that a patient has experienced the draining sinus also should be noted. When located in the lower extremity, draining sinuses often are distal to the osteomyelitis, secondary to the forces of gravity. If the patient has experienced a draining sinus for an extended period of time, the patient will have an increased risk for squamous cell carcinoma (Fig. 2) (5). A biopsy should be obtained of all sinus tracts at the time of surgery. Often, the draining sinus is located in an undesirable location for the incision and does not need to dictate the surgical approach to the bone (Fig. 3). After the source of infection is eradicated, the sinus tract will close.

Limb deformities often accompany osteomyelitis and should not be overlooked when examining radiographs and performing a physical examination. A radiograph (erect-limb radiograph) that is obtained by using teleoroentgenography can be examined to assess for limb length discrepancies and deformities. Occasionally, a single surgical solution can be used to treat both problems (e.g., bone transport used to treat segmental osteomyelitis and to restore the overall length of a shortened limb).

If any question remains regarding the extent of bone involvement shown by the initial radiographs, the patient can undergo magnetic resonance imaging (MRI) with gadolinium contrast enhancement. However, the surgeon must interpret MRI findings with caution because reactive edema in the bone can exaggerate the extent of the infection in the bone (6). Computed tomographic scans can be helpful in determining the presence of an infected nonunion.

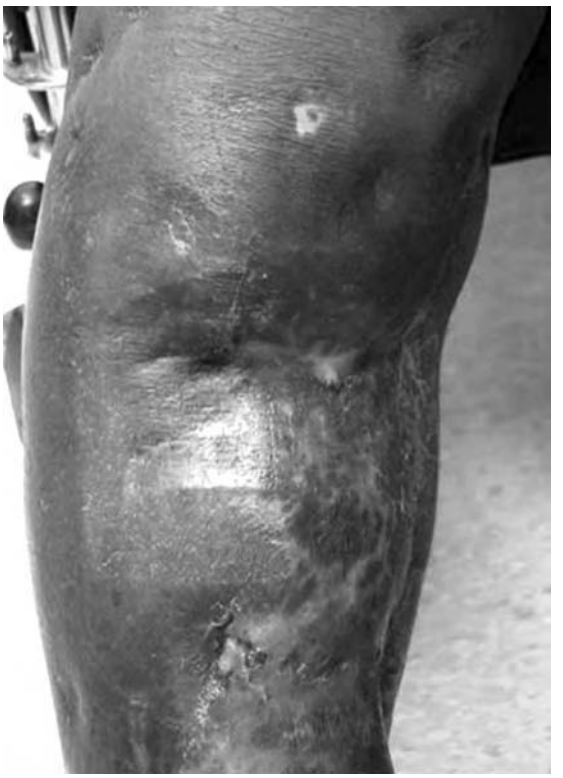

Figure 1 Photograph shows a patient with diabetes who has a poor vascular supply. Severe soft-tissue scarring is visible over the distal tibia.

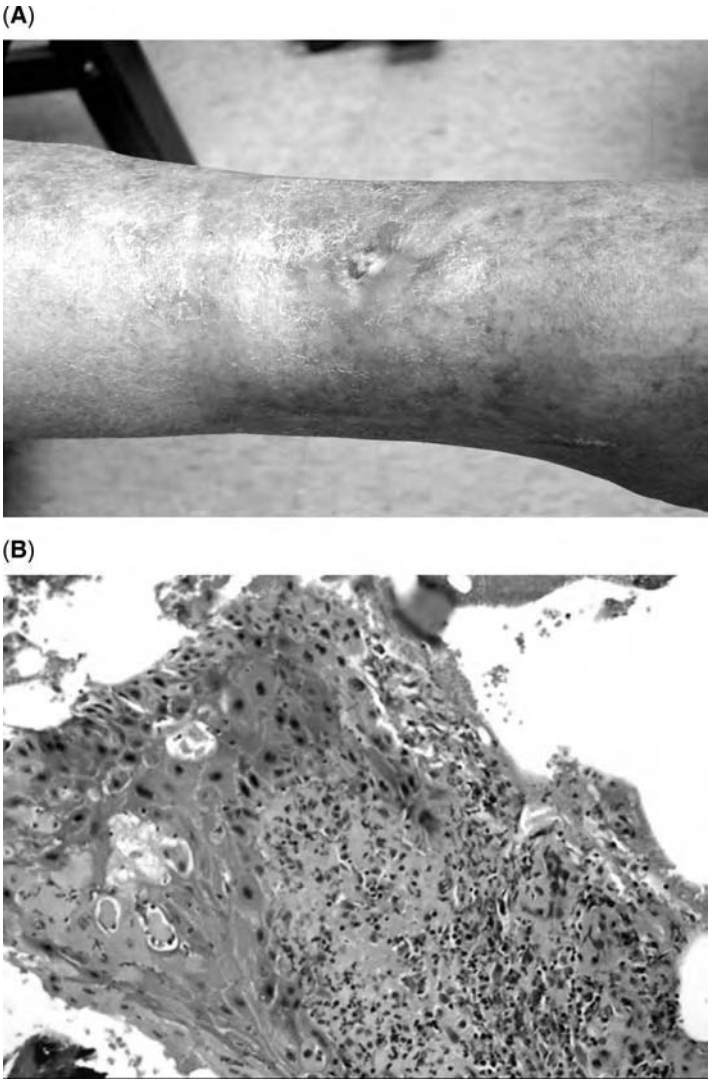

Figure 2 (**A**) Clinical photograph of a patient with a 50-year-old draining sinus tract. (**B**) Pseudoepithelial hyperplasia, a precursor to full-blown squamous cell carcinoma, biopsied from the 50-year-old draining sinus tract.

CLASSIFICATION

The Cierny–Mader Staging System for Osteomyelitis is listed in the clinical evaluation section. Patients with anatomic types I and II osteomyelitis experience inherent bone stability and do not need to be treated with fixation. Patients with type III osteomyelitis are at risk of fracture after debridement and often require supplemental stabilization, such as external fixation. Patients with type IV osteomyelitis experience unstable bones that always require fixation. The treatment guidelines are discussed in the next section.

TREATMENT OPTIONS

Treatment options are based primarily on the Cierny–Mader Staging System for Osteomyelitis. Regardless of the stage of osteomyelitis, debridement of bone is a necessary surgical procedure. After undergoing debridement, the patient must also be willing to undergo the necessary surgical reconstruction of the limb. Some patients or caregivers are unable to handle the often rigorous aftercare associated with circular external fixation or the extended periods of nonweight bearing. Knowing the patient is the key to choosing the surgical reconstruction that is

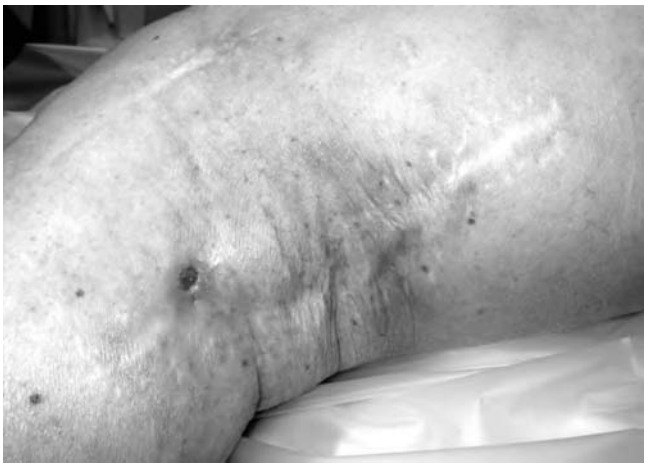

Figure 3 Clinical photograph of a dependent draining sinus that was located at the level of the popliteal fossa of a patient with femoral osteomyelitis.

best in each case (7). At our center, all patients are treated by a team of physicians that includes an internist and an infectious disease specialist. The team approach ensures that the patient is medically optimized and receives the appropriate antibiotic therapy postoperatively with blood level monitoring.

Treatment principles include debridement of all nonviable infected tissue, dead space management, local antibiotic delivery, stabilization of the limb, and future limb reconstruction.

SURGICAL TECHNIQUES

Surgical treatment of osteomyelitis is based on a few basic principles. Biopsy cultures and a biopsy specimen are pathologically examined. All necrotic and nonbleeding bone must undergo debridement. A high-speed burr is used to debride the bone, and continuous cooling irrigation is applied to the burr to decrease the thermal injury. When possible, the debridement is performed with the tourniquet down to allow for the best assessment of bleeding bone. The tibial cortex experiences very little bleeding; therefore, the surgeon must look for the ''paprika'' sign. The Versajet hydrosurgery system (Smith & Nephew, Memphis, Tennessee, U.S.A.) is a helpful tool to use during soft-tissue debridement. It is a hydroscalpel that uses water to cut a fine layer of tissue and vacuum suction to remove the debris (Fig. 4). After all necrotic bone and soft tissue are removed, the wound is irrigated with as much as 9 L of saline (based on the size of the wound). The debridement should be performed by using one set of instruments. After irrigation, all gowns, gloves, and drapes are replaced and clean instruments are used to complete the surgical procedure. The different reconstruction options are outlined in the next section.

All patients are treated with six weeks of antibiotics that are specific to the cultured organism. After the C-reactive protein and erythrocyte sedimentation rate have normalized and the patient is no longer receiving antibiotic treatment, iliac crest bone graft is applied

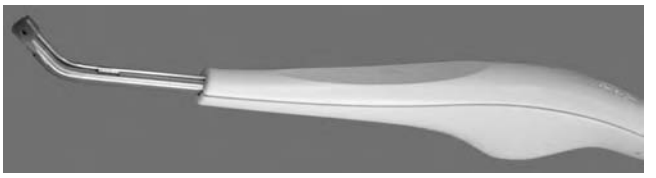

Figure 4 Photograph of a Versajet hydrosurgery system (Smith & Nephew, Memphis, Tennessee, U.S.A.). A fine stream of water at high pressure is used to cut through and debride soft tissue. A suction mechanism on the wand removes the cut tissue, as it is debrided.

to the affected limb during a second surgical procedure (8). For additional protection from infection, separate instruments are used for the bone graft harvest. Many supplementary bone morphogenic proteins (BMPs), such as BMP-7 and BMP-2, can be used to supplement the bone graft. Postoperatively, all patients continue to use an external bone growth stimulator.

Treatment Strategies and Surgical Technique Based on Cierny–Mader Stages

Stage I osteomyelitis is located within the medullary canal (Table 1). This stage often is associated with a draining sinus that has been observed in a clinical setting. If a limb deformity is present, a Palacos cement chest tube rod can be used to deliver high doses of antibiotics to a local setting (Fig. 5) (9). Reamers are used to debride the bone in a sequential manner; the canal is reamed to a diameter that is at least 2 mm larger than the 10 mm diameter of the chest tube rod. A thin canal irrigator is used to irrigate the canal. The antibiotic chest tube rod is inserted into the canal, and a loop of guidewire is left prominent for easy removal. The rod is removed 8 weeks after surgery.

In cases of stage I osteomyelitis with associated deformity, debridement can be accomplished from a cortical window. The size of the cortical window must be less than one-third the diameter of the bone to maintain structural integrity (Fig. 6) (10). A high-speed burr is used to create the window and continuous irrigation is applied. After all the necrotic bone and debris are removed from the canal, antibiotic beads on a wire are packed into the medullary space. Great care is taken not to pack the beads outside of the medullary canal because a dead space will be created in the soft tissue. An alternative to the traditional antibiotic cement beads is calcium triphosphate or calcium sulfate that has been mixed with high concentrations of antibiotics. This alternative circumvents the need for a second operation to remove the cement beads because the calcium triphosphate and calcium sulfate are absorbed by the body. McKee et al. (11) reported a series of bone voids that occurred as a result of osteomyelitis (average volume, 30.5 cc). The average follow-up was 28 months, and 23 of 25 patients experienced eradication of infection. However, eight of the 23 patients experienced a draining sinus that healed two to three months postoperatively. Conway conducted a study of 12 patients with a mean defect volume of 30.2 cc (unpublished results). Calcium triphosphate was used as an absorbable antibiotic depot when treating these patients. Eleven patients experienced eradication of infection at a minimum follow-up duration of nine months (average follow-up duration, one year). Draining sinuses were not observed postoperatively in this series (Fig. 7). Recipes for antibiotic concentrations are listed below:

40 g of Palacos cement + 3.6 g of tobramycin + 1 g of vancomycin
40 cc of calcium triphosphate + 3.6 g of tobramycin + 1 g of vancomycin
OsteoSet-T: Calcium sulfate pellets with 4% tobramycin sulfate

Although the author prefers the use of calcium triphosphate for an absorbable antibiotic depot, using this technique is not recommended for extremely large or uncontained defects, such as those observed in cases of stage III osteomyelitis.

Stage II osteomyelitis, in which the osteomyelitis affects only the cortical surface of the bone, is located in stage IV pressure ulcers (i.e., ischium and sacrum) and the anterior tibia. This stage of osteomyelitis is treated with aggressive debridement and good coverage of soft tissue (Fig. 8). The Vacuum Assisted Closure Advanced Therapy System (Kinetic Concepts, Inc., San Antonio, Tennessee, U.S.A.) is effective for granulation tissue coverage. The thin tissue covering the bone, even with epithelial coverage, is very friable and can break down with minimal trauma. Depending on the location, a gastrocnemius, soleus, or free flap might be necessary.

Stage III osteomyelitis involves the cortical bone and the medullary canal. Aggressive debridement of all necrotic bone often can cause the remaining bone to lack adequate structural stability. The intramedullary dead space is filled with antibiotic cement, and the bone is stabilized. Bone grafting is performed during a second procedure (at a minimum of eight weeks after index surgery and the completion of antibiotic therapy) to enhance bone stability. An absorbable antibiotic depot, such as calcium triphosphate or calcium sulfate, is not used to treat stage III osteomyelitis. In the author's experience, absorbable antibiotic depots have not been particularly successful in treating large, ''uncontained'' defects that remain after debridement. Options that can be used to create bone stability include applying

Table 1 Strategies for Treating Infected Nonunion

Location	Amount of Bone Loss	Strategy
Diaphysis of femur	>5 cm	**Strategy 1** *First stage*: Resection is performed and external fixation applied. Transport is then performed over custom intramedullary rod *(use with caution—recurrent infection is main risk)*. If bone loss is >10 cm, consider double-level transport *Second stage*: Bone graft is applied to docking site **Strategy 2** *First stage*: Resection is performed and external fixation applied. Intramedullary rod with cement spacer is then inserted *Second stage*: Cement spacer is removed and allograft and bone graft inserted *(use with caution—recurrent infection is main risk)*
	<5 cm	*First stage*: Acute shortening is performed and limb is fixated with intramedullary rod (antibiotic coating optional) *Second stage*: Bone grafting is performed *Third stage*: Lengthening over intramedullary nail is performed or intramedullary skeletal kinetic distractor inserted
Diaphysis of tibia	>10 cm	*First stage*: Resection is performed and external fixation applied. Double-level bone transport or ipsilateral fibular transport is performed *Second stage*: Bone graft is applied to docking sites
	4–10 cm	*First stage*: Resection is performed and external fixation applied. Bone transport is performed with circular external fixation (optional lengthening over nail) *Second stage*: Bone graft is applied to docking site
	<4 cm	**Strategy 1** *First stage*: Acute shortening is performed and limb is fixated with antibiotic-coated intramedullary rod *Second stage*: Bone grafting is performed (optional: proximal tibial corticotomy and lengthening over nail or intramedullary skeletal distractor can be inserted after docking site heals) **Strategy 2** *First stage*: Acute shortening is performed and circular external fixation applied. Proximal corticotomy is performed for lengthening *Second stage*: Bone graft is applied to docking site
Diaphysis of humerus	>10 cm	*First stage*: Resection is performed and external fixation applied. Intramedullary rod with cement spacer is then inserted *Second stage*: Reconstruction is performed by using intramedullary rod and allograft or by applying bone graft to vascularized fibula
	5–10 cm	*First stage*: Resection and bone transport are performed with monolateral fixator or Ilizarov apparatus *Second stage*: Bone graft is applied to docking site
	<5 cm	*First stage*: Acute shortening is performed. External fixation is applied or antibiotic-coated intramedullary rod inserted *Second stage*: Bone graft is applied
Intra-articular region	Any amount of bone loss	*First stage*: Joint is salvaged with stable internal fixation. External fixation can be applied across joint. Fusion might be performed if attempts to salvage joint fail

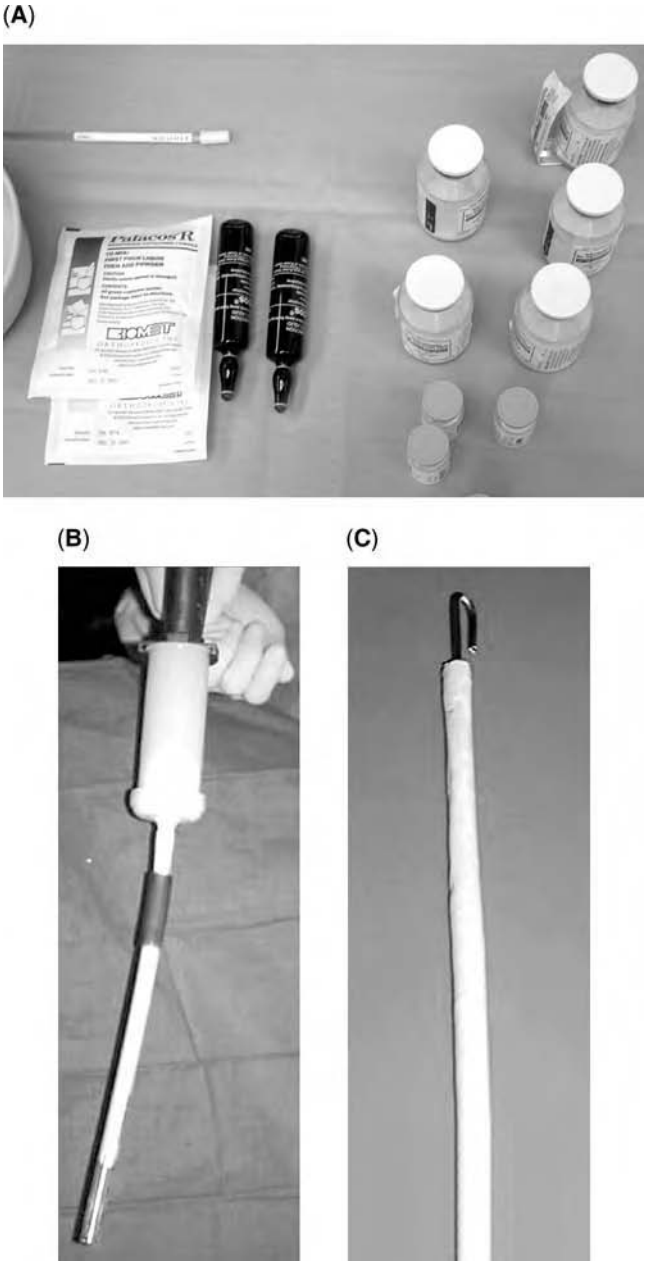

Figure 5 (**A**) Supplies needed to make the antibiotic chest tube rod are shown and include Palacos cement, a 3-mm ball-tipped guidewire, 3.6 g of tobramycin, and 1 g of vancomycin per 40-g package of cement, an extra monomer for mixing the antibiotics and cement powder, and a 40-French chest tube (used to make a 10-mm diameter rod). The cement gun that is used to insert the wet cement into the chest tube is not shown. (**B**) Plastic chest tube is cut off the cement rod before the plastic melts. (**C**) Finished antibiotic chest tube has a curved hook at the inserting end for easy removal.

a circular external fixator or inserting an intramedullary, antibiotic-coated locked rod. An external fixator should be used to stabilize any distal tibial defects. The foot should be included in the construct to achieve maximum stability for bone and soft-tissue healing (Fig. 9). The use of an antibiotic-coated intramedullary rod is an excellent technique to stabilize midshaft defects. It is made from a 10-mm diameter nail that is inserted into a mold and coated with an antibiotic-loaded Palacos cement rod (Fig. 10). The mold allows for a 1-mm circumferential cement mantle and a 2-mm increase in the diameter of the rod. The canal is over-reamed 1 mm more than the coated rod diameter. The intramedullary rod provides antibiotic delivery and bone stability. The author has successfully used an antibiotic-coated intramedullary rod to treat 20 cases of infected nonunions. One complication of using the intramedullary rod is debonding between the cement and rod interface. This debonding has occurred with removal in four cases and during insertion in one case (12).

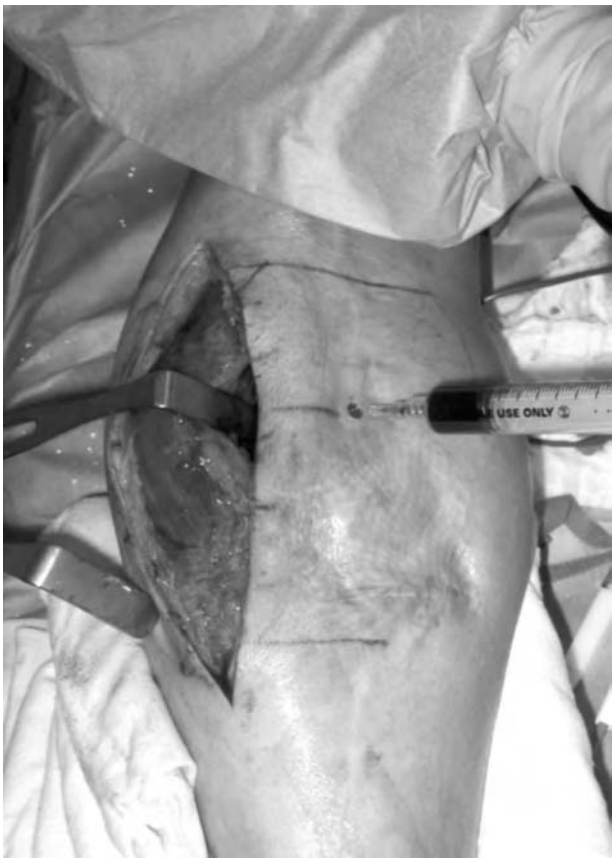

Figure 6 Clinical photograph of a patient with stage I tibial osteomyelitis that is not amenable to a chest tube rod secondary to proximal tibial varus deformity. A cortical window was made in the lateral cortex of the tibia by using a 5-mm burr.

Stage IV osteomyelitis is the most challenging type of bone infection to treat and includes the infected nonunion. Two problems are associated with infected nonunion: the lack of bone stability and the infected bone. These problems often are associated with soft-tissue compromise and usually occur as a result of open fracture. Many options exist to treat infected nonunion. The options are based on the location of the nonunion and the quality of the host. Many of the treatment strategies for segmental infected nonunion of the tibia or femur include resection and bone transport with a circular external fixator, resection and insertion of a cement spacer stabilized by an intramedullary rod, resection and transport over a rod, resection and reconstruction with allograft bone, or, in some cases, amputation. These options differ when treating an intra-articular infected nonunion. The next section outlines the treatment strategy and rationale for treating nonunions.

INFECTED NONUNION OF THE DIAPHYSIS

Diaphysis of the Femur

Preoperative planning for femoral cases is essential because the reconstruction is based on the amount of bone resected. It is wise to prepare for the worst case scenario based on the results of preoperative studies. In such cases, depending on the type of host, many treatment options are available. The femur is made up of a large soft-tissue envelope that is very accommodating with respect to bone coverage but not so amenable to long-term external fixation pins. Patients with obesity have a large soft-tissue envelope and often have low tolerance for external fixator pins. In such cases, external fixation would provide less stability than an intramedullary device secondary to the distance of the frame from the bone needed to accommodate the soft tissues. Patients with obesity would better tolerate bone transport over an intramedullary rod or, in cases of substantial bone loss, allograft. If a patient is treated with bone transport over an intramedullary rod, a custom rod can be used to capture the transported segment after the transport is complete. Immediate insertion of an intramedullary

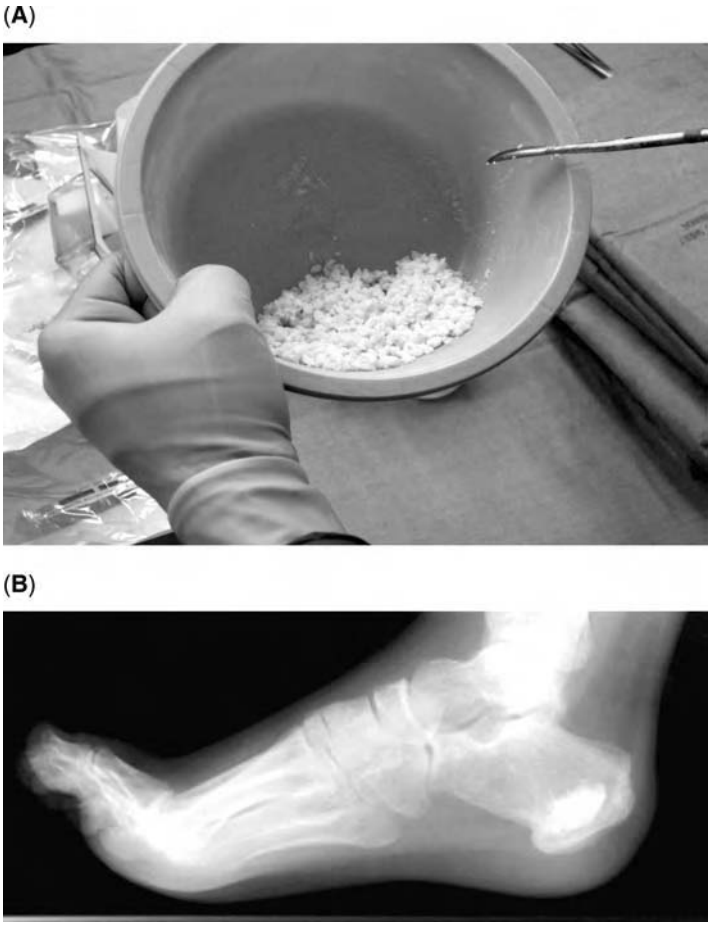

Figure 7 (**A**) Clinical photograph of calcium triphosphate that has been mixed with antibiotic powder before inserting it into a contained osteomyelitis defect. (**B**) Lateral view radiograph shows that the calcium triphosphate completely fills the calcaneal defect after osteomyelitis debridement has been performed.

rod at the time of osteomyelitis resection should be performed with caution. The author prefers to use the antibiotic-coated locked rod in such cases.

Compared with the tibia, the femur is much more accepting of acute shortening when a diaphyseal resection of 5 cm or less is performed. In such cases, the author prefers acute shortening and fixation with an intramedullary rod because of the initiation of immediate bone contact for healing. Bone grafting of the docking site is performed eight weeks after the index surgery and two weeks after the completion of a course of antibiotics. Performing resection and acute shortening of more than 5 cm in the femur, however, can result in severe and occasionally permanent extensor weakness (13). The permanence of the weakness depends in part on the location of the acute shortening. The closer the acute shortening is to the knee, the greater is the effect it will have on the extensor mechanism. A 5-cm lengthening can be

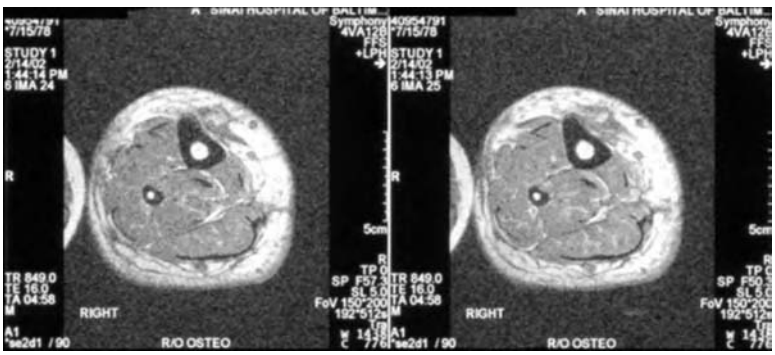

Figure 8 Magnetic resonance imagining findings show isolated, superficial cortical osteomyelitis involvement (as seen in Cierny–Mader stage II).

Figure 9 Clinical photograph of a patient with distal tibial infected nonunion. Note that the foot is included in the frame to increase the stability of the distal fragment.

performed at a later date. The timing of the lengthening can vary based on the severity of the infection. The author prefers to perform lengthening after the active infection is eradicated, often at the same surgical setting as the bone graft to the docking site; the procedure can be performed by lengthening over an intramedullary nail. After the docking site is completely healed, the patient could undergo a procedure to exchange the nail for an intramedullary skeletal kinetic distractor (Orthofix, McKinney, Tennessee, U.S.A.).

Ten-centimeter defects can be treated with double-level transport of the femur over a custom intramedullary rod. The custom rod should have a screw hole to lock the transported

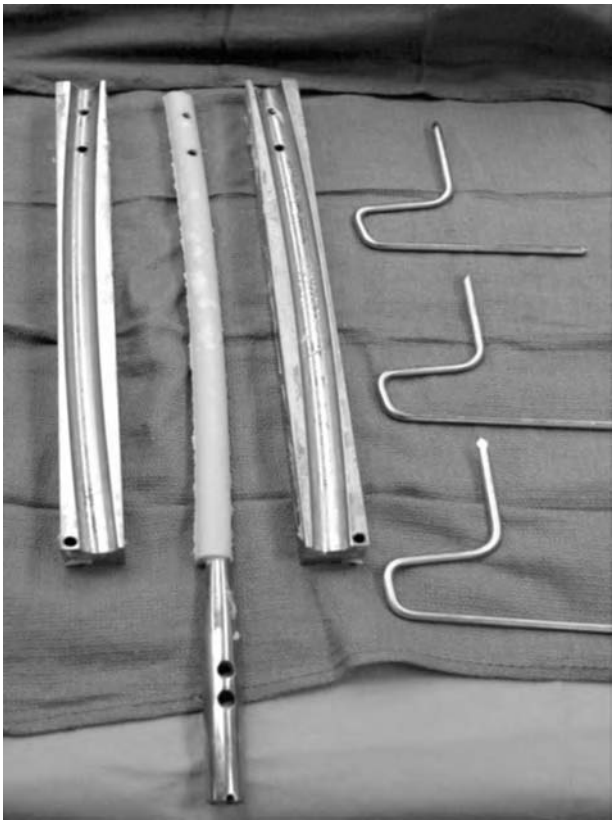

Figure 10 Clinical photograph of an antibiotic-coated rod and the mold used to form the rod coating.

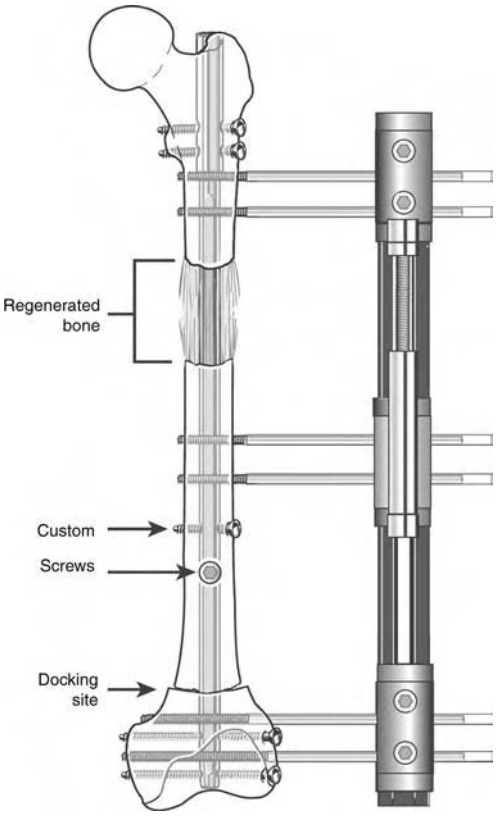

Figure 11 Illustration of a femoral rod bone transport frame for transporting over a rod. Note the custom screw holes that must be created in the rod before insertion. The screw holes are used for locking the transported segment. Two screws should be used to secure the transported segment.

segments in place (Fig. 11). The docking site is grafted with bone when the frame is locked and the fixator is removed. Performing bone transport by using distraction osteogenesis is a valuable technique that is equally effective in B and A hosts (14). The use of bone transport over a nail in the setting of an infected nonunion can be risky with recurrent infection as the major risk. This technique should be used with caution.

Large segmental defects can be difficult to treat with bone transport and can necessitate that a frame remain in place for more than two years. Allograft reconstruction is an alternative method that can be used to treat large segmental defects. A staged procedure is performed to resect and stabilize the bone ends. A femoral rod is then inserted through the allograft using cement, with bone graft along the allograft surface and ends. Cementing the medullary canal of the allograft around the intramedullary rod decreases the risk of allograft failure and infection (Fig. 12) (15).

Diaphysis of the Tibia

Infected nonunions often are located in the tibia. Bone transport is a useful technique that can be used to treat the majority of diaphyseal segmental defects of the tibia, which range in size from 5 to 10 cm. A resection of the infected nonunion or the segmental osteomyelitis can be performed at the same surgical setting as the osteotomy and bone transport as long as the surgeon uses two separate sets of operative instruments and preps the limb a second time. For distal to proximal transport, the distal tibial fixation is small and the foot needs to be included in the frame to allow for adequate healing of regenerated bone. In some cases, a double-level transport is necessary to treat large defects. The level of the osteotomy should be performed as close to the metaphyseal area as possible to have the best potential for healing of regenerated bone. Distraction is begun on postoperative day 7 with the use of a circular external fixator (one-quarter turn, four times per day). Great care must be taken postoperatively to maintain good knee and ankle range of motion. In some cases, administering Botulinum toxin can help the patient perform exercises during physical therapy that will prevent contracture of the knee and ankle (16). After the bone is positioned 1 cm from the docking site, bone graft is added to

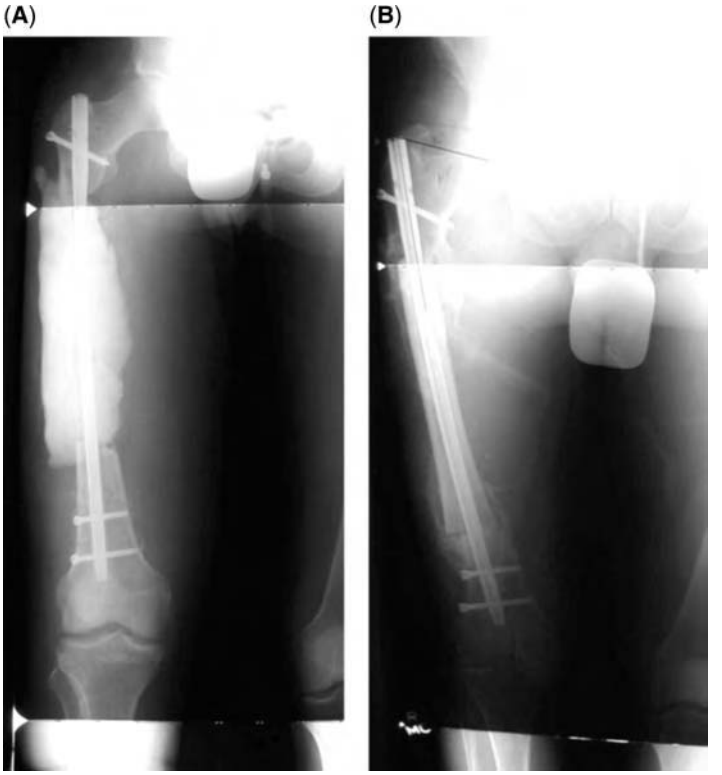

Figure 12 (**A**) Anteroposterior view radiograph of a patient with 24-cm segmental osteomyelitis of the femur. The patient was treated with complete resection and insertion of a cement spacer. (**B**) Anteroposterior view radiograph shows patient after reconstruction was performed with a cemented allograft.

the docking site to aid in healing. The bone ends are resected to clean the bleeding bone, and the frame is used for additional compression of the docking site. When possible, the fibula can be used to help create a synostosis and will allow for greater surface area to exist at the docking site. Currently, the author uses traditional iliac crest bone graft at the docking site, occasionally supplemented with allograft cancellous chips or a concentrated bone-morphogenic protein.

For smaller diaphyseal defects (less than 4 cm), performing acute shortening can promote faster healing and fewer complications at the docking site than performing bone transport (17). Acute shortening is most commonly performed in the distal tibia. Often, after distal acute shortening, the patient experiences a large soft-tissue envelope around the ankle. This does not offer good cosmesis and can make it difficult to wear shoes that are not customized. Doppler ultrasonography is used intraoperatively to detect any occlusion of the arterial blood flow. The best approach for acute shortening is a transverse incision, which will allow for excellent wound closure. This acute shortening can be combined with a proximal corticotomy to restore length by using a circular external fixator at the same surgical setting. When performing acute shortening, the surgeon must always check the pulse and inform the patient that the limb will have poor cosmesis.

Two techniques are also helpful to treat extremely large diaphyseal defects: transport of the ipsilateral fibula and vascularized free fibular transfer. Both techniques require bone graft to be placed proximally and distally on the docking sites and bracing of the limb after the frame is removed until the fibula experiences hypertrophy. Bracing is no longer required after the fibula has experienced hypertrophy for one to two years. The advantage of performing a transfer of the ipsilateral fibula (also referred to as the *fibula pro tibia procedure* (18)) is that it does not require the skills of a surgeon specializing in microvascular surgery (Fig. 13). Cases that have been treated with free vascularized fibular transfer have been reported in the literature and have experienced successful results. Yajima et al. (19), in 1993, reported success in 30 of 33 cases of vascularized fibular grafts that were used to treat lower extremity osteomyelitis and bone defects. The complications included acute ischemia, thrombosed anastomoses, and fibular stress fractures.

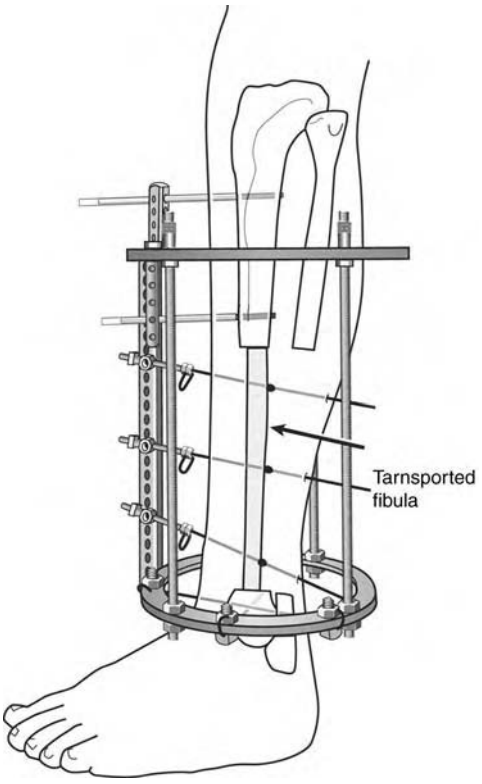

Figure 13 Illustration shows the frame construct for an ipsilateral fibular transfer.

Diaphysis of the Humerus

Small (less than 5 cm) defects can be acutely shortened in the humerus without the patient experiencing any functional deficits. The humerus has this advantage secondary to not being a weight-bearing bone. Intraoperative positioning is critical for this type of case. Our center uses a radiolucent diving board that is placed at the head of the bed. The C-arm is inverted and used as a table. The author prefers to position the C-arm parallel to the bed. A bump is placed under the scapula. To perform a diaphyseal resection, the surgeon can use an anterolateral approach or a posterior triceps splitting approach. Maximum bone contact is essential. In cases that include active infection, acute shortening can be performed with the use of a monolateral or circular external fixator (Fig. 14). Postoperatively, active compression is applied at the rate of 1 mm/day for seven days. The rate can be varied depending on the amount of bone contact. Another option for large segmental defects is performing allograft reconstruction or vascularized fibular transfer after the infection is eradicated. Vascularized fibular transport must be performed by a skilled microvascular surgeon. The humerus requires one to two years to experience hypertrophy, and the procedure is associated with occasional donor site morbidity. The author has had no experience performing vascularized fibular grafts in the humerus. Jupiter (20) reported a series of four atrophic nonunions of the humerus that were treated with vascularized fibular graft; all four nonunions healed.

INFECTED INTRA-ARTICULAR NONUNIONS

Infected intra-articular nonunions are difficult to treat because they often lack a large segment of bone for fixation. The intra-articular fragments can be very small and often have hardware attached to them. Infected intra-articular nonunions are treated with the same basic principles that are used to treat all other infections. When possible, the joint surface should be inspected and irrigated. A postoperative drain should be left in place. If any hardware is present in the limb and still has good purchase, it should not be removed. Often, the bone fragments are small and attempts to remove good fixation can cause the bone to resemble ''swiss cheese.''

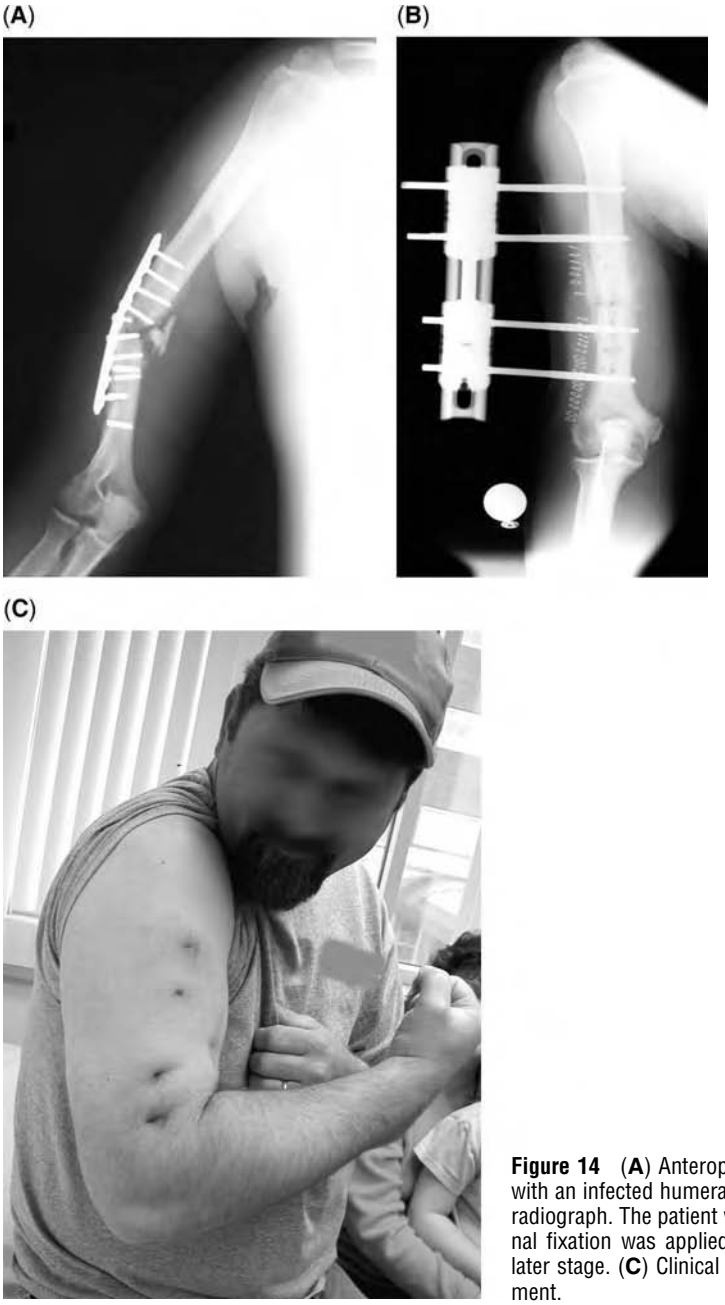

Figure 14 (**A**) Anteroposterior view radiograph of a patient with an infected humeral nonunion. (**B**) Anteroposterior view radiograph. The patient was treated with resection, and external fixation was applied. Bone grafting was performed at a later stage. (**C**) Clinical photograph of the patient after treatment.

Support for the intra-articular fragments can be supplemented with circular wire external fixation that is used in a static mode (Fig. 15). Large defects in the bone or poor purchase of fixation can be supplemented with antibiotic-impregnated bone cement. Postoperatively, range of motion is initiated but no weight bearing is allowed for at least six weeks. In some cases, joint preservation is not possible and arthrodesis, resection of the infected nonunion, and delayed bone grafting must be performed.

COMPLICATIONS

In the author's experience, recurrence of infection is the most common complication that patients experience after undergoing treatment for osteomyelitis. This complication occurs

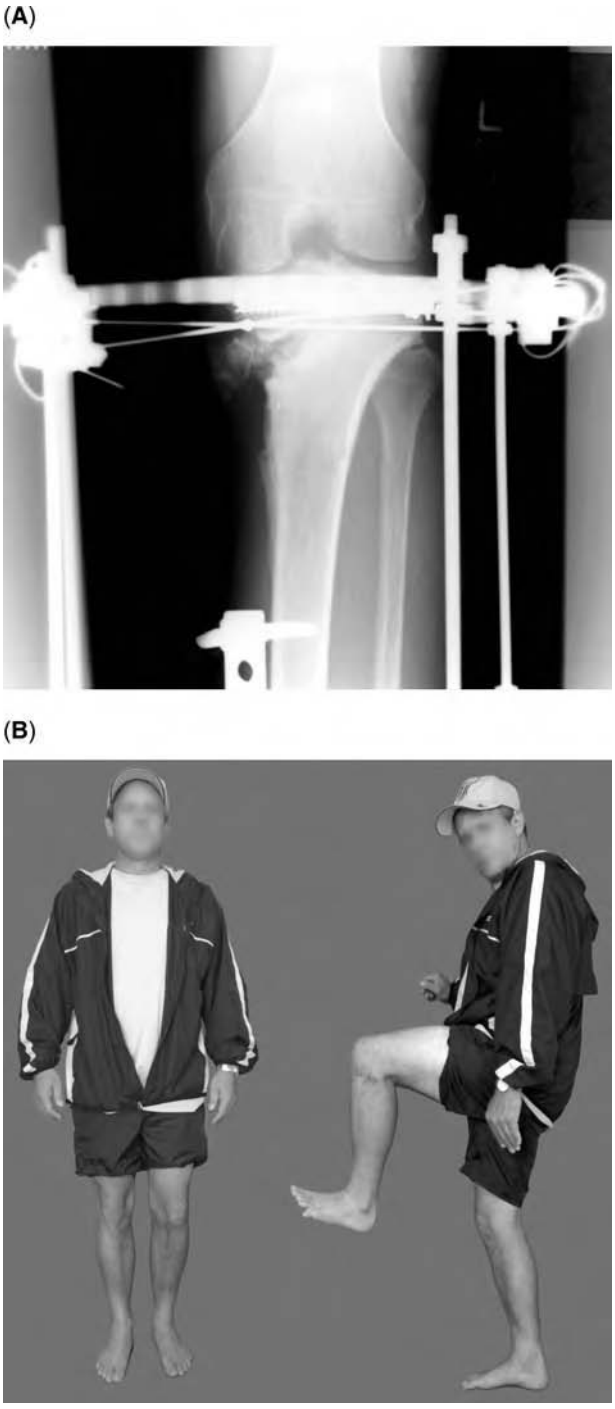

Figure 15 (**A**) Anteroposterior view radiograph of a patient with an infected intra-articular tibial plateau fracture that occurred because of a failed high tibial osteotomy. The radiograph was obtained after the patient had undergone bone grafting. (**B**) Clinical photograph of the patient after completing treatment.

frequently because of inadequate debridement. With increasing surgeon experience, this complication decreases. It is frequently said that osteomyelitis cannot be cured. However, the patient can expect to experience a long, infection-free interval or remission after undergoing segmental resection. Bacteria, however, are becoming increasingly antibiotic resistant, and methicillin-resistant *S. Aureus* has been shown to hibernate intracellularly (21). Recurrence of infection could occur if the patient experiences a change in immune status.

In the author's experience, problems with wound healing and bone healing are the second most common complications experienced by patients after undergoing treatment for osteomyelitis. The majority of patients with osteomyelitis are B hosts, which involves greater susceptibility to wound healing difficulties and delayed bone healing. Delayed bone healing can be avoided by using bone graft and supplementary external bone stimulation. The orthopedic surgeon should also consult a plastic surgeon for all wound healing or wound coverage difficulties.

FUTURE DIRECTIONS

Research focusing on the communication mechanisms of bacteria is currently underway. It is an exciting branch of research that can potentially allow us to turn off the mechanisms that cause bacteria to switch from a dormant state to an active state. If we can control the bacterial signals, we can potentially prevent additional radical surgery and recurrence of infection (22).

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Part V FOOT AND ANKLE

16 Charcot Neuroarthropathy of the Foot and Ankle

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INTRODUCTION

Jean Martin Charcot, in 1883, first described Charcot neuroarthropathy as occurring in patients with tabes dorsalis. Charcot neuroarthropathy was not known to be associated with diabetes until William Reily Jordan noted the connection in 1936 (1). Although two-thirds of patients with Charcot neuroarthropathy have type 2 diabetes, many other causes have been reported [e.g., syphilis (i.e., tabes dorsalis), leprosy, alcoholism, peripheral nerve injuries, spina bifida, folate deficiency (e.g., pernicious anemia), and spinal cord lesions (e.g., syringomyelia, post-traumatic cord injury, meningomyelocele, and disk herniation)] (2).

Charcot neuroarthropathy is a peripheral and autonomic neuropathy that typically presents as a hyperemic event (e.g., a red, swollen foot). The diagnosis of Charcot neuroarthropathy is made clinically after observing a lack of normal foot sensation, presence of ulcerations, and presence of foot deformity because of joint subluxations/dislocations. Many factors contribute to the destruction of the bone and joint in patients with Charcot neuroarthropathy. These factors include loss of protective sensation (peripheral neuropathy), autonomic neuropathy, weight-bearing stress (osseous malalignment/soft-tissue imbalance), trauma (repetitive or incidental), metabolic abnormalities, renal disease/transplant, osteoporosis, and glycosylation of bone proteins and collagen (2,3).

The mechanism(s) for the development of Charcot neuroarthropathy is not completely understood. However, two theories that address the causes of Charcot neuroarthropathy have been described: a neurovascular theory and a neurotraumatic theory (2,3). Both mechanisms likely contribute to Charcot neuroarthropathy. The neurovascular theory is based on increased blood flow to the limb from dilation of the blood vessels because of sympathetic denervation. The loss of vasomotor control allows blood vessels to dilate, thus increasing the peripheral blood flow. The increased peripheral blood flow increases the arteriovenous shunting, which causes hyperemia and bone resorption. Contrary to common misconceptions, most patients (90%) with Charcot neuroarthropathy of the foot have excellent blood supply to the foot (4).

The neurotraumatic theory is based on the patients experiencing an overuse injury because of an absence of protective sensation. Either acute trauma or repetitive trauma can initiate Charcot neuroarthropathy. Absence of protective sensation limits the body's protective mechanisms such as shifting body weight, limiting activity, and muscle guarding (2,3).

The best treatment results for Charcot neuroarthropathy of the foot and ankle are achieved when treatment is initiated during the early stages of the disease. The typical treatment for acute Charcot neuroarthropathy attempts to stabilize the condition by placing the foot in a total contact cast and immobilizing it. Non–weight bearing in a total contact cast can produce osteopenia of the ipsilateral foot and an increased load on the contralateral foot, which can lead to ulceration and Charcot neuroarthropathy in the contralateral foot. Maintaining non–weight-bearing status is difficult for this patient population for various reasons (e.g., obesity, diminished proprioception, and muscle atrophy). A new method for treating acute Charcot neuroarthropathy is to apply a static external fixator that acts like a cast by immobilizing the affected joints and bones (5).

After the Charcot foot is stabilized, the bones and joints halt disintegration and subluxation and coalesce to become fixed in a deformed position. Abnormal osseous prominences are potential areas for ulceration. During gait, the position of the patient's deformed foot and the resultant altered muscle–tendon balance produce aberrant weight-bearing forces that increase the risk for ulceration. When ulcers occur, osteomyelitis should be ruled out as a diagnosis and, if present, removed. If osteomyelitis is concurrent with Charcot neuroarthropathy, the patient has a poor prognosis. Therefore, early recognition and appropriate treatment is paramount to avoid osteomyelitis.

The typical surgical management for Charcot neuroarthropathy includes procedures such as Achilles tendon lengthening, ostectomy, debridement, osteotomy, arthrodesis, and open reduction with internal fixation. Open reduction with plantar plating or rigid internal fixation and an Achilles tendon lengthening are methods that are frequently used for reconstruction (6). Recently, static external fixation has been used for reconstruction of Charcot deformities, but gradual correction with the use of an external fixator is a new treatment option (2,3).

CLINICAL EVALUATION

Charcot deformities of the foot and ankle are vast. Acute, chronic, or acute-on-chronic Charcot neuroarthropathy can be observed at multiple anatomic locations with varying degrees of severity. Typically, the Charcot foot is wider and larger in size than a normal foot and, if untreated, will become even more deformed. The clinical presentation varies depending on the anatomic location and the stage of the disease. Acute Charcot neuroarthropathy might seem to be an infection because it presents as a red, hot, swollen foot (stage 1). Acute Charcot neuroarthropathy can present with or without foot and ankle deformities. A chronic Charcot deformity presents as a severe foot or ankle deformity, typically with ulceration (stages 2 and 3).

Ulcerations, infections, and osteomyelitis commonly are associated with Charcot neuroarthropathy and generally occur as related sequelae. Plantar ulcers commonly occur in patients with Charcot neuroarthropathy of the foot, and the location of the ulcer usually correlates to the anatomic location of the Charcot neuroarthropathy. For example, medial column ulcers of the foot are generally associated with tarsometatarsal Charcot neuroarthropathy that results in a medial column collapse. Tarsometatarsal Charcot deformities typically become stable (by coalescing) and are successfully treated conservatively or with a limited surgical approach (ostectomy or wedge resection with stabilization) (7). However, lateral column ulcers are associated with a more proximal Charcot deformity of the midfoot that typically does not fully coalesce. Instability of the lateral column leads to recurrent ulcers. In such cases, conservative treatment generally is futile and surgical reconstruction often becomes necessary.

Chronic ulcerations eventually lead to deep infection and osteomyelitis. Patients with osteomyelitis, as diagnosed in a clinical setting by probing to bone through the ulcer, undergo surgical debridement to remove all infection. In cases of osteomyelitis, the infected bone should be resected before reconstruction or the infection can be addressed concurrently with reconstruction by using external fixation.

In addition to noting the stage of Charcot neuroarthropathy, the surgeon should thoroughly assess the ulcerations, osteomyelitis, and anatomic location of the deformity. Radiographic assessment aids in determining the stage of the disease and the location of the deformity. Radiographs of the foot and ankle in cases of Charcot neuroarthropathy can be difficult to decipher; the bones of the hindfoot and midfoot are superimposed because of the subluxation/dislocation of these joints. Bone fragmentation and proliferation of new bone during the early and late stages of Charcot neuroarthropathy, respectively, add to the complexity of radiographic interpretation. Radiographs that show the patient during weight bearing should be obtained in all planes to more easily locate the Charcot deformity. Axial view radiographs can be helpful to evaluate Charcot neuroarthropathy of the midfoot, hindfoot, and ankle. Combined Charcot joint deformities are not uncommon. Using normal foot and ankle radiographic reference points and angles provides a quantitative assessment of the degree of deformity present (8). Major ankle, hindfoot, and forefoot axes, angles, and reference points should be used because of the osseous obscurity of the region affected by Charcot neuroarthropathy. Thus, forefoot-to-midfoot or midfoot-to-hindfoot relationships can be assessed and compared with normal relationships that aids in surgical planning.

CLASSIFICATION

Overall, Charcot neuroarthropathy is characterized by identifying the stage and the anatomic location of the deformity.

Stages of Charcot Neuroarthropathy

Eichenholtz (9) defined the stages of Charcot neuroarthropathy based on clinical examination and radiographic findings.

- Stage 1—Development phase, characterized by fragmentation of bones and cartilage, joint effusions, subluxation and dislocation, soft-tissue edema, hyperemia, bone resorption, and intra-articular fractures.
- Stage 2—Coalescence phase, characterized by decreased soft-tissue edema, healing of fractures, and organization of bone fragments.
- Stage 3—Reconstruction phase, characterized by new bone formation and remodeling of bone.

Shibata et al. (10) added another stage:

- Stage 0—Acute phase, characterized by swelling, warmth, joint instability, and normal radiographic anatomy of the foot and ankle.

Location of Charcot Deformity

Authors have described the patterns of Charcot neuroarthropathy based on the anatomic locations of the affected joints in the foot and ankle (2,11,12). The six regions affected by Charcot neuroarthropathy are the metatarsophalangeal, tarsometatarsal, midfoot, ankle, subtalar, and calcaneal regions. Each anatomic location of Charcot deformity has a particular pattern of osseous and soft-tissue deformity.

A patient with Charcot neuroarthropathy with or without ulceration who experiences a collapse that involves the lateral column has a poor prognosis for treatment (12). Various patterns of Charcot neuroarthropathy of the midfoot cause a lateral column deformity. Catanzariti et al. (13) found that ostectomy is successful in healing ulcers of the medial column but not of the lateral column. A collapsed lateral column creates an unstable foot, thereby producing aberrant and apropulsive gait. During gait, the ground reaction force vector ends just proximal to the calcaneocuboid joint and produces a constant deforming force on an already unstable lateral column (14). This pedal instability creates significant treatment difficulties. Catanzariti et al. (13) summarized by recommending that patients with lateral column Charcot deformity require a more complex surgical reconstruction.

TREATMENT OPTIONS

Conservative Treatment Options

Local wound care, total contact casting, custom-molded shoes with inserts, and specialized braces are commonly used to obtain or maintain a closed tissue envelope in patients with Charcot neuroarthropathy of the foot and ankle. The goals of these treatments are to off-load the ulcer and to support the foot and ankle. These conservative treatments can prevent or delay surgical intervention. However, surgical intervention is indicated when off-loading and supportive shoe gear is unable to stabilize the foot and ankle, or when recurrent ulceration is present. Even after surgical intervention, these conservative treatments are implemented.

Surgical Treatment Options

The goals of surgical intervention for the Charcot foot and ankle are to restore alignment and stability, prevent amputation, prepare for a shoe or brace, and allow the patient to be ambulatory. Achilles tendon lengthening, ostectomy, debridement, osteotomy, arthrodesis, open reduction with internal or external fixation, and gradual correction with external fixation

are surgical procedures that should be considered when reconstructing the Charcot foot and ankle. The surgical procedure selection depends on the stage of the disease, the anatomic location of the deformity, the presence or absence of osteomyelitis, the stability of the Charcot deformity, and multiple other factors.

Equinus deformity imparts a major deforming force on the foot and is almost always concurrent with Charcot foot deformities. Surgical strategies to correct equinus deformity of the ankle are based on clinical evaluation. The Silfverskiöld test is the gold standard for determining whether a patient has gastrocnemius equinus deformity or gastrocnemius-soleus equinus deformity. With most Charcot deformities, a severe gastrocnemius-soleus equinus contracture is present; therefore, maximum lengthening is necessary. Percutaneous Achilles tendon Z-lengthening is preferred. However, when possible, gastrocnemius-soleus recession is performed to minimize the loss of Achilles tendon strength (15).

Charcot neuroarthropathy causes the bones of the foot to undergo subluxation and dislocation, which produces a misshapen foot with osseous prominences. Ostectomy is performed to minimize or remove an osseous prominence. When an ulcer and an exostosis are present, the spur can be resected by using a direct or an indirect approach. A direct approach is performed by excising the ulcer and removing the underlying spur through the same incision. The indirect approach is performed by removing the spur from a remote site that is then closed separately from the site of the ulcer excision to avoid cross-contamination. For longstanding and deep ulcers with underlying exostosis, the indirect approach might not be possible because osteomyelitis is typically present. Surgical debridement of the infected bone and a six-week course of intravenously administered antibiotics that are specific to the organisms revealed by bone culture are mandatory.

Although limb preservation is the goal, the patient might undergo amputation of the Charcot foot because of related comorbidities, the severity of the Charcot deformity, or the extent of the infection. Surgical amputation is most commonly performed in patients who have severe Charcot neuroarthropathy of the foot and ankle with extensive osteomyelitis.

Surgical reconstruction typically is performed during stages 2 or 3 of the disease (9). Often, patients present during the later stages (i.e., coalescence and remodeling phase) of Charcot neuroarthropathy because they notice ulcerations or changes to the shape of the foot. It is unusual for patients to present during the acute stages of Charcot neuroarthropathy. During stage 0 or 1, the acute phases, surgical application of static external fixation has been advocated as an alternative to the traditional treatment option of total contact casting. External fixation has been used for early stabilization of acute Charcot joint subluxations and dislocations. Early stabilization with an external fixator off-loads the ulcerations and maintains the anatomic position of the bones, which avoids further deformity and allows for early partial weight bearing (5,16).

The anatomic locations of Charcot deformities have been defined by multiple authors (2,11,12). The exact locations of the deformities, especially in the midfoot joints, are difficult to determine based on plain radiographs because of the osseous superimposition. This distinction is critical because a Charcot deformity in the tarsometatarsal region typically is a stable deformity whereas a Charcot deformity in the midfoot region is an unstable deformity that leads to plantar-central and plantar-lateral ulcerations. Charcot neuroarthropathy in the ankle and subtalar regions also produces an extremely unstable deformity. When multiple locations are affected, the foot becomes severely unstable and difficult to manage.

A Charcot foot that is stable can be placed in a shoe or a brace while off-loading or accommodating osseous prominences. When Charcot deformity leads to an unstable foot, ulcerations are inevitable. Recurrent ulceration, osseous deformity, instability, and infection (e.g., osteomyelitis) are indications for surgical intervention. Eradicating the ulcer and infection and realigning the dislocated/subluxated foot joints with subsequent stabilization are the goals of reconstructing a Charcot foot and ankle. These factors and the patient's medical and surgical histories are critical to formulating an appropriate surgical plan for accurate correction.

An osteotomy is performed in a coalesced Charcot foot that is stable and can undergo osseous realignment. Shortening of the foot typically occurs when a midfoot wedge resection is performed. For example, a dorsally and medially based wedge is typically removed in patients with midfoot rockerbottom deformity. In patients with tarsometatarsal Charcot deformity, a similar type of wedge osteotomy can be performed. External fixation can be used

to gradually correct an osteotomy to realign the osseous segments. A percutaneous Gigli saw technique is preferred for midfoot osteotomies (14).

Arthrodesis is an important surgical step that should be used in all reconstructions of the Charcot foot and ankle (7). Open reduction with internal fixation is the traditional surgical technique that is used to stabilize the deformities. Internal fixation failure and recurrent breakdown of the Charcot joint have caused surgeons to develop new techniques to treat the effects of Charcot neuroarthropathy. Plantar plating and the use of additional hardware were introduced to better stabilize the Charcot foot and ankle (6). Arthrodesis with rigid internal fixation has remained an important treatment that is used to maintain a stable plantigrade foot.

SURGICAL TECHNIQUES (ACUTE VS. GRADUAL CORRECTION)

The surgical principle of obtaining a correction and then maintaining the correction applies not only to fracture reduction but also to deformity correction with the use of the Ilizarov device. The first stage of reconstructing a Charcot foot is to obtain a correction (i.e., acute or gradual soft-tissue correction and osseous realignment), and the second stage is to maintain the correction (i.e., arthrodesis or stabilization). As previously noted, almost all corrections require Achilles tendon Z-lengthening or gastrocnemius-soleus recession.

Acute Correction

Historically, open reduction with internal fixation was the mainstay for treatment of Charcot foot deformities. Large open incisions were made to remove the excess bone and to reduce the fragmented or dislocated bone. In addition, screw fixation or plantar plating was traditionally performed in an attempt to stabilize the Charcot joint. These invasive surgical procedures typically resulted in a nonanatomic correction (e.g., shortening of the foot or incomplete deformity correction) and occasionally resulted in neurovascular compromise, incision healing problems, infection, and the use of casts or boots for non–weight-bearing patients. Although performing open reduction has disadvantages, in cases of tarsometatarsal Charcot deformity, it is advantageous. Typically, Charcot neuroarthropathy of the tarsometatarsal joints is associated with mild-to-moderate deformities because the tarsometatarsal joints are structurally interlocked. Acute realignment achieved by performing a wedge resection and applying internal fixation produces a stable foot.

External Fixation

During the last decade, important advances have been achieved in the technology, preoperative deformity planning, and basic science of external fixation, especially regarding its use for deformity correction. Increased knowledge and improved technology have tremendously expanded the indications and applications of external fixation, including the use of external fixation and deformity-correction principles to treat Charcot neuroarthropathy of the foot and ankle.

External fixators allow for fine-tuning of residual deformity correction outside the operating room. With internal fixation, the precise plan for deformity correction must be obtained at the time of surgery and cannot be altered during the postoperative period. External fixator constructs can allow early weight bearing, which can lessen the severity of disuse osteoporosis, and allow for access to the soft tissues for wound care (16).

The disadvantages of external fixation include the length of treatment, complications such as pin-site infections, and the special surgical expertise required for construction of the frame. Most associated complications are minor and can be addressed nonoperatively. Typically, when operative intervention is required, the treatment can continue while the complication is being treated (16).

External fixation can be used for stabilization of an acute correction or for gradual deformity correction. The initial use of external fixation was for static fixation purposes only after acute correction was obtained with either open osteotomy or arthrodesis (7,17–19). The disadvantages of this approach are the large amount of bone resection required, which shortens the foot, and the large incisions required, which increase the rate of infection and the potential for wound-healing problems.

Gradual Correction

Gradual deformity correction with external fixation is preferred for large deformity reductions of the dislocated Charcot joint(s). Correction with external fixation allows for gradual and accurate realignment of the dislocated/subluxated Charcot joints. One advantage of using an Ilizarov apparatus to gradually correct the deformity is that the technique is minimally invasive, especially for patients with multiple previous incisions. Gradual correction also allows for anatomic correction without loss of foot length or bone mass. External fixation allows for partial weight bearing and limits neurovascular compromise because the correction occurs slowly over a period of time.

A stable or coalesced foot with Charcot deformity will require an osteotomy for correction of the deformity. Osteotomy can be performed by using the percutaneous Gigli saw technique. Midfoot osteotomies can be performed across three levels (i.e., talar neck and calcaneal neck, cubonavicular osseous level, and cuneocuboid osseous level. Performing a proximal osteotomy across multiple metatarsals is best avoided because of the disturbance of the interossei, risk of neurovascular injury, and multiple bones that require stabilization (14).

For an unstable Charcot foot or an incompletely coalesced Charcot foot, correction can be obtained through gradual distraction. Despite the radiographic appearance of coalescence, majority of patients with Charcot deformities can undergo distraction without osteotomy to realign the anatomy of the foot. After realignment, the correction is maintained by creating an osseous fusion with rigid internal fixation that is inserted percutaneously. This two-stage correction is a new technique that was developed by the senior author (Paley). The first stage consists of osseous realignment achieved by performing ligamentotaxis. Distraction and realignment restore the osseous structure and allow for soft-tissue healing. The second stage consists of removing the external fixator while simultaneously performing minimally invasive arthrodesis of the affected joints with percutaneous insertion of internal fixation. We prefer to use multiple large diameter, fully threaded, cannulated, intramedullary metatarsal screws that are inserted percutaneously through the head of the metatarsal by dorsiflexing the metatarsophalangeal joint. Recently, headless screws [e.g., Acutrak Fusion screws (Acumed, Beaverton, Oregon, U.S.A.)] have been used to achieve compression. Typically, three screws are used: medial and lateral column screws and one central screw. These screws span the entire length of the metatarsals to the calcaneus and talus, provide compression across the minimally invasive arthrodesis site, and stabilize adjacent joints. For example, a Charcot midtarsal joint is realigned with gradual external fixation, fused with minimally invasive techniques, and fixated with intramedullary metatarsal screws, which compress the arthrodesis site and stabilize the tarsometatarsal joint. The intramedullary metatarsal screws cross an unaffected joint, the tarsometatarsal joint, thereby protecting the tarsometatarsal joints from experiencing future Charcot neuroarthropathy (Fig. 1).

We have used this gradual distraction technique during the past four years and have achieved good to excellent success. The short-term results are promising considering that neither recurrent ulceration nor deep infection have occurred. Our results have been reproduced by Dr. Guido Laporta who used a similar protocol (oral communication, September 2006). The advantages of our method when compared with the resection and plating method reported by Schon et al. (6) or the resection and external fixation method reported by Cooper (17) are preservation of foot length, soft tissue and osseous anatomy, and cosmesis. Furthermore, our method is much less invasive.

Frame Constructs

A static frame (i.e., a Charcot stabilization construct) should include a distal tibial ring with a closed foot ring. This construct is generally used to treat patients with Charcot neuroarthropathy of the ankle or Charcot neuroarthropathy of the ankle combined with subtalar and/or midfoot Charcot deformity. A forefoot 6×6 butt frame (Fig. 2) should be used when gradually correcting a midfoot Charcot deformity with the use of a Taylor spatial frame (TSF) (Smith & Nephew, Memphis, Tennessee, U.S.A.) (20). The butt frame corrects the forefoot on a fixed hindfoot. In cases in which midfoot Charcot deformity is combined with subtalar joint Charcot deformity that cannot be acutely reduced, a forefoot 6×6 miter frame or forefoot $6 + 6$ frame can be used. The miter or forefoot $6 + 6$ frame can correct both hindfoot and forefoot deformities simultaneously. A forefoot 6×6 butt frame can also be used to correct both hindfoot and

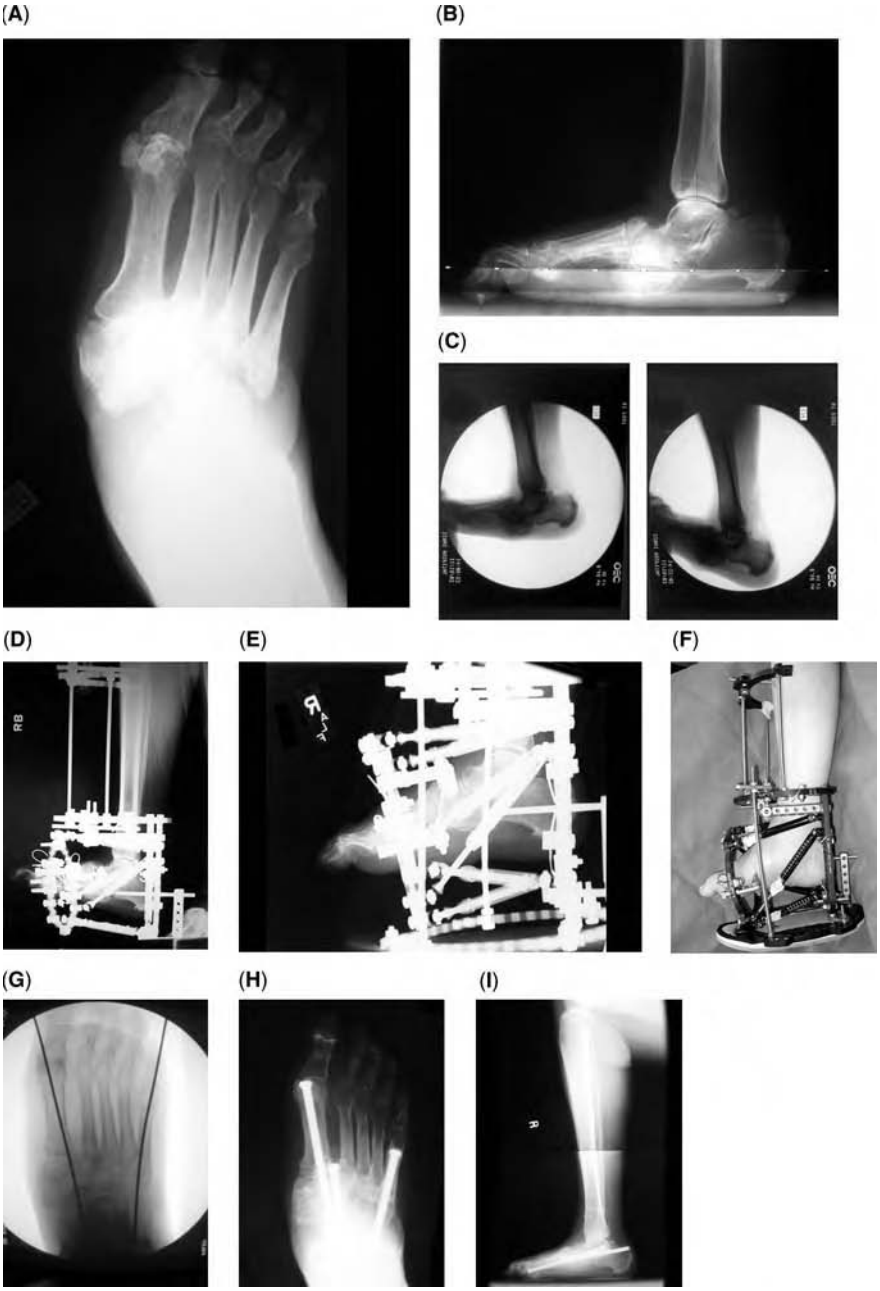

Figure 1 Case of midfoot Charcot neuroarthropathy with hindfoot equinus deformity (Eichenholtz stage 3, stable, without ulceration, or osteomyelitis). (**A**) Anteroposterior view radiograph shows the patient during weight bearing. Note the midfoot abduction. (**B**) Lateral view radiograph shows the patient during weight bearing. Note the dorsal translation midfoot deformity with equinus. (**C**) Percutaneous Achilles tendon Z-lengthening was performed to acutely correct the hindfoot equinus deformity. A lateral view still image obtained by using video fluoroscopy shows the complete correction of the equinus deformity. The hindfoot and ankle were then fixed in the corrected position with the Taylor spatial frame (forefoot 6 $\times$ 6 butt) (Smith & Nephew, Memphis, Tennessee, U.S.A.). (**D**) Lateral view postoperative radiograph shows the initial forefoot position. (**E**) Lateral view postoperative radiograph shows the forefoot position after gradual distraction to realign the forefoot to the hindfoot. (**F**) Clinical photograph shows the final midfoot realignment. Immediately after the external fixator was removed, a minimally invasive fusion of the midtarsal joint was performed. (**G**) Intramedullary metatarsal cannulated screws were inserted percutaneously to stabilize both the medial and the lateral columns of the foot. (**H**) Anteroposterior view radiograph shows the patient during weight bearing. Note the accurate anatomic reduction. (**I**) Lateral view radiograph shows the patient during weight bearing. Note the accurate anatomic reduction. The patient has not experienced a recurrence of ulceration or deformity (four-year follow-up).

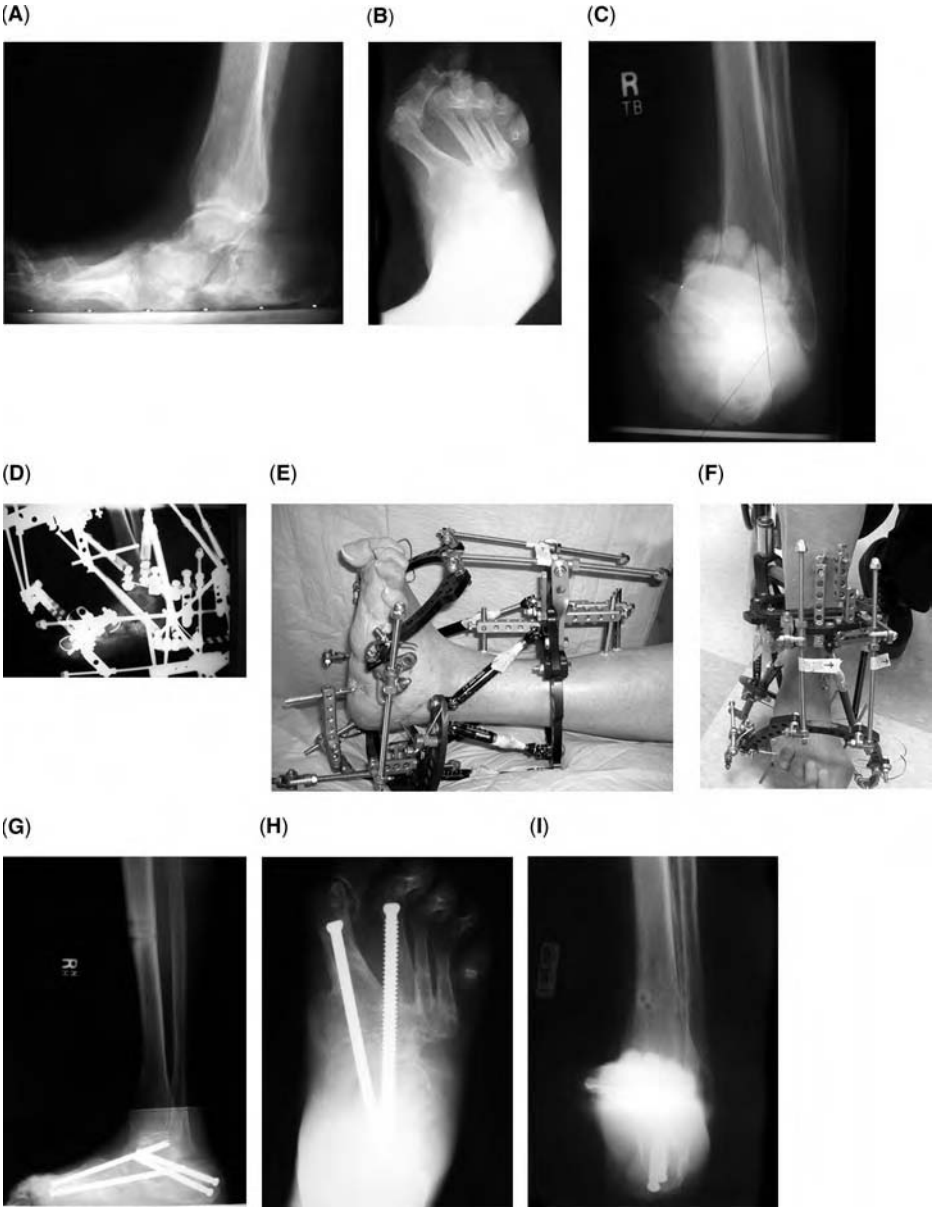

Figure 2 Case of midfoot Charcot neuroarthropathy with hindfoot equinovarus deformity (Eichenholtz stage 3, stable, with superficial ulceration and previously removed osteomyelitis). (**A**) Lateral view radiograph shows rocker-bottom and equinus deformities. (**B**) Anteroposterior view radiograph shows midfoot adduction deformity. (**C**) Axial (Saltzman) view radiograph shows hindfoot varus deformity. A percutaneous Achilles tendon Z-lengthening was performed to aid the hindfoot correction before applying the external fixator. However, complete equinus correction was not achieved acutely. A percutaneous midfoot Gigli saw osteotomy was performed across the neck of the calcaneus and talus. A Taylor spatial frame (long bone) was used for gradual soft-tissue correction of the hindfoot (equinovarus) while the forefoot was gradually corrected with the use of an Ilizarov apparatus through the midfoot osteotomy. (**D**) Lateral view postoperative radiograph shows the midfoot osteotomy with the external fixator in place. The hindfoot equinus deformity was corrected first through gradual soft-tissue realignment. The external fixator was then modified while the patient was in the clinic by backing out the half-pin that crossed the sinus tarsi into the talus so that the half-pin was located only in the calcaneus. The hindfoot varus deformity was then gradually corrected through the subtalar joint. (**E**) Lateral view and (**F**) anteroposterior view clinical photographs show the Ilizarov pusher and puller rods that were used to gradually correct the rockerbottom and adduction midfoot deformities. External fixation was removed after realignment was achieved. Immediately after removal of the fixator, a minimally invasive fusion of the midtarsal and subtalar joints was performed with the use of intramedullary metatarsal cannulated screws inserted percutaneously. (**G**) Lateral view radiograph shows the patient during weight bearing. (**H**) Anteroposterior view radiograph shows the patient during weight bearing. (**I**) Axial view radiograph shows the patient during weight bearing. Note the accurate anatomic reduction. The patient has not experienced recurrence of ulceration or deformity (three-year follow-up).

forefoot deformities simultaneously by stabilizing the ankle and talus with the vertical U-plate (reference ring) and then correcting the forefoot (proximal reference program) and the hindfoot (distal reference program) with two sets of TSF struts (combined anterior and posterior forefoat 6×6 butt frames). A miter frame can also correct a supramalleolar deformity combined with a midfoot Charcot deformity. The TSF can also be combined with Ilizarov parts to create the necessary frame construct (Fig. 3).

Creative frame construction is required because the pedal anatomy and the small size of the foot make it difficult to apply external fixation. Bone segment fixation is important; otherwise, incomplete anatomic reduction or failure of osteotomy seperation occurs. Stirrup wires are external fixation wires that are inserted through the bone and are bent 90° to extend and attach to a ring distant from the point of fixation. This stirrup wire is not tensioned and captures osseous segments that are distant from a ring. Small wire fixation is preferred in the

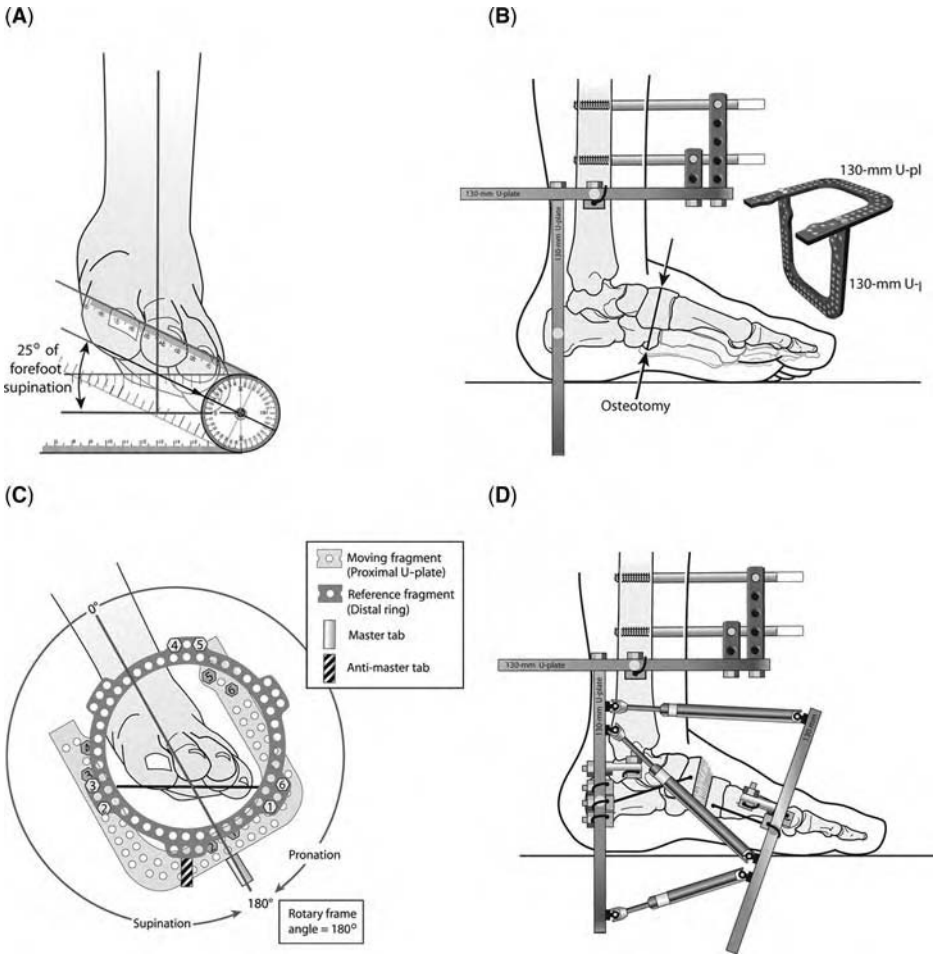

Figure 3 (**A**) Illustration of a forefoot supination deformity of 25°, as measured clinically. No hindfoot or ankle deformity was present. A percutaneous Gigli saw osteotomy at the cuneocuboid osseous level was performed before application of external fixation. Taylor spatial frame (forefoot 6×6 butt) was mounted (Smith & Nephew, Memphis, Tennessee, U.S.A.). (**B**) To fix the tibia, talus, and calcaneus, two U-plates were joined and mounted orthogonal to the tibia in the anteroposterior and lateral planes. With the hindfoot held in a neutral position, the calcaneus was then fixed in a neutral position with two crossing 1.8-mm wires. A talar neck 1.8-mm wire was also fixed to the posterior U-plate. Stirrup 1.8-mm wires were inserted just proximal and distal to the osteotomy and were bent 90° to fixate each of them to their respective external fixation ring. A forefoot ring was then mounted orthogonal to the metatarsals in the anteroposterior and lateral planes with two 1.8-mm crossing metatarsal wires. (**C**) Note the orientation of the rings and the locations of the struts and master tab. The forefoot 6×6 butt frame is shown mounted in a 180° rotary frame offset. Other degrees of rotatory frame offset may be used as well. (**D**) Illustration shows the final correction. Note the slight distraction to allow complete varus correction.

Table 1 Clinical Management

Classification Subgroup	Clinical Evaluation	Surgical Approach	Pearls	Complications/Pitfalls
Eichenholtz stage 0 or 1	Location of Charcot deformity, stability, ulcer, osteomyelitis	Immobilize with total contact cast or static external fixation	Conversion to stage 2 or 3, eradicate osteomyelitis	Nonunion or malunion, malalignment, recurrent ulcer, osteomyelitis, amputation
Eichenholtz stage 2 or 3	Location of Charcot deformity, stability, ulcer, osteomyelitis	Acute correction with internal fixation or gradual correction with external fixation then stabilization	Anatomic realignment, fusion of Charcot joint and stabilization of adjacent joints, eradicate osteomyelitis	Nonunion or malunion, malalignment, recurrent ulcer, osteomyelitis, amputation

foot because of the size and consistency of the bones (Fig. 3). Construction of extremely stable constructs is of great importance when treating a patient with neuropathy (Table 1).

COMPLICATIONS

External Fixation Complications

Patients with neuropathic conditions require close monitoring (i.e., weekly or biweekly) to assess for fixation failure and pin-site infection. Infections around the pin sites of the external fixation device are common and are treated by administering oral antibiotics. The infections rarely require removal of the pins or surgical debridement.

Malalignment

Obtaining proper anatomic alignment in the axial, sagittal, and transverse planes is critical. The severity of Charcot deformities makes it challenging to obtain accurate anatomic correction. Failure to accurately realign the foot and ankle can lead to recurrence of Charcot neuroarthropathy or recurrence of ulceration.

Ulceration/Osteomyelitis

Aggressively debriding the ulceration/osteomyelitis to the level of healthy bleeding tissue should always be performed. External fixation allows for wound healing while the deformity is being corrected.

Fixation Failure

Internal and external fixation (e.g., pin, wire, ring) breakage is not an uncommon complication and can occur when the biomechanical forces of the fixator are exceeded by the lengthening/deformity-correction process or by excessive weight bearing. This complication can be avoided by placing the fixator in a biomechanically advantageous position, using multiple points of fixation, and limiting weight bearing in neurologically compromised patients.

Wound-Healing Problems

Problems with incision healing can be avoided with an atraumatic surgical technique and proper preoperative planning of incision placement. For example, when performing an acute shortening of the midfoot by removing a large bone wedge, a transverse incision should be used to prevent the skin tension that would occur with a longitudinal incision.

FUTURE DIRECTIONS

The aftermath of Charcot neuroarthropathy of the foot and ankle can be devastating for the patient and extremely challenging for a surgeon to manage. Medical advancements to prevent neuropathy and eliminate the disease would be the ideal solution. In the meantime, improvements in technology will provide better fixation options and an enhanced understanding of the metabolic factors of Charcot neuroarthropathy will improve pharmacologic treatment.

ACKNOWLEDGMENT

The authors thank Amanda Chase, MA, and Dori Kelly, MA, for precise professional editing, Joy Marlowe, BSA, for excellent illustrative artwork, and Alvien Lee for photographic expertise.

REVIEW OF LITERATURE

Authors	Methods	Results	Conclusions
Wang et al. (5)	28 patients (retrospective review), open Achilles tendon lengthening, hybrid external fixation, monolateral fixation corrected the medial and lateral columns	No patients had further ulceration, consolidation averaged 3.1 mo, longest follow-up 2 yr	Correct equinus, stabilization of Charcot deformity with external fixation is an effective treatment
Cooper (17)	100 patients (retrospective review), acute open reduction with mostly static external fixation	Mean follow-up 22 mo, no recurrent ulcerations at the primary site, three limbs not salvaged, five patients developed ipsilateral Charcot ankle, stabilization of midfoot in a mean of 4.2 mo	Multiplanar deformities should be corrected gradually with a customized external fixator, external fixation offers various advantages over internal fixation
Farber et al. (18)	11 patients (retrospective review), excision of ulcer, open osteotomy with application of static external fixation	Mean follow-up 24 mo, external fixation with concurrent fusion, average fusion time 57 days and then total contact casting for an average of 131 days, No recurrent ulceration at the surgical site	Treatment good for ulcer resolution and prevention of amputation, fibrous vs. osseous union had no effect on clinical outcome
Simon et al. (7)	14 patients (retrospective review), Eichenholtz stage 1 of the tarsometatarsal joint treated with open reduction and fusion with internal fixation and autologous bone graft	Mean follow-up 41 mo, mean time to weight bearing without assistance was 15 wk, mean time to return to regular shoe gear was 27 wk, no repeat ulceration, gait analysis showed no difference to age-matched diabetic patients	Demonstrated the safety and efficacy of early operative intervention to restore foot alignment and function in cases of Charcot neuroarthropathy of the tarsometatarsal region

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17 Ankle Reconstruction After Trauma

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INTRODUCTION

The ankle is particularly vulnerable to trauma. The bones of the ankle are subcutaneous. The soft-tissue envelope consists of only skin, tendon, and neurovascular structures anterior, lateral, and medial to the joint. Only in the posterior quadrant is there a modest muscular envelope. In addition, the ankle joint does not tolerate deformity or articular incongruity after trauma. Studies have shown that this leads to pain and progressive ankle arthrosis (1–3).

The soft-tissue envelope is a crucial factor in dealing with acute and post-traumatic injury of the ankle. Often the acute injuries are open fractures that require plastic surgery intervention including skin grafts and free flaps. Compromise to the soft tissue is a major factor in determining the outcome of high-energy ankle injuries, particularly with regard to infection (2,4).

Tibial nonunions and failed pilon fractures have been treated with a variety of surgical methods including plate osteosynthesis with bone graft (4,5), intramedullary (IM) nailing (6), and external fixation (7–17). The complexity of a post-traumatic ankle (PTA) can be quite variable and depends on several factors. The ''personality of a fracture'' was a term and concept introduced by Schatzker (18) and its use underscores the complexity of a particular problem and helps organize a treatment approach (Table 1). It is helpful to apply this personality concept to the PTA. The personality of a PTA is determined by a number of factors including joint arthrosis, bone loss, radiographic appearance, and stiffness because they relate to the nonunion biology, deformity, leg length discrepancy (LLD), presence or history of infection, soft-tissue envelope, retained hardware, and patient factors including diabetes, smoking, and neuropathy.

The Ilizarov method has gained many advocates for the treatment of tibial nonunions and failed pilon fractures over the last two decades, particularly hypertrophic nonunions (11,12,14–16,19,20), and nonunions associated with bone loss (13,21–23), infection (24–27), poor soft-tissue envelope (11,16), and ankle fusion (28–31). The classic Ilizarov frame (Smith & Nephew, Memphis, Tennessee, U.S.) has been used to correct all deformity, including lengthening and bone transport (22,32–36) and fusion (28–31). However, deformity correction of translation and rotation can be complex and cumbersome with such a frame requiring lengthy frame modifications.

The Taylor spatial frame (TSF) (Smith & Nephew, Memphis, Tennessee, U.S.) is an evolution of the original Ilizarov frame and uses the same concepts of distraction osteogenesis as the classic frame. However, it can be used with the help of a computer program, to simultaneously correct length and all aspects of deformity including angulation, translation, and rotation. This is accomplished by establishing a ''virtual hinge'' in space around which all deformity is corrected. Circular rings are connected with six struts, which are gradually adjusted by the patient to correct the entire deformity (9,16).

We have used this modern Ilizarov method to comprehensively approach these complex and, in many cases, limb salvage situations. The acute injuries leading to the need for reconstruction range from low-energy ankle fractures to high-energy pilon fractures. A PTA problem can be complex and include numerous factors. These factors will define the personality of the ankle and this will help us establish a tailored and rational treatment approach.

The goals of treatment are a plantigrade foot, optimal leg lengths, bony union, ankle stability, and a mobile painless ankle joint. In many circumstances, the ankle joint is destroyed and an arthrodesis becomes the best option for dealing with pain and/or instability.

Table 1 Summary of Evaluation and Treatment

Classification	Evaluation	Treatment	Technical Pearls/Frame Configuration
Ankle arthritis			
Ankle arthritis, no deformity	Good mobility	Ankle distraction	Hinges at ankle axis
	Poor mobility	Ankle fusion	Screw fixation
Ankle arthritis with deformity	Magnitude and time duration of deformity,	Arthrodesis with I/TSF, acute or gradual	Wedge excision for acute correction; I/TSF for gradual correction; need talus wire
Ankle and subtalar arthritis	MRI and lidocaine joint injections for diagnosis	Tibio-talo-calcaneal arthrodesis with I/TSF	Compression frame with no talus wire
Supramalleolar deformity			
Supramalleolar deformity without arthritis	Check position/mobility of subtalar joint	Supramalleolar osteotomy	I/TSF for acute or gradual correction depending on magnitude and complexity of deformity
Supramalleolar deformity with arthritis	Anticipate correction that can be achieved through ankle fusion	Supramalleolar osteotomy with simultaneous ankle fusion	Two-level I/TSF: acute correction of ankle fusion and gradual correction of osteotomy
Ankle contracture			
Ankle contracture without bony deformity	Distinguish achilles contracture vs. gastrocnemius	I/TSF with hinges at ankle; gradual correction; percutaneous TAL	Axis of ankle from tip medial malleolus to tip of lateral malleolus through the talus
Ankle contracture with bony deformity	Foot may appear plantigrade because of compensation	Supramalleolar osteotomy with simultaneous gradual contracture correction	Two-level frame; gradual correction at both levels.
Ankle contracture with arthritis	Pain and stiffness	Gradual correction of prepared ankle fusion if contracture is large	Prepare fusion; partial correction acutely and use I/TSF to gradually correct rest of contracture
Supramalleolar nonunion			
Supramalleolar nonunion (stiff)	Hypertrophic appearance on X-ray; rule out infection	Do not open nonunion; gradual correction with I/TSF	Osteotomy of fibula needed; then test tibial stiffness
Supramalleolar nonunion (partially mobile)	Normotrophic appearance on X-ray; rule out infection	Minimal exposure of nonunion to open canals and bone graft; then apply I/TSF	Do partial correction in or but do rest gradually and then compress with I/TSF
Supramalleolar nonunion (mobile)	Atrophic appearance on X-ray; rule out infection	Open nonunion site, bone graft and do acute correction; compression with frame	Hold acute correction of nonunion with temporary wires and then apply compression I/TSF
Supramalleolar nonunion (infected)	Check for draining sinus	Resect dead infected bone. Antibiotic beads; no bone graft	Choice of acute shortening of defect or placement of beads. Compression with frame
Supramalleolar nonunion (with arthritis)	CT scan or MRI helpful to diagnose two levels; rule out infection.	Compression of nonunion and ankle arthrodesis	Two-level I/TSF with compression of both nonunion and ankle fusion
Mismatched columns of ankle			
Mismatched columns of ankle (tibia short)	Healed fibula with settling of tibia; distinguish malunion from nonunion	Gradual correction and lengthening of tibia relative to fibula	Do not place a tibio-fibular wire to allow movement of tibia relative to fibula
Mismatched columns of ankle (fibula short)	Valgus deformity with lateral tilt of talus	Gradual lengthening of fibula	Use monolateral frame to lengthen fibula and then insert syndesmosis screws to maintain correct position; need to fix tibia to fibula proximally;

(*Continued*)

Table 1 Summary of Evaluation and Treatment (*Continued*)

Classification	Evaluation	Treatment	Technical Pearls/Frame Configuration
Associated tibial shaft problem			
Associated tibial shaft problem (deformity)	The tibial shaft deformity is usually source of ankle pathology	Correct tibial deformity and ankle pathology simultaneously	Two-level I/TSF; choice of acute or gradual correction at both levels; goal is straight tibia and plantigrade foot
Associated tibial shaft problem (LLD)	Evaluate LLD with blocks and 51 in. erect leg X-ray	Lengthen tibia and approach ankle simultaneously	Two-level I/TSF; gradual lengthening of tibia; acute or gradual correction of ankle; goal with fusion—LLD of 1 cm
Bone loss			
Bone loss (from plafond)	Use long X-ray to calculate the longitudinal defect (X-ray defect + LLD); history of or active infection	Bone transport ankle fusion or acute shortening at ankle fusion followed by gradual tibia lengthening	Two-level I/TSF; acute shortening of no more than 2 cm; monitor pulses; neurovascular risk; fashion surfaces of tibia and talus for good contact
Bone loss (from talus)	History of or active infection; infected talus with osteonecrosis	Tibio-calcaneus fusion; option of simultaneous tibia lengthening or shoe lift	Prepare tibia and calcaneus surfaces for optimal contact; neurovascular risk with acute shortening; option of acute or gradual shortening; two-level I/TSF if tibia lengthening is done

Abbreviations: I/TSF, Ilizarov/Taylor spatial frame (Smith and Nephew, Memphis, Tennessee, U.S.); TAL, tendo-achilles lengthening; LLD, leg length discrepancy; MRI, magnetic resonance imaging; CT, computed tomography.

CLINICAL EVALUATION

In the history, one should obtain information about type of bony and soft-tissue injury, surgical procedures performed, history of infection, and the use of antibiotics. High-energy injuries and open fractures have a higher risk for infection. Information about back pain, perceived LLD, use of a shoe lift, and deformity should be elicited from the patient. The presence of deformity will often lead to the patient's report of a feeling of increased pressure on the medial or lateral part of the foot with a valgus or varus deformity, respectively. A short leg will often lead to complaints of low back pain and contralateral hip pain. If antibiotics are being used to suppress an infected nonunion, an attempt should be made to discontinue these for six weeks prior to surgery in order to obtain reliable intraoperative culture samples. Discontinuation of antibiotics must be done with caution and careful observation, particularly in compromised patients like those with diabetes or on immunosuppressive medications. The current amount of pain, the use of narcotics, and the ability to ambulate with or without support should be noted.

On physical examination, one should look for deformity and LLD with the patient standing still and walking. The inability to bear weight suggests an unstable nonunion. The view from the back is helpful for seeing coronal plane deformity. LLD is evaluated by using blocks under the short leg and by examining the level of the iliac crests. The view from the side is helpful for observing sagittal plane deformity and equinus contracture. The combination of recurvatum deformity above the ankle and equinus contracture of the ankle will lead to a foot translated forward position with an extension moment on the knee (Fig. 1). Range of motion of the ankle, subtalar, forefoot, and toes should be recorded. Rigid compensation for ankle deformity through the subtalar joint is an important factor. This typically occurs when there is long-standing ankle deformity. If this is present, it must be taken into account when correcting the ankle. The condition of the soft-tissue envelope, especially previous surgical wounds and flaps, and neurovascular findings should be recorded. This includes the posterior tibial and dorsalis pedis pulses, foot sensation, and dorsiflexion and plantarflexion motor function of the ankle and toes.

Radiographs should include anteroposterior (AP), lateral, and mortise views of the ankle, Saltzman's view of both feet (37), and a 51-in bipedal erect leg X-ray including the hips

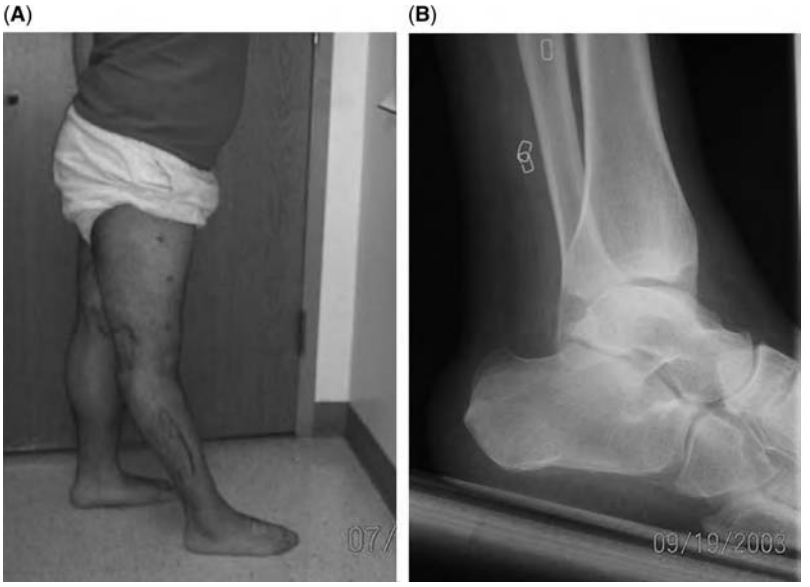

Figure 1 (**A**) Side view showing an equines contracture of the ankle associated with a foot forward position and hyperextension at the knee. (**B**) Lateral radiograph showing the equines contracture of the ankle.

to ankles with blocks under the short leg to level the pelvis (38). LLD as well as a limb alignment can be measured from a standing bipedal 51-in radiograph. The short leg is placed on blocks to level the pelvis and the height of the blocks is recorded. This can be done with the patient using crutches if necessary. These radiographs yield crucial information about LLD, deformity, presence of hardware, arthritis, and bony union. A supine scanogram can also be used to measure length discrepancy but this is not useful for alignment analysis. Computed tomography (CT) scan and magnetic resonance imaging (MRI) can be used as needed for further evaluation. The CT scan can be helpful in getting more information about bony union. The MRI can be helpful for obtaining information about the condition of cartilage in the ankle and subtalar joints and the presence of infection. Nuclear medicine studies can also be used, but we have not found them to be very helpful in this evaluation.

Rotational deformity is best assessed on clinical exam with the patient in the prone position. Thigh–foot axis is used to assess rotational deformity of the tibia. Rotational profile of the femur is used to assess rotational deformity in the femur. CT scan can also be used for this purpose. CT scan cuts at the proximal femur, distal femur, proximal tibia, and distal tibia allow analysis of rotational deformity (38).

Laboratory studies including white blood cell count, erythrocyte sedimentation rate, and C-reactive protein can be helpful for diagnosing the presence of infection. Selective lidocaine injections into the ankle and subtalar joints may be helpful for diagnosing the dominant source of pain.

CLASSIFICATION

The following is a list of PTA personalities that can be addressed with a modular Ilizarov method treatment approach (Table 1):

1. Ankle arthritis
   - with no deformity
   - with deformity
   - with subtalar arthritis

2. Supramalleolar deformity
    - Without ankle arthritis
    - With ankle arthritis
3. Ankle contracture
    - Without supramalleolar deformity
    - With supramalleolar deformity
    - With arthritis
4. Supramalleolar nonunion
    - Hypertrohic (stiff)
    - Normotrophic (partially mobile)
    - Atrophic (mobile)
    - Infected
    - With ankle arthritis
5. Mismatched columns of the ankle
    - Tibia short
    - Fibula short
6. Associated tibial shaft problem
    - Deformity
    - LLD
7. Bone loss
    - From tibial plafond
    - From talus

TREATMENT PRINCIPLES

Features of the Ilizarov Method

The Ilizarov method is particularly useful for addressing this spectrum of PTA pathology. Listed below are versatile features of the Ilizarov method (10,11).

1. Avoids internal fixation in presence or history of infection
2. Allows a minimal incision technique in setting of poor soft tissue
3. Utilizes acute and/or gradual correction of deformity
4. Utilizes opening wedge correction avoiding need for bone resection
5. Useful for large deformity correction
6. Postoperative adjustability for compression or correction
7. Simultaneous lengthening is possible for optimization of LLD
8. Allows multiple level treatment (a modular approach) (Fig. 2)
9. Weight bearing and ankle range of motion are encouraged

These features will be discussed in relation to the spectrum of the PTA personality.

Acute or Gradual Correction

One can employ either acute or gradual correction of a nonunion or malunion. Acute corrections can be performed in conjunction with all methods of fixation including plates, IM nails, and external fixation frames. Gradual correction requires the use of specialized frames.

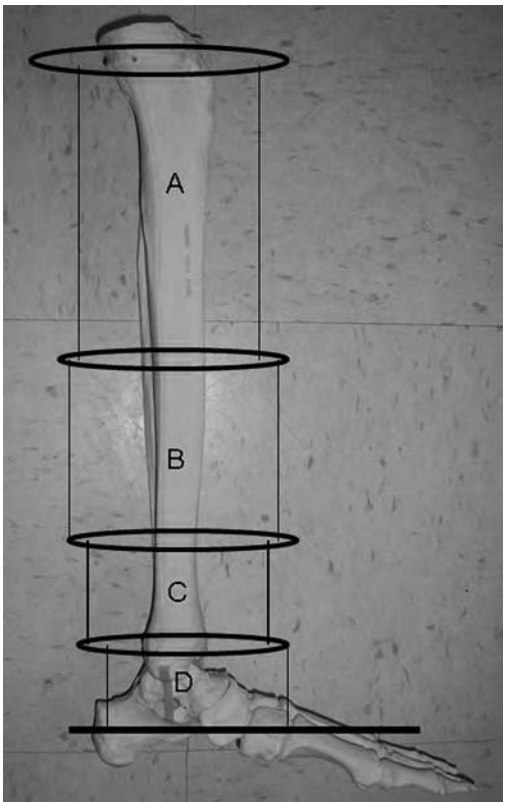

Figure 2 Schematic drawing depicting the various zones of treatment with the Ilizarov method. (**A**) Proximal tibia lengthening and/or deformity correction zone. (**B**) Tibial base consisting of two rings. (**C**) Supramalleolar correction zone. (**D**) Ankle fusion or contracture correction zone.

The personality of the problems helps guide the surgeon toward the best method. For example, a tibial malunion with 15° valgus deformity and 4 cm shortening is best handled with an osteotomy to gradually correct the angular deformity and lengthen the bone with a specialized frame. The Ilizarov method is utilized to gradually correct the complete deformity with distraction osteogenesis. One may choose to perform the deformity correction and lengthening at one level if bone regeneration potential is good. Alternatively, one may choose to perform a double-level osteotomy, one level at the center of rotation and angulation (38,39) for deformity correction, and one level for lengthening in the proximal tibia metaphysis. Gradual correction achieves treatment of shortening and carries less risk of peroneal nerve stretch neuropraxia than if attempted with an acute correction.

The use of plates and IM nails requires an acute correction of angular and translational deformity. Acute corrections are particularly useful for modest deformity correction, mobile atrophic nonunions that are opened and bone grafted, and small bone defects that can be acutely shortened. The principal advantage of acute correction is earlier bone contact for healing and a more simple fixation construct. Acute corrections are generally better tolerated in the femur and humerus and less well tolerated in the tibia and ankle related to issues of neurovascular insult (11,38).

Gradual correction with a specialized frame is useful for large deformity correction, associated limb lengthening, bone transport to treat segmental defects (13,21–23), and for stiff hypertrophic nonunion repair (11,12,14–16,19,20). Gradual correction employs the principle of distraction osteogenesis commonly referred to as the Ilizarov method (10,11). Bone and soft tissue are gradually distracted at a rate of approximately 1 mm per day in divided increments. Bone growth in the distraction gap is called ''regenerate.'' The interval between osteotomy and the start of lengthening is called the latency phase and is usually 7 to 10 days. The correction and lengthening is called the distraction phase. The consolidation phase is the time from the end of distraction until bony union. This phase is most variable and is most affected by patient factors such as age and health. If the structure at risk is a nerve such as the peroneal nerve for a proximal tibia valgus deformity or the posterior tibial nerve for an equinovarus deformity of

the ankle, gradual correction may be the safer option. The correction can be planned so that the structure at risk is stretched slowly (10,11,38). If nerve symptoms do occur, the correction can be slowed or stopped. Nerve release can be employed in select situations based on the response to gradual correction (38).

TREATMENT OPTIONS

Ankle Arthritis

If the ankle is very stiff and painful, then arthrodesis becomes the most predictable option. Although this type of arthrodesis can be done with an Ilizarov frame, it can be successfully performed more simply using screw fixation. There may be some indication for total ankle replacement in a very select group of patients, namely the older patient with no history of infection.

If the ankle has an arc of at least 30° of motion, the arthritis is moderate, and the patient is not interested in pursuing fusion, then distraction arthroplasy may be a good option. This technique was popularized in the Netherlands and involves placing an external fixation frame across the ankle. Encouraging results have been reported at intermediate term follow-up (40). Joint distraction is based on the concept that osteoarthritic cartilage has some reparative activity when there is a release of mechanical stress on the cartilage while intra-articular intermittent fluid pressure is maintained (41). Our current joint preservation approach includes arthroscopic debridement, anterior exostosis removal, and percutaneous tendo-achilles lengthening (TAL) if these are indicated. In addition, we apply an articulated distraction frame to allow ankle range of motion and the ability to correct contracture in addition to the distraction (42) (Fig. 3). This frame is worn by the patient for three months. Weight bearing and ankle motion are encouraged throughout the treatment. Results with this treatment both clinically and radiographically, with an increased joint space, have been encouraging. This treatment does not burn bridges for a possible future need for arthrodesis or ankle replacement. If there is ankle deformity, the distraction can be combined with a supramalleolar osteotomy by adding another level to the frame (Fig. 4).

When there is deformity with its apex at the ankle, correction is done through the fusion. Acute correction is accomplished with removal of a medial or lateral wedge from the plafond for correction of valgus or varus deformity, respectively. Acute correction must be performed with caution and vigilance for neurovascular insult. The Ilizarov frame is very useful in stabilizing flat bony surfaces that lack the congruity of a simple ankle fusion (Fig. 5). Gradual correction is a safer option in setting of large deformity. The prepared ankle fusion site is gradually positioned neutral with the use of a dynamic Ilizarov/TSF.

When both ankle and subtalar joints are affected, there may be an indication to fuse both joints. Selective lidocaine injections of both joints under fluoroscopy can prove useful for preoperative decision making. Both ankle and subtalar joints are prepared for fusion and the Ilizarov frame can be used for compression arthrodesis.

Supramalleolar Deformity

In the absence of symptomatic arthritis, correction of the deformity with a supramalleolar osteotomy is performed (Fig. 6). The goal is to correct the deformity in both the coronal and the sagittal planes. The goal is to achieve a lateral distal tibial angle of 90° and an anterior distal tibial angle of 80°(38,39). The use of the Ilizarov/TSF is particularly useful for a gradual correction of a simple or large oblique plane deformity.

In the presence of symptomatic arthritis, this may be addressed as well. As mentioned earlier, an ankle distraction can be done distal to the supramalleolar osteotomy with the addition of another level of treatment. If the arthritis is severe and symptomatic, the deformity correction can be done simultaneously with an ankle arthrodesis (Fig. 7). Typically, the arthrodesis would be positioned acutely and simultaneous gradual correction of the osteotomy would continue above the ankle. If there is a large ankle contracture, the option of gradual correction through the prepared ankle fusion may also be used.

Ankle Contracture

The usual contracture is equinus. If it is small, the acute correction with TAL may be utilized. If the contracture is large and especially if it is long standing, it may be safer to do the correction gradually. After a percutaneous TAL, an Ilizarov frame is applied across the ankle and hinges

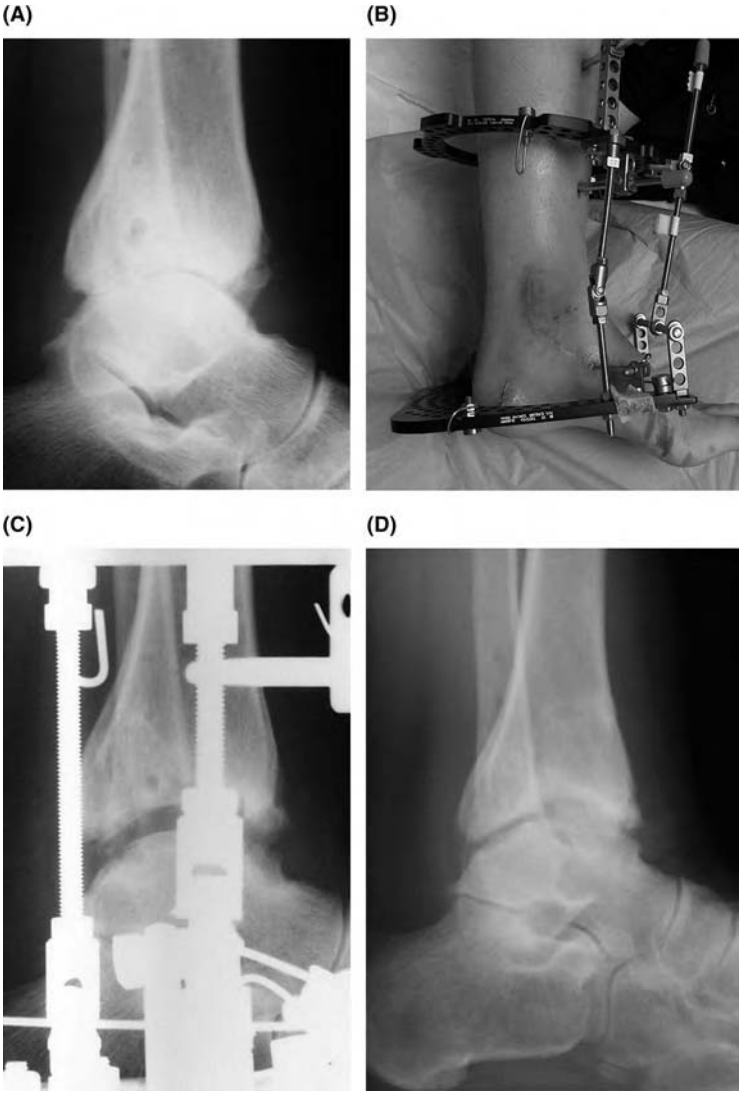

Figure 3 (**A**) Lateral radiograph of ankle showing arthritis with no deformity. (**B**) Side view of an ankle distraction frame with hinges at the axis of the ankle. (**C**) Lateral radiograph showing distraction of the ankle. (**D**) Lateral radiograph of the ankle six months after frame removal showing an increased joint space. Clinical symptoms were significantly improved.

are placed in line on the axis of the ankle (38). This is a doubly oblique plane that passes from the tip of the lateral malleolus to the tip of the medial malleolus through the center of the talus (Fig. 8). The TSF and a virtual hinge may also be used. Correction of the contracture is accomplished by gradually moving the rings parallel to each other. The use of a constrained frame with hinges prevents compression of the ankle articular surfaces during the correction. The posterior tibial nerve is undergoing stretch within the tarsal tunnel during this correction. The gradual approach allows the speed of the correction to be adjusted accordingly and decreases the likelihood of a stretch neuropraxia. If signs and symptoms of stretch neuropraxia are present despite slowing the correction, then a tarsal tunnel release is performed.

Associated supramalleolar deformity may be addressed at the same time. A common presentation is a recurvatum deformity above the ankle combined with an equinus contracture of the ankle. The foot appears plantigrade and anteriorly translated. Acute or gradual correction of both the bony deformity above the ankle and the contracture of the ankle can be accomplished with a two-level Ilizarov/TSF (Fig. 9).

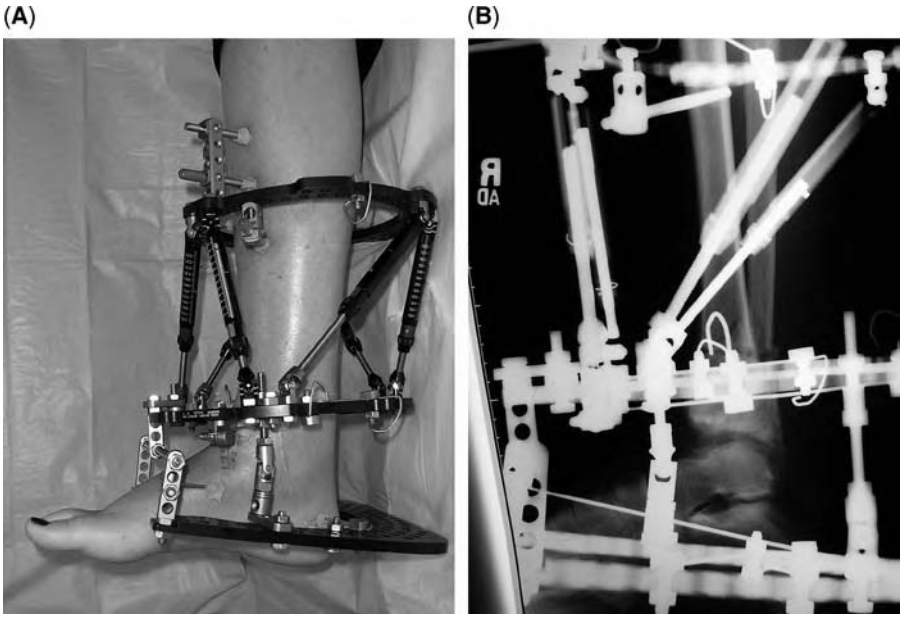

Figure 4 (**A** and **B**) Side view and X-ray of a patient who underwent a supramalleolar osteotomy for deformity correction and simultaneous ankle distraction.

If there is associated arthritis of the ankle, this can be addressed with either a simultaneous fusion or a distraction. This depends on the severity of the arthritis and the physician/patient decision for joint preservation or fusion.

Supramalleolar Nonunion

An excellent application of gradual correction is for a hypertrophic stiff nonunion with deformity. This type of nonunion has fibrocartilage tissue in the nonunion and has the biologic capacity for bony union. It lacks stability and axial alignment. Gradual distraction of this type to achieve normal alignment results in bone formation (Fig. 10). The nonunion acts like regenerate and bony healing occurs. Modest lengthening of no more than 1.5 cm should be done through the nonunion. If additional lengthening is needed, a second osteotomy for lengthening is performed. Several studies have confirmed Ilizarov's success with this technique (11,12,14–16,19,20). The principal advantages are not having to open the nonunion site in the face of poor skin and widened callus and gaining length through an opening wedge correction. This is particularly beneficial to the region above the ankle where the soft-tissue envelope is often compromised. This technique is not useful for mobile atrophic nonunions and less applicable to infected nonunions.

Atrophic nonunions (Fig. 11A) have fibrous tissue at the nonunion site and tend to be mobile. This is often the situation after previous open surgery where surgical exposure may have compromised the bone-healing biology. Treatment needs to be directed toward improving both the biology and the mechanical environment in order to achieve bony union. Normotrophic nonunions have both fibrous tissue and fibrocartilage and are partially mobile (Fig. 11B). Atrophic and even normotrophic nonunions should be exposed, bone ends should be contoured so that there is healthy bleeding bone on both sides with good contact, and IM canals should be opened. Stripping of soft tissue should be performed within moderation. Acute correction of deformity should be followed by bone grafting and stable fixation with compression. This can be accomplished with a plate, IM nail, or a frame depending on surgeon's preference and location. Compression plating of aseptic nonunions has been used successfully (5). In contrast to acute fracture treatment where rigid stability is not necessarily the goal (43), the goal for stabilization of nonunions should be a relatively rigid construct (5,11).

Circular external fixation can also be useful for atrophic and normotrophic nonunions. In the case of an atrophic nonunion, the frame is used for stabilization after acute correction

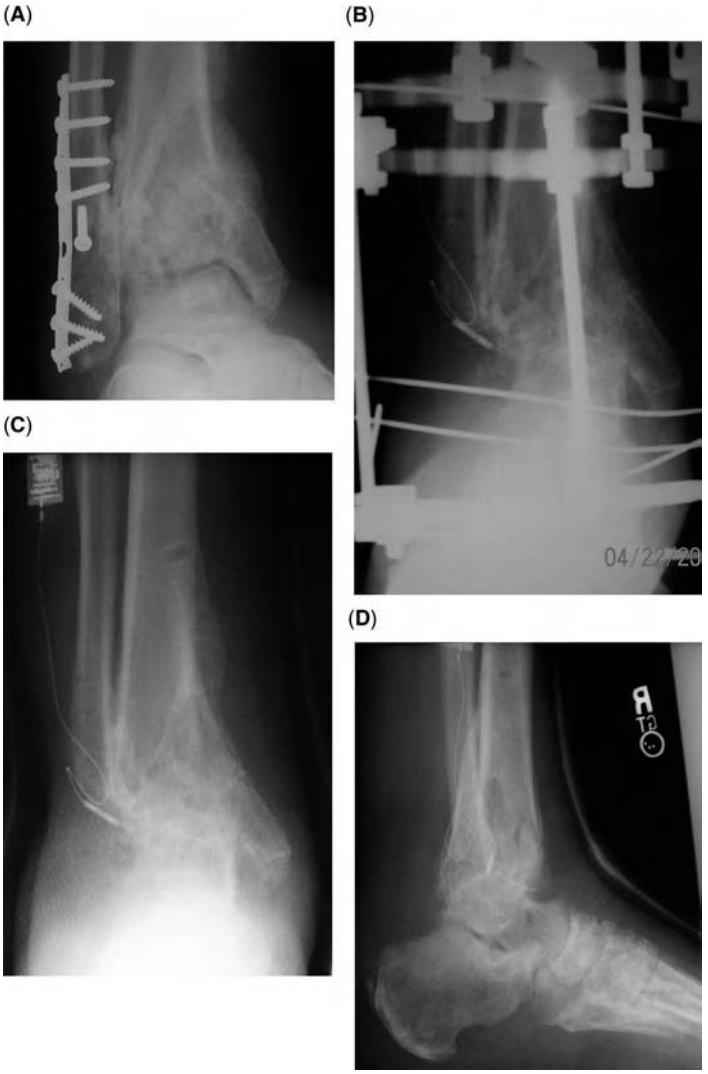

Figure 5 (**A**) Anteroposterior (AP) X-ray showing post-traumatic arthritis of the ankle with varus deformity after a pilon fracture. (**B**) AP X-ray showing the tibio-talar junction under compression in an Ilizarov frame. Acute correction of the deformity was performed. (**C** and **D**) AP and lateral X-ray of the ankle six months after surgery showing a successful ankle fusion.

and an open approach. A positive feature of using a frame is that in addition to the ability to acutely compress the nonunion in surgery, one can add more compression during the postoperative period. The frame is also stable enough to allow full weight bearing right after surgery (11,17,25). Normotrophic nonunions can also be approached in another fashion with the use of gradual correction. The nonunion can be approached in a minimally invasive fashion through 1 to 2 cm incisions. With the aid of intraoperative fluoroscopy, the nonunion can be mobilized with an osteotome and the IM canals can be opened by using a cannulated drill and curettes. Bone graft can then be inserted. The frame is then applied and used to gradually correct the deformity (angulation and translation). Once this is accomplished, axial compression is then performed. Full weight bearing is allowed immediately after surgery. If additional length is needed, an osteotomy for gradual lengthening can be performed at a different site.

Nonunions after tibial pilon fractures can result in metaphyseal nonunion combined with ankle arthrosis (Fig. 12). Infection, poor soft tissue, and retained hardware often complicate these situations. Treatment should be directed toward repair on the distal tibia, correction

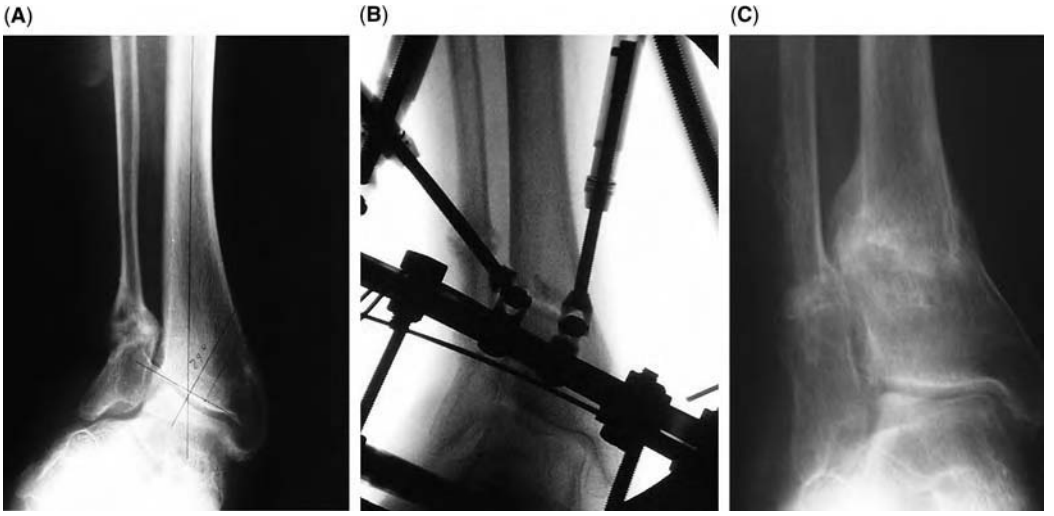

Figure 6 (**A**) Malunion with valgus deformity of the tibial plafond in a patient with rheumatoid arthritis. (**B**) After application of Ilizarov/Taylor spatial frame and percutaneous supramalleolar osteotomy. (**C**) After healing of the opening wedge correction obtained with distraction osteogenesis.

of deformity, and ankle arthrodesis if necessary. This can be accomplished with internal (5) or external fixation (28–30). If bone resection is needed as in the case of infection, then ankle fusion and simultaneous tibia lengthening can be done with the Ilizarov method (11,31).

Infection

Infected nonunions are most complex. Typically, these are atrophic and mobile; however, they can also be stiff and hypertrophic. Infected nonunions should typically be approached in an open fashion. The goals of surgery are to remove all dead bone, open the IM canals, oppose bleeding bone surfaces, and correct the deformity. The patient should ideally have been off all antibiotics for several weeks and multiple intraoperative cultures and pathology specimens are sent to the laboratory at the time of surgery. The nonunion is then mechanically stabilized. With the help of an infectious disease consultant, treatment for chronic osteomyelitis is rendered. This usually consists of culture-specific intravenous antibiotics for six weeks followed by an oral regimen. Removal of dead bone is needed to eradicate infection. Bone graft should not be used at the primary surgery. Antibiotic beads can be used for dead space management and local antibiotic delivery. Several weeks later, the beads can be removed and the nonunion can be bone grafted. The use of absorbable antibiotic beads made of calcium sulfate has been advocated by some in order to avoid the need for removal and subsequent bone grafting (44). If acute shortening is performed and there is little dead space or purulence, antibiotic beads are not used.

Stabilization can be accomplished with a plate, IM rod, or an external frame. All of these methods have been used successfully; however, internal fixation has the disadvantage of adding foreign material to the infected site and is fraught with risk. The use of external fixation is my preferred approach in most cases of infection. It has then advantage of not adding foreign material to the infection site and can be used to treat more complex situations. If debridement of the nonunion results in a bone defect, the frame can be used for *bone transport* (Fig. 13) or acute shortening and gradual lengthening (11,22,27,32).

Staging Treatment

Staging the treatment is an important strategy for nonunion management. In the case of infection, antibiotic beads may be removed after several weeks and bone graft inserted. In the situation of bone debridement resulting in a bone defect, one may choose to do a gradual or acute shortening with a frame. An osteotomy for lengthening can be done several weeks later after the infection is cleared and after the patient and surgeon have decided on the option

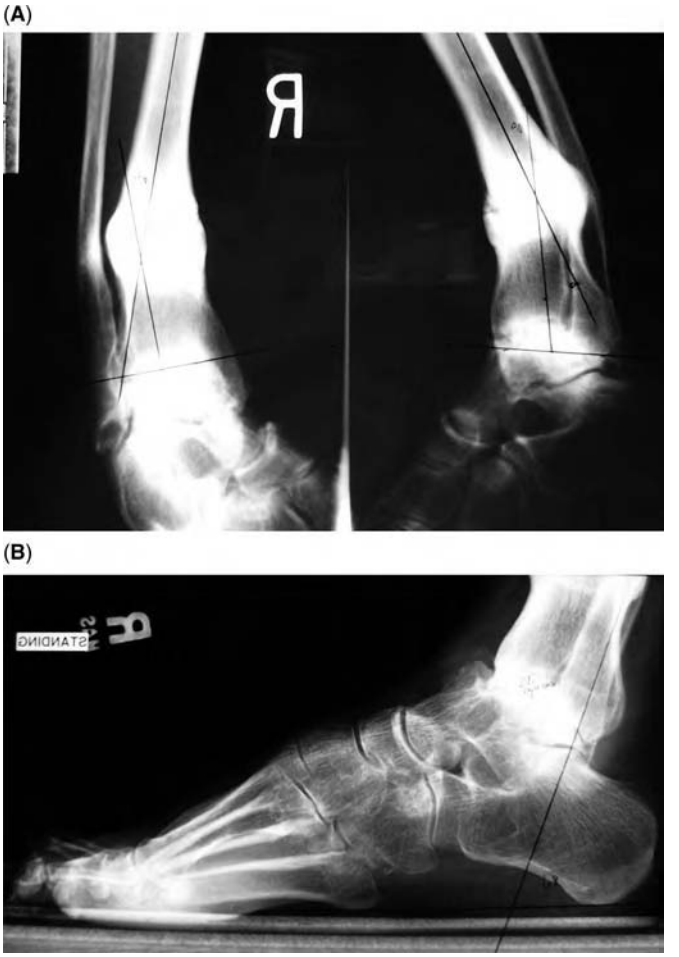

Figure 7 (**A**) Anteroposterior and lateral radiographs showing a patient with a varus, recurvatum malunion with associated ankle arthritis. (**B**) Lateral X-ray of the ankle showing the advanced ankle arthrosis combined with equines contracture of the ankle.

of lengthening versus the use of a shoe lift. This has the advantage of protecting the osteotomy site from contamination. In addition, it is often difficult to predict the precise amount of bone resection needed. Once this is known, the patient and surgeon can make a more informed decision about lengthening.

When bone transport is used to treat a bone defect, the docking site should be prepared when there is about one centimeter of gap. Preparation of the docking site includes debridement of fibrous tissue, realignment of bone ends to maximize bony contact and minimize deformity, and the addition of bone graft. This improves the rate of bony union (22).

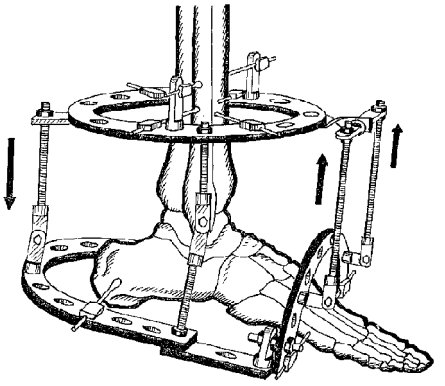

Figure 8 Schematic drawing showing a constrained frame for gradual correction of an equines contracture. Note the hinges at the ankle. *Source*: Courtesy of Arkady Blyakhar.

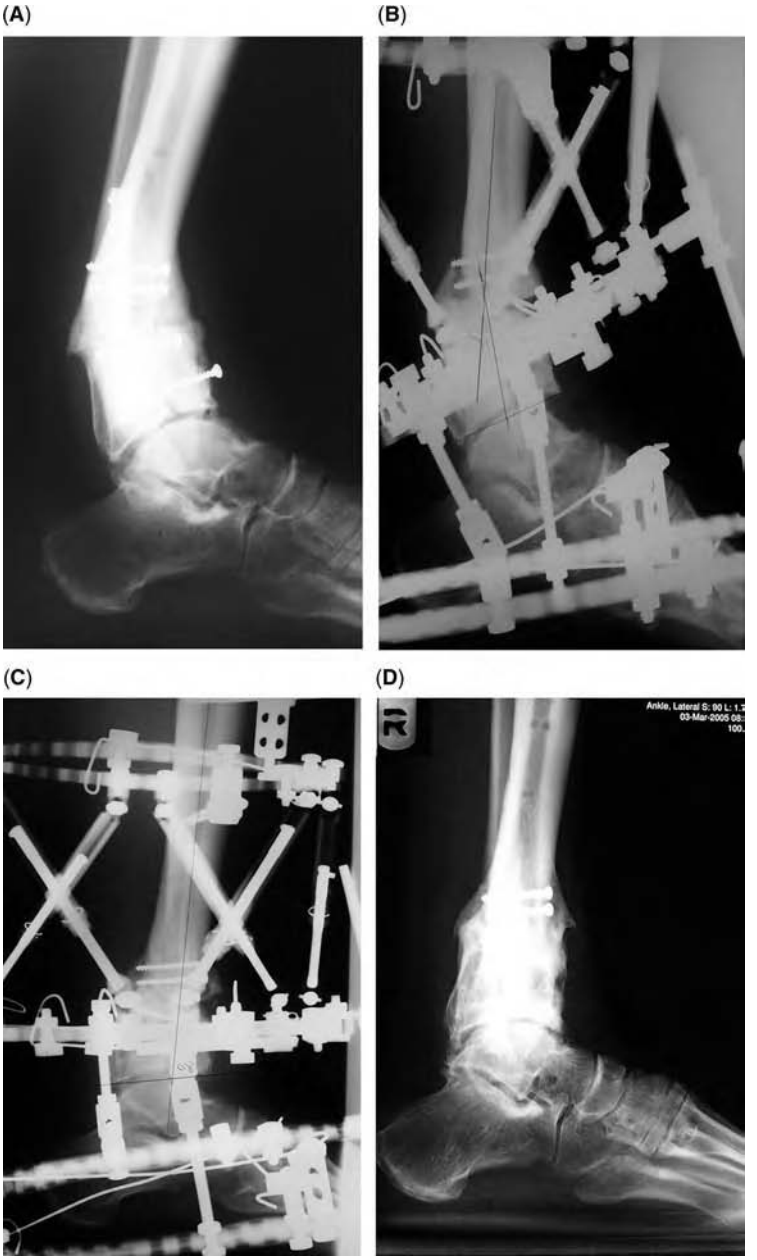

Figure 9 (**A**) Lateral X-ray showing a recurvatum deformity of the distal tibia with equinus contracture of the ankle. (**B**) Postoperative lateral X-ray showing a frame with potential for correction of the supramalleolar deformity and the ankle contracture. (**C**) Lateral X-ray six weeks later showing the correction. (**D**) Lateral X-ray one year later.

If the soft-tissue coverage is poor, flap coverage (24,27), or the use of a vacuum-assisted closure (45,46) device may be needed. A staged approach with the plastic surgeon can be helpful. For example, one may do a debridement of bone and soft tissue and apply a simple frame that allows your plastic surgeon access to the wound. After flap coverage has been accomplished, one can then go back and perform bone transport for a bone defect or elevate the flap after several weeks and bone graft the nonunion site.

Mismatched Columns of the Ankle

The tibia may shorten relative to the fibula. This can be observed as a nonunion or malunion of the tibial plafond. This will lead to abnormal stress transmission across the ankle and premature arthritis (3). In a normal situation, a transmalleolar line will intersect a mid-diaphyseal line of the tibia at 83° (38). Variation from this measurement signifies shortening

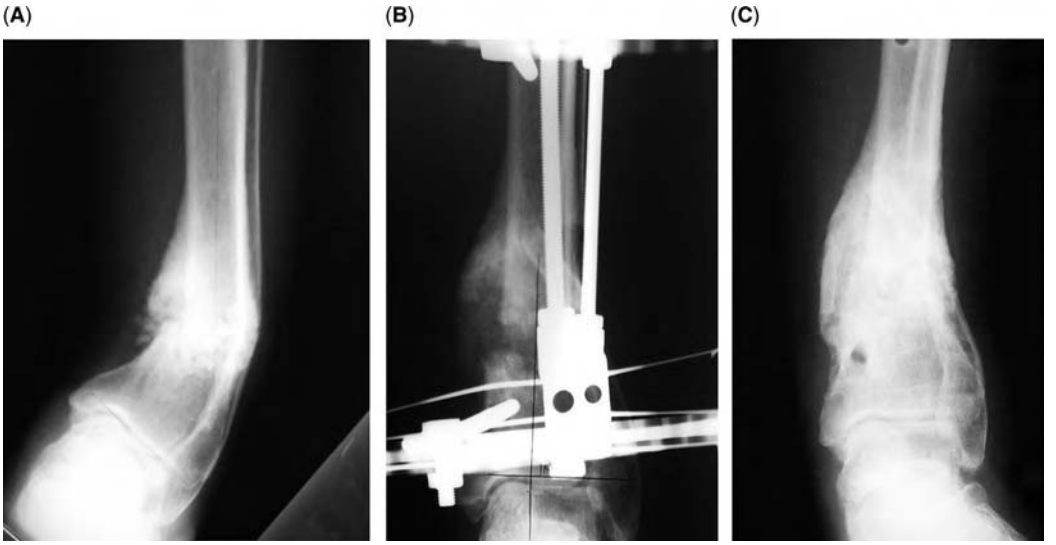

Figure 10 (**A**) Anteroposterior X-ray showing a stiff nonunion with large varus deformity in a blind diabetic patient. This patient had a lateral ulcer at the apex of the fibula deformity. (**B**) Four weeks after application of the Ilizarov frame. The nonunion was not exposed and gradual distraction correction was performed. (**C**) Six months after frame removal showing healed nonunion without deformity.

of the tibia or fibula. This may present after treatment of a pilon fracture with fibula plating and spanning external fixation and percutaneous screws for the tibia. The fibula heals out to length and the tibia settles with relative shortening and varus deformity. Treatment consists of lengthening and correction of the tibial deformity relative to the fibula. This is accomplished with an Ilizarov frame with the distal leg ring fixed to only the tibia and not the fibula. This setup provides the potential for relative lengthening of the tibia through an osteotomy in the case of a malunion or through the nonunion itself (Fig. 14).

Alternatively, the fibula may shorten relative to the tibia. This will occur if the fibula is initially fixed short immediately after the trauma or if it is not fixed and gradually shortens during healing. In this situation, fibula lengthening may be accomplished gradually with distraction osteogenesis. Once the length is correct, syndesmosis screws can be inserted to maintain the length and the frame is removed (47) (Fig. 15).

Associated Tibial Shaft Problem

An example of this would be malunion of the tibia that is associated with ankle arthritis. Recurvatum deformity of the tibia will lead to uncovering of the talus, abnormal forces across the ankle, equinus contracture, and arthritis. Two-level Ilizarov treatment can be used to correct the tibial deformity and achieve ankle fusion with acute or gradual correction (Fig. 16). If there is shortening of the tibia, this can be addressed at the same time as the deformity correction at the apex of the deformity. An osteotomy (48) at the proximal tibia for lengthening can be done if the bone-healing potential at the apex of deformity is not optimal (Fig. 17).

Bone Loss

Bone loss from the tibia plafond is the result of the trauma or subsequent infection. This may be associated with the need for an ankle fusion. If a fusion is needed, then a bone transport ankle fusion is done (Fig. 18). Alternatively, acute shortening and fusion of the ankle is done, and gradual lengthening of the tibia follows (31). In the situation of infection, there is an advantage to delaying the proximal tibia lengthening for a few weeks. This allows treatment of the infection with culture-specific antibiotics, a better appreciation for the amount of lengthening that is needed, and a safer environment for the proximal tibia lengthening with less chance of contamination.

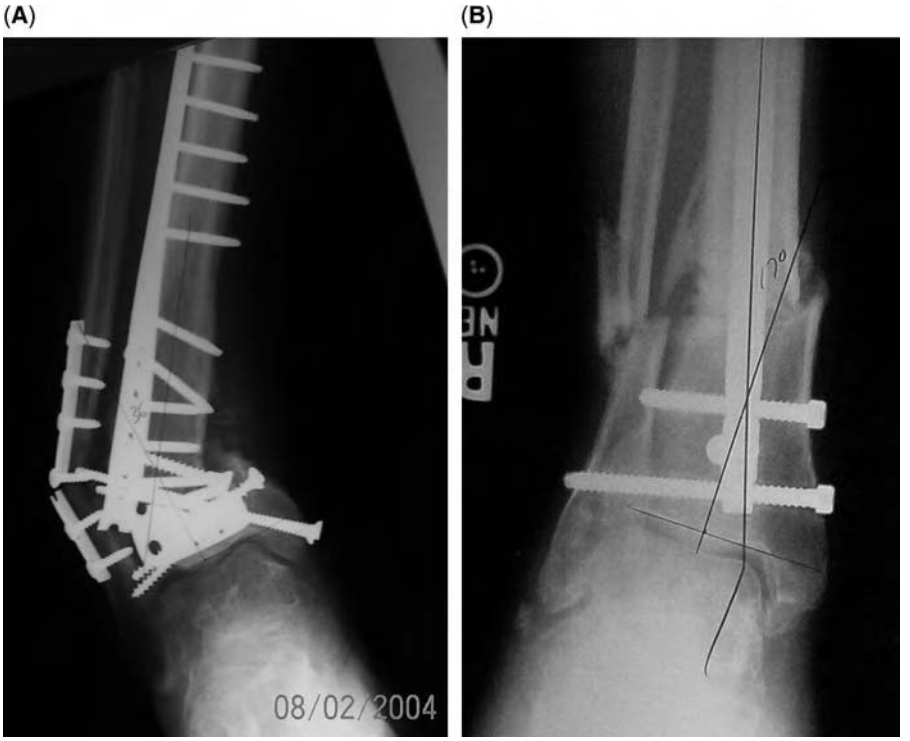

Figure 11 (**A**) Anteroposterior (AP) X-ray of an atrophic mobile nonunion of the distal tibia/fibula with deformity and retained hardware. (**B**) AP X-ray showing a normotrophic partially mobile nonunion with retained intramedullary nail and valgus deformity.

Bone loss from the talus can be the consequence of necrosis and osteomyelitis. This can lead to the need to remove the talus. Ankle fracture in a neuropathic patient can lead to a Charcot ankle with collapse and destruction of the talus. In either situation, a tibia to calcaneus ankle fusion is needed. This will usually lead to about 4 cm of shortening. Because 1 cm of

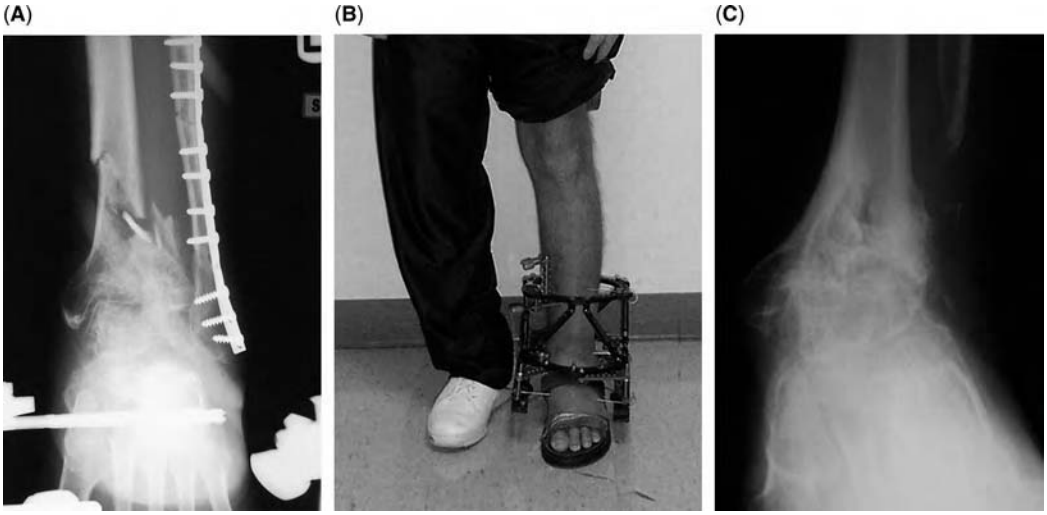

Figure 12 (**A**) Anteroposterior (AP) X-ray showing a pilon fracture nonunion nine months after trauma associated with advanced ankle arthrosis. This patient had been non–weight bearing in ankle spanning frame for nine months. (**B**) Front standing view showing an Ilizarov/Taylor spatial frame being used for simultaneous compression of the plafond nonunion and the ankle arthrodesis. (**C**) AP radiograph nine months after frame removal showing bony union.

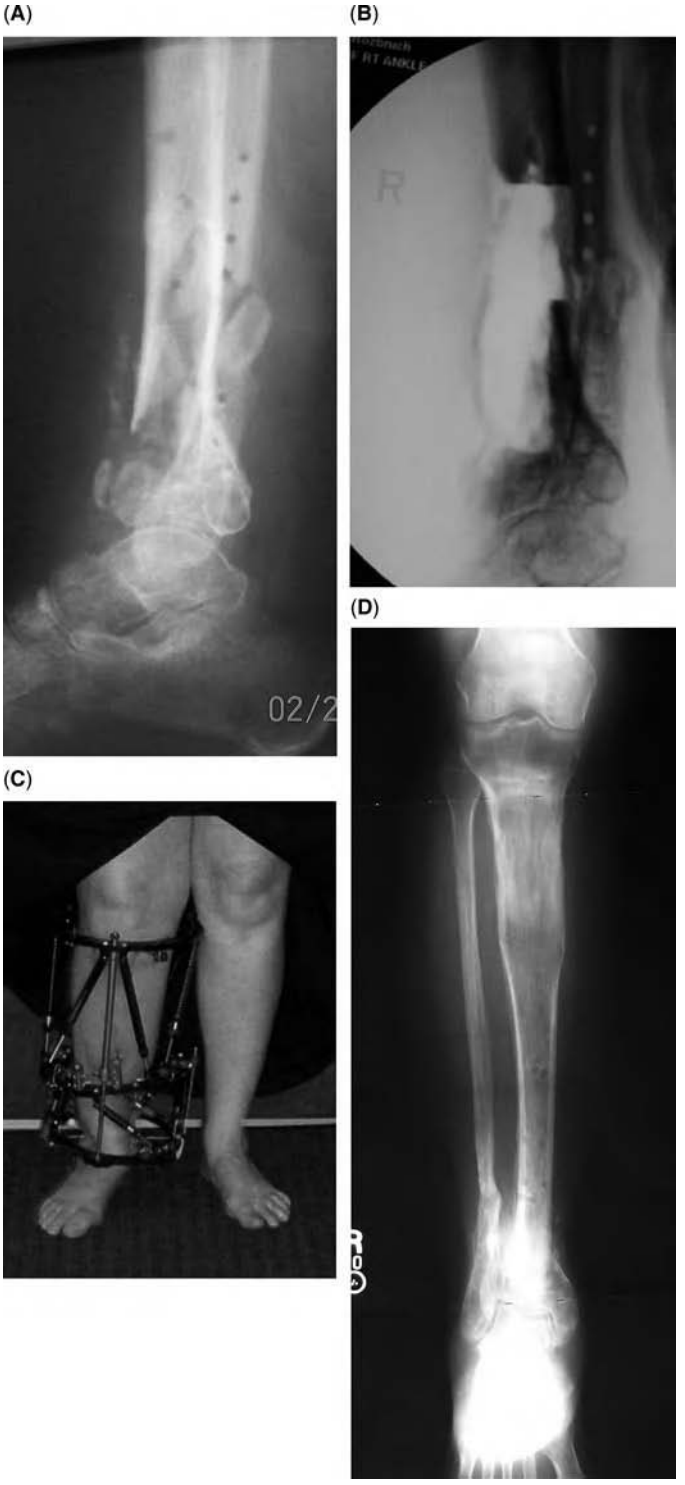

Figure 13 (**A**) Lateral X-ray showing an infected nonunion of the distal tibia. (**B**) Intraoperative X-ray after resection of dead infected bone. Note the 8 cm defect. (**C**) Standing front view showing a bone transport Ilizarov/Taylor spatial frame. (**D**) Anteroposterior radiograph three months after frame removal showing a successful bone transport with 8 cm proximal tibia lengthening and healed docking site at the distal tibia.

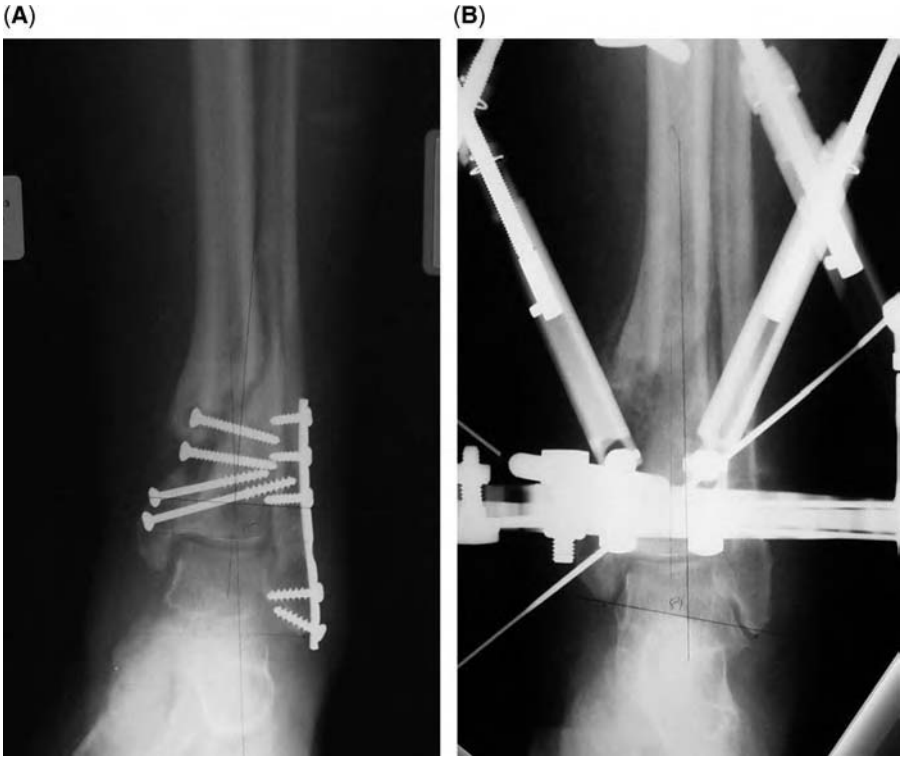

Figure 14 (**A**) Anteroposterior radiograph showing relative shortening of the tibia and a nonunion. (**B**) After gradual lengthening and deformity correction of the tibia with an Ilizarov/Taylor spatial frame.

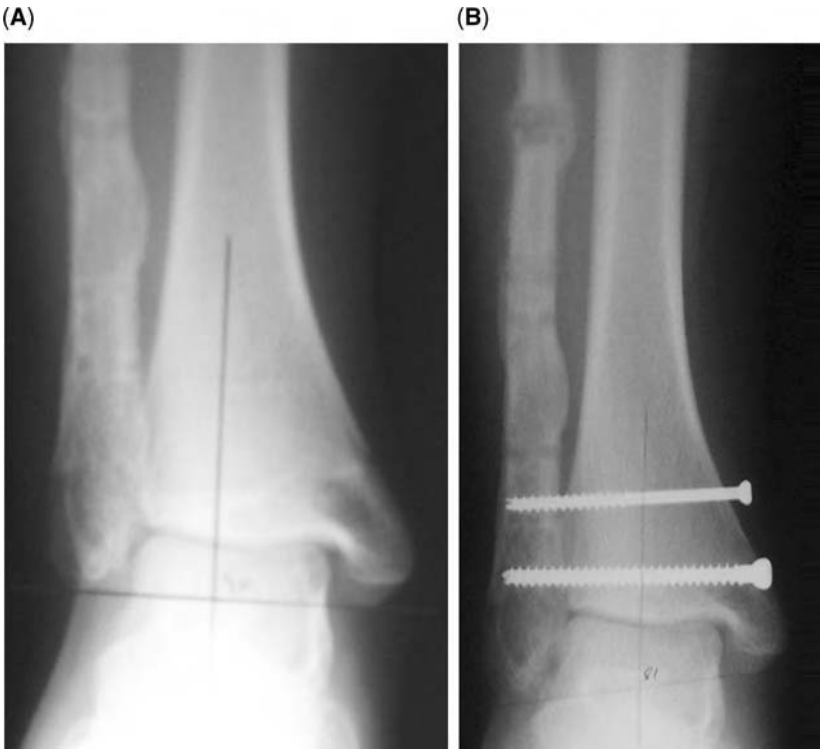

Figure 15 (**A**) Anteroposterior X-ray showing relative shortening of the fibula. (**B**) After fibula lengthening and insertion of syndesmosis screws.

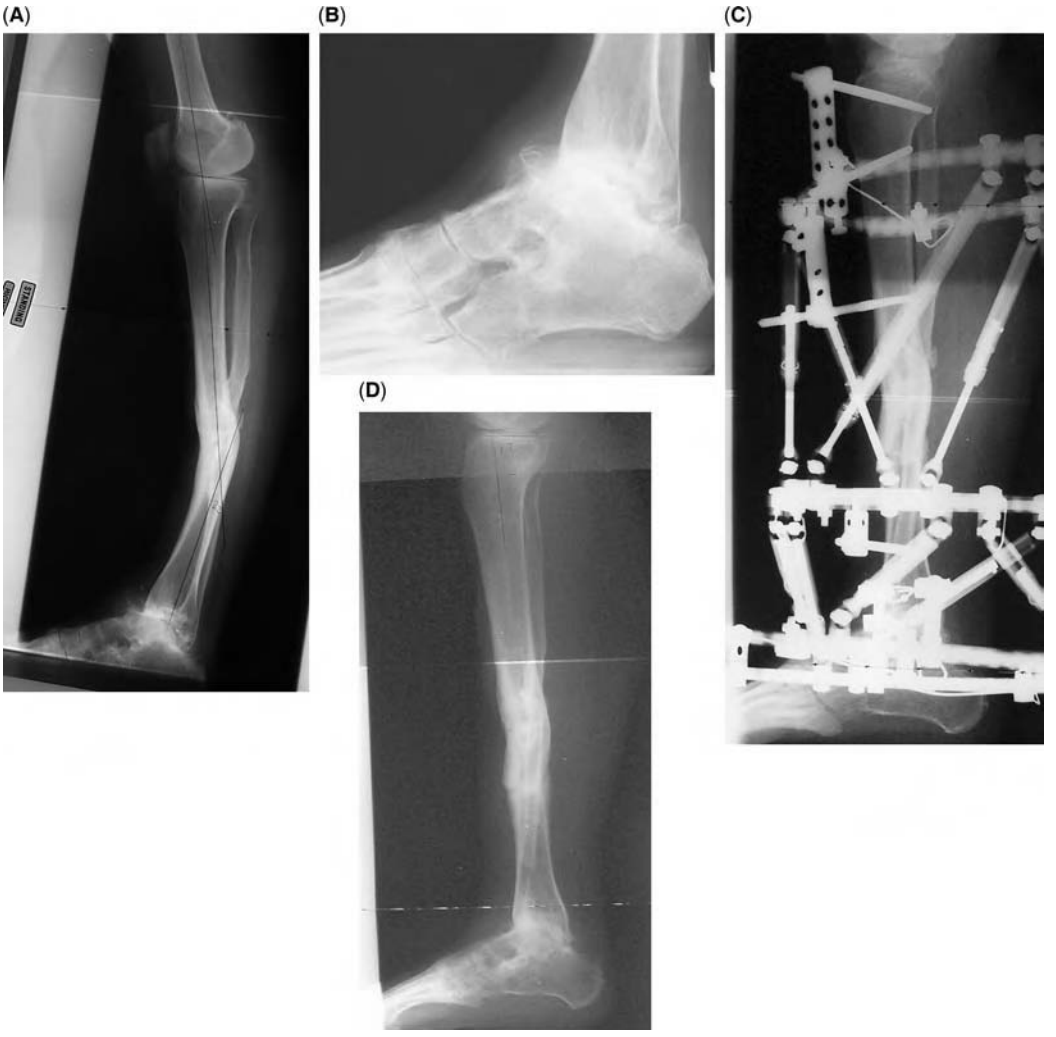

Figure 16 (**A**) Lateral X-ray showing a recurvatum malunion of the tibial shaft associated with arthrosis and contracture of the ankle. (**B**) Lateral X-ray of the ankle. (**C**) Lateral X-ray showing a two-level frame with correction of the tibia deformity and ankle arthrodesis. (**D**) Lateral X-ray six months after frame removal showing bony union and correction of deformity.

shortening is desirable with an ankle fusion, the patient can undergo a simultaneous 3 cm proximal tibia lengthening or use a shoe lift. The tibia to calcaneus contact can be achieved acutely or gradually (Fig. 19). Gradual shortening is safer in terms of neurovascular insult.

SURGICAL TECHNIQUES

The Ilizarov/TSF can have several levels as needed including proximal tibia, middle tibia, talus, and calcaneus. The number of levels used depends on the personality of the PTA (Fig. 2). A base of typically two rings is placed along the axis of the middle tibia. Each ring is usually fixed with two points of fixation with 1.8 mm smooth tensioned wires and/or 6 mm half-pins (Fig. 20). This can be modified to one ring if less fixation is needed. The remainder of the construct depends on the particular situation. A distal tibia ring is used for a supramalleolar osteotomy. A foot ring with hinges is used for ankle distraction or contracture correction. A foot ring with compression across the ankle is used for arthrodesis. A proximal tibia ring is for proximal tibia lengthening or deformity correction.

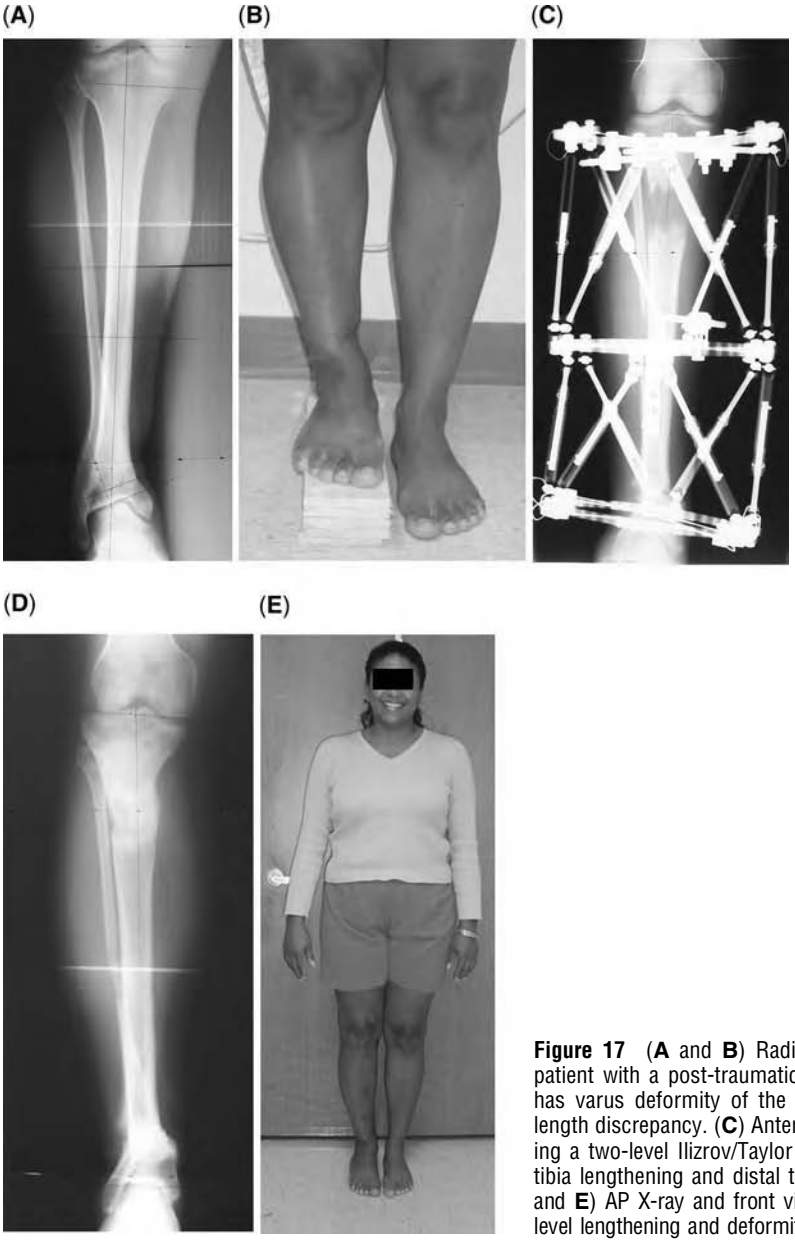

Figure 17 (**A** and **B**) Radiograph and front view of a patient with a post-traumatic growth arrest. This patient has varus deformity of the distal tibia and 6 cm of leg length discrepancy. (**C**) Anteroposterior (AP) X-ray showing a two-level Ilizrov/Taylor spatial frame with proximal tibia lengthening and distal tibia deformity correction. (**D** and **E**) AP X-ray and front view showing successful two-level lengthening and deformity correction.

Ilizarov Frame Considerations

The frame should be applied to the leg so that rings are perpendicular to the bone axis, the rods are parallel to the bone axis, and there is adequate clearance between the soft tissues and the rings especially at the posterior leg. The bone defect edges should be perfectly pointed toward each other to avoid deformity and to optimize contact at the anticipated docking site. If deformity should occur, this can be managed with frame modification and/or a surgical procedure to optimize contact at the docking site.

Taylor Spatial Frame Considerations

Rings are placed on either side of the defect site and the anticipated lengthening site(s). The rings can be placed independently to optimally fit the leg. This is called the rings first method. One ring is chosen as the reference ring for each level of movement, and it is important that

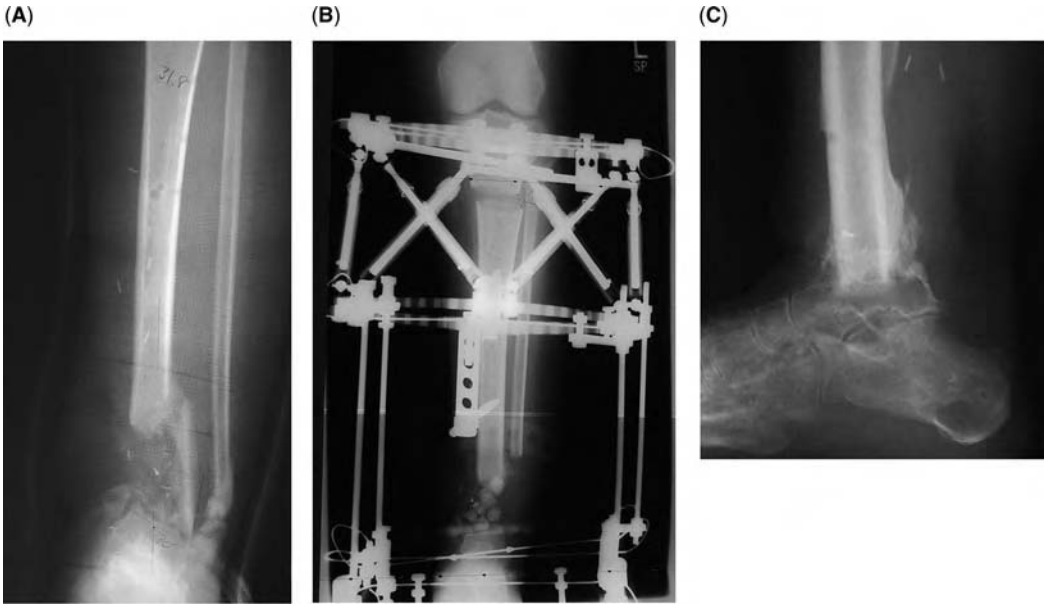

Figure 18 (**A**) Anteroposterior (AP) radiograph showing an infected nonunion bone defect of the distal tibia and articular surface. (**B**) AP radiograph showing a two-level frame for proximal tibia lengthening and gradual shortening of the defect and ankle fusion. (**C**) Lateral X-ray showing a successful ankle fusion. A 10 cm proximal tibia lengthening was performed in this patient.

this ring be placed orthogonal to the axis of the tibia. Mounting parameters are defined by the center of the reference ring and this will define the point in space where the deformity correction will occur. It is important to maintain enough distance between rings so that the struts can fit properly. In this frame, one is limited by the shortest length of strut. The advantages of this

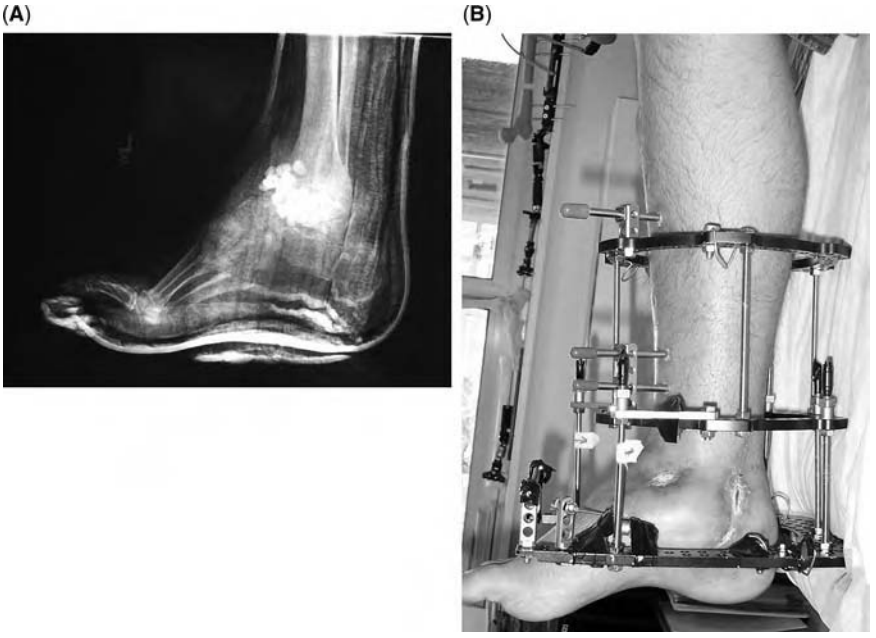

Figure 19 (**A**) Lateral X-ray showing antibiotic beads in defect after resection of the talus. This patient had septic osteonecrosis of the talus requiring resection. (**B**) Side view of the ankle arthrodesis frame applied after removal of the beads and preparation of the tibia and calcaneus for fusion. Gradual leg shortening was done and leg length discrepancy was treated by adjusting a contralateral amputation prosthesis.

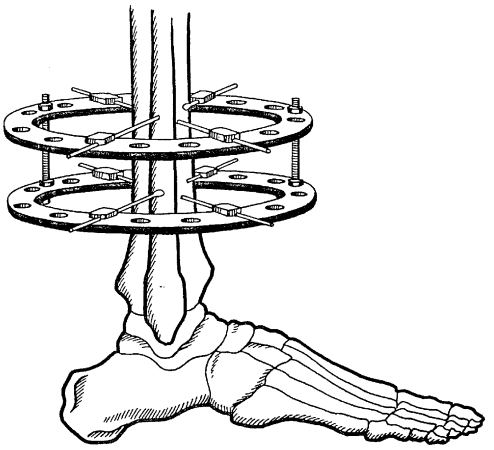

Figure 20 Schematic diagram of two Ilizarov rings used as a tibial base for many ankle corrections. *Source*: Courtesy of Arkady Blyakher.

frame is that the application is easier and the fit on the leg is better when using the rings first method. Also, residual deformity at the lengthening and docking sites can be addressed by using the same frame to correct angulation and translation simultaneously in the coronal, sagittal, and axial planes without major frame modification. This allows precise docking with optimal bone contact and minimizes angular deformity at the docking and lengthening sites (16).

Ankle Distraction

The frame includes a proximal circular ring placed about 8 cm above the ankle joint, a foot ring, and hinges at the ankle joint.

The proximal ring is positioned perpendicular to the axis of the tibial shaft. A temporary smooth K-wire is inserted through the talus from the center of the tip of the fibula and then directed to the center of the tip of the medial malleolus in a proximal and anterior direction. This is then checked under the fluoroscope to ensure proper placement. This is perhaps the most crucial portion of the procedure because this marks the true oblique axis of the ankle joint. This wire will mark the hinge position to allow the talus to move smoothly with in the mortise as it is distracted.

A foot ring is then secured to the hindfoot and midfoot by placing two smooth wires in an oblique fashion through the calcaneus and cuneiforms/cuboid, respectively. These are then tensioned and secured as described above. The foot ring is positioned parallel to the plantar surface of the foot. A transverse midfoot wire is inserted and tensioned to the ring.

Using the previously placed guide wire for the true axis of the ankle joint, two universal hinges are secured to the foot ring attached at points defined by the temporary joint axis wire. The joint axis wire may then be removed. The hinges are then secured to the proximal ring placed on the tibia using threaded rods and short connection plates. These rods should be perpendicular to the ankle in both the coronal and the sagittal planes. A compression/distraction rod is placed anteriorly to control ankle range of motion; thus completing the frame. The ankle joint is then taken through a range of motion under fluoroscopy to ensure smooth symmetric motion of the talus within the mortise. The ankle is then taken through a range of motion under fluoroscopy to check the amount of distraction as well as double check the alignment. Gradual distraction of 1 mm per day in four separate daily adjustments for one week is prescribed. A total of 6 to 7 mm of distraction is achieved.

Supramalleolar Osteotomy

The middle tibia ring block is applied. A distal tibia ring is fixed with two or three tensioned 1.8 mm wires and an anteromedial half-pin (just medial to the tibialis anterior tendon). The rings are applied to match the deformity. TSF struts are used to connect the rings across the deformity. After a percutaneous osteotomy (18) of the distal tibia and the fibula, a gradual correction of the deformity follows as per the Ilizarov method. If there is an ankle contracture, then a foot ring is placed and gradual correction can be done simultaneously. Hinges are

placed at the axis of the ankle as in the situation of an ankle distraction. A pulling rod can be placed anterior or a pushing rod posterior to motor the correction.

Ankle Arthrodesis

The ankle is approached in an open fashion, and the joint surfaces are prepared for arthrodesis. This involves removal of remaining cartilage, fibrous tissue, and correction of deformity. Wedge excision from the tibial plafond may be needed for deformity correction. In cases of an acute positioning, there should be excellent contact and alignment between the tibia and talus (or calcaneus). The position should be held with provisional wires placed from the bottom of the heel (Fig. 21A).

Compression between the middle tibia ring block and a foot ring is necessary. The foot ring is fixed to two oblique calcaneus wires and a midfoot wire. A forefoot wire can be added if the extra stability is needed. A talus wire can be used to protect the subtalar joint from compression in the situation of a tibiotalar arthrodesis (Fig. 21B). This is not needed for to tibio-talo-cancaneal arthrodesis or a tibio-calcaneal arthrodesis (Fig. 21C). Compression across the ankle is performed with longitudinal rods in line with the mechanical axis of the leg (Fig. 21D). If there is a desire for gradual correction through the arthrodesis site, this may be done with TSF struts.

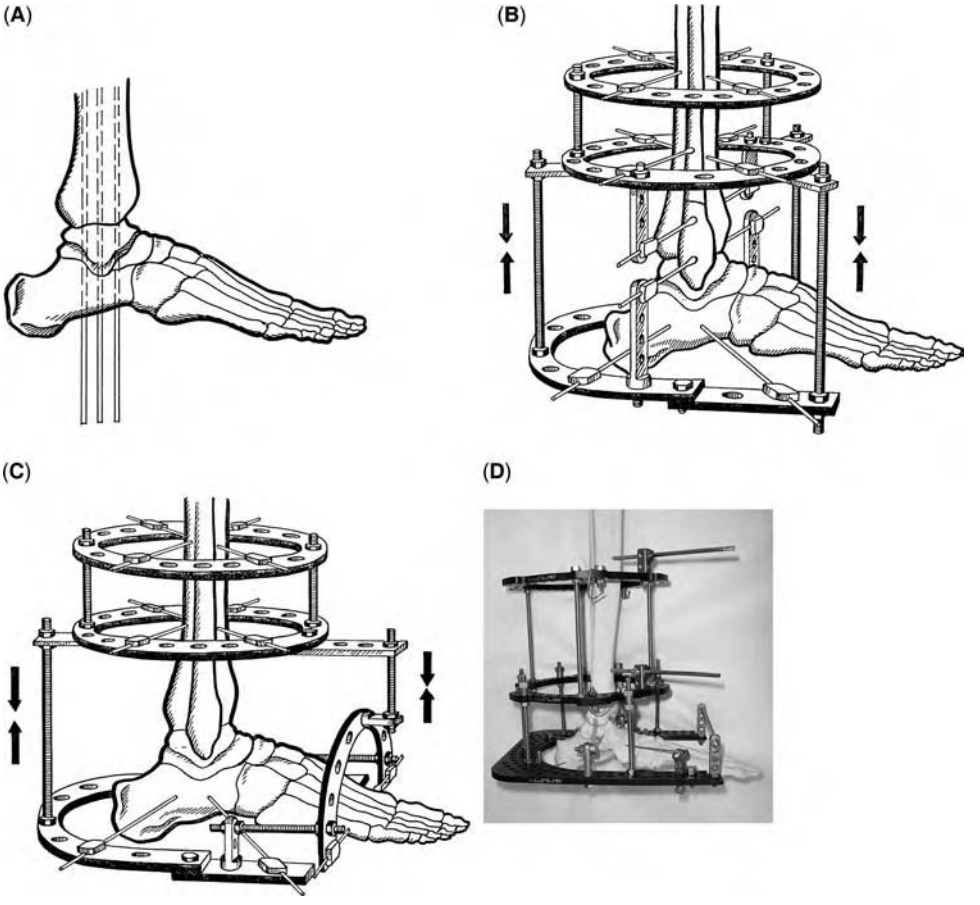

Figure 21 (**A**) Schematic diagram showing provisional fixation of ankle arthrodesis with axial wires prior to application of the Ilizarov/Taylor spatial frame. (**B**) Schematic diagram of an ankle arthrodesis frame. Note the talus wire that serves to prevent compression of the subtalar joint. (**C**) Schematic diagram of an ankle and hindfoot arthrodesis frame. There is axial compression from tibia to calcaneus. There is also option of compression across the talonavicular and calcaneocuboid joints. (**D**) Ankle arthrodesis frame mounted on saw bone. *Source*: Figures A, B, C, Courtesy of Arkady Blyakher.

Bone Transport for Infected Nonunions

All nonviable bone and soft tissue is debrided. Bone is debrided back to healthy appearing bone with open IM canals and with bleeding surfaces. This is best done without the use of a tourniquet. Bone cuts are typically made perpendicular to the anatomic axis of the tibia using a power saw cooled with saline irrigation. A K-wire placed with the help of fluoroscopy is used as a guide for the bone cut. In the adult patient, rings are applied with a combination of 1.8-mm Ilizarov wires and 6-mm hydroxyapatite-coated half-pins. Small-sized wires and half-pins may be used in children. The 1.8 mm wire is placed perpendicular to the axis of the bone in the coronal plane. The ring is attached with about two fingerbreadth spacing between the skin and the ring and the wire is tensioned to 130 kg. The half-pin is then placed setting the ring perpendicular to the sagittal plane bone axis. A ring block is either one or two rings. Each ring block should have a combination of three to four wires and/or half-pins. In situations where there is a very short proximal or distal tibia segment, consideration should be given to extending the fixation across the knee or ankle. This strategy is most commonly used for a short distal tibia segment with extension of the frame to the foot, at least temporarily.

With the monofocal approach, there is one level of activity. A ring block is applied both proximal and distal to the defect. The space between the innermost rings is chosen so that after docking there will be adequate room to approach the docking site for possible bone grafting or wound-revision surgery. Ideally, this space should be greater than 5 cm. Connecting rods or struts are placed between the innermost rings to be used for compression or gradual shortening. The fibula must have a defect that is comparable to the tibial defect. A modest amount of acute shortening may be done. Pulses should be checked to make sure that this does not cause any vascular compromise. Limb shortening will occur.

With the *bifocal* approach, there are two segments with activity. One segment (the defect) is undergoing compression/shortening, and one segment (the bony regenerate) is undergoing distraction/lengthening. This can maintain the length of the limb. A ring block is applied on either side of the bone defect. The space between the innermost rings is chosen so that after docking there will be adequate room to approach the docking site for possible bone grafting or wound-revision surgery. Connecting rods or struts are placed between the innermost rings to be used for compression or gradual shortening. Another ring block is placed on the other side of the anticipated lengthening osteotomy site. Rods or struts are applied across this segment and are set up for lengthening or distraction. The rods are then disconnected in preparation for the osteotomy. The osteotomy is done in a percutaneous fashion (48) using either the multiple drill hole and osteotome technique or the gigli saw technique. Care is taken to perform this osteotomy outside the zone of injury in healthy bone. Ideally, this osteotomy is done in the metaphyseal bone. The proximal metaphyseal location is preferable to the distal metaphysis because of increased bone regeneration potential (10).

In conclusion, the PTA has many faces. Following analysis of the personality of the ankle, one can implement a rational modular treatment approach. The Ilizarov method can be used to comprehensively address the PTA with distraction, correction of bony deformity and soft-tissue contracture, osteotomy, arthrodesis, and lengthening.

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18 Foot Deformity: Osteotomy or Arthrodesis

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INTRODUCTION

Foot deformity may result from a wide range of conditions including post-traumatic, degenerative, infectious, rheumatologic, diabetic, neurological, and congenital disorders. The patient may seek evaluation for symptoms including pain, limited gait, difficulty with footwear, and skin breakdown that may result in limb-threatening infection. Some feet have such extensive soft-tissue compromise or infection that foot salvage is impossible. Nevertheless, deformity correction and foot salvage are often preferred over amputation because of the decreased energy of walking with a natural than a prosthetic leg and body image difficulties that may be associated with amputation.

Deformity in the foot may be corrected with either osteotomy or arthrodesis. Osteotomy can be performed as an acute correction or gradually with external fixation and distraction osteogenesis techniques. Arthrodesis is usually recommended in the presence of arthritis, neuroarthropathy, chronic dislocation or subluxation, and in some cases of joint ankylosis with associated pain. If these conditions are not present, it may be preferable to salvage hindfoot motion to maintain as normal a gait pattern as possible and minimize the potential for progressive degenerative changes in joints adjacent to an arthrodesis. Arthrodesis and osteotomies can also be combined to correct multiple deformities often present in the foot.

CLINICAL EVALUATION

Patient evaluation includes a thorough history and physical examination. Pertinent findings in the history include medical conditions such as diabetes, cardiovascular disease, renal failure, and tobacco, drug, or alcohol abuse. Other important issues include prior trauma, infection, and surgery. It is necessary to ascertain the psychosocial situation of the individual, including available support systems and ability to be compliant with a treatment regimen. Furthermore, occupation, hobbies, and recreational activities are considered in the decision-making process.

Physical examination includes evaluating range of motion of the ankle, subtalar, Chopart's, Lisfranc's, and metatarsophalangeal joints. A neurovascular examination includes testing of motor function and sensation. The soft tissues are evaluated for flaps, skin grafts, scars, incisions, vascular changes, and ulcers. Bony prominences are noted and correlated with sites of pain or ulcer. Joint stability is assessed clinically and may be confirmed with regular and stress radiographs. Foot deformity such as cavovarus, flatfoot, equinovarus, or rockerbottom deformity is noted, and degree of flexibility or joint contracture is assessed. Alignment of the hindfoot, midfoot, and forefoot is evaluated in the seated and standing positions and during gait. Many patients with foot deformity have some degree of equines deformity; this may be caused by several factors such as bony impingement from anterior ankle osteophytes, joint ankylosis or arthritis, flattop talus (which restricts the normal sagittal roll of ankle joint motion), or soft-tissue contracture (such as the joint capsule contracture, gastrocsoleus muscle complex tightness, or posterior leg compartment fibrosis).

The components of deformity should be defined. These include the ankle/supramalleolar region, the hindfoot, and the forefoot. Ankle and supramalleolar deformity will affect the foot as a whole. Correction in this region can be expected to move the foot as a unit. Hindfoot

and forefoot deformity can be in the same direction (hindfoot varus and forefoot supination) or can be compensatory (hindfoot varus and forefoot pronation). In addition, the compensation can be flexible or rigid. One has to use this information to decide which components of the deformity require correction. At times, the hindfoot and forefoot require correction in same direction and the foot is moved as a unit. Other times, the hindfoot and forefoot require correction in different directions and these areas must be moved independently. With flexible compensatory deformity, the fixed deformity is corrected and the flexible component will accommodate.

Radiographic evaluation of foot deformity includes weight-bearing ankle, hindfoot, and foot radiographs. Standing ankle and tibial radiographs may demonstrate deformity of the ankle or leg that may affect the treatment plan. If a rotational deformity of the limb is present on clinical examination, an anteroposterior (AP) ankle radiograph is made in the foot-forward position to evaluate intra-articular wear or malalignment. Lateral ankle radiographs are made in the plane of the ankle malleoli. If a frontal plane deformity is present, the true lateral radiograph is made by angling the beam to match the deformity. The hindfoot alignment view is a weight-bearing radiograph that enables observation of the tibia, ankle joint, and calcaneal tuberosity on a single view; it is the only radiograph that requires a specialized mounting box to angle the radiographic plate 20° from the vertical plane (1). An alternative radiograph is the long axial view, which usually is made nonweight bearing and visualizes the tibia, subtalar joint, and calcaneal tuberosity. A line drawn on the vertical axis of the midbody of the calcaneus should be parallel and approximately 1 cm lateral to the mid-diaphyseal line of the tibia (Fig. 1). Valgus deformity and lateral translation indicate a pes planus deformity; varus angulation and medial translation indicate cavovarus-type deformity.

The weight-bearing AP foot radiograph is measured for talo-first metatarsal angle, navicular coverage, and joint subluxation or arthritis. The lateral foot view is measured for talo-first metatarsal angle, calcaneal pitch, and joint subluxation or arthritis. If the opposite limb is pain free and has no obvious deformity, comparison radiographs may be made for preoperative planning. The oblique nonweight-bearing view may reveal a calcaneonavicular coalition. Computed tomography may be helpful in evaluating nonunion of fracture or previous fusion, tarsal coalition, degenerative joint disease, and tibial rotational deformities. Magnetic resonance imaging may be useful in the diagnosis of osteomyelitis, tumors, or tendon abnormalities.

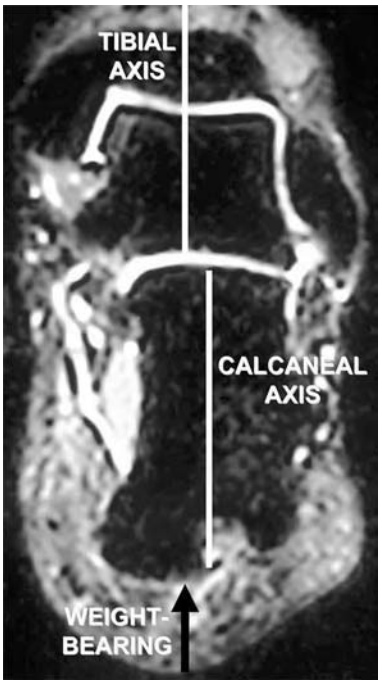

Figure 1 A line drawn on the vertical axis of the midbody of the calcaneus should be parallel and approximately 1 cm lateral to the mid-diaphyseal line of the tibia.

CLASSIFICATION

Deformities are classified on the basis of underlying disease, alignment of hindfoot and forefoot, and location of bony prominences.

The specific disease process may be associated with characteristic deformities. Charcot–Marie–Tooth disease is associated with weakness of the tibialis anterior, peroneus brevis, and intrinsic muscles with intact peroneus longus and tibialis posterior muscles (2); the cavovarus deformity may include heel varus, cavus, plantarflexed first ray, tight plantar fascia, and claw toes. The intact peroneus longus may contribute to hindfoot rigidity and plantarflexed first ray (3), resulting in cavus deformity and secondary contracture of the plantar fascia, and the intact tibialis posterior may contribute to heel varus. Stroke may be associated with equinovarus deformity resulting in weight bearing on the lateral border of the foot. Rockerbottom deformity with plantar bony prominence that may lead to ulceration can result from triple arthrodesis malunion or neuropathic collapse. Rheumatoid arthritis may cause a severe, rigid planovalgus deformity. Post-traumatic or iatrogenic deformity may be varied depending on the injury.

The relationship between hindfoot and forefoot is important in the normal function of the foot. Compensatory deformities in the foot and ankle are common. Hindfoot varus deformity (e.g., cavovarus foot, Charcot–Marie–Tooth disease, or calcaneal fracture) may be compensated by pronation in the forefoot (forefoot valgus) and plantarflexion of the first ray with associated rise of the arch of the foot. In this case, correction of the hindfoot varus alone may aggravate the forefoot valgus unless forefoot malalignment is also corrected.

Hindfoot valgus deformity (e.g., acquired flatfoot or posterior tibial tendon dysfunction) may result in dorsiflexion of the medial column and a forefoot supination deformity (forefoot varus); the medial column dorsiflexion deformity may occur through the spring ligament complex and talonavicular joint, the naviculocuneiform joints, or the first tarsometatarsal joint.

TREATMENT OPTIONS

The goal of foot deformity correction is to achieve a plantigrade foot that fits into functional footwear for ambulation with minimal pain and avoidance of ulceration. The method selected depends on the many factors delineated in the history and physical examination. The decision-making process also relies on individual patient characteristics including age, personality, functional goals, and expectations. The physician and staff must spend sufficient time establishing a relationship with the patient so that the risks and benefits of surgery are understood and the patient feels he or she has made a well-informed decision.

Gradual vs. Acute Correction

Acute correction of deformity with osteotomy or arthrodesis is performed when acute realignment achieves a plantigrade foot with an acceptable degree of shortening (Fig. 2). When correction is achieved with arthrodesis or closing wedge osteotomy, the surgical wound usually is closed without tension. However, if the foot is short and a lengthening is desired, acute correction may be associated with excessive wound tension and increased risk of wound problems or neurovascular compromise. In such cases, gradual correction may allow for soft-tissue lengthening and minimize the potential for wound or neurovascular problems.

Gradual correction can occur through a joint or through an osteotomy (4). Gradual correction through an osteotomy with distraction osteogenesis is indicated when acute correction would result in minimal bone contact at an osteotomy site, potentially increasing the risk of nonunion. A scarred, grafted, or multiply operated foot with compromised soft tissues also is an indication for gradual correction.

Some deformities, such as residual clubfoot, acquired equinovarus, pes cavus, or severe equinovalgus, may be corrected gradually through the deformed joints. This usually is performed in younger patients (less than eight years old), but successful correction can be achieved for some cases of untreated clubfoot, post-traumatic, or neurologic equinovarus deformity in older adolescents and adults (5–9). In these cases in the older patient, correction may result in foot stiffness, and may require limited arthrodesis or tendon transfer to prevent recurrence.

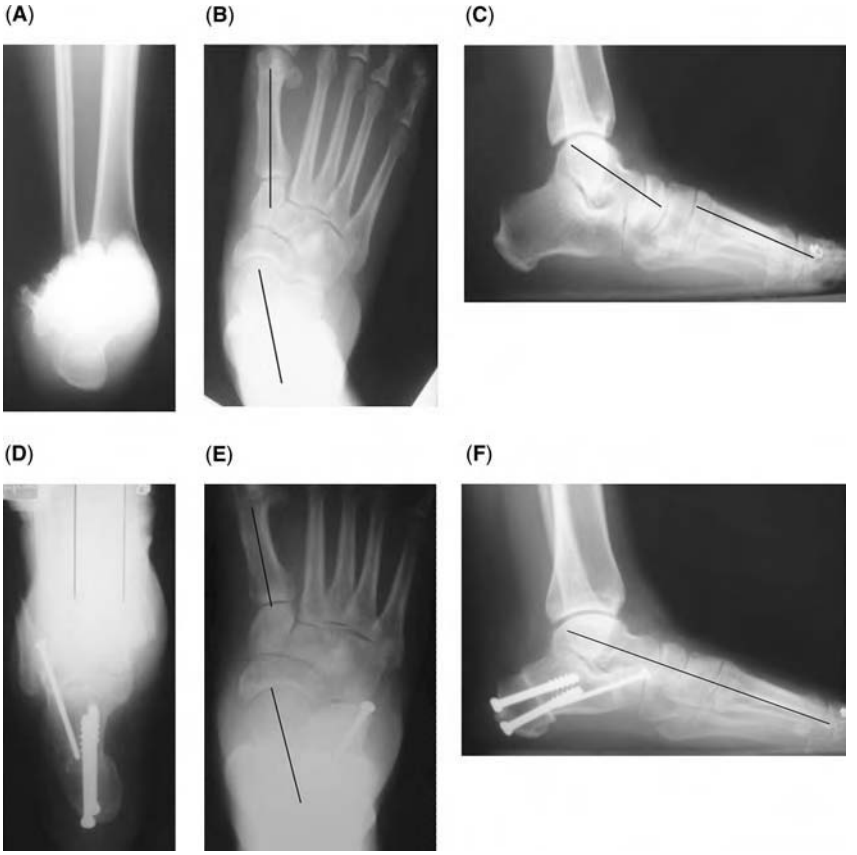

Figure 2 A 45-year-old healthy woman with acquired flatfoot and posterior tibial tendon dysfunction resulting in hindfoot pain. (**A**–**C**) Preoperative long axial, anteroposterior (AP) foot, and lateral foot radiographs show heel valgus, forefoot abduction, and plantarflexed talo-first metatarsal angle, respectively. (**D**–**F**) Treatment included an acute medial displacement calcaneal osteotomy; Evans lateral calcaneal osteotomy with tricortical, iliac crest bone graft; flexor digitorum longus tendon transfer to the navicular; and gastrocsoleus recession. No frame was used. Postoperative axial heel, AP foot, and lateral foot radiographs show union of the osteotomies and correction of the preoperative heel valgus, forefoot abduction, and talo-first metatarsal alignment, respectively.

Arthrodesis vs. Joint Preservation

Arthrodesis is indicated for treatment of degenerative joint disease, ankylosis, and some cases of motor insufficiency, severe foot joint instability, chronic dislocation, or joint malalignment. A more normal gait pattern may be achieved with joint preservation than arthrodesis if pain-free motion is achieved. Joint preservation for deformity correction in the foot is usually accomplished with osteotomies, soft-tissue procedures, and joint distraction. Deformity correction can be achieved or improved with arthrodesis of the midfoot joints that normally have little motion. Arthrodesis of the ankle joint increases stress in the subtalar and talonavicular joints and this may cause progressive degenerative changes (10).

External Fixation

External fixation, with or without internal fixation, may be indicated in some cases of acute osteotomy correction or arthrodesis (11,12). Patients may have poor bone quality or bone stock due to trauma, previous infection requiring bone debridement, osteopenia from prolonged nonweight bearing, failed internal fixation with sizable defects that require bone grafts, or neuropathic arthropathy. In these situations, internal fixation with screws, plates, or nails may fail to provide adequate fixation, but external fixation may provide stability required for union.

The external fixator may be used as a static or holding frame when combined with internal fixation that is sufficient to provisionally stabilize the arthrodesis or osteotomy.

Compression can still be applied across the arthrodesis or osteotomy site to improve stability and healing potential. External fixation may allow the patient to mobilize more safely for bed-to-chair transfer and possibly an earlier return to partial and full weight bearing to minimize further atrophy of the limb.

It is important in these cases that all external fixation wires and half-pins in the foot and leg remain separated from the internal fixation devices in the foot to avoid bacterial contamination of the internal fixation from colonized external fixator pin tracks. This can be difficult in the foot and often requires the use of strategically placed wires with knowledge of Ilizarov techniques. The internal fixation devices must be placed to avoid the external fixation wires, and often are smaller than those used without concomitant external fixation.

Gradual deformity correction with external fixation usually is performed through an osteotomy, but also can be done through severely deformed joints (12). When there is a chronic joint dislocation, gradual deformity correction through the joint can be done as the first stage of a realignment arthrodesis. Correction of severe foot deformities can be done through percutaneous or limited open osteotomies, with maintenance of foot length.

SURGICAL TECHNIQUES

Arthrodesis Techniques

The joints to be fused are exposed and debrided down to healthy, bleeding, cancellous bone, with excision of residual joint surfaces and fibrous debris. Deformity correction involves realignment to restore a neutral relationship between the leg, ankle, hindfoot, and forefoot. In some cases, acute realignment may be facilitated by placing a temporary Kirschner wire orthogonal to the longitudinal axis of the hindfoot and another orthogonal to the longitudinal axis of the forefoot; neutral forefoot to hindfoot alignment is achieved by rotating the forefoot relative to the hindfoot until the Kirschner wires are parallel to each other. If shortening is done, wedge osteotomy may create bone-to-bone apposition at the arthrodesis site; if acute lengthening is done, a structural, corticocancellous bone graft (iliac crest or allograft) may be useful. Fixation is accomplished with internal fixation hardware (a combination of wires, screws, staples, or plates) or external fixation. Alignment and position of fixation devices are evaluated intraoperatively with C-arm fluoroscopic radiographs. Cancellous bone graft (proximal or distal tibial, iliac crest, or allograft) may be indicated to improve potential for union, especially in fusion sites having poor vascularity. Recombinant bone morphogenic proteins are used in selected cases pending further clinical trials.

Osteotomy Techniques

Osteotomy techniques in the foot vary according to location and whether the osteotomy is done for acute or gradual correction of deformity. The primary techniques use multiple drill holes, Gigli saw, microsagittal saw, and osteotome. For acute correction, the microsagittal saw and osteotome may be satisfactory. For gradual correction with distraction osteogenesis, it is important to minimize periosteal damage and dissection, preserve the periosteal blood supply, and minimize thermal necrosis of bone by avoiding use of saws. The osteotomy is preferably done through trabecular, vascularized bone, and sclerotic bone is avoided, if possible, because of poor healing potential.

The multiple drill hole technique is preferred in the calcaneus, which may be approached laterally or medially. The lateral approach to the calcaneal tuberosity is most commonly used and is made posterior to the peroneal tendons. Multiple drill holes are made from lateral to medial with a 3.5 or 4.5 mm drill, and the osteotomy is completed with an osteotome. The sural nerve is at risk for injury or postoperative scarring and neuroma formation. For gradual correction in adults, calcaneal osteotomies are distracted 0.5 to 1 mm per day.

The Gigli saw technique is useful in the midfoot. The osteotomy usually is performed from medial to lateral, but also may be done from lateral to medial. Four incisions are made: dorsomedial, dorsolateral, plantarmedial, and plantarlateral. The Gigli saw is passed plantar to the bony arch, then from plantar to dorsal, and then dorsal to the bony arch, carefully staying close to bone to minimize potential for neurovascular injury. This technique may be used in the region between the talar neck and the tarsometatarsal joints. The Gigli saw also may be useful for open procedures with internal fixation when bone wedges are removed from the

midfoot, as in rockerbottom deformity correction. For gradual correction in adults, midfoot osteotomies are distracted 0.75 mm/day.

Ring Fixator Frame Design and Stability

A well-designed frame should provide stability, allow soft-tissue clearance, and permit weight bearing as soon as possible. Wires are placed carefully to avoid neurovascular structures. Wires or half-pins are not placed near poor skin or open wounds to prevent a simple pin-tract infection from communicating to the open wound. A mature, stable skin graft does not increase pin-tract infections and may be used as a pin site.

Frame stability in the foot is a challenge due to the limits of anatomic size. A minimum of two and preferably three points of fixation are needed in each segment of the foot. This demands carefully planned wire placement and time-consuming methods to fix the wires to the rings. During distraction, an osteotomy must have fixation on either side to prevent joint subluxation.

Pin placement in sclerotic bone is avoided, if possible, because of the greater potential for pin-tract osteomyelitis in avascular bone. Bone with severe osteopenia is better stabilized with Ilizarov wires in multiple planes than half-pins. Spatial frame struts, connecting rods, and rings are placed to allow access to wounds. A footplate with a rocker sole is attached to the foot ring as soon as weight bearing is allowed.

In many complex foot deformity cases, Ilizarov frames with or without struts (Taylor Spatial Frame Struts, Smith & Nephew Inc., Memphis, Tennessee, U.S.A.) do not allow weight bearing. In these cases, a frame modification to a standard foot ring may be performed after deformity is corrected and initial osteotomy healing has occurred. During the frame modification, the osteotomy may require temporary pinning to maintain position. In rare cases, special foot plates can be adapted to these frames that attach to the tibial frame to allow pivot transfers.

Acute Tarsal Tunnel Syndrome and Poor Skin Quality

In the foot, the posterior tibial nerve is the most common nerve at risk during acute or gradual correction. The nerve may be impinged in the tarsal tunnel during correction of equinovarus deformity of the ankle and hindfoot, and tarsal tunnel release may be indicated. It is important to anticipate and plan for a surgical incision for this procedure.

SPECIFIC DEFORMITIES

Pes Planus

The etiology, severity, patient age, and functional status of pes planus deformity will dictate the optimal procedures for correction (13,14). The most common etiology in the adult is acquired flatfoot associated with posterior tibial tendon insufficiency (15). Earlier stages of disease, associated with flexible deformity that is passively correctible, may be corrected with osteotomy techniques and joint preservation or limited arthrodesis. For mild hindfoot valgus without major midfoot abduction, a calcaneal osteotomy with medial displacement (1 cm) of the posterior calcaneal tuberosity may be combined with a flexor digitorum longus tendon transfer to the navicular or medial cuneiform. In cases with midfoot abduction and hindfoot valgus, an opening wedge osteotomy (Evans procedure) of the anterior process of the calcaneus may be combined with the tendon transfer, with or without the medial displacement calcaneal osteotomy (Fig. 2). Associated forefoot varus may be corrected with a medial column (naviculo-medial cuneiform and/or first tarsometatarsal) arthrodesis, especially when there is joint hypermobility or subluxation. Forefoot varus correction also may be achieved with a dorsal opening wedge osteotomy of the medial cuneiform.

All patients should be assessed for a gastrocnemius contracture, which may be corrected with a gastrocnemius recession if there is the inability to reach 0° to 5° of dorsiflexion after deformity correction. The use of a gastrocnemius recession or Achilles lengthening must be carefully considered because push-off weakness may result. There is a prolonged recovery period of approximately 6 to 12 months for complete rehabilitation following these reconstructions. Hindfoot arthrodesis procedures may be indicated in the less active, elderly patient who

desires a pain-free stable hindfoot. Isolated subtalar fusion is indicated for hindfoot valgus deformity with forefoot varus of less than 10°.

In later stages of adult acquired flatfoot, the deformity can be rigid and severe, and a hindfoot arthrodesis may be the surgical treatment of choice. Hindfoot valgus is corrected by realigning the calcaneus under the talus prior to fusing the subtalar joint. Forefoot varus is corrected through the transverse tarsal articulation, and occasionally midfoot. Severe flatfoot deformity associated with peritalar subluxation may be corrected with lateral column lengthening (distraction calcaneocuboid arthrodesis) and flexor digitorum longus transfer, but complication and reoperation rates are high (16). These corrections usually are accomplished acutely, but gradual correction with external fixation may be indicated to reduce the potential for lateral wound dehiscence in cases with marked forefoot abduction, hindfoot valgus, and associated lateral soft-tissue shortening.

A double arthrodesis (talonavicular and calcaneocuboid) or isolated talonavicular arthrodesis with medial displacement calcaneal osteotomy is indicated when hindfoot deformity is associated with forefoot varus exceeding 10° or with marked midfoot abduction. These procedures are considered if there is a reducible subtalar joint free of arthritis, and subtalar motion is limited by the talonavicular arthrodesis. The talonavicular arthrodesis and calcaneal osteotomy procedure usually provides a greater correction of hindfoot valgus than the double arthrodesis. The potential to correct deformity using the double arthrodesis can be improved by performing a distraction bone block arthrodesis through the calcaneocuboid joint, thus lengthening the lateral column of the foot. This method is preferred in the most severe cases when the subtalar joint remains reducible. A triple arthrodesis is indicated in those cases where the subtalar joint is rigidly fixed in valgus with associated forefoot varus or abduction. Double arthrodesis may be complicated by nonunion and residual deformity, and salvage may include acute, gradual, or combined correction depending on the severity of the component deformities (Fig. 3).

With severe cases of adult acquired flatfoot, other aspects of deformity are often present. These include equines contracture, midfoot deformity (naviculocuneiform or tarsometatarsal), ankle arthritis, deltoid ligament insufficiency, and global ankle ligamentous insufficiency. Total ankle replacement may be indicated for treatment of ankle arthritis associated with hindfoot arthrodesis.

Flatfoot in children usually is bilateral and asymptomatic and rarely is surgical correction indicated. A complete discussion of symptomatic flatfoot in children and adolescents is beyond the scope of this chapter (13). Painful flexible flatfoot may be corrected acutely with a medial displacement calcaneal osteotomy or an anterior process calcaneal osteotomy (Evans procedure) as in the adult flatfoot, with or without posterior tibial tendon advancement, flexor digitorum longus tendon transfer to the navicular, plication of the spring ligament complex, and gastrocnemius recession or Achilles tendon lengthening. Rigid flatfoot is often associated with a tarsal coalition. Surgical treatment can include hindfoot arthrodesis or coalition resection, with associated deformity correction. Arthrodesis is generally preferred for large talocalcaneal coalitions or those with an arthritic subtalar posterior facet. Isolated subtalar arthrodesis is usually preferred to a triple arthrodesis except in cases of transverse tarsal joint arthritis or instability. Correction of the flatfoot deformity should accompany the arthrodesis. Valgus deformity may be corrected through the subtalar joint or with a concomitant medial displacement calcaneal osteotomy.

Other causes of flatfoot include post-traumatic deformity or arthritis, degenerative or inflammatory arthritis, congenital deformity, and persistent deformity after triple arthrodesis. Salvage of residual deformity after triple arthrodesis can be achieved with a revision of the triple arthrodesis or V-type osteotomy through the fusion mass. The level of this osteotomy may be modified based on the location of the deformity. Acute correction may be done for less severe deformity, but many cases are best treated with gradual correction (Fig. 4). Acute correction can be done with internal fixation for either the hindfoot or midfoot, with gradual correction for the other component of the deformity.

Cavus Foot

Surgical management of the cavus foot is complex and must be tailored to the specific deformity of the patient. Most cavus feet develop from muscle imbalance and are associated with neurologic disorders in approximately two-thirds of cases (17). Progressive deformities yield

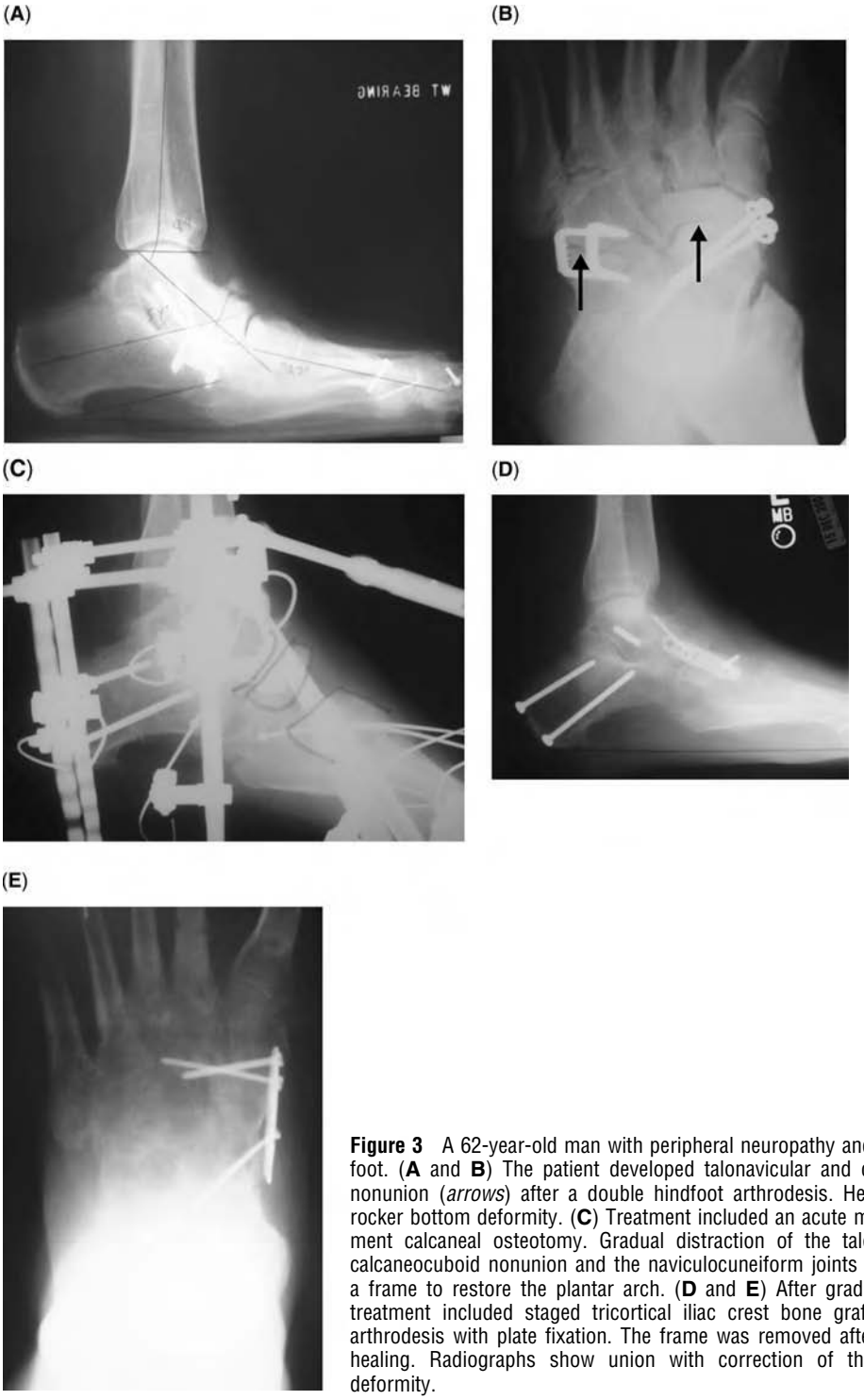

Figure 3 A 62-year-old man with peripheral neuropathy and acquired flatfoot. (**A** and **B**) The patient developed talonavicular and calcaneocuboid nonunion (*arrows*) after a double hindfoot arthrodesis. He had a painful rocker bottom deformity. (**C**) Treatment included an acute medial displacement calcaneal osteotomy. Gradual distraction of the talonavicular and calcaneocuboid nonunion and the naviculocuneiform joints was done with a frame to restore the plantar arch. (**D** and **E**) After gradual distraction, treatment included staged tricortical iliac crest bone graft and midfoot arthrodesis with plate fixation. The frame was removed after radiographic healing. Radiographs show union with correction of the preoperative deformity.

both higher recurrence rates and poorer long-term outcomes with triple arthrodesis (18,19). The majority of cavus deformities involve the forefoot with a plantarflexed first ray as seen in Charcot–Marie–Tooth disease or idiopathic cavus foot. Less commonly, the hindfoot can create cavus deformity, with increased calcaneal pitch noted on a lateral radiograph resulting from gastrocsoleus weakness as observed in poliomyelitis.

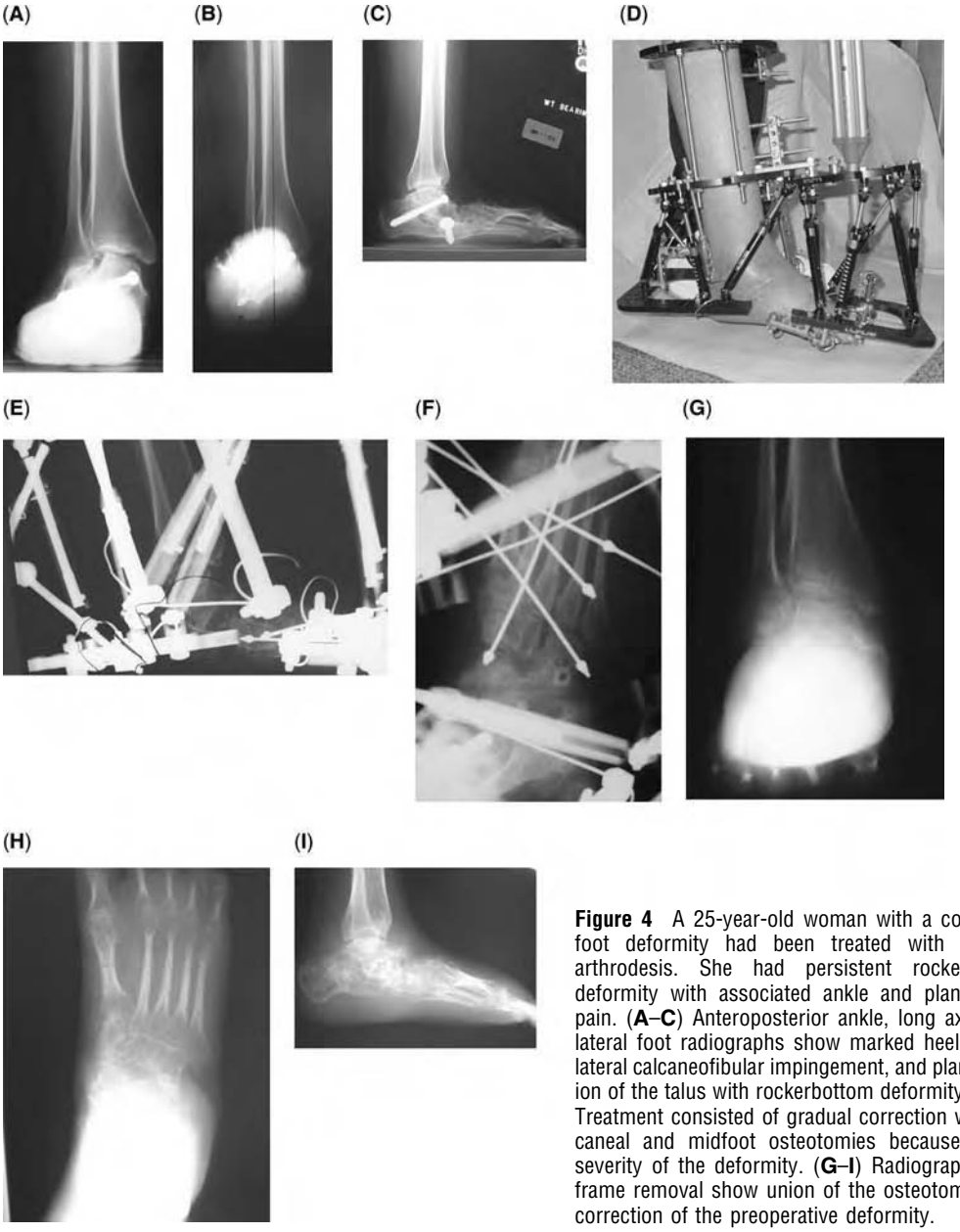

Figure 4 A 25-year-old woman with a congenital foot deformity had been treated with a triple arthrodesis. She had persistent rockerbottom deformity with associated ankle and plantar foot pain. (**A**–**C**) Anteroposterior ankle, long axial, and lateral foot radiographs show marked heel valgus, lateral calcaneofibular impingement, and plantarflexion of the talus with rockerbottom deformity. (**D**–**F**) Treatment consisted of gradual correction with calcaneal and midfoot osteotomies because of the severity of the deformity. (**G**–**I**) Radiographs after frame removal show union of the osteotomies and correction of the preoperative deformity.

Evaluation includes assessment to distinguish between the fixed and flexible components of the deformity (20). Forefoot-driven deformity is determined by the Coleman block test (21) (Fig. 5). The heel and lateral forefoot are supported on a block while the first to third rays are allowed to drop off the medial side. If the heel corrects out of varus to neutral alignment, then surgical correction can be directed at the valgus forefoot. The plantarflexed first ray may be treated with a dorsiflexion closing wedge first metatarsal osteotomy or first tarsometatarsal joint arthrodesis. Hyperextension of the first metatarsophalangeal joint with associated claw hallux deformity is treated with a hallux interphalangeal joint arthrodesis and extensor hallucis longus tendon transfer to the distal first metatarsal (Jones procedure). Lesser claw toes are realigned with extensor tenotomies, metatarsophalangeal joint capsulotomies, and proximal interphalangeal joint condylectomies. During clawtoe correction, extensor hallucis longus and extensor digitorum longus tendon transfers to the midfoot may add ankle dorsiflexion power to lessen foot drop symptoms.

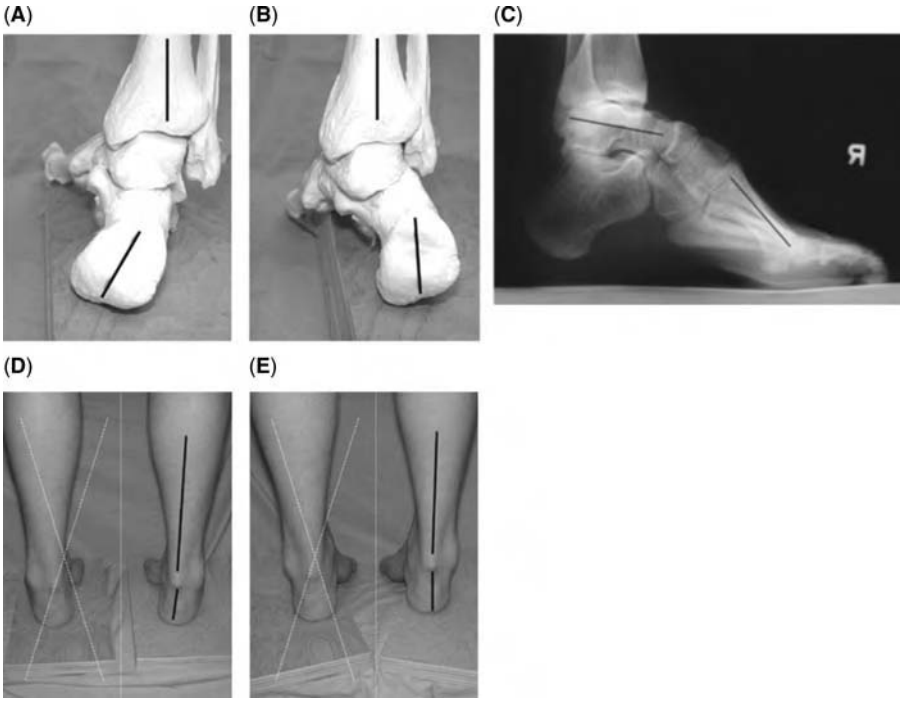

Figure 5 Coleman block test. (**A** and **B**) Foot model simulating cavus foot with flexible hindfoot. With the entire foot on the block, heel varus is noted. When the medial forefoot overhangs the medial edge of the block, the heel corrects out of varus to neutral alignment. (**C**) Lateral radiograph of an adolescent man with Charcot–Marie–Tooth disease. High calcaneal pitch and plantarflexion of the first metatarsal relative to the talus are noted. (**D** and **E**) Coleman block test shows that his hindfoot is flexible and the heel varus deformity corrects to neutral when the medial forefoot overhangs the medial edge of the block.

Muscle imbalance is the key component to the development of the cavus deformity; therefore, tendon lengthening, release, or transfers frequently are added to assist in correction of the deformity and to prevent recurrence. The most common soft-tissue procedure is release of the contracted plantar fascia. In cases of severe deformity, the contracted skin may prevent the deformity from being corrected unless shortening of the lateral column is performed acutely to decrease the risk of wound dehiscence and infection. Such cases may be better managed with gradual correction using distraction osteogenesis.

For rigid hindfoot deformity, lateral closing wedge calcaneal osteotomy (Dwyer procedure) may correct deformity and spare subtalar motion, especially with mild-to-moderate deformities (22). Subtalar or triple arthrodesis is reserved for severe, rigid deformity with subtalar arthritis or marked limitation of subtalar motion. With Dwyer osteotomy, an 8-to-12 mm lateral bone wedge is resected to obtain adequate correction of the varus heel. Closure of the osteotomy may be challenging in the adult population, resulting in incomplete correction (18). Another option to realign the varus heel is the lateral displacement calcaneal osteotomy (23). Cases of failed Dwyer or lateral displacement osteotomy, severe deformity, or failed triple arthrodesis with varus malunion may be salvaged with distraction osteogenesis using an external fixator (Taylor Spatial Frame, Smith & Nephew Inc., Memphis, Tennessee, U.S.A.).

Gradual correction may be preferred for the severe deformity that may be only partially corrected with acute treatment (12) (Fig. 6). In some cases, the hindfoot varus is so severe that the foot is weight bearing on the lateral side of the base of the fifth metatarsal tuberosity. An ulcer under the fifth metatarsal tuberosity is another relative indication for gradual correction with external fixation to minimize the risk of deep infection associated with the larger wounds required for acute treatment. Furthermore, a severe deformity with a medial skin crease may not be amenable to complete correction with acute treatment, and gradual distraction

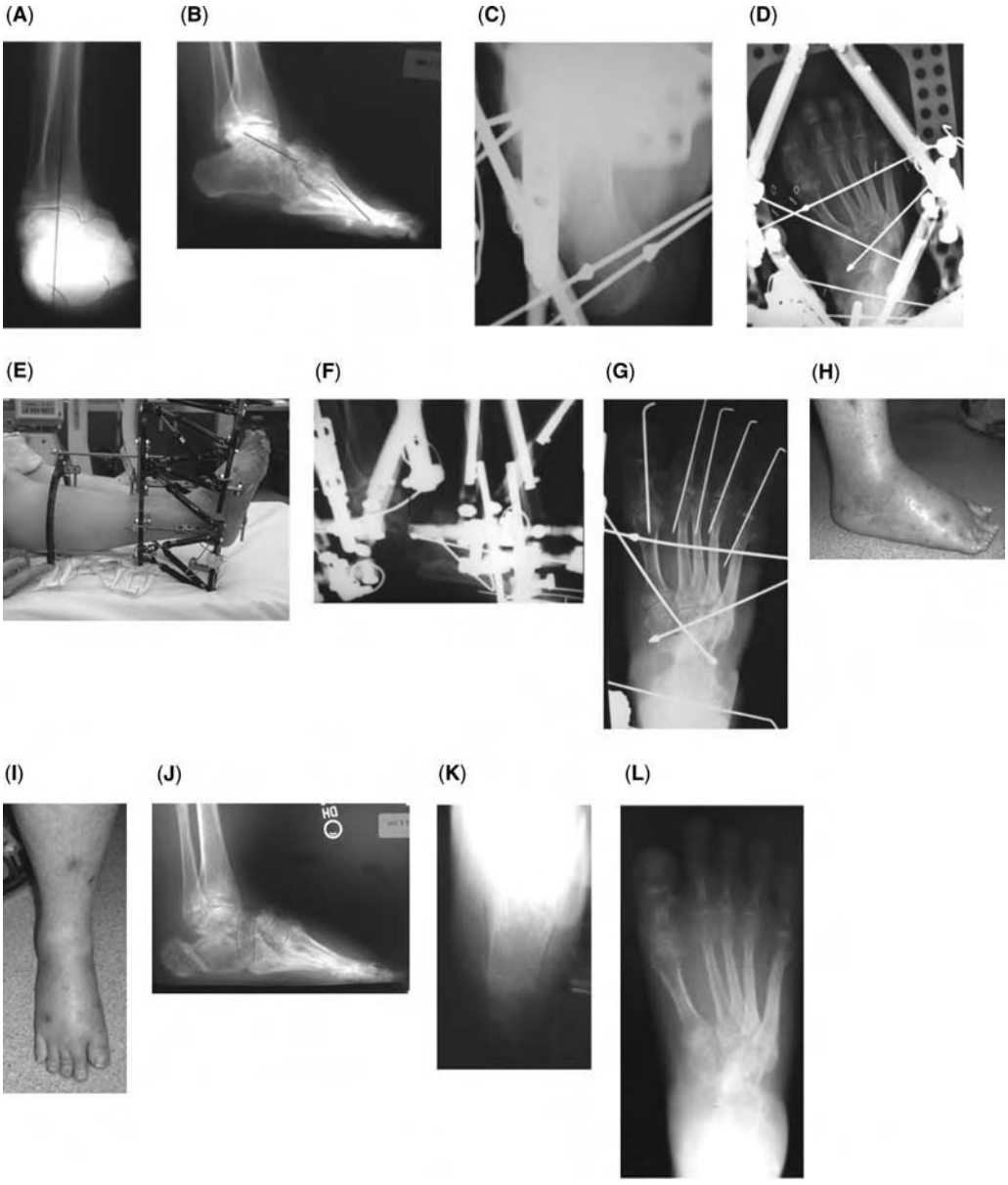

Figure 6 A 50-year-old woman with a cavus foot. She had a history of poliomyelitis and a triple arthrodesis malunion. Treatment included a calcaneal osteotomy to correct the heel varus and a midfoot osteotomy to correct the forefoot adduction and supination deformity. Distraction was done over a six-week period, after which the frame was converted to a holding frame for eight weeks followed by casting for four weeks. (**A**) Preoperative long axial radiograph shows varus malalignment of the calcaneal tuberosity relative to the tibial long axis. (**B**) Preoperative lateral radiograph of the foot shows plantarflexion of the first metatarsal relative to the talus. (**C**) Axial radiograph after calcaneal osteotomy and application of frame. (**D**) Anteroposterior (AP) foot radiograph after application of frame. Forefoot adduction is noted. (**E**) External fixator after application. (**F**) External fixator after distraction of midfoot osteotomy. (**G**) AP foot radiograph after correction of forefoot adduction, before frame removal. (**H** and **I**) Clinical improvement of cavus is noted after frame removal. (**J**–**L**) Lateral, axial heel, and AP radiographs after correction of deformity and healing of calcaneal and midfoot osteotomies.

may enable more complete correction of deformity and minimize risk of dehiscence of the medial soft tissues. In cases of foot deformity associated with a short leg resulting from poliomyelitis, gradual correction of the foot deformity with hindfoot arthrodesis and external fixation may be done with concomitant tibial lengthening (24).

COMPLICATIONS

Complications of surgery include anesthetic problems, infection, neurovascular injury, fixation failure, malunion, nonunion, and thromboembolic disease. Infection is managed with wound culture, debridement, and antibiotics. Consultation with an expert in infectious diseases may be helpful. Wound management may include dressings, vacuum-assisted closure, and immobilization.

Complications of external fixation with distraction osteotomy include pin-tract infection, failure of osteotomy separation, acute tarsal tunnel syndrome, toe contractures, wire breakage or cutout, and fracture (4). Delayed union and recurrent equines contracture may occur, especially with neuromuscular conditions (24). A nonunion rate of 20% has been reported for flatfoot correction with distraction calcaneocuboid arthrodesis, and additional surgery may be required in 71% of cases (16). Malunion may occur as a result of hardware failure, incomplete correction of deformity, or patient noncompliance with weight-bearing restrictions. Patient selection is important because nonunion may be more frequent in smokers and noncompliant patients and may necessitate revision or amputation. Pin-tract inflammation and infection are common, and are treated with pin-site release or pin removal and debridement.

Residual foot pain may occur after complex reconstruction, especially in salvage surgery. There can be multiple causes for this, and evaluation includes assessment for residual deformity and nonunion. Treatment may include bracing, shoe inserts, shoe modifications, or revision surgery. Recurrent deformity may occur following deformity correction performed through soft tissues or joints or in patients with progressive neurological conditions. This emphasizes the importance of patient follow-up after correction.

FUTURE DIRECTIONS

Foot deformity is likely to remain a prevalent problem, especially because of the evolving diabetes epidemic. Advances in the salvage of the severely ulcerated, infected, and deformed foot have progressed tremendously in the past two decades, and likely will continue to evolve.

Acute management may be facilitated, and hardware-related wound problems decreased, with the availability of recently developed low-profile internal fixation plates such as those for the lateral wall of the calcaneus. Specific implants for other regions of the foot may be developed.

External fixation of the foot and ankle is evolving, and new devices specific for the foot may be designed for gradual correction of foot deformity. Modern frames (Taylor Spatial Frame, Smith & Nephew Inc., Memphis, Tennessee, U.S.A.) have advanced the correction of deformity throughout the lower extremity, especially in the foot where complex translational, rotational, and oblique plane deformities are common. Components of these modern frames currently are available for the foot, and devices to facilitate the application of ring fixators to the foot are available (Quantum Medical Concepts, Hood River, Oregon, U.S.A.). Approaches that combine the advantages of internal and external fixation in the foot may be developed. The role of nutritional status and supplementation in the prevention and management of wound problems, infection, nonunion, and other complications remains to be investigated.

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19 Joint Distraction in the Treatment of Ankle Osteoarthritis

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INTRODUCTION

Osteoarthritis of the ankle joint is a progressive and disabling joint disease, occurring often in young people secondary to trauma. There is no cure for osteoarthritis, and current conservative treatment is mainly symptomatic. Options for surgical treatment of patients with a disabling, painful osteoarthritic ankle are limited and far from optimal.

Post-traumatic etiology of ankle joint osteoarthritis is the most common. In such cases, incongruity in the ankle joint frequently occurs, including malpositioning of the talus, widening of the ankle mortise, and shortening of the fibula. Cartilage defects may be present as well. On a molecular level, progression of osteoarthritis is manifested by rupture of the collagen network, loss of matrix molecules—mostly proteoglycans and collagen fragments—and results in loss of the mechanical properties of the cartilage (1). In addition to changes in the cartilage, bony changes such as subchondral sclerosis, osteophytes, and subchondral cysts also occur in osteoarthritis (Fig. 1) (2–4).

We developed a new approach and strategy in the treatment of the osteoarthritic ankle joint. In 1989, we started to perform joint distraction procedure in our clinical practice using Ilizarov fixator. The treatment approach of ankle distraction is based on the concept that osteoarthritic cartilage has some reparative activity when the damaged cartilage is mechanically unloaded, preventing further wear and tear, while the pressure changes in synovial fluid, essential for the nutrition of the cartilage, are maintained.

CLINICAL EVALUATION

Clinical parameters such as pain, functional ability, clinical condition, and ankle joint mobility are evaluated by the use of a standardized questionnaire and physical examination. Joint space width, and subchondral sclerosis are assessed on standardized radiographs of the ankle joint using a digital method (5).

We developed a questionnaire to evaluate pain, functional ability, and clinical condition (6,7). Pain is measured by use of a visual analogue scale, where patients are asked to score the pain between 0, no pain and 10, unbearable pain. The functional ability questionnaire includes assessment of morning stiffness, ability to climb stairs, and walking distance.

The Ankle Osteoarthritis Scale is also used for evaluation of pain and function (8). A good correlation between these questionnaires was shown in our most recent study.

The clinical condition is assessed by physical examination of ankle joint mobility, pain during movement, crepitus, and swelling. Ankle joint mobility is determined by measuring the range of motion and is compared to the contralateral ankle joint.

Position and clinical condition of the other joints in both feet and lower limbs need to be assessed. Great care must be given to the subtalar joint that may be osteoarthritic and painful as well. Ankle joint distraction will not be very helpful in such cases. Equinus position of the foot is another common finding.

Severity of osteoarthritis as seen on radiographs may not correlate with severity of clinical symptoms, making such findings less important in making decisions for joint distraction.

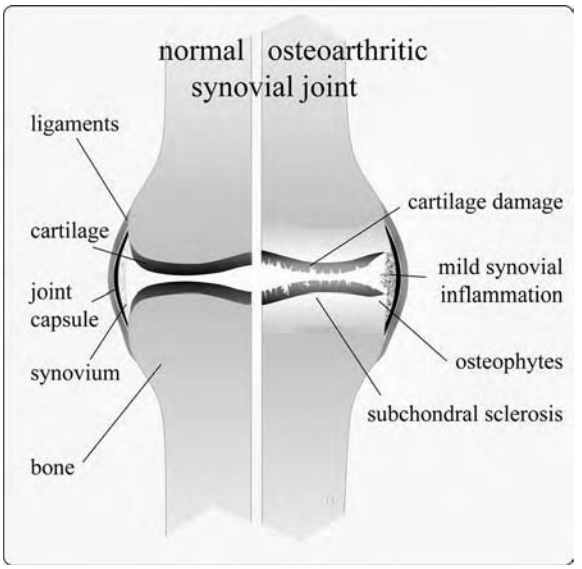

Figure 1 Schematic drawing of a synovial joint. Structures of a healthy joint (*left*) and structural changes occurring in osteoarthritis (*right*) are shown.

To measure joint space width and bone density, we use the Ankle Images Digital Analysis (5). The specific role of the subchondral bone in the process of osteoarthritis is still unclear. Although unambiguously involved, it is still under debate whether the changes in bone are the result of changes in the cartilage, or whether they initiate the process of osteoarthritis (9–12).

The etiology of painful osteoarthritis in the ankle joint may vary greatly. In our studies, osteoarthritis was mostly due to trauma. We excluded patients suffering from systemic diseases such as diabetes, rheumatoid arthritis, hemophilia, etc. This was done for scientific reasons only. Also, we considered previous septic arthritis in the ankle joint as a contraindication for a joint distraction.

TREATMENT OPTIONS

Current treatment of osteoarthritis is aimed at reducing pain, minimizing functional disability, and slowing the progression of the disease. Patients' education with respect to modification of activities and lifestyle is essential (13). When conservative treatment no longer provides sufficient pain relief, surgery is indicated.

The options for surgical treatment are arthroscopic debridement, arthroplasty, and arthrodesis. The results of an arthroscopic debridement are unpredictable (14,15). The long-term results of ankle arthroplasty, especially in young patients, are still uncertain. In our present clinical practice, patients younger than 55 years of age, suffering from late-stage osteoarthritis with a pain score of less than six (on a scale from 0–10), undergo an arthroscopic debridement of ankle joint hoping to gain time before making final decisions for treatment. Patients are informed that the expected benefit is unpredictable, both in quality and in duration (14).

Ankle arthrodesis remains the gold standard of treatment for osteoarthritis of the ankle joint. It is effective in relieving pain, but at the expense of the loss of joint motion. This leads to overloading of adjacent and contralateral joints (16), increasing the risk of developing arthritis in these joints, especially in young people in later years. In our practice, arthrodesis is indicated in elderly patients with painful osteoarthritic ankle joints.

The goal of the ankle distraction is to unload the damaged articular surfaces completely during full weightbearing while maintaining intermittent pressures of the synovial fluid. Intra-articular fluid pressures have been measured and found to change significantly during such axial movements (17). In vitro and animal in vivo studies have demonstrated these fluid pressures to stimulate proteoglycan synthesis in osteoarthritic cartilage (18,19). Joint distraction during a three-month period results in transient periarticular osteopenia because the load is partially transferred through the frame instead of the joint. Subchondral sclerosis does not return after treatment and may have a beneficial influence on osteoarthritis.

The major indication for joint distraction is pain, severe enough to consider an arthrodesis.

In a randomized controlled trial, we demonstrated that the clinical benefit after joint distraction in the treatment of severe ankle osteoarthritis was statistically significantly higher than that after arthroscopic debridement (6). This indicates that the clinical benefit found after joint distraction is not achieved by the preceding debridement alone. We did arthroscopically remove osteophytes if they seemed to prevent a plantigrade position of the foot in the Ilizarov frame.

In our reports, we did not correct any abnormality in alignment or anatomy of bones and soft tissues in the ankle joint region, nor did we do any surgery on intra-articular cartilage. We did not use hinges in the external fixator to permit motion in the distracted ankle joint. Yet, such additional surgery prior to joint distraction may increase the chances for a beneficial outcome and should be considered in planning treatment.

SURGICAL TECHNIQUE

A wire-only ring fixator is used for ankle distraction. It is preassembled before surgery and consists of two rings, one for the proximal and one for the distal part of the tibia, and a foot ring.

Two 1.8-mm Kirschner wires are drilled through the proximal tibia, perpendicular to the longitudinal axis of the tibia. These wires are fixed to the ring under tension (1.3 kN). The proximal ring is then connected with four threaded rods to the distal ring, where two wires are inserted and fixed to the ring in a similar way.

Two olive wires are inserted medially and laterally through the calcaneus at about a 30° angle between them and tensioned at 0.9 kN to a half-ring around the heel. One Kirschner wire is inserted through the talus and fixed to the foot ring with some tension. This wire is important to prevent distraction of the subtalar joint (Fig. 2).

One olive wire is inserted on the medial side through the bases of the first and fourth or fifth metatarsal bones and tensioned (0.5 kN) to the half-ring over the forefoot. The foot ring is connected to the distal tibial ring with threaded rods fixing the foot in a neutral position to allow weightbearing on plantigrade foot (Fig. 2).

Mechanical unloading of the degenerative articular surfaces is achieved by gradually increasing the distance between the tibial and foot frames using four threaded rods around the ankle. The rate of distraction is 0.5 mm twice a day until total distraction of 5 mm is confirmed on a radiograph. This radiograph should be done while standing, if weightbearing is tolerated. The distraction of 5 mm on a radiograph is maintained, doing adjustments if necessary, during the three-month period of treatment. Weightbearing as tolerated is allowed within one week after surgery. Absence of ankle motion during ambulation is compensated by rocker bottom sole. Physical therapy and medication are administered, if needed. The ring fixator is removed after three months, and weightbearing as tolerated is permitted.

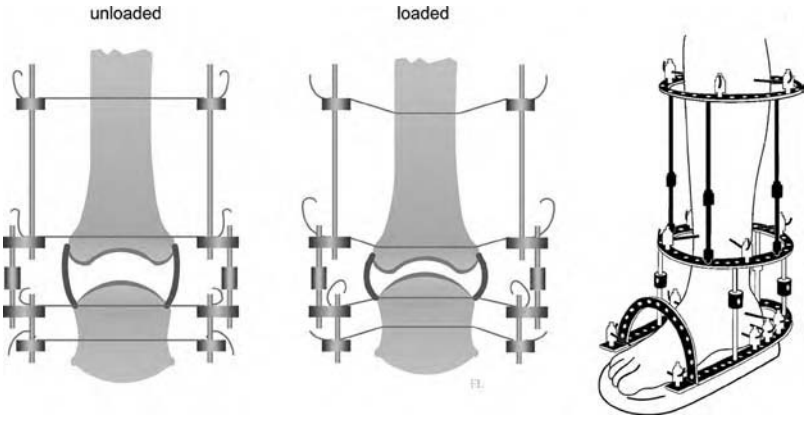

Figure 2 Schematic drawing of Ilizarov ankle distraction.

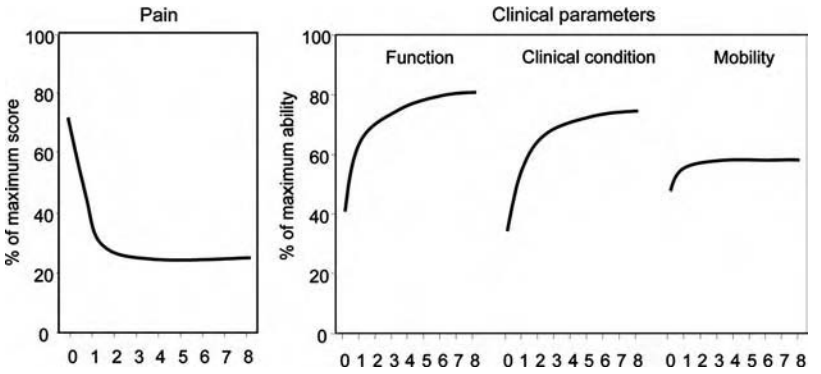

Figure 3 Clinical outcome with respect to pain, function, clinical condition, and mobility in an open prospective study. Values are presented as the percentage of the maximum score (10 points for pain, 30 points for function, 8 points for clinical condition, and the dorsiflexion of the contralateral ankle for mobility), where a decrease in pain and an increase in function, clinical condition, and mobility are beneficial. *Source*: From Refs. 6,7.

OUR RESULTS

Pain, Functional Disability, and Clinical Status

Prospective and retrospective evaluations showed significant pain relief, improvement of daily functioning and clinical status in about 70% of the patients treated with ankle distraction (Figs. 3 and 4) (6,7,17,20). This reduction in pain, improvement of function and clinical status may occur only after a period of months following removal of the external fixator. The improvement of parameters increased over time in the years after joint distraction treatment.

Ankle Joint Mobility

Pain and stiffness of the joint capsule in an osteoarthritic ankle joint may lead to severe restriction of function and disability. A fixed equinus deformity of the foot could contribute to the severity of symptoms and functional impairment. Joint mobility after joint distraction appeared to be

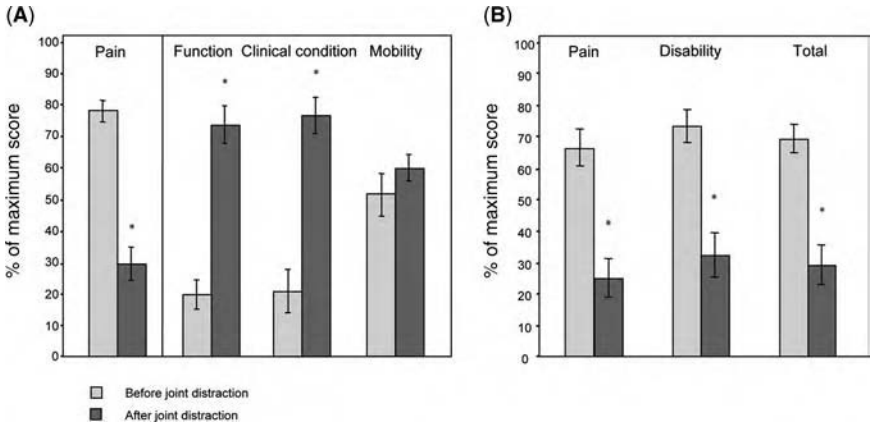

Figure 4 Clinical outcome in a retrospective study, more than seven years after joint distraction treatment. (**A**) Pain, function, clinical condition, and mobility evaluated by the Van Valburg score. Average scores are given before and after treatment. Data are presented as a percentage of the maximum score being 10 points for pain, 30 points for function, and 8 points for clinical condition. Joint mobility (range of motion) is presented as a percentage of mobility of the contralateral ankle. (**B**) Clinical results of joint distraction evaluated using the Ankle Osteoarthritis Score (8). Average scores for pain, disability, and the total AOS score are given before and after treatment. Data are presented as a percentage of the maximum score being 900 mm for pain, and 900 mm for disability (nine questions each), where a decrease in pain and a decrease in disability are beneficial. *Source*: From Refs. 6,7.

improving in the following years; however, this was not statistically significant. Most important is that joint mobility is maintained. Equinus contracture commonly recurred after ankle distraction.

Age

Age did not seem to affect results in our studies (6). In theory, capacity of the body to repair cartilage diminishes with aging (21).

Radiographic Evaluation

In our studies, radiographic evaluation of patients with a joint space narrowing of more than 10% and a subchondral sclerosis before treatment showed gradual widening of the joint space and diminished subchondral sclerosis in the years following joint distraction (6,7). However, no statistically significant relationships between radiographic characteristics and clinical findings were demonstrated.

DOES JOINT DISTRACTION RESULT IN ACTUAL REPAIR?

It is questionable whether repair of cartilage in a severely osteoarthritic joint is possible at all. Although there is no information on the quality of the cartilage, the clinical parameters, combined with the duration of the effect and the progressive improvement with time, suggest the formation of functional tissue.

Animal studies have demonstrated that joint distraction is able to change chondrocyte activity beneficially (19). However, because of the limited follow-up period, actual cartilage repair was not demonstrated. Recently, a study in rabbits showed that at 12-week follow-up after a 6-week joint distraction, histological cartilage repair did not occur (22). We think that chondrocytes may need a longer period of time after joint distraction to be able to repair the cartilage matrix.

However, it is important to recognize that the observed clinical success may not be dependent on cartilage repair, bony changes, and cartilage–bone interactions alone, but may be related to the formation of fibrous tissue. Interposed and molded fibrous tissue within the joint could be the reason for increased joint space and decreased pain.

COMPLICATIONS

Complications of ankle distraction are commonly associated with insertion and maintenance of the wires. A proper technique of positioning and tensioning of the wires is essential for a good outcome. Special care must be given to insertion of the proximal tibial wires in order to avoid a peroneal nerve injury. Position of the forefoot wire should be away from the mid-tarsal joints.

Improperly tensioned or loosened wires could cause pain and pin tract infection, which is the most common complication. It usually responds well to oral antibiotics. If the wire through the talus is infected and the distraction of 5 mm has been achieved, we prefer to remove this infected wire to decrease the risk of septic arthritis.

Lateral translation of the forefoot may occur over the forefoot wire away from the olive. This could happen when an equinus deformity has been corrected with some force.

The forefoot wire could interfere with full weightbearing secondary to discomfort or pain and has a tendency to break. An adequate support of the forefoot with a foot splint fixed to the external fixator is helpful in decreasing pain with weightbearing.

FUTURE DIRECTIONS

Regardless of underlying mechanisms, the improvement of clinical symptoms as well as positive radiological changes after joint distraction validate the concept of joint distraction in the treatment of osteoarthritis. However, further research is required for better understanding and predicting the results of this complex unconventional treatment. Surgical correction of pre-existing deformities, use of hinges to allow movement of the joint during distraction, and the administration of systemic or local agents may further improve the final outcome of joint distraction. Study of these additional interventions will be needed.

The prolonged benefit of joint distraction in cases of ankle osteoarthritis could be used to justify studies on joint distraction in the treatment of osteoarthritis in other joints such as knee and hip. Distraction of these joints, if successful, could also have a great social and economic impact.

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20 An Approach to Contracture and Deformity of the Foot and Ankle

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INTRODUCTION

Contractures of the foot and ankle, while common, remain a challenging problem for the orthopedic surgeon. Rigid soft-tissue contractures may be caused by nerve, skin, or muscle pathology and can be seen in patients suffering from one of a variety of conditions: burns, poliomyelitis, or Charcot–Marie–Tooth disease. Additionally, high-energy trauma may cause bony deformities such as malunion, nonunion, and infection, and congenital malformations may present both soft-tissue and bony pathology. Fortunately, the orthopedic surgeon possesses a variety of techniques with which to address these varied problems: casting, soft-tissue release and transfer, osteotomies, internal and external fixation techniques, arthrodesis, and amputation.

CLINICAL EVALUATION

A thorough preoperative evaluation is always required in order to understand the nature of the deformity and set upon a course of treatment. Classification systems can be useful for this purpose, although not a replacement for the physician's experience and judgment, and careful consideration of the patient's unique medical history and whether the deformity is a result of a chronic condition or trauma. The author's approach to clinical evaluation is to divide foot and ankle contractures first by origin (such as malunion/nonunion, arthritic, burn, neuromuscular, infection, or congenital or developmental) and then, secondly, through radiographic analysis, by type, as either a simple deformity or a complex (combination of angular, rotational, translational, length) deformity.

A simple deformity is in only one plane and involves one tissue, for example, equinus soft-tissue contracture, or angular malunion, or simple nonunion of bone. Complex deformities involve more than one plane, more than one tissue, or a deformity at more than one location. Examples of complex deformities include the equinocavus contracture, rocker-bottom deformity, and most burn contractures. Another important distinction in clinical evaluation is the age of the patient, because children younger than eight years old, due to the plasticity of their bones, often can undergo successful correction of deformities without osteotomy, while older children and adults are more likely to require osteotomy (1,2). The observation of fixed bony deformity during clinical evaluation also will help to classify the deformity and aid in determining whether osteotomy is indicated.

CLASSIFICATION

Simple Deformities

The class of common simple deformities includes the equinus contracture, distal tibial angular deformities, and simple nonunions.

Equinus Contracture

It is almost certainly the most common deformity encountered by the foot and ankle surgeon. The causes of this deformity include burns, trauma, neuromuscular disease, and congenital conditions. The patient may adopt a toe-to-toe gait pattern, a toe-to-heel gait pattern, with excessive knee hyperextension, or excessive external rotation and pronation. Physical examination is the most valuable procedure in diagnosing equinus contracture.

Distal Tibial Angular Deformities

Distal tibial angular deformities are usually the result of post-traumatic malunion. The ultimate goal of treatment is the avoidance of long-term joint degenerative changes. While there are few long-term natural history studies available, biomechanical studies can help guide treatment. Joint contact forces have been shown to increase progressively with increasing angular deformity and proximity to the articular surface. Acceptable limits of coronal and sagittal angulation typically are reported as 10° to 15°, but patients do not tolerate this degree of deformity, especially with distal malunions or limited subtalar motion.

Nonunions

Nonunions are rarely seen in the foot but, because of the increasing occurrence of high-energy pilon fractures, have become more frequently encountered in the distal tibia. Nonunions of the distal tibia (and elsewhere) are not a homogenous group; multiple classification systems have been proposed and can be useful. Classification systems, however, cannot replace an understanding of the personality of each individual nonunion. They may be divided on the basis of geometry (transverse vs. oblique), biologic activity (hypertrophic vs. atrophic), the presence or absence of infection, and the degree of bone loss.

Occasionally, these deformities present with purely varus or valgus angulation, but they commonly appear to have components of malalignment in multiple planes. Although these *multiplanar* deformities can initially be intimidating, it is important to realize that they actually represent a single deformity that occurs on an oblique plane (3). The axis of this oblique plane can be determined trigonometrically, but is determined most easily graphically or with the use of nomograms (Fig. 1). Oblique radiographs taken at orthogonal angles to the calculated axis of the deformity provide confirmation of this analysis.

Complex Deformities

Common complex deformities include complex equinus deformity, rocker-bottom deformity, and cavus deformity.

Complex Equinus Deformities

Complex equinus deformities are frequently encountered as a component of a more complex problem. Neuromuscular disease, high-energy trauma, and burns typically present with

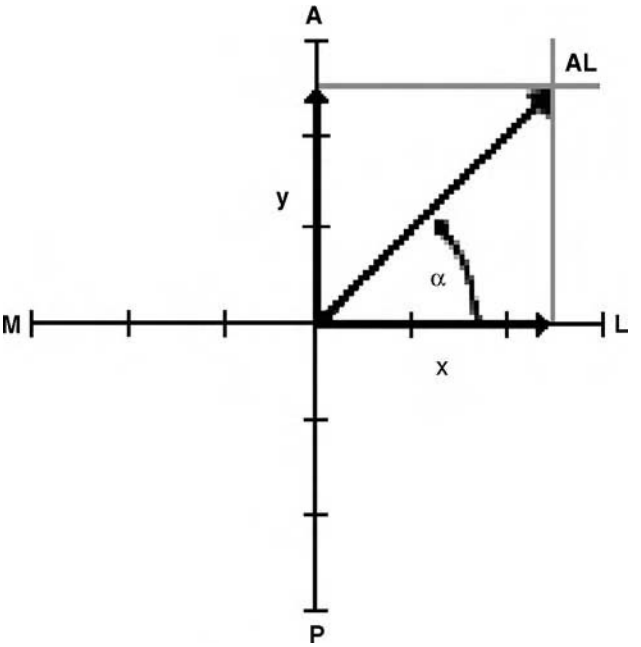

Figure 1 Measurement of oblique plane deformity. The angular deformity in the anteroposterior radiograph is measured and marked on the *x*-axis. Deformity in the lateral radiograph is marked on the *y*-axis. The oblique angulation is the vector sum of these two lines. The magnitude of oblique angulation in degrees is determined by measurement of this line. The orientation of the oblique plane relative to the frontal plane can be measured on the graph (α); 1 mm = 1°.

multiple deformities of the foot. Equinovarus and equinocavus deformities frequently are seen, and equinovalgus deformities have been reported. These complex deformities are more difficult to correct and require more complex strategies for correction.

Cavus Deformities

Cavus deformities can be classified as mild (metatarsal–calcaneal angle of 135–150°), moderate (120–135°), or severe (lower than 120°).

Rocker-Bottom Deformity

Rocker-bottom deformity is occasionally encountered as a result of dorsal soft-tissue contracture (burns) but has been more commonly seen as the result of overcorrection of the forefoot during equinus correction.

TREATMENT OPTIONS

Equinus Contracture

Mild (< 15°) and moderate (15–30°) equinus contractures usually respond to stretching, casting, and bracing. If moderate cases do not respond, then a Hoke percutaneous, Achilles tendon lengthening, and casting is usually successful. Severe (>30°) equinus contractures can be treated with external fixation techniques (Fig. 2).

Distal Tibial Angular Deformities

Multiple techniques are available and offer excellent outcomes. Osteotomy with internal fixation (Fig. 3) and the use of circular frames with corticotomy (Fig. 4) are the two most common techniques and can be used successfully, depending on soft tissue integrity, infection, and the surgeon's judgment.

Nonunions

Nonunions with less than 1 cm of bone loss and no evidence of infection usually can be managed with internal fixation techniques. For patients with compromised soft tissue, osteomyelitis, more extensive bone loss, or hypertrophic pseudarthroses, external fixation with the use of Ilizarov techniques can be more effective (Fig. 5).

Complex Equinus Deformities

More severe corrections typically require more advanced techniques. Severe equinus contractures involve more than a tightened heel cord; all periarticular structures are contracted, including the medial neurovascular bundle. Rapid or intraoperative correction of a severe deformity places the soft tissues, including the neurovascular bundle, at risk. These are treated best with gradual approaches, particularly Ilizarov techniques with a circular fixator (3–7).

Cavus Deformities

Mild cavus deformities occasionally respond to stretching, casts, or soft-tissue release. Moderate midfoot cavus can be treated with soft-tissue release and midfoot osteotomy; however, the recurrence rate is high. Severe cavus deformities are treated more successfully with the Ilizarov technique (Fig. 6).

Rocker-Bottom Deformity

Rocker-bottom feet have been treated classically with capsulotomy and midfoot osteotomies, but severe deformity (metatarsal–calcaneal angle $> 200°$) is well suited to correction with Ilizarov techniques.

SURGICAL TECHNIQUES

Internal Fixation

Surgical principles of internal fixation for simple nonunion include atraumatic handling of the soft-tissue envelope, debridement of fibrous tissue and necrotic bone at the nonunion site, use

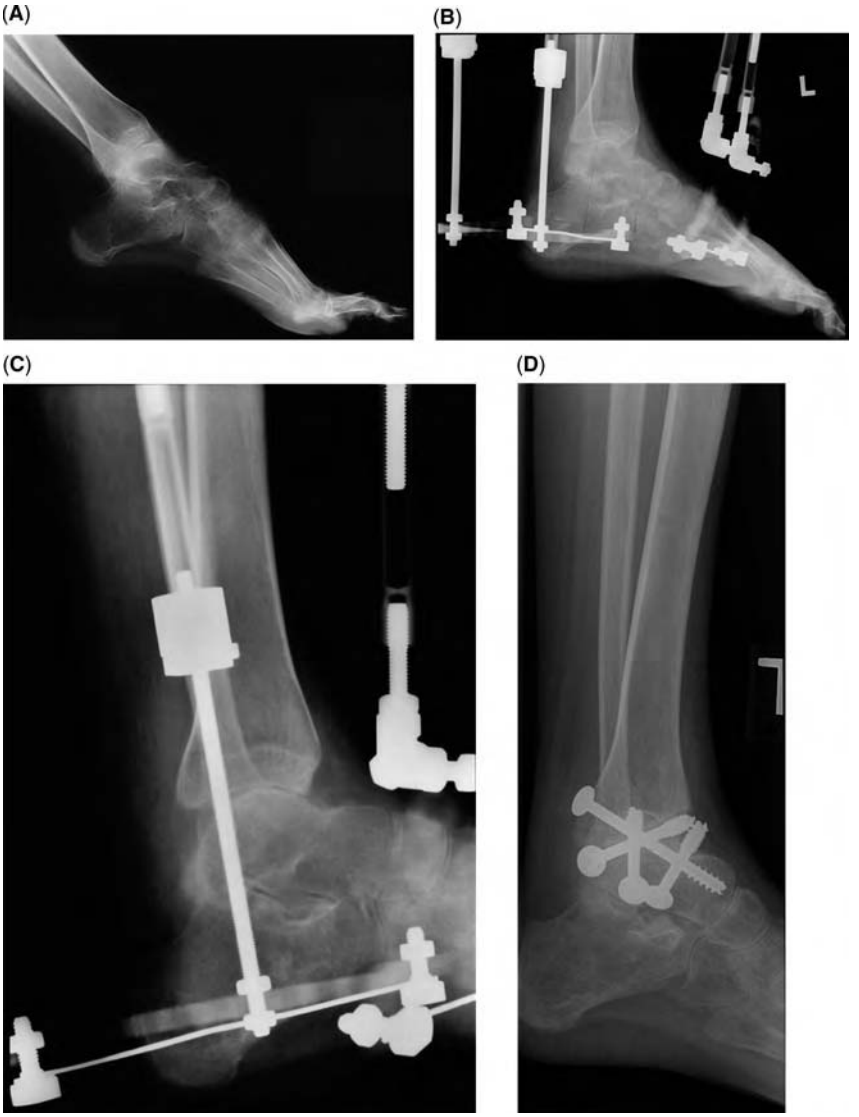

Figure 2 A 22-year-old male paraplegic developed bilateral lower leg contractures eight years after being paralyzed in a motor vehicle accident. Severe equinus contracture of the left ankle (**A**) was treated with Ilizarov external fixation (**B**), three weeks, and (**C**), six weeks. After correction, the left ankle was fused at 90° (**D**).

of autogenous cancellous bone graft, and adequate bony stability with compression provided by plates and screws. Fibular osteotomy usually is required to allow compression and can be performed at the nonunion site or preferably at a midshaft location that allows the intraosseous membrane to provide greater stability. Early stable fixation allows protected weightbearing and range of motion exercises at the ankle, promoting union.

Although internal fixation is an extremely successful technique, there are times when external fixation is better. Patients with previously failed arthrodesis, significant bone loss, infection, or compromised soft tissues are often poor candidates for internal fixation, but these patients often can have ankles fused successfully with the use of an external fixator and Ilizarov techniques (8,9). The technique is similar to that described for nonunion. Modifications to the frame include the use of multiple wires placed through the body of the talus to provide tibiotalar compression and the placement of a ring at the proximal tibial metaphysis for lengthening, if necessary. Fixation of the foot can be used to provide greater stability.

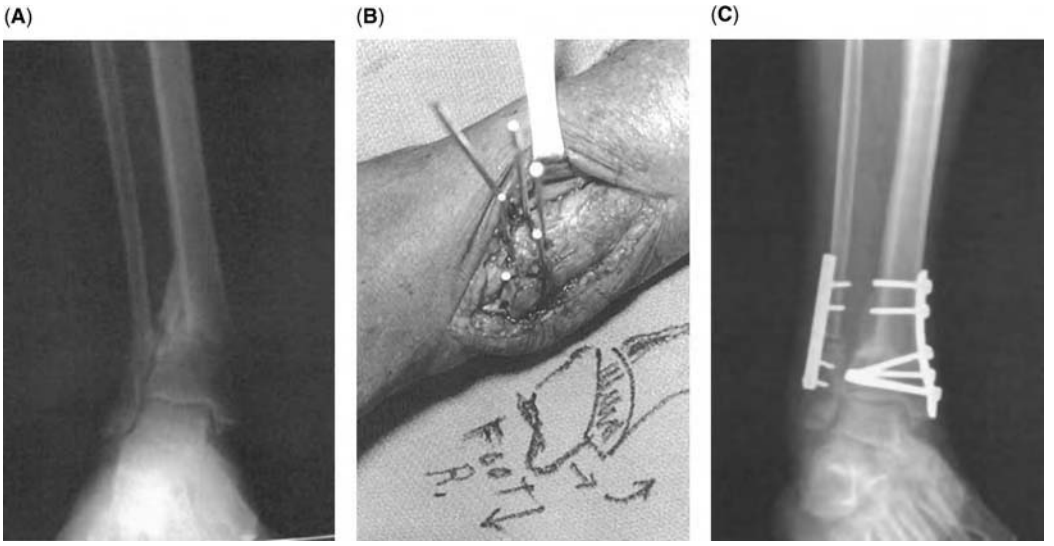

Figure 3 (**A**) Preoperative radiograph of a 60-year-old woman with a two-year history of distal tibial malunion after nonoperative treatment. (**B**) Dome osteotomy for valgus deformity. (**C**) Postoperative view at 12 weeks with the patient weightbearing.

External Fixation

External fixation offers well-recognized advantages and disadvantages. It is particularly valuable for injuries that require lengthening for deformity correction. Percutaneous corticotomy, when used with external fixation techniques, offers the advantage of minimally disrupting the soft-tissue envelope. This minimal disruption makes external fixation especially appropriate for patients who have sustained open fractures or burns or have undergone multiple operative procedures. When placed under tension and in multiple planes, the small wires or pins allow fixation of poor-quality bone and very small fragments that otherwise can be very difficult to control. External fixation with gradual correction also offers the advantage of precise correction that can be modified throughout the postoperative course. Because of the stability provided by circular fixators, motion at the ankle and subtalar joints is possible, and almost immediate ambulation with weightbearing can be encouraged.

Distal tibial angular deformities can be managed successfully with external fixation techniques. A number of unilateral fixation devices are available. Ring fixators, combined with the use of Ilizarov techniques, have been used more widely in deformity correction (3–7). Circular fixators have some biomechanical benefit because of pin placement, flexibility, and the ability to apply immediate and gradual compression. Many surgeons believe circular fixators offer greater versatility and adaptability to match specific deformities.

For the author, the frame used for correction of distal tibial malalignment is similar to the classical equinus frame, with half-pins (hybrid frame) and wires being placed in the distal tibia and joint (Fig. 4). The tibial segment consists of two full rings placed proximal to the deformity. The ring block is fixed to bone, using a hybrid technique with one wire at each ring and multiple half-pins placed anteriorly. A distal ring is placed parallel to the tibial plafond. The ring is usually fixed to the distal tibia with either two wires or three wires and an anterior half-pin. Wire fixation can be achieved through the talus in patients with poor quality or minimal bone at the distal fragment. In patients with severe deformity or minimal bone, a calcaneal half-ring and a metatarsal half-ring can be used to provide additional stability and fixation. When no lengthening is required, hinges are oriented perpendicularly to the plane of maximum deformity.

Following a 7- to 10-day latency period, correction is begun. Distraction is applied with telescoping rods placed on the concave side of the deformity at a rate of 1 to 2 mm/day divided into four increments. If the patient will tolerate it, ambulation with crutches is

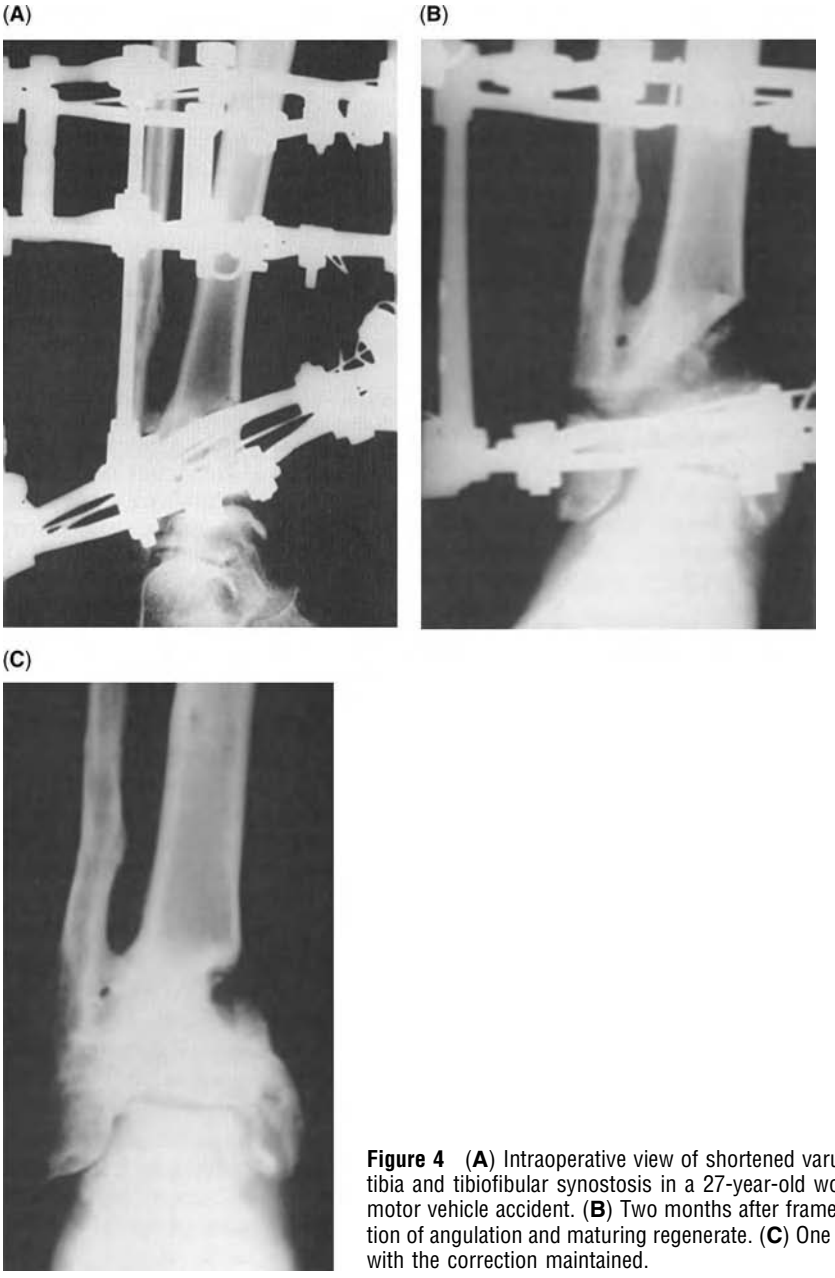

Figure 4 (**A**) Intraoperative view of shortened varus malunion of the distal tibia and tibiofibular synostosis in a 27-year-old woman three years after a motor vehicle accident. (**B**) Two months after frame application with correction of angulation and maturing regenerate. (**C**) One year after frame removal with the correction maintained.

encouraged throughout the correction. The fixator is left in place until evidence of bony union is present clinically and radiographically (Fig. 4). Removal of the frame is performed in the clinic with minimal discomfort for the patient, using a combination of intravenous or intramuscular analgesics and local anesthetics, as necessary. Occasionally, patients will require conscious sedation or general anesthesia.

The frame used for the equinocavus foot uses the equinus frame with modification. Threaded rods are used to connect the calcaneal and metatarsal half-rings medially and laterally. The medial column is distracted at a rate of 1 mm/day. The lateral rod is typically static during correction because the majority of the deformity is medial (Fig. 5). If needed, the lateral column can be distracted. Toe flexion contractures occasionally are encountered. They are best treated with flexor tenotomy and pinning for six weeks.

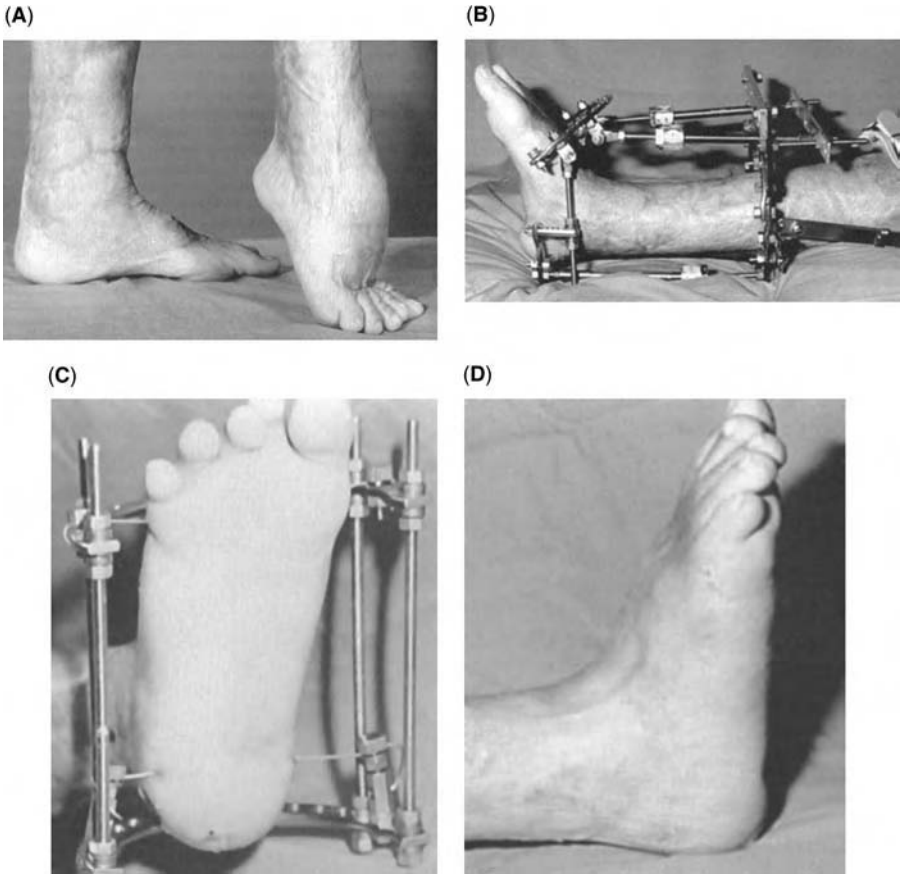

Figure 5 (**A**) Preoperative view of severe equinocavus deformity secondary to burns in an 11-year-old boy. (**B**) Equinus frame with cavus modification and knee bracing to prevent flexion contracture secondary to tendo Achilles lengthening. (**C**) The cavus frame. The calcaneal half-ring is distracted from the metatarsal half-ring with a threaded rod. (**D**) Three and one-half years after surgery, the patient has maintained a plantigrade foot and participates in all sports.

Arthrodesis

Simple ankle arthrodesis typically is performed using well-described internal fixation techniques. Internal fixation is generally a successful technique but is usually undesirable in the case of a previously failed arthrodesis, significant bone loss, infection, or compromised soft tissues.

Intraoperatively, the distal end of the tibia and the talar dome are debrided to bleeding (*paprika sign*) bone. The tibial frame and the foot frame are then compressed 1 to 2 cm. A sterile Doppler device is used to assure that the dorsal pedal and posterior tibial arteries have not collapsed. If extensive bone loss has occurred, it may not be possible to achieve immediate bony apposition, and shortening of the extremity more than 2 cm is undesirable. Postoperatively, the tibia and talus are compressed 1 mm/day until contact. Compression is then continued at the rate of 1 mm/week. This weekly compression can be gradual (0.25 mm every two days) or acute (1.0 mm every seven days). If there is significant shortness (>2 cm) from distal tibial or talar bone loss, the tibia is lengthened proximally (Fig. 6). Weightbearing is encouraged because it promotes healing of the fusion and regeneration, and decreases pain and edema. The frame is removed when there is clinical and radiographic evidence of fusion and proximal regenerate maturity. The fusion typically is protected after frame removal with a walking cast for approximately six weeks.

Osteotomy

Most surgeons are familiar with opening and closing wedge osteotomy techniques. These have been described for the management of distal tibial malalignment, with excellent results via

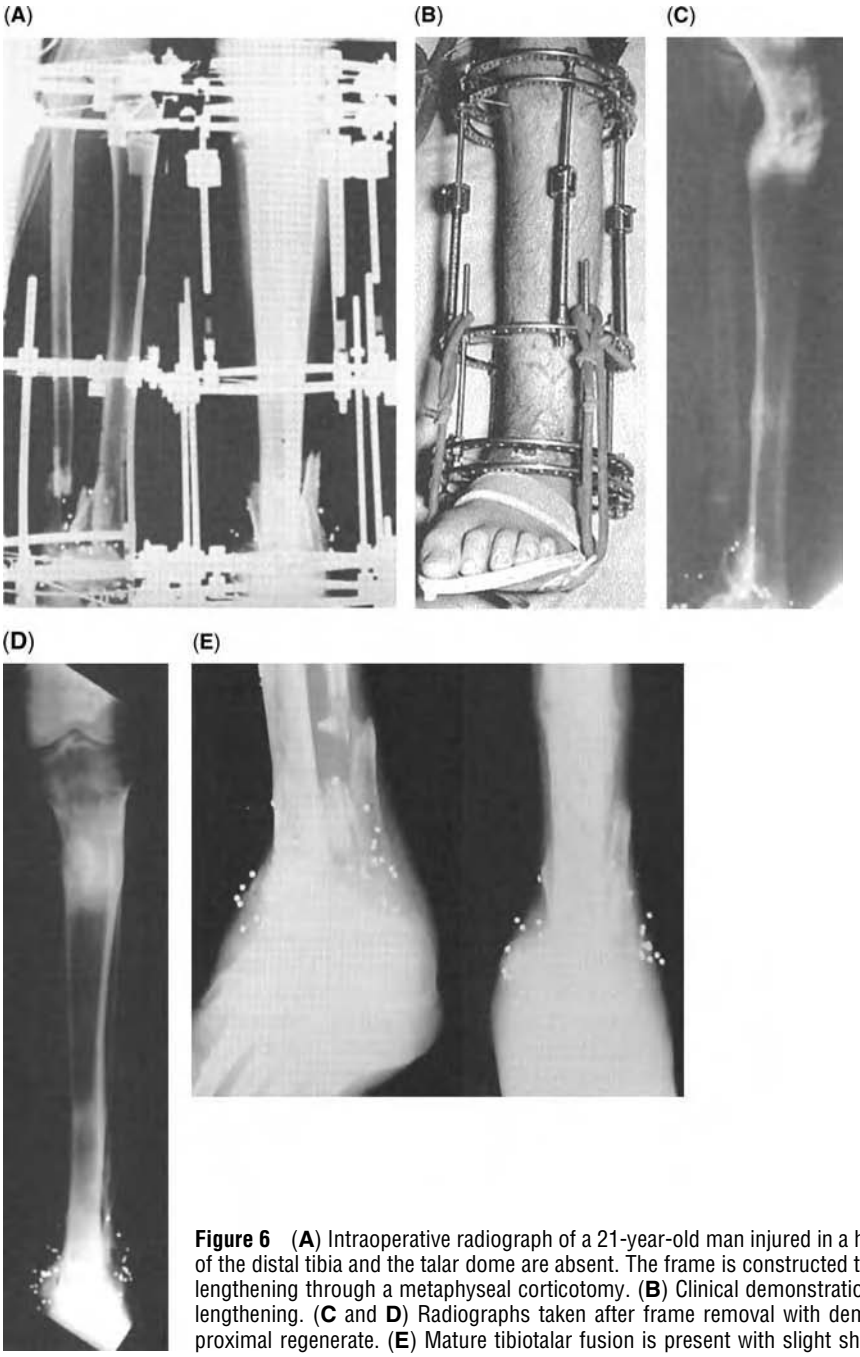

Figure 6 (**A**) Intraoperative radiograph of a 21-year-old man injured in a hunting accident; 6 cm of the distal tibia and the talar dome are absent. The frame is constructed to allow proximal tibial lengthening through a metaphyseal corticotomy. (**B**) Clinical demonstration of the frame before lengthening. (**C** and **D**) Radiographs taken after frame removal with demonstration of mature proximal regenerate. (**E**) Mature tibiotalar fusion is present with slight shortening of the fibula.

either method. The bony apposition in closing wedge osteotomies promotes greater bone contact, healing, and weightbearing, but requires a shortening. Conversely, opening wedge osteotomies can preserve length, but at the expense of stability, and often require the use of bone grafting with its additional morbidity. Additionally, the opening wedge technique requires a soft-tissue envelope that accommodates any potential increase in length. The osteotomy's shape influences stability and available bone surface for healing. Dome, barrel, or ''banana'' osteotomies are technically more demanding to perform but offer relatively more stability and good bone surface contact for healing.

The author prefers to use a closing dome osteotomy because its shape is inherently stable and less length is lost than with a simple closing wedge osteotomy. It is performed in line with

the axis of deformity and can be used for varus and valgus malalignment. The apex of the osteotomy is placed at the apex of the deformity on its concave side, and its base lies along the convex side of the deformity. The osteotomy is centered over a line that bisects the axes of the proximal and distal bony fragments. The shape of the osteotomy is first outlined by the placement of multiple Steinman pins, placed perpendicular to the long axis of the bone along an arc (Fig. 3). Bone cuts then are performed with a narrow, sharp osteotome between adjacent pins. The anterior cortex is removed piecemeal to gain access to the posterior cortex. The entire anterior cortex is scored with the osteotome before the canal is exposed so that ''ridges'' occur along the arc. The pins are used to perforate the cortex and help prevent propagation of fractures away from the desired path of the osteotomy. After removal of the desired bone, the osteotomy is compressed and fixed with a plate and screws. To allow the distal tibial fragment to move, it is usually necessary to osteotomize the fibula. For varus deformities, it is usually necessary to remove a section of fibula to allow for the shortening that accompanies the closing tibial wedge. Although not always necessary, fibular shortening may be necessary for valgus deformities, depending on the degree of shortening required at the tibia and the amount of fibular abutment at the lateral mortise. When possible, the fibular osteotomy site is compressed rigidly with a plate and screws to add lateral stability to the tibial osteotomy and reduce fibular abutment of the peroneal tendons, lateral talus, subtalar joint, or calcaneus. Bone grafting (from the tibial resection) sometimes is necessary if a gap exists at the osteotomy site (Fig. 3). This fibular fixation provides additional stability but is not an absolute requirement and is not always possible due to the soft tissue over the fibula. Postoperatively, patients are placed into a nonweightbearing cast until evidence of bony union is present clinically and radiographically.

Amputation

Many surgeons unfortunately view amputation as a failure on their part. For many patients, amputation offers the best opportunity for an early return to maximal function. Patients can rapidly return to work and participate in everyday activities with the aid of modern prosthetics and aggressive rehabilitation. Many patients are not psychologically and socially capable of enduring multiple surgeries, long treatments with external fixators, extended antibiotic regimens, and the potential for recurrence. Amputation should be openly discussed with patients and their families before the decision to proceed with reconstruction.

Osteomyelitis

Osteomyelitis remains one of the most difficult problems encountered by the foot and ankle surgeon. Treatment of osteomyelitis has focused on two concepts: first, removal of the infection with debridement of all infected, necrotic tissue and avascular bone, and culture-directed antibiotics; and second, reconstruction by providing bony stability, eliminating the dead space, and obtaining soft tissue coverage (10). Classic Ilizarov teaching stated that debridement might not always be necessary. Revascularization of the osteomyelitis center and the resultant biologic stimulation by the new bone at the corticotomy site (''regenerate'') could control the infection without radical debridement because ''osteomyelitis burns in the fire of regeneration'' (7). Unfortunately, although bony union can be achieved with this technique, more recent studies have shown that its success rate is less than 75% (11).

Thorough (radical) debridement may require en bloc resection of bone, which would result in significant bony instability and significant loss of length. Occasionally, when less than 2 cm of bone is excised, it is possible to perform an acute limb shortening, which provides relative bony stability and compression. More often a dead space that requires treatment is present.

Circular fixators, used with Ilizarov techniques, are especially well suited to these situations. They provide excellent stability, do not require the placement of implants at the infection site, allow bone transport and lengthening to eliminate the dead space, and allow access to the soft tissue for monitoring and wound care. Operatively, debridement can be performed before or after frame placement depending on the surgeon's preference. At the author's institution, debridement usually is performed before frame placement, allowing improved access to the debridement site and a rough estimate of the ability to acutely correct angular, rotational, or translational deformity. Because many of these infections are polymicrobial, and intraoperative cultures often do not correspond with cultures taken preoperatively from sinuses, it is important to take multiple

culture samples during the procedure (12). The author routinely sends a minimum of three cultures: one of fluid, one of soft tissue, and one of bone. Whenever possible, the intramedullary canal should be debrided to normal bleeding bone. It is typically sealed at the deformity and often contains poor bleeding bone or sequestrum. Re-establishing the intramedullary canal will remove bacterial load, but its most important function is increasing blood flow to the dead space to provide healing and antibiotics. All sinuses are excised as part of the soft-tissue debridement.

The frame used for treating infection of the distal tibia is similar to the ones previously described for nonunions and arthrodeses. A ring is placed at the proximal tibial metaphysis to

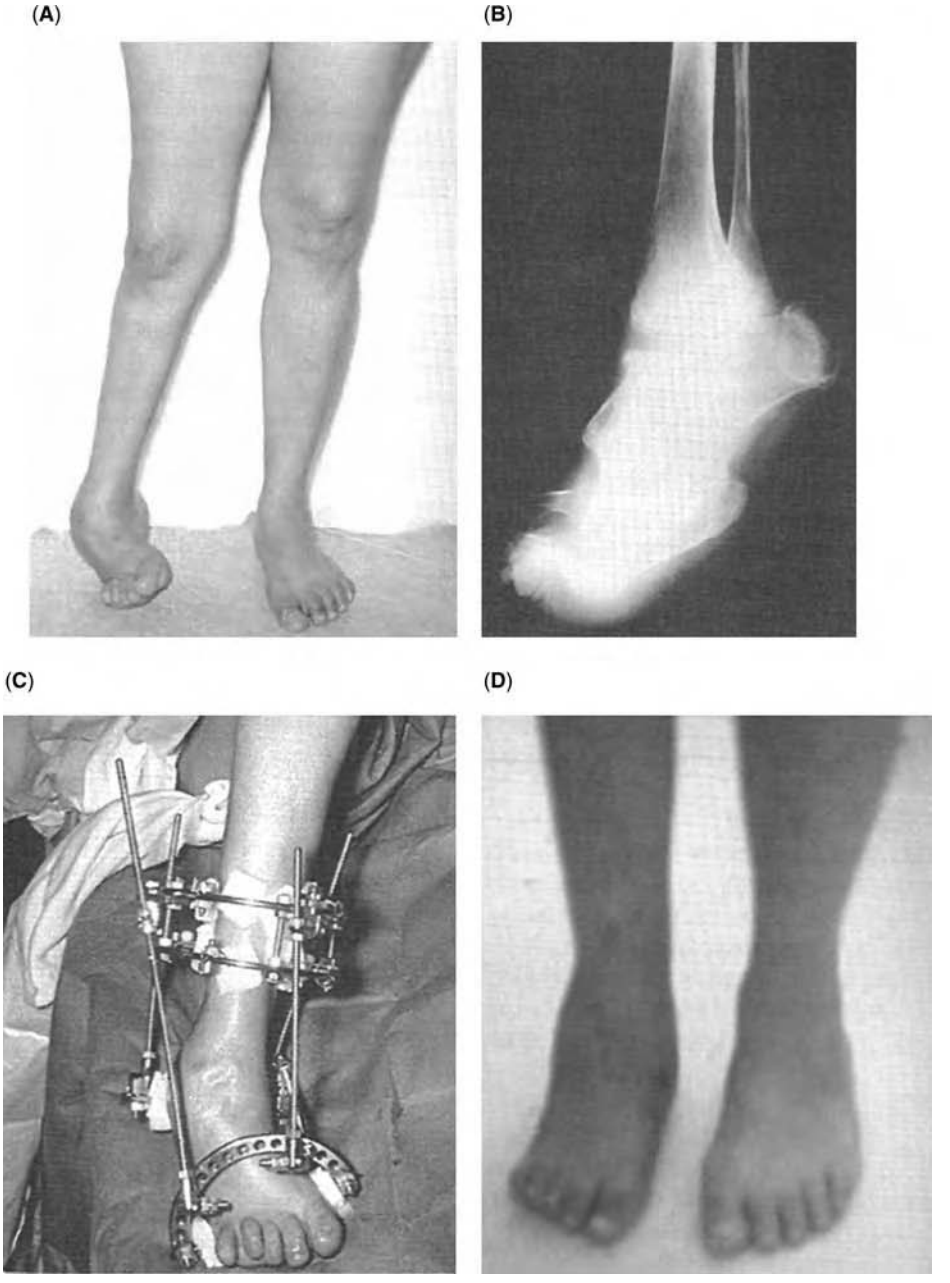

Figure 7 (**A**) Severe equinocavovarus deformity in a 42-year-old woman with a history of poliomyelitis since age 14. (**B**) Radiograph of affected deformity with severe degenerative changes. (**C**) Application of equinus frame with cavus modification and anterior threaded rods designed to derotate the forefoot. (**D**) After frame removal, the patient maintained a plantigrade foot and normal shoe wear.

allow *bifocal osteosynthesis.* Rings are then placed proximal and distal to the debridement site to provide stability. When possible, a distal ring is placed at the distal tibial metaphysis, but extension of the frame to the foot is commonly done to prevent equinus or cavus deformities. The proximal rings are connected with telescoping rods, to apply distraction at a corticotomy performed at the proximal metaphysis. The proximal ring block is then connected to the ring distal to the debridement site, with telescoping rods to apply compression at the deformity. The distal two rings are connected statically (Fig. 6).

Broad-spectrum intravenous antibiotics are administered postoperatively. Ideally, the surgeon should work with an infectious disease expert for culture-specific medications (13). At the author's institution, intravenous antibiotics are continued for two weeks, and oral antibiotics are continued for an additional four weeks. Intravenous antibiotics are routinely completed at home with the help of a home health agency.

Special Patient Populations

Patients with neuromuscular disease and patients with burns to the extremities typically present with the most severe challenges encountered by surgeons. The most difficult part of reconstruction for these patients often is not achieving correction but maintaining it.

Neuromuscular deformity is typically the result of unbalanced muscle actions and is often seen with severe arthritic changes (Fig. 7). After correction, these forces are still present and likely to cause recurrence if no additional steps are taken. Postoperative bracing can occasionally be effective. However, soft-tissue balancing procedures, such as a split posterior tibial tendon transfer or fusion, should be strongly considered. Distraction osteotomies can be performed in the foot as an alternative, although they are technically demanding (3,5).

Burn patients' deformities are similarly prone to recurrence. Unlike patients with neuromuscular disease, however, burn patients are extremely poor candidates for soft-tissue transfer or osteotomy because of their compromised soft tissues. Postoperative bracing and physical therapy may succeed in maintaining corrections, but the patient must be prepared for the repeat application of an Ilizarov device if the deformity recurs (4,14,15).

FUTURE DIRECTIONS

Correction and care of contractures, both simple and complex, will continue to challenge even experienced orthopedic specialists. Given the broad array of injuries and afflictions that lead to contractures of the foot and ankle—from high-energy trauma to poliomyelitis—it is unlikely that their occurrence will decrease or that their treatment significantly improve unless there are major advances to decrease injuries and burns, and only if there is earlier and more aggressive treatment of infection and deformities. We are hopeful that tissue-engineering techniques will revolutionize this and other areas of musculoskeletal surgery. Until then, identical conditions can sometimes be treated with different techniques, with no measurable difference in outcome. Improvement in the treatment of contractures comes with the improvement of the individual physician, as the physician gains knowledge and experience. Most importantly, the physician must work with patients to set appropriate goals; it is usually possible to restore function to a foot but often impossible to provide a completely normal foot or ankle.

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21 Metatarsal Lengthening

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INTRODUCTION

Brachymetatarsia is a congenital foot deformity that presents as a unilateral or bilateral short metatarsal(s), typically involving the fourth toe (Fig. 1A). The prevalence of brachymetatarsia of the lesser metatarsals has been reported to be 1 in 1820 to 1 in 4586 (1,2). The female-to-male ratio has been reported to be 25 : 1 with the prevalence of bilateral short metatarsals reported to be 72% (2,3). According to Tachdjian (4), a short first metatarsal ''Morton's foot'' is more common than a short fourth metatarsal. Takakura et al. (5) found that the prevalence of congenital short first metatarsals is 1 in 10,000 based on Japanese children, which is less common than the short fourth metatarsal. Recessive hereditary patterns have also been described in patients with a congenital short metatarsal (2).

The cause of brachymetatarsia is believed to be premature closure of the epiphyseal growth plate. The etiology of a short metatarsal may be acquired or congenital (Table 1) (6). Acquired short metatarsals are caused by trauma, infection, tumor, Freiberg's disease, radiation, and surgery. In addition, acquired short metatarsals can be associated with skeletal and systemic abnormalities (sickle cell anemia, multiple epiphyseal dysplasia, multiple hereditary osteochondromas, and juvenile rheumatoid arthritis). Surgically induced (iatrogenic) shortening of metatarsals are caused by transphyseal fixation, osteotomies of the metatarsals, and internal/external fixation producing a growth arrest or synostoses between metatarsals. A failed bunionectomy or overaggressive first metatarsal cuneiform arthrodesis can also result in an acquired short first metatarsal. Congenital short metatarsals can be found isolated or associated with systemic syndromes, endocrinopathies, and dysplasias. Polydactyly or syndactyly is also found in combination with congenital short metatarsals. Congenital short metatarsals are typically bilateral and often associated with other skeletal deformities as opposed to acquired short metatarsals that are mainly unilateral in presentation (6,7).

Patients with brachymetatarsia exhibit a dorsally displaced toe and are often bothered by their difficulty with shoeing, pain, and appearance (6,8).

CLINICAL EVALUATION

A thorough patient history is important as brachymetatarsia can coexist with a skeletal or systemic abnormality. When a coexisting medical diagnosis is not identified, a complete medical evaluation with appropriate laboratory analysis should be performed prior to surgical intervention. When performing a comprehensive clinical examination of the foot and ankle, it is important to evaluate the short metatarsal(s) and identify concomitant deformities. With plantarflexion of all the metatarsal phalangeal joints (MTPJ) on a non-weightbearing examination, the metatarsal heads become prominent through the dorsal skin. This positioning of the forefoot allows for an accurate clinical evaluation of the metatarsal parabola. Examination of the integument should also be performed to evaluate the callus patterns.

Typically, the short metatarsal induces a dorsally displaced digit (Fig. 1B and C). This abnormal digital alignment can produce a dorsal digital callus at the proximal interphalangeal joint (PIPJ) and a localized submetatarsal callus from increased plantar pressure beneath metatarsal head. The digit of the short metatarsal is dorsally contracted and dorsally subluxated at the MTPJ. This digit typically demonstrates minimal flexion deformity at the PIPJ or distal interphalangeal joint. The soft-tissue compensatory contracture at the MTPJ should also be evaluated preoperatively by manual plantarflexion of the MTPJ to assess for digital reducibility/realignment. The Kelikian push-up test is performed to simulate weightbearing by loading the

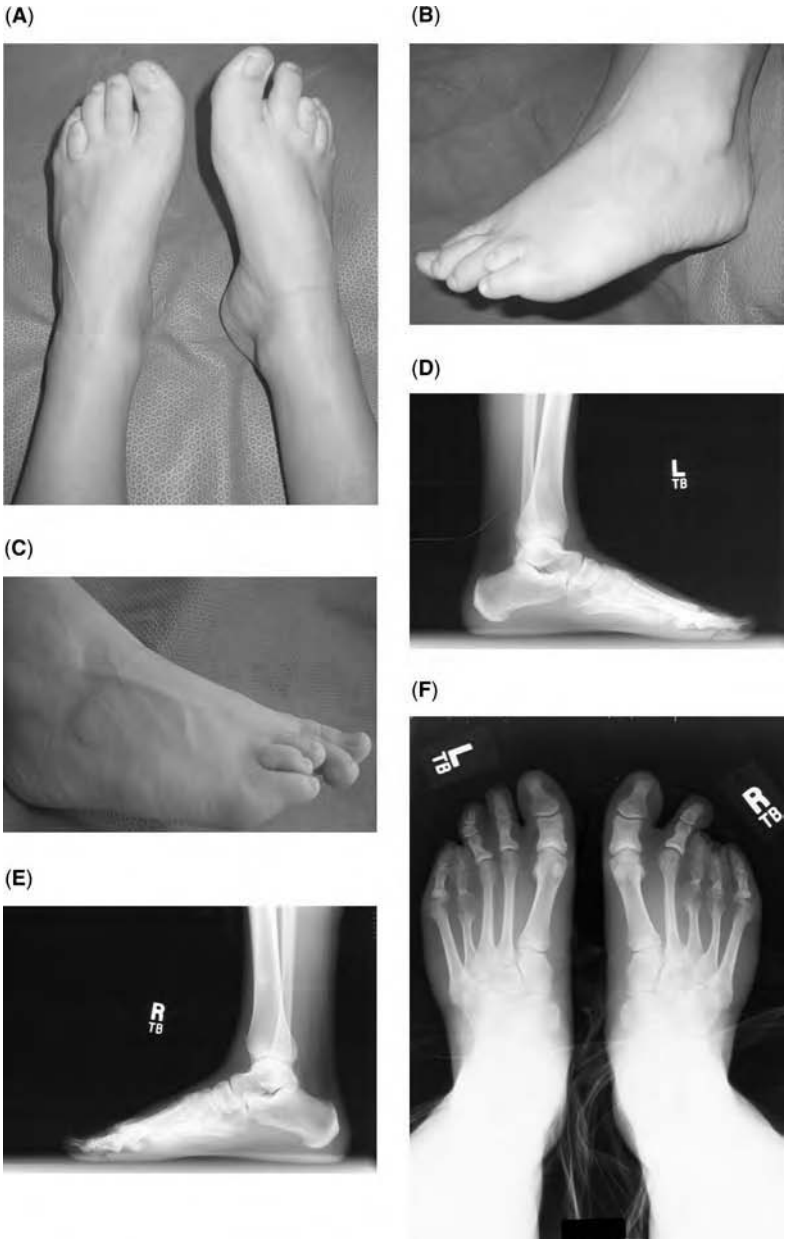

Figure 1 Clinical photos and radiographs of bilateral congenital short fourth metatarsals with a congenital short third metatarsal on the right. (**A**) Front view. (**B**) Side view of the left side. Note the dorsal displacement and the limited flexion deformity at the proximal and distal interphalangeal joints of the fourth digit. (**C**) Side view of the right side. (**D**) Lateral radiograph of left side. Note the increased fourth metatarsal declination and extension contracture of the toe. This increased declination is observed at the level of the distal metaphyseal–diaphyseal junction (region of the growth plate). The dorsal cortex of the short metatarsal is parallel to the adjacent metatarsals. (**E**) Lateral radiograph of the right side. (**F**) Anterior–posterior radiograph. The metatarsal length is abnormal with the bilateral fourth metatarsals and the right third metatarsal being short of the line formed by drawing the metatarsal parabola angle. Note the slight medial bowing of all the three short metatarsals. The transverse plane deviation of the adjacent digits converging toward the short metatarsal can also be observed.

lateral column of a non-weightbearing foot. Only when simulated weightbearing is achieved can the digital reducibility be tested. The joint is then considered reducible, semireducible, and non-reducible. It is considered reducible if the proximal phalanx can plantarflex at the MTPJ so the toe is entirely plantigrade, semireducible if the proximal phalanx cannot plantarflex at the MTPJ so

Table 1 Causes of Brachymetatarsia

Congenital			
Systemic Syndromes	**Dysplasias**	**Endocrinopathies**	**Acquired**
Aarskog syndrome	Acromesomelic dysplasia	Albright's hereditary osteodystrophy	Trauma
Apert syndrome	Achondroplasia	Pseudohypoparathyroidism	Physeal fracture
Brachiodactyly type E	Achrodysostosis	Pseudopseudohypoparathyroidism	Iatrogenic injury
Carpenter's syndrome	Geleophysic dysplasia		Radiation
De Lange syndrome	Metaphyseal chondrodysplasia		Sickle cell crisis
Down syndrome	Thanatophoric dysplasia		Juvenile rheumatoid arthritis
Ectrodactyly	Diastrophic dysplasia		Infection
Grebe syndrome	Multiple epiphyseal dysplasia		Tumor
Hypochondroplasia	Myositis ossificans		Polio
Hajdu–Cheney syndrome			
Hand–foot–genital syndrome			
Jeune's thoracic dystrophy			
Killian/Teschler-Nicola syndrome			
Langer–Giedion syndrome			
Leri–Weill dyschondrosteosis			
Majewski-type short rib-polydactyly			
Mohr syndrome			
Multiple synostoses syndrome			
Orofacial–digital syndrome			
Pfeiffer syndrome			
Poland's anomaly			
Rett syndrome			
Robinow's syndrome			
Rothmund–Thomson syndrome			
Ruvalcaba's syndrome			
Saethre–Chotzen syndrome			
Taybi syndrome			
Turner syndrome			
Weill–Marchesani syndrome			
Warfarin effects			
Werner's syndrome			
X syndrome			
18p syndrome			

the toe is not completely plantigrade, and nonreducible if the proximal phalanx cannot plantarflex at the MTPJ so the toe remains dislocated. When the digit is semireducible or nonreducible, a complete MTPJ release (dorsal capsulotomy, medial and lateral collateral ligament release, extensor tendon lengthening, and plantar plate release) is recommended. A partial MTPJ release (dorsal capsulotomy) should be performed when the digit is reducible. It is not recommended to perform an arthroplasty or arthrodesis because this will result in shortening of an already short ray. However, if a MTPJ release is not adequate, adding a PIPJ arthroplasty or arthrodesis can allow for complete reduction of the digit. In addition, in some cases it maybe necessary to lengthen the combined extensor tendon of the toe. This surgical realignment of the toe, when deemed necessary, should be addressed prior to or at the same time as the metatarsal lengthening procedure. Reducing the contracture prior to lengthening will allow the toe to remain in an anatomical position during and after lengthening.

Weightbearing anterior–posterior (AP), lateral, and medial oblique foot and ankle radiographs allow for accurate surgical planning and assessment of the metatarsal deformity. Obtaining bilateral foot films can also be very helpful for surgical planning. Radiographic assessment of the short metatarsal on AP and lateral views is important to preoperatively plan placement of the external fixation and the osteotomy level. Typically, the radiographic position of short metatarsal is slightly deviated on both X-ray views as compared to the adjacent metatarsals.

On the lateral radiograph an increased declination is observed at the distal metaphyseal–diaphyseal junction (region of the previous growth plate), which presents as a flexion deformity of the distal metatarsal. A congenital short metatarsal adapts while the child is growing. The short metatarsal grows into greater declination than the adjacent metatarsals in an attempt for the short metatarsal to contact the ground in the same plane as the other metatarsals. The

corresponding proximal phalanx of the short metatarsal is dorsally displaced secondary to a shortened soft-tissue sleeve (8). The short metatarsal declination defines the plane of lengthening as the half-pins of the external fixation device are mounted perpendicular to the longitudinal bisection of the metatarsal in the sagittal plane (Fig. 1D).

On the AP radiograph, the metatarsal parabola is defined as the angle formed by connecting the distal articular surfaces of the first, second, and fifth metatarsals. The parabola can be quantitatively measured as an angle formed between a line connecting the distal ends of the first and second metatarsals with a line connecting the distal ends of the second and fifth metatarsals. The angle between these two lines is defined as the metatarsal parabola angle (normal = 142.5°). This angle is used to identify an abnormal length metatarsal. Most times, the best-fit line is drawn to determine the short verses long metatarsals. Bilateral radiographs are helpful when assessing patients with multiple short metatarsals. Short metatarsals are typically associated with short phalanges. First metatarsal protrusion distance (normal = 2.0 mm plus or minus) is defined as the distance between the distal points of the second and first metatarsals on an AP radiograph. The transverse plane deviation of the short metatarsal is also important to assess. Typically, a short third or fourth metatarsal has a slight medial bow. Therefore, it is important to measure the metatarsus quintus abductus angle (fourth intermetatarsal angle) (normal = 8–10). When lengthening a metatarsal with a slight medial bow, an accurate vector of lengthening is critical. If this bowing is overlooked, lengthening can result in encroachment of the metatarsal heads, medial or lateral deviation of the digit, and pain. A short first or second metatarsal usually lacks this observed bowing (Fig. 1E and F).

Preoperative planning is important to identify the amount of metatarsal length necessary to re-establish the metatarsal parabola. Prediction of the needed metatarsal length also provides an accurate calculation of the number of days required for lengthening. By drawing the metatarsal parabola angle and measuring the longitudinal distance between the most distal point on the affected metatarsal head to the metatarsal parabola line, the amount of metatarsal lengthening needed to restore the parabola could be calculated. For example, if the amount of metatarsal length needed to re-establish the metatarsal parabola is 20 mm, the amount of time required to obtain this length would be approximately 40 days, based on the desired distraction rate of 0.5 mm/day. This is useful information to discuss with the patient preoperatively.

However, the rate of distraction may need to be adjusted during the postoperative period; therefore, appropriate patient education is important. The consolidation period typically takes at least the same amount of time as the lengthening phase or longer. Consolidation time is variable, but typically ranges from two to four months. Therefore, prediction of needed metatarsal length provides a time line for the patient's total length of treatment (lengthening phase and consolidation phase).

CLASSIFICATION

With an AP X-ray and measurement of the metatarsal parabola, the amount of lengthening could be determined. If the amount is less than 1 cm, acute or gradual lengthening can be performed. However, if the amount is greater than 1 cm then gradual lengthening is preferred (Table 2).

Table 2 Clinical Management

Classification Subgroup	Clinical Evaluation	Surgical Approach	Pearls	Complications/Pitfalls
Metatarsal < 1cm short	Kelikian push-up test (reducible, semireducible, nonreducible MTPJ)	Digital realignment, bone graft with plate/IM pinning, restore metatarsal parabola	Pinning across MTPJ	Nonunion or malunion, MTPJ stiffness and subluxation, metatarsalgia, neurovascular stretch
Metatarsal > 1 cm short	Kelikian push-up test (reducible, semireducible, nonreducible MTPJ)	Digital realignment, monolateral distraction, restore metatarsal parabola	Pinning across MTPJ, distraction = 0.5 mm/day	Overlengthening, MTPJ stiffness and subluxation, metatarsalgia, malalignment

Abbreviations: MTPJ, metatarsal phalangeal joints; IM, intramedullary.

TREATMENT OPTIONS

Surgery to lengthen a short metatarsal can improve cosmesis, shoeing, and decrease the pain associated with this deformity. Acute and gradual lengthening surgical techniques have been described for correction of brachymetatarsia. Acute lengthening of a short metatarsal with autogenous bone grafting was first described in 1969 by McGlamry and Cooper (9). Since then many other techniques for acute lengthening (one stage) have been reported using interposition of synthetic materials, allograft, step-cut, or oblique osteotomy with distraction and internal fixation (1,3,10,11). Some authors have performed shortening of adjacent metatarsals or proximal phalanxes so as to decrease the amount of metatarsal lengthening needed (12–14).

Gradual lengthening with external fixation (distraction osteogenesis) is preferred for lengthening greater than 1 cm (6). With gradual lengthening, the rate of postoperative lengthening can be adjusted, the patient can bear weight during treatment, and the patient can have input regarding the final length. The risk of neurovascular compromise is less than seen with acute lengthening that can cause a severe soft-tissue stretch (6,7). Gradual lengthening of short metatarsals have been performed with subsequent interpositional bone graft (15,16) and with distraction osteogenesis alone (17–19). Various types of external fixation devices have been utilized to achieve gradual metatarsal lengthening (20–23,26).

SURGICAL TECHNIQUE

This technique provides accurate final anatomical alignment by ensuring the proper plane and vector of metatarsal lengthening. The patient is positioned supine on the radiolucent table with a bump under the hemisacrum to obtain a foot forward position. Based on the reducibility of the MTPJ contracture, partial or complete MTPJ release with or without an arthroplasty or arthrodesis of the PIPJ should be performed to realign the digit prior to the metatarsal lengthening procedure. The initial spread and locations of the pins are determined on preoperative planning with a four-pin Orthofix Mini-M100 external fixator (Orthofix, Verona, Italy) The half-pins are inserted percutaneously under fluoroscopic control, bicortically and perpendicular to the shaft of the metatarsal.

The first half-pin is placed at the most distal region of the metaphyseal–diaphyseal junction. A 1.8 mm wire is used to predrill the hole under fluoroscopic control perpendicular to the bone on the lateral view. A 3.0 to 2.5 mm $\times$ 60/20 mm half-pin is inserted (Fig. 2A). Because the fixator is mounted perpendicular to this first pin, the first pin determines the plane of lengthening. It is important that the direction of metatarsal lengthening will bring the final position of the metatarsal head to the appropriate level in sagittal plane.

The external fixator should be set so that the half-pin clamps are at the smallest distance apart minimizing the pin spread. This ensures that all the half-pins are maintained within the short metatarsal. If the metatarsal is too short the most proximal half-pin can be placed in the tarsus (cuboid) spanning the Lisfranc's joint. The second half-pin is inserted proximally in the base of the metatarsal, parallel to the first pin and just distal to the adjacent metatarsal–cuboid joint. This second half-pin establishes the vector of metatarsal lengthening and determines the final position of the metatarsal head in the transverse plane. According to the preoperative plan, two half-pins are placed in the distal end of the metatarsal and two in the base of the fourth metatarsal staying outside the metatarsal–cuboid joint (Fig. 2B and C). A 5 mm incision is made lateral to the short metatarsal at the proximal metaphyseal–diaphyseal junction between the two half-pin clusters. A small hemostat is used to dissect down to the metatarsal and with a periosteal elevator gently lift the dorsal periosteum medially and laterally. To begin the osteotomy, a 1.8-mm wire is used to drill multiple orthogonal holes into the metatarsal under fluoroscopic control. The level of the osteotomy should be just distal to the proximal half-pin cluster (Fig. 2D). A small osteotome is then used to complete the osteotomy, without producing excessive displacement of the fragments, and avoiding an extension of the osteotomy into the half-pins (Fig. 2E). The Orthofix Mini-M100 external fixator is applied. Next, pin the MTPJ with a 0.062-in. diameter Kirschner wire. This wire should be inserted from the tip of the toe across the phalanges and then under fluoroscopic control across the MTPJ, stopping at the distal external fixation pins (Fig. 2F). It is necessary to perform a release of the dorsal toe contracture to allow for appropriate toe realignment in order to pin the digit. The Kirschner wire is then bent 90° outside the skin and again 90° over the dorsum of the toe. A third 90° bend is needed to attach it with an end clamp to the external

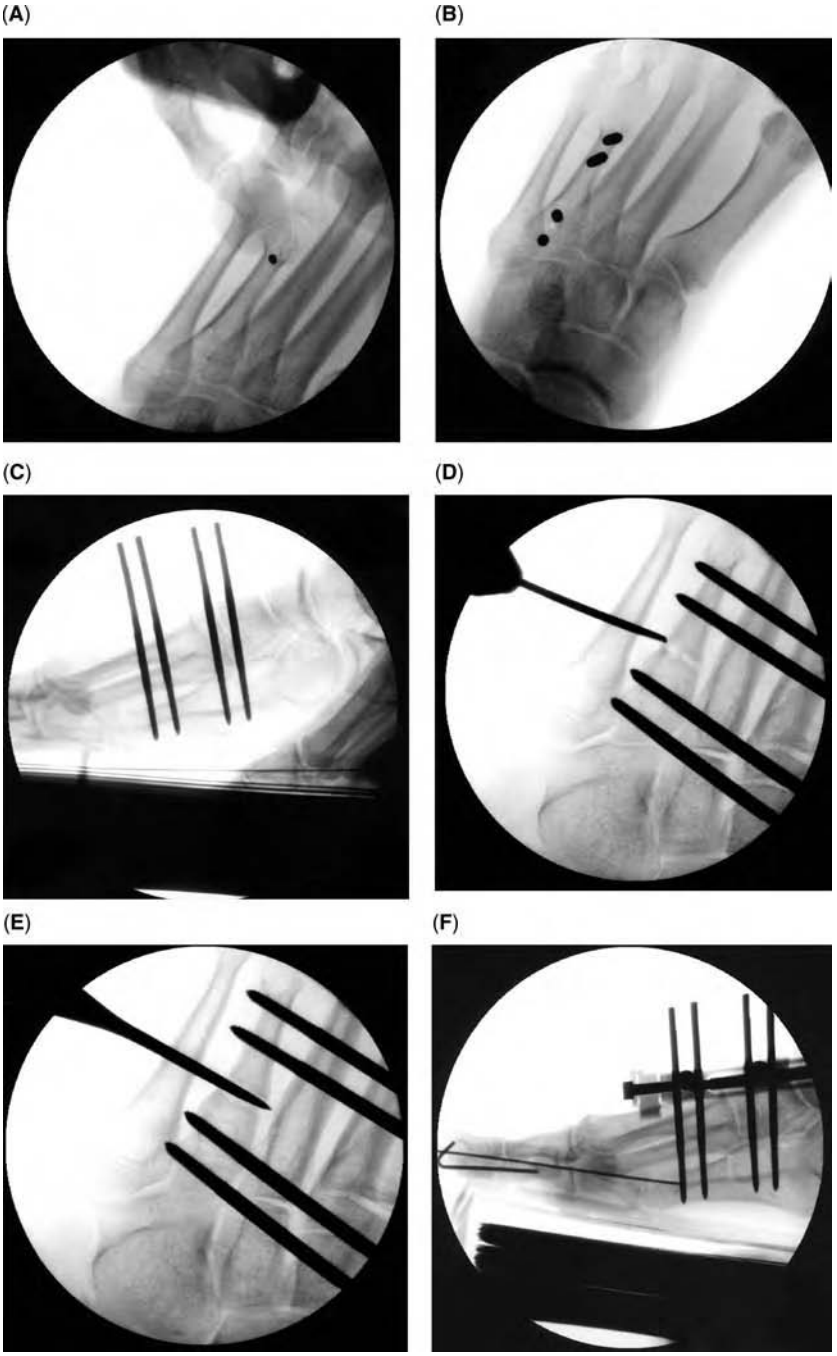

Figure 2 Intraoperative fluoroscopic images detailing surgical technique. (**A**) The first pin is placed at the distal most region of the metatarsal metaphyseal–diaphyseal junction. The half-pin is inserted perpendicular to the longitudinal bisection of the metatarsal (sagittal plane axis) on the lateral view. This pin defines the plane of metatarsal lengthening. (**B**) The second pin is placed at the proximal most region of the metatarsal. Similar to the first half-pin it is inserted perpendicular to the sagittal plane axis of the metatarsal on the lateral view. In addition, the exact medial/lateral position of this second half-pin defines the direction or vector of metatarsal lengthening (two points define a line). This second pin defines the final position of the metatarsal head compared to the adjacent metatarsal heads in the transverse plane. (**C**) Fluoroscopic lateral view confirms all four half-pins are parallel and perpendicular to the longitudinal axis of the short metatarsal in the sagittal plane. (**D**) With fluoroscopic guidance a 1.8-mm wire is used to drill multiple orthogonal holes into the metatarsal at the level of metaphyseal–diaphyseal junction. (**E**) A small osteotome is then used to complete the osteotomy. (**F**) Using a 0.062 in. diameter Kirschner wire the metatarsal phalangeal joint was pinned stopping at the distal pins.

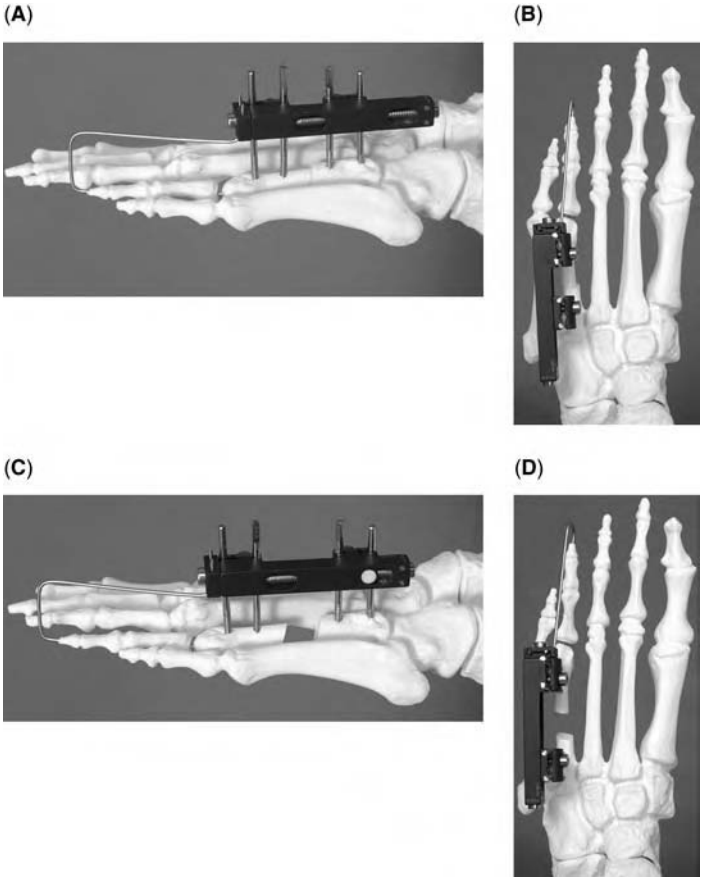

Figure 3 Saw bone demonstration of the frame mounting and technique. (**A**) and (**B**) Front and side view before lengthening. The Kirschner wire is bent so as to attach it with an end clamp into the distal aspect of the external fixator. (**C**) and (**D**) Front and side view after lengthening.

fixator. It is best to orient the fixator so that the distraction knob is proximal and the Kirschner wire clamp is distal (Fig. 3). Traditionally, a separate Kirschner wire was used to stabilize the MTPJ (7). However, this pin can be easily dislodged during the lengthy treatment time. By connecting the digital pin to the external fixation device, the pin is incorporated into the apparatus to form a more stable construct. The percutaneous osteotomy incision is closed and a compressive dressing applied along with toe dressings. Distraction is started after five days, at a rate of 0.25 mm twice a day. When bone is out to length and fully consolidated the frame may be removed (Fig. 4).

It is imperative that four half-pins that are perpendicular to the mid-diaphyseal axis of that metatarsal and parallel to each other be placed in order to achieve accurate sagittal and transverse plane lengthening and thus the final alignment. Because the fixator is mounted perpendicular to the first half-pin, this half-pin ensures the correct plane of bone lengthening. There is almost no room for error in transverse placement of the first half-pin due to the narrow nature of the bone. It is important that the vector of metatarsal lengthening be such that the final position of the metatarsal head is located at the appropriate level in the trasverse plane as determined by the second half-pin.

Bilateral and Multiple Metatarsal Lengthening

Bilateral and adjacent short metatarsals can be lengthened simultaneously (Fig. 5). Hard-soled postoperative shoes protect the forefoot and allow the patient to walk during the postoperative period.

Multiple short and especially adjacent metatarsals within the same foot pose a challenge. The principles of lengthening previously outlined should be maintained while the application of the external fixators are modified. In the case of two adjacent short metatarsals, the first

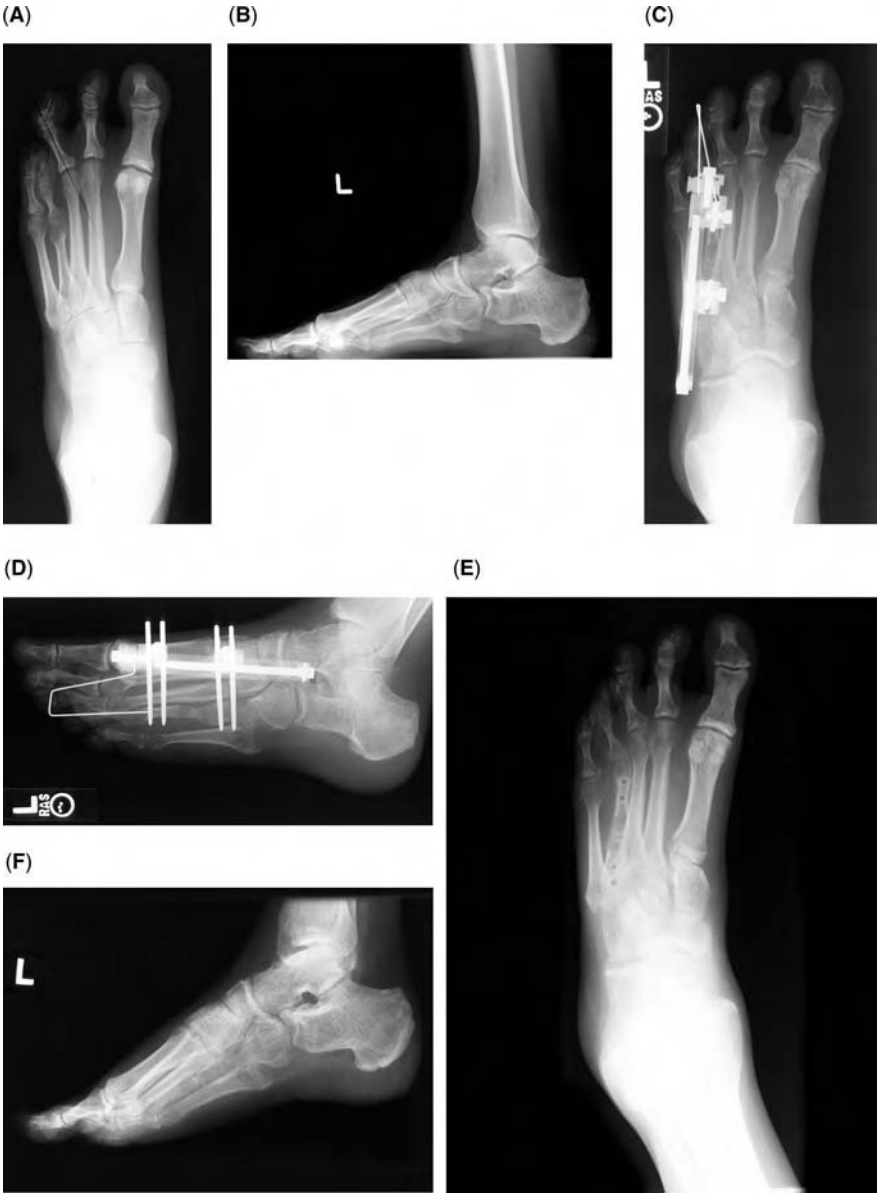

Figure 4 (**A**)–(**F**) A case of congenital short fourth metatarsal: the (A,B) preoperative, (C,D) during lengthening, and (E,F) follow-up radiographs [anterior-posterior (AP), and lateral] are presented.

monolateral fixator is placed in the method outlined in this chapter and the second monolateral fixator is placed oblique (approximately 20–40° angled in the frontal plane) to the first. Technically, this is accomplished by rotating the foot the desired amount of oblique offset to the first fixator half-pins. Insertion of all metatarsal half-pins is accomplished prior to osteotomy and application of either of the two fixators.

COMPLICATIONS

Malalignment

Accurate placement of the first half-pin (most distal) and the second half-pin (most proximal) defines the plane and vector of lengthening in the sagittal and transverse planes, respectively. Sagittal plane malalignment (metatarsal plantarflexion/dorsiflexion) can induce metatarsalgia

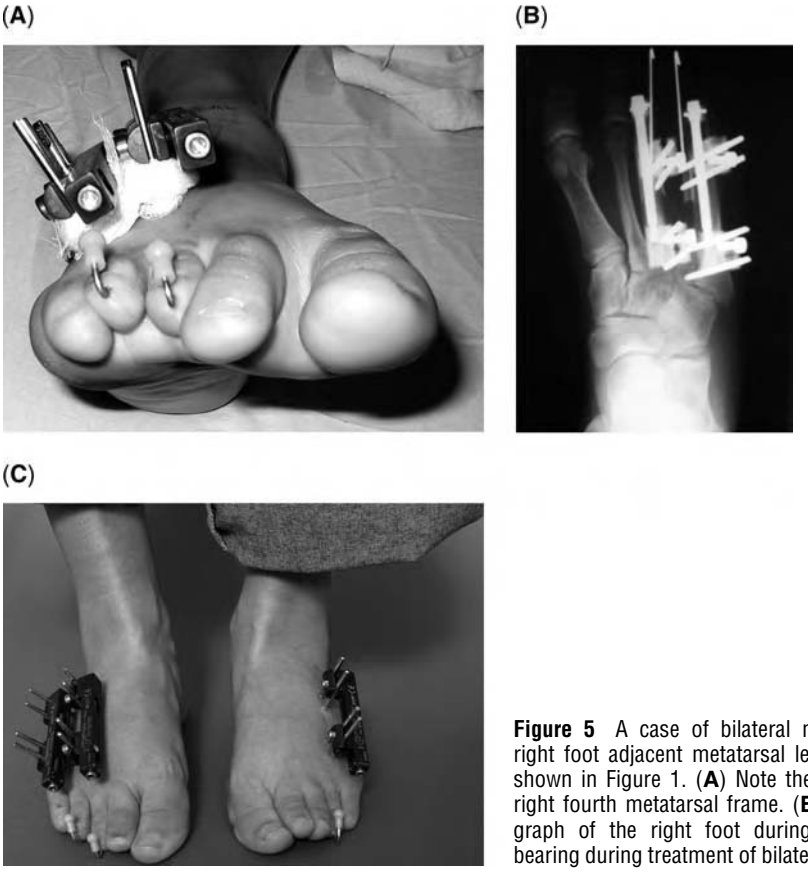

Figure 5 A case of bilateral metatarsal lengthening and right foot adjacent metatarsal lengthening same patient as shown in Figure 1. (**A**) Note the increased obliquity of the right fourth metatarsal frame. (**B**) Weightbearing AP radiograph of the right foot during lengthening. (**C**) Weight bearing during treatment of bilateral metatarsal lengthenings.

and subsequent digital deformity. Creating a plantigrade metatarsal head in the sagittal plane provides the necessary realignment for normal pedal function. Metatarsal head encroachment occurs when the short metatarsal is lengthened such that the head of this metatarsal is not equally spaced between the adjacent metatarsals. This encroachment can induce soft-tissue impingement and subsequent transverse plane digital deformity.

Overlengthening

Inadequate restoration of the metatarsal parabola by overlengthening or underlengthening can produce pain and secondary digital deformities. Follow-up visits every 10 to 14 days are important to monitor the amount of metatarsal lengthening. Also weightbearing radiographs at each visit are critical for determining the amount of metatarsal lengthening gained. The metatarsal parabola angle is the radiographic measure to determine when the lengthening is completed. Routine follow-up is also critical to avoid complications such as premature consolidation and nonunion (5).

Nonunion/Malunion

The technique of a percutaneous minimally invasive osteotomy of the metatarsal is essential for good bone regenerate formation. However, nonunion, malunion, and delayed union of the metatarsal can occur. Deformity can occur if lengthening is performed on an incorrect axis or if pin loosening leads to poor control of the bone. Nonunion and delayed union can occur if lengthening is performed too quickly. Other patient factors may increase the risk of nonunion.

Premature Consolidation

With gradual lengthening, premature consolidation can occur when a patient fails to perform the external fixation adjustments accurately or the rate of distraction is too slow. Premature consolidation can be corrected by performing a repeat osteotomy of the metatarsal, preferably at a different location, with continuation of lengthening (7).

Subluxation

The MTPJ is at risk for subluxation during gradual lengthening of a metatarsal. Patients that require greater amounts of metatarsal lengthening are more susceptible to MTPJ subluxation (24,25). The level of the osteotomy is another contributing factor. The subluxation forces increase when the osteotomy is close to the joint. Soft-tissue rebalancing of the digit and pinning of the MTPJ are important adjunctive steps to maintain a reduced MTPJ during lengthening. Pinning the digit stabilizes the MTPJ throughout the lengthening. Furthermore, attaching this pin to the external fixator helps insure that this important stabilizing pin will not dislodge. Thus, pinning of the digit across the MTPJ is recommended to provide stability and alignment during the lengthening preventing subluxation.

Metatarsalgia/MTPJ Stiffness

Metatarsalgia can result if underlengthening/overlengthening of the metatarsal is performed (2,7). MTPJ stiffness (decreased range of motion), or subluxation of the MTPJ can occur with acute or gradual metatarsal lengthening and especially with overlengthening (21,24,25). Although pinning of the digit is important to prevent MTPJ subluxation during lengthening, it can also increase the risk of postoperative MTPJ stiffness (7,23).

Scarring

Skin incisions can be problematic if hypertrophic. Pin dragging/cutting through the skin with gradual lengthening can increase the amount of scar tissue.

Compliance

Patient education is also important to ensure successful lengthening with external fixation. A patient education class should be preformed with each patient preoperatively. A booklet or handouts should be given and reviewed to improve the patient's understanding, which in turn should improve compliance.

Infection

Pin-tract infections are common with all external fixation devices but typically resolve with oral antibiotics (7).

Acute Lengthening Complications

Donor bone graft site fracture, neurovascular damage secondary to acute lengthening, cast stiffness, and cast sores have been reported with acute lengthening. Urano and Kobayashi (2) had one of 82 patients who underwent acute lengthening developed temporary hypoesthesia; however, none had vascular complications. Avascular necrosis has not been a reported complication. Stress or complete fractures can occur after acute and gradual metatarsal lengthening (2,6,21,24,25). A delayed union rate of 2.5% and a nonunion rate of 2.5% to 63% have been reported for acute lengthening with bone grafting (2,24).

FUTURE DIRECTIONS

As with femoral and tibial lengthening, internal lengthening methods may be developed as an alternative to external metatarsal lengthening. This would have the benefits of avoiding external fixation. The current mechanism of internal lengthening hardware is rotation, and this would be difficult to perform on a single metatarsal. Thus, a different mechanism of lengthening would have to be developed.

REVIEW OF LITERATURE

Authors	Journal, Year	Title	Number of Patients	Results	Conclusions
Baek and Chung	J Bone Joint Surg (Br), 1998	Treatment of congenital brachymetatarsia by one-stage lengthening	21 (34 metatarsals)	Autogenous bicortical iliac crest bone graft used, average gain in length = 14 mm (32% increase)	One-stage lengthening = effective when gradual distraction with Inge retractor for 30 min is performed during surgery
Song et al. (26)	Foot Ankle, Int 2003	Fourth brachymetatarsia treated with distraction osteogenesis	16 (22 metatarsals)	Average gain in length = 16.5 mm (39% increase), average healing index = 72.9 days	Distraction is an effective method, complications = MTPJ stiffness and subluxation
Takakura Y et al.	J Bone Joint Surg, (Br) 1997	Lengthening of short great toes by callus distraction	4 (7 metatarsals)	Distraction is effective for the first metatarsal, excessive lengthening = MTPJ stiffness	Distraction should not exceed 40% of the preoperative metatarsal length
Kawashima T et al.	Ann Plast Surg, 1994	Treatment of brachymetatarsia by callus distraction (callotasis)	3 (4 metatarsals)	Average gain in length = 23.71 mm, average healing index = 112.5 days	Complications = MTPJ subluxation and metatarsal fracture optimal distraction rate = 0.35mm b.i.d.
Kim et al.	J Bone Joint Surg (Br), 2003	The management of brachymetatarsia	12 (35 metatarsals)	Three patterns of metatarsal lengths, shortening and lengthening of metatarsals and phalanges to obtain normalization	By shortening adjacent rays the amount of lengthening is decreased thus one-stage lengthening can be done
Levine et al.	Foot Ankle Int, 1995	Distraction osteogenesis for congenitally short lesser metatarsals	3 (6 metatarsals)	Average gain in length = 15.5 mm, average healing index = 15.5 wk	Distraction was successful, complications = MTPJ stiffness, pin infection, premature consolidation, fracture through regenerate bone

Abbreviation: MTPJ, metatarsal phalangeal joints.

ACKNOWLEDGMENTS

I would like to thank my partners Drs. John Eerzenberg and Dror Paley who have given me the foundation and inspiration for this work. Also special thanks to Alvien Lee for expertise and assistance with the photography.

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Part VI KNEE

22 Osteotomy for Realignment of the Knee

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INTRODUCTION

The treatment of monocompartmental osteoarthritis of the knee is a common indication for knee deformity correction in the adult. Established arthritis in young patients presents a therapeutic dilemma, as physically active patients under the age of 50 years are not ideal candidates for joint arthroplasty. An alternative surgical intervention to minimize pain, increase function, preserve articular cartilage, and delay the need for arthroplasty is desirable. Additionally, congenital or acquired malalignment may result in early arthrosis due to long-standing unequal loads across the knee joint (1). Knee realignment osteotomy may be indicated for patients presenting with deformity, decreased activity level, gait disturbance, pain, or a combination of complaints.

CLINICAL EVALUATION

Patient selection is one of the most important factors for a successful outcome after high tibial osteotomy (HTO). A complete preoperative evaluation often requires more than one office visit to be certain that the patient's expectations match the expected outcome. Although extreme activities such as heavy manual labor and long-distance running are possible after a successful osteotomy, such repetitive impact loading activities will accelerate further degeneration, thereby providing less and shorter duration of pain relief and cartilage preservation. Patients older than 65 years are usually better served with arthroplasty, although older patients with particularly high activity demands may be considered for osteotomy.

A careful evaluation of the location and nature of the pain is critical to the success of a contemplated osteotomy. Pain due to joint overload or early arthritis is often made worse by prolonged activity and may be present at night. Pain that is definitively localized to the joint line of the involved compartment (Fig. 1) is more reliably alleviated by osteotomy than pain that is diffuse or localized to other compartments. One exception to this rule may be mild retropatellar pain (2,3). Mechanical symptoms should alert the examiner to possible meniscal pathology that may require arthroscopy.

A history of inflammatory joint disease is a contraindication to performing realignment osteotomy, as is radiographic evidence of multicompartmental arthritis or chondrocalcinosis. Clinical impression among experienced surgeons is that avascular necrosis and oseteochondritis dessicans are often accompanied by inflammation, which does not respond as reliably to offloading osteotomy. A history of cigarette smoking is a relative contraindication to surgery, as is a history of lateral meniscectomy. Obesity renders the surgery more technically demanding, hampers postoperative mobility, and heightens the risk of implant failure. Despite this, the young, obese patient is often more suitably treated with osteotomy than with arthroplasty. Patients with a history of chronic pain syndrome or those with an excessively introspective personality report less satisfaction postoperatively than patients of a more stoic nature.

Physical examination begins with an assessment of the alignment of the lower extremities along with any leg length discrepancy. Opening wedge osteotomy can be expected to slightly

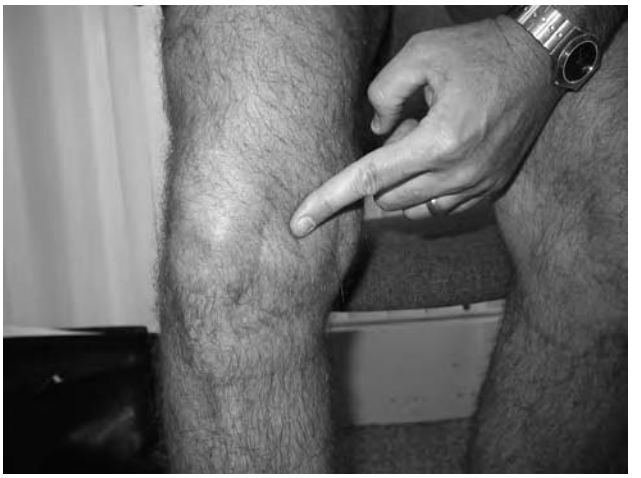

Figure 1 Typical localization of isolated medial compartment pain.

increase the length of the leg, while a closing wedge will have the opposite effect. Gait is observed for the presence of a dynamic, weight-bearing thrust to the knee, indicating ligamentous laxity. Gait analysis has shown that a lateral thrust is associated with a poor outcome unless overcorrection of the mechanical axis at the time of surgery is achieved (4,5). Previous skin incisions are noted in addition to the condition of the soft-tissue envelope surrounding the knee. Severe scarring may prevent closure of the skin following an opening wedge osteotomy. Palpation of the knee should ideally demonstrate tenderness of the involved compartment and minimal tenderness at other locations. Range of motion should be noted, with attention to the presence of any flexion contracture that may be worsened by an opening wedge—-type correction. Conversely, small contractures may be amenable to correction at the time of closing wedge or fixator-assisted osteotomy. Flexion contracture of greater than 10° or flexion of less than 50° is relative contraindications to HTO. The degree of static and dynamic laxity is noted in the coronal and sagittal planes. Coronal and multiligamentous instability will be exacerbated by closing wedge realignment, as this renders the collateral ligaments relatively lax. Similarly, an inadvertent increase in the posterior tibial slope by an opening wedge correction will create flexion deformity and may exacerbate pre-existing anterior cruciate ligament insufficiency.

Radiographic evaluation includes a bilateral, 51-in. standing anteroposterior (AP) X-ray, 45° flexed-knee Rosenberg postero-anterior (PA) view, patellar Merchant views, and a lateral X-ray. Some surgeons prefer single long-leg–standing X-rays to more accurately reproduce the stance phase of gait; however, it has been shown that any static standing radiograph is an imprecise predictor of the correction necessary for HTO (6). Flexed-knee PA views are critical to assess posterior joint wear, which may not be evident on regular AP roentgenographs (Figs. 2A and B).

Evidence of degenerative change in each compartment is assessed. The tibial slope, height of the patella, and any increased joint line obliquity is noted. Presence of osteochondral lesions is identified. The leg lengths are measured on the long-leg view. Other deformity at the hip and ankle, as well as the presence of arthritis in these joints, is sought.

The overall mechanical axis is drawn on the long-leg view from the center of the femoral head to the center of the talus (7). The degree of varus or valgus is determined in the following way. The mechanical axis of the femur, from the femoral head to the medial tibial spine, is drawn. The mechanical axis of the tibia, from the center of the talus to the medial tibial spine, is drawn. The angle formed by the intersection of these two axes at the knee is the amount of deformity present (8). The mechanical lateral distal femoral angle (mLDFA) is then measured by drawing a line along the lateral and medial femoral condyles (7). The angle formed when it crosses the mechanical axis of the femur is the mLDFA. Similarly, the mechanical medial proximal tibial angle (mMPTA) is calculated by drawing a line across the medial and lateral tibial plateaus. The mMPTA is measured at the intersection of this line with the mechanical axis of the tibia. The normal value for each of these measurements is $87° \pm 4°$ (Fig. 3) (7). Values deviating from the norm indicate whether the source of the malalignment is in the tibia, the femur, or is a combination of both. Additionally, the two lines drawn along the joint

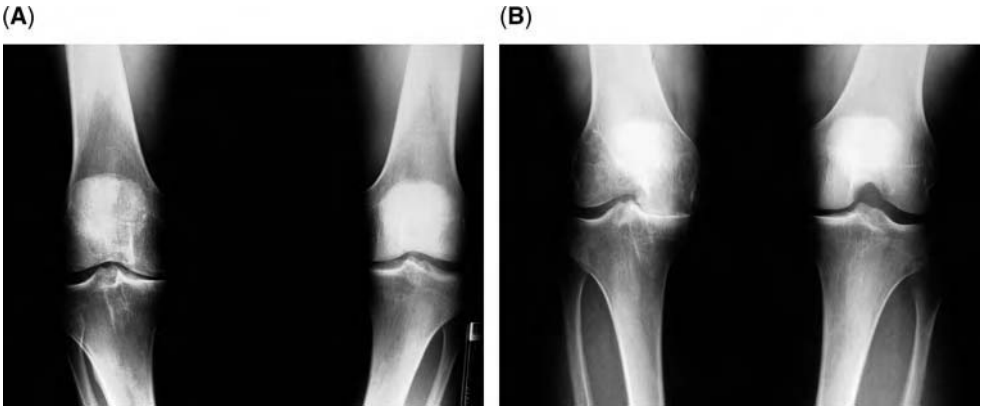

Figure 2 (**A**) Right knee moderate medial joint space narrowing, anteroposterior (AP) view. (**B**) Same knee as 2A, Rosenberg view, showing extensive medial changes not appreciated on AP view.

surfaces of the femur and tibia themselves form the joint line convergence angle (JLCA). The normal JCLA is 2°. An increased value indicates ligamentous laxity of the knee (9).

Once the source of the deformity is identified along with its magnitude, the apex of the deformity should be located. This is important to localize, as a periarticular osteotomy will induce translation of the involved bone if the apex of the deformity is located remote from the osteotomy site (9).

CLASSIFICATION

Knee deformity may be categorized as originating in the tibia, femur, or both. This is determined using the radiographic angles described above. Corrective osteotomy should be generally performed on the bone with the greatest amount of deformity, although small varus deformities originating in the femur may be corrected at the proximal tibia, as long as the resulting joint obliquity is 10° or less (10). Valgus osteoarthritis is best corrected via a medial closing wedge osteotomy of the femur, as lateral opening wedge tibial osteotomy carries a higher rate of peroneal nerve palsy postoperatively (11). Combined deformities exceeding 5° at each site may warrant osteotomies of the distal femur and proximal tibia to avoid joint line obliquity. In the case of deformity resulting from cartilage loss without bony abnormality, varus should be corrected through the tibia and valgus through the femur.

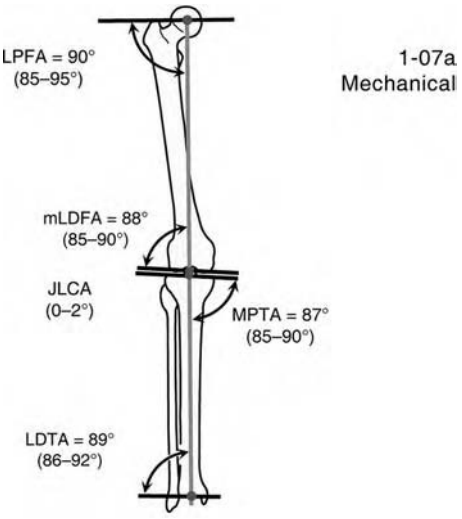

Figure 3 Normal mechanical axis and joint alignment angles. *Source*: Reprinted from Ref. 9.

TREATMENT OPTIONS

Varus deformity associated with medial compartment osteoarthritis is the most common malalignment treated with osteotomy. Varus deformity of the tibia is assessed in three key areas: (1) the degree of deformity present, (2) the ligamentous stability of the knee, and (3) the presence or absence of osteoarthritis.

Selection of an osteotomy technique for varus deformity of the tibia is based on the degree of deformity present and the degree of stability of the knee. The magnitude of acute correction is limited to 15° in most patients. Small-to-moderate deformity (less than 15°) with stable ligaments can be treated with lateral closing wedge HTO.

Knees with moderate deformity (less than 15°) and unstable ligaments benefit from opening wedge HTO, as this will relatively tighten the soft tissues.

Knees requiring deformity correction greater than 15° are best treated with a gradual correction technique using a circular external fixator. The dome osteotomy allows for greater correction magnitude, with less effect on joint line obliquity. It is done below the tubercle, thereby avoiding deleterious effects on the extensor mechanism. As the osteotomy becomes more diaphyseal, however, the healing rate may be expected to become slower. Also, when the osteotomy is away from the periarticular apex of deformity, intentional translation is needed at the osteotomy.

Acute removal of a lateral wedge larger than 15° would render the collateral ligaments unstable and would create excessive deformity of the proximal tibia. In addition, an excessively large wedge removal would cause difficulty approximating and holding the osteotomy surfaces and may risk neurocompartmental compromise.

Acute distraction wedges of greater than 15° may also have deleterious effects of: neurocompartmental compromise, excessive tightening of the patellar ligament with resulting patellofemoral pressure, tightness of the posterior soft tissues with resulting flexion contracture, and delayed union.

The presence of arthritic changes in the involved compartment or the presence of a lateral thrust with gait necessitates overcorrection of the deformity. Several studies suggest that obtaining an adequate amount of overcorrection is the single strongest predictor of a good outcome following osteotomy (5,12–16). The degree to which the weight-bearing line should be shifted remains controversial. Fujisawa studied the amount of correction as related to outcome when advanced medial degenerative changes were present. Based on these studies, the mechanical axis should be placed lateral to the lateral tibial spine, but not beyond a point half the width of the lateral compartment. This area has now become known as the Fujisawa point, and is generally considered to be 62% of the distance from the medial margin of the tibial plateau to the lateral margin (17). Arithmetic calculation provides this linear distance across the plateau and provides a reproducible goal for surgeons to achieve. Knees without evidence of medial arthritis are corrected to neutral mechanical alignment, so that the mechanical axis falls through the medial tibial spine.

SURGICAL TECHNIQUES

A careful preoperative plan is essential to obtaining an appropriate surgical correction and outcome, regardless of the surgical technique selected. Determination of the magnitude of the deformity, its apex, and plane is necessary. Many methods have been described to preoperatively calculate the correction magnitude. Each of these methods is affected by preoperative measurements, intraoperative accuracy, intraoperative simulation of weight bearing, and the ability to maintain the correction achieved postoperatively. It is recommended that two of these methods be employed in each preoperative plan, thereby minimizing calculation error. One method is the paper cutout technique, which is a tracing of a full-length standing AP X-ray and a trial osteotomy performed with scissors on the paper, allowing for the desired amount of correction to be tested. A second method involves adding the magnitude of angular deformity with the desired amount of overcorrection plus the JLCA. This will yield the overall magnitude of the proposed correction.

Closing Wedge High Tibial Osteotomy Technique

A curvilinear incision of 10 cm is made from the fibular head to the anterolateral tibial crest and continued distally. The anterior compartment muscles are elevated subperiosteally while

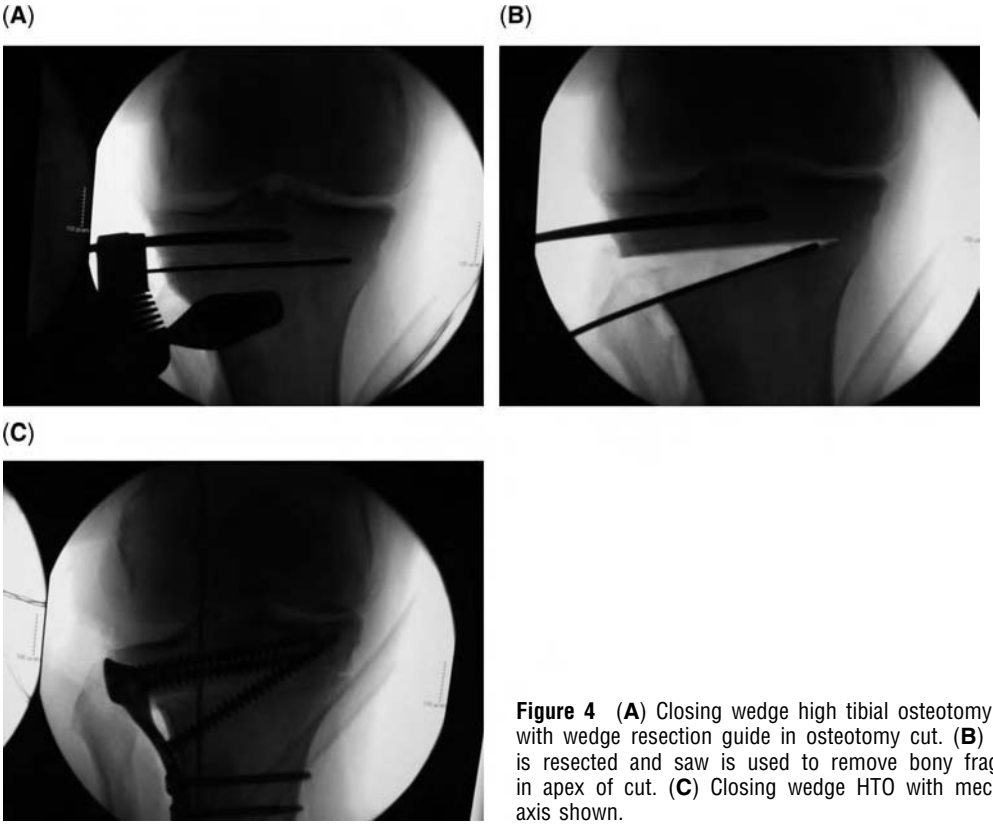

Figure 4 (**A**) Closing wedge high tibial osteotomy (HTO) with wedge resection guide in osteotomy cut. (**B**) Wedge is resected and saw is used to remove bony fragments in apex of cut. (**C**) Closing wedge HTO with mechanical axis shown.

the posterior and anterior tibia under the patellar tendon are also exposed. A Cobb elevator releases the proximal tibiofibular joint (PTFJ) capsule until it easily springs apart. Avoid levering on the joint, however, as this may fracture the fibular neck. The joint may be removed with an osteotome if it is particularly stiff or if the correction needed is large. A transverse cutting jig is placed parallel to the knee joint and a lateral fluoroscopic image confirms parallel placement in the sagittal plane. Osteotomy is cut with a saw under irrigation, stopping 1 cm from the medial cortex. A calibrated oblique cutting guide is then placed in the osteotomy and the desired degree of wedge is cut and removed. Small curettes and the saw are used to remove remnants of the wedge. A 5-hole, L-plate (Zimmer, Warsaw, Indiana, U.S.) is affixed proximally and the gap is slowly closed with an osteotomy clamp (Zimmer, Warsaw, Indiana, U.S.), taking care to prevent soft-tissue interposition anteriorly and posteriorly. Alignment is checked with a cautery cord, held at the centers of the femoral head and talar dome. At the desired amount of correction, the most distal screw is placed. The third most distal hole is used to place an oblique screw across the osteotomy, and the final distal screw is inserted (Figs. 4A–C). The removed wedge is morsellized and the cancellous graft is soaked in autologous platelet concentrate before packing it posteriorly around the osteotomy site. The wound is closed over a drain in the subcutaneous space. The patient is allowed 50% weight bearing with a knee immobilizer for the first month and is then progressed as tolerated.

Opening Wedge High Tibial Osteotomy Technique

A 4-cm vertical incision is made half-way between the anterior and posterior border of the medial tibia, beginning 2 cm distal to the joint line. The tibia is exposed subperiosteally including under the patellar tendon, posteriorly, and distally and retractors are placed in these locations. A saw is positioned with fluoroscopy parallel to the joint and the tibia is scored. A lateral image is checked to ensure that the cut is parallel to the joint in the sagittal plane. The osteotomy cut is made under constant irrigation, stopping 1 cm from the lateral cortex.

A 4.8-mm drill bit is then used to create a ''relaxing hole'' at the apex of the osteotomy cut, so as to distribute the distraction force and prevent fracture into the joint. Two lamina spreaders are then placed, anteriorly and posteriorly in the cut, and the osteotomy is slowly opened, hinging on the lateral cortex. Alignment is then checked with a cautery cord, held at the centers of the femoral head and talar dome. At the desired amount of correction, an allograft corticocancellous wedge is fashioned to fit the distraction gap. The wedge is first coated with autologous platelet concentrate and the excess cancellous bone is packed into the apex of the osteotomy cut. A T-plate (DePuy, Warsaw, Indiana, U.S.) is applied, using the third most distal hole to place an oblique screw across the osteotomy, thereby locking the construct. The most distal screws may be placed percutaneously (Figs. 5A–C). For patients weighing more than 100 kg, a medial tibial locking plate is used (Synthes, Paoli, Pennsylvania, U.S.). The wound is closed over a drain in the subcutaneous space. The patient is allowed 50% weight bearing for the first month and is then progressed as tolerated.

High Tibial Osteotomy with Circular External Fixator Technique

A fibula osteotomy is first carried out if correction greater than 20° or lengthening is planned. This is performed at the distal two-third to one-third of the fibula using a small saw and the incision is then closed. When correcting a varus deformity, an oblique fibula osteotomy is necessary to allow some shortening.

A preconstructed hexapod frame (Taylor Spatial Frame) (Smith & Nephew, Memphis, Tennessee, U.S.) is affixed to the proximal tibia via a 1.8 mm reference wire, parallel to the joint and on the proximal side of the ring. A third ring with a wire across the syndesmosis may be attached just above the ankle to control the fibula and protect the tibia fibula relationship at the ankle. This is most important in the setting of lengthening, large angular and/or rotational deformity correction. A 6-mm hydroxyapatite-coated half-pin secures the distal ring to the tibia, taking care to properly center the leg in the frame. A nonbeaded wire affixes the distal fibula to the third ring when necessary. A second beaded wire is placed on the distal side of

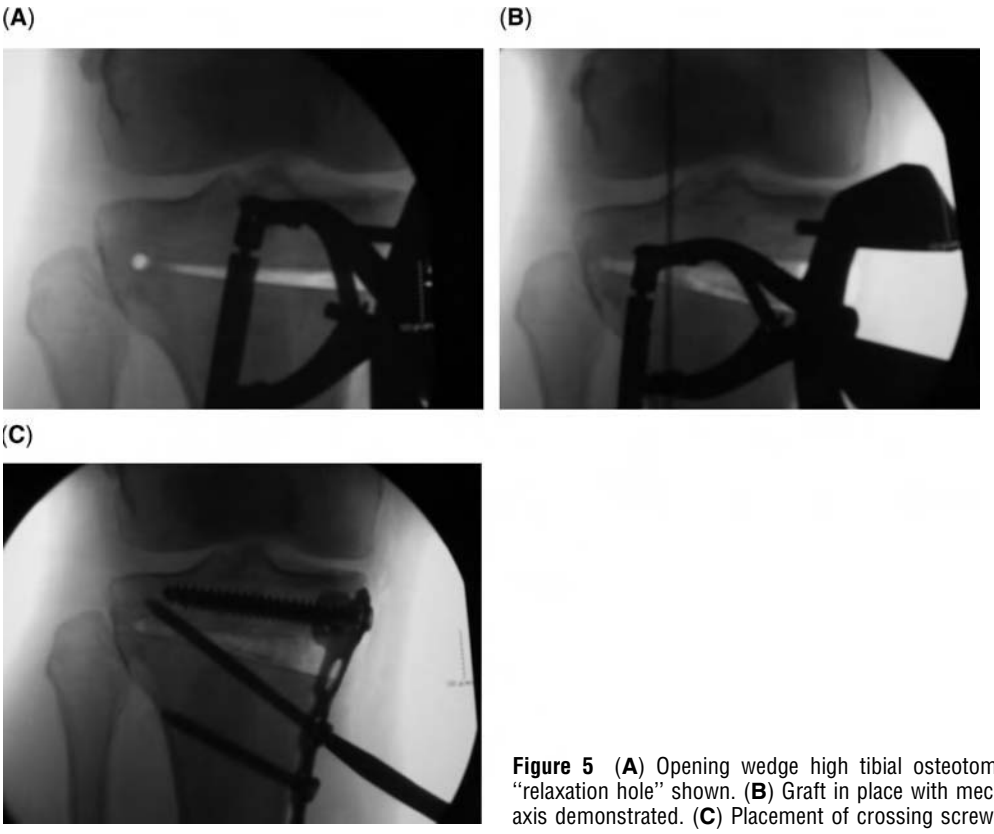

Figure 5 (**A**) Opening wedge high tibial osteotomy with ''relaxation hole'' shown. (**B**) Graft in place with mechanical axis demonstrated. (**C**) Placement of crossing screw.

the proximal ring. A half-pin is affixed proximally using a one-hole Rancho cube. A total of three half-pins are then placed on the middle ring and a second half-pin is placed on the third ring. Three fixation points now secure each ring.

Alternatively, a *rings first method* can be used to apply the Taylor Spatial Frame (Smith & Nephew, Memphis, Tennessee, U.S.). Each ring is applied and fixed to the leg independently on either side of the osteotomy. The rings are then connected with six struts in a specified manner.

Corticotomy is carried out after the struts between the first and the second rings are detached. A 2-cm incision over the crest of the tibia, just distal to the tubercle, is made. The bone is exposed subperiosteally, medially, and laterally. A 4.8-mm drill bit is used to create multiple holes in the tibia. An osteotome completes the corticotomy. Completeness is confirmed under fluoroscopy. The periosteum and skin are closed and the struts are reattached, and osteotomy compressed. The angular deformity correction is carried out post-operatively with the aid of internet-based software program. AP X-rays are taken weekly to judge regenerate bone formation. Full-length standing AP X-rays are taken as the correction nears completion to ensure precise axis correction (Figs. 6A–F). The frame is removed under outpatient sedation when solidly healed, usually 8 to 14 weeks after application, allowing the patient full-weight bearing throughout treatment.

Complications

As with any surgical procedure, a variety of intraoperative, early, or late complications may arise. Pitfalls encountered during closing wedge HTO include failure to maintain a subperiosteal

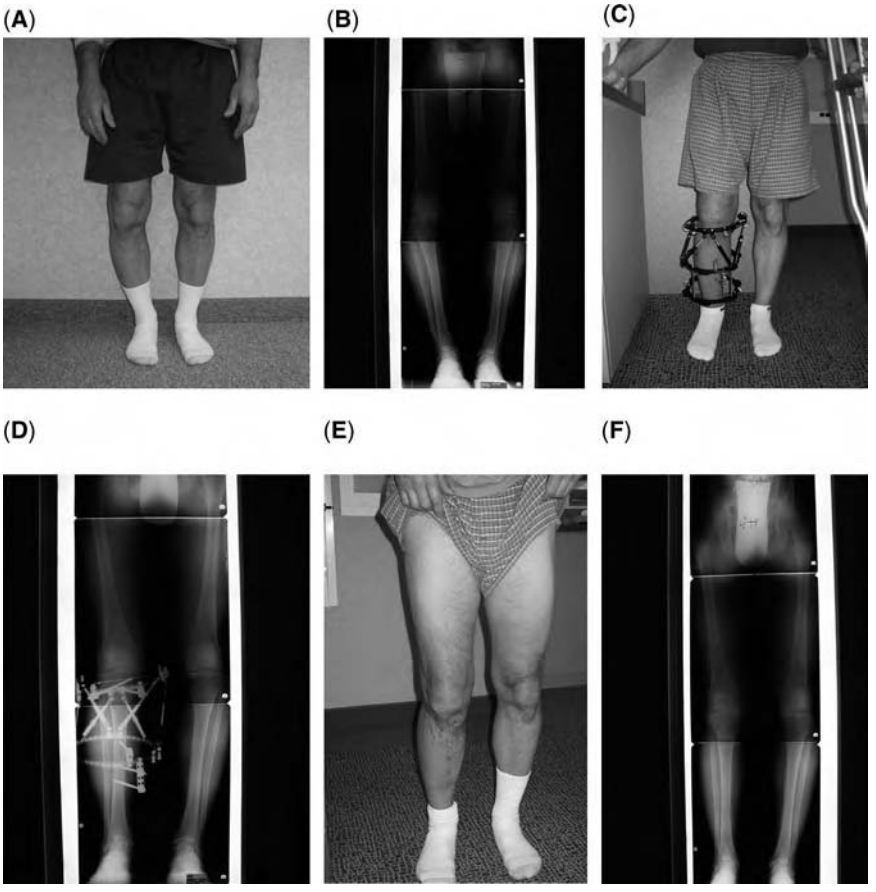

Figure 6 (**A**) Right varus knee, pre-op. (**B**) Standing X-ray, pre-op. (**C**) Hexapod frame with third distal ring for HTO with external fixation. (**D**) Standing X-ray, after correction of deformity. (**E**) Outcome after frame removal. (**F**) Standing X-ray after frame removal.

Table 1 Decision Making in Proximal Tibial Osteotomy

	Stable Ligaments	Unstable Ligaments	Osteoarthritis Present	No Osteoarthritis	Minor Leg Length Discrepancy Present	Major Leg Length Discrepancy Present
Varus $\leq 15^\circ$	Closing wedge	Opening wedge	Overcorrect to 4° mechanical valgus	Correct to 0° mechanical valgus	Opening wedge	External fixator; add distraction for length
Varus $> 15^\circ$	External fixator	External fixator	Overcorrect to 4° mechanical valgus	Correct to 0° mechanical valgus	External fixator; add distraction for length	External fixator; add distraction for length

dissection during mobilization of the anterior compartment muscles. This may cause excessive postoperative pain, mimicking a compartment syndrome. Excessive leverage on the PTFJ may cause a fracture of the fibular neck and concomitant peroneal nerve injury. Failure to mobilize the PTFJ adequately will prevent closure of the tibial osteotomy site. This requires excision of the PTFJ with an osteotome until it is freely mobile. Another cause of incomplete closure of the osteotomy site is interposed soft tissue anteriorly or posteriorly. This is best prevented by reinsertion of retractors at the time of osteotomy closure. The medial apex of the osteotomy site may fracture through the medial tibial cortex as the osteotomy is closed.

Opening wedge HTO may be complicated by intra-articular fracture if the apex of the osteotomy is nearer to the subchondral bone than to the lateral tibial cortex. Utmost care must be taken when distracting the cut and when placing the graft to ensure that no opening in the sagittal plane is occurring, as this will both increase the tibial slope and cause difficulty closing the soft tissues. Complications specific to use of the external fixator include pin-tract infections as well as those associated with conventional osteotomy.

Nonunion occurs at a rate of 0.5% to 1% (18) and may be treated with ultrasonic stimulation, open bone grafting with fixation exchange to a more sturdy construct, conversion to external fixation, or by total knee arthroplasty. Patella baja may be induced by both opening and closing wedge osteotomies as a result of shortening and scarring of the patellar tendon. This may complicate a future surgical exposure for a knee arthroplasty. Early postoperative knee motion may help prevent patella baja (19). Avascular necrosis of the proximal fragment may occur if the thickness of the fragment is less than 2 cm (18). Deep infection is uncommon following HTO, while superficial infections and pin-site infections are seen more regularly. Compartment syndrome is an ever-present risk and is best prevented by meticulous surgical technique and use of a drain postoperatively. Clinically, significant deep vein thrombosis (DVT) is uncommon; however, one study has found a rate of DVT of 58% (20), indicating that prophylaxis is warranted. Major vascular injury is rare but is best prevented by placement of a posterior retractor immediately adjacent to the tibia. Neurologic injury following HTO most commonly affects the peroneal nerve. Osteotomy of the fibula should be undertaken at the junction of the middle and distal thirds of the fibula to minimize the risk (18).

FUTURE DIRECTIONS

Computer-assisted technology that further refines preoperative planning, and the degree of correction obtained intraoperatively may facilitate both opening and closing wedge HTOs in the future.

EDITOR'S ADDENDUM

We prefer to avoid closing wedge proximal tibia osteotomies in order to preserve metaphyseal bone stock and avoid problems with proximal migration of the fibula and lateral collateral ligament laxity. External fixation and gradual correction has become our treatment of choice for even small- and medium-sized deformities. Accuracy of correction and postoperative adjustability are high with this method. This is particularly helpful when there is joint line convergence contributing to the deformity.

For simple tibial deformities (coronal plane varus deformity of less than 10°), we use the monolateral hemicallatosis frame with a loose hinge (EBI, Parsippany, New Jersey, U.S.). No

fibula osteotomy is needed as the proximal tibia fibula joint and lateral tibial cortex act as the hinge for the gradual opening wedge correction.

For complex deformities (coronal plane deformity greater than 10°, oblique plane deformity, flexion contracture of the knee, rotational deformities, lateral collateral ligament laxity, and AP knee instability), we use a two-ring Taylor Spatial Frame (Smith & Nephew, Memphis, Tennessee, U.S.) construct and a gradual correction. A third ring with a syndesmosis wire is used in the setting of lengthening, rotational deformity correction of greater than 15°, and angular deformity correction of greater than 20°.

REVIEW OF THE LITERATURE

References	Title	Number of Patients	Results	Conclusions
(19)	HTO with a calibrated osteotomy guide, rigid internal fixation, and early motion	64 HTOs	67% good or excellent at avg. 8.5 yr. 33% had TKA. No patella baja observed	Use cutting jig, rigid internal fixation, early ROM
(12)	Proximal tibial osteotomy	87 HTOs in 73 patients	If valgus =/> 8° at 1 yr, 5 yr survival = 90%, 10 yr = 65%	Risk failure if < 8° valgus or with obesity
(15)	Survivorship of the high tibial osteotomy.	106 HTOs in 85 patients; 94 were closing wedge, 12 were dome osteotomy	73% 5 yr survival; 51% at 10 yr. Subset < 50 yr and flexion > 120° had 95% 5 yr, 80% 10 yr, 60% 15 yr	Survival improved by age < 50 yr, sufficient valgus correction, no lateral thrust pre-op
(16)	Tibial osteotomy for the treatment of varus gonarthrosis	76 HTOs in 66 patients	10 yr survival 74%. If valgus 8–16° 10 yr survival 90%	Sufficient valgus correction most important for survival
(13)	The influences of biomechanical factors on cartilage regeneration after HTO	58 HTOs in 47 patients. Mechanical axis correction to 74% across plateau	Partial or full medial compartment fibrocartilage coverage in 55%	Sufficient valgus correction most important for cartilage regeneration and functional outcome
(14)	Regeneration of degenerated articular cartilage after high tibial valgus osteotomy	146 HTOs in 115 patients	Partial or full medial compartment coverage in 91%	Valgus >5° correlated with full cartilage regeneration and increased medial joint space
(5)	Relationship between gait and clinical results after HTO	32 HTOs	Only alignment significantly associated with clinical results at 6 yr	Pre-op peak adduction moment does not correlate with clinical or radiographic outcome if sufficient valgus is achieved
(10)	Proximal tibial varus osteotomy for OA of the lateral compartment of the knee	31 varus-producing HTOs in 28 patients	Avg. 9.4 yr, 77% had no or mild pain	Correct to 0° anatomic axis. May operate on tibial side if valgus =/<12° or if resulting joint obliquity = /<10°
(11)	Proximal tibial varus osteotomy	34 varus-producing HTOs	Avg. 11 yr, 88% good or excellent. 9% transient peroneal nerve palsy	Correct to 0° anatomic axis. Good option if valgus deformity is in tibia
(21)	Distal femoral varus osteotomy for OA of the knee	30 medial closing wedge with blade plate	Avg. 8.3 yr, 83% satisfactory	Reliable procedure for lateral OA of the knee

Abbreviations: Avg., average; HTO, high tibial osteotomies; OA, osteoarthritis; TKA, total knee arthroplasty; ROM, range of motion.

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23 Proximal Tibial Osteotomy for Medial Compartment Osteoarthritis of the Knee Using the Taylor Spatial Frame

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INTRODUCTION

Medial compartment arthritis of the knee is a common problem that is typically associated with varus alignment and a weight-bearing axis that lies in the medial compartment. Knee flexion or hyperextension deformity or ligamentous instability may also be present. The young patient who is not an ideal candidate for total knee replacement may benefit from proximal tibial osteotomy for pain relief. The Ilizarov method with the Taylor Spatial Frame (TSF) (Smith and Nephew Inc., Memphis, TN) is a powerful tool for accomplishing proximal tibial deformity correction. A specialized feature of the TSF is its virtual hinge that allows for the simultaneous correction of multiplanar deformities and the stabilization of the ligament deficient knee. The power of the spatial frame lies in its precise control over final limb alignment and in its ability to perform a residual correction. The stability of the ringed construct permits early weight bearing and provides an ideal environment for new bone formation and a rapid healing response. Classic Ilizarov principles are followed to ensure proper frame application while the web-based computer program directs the gradual correction process. Patient involvement is crucial to the success of this technique. Computer-generated schedules and easy-to-read struts greatly simplify adjustments for patients. The most common complication is pin tract infection that is adequately treated with a course of oral antibiotics in nearly all cases. The goal of this chapter is to describe the advantageous role of this method of realignment and the basic technique used to perform the correction.

TREATMENT OPTIONS

Sir Robert Jones, who used a closing wedge osteotomy for the correction of tibia vara in children, first described the operative realignment of the proximal tibia in 1800. Jackson reported success with this osteotomy in adult patients suffering from osteoarthritis associated with varus knee alignment. Coventry refined and popularized the high tibial osteotomy technique, and good results have been obtained using this method (1–4). Closing wedge osteotomies, however, has been associated with problems including an increased incidence of patella baja and loss of tibial inclination—both of which result in a difficult conversion to total knee arthroplasty (5). Other obstacles include tibial bone loss, a lack of accuracy, and the inability to alter residual angular deformity postoperatively (6–10).

Owing largely to the work of Ilizarov, distraction osteogenesis has gained worldwide recognition as a versatile technique for correcting a host of bony abnormalities. The application of this gradual realignment method toward the treatment of medial compartment knee gonarthritis has been undertaken with great success (11–14). With the use of gradual correction techniques, problems leading to difficult conversion to knee arthroplasty typical of closing wedge osteotomy are minimized. The ability to alter the fixator in the postoperative period to improve limb alignment has been one of the most appealing aspects of using external fixators with osteotomies. Both monolateral external fixators and Ilizarov ringed fixators have been designed to perform gradual limb realignment. Unilateral frames are

Table 1 Clinical Decision Making

Proximal tibial deformity	Monolateral Hemicallatosis Frame without Fibula Osteotomy (Gradual Correction)	Two-ring TSF with Fibula Osteotomy (Gradual Correction)	Three-ring TSF with Fibula Osteotomy and Syndesmosis Wire (Gradual Correction)
Varus $<10^\circ$	X	–	–
Varus $>10^\circ$	–	X	–
Valgus deformity	–	X	–
Sagittal plane deformity	–	X	–
Axial plane (rotational) deformity $<15^\circ$	–	X	–
Axial plane (rotational) deformity $>15^\circ$	–	–	X
Angular deformity $>20^\circ$	–	–	X
Need for >1 cm of lengthening	–	–	X

Abbreviation: TSF, Taylor spatial frame.

comfortable and well tolerated by patients. They are light weight and fit under most clothing making them less conspicuous. However, unilateral frames are less versatile than circular fixators and do not allow for the correction of complex deformities (15). We use monolateral frames in conjunction with proximal tibial osteotomy for the gradual correction of varus tibial deformity of less than 10° not associated with other bony or soft tissue abnormalities (Table 1).

When compared with monolateral fixators, Ilizarov ringed external fixators offer increased stability (15). This may decrease the incidence of early loss of correction, a known complication of all methods of tibial osteotomy. The traditional Ilizarov frames allow for the correction of multiple deformities through a common osteotomy site performed as separate steps of a staged procedure. These fixators are technically demanding but have yielded good results avoiding many of the complications of the Coventry osteotomy (14,16).

The TSF has been utilized in the treatment of medial compartment pain associated with varus malalignment. The strut adjustments are easy to perform and the computer printed schedule is relatively simple for patients to follow. The TSF is far more forgiving than the traditional Ilizarov ringed system. The computer can precisely control the Spatial frame's virtual hinge, drastically reducing time spent on preoperative calculations and on intraoperative hinge positioning. A great strength of the TSF lies in its ability to correct large varus deformities and multiplanar deformities. The TSF can simultaneously incorporate and correct varus malalignment with flexion deformity and internal or external rotation deformity. Lengthening or shortening at the osteotomy site may be added as well. Residual deformity remaining at the termination of the correction with a TSF requires only a simple schedule adjustment without any need for further surgery or changes in the frame construct. A residual program can also be run once the desired alignment is obtained to provide compression across the osteotomy for enhancement of bony healing (17). Disadvantages of circular fixation include the bulk of the frames, the need for meticulous pin care of multiple sites, and more discomfort related to the use of tensioned wires.

INDICATIONS AND CONTRAINDICATIONS

The ideal candidate for the proximal tibial osteotomy is a patient less than 60 years old with complaints of isolated medial joint line pain exacerbated by weight bearing activities. Radiographs demonstrate medial compartment arthritis and varus alignment. Patients are counseled that the goal of this surgery is to alleviate the medial pain and slow the progression of further damage to the medial compartment articular cartilage. The most compelling indications for the use of the TSF in conjunction with proximal tibial osteotomy include patients with large and complex deformities about the knee joint, ligamentous laxity, or limb shortening (Figs. 1A–N).

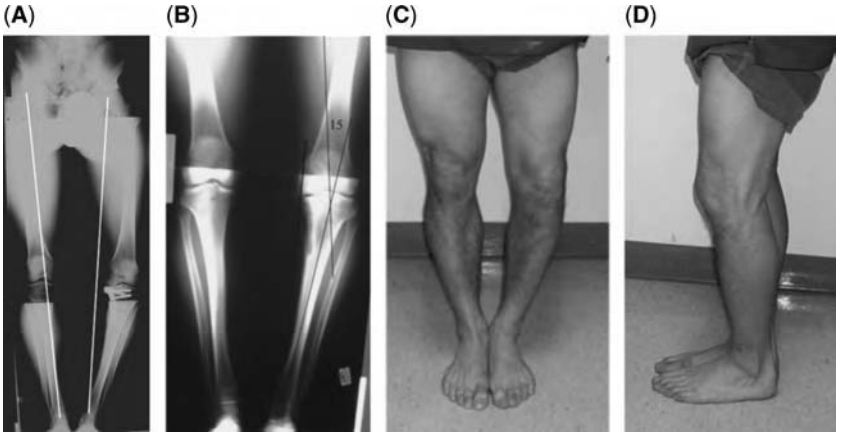

Figure 1 (*Continued on next page*) (**A**)–(**N**) Case example of 37-year-old male with medial compartment arthritis after a medial plateau fracture. (**A**) Erect 51 in. bipedal radiograph showing the center of the hip and ankle joints on one cassette. The left lower extremity has a medial mechanical axis deviation of 37 mm. (**B**) Standing anteroposterior radiograph of bilateral lower zextremities. The left lower extremity has 15° of varus deformity. (Medial proximal tibial angle is 80° and joint line convergence angle is 5°). Advanced medial compartment degeneration is apparent. (**C**) and (**D**) Front and side views demonstrating varus, internal rotation, and flexion deformities. (**E**) Early postoperative photograph before the strut adjustment period. Internal rotation deformity is appreciated with the patella facing forward. (**F**) (See color insert.) The deformity data is entered into the web-based computer program and an image is generated recreating the deformity. (With reference to the stick diagrams, the blue box represents the knee and the green circle the foot.) (**G**) and (**H**) Anteroposterior and lateral computer animation images, respectively, illustrate the deformity with frame in place. (**I**) The prescription indicates the appropriate strut settings for each day during the adjustment period. (**J**) and (**K**) Three months after frame removal the valgus and sagittal alignment is maintained. (**L**) and (**M**) Full correction of varus and flexion deformities. (**N**) Symmetric thigh foot axis indicates correction of the internal rotation deformity.

In patients with knee flexion contracture and flexion deformity of the proximal tibia, the addition of extension at the osteotomy will correct this deformity allowing for full extension. Anterior Cruciate Ligament deficiency associated with medial gonarthritis leads to anterior tibial subluxation. By increasing the posterior proximal tibial angle (18) from 81° to 90°, the instability can be decreased (19). In patients found to have internal or external tibial torsion deformities, these axial malalignments can also be corrected simultaneously through the same osteotomy site further expanding the indication for TSF. Patients with laxity of the posterolateral ligaments will benefit from use of the TSF. While closing wedge osteotomy increases the relative length of the lateral ligaments, a lateral opening wedge osteotomy, that would be used to correct a valgus deformity, affords us the ability to retension these lateral ligaments. This is carried out by lengthening the tibia without performing a fibular osteotomy or securing the proximal tibiofibular joint. The fibula will migrate distally tightening the lateral ligamentous structures. If correcting a varus deformity without lengthening, then a distal transport of the proximal fibula after osteotomy of the middle fibula is needed.

A two-ring TSF is adequate for most moderate deformity corrections (Table 1). We add a third ring just above the ankle with a tibio/fibula syndesmosis wire in the following situations: need for greater than 1 cm of lengthening, coronal/sagittal plane angular deformity greater than 20°, or rotational (axial plane) deformity greater than 15°. This is to achieve greater stability, a longer lever arm for the correction, and to protect the distal tibia-fibula relationship at the ankle.

Patellofemoral compartment arthritis has been considered a relative contraindication for High Tibial Osteotomy (HTO). Although many patients with medial gonarthritis experience anterior knee discomfort related to patellofemoral chondromalacia, many authors note that the presence of Outerbridge grade III to IV changes of the patellofemoral articular surfaces has not affected the final outcome after HTO (1,12,20).

The presence of lateral compartment disease has classically been taught to jeopardize the results after HTO. Miller and Sterett have observed that gradual correction to neutral alignment of the varus arthritic knee containing small areas of grade IV chondromalacia laterally has yielded reliably good results (12).

Figure 1 (*Continued from previous page*).

CLINICAL ASSESSMENT AND PREOPERATIVE PLANNING

All patients are clinically evaluated by history and physical examination. Special attention is directed toward the assessment of leg length, knee ligamentous stability, and rotational alignment as correction of these parameters using the TSF may be added to the varus realignment. Gait is carefully evaluated to assess lower extremity function looking for abnormal kinematics. An erect anteroposterior (AP) 51-in radiograph of both lower extremities on one cassette, standing AP, lateral and merchant views of the knee are obtained. Mechanical axis deviation is determined using the malalignment test (21). Values for the lateral distal femoral angle, medial proximal tibial angle, and joint line convergence angles are measured to localize sources of deformity. Sagittal deformity is evaluated on the lateral radiographs. The Center of rotation of angulation (CORA) is located and the magnitude of deformity measured (21). The CORA is typically situated at or near the joint line. The postoperative goal is to unload the medial compartment. This is reliably achieved by realigning the distal tibia such that the mechanical axis passes through the lateral compartment. Fujisawa et al. described an ideal target point for the mechanical axis—one-third of the distance from the knee center across the lateral plateau (22). Jakob and Murphy modified the position of this point based on the degree of medial articular cartilage damage. The medial third of the lateral tibial plateau is itself divided into thirds. If the extent of degeneration of the medial compartment is minimal, then the goal will be correction

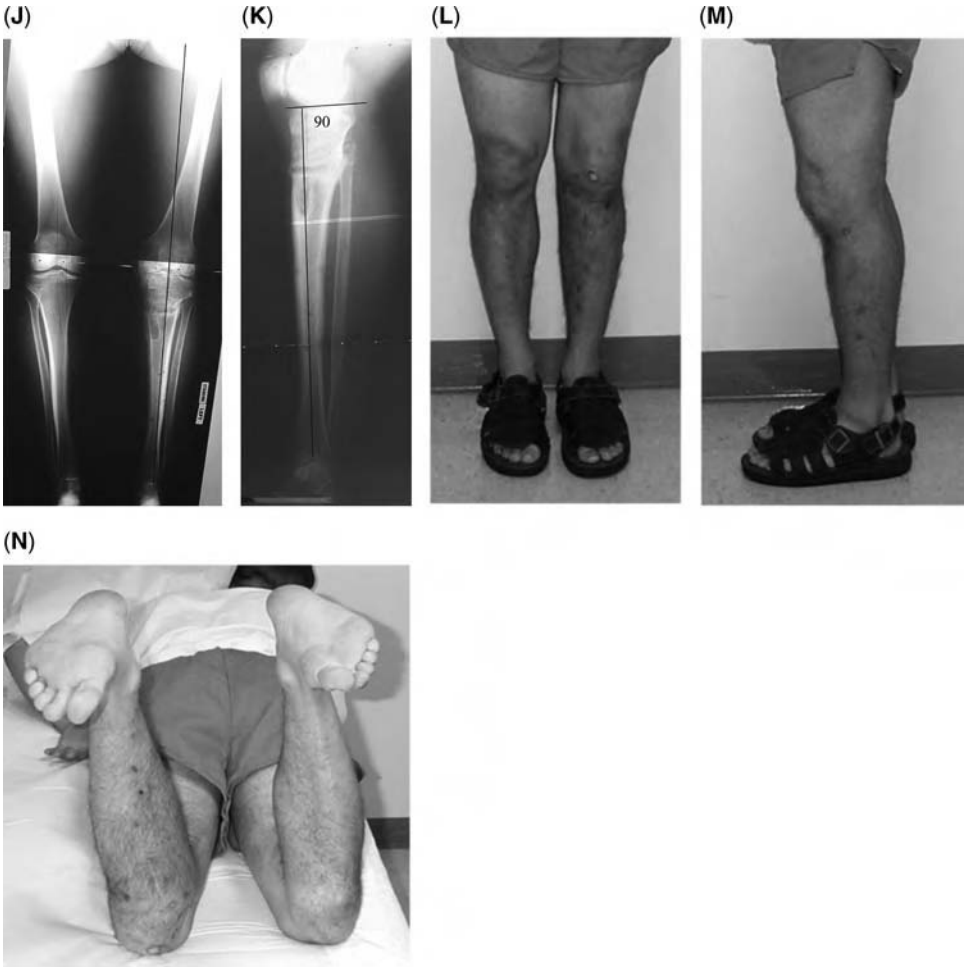

Figure 1 (*Continued from previous page*).

of the mechanical axis through the medial third (one-third of the distance to Fujisawa's point). If the medial compartment damage is more advanced, then the goal will be overcorrection of the mechanical axis through the middle third. With severe arthrosis of the medial compartment, the mechanical axis is shifted to pass through Fujisawa's point (23). We use Fujisawa's point with Jakob's modifications to guide final alignment. To complete the planning, a new mechanical axis line is drawn through the desired point in the lateral compartment and the magnitude of deformity is remeasured in the frontal plane. These values will be entered into the computer to assure that the distal fragment will be well aligned at the completion of the adjustment period.

We often recommend a simple knee arthroscopy prior to osteotomy or in conjunction with the osteotomy. The arthroscopic procedure serves as an excellent tool for evaluating the integrity of the lateral and patellofemoral compartments as well as providing an additional opportunity to treat mechanical symptoms originating from the medial compartment. The extent of degenerative change in the medial compartment encountered at arthroscopy may provide additional information with regard to the patient prognosis. Arthroscopy also provides an opportunity to perform abrasion arthroplasty of the degenerated articular surfaces. It has been observed that eburnated and abraded surfaces in combination with proximal tibial realignment form new cartilage more readily than those covered by fibrillated cartilage (24–26).

Osteotomy of the fibula is required in many patients undergoing proximal tibial osteotomy with use of the TSF. This is primarily because those patients for whom the TSF is indicated undergo large corrections or corrections of multiple deformities that rely on a

mobile fibula. If the lateral ligaments require tensioning, then osteotomy and distal transport of the fibula is performed.

TECHNIQUE

The patient is taken to the operating room and placed supine on a radiolucent operating table. Sheets are placed under the ispilateral pelvis to internally rotate the lower extremity until the patella is pointing directly toward the ceiling. At our center, regional epidural anesthesia is typically used to provide analgesia for the surgery. A dose of prophylactic antibiotics with gram-positive coverage is given in the operating room prior to skin incision. C-arm fluoroscopy is used throughout the procedure to assure ideal positioning of the fixator and allow for the implementation of minimally invasive techniques. The C-arm is positioned on the side of the contralateral leg. If a lengthening, rotational correction, flexion correction, or significant translation of the distal fragment is planned, then a fibular osteotomy is necessary. This is carried out under tourniquet and is performed at the mid-diaphyseal level. When varus is being corrected, an oblique osteotomy is made allowing the fibula to shorten as the cut bone ends slide past one another. If rotation is being corrected, then a transverse fibular osteotomy with a minor resection can be performed allowing the bone ends to translate and shorten. A direct approach is made to the fibula through the interval between the peroneal muscles and the soleus. Care is taken when performing the subperiosteal dissection as the motor branch to the Extensor Hallucis Longus lies close to the anteromedial border of the fibula. The soft tissue is protected with Hohman retractors while the fibular diaphysis cut with an oscillating saw. Frequent pauses are made during sawing and liberal saline irrigation is used to diminish heat production. An osteotome is used to ensure that the medial cortex of the fibula is cut (Fig. 2). The fascia is left open and the skin is closed in layers. The tourniquet is then deflated for the remainder of the operation.

The technique used to apply the TSF is the ''rings first'' method as opposed to using a preconstructed frame. We favor this technique because it frees the rings for ideal placement on the leg with regard to the soft tissues. The tourniquet is not recommended for this portion of the surgery as it is felt that adequate blood flow is needed to cool wires and drills as they pass through the tibia. A review of the anatomic safe zones for wire passage through the tibia should be performed prior to surgery. Using the fluoroscopic AP projection, a smooth 1.8 mm Ilizarov wire is advanced across the proximal tibial metaphysis from lateral to medial parallel to the joint line. This wire will assure proper positioning of the proximal ring. The wire should start 14 mm distal to the lateral tibial plateau in order to remain out of the joint capsule (14,27). Once this wire has been placed, the proximal ring is centered on the leg and positioned parallel to the joint surface (Fig. 3). The wire is tensioned to 130 kg in the standard fashion (Fig. 4). The anterior aspect of the ring should lie between one to two finger breaths from the tibial crest. We prefer to use a two-third ring proximally to accommodate posterior leg swelling and allow knee flexion. Having set the ring in the AP plane, attention is turned to the lateral

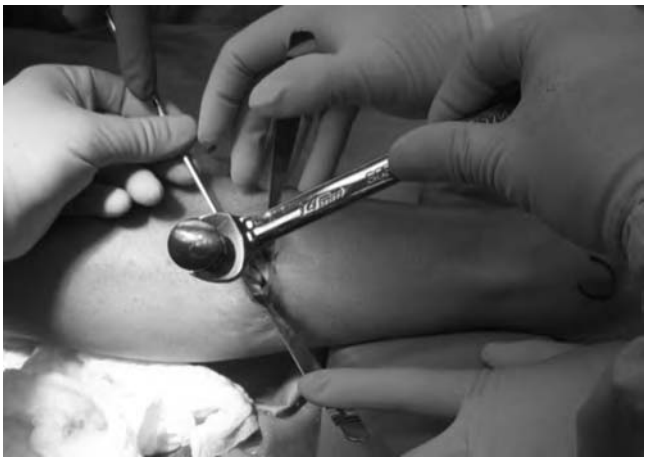

Figure 2 Fibular osteotomy is completed by rotating the osteotomy with a 14-mm wrench.

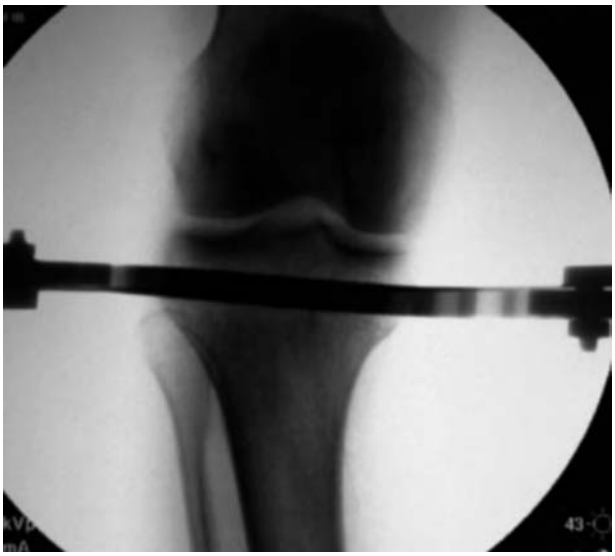

Figure 3 The proximal ring should be mounted parallel to the joint surface on the anteroposterior projection.

projection. The ring is held in a position orthogonal to the proximal tibia in the sagittal plane as seen on lateral fluoroscopy. A second wire is placed through the fibular head exiting the anteromedial tibia. When placing wires through the fibular head great care is taken to avoid damaging the peroneal nerve. The surgeon places the wire by hand onto the fibular head while the foot is observed for signs of movement indicating nerve irritation. The wire is then advanced and fixed to the ring. If the lateral ligaments are to be tensioned, then the fibular head is not captured with this second wire. A cortical threaded half pin is placed from anteromedial to posterolateral. We advocate the use of hydroxyapatite coated half pins when performing this procedure on patients with osteopenia. These pins have been associated with a decreased incidence of loosening, and subsequently, a lower rate of pin site infection (28). The pin is secured to the ring and an additional half pin is placed anterolateral at Gerdy's tubercle to posteromedial (Fig. 5).

Once the proximal ring is secured, the mounting parameters are addressed. The mounting parameters are a set of measurements that inform the computer of the location of the reference ring with respect to the virtual hinge referred to as the origin. The computer will need to know if the reference ring is anterior/posterior to the origin, medial/lateral to the origin, and proximal/distal to the origin and these exact distances in millimeters. For a proximal tibial osteotomy, the proximal ring is used as the reference ring. The distance from the center of this ring to the origin in the coronal, sagital, and axial planes is measured. The origin can be positioned at the CORA of the deformity or at the center of the osteotomy site. If the center of the osteotomy site is used as the origin, then one must typically add lateral translation to the

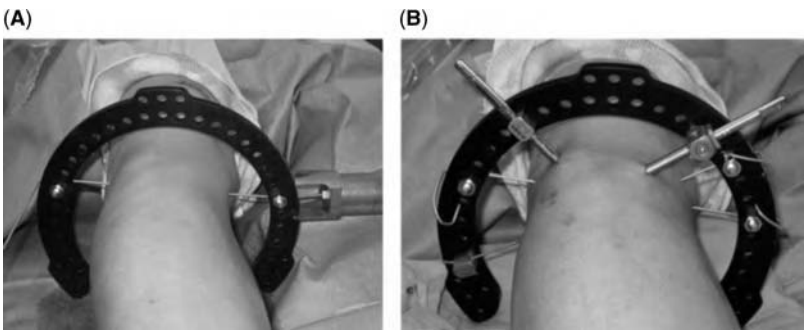

Figure 4 (**A**) Reference wire is tensioned to the proximal ring establishing frontal alignment. (**B**) Typical configuration of proximal ring fixation block with two wires and two half pins.

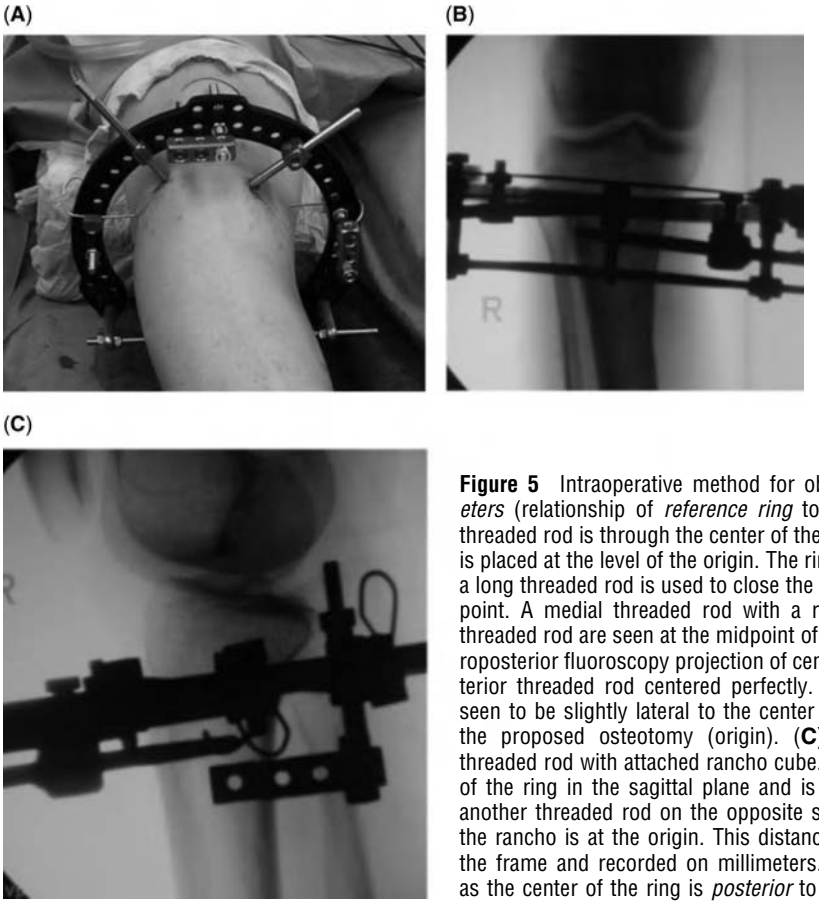

Figure 5 Intraoperative method for obtaining *mounting parameters* (relationship of *reference ring* to the *origin*). (**A**) Anterior threaded rod is through the center of the ring, and the rancho cube is placed at the level of the origin. The ring is open posteriorly, and a long threaded rod is used to close the ring and establish a center point. A medial threaded rod with a rancho cube and a lateral threaded rod are seen at the midpoint of the ring as well. (**B**) Anteroposterior fluoroscopy projection of central threaded rod and posterior threaded rod centered perfectly. The center of the ring is seen to be slightly lateral to the center of the tibia at the level of the proposed osteotomy (origin). (**C**) Lateral image showing threaded rod with attached rancho cube. The rod marks the center of the ring in the sagittal plane and is perfectly overlapped with another threaded rod on the opposite side of the ring. The tip of the rancho is at the origin. This distance is measured directly on the frame and recorded on millimeters. The data will be entered as the center of the ring is *posterior* to the origin.

deformity parameters to ensure restoration of the mechanical axis at the end of the correction. To determine mounting parameters, the center of the ring and the center of the proposed osteotomy site must be localized on the same C-arm image. Closing the proximal two-thirds ring with a threaded rod, and centering a nut on that rod, the AP parameters are established first. This will define the center of the posterior part of the ring. Placing a threaded rod in the middle hole of the anterior master tab, the corresponding center of the anterior aspect of the ring is marked (Fig. 5A). A three-holed rancho cube is placed on the threaded rod at the level of the proposed osteotomy. The C-arm is centered on the ring in the AP position until the ring is seen as a single line. The leg is then rotated under live image intensification until the anterior threaded rod overlaps the nut on the posterior rod (Fig. 5B). This generates a picture well centered on the ring. The distance between the rancho holes and the center of the bone is recorded and a direct measurement is taken on the frame with a ruler. The center of the frame is usually lateral and proximal to the center of the bone. The lateral mounting parameters are taken in a similar fashion. A threaded rod is placed on the medial side of the ring in the center most hole. A second rod is placed on the lateral side in the corresponding hole. A three-holed rancho cube is secured onto the rod at the level of the proposed osteotomy. The C-arm is moved into a lateral position and aligned on the ring making the ring appear as one line. The leg is then rotated under live image until the medial and lateral rods are aligned (Fig. 5C). The distance from the threaded rod line to the center of the bone is recorded using the rancho cube holes as a reference. The actual distance is measured on the frame with a ruler. The center of the frame should be posterior to the origin. These values are recorded for later use in the creation of a schedule for strut adjustments.

Attention is then turned to the distal ring. Some thought should be given to determining the optimal distance between the rings. This will help minimize strut changes that are an inconvenience to the patient and the surgeon. Typically medium struts are used, and they

are set in the middle length position (145 mm). A medial face wire is advanced from lateral to medial across the tibia orthogonal to the long axis of the tibia. Care must be taken not to generate heat while advancing the wire through this cortical bone. Frequent pauses are prudent, and a wet sponge can be used to cool and guide the wire during its insertion. The distal ring is centered on the leg and fastened to the wire. The wire is tensioned to 130 kg. A strut can be used to hold the ring orthogonal in the sagittal plane. Once the ring position is set on the lateral view, two additional half pins are inserted proximal and distal to the distal ring, preferably out of plane, yielding a total of three points of fixation distally (Fig. 6). The six struts are attached to the proximal ring and tightened loosely. The struts are secured to the distal ring without introducing any tension or compression forces to the system. Free rotation of the struts should be possible as the shoulder bolts spin through the ring. The strut lengths are recorded, and all of the struts are detached from the proximal ring to carry out the tibial osteotomy.

The percutaneous tibial osteotomy should be made distal to the tibial tubercle to prevent involvement of the extensor mechanism, but it should be proximal enough that it courses through cancellous metaphyseal bone to ensure reliable regenerate formation. A 1-cm incision is made over the tibial crest just distal to the tibial tubercle. The incision is carried down through the periosteum and onto the crest. A 5-mm elevator is used to gently raise a portion of the periosteum on either side of the tibia. The cortex is predrilled in multiple directions along the same transverse plane with a 4.8-mm drill. A 5-mm osteotome is advanced through the cortical bone of the tibia's medial and lateral faces (Fig. 7). When the osteotome is fully seated through the width of the bone and is engaging the posterior cortex, it is twisted with a 14-mm wrench producing an audible crack as the posterior cortex fails. The distal ring is gently externally rotated with respect to the proximal ring to ensure that the corticotomy is complete (Fig. 8). The bone ends are reduced to their pre-osteotomy position relieving stress on the periosteum and decreasing bleeding (Fig. 9). The struts are reattached to the rings at their previously measured lengths stabilizing the osteotomy site (Fig. 10A and B). The wound is closed with simple sutures, and the pin sites are dressed with xeroform and sterile dressings. An ACE bandage is used to support the forefoot in a neutral position. The epidural is discontinued in the immediate postoperative period to avoid masking early signs of compartment syndrome. A latency phase of 10 days is used prior to the start of frame adjustments.

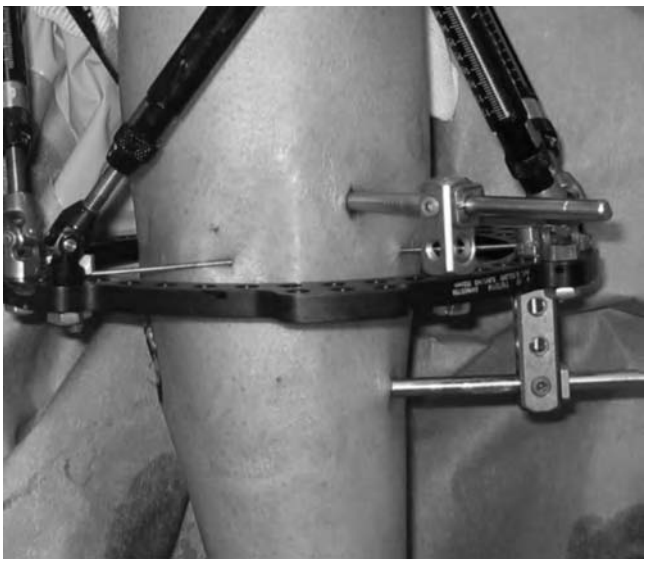

Figure 6 Typical set up of distal ring with one wire and two half pins.

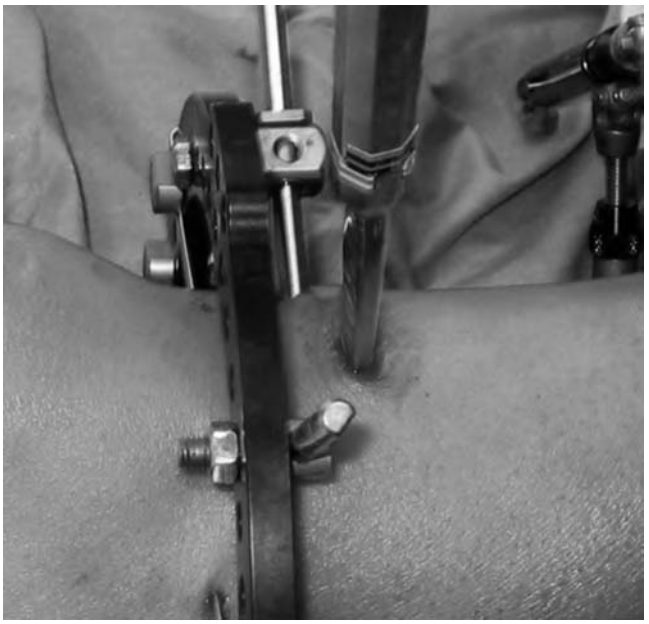

Figure 7 The osteotome is advanced through the small skin incision. The osteotome is twisted with the 14 mm wrench 90° and then rotated back to its original position preventing displacement.

RESULTS

Many authors have used distraction osteogenesis in the context of proximal tibial osteotomy to obtain gradual correction of varus alignment. In a prospective randomized trial comparing closed-wedge high tibial osteotomy with open-wedge osteotomy by hemicallotasis, Magyar noted that undercorrection and overcorrection were relatively common complications of the Coventry type osteotomy. This lack of accuracy led to a broad array of final alignments in the closing wedge group. This was not observed in his opening wedge, hemicallotasis patients whose final alignment was accurately controlled. Furthermore, he reported that rate of loss of correction over the first postoperative year was much higher in the closing wedge osteotomy group (29).

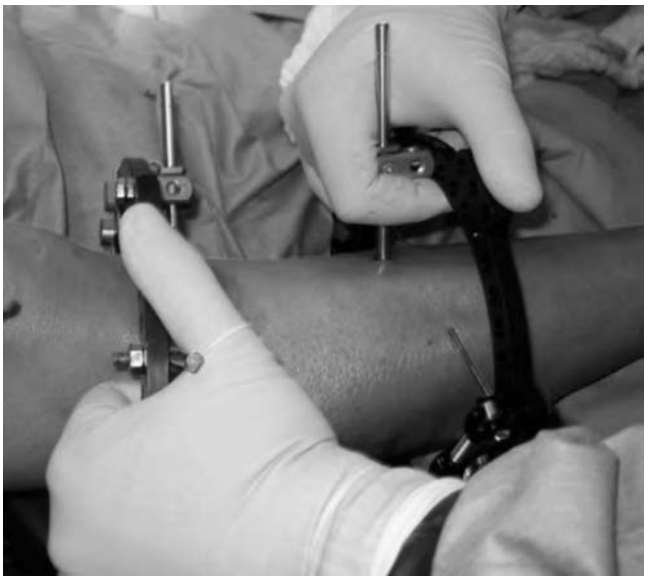

Figure 8 Rotational osteoclasis ensures a complete osteotomy. External rotation is thought to prevent tension on the peroneal nerve.

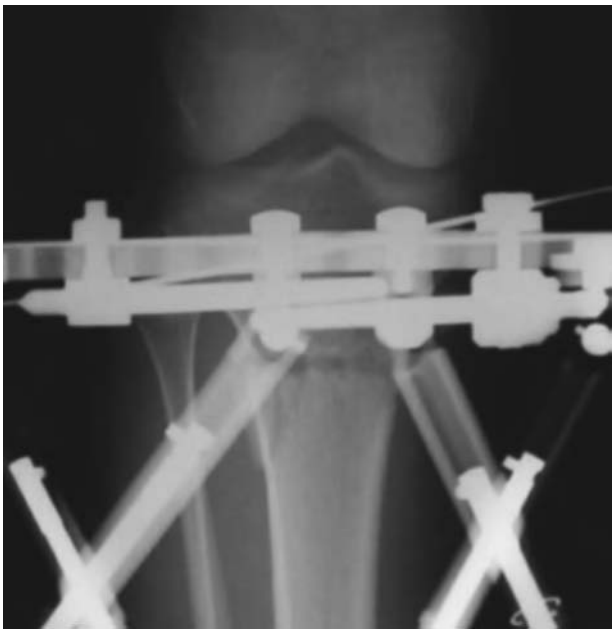

Figure 9 Anteroposterior radiograph demonstrating typical transverse osteotomy in a reduced position.

Nakamura et al. compared 50 knees undergoing proximal tibial osteotomy using either a dome osteotomy technique or a hemicallotasis method. The dome osteotomy was performed above the level of the tibial tubercle using acute correction and compression clamp fixation. In the hemicallotasis group, the osteotomy was performed below the tubercle and correction was achieved gradually with external fixation. Significant postoperative shortening of the patellar tendon and decrease in the sagittal angle of inclination of the tibial plateau was observed in the dome osteotomy group, but not in the hemicallotasis group (5).

Sen et al. looked at 53 patients undergoing high tibial osteotomy fixed with either internal fixation or external circular fixation. They found that those patients stabilized with external fixation demonstrated better results in terms of Hospital for Special Surgery score, alignment of the lower extremity, and preventing the progression of arthritis (14).

In our clinical experience, proximal tibial osteotomy performed in conjunction with the TSF yielded good results in the treatment of pain and dysfunction associated with genu varum. Thirty-four patients with medial mechanical axis deviation and medial compartment degeneration were treated with this technique (unpublished data). Indications for this technique included genu varum greater than 8° and associated deformities including rotational malalignment, shortening, flexion contracture, knee hyperextension, and ligamentous laxity

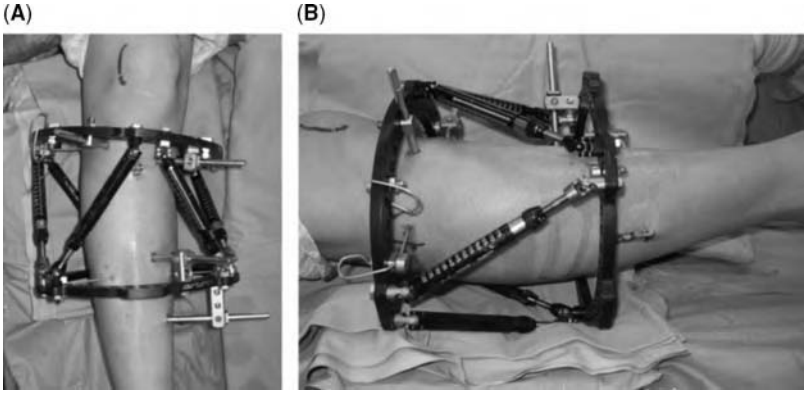

Figure 10 (**A**) Frontal and (**B**) lateral views of final frame position at the completion of surgery.

or instability. In all patients, ideal alignment was restored. short form-36 scores improved an average of 16 points. There was a trend toward less improvement in patients with more severe arthrosis. In those patients with knee instability, there was subjective improvement in all cases. In our experience, we have noticed that the treatment was well tolerated and all patients were satisfied with their results.

COMPLICATIONS

Pin Infection

Pin site infection is a common complication that we encounter when using external fixation. Pin infections are marked by erythema, increasing pain, and drainage around the pin or wire. The vast majority of these respond well to more aggressive local pin care and oral antibiotics. If the infection does not resolve quickly, then broader spectrum antibiotics are added or the pin or wire is removed. More advanced infections are treated with removal of the pin or wire and local bone debridement in the operating room, and intravenous antibiotics as needed. Loose pins and wires are removed and the pin sites debrided even in the absence of infection. Although the possibility exists that chronic osteomyelitis from an infected pin tract could lead to a future infected total joint arthroplasty this complication has not been reported in the literature.

Premature Consolidation

Incomplete corticotomy can complicate proximal tibial osteotomy. A circumferential division of the tibial cortex may be assured by rotating the proximal and distal rings in opposite directions and witnessing uninhibited motion through the corticotomy site. Other methods have been described including acute distraction and angulation at the osteotomy site, but these techniques are more disruptive to the periosteum and not recommended. True premature consolidation of the osteotomy is rare in the adult patient. Once the osteotomy is performed there is a latency period of 7 to 10 days before any correction is attempted. If the latency period is prolonged, then the osteotomy site will consolidate prematurely. Similarly if the correction is carried out too slowly, the osteotomy site may heal preventing further correction.

Patient-Related

The success of any gradual correction system is dependent on the patients' ability to participate in their own care. Patients are responsible for performing their own strut adjustment three times daily at the outset of treatment. The TSF has simplified this process through color coordination and a precise numbering system. Even so, patients have made strut adjustment errors. These mistakes are usually quickly acknowledged and remedied. Patients need to be seen frequently during the adjustment period to avoid errors.

Nonunion

Osseous nonunion can complicate any osteotomy procedure. Causes may include inadequate fixation, lack of weight bearing, smoking and other causes of poor blood flow to the extremity, patient comorbidities, too rapid a correction, poor osteotomy technique, and an osteotomy through diaphyseal bone. Nonunions are treated aggressively with a variety of methods including compression across the osteotomy site, percutaneous periosteal and endosteal stimulation, and additional points of fixation. Nonunions are rare when using the TSF technique. When there is impaired healing, this specialized frame provides ideal circumstances for effective treatment. We have not had any nonunions using this method, but we have successfully treated many referred proximal tibial nonunions with the Ilizarov method.

Deep Vein Thrombosis

Deep vein thrombosis (DVT) is always a concern with surgery of the lower extremity. Treatment is aimed at prevention. Patients are launched into early rehabilitation programs emphasizing immediate mobility to avoid venous stasis. There is no restriction to movement at the ankle, knee, or hip, and frame stability allows comfortable weight bearing early in the postoperative period. While in the hospital, the patients receive subcutaneous low molecular weight heparin. Upon discharge, patients start a one-month course of aspirin despite concerns about its effects on bone healing. With this regimen, we have not had any cases of DVT or pulmonary embolism.

POSTOPERATIVE MANAGEMENT

General

Patients are admitted to the hospital for two to three days. Nonsteroidal anti-inflammatory medications are avoided in all osteotomy patients for fear of adverse affects on bone formation. The patients receive intravenous antibiotics for 24 hours and are then switched to oral antibiotics. The patients are discharged on oral antibiotics for 10 days and oral pain medication. Patients return to the office 10 days postoperatively where sutures are removed and they are educated on how to perform strut adjustments. Patients are seen every two weeks during this adjustment period, and then once monthly during the consolidation period.

Deformity Correction

Correction of the deformity begins after a latency period of 7 to 10 days. The TSF web-based program is used to generate a daily schedule for strut adjustments that the patient will perform at home. The computer requires the input of basic information including the side, the deformity parameters, the size of the rings and length of struts used, the mounting parameters measured during frame application, and rate of daily adjustment. Additionally, a structure at risk is selected and entered into the program to assure gradual correction. For valgus producing osteotomy, the structures a risk are the medial soft tissues as they are in the concavity of the correction and will be stretched the greatest distance. Using this information, a clear and simplified prescription is created for the patient to be followed every day (Fig. 1H). We prescribe that struts 1 and 2 be turned in the morning, struts 3 and 4 in the afternoon, and struts 5 and 6 in the evening for a total movement of 1 mm per day. The duration of the adjustment phase depends on the amount of correction needed and is typically between 14 and 28 days. The length of time in the frame is approximately three months.

Pain Management

Transdermal wires and pins can be irritating, and we encourage patients to use appropriate oral pain medications. This is especially true during the adjustment period. Once the correction is complete, the frame is no longer moving, and the pain level decreases. Severe or atypical pain merits an evaluation for infection or DVT.

Pin Care

The dressings are removed on postoperative day two. Nurses teach proper daily pin care consisting of a mixture of half normal saline and half hydrogen peroxide applied to the pin sites with sterile cotton swabs. Pins and wires are covered with Xeroform dressings at the skin (Fig. 11). Patients are allowed to begin showering on the fourth postoperative day. They are instructed to wash the frame and pin sites with antibacterial soap as an adjuvant form of pin care. Problematic smooth wires can be removed in the office without anesthetic. This is particularly done after the distraction phase or if a wire is painful and infected.

Rehabilitation

Ilizarov stressed the importance of early physical conditioning in conjunction with the application of circular fixators. Early motion increases blood flow to the lower extremity, prevents joint stiffness, and shortens recovery time (11). Physical therapy assists with weight bearing as tolerated ambulation and range of motion exercises for the knee and ankle joints. Crutches are normally needed for the first four to six weeks after surgery. Occupational therapy provides a custom-neutral foot-splint to prevent equinus posturing during sleep. Patients are encouraged to attend outpatient physical therapy where they continue with their rehabilitation programs.

Frame Removal

Fixators are removed when patients are ambulating without pain or have started using an assistive device, and when callus is seen on three corticies around the osteotomy site. This is typically three months after the index surgery. We prefer to remove the frames in the operating room. The removal of hydroxyapatite coated pin can be painful and is best done under

Figure 11 Xeroform dressings act as a barrier to bacterial access into the pin sites and keep the skin soft reducing symptoms of pin irritation.

sedation. We choose to curette and irrigated all half pins sites in an effort to keep pin tracts clean for possible later arthroplasty. Transfixtion wire sites are not debrided unless there is concern over a specific site. At the time of frame removal, bony union and maturation of the regenerate may be evaluated with a stress test under C-arm fluoroscopy. The struts are removed and the rings manually compressed and distracted looking for motion at the osteotomy site. A lack of consolidation will require replacement of the struts and prolonging the time in the frame. Once the fixator is removed, the patients are placed into a hinged knee brace with 0° to 90° of motion. They are allowed 50% partial weight bearing for two weeks then progress to full weight bearing thereafter.

POSSIBLE CONCERNS AND FUTURE OF TECHNIQUE

The addition of cartilage sparing and regenerating procedures to the proximal tibial osteotomy technique may prove to increase the longevity of the procedure. Many authors advocate the routine use of knee arthroscopy with abrasion chondroplasty, microfracture, or subchondral drilling at the time of HTO (24,30). These procedures are easily performed during arthroscopy and add minimal cost to the surgery. Meniscal transplantation and autogenous chondrocyte implantation are expensive techniques that may prolong the integrity of the knee articular surfaces when combined with tibial realignment osteotomy. Further studies need to be undertaken to determine the efficacy of these combined procedures, guidelines for candidacy, and the cost-benefit relation.

In only a few short years, the TSF computer programming technology has improved dramatically. New software for calculating deformity and mounting parameters using digital radiographs is under development. This could simplify the process even further. The role of computer aided navigation in tibial osteotomy using the TSF has yet to be realized. Navigation may have a profound impact on the accuracy of deformity correction. This may be most useful in the setting of acute correction, where the accuracy of bone cuts is essential. There could be advantages of its use intraoperatively when planning gradual corrections as well. Navigation is also expected to decrease fluoroscopy usage and operative time.

SUMMARY

Proximal tibial osteotomy using the Ilizarov method and the TSF has several advantages over the traditional techniques. It is minimally invasive and there is no internal hardware. There is no need for acute deformity correction that should be safer in terms of neurovascular complications. Postoperative adjustability allows a very precise correction. Complex deformity and large deformity can be managed as well as simple deformity correction. The TSF in particular is helpful for simultaneous correction of rotation, angulation, and translation in all planes. Although the standard osteotomy is below the tubercle, this can be modified in order to tension the medical collateral ligament and lateral collateral ligament as needed (31,32). The circular frame allows patients to weight bear as tolerated, having the advantage of diminishing disuse osteopenia, muscle atrophy, and allowing the patient to stand for a bipedal radiograph. Precise analysis of the correction can be done and additional adjustment may be recommended as needed.

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24 Arthrodesis of the Knee

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INTRODUCTION

Knee arthrodesis has been performed since the early 1900s for a variety of indications, most commonly advanced osteoarthritis and infectious arthritis (1–10). Today, however, the most common indications for knee arthrodesis are trauma where the joint is no longer salvageable or following total knee arthroplasty (TKA) infections. Charnley's results with knee arthrodesis for osteoarthritis are impressive (11). The majority of patients returned to work within 12 weeks without a brace. Today, knee arthrodesis is considered a procedure of ''last resort'' or ''better than an amputation.'' This thinking, along with the many revision total knee options and tumor prostheses available, has made successful knee arthrodesis quite a challenge. The typical patient presenting for a knee arthrodesis has experienced as many as three revisions and numerous surgical procedures to preserve knee motion. The patients are typically over 70 years of age with multiple medical problems including osteoporosis. These patients also have a loss of soft tissue anteriorly, as well as no extensor mechanism, with exposed bone and significant bone loss (>4 cm). Although Charnley's study found that patients recovered within 12 weeks, complications such as loss of soft tissue and infection will significantly extend the time to bone union well beyond 12 weeks. With the aging population and the popularity of TKA, the number of patients requiring a knee arthrodesis certainly will increase. The remainder of this chapter will focus upon the clinical evaluation, classification, treatment options, surgical techniques, and complications of knee arthrodesis.

CLINICAL EVALUATION

The initial clinical evaluation includes a thorough history of the patient's medical problems. Obesity, diabetes, and smoking increase the risk of wound complications and ideally need to be managed with a weight-loss program, a proper diabetic regimen, and a smoking cessation program (12). A critical part of the clinical evaluation is the patient's mental condition. The patient must be willing to accept a knee arthrodesis and cooperate with the treatment plan. Some older patients are not mentally capable of handling an external fixator with the associated pin care and adjustments. A successful outcome is more likely with complete patient cooperation.

All previous incisions are noted at a physical examination. The surgical incision must be planned in relation to other incisions in order to avoid wound necrosis (Fig. 1). If wound complications are anticipated, a preoperative plastic surgical consult is obtained. Special flap techniques or tissue expanders can be used for prevention of soft tissue complications (13). A good vascular supply to the leg is crucial. If the peripheral pulse is diminished, then a vascular consult should be obtained to maximize the blood flow to the leg. Also, acute compression intraoperatively can cause additional vascular compromise secondary to extensive scarring; therefore, a thorough preoperative evaluation is needed as a baseline. Any sign of active infection such, as significant erythema, cellulitis, or draining sinuses, is important for surgical planning. Active infection affects the surgical options for fixation and several authors recommend eradication of infection prior to insertion of an intramedullary nail (14–21).

Preoperative radiographs, including teleroentgenogram, are helpful in determining the current limb-length discrepancy (LLD) as well as preoperative sizing of intramedullary devices (Fig. 2). All patients following knee arthrodesis will have LLDs with the average

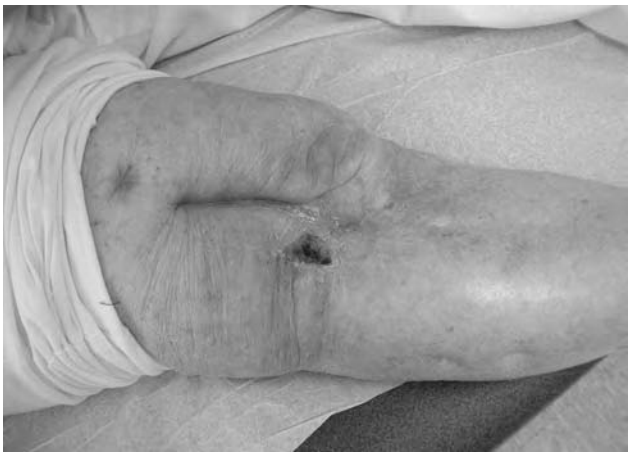

Figure 1 Soft tissue inspection on preoperative physical examination of the knee reveals an open sinus and an inverted anterior incision. There is also a muscle flap covered with a split-thickness skin graft.

ranging from 2.5 to 6.4 cm (22–27). If the LLD following the arthrodesis is anticipated to be greater than 5 cm, then options for limb lengthening simultaneously (28) or following the arthrodesis are presented to the patient in the preoperative discussion.

CLASSIFICATIONS

This classification scheme is based upon the patient's LLD, soft tissue envelope, bony integrity, and patient capabilities. It is summarized in Table 1.

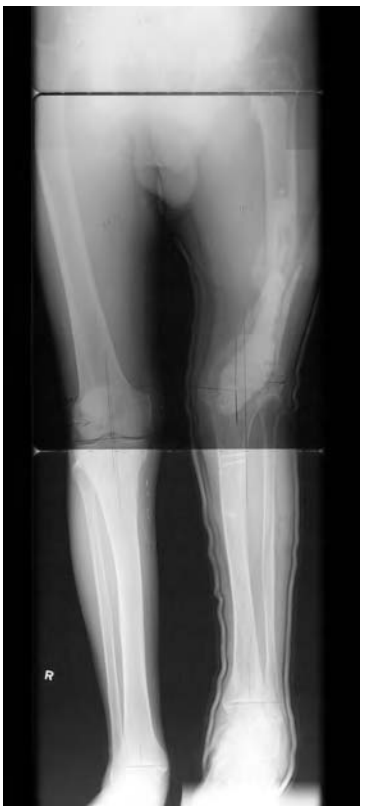

Figure 2 Teloroentenogram demonstrating the leg length discrepancy as a result of fracture during infected total knee arthroplasty component removal.

Table 1 Preoperative Classification for Knee Arthrodesis

Problem	Treatment
Greater than 5-cm lengthenings with noncompressible gap	Long IM rod (antibiotic-coated if infected) with femoral bone transport // staged bone grafting
Greater than 5-cm lengthenings with compressible gap	Long intramedullary (IM) rod (antibiotic-coated if infected with infection eradication) with acute compression of the arthrodesis followed by lengthening over nail (simultaneously or later)/bone grafting
Actively infected, <5-cm limb-length discrepancy (LLD), nonobese, mentally capable of a fixator	Infection eradication and biplanar external fixator/staged bone grafting
Actively infected, <5-cm LLD, obese, not mentally capable of fixator	Infection eradication/long IM rod and bone grafting
Anterior cortical defects: femur and tibia	Infection eradication/long IM rod and bone grafting
Failed knee arthrodesis	External fixation (+infection eradication)/bone grafting

TREATMENT OPTIONS

Outlined below is the rationale for each category.

Noncompressible gap with greater than 5 cm bone defect. These patients often experience complications such as severe bone loss and soft tissue compromise. In these cases, a noncompressible gap is defined by the inability to achieve acute bone approximation secondary to vascular compromise or severe, thick, soft tissue scarring that is not compliant enough for compression. Also, with compression, unless the incision is transverse, a significant amount of shortening will not allow a longitudinal wound to close. For significant soft tissue defects, femoral bone transport can also be used to close the soft tissue defect (Fig. 3). The first surgery is for eradication of infection followed by a second surgery approximately six weeks later to insert the intramedullary rod, perform an osteotomy for bone transport, and apply an external

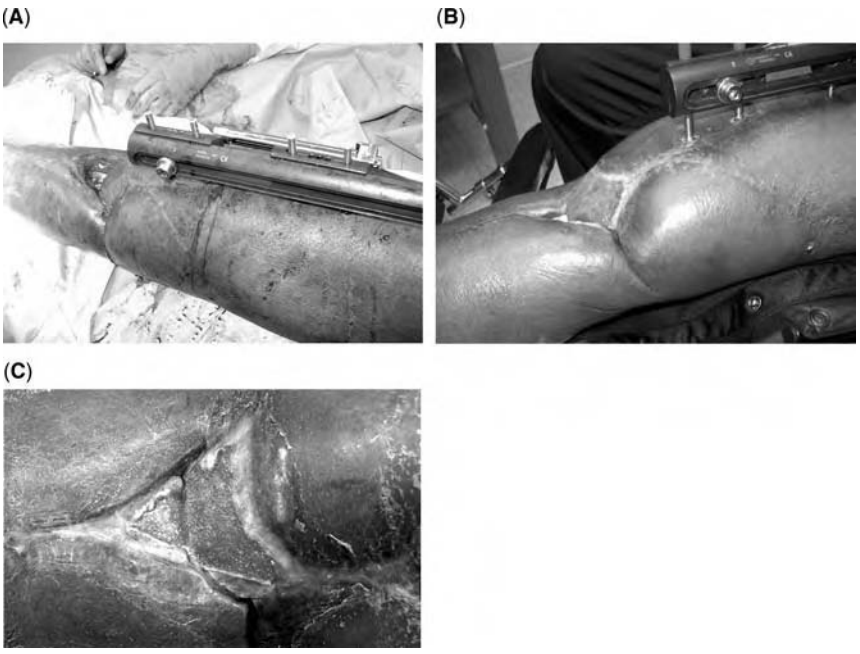

Figure 3 Patient with a noncompressible knee arthrodesis gap secondary to an arterial injury resulting in a vascular bypass graft. This was treated with femoral bone transport over a rod. Bone transport is also an effective way to transport skin and overlying soft tissue as well. (**A**) Postoperatively; (**B**) during transport; and (**C**) at the completion of transport with good soft-tissue coverage.

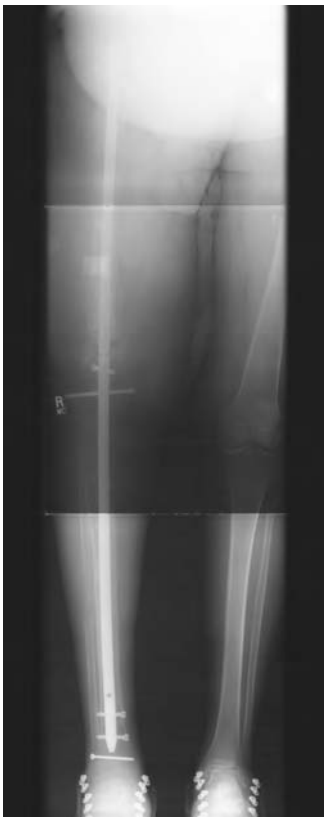

Figure 4 Teloroentenogram demonstrating a custom intramedullary knee arthrodesis rod with interlocking screws in the distal femoral and proximal tibial transported segments.

fixator. If the gap is extremely large (10 cm), the best way to proceed is to use a double-level transport (proximal tibia and distal femur) over a rod. The advantage of the rod is minimal angulation of the regenerate bone formation and, when the gap is eliminated, a large fragment percutaneous locking plate is used to fuse the knee with the addition of a bone graft. The other option for fixation is a custom rod with the locking holes drilled for the transport segment prior to rod insertion. Only touchdown weight bearing is allowed until the regenerate is consolidated (Fig. 4).

Compressible gap with greater than 5 cm LLD. Any patient who has greater than 5 cm LLD is offered the opportunity to correct LLD with the knee arthrodesis. If they wish to consider it as an option, then an intramedullary rod is chosen for the arthrodesis. With lengthening over a nail (LON), the inconvenience of an external fixator is minimized, as well as any resultant deformity from the lengthening. Often, a better quality regenerate forms with LON. The regions available for osteotomies in these cases are the proximal femur and the proximal tibia. Any device other than a monolateral fixator in the region of the proximal femur is cumbersome and awkward for these patients who are often elderly and overweight (Fig. 5). If an infection is present, the procedure can be performed in two stages. The first stage consists of infection eradication and insertion of a cement spacer. In the second stage, the knee arthrodesis is performed with a rod and simultaneous LON. If there are any patient concerns about simultaneous arthrodesis with lengthening, the lengthening can be performed at a later date over the existing nail. Bone grafting is routinely performed at the time of rod insertion to supplement the arthrodesis. Touchdown weight bearing is allowed during the lengthenings postoperatively.

Actively infected, nonobese, below 5 cm LLD, mentally capable of a fixator. This category of patients is best served with an external fixator because the arthrodesis and infection eradication can be performed at the same surgical setting. Performing these functions simultaneously significantly decreases the total treatment time for patients. The best of both infection principles are combined at the initial surgery—good debridement and good bone stability. Patients with significant obesity are not good candidates for external fixation because

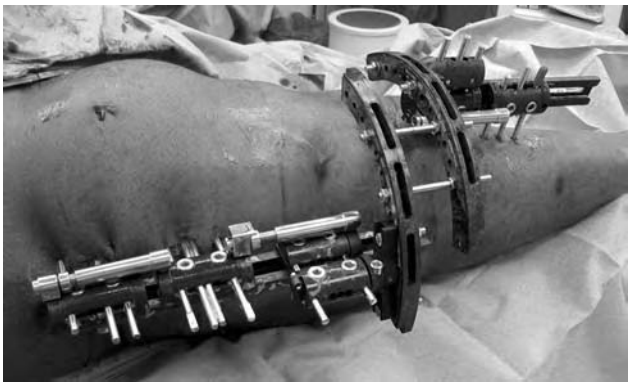

Figure 5 Clinical photo of a patient undergoing a proximal–tibial and distal–femoral lengthening over a nail with Sheffield rings (Orthofix, Verona, Italy) compressing the arthrodesis site.

the longest external fixator pins are 250 mm. If the soft tissue will not accommodate the frame with this length of pin, then the bar will cause skin breakdown and not allow for any swelling. The external fixator allows for active compression at the arthrodesis site and can be used to lengthen by small measures simultaneously (Fig. 6). In the author's experience of four external fixator lengthenings with simutaneous knee arthrodesis, the consolidation of the regenerate did not delay the removal of the external fixator (28).

If there is a large anterior wound at the time of initial application of the external fixator, the author applies the lateral fixator and allows the patient only 50% weight bearing postoperatively. This allows for easier access to the wound anteriorly until the wound is healed (Fig. 7). As soon as the wound is healed and the infection is eliminated, the anterior Orthofix rail (Verona, Italy) is applied at the time of bone grafting. The patient is then allowed full weight bearing. Bone grafting is always done in these cases in a delayed fashion when the infection is eradicated. This is usually two months following the initial surgery. Any residual anterior wounds are skin grafted at the time of bone grafting.

An Orthofix biplanar external fixator is the author's preferred method of knee fusion because of the ability to actively compress postoperatively while simultaneously treating the infection.

Actively infected, obese, below 5 cm LLD, not mentally capable of fixator. These patients are difficult to treat because they are unable mentally or physically to care for a fixator. Often these patients do not have a good support network of friends or family who could assist them with the pin care and fixator turns, if needed. These situations are best treated with long intramedullary arthrodesis rods. The treatment plan occurs in two stages. In the first stage, the infection is eradicated and a cement spacer is inserted. Numerous authors have demonstrated the need to eradicate infection prior to insertion of an intramedullary rod (14,16–19). Postoperatively the patient uses a hinged knee brace locked in extension or a cylinder cast for immobilization. Two months later, after a six-week course of antibiotics and no signs of infection, the cement spacer is removed through a transverse incision and a long intramedullary fusion rod is inserted (Fig. 8). If there is any question as to a residual infection, the bone grafting with iliac crest bone graft can be performed at a later stage. Postoperatively, if there is good bone contact, the patient may bear weight as tolerated.

Anterior cortical defects—femur and tibia. This situation commonly arises when the long stem revision TKA is difficult to extract and the joint surgeon makes an anterior cortical window in the tibia and femur. Although this exposure allows for easier implant removal, it devascularizes a large segment of cortical bone in an already compromised and infected leg (Fig. 9). In these cases, a thorough debridement of the area (often including the anterior cortical segment when it is completely devascularized) is completed and a fusion is performed using an intramedullary rod. This rod bypasses the cortical defects that would otherwise make a fixator application with half-pins difficult. With active infection, a thorough debridement is followed by a cement spacer insertion at the first surgical procedure. After the infection is eradicated, the intramedullary rod and bone graft are inserted during the second procedure.

Failed knee arthrodesis. The majority of cases with failed knee arthrodeses occur for two reasons (excluding inadequate blood supply): inadequate fixation or persistent infection. With adequate bone stock, the Wichita Fusion Nail (Stryker, Mahwah, New Jersey, U.S.A.) has

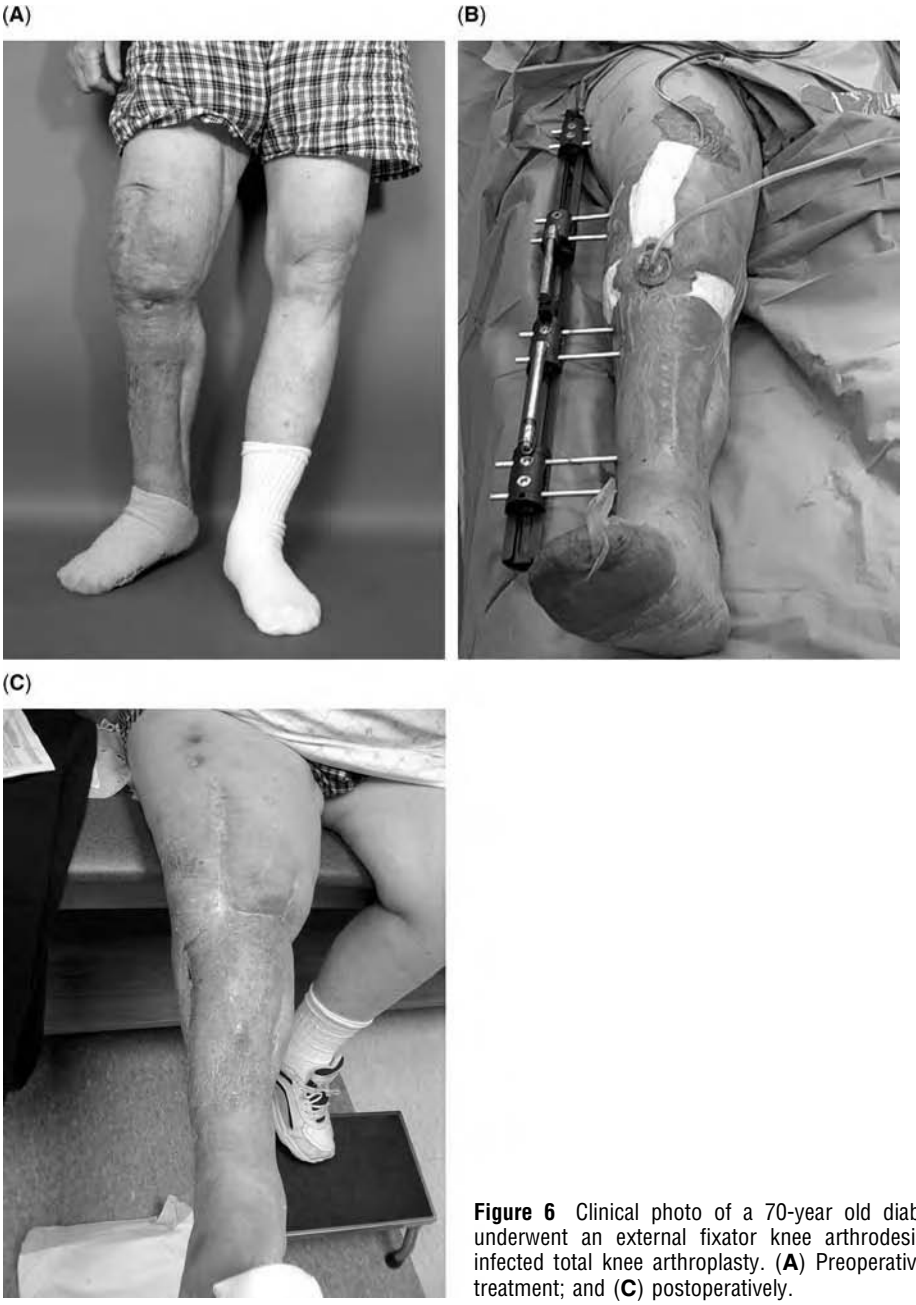

Figure 6 Clinical photo of a 70-year old diabetic male who underwent an external fixator knee arthrodesis following an infected total knee arthroplasty. (**A**) Preoperatively; (**B**) during treatment; and (**C**) postoperatively.

shown very good results as reported by Christie et al. with a 100% fusion rate in 53 knees following failed TKA (29). When large metaphyseal bone loss is experienced, the Wichita Nail does not provide adequate fixation. If the femoral segment and tibial segment each measure 20 cm, it does not provide enough stability given the long lever arm across the knee. The majority of failed knee arthrodeses are secondary to short intramedullary devices inserted through the knee (Fig. 10). There are two ways to revise these arthrodeses from short fusion nails. The first way is a long intramedullary device from hip to ankle, which neutralizes the long lever arm across the knee. The intramedullary device, accompanied with iliac crest bone grafting, is only an option if the patient does not currently have an infection. The device provides a static situation, however, and one cannot obtain active compression from an intramedullary rod. The second way and the author's preferred method is to use a biplanar

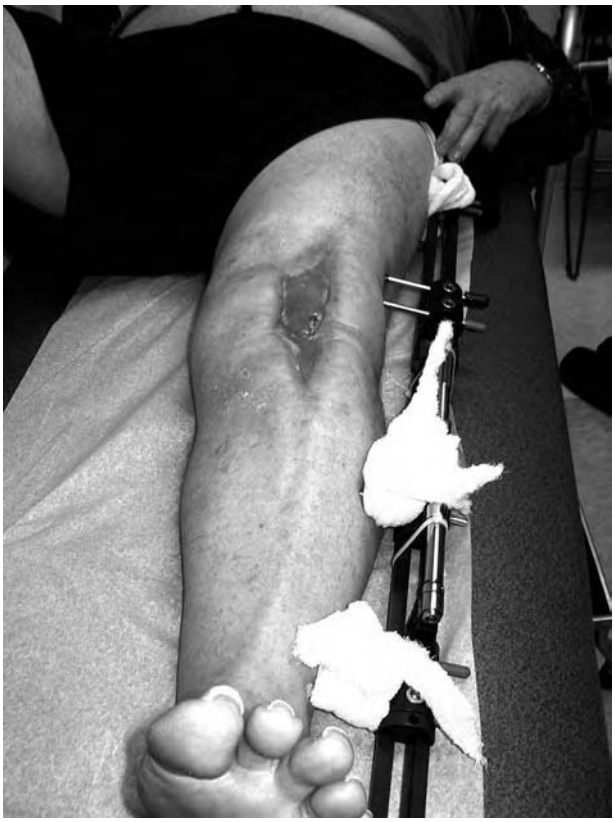

Figure 7 Clinical photo of a patient with a failed knee arthrodesis from a short intramedullary rod with a large anterior soft-tissue defect. Only the lateral fixator was applied and a wound vacuum-assisted closure was used to heal the anterior soft tissues. Following the formation of healthy granulation tissue, a split-thickness skin graft was applied to the wound.

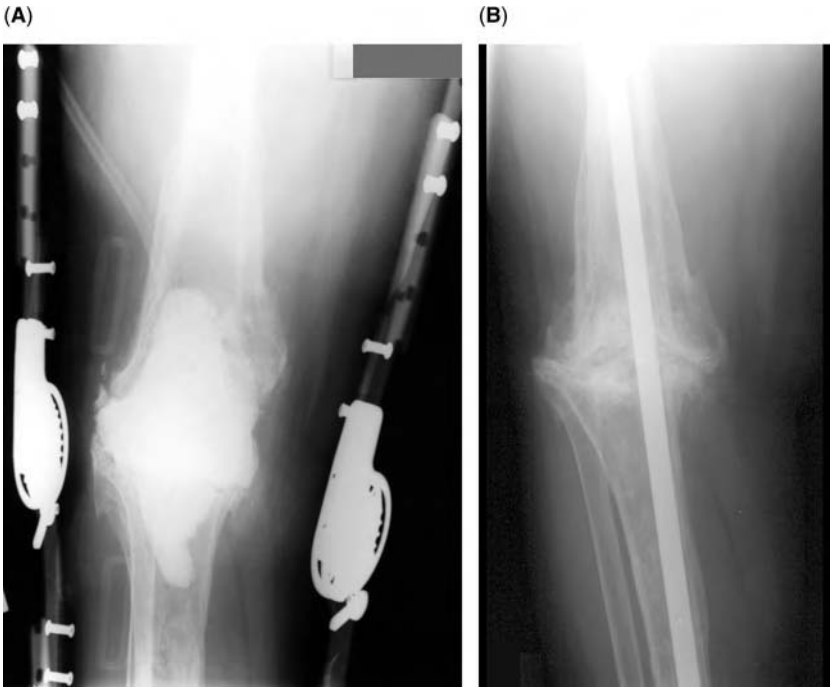

Figure 8 Preoperative (**A**) and postoperative (**B**) radiographs of a patient following an infected total knee arthroplasty. This patient had a spacer placed for eight weeks followed by the insertion of an intramedullary rod and iliac crest bone grafting.

Figure 9 Operative photo of an anterior tibial cortical window fixed with cerclage wires following the removal of an infected long-stem total knee arthroplasty component. Note the complete devascularization of the anterior segment.

external fixator that provides active compression at the fusion site. With external fixation, excellent stability and compression initiate the fusion process at the same surgical setting as the infection eradication. Bone graft is inserted at a second stage when there is no further evidence of infection.

Rand and Bryan (30) reported on a series of 25 patients with failed knee arthrodeses. All initial failures were due to bone loss. Their preferred method of revision arthrodesis was an external fixator. On the first repeat attempt, 5 of 14 united and 4 of 8 united on the second repeat attempt for a final success rate of 9 out of 25 patients. Clearly, a failed knee arthrodesis is one of the more difficult situations.

SURGICAL TECHNIQUES

Because the majority of knee arthrodeses are performed for infection, one Mayo stand is kept separate for the initial debridement. Once these instruments are used and the debridement is completed, the leg is reprepped and new drapes, gowns, and gloves are used and the dirty Mayo stand is removed. The entire limb from iliac crest to the toes is prepared in order to determine rotational alignment. Sterile tourniquets are used to decrease blood loss.

Initial Debridement

Initial debridement is performed in a systematic fashion starting with the bone. The tibial debridement is performed first. This includes using the burr to remove all necrotic bone and expose bleeding bone. A thorough debridement includes reaming the tibial canal to remove any necrotic debris or residual cement. The reamer and burr are cooled with irrigation while they are used to prevent thermal necrosis of the bone. Cement removal instruments are also useful in the debridement of the canals following infection from revision of TKA. The tibia is always debrided first because if an arthrodesis rod is the choice for fixation, the rod

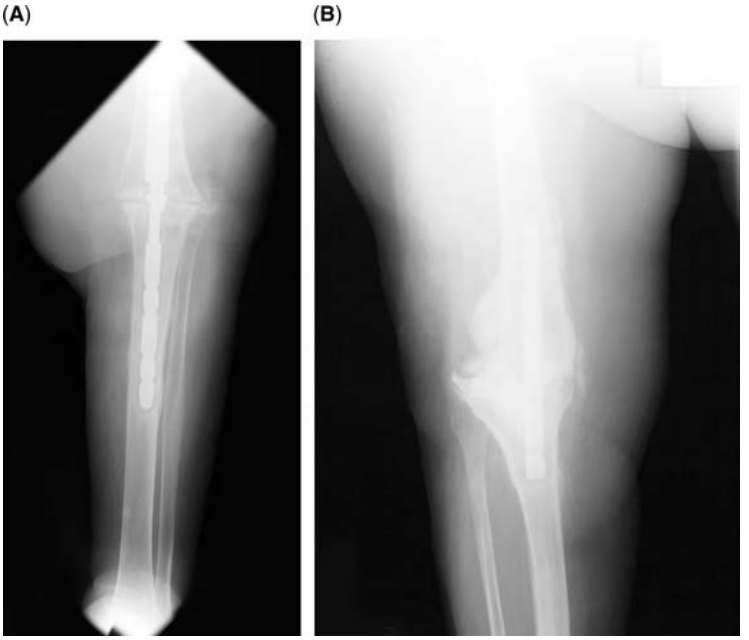

Figure 10 (**A** and **B**) Radiographs demonstrating two failed knee arthrodeses with two different types of short intramedullary devices.

diameter is determined by the last reamer to fit in the tibia. The rod diameter that is chosen is usually 1 mm less than the last reamer used. This prevents over reaming of the femur and a poor nail fit. The femur is reamed to the same diameter as the tibial canal. Once the bone is thoroughly debrided to clean bleeding bone, the soft tissue is addressed with a complete debridement of all necrotic tissue. The author uses a scalpel for the initial synovectomy and then the Versajet (Smith & Nephew, Memphis, Tennessee, U.S.) to safely debride the posterior capsule and the lateral and medial gutters. These areas often are very difficult to debride with a scalpel. During the initial debridement, the posterior capsule is carefully dissected from the posterior femur and tibia. This is necessary to determine whether the defect will be acutely compressible. Once the initial debridement is completed, then 9 L of saline are irrigated through the knee. The drapes and gowns are all replaced following the irrigation and the leg is reprepped. All of the debridement instruments are removed, and Bovie and suction are replaced with new ones. Then the procedure proceeds as a clean procedure.

Long Intramedullary Knee Arthrodesis Rod

Following the initial debridement, if the canals were not reamed, the tibial canal is sequentially reamed 0.5 to 1 mm over the rod chosen (keeping in mind the largest diameter intramedullary rod is usually 13 mm). The femur is reamed after the tibia to prevent over reaming of the femoral canal and a loose fitting nail. The length of the chosen rod is measured from the greater trochanter to 2 cm above the ankle. Great care must be taken when inserting the rod to obtain the proper rotation. The rotation of the leg must be chosen prior to the rod engaging the tibial segment. Any rotation of the tibial segment with the rod engaged in the tibia can result in a fracture. If additional compression is needed at the knee arthrodesis site, a temporary external fixator can be placed prior to insertion of the locking screws. The disadvantage of the intramedullary rod is the inability to obtain additional compression. The Stryker T2 Arthrodesis Nailing System (Stryker Trauma, Mahwah, New Jersey, U.S.A.), however, has the capability to add 1 cm of compression following the locking of the rod proximally and distally.

Long Intramedullary Arthrodesis Rod with Femoral Transport

The intramedullary rod is inserted as described above and the gap at the arthrodesis site is not compressed. A femoral external fixator is applied with three pins proximally and three pins

distally. These pins are inserted using a cannulated wire technique to prevent the pins from contacting the rod or entering the medullary canal. Once these pins are inserted, the femoral osteotomy is performed with multiple drill holes through two incisions—one laterally and one anteriorly. This osteotomy is difficult to perform and fluoroscopy should be used to confirm the osteotomy separation. Postoperative weight bearing is usually touchdown for balance until bone contact and the four cortices are consolidated. Once the segment has been transported, a plate can be added to hold the transported segment in place at the time of docking and bone grafting. The fixator can then be removed. Postoperatively, if the leg is lengthening, the gastrocnemius–soleus complex can become tight with a resultant equinus contracture. It is essential to preserve ankle range of motion with aggressive daily physical therapy and splinting.

Biplanar External Fixation

The author's preferred method for external fixation is the Orthofix Limb Reconstruction System (Orthofix International, Verona, Italy). The initial application starts with a wire inserted perpendicular to the proximal femur at the level just above the lesser trochanter. Another wire inserted is inserted just above the ankle perpendicular to the tibial mechanical axis (Fig. 11). The fixator is mounted in line with the mechanical axis of the leg and checked with the ''Bovie cord test'' to ensure that there is no mechanical axis deviation. There are four clamps on the lateral fixator that are placed on the distal tibia, proximal tibia, distal femur, and proximal femur. Once the lateral fixator is temporarily mounted on the wires, fluoroscopy is used to ensure that the proximal tibial and distal femoral clamps will line up with the bone. Often they are too posterior and ''sandwich'' clamps are needed to ensure that the pins engage the bone. Once this is checked and adjusted, the lateral fixator is mounted and the bone ends are then compressed under direct vision. The pulse is also documented before and after compression to prevent any vascular compromise. Then the wound is closed and the anterior fixator is placed with a similar four-clamp construct. Often, the anterior wound cannot

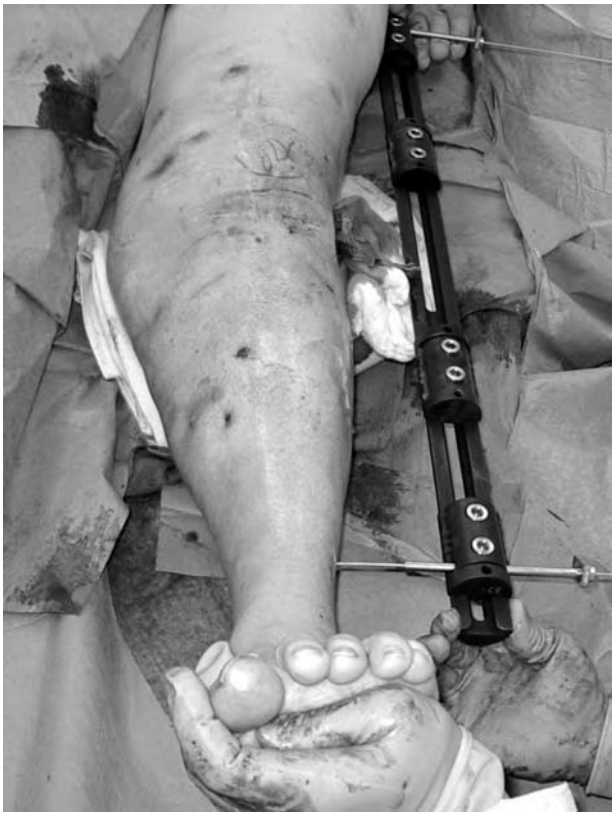

Figure 11 Operative photo demonstrating the initial mounting of the lateral Orthofix fixator to the femur and tibia in line with the mechanical axis of the limb.

Figure 12 Drawing depicting the completed application of the Orthofix Biplanar external fixator.

be closed. As a result, the placement of the anterior fixator is delayed until the wound is healed. A wound vacuum assisted closure (KCI, San Antonio, Texas, U.S.A.) is often used for these anterior wound problems as well as plastic surgical procedures. If only a lateral fixator is placed, the patient is allowed 50% weight bearing. An iliac crest bone graft is also used to supplement the fusion once the wound is healed and can be performed at the same time as the anterior fixator application. Once the biplanar Orthofix external fixator is placed, the patient returns monthly for radiographs and active compression (Fig. 12). Postoperative compression is performed routinely either by the patient or the surgeon depending upon the amount of compression obtained at the time of surgery. The patient is allowed weight bearing as tolerated with the biplanar device. In the author's experience, despite the best bone contact, no fusion has been stable for frame removal in less than six months and all fusions performed after infected TKA require bone grafting.

COMPLICATIONS

Complications following knee arthrodesis are common with rates ranging from 20% to 84% (18,25,31,32) and include recurrent infection, nonunion, peroneal nerve palsy, and thrombophlebitis.

Peroneal nerve palsies are a result of compression at the knee arthrodesis in the presence of scar tissue or excessive compression. These have been reported by several authors (33–35) and the best treatment for this complication is immediate nerve decompression (Fig. 13) (36,37).

Nonunion is a common complication with any method of knee arthrodesis (22,23,30). Rand and Bryan (30) reported on 25 failed knee arthrodeses and the most common reason for the failures was due to loss of bone stock. They were successful with arthrodeses using repeat external fixation in 5 of 14 knees and a second attempt at arthrodesis was successful in four of eight knees. Nonunion clearly is a difficult problem. Bone grafting is routinely used

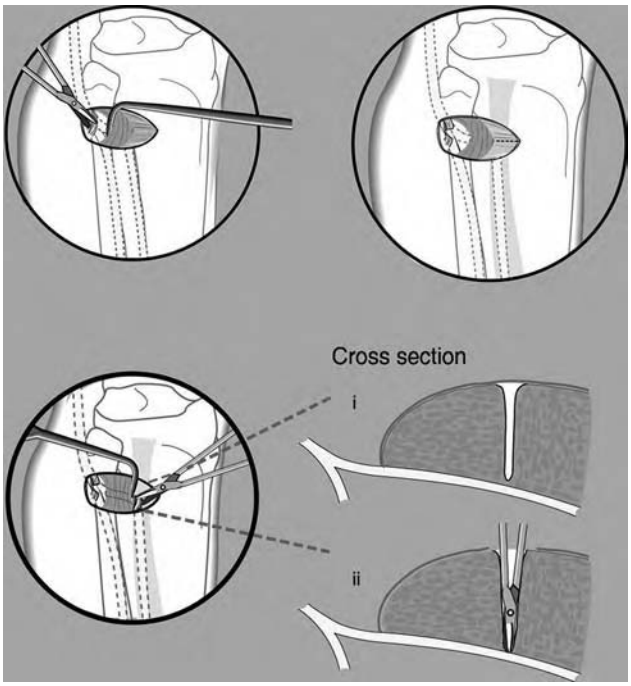

Figure 13 Drawings depicting the decompression of the peroneal nerve. *Source:* From Ref. 38.

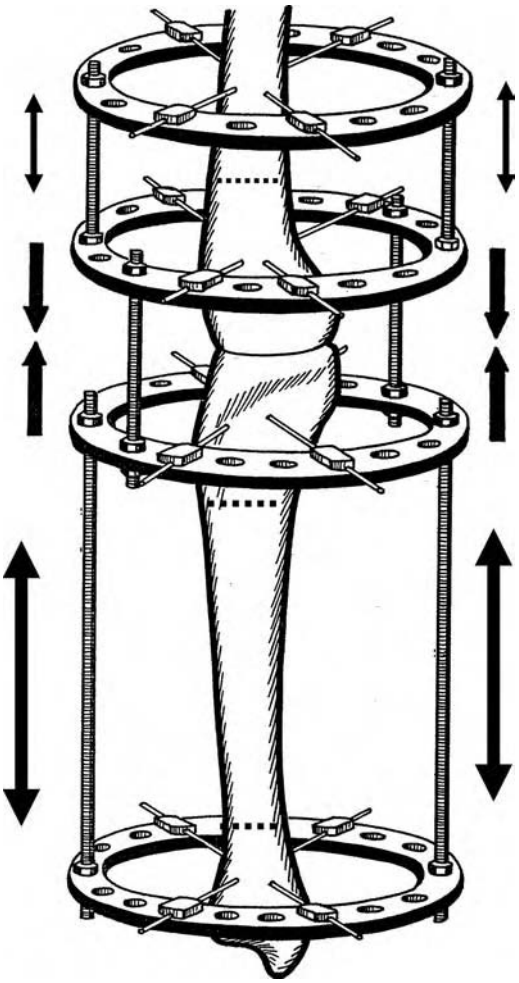

Figure 14 Schematic illustration of an Ilizarov construct for knee arthrodesis and lengthening. Note the compression across the arthrodesis site and distraction at osteotomy sites (*dotted lines*) of the distal femur, proximal tibia, and/or distal tibia. *Source*: Courtesy of Arkady Blyakher.

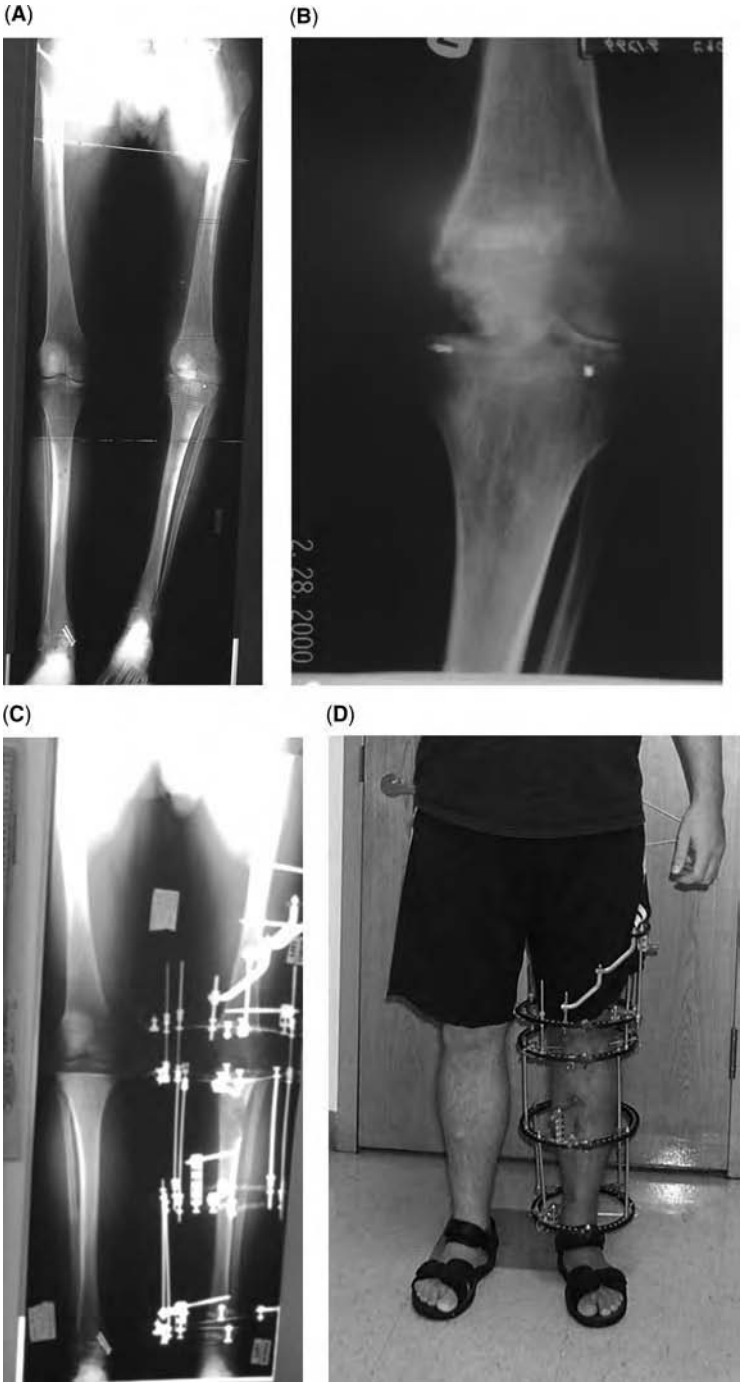

Figure 15 (*Continued on next page*) A 35-year old man with open knee injury and methicillin-resistant *Staphylococcus aureus* septic arthritis and osteomyelitis. (**A**) Preoperative standing bipedal radiograph. (**B**) Preoperative anteroposterior view radiograph of the knee. (**C**) Standing bipedal radiograph obtained at five months shows progression of healing at the knee arthrodesis site and the 4 cm of lengthening regenerated bone in the proximal tibia. (**D**) Standing front view photograph shows the patient at five months with the Ilizarov frame in place. (**E**) Anteroposterior view radiograph of the knee, obtained at 12 months, shows complete bony union of the arthrodesis and lengthening site. (**F**) Standing front view photograph shows the patient at 12 months; no deformity is present, and limb length discrepancy is 6 mm. (**G**) Standing side view photograph of the patient at 12 months. *Source*: Case supplied by S. Robert Rozbruch, Institute for Limb Lengthening and Reconstruction, Hospital for Special Surgery, Weill College of Cornell University, New York, New York, U.S.A. (Case supplied by S. Robert Rozbruch.)

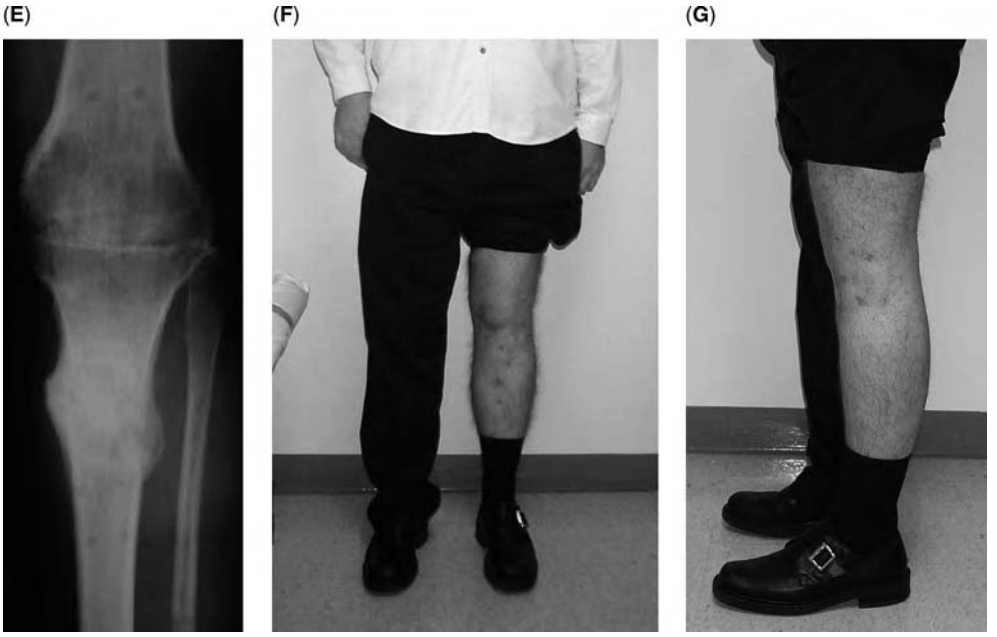

Figure 15 (*Continued from previous page*)

with any initial fixation in order to avoid prolonged disability and pain from a nonunited knee arthrodesis. Prevention of this complication is the best method for treating it.

FUTURE DIRECTIONS

The future holds more challenging cases in knee arthrodesis. Obesity and multiply revised TKAs will limit fixation choices and decrease bone stock. The number of knee arthrodesis procedures is on the rise as the number of failed and infected TKAs increases. Knee arthrodesis is an excellent alternative to an above-knee amputation because it provides a stable, painless, uninfected limb with better energy efficiency than an above-the-knee amputation (39,40). As the patient population undergoing this procedure ages, gait and balance problems will become significant. In this older population, a stable sensate foot is much better for balancing a limb than a prosthetic leg.

EDITOR'S ADDENDUM

We (28) reported a group of young patients with a nonreconstructable knee joint, bone loss, and infection after trauma that underwent knee arthrodesis with simultaneous lengthening. Arthrodesis of the knee with simultaneous limb lengthening was performed through an osteotomy of the tibia and/or femur and the use of an Ilizarov frame (Fig. 14). Bony union of the knee arthrodesis and lengthening sites and good alignment were achieved in all patients (Fig. 15). Mean amount of lengthening was 5.4 cm. Average time in frame was 11 months. LLD after treatment averaged 1.8 cm. Mean duration of follow-up after frame removal was 35 months. At follow-up, infection had not recurred, pain was not present, and assistive devices were not needed for ambulation. Average SF-36 scores improved in all eight categories, and the average American Academy of Orthopaedic Surgeons lower limb modules improved from a mean of 33 to a mean of 68.

We concluded that knee arthrodesis with simultaneous lengthening can be performed successfully using the Ilizarov method. It enables surgeons to optimize limb length during knee arthrodesis. The use of external fixation and the avoidance of internal implants may be advantageous in the presence or history of infection. The Ilizarov frame provides stability that allows weight bearing during treatment.

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25 Contractures of the Knee

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INTRODUCTION

Severe knee contractures are one of the most difficult problems causing gait disturbances (1). Extension contractures of the knee (ECK) cause swing-through dysfunction as well as dampen a small flexion moment in mid-stance (2). Flexion contractures of the knee (FCK) are more problematic because the center of gravity permanently falls posterior to the knee and the leg is functionally shortened. This shortens the stride length and fatigues the extensor mechanism, profoundly disrupting gait mechanics (2–5). Unlike isolated ankle or hip contractures, FCK cannot be well compensated by the other lower extremity joints or the lumbar spine.

Knee contractures can be congenital, acquired, or a combination of the two. Congenital conditions include arthrogryposis, pterygium syndromes, meningomyelocele/sacral agenesis, and tibial hemimelia. Examples of acquired conditions are neuromuscular disorders, trauma, burns, septic arthritis, juvenile rheumatoid arthritis, hemophilia, polio, and fibrosis from intramuscular injections. Combinations of congenital and acquired causes may occur such as in skeletal dysplasias or dwarfism syndromes. Distal femoral or proximal tibial deformities may create or exaggerate a knee flexion/extension deformity. We will limit our scope to the treatment of soft tissue contractures without associated juxta-articular bony deformities. I have found gradual extension of knee contracture by external fixator (GEKCEF) to be an important treatment option for severe contractures in both children and adults, and will discuss the technique in detail.

CLINICAL EVALUATION

Knee

Contractures have either a soft or a hard endpoint. A soft endpoint has the same feel as hyperextending an metacarpal phalangeal joint, whereas a hard endpoint is similar to an elbow's block to overextension. A hard endpoint in an ECK usually indicates an intracapsular cause, whereas a soft endpoint is more commonly due to muscle fibrosis or shortening. In practice, though, ECKs are due to a combination of both intra- and extra-articular causes. An FCK with a soft endpoint will likely have a sizable improvement immediately from soft tissue procedures. If the endpoint is hard, soft tissue procedures are still essential, but the acute improvement will be less.

In lower extremities with a knee contracture and a coronal plane deformity of the femur or tibia, it is difficult to differentiate the contribution of each to the total limb malalignment and to accurately measure the magnitude and location of the bone deformity (6). FCK prevents proper positioning of the limb for radiographic analysis. In these cases, it is easier to correct the FCK first. Once the knee is straight, corrective osteotomies can be accurately planned.

Lack of patellar mobility in ECKs indicates intracapsular fibrosis, requiring release as part of treatment. In FCKs, an immobile patellar may also require release after correction of the contracture to improve active extension. The presence or absence of functional knee extension strength is important for choosing treatment options, particularly in treating ECK.

Hip

Hip deformities and contractures are challenging obstacles to correcting knee contractures, maintaining correction, sitting, and ultimately ambulating postcorrection. Hip flexion

contractures greater than 30°, extension contractures, and rotational abnormalities that prohibit positioning the knee directly anteriorly need to be addressed prior to contracture correction. If the knee is treated first, it will be difficult to maintain knee correction while treating the hip.

Foot and Ankle

With regard to the foot and ankle, the primary question is whether the foot needs to be stabilized while treating the knee. Stability is important under two conditions: when the ankle is at risk of developing an equinus contracture and when the foot is undergoing simultaneous deformity correction, either gradually or acutely.

Soft Tissue

The presence of skin grafts or extensive scars may factor in planning surgical incisions or pin sites. Popliteal space scarring and compromised neurovascular status will limit the extent of acute contracture correction in order to avoid skin necrosis, neuropraxia, or vascular embarrassment (7–9). In these cases, GEKCEF may be a safer option. These patients need to be closely monitored, and the rate of contracture distraction should be adjusted as needed.

RADIOGRAPHIC EVALUATION

In FCKs, anteroposterior (AP) radiographs of the femur, tibia, or knee are difficult to obtain because of the flexed position of the knee. Long leg AP films are essentially meaningless (6). Individual prone tibia and femur posteroanterior films allow each segment to be placed much closer to the film cassette. Extension and flexion lateral radiographs define the knee's range of motion (ROM), and demonstrate any tendency towards tibial hinging or subluxation. Also, distal femoral and proximal tibial joint orientation lines help determine if a deformity is entirely a contracture or has a bony procurvatum/recurvatum component, by measuring the posterior distal femoral and posterior proximal tibia angles (10). Poor bone density indicates the potential for fracture. For GEKCEF, the diameter of the Rancho half-pins should be about 25% to 33% of the diameter of the isthmus of the bone.

CLASSIFICATION

Extension Contractures of the Knee

ECKs can be classified as having intra-articular or extra-articular etiologies, or commonly both. In intra-articular ECKs, the patellar mobility is significantly limited with a hard endpoint. Intra-articular contractures require aggressive releases of the knee joint itself, whereas extra-articular contractures are treated primarily through the quadriceps. Knees with ECKs can further be characterized as those that have functionally active extension against gravity and those that do not. Those that do have active extension against gravity should not be treated with quadriceps lengthening procedures if possible, as the ability to walk without orthotic support may be sacrificed.

Flexion Contractures of the Knee

Children should be considered separately from adults when classifying knees with FCKs. Children's contractures can recur due to growth or from remodeling of extension osteotomies. On the other hand, adults develop intra-articular fibrosis rapidly, even when contractures are initially extracapsular. These contractures are more resistant to correction by simple soft tissue procedures. The size of the contracture naturally dictates treatment. In children, the contractures are grouped as less than 20°, between 20° and 30°, between 30° and 40°, and over 40°. In adults, there are three ranges: less than 20°, between 20 and 30°, and over 30°.

FCKs can also be discriminated as having a soft or a hard endpoint. The presence of a hard endpoint usually indicates the need for a more involved treatment, be it posterior capsular release (PCR), osteotomy, or GEKCEF.

TREATMENT OPTIONS

Extension Contractures of the Knee

In ECKs, both the intra- and extracapsular components need to be addressed. Intracapsular releases can be performed through a lateral and an anteromedial incision, releasing the lateral and medial retinaculum, suprapatellar pouch, and any intra-articular adhesions (11,12). Extracapsular contractures are addressed by Judet type elevating quadricepsplasties (11), quadriceps lengthenings (8), or a combination of the two (8,13,14). Elevating quadricepsplasties involves extraperiosteal release of the quadriceps off the femur and the lateral intermuscular septum. In extensive cases, the vastus lateralis is released from the greater trochanter and the rectus femoris off the pelvis. Quadriceps lengthening procedures can be V-Y lengthenings (8) or a more extensive release of the medial and lateral vasti from the quadriceps tendon, reattaching them more proximally, possibly combined with a rectus femoris tendon lengthening (8,13).

Flexion Contractures of the Knee

FCK can be treated nonoperatively or by soft tissue releases, extension osteotomies, and GEKCEF.

Nonoperative Treatment

An FCK of less than 20° can often be tolerated in patients who can compensate with normal strength hip and ankle function (1). The quadriceps' function is augmented when the body's center of gravity is brought anterior to the knee by leaning forward with the hip locked in extension, and using the plantar flexors to stabilize the ankle (4,5). Bracing with knee-ankle-foot orthoses (KAFOs) or floor reaction ankle-foot orthoses may improve gait efficiency in the face of quadriceps and ankle weakness.

Hamstring Lengthenings

Hamstring lengthenings (HSLs) are indicated when the popliteal angle is greater than the flexion contracture. If the contracture is small and the endpoint is soft, HSL may be useful as the only procedure; otherwise it serves as an adjunct to other procedures. Hip extension strength should be evaluated preoperatively, and if weak, a transfer of the hamstrings tendon to the distal femur may be indicated to prevent a crouched gait (15). In children, FCKs of 20° or less are often responsive to HSL alone. In walking children with cerebral palsy, HSL may be effective with contractures up to 30°. Due to rapid intracapsular fibrosis in adult knees, only relatively recent contractures of less than 20° are indicated for HSL alone. I prefer a percutaneous release of the gracilis and semi-tendinosus just above the knee flexion crease, and open muscle-sparing tenotomies of the semi-membranosus and biceps femoris through 3- to 4-cm incisions at the junction of the middle and distal thirds of the thigh.

Posterior Capsular Releases

PCR are usually done in conjunction with hamstrings and/or gastrocnemius tenotomies. In adults, posterior knee releases are indicated for most contractures under 20°. In children, a posterior release is necessary for contractures of less than 20° with a hard endpoint or without hamstring tightness. It is usually necessary as an adjunct to HSL for contractures between 20° and 30° with a soft endpoint and hamstring tightness.

Supracondylar Femoral Extension Osteotomies

Extension osteotomies of the distal femur are indicated for children with contractures up to 45°, and are even suitable for contractures as small as 20° to 30° with a hard endpoint. Although larger contractures, up to 90° (16,17), have been treated with extension osteotomies, this can lead to neurovascular stretch injury, nonphysiologic weight-bearing on the posterior femoral condyles, and unacceptable recurrence rates of 0.9° per month (16,18,19). Neurovascular injury can be avoided by shortening the femur, although this exacerbates the limb shortening caused by the angular osteotomy. Attention to alignment is important to avoid recurvatum or coronal plane deformities (20). In adults, concerns of neurovascular injury, femoral shortening, and posterior femoral condyle weight bearing limit the indications to contractures of 30° or less. A 90° condylar blade plate through a lateral approach to the distal femur is used for fixation. With the knee in full extension, the blade is inserted parallel to

the distal femoral condyles on the AP image view, and the plate portion is in line with the tibial shaft in the sagittal plane. Either a transverse or isosceles triangular closing wedge osteotomy is performed, shortening the femur as little as possible.

Gradual Extension of Knee Contracture by External Fixation

GEKCEF is an excellent tool for correction of knee flexion contractures of essentially any size (21–23). Gradual rather than acute correction reduces the risk of neurovascular injury. Tibial subluxation can be addressed during contracture correction. No femoral shortening is required, even with large deformities. In pterygia, the contracted skin and fascia can be gradually stretched, essentially resolving the soft tissue web once the knee is fully extended. Its main drawbacks are its technical difficulty, patient/family acceptance, and inconvenience. Indications, therefore, are contractures over 40° in children and 30° in adults, and it may be combined with a posterior capsule and/or tendon release. After GEKCEF, there is usually a loss of motion compared to the preoperative total arc of motion (TAM). Intensive physical therapy can restore most or all of the motion (24). Stiff knees, with a hard endpoint and low TAM, require a slower rate of correction for patient comfort and to reduce chances of periarticular fracture. They should be held in the corrected position longer prior to frame removal, and managed in extension bracing for an extended period of time. To improve on the preoperative TAM, a correction of knee extension contracture can be performed after the gain in knee extension has been stabilized.

Knees with flexion contractures and subluxation of the tibia on the femur (posterior or rotatory) are best treated by acutely reducing the subluxation with an external fixator, followed by gradual extension of the knee. Instability of the collateral or cruciate ligaments is unusual after correction (25). The posterior cruciate ligament can be transected as part of an extensive posterior release, without causing clinical knee instability postcorrection. Any significant ligamentous repair should be done after contracture correction rather than before.

SURGICAL TECHNIQUES

Since 1999, we have treated 22 FCK with GEKCEF, 12 of which were arthrogrypotic knees. Below is our treatment protocol.

Approximation of Knee Center of Rotation

The true center of knee rotation cannot be defined, but an axis can be approximated, running through the origins of the medial and lateral collateral ligaments (26). Under image intensification, the knee is positioned with the femoral condyles overlapping in the lateral projection (approximately 3° of external rotation), and the intersection point of the posterior femoral cortex and the widest anterior-posterior dimension of the femoral condyles is identified (Fig. 1) (23). In pediatric cases, the immature posterior femoral condyles are visualized by an arthrogram; the axis point is often at the level of the physis. A 1.8-mm Ilizarov transfixation wire is then drilled from medial to lateral, protruding 3 to 6 cm from either side of the skin. Posterior and/or proximal positioning of the hinge should be avoided because this exacerbates the tendency towards posterior tibial subluxation and anterior tibiofemoral impaction at terminal extension.

Posterior Knee Release

The leg should be sterilely prepped to the level of the anterior superior iliac spine. No tourniquet is used. Medial and lateral incisions, 5 to 8 cm in length, are centered over the palpable posterior femoral condyle, parallel to the ground (Fig. 2) (27). Beginning laterally, the iliotibial band is incised, and its posterior half is released. The biceps femoris tendon is isolated and transected after verifying the safety of the common peroneal nerve. Soft tissues are elevated off the posterior joint capsule, until a finger can be passed to the capsule's midpoint. The lateral head of the gastrocnemius is isolated and transected. A capsulotomy along the joint line extends from the posterior half of the lateral collateral ligament to the midpoint of the posterior capsule (Fig. 3). If the posterior geniculate artery is cut or avulsed, the wound is packed for five minutes, obtaining hemostasis.

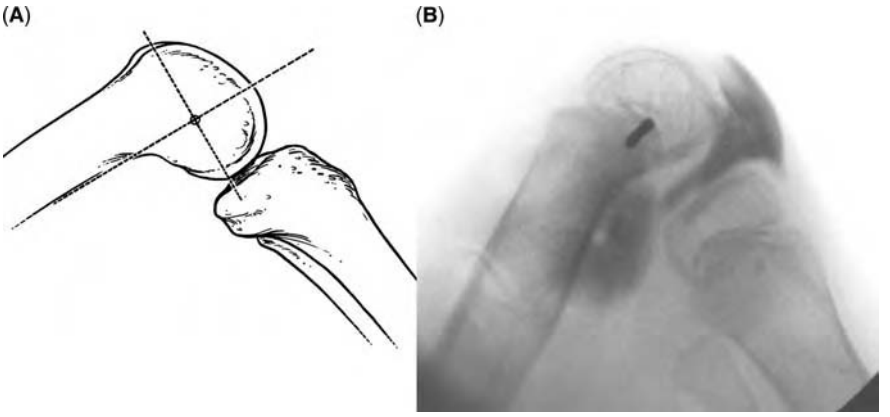

Figure 1 (**A**) Knee flexion-extension axis, at the junction of the posterior femoral cortex and the widest anterior-posterior dimension of the femoral condyles. (**B**) Axis in a 3-year old. An arthrogram is used to overlap the posterior and distal femoral condyles.

Medially, the vastus medialis is retracted anteriorly, and the semitendinosus and gracilis tendons are transected. Deep to these structures, the semimembranosus is identified and its fascia transected, leaving the muscle belly intact. The soft tissues are gently and bluntly elevated off the medial aspect of the posterior capsule. The medial head of the gastrocnemius is transected. The capsulotomy is completed posteriorly and is extended anteriorly to incise the posterior half of the medial collateral ligament, staying cephalad to the attachments of the medial meniscus (Fig. 4). If the posterior cruciate ligament can be felt as a taut band in the intracondylar notch, it should be released. Only the skin is closed with bioabsorbable suture. We have not found a drain to be necessary.

Ilizarov Frame Application

A femoral ring block (two full rings connected by four threaded rods) is attached to the femur. An Ilizarov universal joint is aligned on either side with the knee axis wire. A tibial ring block is positioned by connecting it to the universal joints with threaded rods. The tibial block is then affixed to the tibia, initially with transverse wires. The wire to the proximal tibial ring is positioned so that as it is tensioned, it pulls the tibia slightly anterior on the femur; this will prevent posterior tibial subluxation during contracture correction (Fig. 5). After the tibial block is well secured to both the tibia and the universal joints, the knee is distracted on the tibial side by 5 to 10 mm, to avoid impingement and articular cartilage damage during correction. The motor for the frame can either be a posterior distraction or an anterior compression rod; the latter is more convenient for seating.

In some cases, full rings could not be applied at the distal femur and proximal tibia. In small children with contractures greater than 60°, the full rings will impinge against each other

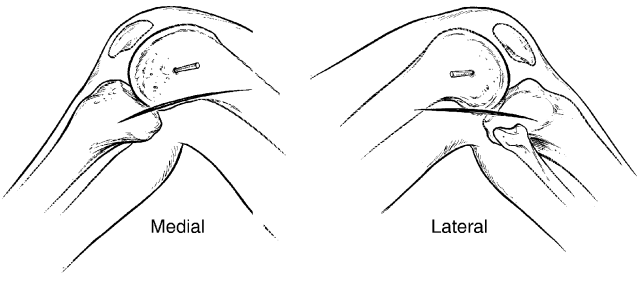

Figure 2 Incisions for posterior knee release, centered over the palpable posterior femoral condyles, parallel to the ground.

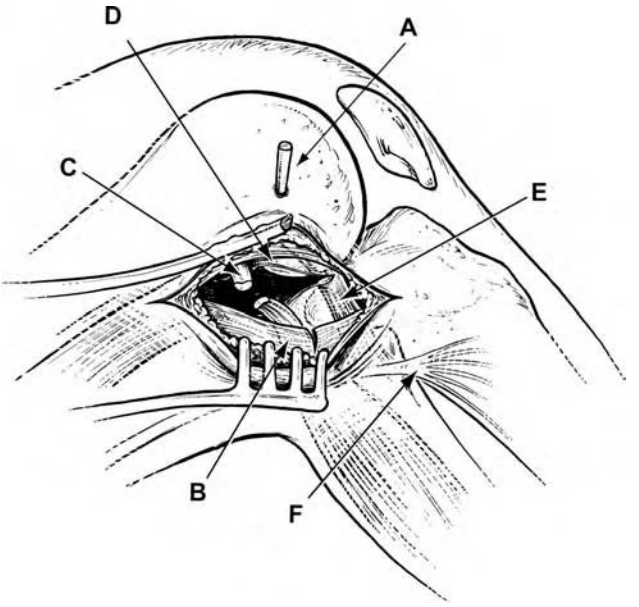

Figure 3 Posterior knee-release, deep dissection, lateral aspect. (**A**) Knee axis wire, (**B**) iliotibial band, posterior half transected, (**C**) transected lateral gastrocnemius, (**D**) cut posterior-lateral capsule, with posterior femoral condyle exposed, (**E**) lateral collateral ligament, posterior-half transected, (**F**) peroneal nerve.

posteriorly. Similarly, with pterygia, the soft tissue web will interfere with application of the full rings. Instead, a half or two-third ring is used as a ''drop ring'' either in addition to, or as substitution for, the periarticular full rings, so as to provide better support for the hinges (Fig. 6).

Postoperative Care

Correction is begun approximately one week after surgery. A modified method of similar triangles is used to calculate a rate of correction between 1° and 2° per day, adjusted to the patient's tolerance (28). After full extension is achieved, the frame is maintained as a holding frame for an additional time. The frame is subsequently removed in the operating room, the leg measured for a KAFO with locking knee hinges. It is then cast in full extension, molded to prevent posterior tibial subluxation. The knee often has initial ligamentous laxity, which usually resolves by the time

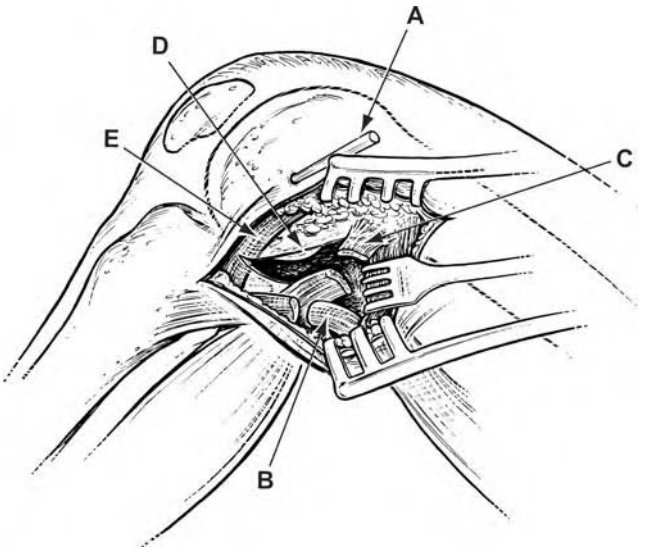

Figure 4 Posterior knee release, deep dissection, medial aspect. (**A**) Knee axis wire, (**B**) transected medial hamstrings, (**C**) transected medial gastrocnemius, (**D**) cut posterior-medial capsule, with posterior femoral condyle exposed, (**E**) medial collateral ligament, posterior-half transected.

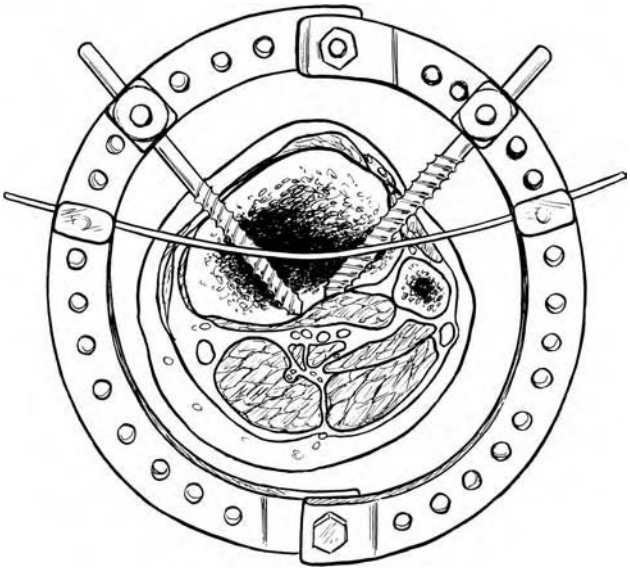

Figure 5 Tensioning the proximal tibia anteriorly. An Ilizarov wire is attached anteriorly to the ring prior to tensioning the wire.

of cast removal. The cast is removed in two weeks and full-time KAFO use is begun immediately, as is physical therapy to regain knee motion (Fig. 7).

COMPLICATIONS OF GRADUAL EXTENSION OF KNEE CONTRACTURE BY EXTERNAL FIXATION

The most common problems, other than those routinely expected with long-term fixators (e.g., minor pin tract infection), were posterior tibial subluxations, recurrences of contracture, fractures, and stiff knees.

Tibial Subluxation

Tibial subluxation is prevented by accurately placing the knee axis wire, erring on the side of placing the wire slightly inferior and anterior, and tensioning the tibia anterior in relation to the femur (Fig. 4). Any subluxation at frame removal does not lead to persistent joint subluxation, but in fact leads to contracture recurrence. If subluxation is identified while the frame is still on, the lateral knee radiograph is evaluated for a shift in the frame's axis in comparison to the knee's axis. The knee is then flexed until the tibia aligns well with the femur, followed by

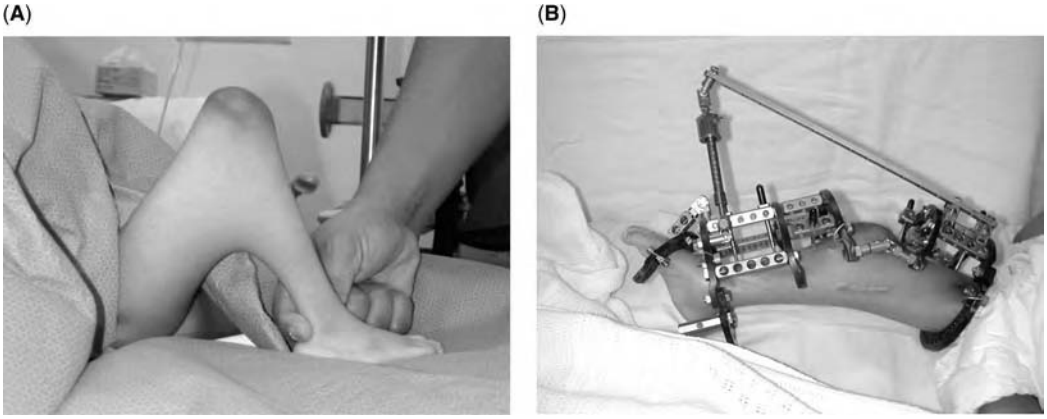

Figure 6 (**A**) Four-year-old girl with sacral agenesis and bilateral 120° contractures and popliteal pterygia. (**B**) Ilizarov frame with distal femoral and proximal tibial drop rings, midcorrection.

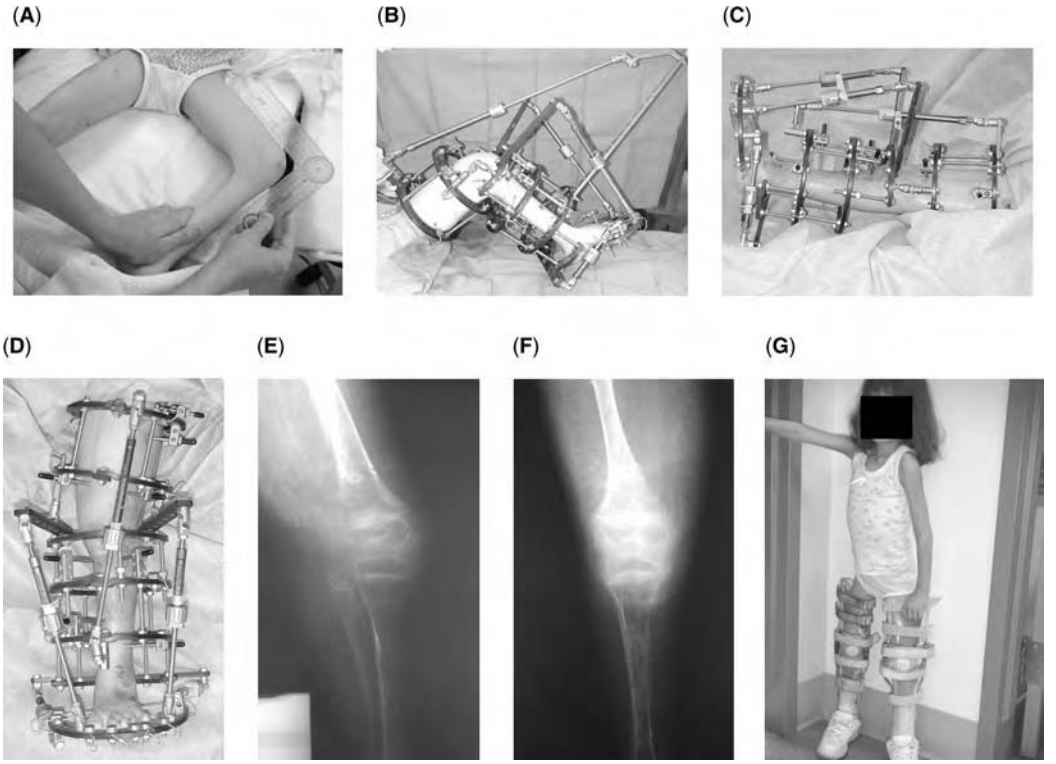

Figure 7 (**A)** Four-and-half-year-old girl with arthrogryposis, with bilateral 80° flexion contractures and clubfoot deformities. (**B)** Application of Ilizarov frame after posterior knee-release, and hind and midfoot osteotomies, for gradual correction of knee and foot deformities. (**C)** and (**D**) Side and front views on completion of correction. (**E)** and (**F**) Lateral and anteroposterior radiographs immediately after frame removal. The knee joint is still distracted. (**G)** At six years of age after correction of both knees.

realigning the axes. The joint is further distracted or the tibia further tensioned anteriorly, prior to resumption of contracture correction.

The use of the Taylor Spatial Frame (Smith & Nephew, Memphis, Tennessee, U.S.) may also prove to be useful for the correction of joint subluxation.

Recurrence of Contracture

Two patients with severe pterygia and flexion contractures over 90° developed acute recurrences. This complication is best prevented by obtaining a well aligned, non-subluxated knee prior to frame removal, then casting it for two weeks, to allow for stabilization of the ligaments. KAFO usage is continuous initially, then only at nighttime as the patient gains knee extension strength. Acute recurrences need to be treated with reapplication of the Ilizarov frame.

Fractures

Fractures could occur at different times during treatment. The perioperative fractures can be avoided by using half pins with widths not greater than 25% to 33% of the diameter of the bone's isthmus, avoiding unicortical pins, and being aware of any limitations of hip ROM, which could lead to excessive stress on the femur with positioning. Periarticular bending fractures occur during contracture correction, and are minimized by ensuring an adequate posterior release and initial joint distraction of about 10 mm. Tight clusters of metaphyseal pins seem to weaken bone and should be avoided to prevent fractures during frame removal. The limb should be gently handled until it is protected in a cast.

Knee Stiffness

Two stiff knees (TAM $< 20°$) occurred in cases where a strict protocol of ROM in a frame during physical therapy was followed. This was accomplished by unlocking the frame in

Table 1 Clinical Management

Classification Subgroup	Clinical Evaluation	Surgical Approach	Pearls	Complications/Pitfalls
Extension Contractures of the Knee				
Intra-articular	Poor patellar mobility	Intra-articular release	ECKs often are a combination of intra- and extra-articular causes	Recurrence of fibrosis
Extra-articular, good extension strength	Soft endpoint, antigravity strength	Judet's quad'plasty		Skin necrosis, hematoma
Extra-articular, poor extension strength	Soft endpoint, weak extension	V-Y quads lengthening	Will need orthosis for walking	Knee extension weakness
Flexion Contractures of the Knee				
Children				
Less than 20°, soft endpoint	Popliteal angle greater than FCK	Nonoperative management, or HSL	Long sitting post-op for stretching	Hamstrings weakness with HSL
Less than 20°, hard endpoint	Rule out bony flexion deformity	PCR, possible HSL	Immobilize 6 wk	Skin necrosis
20–30°, soft endpoint	Check hip extension strength	HSL, possible PCR	Protect heels in cast/ brace	Neuropraxia, skin necrosis
20–30°, hard endpoint	Rule out bony flexion deformity	PCR possible HSL, or SFEO	Immobilize 4–6 wk if PCR	Neuropraxia, skin necrosis
30–40°, soft endpoint	Evaluate knee extension strength	SFEO with possible HSL	ROM early (2–4 wk)	Neuropraxia, recurrence
30–40°, hard endpoint	Evaluate for knee subluxation	SFEO or GEKCEF	ROM early (2–4 wk) if SFEO	Recurrence with growth
Greater than 40°	Evaluate for knee subluxation	Soft tissue release, PCR and GEKCEF	Tension tibia anteriorly, distract joint	Fracture, stiff knee, subluxation, recurrence
Adults				
Less than 20°, soft endpoint	Popliteal angle greater than FCK	HSL usual not helpful alone, PCR	Careful not to over lengthen hamstrings	Hamstrings weakness, lack of correction
Less than 20°, hard endpoint	Rule out bony flexion deformity	PCR, possible HSL	Immobilize 4–6 wk	Neuropraxia, skin necrosis
20°–30°, soft endpoint	Evaluate knee extension strength	SFEO with possible HSL	Need walking brace post-op?	Neuropraxia, skin necrosis
20–30°, hard endpoint	Evaluate for knee subluxation	SFEO or GEKCEF	ROM early (2–4 wk) if SFEO	Neuropraxia, skin necrosis, vascular injury
Greater than 30°	Evaluate for knee subluxation	Soft tissue release, PCR and GEKCEF	Tension tibia anteriorly, distract joint	Fracture, stiff knee, subluxation, recurrence

Abbreviations: FCK, flexion contractures of the knee; ECK, extension contractures of the knee; HSL, hamstrings lengthening; PCR, posterior capsular release; SFEO, supracondylar femoral extension osteotomy; GEKCEF, gradual extension of knee contracture by external fixator; ROM; KAFO, knee-ankle-foot orthoses; FCK, flexion contractures of the knee; ECK, extension contractures of the knee.

order to work on knee flexion. In our opinion, an inflammatory response is stimulated by the contracture distraction, and is exacerbated by forced ROM in the frame, resulting in pericapsular contracture and stiffness. We stopped prescribing ROM therapy while in the frame; instead we keep the knees fully extended for a relatively short period (two to four weeks) prior to frame removal, with the knee well distracted. Our long-term results show an eventual restoration of TAM (24).

FUTURE DIRECTIONS

GEKCEF has proved to be an important treatment option in both children and adults. Children with pterygium remain a challenge, with frequent recurrences occurring in most series (22,29).

Protocols to prevent recurrence need to be established for this relatively rare condition. Can correction be obtained without significant soft tissue release (29)? Does the abnormal posterior fascia need to be resected from the proximal thigh to the distal calf (30)?

Another area of study is the treatment of the patient with a stiff knee (low TAM), or prevention of knee stiffness post-GEKCEF. Future work may yield substances that can be given systemically or injected locally to prevent joint or muscular fibrosis during treatment.

REVIEW OF LITERATURE

Authors	Journal, Year	Title	Number of Patients	Results	Conclusions
Dias	J Pediatr Ortho, 1982	Surgical management of knee contractures in myelomeningocele	15 ECK, 23 FCK	V-Y quad'plasty, 13/15 knees good or excellent; radical flexor release 23/25 good or excellent	ECK release early, need KAFO to walk
Hosalkar, Jones et al.	J Bone Joint Surg, Br 2003	Quad'plasty for knee stiffness after femoral lengthening in congenital short femur	5	Includes modified V-Y quad'plasty; good or excellent in 4/5	No increased extensor lag; continuous passive motion machine for 1 wk
Ali, Villafuert, et al.	Clin Orthop, 2003	Judet's quad'plasty surgical technique and results in limb reconstruction	10	Nine good or excellent, one fair	7 of 10 needed manipulation under anesthesia
Hollister, Jatana, et al.	Clin Orthop, 1993	The axes of rotation of the knee	Six cadaver knees	Flex/Exten axis is anteriosuperior to postero-inferior from medial to lateral	Distal articular femur is cone shaped
Murray, Fixsen	J Pediatr Orthop, 1997	Management of knee deformity in classical arthrogryposis multiplex congenita	FCKs 19 knees in 10 patients, PCR and serial casting	Recurrence in 6 of 19 knees, needing further surgery, including knee fusions	Poor long-term outcome, few remained ambulatory
DelBello, Watts	J Pediatr Orthop, 1996	Distal femoral extension osteotomy for knee flexion contracture in patients with arthrogryposis	22 knees	Good initial correction, improved ambulation; eight knees needed at least one repeat osteotomy	Remodeling occurs at 0.9°/mo in growing children
Asirvatham, Mukherje, et al.	J Pediatr Orthop, 1993	Supracondylar femoral extension osteotomy: its complications	100	72% improved gait, 10% post-op deformities, 9% neurovascular injury	Internally fix osteotomy, cast in 30° flexion
Herzenberg, Davis et al.	Clin Orthop, 1994	Mechanical distraction for treatment of severe knee flexion contractures	14 knees in 10 patients	Five knees good or excellent ROM, two knees fair, three knees poor	Rebound phenomena on frame removal
Brunner, Hefti	J Pediatr Ortho B, 1997	Arthrogrypotic joint contracture at the knee and foot: correction with a circular frame	13 knees in 7 patients	Six of seven patients had improved ambulation	Recurrences seen in all patients, especially under 10 yr old

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Part VII HIP

26 Ilizarov Hip Reconstruction

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INTRODUCTION

Children, adolescents, and young adults with late sequelae of neonatal hip sepsis (1,2) or chronic hip dislocation from dysplasia often present with the clinical problems of pain, Trendelenburg gait, lower extremity length discrepancy, and hip instability, all related to absence of part or all of the femoral head and neck and proximal migration of the femur.

Reconstructive procedures for this difficult problem including trochanteric arthroplasty, hip arthrodesis, pelvic osteotomy, and femoral osteotomy have not been satisfactory (1).

A proximal femoral subtrochanteric osteotomy (pelvic support osteotomy) has also been described. Support is achieved by valgus osteotomy of the proximal femur, placing the upper end of the femur against the lateral aspect of the pelvis. In addition, the valgus angulation improves hip biomechanics by improving the mechanical efficiency of the abductor musculature (3–5).

This approach also has shortcomings. The optimal extent of angulation is difficult to achieve. If the angle is too small, there will be insufficient improvement of hip biomechanics. If the angle is too large, the result will be excessive knee valgus, fixed pelvic obliquity, and impingement pain when the patient adducts the lower extremity to a neutral position. Also, lower extremity length discrepancy is not addressed by the pelvic support osteotomy.

Ilizarov combined pelvic support osteotomy with a second separate distal femoral varus lengthening osteotomy (Fig. 1A–E). Furthermore, he emphasized the importance of not only valgus but also extension at the proximal osteotomy site. This procedure is called the Ilizarov hip reconstruction (IHR) (2,7–9).

The proximal femoral subtrochanteric osteotomy (pelvic support osteotomy) as a treatment for instability of the hip has a long history in orthopedic surgery. This history has been outlined by Hass (3). The concept was to achieve support for the pelvis from the proximal femur. The resulting increased stability of the hip was due to actual support of the pelvis on the osteotomized proximal femur. In this type of reconstruction, the hip joint is not directly approached. Hass reported that in 1917, Lorenz described an oblique subtrochanteric valgus osteotomy in which the distal femoral segment was positioned into the acetabulum. In 1918, Von Bayer described a subtrochanteric osteotomy in which the acetabulum rested on the lesser trochanter. In 1922, Schanz described a lower subtrochanteric osteotomy at the level of the ischial tuberosity.

Milch (4,5,10,11) expanded the concept and popularized the pelvic support osteotomy in the United States during the mid-20th century. He advocated subtrochanteric valgus osteotomy to improve hip mechanics but cautioned against excessive valgus. Excessive valgus at the osteotomy leads to abutment of the proximal femur against the pelvis and even pelvic tilt when the patient tries to bring the involved extremity into a neutral abduction/adduction position. This is not desirable because it limits adduction and results in pain. Thus, there were two competing goals. Although excessive subtrochanteric valgus would improve hip stability, it would also cause valgus malalignment of the knee and abutment of the proximal femur against the pelvis as the patient attempted to bring the hip into a neutral abduction/adduction position. The compromise is a lesser degree of abduction than would be ideal to stabilize the hip and eliminate the Trendelenburg gait.

The optimal level for pelvic support osteotomy has been controversial. Although some authors have recommended a proximal osteotomy with insertion of the lesser trochanter into the acetabulum, others have preferred a longer proximal segment. We favor a more distal

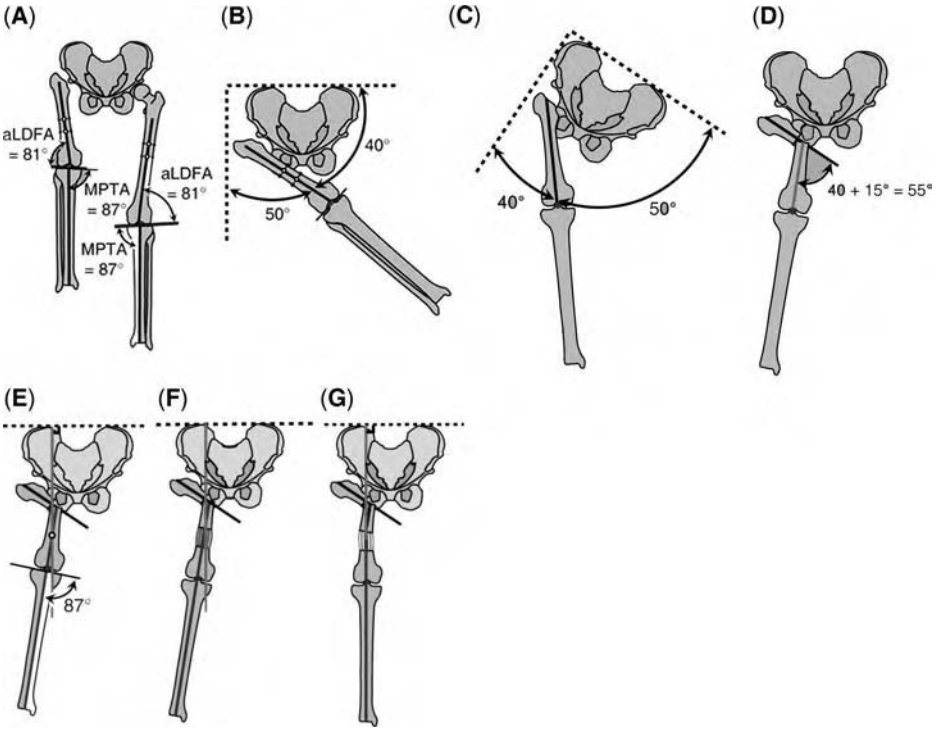


Figure 1 Schematic diagrams of surgical planning. (**A**) This is a schematic representation of a bipedal stance standing radiograph showing lower extremity length discrepancy. A block is placed under the short lower extremity to make the horizontal line of the pelvis parallel to the floor. The lower extremity length discrepancy is measured from this radiograph. (**B**) Maximum adduction is 50 degrees. The level of the poroxinal osteotony should be located at point where the femur crosses the ischial tuberosity. (**C**) This is a schematic representation of a single lower extremity stance standing radiograph showing 40° of adduction and Trendelenburg sign. The adduction of the femur relative to a horizontal pelvis line represents the pelvic drop associated with Trendelenburg sign. (**D**) Calculation of desired angle of proximal osteotomy. Fifteen degrees is added to the amount of adduction seen in the previous figure. (**E**) Determination of distal osteotomy level and degree of varus correction required. This is the intersection between the proximal and distal mechanical axis lines. The proximal mechanical axis is a perpendicular line to the horizontal line of the pelvis that passes through the proximal femur osteotomy. The distal mechanical axis line is a line from the middle of the talus that passes through the center of the knee. (**F**) The femur is first lengthened at the distal osteotomy. The varus re-alignment then follows. (**G**) Schematic diagram of a radiograph obtained at the end of distraction showing both proximal and distal osteotomies. Note lengthening and varus correction at distal osteotomy to achieve equal lower extremity lengths and 90° pelvic-mechanical axis. *Source*: From Ref. 6.

osteotomy, similar to that recommended by Schanz, as reported by Hass (3). The specific area of weight bearing is not absolute. Although this likely varies with the level of the osteotomy, the aim is to achieve a soft-tissue interpositional weight-bearing surface between the apex of the proximal femoral osteotomy and the pelvis. With a distal osteotomy, this weight bearing likely occurs at the inferior aspect of the pelvis, near the ischial tuberosity.

IHR (2,7–9,12–14) is a combination of the pelvic support osteotomy and a second distal femoral osteotomy to correct lower extremity length discrepancy and to realign the extremity (Fig. 1A–E). The proximal femoral osteotomy was designed to eliminate hip adduction. If the hip cannot adduct, Trendelenburg sign and gait cannot occur because the pelvis cannot drop. To achieve this, overcorrection of the valgus osteotomy is required. Based on experience, 15° of overcorrection was performed (2,6). This places the extremity in a fixed abduction position relative to the pelvis. This was one of the problems with previous pelvic support osteotomies and led to problems with the knee joint. To address this problem, Ilizarov introduced a second femoral osteotomy more distal than the first. This second femoral osteotomy was designed to realign the knee joint and to correct the lower extremity length discrepancy.

With the addition of this second femoral osteotomy, Ilizarov solved two of the problems that had not been previously addressed. Furthermore, Ilizarov emphasized extension of the

proximal femoral osteotomy to correct the fixed flexion deformity of the hip and to permit locking of the hip joint. The biomechanics of the hip are significantly improved by these corrections. The valgus alignment of the proximal femur positions the greater trochanter and the abductor muscle insertion laterally and distally. The lateralization increases the length of the abductor lever arm, while the distal shift tensions the previously redundant muscle. The valgus alignment also creates a fulcrum at the medial end of the pelvic support. The lower the level of the proximal osteotomy, the more medial the fulcrum. Medialization of the fulcrum decreases the abductor force needed to balance the weight of the body in single lower extremity stance. The net effect is a marked improvement in the function of the hip abductor mechanism. Extension of the osteotomy also contributes to this by stabilizing the hip in the sagittal plane during single lower extremity stance. If there is any fixed flexion deformity present, the pelvis unlocks itself from the ''pelvic support'' and the fulcrum is lost, destabilizing the hip and the abductor lever arm. Finally, equalization of lower extremity length discrepancy is also important to improve gait mechanics. With a lower extremity length discrepancy and without use of a shoe lift, the pelvis is tilted. This alters the abductor lever arm and leaves room for adduction of the femur on the pelvis in single lower extremity stance. Therefore, without equalization of lower extremity length, pelvic drop cannot be prevented (Fig. 2) (2,6).

CLINICAL EVALUATION

A thorough history of early childhood is needed. This will establish the etiology of the condition that may be dysplasia with chronic hip dislocation or early childhood sepsis. It is important to understand the source of the pain. Fatigue pain related to the inefficient hip biomechanics is better treated with this approach than arthritic intra-articular pain.

Physical examination should concentrate on evaluation of gait, leg lengths, and range of motion. A Trendelenburg sign confirms the weakness of the hip. Range of motion of the hip (flexion, flexion contracture, abduction, adduction, external rotation, and internal rotation in both supine and prone positions) should be carefully recorded. The patient with a ''flail'' hip that has a lot of motion that is not painful will be the best candidate for IHR. The patient's ability to adduct the hip combined with weakness of hip abduction strength is a key contributing factor to the Trendelenburg gait. With IHR, the adduction is eliminated and greater ability to abduct the hip ensues. In addition, the inability to lock the hip into extension also contributes to the abnormal gait. With IHR, extension at the proximal osteotomy helps the patient lock the hip into extension during gait.

A patient with a stiff painful hip will not be helped with this type of extra-articular reconstruction. In this case, an intra-articular operation such as an arthrodesis (15), total joint arthroplasty, or resection arthroplasty (2,6,10,11,16) may be needed. A resection arthroplasty can potentially be combined with an IHR (2,6,10,11,16). The resection arthroplasty can be used to improve mobility and relieve intra-articular pain but will create an unstable hip. IHR can then be used to stabilize the hip.

Gait analysis should be done. Typical findings are stance time asymmetry, and decreased ground reaction force. In other words, the patient spends less time on the abnormal lower extremity and puts less weight on it while walking (17).

Leg length discrepancy (LLD) and limb deformity should be analyzed (6) by looking at the patient with legs, iliac crests, and back exposed. Blocks should be placed under the affected leg until the pelvis becomes level and the back looks straight. This gives a good estimate of the total LLD, which may be a combination of proximal migration of the hip and shortening from growth arrest of the proximal femur. The same amount of block should be used for the anteroposterior standing radiograph (teleroentgenogram) of both lower extremities discussed next.

Preoperative radiographs include an anteroposterior radiograph of the pelvis, anteroposterior standing radiograph (teleroentgenogram) of both lower extremities, single lower extremity stance anteroposterior radiographs of both lower extremities, anteroposterior standing radiograph of the affected side, and maximum adduction cross-legged anteroposterior radiograph of the pelvis (patient placed supine with lower extremities adducted; involved hip flexed and adducted over top of uninvolved hip). Proximal migration is calculated from the difference between the distances from a transverse line through the sacroiliac joints to the tip of the greater trochanter of both sides. Lower extremity length discrepancy is calculated from the teleroentgenogram. The pelvic mechanical axis angle was measured between a line

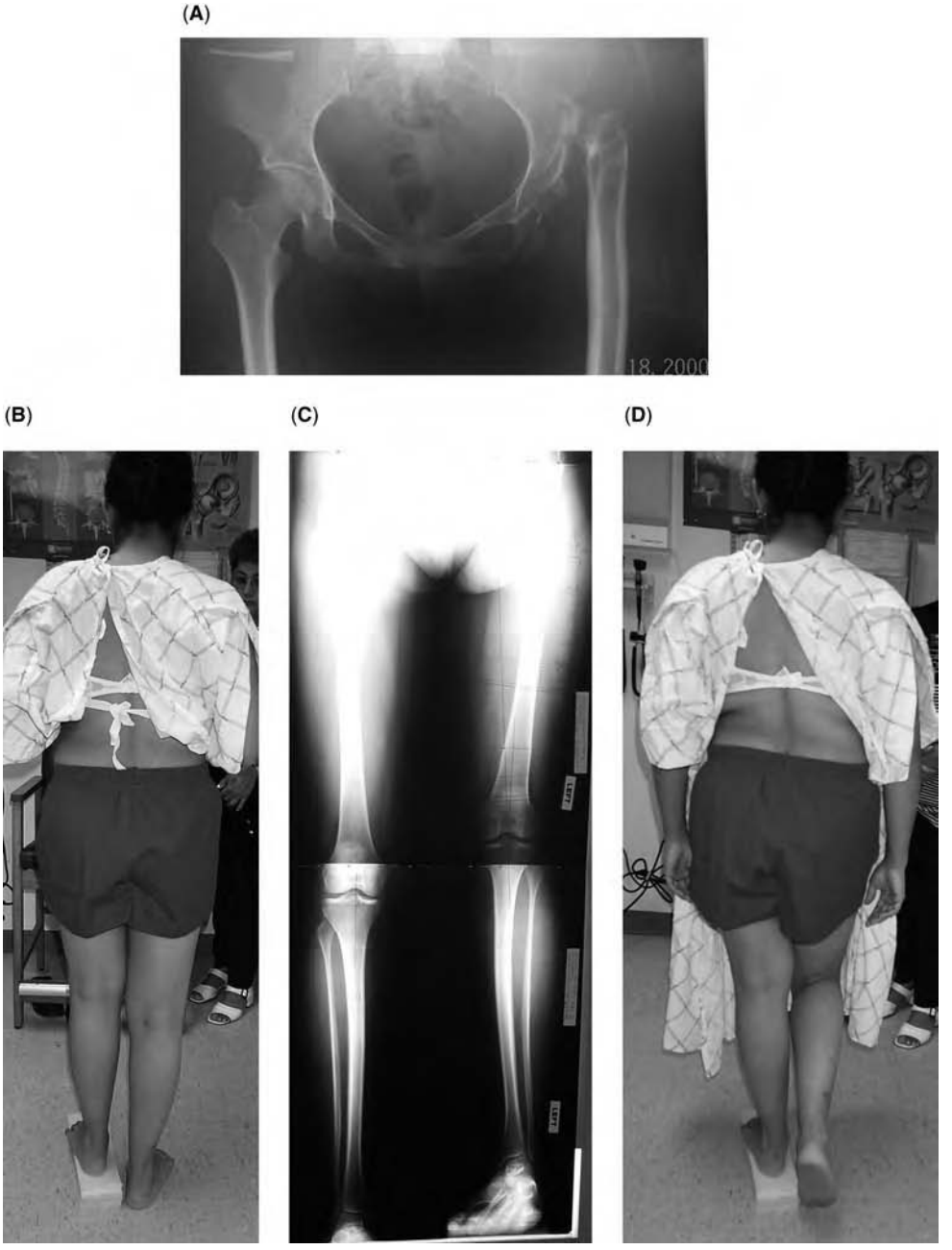
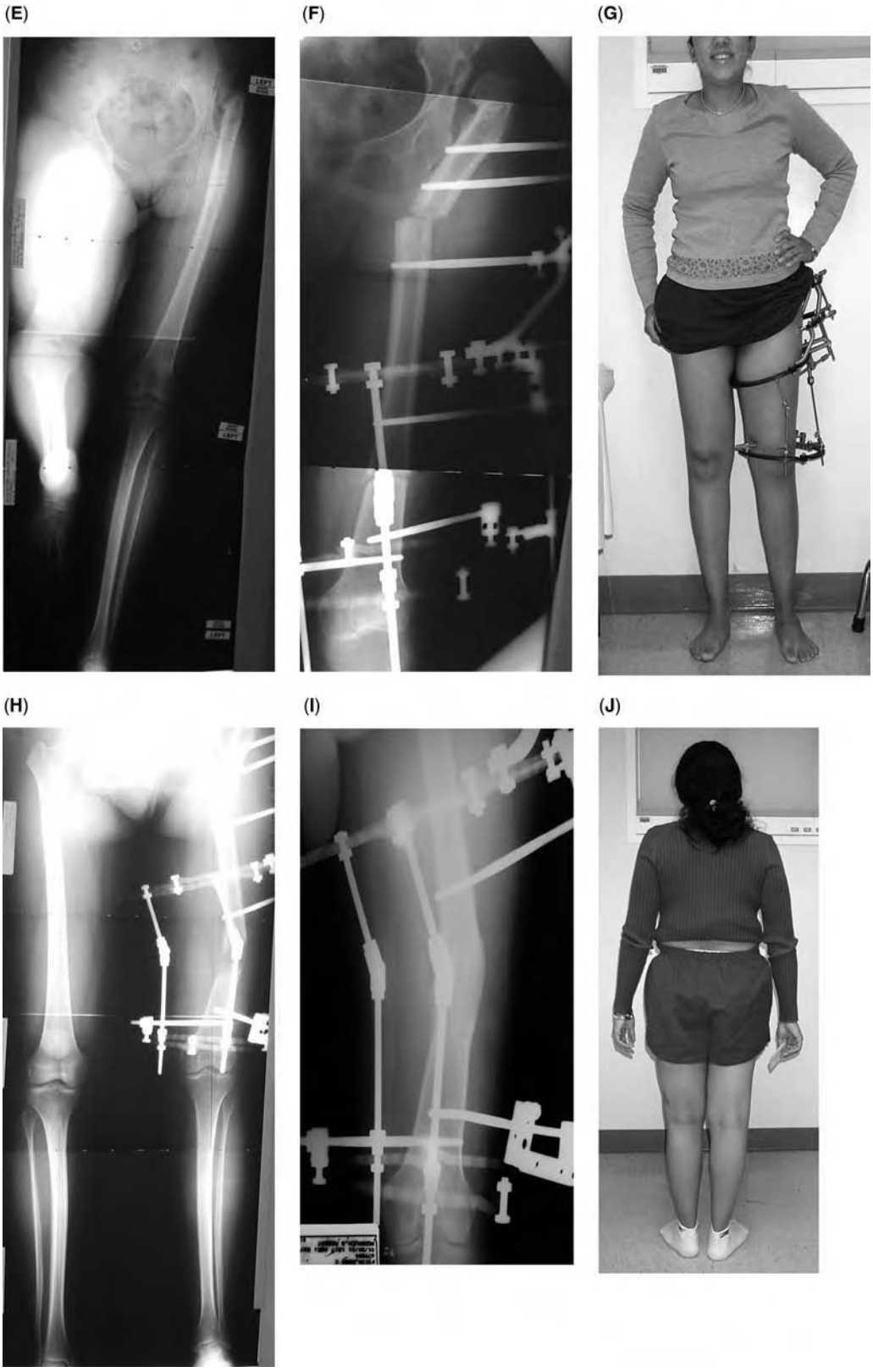


Figure 2 (*Continued on next page*) Images of a 22-year old female patient with residual sequelae of neonatal hip sepsis. (**A**) Preoperative AP Pelvis X ray showing an absent left femoral neck and head and proximal migration of the femur. (**B** and **C**) Preoperative back view and 51-in. bipedal standing X ray showing 5 cm leg length discrepancy. (**D** and **E**) Preoperative back view and 51 in. bipedal standing X ray showing positive Trendelenburg sign. (**F**) Immediate postoperative X ray showing acute valgus angulation at the proximal osteotomy and no displacement at the distal osteotomy. (**G** and **H**) Front view and 51 in. bipedal standing X ray at the end of the distraction showing normal alignment and equal leg lengths. (**I**) Close-up view of the distal femur osteotomy showing the lengthening and the varus correction. (**J**–**L**) Back view, 51-in. bipedal standing X ray, and front view 12 months after frame removal showing equal leg lengths and normal alignment. (**M**) Front view showing an absent Trendelenburg sign. (**N**–**P**) Clinical photos 12 months after frame removal showing hip motion and abductor strength.



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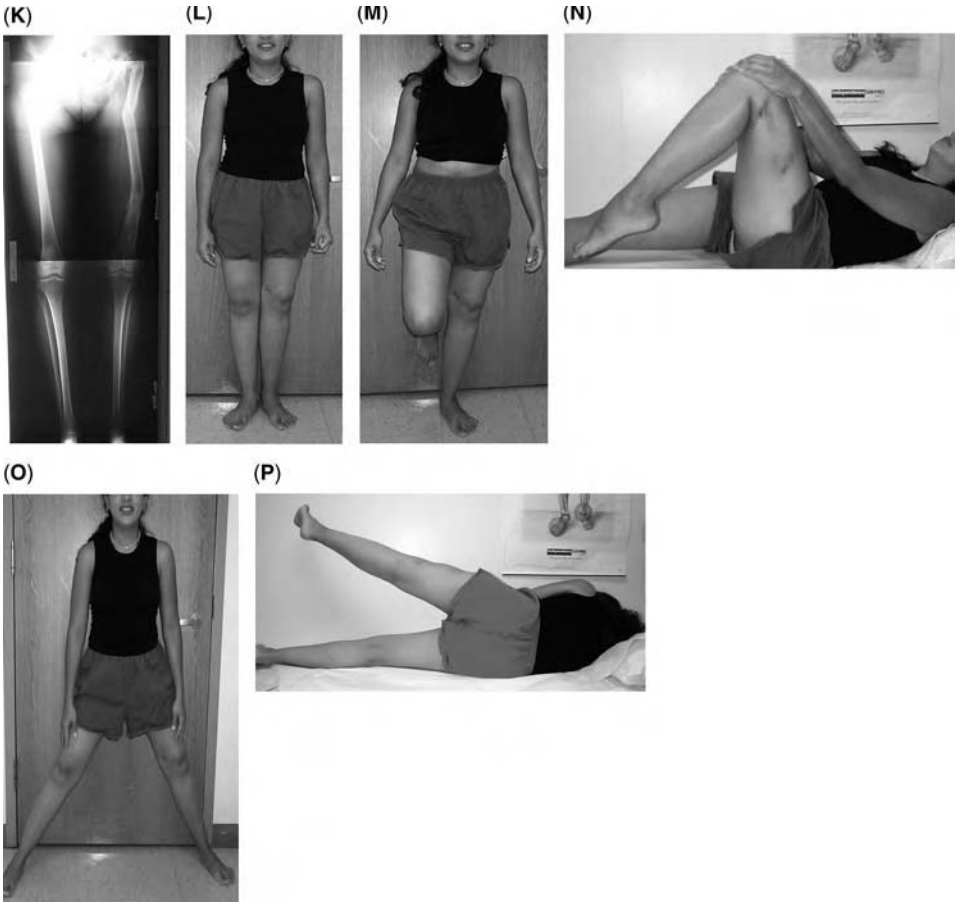

Figure 2 (*Continued from previous page*)

through the inferior end of the two sacroiliac joints and a mechanical axis line that was extended proximally from the ankle joint center through the knee joint center (2,6).

Trendelenburg gait is one of the hallmarks of this condition. Younger children often do not manifest a pelvic drop with gait because of their lower weight and shorter stride length. As they become adolescents and increase in height, lower extremity length, and weight, the pelvic drop becomes more apparent. With time, this is associated with increased pain and fatigue while walking, especially toward the end of the day. We found IHR to be very effective in eliminating Trendelenburg gait and sign in these patients. No other treatment method except arthrodesis has been able to successfully address this aspect of the problem. In contrast to arthrodesis, acceptable painless hip range of motion is preserved. If the patient presents with a painful stiff hip, arthrodesis may be a better option. IHR is best suited for the patient with an unstable hip that is mobile and associated with a lower extremity length discrepancy and Trendelenburg gait.

CLASSIFICATION

We classify patients by age, nature of their symptoms of pain, hip mobility, and radiographic appearance. Children with growth remaining will have greatest tendency to remodel the proximal osteotomy, which decreases its efficacy. Adolescents and young adults with no significant growth remaining are ideally suited for this surgery. Adults may be better served with a total hip replacement (THR) (18).

Hips with infantile hip sepsis are classified based on an AP pelvis X ray according to the Hunka classification (19): Type I, absent or minimal femoral head changes; Type IIa, deformity of femoral head with intact physis; Type IIb, deformity of femoral head with premature physeal closure; Type III, pseudarthrosis of the femoral neck; Type IVa, complete destruction of the capital

Table 1 Ideal Candidate for Ilizarov Hip Reconstruction

Factors Favoring Choice of IHR	Factors Against Choice of IHR
Fatigue pain or no pain	Painful hip with movement
Trendelenburg gait	Abduction contracture of hip
Mobile hip	Stiff hip
Absent femoral head and neck on X ray	Arthrosis of hip joint with head in acetabulum
Adolescent or young adult age	Young child (osteotomy will remodel)
LLD	Adult (may be better served with THR)

Abbreviations: IHR, Ilizarov hip reconstruction; LLD, leg length discrepancy; THR, total hip replacement.

femoral epiphysis with stable neck fragment; Type IVb, complete destruction of the capital femoral epiphysis with unstable small neck fragment; Type V, complete destruction of head and neck to the intertrochanteric line with dislocation of hip. While this is not a specific classification for chronically dislocated hips from dysplasia, it can be used in a similar fashion.

Hunka types IV and V are usually the most unstable (with proximal migration), most mobile, and least painful hips. These are the best candidates for IHR.

We classify hips into stiff or mobile. As mentioned, the mobile hip is best suited for this reconstruction. We classify the nature of hip pain with three types: no pain; fatigue pain; and acute pain with hip movement. Fatigue pain represents inefficiency of the hip abductors. Acute pain represents intra-articular mechanical pain. Patients with fatigue pain are best suited for IHR (Table 1).

TREATMENT OPTIONS (TABLE 2)

Reconstructive procedures for this difficult problem including trochanteric arthroplasty, hip arthrodesis, pelvic osteotomy, and femoral osteotomy have not been satisfactory. Full discussion of these is beyond the scope of this chapter.

Hip arthrodesis (15) may be the best option when there is arthrosis, pain, stiffness, and a reduced hip joint with a femoral head in the socket. However, hip arthrodesis may be avoided in patients who are good candidates for IHR. This is beneficial because hip arthrodesis has been associated with pathologic conditions of the ipsilateral knee, contralateral hip, and back. Callaghan et al.(15) reviewed 28 patients with hip arthrodesis at a mean of 35 years of follow-up. They observed ipsilateral knee pain in 16 (60%), back pain in 16 (60%), and contralateral hip pain in seven (25%). In addition, hip arthrodesis is technically most difficult when there is no femoral head and neck and where there is proximal migration.

Pelvic support osteotomy is not ideally suited for young children. Although there is no contraindication to performing it at this age, one should expect to have to repeat the pelvic support osteotomy at or near skeletal maturity. Because the amount of lower extremity length discrepancy requires two lengthenings in most cases or one lengthening and an epiphysiodesis, the pelvic support osteotomy should be reserved for the second lengthening (2). The femur can be lengthened at a younger age with extension of the external fixation to the pelvis. Extension of the external fixation to the tibia should also be considered if rotatory subluxation of the tibia is identified (20).

Table 2 Treatment Options

Surgical Option	Best Candidate
IHR	Adolescent; mobile hip; no pain, LLD, Trendelenburg gait; unstable hip; no femoral head or neck
IHR with femoral head resection	Adolescent; stiff hip; pain, LLD, Trendelenburg gait; stable hip; femoral head or neck and arthrosis
Hip arthrodesis	Same as above
Femur lengthening with spanning external fixation across the hip	Child; mobile hip; no pain, LLD, Trendelenburg gait; unstable hip; no femoral head or neck
Total hip replacement	Adult; stiff hip; pain, small LLD, Trendelenburg gait; stable hip; Femoral head or neck and arthrosis
Total hip replacement after hip distraction	Adult; stiff or mobile hip; pain, LLD from proximal migration, Trendelenburg gait; unstable hip; no femoral head

Abbreviations: IHR, Ilizarov hip reconstruction; LLD, leg length discrepancy.

IHR is most suitable for skeletally mature adolescents and young adults. Older adults may be best treated with THR. If the hip is proximally migrated, a gradual hip distraction can be used to pull the femur distally prior to insertion of a THR (18). Conversion of IHR to THR can be performed in later life if needed. Schiltenwolf et al. (21) reviewed their long-term follow-up of 24 patients with painful congenitally dislocated hips who underwent subtrochanteric valgus osteotomies without femoral head resection. Most patients maintained improved pain, gait, and hip abduction and extension. Four of these patients underwent THRs without difficulty. If necessary, the proximal femoral deformity could be straightened with an osteotomy and use of a long-stem prosthesis.

SURGICAL PLANNING

IHR involves a proximal femoral osteotomy for valgus and extension and a distal femoral osteotomy for lengthening and limb realignment.

The level of the proximal femoral osteotomy was determined from the maximum adduction anteroposterior cross-legged radiograph of the pelvis. The affected lower extremity and hip are maximally adducted over the normal side. Some flexion of the affected side is necessary to accomplish this. The osteotomy was performed at the level at which the femoral shaft crossed the ischium. This is similar to the osteotomy level proposed by Schanz, as reported by Hass (3). The amount of valgus angulation at the proximal osteotomy was determined by adding 15° to the femoropelvic adduction measured on the anteroposterior standing radiograph of the affected side rather than the maximum adduction anteroposterior radiograph of the pelvis (Fig. 1A–C). The overcorrection of 15° was determined based on experience and was used to compensate for fatigue of the abductor muscles. Extension at the proximal osteotomy site was equal to the amount of fixed flexion deformity of the hip. Finally, internal rotation at the osteotomy was determined at the time of surgery as the amount of hip external rotation that occurs with maximum adduction of the hip. As the hip is maximally adducted, external rotation of the femur occurs. This amount of external rotation of the hip is corrected with internal rotation at the proximal femoral osteotomy site.

The goal of the distal femoral osteotomy is to lengthen and realign the lower extremity (Fig. 1D and E). A perpendicular line from a horizontal pelvic line that passes through the proximal osteotomy should ultimately pass through the center of the knee and the center of the ankle. Distal femoral osteotomy is needed to achieve this. The level of the distal femoral osteotomy was determined based on the amount of valgus of the proximal femoral osteotomy. A paper tracing of the planned correction was performed. A perpendicular line was drawn to the pelvis through the region of the apex of angulation of the proximal osteotomy and extended distally. A distal line was drawn from the center of the ankle through the center of the knee joint and extended proximally. The intersection of these two lines is the level of the second center of rotation of angulation, which corresponds to the level of the hinges controlling the varus of the distal osteotomy. The level of the distal femoral osteotomy was either at this level or slightly more distal. In the latter situation, the distal femur will translate medially with varus angulation. The magnitude of the varus alignment of the distal osteotomy is equal to the magnitude of angulation measured on the preoperative drawing between the intersecting distal and proximal axis lines, described above.

SURGICAL TECHNIQUE

Preoperatively, an Ilizarov external fixation frame was constructed with two rings connected by a hinge at the anticipated level of the distal femoral osteotomy. The magnitude of varus angulation of the distal osteotomy was fixed in the hinges. A femoral arch was connected parallel to the proximal ends of the two rings. The frame was sterilized before surgery.

The patient is positioned on a radiolucent operating table with sheets placed under the sacrum to maintain a level pelvis and to avoid any rotation. Using an image intensifier, a 1.8 mm Ilizarov wire, and a surgical marking pen, a line is marked across the inferior edge of the two sacroiliac joints. This is called the *horizontal line of the pelvis*.

The involved extremity is then maximally adducted and crossed over the contralateral extremity. The involved hip flexes and adducts over the uninvolved hip. This places the

proximal femur in an adducted and flexed position. In this position, the femur will externally rotate because maximal adduction of the hip results in some flexion and external rotation. This is also a unique function of the pathological anatomy in this region. This rotation must be taken into consideration during pin placement.

The proximal femur is now in the position it will assume after the proximal osteotomy. This is the pelvic support position. Observe that the proximal femur cannot adduct further. This is important for elimination of the Trendelenburg gait. The first proximal pin is then inserted. Because the upper femur is in its postosteotomy position, it has moved relative to the overlying skin and subcutaneous tissues, such that a pin placed in this position will enter the skin over the underlying bone without tethering or displacing the skin after the osteotomy. The 6 mm threaded half-pin is inserted parallel to the horizontal line of the pelvis from the lateral side, parallel to the floor. The pin is connected to a free femoral arch. The arch is kept perpendicular to the floor with the extremity crossed, automatically imparting extension to the osteotomy. For more or less extension, the arch can be tilted in a posterior or anterior direction, respectively.

A second 6 mm threaded half-pin is inserted anterolaterally into the proximal fragment, parallel to the same femoral arch. The proximal arch is then fixed to the proximal femur at the correct angle. The extremity is then uncrossed and placed parallel to the other extremity with the patella facing forward, or anteriorly. The skin at the upper end of the thigh is now tented over the upper two pins. This corrects after the proximal osteotomy.

In the supracondylar region of the distal femur, a 1.8 mm Ilizarov wire is inserted parallel to the knee joint line. It is fixed and tensioned to the distal ring of the preconstructed frame. A 6 mm threaded half-pin is inserted from the posteromedial direction on the distal ring. A second half-pin is inserted from the proximal ring on the lateral side. The proximal femoral osteotomy is then performed. Through a 1 cm lateral incision, multiple drill holes are made using a 4.8 mm drill bit at the planned level of osteotomy. An osteotome is then used to complete the osteotomy. All procedures are performed under fluoroscopic guidance.

After completing the proximal femoral osteotomy, the distal femoral fragment is rotated until the distal wire is rotationally parallel to the first half-pin that was inserted. The distal fragment is then displaced medially by manipulating the arches before connecting them. Finally, the extremity is abducted to make the distal arch parallel to the proximal arch. In doing this, the proximal arch is also flexed to effect the extension correction. Two threaded rods with conical washers at each end are connected between the two arches. It is very important not to lose contact between the femoral segments during the medial translation. If contact is lost, the osteotomy will be unstable. One more 6 mm threaded half-pin is inserted from a posterolateral direction into the distal segment and another half-pin is inserted from the distal arch.

If there is any residual tenting of the skin by the proximal pins, a temporary third small pin is inserted, allowing removal and reinsertion of each of the proximal pins to allow skin tenting to equilibrate. The pin is removed, and a new incision is made at the level of the pin. The pin is reinserted in the previously drilled and tapped bone hole. This is preferable to creating long deep scars from pin site releases.

The distal femoral osteotomy is then made at the planned level in the same percutaneous manner described above. Next, the distal femoral wire is removed. A third 6 mm threaded half-pin is added proximally and distally in larger children and adults.

Some patients have rotatory instability of the knee associated with this condition. They demonstrate a tendency to subluxate the knee during lengthening. If this was recognized preoperatively, extension of the external fixation to the tibia with knee hinges is considered. The knee hinges should be located at the intersection of Blumenstat line and the posterior cortex of the femur. Two tibial half-pins are connected to a single half-ring suspended from the hinges with threaded rods.

Above is our technique described using the Ilizarov frame (2). IHR can also be performed using monolateral external fixation (13).

COMPLICATIONS AND EXPERIENCE

We published our experience (2) using IHR to treat eight consecutive patients with Hunka type IV and V hip deformities after infantile hip sepsis. The patients' mean age at surgery was 11.2 years (range, 7.8–14.2 years). All hips were unstable; the mean proximal migration was 3.4 cm

Table 3 Complications in Our Series

Patient No.	Complication	Treatment
1	Superficial pin infection	Oral antibiotic
	Knee stiffness, flexion to 20°	Soft-tissue release
2	None	None
3	Premature consolidation	Repeat osteotomy
4	Premature consolidation	Repeat osteotomy
	Knee stiffness	Soft-tissue release
5	Superficial pin infection	Oral antibiotic
6	Proximal migration of femur	Increase valgus at proximal osteotomy
	Knee subluxation	Extension of frame across the knee
7	None	None
8	Superficial pin infection	Oral antibiotic

Source: From Ref. 2.

(range, 1–5.5 cm). Proximal osteotomy was performed at a mean of 7.2 cm (range, 5.5–9.2 cm) distal to the tip of the greater trochanter. The mean valgus angulation was 44° (range, 16–70°) and the mean extension angulation 19 (range, −5–30°). The mean distal femoral lengthening was 5.7 cm (range, 4.4–7 cm), and the mean varus angular correction was 10° (range, 0–23°). The mean time in the Ilizarov frame was 4.7 months (range, three to seven months). Outcomes were clinically and radiographically evaluated. The clinical evaluation included gait analysis and the use of a modified Harris hip score.

At a mean follow-up of five years (range, 1.9–9.8 years), lower extremity length discrepancy improved from a mean of 4.6 cm (range, 0.6–6.4 cm) to 0.8 cm (range, 0–1.2 cm). Modified Harris hip score improved from a mean of 51 points (range, 21–67 points) to 73 points (range, 64–79 points) ($p = 0.007$). All extremities were well aligned with a pelvic mechanical axis angle of 89° (range, 84–94°). The mean mechanical axis deviation was 0.2 mm lateral (range, 1.6 mm medial to 2.3 mm lateral). Stance time asymmetry improved from a mean of 16% (range, 12–19%) to 5.4% (range, 1–12%) ($p = 0.0037$). The mean ground-reaction-force (second peak) improved from 102% of body weight (range, 94–107% of body weight) to 122% of body weight (range, 114–134% of body weight) ($p = 0.0005$).

Our conclusions from the study were that IHR can successfully correct Trendelenburg gait and simultaneously restore knee alignment and correct lower extremity length discrepancy. When performed on a young patient, remodeling of the proximal osteotomy and additional development of lower extremity length discrepancy is expected and the procedure may need to be repeated.

Our experience with complications is summarized in Table 3.

FUTURE DIRECTIONS

The early effective treatment of infantile hip sepsis and developmental dysplasia of the hip will prevent most cases of severe hip destruction and the need for the types of hip reconstruction discussed in this chapter. Effort at making a timely diagnosis and instituting aggressive treatment will helpful in this regard. This is a public health issue that needs to be improved particularly in developing countries.

Surgical techniques may be further improved by improving the accuracy of the deformity correction. The use of computer assisted surgery including computer navigation and the Taylor Spatial Frame (Smith and Nephew, Memphis, Tennessee, U.S.) hold promise in this arena. This is discussed in another chapter.

Monolateral external fixation is usually more comfortable than circular frames for the patient around the hip and femur. However, it is generally less versatile than circular external fixation. Inan and Bowen (13) reported on the use of monolateral external fixation for IHR. Further improvements in the stability and versatility of monolateral frames will expand its use for IHR.

As THR becomes more durable, its use in younger patients will increase. Improvements in prosthetic biomaterials will likely decrease wear debris and increase the longevity of THR.

The combination of hip distraction and staged THR (18) may then be considered in an even younger patient.

REVIEW OF LITERATURE

References	Methods	Results	Conclusions
Rozbruch SR, Paley D, Bhave A, Herzenberg JE. JBJS-A2005 (2)	Retrospective review of eight patients with late sequelae of infantile hip sepsis who underwent IHR	Corrected LLD; significant improvement in modified Harris hip score and gait analysis	Good procedure to improve gait, LLD, and deformity. Young patient will remodel osteotomy and may need repeat procedure
Choi H et al. Clin Orthop Related Research, 2005 (1)	Retrospective review of 43 patients with severe sequelae of infantile septic arthritis of hip; four hips treated with IHR	In subgroup of hips with absent head/neck, satisfactory results obtained in 5/10 who had trochanteric arthroplasty and 4/4 who had IHR	IHR is better suited than trochanteric arthroplasty for older children with absent head/neck where previous reconstructive surgery was unsuccessful
Kocaoglu M et al. Acta Orthop Scand 2002 (14)	Retrospective review of IHR used for treatment of neglected high dislocation of hip in 14 patients with mean age 20	Satisfactory outcome achieved. Pain relief and correction of LLD and deformity in all. 11/14 had negative Trendelenburg sign	IHR successful treatment for neglected hip dislocation from developmental dysplasia, paralytic hip dislocation, and proximal femoral focal deficiency
Inan M et al. JBJS-A 2005 (12)	11 patients with congenital dislocation of hip were treated with IHR. Gluteus medius muscle was evaluated with magnetic resonance imaging	Functional and painless hip achieved in all 11 patients. 5/11 had persistent Trendelenburg sign. Muscle volumes were restored 43–89% of normal	Results of Trendelenburg test correlated with restoration of muscle volume and age over 31. No correlation with muscle length. Overall good results but better in younger adults
Inan M, Bowen RJ Clin Orthop Relat Res 2005 (13)	Review of 16 patients who underwent IHR with a monolateral external fixator; mean age 25	Harris Hip score improved (). Four patients had persistent Trendelenburg sign. All achieved pain-free hip, no LLD, and normal knee motion	Monolateral external fixation worked well for IHR

Abbreviations: IHR, Ilizarov hip reconstruction; LLD, leg length discrepancy.

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27 Proximal Femoral Osteotomy

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INTRODUCTION

In the early part of the 20th century, proximal femoral osteotomy and realignment found popularity in adult patients for the treatment of hip osteoarthritis. In more recent times, this procedure has been reserved for young patients with symptomatic hip disease in whom total joint arthroplasty has offered an inadequate solution. Problems with rapid wear and loosening leading to early failure of joint replacements are well documented in this patient population. Intertrochanteric osteotomy found some utility in providing temporary relief of pain in this challenging group of patients. However, the current use of newer biomaterials with improved wear properties seems to offer an arthroplasty solution with longer-term benefits. These modern joint arthroplasty techniques have narrowed the indications of this once common procedure.

In contemporary orthopedics, proximal femoral osteotomy finds great utility for adults in the treatment of hip deformities. Common deformities include a varus or valgus neck shaft angle, rotational malalignments, bone defects, and leg length discrepancy. These deformities can be acquired as in the case of proximal femur fracture malunions and nonunions, or congenital (developmental) as in the cases of fibrous dysplasia, coxa vara, and developmental dysplasia. Regardless of the etiology, these patients with femoral deformity are at an increased risk for the development of pain and arthritis in the affected hip over time due to wear of the abnormal joint from malalignment. Once arthritis has been established, the problem is further complicated by the presence of femoral deformity. Standard hip replacement techniques and implants often cannot be used in the setting of proximal femoral deformity. One stage femoral osteotomy and total joint arthroplasty is technically demanding and associated with complications including intraoperative fracture and osteotomy nonunion (1–3). The benefits of early proximal femoral osteotomy to correct deformity are two-fold. In the deformed nonarthritic hip, the realignment will often reduce symptoms, if present, and will prevent further joint degeneration. In the deformed hip with arthritic changes, restoration of normal alignment will often decrease pain and improve function. Moreover, if the relief of symptoms is incomplete and the patient goes on to require hip replacement surgery, then the arthroplasty procedure has been simplified by the restoration of a more normal anatomy.

CLINICAL EVALUATION

The patient history will help differentiate acquired from congenital (developmental) deformities. In general, acquired deformities are easier to correct acutely as the surrounding soft tissues have a memory of their preinjury length. If the correction of a congenital deformity involves significant stretching of the soft tissues, then this may warrant a gradual approach.

Patients with underlying metabolic bone disorders must be identified and optimized prior to operative intervention.

A thorough examination is needed to uncover all aspects of the deformity. The hip, knee, and ankle joints are examined looking for deformity and range of motion. Hip joint contractures may be accommodated through the osteotomy. For example, if a patient has a hip flexion

contracture, then an extension osteotomy of the proximal femur will reposition the femur with respect to the pelvis and will decrease the lumbar spine compensation. The rotational profile of the lower extremity, including hip internal and external rotation and thigh foot axis, is documented. Femoral retroversion and anteversion may cause hip pain and lead to an altered gait. Limb length discrepancy is measured using blocks to level the pelvis and later with radiographs. The quality of the patient's skin and position of previous incisions are noted to assist with safe surgical approaches. Clinical signs of infection are sought.

Laboratory studies may be indicated. In the case of a bony nonunion, an infection must be assumed until proven otherwise. Standard white blood cell count, erythrocyte sedimentation rate, and C-reactive protein are obtained in these cases. If osteonecrosis is present, then an investigation of the etiology may be indicated.

IMAGING STUDIES

The evaluation relies heavily on appropriate, high quality radiographs. A standing anterior-posterior (AP) pelvis X-ray is ideal for measuring the neck-shaft angle and assessing the hip joint integrity. The cross-table lateral X-ray will provide information about sagittal plane deformity. The erect bipedal 51-in. radiograph that extends from the top of the iliac crests to below the ankle joints gives excellent information about deformity and leg length discrepancy. Long X-rays will help identify compensatory deformities, which are common in patients with chronic deformities of the hip. A distal femur deformity may have developed to compensate for the proximal deformity; the knee joint may have developed a contracture or the tibia and ankle may have developed compensatory deformities. These other compensatory deformities and contractures may be missed if the initial evaluation is focused only on the hip. Failure to recognize the presence of the secondary deformities can lead to poor planning and results. All implanted hardware and prostheses, and their relationship to the proximal femur should be noted.

Advanced imaging can be helpful and should be used as needed. Computed tomography (CT) scan of the hip can confirm the presence of a nonunion. CT scan version studies will help evaluate femoral anteversion and retroversion. Magnetic resonance imaging (MRI) is an excellent means of assessing the hip joint condition and detecting areas of osteomyelitis, osteonecrosis, and early arthrosis. One should keep in mind the sensitivity of MRI and its tendency to overestimate the extent of bony infection.

INDICATIONS

The basis for performing a proximal femoral osteotomy varies. In the presence of deformity, the goal is to correct the deformity and in doing so realign the hip and lower extremity. This may include frontal, sagittal, and rotational corrections and perhaps even lengthening through the osteotomy. Proximal femoral osteotomy is commonly utilized in the treatment of nonunions of hip fractures. Both femoral neck and intertrochanteric fracture nonunions respond positively to valgus-producing realignment osteotomies. Malunions of hip fractures, including intertrochanteric type and unreduced slipped capital femoral epiphysis (SCFE), are other common indications for osteotomy. Common congenital (developmental) conditions that benefit from osteotomy include coxa vara, Shepard's crook deformity from fibrous dysplasia, femoral anteversion, developmental dysplasia of the hip, and hip flexion contracture from achondroplasia. Less frequently, proximal femoral osteotomy is performed in adults for the treatment of hip arthritis and osteonecrosis.

Femoral Neck Nonunion

In this instance, the fracture fails to heal despite an adequate blood supply. Weight-bearing forces across a vertically oriented fracture line produce shear stresses at the fracture site that favor the production of fibrous tissue. A Pauwels valgus producing intertrochanteric osteotomy reorients the fracture site into a more horizontal position. Axial loading in this situation encourages osteogenesis and fracture union. Authors have reported great success with this approach to femoral neck nonunion (Fig. 1) (4,5). Although a fixed angle blade plate has

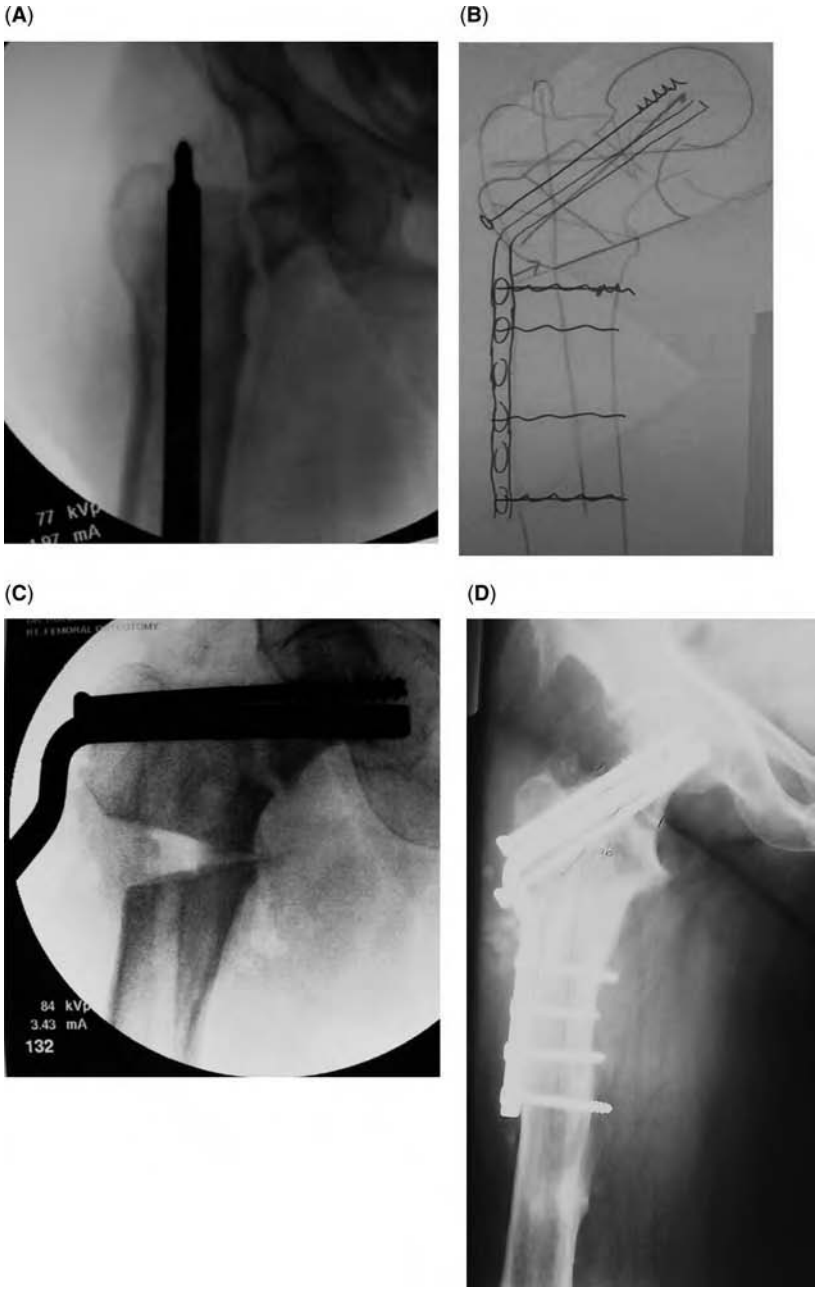

Figure 1 (**A**) Nonunion of femoral neck stress fracture. (**B**) Preoperative planning. This helps plan translation and osteotomy location. (**C**) A compression lag screw is used across the nonunion site. A wedge has been removed from the intertrochanteric region. When the plate is secured to the femoral shaft, the cut surfaces will be well opposed and excellent bony contact obtained. (**D**) A lateral closing wedge osteotomy was created at the level of the lesser trochanter and stabilized with a fixed angle blade plate. A valgus orientation of the proximal femur and improved orientation of the femoral neck nonunion have been achieved.

traditionally been used to stabilize this osteotomy, sliding hip compression screw has been used successfully as well (4,6).

Pertrochanteric Nonunion

Pertrochanteric hip fractures typically do not disturb the blood supply to the femoral head and tend to heal predictably. Nonunions of this common fracture pattern are usually the result of a

combination of varus malalignment with loss of fracture compression and inadequate stability of fixation. Treatment is aimed at correcting the varus neck shaft angle to a normal or slight valgus orientation and improving the stability at the fracture site often with a fixed-angle device. Often, repair of the nonunion with revision of fixation will treat this problem (7,8). Alternatively, osteotomy can reliably restore alignment, length, and heal the nonunion (Fig. 2) (9).

Intertrochanteric Malunion

When the fracture collapses into varus angulation and then goes on to bony union, a malunion results. The hallmark of this malunion is a varus neck-shaft angle with shortening of the ipsilateral femur, shortening of the abductor musculature or lever arm, and often trochanteric-pelvic

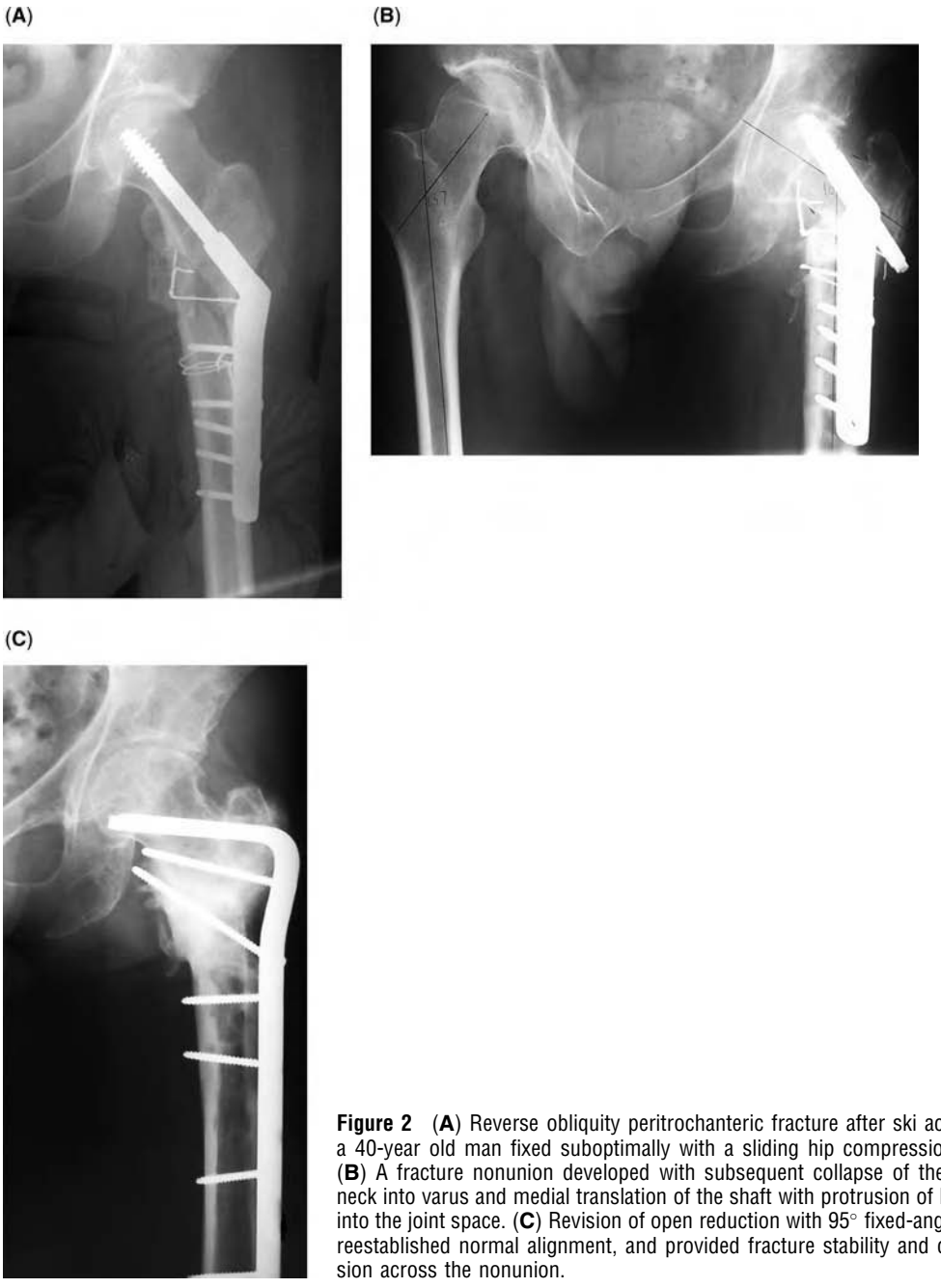

Figure 2 (**A**) Reverse obliquity peritrochanteric fracture after ski accident in a 40-year old man fixed suboptimally with a sliding hip compression screw. (**B**) A fracture nonunion developed with subsequent collapse of the femoral neck into varus and medial translation of the shaft with protrusion of hardware into the joint space. (**C**) Revision of open reduction with 95° fixed-angle device reestablished normal alignment, and provided fracture stability and compression across the nonunion.

abutment and a Trendelenberg gait with poor hip motion. This patient is at an increased risk of the development of hip arthritis. Intertrochanteric osteotomy serves to realign the hip joint, restore normal abductor mechanics, and reestablish equal leg lengths.

Slipped Capital Femoral Epiphysis Malunion

SCFE is a common fracture seen in the adolescent population. In many cases in-situ pinning of the displaced fracture is performed because this provides a reduced risk of osteonecrosis. When a displaced slipped epiphysis heals in situ, a fracture malunion is the result. This deformity is often characterized by coxa vara, femoral shortening, and retroversion of the femoral neck with a significant loss of hip motion. A valgus rotational proximal femoral osteotomy serves to correct the varus and reestablish normal rotation both of which reorient the femoral head in the acetabulum theoretically offering some protection from the development of arthritis. This procedure will also equalize limb length and abductor tension helping to normalize gait (Fig. 3). Maussen found that in patients with significantly displaced chronic SCFE treated with intertrochanteric osteotomy, hip degeneration was not prevented (10).

Fibrous Dysplasia

The Sheppard's crook deformity of the proximal femur has long been recognized as a hallmark of fibrous dysplasia. Repeated microfractures of the femoral neck (Fig. 1A) heal in a progressively more varus orientation. Significant shortening of the femur, trochanteric-pelvic abutment, and shortening of the abductor lever arm occur concomitantly. Rotational deformity may also be present. Patients complain of limb shortening, hip stiffness, and the inability to abduct the lower extremity, which is especially troublesome for women of childbearing age. Pain may be present as well. These patients are at risk for progression of the deformity, fracture of the femoral neck, and joint degeneration. Valgus-producing proximal femoral osteotomy serves to prevent progression of the deformity and the development of a fracture, reestablish a more normal femoral head-acetabular relationship, lengthen the extremity, tension the abductors, and greatly improve hip range of motion in abduction (Fig. 4).

Developmental Dysplasia of the Hip

Adults with hip dysplasia often have both acetabular and femoral deformity. The femoral neck assumes a valgus and anteverted orientation while the acetabulum is shallow with varying degrees of uncovering of the femoral head ranging from mild to a frank dislocation. In select patients, surgery is indicated to improve femoral head coverage or reduce the hip joint. A varus-producing proximal femoral osteotomy with derotation of the anteverted neck will improve femoral head orientation. Often this is combined with a periacetabular osteotomy to improve superolateral and anterior head coverage.

Proximal Femoral Lengthening

Although most femoral lengthening procedures are preformed through distal metaphyseal or diaphyseal osteotomy sites, osteotomy of the proximal femur provides another good option. When a patient has a distal femoral nonunion and shortening, then a proximal osteotomy offers an excellent osteogenic site (Fig. 5). Similarly in cases of distal femoral nonunion with bone loss, proximal osteotomy allows for bone transport to fill the defect (Fig. 6) (11).

Osteoarthritis and Osteonecrosis

In cases of arthritis and osteonecrosis with focal defects in the weight-bearing area, the goal of the femoral osteotomy procedure is to alter the contact point across the articular cartilage during weight bearing. When there is focal arthritic change or necrosis, the compromised area of the femoral head may be rotated away from the maximum weight-bearing area. This can be accomplished with combinations of a varus or valgus and flexion or extension osteotomy. This will decrease stress and pain at the degenerated area of articular cartilage. This area of damaged cartilage has been shown to undergo a reparative process through which new

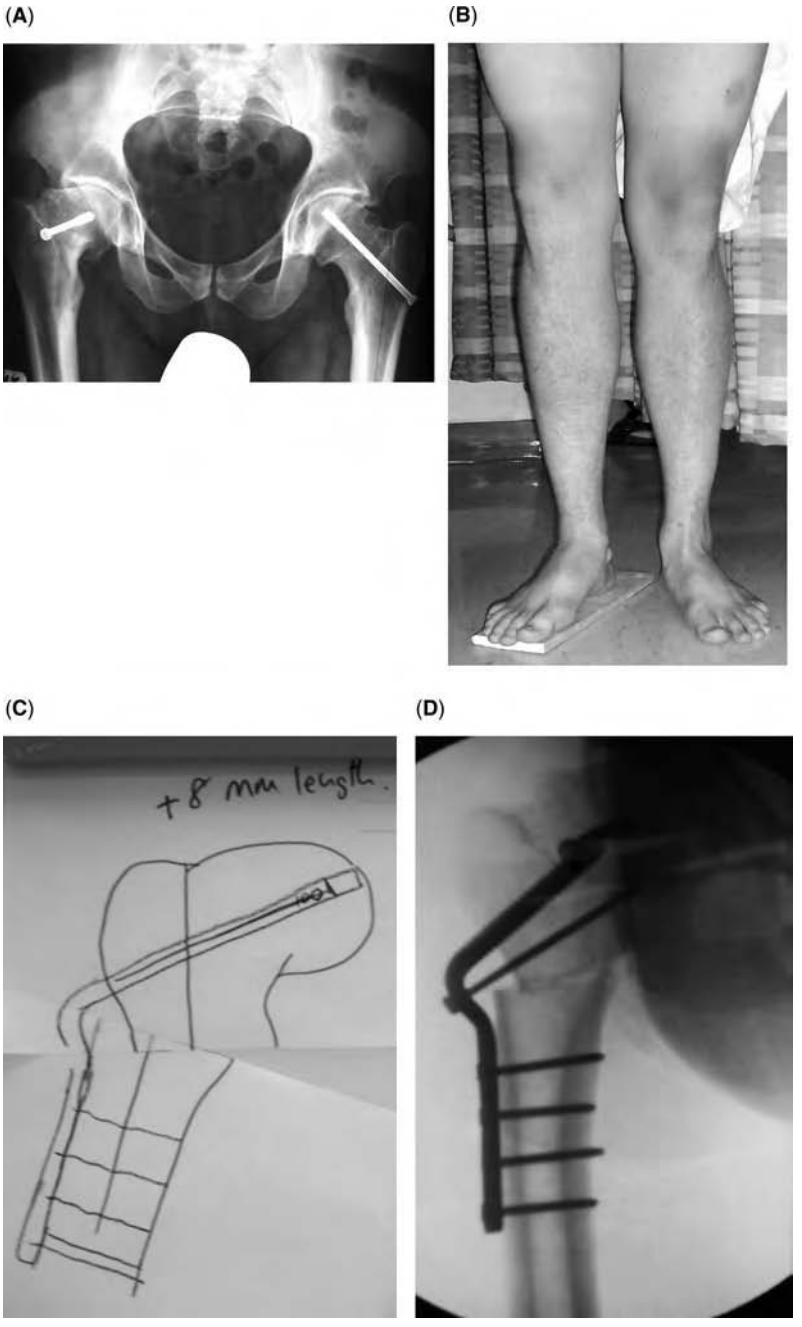

Figure 3 (**A**) The right hip shows a varus and shortening deformity typical of a SCFE malunion. (**B**) This patient had shortening and external rotation deformity from retroversion of the femoral neck and head. (**C**) This diagram is a preoperative planning cutout of a valgus producing osteotomy. In this example, a wedge of bone was not removed and 8 mm of length will be obtained with the correction. Note the medial translation of the proximal fragment. (**D**) A valgus rotation osteotomy has corrected this multiplaner deformity and improved hip biomechanics.

collagen is created (Fig. 7). A varus or rotational osteotomy with or without vascularized bone grafting has been used successfully in this manner (12–15).

TREATMENT OPTIONS

Proximal femoral osteotomy can be stabilized using either internal or external fixation. Deciding between these devices is often the surgeon's preference; however, each has distinct

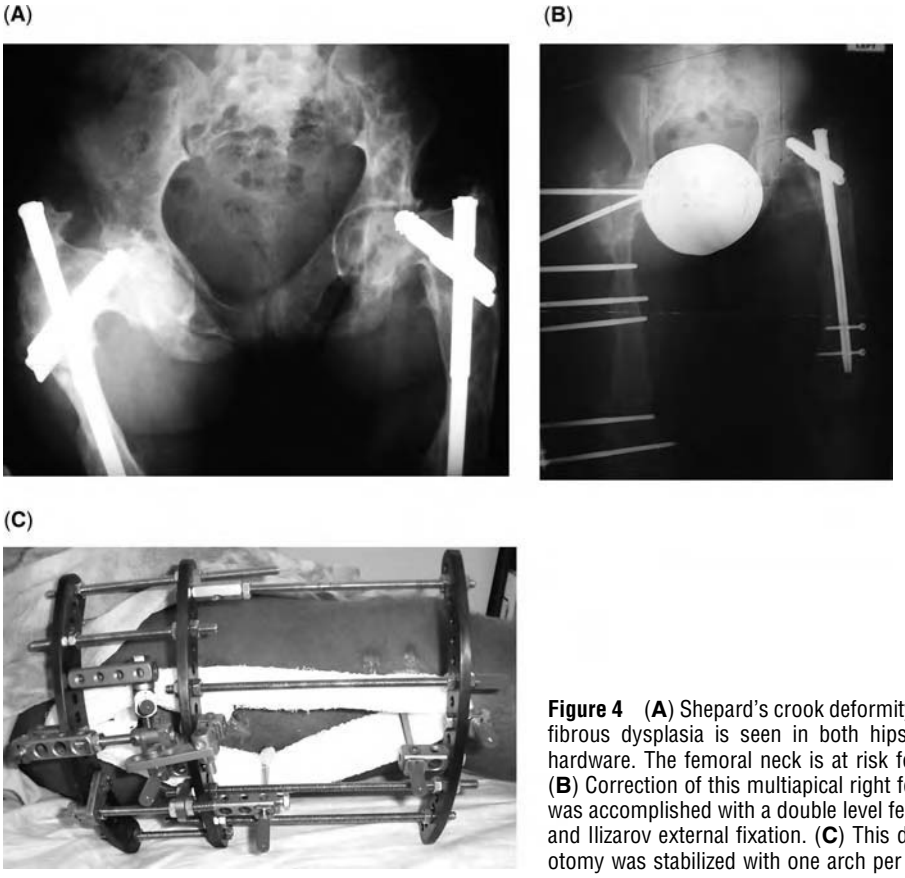

Figure 4 (**A**) Shepard's crook deformity associated with fibrous dysplasia is seen in both hips with ineffective hardware. The femoral neck is at risk for frank fracture. (**B**) Correction of this multiapical right femoral deformity was accomplished with a double level femoral osteotomy and Ilizarov external fixation. (**C**) This double level osteotomy was stabilized with one arch per bone segment.

advantages. In the presence of infection, external fixation provides stability without permanent internal hardware. When the skin quality is very poor or the patient has compromised ability to heal a large incision, external fixation with a percutaneous osteotomy provides a great solution. When multiple osteotomies are being performed in the femur simultaneously, external fixation can stabilize the entire bone and avoid a huge incision (Fig. 4). External fixation is optimal when performing a bony lengthening or bone transport through a proximal femoral osteotomy (Figs. 5 and 6). External fixation allows for postoperative adjustability including the correction of residual deformity. Most patients are allowed weight bearing as tolerated on ambulation after external fixation surgery.

Internal fixation has the advantage of avoiding the discomfort of external pins and allowing the osteotomy to be made proximal to the lesser trochanter where half pins cannot be bicortical. In instances where hardware needs to be removed, an open approach to the proximal femur is often needed. In these instances, inserting a blade plate does not add to the morbidity of the procedure and provides a cure with one surgery (Fig. 1). Internal hardware is well tolerated around the hip, and wound closure is rarely difficult. Although we do not routinely use intramedullary nails to stabilize a proximal femoral osteotomy, this is a viable technique.

The proximal osteotomy can be made either in an intertrochanteric or subtrochanteric location. Some authors speculate that a subtrochanteric location is associated with a higher risk of osteotomy nonunion when compared with an intertrochanteric site (4,6). A subtrochanteric site simplifies correction of multiplanar deformities, is necessary for lengthening, and does not disturb the relationship of the greater and lesser trochanter. When using an external fixator, the location of the osteotomy needs to be subtrochanteric because the pins have to be distal to the lesser trochanter to avoid intracapsular pin placement. In general, we prefer subtrochanteric osteotomy and have not had difficulty obtaining union.

Radiographic planning is a crucial portion of surgical preparation. The neck-shaft angle is measured on both sides using the AP pelvis X-ray. Anatomic and mechanical axis planning

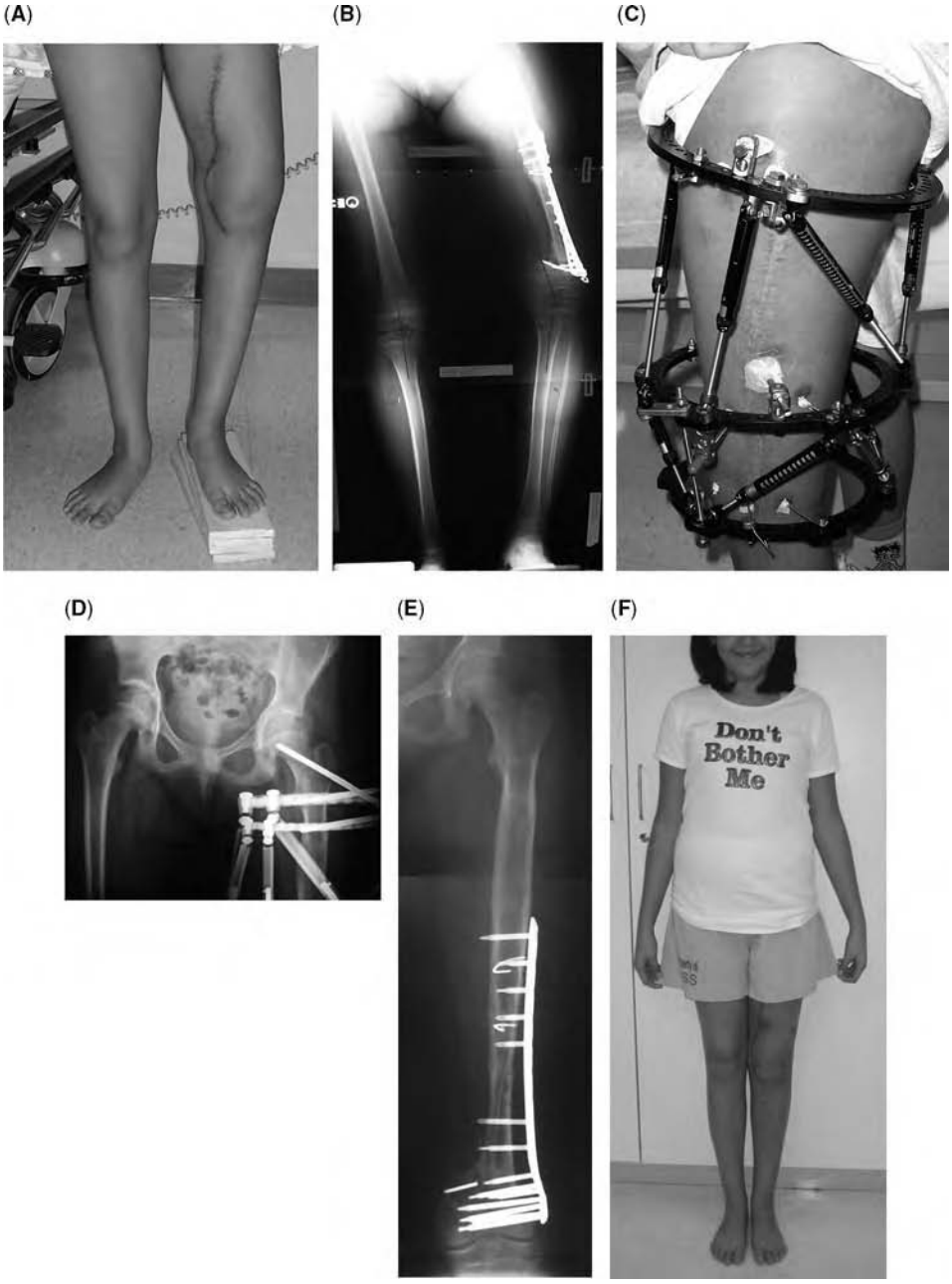

Figure 5 (**A**) This 11-year-old female underwent resection of distal femoral osteogenic sarcoma and reconstruction with a vascularized fibular graft and internal fixation. She has a mobile nonunion in 30° of varus with failed hardware and 7 cm of shortening. (**B**) This 51-in. erect, bipedal radiograph demonstrates persistent nonunion of the distal graft-host junction along with varus deformity and shortening. (**C**) The patient underwent Ilizarov reconstruction of the limb with a two level TSF. Lengthening was done at the proximal level and nonunion repair and correction of deformity was performed at the distal level. (**D**) Regenerate formation of 7 cm can be seen at the lengthening site in the proximal femoral diaphysis. A half pin was placed up the femoral neck to prevent a stress fracture in that area. (**E**) After frame removal, the patient sustained a fracture above the nonunion through the old free fibula graft. This was fixed with a percutaneously placed submuscular locking plate. (**F**) Six months after removal of external fixator, the patient is pain free and walking without support. The deformity and shortening have been corrected.

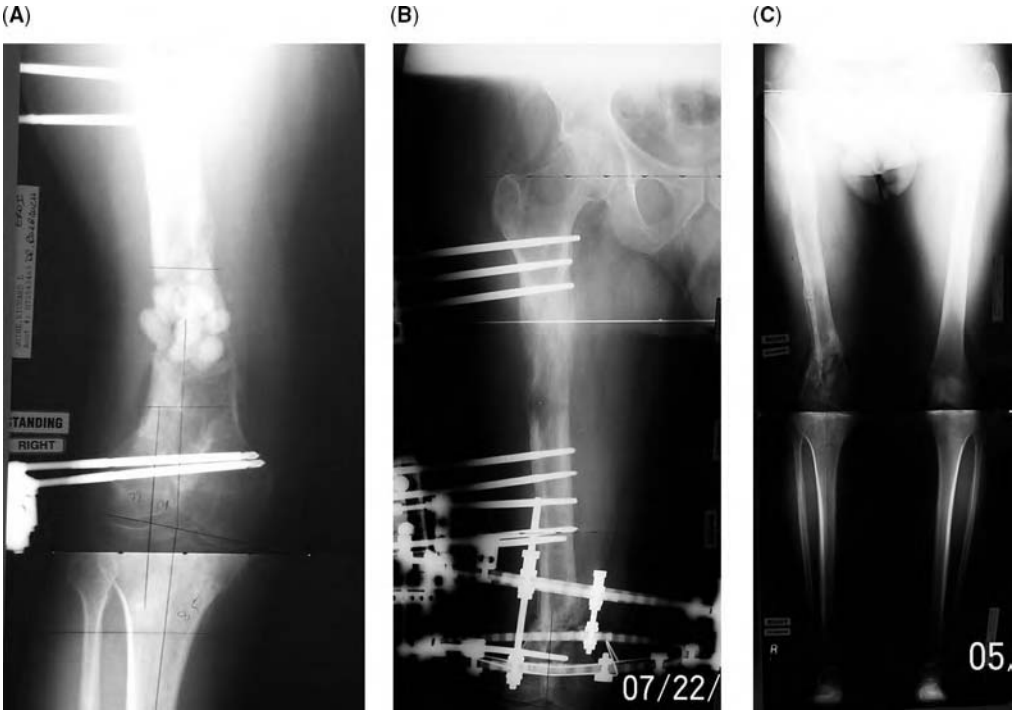

Figure 6 (**A**) Infected nonunion of the distal femur with bone loss initially treated with spanning external fixation and antibiotic beads. This patient has a bone defect of 4 cm and a LLD of 4 cm resulting in an 8 cm total longitudinal deficiency. Also note valgus deformity. (**B**) Proximal femoral osteotomy was performed and bone transport and lengthening were achieved. (**C**) Successful bone transport with healed nonunion, equal leg lengths, and no deformity.

on the long standing X-ray will help localize the apex and magnitude of the deformity. Because the apex of the deformity is not at the level of the osteotomy in most cases, compensatory translation is needed. Typically, medial translation of the proximal fragment is combined with a valgus osteotomy (Figs. 3 and 7) and lateral translation of the proximal fragment is combined with a varus osteotomy. Leg length discrepancy is measured and an estimate can be made of how the proposed surgery will affect leg length. This will also help determine if a lengthening procedure is indicated. Small amounts of length can be obtained with acute valgus producing correction. The extent of expected lengthening, the fit of the plate, and the necessary translation can be planned with cutout drawings (Figs. 1 and 3).

Preoperative planning demands a review of all the information obtained from the history, physical examination, and imaging studies. The radiographically measured degree of deformity is correlated with the clinical examination. A patient may have femoral retroversion, which produces an external rotation deformity, but walk with a normal foot progression angle because he has internal tibial torsion. Correcting the femoral malrotation would uncover the tibial deformity and lead to an internally rotated foot progression angle. One needs to decide in advance whether to disregard the femoral retroversion or correct it along with the tibial torsion. This example reminds us that the goal is to optimize the patient's function, devise a good plan that may call for compromise, and not to simply treat the X-ray. If using a blade plate, then determine the optimal position for the blade in the femoral neck on X-ray. Templates are available to simplify this process. Plan the level of the osteotomy based on bone quality and presumed healing capacity and location of the center of rotation of angulation of the deformity. Consider the best approach with regard to skin condition. Have the correct instrumentation available to remove old hardware. Involve the patient and the family in the decision process and provide them with realistic expectations from the surgery.

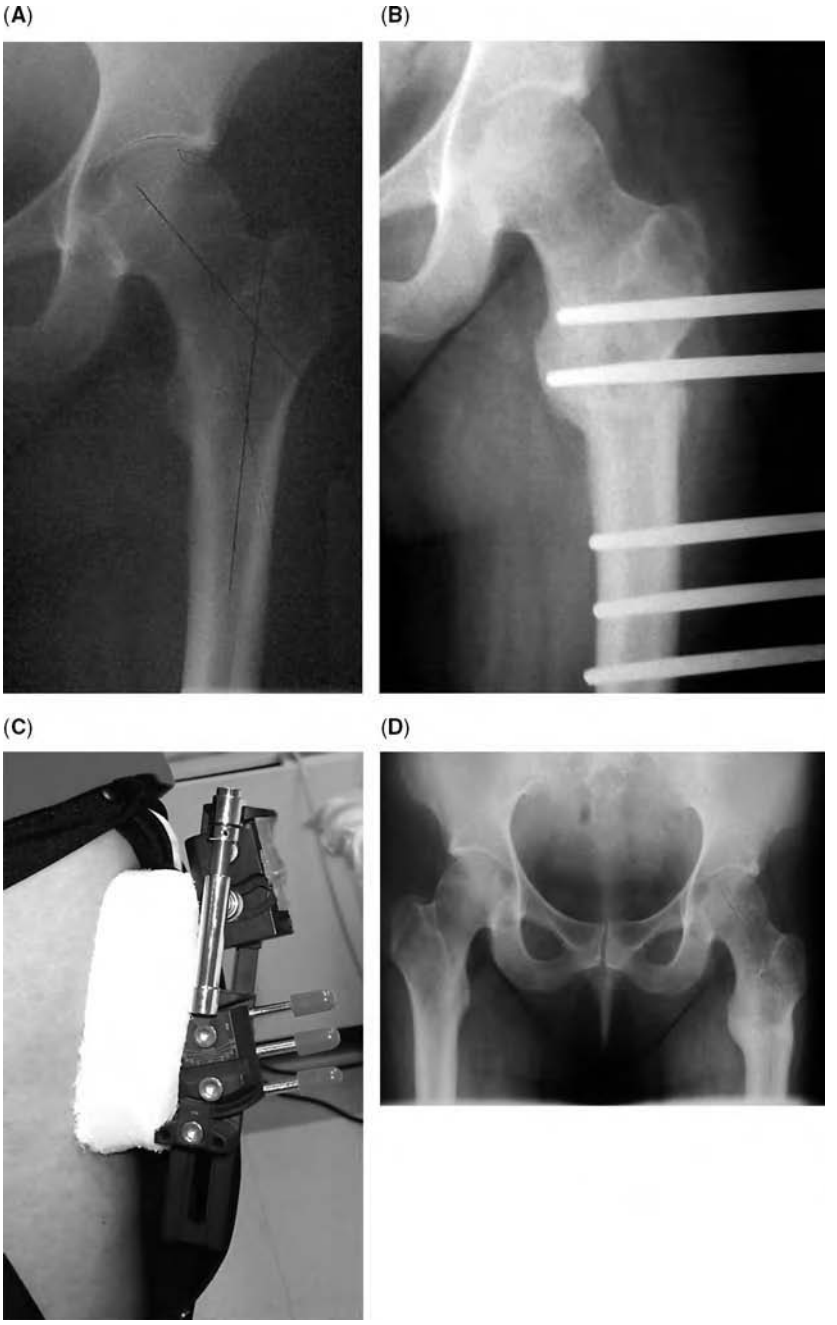

Figure 7 (**A**) This 46-year-old woman had painful articular cartilage lesion of the superolateral aspect of her femoral head with a normal neck-shaft angle. (**B** and **C**) A valgus-producing proximal femoral osteotomy was performed and stabilized with an external fixator. The external device allowed for precise control over the final orientation of the femoral head. The lesion was rotated out of the weight-bearing area of the hip. Although this correction was performed acutely, gradual correction remains an option with this versatile fixator. Note the medial translation of the proximal fragment. (**D**) Six-month follow-up reveals a healed osteotomy with a laterally shifted articular lesion.

SURGICAL TECHNIQUE

Intraoperative Details

Have available in the room: the patient's X-rays, a sterile goniometer, Steinman pins to mark and judge rotation, C-arm Fluoroscopy, a radiolucent operating room (OR) table, and a broken

hardware removal set. We prefer the Jackson flat table. The patient is positioned supine with a bump under the ipsilateral buttock.

Internal Fixation

The implant is typically a fixed angle device such as a 95° or 130° angled blade plate (Synthes, Paoli, PA), which is inserted through an open approach with an acute correction. The lateral approach to the proximal femur is utilized. If correcting rotation, I will place a Steinman pin into the proximal femur posteriorly at the level of the lesser trochanter. A second pin is then placed in the distal femur at an angle that mimics the deformity such that when the deformity is corrected the pins will be parallel. The guide wire for the blade plate is inserted into the femoral neck and head in the predetermined ideal location. The seating chisel is advanced over the wire taking care to enter the bone at the ideal angle in the sagittal plane. Any planned flexion or extension would be set at this time. For valgus-producing osteotomy, the blade plate can be inserted before completing the osteotomy. The plate is then used to help obtain the correction. For varus-producing osteotomies, the bone is cut before the implant is inserted (due to impingement of the plate on the femoral shaft) and the seating chisel is used to help reduce the proximal fragment. The osteotomy is typically made at the inferior aspect of the lesser trochanter. With a valgus-producing osteotomy, a small wedge of bone can be removed to improve bone contact at the osteotomy site (Fig. 1). More length can be obtained if no bone is removed (Fig. 3). A compression device can be utilized and the screws are then inserted through the side plate. The wound is closed in layers over a drain.

Figure 8 Clinical picture of the boy in whom hip deformity was corrected with a two arch Ilizarov frame and a percutaneous osteotomy.

External Fixation

An external fixator is mounted percutaneously and can be combined with a percutaneous osteotomy. Although gradual correction of deformity is possible, an acute correction is more typical. When using external fixation, all half pins are inserted percutaneously. The femur is predrilled bicortically and the pins are hand-inserted to reduce the risk of bone necrosis. Each bony segment is stabilized with two to three pins. One half pin can be inserted centrally into the femoral neck and head, taking care not to enter the joint (Fig. 5D). Additional one to two pins are placed around the level of the lesser trochanter remembering that bicortical pins cannot be used proximal to the lesser trochanter. Two to three pins are placed in the shaft of the femur for stability. If using Ilizarov frame, then one femoral arch is attached to each segment to mimic the deformity. A percutaneous osteotomy is made and the arches are manipulated to place the femur in the desired alignment. If the frame truly mimicked the deformity, then the reduction will be obtained by making the arches parallel. These are then stabilized with one another while the reduction is held manually (Figs. 4 and 8). The same correction can be obtained using a monolateral fixator (Fig. 7). Again, the frame is mounted in the deformed position and then the correction is accomplished either acutely or gradually after the osteotomy.

Osteotomy Technique

We prefer to make our osteotomy with an osteotome after predrilling the site. Typically we do this percutaneously, but we use the same technique in conjunction with an open exposure. C-arm fluoroscopy is brought into the AP projection and centered over the proximal femur. We make our bone cut directly below the level of the lesser trochanter. The proposed site is predrilled with three to four bicortical passes of a 4.8 mm drill bit. The image ensures that the drill maintains one plane. An 8 mm osteotome is then passed from lateral to medial through the cortical bone. Multiple passes and increasing width osteotomes are used until the

Table 1 Decision Making About Proximal Femoral Osteotomy

Diagnosis	Deformity	Technique and Implant	Pearls	Complications
Femoral neck nonunion	Varus LLD	130° blade plate wedge resection use compression technique	Retain screws while seating blade to prevent displacement	ON poor positioning of blade in head NU of osteotomy
Intertrochanteric nonunion and malunion	Varus LLD	95° blade plate stimulate/mobilize NU w/osteotome	Hardware removal creates bone loss in the head making blade positioning crucial	Poor positioning of blade in head
Fibrous dysplasia	Multiapicaldeformity	External fixation recruit skin to prevent necrosis resolve CORAs to minimize number of osteotomies	Apply each segment of frame to mimic the deformity. After osteotomy and reduction the rings are parallel. Release skin under tension	Skin necrosis pin loosening
Hip dysplasia (reduced hip joint)	Valgus anteversion consider periacetabular osteotomy	Internal or external fixation Ex Fix allows percutaneous technique	May need trochanteric advancement	Trendelenberg gait from proximal relocation of the greater trochanter
Coxa Vara	Varus retroversion or anteversion LLD Trendelenberg	Internal or external fixation Ex Fix if lengthening needed	Correct rotational deformity	Depends on etiology osteonecrosis osteoarthritis
Femoral shortening requiring proximal lengthening	LLD hip and knee stability	External fixation proximal lengthening	Circular fixation offers excellent control and can be well tolerated, may use monolateral frame	Discomfort loss hip/knee motion varus deformity
Distal femoral bone defect	Bone transport	External fixation monolateral rail works well	Bone graft docking site	Flexion and varus of transport fragment

Abbreviations: ON, osteonecrosis; CORA, center of rotation of angulation; LLD, leg length discrepancy; Ex Fix, external fixator; NU, nonunion.

Table 2 Review of Literature

Authors	Journal and Year	Title	Design	Results	Conclusions
Bartonicek, Skala-Rosenbaum, Dousa	J Orthop Trauma 2003	Valgus intertrochanteric osteotomy for malunion and nonunion of trochanteric fractures	Retrospective clinical study 15 patients 120° blade plate	14 healed Varus corrected average lengthening achieved = 2 cm Harris hip score improved 19 pts	Effective and reliable procedure restores hip function corrects deformity
Wu, Shih, Chen, et al.	J Trauma 1999	Treatment of femoral neck nonunions with a sliding compression screw: comparison with and without subtrochanteric valgus osteotomy	Prospective study 32 patients osteotomy if shortening >1.5 cm F/U minimum 2 yr	All nonunions healed 3 complications in osteotomy group (18%): ON femoral head (2), Osteotomy site nonunion (1) no complications nonosteotomy group	Sliding hip compression screw is effective Rx for fem neck nonunions w/ <1.5 cm shortening adding osteotomy will improve leg length
Hartford, Patel, Powell	J Orthop Trauma 2005	Intertrochanteric osteotomy using a dynamic hip screw for femoral neck nonunion	Retrospective 8 patients F/U 2 yr average	Nonunion healed in all patients alive at F/U (7) Harris hip score improved 49 pts new (no suggestions) gait in 4 patients (medialization of femoral shaft)	IT osteotomy effective means achieve valgus and heal NU avoid NU seen w/ subtroch osteotomy altered biomechanics hip additional rotational control of blade plate not necessary
Ali, Saleh	Injury 2002	Treatment of distal femoral nonunions by external fixation with simultaneous length and alignment correction	Retrospective 15 patients F/U 2 yr minimum 5 infected	14 cases united 5 cm lengthening average	External fixation advantages include ability to span knee and increase stability, simult. lengthening, compatible w/ infect and poor skin
Papagel-opoulos, Trousdale, Lewallen	CORR 1996	Total hip arthroplasty with femoral osteotomy for proximal femoral deformity	Retrospective 31 patients F/U 4.6 yr average	Harris hip score improvement complications: intraop femur Fx (7) NU osteotomy (4) aseptic stem loosen-ing (4), hip instability (4)	Simultaneous femoral osteotomy to correct deformity and THA is technically demanding w/ associated complications

Abbreviations: Pts, patients; Fx, fracture; ON, osteonecrosis; Rx, treatment; NU, nonunion; THA, total hip arthroplasty.

osteotomy is complete. Rotational osteoclasis is the final maneuver to ensure free movement of the cut bone ends. Alternatively, a power saw combined with cooling irrigation fluid may be used. This is especially helpful if one is planning to excise a wedge of bone (Fig. 1).

POSTOPERATIVE CARE

Drains are usually removed on the first postoperative day. Weight bearing as tolerated is typically allowed immediately after external fixation. Partial weight bearing precautions (30 lbs) are usually used for six weeks after internal fixation. Wound care is routine. Pin care for external fixation starts on the second postoperative day and showering begins after the fourth postoperative day. The first office visit is at two weeks post surgery to remove sutures. This is followed by regular monthly visits with X-rays until bony union is observed. A shoe lift may be indicated for limb length inequality or a later limb lengthening procedure made be planned if indicated.

COMPLICATIONS

Major complications include infection, neurovascular injury, nonunion, inability to obtain or maintain a full correction, persistence of postoperative pain, and continued degeneration of hip articular cartilage. Other complications include deep vein thrombosis and painful hardware. With regard to external fixation, complications include pin infection, fracture above or below the frame and fracture through a screw hole after frame removal, stiffness of adjacent joints, and septic arthritis if pins communicate with the joint.

FUTURE AND CONTROVERSIES

When proximal femoral osteotomy is used for the correction of congenital and acquired deformities and repair of hip fracture, results of nonunions have been favorable. Hip range of motion, gait, pain, leg length discrepancy, and patient satisfaction are improved. If arthritis develops, then future joint replacement is often facilitated. Simultaneous femoral osteotomy and total hip arthroplasty is a technically demanding procedure that has yielded acceptable results for complex hip reconstruction with deformity. Proximal femoral osteotomy will continue to find applications in the correction of deformity in adult patients. Computer navigation promises to greatly advance the technical accuracy of all osteotomy procedures and will undoubtedly have a profound impact on how we execute forthcoming proximal femoral osteotomies.

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28 Articulated Hinged Distraction of the Hip

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INTRODUCTION

Degenerative arthritis of the hip in young individuals, whether from osteonecrosis or chondrolysis, is a difficult therapeutic problem. The ideal treatment for arthritis would allow for painless range of motion (ROM) without sacrificing future mobility of the joint. In the elderly, total joint arthroplasty is a reasonable solution for arthritis; however, in young individuals total joint arthroplasty is associated with significant problems, including polyethylene wear, loosening, and the need for multiple revisions (1–3). Fusion of the hip joint does relieve pain (4–6,23), but at the expense of ROM and deleterious long-term consequences on surrounding joints (4,6–11). Articulated hinged distraction (AHD) utilizes external fixation to reduce joint forces and allows for the repair of articular cartilage (8,12). AHD takes advantage of the intrinsic capacity of cartilage cells to regenerate in young individuals.

The concept of AHD is that the neutralization of forces across the hip joint creates a suitable environment for cartilage repair. This has been termed arthrodiastasis. In degenerative arthritis of the hip there is a cycle of injury to the articular surface. The initial injury causes inflammation, which in turn creates more injury to the articular surface (13). Even at rest this cycle continues because the hip joint is constantly experiencing significant compressive force due to sustained muscle contracture (14). AHD neutralizes both muscle and weight bearing forces and creates a better environment for the repair of articular cartilage (14). Movement of the hip with the distractor in place causes circulation of synovial fluid, which is thought to encourage fibrous repair of articular cartilage.

The principle behind AHD has been supported in animal models (8,12). In a dog model, the excision of cartilage from the tibiotarsal joint was followed by application of a hinged external fixator. Later histologic analysis revealed the formation of callus, whose macroscopic and microscopic appearance was similar to normal articular cartilage.

A number of case series have shown successful application of AHD to patients. AHD has been used successfully in the elbow and knee in human subjects (15,16). Several series have shown successful use of AHD, especially in young individuals (14,17–19). AHD provides an ideal environment for self healing and does not prevent the future use of less conservative measures, such as arthrodesis or total joint arthroplasty.

CLINICAL EVALUATION

A thorough history of the hip disability and comorbidities, physical examination, and accurate interpretation of radiographic modalities are essential for successful use of AHD. Accurate diagnosis of etiology and stage of disorder dictate treatment and will help predict prognosis.

The history of the hip pain and disability should help the surgeon arrive at one of the following etiologies: Legg-Calve-Perthes (LCP) (19), secondary osteonecrosis of the femoral head, or chondrolysis of the hip joint. In addition, hip distraction can be utilized to protect a hip after labral repair or femoral head fracture. Identification of comorbid conditions is also important. In osteonecrosis of the femoral head for example, when there is concomitant Down's syndrome or epiphyseal dysplasia the outcome of AHD is inferior compared to when there is osteonecrosis alone.

In the physical examination, the following should be noted: neurovascular status of the affected extremity, ROM of hip and knee, pelvic obliquity, gait, and the presence of the Trendelenburg sign. Decreased ROM may be caused by a soft tissue contracture. Surgical release can be accomplished at the same time as placement of the distraction device. Decreased ROM may also be caused by partial or complete subluxation, which changes the treatment options. Quantification of the pain using a visual analog scale for future comparison proves useful in children.

Initial radiographic assessment should include the following views: anteroposterior, frog leg lateral, and a von Rosen view. The von Rosen view is taken with the patient supine and the hip slightly flexed, abducted 45° and maximally internally rotated. The purpose of the von Rosen view is to show potential coverage of the femoral head by the acetabulum. These views should be reviewed to determine the stage of specific disease, joint space, fluency of Shenton's line, percent coverage, and whether the femoral head is reduced and/or reducible. If there is a concern about anterior coverage of the femoral head, a false profile view can be obtained. A false profile view is obtained with the patient standing so the beam passes through the hip as the patient stands at a 65° angle from the plate.

Complete dislocation is a contraindication for distraction arthroplasty. But subluxation, if reducible, may be treated with AHD. A computed tomography scan can be helpful to identify acetabular or femoral cysts and degree of collapse of the femoral head articular surface.

CLASSIFICATION

There is no formal classification system for the application of AHD about the hip. Three etiologies of hip pain and disability that hip distraction has been used for are LCP disease, primary and secondary osteonecrosis, and chondrolysis. The decision to treat each of these conditions with hip distraction relies on various parameters. In addition, the prognosis after treatment with AHD is different for each etiology.

In one series looking at the use of AHD for LCP disease, the majority of cases in which the hip distractor was used were Catterall IV and Herring C grading (18). Analysis of the radiographs for staging may be helpful. The modified Waldenstrom classification divides LCP into four radiographic stages. The stages are initial, fragmentation, reossification, and residual. It is best to wait for the disease to progress to fragmentation phase before treating with AHD. If the distractor is placed too early, the hip will still deteriorate after removal of the device. With appropriate use, the prognosis of LCP after treatment with hinge distraction is usually good to excellent (19).

Osteonecrosis from various etiologies has also been addressed with AHD. One series of patients (14) with predominantly grade IV osteonecrosis as per the classification of Ficat and Arlet (21,22) had good results.

AHD has also been used in patients with chondrolysis. There is no study to identify the appropriate joint space narrowing when distraction should be used. Progressive joint space narrowing with continued pain and failure of conservative treatment is a reasonable point to consider AHD. Aldegheri et al. (14) found good results in 14 of 15 of these patients. In a smaller series by Thacker et al. (19), all three patients with chondrolysis treated with AHD had excellent results.

TREATMENT OPTIONS

Patients being considered for hip distraction should have first tried nonoperative treatment, which consists of activity modification, physical therapy, and nonsteroidal anti-inflammatory drugs (NSAIDs).

For LCP, surgical options consist of femoral osteotomy, pelvic osteotomy, a combination of both, or AHD. For younger patients with LCP who need surgical intervention, femoral osteotomies are the first choice in treatment. However, if the femoral osteotomy fails and pain is still present with progression of disease, AHD is recommended. In older patients with LCP, AHD may represent the first line of treatment. Treatment should begin when radiographic evaluation reveals that the patient's proximal femur is in the fragmentation phase (Fig. 1). Patients with LCP should have the distractor in place for four to six months. The exact length

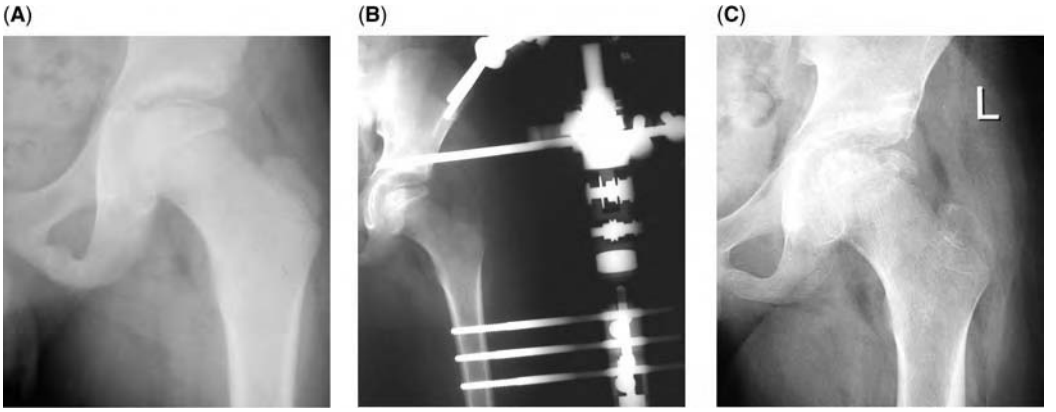

Figure 1 A 10-year-old patient with Legg-Calve-Perthes of the left hip. (**A**) Fragmentation stage with early collapse. (**B**) The distractor is in place. Note rounding of the collapsed femoral head. Placement of the screws directly above the acetabulum is in 10° of abduction and distal screws are parallel to the screws directly above the acetabulum. (**C**) After removal of the distractor showing resolution and healing of the femoral head.

of time depends on the rate of healing of the femoral head. Once the femoral head has healed, the distractor may be removed.

In secondary osteonecrosis, AHD can also be used as a first line of treatment once the femoral head has begun to fragment. Like LCP, patients with secondary osteonecrosis should have the distractor in place for four to six months depending on femoral head healing.

Currently there are few surgical options for patients with chondrolysis. Once a patient has failed NSAIDs and physical therapy, he or she becomes a candidate for AHD. The patient's symptoms, physical examination, and X-rays should be followed regularly. If joint space narrowing and pain continues, AHD should be considered. Patients with chondrolysis should have the distractor in place for approximately three months.

SURGICAL TECHNIQUE

Placement of the hip distractor can be divided into three components: identification of the center of the femoral head, placement of the pelvic side of the distractor, and lastly placement of the femoral side of the distractor. Before induction of anesthesia, all components should be assembled. Preanesthesia assembly decreases anesthesia time and allows the surgeon to identify any missing or defective components. Contracture releases, if necessary, should be done before pin placement.

Identification of the center of the femoral head begins with proper positioning of the patient and fluoroscope in order to be able to obtain adequate anteroposterior and lateral hip views. The patient should be placed supine with a small bolster under the affected hip on a radiolucent table. The patient should be positioned so the affected hip is on the side edge of the table. Multiple anteroposterior and lateral projections as well as "live" fluoroscopy may be necessary.

The center of the femoral head is judged to be the center of hip rotation. Under fluoroscopic guidance, the center of the femoral head is found and marked. If the femoral head is collapsed and does not appear as a true sphere, the point should be at the center of the femoral head as though the head were complete and intact. A line from the top of the greater trochanter to the center of the femoral head in neutral hip position is marked using a 2.0-mm Kirschner wire (K-wire). This newly marked point is the center of rotation of the hip, and the center of the fixator hinge should be placed at this point (Fig. 2). The cannulated center of the pivot body is the center of rotation of the fixator. The pivot body should be threaded over the 2.0-mm K-wire.

Attention can now be turned to the three proximal half pins directly above the acetabulum. These pins are inserted 10 to 15 mm above the joint line. Prior to drilling for the first proximal half pin, the degree (if any) of abduction should be determined. If abduction of

Figure 2 Kirschner wire in the center of rotation of the femoral head.

10° is preferred, pin insertion must be angled accordingly. The fixator should be positioned away from the patient to anticipate swelling of the soft tissue that may occur during treatment.

The placement of the first pin is guided by the proximal clamp of the hip distractor. Fluoroscopy is necessary to prevent overpenetration of screws. Judet views are useful to confirm placement. The first pin placed should be the most anterior because bone stock and the space are most limited anteriorly. The hip distractor will guide placement of the second and third pins. It is important to place the pins with the distractor and to check the positions of the hinge. Three pins are recommended to improve strength and distribute the load. In most cases, there will be two pins applied anterior to the distraction post and one placed posterior to the post. Because there is a concern of violating the sciatic notch posteriorly, a 2.0-mm K-wire can be used to determine if it is safe to place a posterior pin. Supplemental pins are then utilized to support the proximal construct. Either two half-pins are applied through the tables of the ileum and utilizing pins to bar attachments are connected to the three supracetabular screws or one half-pin can be inserted anterior to posterior at the level of the anterior-inferior iliac spine and similarly connected.

Attach the distal femoral component. Be certain the distal pins are appropriately spaced from the center of the distractor and that the entry is perpendicular. Pins should have a bicortical purchase and be parallel to the pins that are directly proximal to the acetabulum. Adjust the position of the femur to the desired hip abduction. Predrill and then insert the distal pin. After placement of the first pin, use the fixator as a guide to insert the second and third pins (Fig. 3).

Once the hip distractor is in place, the distraction can be performed. Distraction may be done acutely. However, if the surgeon encounters any binding, gradual distraction is recommended. Distraction is typically done between 5 and 8 mm. A landmark that can be used to determine the amount of distraction needed is the restoration of Shenton's line. If the component being used has proximal and distal means of distraction, the proximal distractor should be utilized. The hinge and the femoral head should move together in order to maintain the hinge at the center of hip rotation. After appropriate distraction has been attained, the distractor should be locked. Xeroform and gauze should be used to dress pin-insertion sites.

The following problems may be encountered: (i) hinged placement is too proximal or distal prior to application of femoral pins, (ii) as distraction occurs, the hinge moves distal to the center of the femoral head, and (iii) after the frame is applied, the hinge is noted to be too proximal.

The solution to the first problem—poor positioning of the hinge prior to application of femoral pins—is adjustment of the proximal distractor. In the event that the hinge is too distal and there is no more room to shorten the distractor, this can be solved by doing a distraction at the femoral clamp after the femoral pins have been inserted. This will effectively bring the femoral head down to the level of the hinge.

Figure 3 Schematic representation of final pin placement. The squares along the iliac crest, just above the acetabulum and on the diaphysis of the femur, are the appropriate zones for pin placement.

The second problem—movement of the hinge after distraction—will interfere with smooth flexion and extension. This occurs because the pins may bend prior to the force causing the hip to distract. This will constantly change the position of the hip in relation to the hinge prior to distraction. The solution is to distract distally on the femoral clamp. This effectively brings the femoral head down to the level of the hinge.

The third frequently encountered problem is noting that the hinge is too distal after the frame is already in place. The solution is to compress through the proximal distractor and then distract from the distal end. Once the hinge is aligned, distraction from the proximal device may continue (Fig. 4).

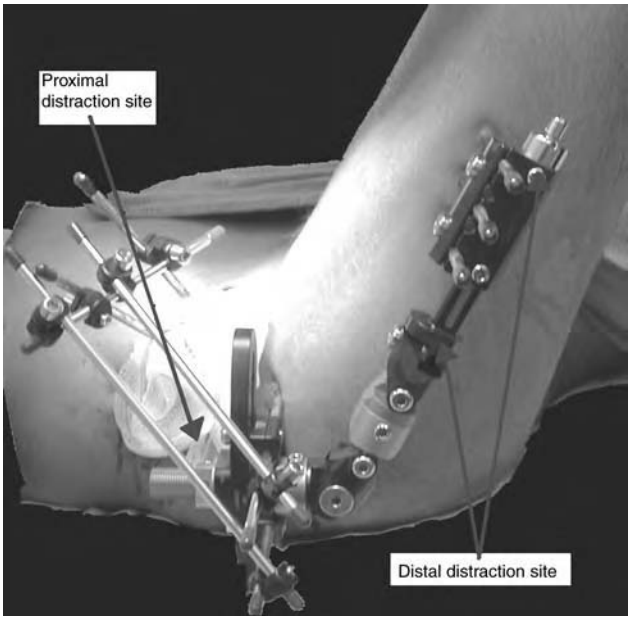

Figure 4 Note the two locations where distraction and compression can take place. If the center of the distractor is too distal, the distractor can be distracted distally and compressed proximally until the distractor is centered appropriately.

REVIEW OF LITERATURE

Authors	Journal, Year	Title	Number of Patients	Results	Conclusions
Thacker, M; Feldman, D	J Pediatric Orthop, 2005	Hinged distraction of the adolescent arthritic hip	11	7—excellent outcome; 3—good outcome; 1—failed distraction	Distraction is effective in eliminating pain, improving function, and preventing degeneration
Aldegheri, R; Trivella, G; Saleh, M	Clin Orthop Rel Res, 1994	Articulated distraction of the hip	80	Poor results in patients older than 45 years or having inflammatory arthropathy. Satisfactory results were achieved in more than 70% of patients aged below 45 years old	Younger patients without inflammatory arthropathy tend to benefit the most from AHD
Segev, E; Ezra, E; Wientroub, S; Yaniv, M	J Pediatric Orthop B, 2004	Treatment of severe late onset Perthes' disease with soft tissue release and articulated hip distraction: early results	16	Postoperatively, all patients had improved ROM, decreased pain and were able to walk without support. Most patients had radiographic improvement of Shenton's line and saddle shape deformity	AHD improved clinical and radiographic parameters in patients with avascular necrosis of the femoral head. The improvement was greatest in LCP
Kucakkaya, M; Kabukcouglu, Y	J Pediatric Orthop, 2000	Avascular necrosis of the femoral head in childhood: the results of treatment with articulated distraction method	11	Avascular necrosis healed, but there were five aspherical congruencies and two aspherical incongruencies	Poor results, but unable to make generalizations secondary to small number of patients
Canadell, J; Gonzales, F; Barrios, RH; Amillo, S	Intern Orthop, 1993	Arthrodiastasis for stiff hips in young patients	9	ROM improved from 20° to 65°. Articular space widened by 2.8 mm and pain was present in only 3 patients at follow up	AHD improves clinical and radiographic findings in young patients with stiff hips

Abbreviations: AHD, articulated hinged distraction; LCP, Legg-Calve-Perthes; ROM, range of motion.

COMPLICATIONS

Complications that have been encountered in the literature with AHD include: pin site infections, pin loosening, hardware failure, painful knee effusion, pain that necessitated placement of an epidural catheter, abduction contracture after removal of external fixator, and injury to a collateral of the superior gluteal artery (14,18,19). Some common difficulties after AHD are pain interfering with rehabilitation and development of a flexion contracture.

The number of pin-site infections varies between different series from 1 out of 11 to 13 out of 16 (23). Meticulous technique of pin insertion, should decrease the number of pin problems. Postoperative management including daily showering, prophylactic antibiotics, and dressings over the pins to prevent skin movement, are important as well. If there is increasing drainage, redness, or pain at a pin site, oral antibiotics should be started immediately. True infections, which are deep, associated with fever, and possibly have osseous changes, should be treated aggressively with intravenous antibiotics and possible pin exchange.

Pin loosening is reported in a number of series. With the use of hydroxyapetite-coated pins, this is becoming a rare complication. The pins should be changed if they do become loose.

In one series, a gluteal artery collateral was damaged by pin placement and intraoperative dissection was carried out to coagulate the vessel (17).

Difficulties that are present after surgery include pain that interferes with rehabilitation and development of a flexion contracture. In some instances, rehabilitation may include continuous passive motion (CPM) machines. Postsurgical pain may interfere with CPM acutely after surgery. One solution for this problem is the use of an epidural catheter (19). Once the acute pain has resolved, the epidural catheter can be removed. The longest the catheter should be in place is 72 hours.

Contractures may occur during or after fixator placement. Postsurgical development of a flexion contracture can be treated by locking the distractor in extension at night. Kukukkaya et al. (13) encountered an abduction contracture after fixator removal. They attributed this to impingement between the femoral head and the inferior region of the acetabulum. After resecting the osseous fragment, full hip ROM was restored.

Clinical improvement is not always mirrored by improvement in radiographic appearance. However, joint space should improve during and after placement of distractor. Two major determinants of a good outcome are correct application of the device and closely supervised rehabilitation.

FUTURE DIRECTIONS

Randomized studies comparing AHD with other modalities for treating hip arthritis in the young are needed. Most of the current literature are case series that, although informative, lack controls needed to draw more concrete conclusions.

In addition, AHD may be used with other medical and surgical interventions to improve outcomes. For example, AHD with the concomitant use of either bisphosphonates or free fibular grafts might provide new and improved treatment for aseptic necrosis.

Lastly, future studies at the cellular level may elucidate how hip distraction affects the joint environment and potential for cartilage repair. Further studies may provide information on what degree of distraction is optimal.

Currently AHD appears to provide a means for relieving hip pain caused by aseptic necrosis and chondrolysis that does not interfere with the future use of other surgical interventions.

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Part VIII PEDIATRIC

29 Lengthening Reconstruction Surgery for Congenital Femoral Deficiency

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INTRODUCTION

Congenital femoral deficiency (CFD) is a spectrum of severity of femoral deficiency and deformity. Deficiency implies a lack of integrity, stability, and mobility of the hip and knee joints. Deformity refers to bone malorientation, bone malrotation, and soft tissue contractures of the hip and knee. Both deficiencies and deformities are present at birth, nonprogressive, and of variable degree.

CLASSIFICATION

Existing classifications of congenitally short femora and proximal femoral focal deficiencies are descriptive but are not helpful in determining treatment. A recent longitudinal follow-up of different classification systems (1) showed that they were inaccurate in predicting the final femoral morphology based on initial radiographs. The Paley classification system (Fig. 1) is based on the factors that influence lengthening reconstruction of the congenitally short femur (2).

Paley Classification of Congenital Femoral Deficiency (Fig. 1)

Type 1: "intact femur" with mobile hip and knee

a. Normal ossification proximal femur
b. Delayed ossification proximal femur

Type 2: "mobile pseudarthrosis" with mobile knee

a. Femoral head mobile in acetabulum
b. Femoral head absent or stiff in acetabulum

Type 3: "diaphyseal deficiency" of femur

a. Knee motion 45° or more
b. Knee motion less than 45°
c. Complete absence of femur

Type 4: "distal deficiency" of femur

Knee joint mobility/deficiency rather than hip joint mobility/deficiency is the most important determining factor for functional outcome and reconstructibility in cases of CFD. Paley Types 1 and 2 CFD are the most reconstructible. A wide spectrum of hip and knee dysplasia and deformity exists in Type 1 cases. Because Type 1 is the type most amenable to lengthening, it merits subclassification according to factors that require correction before lengthening can be performed.

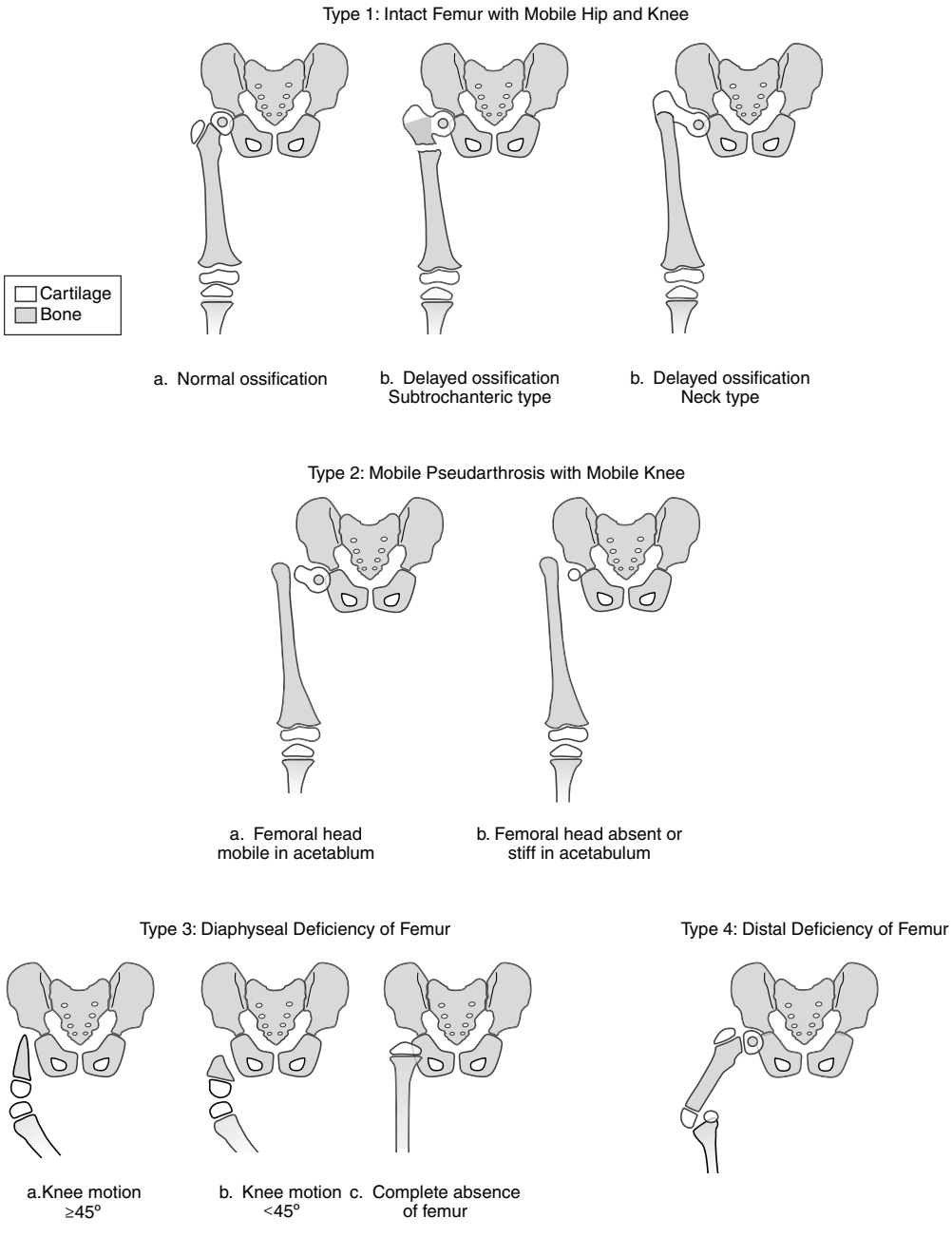


Figure 1 Paley classification of congenital femoral deficiency. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

TYPE 1 CONGENITAL FEMORAL DEFICIENCY: INTACT FEMUR

Hip and Knee Considerations

Type 1 CFD is the type that is most reconstructible. Before lengthening, significant bone deformities and soft tissue contractures of the hip and knee should be reconstructed. At the hip, if the acetabulum has a center edge (CE) angle of more than 20°, the neck shaft angle is more than 110°, and the greater trochanter is not significantly overgrown such that the medial proximal femoral angle (MPFA) is not less than 70°, no hip surgery is required before the first lengthening. At the knee, if the fixed flexion deformity (FFD) is less than 10°, the patella tracks with no subluxation laterally, and no evidence of significant rotary subluxation or dislocation

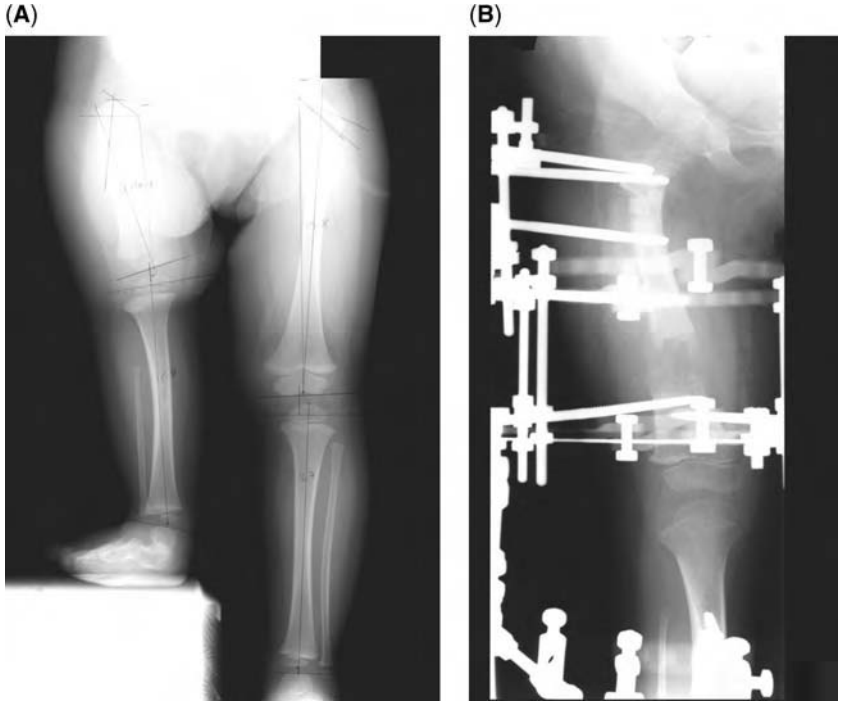


Figure 2 (**A**) Radiograph of patient with Paley Type 1a congenital femoral deficiency. The femoral neck is ossified and has significant coxa vara. (**B**) Radiograph shows initial lengthening with concurrent proximal subtrochanteric osteotomy for coxa vara correction. The lengthening is performed through the distal osteotomy site, and the external fixator bridges the knee joint to prevent subluxation and dislocation.

of the tibia on the femur is present, the knee does not require surgical reconstruction before lengthening (Fig. 2). If, however, any of these criteria are not met, the hip and/or knee should be reconstructed before the first lengthening.

Acetabular Dysplasia

It is very common for even mild cases of CFD to include acetabular dysplasia, which predisposes the femoral head to subluxation during lengthening. A CE angle less than 20° before femoral lengthening is an indication for pelvic osteotomy. The acetabular dysplasia associated with CFD is not like that associated with developmental dysplasia of the hip. The deficiency is not predominantly anterolateral. The deficiency is more superolateral, often with a hypoplastic posterior lip of the acetabulum. Therefore, Dega osteotomy is the method we prefer over the Salter osteotomy or Millis-Hall modification of the Salter osteotomy (combining innominate bone lengthening with the Salter by using a trapezoidal instead of a triangular graft), because improved coverage can be gained with Dega osteotomy (3). Dega osteotomy is best performed when the patient is two years of age but can be performed in older patients if the triradiate cartilage remains open (Fig. 3).

Proximal Femoral Deformities

The proximal femoral deformity of CFD is not a simple coxa vara in most cases. It is a complex combination of bone deformities in the frontal, sagittal, and axial planes, combined with soft tissue contractures affecting all three planes. The severity of these deformities is often mild to moderate in Type 1a cases but is usually severe in Type 1b cases.

In more severe cases, the obvious coxa vara is associated with an abduction contracture of the hip. If the coxa vara is corrected on its own, the abduction contracture will be uncovered. This contracture will prevent full valgus correction and/or will prevent the hip from returning to a neutral position relative to the pelvis. The abduction contracture causes a fixed pelvic tilt,

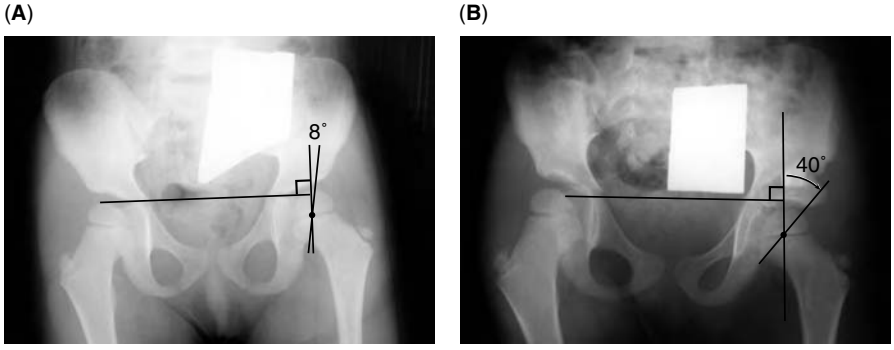


Figure 3 (**A**) Three-year-old female patient with Paley Type 1a congenital femoral deficiency and concurrent right hip dysplasia shown by the diagonal acetabular sourcil and a center edge angle of 8° (left center edge angle = 25°). (**B**) Postoperative radiograph after Dega osteotomy shows corrected dysplasic acetabulum.

which makes the limb length discrepancy (LLD) appear less than it was before surgery. In the presence of an open growth plate or a nonossified neck/subtrochanteric segment, the abduction contracture leads to recurrence of the coxa vara through these cartilaginous structures.

FFD of the hip usually accompanies severe coxa vara. The magnitude of the FFD often is masked by extension deformity in the bone of the proximal femur. External rotation deformity of the distal relative to the proximal femur (retroversion) is always present because of a combination of bony torsion and contracture of the piriformis muscle. The correction of these deformities is accomplished with a new surgical procedure, the *superhip procedure* (SUPER is an acronym for Systematic Utilitarian Procedure for Extremity Reconstruction).

SUPERHIP PROCEDURE (FIG. 4)

Step 1: Incision and Reflection of the Anterior Flap

A long, slightly concave anterior incision is made over the posterolateral border of the femur, from the iliac wing (starting 4 to 6 cm posterior to the anterior superior iliac spine) to one-third the way down the femur. A second, S-shaped incision is made from the lateral side of the patellar tendon, extending proximally in line with the intermuscular septum at the level of the knee joint, leaving a bridge of intact skin between the two incisions. The subcutaneous tissues are dissected off the fascia lata, reflecting the flap of skin and subcutaneous tissues anteriorly. The limit of the dissection of the proximal incision is the interval between the tensor fascia lata (TFL) and the sartorius. Distally, the fascia lata is exposed from the patella to the intermuscular septum. In very short femora, the two incisions are connected and the anterior flap is reflected as one (Fig. 5A and B).

Step 2: Reflection of the Fascia Lata and Tensor Fascia Lata Muscle

The fascia is split longitudinally at the TFL-sartorius interval, with care taken to stay on the TFL side to avoid injury to the lateral femoral cutaneous nerve. The fascial incision is extended distally to the lateral border of the patella, ending at the tibia. The posterior split of the fascia lata starts distally and posterior at the intermuscular septum and extends proximally up to the interval between the TFL and gluteus maximus. If knee ligamentous reconstruction is not required, the fascia lata is cut distally at the tibia and is reflected proximally. If ligamentous reconstruction with use of the fascia lata is planned, the fascia lata is cut proximally and reflected distally. To reflect the fascia lata, it has to be dissected free of all the overlying subcutaneous tissue, including that under the skin bridge between the two incisions. At the proximal end, the TFL muscle is reflected proximally and posteriorly on its posterior pedicle. Its anterior vascular pedicle (terminal branch of the lateral femoral circumflex vessels) can be cauterized and cut. Care should be taken to separate the tensor muscle from the underlying gluteus medius. The two muscles might be adherent to each other. The gluteus inserts on the greater trochanter, but the tensor passes over the trochanter (Fig. 5C).

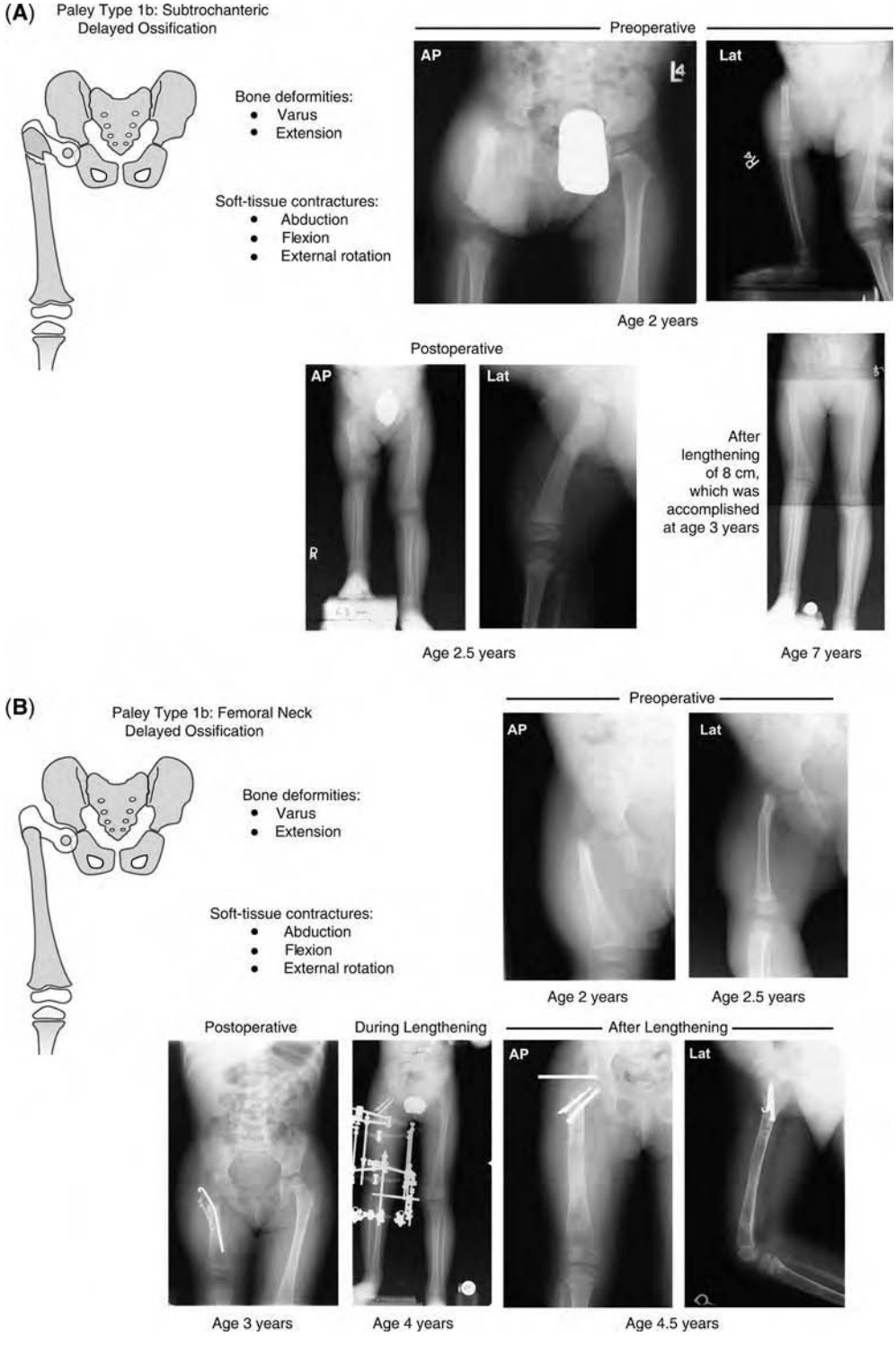


Figure 4 Radiographic examples of superhip procedure for Paley Type 1b congenital femoral deficiency. (**A**) Subtrochanteric delayed ossification. (**B**) Femoral neck delayed ossification. *Abbreviations*: AP, anteroposterior; Lat, lateral. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

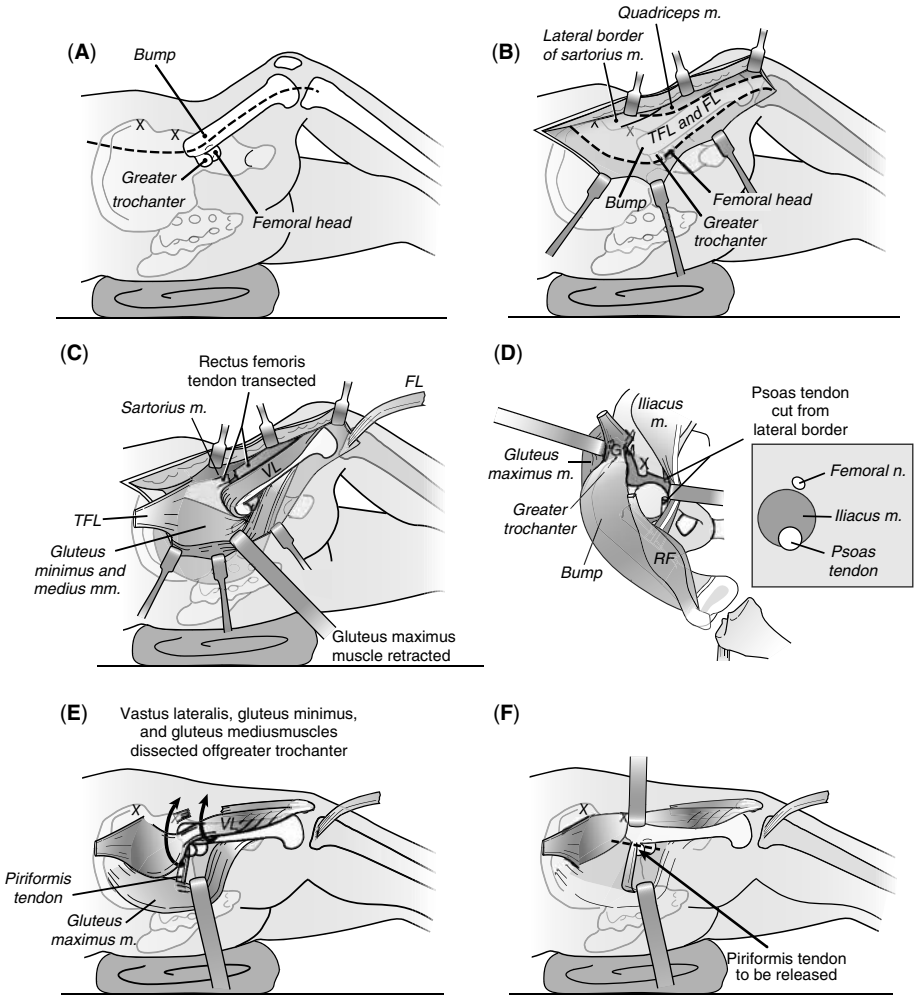


Figure 5 Superhip procedure. (**A**) Incision. (**B**) Reflection of anterior flap. A point 4 to 6 cm posterior to the anterior superior iliac spine is marked on the skin, and the lateral ''bump'' is marked on the skin. These two points are connected with a curvilinear line that extends distally on the posterior margin of the vastus lateralis muscle belly. The second incision is a distal ''S'' incision that begins at the level of the lateral intramuscular septum on the side of the thigh and proximally at the level of the superior pole of the patella and extends to the lateral margin of the patella tendon to the tibial tubercle. The anterior flap is dissected off the deep fascia to the midline of the thigh. (**C**) Reflection of fascia lata and tensor fascia lata (TFL) muscle. The anterior and posterior margins of the fascia lata are dissected as described, with the fascia lata being released proximally at the musculotendinous junction of the TFL muscle. The fascia lata is reflected distally to its insertion on Gerdy's tubercle of the proximal tibia. The TFL muscle is dissected off the gluteus minimus and medius and reflected proximally. (**D**) Hip flexion contracture release. After the TFL muscle is reflected proximally, the dissection is continued medially under the sartorius muscle. The rectus femoris tendon is the first structure identified as it inserts on the anterior inferior iliac spine. This tendon is released, and the psoas muscle and tendon are then identified. Before release of the psoas tendon, the femoral nerve, which is adjacent to the psoas tendon, is identified and decompressed. (**E** and **F**) Release of abduction and external rotation contracture. The confluent tendinous portions of the hip abductor muscles (gluteus minimus and medius muscles) and the vastus lateralis muscle are sharply dissected off the cartilaginous greater trochanter, creating a continuous musculotendinous sling. This release resolves the abduction contracture and allows access to the piriformis tendon. *Abbreviations*: m, muscle; FL, fascia lata; mm, muscles; VL, vastus lateralis; RF, rectus femor. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

Step 3: Hip Flexion Contracture Release

The dissection of the flexors is performed posterior to the sartorius to find the anterior inferior iliac spine. The rectus femoris tendon insertion is identified at the inferior spine. The direct and reflected head of the rectus femoris tendons are transected. Just medial to the rectus are sthe iliopsoas muscle and tendon. The psoas tendon is cut, leaving the intact iliacus muscle

fibers to bridge the gap. Care should be taken to avoid injury to the femoral nerve, which lies anterior to the medial edge of the iliacus muscle (the psoas tendon lies posteromedial to the iliacus muscle). Although the sartorius muscle might contribute to the flexion contracture, it is usually not tight. The anterior fascia of the thigh and the sartorius fascia might be tight. They might need to be released. The lateral femoral cutaneous nerve should be identified and protected before releasing these fasciae. The remaining flexion contracture is from the gluteus medius and minimus muscles and is addressed in the next step of the procedure (Fig. 5D).

Step 4: Abduction and External Rotation Contracture Releases

The abductor tendons (gluteus medius and minimus) insert into the greater trochanter and extend distally to become confluent with the quadriceps origin on the greater trochanter. The tendinous portions of both muscle groups can be sharply dissected and reflected together, maintaining the longitudinal continuity of the musculotendinous units. Neither muscle group can retract or shorten. With this step, the posterior aspect of the glutei is identified and followed to the posterior border of the greater trochanter. The posterior border of the vastus lateralis at the intermuscular septum is similarly identified and dissected free of the femur subperiosteally. This line is continued proximally along the posterior aspect of the greater trochanter. Because the tendinous covering of the trochanter is thin, it is important to peel a thin layer of cartilage with the flap. The flap of the conjoint gluteus–quadriceps tendon is sharply dissected and reflected from posterior to anterior off the trochanter and then anteriorly off the intertrochanteric line, leaving the anterior hip capsule intact. During this release, the piriformis tendon should be identified and released from its trochanteric insertion. This permits the femur to rotate internally. Once the abductor–quadriceps unit is free of the trochanter, the extension of the hip capsule to the acetabulum of the hip is evident. It is important not to release this capsule from the trochanter because doing so can lead to lateral subluxation. Because of the flexion contracture, femoral retroversion, and extension of the proximal femur, the greater trochanter is usually located very posterior and medial to the more prominent lateral "bump" (Fig. 5E and F).

Step 5: Proximal Femoral Osteotomy and Fixation

An arthrogram of the hip is next obtained. With the abduction, flexion, and rotation contractures all released, the femoral head and neck can be placed in neutral orientation to the pelvis by extending and maximally adducting the hip joint (Fig. 6). The fixation can be performed in one of two ways: (i) by using the hip plate method and (ii) by using the Rush rod method.

Hip plate method

With the hip plate method, the preferred implant is the pediatric sliding hip screw (Smith & Nephew, Memphis, Tennessee, U.S.A.). The first step is to place a guidewire from the tip of the trochanter to the center of the femoral head (Fig. 7A and B). A second reference wire is drilled up the femoral neck into the center of the femoral head. The neck reference wire should be at 45° to the trochanteric reference wire (Fig. 7C). The lag screw is inserted. The side plate is applied to the lag screw. With the use of a saw, a triangular segment of bone is removed from the side of the femur by making one cut parallel and one perpendicular to the plate. The width of the perpendicular cut is the same as the width of the shaft of the diaphysis of the femur

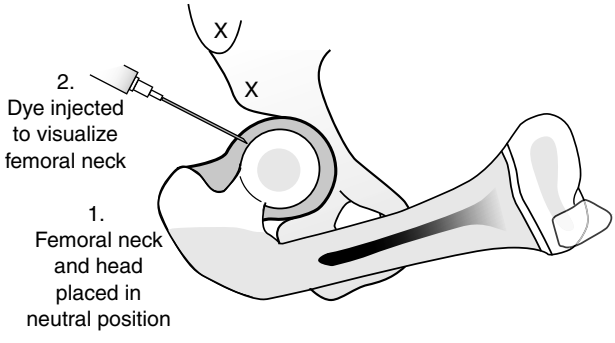


Figure 6 Arthrography performed, and hip placed in neutral position. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

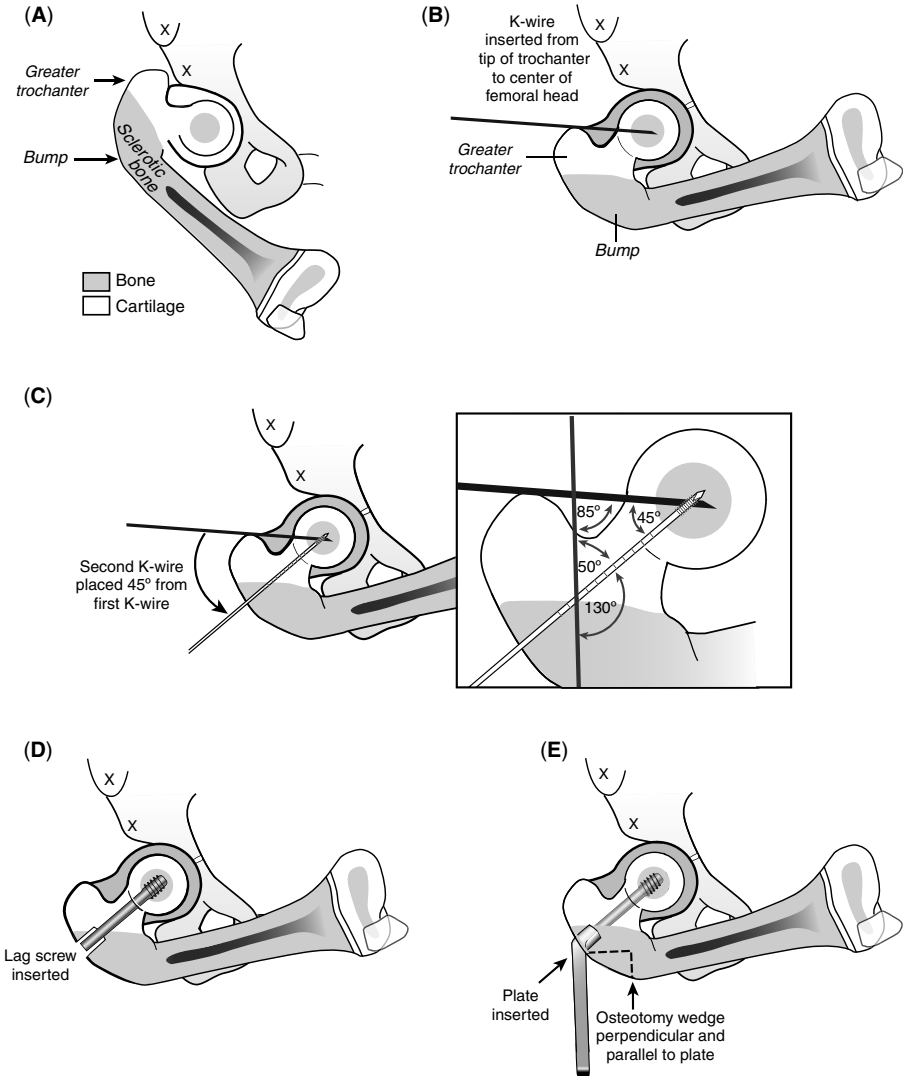


Figure 7 (**A** and **B**) After arthrography of the hip, a K-wire is placed from the tip of the greater trochanter to the center of the femoral head. (**C**) Second guidewire is placed into the femoral neck 45° from the initial guidewire. This will produce a 130° neck shaft angle after the osteotomy is completed. (**D**) Second guidewire is overdrilled, and the pediatric lag screw is inserted. (**E**) Side plate is applied and used as a guide for the initial bone cuts. A saw is used to remove a triangular segment of bone. The width of the perpendicular cut equals the diameter of the femoral diaphysis. (**F** and **G**) Second osteotomy is started at the parallel cut and directed distally in an oblique fashion. The third osteotomy is directed perpendicular to the long axis of the distal femoral segment and positioned to remove a 1- to 2-cm segment. (**H**–**K**) Compression screw is inserted into the pediatric lag screw. The femur is reduced to the plate and secured with screws. The most proximal screw should cross the osteotomy site into the medial buttress or up the femoral neck. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

(Fig. 7D and E). Next, an osteotomy is made from the lateral end of the parallel cut, obliquely across the femur. A second osteotomy is made perpendicular to the femur 1 to 2 cm distal to the first cut. The segment that is removed is used as bone graft for the Dega pelvic osteotomy (Fig. 7F and G). The compression screw is next inserted to join the plate to the proximal femur. The distal femur is internally rotated and reduced to the plate. The plate is secured with screws. The most proximal screw crosses the osteotomy line into the medial buttress of bone left after removal of the triangular segment. Occasionally, it can be advanced parallel to the lag screw into the femoral neck and head. If this screw cannot be oriented to go across the femoral neck, a second screw should be inserted across the femoral neck for rotational control. This second head and neck screw is required in the hip with Type 1b CFD with delayed

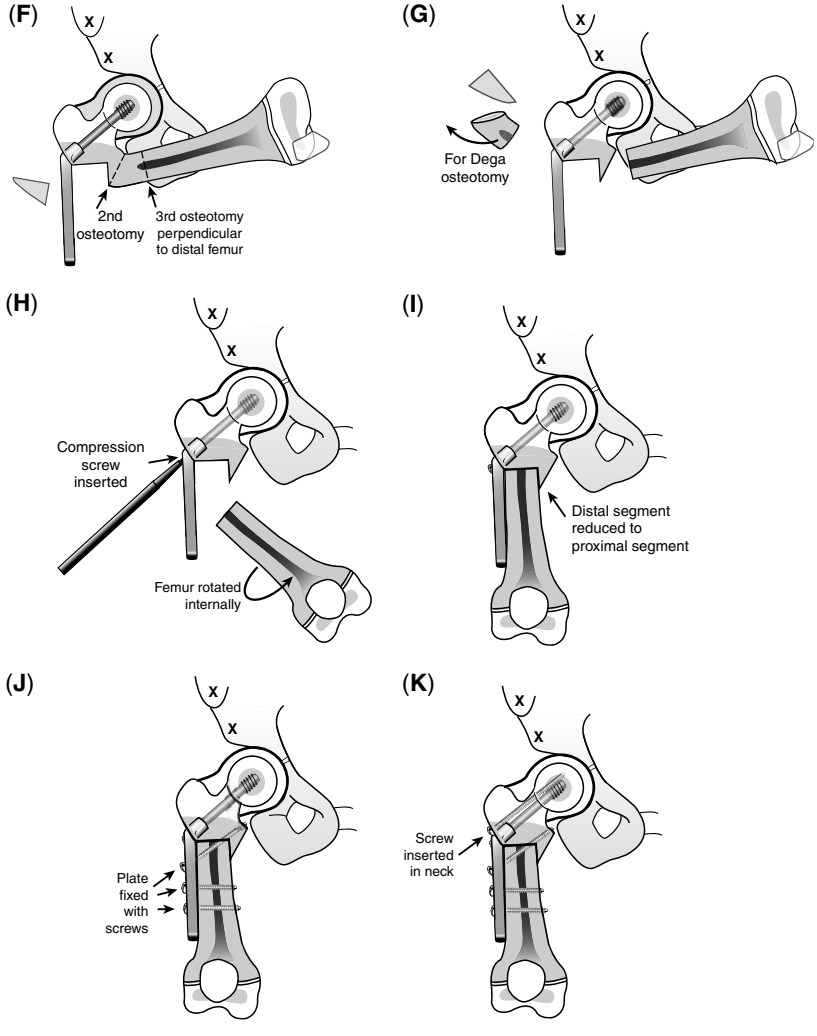


Figure 7 (*Continued*)

ossification of the femoral neck. With the subtrochanteric delayed ossification type, this is not necessary. With the subtrochanteric delayed ossification type, the lag screw does not need to cross the proximal femoral physis (Fig. 7H–K).

Rush rod method

With the Rush rod method, the first step is to place a guidewire from the tip of the greater trochanter to the center of the femoral head. A second reference wire is drilled at an 85° angle to the first wire, to create an MPFA of 84°. The second wire should be drilled from the lateral aspect of the femur to exit through the lateral aspect of the piriformis fossa (Fig. 8A). The wire passing through the piriformis is overdrilled with a cannulated drill. The diameter of the cannulated drill depends on the diameter of the Rush rod to be used for fixation. For a 1/8″ (3 mm) Rush rod, a 3.2-mm cannulated drill is used. Parallel and perpendicular cuts are made in the femur relative to the Rush rod, removing a triangular segment (Fig. 8B and C). A second osteotomy is performed 1 cm distal to the location of the triangular osteotomy. This creates a medial buttress of bone. A third osteotomy is performed 1 to 2 cm distal to the second osteotomy, removing a segment of bone. The medullary canal of the distal femur is drilled open with a solid drill bit of the same diameter as that which was used for the proximal end. The Rush rod is inserted into the proximal segment. The distal segment is reduced to the proxima segment and the Rush rod advanced across the osteotomy (Fig. 8E and F). For additional rotatory control,

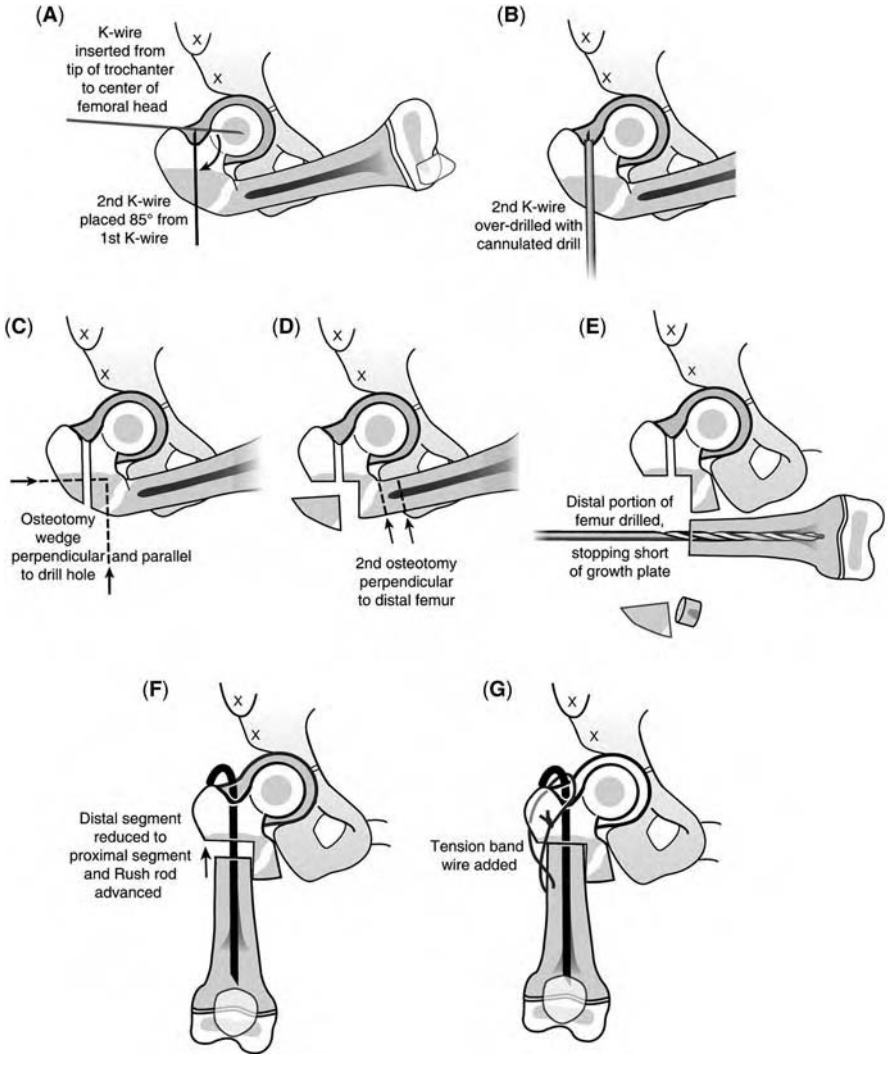


Figure 8 (**A**) Initial K-wire is placed from the tip of the greater trochanter to the center of the femoral head. The second guidewire is drilled from the lateral aspect of the femur, creating an 84° medial proximal femoral angle. (**B** and **C**) Second guidewire is overdrilled, creating the path for the rush rod in the proximal segment. Two cuts are made parallel and perpendicular to the rush rod path, removing a triangular segment of bone. (**D**) Second and third osteotomies are performed distal to the triangular osteotomy. These two osteotomies create a medial buttress proximally and remove a 1- to 2-cm segment of bone distally. This bone segment is later used for the Dega osteotomy of the pelvis. (**E**) Distal femoral canal is reamed with the same size drill as used in the proximal segment. (**F**) Rush rod is inserted from the proximal starting hole and advanced across the osteotomy site, reducing the femur. (**G**) Tension band wire is added for rotational control. The femur should be internally rotated and the anteversion of the hip confirmed with the use of fluoroscopy before the wire is tensioned. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

a tension band wire is added. A 1.8-mm K-wire drill hole is created in the lateral cortex of the distal femur, from anterior to posterior, relative to the patella forward. An 18-gauge wire is then threaded through this hole and twisted in a figure eight fashion around the proximal end of the Rush rod (Fig. 8G). The plate method is most commonly used for hips with Type 1b CFD with delayed ossification of the femoral neck. The Rush rod method is most commonly used for hips with Type 1a or 1b CFD with subtrochanteric delayed ossification.

Step 6: Pelvic Osteotomy

Because of the extensive exposure, it is not necessary to split the iliac apophysis to perform the Dega osteotomy. The ilium is exposed by elevating and retracting the hip abductor muscles

from the lateral wall of the ilium, starting anterior at the anterior inferior iliac spine and working posteriorly. The dissection is continued back to the sciatic notch and distally toward the ischium. The dissection should not cross the triradiate cartilage. The osteotomy is curved along the lateral cortex from the anterior inferior iliac spine to triradiate cartilage posteriorly. The osteotomy is inclined toward the triradiate cartilage medially and should start 2 cm above the joint. The osteotomy is levered distally to cover the femoral head. The bone graft obtained from the femur is used to hold the osteotomy open and to level the sourcil (Fig. 9).

Step 7: Tendon Repair and Closure

The conjoint abductor–quadriceps tendon is sutured directly into the cartilaginous greater trochanter with absorbable suture. This is performed with the femur in neutral abduction. The TFL is also sutured to the greater trochanter to augment the abduction strength of the hip. The incision is closed in layers, including Scarpa's fascia and subcuticular and skin layers. A suction drain is used because of the large anterior flap and is left in place until the draining stops, which can take several days. The patient is placed in a spica cast in full hip extension and neutral abduction and rotation for six weeks (Fig. 10).

INSTABILITY OF PATELLA OR TIBIA AND KNEE FLEXION CONTRACTURE

Instability of the patella necessitates performing a stabilizing–realigning procedure before lengthening. Isolated anteroposterior instability of the tibiofemoral joint without knee joint dislocation or rotatory subluxation does not need to be addressed before lengthening. Isolated subluxation or dislocation of the patella should be treated before lengthening. This reconstruction is based on a combination of elements from the Langenskiöld procedure (3) (designed for congenital dislocation of the patella), the MacIntosh procedure (4) (extra-articular reconstruction for anterior cruciate deficiency), and the Grammont procedure (5) (designed for recurrent dislocation of the patella). This knee stabilization procedure can be performed at the same time as the pelvic osteotomy or superhip procedure. When knee stabilization is performed together with a superhip procedure, the fascia lata is reflected from proximal to distal and is used for the ligamentous reconstruction.

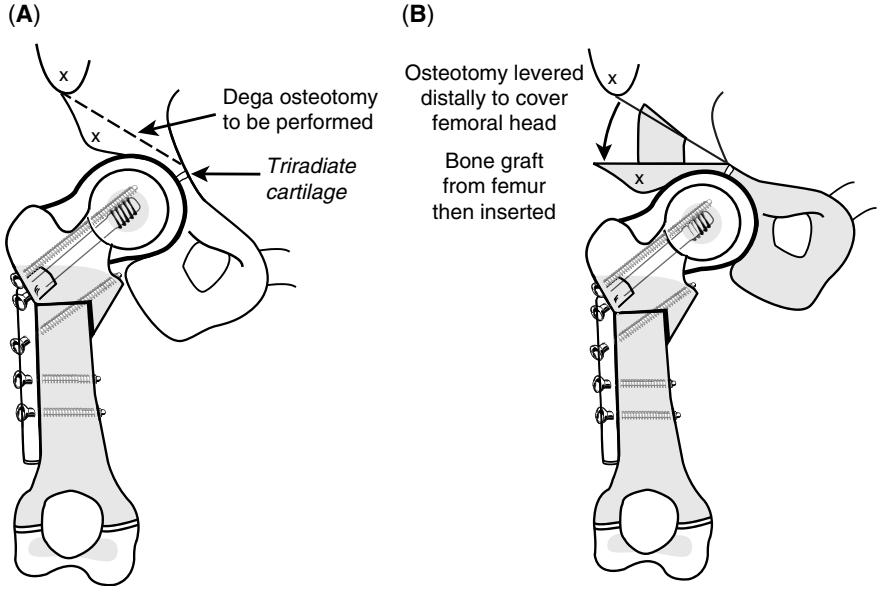


Figure 9 (**A** and **B**) Dega osteotomy is performed, exposing the outer table of the iliac crest under the abductor muscles without splitting the iliac apophysis. The osteotomy curves from the anterior inferior iliac spine to the triradiate cartilage along the lateral cortex of the iliac crest. The bone segment from the femur is used as the bone block for the Dega osteotomy. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

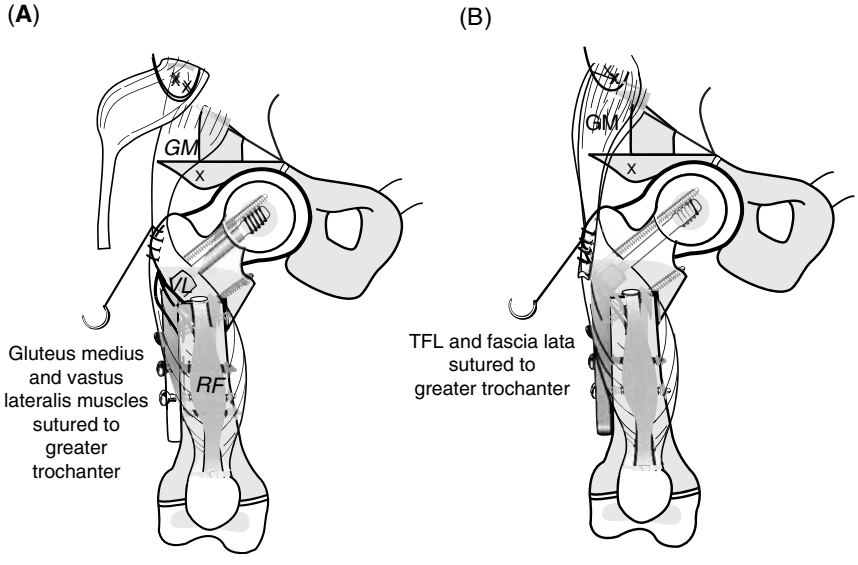


Figure 10 (**A** and **B**) Conjoint abductor-quadriceps tendon is sutured to the cartilaginous greater trochanter. The tensor lata muscle also is sutured to the greater trochanter to augment hip abduction strength. *Abbreviations*: GM, gluteus medius; VL, vastus lateralis; RF, rectus femoris tendon; TFL, tensor fascia lata. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

SUPERKNEE RECONSTRUCTION SURGICAL TECHNIQUES

The superknee procedure is a combination of patellar realignment with extra- and intra-articular knee ligament reconstruction. When needed, a posterior capsulotomy and knee flexor tendon release also are performed. Typically, the superknee consists of extra-articular or combined extra- and intra-articular anterior cruciate ligament (ACL) reconstruction (MacIntosh procedure), the reverse MacIntosh (Paley knee reconstruction) procedure (2), a posterior cruciate ligament (PCL) extra-articular reconstruction, the Grammont patellar tendon realignment, a lateral release of the patella, and a Langenskiold procedure (2) for patellar reduction. In cases with actual posterior dislocation, intra-articular PCL reconstruction using the fascia lata is performed instead of or in combination with extra-articular PCL reconstruction.

Step 1: Fascia Lata Harvest

The knee is exposed through a long S-shaped incision. The anterior margin of the fascia lata and the posterior margin, where it blends with the intermuscular septum, are incised longitudinally. The fascia lata is transected as proximally as possible and reflected distally until its insertion onto the tibia (Fig. 11A and B).

Step 2: Preparation of Fascia Lata For Ligamentization

The fascia lata should be split into two longitudinal strips to make two ligaments. A Krackow whipstitch (8) is used to run a nonabsorbable suture from the free end of the fascia lata toward the Gerdy's turbercle in a tubular fashion (Fig. 12).

Step 3: Lateral Release and Grammont Patellar Tendon Realignment

In all cases, lateral release is performed. When patellar maltracking is more significant, a Grammont procedure is performed to medially transfer the patellar tendon. When fixed subluxation or dislocation is present, the modified Langenskiöld procedure is performed. The lateral capsule should be cut to, but not through, the synovium. The vastus lateralis

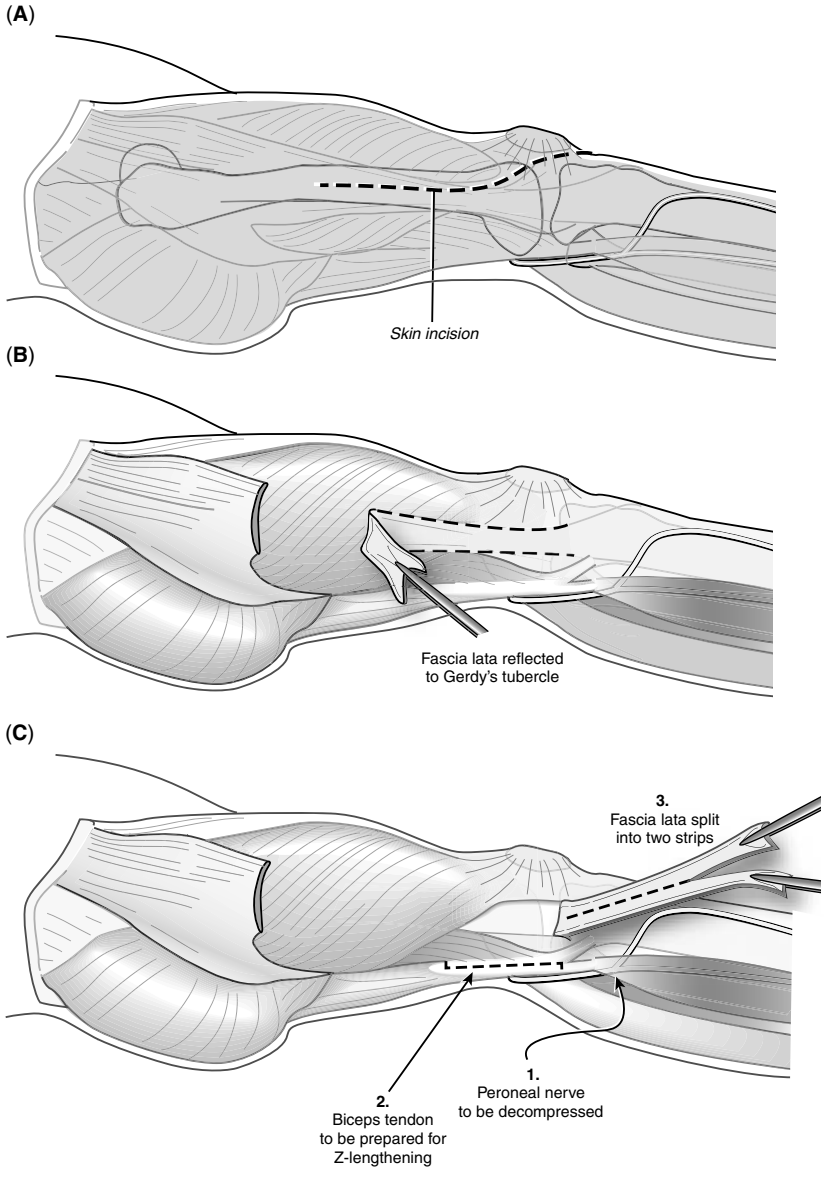


Figure 11 (**A** and **B**) S-shaped incision is performed on the lateral aspect of the distal thigh. The anterior and posterior margins of the fascia lata are identified and dissected proximally. The tensor fascia lata (TFL) is transected just distal to the TFL muscle belly. (**C**)The TFL is split longitudinally into two equal strips of tendon. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

should be elevated off the intermuscular septum. If the patella is nonetheless tethered laterally by the vastus lateralis, its tendon is released from the patella and transferred centrally to the quadriceps tendon under minimal tension. The lateral release is extended distally to the lateral aspect of the patellar tendon. If the Grammont patellar tendon medialization is to be performed, the incision should be extended past the tibial tuberosity along the crest of the tibia, incising the proximal periosteum. A parallel periosteal, para tendinous deep incision is made medial to the patellar tendon. Sharp dissection is performed with a Beaver blade scalpel to elevate the patellar tendon off the tuberosity without peeling off cartilage. The periosteal extension of the tendon is elevated with the tendon so that the detached tendon remains tethered distally. The patellar tendon can then be displaced and sutured medially (Fig. 13).

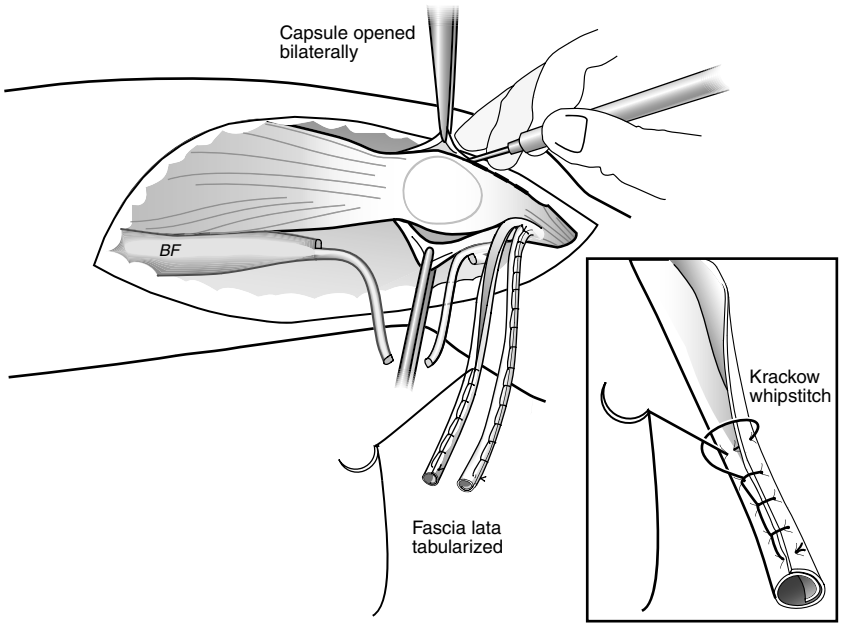


Figure 12 Tendons are then prepared with a Krackow whipstitch to form a tubular graft. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

Step 4: Macintosh Intra- and/or Extra-Articular Anterior Collateral Ligament Reconstruction

The lateral collateral ligament (LCL) is identified. A tunnel is made under this ligament without entering the knee joint (Fig. 14A). Another tunnel is made subperiosteally, from anterior

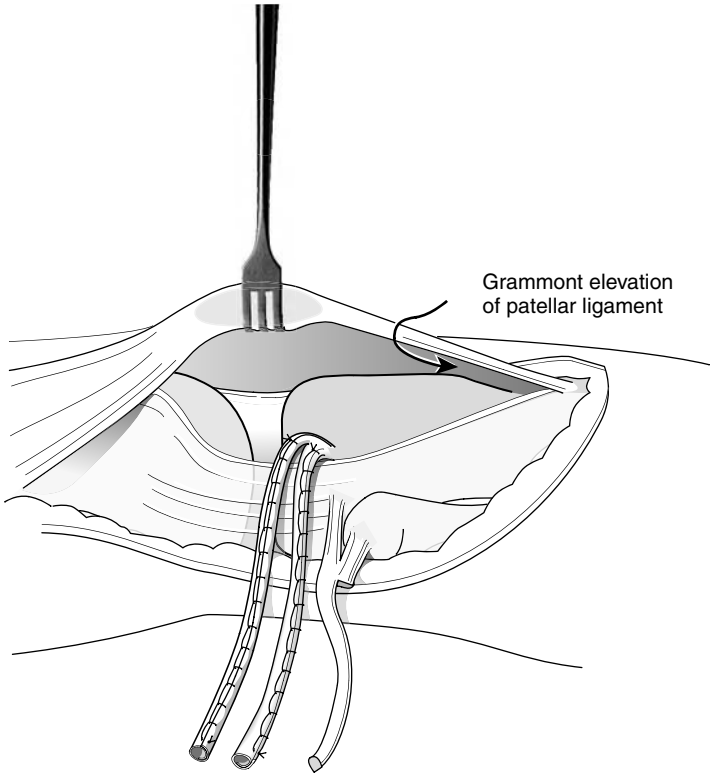


Figure 13 Grammont patellar tendon medialization is performed by incising the medial and lateral borders of the patella tendon past the tibial tubercle. The patella tendon is elevated off the tibial tubercle apophysis with an extension of periosteum that remains intact distally. The patella tendon can then be shifted medially. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

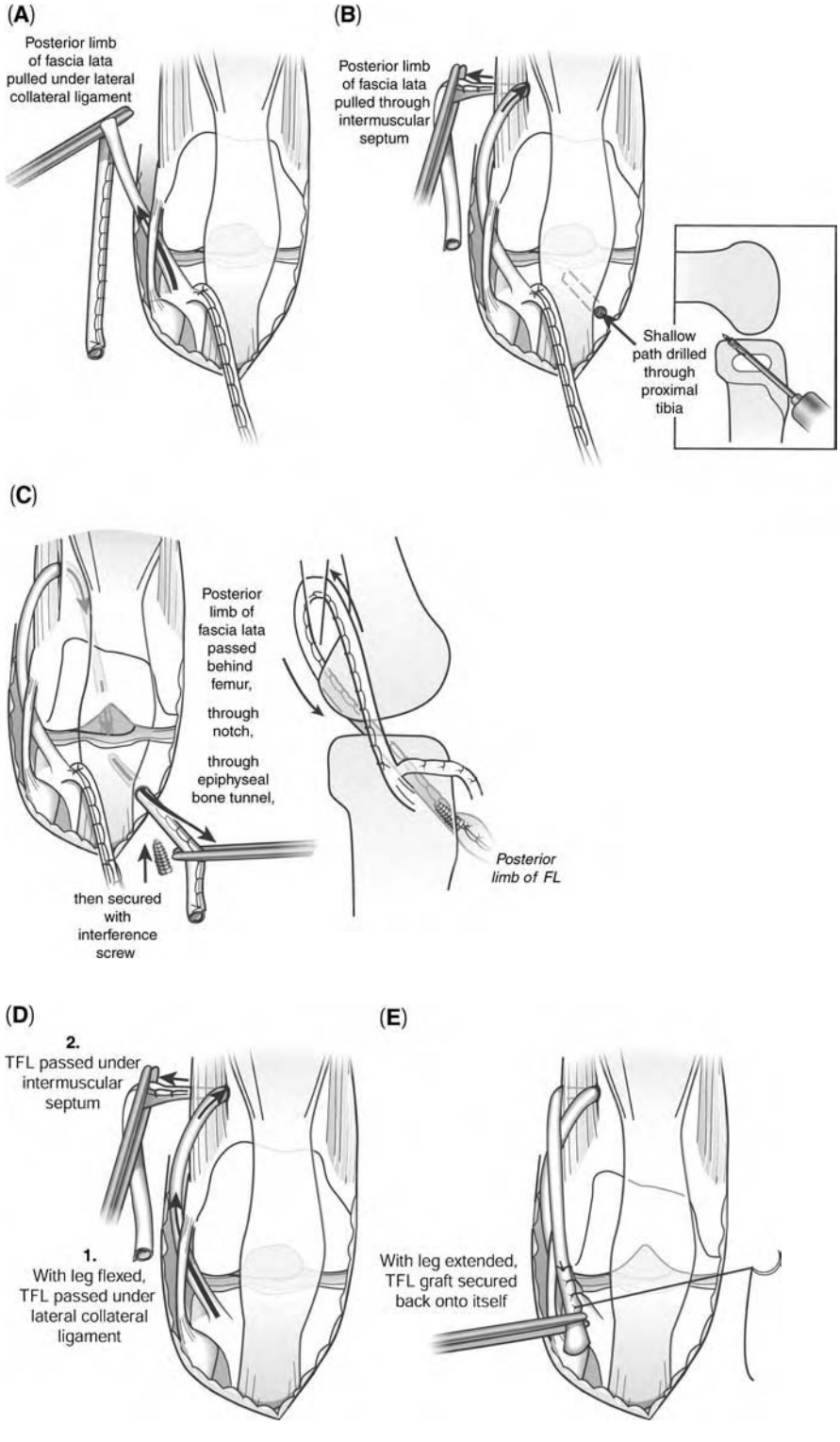


Figure 14 (*Caption on facing page*)

Figure 14 (*Figure on facing page*) (**A**) Lateral collateral ligament (LCL) and the distal aspect of the posterior intramuscular septum are identified. The posterior limb of the tensor fascia lata (TFL) graft is passed under the LCL. (**B**) Posterior limb is passed through a subperiosteal tunnel under the lateral intramuscular septum. The graft enters the subperiosteal tunnel from the anterior aspect and heads distally toward the posterior knee joint capsule. Bony tunnel is created in the proximal tibial epiphysis (inset). A wire is placed into the epiphysis medial to the patella tendon. The wire is directed toward the lateral femoral condyle and exits the tibial epiphysis at the midpoint of the ossification center. This wire is overdrilled with the appropriate size cannulated drill, depending on the graft size. (**C**) Suture passer is inserted into the bony tunnel and retrieved at the posterior aspect of the knee with a curved clamp. After the graft has been passed under the LCL and through the subperiosteal tunnel, it is pulled through the posterior joint capsule and out the tibial epiphyseal tunnel. The graft is secured with a headless bioabsorbable interference screw. (**D** and **E**) Alternatively, instead of a combined intra- and extra-articular repair, an isolated extra-articular reconstruction can be performed. The graft is then tensioned with the limb in full extension, folded back onto itself, and secured with nonabsorbable suture. *Abbreviation*: FL, fascia lata. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

and proximal to distal and posterior over the lateral intramuscular septum of the femur, directed toward the posterior knee joint capsule. A hole in this capsule is made with a curved clamp. The posterior limb of the fascia lata is passed under the LCL. A bony tunnel is made in the proximal tibial epiphysis by using an ACL reamer over a wire. The wire is inserted from anteromedial to the patellar tendon to exit in the center of the tibial plateau. The diameter of the reamer is based on the outer diameter of the ligamentized posterior limb of the fascia lata (Fig. 14B). A suture passer is passed through the tibial epiphyseal tunnel, out the posterior capsule of the knee. The suture passer is retrieved from the posterior aspect of the knee with a curved tonsil clamp. The suture attached to the fascia lata is pulled through the knee and the bony tunnel via the suture passer. It is fixed to the tunnel by using a bioabsorbable headless screw (Arthrex, Inc., Naples, Florida, U.S.A.) (Fig. 14C). If only extra-articular ACL repair is needed, the fascia lata is looped back and sutured to itself and no tunnel is made (Fig. 14D and E). The ACL graft should be tensioned in extension to prevent a flexion contracture. The extra-articular portion self-tensions as the knee flexes. To prevent loosening, the graft can be reinforced and retensioned after fixation by passing a nonabsorbable suture through bone at the point at which the graft loops over the intermuscular septum. We currently prefer to use FiberWire (Arthrex) not only to anchor the ligament but also as a ligament augmentation device used in the Krackow ligamentization stitch described above. We prefer to combine extra-articular PCL reconstruction with an intra-articular ''over the top'' ACL repair.

Step 5: Extra-Articular Posterior Collateral Ligament Reconstruction (Reverse MacIntosh)

The anterior skin flap is elevated off the knee and dissected and reflected medially until the entire vastus medialis can be visualized. The medial intermuscular septum (very rudimentary) and the adductor magnus tendon are located posterior to the vastus medialis muscle. The anterior limb of the fascia lata is passed first under the patellar tendon and through a medial capsule tunnel. The graft is then passed through a subperiosteal tunnel around the adductor magnus tendon. It is then sutured to itself with nonabsorbable suture (Fig. 15). This extra-articular ligament should be tensioned with the knee in 90° of flexion to prevent an extension contracture. If intra-articular PCL reconstruction is preferred, the lateral head of the gastrocnemius muscle is released from the femur after the peroneal nerve is protected. The posterior epiphysis of the proximal tibia is identified to the midline. An anterior to posterior drill hole is made through the epiphysis, and the anterior limb of the fascia lata is passed from anterior to posterior, exiting near the midline posteriorly. Another drill hole through the medial distal femoral epiphysis is made, oriented from anteromedial to posterolateral. The ligamentized fascia lata is passed through the posterior capsule and into the medial femoral epiphysealis tunnel. It is fixed in place with a Biotenodesis absorbable screw (Arthrex).

Alternative Step For Patellar Realignment: Langenskiöld Reconstruction

If the patella is unstable with fixed lateral subluxation or dislocation, a modified version of the Langenskiöld procedure is performed before the ligament reconstruction (4). First, the capsule is separated from the patella and synovium medially and laterally. The synovium is also

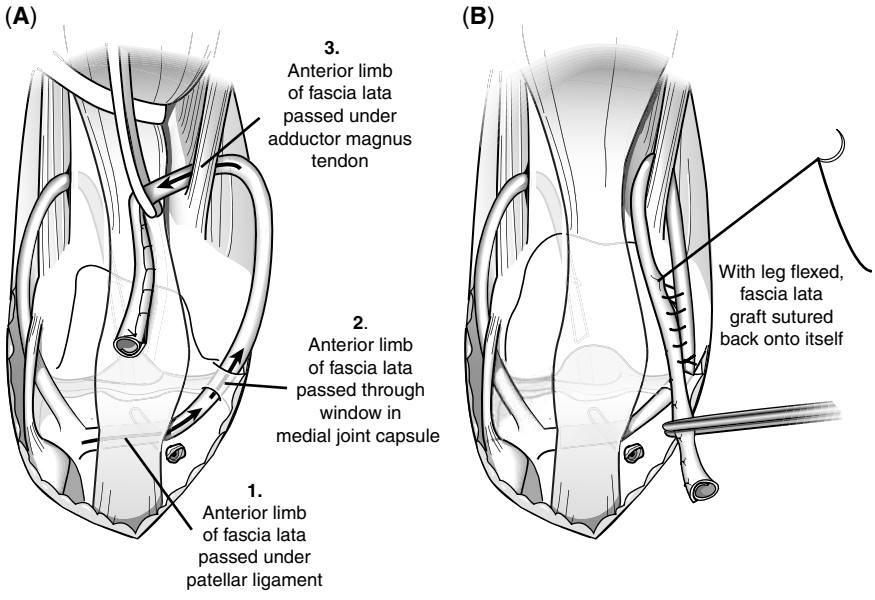


Figure 15 (**A** and **B**) Reverse MacIntosh (extra-articular posterior collateral ligament) is performed by passing the anterior limb of the tensor fascia lata graft under the patella tendon and through a window created in the medial joint capsule. The graft is then passed through a subperiosteal tunnel under the adductor magnus tendon, looped back onto itself, and secured with nonabsorbable suture. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

dissected free from the quadriceps muscle proximally and the patellar tendon distally (Fig. 16A and B). Once the synovium is completely free from its overlying tissues, it is cut from the patella circumferentially. The quadriceps and patellar tendons are left attached to the patella proximally and distally. The synovium is then a free layer with a patella-sized hole in it (Fig. 16C and D). This hole is sutured closed in a longitudinal direction, leaving the patella extra-articular. The medial gutter synovium and the synovium under the vastus medialis are dissected free from the medial capsule and retinaculum. The medial capsule is cut transversely, parallel to the joint line at its distal end. The patella tendon is elevated from the apophysis sharply after incising the medial and lateral borders of the tendon distally into the periosteum. The patella tendon is shifted medially at least 1 cm (Grammont procedure, Step 3) and is sutured in position at the end of the soft tissue reconstruction (Fig. 16E and F). A longitudinal incision is made in the synovium more medially, with the knee in extension. The patella is inserted into this new hole, and the synovium is sutured to the patella circumferentially. The medial capsule is advanced over the top of the patella and is sutured to its lateral border. The lateral capsule is left open (Fig. 16G–K). If the reverse MacIntosh procedure is performed, the fascia lata should not be fixed in place until after the Langenskiöld repair is completed (Fig. 16L and M).

Alternative Step: Knee Flexion Contracture Release

If a knee flexion deformity of more than 10° is present, it can be treated by posterior capsular release. To avoid direct surgical and indirect stretch injury to the peroneal nerve, this nerve is identified and decompressed at the neck of the fibula. Next, the biceps tendon is Z-lengthened and the lateral head of the gastrocnemius muscle is released from the femur. The lateral capsule is identified and incised at the level just proximal to the joint line. The posterior capsule is dissected free of the posterior popliteal fossa contents, from a lateral to medial direction (Fig. 17A and B). A similar dissection is performed from the medial side after taking down the medial head of the gastrocnemius muscle. To expose the medial side, the anterior skin flap is reflected medially as thick as possible to preserve its circulation. The medial and lateral dissections are connected. Great care must be taken to avoid injury to the popliteal vessels, which lie adjacent to the medial half of the posterior capsule. The entire posterior capsule is then divided under direct vision from both sides (Fig. 17C and D). The knee FFD can be corrected by extending the knee.

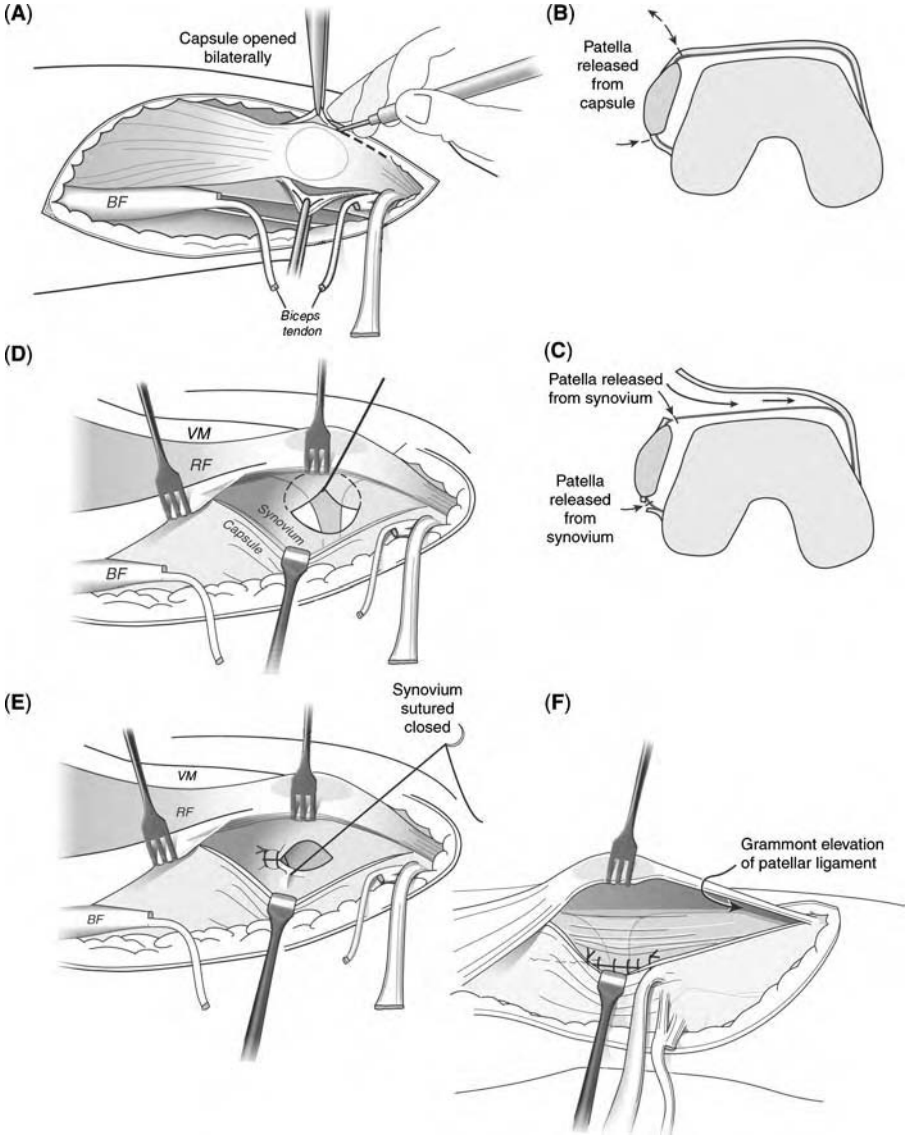


Figure 16 (**A** and **B**) Initial step in the modified Langenskiöld reconstruction is to perform a medial and lateral capsulotomy. The knee joint capsule is dissected away from the synovium medially and laterally. The synovium also is dissected free from the quadriceps tendon and the patella tendon. (**C** and **D**) Synovium is released from the patella circumferentially, leaving the quadriceps and patella insertions intact. (**E** and **F**) Hole in the synovium is closed longitudinally with absorbable suture, leaving the patella with the quadriceps and patella attachments extra-articular. The Grammont elevation and Grammont medial patellar tendon shift are then performed (see Step 3 and Fig. 13). (**G**–**K**) Knee is positioned in full extension, and the new position for the patella is marked on the synovium. A longitudinal incision is created in the synovium. The synovium is sutured to the patella in a circumferential fashion. The medial capsule is advanced and sutured onto the lateral side of the patella. The lateral capsule is not repaired. (**L** and **M**) If an extra-articular posterior collateral ligament (reverse MacIntosh) is performed, the Langenskiöld reconstruction is completed before the ligamentous reconstruction. Tensor fascia lata graft passes through the advanced medial capsule is sutured onto itself. *Abbreviations*: m, muscle; VM, vastus medialis; RF, rectus femoris tendon; BF, biceps femoris muscle. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

The collateral ligaments are left intact. The biceps femoris Z-lengthening is repaired (Fig. 17E and F). In addition, if a muscular contracture of the medial hamstrings is present, a small posteromedial incision can be made to recess the semitendinosis and semimembranous tendons.

The above hip and knee procedures are termed "preparatory surgery for lengthening" and must be performed before femoral lengthening. Preparatory surgery prevents complications that

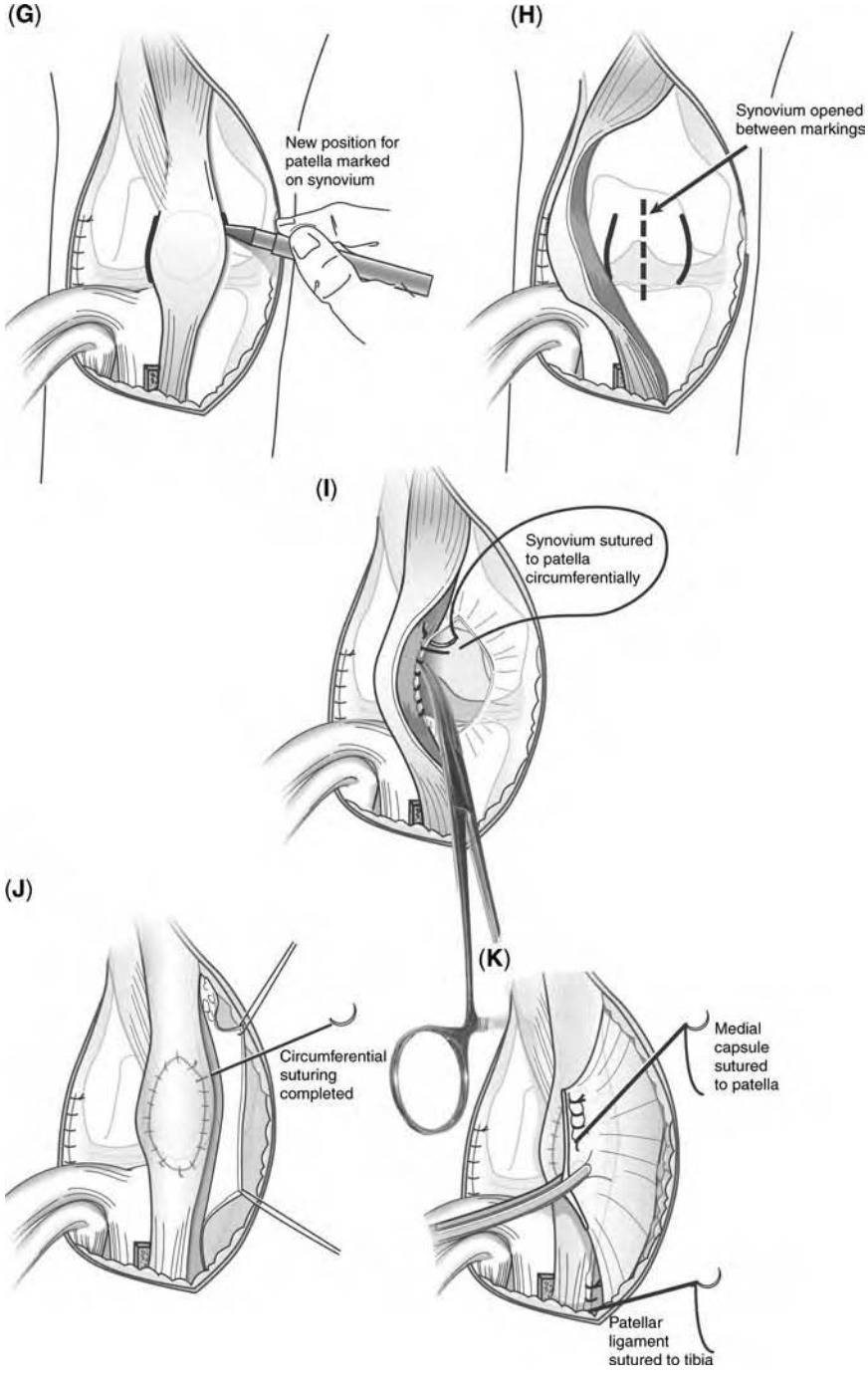
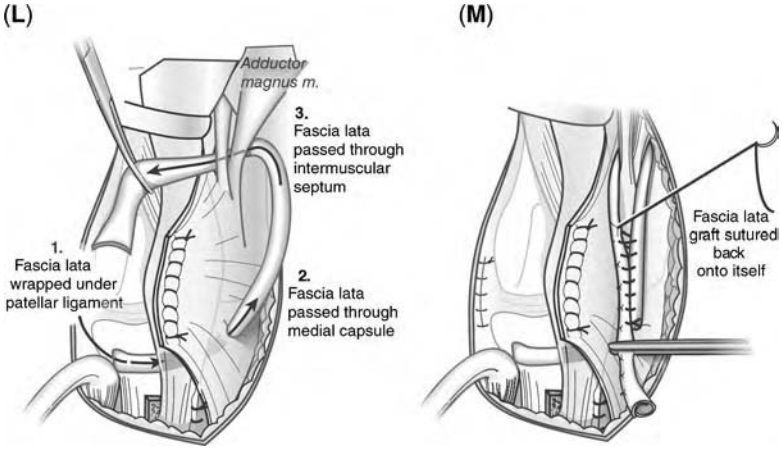


Figure 16 (*Continued*)

could occur during the lengthening process. The pelvic osteotomy prevents hip subluxation/dislocation. The proximal femoral reconstruction (superhip) prevents worsening of the coxa vara deformity, proximal migration of the femur, and dislocation of the hip. The patellar realignment prevents patellar dislocation and extension knee contracture. The ACL–PCL reconstruction prevents knee subluxation/dislocation and late problems of knee instability in adolescence. The fascia lata excision prevents posterolateral rotatory subluxation of the knee, knee contractures, increased pressure on the joint and physis, and valgus deformity of the knee.



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FEMORAL LENGTHENING OF TYPE 1 CONGENITAL FEMORAL DEFICIENCY

Choice of Osteotomy Level for Lengthening of the Congenitally Short Femur

With Type 1 CFD, it is preferable to lengthen the femur by using a distal femoral osteotomy. Distal osteotomies have a broader cross-sectional diameter for better bone formation and are less prone to the deforming forces from the adductors and hamstrings. Distal osteotomy lengthening is closer to the knee joint and therefore has greater effect on knee range of motion and on knee subluxation. Proximal osteotomies have less effect on knee range of motion but are more prone to poor bone formation. Also, when using the external-fixator-only method of bone lengthening, a higher rate of fracture after removal of external fixation is encountered in the proximal lengthening groups when compared with the distal lengthening groups. Proximal osteotomies should be reserved for the technique of lengthening over nails (LON) because the nail prevents deformation of the proximal femur, both during and after removal of fixation, prevents fracture, and allows early removal of the external fixator despite incomplete consolidation of the regenerate bone.

Associated deformities of the hip and knee need to be considered when choosing the level of osteotomy. The external rotation deformity of the femur associated with CFD should be corrected at a proximal osteotomy level. The quadriceps muscle is in a normal relationship to the knee joint and originates distal to the level of a proximal femoral osteotomy. Therefore, a proximal femoral internal rotation osteotomy does not disturb the quadriceps orientation relative to the knee joint. In comparison, a distal osteotomy leaves the bulk of the quadriceps muscle attached proximally in a lateral position and rotates the knee medially, increasing the effective Q angle and the tendency to laterally displace the patella (Fig. 18). A varus deformity of the hip or the proximal diaphysis is corrected by using a proximal osteotomy, and a valgus deformity of the distal femur is corrected by using a distal osteotomy. These deformities can be addressed at the time of the lengthening by performing acute correction with either a proximal or distal osteotomy, as noted.

The proximal femoral osteotomy should be used for varus correction or derotation. Lengthening should not be performed through the proximal osteotomy because of the poor regenerate bone formation. In cases in which a proximal femoral osteotomy is performed, a distal osteotomy is also performed for lengthening.

The distal femoral osteotomy is used to correct the distal femoral valgus deformity (Fig. 19). This region of the femur has a wider cross-sectional area, producing a regenerate bone that is wider, stronger, and subjected to less bending forces than in the proximal femur.

In older children with wider medullary canals (>7 mm), the LON technique can be performed (9,10). A proximal osteotomy can be used for lengthening with this technique because it involves little risk of refracture with a rod in the medullary canal. A greater trochanteric starting point is used, along with an appropriate size Rush rod with a proximal bend (9). Fixator-only lengthening is the method used for the first lengthening. The LON technique or fully

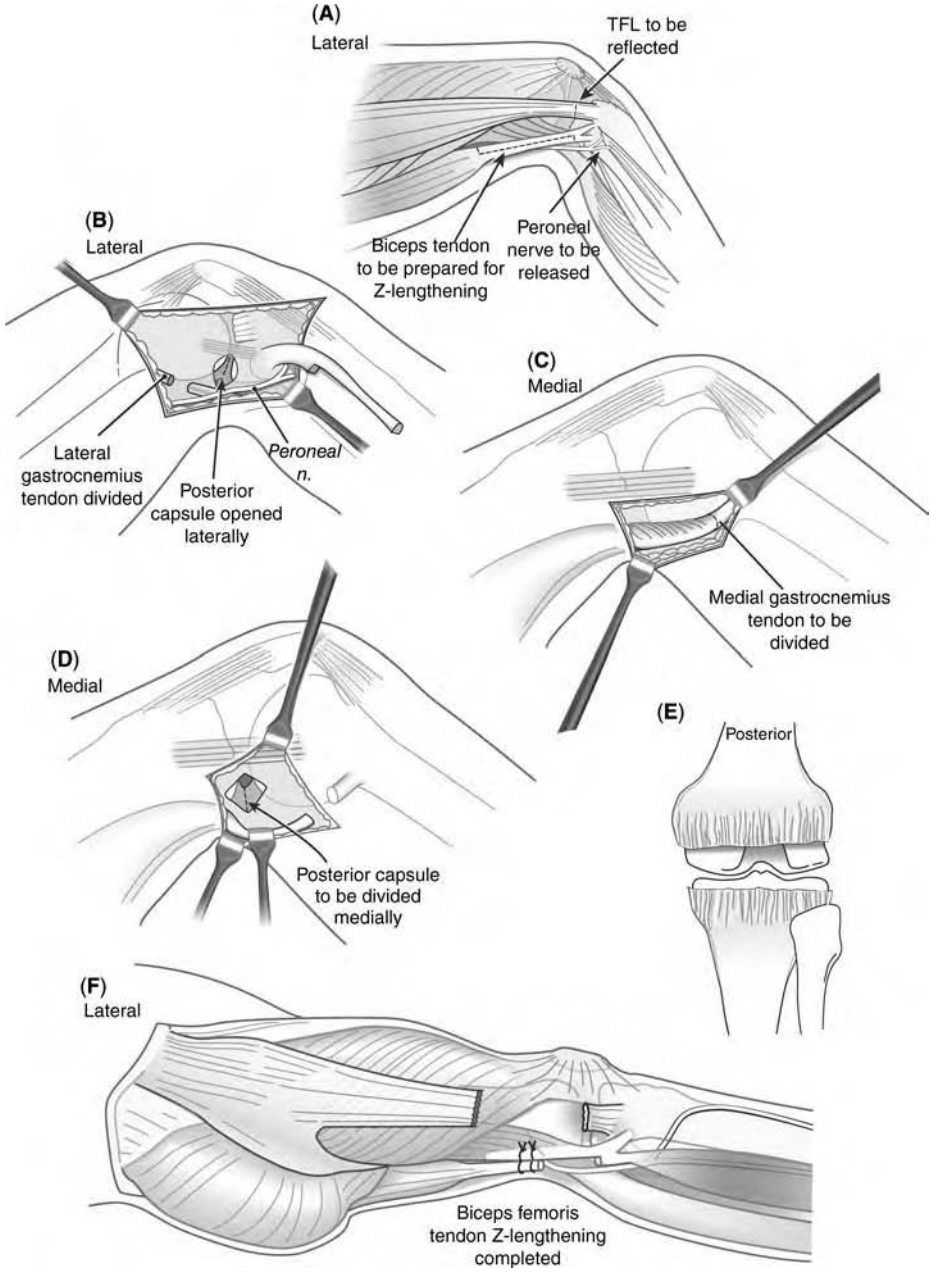

Figure 17 (**A** and **B**) Knee flexion contracture release is performed through the same S-shaped lateral incision as previously described. The peroneal nerve is identified and decompressed, and then a Z-lengthening of the biceps femoris tendon is performed. The lateral collateral ligament and lateral gastrocnemius tendon are identified. The lateral head of the gastrocnemius muscle is released, and the posterior joint capsule is identified. The posterior joint capsule is incised proximal to the joint line. (**C** and **D**) Medial side of the knee joint is exposed either by dissecting the anterior flap medially or through a separate medial incision. After the exposure is completed, the medial gastrocnemius muscle is released and the posterior capsule identified. Again, the posterior capsule is dissected free from the popliteal fossa contents and incised proximal to the joint line. The posterior capsule is divided under direct visualization from both the lateral and medial dissections. (**E** and **F**) Knee flexion deformity can then be corrected by gently extending the knee. The biceps femoris tendon lengthening is then repaired. *Abbreviation*: n, nerve. *Source*: Figure copyrighted to the Rubin Institute for Advanced Orthopedics.

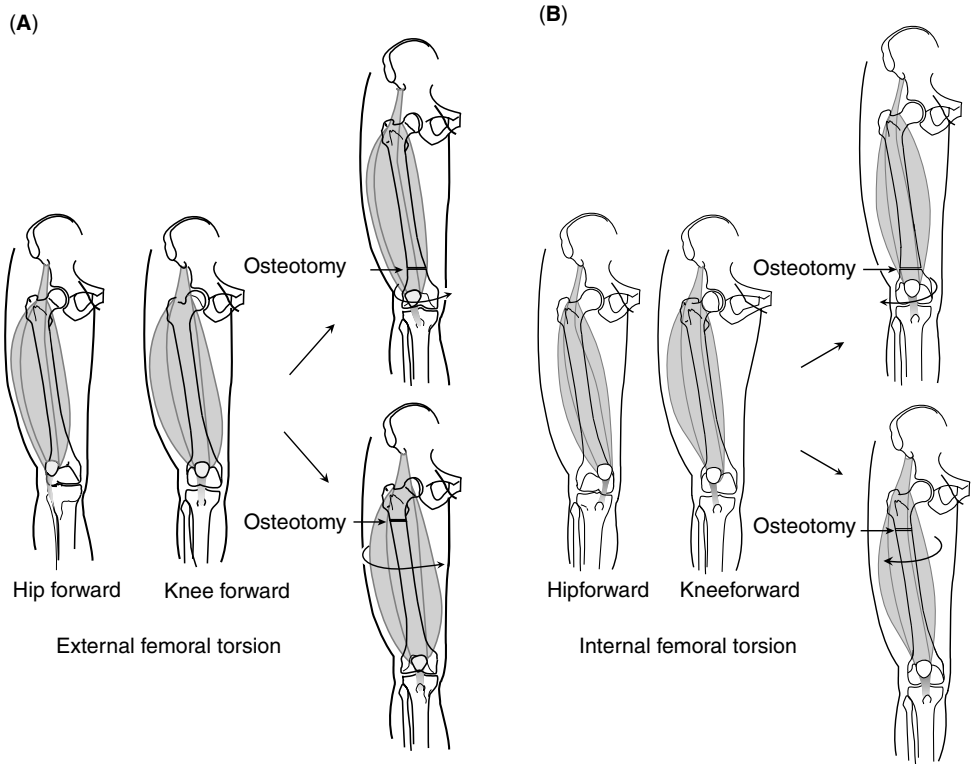

Figure 18 (**A** and **B**) Congenitally short femur with marked external rotation deformity. Femur is derotated through a proximal osteotomy (*lower right*). Femur is derotated through a distal osteotomy (*upper right*). Modified from Ref. 7.

implantable lengthening with an intramedullary lengthening device [e.g., intramedullary skeletal kinetic distractor (ISKD) (11)] is often the method for the second or third lengthening if the anatomic dimensions permit. The LON method allows the fixator to extend across the knee joint with hinges, as does the fixator-only method. The implantable lengthening nail method must rely on splinting the knee in extension between therapy sessions to prevent knee subluxation. The ISKD lengthener, currently the only implantable lengthening nail approved by the U.S. Food and Drug Administration, often lengthens faster than desired. This increases the risk of subluxation. To use this device safely, we limit the total amount of lengthening to no more than 5 cm. Most knee subluxations do not occur before 4 cm of length has been achieved. With external fixation-only lengthening, lengthening 8 cm and more can be performed safely as long as the knee range of motion can be maintained from full extension to more than 45° of flexion. With implantable lengthening, more frequent, shorter lengthenings are accepted as a tradeoff for the convenience and advantages of not wearing an external fixator. LON offers the advantages of both, by shortening the external fixation time while permitting as much lengthening with knee protection as with the external-fixation-only method.

Soft Tissue Releases for Lengthening

Soft tissue releases are essential in conjunction with lengthening to prevent subluxation and stiffness of the knee and hip. If a superhip or superknee reconstruction has already been performed, soft tissue releases that were addressed at the previous surgery do not need to be repeated. Soft tissue releases are performed at the time of the index procedure.

Before surgery, the range of motion of the hip and knee should be evaluated and the presence of contractures identified. The muscle lengthening tests are the straight leg raising test (popliteal angle) for the hamstrings, the prone knee flexion test (Ely test) for the rectus femoris, and hip abduction range for the adductors. If a popliteal angle of more than 0° or a prone knee bend less than the supine knee bend (or pelvic flexion with prone knee

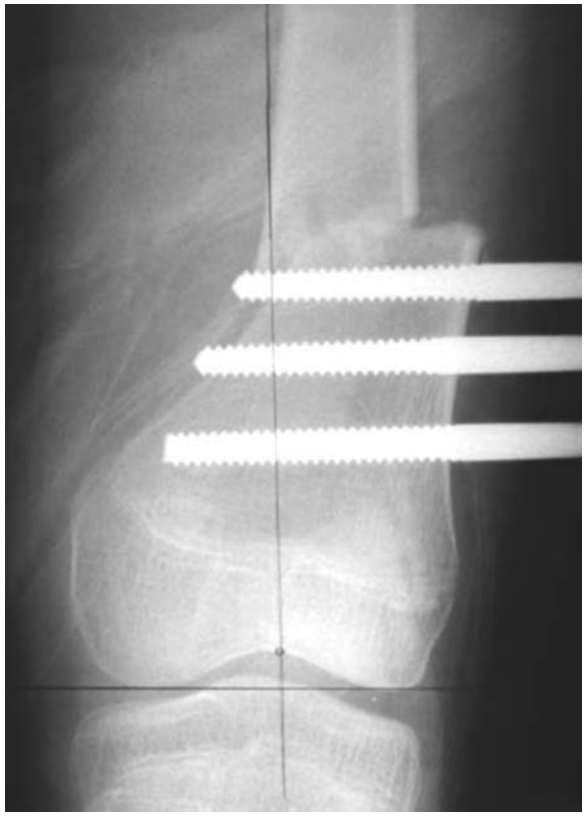

Figure 19 Radiograph shows acute valgus correction performed at the osteotomy site for lengthening.

flexion-positive Ely test) is present, the hamstrings and rectus femoris, respectively, are tight and will lead to contractures during lengthening. The medial and lateral hamstrings should be recessed proximal to the knee, to reduce the popliteal angle to 0°. The rectus femoris tendon should be released off the anterior inferior iliac spine through a small anterior inguinal incision. Finally, if hip abduction is limited, especially with proximal lengthening, percutaneous adductor tenotomies should be performed of the adductor longus and gracilis tendons. During lengthening, if a severe adduction contracture develops or the hip begins to subluxate, a more extensive open adductor release (including the adductor brevis) is performed. Woolf and Gross (12) have also recommended distal release of the adductor magnus tendon.

If the fascia lata has not been excised, a release is performed. The entire fascia lata, including the posterior intermuscular septum, is transected at the level of the proximal pole of the patella. The lateral biceps can also be safely recessed through the same incision, if needed. The incision for this combined release of the biceps and fascia lata should be made laterally over the top of the intermuscular septum. The proximal TFL is occasionally released as it passes over the greater trochanter. This is rarely performed at the index procedure. This procedure is usually performed in a delayed fashion to treat hyperlordosis of the lumbar spine, hip flexion contracture, and hip abduction contracture resulting from the lengthening. The proximal fascia lata can be released through the same small incision used to release the rectus femoris tendon. For distal femoral lengthening, it is not necessary to release the hip adductors. Again, proximal femoral lengthening usually requires hip adductor release in a delayed fashion unless a contracture is present at the index surgery.

Botulinum toxin is injected in the quadriceps muscle at the time of surgery. This reduces muscle spasm and pain, allowing improved knee flexion range of motion during physical therapy (13).

Knee Instability Consideration

All cases of CFD can be assumed to have hypoplastic or absent cruciate ligaments, with mild-to-moderate anteroposterior instability. Some knees also have mediolateral and torsional

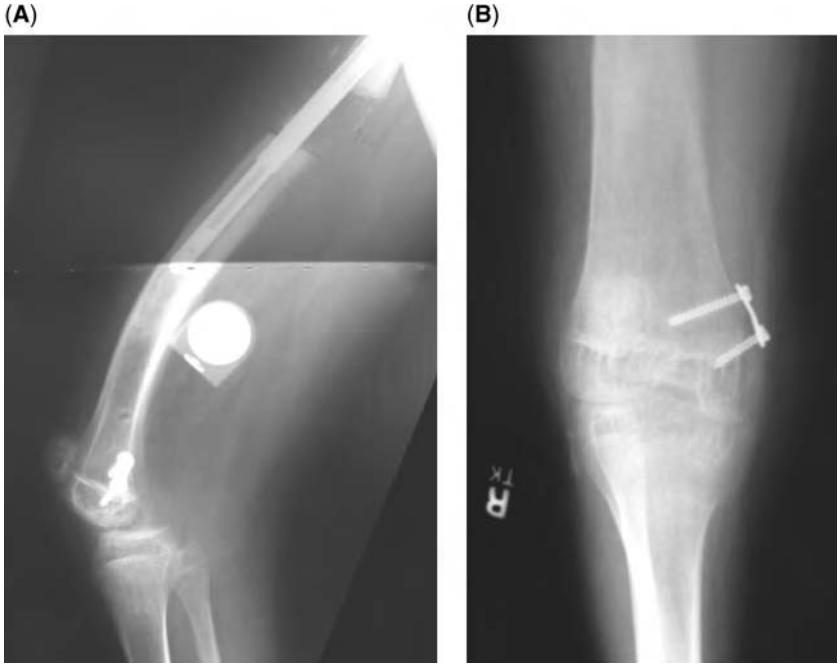

Figure 20 Patient with congenital femoral deficiency undergoing femoral lengthening with an intramedullary lengthening nail. Lateral view (**A**) and anteroposterior view (**B**) radiographs show knee flexion contracture with concurrent posterior-lateral subluxation. Aggressive therapy and soft tissue releases can be used to solve this complication. If an external fixator is being used for lengthening with this complication, lengthening is halted immediately. Fortunately, the intramedullary nail had completed the programmed 5 cm of lengthening at the time this radiograph was obtained.

instability. Despite this, the knee is shown to have normal tracking preoperatively, leaving no indication to perform ligamentous reconstruction. The significance of this knee instability is the tendency of the knee to subluxate with lengthening. Knee subluxation with lengthening is usually posterior or posterolateral (posterior plus external rotation of the tibia on the femur) (Fig. 20). Rarely, this subluxation can be anterior. Knee extension maintains a reduced knee position. Therefore, to prevent posterior subluxation, some authors recommend splinting the knee in extension throughout the distraction phase (14). This promotes knee stiffness while protecting the knee from subluxation. The knee should be protected by extending the fixation to the tibia with hinges. The hinges permit knee motion while preventing posterior and anterior subluxation. This hinge configuration is easily performed with the Ilizarov circular fixator (Fig. 21). More recently, a technique to extend the fixation to the tibia with a hinge by using a monolateral external fixator has been developed (Fig. 22).

A less common pattern of knee instability is anterior subluxation/dislocation of the tibia on the femur. This occurs when the knee is fully extended. This instability pattern is partly caused by an anterior deficiency of the distal femur. Although one treatment option is extension osteotomy of the knee, the superknee procedure is our recommended treatment to avoid loss of knee flexion and to provide the most normal anatomy and stability of the knee.

Distal Femoral Lengthening: Ilizarov Fixator Technique

The necessary soft tissue releases are performed first, as described above. If a proximal femoral derotation, valgus, and/or extension osteotomy is needed, the proximal pin is inserted into the proximal femur with the hip in the position in which it will lie after the correction. For example, with correction of varus, flexion, and external rotation, the femur should be externally rotated and crossed over the other thigh to adduct and flex the hip. This places the hip in the true neutral position. The first half-pin is inserted from lateral to medial in the frontal plane, parallel to the line from the tip of the greater trochanter to the center of the femoral head (proximal femoral joint orientation line) (Fig. 23). The plan is to attach the proximal arch

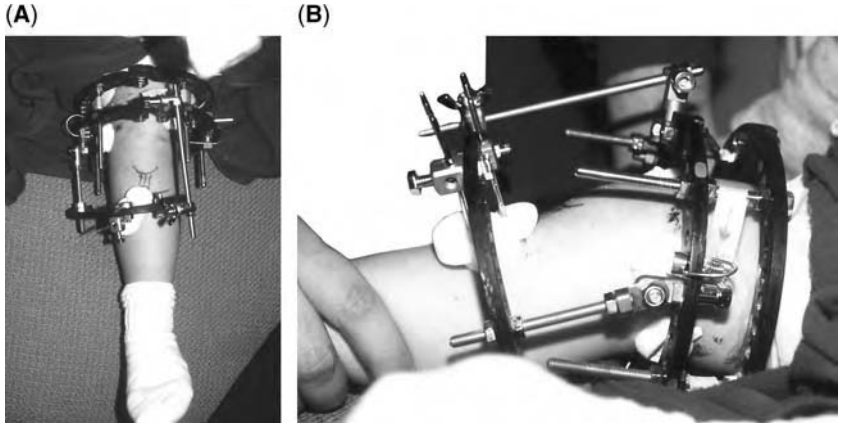

Figure 21 (**A** and **B**) Clinical photographs show an Ilizarov external fixator being used for femoral lengthening in a patient with congenital femoral deficiency. Note the distal femoral ring bridged across the knee joint with hinges to prevent subluxation and dislocation.

parallel to the proximal femoral joint orientation line, the middle ring perpendicular to the mechanical axis of the femoral diaphysis (7° to the anatomic axis), and the distal ring parallel to the knee joint line. After the osteotomies, when all the rings and arch are parallel, the mechanical axis of each segment will be aligned and the joint orientation of the hip and knee will be parallel. A second proximal half-pin is inserted on the proximal arch from 30° anterolateral to

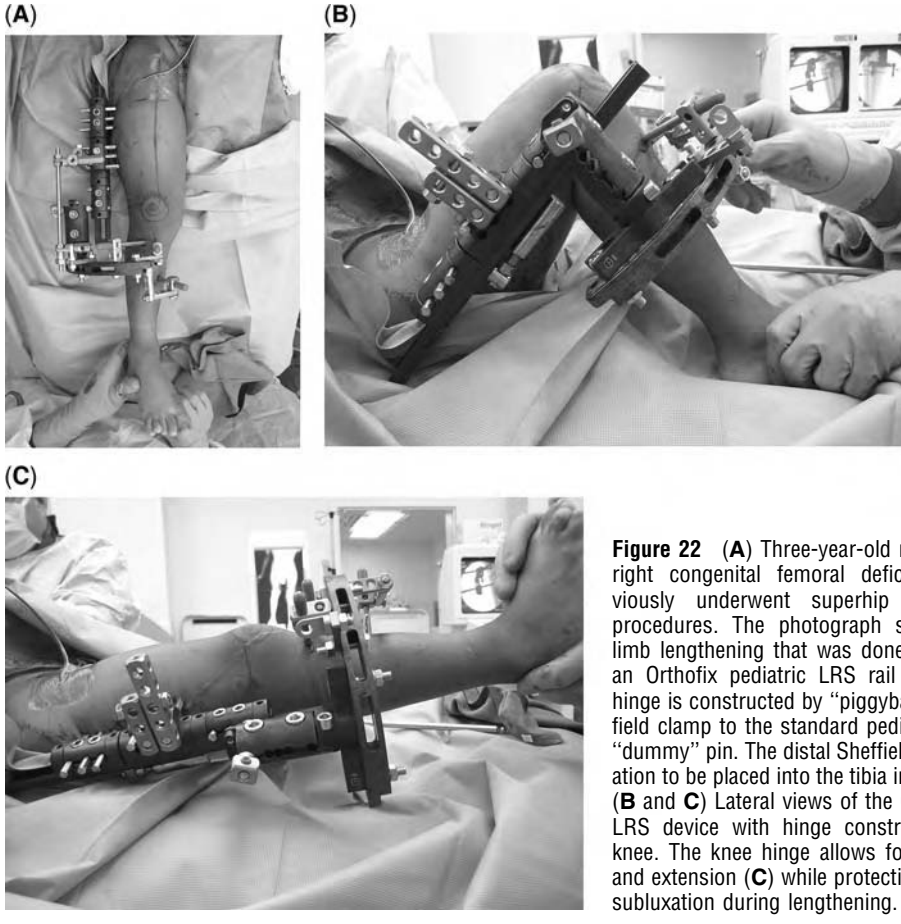

Figure 22 (**A**) Three-year-old male patient with right congenital femoral deficiency who previously underwent superhip and superknee procedures. The photograph shows the initial limb lengthening that was done with the use of an Orthofix pediatric LRS rail system. A knee hinge is constructed by ''piggybacking'' the Sheffield clamp to the standard pediatric clamp via a ''dummy'' pin. The distal Sheffield arch allows fixation to be placed into the tibia in multiple planes. (**B** and **C**) Lateral views of the Orthofix pediatric LRS device with hinge construct bridging the knee. The knee hinge allows for full flexion (**B**) and extension (**C**) while protecting the knee from subluxation during lengthening.

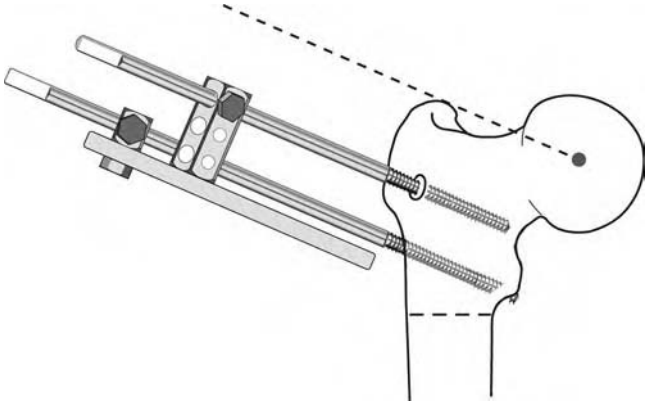

Figure 23 First pin is inserted in the frontal plane from lateral to medial. The pin is directed parallel to the proximal orientation line from the tip of the greater trochanter to the center of the femoral head. *Source*: Modified from Ref. 7.

the first pin. The proximal arch is perpendicular to the floor with the limb crossed over and externally rotated. This allows correction of the varus, flexion, and external rotation deformities. Two Ilizarov rings are applied to the distal femoral reference wire, parallel to the knee joint. For young children, arthrograms are obtained for the purpose of outlining the cartilaginous femoral condyles. This also allows visualization of the posterior femoral condyles in the sagittal plane for hinge placement. Conical washers or hinges are used between the two distal rings because of the valgus of the distal femur. The rings are at the valgus deformity angle to each other. A lateral half-pin is inserted into the midsegment of the femur and attached to the middle ring. This pin is at 7° to the shaft of the bone. The proximal subtrochanteric osteotomy can then be performed. The osteotomy is performed percutaneously by creating multiple drill holes and completing the osteotomy with an osteotome. The osteotomy is internally rotated, laterally translated, and then angulated into valgus and extension to correct all components of the deformity. The order of correction is important to achieve the necessary displacement without loss of bone-to-bone contact and stability (Fig. 24). Two additional half-pins are inserted and fixed onto the distal ring, one from posteromedial and one from posterolateral between the quadriceps and the hamstring muscles. An additional pin is placed in the middle segment. In small children, all half-pins are inserted by using the cannulated drill technique. This technique permits very accurate placement of large diameter pins in narrow bones to avoid eccentric placement. Eccentric placement of drill holes and half-pins in the femoral diaphysis leads to fracture. Next, the distal femoral osteotomy is performed percutaneously, with multiple drill holes and an osteotome. The previously placed reference wire is removed to avoid tethering of the quadriceps and fascia lata.

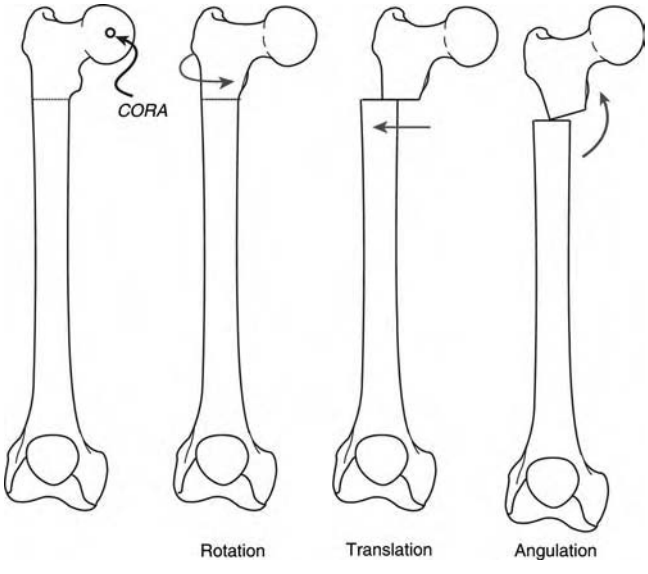

Figure 24 Sequence of deformity correction is essential to obtain the correction and maintain stability and bone contact. *Source*: Modified from Ref. 7.

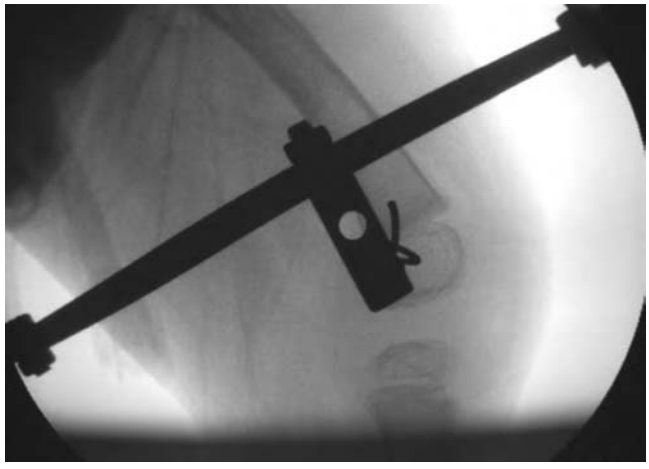

Figure 25 Intraoperative fluoroscopic view of Ilizarov hinge placement at the point of intersection of the posterior femoral diaphyseal line and the distal femoral physis.

Knee Hinges

The last step is to extend the fixation to the tibia by using hinges placed at the center of rotation of the knee. The center of rotation of the knee is located at the intersection of the posterior femoral cortical line and the distal femoral physeal line (15) in the plane in which the two posterior femoral condyles are seen to overlap on the lateral view (Fig. 25). For younger children, an arthrogram of the knee allows visualization of the posterior femoral condyles. The distal femoral ring, which is parallel to the knee, must appear to be perpendicular to the X-ray beam. The medial and lateral skin is marked at the location of the planned hinge placement. Two threaded rods with female hinges attached are dropped from the distal femoral ring to the marked level of the knee hinge. A single half-ring is attached to two threaded rods and attached to the hinges. This half-ring is oriented perpendicular to the tibia with the knee in full extension. The first half-pin is inserted from anterior to posterior into the tibia. After this pin is secured to the proximal tibial half-ring, the knee is flexed and extended through a range of motion. If this range feels frictionless (perform a drop test: drop the tibia to see whether it flexes without any catch), second and third tibial half-pins are added. Finally, a removable knee extension bar is inserted between the distal femoral ring and the tibial half-ring.

Distal Femoral Lengthening: Orthofix Fixator Technique

Start by identifying the center of rotation of the knee joint (see description above). A 1.8-mm wire is drilled into the lateral edge of the physis at the intersection of the posterior cortex of the femur, with the physis in line with the plane of overlap of the posterior femoral condyles (Fig. 26). The Orthofix (McKinney, Texas, U.S.A.) LRS rail system (pediatric or adult) is lined up with the hinge axis through its most distal clamp hole. A commercially available sandwich clamp is used. (If this is unavailable, one can fashion a sandwich clamp by using two pin clamp lids held to the pin clamp body by a 30-mm Ilizarov bolt and the convex conical washer for centering in the hole.) The sandwich clamp provides a second layer of pin holes more anteriorly (Fig. 27). The fixator bar is aligned with the shaft of the femur and the most proximal half-pin inserted into the proximal femur. The distal-most pin is then drilled one hole proximal and anterior to the center of rotation reference wire. For ease of application, an LRS without sandwich clamps is used to place the remaining pins (three proximal and three distal). If distal femoral valgus is present and is to be corrected acutely, a swivel clamp is used for distal pin placement. When using a pediatric LRS, the three-hole pin clamp contains only two half-pin sites because one hole is occupied for the knee center of rotation wire. A third pin is added by using an Ilizarov cube connected to the two pins (Fig. 28). Before reapplying the sandwich clamps, the osteotomy is performed and the distal valgus is corrected acutely. After the correction, the fixator can be exchanged for another fixator with straight clamps and sandwich attachments. All the pins should be in the upper deck of the double-decker sandwich clamp distally. The only pin in the lower deck is the knee axis pin. The knee axis pin is a dummy pin that does not enter the patient's limb. It is a segment of pin that protrudes laterally from the

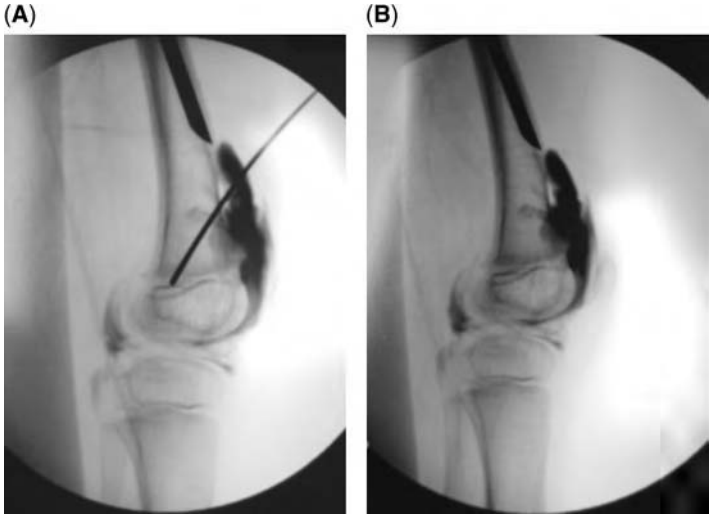

Figure 26 (**A** and **B**) Intraoperative fluoroscopic images show arthrography of the knee. The lateral view is obtained, and the posterior aspects of the femoral condyles are superimposed to create the perfect lateral view. The hinge reference wire is inserted at the intersection of the posterior femoral cortical line and the distal femoral physis. This marks the center of rotation of the knee joint.

clamp. A Sheffield clamp from Orthofix is applied to this pin to act as a hinge. It is locked in place by placing a cube lateral to it with a setscrew to prevent it from moving laterally. Conical washers are used between the Sheffield clamp and the LRS to reduce friction (Fig. 29). The Sheffield clamp is left partially loose to permit motion. A one-third Sheffield arch is attached to the clamp, arching toward the tibia. An anteroposterior pin is inserted, and the drop test is performed (see above). If friction occurs during the drop, the Sheffield clamp should be loosened. If friction persists, adjust the connection of the tibial pin to the Sheffield arch. If friction further persists after the adjustment, the axis pin might need to be bent slightly to alter the axis of rotation. Once the drop test is negative, two more oblique pins are inserted into the tibia and connected to the Sheffield arch by using cubes (Fig. 30). A removable knee extension bar is fashioned from Ilizarov parts to be used especially at nighttime. The easiest way to accomplish this is to apply a cube to the protruding ends of the distal pins and build off this cube with a post (Fig. 31). If an unstable hip is present, a hinge axis pin for the hip can also be placed and attached to the proximal clamp. The same Sheffield clamp and arch arrangement are used. Three pins are placed in the pelvis, one through the anterior–inferior and one through the anterior–superior spines extending posteriorly. One is inserted laterally. These are fixed to the Sheffield clamp to prevent proximal subluxation of the hip during lengthening (Fig. 32). As one can see, the principles applied when using monolateral fixation are the same as those for circular fixation (i.e., hinge fixation across joints when a joint is at risk).

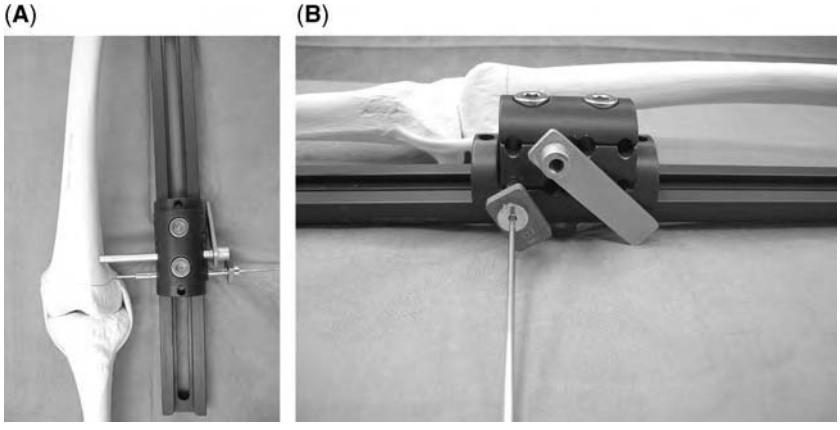

Figure 27 (**A** and **B**) Bone model shows LRS sandwich clamp placed distally, with the most distal hole containing the hinge axis wire. The first distal half-pin is placed on the anterior row one hole proximal to the hinge axis pin.

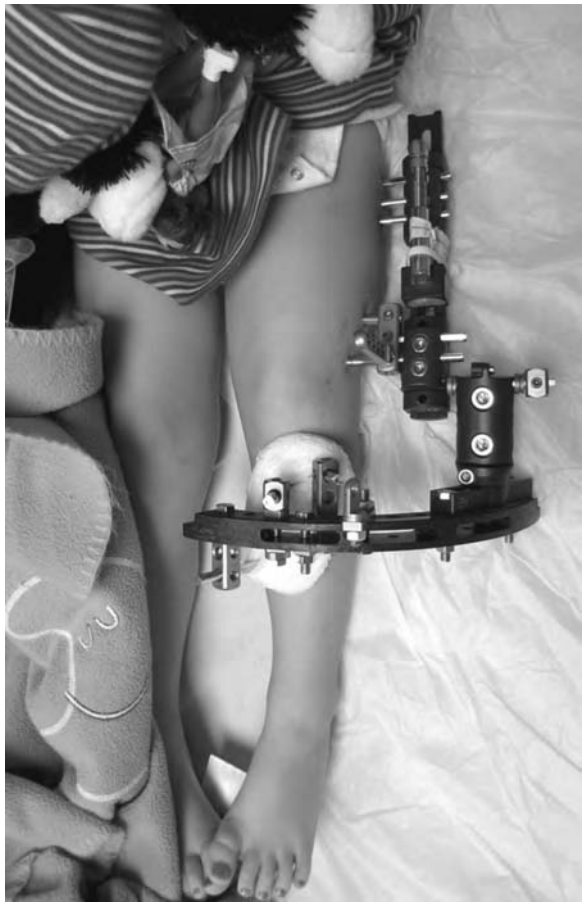

Figure 28 Example of pediatric Orthofix rail with a three-hole cube placed on the distal half-pins to allow a third half-pin to be inserted into the distal fragment.

REHABILITATION AND FOLLOW-UP DURING LENGTHENING

Femoral lengthening requires close follow-up and intensive rehabilitation to identify problems and maintain a functional extremity. Follow-up is usually conducted every two weeks for radiographic and clinical assessments. Clinically, the patient is assessed for hip and knee range of motion, knee subluxation, nerve function, and pin site problems. Radiographically, the distraction gap length, regenerate bone quality, limb alignment, and joint location are assessed.

Physical therapy is begun within one or two days after surgery and should continue daily throughout the distraction and consolidation phases. Physical therapy is briefly discontinued

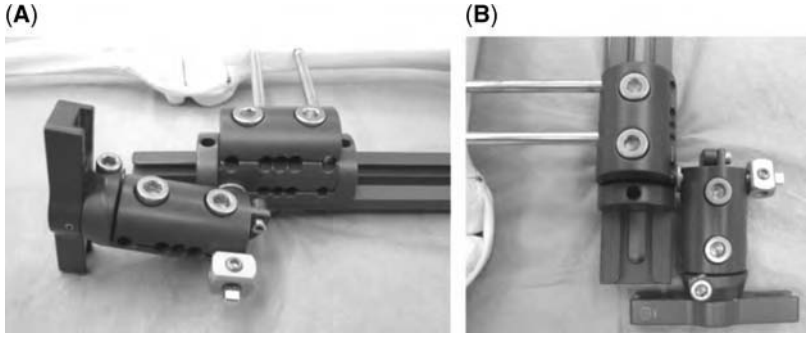

Figure 29 (**A** and **B**) Bone model with a dummy pin inserted into the distal posterior hole of the LRS sandwich clamp that replaces the hinge axis wire. A Sheffield clamp is attached to the hinge dummy pin to create the knee hinge.

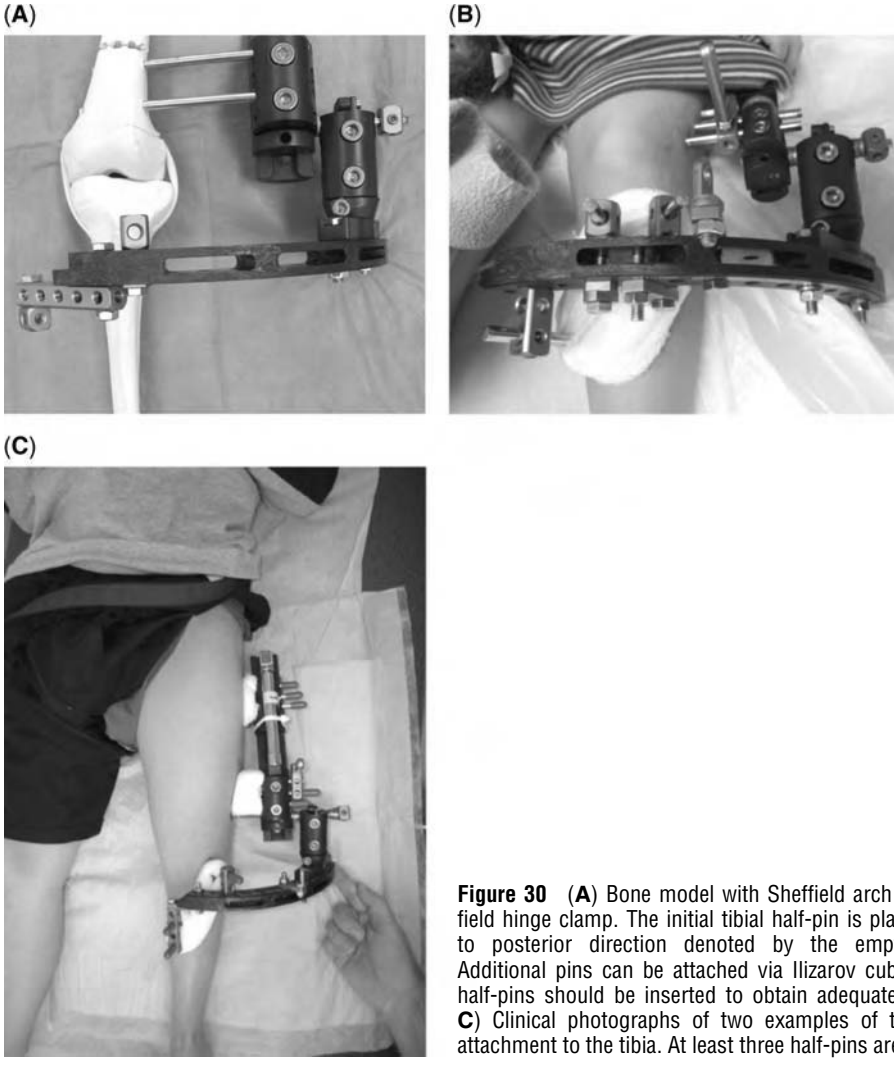

Figure 30 (**A**) Bone model with Sheffield arch attached to Sheffield hinge clamp. The initial tibial half-pin is placed in an anterior to posterior direction denoted by the empty Ilizarov cube. Additional pins can be attached via Ilizarov cubes. At least three half-pins should be inserted to obtain adequate stability. (**B** and **C**) Clinical photographs of two examples of the Sheffield arch attachment to the tibia. At least three half-pins are used for fixation.

after removal of the external fixator to avoid a fracture through the regenerate bone or a pin hole. Once the bone is strong enough, physical therapy is continued. During the distraction phase, one or two formal sessions with a therapist are required each day (45–60 minutes each session). In addition, at least two home sessions per day (30 minutes each) are recommended. The more physical therapy is performed, the better the functional result will be. The philosophy of therapy for

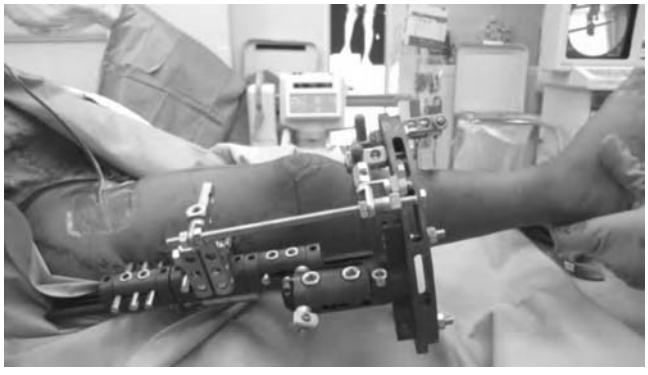

Figure 31 Clinical photograph shows a completed Orthofix external fixator for femoral lengthening in a patient with congenital femoral deficiency. The knee extension bar is constructed by building Ilizarov cubes from the half-pins to the Sheffield arch. Sockets are used to connect the extension bar to the frame, which allows for easy removal of the bar during physical therapy.

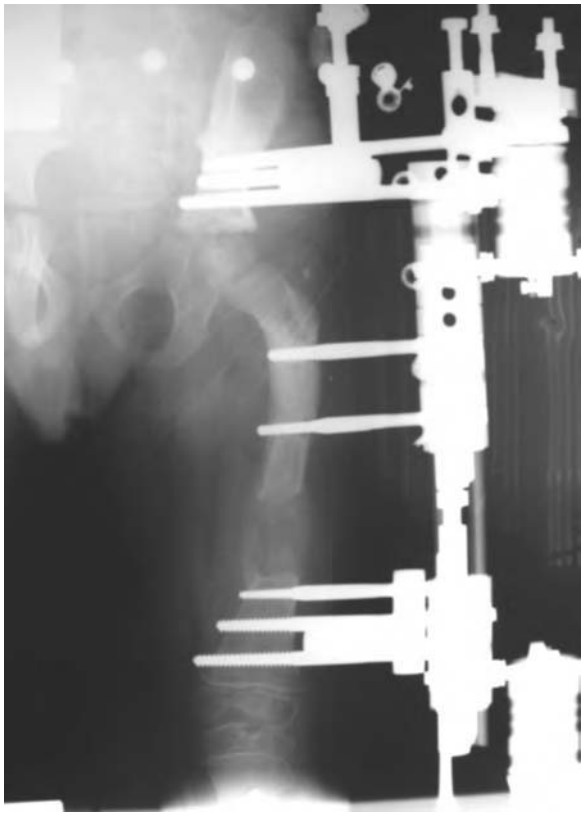

Figure 32 Radiograph shows a monolateral Orthofix external fixator bridging across the hip joint to prevent subluxation. The hip hinge is created by using a dummy half-pin at the center of hip rotation, which is connected to a Sheffield clamp and arch.

lengthening is very different from that for other orthopedic surgical procedures. With most orthopedic procedures, the patients are at their worst after surgery and recover gradually. Patients undergoing lengthening are at their best one week after surgery. Thereafter, because of the distraction, the muscles become tighter and the range of motion of the joints is more limited. It is not until the consolidation phase that the usual pattern of rehabilitation and recovery occurs. One can think of the lengthening surgery ending at the end of the distraction phase: a surgical procedure that can be measured in months rather than hours. In the absence of a therapy program, we will not even consider performing femoral lengthening.

The majority of the therapy time should be spent obtaining knee flexion and maintaining knee extension. Passive exercises are most important during the distraction phase, and passive plus active exercises are most important during the consolidation phase. Hip abduction and extension are the two important hip exercises. Strengthening exercises should be focused on the hip abductors and the quadriceps. Electric stimulation is applied to the quadriceps muscle. Upper extremity strengthening is helpful for use of walking aids and transfers. Weight bearing is encouraged and allowed as tolerated.

Knee flexion should be maintained at more than 45°. If knee flexion is 40° or less, the lengthening should be discontinued or at least slowed and knee rehabilitation should be increased. If, after a few days, the knee flexion improves, lengthening can resume. Our motto is "never sacrifice function for length." More length can be obtained with subsequent lengthenings, but a new knee joint cannot be created. Therefore, preserving the knee joint and its motion is most important. Flexion contracture might develop during lengthening. To prevent this, a knee extension bar can be used at night and part-time during the day. FFD of the knee places it at risk for posterior subluxation. Subluxation of the knee can be suspected clinically based on a change in shape of the front of the tibia relative to the patella. Posterior subluxation of the tibia presents with a very prominent patella and a depression of the tibia relative to the patella (ski hill sign). Extension of the external fixation across the knee with hinges prevents posterior subluxation.

Hip motion can become more limited with lengthening. Adduction and flexion contractures are the most significant because they lead to hip subluxation and dislocation. Release of

the adductors, rectus, and TFL during lengthening might need to be considered to allow further lengthening. Usually, the TFL and the rectus femoris tendon are addressed during the preparatory surgery.

Nerve injury from surgery or distraction is unusual with femoral lengthening. To avoid peroneal nerve injury from the pins, the posterolateral pin should not enter posterior to the biceps tendon. During distraction, if the patient complains of pain in the dorsum of the foot or requests frequent massaging of the foot, referred pain from stretch entrapment of the peroneal nerve is most likely the cause. More advanced symptoms include hyper- or hypoesthesia in the distribution of the peroneal nerve and weakness of the extensor hallucis longus muscle. Quantitative sensory testing, if available, is the most sensitive test to assess for nerve involvement. If the nerve problem is identified early, it can be treated by slowing the rate of distraction. If, despite slowing the distraction, symptoms continue or motor signs develop, the peroneal nerve should be decompressed at the neck of the fibula. This release should include transverse fasciotomy of the lateral and anterior compartments and release of the intermuscular septum between these compartments (7).

Hypotrophic regenerate formation requires slowing the distraction rate. Overabundant bone formation can lead to premature consolidation and requires increasing the distraction rate for a few days. A mismatch between the increase in the distraction gap from one visit to the next and the number of millimeters of distraction performed during the same time period is a sign of an impending premature consolidation. Radiographs are used to assess joint location. A break in Shenton's line or increased medial–lateral head-teardrop distance indicates subluxation of the hip. In the knee, posterior or anterior subluxation can be monitored on the lateral radiographic view with full knee extension (16). Limb length equalization should be based on full-length standing radiographs. Limb alignment is assessed for the femur and tibia separately and in combination. Separately, the joint orientation of the knee should be measured by using the malalignment test (7). Axial deviation from lengthening (procurvatum and valgus for distal femoral lengthening and procurvatum and varus for proximal lengthening) is identified and corrected at the end of the distraction phase, when the regenerate bone is still malleable. When malalignment of the femur and tibia is present, the femoral malalignment is corrected to a normal distal femoral joint orientation. The femur is not over- or undercorrected to compensate for the tibial deformity. The tibia should be corrected separately, either during the same treatment or at a later treatment.

Complete failure of bone formation is very unusual. Partial defects are not uncommon. The most common location of regenerate defects is lateral. Dynamization of the fixator should be performed, and bone growth stimulators can be used. Resection of the fibrous tissue in these defects and cancellous bone grafting might become necessary to reduce the external fixation time and prevent fracture after frame removal.

Once the regenerate bone is deemed healed, the frame can be removed from the femur and tibia. Previously, we applied a one-legged spica cast to prevent fracture. Despite application of the cast, we experienced a 34% fracture rate of the femur after removal, compared with a 9% fracture rate for all other bones and for noncongenital femoral lengthening. Because of this high fracture rate, prophylactic Rush rod placement in the femur is now performed at the time of external fixation removal. This new protocol has virtually eliminated the complication of refracture after lengthening. Because a rod is being inserted into the femur at the time of removal of the external fixator, intramedullary infection is a concern. Despite this concern, we have observed only two cases in which local deep pin bone infection occurred. The infections were easily treated by curettage of the lateral cortex of the bone combined with removal of the Rush rod and administration of antibiotics. Prophylactic rodding permits continuation of knee mobility after removal. Formal physical therapy is delayed for a month, but the patient is permitted gentle knee range of motion. This is to protect from osteoporotic stress fractures through the tibial pin holes. The prophylactic rod placement also permits weight bearing with a removable spica cast immediately after frame removal.

DIFFERENCES IN TREATMENT OF TYPES 1a AND 1b CONGENITAL FEMORAL DEFICIENCY

In general, Type 1a CFD (normal ossification) has less hip, proximal femoral, and knee deformity, deficiency, and discrepancy than does Type 1b (delayed ossification). Most Type 1a cases do

not require the complex superhip reconstruction. Approximately one-half of the Type 1a cases do require pelvic osteotomy before lengthening. All CFD Type 1a cases require extension of the fixator across the knee to protect the knee joint. The distinction between Types 1a and 1b should be made while the patient is in infancy because the natural history of Type 1b is to ossify. Therefore, adult Type 1b cases generally appear to be severe Type 1a cases. The strategy of treatment for Type 1b is to correct all the associated deformities, which will allow the proximal femur and hip to be more normally oriented and accept more axial loading. The response to the anatomic change is ossification of the proximal femur and conversion from Type 1b to Type 1a. We do not perform lengthening in Type 1b cases until they convert to Type 1a. This conversion usually occurs within two years of the superhip procedure. Our preference is to perform the first lengthening when the patient is between the ages of two and four years. Patients with Type 1a CFD typically undergo their first lengthening at the age of two years, whereas patients with Type 1b CFD typically undergo lengthening closer to the age of four years.

TREATMENT OF TYPES 2 AND 3 CONGENITAL FEMORAL DEFICIENCY

Detailed discussion of the treatment of Types 2 and 3 CFD are complex and beyond the scope of this textbook. For the purposes of this text, we provide only a summary of our strategy for Types 2 and 3.

Type 2 Congenital Femoral Deficiency

With Type 2 CFD, the deciding factor is the presence or absence of a mobile femoral head in the acetabulum. Despite magnetic resonance imaging, arthrography, and other studies, often, the only definitive way to determine the presence or absence of a mobile femoral head is to open the hip joint capsule and examine the femoral head for mobility. If the femoral head does not move in the acetabulum, it should not be joined to the femoral shaft. If it is joined to the femoral shaft, effectively, arthrodesis of the hip is created. If the femoral head is mobile, it can be connected to the remainder of the femur by a complicated reconstruction of the femoral neck, which we call ''superhip 2.'' This converts Type 2a to Type 1a, and further treatment is as for Type 1a. If no femoral head is present or if it is stiff in the acetabulum, only the superhip soft tissue release is performed to release the flexion contracture of the hip and excise the fascia lata. Serial lengthenings with fixation to the pelvis are performed until skeletal maturity, at which time the last lengthening is combined with a pelvic support osteotomy. The only difference between pelvic support osteotomy in cases of CFD and that for other conditions (17) is that the fixation is extended to the pelvis.

Type 3 Congenital Femoral Deficiency

Type 3a CFD can be treated like Type 2b, with hip release, serial lengthenings, and pelvic support osteotomy, or can be treated by prosthetic fitting options, including prosthetic reconstruction surgery. Prosthetic reconstruction surgery refers to Syme amputation and rotationplasty. For Type 3b CFD, which includes a stiff knee joint ($<45^\circ$ of motion), we are more likely to recommend prosthetic reconstruction surgery. In Type 3a cases, when the knee joint has a functional range of motion, converting Type 3a to Type 2b can be considered. The extent of treatment required for reconstruction in such cases entails four or more lengthenings and should not be considered lightly. Rotationplasty provides more predictable functional results than does lengthening for such severe cases. When rotationplasty is chosen, our preference is to use a modification of the rotationplasty described by Brown (18). This includes fusion of the distal femoral remnant to the pelvis. Paley's modification is to perform the fusion to the inferior iliac surface of a Chiari osteotomy instead of to the lateral ileum as described by Brown. This medializes the limb and provides a broader surface for fusion. The femoral segment is fixed to the pelvis by using screws crossing the physis to produce a distal femoral epiphysiodesis. Distal femoral epiphysiodesis is required to prevent distal migration of the rotated knee joint (functionally now the hip joint). When Syme amputation is chosen, the superhip approach to eliminate contractures around the hip, combined with knee flexion contracture release, improves the posture of the short thigh and knee joint to enable better prosthetic fitting.

AGE STRATEGIES (TABLE 1)

The majority of Type 1 CFD cases require at least two lengthenings (Table 1). Because the expected discrepancy at skeletal maturity increases, the number of lengthenings required to equalize LLD increases. Our preferred strategy is to perform the first lengthening when the patient is between the age of two and four years. We have found that children between the age of 4.5 and 6 years are often not at the optimal age psychologically to deal with limb lengthening. Their cognitive level is insufficient to understand why their parents allowed someone to do this to them, despite that they are beginning to be more independent and might seem to be mature enough to handle the process. The younger children do much better because their cognitive level accepts everything their parents decide without question. Children between 4.5 and 6 years of age seem to understand too little and too much at the same time. They do not connect their recognition that they have a short limb with the solution of limb lengthening. Beyond the age of six years, children enter the age of reason and begin to understand that they are different from other children and that they have a problem for which there is a solution. They learn to accept the solution by reason rather than by faith. Their cooperation is voluntary rather than coerced. The amount of lengthening that can be attained in the femur at any one stage usually is between 5 and 8 cm. This lengthening amount seems to be independent of the initial length of the femur and age of the child. Generally, 5 to 8 cm can be safely attained in toddler (age two to four years) femora as well as in older children and adult femora, despite that the percentage of lengthening is greater in the toddler. Combined femoral and tibial lengthenings allow greater total lengthening amounts. Tibial lengthening of 5 cm or lower can be combined with a 5-cm femoral lengthening. Femoral lengthening in children younger than six years can be associated with sustained growth stimulation (19). By beginning lengthening at a young age, we are able to reduce one or more levels of prosthetic/orthotic need. This means going from a knee-ankle-foot orthosis to an ankle-foot orthosis and shoe lift, or from an ankle-foot orthosis and shoe lift to a shoe lift only, or from a shoe lift to no lift. The complication rate in this young age group is no higher than in older children, in our experience.

Adults (age 18–60 years) with CFD whose parents refused prosthetic reconstruction surgery for them when they were children have undergone limb lengthening. We were able to successfully equalize their limb lengths with one or two lengthenings, depending on the discrepancy (the most severe case underwent 25 cm of equalization two LON treatments). Therefore, adult CFD residua are not contraindications to treatment.

Table 1 Treatment Strategies and Timing of Reconstructive Stages in Management of Congenital Femoral Deficiency

Predicted LLD at maturity	Number of procedures	Timing and amount of lengthening
≤6 cm	1 Lengthening	Age > 6 yr
7–12 cm	2 Lengthenings	Age 2–4 yr, ≤6 cm; age 8–14 yr, < 8 cm
	1 Lengthening	Age 2–4 yr, ≤6 cm or age 6–10 yr, < 8 cm + epiphysiodesis of < 5 cm
12–16 cm	2 Lengthenings	Age 2–4 yr, < 7 cm or age 6–8 yr, 6–8 cm + age 10–14 yr, 8 cm
16–20 cm	2 Lengthenings	Age 2–4 yr, ≤6 cm or age 6–8 yr, < 8 cm + age 10–14 yr, 8 cm + tibia < 5 cm during one femoral lengthening
	3 Lengthenings	Age 2–4 yr, ≤6 cm + age 8–10 yr, 6–8 cm + age 10–14 yr, 8 cm
	2 Lengthenings	Age 2–4 yr, ≤6 cm + age 10–14 yr, 8 cm + epiphysiodesis of < 5 cm
21–25 cm	3 Lengthenings	Age 2–4 yr, < 5 cm + age 8–10 yr, 6–8 cm + age 12–14 yr, 10–12 cm
	3 Lengthenings	Age 6–8 yr, < 8 cm + age 10–12 yr, 8 cm + age 12–16 yr, 8–12 cm + tibia < 5 cm during one femoral lengthening
	2 Lengthenings	Age 6–8 yr, < 8 cm + age 10–12 yr, 8 cm + epiphysiodesis of 5 cm + tibia < 5 cm during one femoral lengthening + epiphysiodesis of < 5 cm
> 25 cm	3 Lengthenings 4 Lengthenings	+ epiphysiodesis of 5 cm

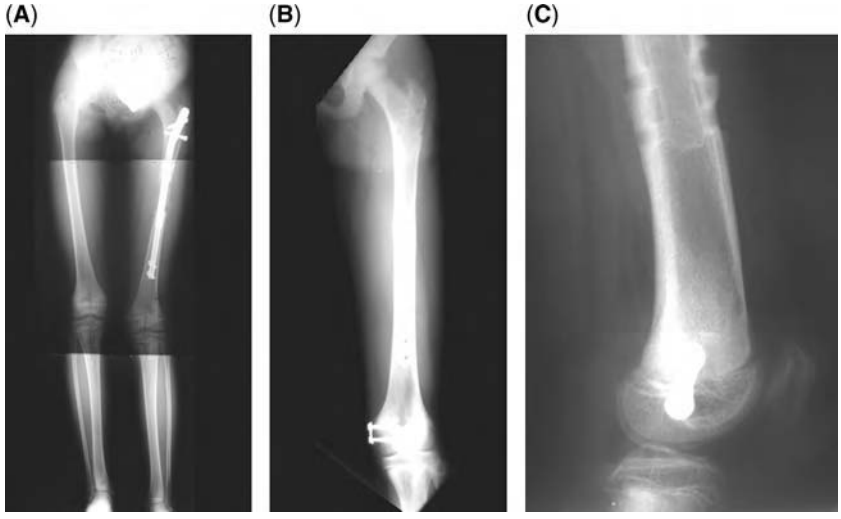

Figure 33 (**A**–**C**) Radiographs of a 10-year-old female patient undergoing femoral lengthening with an intramedullary lengthening nail and concurrent genu valgus secondary to congenital femoral deficiency. At the time of intramedullary skeletal kinetic distractor nail removal, a distal femoral hemiepiphysiodesis is performed with the Orthofix eight-Plate device for gradual correction of the valgus deformity.

ROLE OF EPIPHYSIODESIS AND HEMIEPIPHYSIODESIS

Epiphysiodesis is used as an adjuvant method to equalize LLD. It should be calculated into the total strategy of equalization surgeries. Epiphysiodesis should be used for 5 cm or less of LLD equalization. Judicious use in some cases might avoid the need for one lengthening (e.g., predicted LLD = 11 cm; plan 6 cm of lengthening before age four years and 5 cm of epiphysiodesis near puberty). Calculation of the timing of epiphysiodesis can be achieved by using the Anderson and Green method (20) or Moseley (21) method or more simply and accurately by using the multiplier method (22,23).

Hemiepiphysiodesis is very useful to correct the valgus deformity of the knee from distal–femoral or proximal–tibial origins. We currently prefer to use the Orthofix eight-Plate device, developed by Dr. Peter Stevens of the University of Utah, rather than Blount staples. Correction of the valgus deformity of the femur permits internal lengthening of the femur with an intramedullary lengthening device (ISKD) after the knee deformity is corrected (Fig. 33).

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30 Congenital Leg Deformities: Tibial Hemimelia

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INTRODUCTION

The first correct description of the so-called tibial hemimelia was given by Billroth (1) according to Dankmeijer (2). The longitudinal tibial reduction defect is a very rare disease with an incidence of 1/1,000,000 per live birth (3,4).

Many different operative techniques have been tried in the treatment of this disease (5–11). Primary amputation (12–16) has been recommended, especially for the severe forms because the functional results of the reconstructions were often disappointing (17,18). In addition, a high rate of secondary amputations has been reported (19). The failure of primary treatments was for the following reasons:

1. Delayed reconstructive operations in the late childhood or even in the adolescence remove the potential for physiological and adaptive growth of a young organism.
2. The inability to identify anatomical structures (e.g., cartilage anlage) preoperatively because of insufficient diagnostic testing.
3. The strategy of functional remodeling potential of apparently useless anatomical structures, which are reconstructed into useful structures, was not used.

The following section will present how to use with the help of modern diagnostic tools approaches (custom made operations), physiologic adaptive growth potential. We have utilized and reconstructed existing anatomical structures for treatment of the tibial reduction deficiencies with new operative approaches.

CLINICAL EVALUATION

The clinical symptoms of the tibial reduction deficiencies depend mainly on the degree of the malformation, which can be quite variable.

Generally, the disease is characterized by the deformity at knee and upper ankle joint frequently combined with knee joint contractures. Frequently, active knee extension is not possible. This can be caused by a contracture as well as by the absence of activity of the anlage of the quadriceps muscle (9). Frequently, a knee joint with absent collateral and cruciate ligaments and, furthermore, with distinct, passive multidirectional instability is found. Then the complete lower limb is affected and displays a shortening and a bowing, which correlates specifically with an agenesia of the tibia (Fig. 1). The fibula can be fairly normally developed but appears frequently hypoplastic, dysplastic, or bowed (Figs. 2 and 3). The head of the fibula can be palpated under the skin at the dorso-lateral side of the femoral condyle. It displays all grades of displacement from subluxation to dislocation. The fibular head is always located proximal and dorsal but never ventral to the femoral condyles (20). In the ankle joint, an extreme varus deformity is present, sometimes in combination with foot equines or increased adduction and supination of the forefoot. The lateral malleolus is prominent and may protrude at the back of the foot. Associated foot malformations are frequently detected. These include clubfoot, equines foot, polydactyly, hypoplasia, aplasia, agenesia of digits and rays, as well as synchondrodesis and synostosis of two or more bones frequently between talus and calcaneus (20–22). The reduction defects of the rays start at the medial side of the foot. The femur may also be developmentally affected. Various morphological alterations include bifurcation, duplication of distal femur, distal hypoplasia, and dysplasia (23,24). The patella

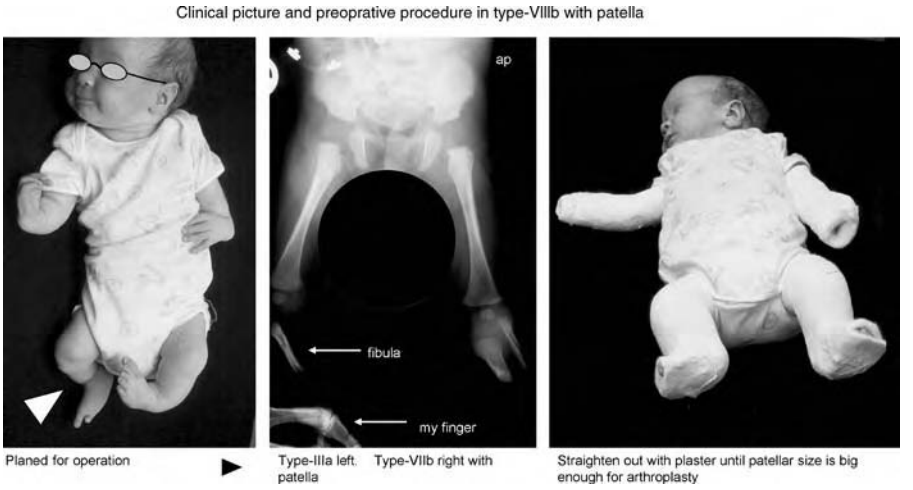

Figure 1 A 15-month-old boy with malformation of all four extremities with radial clubhands, left tibia type IIIa, and right tibia type VIIb without patella. The right leg is intended for surgery. The score is col/fel+/pal/tiVIIb(+)/fil/peIII (2/2+2/3/0+1/2/0 = 12 total). This is a type VIIb class 4 (Tables 1 and 2).

structure ranges from a largely normal formation to agenesis including intermediate forms of hypoplasia and aplasia (2,25–27).

Congenital dysplasia of the hip in this context has been described by various authors (20,21,28–30). The many other associated congenital malformations and the genetics are beyond the scope of this chapter (31,32). These affected children have normal or above average intelligence (15,33).

The neurological, psychomotor, and psychological development is described as normal (34–37).

It is important to recognize all malformations and abnormal function not only from the affected leg but also from the other extremities and the rest of the skeleton. For this purpose, all modern diagnostic tools should be used including sonography, magnetic resonance imaging (MRI), three-dimensional CAT scan, and orthography. With the recognition of all this data, including the intraoperative diagnosis of the local anatomy, comes the possibility of organizing an individual functionally-motivated treatment plan.

CLASSIFICATION

We have not found the previous classifications of tibial malformations to be useful in guiding modern treatment. Definitions are imprecise, diagnosis is made from only X-ray, complete types of malformations are ignored (30,38), and the descriptions are often too complex for

Figure 2 X-ray series of a six-year-old girl with diastasis of the left tibia and fibula distally with subluxated foot (type II). Left picture shows the situation before operation. Right pictures show the situation after reposition of the foot and plasty of the syndesmosis ventrally and dorsally by periostal flaps.

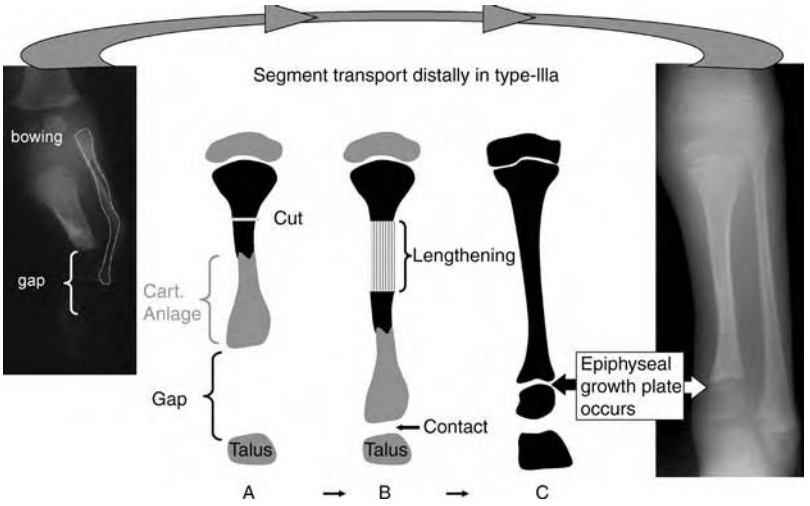

Figure 3 Schematic drawing of procedure for the treatment of tibial reduction defect type IIIa. The gap between the cartilage anlage of the distal tibia and the talus is closed by bone transport from proximal to distal. Left X-ray shows the situation before surgery in a three-year-old boy. Right X-ray shows a complete maturation of the cartilage anlage with epiphyseal growth plates and a stable ankle joint configuration four years after operation.

useful clinical application (41) (Fig. 4) (Table 1). A more precise preoperative diagnosis can be obtained with sonography and MRI. This combined with an experienced intraoperative analysis of anatomic structures will ensure a precise description of the pathological-anatomical situation, which can then be used to guide modern treatment. A new classification for tibial malformations, which is used to guide treatment, is presented reflecting the severity of pathological-anatomical conditions (Fig. 4). In this classification, features of entire lower extremity are included (coxa-femur-patella-tibia-fibula-pes) but special attention is directed

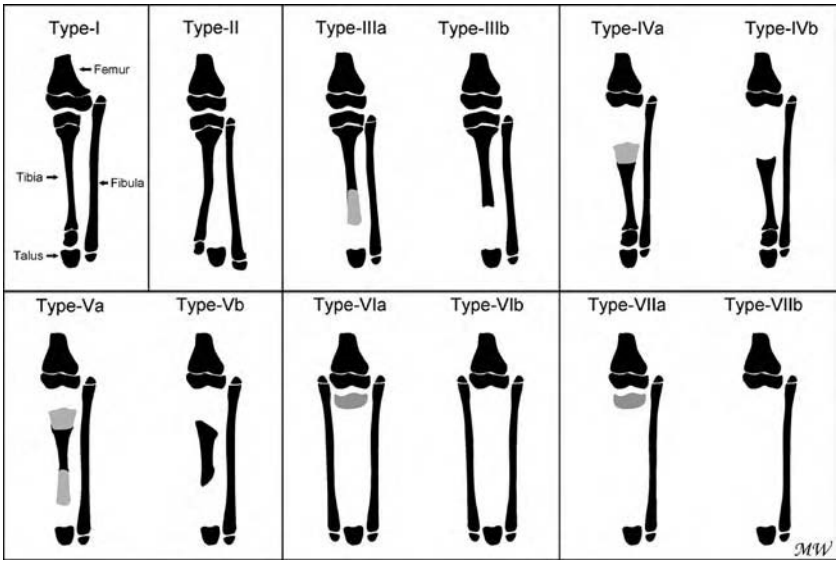

Figure 4 Classification of tibial reduction defects in seven types and five subgroups (a = with cartilage anlage and b = without cartilage anlage) according to the severity of the malformation. Black color = bone, gray color = cartilage. The pictures represent a higher maturation level of the lower leg instead of the situation immediately after birth because of the possibility for more detailed illustrations. Type 1: hypoplasia; type 2: diastasis; type 3: distal aplasia; type 4: proximal aplasia; type 5: biterminal aplasia; type 6: agenesis with double fibula; and type 7: agenesis with single fibula.

Table 1 Classification and Scoring System of the Tibial Reduction Defects of the Leg

Anatomic Region	Type	Skeletal Defect	Score	Type	Muscle Function	Score
COXA (co)(hip)						
Co	I	Normal	2	+	Existent	2
Co	II	Dysplasia	1	(+)	Partly absent	1
Co	III	Subluxation	0	–	Absent	0
FEMUR (fe)						
Fe	I	Normal	2	+	Existent	2
Fe	II	Distal hypoplasia	1	(+)	Partly absent	1
Fe	III	Distal dysplasia	0	–	Absent	0
PATELLA (pa)						
Pa	I	Normal	3			
Pa	II	Aplasia	1			
Pa	III	Agenesis	0			
TIBIA (ti)						
Ti	I	Hypoplasia	22	+	Existent	2
Ti	II	Diastasis	20	(+)	Partly absent	1
Ti	III	Distal aplasia		–	Absent	0
Ti	a	With cartilaginous anlage	18			
Ti	b	Without cartilaginous anlage	8			
Ti	IV	Proximal aplasia				
Ti	a	With cartilaginous anlage	16			
Ti	b	Without cartilaginous anlage	6			
Ti	V	Biterminal aplasia				
Ti	a	With cartilaginous anlage	14			
Ti	b	Without cartilaginous anlage	4			
Ti	VI	Agenesis with double fibula				
Ti	a	With cartilaginous anlage	12			
Ti	b	Without cartilaginous anlage	2			
Ti	VII	Agenesis with single fibula				
Ti	a	With cartilaginous anlage	10			
Ti	b	Without cartilaginous anlage	0			
FIBULA (fi)						
Fi	I	Normal	2			
Fi	II	Hypoplasia	1			
Fi	III	Dysplasia	0			
PES (pe)(foot)						
Pe	I	Normal	2			
Pe	II	Rays 3–4	1			
Pe	III	Rays 1–2	0			

to the tibia. The tibial malformations are divided into seven main groups and eight subgroups. The *cartilaginous anlage* is marked in the subgroups with "a" when it exists and with "b" when it is absent (Table 1). The pathological-anatomical terms are used in respect to their original definitions. Because these terms do not differentiate between cartilaginous and osseous anlages, a correlation to one of both had to be made. We chose for all definitions to be related to the *osseous anlage*. Thus, hypoplasia is defined as complete but underdeveloped osseous part of the skeleton. Aplasia is defined as partial loss of an osseous part of the skeleton—either proximal or distal. For example, aplasia exists proximally or distally with or without a cartilaginous anlage.

The presence of a cartilaginous anlage was assigned little importance in the previous orthopaedic literature, but we have found it to be critical for treatment. If the cartilaginous anlage is in contact with the corresponding joint, the anlage will mature completely to an osseous structure with growth plates in the context of weight bearing and functional loading. The terms partial or complete aplasia frequently found in the literature are confusing descriptions. The complete absence of a skeleton bone is defined as *agenesis*. The cartilaginous anlage of an osseous agenesis is normally proximally located.

Additional description with proximal, distal location or complete anlage seems to be sufficient in this context. The character of additional affected structures of the complete leg is of tremendous significance with respect to the treatment of the tibial reduction deficiency. Deformity of the hip, poor function of the quadriceps, or an absent patella are critical factors for planning reconstructive surgery. Thus, the system of the classification includes the complete extremity to define the extent of the malformation. This is reflected in a scoring system, which will allow comparison of different forms of treatment and a definition of basic principles of treatment in relation to the extent of the malformation (Tables 1 and 2). Both the classification and the scoring system are based on the main practical aspects of the pathoanatomy. Definitions are as follows:

There are three skeletal types for hip, femur, patella, fibula, and foot. There are three grades of muscle function for each of these regions (Table 1). The scoring system was developed with treatment and prognosis potential in mind. The higher number should represent better treatment and prognosis potential. The minimum is 0 and the maximum is 39. Because the patella is so important for reconstructive surgery, it is assigned a higher score of a maximum of three points. For the hip, femur, and fibula, a muscle function maximum score of two points is assigned. The importance of the cartilage anlage especially for reconstructive surgery is illustrated in a 10-point difference between a and b of the same type and an 8-point difference between different tibial types with cartilage anlage (e.g., type IVa = 16) versus without (e.g., type IIIb = 8). Abbreviations are used for the anatomic regions: coxa (co), femur (fe), patella (pa), tibia (ti), fibula (fi), and pes (pe). The three types of muscle function are described as +, (+), and − corresponding to existent, partially absent, and absent respectively. Quadriceps muscle function related to the femur. Muscles that move the hip joint are related to coxa, those that move the knee joint are related to the femur, and those that move the foot are related to the tibia.

Example 1: Distal diastasis of the tibia with four-ray foot and normal hip joint, normal femur, normal patella, hypoplasia of fibula and normal muscle function is abbreviated as coI+/feI+/paI/tiII+/fiII/peII with a score of 35 points (2+2/2+2/3/20+2/1/1).

Example 2: Tibial agenesis without cartilaginous anlage with singular hypoplastic fibula, dysplasia of the hip, missing function of the quadriceps muscle, femoral hypoplasia, agenesis of the patella and a two-ray foot is abbreviated as coII+/feIII (+)/paIII/tiVIIb

Table 2 Five Different Classes of Tibial Reduction Defects of the Leg Based on the Score

Class	Score
5	0–7
4	8–15
3	16–23
2	24–31
1	32–39

(+)/fiII/peIII with a score of 7 points (1+2/1+1/0/0+1/1/0). According to the scoring system, five classes of defects are defined (Table 2): Example 1 would be called tibia defect type II/class1 and example 2 would be called tibia defect type VIIB/class 5. In this way, the focus is on the tibial deformity but the entire lower extremity is also factored into the equation.

Seven main types of tibial defects have to be recognized and the additional defects of the whole leg can be summarized in the five-class system. With this new classification and scoring system the tibial defects are defined precisely, the pathological anatomical terms are correctly used, and the entire lower extremity and its function are included. In summary, this system allows a simple but comprehensive treatment-relevant classification to be implemented for outcome comparisons of specific tibial deficiency types among authors. For daily clinical use, the tibial classification alone is preferred.

TREATMENT OPTIONS

The type of operative treatment should be oriented on the requirement of the patients and/or their parents. It is the responsibility of the surgeon to explain all treatment options.

During these discussions, he should discuss the possibilities of conservative treatment by orthotic devices (external joint stabilization) and orthoprosthetic devices (external joint stabilization and artificial equalization of length defects). With respect to the operative methods, he should differentiate between amputation techniques, techniques that improve the ability to wear support devices and (re)constructive techniques.

The medical advice to the parents should start as early as possible to use the best of all options. The surgeon should point out that a lifelong orthotic or orthoprosthetic fitting of the patient could be recommended only when a good function of the joints exists (tibia type I–II).

It should be pointed out that slight operative treatments could be necessary to avoid abnormal growth of the extremity to maintain the ability to use prosthetic devices. In mild cases (tibia type III and IV) the growth potential of the affected leg can, specifically when a cartilaginous anlage is present, be stimulated in that way that only a few secondary lengthenings or corrective operations are required. In severe cases (tibia type VI–VII), the basic decision has to be made between an amputation under the femoral condyles with an above knee amputation prosthetic fitting or complex repetitive (re)constructive operations. The amputation in the early childhood has on the one hand the advantage of a definitive solution with one single operation and on the other hand the disadvantage of the loss of lower leg and foot.

The (re)constructive surgeries include several general approaches. These include joint forming techniques, muscle/tendon surgery to improve active joint movement, and lengthening for treatment of leg length discrepancies.

The aim of all operations is the improvement of the affected extremity. Rather than a reconstruction, this can be more accurate though as a new construction (de novo creation) using anatomic structures that were without previous function.

The advantage of the constructive operations is the preservation of the extremity. The disadvantage is the requirement of frequent surgery—mostly lengthening operations during childhood and one or more complex primary operations.

If parents insists on a definitive solution with a minimum of surgery and prefer a prosthetic fitting, an amputation in mild to moderate cases is reasonable. In cases of defects at the distal lower leg with an unstable foot, amputation techniques (14,16) to maintain maximum length have been described.

In bilateral cases, amputation techniques should be used with reservation because energy consumption with gait is higher compared with unilateral amputation. Constructive surgery is preferred in bilateral situations because the length discrepancy is much smaller, and fewer operations are typically necessary. In bilateral cases and concurrent malformations of the upper extremities, it has to be considered that the feet are required as substitution or complement for hands. The results of constructive surgery should be measured against those of amputation with prosthetic wear. If parents insist on the preservation of the affected limb of their child by constructive surgery, even if the expected functional result is expected to be inferior to amputation, their decision should be respected. If the function of their children's limb is more important for the parents and the expected functional result between amputation and constructive surgery is the same, an amputation should be recommended because the

amount of surgery needed is less. We have been confronted with parents who insist on amputation even though constructive surgery is expected to yield a better result because the effort and the frequency of surgery will be a great strain for the child and family. We have not observed greater long-term psychological disturbance in children who have undergone the reconstructive surgical treatment. The surgeon should give his recommendation in a neutral way and should respect the decisions of the parents.

SURGICAL TECHNIQUES

In this section, new constructive techniques are presented with respect to the different types of defects. The variety of soft-tissue techniques to improve muscle function is beyond the scope of this chapter.

The treatment is separated into preoperative, operative, and postoperative phases. The preoperative phase serves as preparation for the surgery. An example is diminishing joint contractures with casts and orthotic devices (Fig. 1).

The postoperative phase includes an acute and postacute times. The acute time is when the patient is with ring fixateurs to stabilize the constructed joints (Fig. 5). The postacute time is after removal of the fixators when the patient is braced to treatment to prevent recurrence of deformity or loss of correction (Figs. 6 and 7). Physical therapy is used during the various phases. The success of the treatment depends significantly on the intraoperative diagnosis of the pathological alterations by the surgeon. Variations of the pathologic-anatomy require modifications of specific therapy. The following are general guidelines:

Hypoplasia of tibia (type I) is characterized by the complete osseous anlage of the tibia with its adjacent joints however it is short and there is increased length of the fibula. The therapy is focused on lengthening of the tibia. The lengthening procedure can be carried out in two different ways:

1. Mounting of the ring fixator only at the tibia, not including the fibula. Following the tibial corticotomy, the lengthening of the tibia is performed gradually until the length matches the fibula. This technique results in a tremendous stress of the soft tissue on the ends of the fibula resulting in subluxation of joints, particularly the knee joint. Should this be noted, the distraction has to be stopped. The risk for subluxation can be reduced by performing soft tissue releases [e.g., Z-plasty of the lateral collateral ligament (LCL)]. If signs of a subluxation are recognized, the frame can extend over the joint level.
2. Mounting of the ring fixator at the tibia, not including the fibula and by extending the frame across the adjacent joints. The distraction is carried out between thigh and foot with a simultaneous corticotomy of the tibia. This protects the knee and ankle from excessive pressure and subluxation. This technique is appropriate specifically for significant differences in length between tibia and fibula. However, it has the disadvantage of a rigid immobilization of the knee and ankle joints. This problem can be lessened by using an iso-

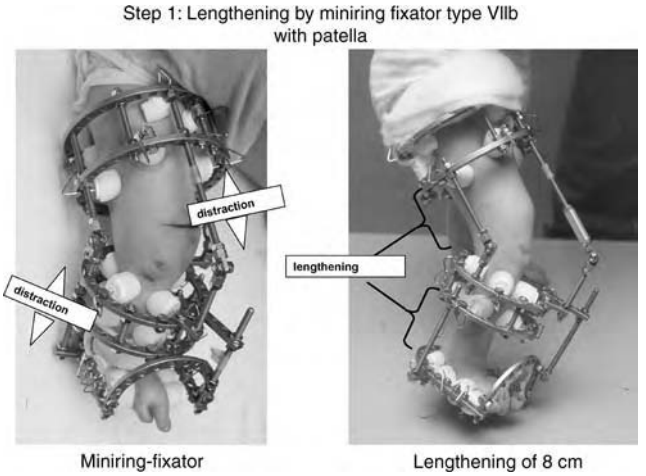

Figure 5 The first step of the surgical procedure is a soft tissue lengthening of the leg as a prerequisite for the transposition of the fibular head under the femoral condyles and the foot under the distal end of fibula.

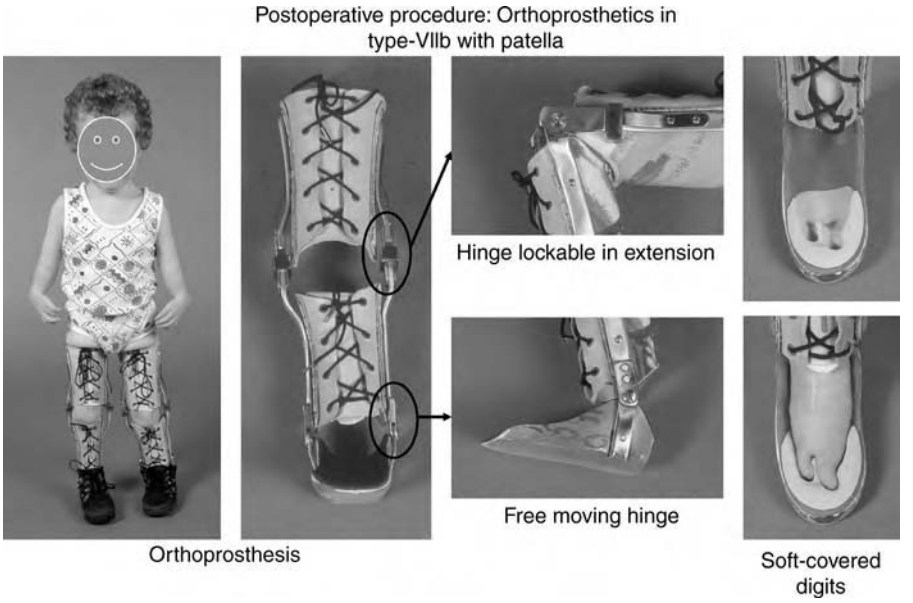

Figure 6 After removal of the frame, the leg(s) is (are) fitted in an orthothic device with lockable knee hinges in full extension for walking. After strengthening of the upper leg muscles, the hinges can be unlocked. The ankle joint can be fitted in the orthotic device with or without ankle hinges depending on the severity of the malformation. Also a two-ray foot can be sufficiently fitted. A night brace for preventing a growth disturbance is used.

metric mechanical hinge at the joints mounted either following the end of the distraction or at the beginning of distraction. The disadvantage of the latter variation is a higher risk of developing contractures during the lengthening, which must be avoided by intensive painful physiotherapy. Removal of the fixateur follows after the consolidation of the callus. If necessary, the lower leg is splinted by brace during final callus maturation.

Figure 7 X-ray series shows the hypertrophy of the fibular head stabilized after an anterior cruciate ligament plasty. The six-year-old girl is able to walk and run with the use of orthotic devices on both similarly affected legs three years after surgery. The function of the absent quadriceps muscle is compensated by rubber bands that allows active flexion and gives passive extension. The score of the legs are characterized each by coII/feIII (+)/paIII/tiVIIb (+)/fiI/peII, (=7 points). This is type VIIb class 5 (Tables 1 and 2).

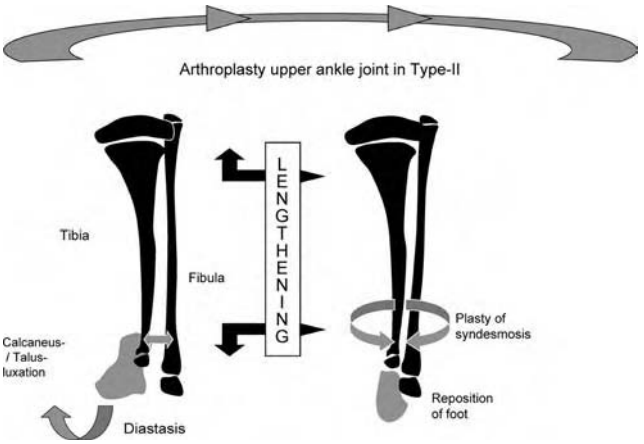

Figure 8 Schematic of surgical procedure for treatment of tibial reduction defect type II. Left picture shows the situation before operation with diastasis of tibia and fibula distally with a subluxated foot. Right picture shows the situation after surgery with soft tissue lengthening between lower leg and foot, reposition of the foot and plasty of the syndesmosis ventrally and dorsally.

Distal diastasis of the tibia (type II) is characterized by an insufficiency of the syndesmosis and a separation of tibia and fibula. There is additionally a proximal subluxation/migration of the foot between both the bones. The degree of this migration can vary leading to different clinical situations. A patient may walk on the hyperplastic distal fibula or weight bear on both bones (Figs. 2, 8, and 9). The principle of the technique is the elimination of the subluxation of the foot by distraction, the acute correction of the diastasis as well as the repositioning of the foot with coaptation of the tibial joint surface to the talus at the ankle. The necessary distraction can be done by a ring fixator at the lower limb with integration of the foot. Then, the diastasis can be corrected in a single-step operation using a bone forceps followed by a reconstruction of the syndesmosis using a ventral and dorsal periosteal flap (or peroneal tendon plasty, or artificial tendons). An olive wire secures the syndesmosis during the healing process. The reposition of the ankle joint is secured with an axially placed Ilizarov wire that can be removed after six weeks. After the removal of the wire, the ankle joint can be mobilized with the protection of isometric ankle hinges for another six weeks. The lower limb will be fitted in an orthosis with a movable foot as long as there is sufficient long-lasting stability of the ankle joint.

Distal aplasia of the tibia with cartilaginous anlage (type IIIa) is characterized by a proximally ossified tibia and a distally attached cartilaginous anlage. Although there is no contact with the talus, there is a preformed joint part that matches the talus (Figs. 3 and 10).

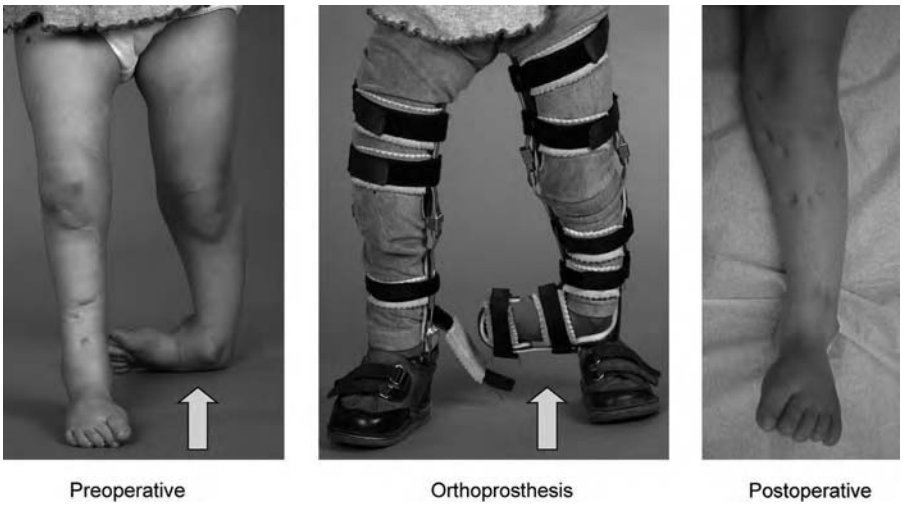

Figure 9 Left two pictures show the left leg before surgery including orthoprosthetic care. Right picture shows postoperative result. The girl has also gone through a surgical procedure of the right leg affected with a tibia type IIIb. According to the score the left leg is characterized by col/fel+/pal/tiII (+)/fil/pel (2/2+2/3/20+1/2/2 = 34 total). This is a type II class 1 (Tables 1 and 2).

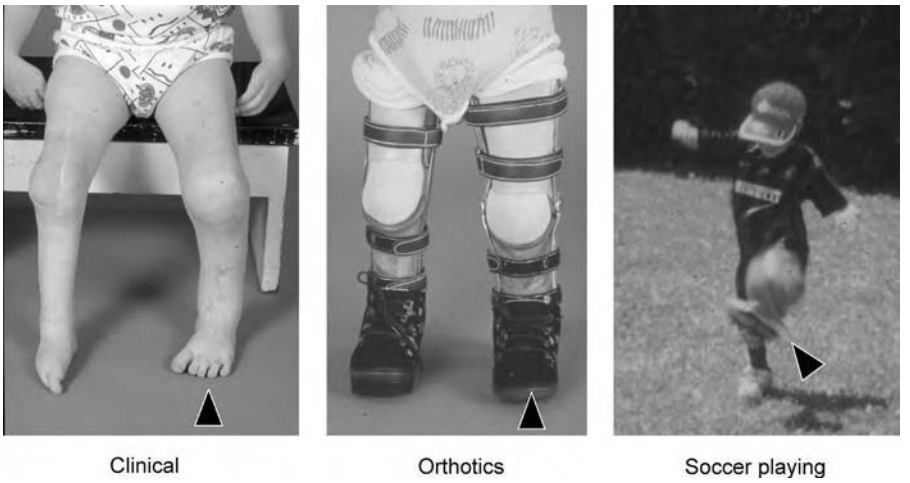

Figure 10 Postoperative clinical results including orthoprosthetic care. All four extremities are malformed. The boy is able to play soccer, ski, and skateboard.

The principle of therapy is to put the tibia and talus in contact. With weight bearing, the cartilaginous anlage and the talus can be expected to grow and mature together. For this purpose, a ring fixator is mounted at the lower limb integrating the foot and an osteotomy of the tibia is performed at its osseous part. The distal osseous part of tibia including the attached cartilaginous anlage will be transported distally until there is contact with the talus. Preoperatively, the defect distance can be measured with MRI. The correct docking of the joint components can be demonstrated with arthrography. The "new" ankle joint will be moved after placement of isometric mechanical hinges on the frame six weeks before removal. The lower limb is then fitted in a lower leg orthosis with a movable foot during childhood.

Distal aplasia of the tibia without cartilaginous anlage (type IIIb) is characterized by a proximally ossified tibia without a distally connected attached cartilaginous anlage (Figs. 11–13). The principle of treatment is insertion of the fibular head into the proximal part of the tibia and placement of the foot underneath the distal end of the fibula.

To stabilize the centralization of the foot, a malleolus plasty can be performed (39). The treatment starts with the mounting of a fixator from the thigh to the foot for soft tissue distraction. After this, a transposition of the fibular head underneath the proximal tibia and the foot underneath the distal fibula end is possible. The origin of the LCL has to be separated proximally before the lengthening to avoid an epiphyseolysis of the proximal fibula. The

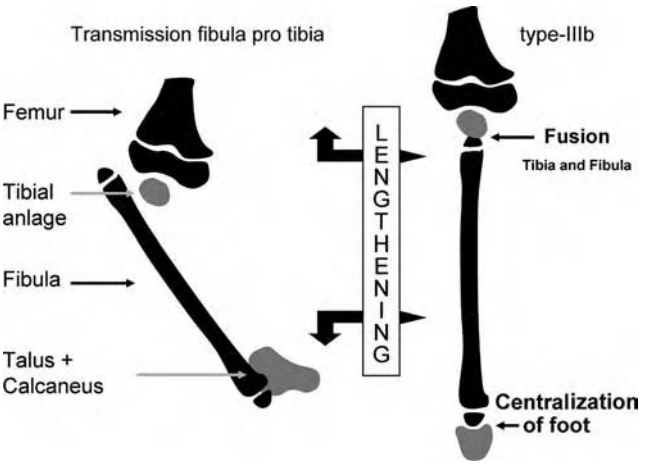

Figure 11 Schematic drawing of operational procedure for treatment of tibial reduction defect type IIIb. Left picture shows the situation before surgery with subluxation of fibula and foot. Right figure shows the situation after surgery with soft tissue lengthening across the knee and ankle, reposition and centralization of the foot as well as chondrodesis between fibular head and tibial anlage.

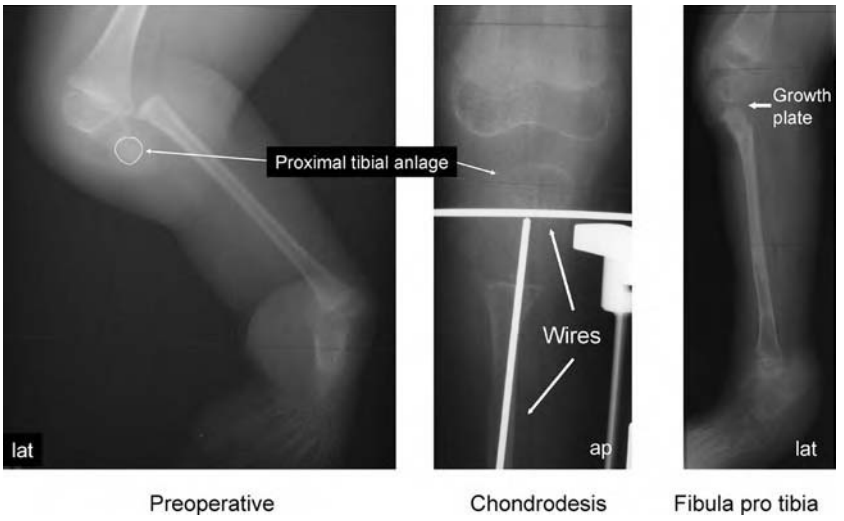

Figure 12 X-ray series of a six-year-old girl with tibia type IIIb. Left picture shows the situation before surgery. Middle picture shows the fixation of the tibial anlage and fibular head by an axial fibular wire and a transverse wire through the tibial anlage. Right picture shows the situation after surgery and removal of the ring fixator. Note the open growth plate of the former fibula head after the osteo-chondro-synthesis between fibular head and tibial anlage.

Achilles tendon must also be lengthened with a Z-plasty. When the corresponding length of distraction is achieved, the fibula head is moved under the proximal tibia. During this procedure, LCL is divided and then inserted medially and laterally on to the tibia. The periosteum of the tibial end is opened like a door and the cartilaginous top of the fibula is fixed to the tibia to accomplish an osteo-chondro-synthesis. The periosteal and perichondral flaps are sutured ventrally and dorsally at the fibular head. An axial intramedullary Ilizarov wire from the fibula stabilizes the proximal epiphysis (Fig. 12). A transverse Ilizarov wire through the proximal tibia secures the osteo-chondro-synthesis. This knee arthroplasty, secured by isometric mechanical hinges, can be moved while a stable fixation of the osteo-chondro-synthesis is present. In the case of a bilateral malleolus plasty, the ankle joint is immobilized for six weeks during healing. Then ankle joint is mobilized with the stability of isometric mechanical hinges mounted on the frame. The fixator is retained until the osteo-chondro-synthesis unites—usually three months. After removal of the fixator, the leg is fitted in a knee ankle foot orthosis (KAFO) with movable knee and ankle joints.

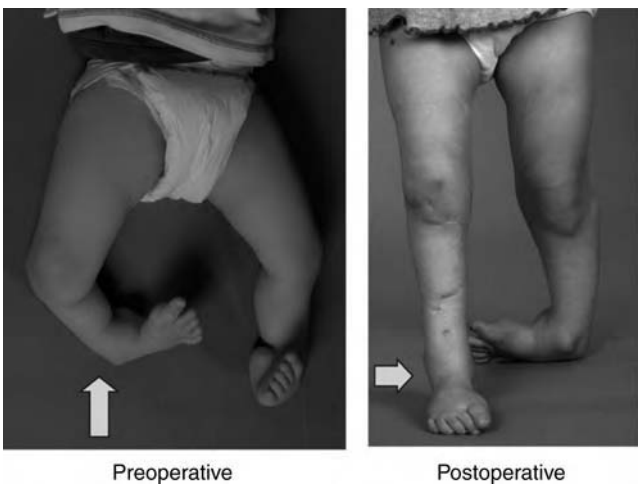

Figure 13 Left figure shows the right leg before surgery showing the subluxation of the fibula proximally and the foot distally. Right picture shows postoperative result. The six-year-old girl has also gone through an operational procedure of the left leg affected with a tibia type II. The right leg is characterized by col/fel+/pal/tiIIIb(+)/fil/pel (2/2+2/3/8+1/2/2 = 22 total). This is a type IIIb class 3 (Tables 1 and 2).

Proximal aplasia of the tibia with a cartilaginous anlage (type IVa) is characterized by a distally ossified tibia and a proximal cartilaginous anlage without contact to the femoral condyles. The proximal end of the cartilaginous anlage has the potential to fit to the distal femur.

The principle of this treatment is to bring the proximal cartilaginous anlage of the tibia in contact with the femoral condyles. With weight bearing, the cartilaginous anlage and the distal femur articular surface can be expected to grow and mature together. For this purpose, a ring fixator is mounted across the knee and tibia. Osteotomy of the bony part of the tibia is performed. The proximal osseous part of the tibia with its cartilaginous anlage is transported proximally until contact is made with the femoral condyles. In the case of a short distal tibial segment, frame extension to the foot is recommended. Preoperatively, the defect distance can be measured with MRI. The correct docking of the joint components can be demonstrated with arthrography. The ''new'' knee joint will be moved after placement of isometric mechanical hinges on the frame six weeks before removal. The lower limb is then fitted in a KAFO with a movable knee and ankle joints during childhood.

Proximal aplasia of the tibia without cartilaginous anlage (type IVb) is characterized by a distally ossified tibia without a connecting cartilaginous anlage. This may occur with or without a patella.

If a sufficiently developed patella exists, it will be used as a substitution for the tibial plateau (compare to type VIIb, Figs. 1,5,6 and 14–17). If no sufficient patella exists, then the fibular head will be transposed under the femoral condyles to induce a transformation of the fibular head into a ''functional tibial plateau'' (compare type VIIb without patella, Figs. 7,18 and 19). An anterior cruciate ligament (ACL) plasty by use of the LCL or other soft tissue structures attached to the fibular head helps stabilize this ''new knee joint.'' A synostosis of the fibula to the tibia is performed. The weight bearing on the fibula results in a hypertrophy of the fibula with formation of a ''tibial plateau'' (form follows function). The proximal fibular epiphysis substitutes hereafter the proximal tibial epiphysis. The distal tibia completes its maturation due to normalization of weight bearing. The treatment starts with the application of a ring fixator from the thigh to the foot across the knee and ankle. Soft tissue distraction of the proximal fibula beyond the proximal tibia is performed. In a simultaneous step, the LCL is released during this operation to avoid excessive tension and an epiphyseolysis of the fibular head and to utilize the LCL as a substitute for the ACL. Once enough distraction is achieved, a surgery to place the fibula head centrally between the femoral condyles is performed. The approach to the femoral condyles is performed with an S-like incision over the knee region. Attention should be directed to preserve appropriate parts of the fascia or capsule for use

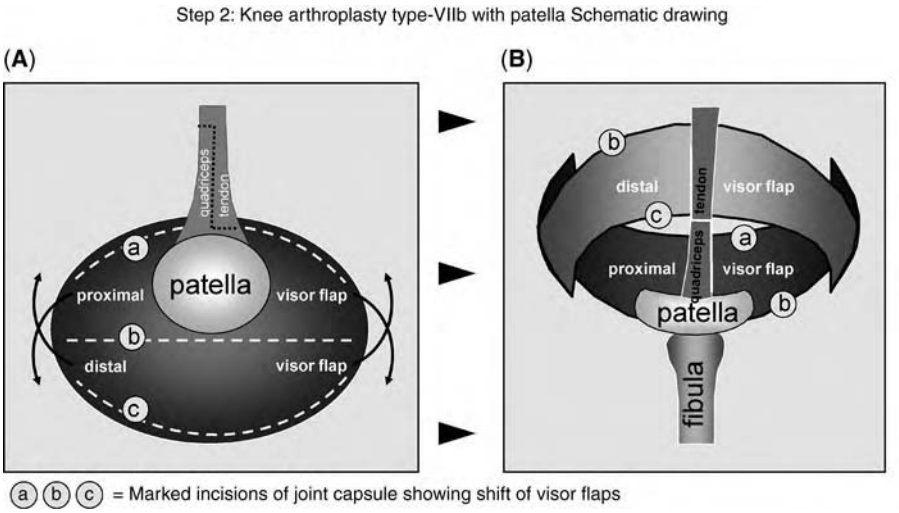

Figure 14 Left figure: schematic figure of the incisions (*dotted lines* a, b, and c) required for creation of two visor flaps based medially and laterally. Right figure: crossed flaps for repositioning of the patella forming a patello-fibular chondrodesis. The fibula functions as the tibia and the patella functions as a ''tibial plateau.'' Note the Z-plasty of the quadriceps tendon.

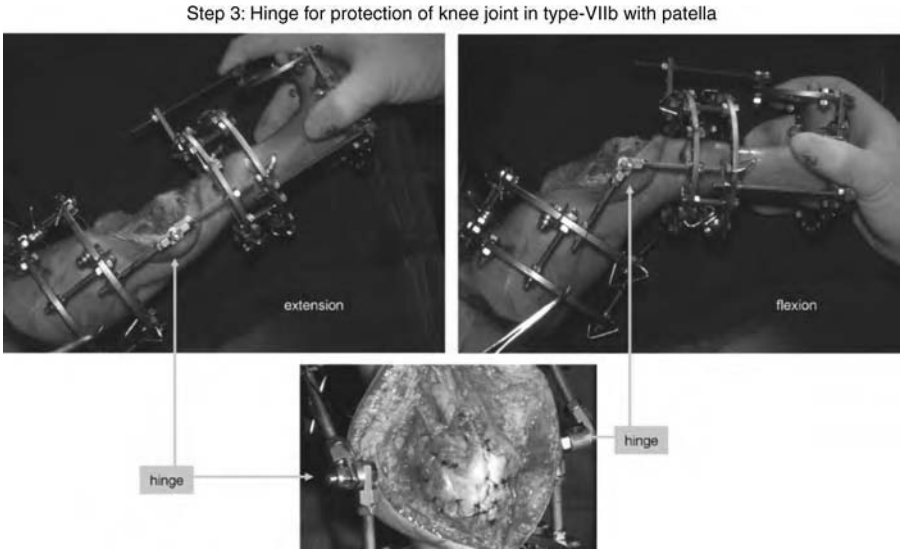

Figure 15 Mechanical hinges at the isometric point (*arrows*) permit early mobilization while providing stability and protection to the knee arthroplasty.

to construct a lateral knee ligament. When there is no patella the opening of the knee capsule has to be performed for adjustment of the fibular head. If a sufficient patella is present, it is utilized as ''tibial plateau substitute'' (compare the technique described for type VII). The LCL (or capsular structures, fascia lata, biceps tendon) is passed through a tunnel in the lateral femoral condyle as central replacement of the new ACL. Growth disturbances of the distal femur have not been a problem. The fibula will be osteotomized at the level of the proximal end of the tibia and a synostosis with the tibia is performed.

An axial intramedullary Ilizarov wire through the fibula secures the proximal epiphysis of the fibula and, additionally, the synostosis with the tibia. Hinges at the knee allow early-protected knee motion. At the beginning, the range of motion is limited (e.g., ex/flex: 0°/45°) and can be improved after the ACL reconstruction has healed. The fixator remains in place for about three months. After removal of the fixator, a KAFO with movable knee and ankle joints is used.

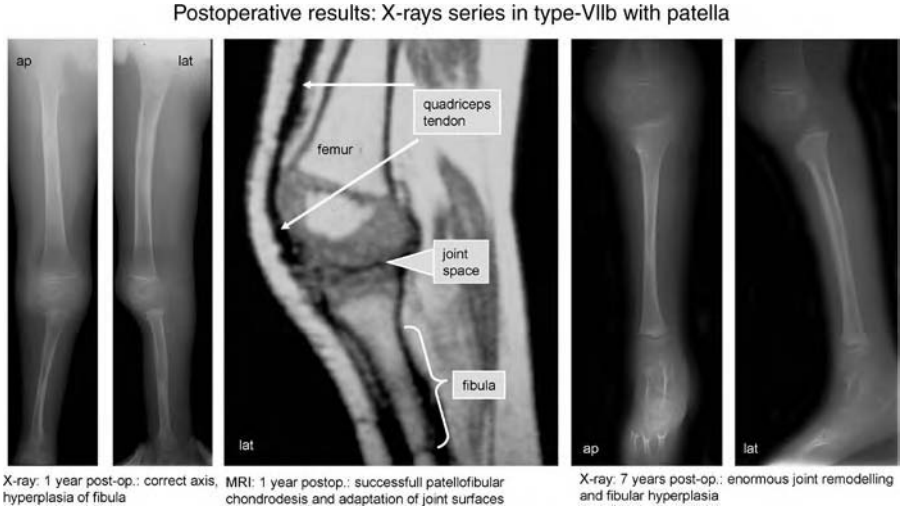

Figure 16 The radiographic pictures show the tremendous hypertrophy of fibula and patella (form follows function). In the magnetic resonance imaging (MRI), the articulating joint partners are visible and congruent.

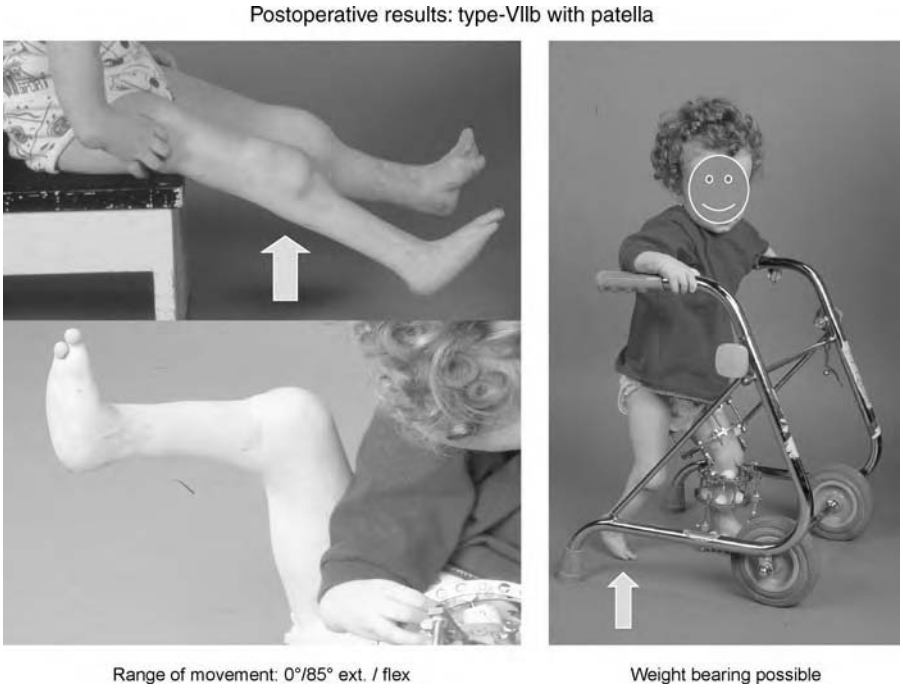

Figure 17 One year postoperative, the results show a good range of motion with possible weight bearing of the right leg without orthotic device. The left leg is still under treatment.

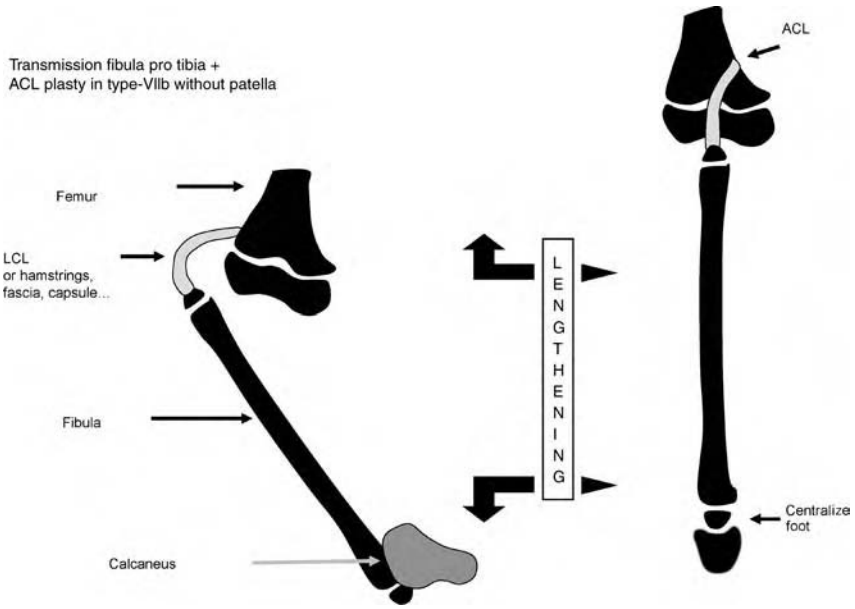

Figure 18 Schematic drawing of the surgical procedure for the treatment of tibial reduction defect type VIIb without patella. Left picture shows the situation before surgery with subluxation of the fibula and foot and preparation of the lateral collateral ligament (LCL) for use as the anterior cruciate ligament (ACL). Right picture shows the situation after surgery with soft tissue lengthening between upper leg and foot, ACL plasty between the fibular head and femur as well as centralization of the foot.

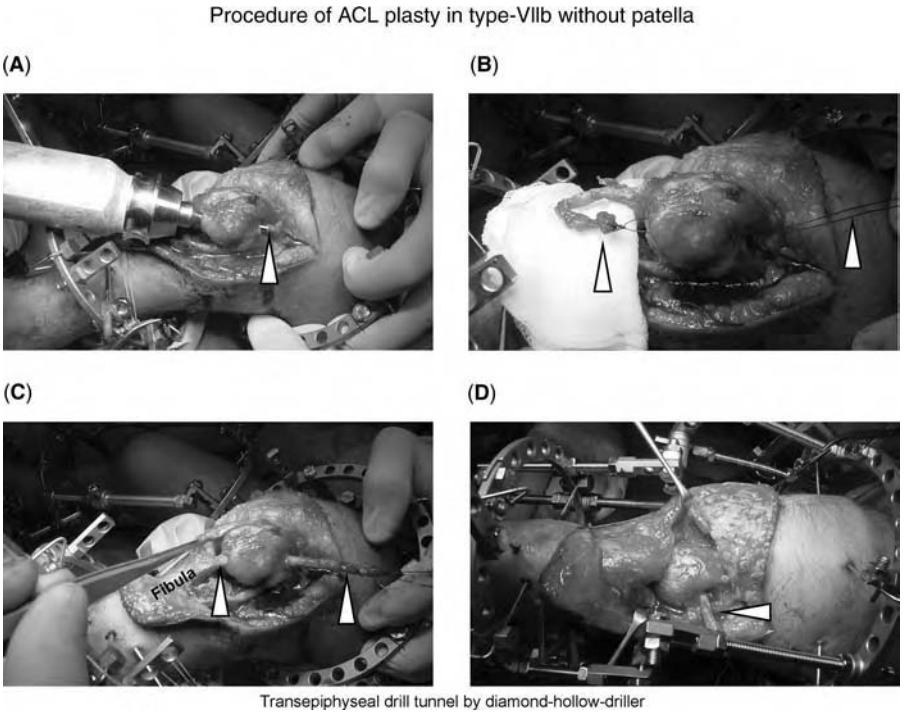

Figure 19 Technique of anterior cruciate ligament plasty in four steps (**A**–**D**). (**A**) A transepiphyseal drill tunnel from the apex of the dysplastic distal femoral epiphysis to the lateral aspect of the femur is made. (**B**) The lateral collateral ligament (LCL) is pulled through the tunnel. (**C**) The fibular head is opposed to the femoral epiphysis and a isometric mechanical hinge on the frame stabilizes the position of the fibular head in place. (**D**) With adequate tension the LCL is sutured to fascia lata or to itself by way of an additional drill hole in the lateral femur.

Biterminal aplasia of the tibia with cartilaginous anlage (type Va) is characterized by a centrally ossified tibia and a proximally and distally attached cartilaginous anlage without contact to femoral condyles and talus, respectively, but each with a preformed joint part (Figs. 20 and 21).

The principle of this technique is to bring the cartilaginous anlage of the tibia in contact proximally with the femoral condyles and distally with the talus. Weight bearing of the tibia then leads to a maturation of the cartilaginous anlage including the proximal and distal growth plates that will lead to longitudinal growth.

For this purpose, a ring fixator is mounted at the lower limb with extension to the foot. After the Z-plasty lengthening of LCL, soft tissue distraction to pull the fibula distally is the first step. Then, the tibia is cut in its osseous part. The proximal part of the tibia with its attached cartilaginous anlage is transported proximally until contact with the femoral condyles is achieved. The distal osseous part of the tibia with its attached cartilaginous anlage is transported distally until contact with the talus is achieved. Preoperatively, the defect distance can be measured by MRI. Arthrogram can be used to see the docking sites. Hinges at the knee and ankle protect these joint and allow motion. The leg is fitted in a KAFO after frame removal.

Biterminal aplasia of the tibia without cartilaginous anlage (type Vb) is characterized by a centrally ossified tibia without a proximally or distally attached cartilaginous anlage. This aplasia appears with or without a patella.

There are two options for the treatment: (i) the remaining tibia is resected and the technique used for tibial agenesis is used (see type VIIb) and (ii) the remaining tibia is approached with the same treatment used for type Va. However, it should be noted that in this situation, no growth zones and no joint partners might be available. This would necessitate multiple lengthening procedures.

Agenesis of the tibia with cartilaginous anlage and a double fibula (type VIa) is characterized by a completely absent tibia except for a proximally located cartilaginous anlage.

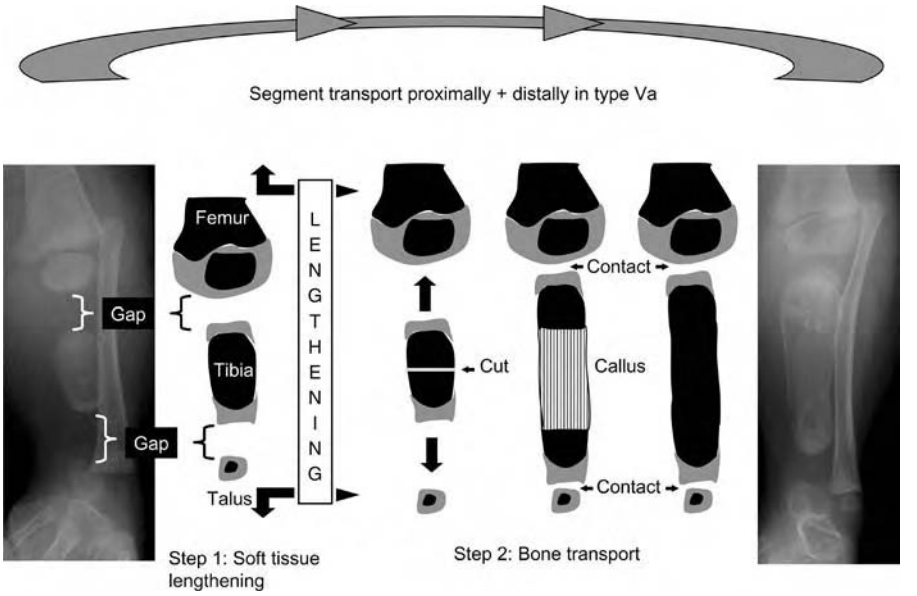

Figure 20 Schematic drawing of surgery for treatment of tibial reduction defect type Va. The gap between the cartilage anlage of the proximal tibia and the femoral condyles as well as distal tibia and the talus is closed by bidirectional bone transport. Left X-ray shows the left lower leg before surgery in a 10-month-old girl. Right X-ray shows the lower leg one year after removal of the frame with good further maturation.

Treatment selections should be based on the growth potential of this cartilaginous anlage. If this anlage is considered rudimentary without substantial function or potential as a joint, then it should be resected and the technique described for type VIb should be used. If however the cartilaginous anlage is considered substantial enough to be a joint partner with lateral stability, the reconstruction can be used. After maturation of the cartilaginous anlage once the medial fibula is transposed under it with a chondrodesis, it can become a functional

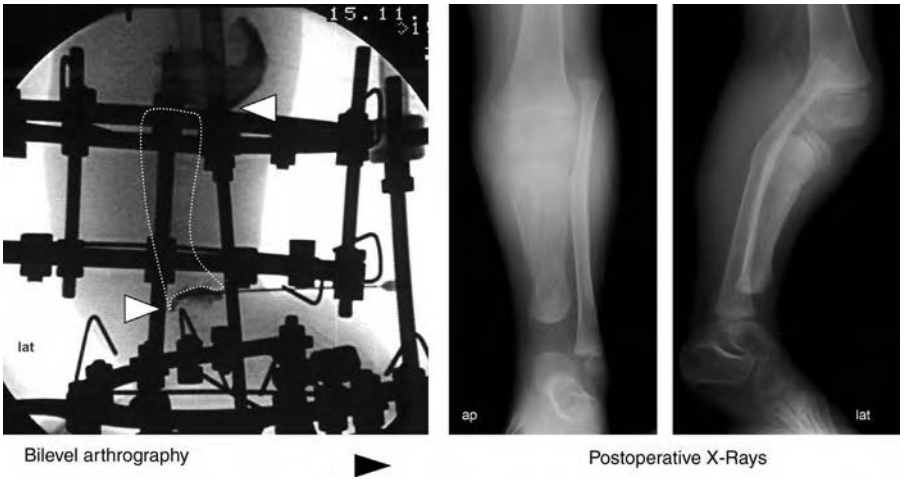

Figure 21 Left X-ray shows the two level arthrography (*arrows*, *dotted line* = circumference of tibia) to detect the docking of the transported bone segments. Right X-ray shows a further maturation of the cartilage anlage with a visible epiphyseal growth plate three years after surgery. Additionally, an over growth of the fibula occurred and the same procedure had to be repeated. This leg is characterized by col/fel+/pal/tiVa+/fil/pel (2/2+2/3/14+2/2/2 = 29 total). This is type Va class 2 (Tables 1 and 2).

joint. The medial fibula should articulate distally with the talus. If not, a technique appropriate for a diastasis should be chosen (see type II). If the level of the lateral fibula is correct and the lower limb not shortened, then transposition of the medial fibula underneath the cartilaginous anlage can be performed by a shortening osteotomy. In the case of shortening of the lower leg with proximal overgrowth of the fibula, an equalization by a soft tissue distraction using a ring fixator should be performed first. For this, one should use the same technique that is applied for type VIIb.

Agenesis of the tibia without a cartilaginous anlage and a double fibula (type VIb) is characterized by a totally absent tibia. This type of agenesis occurs with or without a patella.

If no sufficient patella exists, then the heads of both fibulae are utilized as joint partners to the femoral condyles. The corresponding LCL's are used as an ACL and a posterior cruciate ligament (PCL). The talus is transposed under the medial distal fibula. The lateral fibula distally functions as a lateral malleolus.

The treatment is initiated with the mounting of a ring fixator at the thigh, the lower limb, and the foot spanning the knee and ankle to enable a soft tissue distraction. The medial fibula is gradually pulled under the medial femoral and the lateral fibula under the lateral femoral condyle. Simultaneously, the LCLs should be released both medially and laterally to avoid an epiphyseolysis of the proximal fibular epiphysis and to utilize the ligaments as substitution for the cruciate ligaments. Once the corresponding distance of distraction is obtained, the surgical transposition of the two fibular heads under both femoral condyles follows. The approach is performed with a S-shaped incision. Remnants of fascia and capsule should be used to construct LCLs. If no sufficient patella is present, then the incision of the knee capsule for transposition of the fibular heads is carried out ventrally. The LCL (or capsular structures, fascia lata, biceps tendon) of the lateral fibula is utilized as a substitution of the PCL by passing it through a drill hole in the medial femoral condyle. If a sufficient patella is present, it will be used as a ''tibial plateau substitution.''

The medial fibula should articulate distally with the talus. If not, it should be approached as described for a diastasis (see type II). If the level of the lateral fibula is accurate and the lower limb not shortened, then the transposition of the medial fibula underneath the cartilaginous anlage can be performed with a shortening osteotomy.

Agenesis of the tibia with a cartilaginous anlage and single fibula (type VIIa) is characterized by a totally absent tibia except a mostly proximal cartilaginous anlage.

The choice of treatment is based on the quality of the cartilaginous anlage. If this anlage is considered rudimentary without substantial function or potential as a joint, then it should be resected and the technique described for type VIIb should be used. If, however, the cartilaginous anlage is considered substantial enough to be a joint partner with lateral stability, the reconstruction can be used. After maturation of the cartilaginous anlage once the medial fibula is transposed under it with a chondrodesis, it can become a functional joint. The fibula should articulate distally with the talus. If this is not the case, the talus should be transposed under the distal end of the fibula.

Agenesis of the tibia without a cartilaginous anlage and single fibula (type VIIb) is characterized by a completely absent tibia with (Figs. 1, 5, 6, and 14–17) or without a patella (Figs. 7, 18, and 19). If a sufficient patella is present, the principle of the treatment is a transposition of the fibular head into the caudally shifted patella. The caudal transposition of the patella is performed using transverse double visor flaps of the knee capsule shifted toward each other. The patella is converted into a ''tibial plateau'' and the fibula into a ''tibia'' (form follows function).

The LCL can be used as additional anchor for the chondrodesis between the fibula head and patella or as substitution of the ACL by passing it through a drill hole in the patella and through a drill hole in the epiphysis in the medial femoral condyle. The foot is transposed under the distal fibula end. This technique is characterized by four steps:

1. Application of a mini-ring fixator on the front thigh to foot spanning the knee and ankle to distract the soft tissue envelope (about 80 mm). This allows enough length for eventual transposition of the fibula and foot.
2. Operative knee arthroplasty (40).
3. Mounting of an isometric hinge on the fixator to protect the knee arthroplasty and to allow immediate postoperative physiotherapy.
4. Transposition of the foot with or without a malleolus plasty.

On the thigh, a proximal medial open half ring is connected with a distal full ring and attached to the femur with four olive wires. Two full rings are applied to the lower leg and fixed at the bones with four olive wires. The knee is spanned with hinges. The pre-existing flexion contracture at the knee level is integrated in the mounting with locked hinges. The horseshoe-like mounting of the foot is fixed with two-crossed calcaneal and two opposed metatarsal olive wires and, then, connected with rods across the ankle. Using distraction rods, the soft tissue is distracted to reduce the subluxation of the fibula and foot.

The open approach is performed by using an S-like skin section beginning proximal-lateral (using the scar of the former incision that was used to release the LCL), crossing the region knee horizontally to the medial side and then proceeding in a distal-medial direction. Following this step, the Latex-covered LCL is prepared and the Latex glove is removed. Then, a Z-plasty lengthening of the quadriceps tendon is done. Following this step, the knee joint capsule is cut three times in a horizontal position. The first incision is carried out at the proximal fold of the capsule, the second one is made directly at the distal patella pole, and the third incision follows the distal fold of the capsule. In this way, two visor flaps are created that are attached laterally and medially. The proximal visor flap, which includes the patella, is shifted distally and the distal visor flap is transposed proximally. The capsule flaps are sutured with one another at the contact points (Fig. 14).

This allows the patella to be positioned under the distal femur articular surface. The head of the fibula is prepared and opposed to the ventral surface of the patella. During this process, the perichondrium of the head of the fibula is incised in an H-like manner resulting in two door-like perichondral flaps. In the same manner but rotated 90°, the perichondrium of the patella is centrally incised. The perichondrium and the capsule tissue of fibula and patella, respectively, are sutured together after the cartilage surface of patella and fibula are placed in contact with each other. The patella is converted into a ''tibial plateau'' and the fibula into a ''tibia'' (form follows function).

This is stabilized with an axial intramedullary Ilizarov wire inserted retrograde from the fibula up to the patella. The patella is stabilized with a horizontally oriented Ilizarov wire and this is attached to the ring. The chondrodesis is secured in this manner.

If a simultaneous ACL plasty is intended using the LCL (or soft tissue structures attached to the head of the fibula), then a tunnel is drilled through the center of the patella and the ligament is passed through the tunnel. Another tunnel is drilled in the femur and the new ACL is secured under tension. The Z-plasty of the quadriceps is sutured in the lengthened position (Fig. 14). Two hinges are positioned on frame at the isometric axis of the constructed knee joint to protect and stabilize the chondrodesis between fibula and patella and the knee arthroplasty. Intraoperatively, the optimal range of knee motion is determined and set up with a mechanical locking device on the frame. Full weight bearing in the fixator with a locked knee joint is possible. The range of motion is gradually increased; however, it is most important to maintain full extension.

In a third operation, the foot is transposed. During this procedure, the distal end of the fibula is opposed to the talus or even to a different tarsal bone (it depends on the available anatomical structures) and is stabilized with an axial wire placed retrograde from the foot into the fibula. The ankle is also stabilized with the Ilizarov frame across the ankle. The aim of the transposition is a permanent stability of the foot. If the foot is flexible, then there is greater risk for a future subluxation. Motion of the ankle is best assured by centering the tip of the fibula on the neck of the talus and by stabilizing the new ankle with a bilateral malleolus plasty (39).

After removal of the fixator, a KAFO with hinges at the knee and ankle is fitted. The knee hinge is locked in extension for ambulation (Figs. 6 and 7).

If the patella is absent, the principle treatment is a transposition of the fibular head to the femoral condyle to induce a transformation and hypertrophy of the head of the fibula into a tibial plateau. A cruciate plasty is done to maintain stability. Transposition of the foot is also performed.

COMPLICATIONS

My experience of conservative and operative treatments is based on 67 patients and 102 extremities with tibial reduction deficiencies. All new surgical techniques in this chapter are based only on treatment of my own patients. There are many complications from conservative treatment (orthoprosthetic care) including poor fitting, poor patient selection, and loss of

precious time for constructive operative techniques. There are also complications that arise from amputation treatment.

The focus is on complications of our new surgical approaches and their treatment. We differentiate between complications from the surgery and those that arise from anatomic peculiarities that complicate the operation. The following are examples of such anatomic peculiarities that make surgery more challenging:

1. Atypical alignment of tendons, which have to be oriented in a new manner (e.g., diastasis of the tibia with intraarticular position of the extensor tendons).
2. Fusion of the knee capsule with the cartilaginous tissue of the femoral condyles. This requires a careful dissection of the layers.

To avoid a new fusion, a glove (Latex free) can be put temporarily between the layers of the affected tissues for its later removal (at a second stage surgery).

Aside from the typical complications associated with the use of external fixation, specific postoperative complications are as follows:

1. The soft tissue distraction of the fibula leads to great tension mediated by the LCL and the epiphyseal growth plate of the proximal fibula could become avulsed. The release of the fibula head from its capsule and the LCL helps avoid an epiphyseolysis.
2. A fusion of the prepared LCL to the surrounding soft tissues may be carried out by covering it with a Latex-free glove finger.
3. Deformity may occur. Only after the maturation of the epiphyseal growth plates, with time and weight bearing, can the orientation of the growth plates be recognized. This may require an osteotomy that can be used to correct deformity and achieve additional lengthening.
4. If the centralization of the foot does not achieve sufficient stability or if the orthotic devices are not worn regularly, a subluxation of the foot may arise. This would require a repeat foot transposition with the possible necessity of a bilateral malleolus plasty (39).
5. Daily intensive physical therapy is of tremendous importance for a child with a joint arthroplasty. When this intensive training is ignored, new contractures may arise. These may require reapplication of a frame.
6. Spanning frames can prevent growth at growth plates. This can lead to growth arrest. To avoid this problem, the frame should be loosened every six weeks to relieve the resulting growth tension.

FUTURE DIRECTIONS

Effort should be focused on the education of colleagues who are interested in these problems. The goal is to initiate therapy early and to avoid unnecessary amputation.

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31 Fibular Hemimelia

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INTRODUCTION

Fibular Hemimelia is the most common long bone congenital absence or hypoplasia. It was first described by Gollier in 1698 (1–6). Fibular hemimelia constitutes a spectrum of abnormalities associated with this deficiency including rotational abnormalities (7), angular abnormalities of both the femur and the tibia, shortening of the limb, and abnormalities of the ankle, hindfoot, and the forefoot (1,2,8–12). The abnormalities of the femur include variable degrees of shortening, retroversion, and distal femoral valgus. The shortening is usually at least 5%, but can be greater. The abnormalities of distal femoral valgus and retroversion have been previously described.

There are associated abnormalities of the knee, which include anteroposterior (AP) instability and abnormalities of the patella (13–16,16a). Abnormalities of the patella include instability of the patella, which may be due to abnormal formation of the patellofemoral joint, lateral instability, and lateral distal femoral hypoplasia.

The AP instability is due to either absence or abnormality in the cruciate ligament (14). Most commonly, instability of the knee presents itself as posterior subluxation of the tibia on the femur during limb lengthening. Less commonly, anterior subluxation can also be seen. Instability is not usually apparent clinically until such time limb lengthening is attempted. Abnormalities of the leg include variable degrees of fibular hypoplasia, shortening of the leg, and apex anteromedial angular deformity of the tibia (procurvatum and valgus).

CLINICAL EVALUATION

Evaluation of the involved infant should include assessment, both above and below the knee. The percentage of limb shortening is relevant in that the percentage will not change, but the number of centimeters or inches will increase over time with growth. This will ultimately determine how many procedures may be required, if reconstruction is selected.

Even if there is no significant shortening in the thigh segment, there will be retroversion (7) and distal valgus (17), which will need to be addressed. The usual knee abnormalities include patellar hypoplasia and AP instability to testing.

In fibular hypoplasia or aplasia, there is shortening below the knee. The amount of shortening is not related to the degree of fibular absence (18). In some patients, even though a fibula may not be appreciated clinically or radiographically, the fibula can appear later (Fig. 1). There may be a fibular anlage, apparent only by palpation. If this is causing lateral tether, excision is recommended (Fig. 2). Occasionally, there is a dimple overlying the tibia as one may see overlying the femur in cases of congenital femoral deficiency.

Anteromedial bowing (valgus and procurvatum) is always present. The degree of this is variable. If a dimple exists, it is directly overlying the tibial deformity. Elimination of this deformity is usually permanent and can be combined with limb lengthening or amputation.

Mobility of the ankle joint is usually decreased compared to the contralateral side, if this side is normal. The ankle joint can be horizontal, spherical, or valgus in shape (12,18–21), but is always accompanied by decreased dorsiflexion in the author's experience. A valgus ankle position can often be clinically appreciated by the foot position. Hindfoot mobility may also be diminished due to the presence of a tarsal coalition, usually talocalcaneal (18,22). Although this is often not appreciable in newborn radiographs due to the lack of visible ossification, this can usually be appreciated by simple palpation of the hindfoot. An immobile hindfoot usually identifies a talocalcaneal coalition, despite being unable to identify it early by traditional radiography.

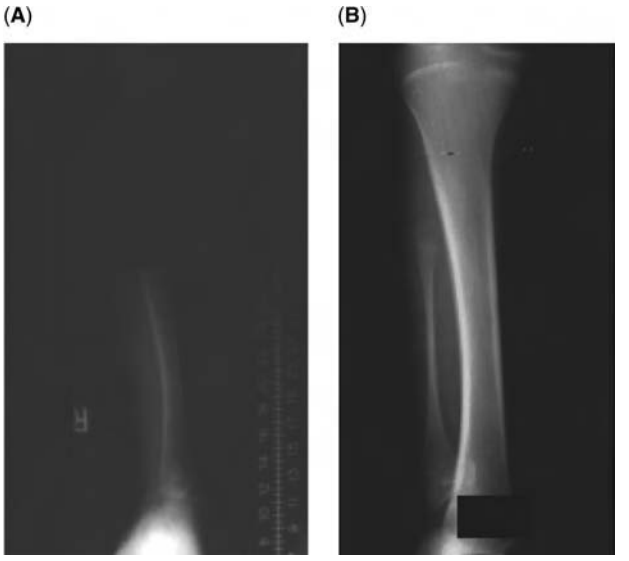

Figure 1 (**A**) Toddler X-ray with no visible fibula. (**B**) Subsequent X-ray with miniature fibula.

Although it is less common, a study by Caskey and Lester (10) identified a clubfoot, rather than an equinovalgus variation, in 23 cases in 147 limbs (6.49%). In their patients, tarsal coalition was the norm, rather than the exception.

Forefoot abnormalities are apparent clinically as missing lateral rays. A decreased number of toes may be associated with variable metatarsal deficiency.

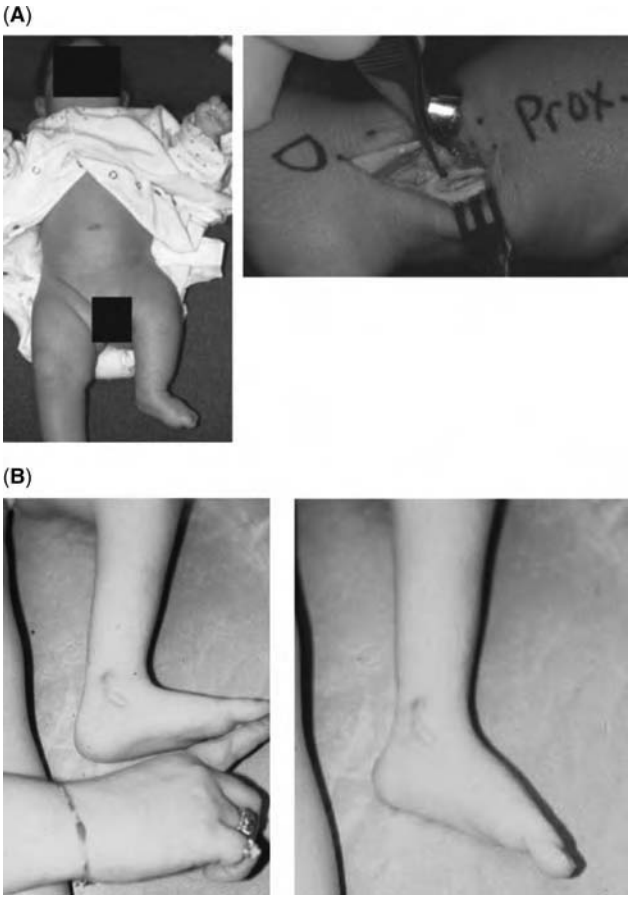

Figure 2 (**A**) Picture of infant with hypoplastic femur, fibular hemimelia with no fibula, and subsequent deforming anlage resection in the operating room. (**B**) Subsequent ankle motion.

Long supine radiographs with the unossified patella (palpable) anterior will delineate the basic problems of any significant femoral deformity, leg shortening, and valgus tibial angulation. Lateral radiographs of the leg will delineate the degree of procurvatum of the tibia. AP and lateral radiographs of the ankle will show ankle morphology. AP and lateral radiographs of the foot will show toe and metatarsal abnormalities and may help delineate hindfoot problems.

CLASSIFICATION

There have been several classification systems used to describe the spectrum of limb hypoplasia (2,18,23,24,24a). The most common are those of Achterman and Kalamchi (23), Coventry and Johnson (2), and recently, Stanitski and Stanitski (18). With the exception of the Stanitski classification and the Birch system (24), the other classification schemes do not address the ankle joint or distal tibial epiphyseal morphology, the subtalar joint or intrinsic foot abnormalities (18).

Briefly the Stanitski classification designates first I, II, or III delineating nearly normal (I), miniature (II), or absent fibula (III). Horizontal (H), spherical (S), or valgus (V) delineate the ankle joint shape. A small subscript "c" delineates a coalition. The number of rays (1–5) is then expressed numerically (18).

The proposed term of postaxial hypoplasia (9), although descriptive, is nonspecific and really does not describe the amount of fibula present or the involvement of the limb.

Abnormalities of the foot include abnormalities of the hindfoot, as well as abnormalities of the forefoot. In terms of the hindfoot, tarsal coalition of the talocalcaneal variety is relatively common (22,25). Although it is very hard to diagnose this radiographically in a newborn infant, one can usually diagnosis this fairly easily, clinically by palpation of a rigid hindfoot in an infant. The involved patient may be missing one or more lateral rays of the forefoot. Coalition in itself is not a reason for amputation, but takes careful explanation to the parents that this will never be a normally mobile hindfoot.

As opposed to what is often suggested in the literature, it is possible to see individuals with no fibula, a horizontal ankle joint, and five toes, including five metatarsals (Fig. 3).

The genetic abnormality that results in the occurrence of fibular hemimelia is unknown. It has been suggested that in humans, as in several quadruped species, fibular development is unstable developmentally because it is undergoing regression.

TREATMENT OPTIONS

Treatment options basically come down to ablation or reconstructive attempts (Fig. 4). The decision should be made as early as possible for the psychological benefit of the patient and family, as well as the physical benefit of the child. In general, one should try to avoid multiple reconstructive attempts followed by amputation. This tends to be the worse of either choice, both psychologically and physically.

The decision for reconstruction is very complex. Lengthening is never an easy process for the physician, patient, or caregiver (Figs. 5 and 6). It is made easier by listing the existing problems. All patients with fibular hypoplasia or aplasia have limb shortening, potential knee or ankle instability, and excessive femoral retroversion and distal femoral valgus (17,18). There may also be significant femoral shortening and possible nonunion, hip, ankle, and foot abnormalities.

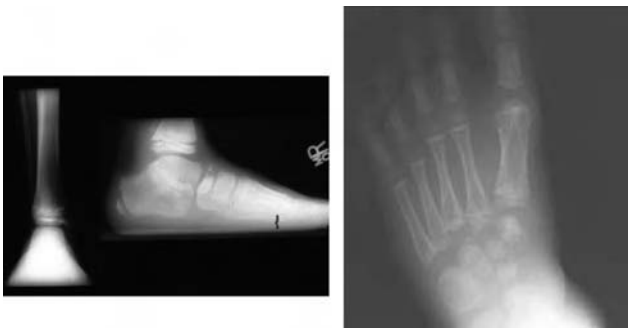

Figure 3 X-rays of foot and ankle of patient with absent fibula and five metatarsals and toes.

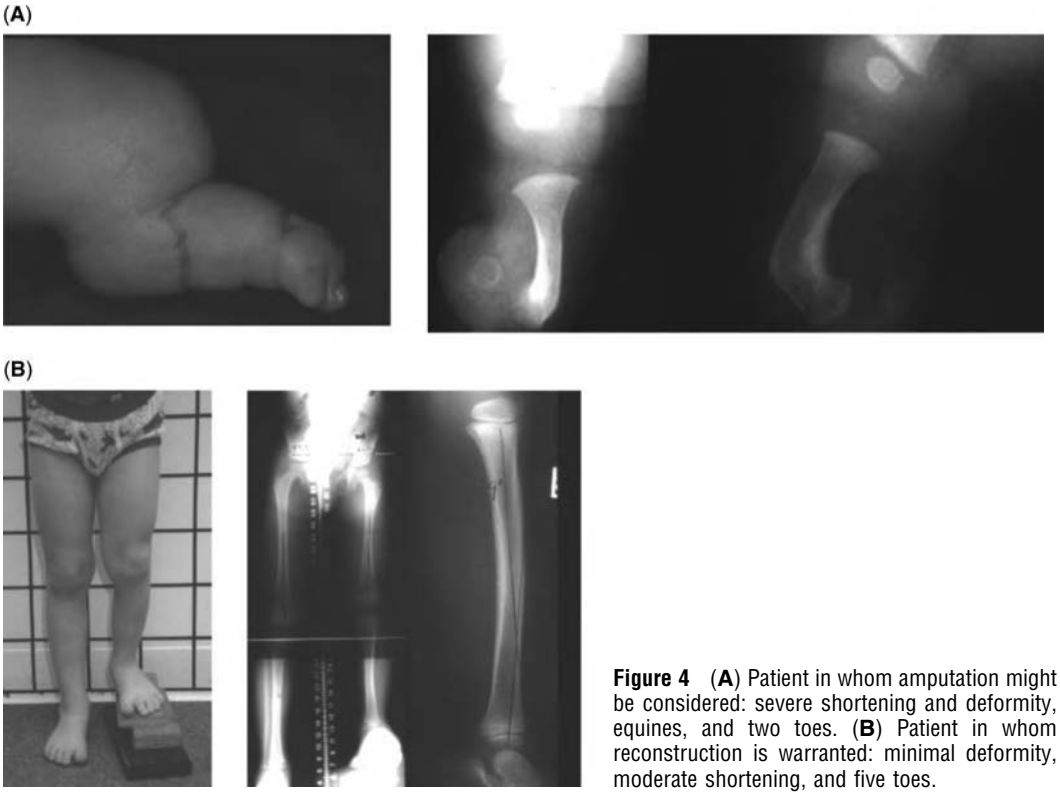

Figure 4 (**A**) Patient in whom amputation might be considered: severe shortening and deformity, equines, and two toes. (**B**) Patient in whom reconstruction is warranted: minimal deformity, moderate shortening, and five toes.

The author does not believe it is possible to create a ''normal'' ankle. To do so in a valgus ankle would malorient the distal tibial physis. In the past, physeal malorientation has resulted in eventual recurrence of the problem (26). It is possible to manage ''an uncontrollable'' ankle by fusion. This allows further lengthening to proceed without concern for equinus or valgus.

The issues that are of particular importance are associated abnormalities above the knee (hip, femoral shortening, and femoral nonunion) and the ultimate nature of the foot (18). The latter is controversial in that the designation of a ''good'' foot is not universally agreed upon.

Resection of a detrimental fibular anlage will require temporary use of an articulated ankle–foot orthosis and a shoe lift (Figs. 7 and 8), prior to lengthening attempts. This is useful in order to maintain ankle motion and avoid valgus.

SURGICAL TECHNIQUES

In the past, amputation was routinely recommended for this disorder (1,2,9,27,28).

A relatively recent study by McCarthy et al. still recommends amputation as being more cost effective and having superior clinical results than limb lengthening. This study evaluates 30 limbs and cites the incidence of fewer operations, fewer potential complications, and more active patients than limb lengthening. The study, however, fails to take into consideration parental refusal of amputation and the lifelong expense of prosthetic fitting and prosthetic care.

There is also difficulty truly assessing the virtues of amputation versus lengthening. That which should be considered, and has not been fully discussed in many articles favoring amputation is the nature of the limb. Apples were being compared with oranges in that ''fibular hypoplasia'' is a global, nonspecific term. Not clearly delineated were the nature of the limbs in the series. Not all patients with fibular hemimelia are alike.

If amputation is selected, it is probably best performed early for a number of potential reasons. If this is done prior to walking age, it does not interfere with the child's physical development. Also, early amputation as with other procedures have resulted in no-specific memory of the operation or hospitalization. The child amputee will act as though this is the norm for him or her. Cosmetically, it is no doubt more preferable for a male than for a female.

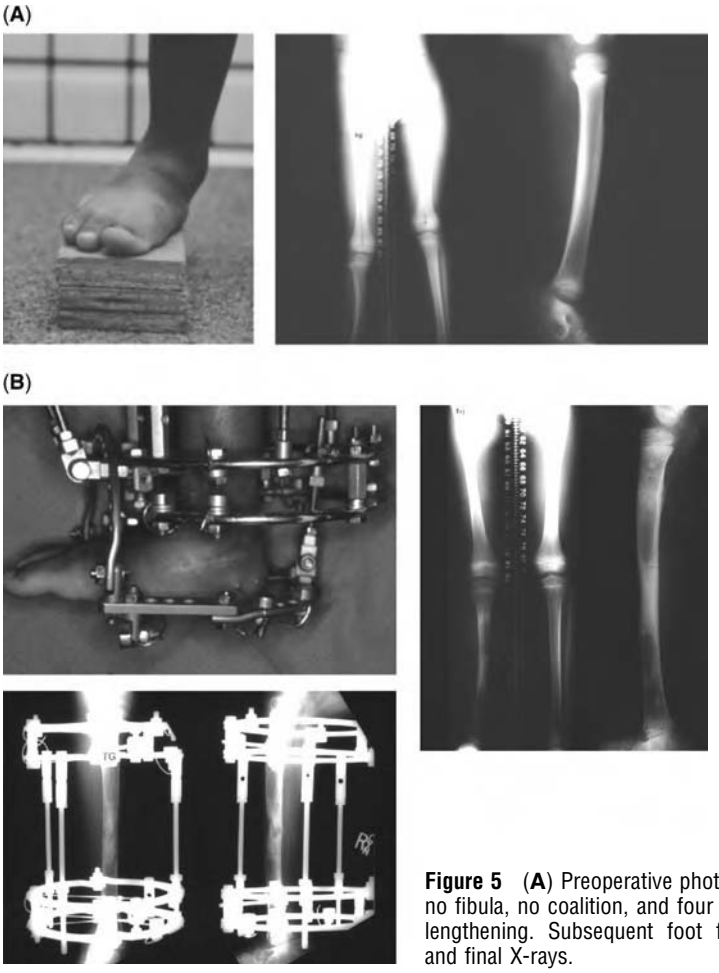

Figure 5 (**A**) Preoperative photo and long radiograph of patient with no fibula, no coalition, and four toes. (**B**) Photo in foot frame during lengthening. Subsequent foot frame removal during consolidation and final X-rays.

In general, the type of elective amputation in the growing child should ideally be through a joint and not through a bone. This avoids the potential of stump overgrowth and possible revisions. There is some argument of a Boyd versus a Syme amputation (8,13,30–32). Without exploring the relative pros and cons, both are endbearing and hence do not require prosthetic donning in order to go to the restroom, for example, in the middle of the night. Prosthetic adjustment can easily address length concerns. In either case, the apex anteromedial tibial deformity can be eliminated at the same surgical setting as the amputation and easily fixed temporarily with an intramedullary device, such as a smooth K-wire of appropriate caliber. This can easily be removed in the office when the tibial osteotomy is healed.

In the past, amputation has also been performed because of the perceived ability of not being able to create a ''good'' foot. There is some argument about what constitutes a ''good'' foot. Generally speaking, the author believes this would be a foot with a suitable weightbearing surface, which functions in normal daily activities and is painless to the individual involved. If the foot is missing lateral rays, the foot is always more narrow than the normal foot and often requires modification in shoewear, such as the addition of shoe orthoses. This, in itself, is not a good reason for amputation of a limb. Although amputation is certainly not something to be discarded entirely, it is probably relegated currently to those feet in which limb lengthening could not preserve a good weight-bearing foot surface or a reasonable ankle, hindfoot, and/or forefoot.

Even an ankle fusion if painless, although it does not move, should not be an indication for amputation and frequently facilitates lengthening because there is no need to control the foot position (Fig. 8).

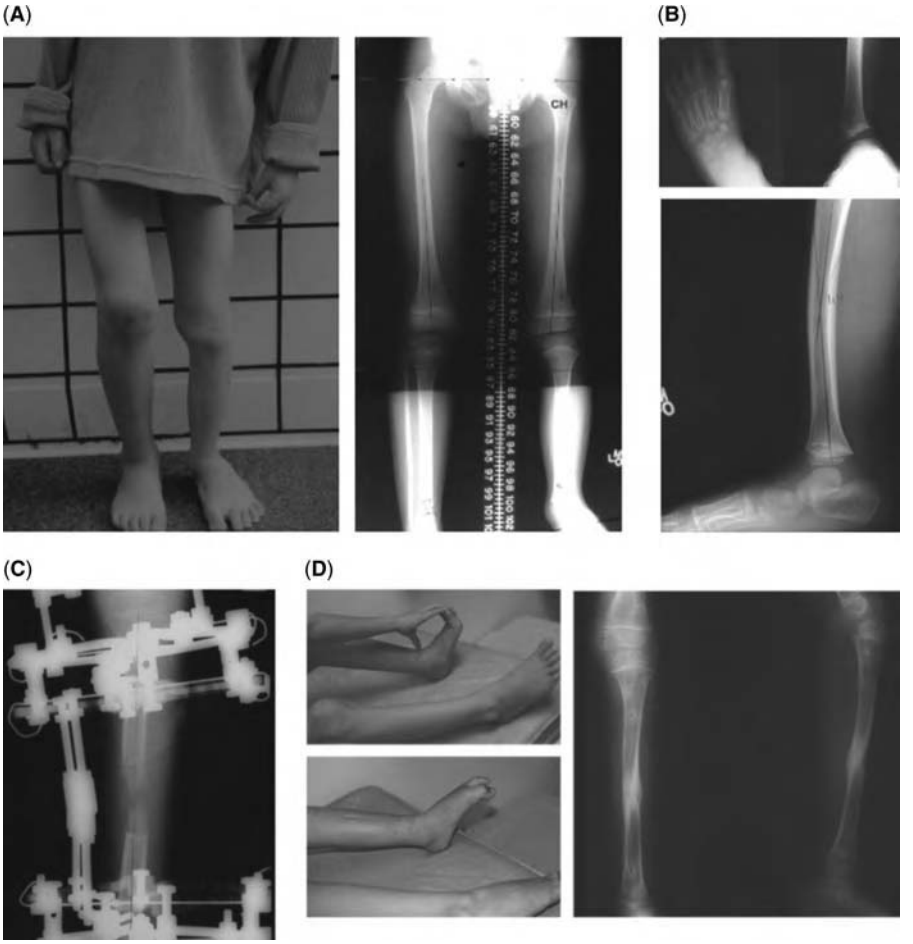

Figure 6 (**A**) Preoperative photo and radiographs of boy without fibula or coalition and five toes. (**B**) Preoperative radiographs demonstrating tibial and ankle deformity and five toes. (**C**) Radiograph during lengthening after correction of tibia valga. (**D**) Final photos showing ankle ROM and X-rays demonstrating elimination of valgus and procurvatum.

Limb lengthening can be complicated by psychological or physical factors (11,18,19,25,29,33,34). These have been well-described elsewhere and are not specific to fibular hemimelia. Prior to lengthening, attempts to create a horizontal ankle joint will cause malorientation of the distal tibial physis unless an intraepiphyseal osteomy is performed to correct this otherwise triangular-shaped distal tibial epiphysis (12,34a). Attempts at distal tibial osteotomy proximal to the distal growth plate by malorienting the growth plate will be fraught with recurrence similar to attempts to extend the distal femur in cases of myelodysplasia with knee flexion (26).

If in fact lengthening is elected, in general one must protect a mobile ankle and be very wary of the potential of knee subluxation. In the event of a mobile ankle, circular fixation can be removed from the foot once lengthening has been achieved.

Fixation is generally recommended due to the ability to easily extend the apparatus to the foot (Fig. 9) (11,18,34). After the lengthening has been completed, the foot portion of the frame can be removed during consolidation. Generally speaking, foot and/or ankle positions are not affected by consolidation, but rather by stretch during the lengthening process. If an ankle fusion has been performed, a uniplanar fixator can easily be used to lengthen a straight tibia without concerns for the ankle or foot as mentioned above.

The potential of knee subluxation, usually but not always posterior, is a concern. Care must be taken to maintain as much active and passive knee motion as possible, although it is never ''normal'' during lengthening. In cases of potential anterior knee instability (the risk of posterior subluxation), night knee extension splinting is helpful. In the case of decreased knee motion, lengthening can be slowed or stopped temporarily to regain motion.

Figure 7 Prior to lengthening, maintenance in articulated AFO and shoe lift.

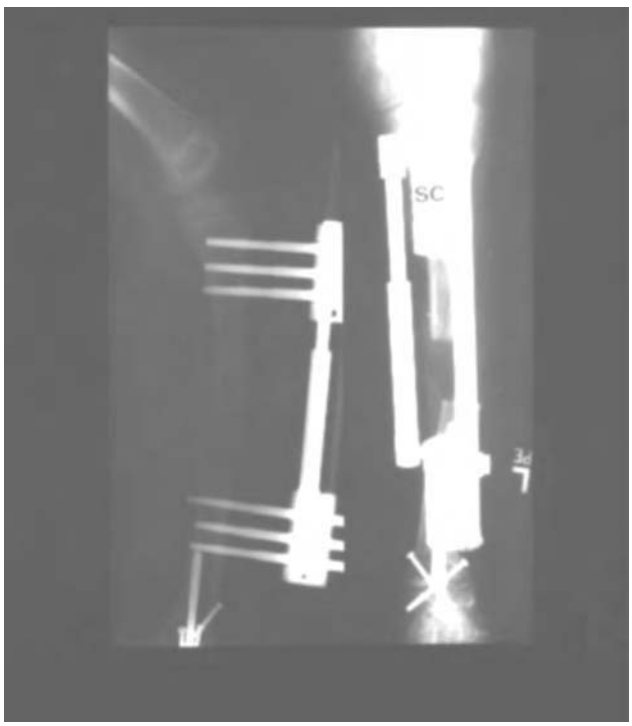

Figure 8 Monolateral device used to lengthen patient with prior ankle arthrodesis.

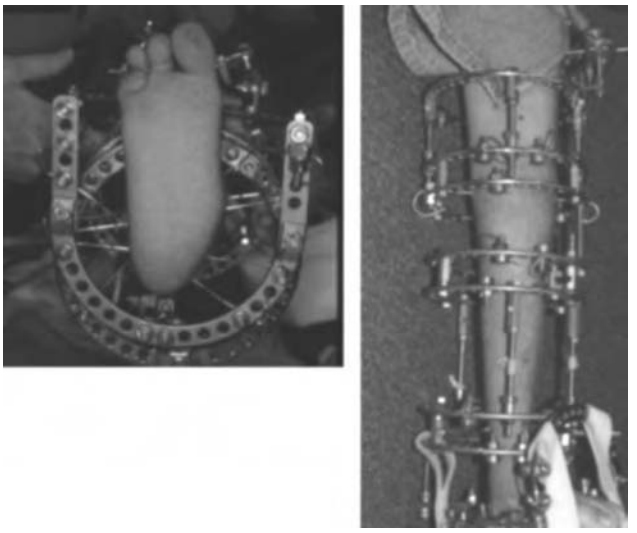

Figure 9 Foot frame in patient with potentially unstable ankle during lengthening.

If lengthening is chosen, as opposed to amputation, the treating physician must have a plan which includes: number and timing of the lengthening, timing of adjunctive procedures, such as contralateral epiphyseodesis or acute shortening and angular or rotational corrections of the ipsilateral femur +/−lengthening, and finally, "bail-out" options, should the goals not be realized. Any of the above can be combined with shortening options, such as epiphyseodesis or acute femoral shortening, in order to decrease the amount of lengthening necessary in the involved leg.

COMPLICATIONS

Complications of lengthening and fibular hemimelia in general are not specific to the condition with the exception of potential worsening of foot and/or ankle position (Fig. 10). Complications of tibial and/or femoral lengthening have been well described elsewhere. Intrinsic problems are related to the soft tissue and tend to be worse in congenital versus acquired problems.

Bone generation problems can generally be avoided, particularly in children, by vigilant radiographic assessment. Any narrowing of the regenerate bone by radiographs suggests that at least slowing the lengthening process is advised. Premature consolidation of the tibia, in the author's experience, has never been a problem even in a young child. Decrease in the ankle motion should be accompanied by slowing or temporarily stopping the lengthening and by vigorous physical therapy. Adjunctive tendon lengthening(s) may be required if motion cannot be restored. Knee flexion at night should be avoided by splintage that is removed during the day.

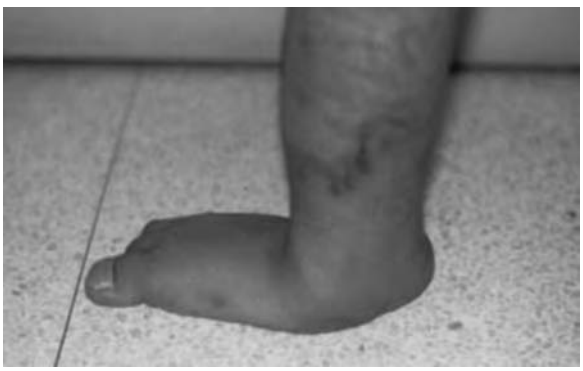

Figure 10 Worsening of foot and ankle position possible in fibular hypoplasia if this is not addressed during lengthening.

Table 1 Clinical Decision Making

Classification subgroup	Clinical Evaluation	Surgical Approach	Pearls	Complications Pitfalls
Horizontal ankle	Assess percentage of limb versus leg shortening	Number of lengthenings depends on shortening. Circular fixator to foot temporarily	Correct femur acutely with uniplanar device. Temporarily extend fixator to foot	Potential ankle instability
Spherical ankle	Percentage of limb shortening, ankle ROM	Same as above	Same as above	Potential ankle instability magnified by lengthening
Valgus ankle	Assess position of the foot related to the leg	Same as above. May need to lengthen heel cord if fusion performed, uniplanar fixator used	Address ankle instability	Will worsen ankle position if foot/ankle are not controlled during lengthening
Tarsal coalition	Assess hindfoot position	Circular fixation to foot	Will have decreased foot height (add to LLD prediction)	Worsening of lateral foot position

Abbrevition: ROM, range of motion.

FUTURE DIRECTIONS

As with all congenital limb shortening conditions, elimination would be the ultimate goal. This would obviate the need for complication-free lengthening.

To do this, it would be necessary to know the genetic causes of the problem and to be able to alter either the gene and/or the fetus. It would be advantageous to be able to stimulate growth of an isolated limb segment (e.g., the leg) and not just the entire organism.

Proper education for the parents as to the spectrum of the disease and the assets and liabilities in their child will avoid the immediate automatic response, which traditionally has been amputation and the correct option for treatment can be chosen.

REVIEW OF LITERATURE

Authors	Methods	Results	Conclusions
Achterma and Kalamchi (23)	Retrospective review of 81 patients from Dupont Institute	97 limbs in 81 patients, boys $>$ girls. Tarsal coalition more common with less fibula	Divided into I, IIa, and IIb dependent upon the amount of fibula present. Classic article, classification often quoted
Caskey and Lester (10)	Retrospective review of 23 clubfeet in 147 limbs (123 patients)	Equinovarus feet (+/−missing lateral rays) possible with fibular hemimelia	Clubfeet less common than valgus feet, but occurred with fibular hemimelia
Stanitski and Stanitski (18)	23 limbs in 32 patients	Attempt at functional classification	Ablation versus reconstruction decision can be made before walking age based on the foot and ankle
Stevens and Arms (35)	Reviewed 20 patients	Diagnosis involves entire limb, not just below the knee	Entire limb needs to be considered in this condition
McCarthy et al. (29)	33 limbs in 25 patients reviewed comparing outcomes of amputation versus lengthening	Ages of amputation versus lengthening different. Amputation outcomes better in terms of activity, pain, and the number of procedures	Even though good results can be obtained with limb lengthening, early amputation led to more active, pain-free children who have fewer procedures, complications are rare and as a group are more satisfied

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32 Growth Arrest

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INTRODUCTION

A growth arrest is an unexpected premature interruption of the longitudinal and/or latitudinal growth of a long bone. Defining the underlying etiology of the growth plate disturbance and the natural history of the problem is important when designing a treatment plan. A growth arrest is most frequently reported as a complication of a physeal fracture. Premature growth arrest is more common in the lower extremities, with post-traumatic arrest more frequent distally (1,2). A premature complete physeal arrest will result in only a retardation of bone length with no angular deformity. If the premature growth arrest involves only a portion of the growth plate, both an angular deformity and a limb-length discrepancy (LLD) will result. Other causes of growth plate disturbance are outlined in Table 1 (3–5).

THE PHYSIS

The physis is a highly organized, complex, and dynamic structure, located at the end of long bones. It is responsible for the lengthening of long bones during growth. The physis is located between the metaphysis and the undifferentiated epiphyseal cartilage. The major cellular component of the physis is cartilage. It assumes its characteristic cytoarchitectural arrangement by the fourth prenatal month and its basic cellular zones are well defined by birth. Detailed descriptions of the complex biophysical and molecular interactions involved in bone development are beyond the scope of this chapter. The readers are directed to a superbly written review on the normal growth plate and bone development by Forriol and Shapiro (6). The cartilage is arranged into three zones—the resting or germinal zone, the zone of proliferating chondrocytes, and a zone characterized by cellular hypertrophy. The hypertrophic zone is primarily responsible for longitudinal bone growth (6). The development and longitudinal growth of the physis is regulated by the interaction of various extrinsic and intrinsic molecular factors that influence the cellular activity of the chondrocytes in the hypertrophic zone (6) (Table 2). The extracellular matrix is composed of collagen, proteoglycans, and glycoproteins. The matrix components are major players in providing strength and stability (collagen) against shear forces experienced by the cartilaginous scaffold, and in resisting compression and deforming forces (proteoglycans). The glycoproteins organize the extracellular matrix. Although the basic cellular makeup of the growth plate is similar at all locations, major differences among the various growth plates do exist. Differences may include the cellularity of the various zones, the cell turnover rates, and the overall height of the physis. These factors may ultimately influence the physis' response to injury.

Physeal fractures typically occur through the hypertrophic zone, but can involve the germinal and proliferative zones.

The perichondrial ossification groove of Ranvier encircles the physis. Chondrocyte progenitor cells (resting cell layer) found in the groove of Ranvier are responsible for the increase in the transverse diameter of the physis. This ring contains the fibrocartilaginous ring of LaCroix. The ring of LaCroix is a band of fibrous tissue that merges with the periosteum of the bone to contribute to the latitudinal growth of the growth plate by appositional addition of chondrocytes and to provide mechanical stability and protection from compression, tension,

Table 1 Etiologies of Growth Arrest

Congenital causes
Metabolic or hematological abnormality
Skeletal dysplasias
Osteopetrosis
Sensory neuropathy
Blount's disease
Madelung's deformity
Acquired causes
Trauma
Burns
Electrical injury
Frostbite
Physeal fracture
Infection
Osteomyelitis
Poliomyelitis
Purpura fulminans
Septic arthritis
Systemic infection (e.g., meningococcemia)
Tumor
Multiple osteochondromatosis
Enchondromatosis
Therapeutic irradiation
Metabolic or hematological abnormality
Hypervitaminosis A
Vitamin A toxicity
Sickle cell anemia
Microvascular ischemia
Disuse
Insertion of metal (smooth vs. threaded pins)

and shear loads experienced at this weak area of the physis. Damage to either of these structures may also result in a growth disturbance.

PHYSEAL BRIDGE (PHYSEAL BAR)

Premature growth arrest—partial or complete—may occur when there is contact between the bone of the epiphysis and the metaphysis, and a bridge of bone (also known as a physeal bridge or a physeal bar) forms between these two anatomic structures. A physeal bridge may begin to form as early as one to two months after injury but may not be become clinically

Table 2 Factors That Influence Chondrocytes Activity in the Hypertrophic Zone

Intrinsic factors
Indian hedgehog
Runx2 and *Runx3*
Parathyroid hormone-related protein
Bone morphogenetic proteins
Fibroblast growth factors
Vascular endothelial growth factor
Matrix metalloproteinases
Cell cycle proteins
Cyclins, cyclin-dependent kinases, CD kinase inhibitors, activating transcription factor 2
Extrinsic factors
Growth hormone
Insulin-like growth factor I/II
Thyroid hormone (T3, T4)
Vitamin D3
Glucocorticoids
Estrogen

Source: From Ref. 6.

apparent until years later (7,8). All bone bridges are a result of damage to the epiphyseal growth plate cells. It has been theorized from numerous animal studies that the development of physeal bone bridges is initiated by neural and vascular insults to the epiphysis (9–12). All these investigators (9–12) reported that normal response of the epiphysis and the metaphysis to injury is through the formation of reactive inflammatory fibrous tissue, and it may facilitate the formation of a physeal bridge, should this tissue vascularize. The cartilage along these transphyseal vessels undergoes calcification, and may foreshadow the eventual chondro-osseous transformation leading to the development of a physeal bar (13).

Certain sites are prone to post-traumatic growth arrest. Although distal femoral physeal fractures are uncommon (1.4% of all physeal fractures), there is a high prevalence of subsequent growth arrest (14). This also occurs with distal tibial physeal fractures. A likely explanation is that this is secondary to the complex geometry of the growth plate. There is a central physeal undulation in the distal femur and an anteromedial undulation in the distal tibia (Kump's bump). Physiological physeal closure begins in these areas (15). It is theorized that injury to the germinal and proliferative zones of the growth plate occurs at the undulations. Other factors to consider include the age of the child, the type of injury, the force of injury, compression, displacement, and infolding of the periosteum (16).

CLINICAL EVALUATION

Physeal bars may begin to develop as soon as one to two months after the cellular components of the growth plate have sustained an insult (7,8), but the clinical manifestations may not become apparent for many years, often not until the adolescent growth spurt. The development of an angular deformity or a relative shortening of the involved extremity during this time ultimately results in a visit to the orthopedic surgeon's office. A detailed clinical history, physical examination, and routine roentgenograms will localize the involved physis (10) and aid in determining a treatment plan. Erect leg X-rays should be used to assess LLD and angular deformity in the lower extremity. Joint orientation angles will be helpful in diagnosing deformity in the coronal and sagittal planes. The extent of growth plate damage and age of the child (growth remaining) are important factors in choosing a treatment approach.

DIAGNOSTIC IMAGING EVALUATION

Diagnostic imaging studies are essential for diagnosing the presence of a physeal bar and then determining its size and location. Table 3 summarizes the current diagnostic imaging options available and the advantages and disadvantages of each imaging technique. Although the plain radiograph remains the standard initial diagnostic imaging study used to confirm the presence and site of the physeal bridge, magnetic resonance imaging (MRI) is now the gold standard used to evaluate and map the physeal bridge, and to determine the extent of damage to the growth plate (Fig. 1).

Park or Harris lines, also called growth recovery lines, represent transverse rather than longitudinally oriented bone trabeculae (17). These lines form during a period of slowed growth secondary to illness or injury. If normal growth resumes, the line is parallel to and grows away from the physis. If the growth recovery line is not parallel to the physis, this indicates a tethering of growth. Small, peripheral bone bridges produce the most characteristic growth recovery lines (15).

Advances in MRI techniques now allow for detailed information to be obtained about the growth plate. Harcke et al. (18) demonstrated that a field-echo pulse sequence with repetition time of 700 msec, echo time of 20 msec, and a flip angle of 40° optimized the resolution of the growth-plate cartilage. They also stated that in the coronal and sagittal images of the normal knee, a ''drop-out'' sign or a discontinuity of the physeal cartilage may be present and should not be mistaken for premature closure of the physis. Ecklund and Jaramillo (15) recommended that fat-suppressed three-dimensional (3D) spoiled gradient-recall echo sequences best demonstrated a growth disturbance and associated abnormalities in children. In addition to these two MRI techniques, Petersen (14) recommended that a T1-weighted gradient with water excitation sequence also be obtained. This sequence provides a true anatomic image based on volume data and provides focused information on the physis and the physeal bridge.

Table 3 Diagnostic Imaging for Evaluation of the Growth Plate

Radiographic evaluation	Advantages	Disadvantages	General comments
Plain radiograph	Identifies the presence of a bony bridge	Improper orientation of the physis relative to the X-ray beam may lead to errant interpretation	Harris growth arrest lines may be the earliest sign of physeal bridge formation Harris growth arrest lines that are not parallel to the physis indicate the presence of a physeal injury
Bone age	Estimate skeletal age Determine potential for growth		Skeletal age dictates the treatment options
Scintigraphy	A physeal bar (or postepiphysiodesis) will have focal decreased uptake	Requires the intravenous injection of a bone seeking radiopharmaceutical Time consuming as patient must be imaged at the time of injection, immediately after the injection, 2–4 hr after the radiotracer administration and at 24 hr after the injection	Distribution of the tracer in bone relates to blood flow, bone turnover, and growth
Scanogram	Documents radiographically the limb length discrepancy	Technical errors in positioning the patient	Serial films may be used to document changes in limb length discrepancy
Computed tomography (CT) scan	Two and three dimensional multiplanar reconstructions are easily obtained Spiral (or helical) CT is very fast at data acquisition Defines the location, duration and size of the physeal bar in the sagittal plane and coronal plane Decreased dose of radiation	Spiral CT may be blurry. If the bar is too small, it may be missed. If the bar is oriented perpendicular to the plane of the CT it may be missed Cannot provide information on the status of the physeal cartilage and surrounding ligaments	
MRI (7)	Defines the location, duration, and size of a physeal bar Identifies a healthy vs. injured physis	Patient may need to be sedated	Harris growth arrest lines can be seen 6 to 7 wk earlier than on normal radiographs Preferred preoperative imaging technique to map a physeal bar MRI immediately postop. to detect incomplete resection MRI recommended 6 mo after surgery to detect bridge recurrence, migration, and necrosis of the interposition material (if fat graft is used) and to evaluate the remnant physis and its repair potential (8)

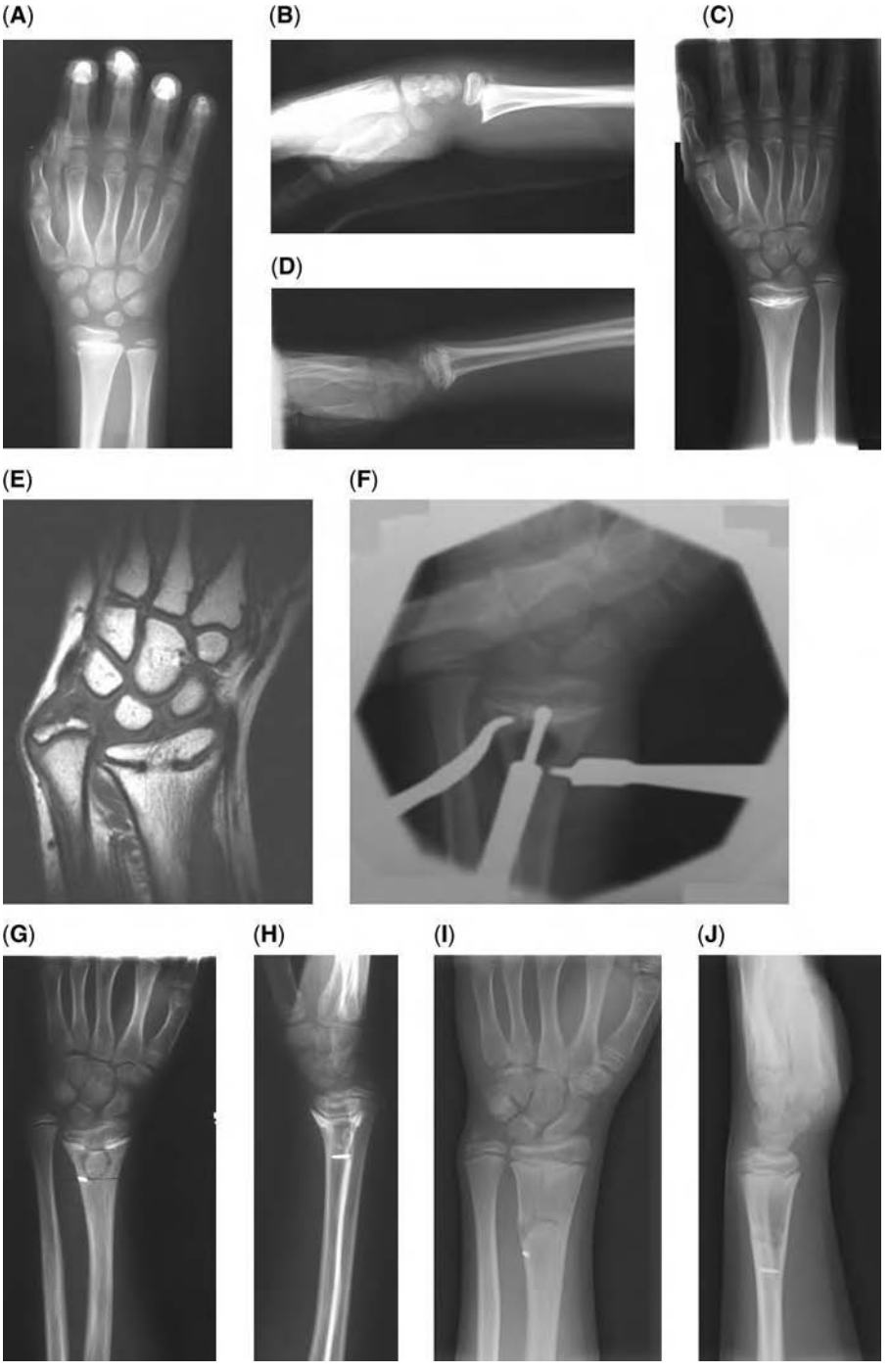

Figure 1 (*Caption on next page*)

MRI is ideal for assessing growth arrests (7,15,19,20). T1-weighted images reveal growth recovery lines and large physeal bridges that have high signal intensity (15). Smaller bridges have variable signal intensity. Intermediate and T2-weighted images demonstrate the cartilage extensions from the growth plate into the metaphysis, which are common after physeal injury (15). In gradient-recalled images, bone bridges are seen as low-signal-intensity interruptions in the high-signal-intensity physeal cartilage (15). The volumetric fat-suppressed 3D-spoiled

gradient-recalled echo sequence allows accurate mapping of the bone bridges (15,20). These bridges may not be discrete confluent areas; irregular nonconfluent areas of bridging occurs (20).

CLASSIFICATION

Peterson (10) proposed a classification system according to the location of the physeal bridge responsible for partial growth arrests (Table 4). Both the size and the location of the physeal bridge may ultimately influence the clinical deformity. This classification system contributes to the overall management of the physeal bridge by conveying both diagnostic and prognostic information.

TREATMENT OPTIONS

All skeletally immature patients who have sustained an injury to a physis should be followed closely through to skeletal maturity. The presence of a physeal bridge may not be a precursor to the development of an angular deformity of a long bone or a LLD, nor is its presence an indication for surgical intervention. Johnson and Southwick (21) reported that growth could continue normally despite the presence of a physeal bridge if the lesion is small. In such instances, continued growth can disrupt an immature bony bridge, thereby permitting longitudinal growth to be re-established.

The growth plates of the distal femur and proximal tibia account for 60% to 70% of the growth of the limb. The impact of permanent growth plate injury is more significant in these compared to slower growing physes.

Treatment options may involve a combination of orthotics, shoe wear modifications, and/or surgical intervention. Four important factors must be considered when formulating a treatment plan for a patient with a documented physeal bar. First, the etiology of the growth arrest should be considered. The results of resecting a physeal bridge are less predictable post-sepsis than when the growth arrest has occurred as a complication of a fracture (Fig. 2). Second, the amount of growth remaining must be considered. Ideally, the patient should have at least two years or 2 cm of growth remaining (7). Third, the physeal bar must be mapped to evaluate both the location and the extent of involvement. The location of the physeal bar ultimately influences the probable success of surgical resection. The location of the physeal bar will also dictate the potential angular deformity and limb length deficiency that may arise. If the physeal bar is eccentrically located, the tethering effect will result in an angular deformity. If the physeal bar is centrally located, then the growth in the periphery will cause cupping of the metaphyseal bone and a tethering in the central region, and a LLD will occur. Surgical resection of the physeal bar is recommended when the physeal bridge involves 50% or lower of the physeal area (7,8). However, in very young children, resecting physeal bridges of over 50% of the physeal area could be considered because the alternatives (e.g., repeated corrective osteotomies and lengthening) are undesirable (7,8). Lastly, the presence of angular deformity must be assessed both clinically and radiographically. Often an angular deformity is present in the involved limb leaving a decision as to whether a corrective osteotomy should be done with

Figure 1 (*Continued from previous page*) A case of a 12-year-old male who sustained a displaced Salter-2 fracture of the left distal radius at age eight. (**A** and **B**) The fracture was reduced under conscious sedation and a long arm cast was applied. Follow-up examination at eight weeks showed normal alignment. The patient returned to the clinic 30 months post injury with a complaint of prominence of the ulna. Radiographic examination (**C** and **D**) shows a shortened radius with a Type II physeal arrest. An magnetic resonance imaging (**E**) showed a central growth arrest with less than 50% of the physis involved. A dorsal radial metaphyseal exposure was used. Using multiple drill holes, an oval section of metaphyseal bone was removed to achieve visualization. Using the image intensifier, the center of the growth arrest was identified and a bur was used to remove the bar (**F**). This was extremely thick sclerotic bone. A side-cutting bit was used to expand the hole back to normal growth plate. This was done circumferentially using a 70° arthroscope to visualize the parts of the growth plate, which were not visible by direct vision. Bone wax was applied to the epiphyseal and adjacent metaphyseal bone. Cranioplast was then used to fill the defect and a 5 mm titanium wire was advanced into the distal radial metaphysis to measure subsequent growth. X-rays taken three weeks postoperatively show the early appearance (**G** and **H**). At last follow-up (26 months post surgery, 13 years old), X-rays show continued growth of the distal radius with no demonstrative angulation noted (**I** and **J**).

Table 4 Classification of Growth Arrest

Type of growth arrest	Location	Features
Complete		Global involvement of the physis
Partial type 1	Peripheral	Variable sized bridge along the margin of the physis Zone of Ranvier is damaged Overgrowth of periosteum with extension farther toward the physis than normal Associated with severe angular deformity
Partial type 2	Linear	A longitudinal structure involving the middle of the physis Connecting and involving two separate segments of the physis There is normal physeal tissue on either side Most commonly a consequence of Salter-Harris type III or IV fractures
Partial type 3	Central	Most severe type of injury Variable-sized osseous bridge forms within the central portion of the physis and is completely surrounded by normal physis Characterized by a conical extension of the epiphyseal ossification center into the metaphysis Acts as a central tether Peripheral zone of Ranvier usually is uninvolved Major effect is retardation of longitudinal growth; and tenting of the physis with eventual distortion of the articular surface

Source: From Ref. 10.

the bridge resection. Although absolute indications for the simultaneous resection of a physeal bridge and performance of a corrective osteotomy cannot be found in the literature, Williamson and Staehli (22) have reported that the spontaneous correction of angular deformities is unpredictable. It is currently recommended that angular deformities over 20° be corrected surgically with an osteotomy because they will probably not correct spontaneously (4). The corrective osteotomy may be performed at the same time as the physeal bridge is resected or it may be done at a later date. Williamson and Staehli (22) recommended corrective osteotomy at the time of bridge resection for all knee and ankle deformities over 10°, especially when the area of the growth arrest is over 25% of the physis. Ogden (4) suggested that corrective osteotomy not be performed at the same time, preferring to wait a few months to see if the physeal bridge has been successfully resected. If there is evidence of recurrence of the physeal bridge, then it is recommended to again resect the physeal bridge and perform a corrective osteotomy (Fig. 3) (4). If the patient is older, has less than two years of growth remaining or less than 2 cm of growth remaining, and has a mild angular deformity, the growth of the injured physis may be arrested surgically (10).

The presence of a LLD of 2 cm or less rarely results in functional impairment and can be treated nonoperatively with a shoe lift if needed. If a LLD is anticipated to be over 2 cm at skeletal maturity, it may be limited with an epiphysiodesis of the contralateral bone (23). If the anticipated LLD is greater or the patient is close to skeletal maturity, consideration should be given to lengthening the involved limb.

The decision to subject the patient to a one-stage procedure (a combination of physeal bar resection and corrective osteotomy) versus potentially a two-stage procedure (initial resection of the physeal bar followed by a corrective osteotomy at a later date) must be determined by the location and size of the physeal bridge and the location and degree of angular deformity and/or LLD.

SURGICAL OPTIONS FOR PHYSEAL BAR RESECTION

The ultimate goal of surgery is to provide an environment to promote the recovery of the growth potential of the involved epiphysis and hence correct both LLD and angular deformity if present. The common underlying surgical theme when treating physeal bars includes the complete removal of the bone bridge while preserving as much of the healthy portion of

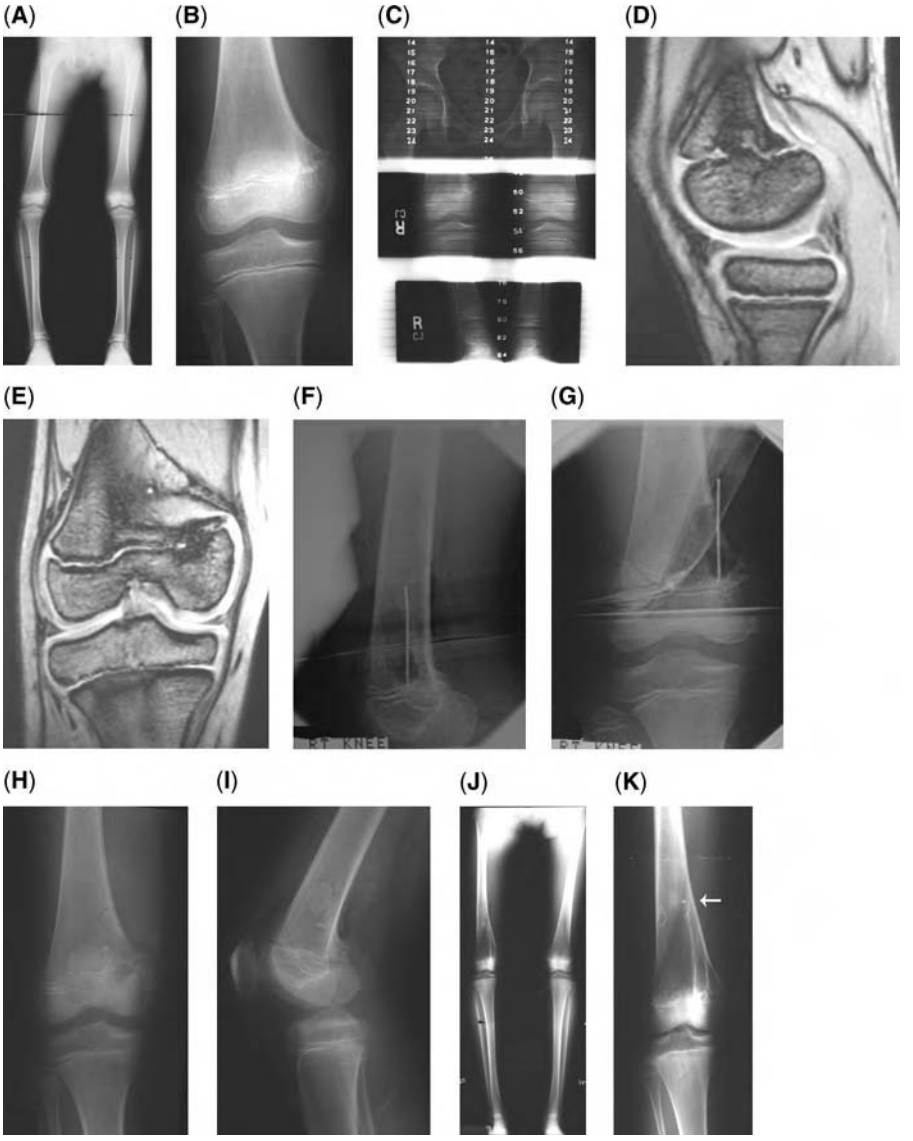

Figure 2 A case of a 16-year-old female with osteomyelitis and septic arthritis as a neonate. She was treated successfully with antibiotics. Mother noticed a limp and slight bowing of the right leg at age eight and brought the patient into the clinic. Radiographs taken at that time (**A**–**C**) show focal premature closure of the posteromedial aspect of the distal femoral growth plate. A varus deformity is noted without LLD. The patient was sent for an MRI to obtain a more detailed view of the physeal arrest (**D** and **E**). The physeal bar was removed surgically (**F** and **G**). The bar was removed with a burr and curettes. A local interpositional fat graft was used. Postoperative radiographs (**H** and **I**). Clinical and radiographic examination three years postoperative, show anatomical symmetrical alignment about the knee with the distal femoral physis continuing to grow. Note the column of fat in the metaphysis (**J** and **K**). Five years postoperatively, her angular deformity and leg length discrepancy began to recur. To prevent progressive leg length discrepancy and angular deformity, bilateral distal femoral epiphysiodeses were performed. On last follow-up (at skeletal maturity), she had a leg length discrepancy of 1.6 cm and a distal lateral femoral angle of 92°, compared to 87° on the left side.

the physis as possible, and then filling of the cavity with interpositional material to prevent recurrence of the bridge. The surgical approaches and the options available are dependent upon the amount of growth remaining, the underlying etiology of the growth arrest, the location and size of the physeal bar, and finally the presence of an angular deformity and/or LLD. The surgical options are reviewed in Table 5.

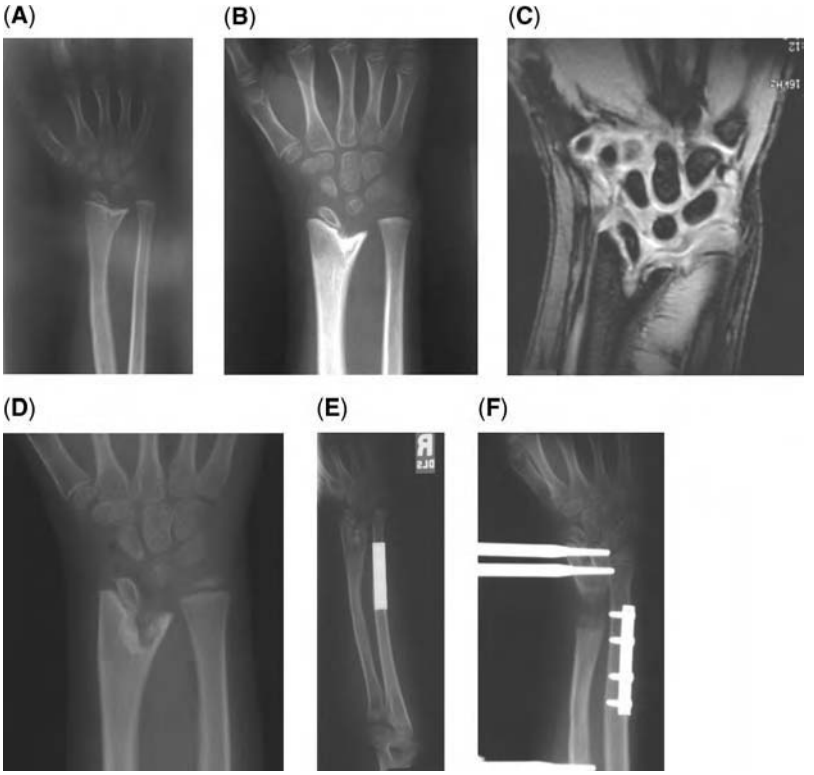

Figure 3 A four-year-old female with a Salter Harris type IV fracture of the right distal radius. Plain films at the time of diagnosis showed the fracture. (**A**) The fracture was treated with a short arm cast for five weeks. Follow-up films seven months after initial injury show a possible Type I peripheral growth arrest. (**B**) An MRI was obtained, which demonstrates growth failure and extension of the physeal cartilage into the metaphysis without an obvious bone bridge. (**C**) X rays taken immediately after physeal bar resection and fat graft interposition show satisfactory resection (**D**), but growth did not resume. X-rays taken after the second attempted bar resection and fat graft interposition. Ulnar shortening and radial osteotomy was done. (**E**) Growth did not resume and the patient was treated with a radial lengthening (**F**), and the plan is to do future lengthenings.

A technique that can be employed toward the end of growth is epiphyseal distraction or hemichondrodiatasis (24–26). Canadell and de Pablos (25) reported breaking bony bridges by physeal distraction, although the exact extent of growth plate involvement was not reported. Aldegheri et al. (24) note that the best results are achieved in post-traumatic cases when the bone bridge occupies less than 20% to 30% of the epiphyseal plate. Angular deformities, commonly associated with eccentric bridges, can be also addressed at the time of physeal distraction (26,27). Canadell and de Pablos (26) report 45 cases of physeal distractions of the lower extremity using a modified Wagner apparatus with a bony bridge present. Resection of the bridge was not performed (all cases $< 50\%$ growth plate involvement) and the physis was distracted. The advantages of this technique include correction of both length and angular deformities. The primary complication is a premature closure of the distracted growth plate (28). Zarzycki et al. (27) have reported that in 24 femoral and 16 tibial physeal lengthenings, the growth plate resumed its activity after the lengthening treatment and fused at the same time as the opposite limb. Their technique utilized a distraction rate of 2×0.25 mm per 24 hours with an overall mean femoral treatment time of 170.7 days (mean healing index of 38.1 days/cm) and an overall mean tibial treatment time of 155.1 days (mean healing index of 35.7 days/cm).

It is suggested that symmetrical physeal distraction be performed first. Once the physis has been shown to be open, the angular deformity can be corrected (25). Canadell and de Pablos advise that if Langenskiold's technique of interpositional insertion is utilized, it not be combined with physeal distraction in one stage.

Table 5 Surgical Options for Physeal Bar Resection

Location of physeal bridge	Surgical technique	Pearls
Type 1 (peripheral)	Approach directly Remove the dense sclerotic bridge from outside to inside until normal physis is visualized all along the periphery of the cavity. Exposed physis should be an opaque white and should be evident out to the cortical edge	Remove overlying periostium
Type II (linear)	Approach through an extraphyseal bone tunnel	Preoperative MRI and reconstruction, intraoperative fluoroscopy
Type III (central)	Approach through a surgically created defect in the metaphysis The transmetaphyseal approach requires removal of a window of cortical bone and internal cancellous metaphyseal bone before the physeal bridge is reached After removal of the entire bridge, the normal physis must be visualized circumferentially in the cavity	Small dental mirror, headlamp arthroscope

To monitor the successful resection of the physeal bridge, it is suggested that metal markers (titanium is ideal for MRI) be placed in the metaphysis and epiphysis. Postoperatively, the distance between the markers can be measured to determine if growth has been restored or if a secondary tether has developed (29). Postoperative MRIs can prove useful if plain radiographs fail to show an open physis. Early recurrence could be treated with physeal distraction and could eliminate the need to surgically resect the physis for a second time.

INTERPOSITION MATERIAL

Over the years, a variety of materials have been used to fill the cavity created by excision of the physeal bar. These materials include autogenous fat, polymethylmethacrylate (PMMA), bone wax, hyaline cartilage (approved for experimental use only), muscle, and silicone rubber. The two most commonly used interpositional materials today are autogenous fat and PMMA.

Autogenous fat is the most commonly used material. Langenskiold (30) reported the use of autogenous fat obtained from the buttocks because of its firm and globular consistency. Autogenous fat may also be obtained locally from the edges of the incision to fill the defect. The major disadvantage of using fat is related to its inability to provide hemostasis within the cavity that is left after resection of the physeal bar. As a result of the lack of hemostasis, fat interposition grafts have a tendency to float away from the physeal edges when the tourniquet is released. It is ill-advised to attempt to obtain stability for the autogenous fat interposition graft by closing the local periosteum over the cavity. This predisposes to the development of new bone formation peripherally, which may act as a secondary tether postoperatively. To prevent the migration of the fat graft caused by bleeding within the cavity, Tachdjian (31) reported that the fat graft may be stabilized within the cavity by suturing it to the surrounding epiphysis and metaphysis. Ogden (4) recommended that the sides of the cavity created by resection of the physeal bar, should be relatively flat and smooth. He also suggested that when fat is used as the interposition graft, the metaphyseal and epiphyseal bone should be undermined away from the physeal edges to reduce the likelihood of physeal bridge recurrence. Khoshhal and Kiefer (8) report using bone wax on the osseous surfaces to decrease bleeding and hematoma formation. The use of fat in the cavity left after physeal bar resection may predispose the bone to fracture. Autogenous fat does not provide the bone with any immediate intrinsic stability (4). Several authors (32–34) reported that in experimental studies, the fat implants revascularize and survive. It remains unknown if this occurs in children as well. Peterson (10) reported that fat interposition grafts may shrink and/or undergo necrosis. This may predispose to the recurrence of a physeal bar.

Methylmethacrylate is the second most-commonly used interpositional material. This radiolucent, inert material has been implanted since the 1940s. There have been no reports in the literature of this material causing a rejection reaction nor has it been implicated as a carcinogenic material to humans. Because PMMA is easy to mold, it provides good hemostasis because it can fill the cavity created by the resection of the physeal bar. PMMA is also able to provide structural stability to the weakened bone because it forms a solid substance that is capable of completely filling the cavity. Several suggestions have been reported in the literature to improve the adherence of the PMMA plug to the epiphysis and prevent its migration. Peterson (10) suggested creating drill holes in the epiphysis; and Khoshhal and Kiefer (8) suggested that the epiphyseal walls be undermined with a right-angled curette to help anchor the PMMA to the epiphyseal walls. The authors will occasionally anchor the PMMA with an epiphyseal K-wire or screw. The metaphyseal cavity should be filled with either autograft or allograft cancellous bone. Autograft bone can be supplemented with allograft cancellous bone if needed.

Postoperatively, the use of PMMA as the interpositional material allows for a shorter period of immobilization. The authors suggest that PMMA be the interpositional material of choice when a large void is left following resection of a physeal bar.

Several other materials have been used experimentally and are not yet approved for routine use in humans. These materials include silicone rubber and hyaline cartilage. Silicone rubber is a biologically inert substance. However, the need to sterilize both the monomer and the catalyst the night before the surgical procedure, make it impractical to use. Although currently approved for experimental implantation only, hyaline cartilage has been shown to be superior to any other material currently being used for interposition grafting. Potential autogenous sources include a rib or sternal cartilage.

POSTOPERATIVE CARE

The decision to allow range of motion of the involved joint and/or weight bearing is made on an individual basis. Several factors must be considered, including: the location of the physeal bar, the size of the cavity, the age of the patient, whether or not an osteotomy was performed to correct an angular deformity and the method of fixation, and the interpositional material used.

The patient is allowed to begin range of motion exercises of the involved limb immediately; if fat is used to fill the cavity, a period of partial weight bearing is recommended. If PMMA is used to fill the cavity, a supportive splint or brace may be used for comfort; however, weight bearing should be encouraged as soon as possible.

When a corrective osteotomy has been performed concomitantly with the resection of a physeal bridge, the decision to allow the patient to bear weight is dependent upon the type of fixation (internal vs. external) and the type of angular correction (acute vs. gradual).

COMPLICATIONS

Following the resection of a physeal bridge, it is imperative that all patients be subjected to regular follow-up until they reach skeletal maturity. Regular clinical and radiological examinations, including scanograms, are necessary to ensure that physeal growth is maintained following resection of the physeal bridge. Several factors have been documented in the literature to explain why the surgical excision of a physeal bridge is fraught with unpredictable results. Results of treatment reported in the literature are 15% to 38% poor and fair (22,35–37). The most common complication is recurrence of the physeal bar. Several factors been linked to the reformation of a physeal bar following resection. One of the major factors is related to the size of the initial physeal bridge and hence the amount of healthy physis remaining. When the physeal bridge involves over 50% of the physis, there is a high probability that it may recur. Even with appropriate indications, there may be a recurrence of the physeal bridge. Recurrence may be due to incomplete resection at the index procedure. Hasler and Foster (29) reported that if the volume of interposition material used is inadequate or, as in the case of fat interposition grafts, the graft shrinks or becomes dislodged from the cavity, the risk of physeal bar recurrence is increased. It is imperative that the interposition graft material fills the entire defect in the epiphysis and metaphysis, and the ossification center. The graft should abut the exposed physis completely. Failure to do so increases the risk of recurrence.

If a cortical metaphyseal window was created to excise the physeal bar, it should be replaced. The metaphyseal periosteum and cortical bone that originally crossed the involved growth plate should not be reinserted or reattached.

Should a physeal bar recur, a repeat surgical resection may be considered. The surgical indications for primary resection as described earlier in this chapter apply here as well (at least 2 cm of growth remaining and $< 50\%$ of the physeal area). If the recurrent physeal bar is to be surgically resected, the angular deformity, if any, as well as any LLD that may be present, should be assessed. If an angular deformity is present, surgical realignment at the time of excision of the recurrent physeal bar should be performed. When insufficient growth remains and an angular deformity and/or a significant LLD is present, gradual correction of the deformity can be performed (38).

Physeal bridges, which occur secondary to osteomyelitis or congenital physeal abnormalities, such as Madelung deformity, may behave quite differently from those, which occur as a result of a traumatic injury to physis. The prognosis for recovery of growth function following resection of the physeal bridge in these cases is unpredictable and poor. Despite the resection of the physeal bar in these cases, the remaining ''healthy'' physeal cells may not be capable of responding to the cellular growth signals and, hence, limited or no growth may occur.

It is not uncommon for growth to resume following resection of a physeal bar and then for it to cease unexpectedly. This phenomenon should not be considered a complication but rather a normal physiological response to physeal injury (4,35,37). Should premature closure of the involved physis occur, an epiphysiodesis of the contralateral epiphysis can be performed. Similarly, if a physeal bridge recurs when the patient is nearing skeletal maturity or the entire physis closes on the injured side but remains open on the contralateral side, an epiphysiodesis of the contralateral side should be considered. Peterson reported in 1988 (39) that the ipsilateral physis at the opposite end of the bone may overgrow in response to the lack of longitudinal growth from the injured physis.

Aside from the above noted complications, there is a small risk of fracture if excessive bone is resected during resection of the physeal bar. Other possible complications include wound infection (between 1% and 3%), problems with wound closure, and delayed union. Several peer-reviewed studies have reported on the outcome of physeal bar resections, including complications. The results of these studies have been summarized in Table 6.

FUTURE DIRECTIONS

Over the last two decades, we have witnessed major advances in defining the dynamic interactions of the cellular and molecular pathways and physical forces that are responsible for the development and growth of long bones. Together with the recent advances in diagnostic imaging techniques such as MRI and improved ultrasound resolution, new biological treatment strategies and tools are being proposed, which may improve the success of the index procedure in the treatment of physeal growth arrest.

Recent emphasis has been focused on identifying and elucidating the key cellular mediators and growth factors involved in chondrogenesis and osteogenesis. Theoretically, the local application of chondrogenic growth factors, if placed in the correct biological environment, may be able to restore growth activity to the damaged physis and/or augment the growth potential of the surrounding ''healthy'' physeal chondrocytes (41). The main obstacle for use of specific growth factors is the delivery of these short-lived cytokines to the desired anatomic site (42). In 2002, Lee et al. (41) successfully demonstrated the expression of a functional IGF-1 protein after the direct transfer of *IGF-1* gene (using direct adenovirus-mediated gene transfer) into autogenous tibialis anterior muscle interposition grafts placed into a proximal tibial physeal defect in rabbits. The IGF-1 protein that was expressed in the interpositional muscle graft had a significant positive/corrective effect on the angular deformity caused by the physeal destruction. Johnstone et al. reported the successful gene transfer and expression of a functional bone morphogenetic protein-1 protein that was capable of promoting the outgrowth of the physeal cartilage adjacent to the site of the physeal bar (43).

Advances in experimental cell biology have also resulted in the successful transfer of autogenous iliac crest physeal chondrocytes (44,45); cultured chondrocytes (44,46); and mesenchymal stem cells (47) into physeal defects in the proximal tibia of rabbits. All of these

Table 6 Review of Literature (Physeal Bar Resections)

Authors	Journal, Year	Title	Number of patients	Results	Conclusions
Birch (40)	Instr Course Lect, 1992	Surgical technique of physeal bar resection	36	36 physeal bar resections 12 successful 20 failures 4 indeterminate	
Peterson (10)	J Ped Orthop, 1984	Partial growth plate arrest and its treatment	98	14 year follow-up Average growth was 84% of the contralateral side 13% of patients had only the index procedure 87% had a second procedure for LLD and angular deformity Recurrence of physeal bar was 18% Infection rate was 3%	
Williamson and Staheli (22)	J Pediatr Orthop, 1990	Partial physeal growth arrest: treatment by bridge resection and fat interposition	28	22 resections followed for 2 yr 11 had excellent results 5 had good results 2 had fair results 4 had poor results Overall mean growth was 83% with 98% in the excellent group and 96% in the good group	Physeal bar resection is an effective method of treatment Poor results occurred in patients with large bridges (40–65% of the physeal area) Fat is a good interposition material Correct any significant deformity at the time of the physeal bar resection if the area of growth arrest is >25% of the physis
Hasler and Foster (29)	Clin Orthop Relat Res, 2002	Secondary tethers after physeal bar resection	22	24 physeal bar resections with fat interposition graft 14 patients had fair or poor results 5 patients had premature arrest of the affected physis with a postoperative growth period <1 yr 8 patients had bar recurrence 4 patients had bridge recurrence 3 had graft dislocation associated with bridge recurrence	MRI is useful to explain failures of the index procedure

studies were able to demonstrate the successful recovery of growth as witnessed by the successful correction of angular deformity and LLD.

More recently, Yoshida et al. (48) reported that *Runx2* and *Runx3* were essential for the differentiation of mesenchymal stem cells into chondrocytes. *Runx2* also regulates limb growth by organizing chondrocyte maturation and proliferation through the induction of Indian Hedgehog expression. Ultimately, the future biological treatment strategies may see a blending of the research developments described above. One possibility may see adenoviral-*Runx2* and *Runx3* (or other growth factors) vectors transfected into cultured mesenchymal stem cells to augment their ability stimulate growth at the site of the injured physis.

CONCLUSIONS

Despite the experimental advances being made in developing new biological treatment strategies to address physeal growth arrests, surgical excision and placement of an inert interpositional graft remains the gold standard. We continue to rely on the growth potential of the surrounding ''healthy'' chondrocytes to respond appropriately to correct angular deformity and LLD. It is not uncommon for subsequent surgical procedures to be required to correct an angular deformity or an LLD resulting from a growth plate injury.

EDITOR'S ADDENDUM

Treatment options for growth arrest include physeal bar resection, physeal distraction, growth plate closure, contralateral epiphysiodesis, and limb lengthening. The young child with a small physeal bar and a lot of growth remaining is the best candidate for physeal bar resection.

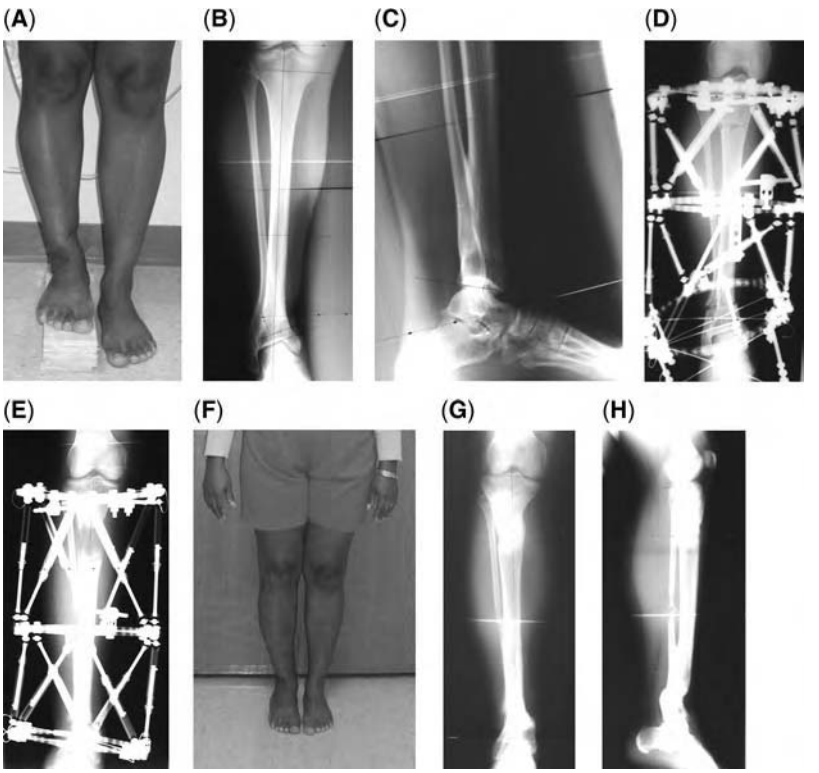

Figure 4 Twenty-five-year old woman who sustained a distal tibia growth plate injury at a young age. She presented with a 6 cm LLD (**A**) and ankle deformity (varus procurvatum oblique plane deformity). (**B** and **C**) A double level osteotomy was performed. (**D**) The deformity was corrected gradually at the distal osteotomy and lengthening was performed at the proximal tibia. (**E**) At 12 months, she had equal leg lengths and correction of her distal tibia deformity. (**F** and **H**) *Source*: Courtesy of S. Robert Rozbruch, MD.

Even if the physeal bar is large, an attempt at resection is still appropriate even if the success is less predictable. If LLD and/or angular deformity still result, these can be treated in the older child with osteotomy for lengthening and deformity correction. The older child with a large physeal bar and little growth remaining is a poor candidate for physeal bar resection. The older child with a small bar may be a good candidate for physeal distraction. We do not have experience with this technique. These children often have LLD and angular deformity. One can predict the LLD at maturity if the injured growth plate were to contribute no additional growth. We approach these cases by closing the injured growth plate and performing an osteotomy for gradual correction of deformity and lengthening. LLD can be equalized by temporarily overlengthening the injured limb or performing a contralateral epiphysiodesis. If a large amount of lengthening is needed, then a bifocal approach can used—one level for deformity correction and one level for lengthening (Fig. 4).

ACKNOWLEDGMENTS

The authors would like to thank Aaron Littleton for his help in preparing this manuscript.

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33 Evaluation of Leg-Length Discrepancy, Epiphysiodesis, and Other Treatment Options

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INTRODUCTION

Leg-Length Discrepancy

Leg-length discrepancy (LLD) is common; 23% of the general population has a LLD of 1 cm or more (1). The prevalence of LLD requiring a corrective device, such as a lift, is approximately 1 in 1000 (2). This chapter outlines the etiology, functional effects, assessment, and treatment of LLD by epiphysiodesis and other treatment options.

The functional effects of LLD on gait have been studied by several authors. The discrepancy that resulted in gait asymmetry exceeding normal was 2.0 cm (range 1.1–3.2 cm) and percentage of limb-length difference was 3.7% (2.2–5.1) (3). Minor leg-length inequalities (<2 cm) do not seem to alter the kinematics or kinetics of gait. Larger inequalities are associated with greater mechanical work (4), and equalizing limb length improves the symmetry of gait (5).

The effect of gait asymmetry on back pain, scoliosis, and knee and hip arthrosis is less well defined. There does not appear to be an increase in the incidence of back pain in patients with small leg-length inequalities (6). Despite this report, clinical experience has shown that there are adult individuals who have gait disturbance, feeling of asymmetry, and hip and back pain with LLD of less that 2 cm.

It has been reported that the incidence of scoliosis is higher in patients with leg-length inequality (7), although one would expect that if the leg-length inequality caused the scoliosis, the scoliosis would compensate for the inequality (with the convexity of the curve toward the short leg). However, in one-third of the cases the opposite is true (8). If a patient with inequality stands with both knees extended, he or she will have a lumbar curve, but often this is only a flexible compensation for the leg-length inequality. Examining the patient with a lift or while sitting will eliminate the curve.

The effects of leg-length inequality on the lower limbs are largely speculative. Tjernstrom and Rehnberg (9) reported few lower extremity complaints in patients with a leg-length inequality. The longer limb will place the hip in slight adduction, about 4° to 5° for a 2-cm leg-length inequality, but only with equal weight on each limb and symmetric joint positions. There is no evidence that leg-length inequality leads to early hip or knee arthrosis (10).

Angular Deformities

The identification and assessment of angular deformities is routinely performed by the orthopedic surgeon. The language is becoming more uniform, allowing for a common terminology for communication and research (11). However, it is still not clear what degree of deformity could result in physical impairment. In tibial malunions, there seems to be a trend toward increased osteoarthritis and subtalar stiffness with varus of greater than 6° (12). Other authors felt that greater than 5° of malalignment about the knee in the coronal plane leads to a significant increase in the progression of osteoarthrosis depending on the underlying disease conditions and activity level (13).

The mechanical axis deviation is affected by several factors including magnitude of the angular and translational deformity as well as the location of the deformity within the bone (11). Thus an analysis with regard to only angular deformity may be incomplete.

CLINICAL EVALUATION

Treatment objectives need to be carefully considered. The decision to treat depends not only on the limb deformity, but also on the physical impairment, which may or may not be linked. Some patients may actually benefit from a small leg-length inequality, such as children with hemiplegic cerebral palsy, polio, other neuromuscular disorders, and in patients with knee or ankle fusion, who can use the leg-length inequality to aid in clearance in the swing phase of gait (14). In children, any expected changes with growth need to be predicted. Usually, the goals are to obtain leg-length equality within 1 cm, alignment in the sagittal and coronal planes within 5° of normal, and symmetrical rotational alignment.

There are other less common indications for limb equalization techniques. Prosthetic fitting can be improved with limb equalization techniques. Shortening, usually by epiphysiodesis, can be performed for patients who underwent an amputation if the residual limb is too long to accommodate a prosthetic foot. Usually, a minimum of 6 cm of leg-length inequality (at maturity) is needed to accommodate ankle/foot or knee prosthesis, and 10 to 12 cm is ideal. For standard prosthetic fitting, a residual limb length of 12 to 15 cm in the femur or tibia is necessary (15).

Numerous radiographic and clinical measurements have been used to assess leg-length inequality. The Galeazzi test, typically used to assess hip dislocation, can also be used as a quick method to assess any disorder that results in significant leg-length inequality. The examination is performed with the patient supine and the hips and knees flexed. The test is positive if the height of the knees is asymmetrical (Fig. 1). It is also helpful in assessing whether the leg-length discrepancy is primarily from the femur or the tibia and in assessing leg-length in someone with knee or hip flexion contractures.

Clinical assessment should include determination of a level of the pelvis with the patient standing. Care must be taken to ensure that the patient is standing with both knees and hips extended and in neutral abduction/adduction. A set of blocks of varying heights can be placed under the shorter leg and the position of the pelvis examined to estimate the amount of leg-length inequality. This is a more accurate method of determining leg-length inequality than using a tape measure, but is still not as accurate as radiographic measurement (16).

Various radiographic measurements have been used to determine leg-length inequality. The teleroentgenogram is a single supine radiograph taken with a radiolucent ruler. This is subject to magnification error of 5% to 10% at the outer border of the film and has the disadvantage of missing deformity that is related to weight bearing such as joint line convergence related to ligamentous laxity and/or loss of cartilage height (10). A 51 in. erect leg bipedal anteroposterior radiograph with blocks under the short leg to level the pelvis allows direct measurement of length and deformity. Orthoradiography incorporates three separate exposures (of the hip, knee, and ankle) in an effort to avoid magnification errors. Scanography uses a similar technique, but exposure size is reduced and all three exposures are on one film

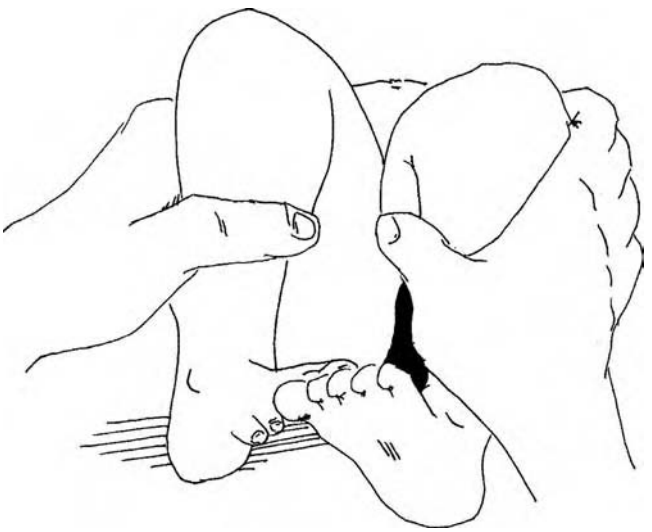

Figure 1 The Galeazzi test.

cassette (16). Both orthoradiography and scanography are subject to movement errors, and angular deformities cannot be assessed. All these techniques are inaccurate if the patient has knee or hip flexion contractures, or if they are simply flexing the knee or hip asymmetrically at the time of exposure. Lateral radiographs, or separate (prone) radiographs of the femur and tibia with a ruler, can be obtained to assess leg-length in patients with knee flexion contractures.

The use of computed tomography scan to assess limb length has increased. It uses less radiation and is more accurate than conventional radiographic techniques in patients with knee or hip flexion contractures (17).

Once the current leg-length inequality has been measured in children with open growth plates, a prediction of the ultimate leg-length inequality at skeletal maturity will be needed to determine treatment. Several methods are typically used to do this: the arithmetic method, the growth remaining curve, the Moseley straight-line graph (18), and the multiplier method (19).

The simplest is the arithmetic method (20). This method assumes growth of the distal femur to be 1 cm per year, the growth of the proximal tibia to be 0.6 cm per year, and that boys reach skeletal maturity at 16 years of age and girls at 14.

The growth-remaining graphs relate chronological age to limb length to determine a child's growth percentile. Using this, the remaining growth of the tibia or femur can be determined graphically.

In an effort to combine this information into one graph, Moseley (18) incorporated the same data into one straight-line graph. The advantage of this technique is that several measurements can be plotted on one graph. The Moseley straight-line graph relies on determination of the bone age as estimated from a left wrist film. When these three techniques were evaluated, there was little significant difference between them (21).

The multiplier method simply takes the current limb-length inequality and multiplies it by a constant listed in a table by chronological age for congenital LLD. A different formula is used for an acquired LLD. Timing of the epiphysiodesis can then be estimated by use of an arithmetic formula to determine limb inequality at maturity (19). It was shown to be as accurate as other methods for determining limb length at maturity and can accurately estimate the timing for epiphysiodesis.

There are several sources of error in determining leg-length inequality at skeletal maturity. Error can occur from radiographic technique, patient position, determination of skeletal maturity, and measurement error. Additionally, growth inhibition may not be constant with age and can change with time, leading to error in the prediction of LLD at maturity. In order to minimize error, it is better to use at least two prediction methods with radiographs taken at two different visits.

CLASSIFICATION

Limb-length inequality is typically classified either by etiology or degree of inequality. There are numerous causes of limb-length inequality. In general, they can be divided into two broad categories: congenital and acquired. Congenital limb-length inequality can be from a limb hypoplasia syndrome such as fibular hemimelia, from a hemihypertrophy syndrome such as Klippel-Trenaunay-Weber syndrome, or from a skeletal dysplasia. Acquired causes include anything that injures or slows the growth of the physis such as a bony bar due to trauma or infection, shortening from a femur fracture with comminution or overriding bone fragments, and any systemic condition that results in asymmetric innervation or vascularization.

Limb-length inequality is also classified by the magnitude of inequality at the time of skeletal maturity. The decision to shorten versus lengthen a limb will depend on the patient's actual or predicted height and the degree of leg-length inequality.

TREATMENT OPTIONS

Treatment of significant leg-length inequality includes the nonoperative approach of shoe lifts/orthotics and the surgical approaches of epiphysiodesis, shortening, and lengthening. These methods can be used alone or in combinations to achieve equalization of leg-lengths.

General guidelines for treatment of a LLD at maturity are: 0–2 cm LLD, no treatment or use a lift; 2–5 cm, perform a contralateral epiphysiodesis or shortening (if skeletally mature); >5 cm LLD perform a lengthening. For very large LLD (>20 cm) consider amputation or prosthetic options.

Orthotics/Shoe Lifts

Often, leg-length equalization can be accomplished with a shoe lift. Usually about two-thirds of the leg-length inequality is corrected with the lift. Up to 1 cm can be inserted inside the shoe. Larger leg-length inequalities need to have the heel and sole built up on the outside of the shoe. This needs to be done for every shoe worn and limits the type of shoe that the patient can wear. Leg-length inequalities beyond 5 cm are difficult to treat with a shoe lift. The shoe looks unsightly, and often the patient complains of instability with such a large lift. A foot-in-foot prosthesis can be used for larger leg-length inequalities. This is often done as a temporary measure for very young children with significant leg-length inequalities. The prosthesis is bulky, and a fixed equinus contracture may result.

Epiphysiodesis

Epiphysiodesis is a simple, outpatient procedure that inhibits growth with few complications (22). However, the final leg-length inequality and the degree of growth inhibition need to be predicted and are subject to errors. Additionally, it effectively shortens the longer leg and is a procedure that is usually performed on the normal side, both of which may be unappealing to the patient and family. Epiphysiodesis can be used in conjunction with limb lengthening for a large leg-length inequality, thus avoiding a second or third lengthening. Hemiepiphysiodesis techniques are performed to impart angular correction in the growing child.

Shortening

This option has become less popular among surgeons and patients. However, it still could be indicated in patients who are willing to spare height and who may not be good candidates for lengthening. Shortening techniques can be used after skeletal maturity to achieve leg-length equality. Shortening can be performed in the proximal femur using a blade plate or hip screw, the middiaphysis of the femur using a closed intramedullary technique or in the tibia.

Lengthening

Lengthening is extensively discussed in other chapters throughout this book.

TREATMENT METHODS

Phemister (22) first described his technique for epiphysiodesis. He removed a section of the epiphysis, then rotated it 90° and placed it back creating a bone block. Currently, other techniques have been developed, such as the percutaneous drill epiphysiodesis and epiphysiodesis with staples and screws. The percutaneous drill epiphysiodesis is performed with the aid of image intensifier and has been reported to result in physeal closure in 85% to 100% of patients (23), with few complications. Scott et al. (24) compared the Phemister technique to the percutaneous technique and found the results to be similar. They preferred the percutaneous technique due to the ease of the procedure and decreased morbidity. It is a timed procedure that is indicated in patients with a documented history of congenital LLD, with matching chronological and bone age, with no angular deformities. Staple epiphysiodesis is more invasive. It has the advantage of allowing for growth after staple removal, so it may be better suited for younger patients. New shape memory alloy staples have been used to prevent dislodgement (Fig. 2A) especially in osteoporotic bone. These staples have a temperature-dependent shape, opening when cooled at the time of insertion and closing upon reaching body temperature after insertion. Percutaneous epiphysiodesis using transphyseal screws have been used in an effort to provide a reversible method of inhibiting growth with less morbidity. This technique has been used primarily in the ankle for correction of angular (valgus) deformity but can also be used about the knee for treatment of leg-length inequality or angular deformities (Fig. 2B,C) (14,25). Screw removal can be difficult; therefore, the screw head should not be deeply buried

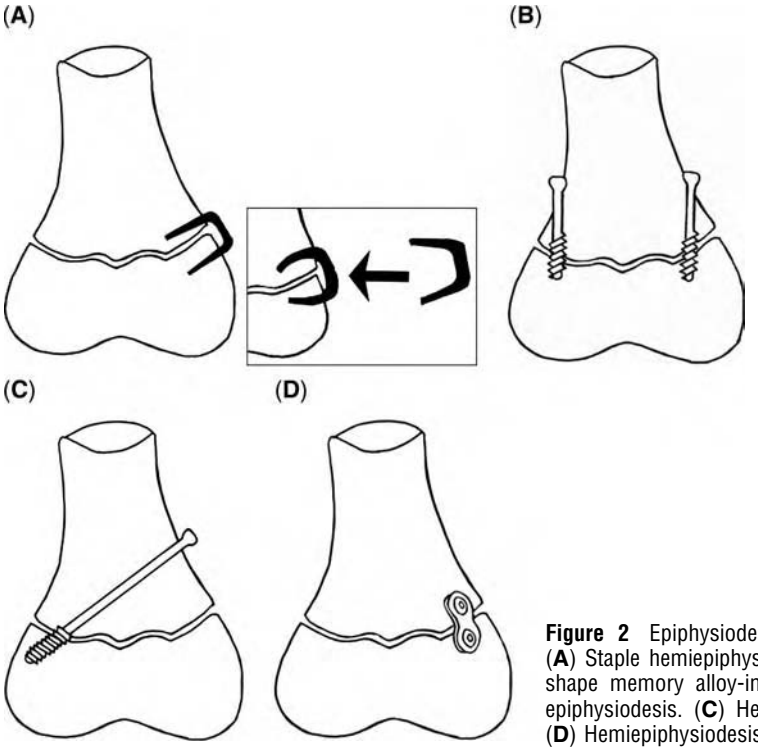

Figure 2 Epiphysiodesis, different types of fixation. (**A**) Staple hemiepiphysiodesis (with a types of fixation shape memory alloy-inset) epiphysiodesis. (**B**) Screw epiphysiodesis. (**C**) Hemiepiphysiodesis with a screw. (**D**) Hemiepiphysiodesis with a plate.

at the time of insertion. A plate has been developed as another method to alter physeal growth (Fig. 2D).

COMPLICATIONS

Error in correction of LLD can occur, especially if there is a large difference in the chronological age and bone age, making prediction difficult. Angular correction is also subject to over- or undercorrection, and careful monitoring is needed in all patients. If a staple or screw hemiepiphysiodesis is performed and the patient is fully corrected prior to skeletal maturity, the hardware can be removed. Some patients may undergo a rebound phenomenon in which some of the deformity recurs. Therefore, slight overcorrection may be indicated prior to hardware removal. We attempt to time our hemiepiphysiodesis so that angular correction coincides with skeletal maturity.

Staples can be prominent or may back out (26). Knee stiffness may occur after surgery. This can usually be prevented with a knee range of motion exercise program after surgery.

FUTURE DIRECTIONS

Other methods of lengthening, such as the use of transplanted cells (cultured stem cells or chondrocytes) or manipulation of the rate of growth via growth factors, may hold promise for less-invasive methods of limb-length equalization and angular correction (27).

REVIEW OF LITERATURE

Authors	Journal, Year	Title	Number Patients	Results	Conclusions
Aguilar, Paley D, Paley J, et al.	J Pediatr Orthop, 2005	Clinical validation of the multiplier method for predicting limb length at maturity	60	Mean error of predictions for LLI was 2.5 cm (using chronological age)	Results are similar to Moseley method

(*Continued*)

Authors	Journal, Year	Title	Number Patients	Results	Conclusions
Bowen, Leahey, Zhang, MacEwen	Clin Orthop, 1985	Partial epiphysiodesis at the knee to correct angular deformity	7 patients (13 extremities)	Average correction was 5°/yr for the tibial and 7°/yr for the femur	Theory for hemiepiphysiodesis prediction is accurate
Moseley CF	J Bone Joint Surg, 1977	A straight-line graph for leg-length discrepancies	Case examples	A graphic method is presented to record and interpret leg-length discrepancy	More accurate than growth-remaining method, particularly in cases of growth inhibition
Scott, Urquhart, Cain	Orthope-dics 1996	Percutaneous versus modified Phemister epiphysiodesis of the lower extremity	24	No difference in results, continued growth of the physis occurred in 12% to 15%, and no angular deformities	The percutaneous method is preferred due to ease of surgical procedure
Terry, Winell, Green, et al.	J Pediatr Orthop, 2005	Measurement variance in limb-length discrepancy: clinical and radiographic assessment of interobserver variability	16	Variance ranged from 0.13 to 1.1 cm for intra and inter observer measurements of LLI	Direct measurements of the slit scanogram in the most accurate method

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34 Congenital Pseudarthrosis of the Tibia

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INTRODUCTION

Congenital pseudarthrosis of the tibia (CPT) refers to nonunion of a tibial fracture that develops spontaneously or after trivial trauma in a dysplastic bone segment of the tibial diaphysis. The pseudarthrosis usually develops during the first two years of life; however, there are reports of cases in which fractures developed before birth and reports of late-onset pseudarthrosis. The etiological nature of this condition is unclear; however, there is a strong association between CPT and neurofibromatosis Type I (NF-I). CPT develops in about 5.7% of patients with NF-I. On the other hand, 40% of patients with CPT were found to have NF-I. Electron microscopy and histopathological studies showed that the main pathology of CPT is hyperplasia of fibroblasts with the formation of dense fibrous tissue. This invasive fibromatosis is located in the periosteum and between broken bone ends and surrounds the tibia causing compression, osteolysis, and persistence of pseudarthrosis (1–3).

Classification of CPT has been attempted based on time of onset, mobility, and radiological appearance. It is well known that CPT may be preceded by cystic and hourglass constriction of the bone or by fatigue-like fracture of the tibia. Difficulties have arisen because the condition includes several different clinical and pathological entities, each having a different history and prognosis. The use of a classification system that is based on the radiological appearance of the tibia can become confusing in that the pattern can change from one variety to another during treatment. Thus, what may first appear as a cystic lesion with an apparently good prognosis may, after one or two attempts at grafting, appear as a dysplastic lesion with a less-favorable prognosis (4,5).

To date, there is still no clear, universally accepted classification system based on both radiological and clinical appearance. This indicates that none of the proposed classification systems are based on a totally accurate concept of the cause, natural history, and treatment result.

CPT remains one of the most perplexing and challenging orthopedic problems. This is because of difficulty of obtaining union and, in cases that achieve union, difficulty of maintaining union. Frequently, the end result of this scenario is a frustrated child who has undergone several failed operations and has a limb that is short, deformed, and almost functionless. This experience led some authors to advise amputation if the third operation failed to achieve bone union and a functioning limb (5,6).

The difficulty in treating this condition occurs because of two factors. The first is biologic: poor healing ability of the dysplastic segment of bone. The second factor is mechanical: technical difficulty to fix small and osteopenic bone fragments in children without damaging the distal tibial physis or ankle joint.

A multitude of treatment protocols have been described and have varying degrees of success (7). Most of these treatments focused on stimulating the healing process by using different bone grafting techniques. The graft materials most commonly used included osteoperiosteal graft, massive onlay graft, autogenous iliac crest bone graft (ICBG), and vascularized bone graft (1,8–10). The fixation methods also varied widely between cortical fixation using bone or metal plates, intramedullary (IM) rods of different types, external skeletal fixation, and lastly external skeletal fixation augmented by IM rod (1,11–15).

Whatever the treatment method used, the natural history of CPT is that the affected bone segment remains atrophic and poorly consolidated with a tendency to refracture until skeletal maturity. At skeletal, maturity the bone quality, for unclear reasons, becomes almost normal, hypertrophy of the tibia occurs, and then the bone ceases to refracture (1,5).

CPT continues to pose a difficult therapeutic challenge in both achievement of bone healing and maintenance of this healing.

Paley (15) theorized that the pathology of congenital pseudarthrosis is not bony but rather periosteal in origin. Anatomically and physiologically, this theory is supported by the following observations: thickening with hamartomatous transformation of the periosteum, appearance of strangulation of the bone with atrophic changes followed by avascular changes (the anterolateral bowing of the bone is due to the dominant muscular forces on the leg from the eccentrically located posterolateral muscle masses), and failure of remodeling of pin holes leading to stress fracture (remodeling is a periosteal function). This theory was considered by Codavilla a century ago. In 1975, Cambras from Cuba used the periosteum and bone from the mother to treat CPT. Cambras considered the periosteum to be the source of the pathology (personal communication). Most recently, pathologic analysis by Hermanns-Sachweb et al. confirmed that pathologic periosteum is the cause of CPT (16). Their most striking finding was that neural cells formed a tight sheath around the periosteal arteries reminiscent of the way Schwann cells in the periosteum cause narrowing or obliteration of the periosteal vessels. The periosteum undergoes hypoxemic degeneration resulting in the formation of a thick fibrous cuff. This leads to impaired oxygen and nutrient supply to the subperiosteal bone and the atrophic bone changes that are observed. As will be described below, Paley has used periosteal grafts successfully in the treatment of CPT. Paley's method has been independently corroborated by Grill from Austria and Kocaoğlu from Turkey.

CLINICAL EVALUATION

The management of a case of CPT starts with general clinical evaluation for presence of NF-I and its consequences that might affect the quality of the patient's life and may temper the enthusiasm to achieve bone healing. In some cases, the treating surgeon would resort to other treatment methods such as amputation and prosthetic replacement (2,5).

The patient's history of the onset of fracture and previous surgical treatment has important prognostic and therapeutic implications.

The affected limb of the patient is examined for mobility or stiffness of the pseudarthrosis, presence of deformity, nearby joint stiffness, previous operative scars, quality of soft tissues, and neurovascular abnormalities.

Radiological examination of the affected leg in the anteroposterior and lateral views is used to determine the level of CPT, quality of bone, presence of retained hardware, and condition of nearby joints. Full-length standing radiographs of both lower limbs are obtained to determine leg-length discrepancy, mechanical axis deviation, and presence of deformities (17).

CLASSIFICATION

A comprehensive classification system should serve therapeutic guidelines, provide prognostic indicators for the condition under study, and consider all the factors that influence treatment options. Meanwhile, the classification should be easy to recall and apply. Recognizing these facts, we developed our classification based on the clinical observations and results of the treatment of 17 patients with CPT and our review of literature.

Our classification (Table 1) considers the patient's condition at the time of treatment and is based on three parameters: (i) Did the patient have any surgical treatment for the pseudarthrosis or not? Previously failed surgical treatment negatively affects the prognosis due to less favorable soft tissue condition and bone quality and possibly retained hardware. (ii) Is the pseudarthrosis of the mobile or stiff type? Mobile pseudarthrosis means presence of intervening abundant fibrous tissue, which requires operative excision and implies a poorer prognosis than the stiff type. (iii) Plain X-ray examination of the affected leg for the radiological typing of CPT: atrophic with narrow bone ends or hypertrophic with wide bone ends. Based on these findings, three types of CPT were recognized (Fig. 1).

Table 1 El-Rosasy-Paley Classification for CPT

Type I	Atrophic bone ends (based on radiographic examination) Mobile pseudarthrosis No previous surgical intervention
Type II	Atrophic bone ends (based on radiographic examination) Mobile pseudarthrosis Previous surgical intervention with or without retained hardware
Type III	Broad bone ends (based on radiographic examination) Stiff pseudarthrosis With or without previous surgical intervention

Patients with Type I CPT usually present early in life, before two years of age, and have a good prognosis with treatment. Type II pseudarthrosis presents usually in older children after experiencing failed surgical treatment, refracture, or an osteotomy to correct a bowed tibia. Type II cases have a relatively poorer prognosis. Type III cases are usually of the late onset type, which develops as a stress fracture of a congenital dysplastic tibia or in a healed tibia after previous bone grafting and has the best prognosis.

TREATMENT OPTIONS

Whatever the method used to treat CPT, certain requirements should be fulfilled to ensure good results: (i) Complete excision of the soft tissue fibromatosis at the site of pseudarthrosis with minimal bone resection. (ii) Stimulation of bone healing by insertion of autogenous ICBG. (iii) Proper fixation of the bone fragments until bone healing is observed radiologically. (iv) Maintenance of bone healing until complete bone consolidation and hypertrophy at the age of skeletal maturity (1,9,12–14,18) (Table 2).

Our preferred method of fixation is to use Ilizarov external fixator augmented by IM rod. The Ilizarov external fixator facilitates fixation of small bone fragments and simultaneous compression of pseudarthrosis, gradual correction of deformities, leg lengthening, and bone transport. The use of an IM rod augments the fixation of osteoporotic bone and prevents axial deviation of bone fragments during compression-distraction. The IM rod remains in the bone indefinitely after fixator removal to act as internal splint and to protect the tibia against refractures.

Table 2 Review of Literature

Reference Number	Title	Number of Patients	Results	Conclusions
(15)	Treatment of CPT using the Ilizarov Technique	15 (16 cases)	94% united, 31% ReFx, No IM rod	Correct angular deformities, maximize cross section of bone
(11)	CPT associated with neurofibromatosis-1: treatment with Ilizarov device	21	66.7% united, 23.5% ReFx, 33.3% nonunion, 2 IM rods	CPT resection, + Ilizarov device, + postoperative compression
(3)	CPT: Part I: European pediatric orthopedic society multicenter study of congenital pseudarthrosis	340	Ilizarov Tx, + ICBG, 75% healing; vascularized fibula, 61% healing	Ilizarov Tx and ICBG or vascularized fibula, + prophylactic IM rod
(14)	CPT: results of technical variations in the Charnley-Williams procedure	23	87% good, 13% unsatisfactory	Pseudarthrosis, excision + autogenous graft, + fibular surgery, + dual IM rod
(12)	Use of an IM rod for the treatment of CPT. A long-term follow-up study	21	76% good, 57% ReFx, 23.8% amputations	Pseudarthrosis, excision + autogenous graft, + IM rod

Abbreviations: CPT, congenital pseudarthrosis of the tibia; ReFx, refracture; IM, intramedullary; ICBG, iliac crest bone graft; Tx, treatment.

Table 3 Clinical Management

Classification Subgroup[a]	Clinical Evaluation	Surgical Approach	Pearls	Complications/Pitfalls
Type I	Signs of NF-1, fibular integrity	Fibromatosis resection, ICBG, dual IM rod, Ilizarov device	Maximize bone contact, Dual rodding	Tenuous union
Type II	Soft tissue condition, bone loss, retained hardware	Acute bone contact, ICBG, IM rod, Ilizarov, ± Limb lengthening	Adequate graft, postoperative compression	Refracture
Type III	Deformity analysis, fibular integrity	Preconstructed frame, distraction-compression	No residual deformity, delayed IM rod	Peroneal nerve palsy

[a]Pseudarthrosis classification followed the system described by El-Rosasy et al. (13). Abbreviations: NF-1, neurofibromatosis-1; ICBG, iliac crest bone graft; IM rod, intramedullary rod.

SURGICAL TECHNIQUES

Based on the classification of CPT, treatment strategies are described for each type of pseudarthrosis (Table 3).

Treatment of Type I CPT

Surgery is performed soon after the tibia fractures to avoid complications that result from delayed treatment (e.g., deformity, joint contracture, and limb shortening) (Figs. 2 and 3). The pseudarthrosis is approached through a standard anterior approach to the tibia. The fibrous tissue hamartoma is carefully dissected from surrounding muscles and excised subperiosteally from the bone ends back to the limits of apparently healthy periosteum. Minimal trimming of the tapered bone ends is done to allow identification of the medullary cavity (aggressive bone resection should be avoided to minimize bone loss). The proximal bone end is drilled in the coronal plane using a thin Kirschner wire (K.W.) starting from distal to proximal for a distance of 2 to 3 cm. Using a thin osteotome, the tip of the proximal end is carefully split along the drill holes. A small drill bit is used to open the medullary canal of the distal fragment. The thickest possible K.W. is then inserted into the medullary canal of the distal fragment. If the ankle joint is to be transfixed, the foot should be held by an assistant in the neutral position and its position is monitored by image intensifier control. One of the authors (Dror Paley) has modified this method so that the wire is inserted disto-proximally from the medial malleolus to spare the ankle joint. The tip of the distal fragment is carefully wedged into the split end of the proximal one. This bone invagination increases bone contact, triples the cross sectional diameter of the tibia, stabilizes the pseudarthrosis, and realigns the tibia. The K.W. is advanced through the medullary canal of the proximal segment until just

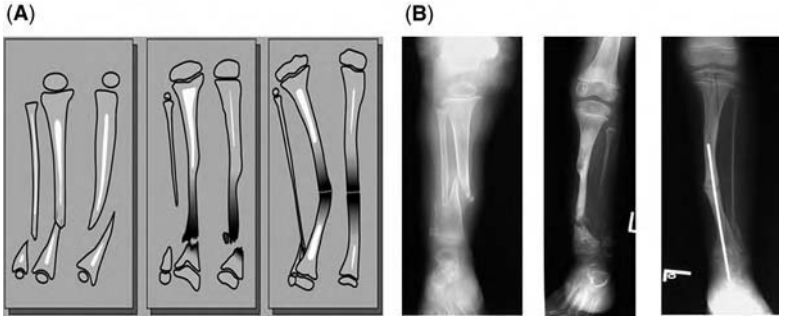

Figure 1 (**A**) Schematic representation and (**B**) radiographic examples of the El-Rosasy-Paley classification of congenital pseudarthrosis of the tibia. Type I: atrophic, mobile pseudarthrosis, and virgin type. Type II: atrophic, mobile pseudarthrosis, and previous surgical treatment. Type III: wide bone ends, stiff pseudarthrosis, and± previous surgical treatment.

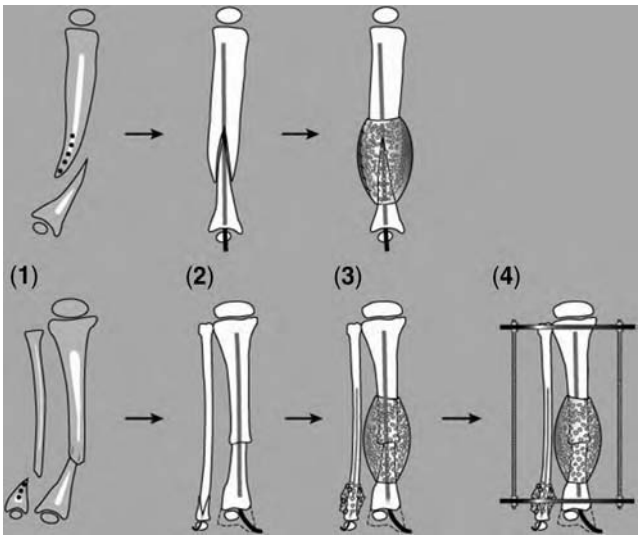

Figure 2 Type I treatment—steps in treatment. (**1**) Longitudinal splitting of the proximal tibial and distal fibular fragments. (**2**) Invagination of bone ends and insertion of intramedullary rods into the fibula and tibia through the medial malleolus to allow free ankle motion. (**3**) Insertion of iliac crest bone graft around the pseudarthrosis. (**4**) Application of the Ilizarov external fixator.

before reaching the proximal tibial physis. The K.W. should span the whole tibia and should be followed under image intensifier to avoid penetration into the knee joint or false passage outside the bone medullary cavity. Autogenous ICBG is harvested and packed all around the pseudarthrosis. Periosteum harvested from the undersurface of the iliacus muscle at the same time and from the same site as the cancellous bone harvest and is wrapped around the pseudarthrosis site. This periosteal graft is meshed using a skin graft mesher to allow it to expand. The periosteal graft should be placed with its cambium layer toward the bone. The wound is closed in layers.

The fibular pseudarthrosis is dealt with in a way similar to the tibia. The fibrous tissue at the pseudarthrosis is excised. A suitably sized K.W. is inserted retrograde into the distal segment. The narrow tip of the proximal bone is wedged into the distal fragment, which is short and broad. The K.W. is advanced into the medullary canal of the fibula till reaching the upper physis. Autogenous ICBG is packed around the fibular pseudarthrosis, and the wound is closed in layers.

A two-ring pediatric Ilizarov external fixator is applied with a foot piece attached to control the foot position. No lengthening osteotomy is done for these patients due to minimal shortening (1–2 cm), which will be compensated for during growth. The pseudarthrosis is compressed for a few millimeter during the following postoperative days. The fixator remains on until complete healing is noted radiographically (average duration, four to five months). The IM rod is left as internal splint; however, it should be followed up regularly so that when

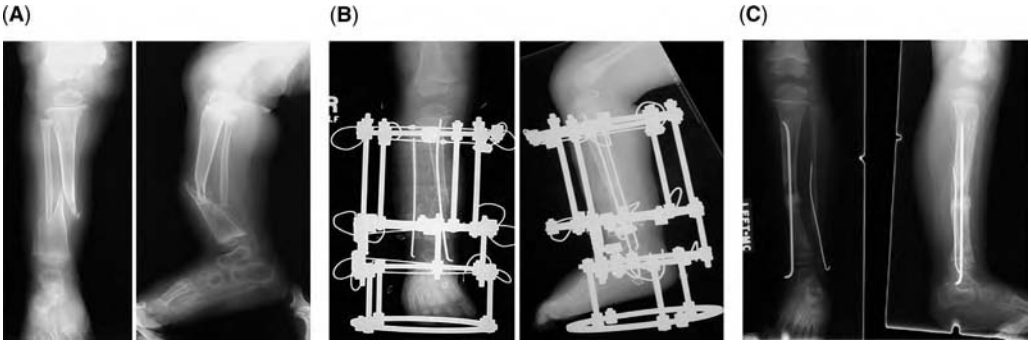

Figure 3 (**A**–**C**) Type I pseudarthrosis treatment. (**A**) Anteroposterior and lateral view radiographs of Type I congenital pseudarthrosis of the tibia in a six-month-old child. (**B**) Immediate postoperative radiographs showing dual rodding of tibia and fibula, bone grafting, and Ilizarov frame application. (**C**) Follow-up radiographs, two years postoperative showing consolidation of the pseudarthrosis and distal migration of the rod due to tibial growth.

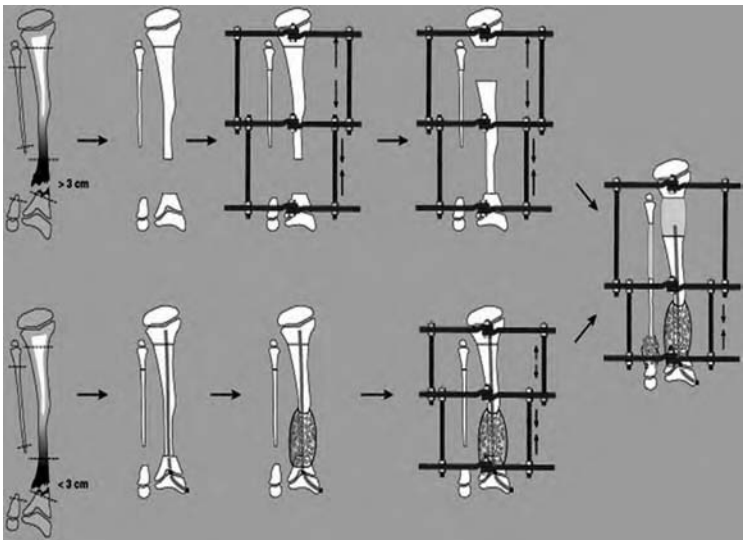

Figure 4 Type II treatment. (*Top row*) Resection of dead bone ends, proximal lengthening osteotomy, bone transport for large bone defect, insertion of intramedullary (IM) rod and iliac crest bone graft (ICBG) at the completion of bone transport; (*bottom row*) resection of dead bone ends, proximal lengthening osteotomy, acute shortening for small bone defect, and insertion of IM rod and ICBG.

it is outgrown by the tibia, it can be replaced by a longer rod to avoid tibial fracture at the tip of a short rod.

Treatment of Type II CPT

The surgical approach to the pseudarthrosis depends on the preoperatively estimated amount of acute shortening of the tibia after debridement of the pseudarthrosis (Figs. 4 and 5). If the resulting bone defect is less than 3 cm, then a transversely placed skin incision is used that is centered on the pseudarthrosis. If the estimated acute shortening is more than 3 cm, the preferred approach utilizes a Z-shaped skin incision. After acute tibial shortening, the limbs of the Z-incision can be transposed to increase the width of the incision at the expense of the length. This incision also allows exposure of the tibia and fibula from the same skin incision and good skin closure at the end of operation (19). All retained hardware is extracted. Fibrous tissue at the site of CPT is excised to leave a healthy soft tissue bed. Debridement, rather than excision, of dead bone is performed till bleeding end is reached. An IM rod of suitable size is inserted. The choice of the type of the IM rod depends on the size of the tibia.

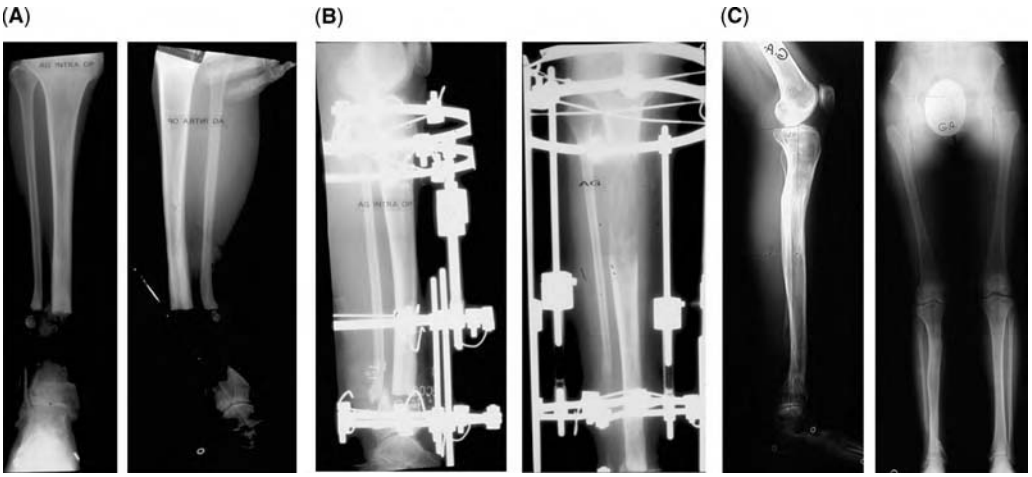

Figure 5 (**A**–**C**) Type II pseudarthrosis treatment. (**A**) Anteroposterior (AP) and lateral view radiographs of Type II pseudarthrosis in a 14-year-old girl after resection of bone ends. (**B**) Acute shortening and proximal osteotomy for lengthening of 10 cm. (**C**) AP and lateral views 11.5 years postoperatively show consolidation of CPT, good alignment, and equal leg lengths.

Immediate bone contact is obtained by acutely shortening the leg. This depends on the size of the bone defect after debridement and distal blood flow after acute leg shortening, which is monitored by Doppler ultrasound throughout the operation and postoperatively. If the bone defect is so great that it would kink the blood vessels and cause lack of blood flow to the foot, then partial acute shortening is done within the tolerance of the circulation, followed by gradual shortening in the following postoperative days at a rate of 2 to 3 mm/day until adequate bone contact is achieved.

Autogenous ICBG is harvested and packed around the bone ends. A three-ring Ilizarov external fixator is then applied with the limb aligned. Proximal metaphyseal osteotomy for limb lengthening is done percutaneously.

Postoperative compression of the pseudarthrosis is performed at a rate of one millimeter, twice weekly till complete bone contact and early bone healing is achieved. After consolidation of the pseudarthrosis is confirmed radiologically, the fixator is removed. The IM rod is left in place and eventually replaced by a longer one when the tibia outgrows the rod.

Treatment of Type III CPT

This type of CPT is amenable to closed distraction-compression without open surgery (Figs. 6 and 7). Preoperative analysis of the deformity is necessary so that a preconstructed Ilizarov external fixator is prepared the day before operation and sterilized. If the fibula is intact, then a percutaneous fibular osteotomy is done before fixator application; the pseudarthrosis itself is not disturbed. The preconstructed fixator is applied under image intensifier control for accurate hinge placement. The hinge is placed at the level of the apex of deformity on the convex side, with the hinge axis perpendicular to the plane of maximum deformity. Gradual deformity correction is started postoperatively at a rate calculated according to the method described by Herzenberg and Waanders (20). After complete deformity correction, the hinges and the distractor are replaced by straight rods and axial compression of the CPT is done until bone consolidation is achieved. The fixator is removed and a well-fitting above-knee cast is applied. As a secondary procedure, an IM rod is inserted percutaneously to protect against refractures.

After Treatment

The limb is fitted into knee-ankle-foot orthosis with a drop lock at the knee and solid ankle and gradually changed into patellar tendon-bearing orthosis near skeletal maturity when the tibia is well developed and thick enough to resist refracture. The patient is seen at three months, six months, and then yearly. Plain X rays are obtained to determine bone quality, evidence of refracture, gradually developing deformity, and length of the IM rod relative to the length of the tibia. Regular follow-up of the patient is necessary until skeletal maturity when the tibia attains normal size and good bone quality (12,13).

Results of Treatment

In our series of 17 cases of CPT, 36 operations were performed (17 procedures for the original pseudarthrosis and 19 for refractures). It was possible to obtain union of the pseudarthrosis with

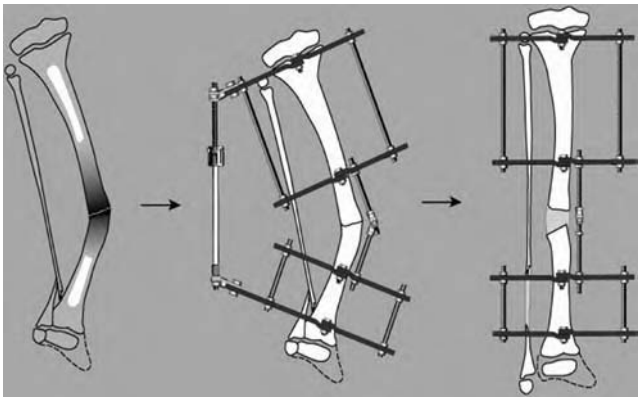

Figure 6 Type III treatment. Application of a preconstructed Ilizarov external fixator (the hinge is placed at the level of the deformity apex on the convex side with its axis perpendicular to the plane of maximum deformity). Gradual distraction of the pseudarthrosis for deformity correction is followed by axial compression until bone consolidation is achieved.

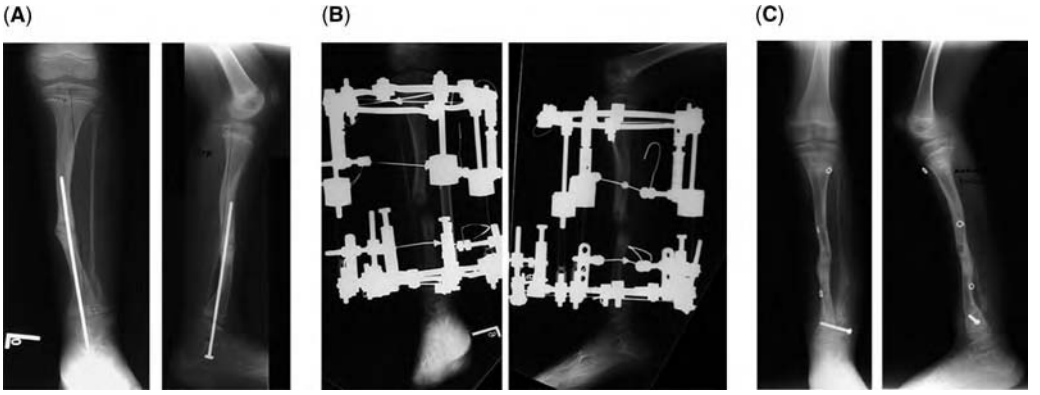

Figure 7 (**A**–**C**) Type III pseudarthrosis treatment. (**A**) Anteroposterior and lateral view radiographs of Type III pseudarthrosis with intramedullary rod previously applied and stress fracture. (**B**) Postoperative radiographs after application of Ilizarov external fixator for distraction of the pseudarthrosis and deformity correction. (**C**) Follow-up radiographs show consolidation of the pseudarthrosis and good alignment.

every treatment. The refracture rate was 68% when only the Ilizarov device was used; however, this rate was reduced to 29% when the IM rod was combined with the Ilizarov device. Patients, who were followed until skeletal maturity, have had their tibiae healed with normal cross sectional area and have experienced no refracture afterwards (Fig. 5). The best results were obtained in Type III and Type I cases; less-favorable results were obtained in patients with Type II, who had experienced several previously failed operations.

COMPLICATIONS

The most dreaded complication in the treatment of CPT is refracture of the tibia after it is healed. Refracture can occur at the site of previous pseudarthrosis, regenerate bone, or pin site hole. Predisposing factors to refracture are residual deformity, tenuous healing of pseudarthrosis with hourglass tibial constriction, pin site hole in the bone, stress riser at the tip of a short IM rod, and premature removal of external fixator before consolidation of regenerate bone (1,6,14,15). Treatment of refracture follows the same protocol as treatment of the original pseudarthrosis.

Other complications, such as peroneal nerve palsy during lengthening and deformity correction, may necessitate nerve decompression at the level of head of the fibula. Valgus deformity of the ankle is corrected by supramalleolar osteotomy and acute deformity correction, Equinus contracture of the ankle is corrected by combined soft tissue release and distraction of the joint; supramalleolar osteotomy is used to correct equinus deformity if the ankle is ankylosed and not amenable to arthrodiastasis. Leg-length discrepancy is corrected by leg lengthening if the discrepancy was of clinical significance; a proximal metaphyseal osteotomy is utilized to equalize leg length during treatment of the CPT or afterwards (13).

FUTURE DIRECTIONS

The introduction of distraction histogenesis, based on the principle of tension stress effect, has revolutionized the treatment of several challenging orthopedic problems including CPT. However, the shortcoming of this technique is that the natural history of CPT is not modified and the affected tibia will remain dysplastic and prone to fractures until skeletal maturity. Realizing that the primary problem in the treatment of CPT is a biological one, where the osteogenic power at the pseudarthrosis site is lacking, future investigations should focus on finding out treatment modalities that locally stimulate osteogenesis and bone repair. Of great promise is the application of local gene therapy in which mesenchymal stem cells are genetically modified to both produce and respond to osteoinductive growth factors with the goal of developing a tissue engineering strategy for new bone formation and fracture healing. Should a successful clinical program of local gene therapy be established, this treatment modality

would modify the natural history of CPT, reduce the tendency to refracture, and may obviate the need for open bone grafting procedure (21).

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35 Congenital Pseudoarthrosis of the Tibia Redefined: Congenital Crural Segmental Dysplasia

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INTRODUCTION

Congenital pseudoarthrosis of the tibia is a rare disorder with an incidence of 1 per 140,000 live births and associated with neurofibromatosis type-1 (NF-1) in 50% of the cases. The first descriptions of this disease were made by Hatzöcher in 1709 and Sir James Paget in 1891 (1–5). The name ''congenital pseudoarthrosis of the tibia'' does not reflect the facts that pseudoarthrosis of the tibia does not occur until the first years of life in 90% of cases (6–8) and appears with a pseudoarthrosis of the fibula in 60% of all cases. In addition, even an isolated fibula pseudoarthrosis could occur in 19% of patients (9).

Our research of the etiology and pathogenesis of this disease has led to a new outlook with respect to definition, classification, and therapy (10). The morphological investigations (light microscopy, transmission electron microscopy, and immunohistochemistry) of the area of tibial pseudarthrosis revealed cells of neurogenic origin surrounding the vessels in the thickened periosteum, comparable to those cells forming the neural myelin sheath. This phenomenon leads to a constriction of the vessels and reduction of the perfusion in periosteum and bone (Fig. 1). It results in a degenerative fibrosis of the periosteum, giving rise to a vicious cycle and decreases the vascular perfusion of the bone. The consequence is fracture of the weakened bone and a subsequent nonunion, which was demonstrated in a remarkable animal experiment by Wright et al. (11). This data correlates with our clinical findings and those of other authors (12) showing that the radical resection of periosteum and scar tissue has a positive effect on the healing process.

Furthermore, we have shown an improved rate of healing with resection of the sclerotic bone. We suggest that this disease has to be redefined as a defect, which affects an entire limb segment including tibia, fibula, and periosteum. We propose that the new term ''congenital crural segmental dysplasia (CCSD)'' is more appropriate because it explains the character and the pathological anatomy of this entity.

With respect to etiology, there is a histological similarity between CCSD and cutaneous neurofibromas suggesting that the gene coding for neurofibromine could be an important factor. The CCSD could represent another localized type of NF-1. The proliferation of the neural cells could be the result of their NF-1 genotype. The neurofibromine protein, in its normal configuration, impairs the guanine nucleotide-binding proteins (so called G-proteins) resulting in a reduced cellular proliferation (13). Mutations of the gene coding for neurofibromine lead to an uncontrolled cellular growth (14).

Interestingly, the activation of somatic mutations of the α-subunit stimulating G-proteins leads to another form of osseous malformation—fibrous dysplasia (15), which also seems to be related to CCSD. Schotland et al. (16) suggested a genetic relationship between fibrous dysplasia and NF supposing that the neurofibromine-G-protein complex could be the missing link between both diseases and in our opinion also to CCSD.

Based upon this information and our clinical experience, we came up with a new classification (Table 1, Fig. 2) and treatment approach.

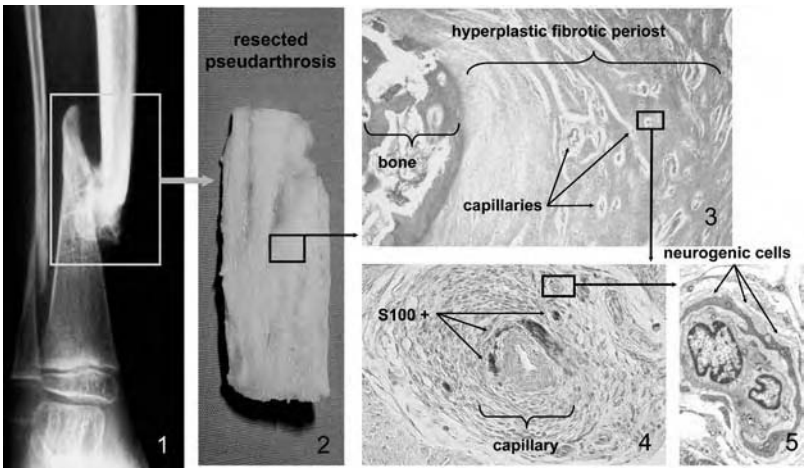

Figure 1 Illustration of the pathological anatomy shown for congenital crural segmental dysplasia-type IVb. *Source*: ©Michael Weber.

CLINICAL EVALUATION

The clinical diagnosis of CCSD is concentrated mainly on the identification of three different clinical forms: (i) postpartum without pseudarthrosis (90%), (ii) postpartum with pseudarthrosis (10%), and (iii) forms 1 and 2, which have had unsuccessful prior surgery.

The diagnosis of CCSD should be considered in children, who in the absence of trauma present with a fracture or nonunion or procurvatum and varus deformity of the lower leg, especially if the deformity is increasing. Further verification of the history is of importance especially in those cases, which have had previous unsuccessful surgery and now have deformities that no longer correspond to the characteristic pattern of CCSD. The diagnosis is also supported by the presence of NF (incidence of 50%).

Our own studies demonstrated that the magnetic resonance imaging of the affected lower leg could detect the thickened periosteum and confirm the diagnosis. It is also useful in planning the extent of surgical resection of the lower leg segment (17). The extent of bony sclerosis, which was considered to be a prognostic factor for the rate of healing by other authors (6,18,19), is also used to plan surgical resection. We propose a new radiological measurement of the extent of sclerosis, independent of different tibial lengths and magnification factor, and a method to determine the extent of resection necessary for a successful healing process (Fig. 3). We measure the extension of sclerosis of the tibial diaphysis in the longitudinal and transverse directions and calculate a ratio to the entire tibial length and width. The length of the diaphysis and the width of the sclerosis are not compared with each other, but rather the percentage of the sclerosis in relation to the diaphyseal length. This percentage is calculated according to the following formulas:

$$\textit{Length extent of sclerosis (\%)} = \textit{length of sclerosis/length of bone} \times 100$$

$$\textit{Width extent of sclerosis (\%)} = \textit{width of sclerosis/width of diaphysis} \times 100$$

The extent of sclerosis of the affected tibia is measured on the preoperative radiograph as shown in Figure 3. The ratio between sclerosis and length of bones or sclerosis and diaphyseal width is calculated. The extent of sclerosis is divided by length of the bone or width of diaphysis resulting in comparable data. A/B and A′/B′ are proportions for the proximal or distal extension of the longitudinal diaphyseal sclerosis. C/D and C′/D′ are the ratio of sclerosis and diaphyseal width proximally and distally of the pseudarthrosis. The ''normal'' ratio between cortical thickness and diaphyseal width is determined in a distance to the pseudarthrosis near the proximal and distal metaphysis (G/F or G′/F′). Afterwards, this normal ratio is compared to the extents of sclerosis in the pseudarthrosis area. Thus, an objective measurement is available, which describes the extent of the sclerosis and, furthermore, allows a comparison between the degree of sclerosis before and after an operative salvage procedure. After the resections of the pseudarthrosis, the same measurements are carried out (Fig. 3B). I/J (proximal) and I′/J′ (distal) correspond to the value of the longitudinal extension of the remaining diaphyseal sclerosis after resection. L/K (proximal) and L′/K′ (distal) correspond to the value for the ratio between the sclerotic thickness and width at the end of the resection.

Table 1 Classification and Score System of Congenital Crural Segmental Dysplasia

Type			Score
I	Indifference stage, procurvatum-varus deformity without progress, no fractures		32
IIa	Prefracture stage, progressive deformity, and no fibular fracture		29
IIb	Prefracture stage, progressive deformity, and fibular fracture		26
IIIa	Prefracture stage, cystic lesion, and no fibular fracture		23
IIIb	Prefracture stage, cystic lesion, and fibular fracture		20
IVa	Tibial pseudarthrosis, sclerosis <30% (20%), and no fibular fracture		17
IVb	Tibial pseudarthrosis, sclerosis <30% (20%), and fibular fracture		14
Va	Tibial pseudarthrosis, sclerosis >30% (20%), and no fibular fracture		11
Vb	Tibial pseudarthrosis, sclerosis >30% (20%), and fibular fracture		8
VIa	Tibial pseudarthrosis, sclerosis >50% (35%), and no fibular fracture		5
VIb	Tibial pseudarthrosis, sclerosis >50% (35%), and fibular fracture		2
Leg length discrepancy in comparison to the healthy leg	0–20%		2
	21–40%		1
	>41%		0
Ankle joint contracture		No	2
		Mild	1
		Severe	0
Knee joint contracture		No	2
		Mild	1
		Severe	0
Location of pseudarthrosis third of lower leg		Medially	4
		Medially/distally	2
		Distally	0
Deformity of ankle joint in comparison to standard		0–5°	2
		6–10°	1
		>11°	0
Osteoporosis		No	2
		Mild	1
		Severe	0
Number of previous surgeries		0	2
		1–4	1
		>4	0
Child compliance		Good	2
		Moderate	1
		Bad	0
Parents compliance		Good	2
		Moderate	1
		Bad	0
Muscle function		Good	2
		Moderate	1
		Bad	0

The question is whether the relative extent of the sclerosis before and after the operation is related to a success of therapy. Our investigations demonstrated that the longitudinal extension of the sclerosis seems to have a significant effect on the prognosis. A clear relationship between extensive preoperative sclerosis and tendency for a poor healing process was demonstrated.

It is our impression that if the longitudinal sclerosis includes more than 30% of the bone, a lower rate of bony union can be expected.

CLASSIFICATION

A classification is most useful if it represents the severity of the disease, the basis for treatment, and a prognostic relevance. Classifications according to Andersen (6,20), Boyd (19,21), and Crawford and Bagamery (22) are somewhat deficient.

We developed a new classification, which is based on the severity of the CCSD and independent from the point in time of the evaluation and allows the surgeon to make a decision regarding treatment (Fig. 2; Table 1). *Type I* is a stage of *indifference*. It is characterized by the typical procurvatum—varus deformity of the lower leg without fracture or pseudarthrosis

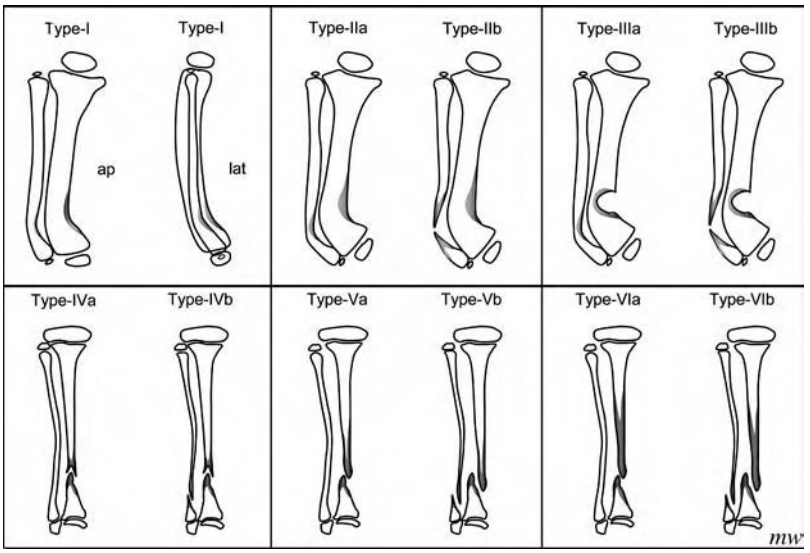

Figure 2 Diagram of the different congenital crural segmental dysplasia types. *Source*: ©Michael Weber.

and without progression of the deformity. *Types II and III* are the stages of *prefracture*. This definition is important because of the impending danger of a fracture. The stage of prefracture is subdivided with respect to an increasing risk of fracture in type II with an increasing deformity and type III with a cystic lesion. The *types IV to VI* describe the stage of *pseudoarthrosis* of the tibia. The differences among types IV, V, and VI are in the extent of the longitudinal sclerosis. This classification is based on results of our pathological and anatomical research and our

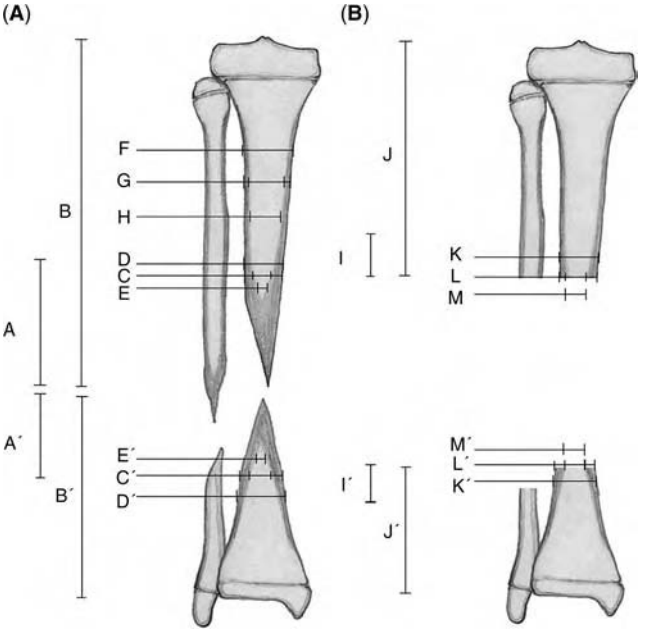

Figure 3 (**A**) Illustration to the method for evaluation of the preoperative extension of sclerosis. A, length of the diaphyseal sclerosis proximal of the pseudarthrosis; A′, length of the diaphyseal sclerosis distal from the pseudarthrosis; B, length of the tibia proximal of the pseudarthrosis; B′, length of the tibia distal of the pseudarthrosis; C, width of the cortical sclerosis at the level of the pseudarthrosis, proximal; C′, width of the cortical sclerosis at the level of the pseudarthrosis, distal; D, width of the tibial diaphysis at the level of the pseudarthrosis, proximal; D′, width of the tibial diaphysis at the level of the pseudarthrosis, distal; E, width of the medullary canal at the level of the pseudarthrosis, proximal; E′, width of the medullary canal at the level of the pseudarthrosis, distal; F, width of the diaphysis near the metaphysis, proximal; G, width of the cortical sclerosis near the metaphysis, proximal;[a1] H, width of the medullary canal near the metaphysis, proximal. (**B**) Illustration to the method for evaluation of the postoperative extension of the sclerosis. I, length of the diaphyseal sclerosis proximal of the resection; I′, length of the diaphyseal sclerosis distal of the resection; J, length of the tibia proximal to the resection; J′, length of the tibia distal to the resection; K, width of the diaphysis at the level of resection, proximal; K′, width of the diaphysis at the level of resection, distal; L, width of the cortical sclerosis at the level of resection, proximal; L′, width of the cortical sclerosis at the level of the resection, distal; M, width of the medullary canal at the level of resection, proximal; M′, width of the medullary canal at the level of resection, distal. *Source*: ©Michael Weber.

Table 2 Six Different Classes of Congenital Crural Segmental Dysplasia According to the Scores

Class	Score
6	0–9
5	10–18
4	19–27
3	28–36
2	37–45
1	46–54

radiological measurement method. This classification is unique in that for types I to III, the pathological dynamics and the progression of the disease are included.

The severity of CCSD increases from types IV to VI. The extent of the sclerosis and, consequently, the percentage of the resected sclerotic bone in relation to the entire tibial length is an essential factor for the classification and prognosis of the disease.

The importance of the fibula is recognized in this classification. A fibula with (b) or without (a) fracture/pseudoarthrosis is indicated as a subtype. The implication of a persisting fibular fracture/pseudarthrosis with hypoplasia, dislocation of the lateral malleolus including a subluxation of the distal talo-fibular joint, and an increasing valgus deformity of the ankle is only scarcely considered in the literature.

Other important factors were considered in the formation of our scoring system (Table 1). The number of previous operations resulting in scaring of the soft tissue and additional osseous defects, the extent of length deficiency, the existence of contractures, and osteoporosis have a significant effect on further complex reconstructive operations.

The psychosocial environment of the child and his or her parents as well as compliance are important factors in success of reconstructive procedures, which demand a high degree of the participation from the involved persons.

These structural conditions and the compliance of child and parents are integrated together with the new classification in a scoring system (Tables 1 and 2).

TREATMENT OPTIONS

The mode of treatment depends on the presence of a fracture of the tibia or fibula and on the dynamics of the deformity (increasing, constant, or a decreasing bowing). It is of no importance whether the bone is recently fractured or if a pseudarthrosis is already present.

Conservative treatment is indicated only when there is no fracture and no progression of the deformity (CCSD type I), because only in these cases an improvement of the deformity can be potentially anticipated. This therapy is focused on bracing. The apex of the deformity is typically located in the middle to the distal third of the lower leg and the use of an ankle–foot orthosis is usually sufficient. If the deformity is located more proximally, a knee–ankle–foot orthotic device with hinges at the knee is recommended. We also use a pad insert to push against the deformity (Fig. 4) to help prevent progression. During the night, the patient wears a removable long leg posterior cast with a similar pad insert and the knee and the ankle joints are held in a neutral position. If increasing deformity is observed despite this bracing, then a healthy periosteum grafting should be performed to avoid an impending fracture. The operative treatment of the disease according to their severity is described below.

We disagree with a frequently expressed opinion that a surgery should not be performed before the end of puberty. Poor results are probably originated from insufficient removal of periosteum or sclerotic bone and/or inadequate osteosynthesis. The delay of surgical treatment until puberty leads to the development of significant problems, especially in cases with distal fracture/pseudarthrosis, such as hypoplasia (foot, entire leg), muscle atrophy, increasing leg length discrepancy, osteoporosis, and contractures. These problems accumulate during the further course of the disease and may significantly complicate or even completely prevent a surgical reconstruction with the consequence of an amputation as the end result. Alternative therapeutic modalities such as pulsating electromagnetic fields (23,24) have been used; however, their therapeutic success is not yet proven (25,26).

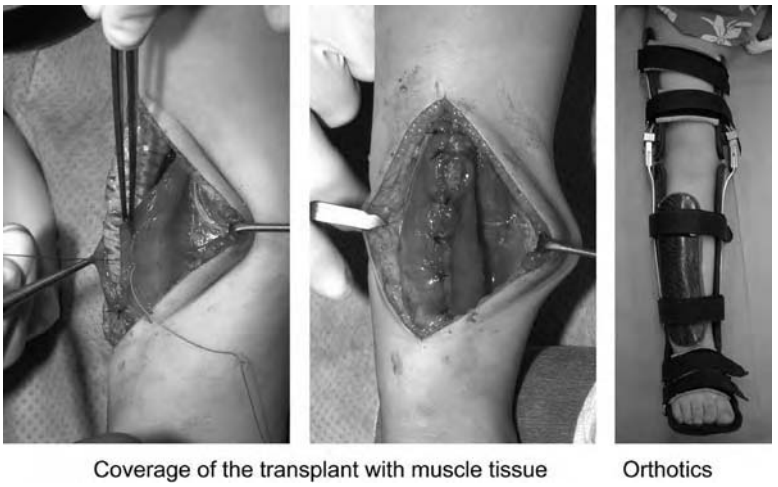

Figure 4 Covering of the periosteal graft with local muscle tissue (left two figures). Orthoprosthetic care (*right*). *Source*: ©Michael Weber.

SURGICAL TECHNIQUES

The type of the operation depends on the severity of CCSD (type II–IV). Ring fixators are favored for reconstructive techniques due to their versatility and stability.

The *CCSD type IIa* (Fig. 2, Table 1) is a progressive deformity of the lower leg without fibular fracture despite conservative orthotic treatment.

The principle of treatment is prevention of a fracture. A complete resection of the affected periosteum and transplantation of nonaffected autogenous periosteum with or without autogenous bone graft of the tibia and fibula is performed (Figs. 4–6). During this procedure the affected periosteum is resected in its entire circumference and longitudinal extension. The sclerotic bone is drilled with a diameter of 2 mm in multiple locations, the drilled bone is left in situ and if required additionally augmented with autogenous and/or homologous bone graft. This area will be enveloped with healthy meshed periosteum (from the iliac crest) and fixed with fibrin glue (Fig. 5). If no sufficient size of periosteum is available to cover the bone completely, the concave side of the procurvatum and varus deformity should be covered with a healthy periosteum. The best source of periosteal tissue is a contralateral iliac crest, where simultaneously autogenous bone graft is harvested.

The periosteum is then covered with muscle and the skin is closed in layers (Fig. 4). Our experience with this technique is promising in a few treated children (Fig. 6); however, the data is preliminary. The child should avoid weight bearing without a brace at all times (Fig. 4).

The *CCSD type IIb* (Fig. 2, Table 1) is a progressive deformity of the lower leg with fibular fracture despite a conservative orthotic treatment.

The principle of the treatment is nearly identical with type IIa. The only difference is that the fibular fracture must be fixed to avoid an atrophy of the fibula and a dislocation of

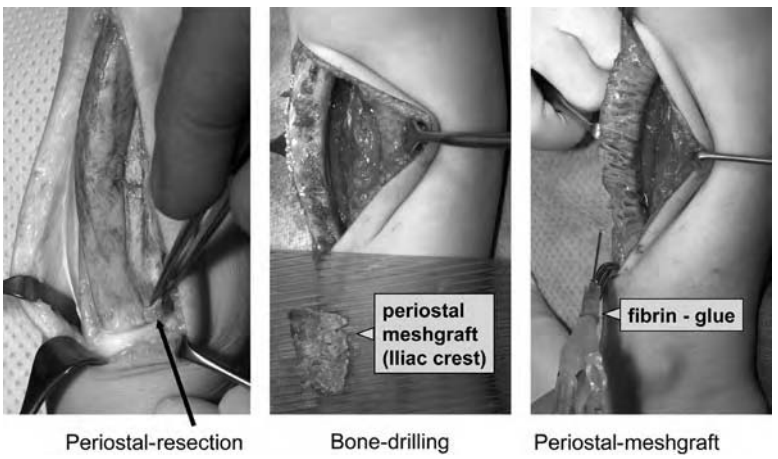

Figure 5 Resection of the pathological periosteum and substitution with autogenous meshed healthy periosteum. *Source*: ©Michael Weber.

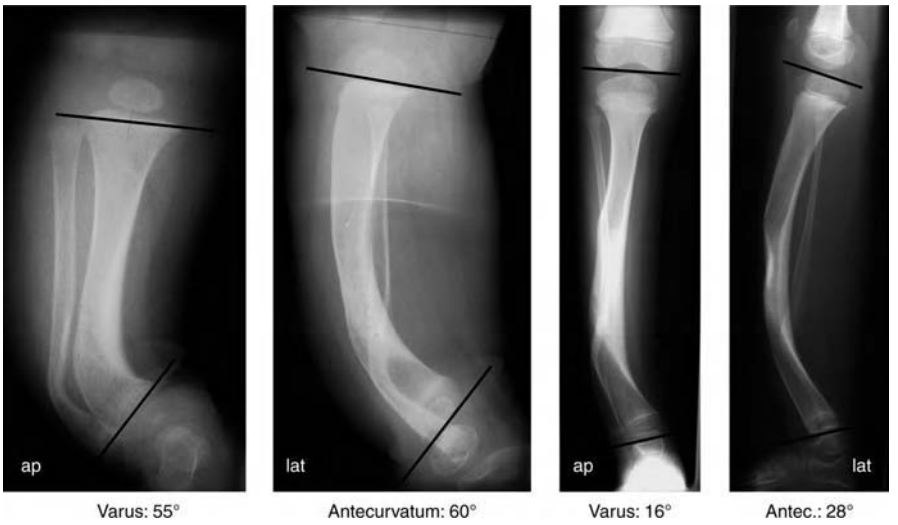

Figure 6 Radiologic course up to four years after the periosteal transplantation with enormous improvement of the deformity. *Source*: ©Michael Weber.

the lateral malleolus. Initially, the pathological part of both fibular ends should be amply resected. Afterwards, two combined operative techniques are applied.

1. The autograft or allograft bone is inserted for the augmentation of the fracture/pseudarthrosis. After that, bone graft and fibular ends are wrapped with a healthy periosteum. This wrapping using nonaffected periosteum serves as a cover for the bone graft. If the fibula is of sufficient size, a central intramedullary wire should be inserted through the fibular fragments. This wire can be buried subcutaneously or connected to the fixator ring.
2. We recommend an own preparation technique using a unicortical graft with cancellous bone from the iliac crest (Fig. 7). The cortical graft is multiple incised with a saw on the cortical side to allow this piece to bend forming a roll (Figs. 7.4–7.7). The fibular fragments are wrapped around with this roll using absorbable sutures (Fig. 7.7). Thus the fibula fragments are well splinted. Because the cancellous side of the graft is located at the outside, good viability of the transplant due to surrounding soft tissue vessels is expected. The integration of the graft is improved also due to the attachment of autogenous cancellous bone graft at the ends of the transplants (see above). This wrapping technique was developed specifically for the augmentation of the docking site following the tibial bone transport. This cortical-cancellous graft is bent to a half tube and wrapped with absorbable sutures around the docking site forming a cylinder (Fig. 7.7). Fibular atrophy complicates the preparation of a conforming cylinder of a corresponding small radius. In this situation, several cortical-cancellous streaks are placed like planks of a barrel surrounding the position of augmentation.

The *CCSD type IIIa* (Fig. 2, Table 1) is defined as prefracture stage with distinct cystic lesion without fibular fracture.

The entire cystic lesion including sclerotic tibial segment should be resected and bone transport using the cable technique should be performed (see type IVa). The intact fibula should not be cut. This will increase the risk of a pseudarthrosis.

The *CCSD type IIIb* (Figs. 2 and 8, Table 1) is defined as prefracture stage showing distinct cystic lesion and a fibular fracture.

The cystic lesion including sclerotic tibial segment should be resected as in type IIIa, and the reconstructive procedure is determined by the extent of the resected segment. An acute compression of the bone ends with a simultaneous proximal lengthening of the lower leg

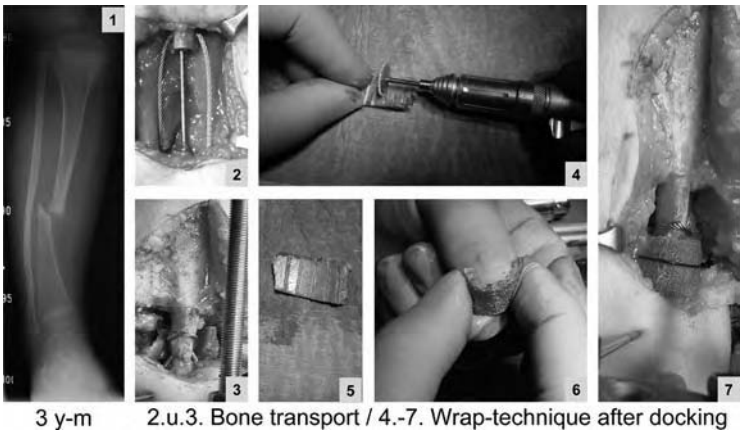

Figure 7 Wrapping technique for the augmentation of the docking site following internal bone transport using the cable technique demonstrated at type IVa. *Source*: ©Michael Weber.

should be performed if less than 30% of the length of the lower leg (about 5 cm in a child) or 20% (about 3 cm in adults) are affected (compare CCSD type IVb, Fig. 9, Table 1). If more than 30% of the length of the lower leg (20% in adults) is affected, a segmental bone transport using the cable technique is performed (compare CCSD type IVa, Figs. 10–12). The fracture of the fibula is repaired as described for CCSD type IIb.

The *CCSD type IVa* (Fig. 2, Table 1) is defined as tibial pseudarthrosis with osseous defect after resection of pseudarthrosis/sclerosis less than 30% (about 5 cm) or 20% (3 cm after growth arrest) and intact fibular.

The aim of the operation is to resect the pseudarthrosis and the complete sclerotic tibia, and to fill the defect in an intact fibula by internal bone transport of the tibia using the cable technique (27–30). The ring fixator is preferred for fixation of the lower leg. Depending on the age of the child or the dimension of the lower leg, a mini-ring-fixator or a conventional ring-fixator is used. If additional deformities are present or are expected due to segmental transport, application of a Taylor-Spatial-Frame is reasonable since it allows a correction without additional remounting. Depending on the length of the lower leg at each fragment, one ring (or better, two rings) is applied and stabilized with at least four wires per fragment. Use of additional wires or screws is reasonable because the fixator is expected to last for a long time and carry a substantial load. Depending on how far the tibial fracture/pseudarthrosis is distally located, the foot will be integrated into the lower leg montage in a typical (horseshoe-like) way. This improves stability and avoids a secondary dislocation of the distal fragment

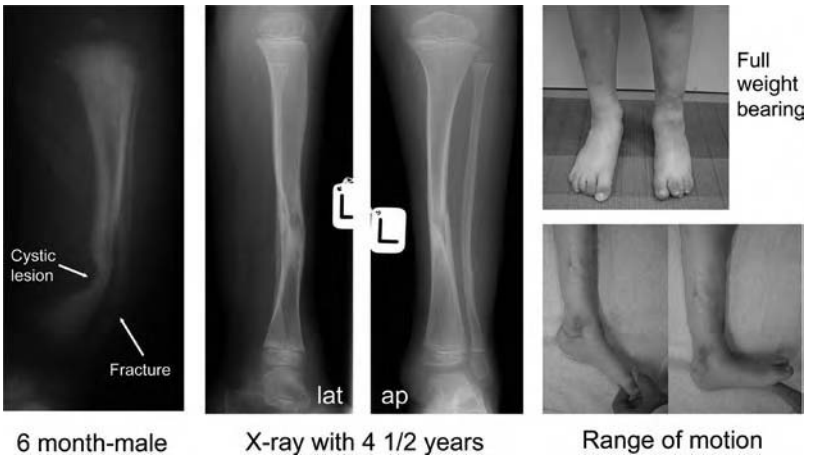

Figure 8 Clinical course of the type IIIb treatment with resection osteosynthesis and proximal lengthening. Primary operation of a six-month-old boy. Operation technique see Figure 9. *Source*: ©Michael Weber.

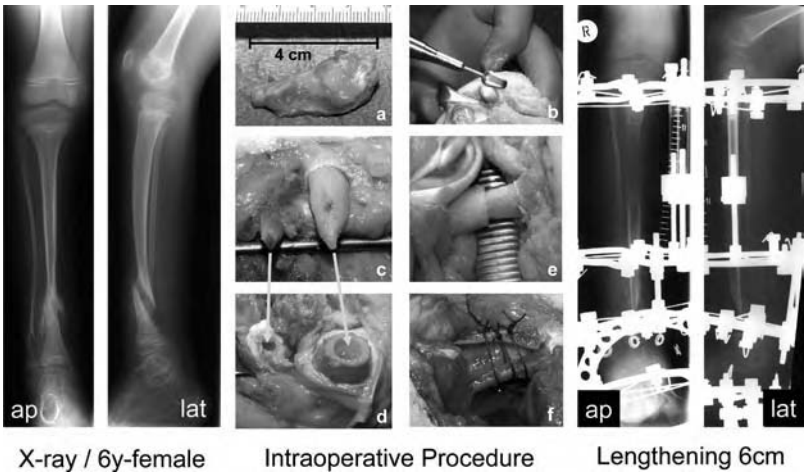

Figure 9 Technique of the resection osteosynthesis with proximal lengthening shown at type IVb. The dowel technique is demonstrated in its sequence: (**A**) resected pseudarthrosis, (**B**) piercing of the proximal ends of the resection with the air drill, (**C**–**E**) insertion of the pierced bone ends into the corresponding distal ends of the resection with the dowel-technique, and (**F**) wrapping technique of the docking site. *Source*: ©Michael Weber.

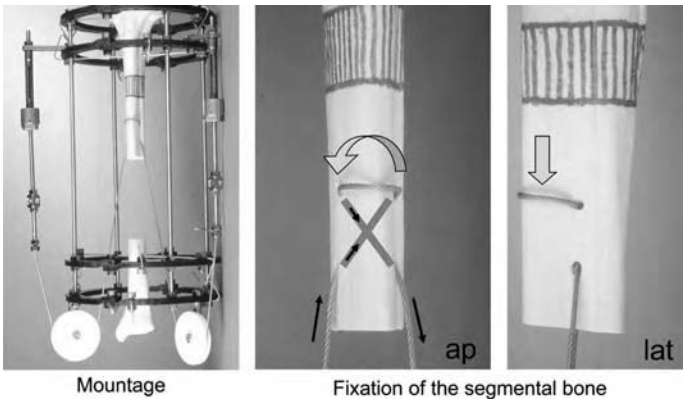

Figure 10 Cable technique and ring fixator mounting technique demonstrated with a saw bone. *Source*: ©Michael Weber.

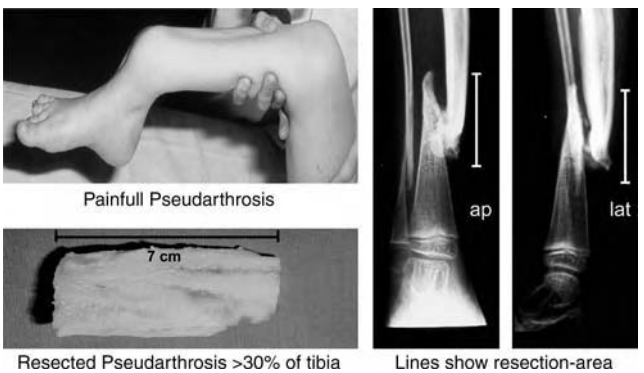

Figure 11 Clinical photographs of a five-year-old boy with type Vb. The resection distance of 7 cm requires a bone transport (see Fig. 12). *Source*: ©Michael Weber.

during treatment. When the ring-fixator is applied and the rings are fixed temporarily, the fracture/pseudarthrosis can be resected. For this purpose, an s-like anterior approach is selected and the pseudarthrosis including the pathological periost is prepared. The length of the resection distance is measured before operation on the x-rays (17). The corresponding segment including the pathological periosteum will be removed. If a distinct thickening of the periost is still present at the borders of the resection (the macroscopic sign of pathology), a subsequent resection up to the region where periosteal thickness appears normal is required. The same is necessary for the osseous borders of the resection. If the sclerosis is clearly visible at the incision border and the medullary canal hardly identifiable, a subsequent resection is necessary as long as reddish bone marrow predominates the area of the incision borderline. Retrospectively, failed operations are usually characterized by insufficient resections.

For fixation of the transported segment with the cable, two crossed but not reconvening drillholes are applied at its ending. The cable (1.5 mm in child, 1.8 mm in adult) is pulled through the drillholes to form a loop over the bone. This technique of fixation avoids a cut through the cable during the transport (Fig. 10). The ends of the cables are each plugged into a Hemovac drain and, using the appending Trokar medially and laterally, lead out through the soft tissue at the distal tibial fragment. It is important that the cable be located in the median level and in alignment with the distal fragment, otherwise, a deviation of the bone segment may occur. The deviation can be avoided by two intramedullary Ilizarov wires which splint the transport (in small anatomical situations, a single wire is enough). The wires are routed from the outside through the proximal fragment into the distal fragment. Insertion of the wires must be performed at the proximal fragment drillhole at a smallish angle. When the wire arrives intramedullarly, it must be driven forward by a hammer because the wire is bending intramedullarly. The proximal end of the wire can be connected to the ring fixator, or better, subcutaneously bent to the bone. The advantage is that the wires, even after removal of the fixator, effect an intramedullary stabilization of the bone. Care should be taken to ensure that the wire ends are proximally correctly bent so that the wires do not cave in intramedullarly during transport or subsequent growth, and do not cause a decubitus at the orthesis. With a short additional cut, the transport segment is proximally prepared for corticotomy. Care should be taken during this procedure to be sure the Ilizarov wires are not damaged. The osteotomy at the transport segment tends to dehisce; therefore, an Ilizarov wire transfixating the osteotomy should be inserted contrary to the transport direction and fixed from the outside at the ring. Before start of the callus distraction, this wire can be extracted easily by a hammer blow.

The transmitted cable ends are applied to the distractors mounted at the fixator and are only slackly tensioned (to avoid dislocation of the osteotomy gap). The position of the rings and the fragments can be now controlled, corrected, and the rings mounted firmly with at least three thread rods. The foot will be integrated in a neutral position into the lower leg montage. Following a latency period (10 days), the transport of the bone segment starts with 0.5 mm a day. When the callus formation is assured, the rate of distraction can be increased to 1 mm a day. Following the docking of the fragments, generation of an autologous (or, if necessary, a homologous) spongy plastic is absolutely recommended. For this purpose, the docking area is uncovered and, if necessary, intervening soft tissue removed. If a gap is still present between the fragments, it can be closed acutely by a continuing distraction of the cables. Starting from the contralateral iliac crest, the apophysis is dissected in the required length twice transversally and once longitudinally, and both parts flapped laterally and medially. The bone as well as the periost now is prepared. If necessary, the medial and lateral sheet of the periosteum can be carefully removed (e.g., for periosteal transplantations). The iliac crest is removed bicortically with sharp chisels in such a way that an osseous margin remains at the apophyseal border of the incision. Both apophyseal parts with their appending soft tissues are sutured together over this osseous margin before additional spongious osseous material from the iliac crest medullary space can be gained (even so, blood may be taken from the iliac crest with a syringe for the subsequent spongiosa transplantation, specifically in the case of a homologous spongiosa transplantation). The preserved periosteum can be meshed for the augmentation (1:3). The bicortical iliac crest bone is split in the spongious region resulting in two corticospongious transplants. Using a fine saw, the sides of the corticalis are given several parallel chinks, without cutting the corticalis completely (Fig. 7). Using this technique, the iliac crest transplants can be bent, placed around the docking position, and fixed with circular absorbable threads before any autologous (or even homologous) spongiosa is placed around the

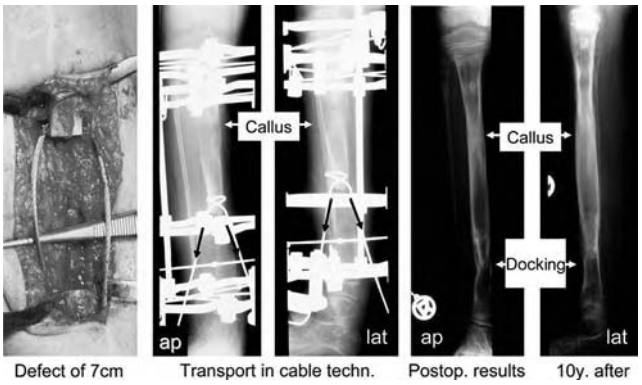

Figure 12 Same patient as in Figure 11. Intraoperative and postoperative photographs of the cable technique. *Source*: ©Michael Weber.

docking position. Due to this wrap technique, the surface of the spongiosa is more easily accessible for the surrounding soft-tissue vessels compared to a corresponding layer of corticalis. Furthermore, a cylinder develops due to the molding that shields the docking zone and protects against further mechanical stress. In all cases, a consolidation of the docking zone could be achieved. The periost can be used as an additional layer, placing it around the transplants and fixing with a fibrin glue. When a distinct fibular bending is present, the pathological periost should be removed by the same approach and augmented with the autologous periost from the contralateral iliac crest. In no instance should a correction osteotomy of the intact fibula be carried out. The deformity equalizes on its own during further growing. With increasing consolidation of the callus, the foot montage, which was already supplied with isometric placed hinges during the primary montage, can be opened and the foot mobilized at the axis of the upper ankle joint. If a contracture preexisted or a long lasting immobilization in the upper ankle joint preceded, a distraction in the upper the ankle joint of 5 mm in length with subsequent continuous extension and flexion is recommended for improvement of the ROM. After the consolidation of the callus, the fixator is removed. Until fitted with a prosthesis, the lower leg is immobilized in a Sarmiento-Brace.

The *CCSD type IVb* (Fig. 2, Table 1) is defined as tibial pseudarthrosis with fibular pseudarthrosis with osseous defect after resection of pseudarthrosis/sclerosis less than 30% (about 5 cm) or 20% (3 cm after growth arrest).

The principle of the operation is the resection of the sclerotic tibia and fibula and the osteosynthesis of the proximal osseous ends of tibia and fibula with their corresponding distal fragments. The resulting length deficiency is equalized by a proximal lengthening of the lower leg following a corticotomy of tibia and fibula.

There are two salvage procedures for the resection osteosynthesis.

1. *The end-to-end technique*: The ends of the fragments are placed directly in contact at the resection margin. In the tibia, this is normally a trouble-free procedure. In contrast, at the fibula, which appears frequently atrophic, this apposition is difficult to achieve. A bone grafting of this site (as shown before) is important for the healing process of both bones.

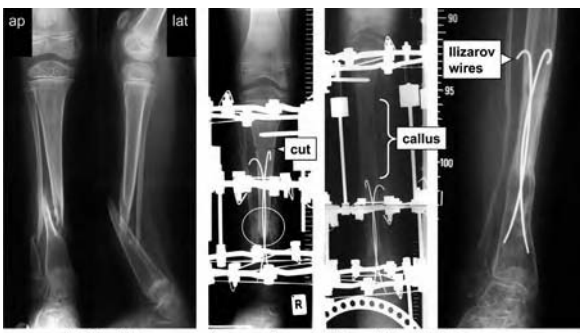

Figure 13 The intramedullary Ilizarov-wire-guidance of the docking site stabilizes the tibia even after the removal of the fixator. The circle marks the docking site (dowel technique) with bone grafting. *Source*: ©Michael Weber.

2. *The dowel technique*: Pointy proximal ends of the fragments are put into the appropriate open medullary canals of the corresponding fragments (Figs. 9 and 13). Some remnants of the sclerosis at the proximal fragments may remain due to the fact that they can be inserted more easily. This technique provides the solid inner splinting of the bones. This is of particular advantage in cases of small osseous structures in young children when central leading wires cannot be inserted and, specifically, in the presence of an atrophic fibula. The healing will also be enhanced by additional bone grafting (see above).

The *CCSD type Va* (Fig. 2, Table 1) is characterized as tibial pseudarthrosis without fibular pseudarthrosis with a resulting osseous defect after resection of the pseudarthrosis/sclerosis of more than 30% of the length of the lower leg. The principle and procedure of the treatment are the same as described for CCSD type-IVa.

The *CCSD type Vb* (Fig. 2, Table 1) is characterized as tibial pseudarthrosis with fibular pseudarthrosis, with resulting osseous defect after resection of pseudarthrosis/sclerosis of more than 30% of the length of the lower leg (20% after growth arrest). The principle of the treatment is a complete resection of the tibial pseudarthrosis with their sclerosis and an internal bone transport using cable technique (Figs. 11 and 12). Splinting of the medullary canal with an Ilizarov wire and bone grafting using cortical-cancellous bone (see wrapping technique above) is recommended for the treatment of the fibula. A bone transport is carried out as described for CCSD type IVa.

The definition of *CCSD type VIa* (Fig. 2, Table 1) is a tibial pseudarthrosis without fibular pseudarthrosis and a longitudinal tibial sclerosis of more than 50%. The principle and procedure of the treatment are the same as described for CCSD type-IVa.

The definition of *CCSD type VIb* (Fig. 2, Table 1) is a tibial pseudarthrosis with fibular pseudarthrosis and a longitudinal tibial sclerosis of more than 50%. The principle of treatment and procedure are described in CCSD type Vb.

It is difficult to stabilize a distal fragment of the tibia when pseudoarthrosis is located in close proximity to the ankle joint. In these cases the foot should be included in the frame as well.

Our results and the European Pediatric Orthopaedic Society multicenter-study (12) demonstrated that conventional surgical techniques such as K-wire, plate-osteosynthesis and unilateral fixators are inappropriate for the treatment of CCSD due to the low rates of healing. Even the fibula transfer is an inferior technique compared to the ring fixator method. Children treated ineffectively end up with secondary defects, osteomyelitis, scarring, shortenings, and increasing painful pseudarthrosis. Those children are hospitalized and psychologically altered by often more than 10 unsuccessful operations so that neither the child nor the parents agree to another reconstructive procedure (31,32). These situations sometimes require an amputation as ultimate treatment.

Acute shortening with proximal lengthening or internal bone transport should be the method of choice for treatment of bone defects. In respect to the technique of the fibular transfer, each surgeon should respond to the question of why the healthy leg should be subjected to the substantial donor problem. There are also other disadvantages such as difficulties of microanastomosis (especially in children), persisting length deficiency, smaller calibers of the transplanted fibula, high rate of fractures, and revision intervention for lengthening. In our opinion, CCSD can be treated much more successfully using the described methods of the bone transport and distraction.

The fibular transfer should be reserved only for those cases, where the sclerosis is so pronounced that a bone transport is not possible, and should be performed using ring fixators to ensure the highest stability after resection of sclerosis as explained before. A long-lasting dynamization phase in the ring fixator allows for adaptation of the fibula and the avoidance of a fracture. The lengthening of the lower leg should be carried out only after a sufficient hypertrophy of the fibular transplant has occurred.

In cases of multiple unsuccessful procedures, the child and the parents could request an amputation (33–45). The described scoring system gives a better base for such a decision. The system includes main pathological changes in CCSD and legitimizes a decision for amputation if those changes are graded in class 6. Three of my patients, who were treated unsuccessfully more than eight times each alio loco, together with their parents refused further reconstructive surgeries so that an amputation became inevitable. These patients scored less then 9, which put them into a class 6 category (Tables 1 and 2). In these cases an own amputation technique

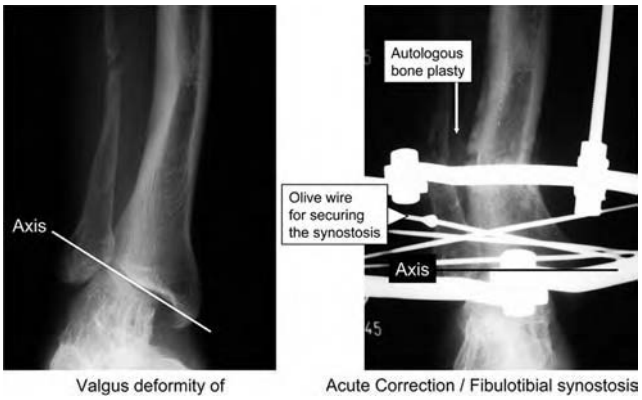

Figure 14 Dome osteotomy demonstrated at type Vb. *Source*: ©Michael Weber.

was used to prevent the problems of conventional amputation techniques such as stump piercing, reduced endbearing. The principle of treatment is the amputation of the lower leg with a concurrent stump plasty using the ipsilateral nerve and vessel pedciculled calcaneus. The method eliminates the mentioned problems and gives rise for a full weight bearing stump besides other advantages compared to conventional amputational techniques (31,32).

Braces (Fig. 4) are recommended in all cases after frame removal to prevent refractures or recurrence of a pseudarthrosis.

OPERATIVE TREATMENT OF OTHER PROBLEMS TYPICAL FOR CONGENITAL CRURAL SEGMENTAL DYSPLASIA

An *ankle joint* problem in CCSD is characterized by a disturbance of growth of the lateral tibial epiphysis and consecutive valgus position of the foot. There are different techniques available to correct this problem depending on the patient's age:

1. Temporary epiphysiodesis of the medial distal growth plate of the tibia until the normalization of the wedge-shaped distal tibia epiphysis occurs.
2. Dome osteotomy for acute correction of the joint line valgus after completion of the growth (Fig. 14).
3. Tibio-fibular synostosis proximally to the growth plate for the stabilization of the ankle joint if the *fibular pseudarthrosis* cannot be healed by the above described methods because of the severe atrophy of the proximal fragment. For this purpose, a synostosis procedure is performed using bone grafting and screws (Fig. 15). Following consolidation, the screws are removed.

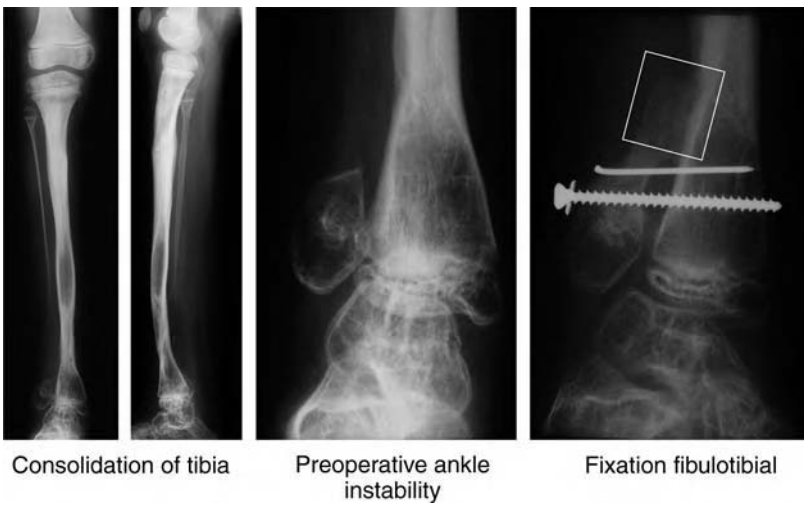

Figure 15 Stabilization of the ankle joint by fibulo-tibial synostosis demonstrated for type IVb. The frame shows the site of the bone grafting. *Source*: ©Michael Weber.

In cases of ankle deformity due to a short distal fibula, it should be pulled down in order to restore the fibulo-talar congruity of the joint along a leading wire using a cable technique, and as a second step, repair of the fibular pseudarthrosis or the tibio-fibular synostosis should be performed. In cases with intact fibula and short tibia, it is important to pull the tibial fragment distally until the fibulo-talar joint congruity is achieved before transfixing fibula and tibia in the ring fixator.

Contractures originate from immobilization or the orthoprosthetic treatment of the adjacent joint of the lower leg and could be treated using ring fixators with established techniques for treatment of contractures.

COMPLICATIONS

Bone transport is advisable in the case of a bony resection of more than 30% in children (about 5 cm) and 20% after growth arrest (about 3 cm) because an acute compression over such distances causes a compromise of the blood flow, lymphedema, persistent soft tissue bulging, and compromises the healing process and the cosmetic result. The difference in our recommendation for acute compression distances between children and adults originates in the higher tolerance of the soft tissues and an improvement of the soft tissue bulging with the further growth of the child. The above numbers are based on our experience and could differ in every situation. For example, the amount of acute compression tolerated could be less in the presence of scars after previous surgeries.

Rates of healing can be compromised when insufficient resection of sclerotic ends is performed in order to avoid bone transport. The mode of treatment can be compared with a tumor resection. Insufficient resection will lead to failure and recurrence (8). In this context, high rates of refractures at the docking zone have to be mentioned. However, these refractures do not represent new real fractures but rather pathological fractures following inadequate trauma due to an absent primary healing. All fractures in the docking region after a minimal trauma should be classified as primary insufficiently healed CCSD.

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36 Blount's Disease

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INTRODUCTION

Erlacher originally described the clinical and radiographic findings typical of Blount's disease in the German literature in 1922. The first detailed English report by Walter Blount was published in 1937 (1), followed by another extensive study by Langenskiold from Finland in 1952 (2).

Two clinically distinct forms, i.e., early (infantile) (Fig. 1) and late onset (adolescent) (Fig. 2) of Blount's disease exist based on the onset of deformity, before or after the age of four years. Although key clinical and radiographic differences exist between these two categories, there are several similarities including predisposition in obese, black children and histologic findings at the proximal tibial growth plate (3,4).

The pathogenesis of genu varum in Blount's disease is based on the Heuter-Volkman principle of increasing compressive forces causing growth inhibition. Excessive pressure at the proximal medial tibial cartilaginous epiphysis causes altered structure and function of the chondrocytes along with delayed ossification of the epiphysis (5). This view is supported by the clinical experience of an increased incidence of Blount's disease in obese children with physiologic genu varum, who start walking at an early age (6). Finite element analysis has also demonstrated the compounding effects of obesity and genu varum on the compressive forces generated at the medial segment of the proximal tibia (7). However, this hypothesis does not fully explain the unilateral or asymmetric involvement of the limbs in Blount's disease. Also, there are reports of documented normal radiographic alignment of the knees of patients, who subsequently developed late onset form of the Blount's disease (4,8).

The relative inhibition of the posteromedial portion of the proximal tibial growth plate leads to asymmetric growth, thereby causing a three-dimensional deformity of the lower leg including varus, procurvatum, and internal tibial torsion (6,9,10). This entity can lead to progressive deformity with gait abnormality, leg length inequality, and premature arthritis of the knee (11–13).

CLINICAL EVALUATION

History and Physical Exam

Clinical evaluation should include a thorough history including presence of pain, functional limitations, as well as age of onset of independent walking age when deformity was first noted and prior treatment modalities such as bracing or surgery (Fig. 3). The psychosocial assessment of the patient and family support are crucial prior to embarking on a lengthy limb reconstruction.

Physical exam involves assessment of weight and height as well as any general dysmorphic features suggestive of secondary causes of genu varum including rickets, bone dysplasias such as metaphyseal chondrodysplasia and endocrinopathies such as Prader-Willi syndrome. Evaluation of leg-length discrepancy, especially with the patient standing with a lift under the short leg to level the pelvis is critical. In severe forms of Blount's disease, the tibia subluxates laterally and allows the medial femoral condyle to fall into the posteromedial depression in stance (10). Gait abnormalities include the presence of varus thrust during single leg stance, which suggests excessive shear forces generated at the knee joint. The relatively long fibula may cause functional laxity of the lateral collateral ligament and further add to the dynamic varus. Varus and valgus instability should be assessed with the knee in full extension and 20° of flexion. Because there is frequent association of procurvatum of the proximal tibia (9), mild flexion deformity of the knee

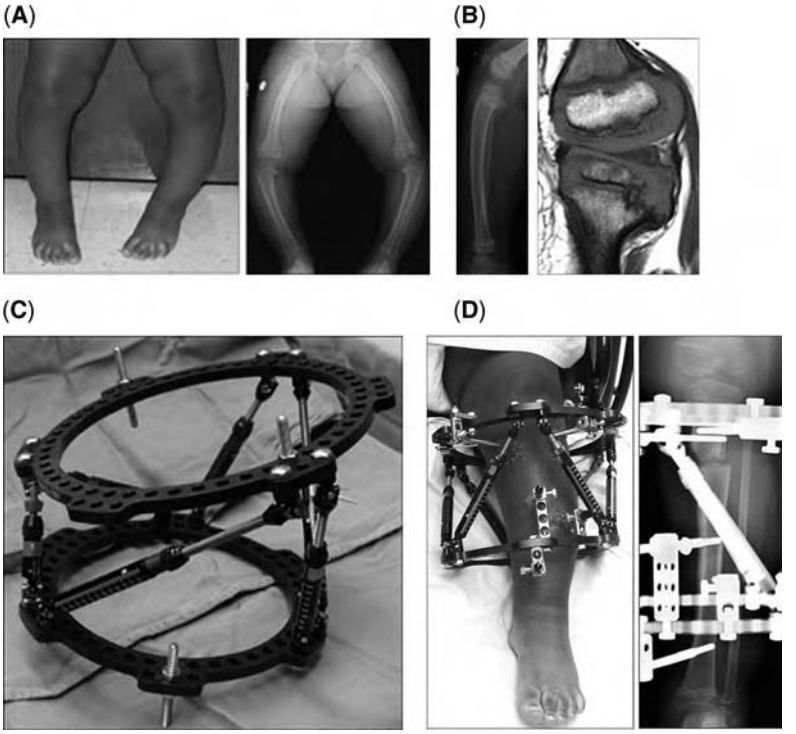

Figure 1 (**A**) Clinical photo and standing radiograph of a three-year-old female with bilateral early onset Blount's disease. (**B**) Lateral radiograph and sagittal magnetic resonance imaging of the same patient demonstrating proximal tibial procurvatum and changes in the posterior portion of the medial tibial growth plate. (**C**) Preconstructed external fixator for correction of varus, procurvatum, and internal tibial torsion of left leg. (**D**) Clinical photo and radiograph following gradual correction of the triplanar tibial deformity.

may be present. Often this bony deformity is mistakenly labeled as a ''flexion contracture'' of the knee, and the sagittal plane deformity is not addressed at the time of skeletal realignment (Fig. 3B). Rotational profile of the lower limb including the thigh-foot angle assessed on prone exam needs to be routinely evaluated because internal tibial torsion is quite common, especially in the early onset form of Blount's disease.

Imaging Studies

Langenskiold (2) described six radiographic stages of the early onset Blount's disease based on patient's age and degree of proximal tibial deformity. Although useful, there is significant interobserver variability with this classification (14). There are other radiologic parameters such as metaphyseal-diaphyseal angle (15,16), as well as contribution of the varus deformity by the femur and tibia (17) that can aid the clinician in differentiating physiologic bowing from early onset Blount's disease.

The key to the identification of hypophosphatemic rickets and metaphyseal chondrodysplasia is the radiographic appearance of the physis. Although both of these disorders are characterized by widening of the physis (18), appropriate blood work aids in confirming the diagnosis of rickets.

Standing full-length anteroposterior (AP) radiograph (teleroentgenogram) (Fig. 1A) of the entire length of both lower extremities with the patella pointing forward is crucial for analysis of frontal plane alignment. However, this study can be challenging to perform in the Blount's population with the large size of the patients compromising adequate visualization of the femoral heads, along with the tendency for the radiology technician to position the leg with foot forward rather than patella forward position. In young children, prior to sufficient ossification of the patella, it is useful to place a metal marker over the center of the patella to confirm its forward position. Also, no more than 60% of the proximal fibula should be seen to overlap the adjacent tibia on a true AP radiograph centered at the knee, irrespective

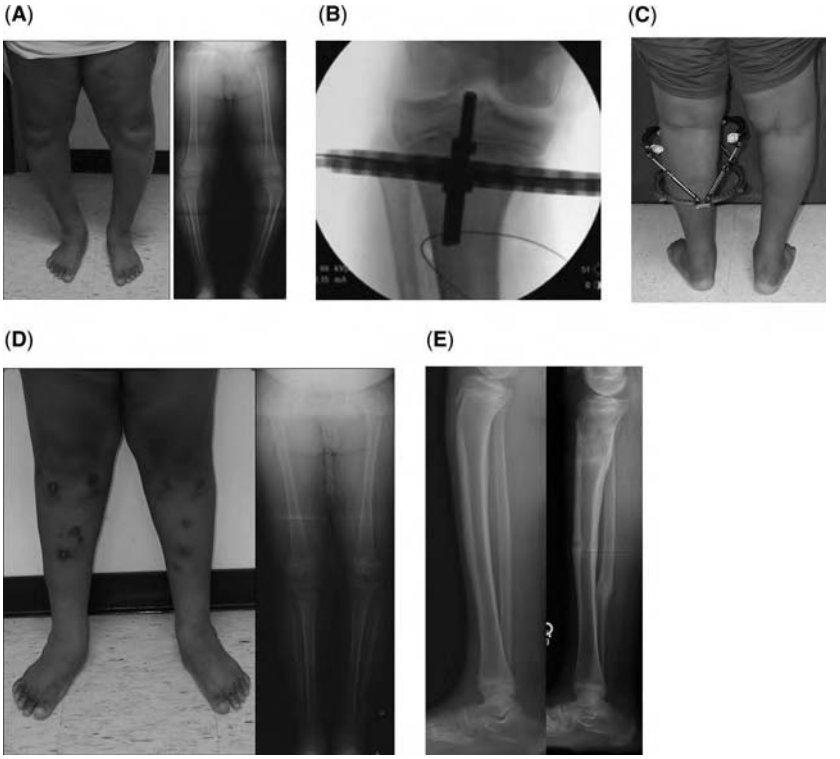

Figure 2 (**A**) Clinical photo and standing anteroposterior (AP) radiograph of a 12-year-old obese male with bilateral late onset Blount's disease. He underwent staged realignment of both legs few months apart. (**B**) Intraoperative fluoroscopic AP view of the proximal tibia after centering of the proximal ring and percutaneous passage of the Gigli saw at the level of the planned osteotomy site. (**C**) Clinical photograph during the consolidation phase of realignment of the left sided deformity. Note the posterior opening of the proximal ring to allow knee flexion. (**D**) Clinical and radiographic appearance following bilateral staged tibial osteotomies with gradual correction. The mechanical axis has been restored to normal. (**E**) Pre- and postoperative lateral view radiographs of the tibia showing adequate correction of the proximal tibial procurvatum deformity.

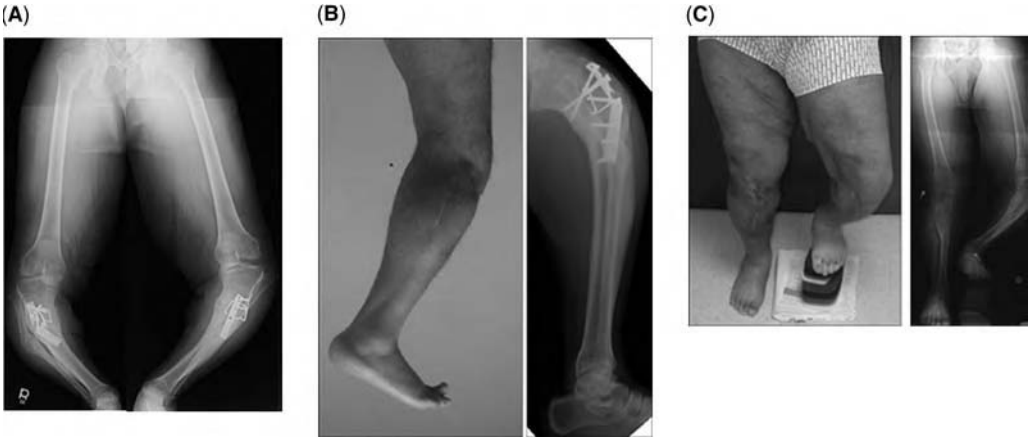

Figure 3 (**A**) Standing anteroposterior view radiograph of a 24-year-old male with bilateral painful knees, who had previously undergone multiple surgeries for correction of Blount's disease. Bilateral multiapical deformities of the tibia with severe shortening are seen. Note the broken hardware on the right and proximal tibiofibular joint incongruity on the left side. (**B**) Clinical and lateral view radiograph demonstrating the flexion deformity secondary to the uncorrected procurvatum, and not a ''knee contracture.'' (**C**) Clinical and radiographic appearance with a 15 cm lift under left leg, following realignment of the right tibia with bilateral hardware removal. He is awaiting similar reconstruction of the left side.

of the patient's age (19). One should systematically evaluate for mechanical axis deviation by performing the malalignment and malorientation tests (20). Despite being referred to as ''tibia vara,'' Blount's disease may have other sources of medial axis deviation arising from the distal femur (9,21,22) and intra-articular deformity creating dynamic varus malalignment. Full-length AP and lateral views of the tibia are needed to fully assess the presence of biplanar deformities in the proximal and distal ends of the tibia (Figs. 1 and 2E). A scanogram and skeletal age assessment are useful for comprehensive analysis of current and future leg-length discrepancy.

Magnetic resonance imaging (MRI) is a useful tool for preoperative analysis of patients with advanced stages of early onset Blount's disease (Figs. 1 and 4A). Intra-articular changes such as posteromedial depression of the tibial plateau and hypertrophy of the medial meniscus are commonly seen. Plain radiographs may overestimate the ''depression'' of the medial plateau (23), which may be due to failure of endochondral ossification of the medial tibial epiphysis in response to increased compressive forces (5). Presence of bony bars and irregularities of the posteromedial proximal tibial growth can be well visualized with computed tomography (CT) and MRI scans (24) (Figs. 4A and C). Although advanced imaging is not indicated routinely in late onset Blount's, changes in adjacent distal femur epiphysis and physis have been described in advanced stages of the disease using MRI (25). Arthrograms are also helpful in delineating the articular surfaces, especially in early-onset disease (23) and are typically done under the same anesthetic as the definitive surgical procedure.

CLASSIFICATION

Traditionally, Blount's disease has been classified on the basis age of onset alone (1,3,26). While this is somewhat useful in determining prognosis and guiding treatment, one should avoid a

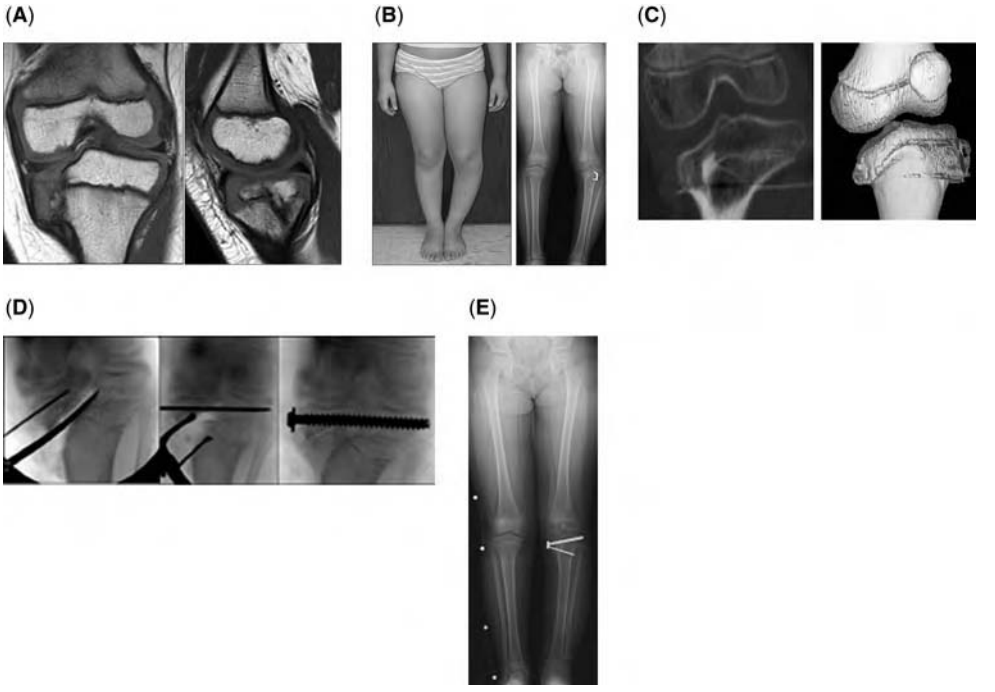

Figure 4 (**A**) Magnetic resonance imaging of a five-year-old with left-sided early onset Blount's disease. Note the unossified medial proximal tibial epiphysis and irregularity in the posterior third of the proximal tibial growth plate. This patient underwent a proximal tibial osteotomy with acute correction and normalization of the mechanical axis. (**B**) She had recurrent deformity, which was unsuccessfully treated with a single anterolateral extraperiosteal staple to attempt correction of the proximal tibial varus and procurvatum. (**C**) Follow-up computed tomography scan three years postosteotomy revealed a bony bar across the medial tibial physis with depression of the medial plateau. (**D**) Fluoroscopic views demonstrating the technique of medial tibial plateau elevation with internal fixation and use of a structural allograft. (**E**) Standing radiograph showed satisfactory healing of the tibial plateau osteotomy and improved limb alignment.

''cookbook'' approach when dealing with this disorder. From a management standpoint, it is helpful to create a ''problem list,'' so that each issue can be individually addressed and the most comprehensive treatment strategy utilized for the specific patient. This approach takes into account all clinical and radiographic information available, enabling the surgeon to methodically think through the various ''problems'' and assign the best-fit solution to each. Because Blount's disease affects children, one needs to consider not only the current deformity and leg-length discrepancy, but also anticipated discrepancy at skeletal maturity, with and without treatment. The key issues that can influence decision making include unilateral versus bilateral deformity, magnitude of tibial and femoral deformity in all three planes, leg length discrepancy, years of growth remaining, as well as condition of skin and soft tissues related to obesity and prior surgeries.

TREATMENT OPTIONS

The goal of treatment in Blount's disease is to attain a normally aligned lower extremity with normal joint orientation (20) and equal leg lengths at skeletal maturity. Observation with repeat clinical and radiographic exam is recommended in the child under two years of age, especially if the diagnosis is uncertain and the varus deformity is mild. Although the clinical effectiveness and feasibility of bracing is debatable, some advocate the use of a knee–ankle–foot orthosis in such cases (27,28).

As established by the basic science studies (5,29) and validated clinically (30), the inhibitory effect of compressive forces on the growth plate is not an all-or-none phenomenon. Thus, if the medial compartment of the knee joint is ''unloaded'' by realignment of the axis deviation laterally, the medial proximal tibial physis may respond by resuming normal growth. This is most applicable in early onset Blount's. Numerous studies have shown that the incidence of recurrent deformity is least with realignment osteotomies performed prior to four or five years of age, before the onset of advanced pathologic changes in the proximal tibial growth plate (28,31–34).

Besides proximal tibial metaphyseal osteotomy, with either acute or gradual correction, several other realignment strategies are available. These include lateral hemiepiphyseodesis around the knee (35), distal femoral osteotomy (34), medial tibial plateau elevation (10,13, 36–41), resection of physeal bony bar (26,34,42), and gradual asymmetric proximal tibial physeal distraction (43). Often, two or more of these surgical modalities are applied at the same time or in a staged manner. Examples include lateral hemiepiphyseodesis of the proximal tibia and fibula along with medial plateau elevation in an older child with severe form of early onset Blount's (Fig. 4). If associated metaphyseal deformity is also present, one may need to perform a second osteotomy for correction at this site as well as gradual lengthening to equalize limb lengths at maturity. This can be performed at the same time as the plateau elevation or subsequently. Similarly, an adolescent who has two different sites of deformity i.e., the distal femur and proximal tibia, may need a two-level osteotomy or a combination of osteotomy at one location and lateral hemiepiphyseodesis at the other. The location and timing for the hemiepiphyseodesis is based on the magnitude of deformity and growth remaining. This option is typically employed for mild to moderate varus malalignment in late onset disease, especially if both extremities are involved.

If gradual correction with external fixation is being contemplated, a candid preoperative discussion regarding need for frequent follow-up visits, including radiography and physical therapy needs to be held. Visual display of a model of the external fixator, education handouts, as well as opportunity to meet with other patients and family members, who have gone through similar procedures can be helpful in allaying the anxiety of the patient and caregivers. The concept of individualizing treatment based on a comprehensive analysis of the limb deformity, amount of growth remaining, psychosocial status of the patient, and ability of the surgeon to execute a well-outlined treatment plan with precision and safety is crucial in achieving a successful outcome.

SURGICAL TECHNIQUES AND PITFALLS

Angular and rotational correction in Blount's disease can be accomplished by acute correction. A variety of techniques have been proposed including closing wedge (39,44), opening

wedge (20), dome (45,46), and inclined (47) osteotomies, each with its own advantages and disadvantages. Different fixation methods have been proposed including cast only (33), smooth pins and wires (33), interfragmentary screws (47), plates and screws (39,48), and external fixators (31,49–51). Irrespective of the type of osteotomy and fixation device, there is a significant incidence of neurologic injury and compartment syndrome with acute correction (28,33,52–54). In order to avoid these major sequelae, a prophylactic anterior compartment fasciotomy, with or without peroneal nerve decompression and insertion of a suction drain should be considered in majority of cases treated with acute correction. Vigilant postoperative management with frequent clinical exam and avoidance of predisposing factors such as tight casts and dressings, epidural analgesia and excessive sedation is necessary.

One needs to determine the location of the apex of the deformity, i.e., the center of rotation and angulation (CORA) (20), and determine whether it is feasible to perform the correction at that site. The tibial osteotomy is typically made in the metadiaphysis, distal to the tibial apophysis and insertion of the patellar tendon. The location of the osteotomy being away from the CORA necessitates appropriate lateral and often anterior translation of the distal fragment, in order to realign the anatomic axis of the tibia. Disregarding this principle creates a secondary translational ''dog-leg'' deformity at the metadiaphysis as well as valgus malorientation of the ankle. Some of the ''classic osteotomies'' such as barrel vault dome osteotomy, popularized by Maquet, may not be applicable in Blount's because being an inverted (''sad face'') arc, the center of rotation in a Maquet osteotomy is significantly distal to the CORA, and thus induces a marked translational deformity with angular correction. On the other hand a ''smiling face'' dome osteotomy (20,45) will have its center of rotation closer to the CORA and will induce appropriate translation and avoid injury to the proximal tibial apophysis. Another common surgical error is to attempt realigning the mechanical axis with a proximal tibial osteotomy while disregarding associated varus malorientation of the distal femur, creating knee joint obliquity. If there is recurrence of the deformity requiring further intervention, the surgical procedure can be complicated by such iatrogenic deformities (Fig. 3).

Although there is no one ''best way'' to perform limb realignment, we prefer to correct most cases of early and late onset disease requiring tibial osteotomy with gradual correction using distraction osteogenesis. This allows a comprehensive means of safe and controlled correction of all deformities including leg-length discrepancy. It also facilitates early mobilization of the patient and the opportunity to analyze limb realignment based on weight-bearing radiographs and avoids large incisions and a prolonged period of cast immobilization. A variety of monolateral (55,56) and circular fixators (57–60) have been described for gradual correction of Blount's disease (Table 1). Although there are distinct advantages of monolateral fixators such as a lower profile, better patient tolerance, and possibly shorter learning curve for the surgeon, there are disadvantages as well. These include constrained sites for pin placement, overlying hardware compromising radiographic visualization of the underlying bone, and lack of versatility with regard to multiplanar correction and limited options for correcting residual deformity during gradual realignment. Currently, we prefer to use a circular fixator such as the Taylor Spatial Frame (Fig. 1C). The two-ring external fixator is placed on the limb to

Table 1 Literature: Distraction Osteogenesis Treatment for Blount's Disease

References	Early/Late Onset	Patients (Limbs)	Device	Major Complications
55	Late	17 (27)	Modified Wagner frame	Undercorrection (1); loss of correction (1)
56	Late	11 (14)	Orthofix T-Garche	Peroneal palsy (1); undercorrection (1)
58	Late	8 (12)	Ilizarov frame	Premature consolidation (1)
60	Late	17 (25)	Ilizarov frame	Delayed union (1); premature consolidation (1); residual LLD (2); residual deformity (1)
57	Both	45 (69)	Ilizarov frame	Residual deformity needing surgery (6); loss of knee mobility (10)
59	Both	19 (22)	Taylor Spatial Frame™	Intractable pin infection (1); delayed union (1)

match the deformity in all planes and correction is performed utilizing the internet-based computer software program. The author prefers to preconstruct the frame and use the ''chronic deformity'' mode, with the ''residual deformity'' modes reserved for final adjustment, if required. A preconstruction of the frame allows the surgeon to think through the surgical steps and choose appropriate ring and strut sizes beforehand. This exercise also minimizes the need for postoperative strut changes. Another technique for deformity correction of the knee using the ''rings first total residual'' mode is described in other chapters. In order to allow sufficient knee flexion, we either use a two-third ring proximally, or cut the posterior third of the ring during the consolidation phase of treatment (Fig. 2C). The surgical technique involves use of a radiolucent table, intraoperative flouroscopy with free draping of the entire lower extremity, and use of a sterile thigh tourniquet. The tourniquet time is kept to a minimum by using it only for preparation of the osteotomy cuts. A 1-cm piece of the fibula is resected at the junction of middle and distal third of the shaft in most cases. The Gigli saw is passed on a heavy suture using two small transverse incisions in the proximal tibial metaphysis (Fig. 2B) (20). Other percutaneous techniques such as a multiple drill-hole osteotomy that minimizes thermal necrosis and can be safely performed through small incisions with preservation of the periosteal sleeve can also be utilized. Power saw osteotomies of the proximal tibia should be avoided. The tibial osteotomy is not completed at this stage, and the preconstructed frame is appropriately centered and mounted on the lower leg (Fig. 2B). Typically, a reference wire and two appropriately sized hydroxy-apatite coated half pins are attached to the proximal ring and three half pins are mounted off the distal ring in different planes. Following fixation of the frame to the tibia the struts are temporarily disengaged from one of the rings, and the osteotomy completed.

The decision to place a distal fibulotibial transfixation wire or screw to prevent proximal migration of the fibula depends on several factors. For cases requiring less than 2 cm of lengthening, we typically osteotomize but do not transfix the distal fibula. In adolescent cases where mainly angular correction is planned and the fibula is relatively long and possibly contributing to the lateral collateral laxity at the knee, the fibular osteotomy is not done with the intention of ''tightening'' the lateral collateral ligament with angular correction. In order to avoid proximal migration of the intact fibula at the ankle mortise, the distal fibula is typically transfixed at the level of the syndesmosis. Whether this ''retensioning'' leads to a lasting correction of the proximal fibula is unknown at this time.

There is no consensus in the literature regarding the ideal alignment of the lower extremity following surgical reconstruction for Blount's disease. Some advocate normalization of the mechanical axis (2), while others feel that some degree of overcorrection should be attempted (28,31,37). A major pitfall in these and other studies is that the assessment of surgical correction is based on non–weight-bearing radiographs of the knee, often with an overlying cast, without visualization of the entire femur and tibia. Given the possibility of recurrent deformity with several years of growth remaining, as well as increased genu valgum in the younger child, we typically aim for slight over correction in early onset and normalize the mechanical axis in late onset form of Blount's.

The area of functional and structural abnormality acting as a ''spot weld'' is most often at the posteromedial portion of the proximal tibial growth plate (Figs. 4A and C). Although asymmetric physeal distraction can address the deformity at this level (43), this technique has not gained popularity because of concerns of intra-articular pin placement, associated discomfort, inability to use in young children and unpredictable future growth at that physis. Few authors have reported on physeal bar resections with mixed results (2,36,42). The technique of hemiepiphyseodesis is well described in the standard texts and involves either ablation of the lateral half of the distal femur and/or proximal tibial and fibular growth plates or temporary cessation with extraperiosteally placed hardware such as staples or plates. The hemiepiphyseodesis option offers the advantages of being technically straightforward with rapid postoperative recovery and thus is quite appealing for mild cases of bilateral adolescent Blount's. However, the need for close follow up is crucial to monitor angular correction and leg length-discrepancy and avoid under or over correction at skeletal maturity.

Medial tibial plateau elevation is recommended in a select group of individuals over six years of age with advanced stages of early onset Blount's disease and dynamic varus instability (Fig. 4). Radiographic confirmation of significant depression of the posteromedial tibial plateau is done preoperatively with advanced imaging studies such as MRI and three-dimensional CT scans (38). Several authors have described either an intraepiphyseal (10,13,40) or

transepiphyseal osteotomy (36–39,41) hinging at the articular cartilage of the intercondylar notch with insertion of a structural bone graft. Care should be taken to simultaneously correct the posterior depression of the medial plateau by inserting a larger portion of the graft posteriorly. It is imperative to perform a lateral proximal tibial and fibular epiphyseodesis at the same time to prevent recurrent deformity. However, the epiphyseodesis will lead to future leg-length discrepancy in a young child. The shortening may be addressed by an appropriately timed contralateral epiphyseodesis (41) or a metaphyseal tibial lengthening, especially if a secondary metaphyseal tibial deformity exists (36,38).

FUTURE DIRECTIONS

There is a growing epidemic of childhood obesity, especially in the western world (61), which is probably going to impact the incidence and severity of disorders such as Blount's disease. Besides employing means to avoid obesity in children, screening high-risk groups, such as has been done for other pediatric conditions, like developmental hip dysplasia and adolescent idiopathic scoliosis, may be beneficial. Although there have been significant advances in imaging, deformity analysis and limb realignment strategies, the underlying etiology of Blount's disease still remains an enigma. Why only certain individuals get this disease, what explains unilateral or asymmetric limb involvement, and why certain children following appropriate realignment have recurrent deformities, remains largely unknown. The genetic and molecular basis of Blount's disease along with as of yet unidentified environmental factors that may be responsible for this disorder need further investigation.

ACKNOWLEDGMENT

The author appreciates the technical assistance of Dr Caixia Zhao and Emily McClemens (PA-C) in preparation of this manuscript.

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Part IX TUMOR

37 Reconstruction after Tumor Resection

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INTRODUCTION

Prior to the 1970s, amputation was the only choice of treatment for sarcoma of the extremities. Limb salvage surgery became possible with advances in medical imaging, chemotherapy, and implant technology (1–4). Modern chemotherapy helps to minimize surgical margins, allowing preservation of the maximum amount of healthy tissue without compromising limb function (4,5). Limb salvage surgery becomes even more important nowadays when a long-term survival is anticipated in patients with sarcoma, and especially with osteosarcoma (1,5–7).

Prostheses, autoclaved autografts, allogenic osteoarticular grafts, or combination of prosthesis and intercalary reconstruction with allografts, autografts (free or vascularized), and spacers normally achieve joint reconstruction in limb salvage surgery. However, the limb function often remains limited and deteriorates over time. Complications such as infections, nonunion of grafts, and bone resorption could eventually lead to amputation (1–4).

An ideal reconstruction method should have biological affinity, resistance to infection, and sufficient biological strength and durability. This should subsequently reduce the incidence of complications. Distraction osteogenesis, established by Ilizarov (8), allows growth of living bone tissue with excellent biomechanical properties. We have used the Ilizarov method reconstructive procedures after tumor excision and have established a link between bone tumor surgery and distraction osteogenesis (7,9,10).

Clinical Evaluation

In cases of juxta-articular osteosarcoma, normal function of the limb could be preserved using the method of distraction osteogenesis in combination with effective chemotherapy and preservation of the epiphysis, joint surface, ligaments, and neurovascular structures. The bony defect is reconstructed with the patient's own living bone tissue regenerated by distraction osteogenesis. This technique is the most conservative method of limb salvage surgery presently available for juxta-articular osteosarcoma in both pediatric and adult patients (7,9–12).

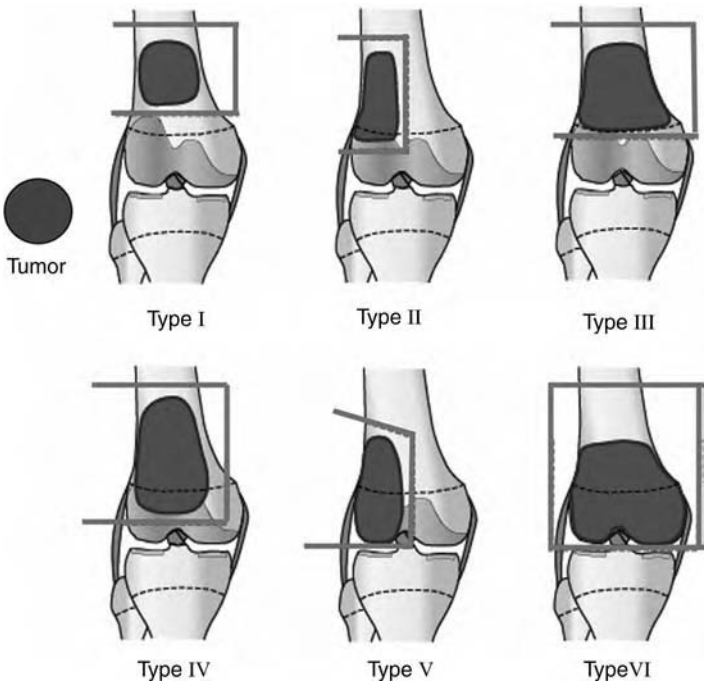
Effective preoperative chemotherapy and adequate radiological evaluation by magnetic resonance imaging are necessary to preserve the joint and healthy structures as much as possible for high-grade malignant tumors. To facilitate decision-making for reconstruction in cases of juxta-articular osteosarcoma around the knee joint, we developed a new system for classification of tumor excision as described below (see the section Classification). Cases indicated for joint-preserving tumor excision should have tumors of Types I to IV according to our classification system, not more than 15 cm in length (resulting in a treatment time of less than one year), and with a good response to chemotherapy. Distraction osteogenesis can be applied even in cases with Types V and VI tumors combined with arthrodesis.

The crucial condition for the success of this technique is the correct indication. This treatment offers good prospects for patients with malignant bone tumors, especially osteosarcoma, which responds well to chemotherapy. Early detection of osteosarcoma should allow larger numbers of patients to be good candidates for treatment using this technique with a good prognosis for survival and limb function. Lung metastases should be considered a relative contraindication for use of this technique.

Classification

Table 1 Resection Types and Decision-Making Regarding Reconstruction wtih Distraction Osteogenesis

Type I	Tumor has metaphyseal location within 2 cm from the growth plate. This tumor can be excised and the bone defect restored by diaphyseal reconstruction using distraction osteogenesis.
Type II	Tumor has metaphyseal location with extension to less than half of the epiphyseal growth plate. Excision of half of the growth plate is mandatory. There is a possibility of joint preservation and reconstruction of the bone defect by distraction osteogenesis (metaphyseal reconstruction). Joint deformity due to unequal growth may develop.
Type III	Tumor has metaphyseal location with extension to the whole growth plate. For these patients, the growth plate is fully excised, and bone reconstruction can be achieved through distraction osteogenesis (metaphyseal reconstruction). However, young patients may develop a limb length discrepancy.
Type IV	The metaphyseal tumor extends through the growth plate into part of the epiphysis at least 10 mm from the joint line. Joint preservation and bone reconstruction by distraction osteogenesis (subarticular reconstruction) is still possible, but limb shortening is anticipated in children.
Type V	The tumor extends into less than half of the epiphysis. There is no possibility for preservation of the whole joint. Options for reconstruction include epiphyseal reconstruction or arthrodesis.
Type VI	The tumor extends into more than half of the epiphysis. For these patients, the available options for reconstruction include arthrodesis using distraction osteogenesis.



Treatment Options

Reconstruction techniques using distraction osteogenesis depend on the location of the tumor and the amount of the excised bone (Table 1).

Joint preservation reconstruction includes diaphyseal (Fig. 1), metaphyseal (Fig. 2), subarticular reconstructions (Fig. 3), and epiphyseal reconstruction. The function of the adjacent joint can be normalized permanently, which is considered to be the best type of conservative surgery at present, i.e., joint preservation and reconstruction using distraction osteogenesis. Joint sacrificing reconstruction refers to arthrodesis (7).

Skeletal defects are restored by shortening-distraction (Fig. 4), bone transport (Fig. 5), or these methods combined with intramedullary nailing. Distraction combined with intramedullary nailing should be performed to reduce external fixation time, if feasible (7).

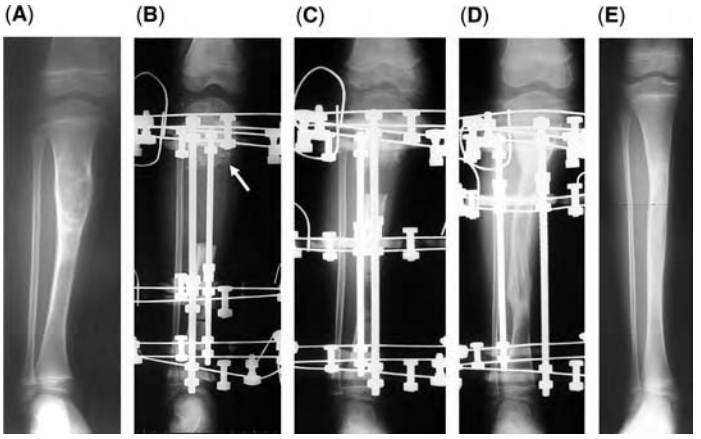


Figure 1 Type 1 diaphyseal reconstruction. (**A**) A six-year-old boy showed a recurrent juvenile adamantinoma in the midshaft of the right tibia. A 7.2 cm en bloc tumor excision was performed. (**B)** The diaphyseal defect was reconstructed by bone transport using an Ilizarov apparatus. Allogenic cancellous bone was grafted to the expected docking site. (**C**) Radiograph during bone transport. (**D**) Radiograph after completion of transport. The cancellous allograft was compressed. Good regeneration was seen. (**E**) Full remodeling of the tibial midshaft was seen six years after the operation. Neither tumor recurrence nor deformities were observed.

Reconstruction by distraction osteogenesis has clear advantages over other techniques. This reconstruction is biological and is expected to be permanent, without late complications associated with endoprostheses or allografts. This technique is not only versatile and applicable for most tumor topographies but also costs less than the other methods in general use. Moreover, the growing part of the bone could be preserved ensuing normal limb growth in some cases. And, when shortening does occur, this can be resolved with limb lengthening.

In our experience, postoperative chemotherapy had no significant hazardous effects on bone regeneration by distraction osteogenesis. We have previously confirmed that although

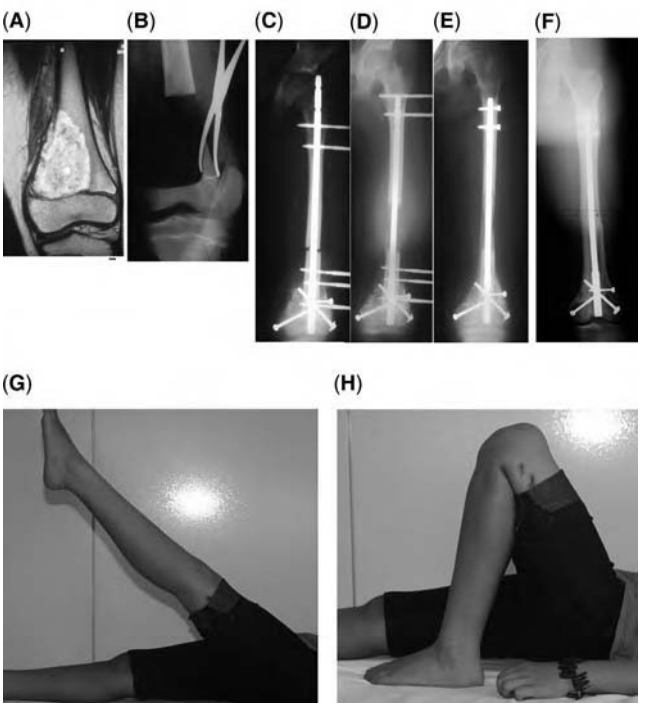


Figure 2 Type 2 metaphyseal reconstruction (**A**) A nine-year-old girl showed conventional osteoblastic osteosarcoma in the left distal femur. (**B)** The epiphysis and lateral metaphyseal cortical wall were preserved as preoperative chemotherapy was shown to be very effective by means of radiological evaluation tools. (**C)** The bone defect measuring 7 cm after tumor excision was shortened acutely and an intramedullary nail was inserted in a retrograde manner. A unilateral fixator was set with half-pins, which were inserted posterior to the intramedullary nail. Then, a multiple drill hole osteotomy was performed in the diaphysis. (**D**) Lengthening of 9 cm was performed (2 cm overlengthening) because leg length inequality was anticipated in the future. (**E**) After completion of distraction, the fixator was removed and proximal transfixation screws were inserted. A femoral extender device was placed on half-pins to maintain the distracted length. (**F**) The periarticular structure of the distal femur was well remodeled eight years after the operation. (**G**) The patient is able to extend the knee joint perfectly. (**H**) The patient is able to flex the knee joint without any problems and enjoy full athletic activities.

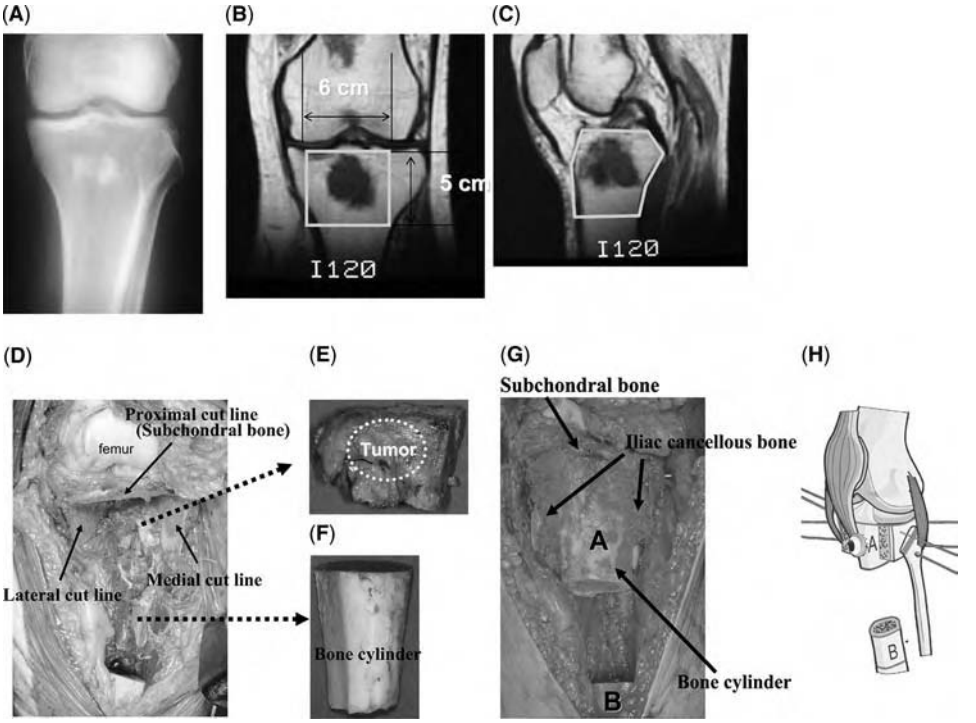


Figure 3 Type 4 subarticular reconstruction. (**A**) A 43-year-old woman presented with conventional osteoblastic osteosarcoma in the proximal tibia. (**B**) Resection margin on T1-weighted magnetic resonance imaging (MRI) (*coronal view*). (**C**) Resection margin on T1-weighted MRI (*sagittal view*). (**D**) Intraoperative view after tumor resection. The articular surface was preserved together with lateral and medial cortical walls, which was beneficial for stabilization of the subarticular part. (**E**) Resected tumor specimen. (**F**) Bone cylinder taken from the diaphysis. (**G**) The bone cylinder was fixed to remained epiphysis with lateral and medial cortical walls. (**H**) This scheme shows the patella tendon reattached to the bone cylinder with a spike washer and a screw. Insertion of multiple wires was required to stabilize the periarticular structure. (**I**) Radiograph before bone transport. (**J**) Radiograph after bone transport. An iliac bone graft was performed at the docking site immediately after completion of transportation. (**K**) Excellent bone formation and remodeling (corticalization) were observed nine years after surgery. (**L**) The patient is able to extend the knee joint. (**M**) The patient is able to flex the knee joint and enjoy full athletic activities.

chemotherapy decreases the regional blood flow, distraction osteogenesis increases regional blood flow and overall, it can be maintained within or above the normal range (13), leading to good bone regeneration and protection against infection. Jarka et al. evaluated the effects of methotrexate on distraction osteogenesis in rats (14). They showed through radiological, histological, and chemical tests that methotrexate did not have a serious detrimental effect on distraction osteogenesis. Recently Subasi et al. studied the effects of a chemotherapeutic regimen consisting of high-dose methotrexate and citrovorum factor rescue, adriamycin, and a combination of bleomycin, cyclophosphamide, and dactinomycin on distraction osteogenesis in a rabbit model (15). Their results were that the use of chemotherapeutic agents had no significant negative effect on distraction osteogenesis. However, care should be taken in extrapolating data obtained in animal models to humans (16). Accumulation of clinical data will confirm the chemotherapeutic effects on distraction osteogenesis in more detail.

Surgical Techniques

The operative technique consists of adequate en bloc tumor excision with preservation of the epiphysis, articular surface, and the maximum amount of healthy tissue. Then, an external fixator is applied. Wires or half pins are usually put in place, two to three for each ring and three to five for each periarticular ring.

Bone regeneration is accomplished by either bone transport or shortening distraction techniques.

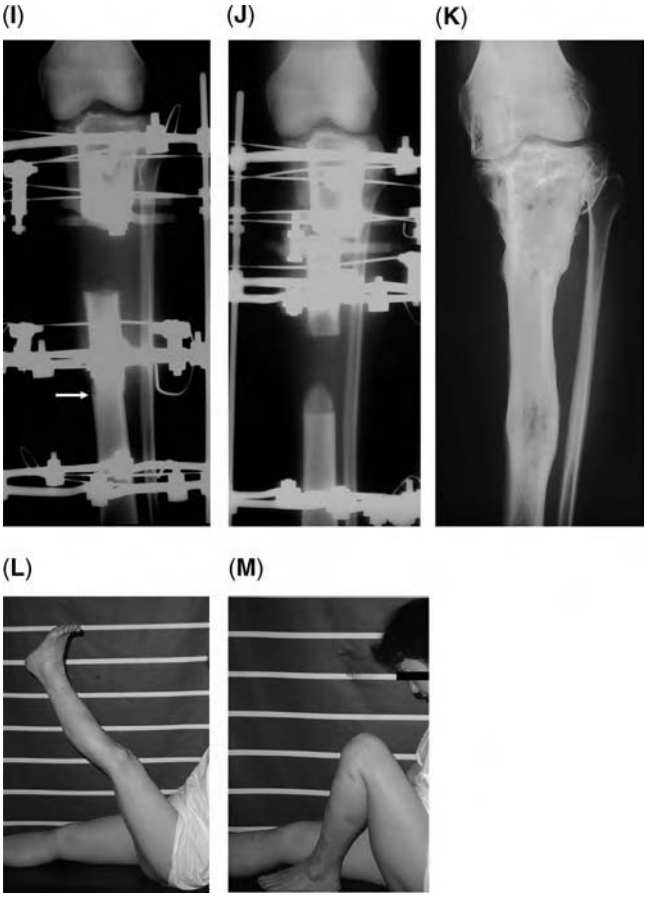


Figure 3 (*Continued*)

Shortening Distraction

Acute shortening of up to 10 cm in the femur and 5 cm in the lower leg is possible without compromise of wound closure and neurovascular insult. Caution must be exercised. Distraction can be carried out at the same site, or at a separate osteotomy site in cases of metaphyseal lesions in order to obtain good joint motion (Fig. 4). When thick portion of the epiphysis remains after tumor excision, a unilateral fixator can be used for distraction. When the

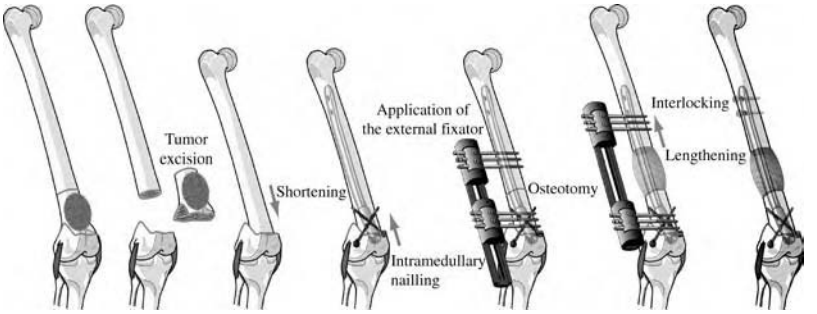


Figure 4 Shortening-distraction: Defects of less than 15 cm of the femur and 5 cm of the tibia can most often be shortened. Then, distraction is initiated at a separate diaphyseal osteotomy site after stabilization of the periarticular part. When a thick epiphysis remains after tumor excision, a unilateral fixator can be used for distraction. When the epiphysis is thin, a ring fixator, such as an Ilizarov apparatus, is convenient for stabilization of periarticular structures.

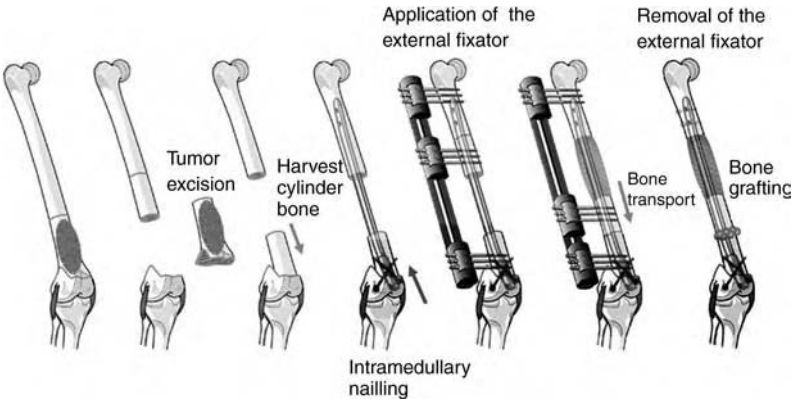


Figure 5 Modified bone transport: The periarticular structure should be stabilized immediately to prevent joint contracture. When it is impossible to shorten the defect, a bone cylinder is taken from the diaphysis and fixed to the remaining epiphysis. The newly created diaphyseal defect is reconstructed with bone transport. The external fixator should be removed after maturation of the regenerate to prevent turning back of the transported segment. Use of a ring fixator is recommended when the stability between the epiphysis is preserved and the bone cylinder is insufficient.

epiphysis is thin, a ring fixator, such as an Ilizarov apparatus, is convenient for stabilization of periarticular structures.

Modified Bone Transport

The periarticular structure should be stabilized immediately to prevent joint contracture. When only a thin epiphysis or articular surface is preserved, it is impossible to shorten the defect. A free bone cylinder should be taken from the diaphysis and fixed to the remaining epiphysis in order to stabilize and reconstruct the periarticular structure (Fig. 5). The newly created diaphyseal defect is restored by bone transport. The external fixator should be removed only after maturation of the regenerate to prevent springing back of the transported segment.

When bone transport is completed, bone grafting at the docking site is necessary to promote union at this site. In pediatric cases, cancellous allograft is used in advance at the docking area prior to initiating the transport (Fig. 1). Intramedullary nailing could be used, if feasible, to reduce the external fixation time. In these cases, a relatively thin intramedullary nail should be inserted allowing introduction of wires and half-pins.

Achieving stability of the periarticular structure is essential in order to begin immediate postoperative rehabilitation, including joint mobilization and muscle exercises. When joint stability is insufficient despite use of multiple wires or screws, additional fixation by spanning the joint should be performed.

Full weight bearing is usually allowed on the next day after surgery. It is delayed for three and four weeks in cases when soft-tissue repair has been performed, such as patella tendon or collateral ligament repair.

Distraction is initiated approximately one to two weeks after the operation using the rate of 0.25 mm two or four times a day, which could be adjusted according to the degree of bone formation. In order to obtain good regeneration, the rate of distraction should be slower in patients with high-grade tumors who have undergone pre- and postoperative chemotherapy because the bone marrow is suppressed by the chemotherapeutic agents. A distraction speed of 0.5 mm/day is maintained for three to six weeks until good bone formation is recognized on radiographs (Figs. 3–5).

Our Clinical Experience

Thirty-six patients have undergone reconstruction with distraction osteogenesis after en bloc tumor excision at Kanazawa University Hospital. External fixation index (EFI), obtained by dividing external fixation time by the length of the regenerate, was 44 days/cm in the

aggressive benign tumor group ($n = 8$), 44 days/cm in the low-grade tumor group ($n = 4$), 21 days/cm in the low-grade tumor combined with intramedullary nailing group ($n = 4$), 47 days/cm in the high-grade tumor group ($n = 15$), and 24 days/cm in the high-grade tumor combined with intramedullary nailing group ($n = 5$). Intramedullary nailing can reduce the external fixation time, resulting in a reduction of EFI.

Complications

There were 16 complications in our series (three fractures after frame removal, two deep infections, two equinus contractures, two skin invaginations, three delayed consolidations, one premature consolidation, one peroneal nerve palsy, one skin necrosis, and one subluxation of the fibular head). Nine complications were treated with additional surgery (reapplication of external fixator for one fracture, resection-shortening-distraction for one deep infection, skin plasty for two skin invaginations, bone grafting for three delayed consolidations, repeat osteotomy for one premature consolidation, and reduction-fixation for one subluxation of the fibular head).

Four (50%) of eight patients in the benign tumor group, one (13%) of eight in the low-grade tumor group, and 9 (45%) of 20 in the high-grade tumor group had superficial wire- or pin-tract infection. More than 90% of patients with wire- or pin-tract infection were treated with intravenously administered antibiotics. Wires or half-pins were removed in four patients. In the high-grade tumor group, eight patients had wire- or pin-tract infection after completion of postoperative chemotherapy and one other patient had such infection immediately after surgery, before initiation of postoperative chemotherapy. All infections were controllable conservatively. There were no cases of infection during the postoperative chemotherapy period alone in the high-grade tumor group.

Future Directions

Further development of this innovative procedure would address the promotion of faster bone-formation to reduce treatment time. The results of our experiments in rabbits indicated that injection of osteoblast-like cells into the center of distraction regenerate seems to shorten the consolidation phase (17). In addition, bioengineering with bone morphogenic proteins and other growth factors and gene therapy for osteoinduction have provided promising results and these procedures may be feasible for orthopedic surgery in the near future (18).

Bone transport with frozen devitalized bone has now been shown to be feasible in both the rabbit model and in a human (19). In the near future, it may be possible to reconstruct extensive defects by bone transport utilizing devitalized bone or artificial bone substitute combined with some growth factors or osteoprogenitor cells and an internal lengthening device.

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38 Reconstruction in Ollier's Disease

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INTRODUCTION

Multiple enchondromatosis, or Ollier's disease, is a nonhereditary condition, characterized by the presence of benign cartilage lesions within the long bones. The term Ollier's disease refers to unilateral involvement only. The disease is a dyschondroplasia rather than a true neoplasm. When an association with multiple hemangiomas is present, the syndrome is known as Maffucci's disease.

Multiple enchondromatosis appears in early childhood with deformities and distortion of the longitudinal growth of the bones, mainly affecting the legs.

Ollier's disease probably is attributable to a failure of the normal enchondral ossification resulting from the proliferation of ectopic islands of chondroid tissue, or to the incapacity of the epiphyseal plate to become mature, causing residual chondroid proliferation in the bones. As a consequence, bowing and growth distortion occur with resulting deformities and leg length discrepancy (LLD) (Fig. 1).

Conventional surgical treatment is not feasible in many patients with Ollier's disease, because the disease weakens the affected bone, as well as making it deformed and short. Bone stabilization is very difficult to obtain using internal fixation in these cases.

The Ilizarov method is very effective as far as mechanical fixation is concerned. It allows gradual correction of the anatomic and the mechanical axis of a limb with simultaneous lengthening (1–3).

CLINICAL EVALUATION

Lesions appear and grow before puberty, usually until the second decade of life (4). Sometimes the first symptom could be swelling of the fingers in a young child (5). Spontaneous remodeling into a normal bone could be observed frequently. Fractures through affected areas also could occur (4).

Enchondromas are usually located in metaphyseal areas in close proximity to, or in continuity with growth plate, causing various deformities and LLD. Patients with enchondromatosis are at risk of potential malignant transformation (6). Also the association of enchondromatosis (especially in Maffucci's syndrome) with chondrosarcomas and other malignancies has been reported (7). Some patients can develop a severe deformity, which is more common at the knee and ankle. Also patients can present with decreased knee mobility at the onset of the disease.

Plain radiographic examination is used to evaluate chondromas and deformities. However, the panoramic radiograph is the most important examination for determining the mechanical and the anatomic axis, as well as the amount of LLD accurately.

The authors treated 10 patients with Ollier's disease (three males, seven females) who were 6 to 40 years of age (mean age, 16 years). The left side was affected in six patients and the right side was affected four. The preoperative LLD ranged from 2 to 18 cm with a mean of 6.9 cm.

A varus and antecurvatum deformity of the femur was observed in four patients, a valgus deformity of the femur in two patients, a valgus and retrocurvatum deformity of the

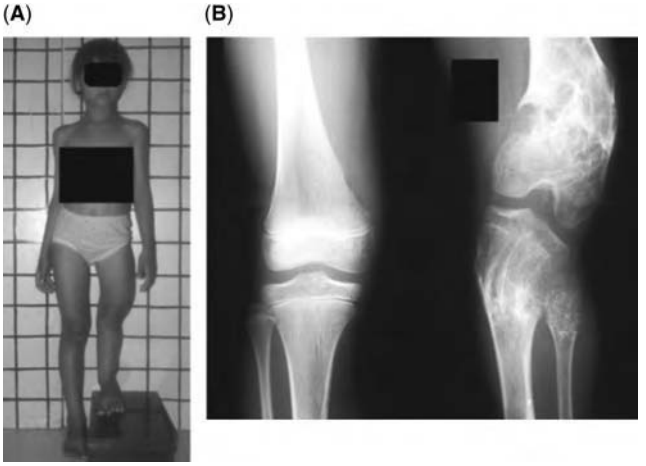

Figure 1 (**A**) Photograph of an 11-year-old girl taken at the onset of the disease. The patient presented with a lower limb discrepancy on the left of 18 cm, a varus and antecurvatum deformity of the femur, and an externally rotated valgus tibia with retrocurvatum. (**B**) Initial radiograph showing shortened femur and tibia on the left and chondromas (in the proximal and distal femur as well as in the tibia).

tibia in four patients, and a valgus and antecurvatum deformity was observed in one patient. Three patients had decreased knee mobility at the onset of the disease. In six patients, previous operations using other techniques failed to correct bone and joint deformities (Table I).

TREATMENT OPTIONS

The classical treatment is bone curettage and grafting of the femur and tibia lesions, which may result in even more severe deformities. Osteotomies are done to correct deformities. However, bone stabilization is difficult to obtain using internal devices. The presence of weak bone and enchondromas preclude internal fixation. The presence of multiple areas of abnormal bone and the difficulty in obtaining normal bone for grafting could be also a problem.

Table 1 Clinical Experience in a Series of Patients with Ollier's Disease[a]

	Age (Years)[b]	Site	Leg Length Discrepancy Before and After the Ilizarov (cm); (Combined)[b]	Angular Deformity Before and After the Ilizarov (Degrees); (Combined)[b]	Prior Surgery[b]	Duration of Treatment (Months)[b]
1	9	Femur/tibia	5.0/0.0	9/0	Curettage and grafting	10
2[c]	11	Femur/tibia	18.0/10.0	20/0	–	17
3	40	Tibia	2.0/0.0	10/0	–	6
4[d]	10	Femur/tibia	13.0/4.0	70/6	Curettage and internal fixation	11
5[c]	12	Femur/tibia	8.5/4.0	20/5	Curettage and grafting	22
6	28	Tibia	3.0/0.0	28/0	Curettage + osteotomy and internal fixation	8
7[c]	9	Femur	4.0/1.0	10/0	–	17
8[c]	23	Humerus/tibia/femur	11.0/2.0	13/0	–	15
9	12	Humerus/femur	2.0/0.0	0/0	Curettage	18
10	6	Ulna/femur	3.0/0.0	0/0	Curettage and internal fixation	10

[a]Patients who were treated with the Ilizarov method.
[b]A satisfactory outcome was seen in all patients.
[c]Resulted in reuse of the Ilizarov.
[d]Resulted in reduced knee mobility.

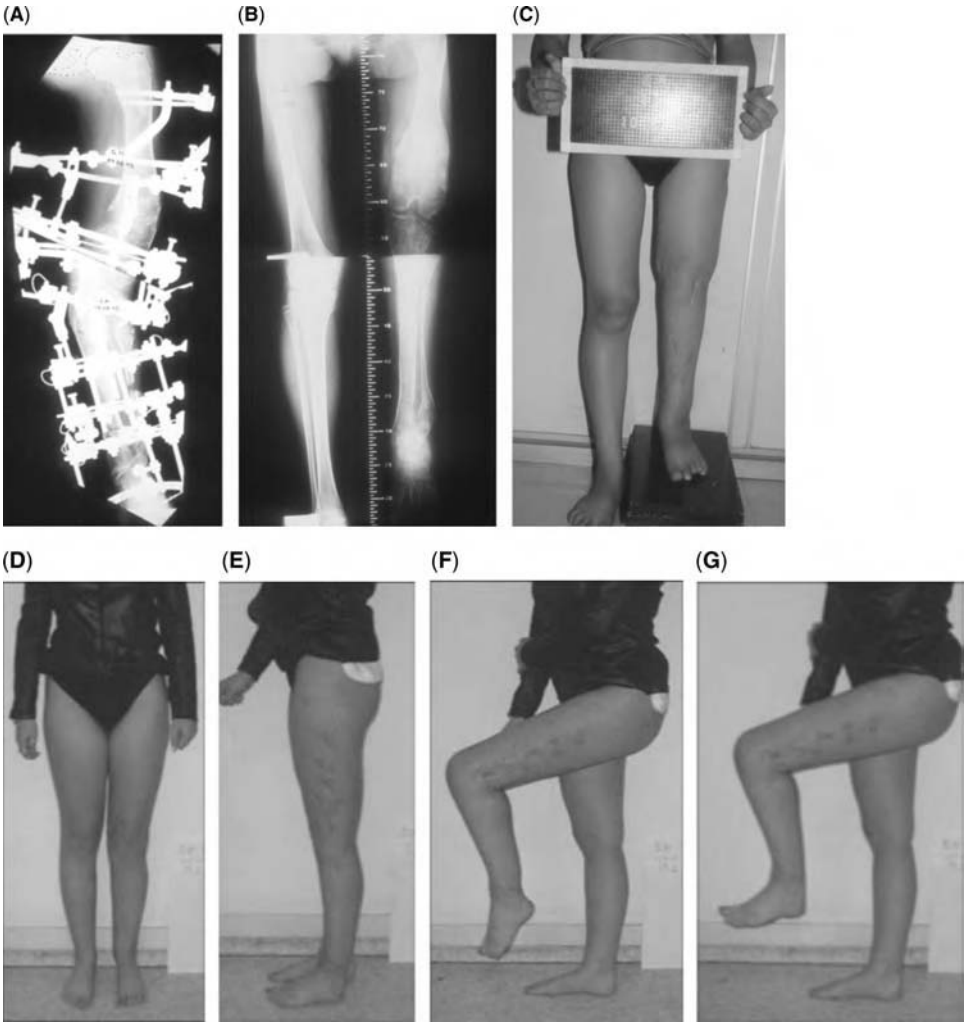

Figure 2 (**A**) Plain radiograph showing the Ilizarov device in the lower limb (at the beginning of deformity correction). (**B**) Radiograph showing the correction of deformities (and the partial correction of the leg length discrepancy). Bone neoformation is seen within the chondromas. (**C**) Photograph of the patient obtained after the first stage of the treatment. The axis of the left limb is fully corrected and discrepancy is partially corrected. (**D**), (**E**), (**F**), (**G**) The final result of elongation.

Due to these difficulties, the authors indicate the use of the external Ilizarov apparatus for the following purposes:

1. For correcting the deformities by performing the appropriate osteotomies
2. For correcting the limb length discrepancy by lengthening the bones
3. For stimulating the conversion of abnormal cartilage of the chondromas into normal bone using the distraction, with no curettage of the lesion (8–10)

The circular Ilizarov frame is used to provide bone fixation and stabilization (11–13). Because bone weakness induced by chondromas is marked, a greater number of wires and olive wires should be used to secure the bone in comparison with stabilization of normal bones. An attempt to place pins and wires in normal bone is made; however, this is often not possible. The osteotomy should be conducted using minimally invasive technique and can be done in normal bone or bone affected with Ollier's disease. In both situations, consolidation is expected to occur. The correct position of hinges is essential for the correction of deformities in order

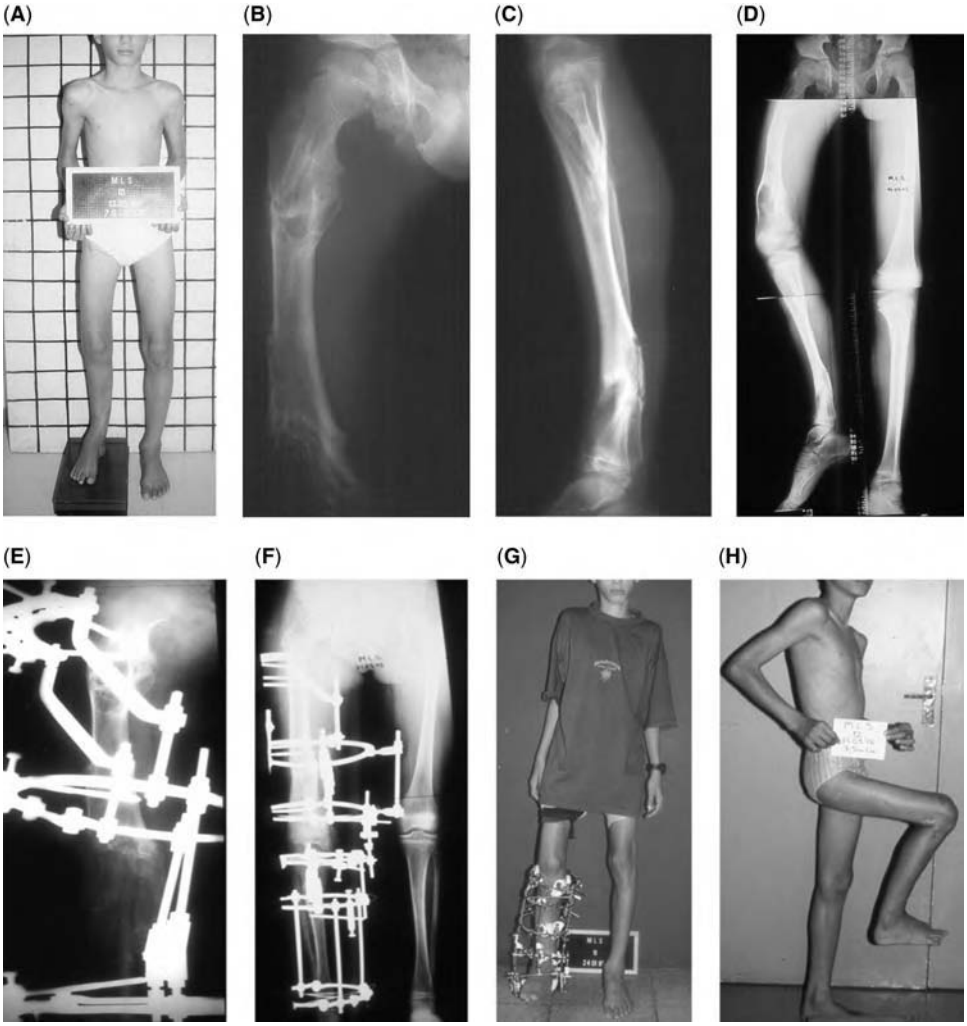

Figure 3 (**A**–**D**) Patient and Initial radiograph of a 12-year-old boy (showing limb length discrepancy, chondromas, and the varus and antecurvatum deformity of the right femur, and the valgus, retrocurvatum, and external rotation of the tibia). (**E** and **F**) Radiograph obtained during the treatment. (Deformities are corrected and chondroma cartilage is converted into apparently normal bone.) (**G**) Photograph of the patient during the elongation. (**H**) Final photograph of the patient (showing that the deformities and the discrepancy are corrected). (Good knee and ankle range of motion is shown).

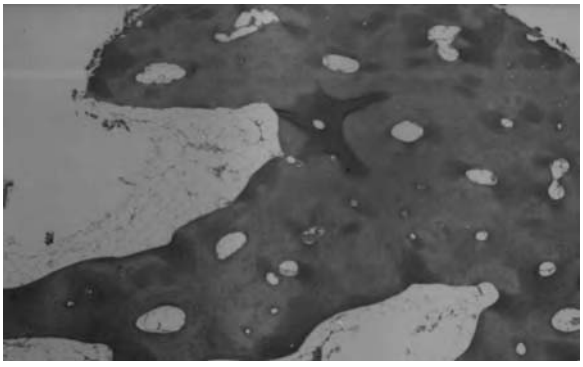

Figure 4 Photomicrograph (Hematoxylin and eosin-X200). Biopsy of the elongated bone presenting typical lamellar and marrow-bone. Cartilage lobuli, present in the enchondromatosis, are not shown in this area.

to restore anatomical and mechanical axis of the limb. Use of a monolateral device could be more comfortable for the patient when correcting deformity of the humerus and femur (Fig. 2).

Latency period should be about five to seven days to avoid premature consolidation. The lengthening rate is on an average 1 mm per day, increments of 0.25 mm every six hours (Fig. 3).

EXPERIENCE AND COMPLICATIONS

We used the Ilizarov device on eight femurs, seven tibias, two humeri, and one ulna.

In all the patients, cartilage was converted into apparently normal bone after surgery, as shown on the imaging studies. Deformities were corrected in eight patients. In two the full correction of the varus femoral deformity was not obtained (6° and 5° of deformity). In one patient, a varus procurvatum oblique plane deformity was seen in the femur. Satisfactory results were obtained in all patients. The final mean LLD was 2.1 cm (Table 1). When corrections are done in young children, there is some recurrence of deformity and LLD after long-term follow-up.

One patient had a markedly reduced mobility of the knee with a final range of 0° to 70°, which persisted even after physiotherapy. In four patients, edema and superficial infection were treated. In one patient, a femoral fracture occurred after removal of the device. This was successfully treated by the reapplication of the Ilizarov frame.

A biopsy of the lengthened area was done in two patients, when the Ilizarov apparatus was removed, 12 and 15 months after its application. The area corresponded to the diaphyseal region of the regenerate. In both cases, the histological diagnosis was normal mature bone (Fig. 4). The radiographic appearance (magnetic resonance imaging and X ray) of the elongated bone did not change over time, even at 10-year follow-up. We have seen no situations of conversion to malignancy.

FUTURE DIRECTIONS

According to imaging studies and biopsy results, abnormal cartilage could be successfully converted into a normal mature bone during distraction osteogenesis (14). This phenomenon should be investigated further and possibly used for treatment of the disease in order to prevent further deformities in children as well as to eliminate the risk of malignant transformation in adults.

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Part X UPPER EXTREMITY

39 Humeral Lengthening and Realignment

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INTRODUCTION

Of all the bones that can be lengthened in the upper body, lengthening of the humerus is perhaps the easiest and most well tolerated. While there are risks, particularly for the radial nerve, in general the complication rate is low, regenerate bone healing is predictable, physical therapy requirements are not excessive, and results are gratifying.

CLINICAL EVALUATION

Prior to lengthening the humerus, patients should undergo a thorough orthopedic evaluation of the entire upper extremity, with special emphasis on joint range of motion/stability, including shoulder, elbow, and wrist range of motion. One should also assess function and document the preoperative neurologic examination of the hand. Radiographs should include anteroposterior and lateral views of the humerus, and if indicated, lateral flexion and extension views of the elbow.

CLASSIFICATION

There are two basic scenarios where humeral lengthening is indicated: bilateral humeral lengthening in dwarfism and unilateral humeral lengthening for length discrepancy after a growth disturbance. Additionally, there may be an element of angular/rotational deformity that requires simultaneous treatment. Angular/rotational deformity without length discrepancy is also an indication for external fixation techniques for the humerus because the correction can be gradual rather than acute, with potentially less risk of acutely stretching neurovascular structures.

Achondroplasia

The most common indication for bilateral lengthening is in achondroplasia, followed by hypochondroplasia. The indication in achondroplasia is most compelling because it truly facilitates important activities of daily living such as personal hygiene after toilet, driving a car comfortably, and reaching for objects. Many older achondroplastic dwarfs have a difficult time reaching the perineum. The typical flexion deformity of the elbow may actually aid in reaching around, but the short length is an obstruction. Young people with achondroplasia typically do not have this problem because they still have enough flexibility in the thoracolumbar spine to aid in reaching, and they are usually not yet obese. However, with aging comes stiffness and stenosis of the thoracolumbar spine in adulthood and often trunkal obesity. These factors together with shortened arms block access to the perineum. We have also seen this problem in some youngsters, both obese and nonobese. Short arms make it difficult to block a fall, and we have seen one achondroplastic child with recurrent subdural hematomas from frequent blows to the head because his arms were too short to break the force of his falling. Leg lengthening in achondroplasia is also very helpful from a functional standpoint, but arm lengthening benefits the patient by giving him or her greater reach, while both sitting

and standing. Driving a car with short arms places the head and face dangerously close to the air bag, which should be disabled.

Growth Disturbance

Neonatal osteomyelitis of the upper humerus and septic arthritis of the shoulder often lead to severe shortening of the humerus, typically with well-preserved shoulder and elbow function and stability. ''Humera vara'' is the term that has been coined to describe this (analogous to coxa vara in the hip). In actuality, the typical deformity seen clinically is often more complex: In addition to varus, there may also be internal rotation and apex anterior angulation of the upper humerus. The humerus may be lengthened up to 10 cm. Lengthening improves reach, function, and cosmesis. Correction of humera vara improves shoulder range of motion and decreases abduction impingement on the acromion. The ideal solution for humera vara with humera breva is a double-level treatment: acute correction of the varus, flexion, and internal rotation at the surgical neck and simultaneous or staged gradual lengthening through a corticotomy at the lower end of the deltoid tuberosity.

Angulation Plus Shortening in Developmental Disturbances

Ollier disease and unicameral bone cysts cause a disturbance of growth of the upper humerus and sometimes the lower humerus as well. In addition to shortening, there may be large angulations. While it is true that angular deformity in the diaphysis of the humerus is well tolerated (because of the large range of motion of the shoulder joint), there can be significant cosmetic issues when angulation is greater than 40°. Distal deformities in the sagittal plane can cause apparent elbow range of motion (ROM) issues. Distal angulation in the frontal plane causes cubitus varus and valgus, which is cosmetically problematic.

TREATMENT OPTIONS

If the treatment is simple lengthening, then a monolateral fixator is quite sufficient. The pediatric LRS rail system (Orthofix, McKinney, TX, USA) works well, though in larger adults, the adult LRS rail system may also be appropriate. For angular deformity correction, one can choose acute correction, but the radial nerve is at risk. Alternatively, one can apply two 2/3 Taylor Spatial Frame (TSF) (Smith & Nephew, Memphis, TN, USA) rings with reasonable comfort for the patient and then gradually correct angulation, length, and rotation. The distal TSF ring is open anteriorly to allow elbow flexion, while the proximal ring is open medially for better clearance against the chest. Only the reference ring needs to be perpendicular to its bone segment, so it is possible to make the second (moving) ring nonorthogonal for better patient fit and comfort. We usually choose the distal ring as our reference ring. The extra small Multi-Axial Correcting (MAC) (EBI, Parsippany, NJ, USA) device may allow monolateral fixation (for better patient comfort) and still allow accurate deformity correction.

SURGICAL TECHNIQUES

Bilateral Humeral Lengthening

This can be done with two teams and two image intensifiers to save time in the operating room, although the image intensifiers may collide over the midline. Position the patient on a radiolucent table so there is no metal anywhere near the shoulders. The image intensifier itself is used as the operating surface and should be totally included in the sterile drapes. Begin by inserting a 1.8-mm wire perpendicular to the long axis of the humerus (medial humeral line) (Fig. 1A–B) and just above the olecranon fossa. The pin position should be biased in the sagittal plane slightly posterior to the midline. If the wire position is satisfactory, then drill over it with a 4.8-mm cannulated drill and insert a 6-mm external fixation pin in the hole. Use the assembled LRS rail system as a drill guide. For the pediatric LRS rail system, use the close spacing (near-near) of the three holes for the next pin and, in the proximal pin clamp, use the no. 1 and no. 3 holes (far-far). The proximal pins are placed just proximal to the deltoid tuberosity (deltoid muscle insertion). If they are placed more proximal than this, the axillary nerve is at risk. The remainder of the pins may also be placed with the cannulated wire/drill

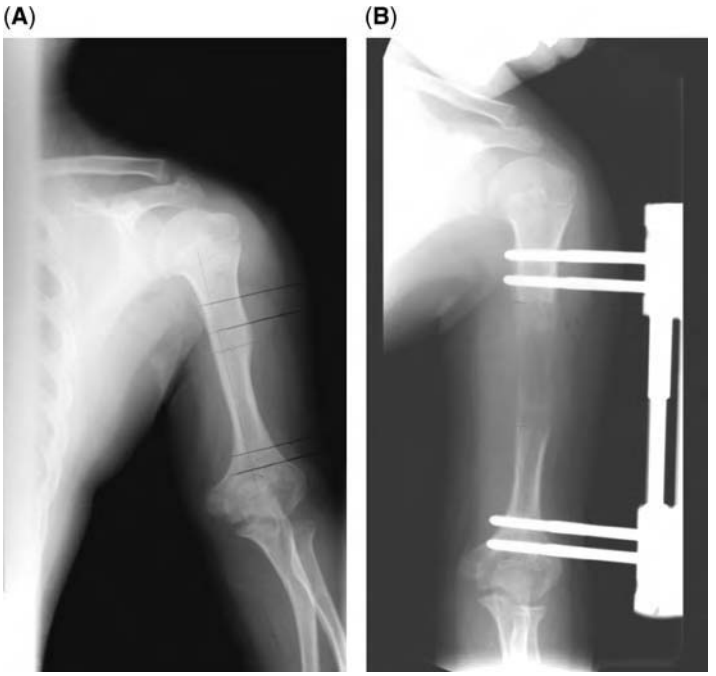

Figure 1 (**A**) Achondroplastic humerus. The long axis of the humerus is parallel to the medial cortical line on the anteroposterior view. The external fixator should be mounted with pins perpendicular to this line. (**B**) The external fixator pins have been placed perpendicular to the medial humeral line. The distal most pin is just proximal to the olecranon fossa.

technique for greatest accuracy. The pin that is most at risk for damaging the radial nerve is the one that is just above the first pin that was placed above the olecranon fossa. That pin should be placed with utmost care. There can be a challenge in applying a monolateral straight fixator to an achondroplastic humerus: The humerus is slightly serpentine in the sagittal plane. Draw a straight line on the lateral view of the humerus to identify proper pin placement (Fig. 2).

Bilateral Humeral Lengthening with Correction of Elbow Flexion Deformity

In achondroplasia with significant elbow flexion deformity, the osteotomy is placed just proximal to the pin placed above the olecranon fossa and is biased anteriorly, so it is close to the CORA (center of rotation of angulation) of the deformity. The second distal pin is applied through the center of rotation of the elbow, below the olecranon fossa. The proximal two pins are applied at the deltoid tuberosity. This pin placement mimics the serpentine angulation in the sagittal plane, so that after the corticotomy, the bone is acutely corrected (extended) and the elbow flexion deformity is corrected. This very distal osteotomy is done through a midline triceps splitting 1-cm incision using multiple drill holes and an osteotome. This technique has the distinct disadvantage of limiting the amount of length that can be achieved because the patient invariably develops an elbow flexion contracture from lengthening across the biceps and brachioradialis muscle. Additionally, the distal pin is very close to the elbow joint, so it inhibits elbow ROM. An alternative approach is to lengthen the achondroplastic dwarf in the usual fashion through a deltoid osteotomy for 7 cm. One year later, a second treatment is done through a distal osteotomy to primarily correct the flexion deformity and also gain some modest additional length, typically 3 cm.

Unilateral Humeral Lengthening

Use a monolateral fixator as described above. The osteotomy is done through a deltoid level corticotomy. Make a 1-cm incision at the anterolateral bare spot where the deltoid tendon ends. The periosteum should be minimally disturbed. Make two or three drill holes with a 4.5-mm drill bit and then use a small osteotome to complete the osteotomy. The round-shaped humeral bone generally breaks easily. Despite the relative proximity of the radial nerve, it is rare to surgically injure this nerve (Figs. 3A–E, 4A–H).

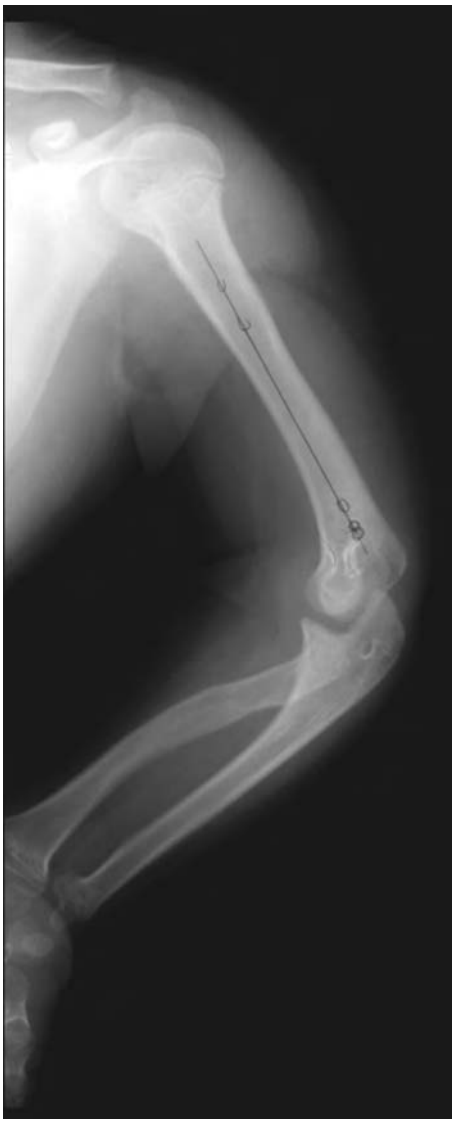

Figure 2 Achondroplastic humerus, lateral view. The bone has a scoliotic s-shaped appearance. When using a monolateral external fixator, you must plan out the pin placement to accommodate this curve. The first pin (placed just above the olecranon) should be inserted slightly posterior, so that the one above it will be contained within the bone. Inserting the proximal pins slightly posterior to the midline also helps keep the distal pins in bone.

Unilateral Humeral Lengthening with Deformity Correction

For angular deformities, we prefer to use the TSF. Usually the distal ring is a 2/3 ring open anteriorly to allow for elbow flexion. It is designated as the "reference ring" and is placed perpendicular to the distal segment. Use the same pin configuration as described for the bilateral humeral lengthening above. It is also possible to suspend pins below the reference ring by shooting them up the medial and lateral columns from the epicondyles. The proximal "moving ring" is a 2/3 ring open medially for better clearance from the chest wall. When calculating the rotary frame offset, the master tab on the proximal ring is usually directly anterior, so the anti-master tab of the distal reference ring is directly anterior, making a rotary frame offset of zero degrees (Fig. 5A–G).

COMPLICATIONS OF HUMERAL LENGTHENING

Pin Site Infection

This is the bane of external fixation treatment, but for some reason, the arm seems to be less troubled by pin infections than the leg. Hallmarks of infection are pain, tenderness, redness, and drainage from the pin site. Systemic symptoms such as fever are uncommon. All patients are given a prescription for an oral antibiotic before discharge from the hospital with instructions

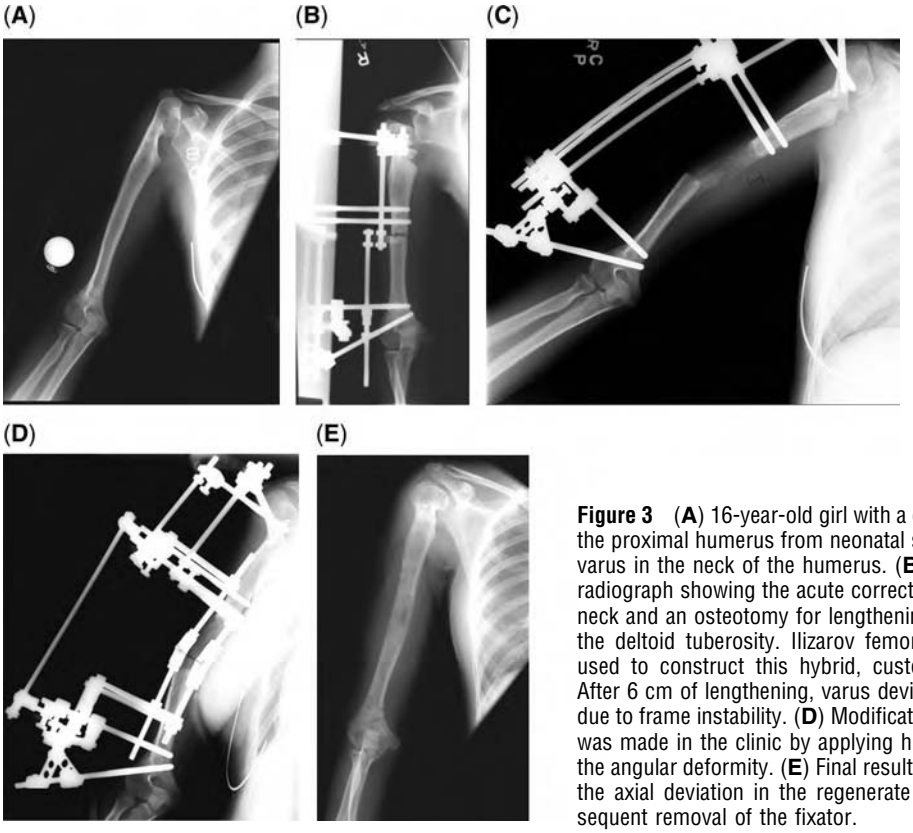

Figure 3 (**A**) 16-year-old girl with a growth arrest of the proximal humerus from neonatal sepsis. Note the varus in the neck of the humerus. (**B**) Postoperative radiograph showing the acute correction of the varus neck and an osteotomy for lengthening at the end of the deltoid tuberosity. Ilizarov femoral arches were used to construct this hybrid, custom fixator. (**C**) After 6 cm of lengthening, varus deviation developed due to frame instability. (**D**) Modification of the frame was made in the clinic by applying hinges to correct the angular deformity. (**E**) Final result after correcting the axial deviation in the regenerate bone and subsequent removal of the fixator.

on self-diagnosis and treatment of pin infection. This minimizes delay of treatment even if it leads to overtreatment in some cases. If the signs and symptoms subside, the doctor is never notified. If symptoms do not clear, then the patient is instructed to call after 48 hours. At this point, a culture is taken and, if indicated, the antibiotic changed according to the specific sensitivities of the offending organism. If there is any necrotic tissue around the pin site, it is debrided in the office. Prevention of pin infection includes minimizing pin skin motion, especially in the proximal arm, where there is more ample soft tissue. This can be achieved by dressings that apply direct pressure to the skin. Daily pin care consists of showering with soap daily and drying the pin site with a clean towel. Any crust or scab at the pin site should be picked off with forceps. Cleaning of pins with swabs and saline or peroxide is avoided if the pin site is quiet. This avoids intentionally irritating a nonproblematic pin site. The key to cleaning the pin sites is as for any wound: removal of any nonvital tissue around the pin site and washing the wound with water (shower) (Fig. 6).

Nerve Complications

Nerve problems in humeral lengthening can be divided into acute (surgical trauma) and gradual (distraction related). The predisposing factor is a tight fibro-osseous tunnel for the radial nerve because it winds around the humerus. Additional points of entrapment of the radial nerve have been well described in the hand literature. Previous scarring and trauma may be a predisposing factor. Injury to the axillary nerve can occur with very proximal pin placement in the upper humerus.

It is essential to see the patient every two weeks during the distraction phase. If available, quantitative sensory nerve testing [Pressure Specified Sensory Device (PSSD) test] is performed at the time of the follow-up. This allows detection of impending nerve problems even before they are noticed clinically. If they are detected early and the rate of lengthening is slowed, the nerve symptoms may abate and the lengthening can be continued at a slower rate. If despite slowing the lengthening the nerve problem persists, the nerve needs to be

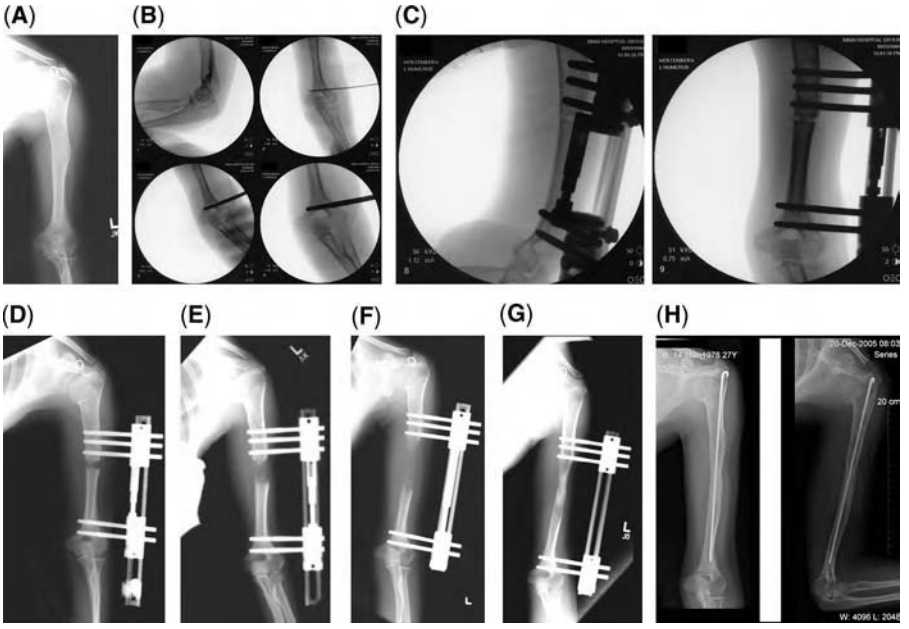

Figure 4 (**A**) 27-year-old woman who has 8 cm arm shortening in her left humerus secondary to growth arrest from neonatal sepsis of the left shoulder/proximal humerus. The distal end of the deltoid tuberosity is the ideal level for lengthening. (**B**) Intraoperative image intensifier views showing the sequence for placing the most distal half-pin. Start with a 1.8-mm wire drilled just above the olecranon fossa on the anteroposterior view and just behind the midline on the lateral view. Next, drill over the wire with a 4.8-mm cannulated drill bit, and then insert the 6-mm half-pin. (**C**) Final image at surgery. The external fixator is aligned with the medial humeral line. (**D**) Sixteen days postoperatively, the distraction gap is evident. (**E**) Six weeks postoperatively, the distraction gap measures 32 mm and new bone formation is evident. (**F**) At the end of distraction, the gap measures 75 mm. (**G**) Eleven months since the original surgery. New bone is mature but has a wasp-waist appearance in the middle of the bone. This would be at risk for fracture if the frame is removed. (**H**) Frame was removed and a Rush Rod inserted to stiffen the bone and prevent fracture.

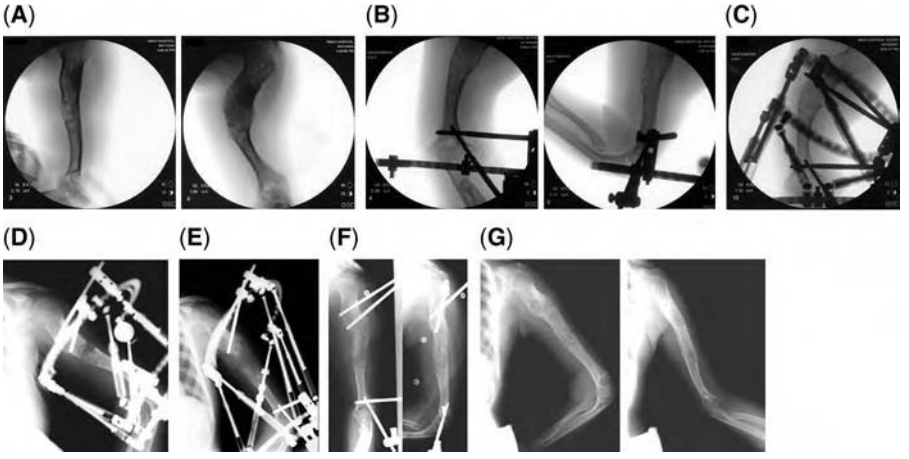

Figure 5 (**A**) 15-year-old girl with Ollier disease. There is a 60° oblique plane deformity of the humerus. With the arm rotated 90° to the plane of the deformity, it appears straight. There is also about 10 cm of shortening. (**B**) Intraoperative films made to document the centering of the distal spatial frame reference ring. The ring is perpendicular to the distal humeral segment. (**C**) At the completion of surgery, the frame mimics the valgus deformity of the humerus. The osteotomy for lengthening has been done at the level of the distal deltoid tuberosity. (**D**) After two weeks of distraction, there is already some new bone evident in the gap. (**E**) Ten weeks postoperatively, the valgus is corrected and the gap now measures 75 mm on the concave side of the deformity. (**F**) Five months after surgery, the frame is ready to be removed. Length and alignment have been restored. (**G**) Follow-up films show excellent maturation and remodeling of the new bone. Despite the fact that the osteotomy was made through an Ollier lesion, the new bone appears normal.

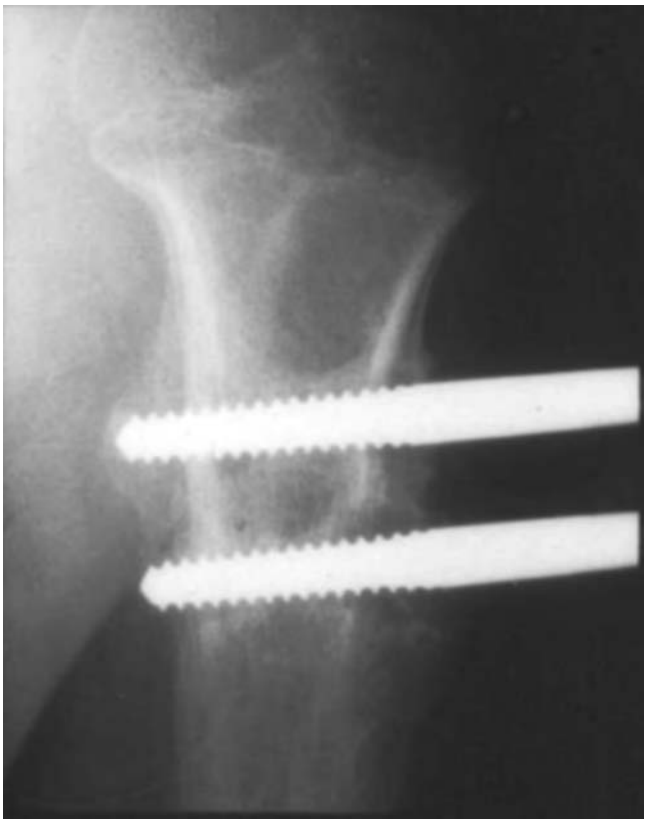

Figure 6 Osteolysis and periosteal new bone formation around pins. This indicates loosening and is frequently accompanied by infection.

surgically decompressed. Decompression of the radial nerve is done through a long incision that starts in the diaphysis laterally and extends anteriorly along the course of the radial nerve into the antecubital fossa. After the decompression, lengthening is able to continue at the same rate. The advantage of early therapeutic nerve decompression is that it permits the goal of lengthening to be achieved.

Premature Consolidation

We have observed premature consolidation of the humerus when treating two conditions: achondroplasia and Ollier disease. Patients with these conditions tend to heal regenerate bone faster than normal. Premature consolidation that is diagnosed radiographically is treated by lengthening at a faster rate (1.5 mm/day) for a few days. Sometimes this increased tension against resistance will lead to a sudden break through the regenerate bone. This is painful, causes an acute diastasis of the bone ends, and may also cause a radial nerve neurapraxia. Acute fracture of the regenerate bone should be treated by backing up the distraction and waiting a day or two before restarting the lengthening. If premature consolidation is diagnosed, the bone may be reosteotomized at a new level. It is preferable not to osteotomize through the regenerate bone because consolidation may become delayed. Also, it is technically difficult and involves greater hemorrhage to perform a repeat corticotomy through the previous regenerate bone.

Joint Contracture

Lengthening the humerus at the level of the deltoid tuberosity rarely causes any significant contracture of the elbow or shoulder. However, lengthening at the supracondylar level causes a very rapid elbow flexion contracture. If physical therapy does not resolve the contracture, then lengthening should be stopped. Another useful trick is to apply a forearm cast and attach an Ilizarov or TSF ring to the cast. This is then rigged to the humeral fixator with lengthening struts (easier in the case of a circular device) to distract the elbow joint into extension (Fig. 7).

Figure 7 Clinical photograph of the patient depicted in Figure 5A–G. During lengthening, an elbow flexion contracture developed. This was successfully managed by fabricating a custom forearm splint that incorporated spatial frame struts linked to the humeral frame to provide distraction of the elbow.

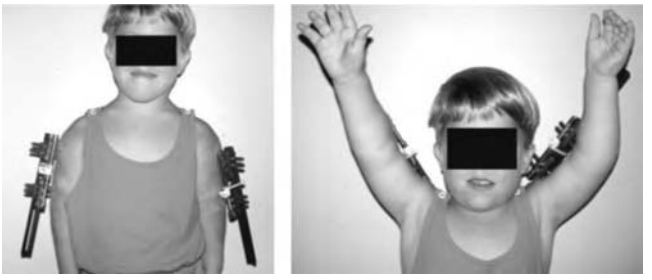

Figure 8 Humeral lengthening with external fixation is well tolerated by patients and is highly successful.

Postoperative Management

Clinical follow-up is recommended every 10 to 14 days during the distraction phase. During these visits, radiographs are obtained and sensory nerve testing may be performed in addition to the clinical evaluation by the physician. During the consolidation phase, radiographs are obtained monthly. The external fixator is dynamized during one of these visits near the end of the consolidation phase. Dynamization is the process of increasing the weight bearing taken by the bone and lessening the load taken by the external fixator. Dynamization and loading can also be accomplished by having the patient walk with a cane in the affected hand. The rods or bar of the external fixator are unloaded to allow dissipation of the lengthening forces so that the bone takes more of the weight-bearing loads. This helps the bone strengthen and thicken in preparation for removal of the external fixator.

At the end of the distraction phase before all adjustments are stopped, it is very important to ensure that the bone is straight. Measurements of joint orientation angles are performed on the radiographs to detect axial deviation. If present, the external fixator is adjusted to correct the angles to normal. If this step is skipped, the bone will heal malaligned. We routinely examine for this and correct the joint orientation angles to normal. Axial deviation can be difficult to adjust and may require creative fixator modification including mounting another more versatile fixator on top of the original pins.

FUTURE DIRECTIONS

Humeral lengthening is well tolerated with external fixators (Fig. 8). Limb lengthening has been gradually headed toward the use of internal lengthening nails. It remains to be seen if the humerus will yield to this new concept. Lengthening over nails may be a viable option as well.

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40 The Use of a Hinged External Fixation of the Elbow

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INTRODUCTION

Hinged external fixation of the elbow is used to maintain alignment of the joint and permit motion. The fixator serves as temporary scaffolding, permitting the motion of flexion and extension, while preventing dislocation. The concept of an articulating fixator about the elbow is based on the fact that normal ulnohumeral kinematics approximates a simple hinged joint. Therefore, recreating the anatomic axis of rotation with a hinged fixator allows for aligned ulnohumeral motion, thereby reducing the risk of excessive and potentially disruptive loads on healing bone or ligaments. In addition to neutralizing the joint reaction loads, distraction can also be applied for the theoretical benefit of providing separation of several millimeters after inserting the resurfacing material following distraction arthroplasty.

The most common settings for the use of this device are:

1. Acute instability following complex dislocations with fractures.
2. Delayed treatment of complex dislocations with fractures.
3. Distraction–interposition arthroplasty.
4. Adjunctive stabilization after contracture release.

Irrespective of the device used, accurate alignment of the fixator axis with the anatomic axis of the elbow is mandatory. Safe placement of the external fixation pins, avoiding neurovascular structures is mandatory. Common difficulties include cellulitis around the pin tracks, pin loosening, and loss of reduction after frame removal.

The purpose of this chapter is to provide useful guidance for the use of hinged fixators at the elbow. This chapter is not an exhaustive review of the results of treatment for these complicated cases. Instead, we have attempted to assemble a summary of our practical experience including some illustrative cases.

SPECIFIC INDICATIONS AND FIXATOR APPLICATION TRAUMA

Complex Acute Dislocations with Fractures

One of the most challenging conditions to treat in the elbow is the complex dislocation (1–5). The term *complex* refers to the combination of a dislocation and a consequential fracture of the coronoid, proximal ulna, and/or radial head (6). Importantly, this injury should neither be confused with a simple dislocation nor should the fracture be the only focus of treatment. This injury pattern is inherently unstable unless both the fracture and dislocation are adequately addressed. An inadequate surgical attempt causes continued instability and loss of fracture fixation. Hinged external fixation can maintain the reduction of both the fracture and the dislocation, while permitting motion. The most unfortunate outcome of these injuries is failure to maintain the elbow joint reduced after surgical intervention. The decision to use external fixation depends on the adequacy and stabilizing capacity of the internal fixation, age of injury, and adequacy of repair. There are also some patients, due to age or infirmity that would not tolerate these devices and a less aggressive approach may be more appropriate.

The radial head, medial coronoid (proximal ulna), and lateral ligament complex—all require direct repair or restoration. The need for hinged external fixation is a contingency decision that should be based on:

- Security of the repairs—bony and ligamentous.
- Immediate stability as judged on the table.

If the fixation of the coronoid is a wire suture or single screw, this should be regarded as tenuous for at least six to eight weeks. Hinged external fixation can be used to neutralize the load on these repairs and to preclude catastrophic redislocation and mechanical failure of the repairs.

Delayed Treatment of Complex Dislocations with Fractures—Complex and Complicated

If treatment has been delayed and the joint dislocated for more than two to three weeks, the necessity of hinged fixation to supplement the repair becomes more imperative. In these settings, the delay may occur after an initial attempt at surgical stabilization, but subsequent dislocation occurred and may have not been noted (Fig 1, 2A and B).

The added time after injury or attempted surgery increases the stiffness and overall scarring around the elbow. The dislocation becomes fixed and in order to reduce the ulnohumeral joint, the entire distal humerus must be released and the surfaces of the joint cleared of fibrotic tissue to achieve reduction. This more-extensive stripping further destabilizes the joint in the immediate postoperative period and the risk of subsequent redislocation is very high without external fixation to protect the reduction and healing tissues.

There are instances where the joint has been dislocated for months. In this instance, the patient and surgeon should be prepared to consider interposition arthroplasty, because the surfaces may be too damaged to support open reduction alone. The long-term results of these cases can be disappointing (see Fig. 2B).

Distraction Arthroplasty for Post-traumatic or Inflammatory Arthritis

Distraction–interposition arthroplasty for arthritis is controversial but often the only alternative given the state of implant arthroplasty for the elbow in the younger active patient (7). There is not enough data or followup at this time, but the senior author has used this technique in younger patients with single-joint painful elbow arthritis with success (Fig 3). This group of patients is small and the advent of newer agents for the treatment of inflammatory arthritis has changed the course and pattern of disease.

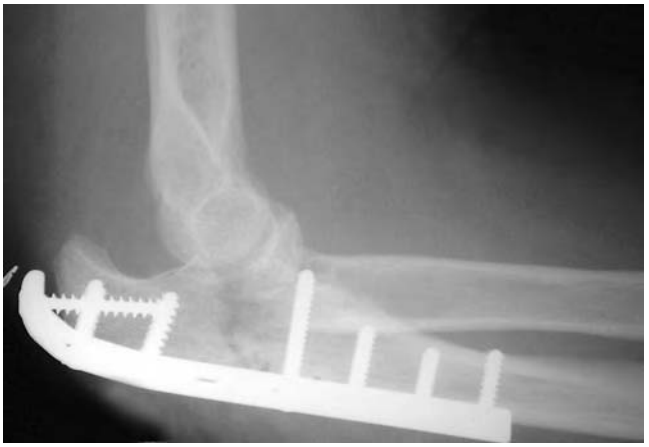

Figure 1 A 63-year-old patient several weeks after repair of fracture of ulna and radial head. The dislocation of the elbow persists. *Source*: Robert N. Hotchkiss MD, Hospital for Special Surgery, New York, New York, U.S.A.

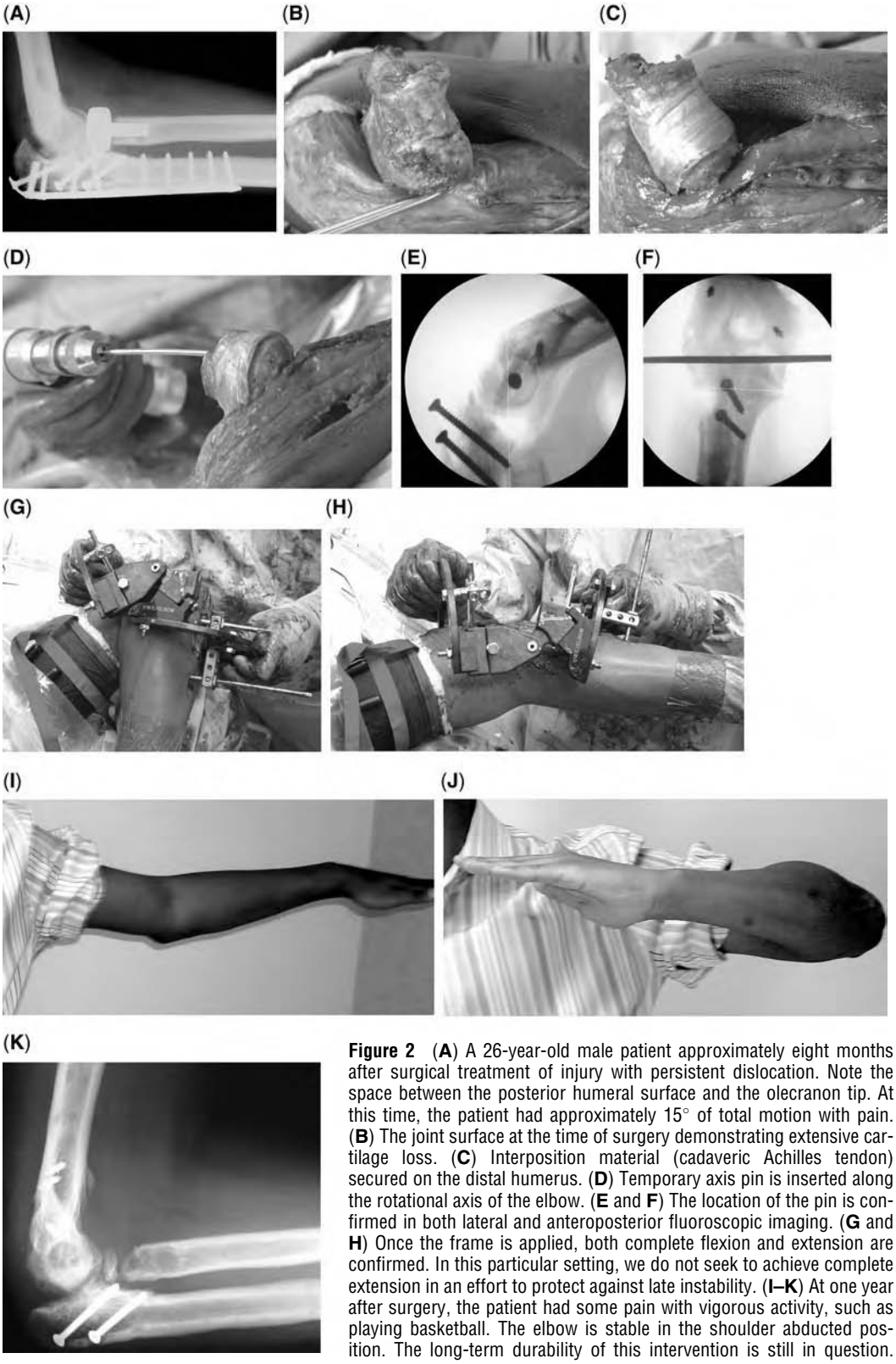

Figure 2 (**A**) A 26-year-old male patient approximately eight months after surgical treatment of injury with persistent dislocation. Note the space between the posterior humeral surface and the olecranon tip. At this time, the patient had approximately 15° of total motion with pain. (**B**) The joint surface at the time of surgery demonstrating extensive cartilage loss. (**C**) Interposition material (cadaveric Achilles tendon) secured on the distal humerus. (**D**) Temporary axis pin is inserted along the rotational axis of the elbow. (**E** and **F**) The location of the pin is confirmed in both lateral and anteroposterior fluoroscopic imaging. (**G** and **H**) Once the frame is applied, both complete flexion and extension are confirmed. In this particular setting, we do not seek to achieve complete extension in an effort to protect against late instability. (**I–K**) At one year after surgery, the patient had some pain with vigorous activity, such as playing basketball. The elbow is stable in the shoulder abducted position. The long-term durability of this intervention is still in question. The x-ray of the joint does not show subsequent complete dislocation. *Source*: Robert N. Hotchkiss MD, Hospital for Special Surgery, New York, New York, U.S.A.

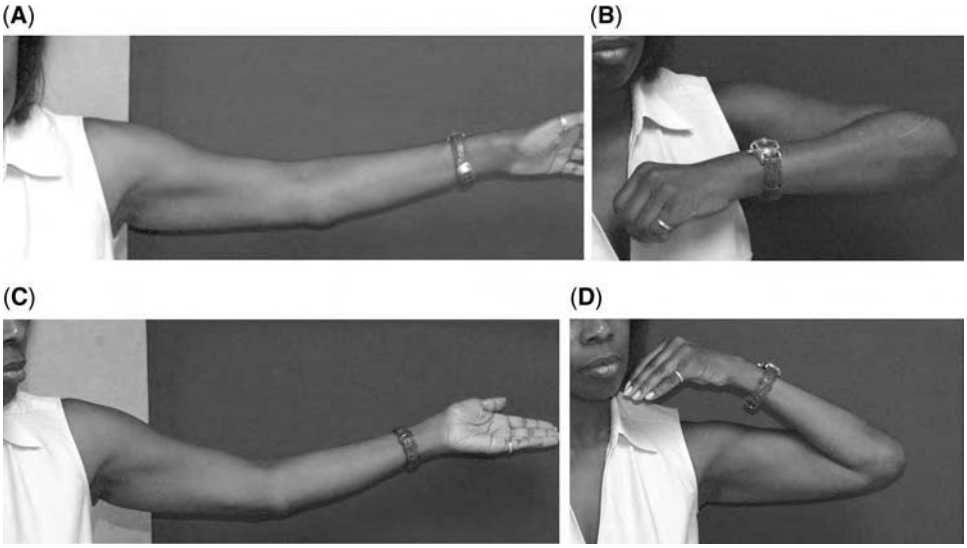

Figure 3 A patient in her mid-30s with rheumatoid arthritis requiring anti-tumor necrosis factor medication. Given her age and level of activity, she was not a candidate for implant total elbow arthroplasty. The images show her elbow motion and stability nearly eight years after interposition arthroplasty. She had little pain and only on occasion of lifting objects greater than 20 lb. *Source*: Robert N. Hotchkiss MD, Hospital for Special Surgery, New York, New York, U.S.A.

Adjunctive Stabilization Following Post-traumatic Contracture Release (Without Interposition)

Because greater experience has been gained in treating the stiff elbow, the use of hinged external fixation for adjunctive stabilization or as an instrument for passive motion has become more limited (8,9). The proper surgical exposure and capsular excision usually permits immediate passive motion therapy without fear of dislocation. In addition, the use of the fixator for passive mobilization is usually not required. However, there are instances where complete ankylosis with bone draped where the ligaments were once present, requires a radical and extensive dissection, rendering the elbow acutely unstable (Fig 4). In this setting, hinged fixation permits the needed tissue resection, while protecting the joint from instability.

APPLICATION OF HINGED FIXATION

Preoperative Preparation

Most of the acute cases present in a dramatic fashion and provoke a natural impulse to proceed to immediate surgery for needed treatment. However, as noted above, an inadequate attempt that results in continued instability, redislocation, and subsequent delay, can severely compromise the final result. It is our recommendation that the surgeon should carefully review the case and make certain that the right team, equipment, and combined skills are available before proceeding to the operating room. We do not prefer to start these difficult cases in the middle of the night with surgical support staff that is unfamiliar with the long list of equipment and the variety needed. In our own practices, we will delay surgery in order to ensure an optimal setting.

Equipment

1. Internal fixation sets of a variety of sizes.
2. C-arm or fluoroscopy imaging.
3. Suture anchors.
4. External fixation equipment with spare or extra drill bits.
5. Radial head replacements and associated instrumentation.

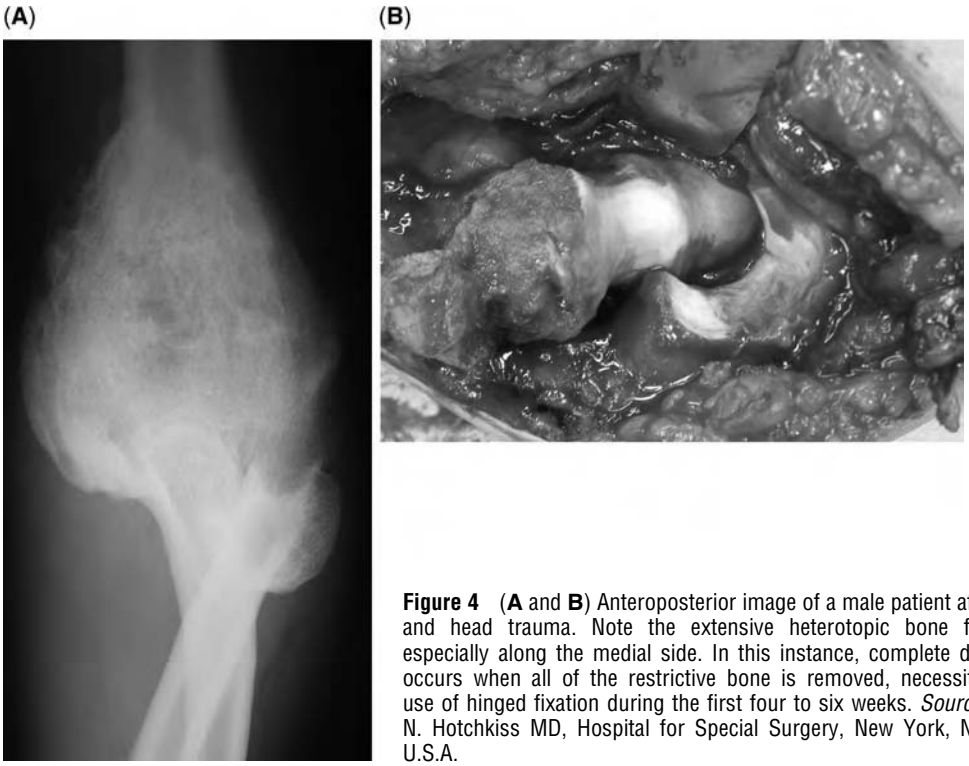

Figure 4 (**A** and **B**) Anteroposterior image of a male patient after elbow and head trauma. Note the extensive heterotopic bone formation, especially along the medial side. In this instance, complete dislocation occurs when all of the restrictive bone is removed, necessitating the use of hinged fixation during the first four to six weeks. *Source*: Robert N. Hotchkiss MD, Hospital for Special Surgery, New York, New York, U.S.A.

Position of the Patient

The optimal position for delayed operative treatment may be the prone position (Fig. 5). This position permits a simple posterior incision and a wide exposure of the joint. This position also makes application of the hinged fixator somewhat easier because there is a tendency for the joint to rest in a position of reduction rather than posteriorly subluxated or dislocation. The drawbacks of the prone position is that it takes greater preparation to position the patient and care must be taken to check the legs, back, and head for pressure points. If the anesthesia team has experience with surgery of the spine, they can often assist in safely positioning the

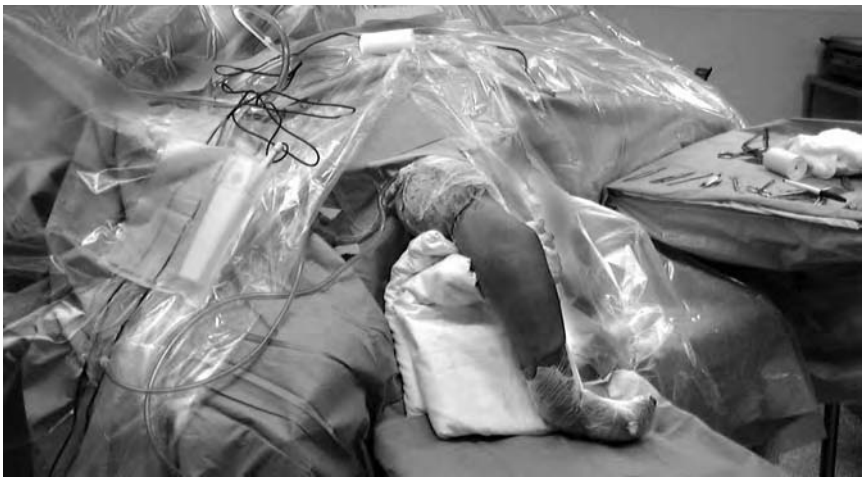

Figure 5 Prone position in operating room. *Source*: Robert N. Hotchkiss MD, Hospital for Special Surgery, New York, New York, U.S.A.

patient. A Foley catheter should also be placed. We tend to use the prone position in cases, where the use of a hinged fixator is likely.

Anesthesia

The advent of the infraclavicular block has been very helpful to both the intraoperative care and the postoperative pain management. The indwelling catheter can be used to bathe the proximal brachial plexus in bupivicaine for two days. The use of intermittent brachial plexus anesthesia reduces the consumption of narcotics, often overcoming the severe acute pain.

Surgical Exposure

Again, the indication for operative treatment is the inability to achieve and maintain stable reduction with the associated radial head fractures and/or coronoid fractures. The lateral approach of the elbow in most cases is best for open reduction, inspection of the joint, repair or replacement of the radial head, and repair of the posterolateral ligament complex.

The posterior or medial incision is generally needed for ulnar nerve exposure, especially if the frame used has a medial pin. For frames where the external pins are exclusively lateral, great care must be exercised and the radial nerve avoided during pin placement. If needed, expose the radial nerve for safety. The other indication for the medial approach to the joint is for open reduction and internal fixation of the medial facet fracture of the coronoid.

Application of the Fixator

Application of a hinged elbow external fixator can be demanding. The most critical point is correct placement of the axis pin (10). This pin must be collinear with center of rotation of the elbow joint to minimize resistance to motion and ultimately half-pin loosening. The anatomic axis of rotation lies at the center of the capitellum and trochlear spool, which is usually

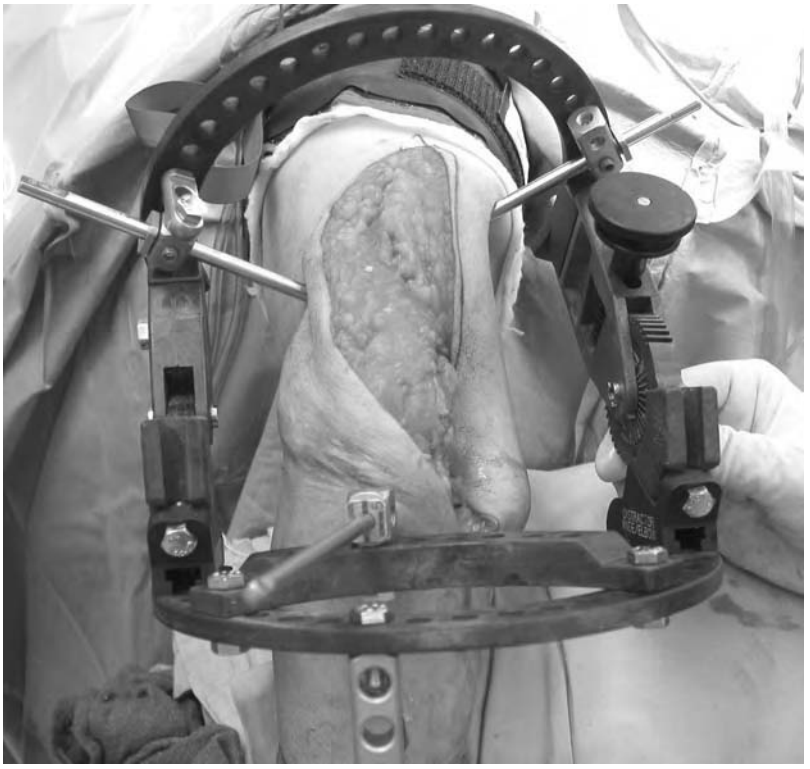

Figure 6 Note position of humeral pins in the anterior medial position (2 o'clock) and the posterolateral (10 o'clock) *Source*: Robert N. Hotchkiss MD, Hospital for Special Surgery, New York, New York, U.S.A..

Table 1 List of Commercially Available Hinged Elbow External Fixators

Hinge Fixator	Type	Motion (in frame)	Features	
Universal Compass Hinge (Smith & Nephew)	Bilateral	Active or passive	Passive/active motion using clutch and worm gear	
				Flexible pin distances and frame configuration using SmithNephew ex-fix system
OptiROM Elbow Fixator (EBI)	Unilateral	Active	Multiple adjustable linkages	
Orthofix Elbow Fixator (Intavent Orthofix Ltd.)	Unilateral	–	Linked components with central connecting units	
Dynamic Joint Distractor II (Stryker Howmedica Osteonics)	Uni-/bilateral	Active only	Simplified frame construction with integrated hinge	
				Compatible with Hoffman II compact couplings

determined from anatomic landmarks (11). Medially, this point lies just distal and anterior to the epicondyle, and laterally just slightly distal to the epicondyle (Fig 6).

If the distal end of the humerus is adequately visible, the axis pin should be confirmed by fluoroscopy. The pin should be slowly advanced and the position confirmed on lateral and anteroposterior (AP) views. The true lateral view should show the pin as a dot within the center of the trochlear spool, while the AP view should show it traversing parallel to the joint, along the normal valgus angulation of the distal humerus (Fig 2E and F). With unilateral frames, the axis pin needs only be inserted far enough to ensure proper orientation and stable bone purchase. With the multiplanar frames, a single axis pin can be advanced across the distal humerus or two pins can be placed from both the medial and lateral sides. Care must be taken on the medial side to protect the ulnar nerve during advancement of the axis pin.

The humeral portion of the frame is attached first, using the axis pin in the distal humerus as the guide. The external fixator frame is assembled around the axis pin and attached to the skeleton with half pins. These half pins should be placed without impaling any major muscle-tendon units or jeopardizing neurovascular structures. The humeral pins are usually placed first. Lateral pins are more easily placed due to patient positioning. The most proximal lateral pin may lie near the course of the radial nerve, which should be protected. If pins are placed medially, they should be placed through an open incision to protect the ulnar nerve.

All half pins should have bicortical purchase. We have been using hydroxyapatite-coated pins. Most adults tolerate a 6.5-mm tapered pin in the humerus and ulna.

Once the humeral portion of the frame is secured, the elbow must be reduced into position and the ulnar portion of the frame attached. The advantage of the prone position is quite clear during this stage, because a single assistant is usually able to hold the joint in position as the half pins are placed in the ulna. The optimal pin placement in the ulna is along the dorsal subcutaneous border. Some of the fixator frames require a more medial–lateral approach to pin placement in the ulna. With these devices, care should be taken to incise the skin and soft tissue, spread to bone, minimizing the soft-tissue injury.

After ensuring that all the connections are secure, the axis pin is removed and the elbow is taken through a range of motion under dynamic fluoroscopy to evaluate for concentric reduction and stability. The frame should permit easy flexion from 120° to 130° of flexion. Extension should be limited in these cases from 20° to 30° short of full extension. This is a practical limit to establish and expect in these injuries. Regaining adequate and functional flexion with a stable durable repair is more valuable than risking all for the last 20° to 30° of extension, which is an inherently unstable portion of the arc of elbow motion.

Before closing skin, all half pins should be checked for proper skin clearance and the ulnar nerve should be clearly unobstructed in flexion and extension of the elbow. Fine-tuning of the frame to achieve the desired amount of distraction and varus/valgus angulation is done at the conclusion of frame application.

Postoperative Care

Postoperative care for first two weeks after surgery requires diligence. We usually try to establish some motion in the first two days following surgery, even if the actual range is 30° total. At the first postoperative visit, the wound and pin sites must be inspected for redness, skin impingement, or impending infection.

At least 40% of patients will require an oral cephalosporin treatment for pin-tract redness and infection. Draining pus is not common, but if this should occur, the pin may need to be removed and exchanged to an alternate location in the operating room.

The nutbolts and connectors of the fixator should also be inspected. Some can loosen and require tightening.

X rays should be taken to assure a complete and proper reduction of the joint. If there is any suspicion of loss of position, the patient should be returned to the operating room. Loss of reduction will lead to a failure of the entire operation.

Hinge Removal

Removal of the fixator should be performed under regional anesthesia. This allows the surgeon to examine for range of motion, stability, and overall stiffness. We usually gently manipulate the joint to improve flexion and extension. Gentle sustained load can be applied but care must be taken. The pin sites and any previous screw holes are weak points in the bone and excessive load could result in fracture.

Postremoval Physical Therapy

There are many textbooks and protocols that apply to each of these conditions and patients. Each patient's rehabilitation program with these complex conditions must be individualized and monitored closely. If the therapy program is too aggressive, more pain and potential instability can occur. If left alone, many patients can become quite stiff. Following hinge removal, it is not uncommon to see the patient two to three times in the first six weeks to supervise their progress.

SPECIFIC FIXATORS

The OptiROM Elbow Fixator (EBI, Parsippany, New Jersey, U.S.A.) is a unilateral frame that is based on multiple stout adjustable linkages, which allow for many degrees of freedom. The axis pin is placed from lateral to medial. The humeral portion of the frame can then be applied with appropriate half pins. Alternatively, the humeral pins can be placed and secured to the frame before the axis pin is inserted. The axis guide ring can be adjusted to lie over the axis pin. The axis ring should slide easily over the guide pin for several centimeters to ensure proper alignment of the frame with the elbow axis of rotation. An additional benefit is that the ulnar half pins can be inserted either from dorsal to volar (preferred) or lateral to medial and locked to the frame with the elbow in an unreduced position. Reduction of the ulnohumeral joint can be done with the frame in place and the universal joints tightened to maintain reduction. This avoids the sometimes-difficult task of placing the ulnar pins, while maintaining perfect reduction of the elbow. A new addition to the EBI frame allows for static progressive ROM with application of torque through an adjustment screw.

The Dynamic Joint Distractor II (Stryker Howmedica Osteonics, U.S.A.) is based on the same concept as its predecessor, the Mayo Dynamic Joint Distractor (2). The frame can be applied in a unilateral or bilateral configuration. The axis pin is placed with the use of a humeral axis guide by open technique on the medial side and percutaneously on the lateral side. This device clamps the center points on the medial and lateral sides and the axis pin is placed through the cannulated guide. The frame is applied over the axis pin and pin guides are used to place the humeral pins. The half pins are connected to the frame with standard Hoffman II Compact clamps. The ulnar pins are placed from lateral to medial in a percutaneous fashion, which has the disadvantage of impaling the common extensor muscles. There is a distraction–compression device built into the frame that can be progressively adjusted. If more stability is required, dual medial and lateral frames can be applied. The major advantages of the frame are low profile and relative ease of application.

The Universal Compass Hinge (Smith & Nephew, Memphis, Tennessee, U.S.A.) is a multiplanar fixator, which allows for incremental passive joint range of motion. The frame is composed of radiolucent 1/2- and 5/8-rings and preassembled prior to application. The humeral half pins are placed in both medial and lateral multiplanar positions. This frame can also be applied in a unilateral fashion, either medial or lateral, but this configuration is not recommended by the manufacturer and there are no specific instructional materials explaining this configuration. The ulnar pins are inserted from the dorsal surface of the ulna in a dorsal to volar direction. The frame has a self-telescoping mechanism to allow for a 20° arc of varus/valgus adjustment. Distraction is independent of the varus/valgus alignment. Additionally, a precision worm gear permits motion within a specified range and can be ''ungeared'' for active and passive motion or kept locked for incremental gear-driven passive motion. The patient or occupational therapist can easily operate the gear. The disadvantages of the Compass Hinge are that its application can be technically demanding and there is less room for adjustment if the frame is placed with the elbow subluxated. Patient comfort is at times an issue because the frame impinges against the chest wall.

SUMMARY

The use of hinged external fixation about the elbow can be extremely valuable for complex and complicated trauma. The use of these devices requires a complete understanding of the axes of alignment of the joint, the soft-tissue anatomy, and the requirements for success. Similar to other surgical devices, success is dependent on when and how to apply the fixator. Current indications include complex acute and recurrent instability after trauma and ligament repair, delayed treatment of complex dislocations with fractures, distraction interposition arthroplasty, and postoperative control of joint stability after radical resection of ankylosing contracture. Careful monitoring of the patient during the time in the fixator and in the weeks following removal is also crucial for an optimal outcome.

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41 Forearm Lengthening with Hybrid Circular Frame

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INTRODUCTION

The discrepancy in length between the two upper extremities does not produce a significant functional deficit. For this reason and for fear of functional loss, lengthening of the forearm has rarely been performed for shortening of the forearm and is usually performed for discrepancies in lengths between the radius and ulna and forearm/wrist deformity.

Several limb-lengthening techniques have been described by many authors. In this chapter, we want to report the Lecco Ilizarov Unit's experience for forearm lengthening using the hybrid (fixation with both skinny wires and half pins) circular frame of Ilizarov.

Use of the Ilizarov method has enabled correction of limb-length discrepancy as well as deformity in all of the limb segments. This technique of lengthening relies on bone regeneration rather than bone grafting (1–10).

CLINICAL EVALUATION

In their extreme variability the different forearm deformities, require surgical strategies often customized for any single patient. These techniques result in both an aesthetic and functional corrections to improve quality of life (11).

Lengthening of the forearm is primarily indicated for discrepancies between the lengths of the radius and ulna (5,12). The present authors believe it is also useful for lengthening of short, one-boned forearms. The typical patient with a radial club hand is usually treated by centralization of the wrist and sometimes pollicization of the index finger. The patient is then left with a forearm that is less than one-half the length of normal. This begins to present a problem only in adolescence when their humerus is fully grown. The long humerus bone keeps them from reaching their mouth and hair. Patients have difficulty with perineal care, cutlery usage, and dressing. Lengthening short forearms is functionally, psychologically, and cosmetically beneficial. Optimally, it would be preferable to perform two smaller lengthenings, one at age eight and the other after maturity, than one large lengthening (1,3,4).

The specific configuration of the Ilizarov apparatus used depends upon whether one or both bones of the forearm are to be lengthened.

A scanogram and comparison radiographs of the contralateral wrist and forearm are done to assess the limb-length inequality and malalignment (2).

CROSS-SECTIONAL ANATOMY OF THE FOREARM FOR WIRE/PIN PLACEMENT

Forearm external fixation requires a perfect anatomic knowledge of this complex region to avoid the risk of vascular and nerve injury (13). In detail, we are describing the principle sites of bone fixation in the right forearm (14).

Cross section at the Level of the Radial Head

Fixation of the proximal radioulnar joint is done with a wire inserted from anterolateral to posteromedial, keeping the forearm in complete supination; the insertion angle is approximating 25° to the coronal plane.

Isolated fixation of the radius is difficult at this level because of the anteromedial vessels and the medial ulnar. It can be done with a 5-mm half pin inserted from posterolateral to anteromedial at an angle of 20° to the sagittal plane.

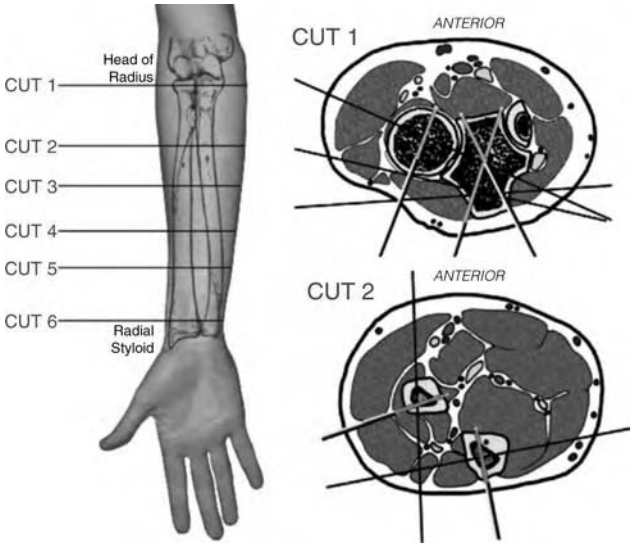

Figure 1 Cross section in the right forearm. Cut 1: proximal radioulnar joint; Cut 2: proximal third. *Source*: From Ref. 15.

Isolated ulnar fixation is much simpler and can be done with one transverse wire and a second wire from anterolateral to posteromedial, posterior to the ulnar nerve. Fixation with half pins can be done posteriorly at an angle of 20° to the sagittal plane (Fig. 1, Cut 1).

Cross section at the Level of the Proximal Third of the Forearm

Isolated ulnar fixation can be done with a transverse wire (almost parallel to the coronal plane) and a half pin from posterior to anterior, perpendicular to the wire.

Isolated radial fixation can be performed with a wire from anterior to posterior and a half pin from posterolateral to anteromedial, angulated 20° to the coronal plane (Fig. 1, Cut 2).

Cross section at the Level of the Proximal Mid-Third

Isolated radial fixation can be carried out with a wire directed from anterior to posterior. A half pin can be inserted from posterolateral to anteromedial at an angle approximating 20° to the coronal plane, a second half pin can be added proximally to the first from anterolateral to posteromedial at an angle approximating 25° to the coronal plane. Fixation of the ulna can be performed with a wire from anteromedial to posterolateral, angulated 20° to the coronal plane, and a half pin from posterior to anterior (Fig. 2, Cut 3).

Cross section at the Level of the Distal Mid-Third

Isolated radial fixation can be carried out with a wire directed from anterolateral to posteromedial angulated 30° to the sagittal plane. A half pin can be fixed in a posterolateral position, perpendicular to the previous wire. Fixation of the ulna can be performed with a wire from anteromedial to posterolateral, angulated 20° to the coronal plane, and a half pin from posteromedial to anterolateral, angulated 10° to the sagittal plane (Fig. 2, Cut 4).

Cross section at the Level of the Proximal Distal Third

Isolated radial fixation can be carried out with a wire directed from anterolateral to posteromedial, angulated 40° to the coronal plane. A half pin can be inserted from a posterolateral position, perpendicular to the previous wire. Ulna fixation is performed with a wire from anteromedial to posterolateral angulated 40° to the coronal plane and a half pin from posteromedial to anterolateral, angulated 15° to the sagittal plane (Fig. 3, Cut 5).

Cross section at the Level of the Distal Radioulnar Joint

Ulnar fixation is performed with a wire directed from anteromedial to posterolateral angulated 45° to the sagittal plane and a half pin directed from posteromedial to anterolateral,

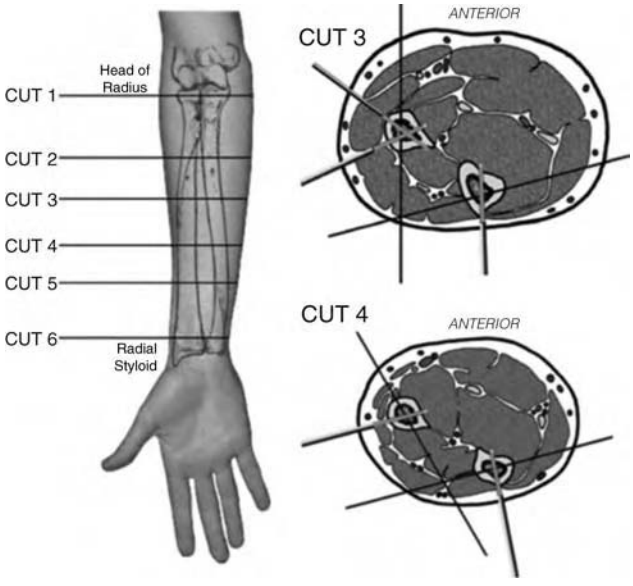

Figure 2 Cross section in the right forearm. Cut 3: proximal mid-third; Cut 4: distal mid-third. *Source*: From Ref. 15.

perpendicular to the previous wire. The radius can be fixed with one wire directed from anterolateral to posteromedial angulated 45° with the coronal plane and a second wire inserted from anterior to posterior, between the flexor carpi radialis and the median nerve, using the open technique: small incision, dividing with a mosquito the underlining soft tissues till the bone surface and, introducing the wire. A half pin is inserted from posterolateral to anteromedial, perpendicular to the first wire. Fixation of the two bones is done with a wire from anterolateral to postero medial, angulated 20° to the coronal plane (Fig. 3, Cut 6).

CLASSIFICATION

Forearm deformities with upper limb discrepancy, include several congenital and acquired pathologies that require lengthening (16); practically, we divide this pathologies into six categories (Table 1) (Fig. 4). Clinical examples are radial clubhand, Madelung's deformity, Ollier's disease, and post-traumatic growth arrest.

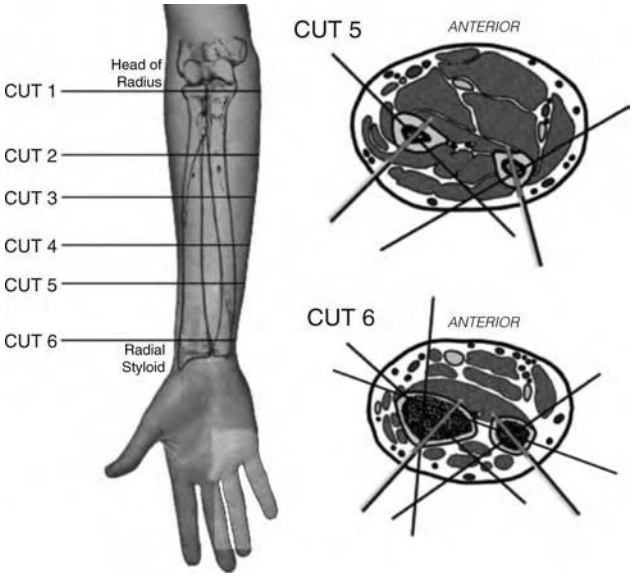

Figure 3 Cross section in the right forearm. Cut 5: proximal distal third; Cut 6: distal radio ulnar joint. *Source*: From Ref. 15.

Table 1 Classification of Forearm Deformity into Six Types

Type 1	Shortening of the radius alone
Type 2	Shortening of the ulna alone without dislocation of the radial head
Type 3	Shortening of the ulna with dislocation of the radial head
Type 4	One-bone forearm (ulnar or radial hemimelia)
Type 5	Shortening of the radius and ulna to the same proportion
Type 6	Shortening of the radius and ulna to different proportions

TREATMENT OPTIONS

Our strategy is to preassemble a frame for any specific deformity and then apply it to the forearm with one proximal and one distal reference 1.8 mm wire, inserted perpendicular to the mechanical axis of each bone segment. Wires are tensioned 110 Kg. Impingement of the soft tissues and the joints are avoided. The fixation is then completed by adding additional wires and half pins. A percutaneous low-energy multiple drill-hole osteotomy is then done at the appropriate ulna and/or radius site. After 8 to 10 days latency, distraction is commenced in order to achieve lengthening and deformity correction (2).

SURGICAL TECHNIQUES

The surgery is done with the patient supine, and the upper extremity resting on a radiolucent hand table. An intraoperative fluoroscopy unit is used. During the surgery, any twitching of the wrist and finger, after the insertion of a wire or half pin, especially close to the radial neck, is highly suggestive of the dangerous proximity to a nerve and, therefore, must be immediately changed.

Based on our classification (Table 1, Fig. 4), a lengthening technique for each type of forearm shortening and deformity is presented:

Type 1: Lengthening of the Radius Alone (Figs. 5 and 10)

One anteroposterior (AP) 1.8 mm wire is inserted into the radius at the mid-forearm level (Fig. 2, Cut 3) and one anterior–posterior in the distal radius (Fig. 3, Cut 6). Two same-sized

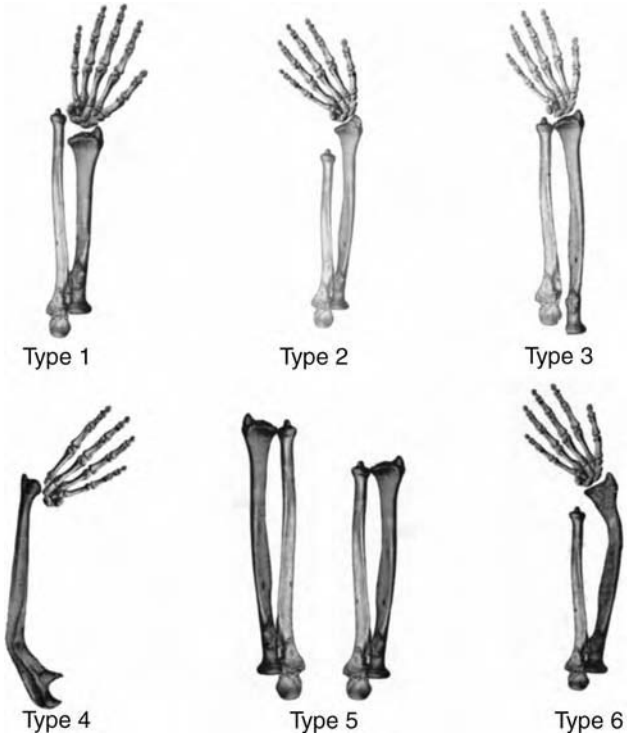

Figure 4 Classification of shortening of the forearm deformities. Type 1: shortening of the radius only; Type 2: shortening of the ulna only; Type 3: shortening of ulna with dislocation of the radial head; Type 4: ulna only with radial club hand (radial hemimelia); Type 5: shortening of both bones to the same proportion (left forearm normal, right short); Type 6: shortening of both bones to different proportions.

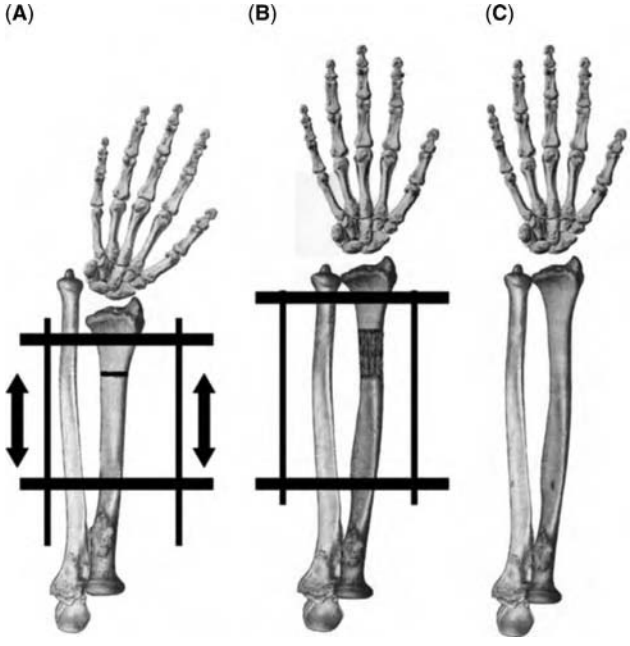

Figure 5 (**A**) Preassembled frame for isolated radius lengthening, after a latency of 8 to 10 days lengthening is started with a rhythm of 0.25 mm twice a day. (**B**) Reaching of the desired length with presence of a good new bone formation. (**C**) Consolidation of the new bone with corrected position of the hand.

rings (leaving one to two fingerbreadths between skin and ring) are chosen and connected with two threaded rods of the same length, one medially and one laterally. These are centered on the forearm wires and then fixed and tensioned to 110 kg. The alignment of the threaded rods should be parallel to the ulna, assuming there is no deformity. A second 1.8-mm wire is added at the distal radius inserted from anterolateral to posteromedial, fixed, and tensioned. Finally, three 5 mm half pins are inserted, after a 3.2 mm predrilling: one is added on the proximal surface of the proximal ring directed from anterolateral to posteromedial, one connected on the distal surface distal of the proximal ring directed from posterolateral to anteromedial; the last half pin is placed off the proximal surface of the distal ring with a dorsal to volar direction. (Fig. 3, Cut 6). In this way, we have obtained very good stability at each level of fixation. Then, the threaded rods are removed and the distal radius osteotomy is performed percutaneously under the guidance and protection of periosteal elevators. A 1-cm dorsal incision is made over the distal radius and percutaneous periosteal elevation is performed. Then multiple low-energy 3.2-mm bicortical bone drills are made and a 3 mm osteotome is used to cut around the radial cortex, following the holes previously made. The osteotome is twisted and a transverse osteotomy is obtained. Finally, four threaded rods and locking nuts are applied between the two rings anteromedially, anterolaterally, posteromedially, and posterolaterally. This permits an unobstructed view of the radius and ulna on AP and lateral roentgenograms.

Type 2: Lengthening of the Ulna Alone (Fig. 7)

A similar frame design is used for lengthening of the ulna, with pin placement in the ulna alone, distally with one 1.8-mm wire and one 5-mm half pin (Fig. 3, Cut 5). Proximally, the fixation involves both the radius and. ulna with one wire and with two half pins in the olecranon (Fig. 1, Cut 1). It is very important in both the ulna and radius proximally to avoid pulling the radial head distally away from the capitellum. A 5/8 ring opened anteriorly (C-shaped) at the elbow is used in the proximal ulna. The optimal site for the corticotomy is in the proximal metaphysis of the ulna. After an 8 to 10 days latency period, lengthening is started with a rhythm of $\frac{1}{4}$ mm twice a day.

Type 3: Lengthening of the Ulna with Reduction of Congenital Dislocation of the Radial Head (Fig. 8)

The preconstructed frame consists in a distal full ring and a proximal 5/8 ring connected by two threaded rods. The radius is transfixed to the ulna distally with a 1.8-mm Kirschner (K) wire, while the ulna is fixed with one wire and one half pin (Fig. 3, Cut 6). Proximally, the

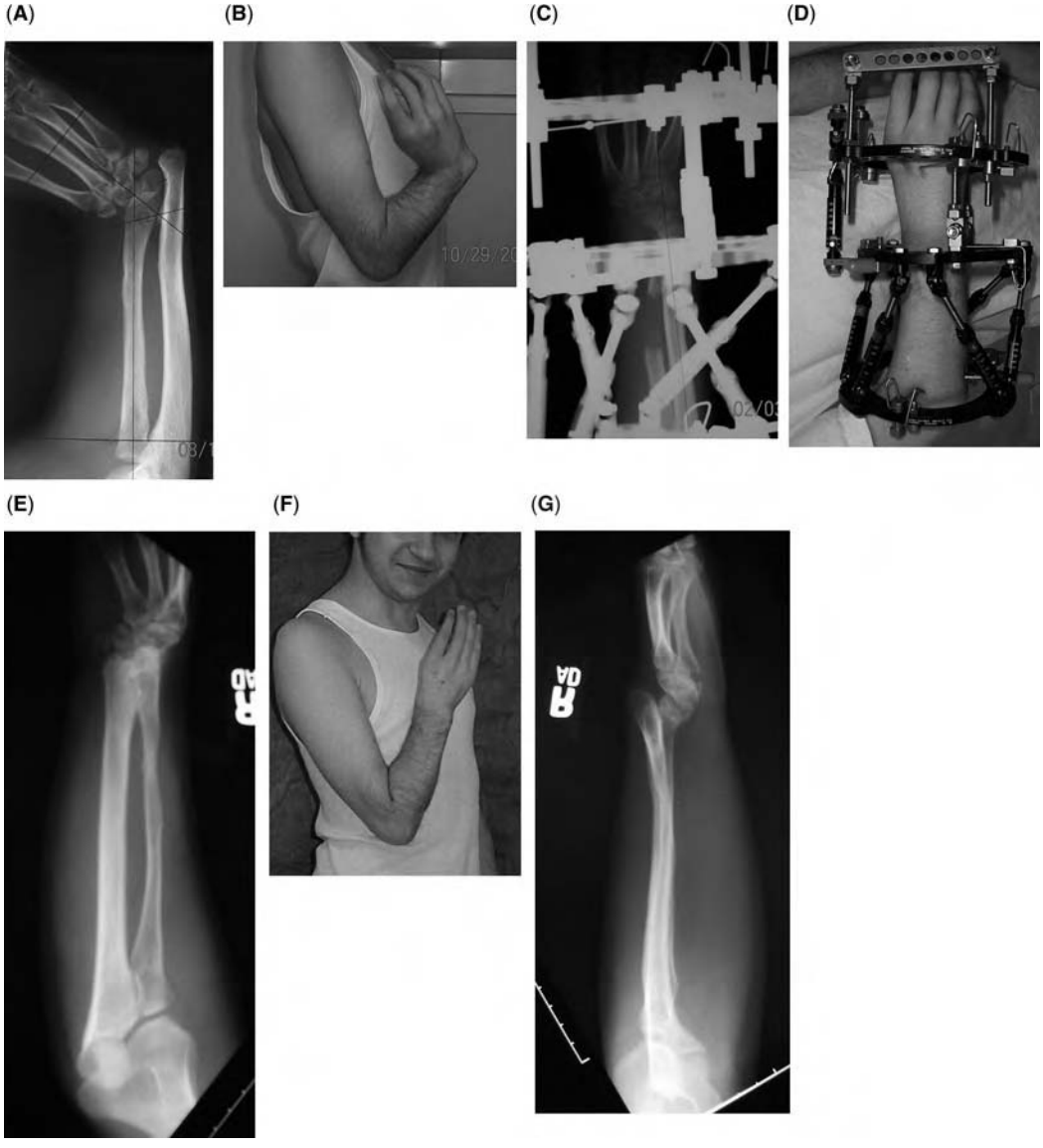

Figure 6 Fifteen-year-old boy with Type 1 forearm deformity—radial shortening combined radial deviation of wrist. (**A** and **B**) Preoperative X ray and clinical photo. (**C** and **D**) X ray and clinical photo after lengthening of radius and correction of wrist deformity with an Ilizarov/Taylor Spatial Frame. (**E–G**) X rays and clinical photo nine months after frame removal. *Source*: Case supplied by S. Robert Rozbruch, MD.

radius is not fixed and the ulna is fixed with one wire and two 5-mm half pins in the olecranon (Fig. 1, Cut 1).

In this situation, the ulna is osteotomized through the level of its deformity rather than at its proximal metaphysis. After the application of a third rod, lengthening of the ulna will lead quite always to reduction of the radial head. Sometimes final reduction may be assisted by the insertion of an appropriately directed olive wire to pull the radial head into the joint.

After an 8 to 10 days latency period, the ulna lengthening is started.

Type 4: Lengthening of the Ulna in Radial Aplasia (Figs. 6, 9, and 10)

Correction of the radial club-hand deformity should ideally have been performed prior to lengthening. Lengthening and deformity correction of the radial club hand can be accomplished with the Ilizarov method.

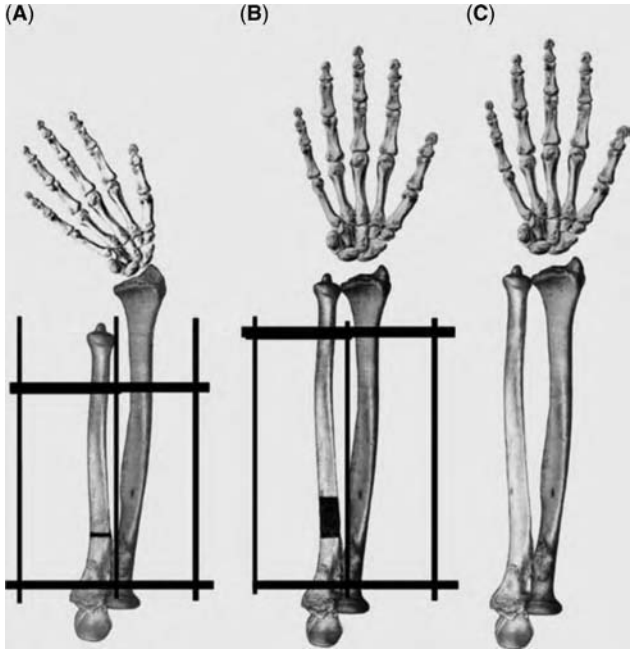

Figure 7 (**A**) Preassembled frame for isolated ulna shortening without radial head dislocation the osteotomy site is in the proximal ulnar metaphysis. (**B**) Reaching of the desired length with presence of a good new bone formation. (**C**) Consolidation of the new bone with corrected position of the hand.

An Ilizarov circular frame for the correction of radial club hand spans three levels. A proximal half ring is fixed at the level of the olecranon with one 1.8 mm diameter K inserted transversely through the olecranon and two 5 mm half pins (Fig. 1, Cut 1). The wire is inserted from medial to lateral to preserve the ulnar nerve, and fixed and tensioned on the half ring, the first half pin is positioned posteriorly and perpendicular to it. The second pin is placed a 45° posterolaterally between the wire and the first pin. It is possible to use 3-mm K-wires instead of half pins: they should penetrate both cortices, but transfixion of the opposite soft tissue compartment is to be avoided.

The distal ulnar ring is fixed with one wire and one or two 5-mm half pins placed several centimeters apart. The pins are inserted from medial and posteromedial aspects at 45° to the

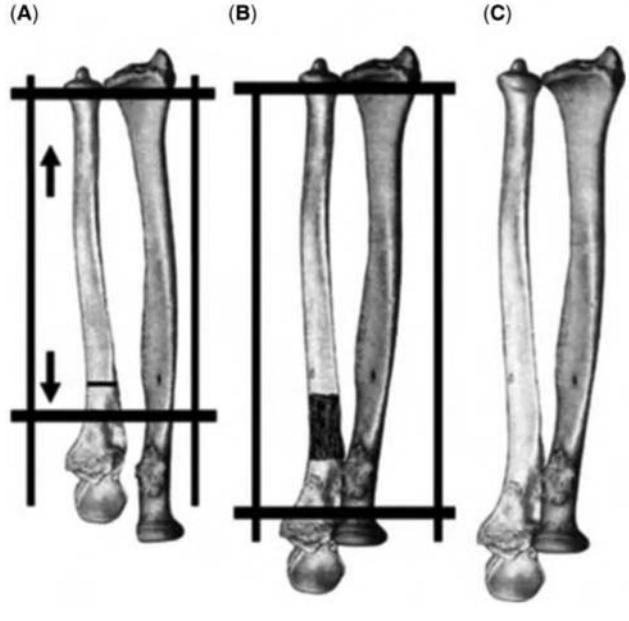

Figure 8 (**A**) Mounting for congenital radial head dislocation with shortening of the ulna and osteotomy site for ulnar lengthening. (**B**) Reaching of the desired length with presence of a good new bone formation and spontaneous reduction of the radial head dislocation. (**C**) Consolidation of the new bone.

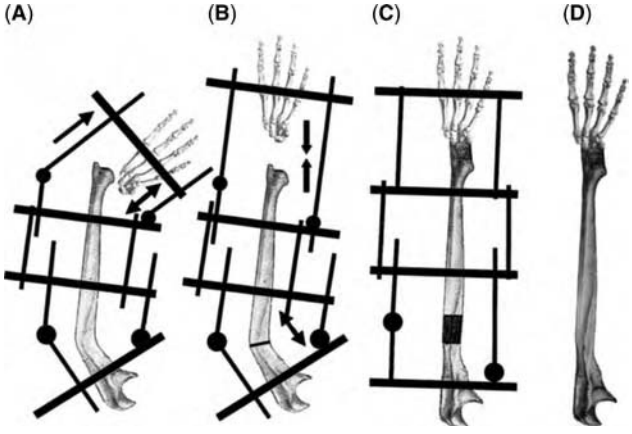

Figure 9 (**A**) Preassembled circular frame to correct radial hemimelia with radial club hand, progressive wrist distraction is started the day after the operation at the rhythm of 0.5 mm three times per day. In this step, no ulnar osteotomy is done. (**B**) Once the hand has reached the corrected position, a compression between carpal bones and distal ulnar epiphysis is performed until achieving bony contact. Then a small incision wrist arthrodesis and an ulnar osteotomy for lengthening are performed. (**C**) End of ulnar lengthening and correction and functional arthrodesis of the wrist. (**D**) Final result.

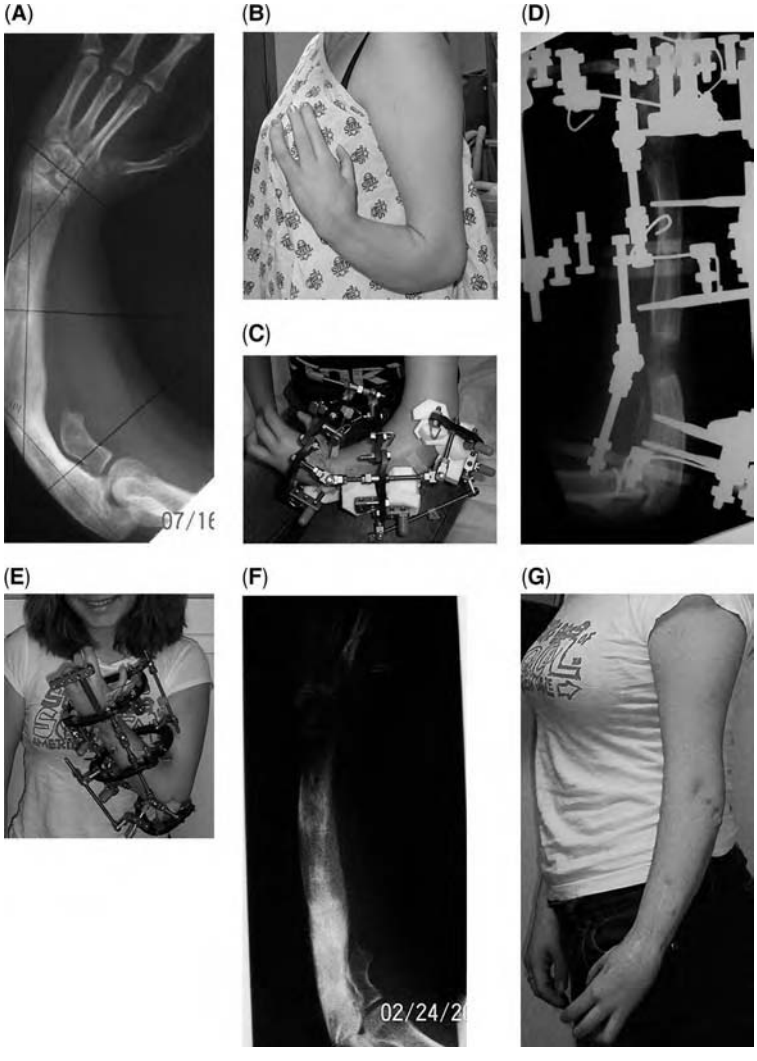

Figure 10 Fourteen-year-old girl with Type 4 forearm deformity—single bone forearm with radial clubhand (radial hemimelia). (**A** and **B**) Preoperative X ray and clinical photo. (**C**) Postoperative photo showing an Ilizarov frame matching the deformity and set up for 2-level correction. (**D** and **E**) X ray and clinical photo after lengthening and correction of deformity. (**F** and **G**) X ray and clinical photo six months after frame removal. *Source*: Case supplied by S. Robert Rozbruch, MD.

coronal plane. The wire is inserted from anteromedial to posterolateral, also at 45° to the coronal plane.

The half pins may again be substituted by 3-mm wires, placed in a triangular configuration to increase stability. Finally, a 5/8 or full circular ring, which will be used to correct carpal deviation, is applied at the level of the metacarpals with one transverse wire. The fixation of the metacarpals may be augmented by one or two half wires. These should be placed at 45° to the plane of correction of the deformity and the last ring should be fixed to the distal ulnar ring with two strategically placed hinges. A ''motor'' to correct the deformity can be placed in a convenient position between the distal two rings. Carbon-fiber rings facilitate imaging in these short, deformed arms and are much less heavy than metal rings.

Once the hand has reached the corrected position, wrist arthrodesis can be achieved by decortication of the articular surface through a 3-cm dorsal skin incision, followed by compression in the frame. It is equally possible to achieve arthrodesis of the wrist by compression in the Ilizarov frame only. If the articular surfaces are congruent, we prefer the latter method. A solid arthrodesis is achieved with either method in two to three months, while lengthening of the ulna takes place.

The ulnar osteotomy is needed to lengthen the forearm, because a second step at the time of wrist fusion can also be used to correct coexisting ulnar deformity. The osteotomy site should be in the proximal ulnar metaphysis, where the osteogenesis is more reliable and where the deformity is usually located. Hinges are used to correct the proximal ulnar angular deformity and also to obtain ulnar lengthening.

In the previously centralized wrist, the two-ring apparatus is applied. Because these are often extensive lengthenings, an additional wire in the metacarpals to prevent a wrist flexion contracture is used if there is no preexisting wrist arthrodesis.

Type 5: Lengthening both the Radius and Ulna to the Same Degree

This type of forearm deformity is rarely treated because generally the function of the elbow and the wrist is fully maintained. Only in selected cases, when the limb-length discrepancy is about 50%, the forearm lengthening can be performed. In these cases two separate frames, one within the other may be used to fix each bone separately and allow some supination and pronation. The description of this technique will be described in Type 6 deformity.

Type 6: Lengthening of Both the Radius and Ulna to Different Degrees (Figs. 11 and 12)

Lengthening of the forearm is primarily indicated for length discrepancies between the radius and ulna. In this situation, we observe a discrepancy in length between the two bones, mainly

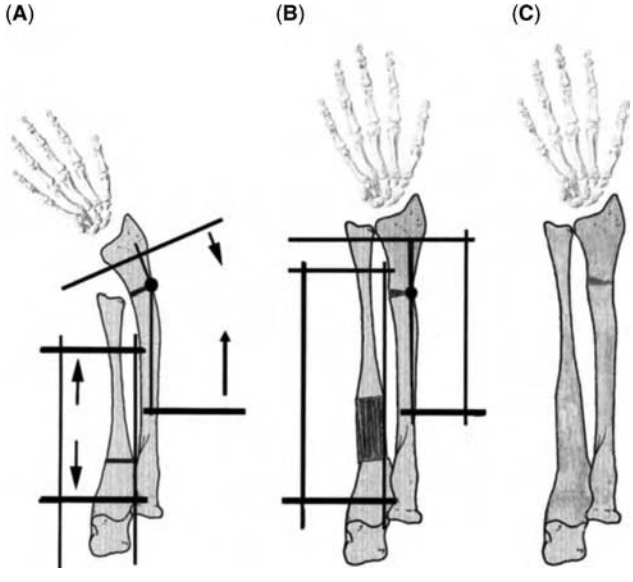

Figure 11 (**A**) Differentiated ulnar-radial lengthening-correction with two separate frames. (**B**) End of treatment with radial correction, ulnar lengthening, and ulnar club hand correction. (**C**) Consolidation of the new bones after removal of the frames.

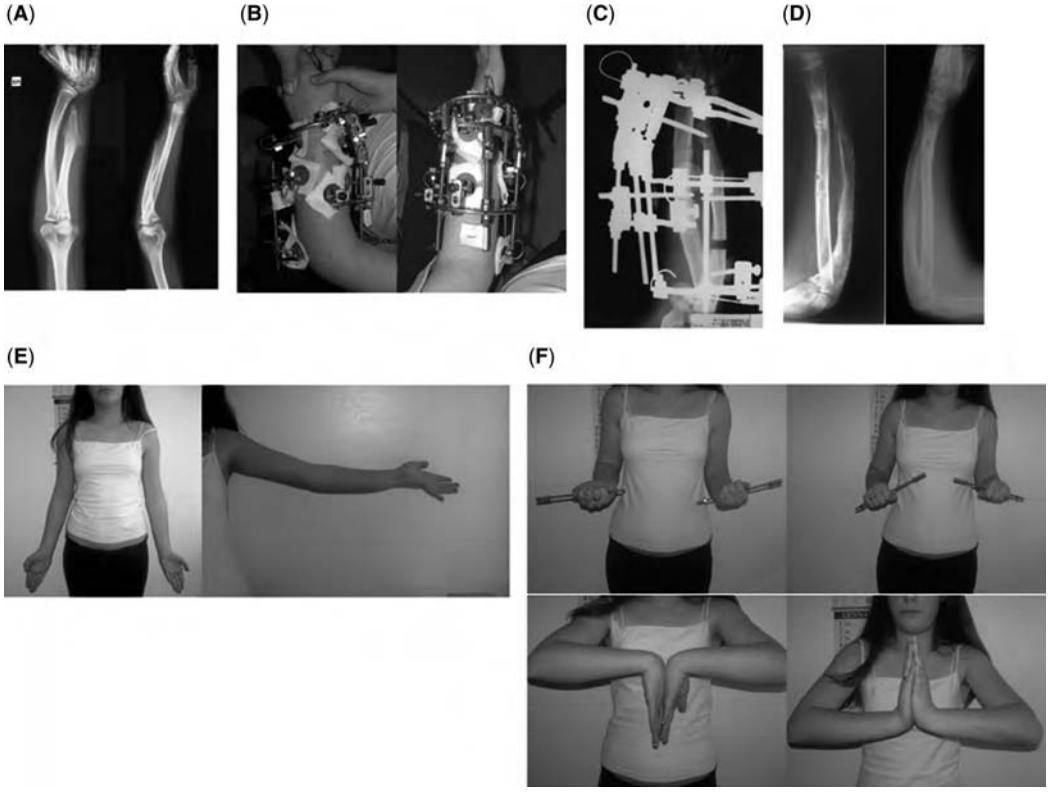

Figure 12 (**A**) Type 6 deformity of the left forearm in a 12-year-old female with ulnar club hand. (**B**) Differentiated ulnar and radial correction with two separate Ilizarov frames. (**C**) X ray of the left forearm at the beginning of the treatment. (**D**) Final X rays showing radial correction, ulnar lengthening, and wrist placed in a functional position. (**E** and **F**) Elbow and wrist function is demonstrated.

involving the ulna, with a distal radius varus deformity and consequentially an ulnar club hand. For these deformities, we prefer performing a differential lengthening of the two bone using two separate frames.

The radius frame consists of a distal ring perpendicular to the mechanical axis of the distal radial metaepiphysis and a half ring placed at the proximal mid-third perpendicular to the proximal radial mechanical axis. Two strategic hinges, one volar and one dorsal are placed on the convex site of the deformity, with the rotation plane tangent to the cortex and perpendicular to the coronal plane.

The connection to the bone is obtained distally with one 1.8-mm K wire and two 5-mm half pins (Fig. 3, Cut 6). Proximally, the half ring is fixed to the bone with one wire and two half pins (fig. 2, Cut 3). A percutaneous dorsal distal-radial osteotomy is performed at the level of the deformity. A motor device is added to the radial side (convexity) of the frame.

The ulnar premounting is simpler and spans one proximal and one distal half ring connected by two threaded rods. The ulna is fixed proximally with one wire and two half pins in the olecranon (Fig. 1, Cut 1) and at the level of the distal third with one wire and two half pins (Fig. 2, Cut 4). A proximal ulnar osteotomy is done and a third threaded rod is added.

After a latency of 8 to 10 days, a 0.25 mm twice a day lengthening on the three threaded rods in the ulna and a radial 0.5 mm twice a day compression on the motor of the radius frame is started.

The correction continues till a complete radial deformity correction is obtained.

The ulnar lengthening is continued until correct wrist alignment is reached (Fig. 9).

AFTER TREATMENT

Postoperatively, the forearm is elevated. Physical therapy begins the day after surgery. This involves exercises for the fingers, wrist, elbow, and shoulder. Initially, the most important

exercises are passive range of motion, and as the pain decreases, active exercises are also encouraged. It is important to try and maintain full excursion for each joint not fixed in the apparatus.

After 8 to 10 days, lengthening begins at the rate of 0.25 mm two times daily. One-quarter millimeter is equivalent to a one-quarter turn of the nuts. The threaded rods are of 1 mm pitch.

The first postoperative roentgenograms are performed one week after the beginning of distraction. One looks for diastasis at the level of the corticotomy. Thereafter, alternate week roentgenograms and follow-up exams are performed.

The patient is encouraged to participate in a daily physical therapy program and to perform the exercises instructed several times per day while at home, school, or work.

A plastic wrist and finger extension-resting splint is made for nighttime use. The splint can be attached to the apparatus and removed easily by the patient. An elbow extension splint is also made to be attached or removed from the apparatus and is predominantly for nighttime use.

If significant pain develops as the lengthening proceeds, the lengthening rate should be reduced to 0.25 mm per day. If significant contractures develop that cannot be kept in check by physical therapy or by splinting, the lengthening rate should also be stopped for four to five days. Regenerate new bone formation should be seen on X ray by the second to third week. If hypotrophic new bone formation appears, an accordion procedure may be started (shortening 0.5 mm twice a day for five days, stop five days, restart the lengthening 0.25 mm a day for 20 days).

If a flexion contracture of the elbow, fingers, or wrist appears to be developing, the extension splints should be worn for up to 12 hours per day. This is best tolerated on a two-hour-on, two-hour-off basis and then during the night. Patients frequently will require oral analgesics and sleeping medication during the distraction phase.

Pin-tract infection prevention is started from the first day. Sterile foam sponges are applied to each pin site, and rubber stoppers or clips are applied on each wire or half pin to keep these sponges in place and to reduce the skin-pin motion. An antiseptic solution or antibiotic powder or ointment may be applied to the sponges on a daily basis. After approximately four to six weeks, the sponges may be discontinued if the pins appear to be well tolerated. Patients may shower initially once per week when they change their sponges. After that, once the sponges have been discontinued, they may shower and swim on a daily basis. Full functional activities to their tolerance level are encouraged.

Once the distraction is completed, the apparatus is left in place and monthly roentgenograms are taken. Bony consolidation proceeds and the process of neocorticalization closely follows. Once there is a sufficient new cortex, the apparatus may be removed. A plaster cast may be needed for protection for several weeks. A removable orthosis can be used for protection. Once the bone is considered to be strong enough to resist all physiologic forces without protection, active physiotherapy is resumed to regain wrist, finger, and elbow range of motion.

COMPLICATIONS

These can be divided into those that are temporary and those that are permanent (17).

Superficial wire and pin-site infections may be avoided by maintaining the wire tension adequately and by stabilizing the skin around the wires. If inflammation does occur, it usually responds well to basic pin-site care and oral antibiotics, and it rarely progresses to deep infection, osteomyelitis, or septic arthritis. After removal of the frame, fracture of the regenerate bone may result from relatively trivial trauma. This can usually be dealt with by plaster cast or by application of another Ilizarov apparatus. Paresthesia in the distribution of the ulnar nerve due to irritation at the time of wire or pin insertion usually resolves spontaneously with time.

Flexion contractures of the fingers tend to be permanent and occur to some degree in all patients. Full cooperation with physiotherapy during treatment will help to minimize this problem, but if a contracture progresses despite intensive exercise, ulnar lengthening should be slowed or stopped altogether. If further lengthening is needed, it is advisable to wait at least a year before distraction is resumed.

FUTURE DIRECTIONS

We feel that the Ilizarov technique, especially in its new configuration with half pins (the so called Western hybrid method), is a powerful way to correct deformity in one or both bones.

Furthermore, multiple levels of fixation allow multiple levels of correction. Other applications include bone defects, nonunions, and congenital pseudarthrosis.

In conclusion, the Ilizarov method is a reliable, successful, and safe method and it is the gold standard to treat forearm-length discrepancy and deformity problems, preserving a satisfactory function of the upper limb during the treatment.

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42 Congenital Hand Deformity

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INTRODUCTION

In cases of congenital hand deformity, the length of metacarpals and phalanges are often too short to achieve pinching or grasping. Lengthening of the digits has been used to improve this function of the hand (1,2). In cases involving a very small palm and absence of fingers, metacarpals have been lengthened to create a larger palm, making it more functional for use in bilateral hand activities (3). An improvement of hand appearance also has been a beneficial psychological factor for both the patient and the family.

There are different surgical techniques available for correction of congenital hand deformities. In this chapter we will concentrate on lengthening technique strategies for metacarpals and phalanges using methods of distraction osteogenesis. This method obviates the need for grafting.

CLINICAL EVALUATION

Congenital hand deformities are diagnosed immediately after birth and strategies for treatments are developed from that time. Because the conditions of hands in cases of congenital deformity are different in each case, various combinations of treatment modalities, including bone lengthening, are used (4). Thorough evaluation of the functional abilities of hands is very important. Often in cases of congenital hand deformities, fingers are fused skeletally or cutaneously, and muscles and tendons that contribute to hand functions are hypoplastic (5). Depending on the clinical and radiological findings, other operations may have to be performed for the improvement of hand function prior to or simultaneously with lengthening. Also, it is very important to ensure a good vascular supply and adequate soft-tissue coverage for the lengthened digit.

The length and the condition of the bone are important factors for bone lengthening (6). These factors are closely related to the particular diagnosis and patient's age. The lengths of the phalanges and the metacarpals tend to be short and sometimes the bone length is crucial in planning and choosing the lengthening device. The technique for lengthening smaller bones is more difficult. However, it is possible to lengthen the bones of children younger than five years of age, whose metacarpals and phalanges are shorter than 10 mm in length.

Bone growth after lengthening could also affect the result of treatment (7). In some cases, the bone growth of metacarpals and phalanges is different in each bone of the same hand. When bone lengthening is used for young children, the growth potential of the bone should be carefully considered.

CLASSIFICATION

We use the following classification: failure formation-transverse arrest (symbrachydactyly), failure formation-longitudinal arrest (hypoplastic thumb, hypoplasia of the little finger), amniotic (constriction) band syndrome, and other diagnoses (8).

Table 1 Clinical Decision Making for Congenital Hand Deformity

Classification Subgroup	Clinical Evaluation	Surgical Approach	Pearls (Other Surgery)	Complications/Pitfalls
Transverse growth arrest (symbrachydactyly)	Pinching or bilateral activity	Multilateral or monolateral	Web plasty separation of fingers	Unequal bone growth after lengthening in younger children
Longitudinal growth arrest (hypoplastic thumb, hypoplasia of little finger)	Pinching	Monolateral	Correction osteotomy	Unequal bone growth after lengthening in younger children
Amniotic (constriction) band syndrome	Grasping or pinching	Multilateral or monolateral	Separation of fingers, resection of amniotic	Bony prominence of fingertip

Source: From Refs. 7 and 10.

Patients with symbrachydactyly and amniotic band syndrome tend to be treated using bone lengthening at a younger age (7). With these two conditions, bone lengthening is required to improve hand function at an early stage of treatment because the fingers are too short or completely absent. However, the bone growth after lengthening is different between these two conditions. In cases of amniotic band syndrome, the lengthened bones usually grow at the same rate as the other bones in the hand. In cases of symbrachydactyly, the bone growth is different not only for the lengthened bones, but also for the other bones in the hand, thus causing the hand shape to change undesirably. In cases of symbrachydactyly involving children under five years of age where bone lengthening is needed, several subsequent lengthening procedures are required after the first lengthening. Young patients belonging to the failure formation-longitudinal arrest group should be carefully assessed because the growth potential of bones is different in each case.

TREATMENT OPTIONS

Two lengthening techniques can be used depending on the original bone length. In general, if the bone is longer than 20 mm the ''monolateral device'' (one side of the digit) is indicated. If the bones are shorter, it is more difficult to attach and use the lengthening device. In cases where the bone is shorter than 20 mm, a ''multilateral device'' (both sides of the digit) is used. The monolateral technique is preferable for patient comfort and ease of pin care. The most important considerations for both treatment options are accurate bone measurement and careful planning before the operation, such as the location of the osteotomy, pin diameter, lengthening method, and appropriate apparatus. It is essential to protect the growth plate, which is often very close to the osteotomy. We recommend doing precise drawing from radiographs and using it for preoperative planning.

SURGICAL TECHNIQUES

Distraction Apparatus

There are many kinds of distraction devices for the hand. One of them is the Ikuta distraction apparatus for small bones such as metacarpals and phalanges (9,10). There are two different sizes of Ikuta apparatus, large and small, which allow the use of wires and screws for lengthening to accommodate various bone sizes. The pin clamps accommodate wires and screws under 2.0 mm in diameter. Kirshner wires (0.7, 1.0, 1.2, 1.5, and 2.0 mm diameters), and 1.8-mm Shantz screws (Aesculap, Melsungen, Germany) are normally used with this device. Distraction is achieved by turning the knobs. A one-third turn on the rod is equal to 0.25 mm of distraction.

Monolateral Technique

The monolateral lengthening technique (Fig. 1) is indicated for longer bones. Larger diameter wires, utilized as half pins, and Shantz screws are used for this technique, providing greater stability. The skin incision is made on the dorsal side of the bone, exposing it for the osteotomy. The periosteum is carefully elevated. After exposing the bone, the screw holes are made in it according to the preoperative planning. If the bone is not long enough to allow placement of

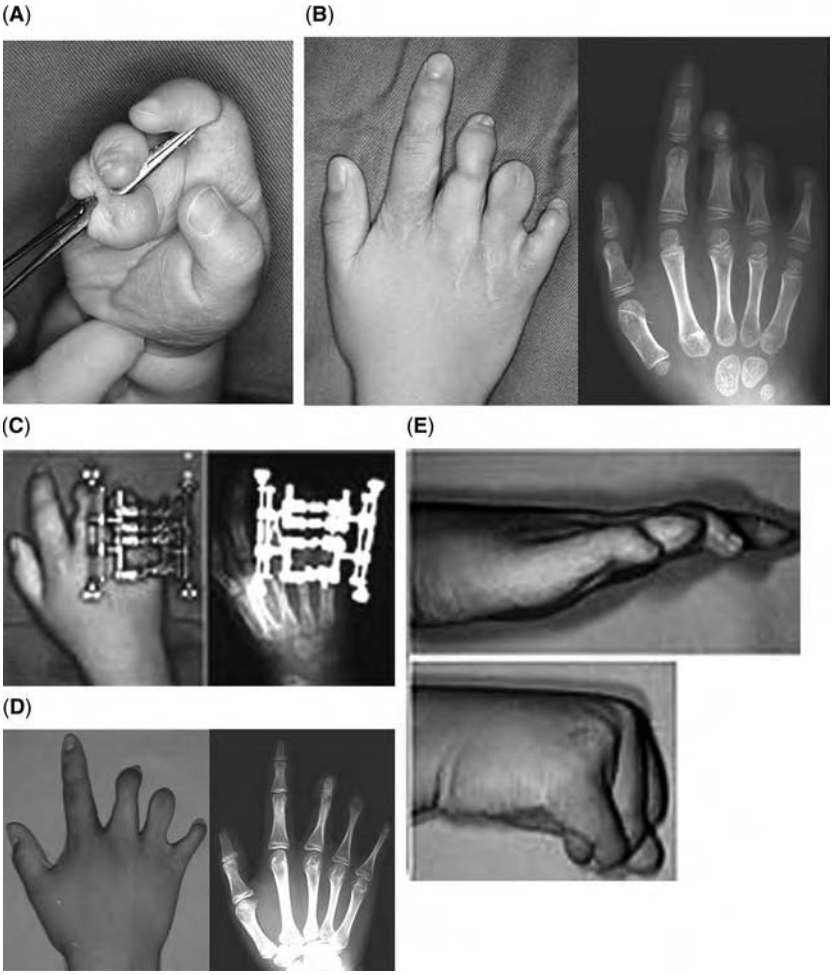

Figure 1 Monolateral technique for lengthening in the case of a five-year-old boy with shortened ring and little fingers due to amniotic band syndrome of the right hand. (**A**) A photograph of the right hand at six-months-old on the first visit to the clinic. Separation of middle, ring, and little fingers and web plasty were performed when the patient was one year of age. (**B**) A prelengthening photograph and a radiograph of the right hand. (**C**) Bone lengthening of ring and little proximal phalanges using the monolateral technique. Apparatus fixation was made across the metacarpophalangeal joints. (**D**) A photograph and a radiograph of the right hand at 110 months after the lengthening. (**E**) Grasping action of the right hand. The patient reported that he could grasp larger objects after lengthening. *Source*: From Ref. 7.

all fixation pins, it may be necessary to span the device across the joint (Fig. 2). The pins are then inserted on the dorsal side of the bone either in a sagittal plane or offset 45° between the dorsal and lateral sides of the bone, while maintaining a perpendicular orientation to its axis. The device should be assembled and temporarily connected to the pins on the hand. It is done to check if the length of pins is adequate to keep apparatus close to the hand and at the same time to have clearance between the skin and the device. The osteotomy is done at the point selected preoperatively. It is preferable to use an osteotome after multiple drill holes are made, but an oscillating saw can be used if there is concern of fracturing into the pin site. The frame is attached and care is taken to avoid displacement of the osteotomy. The operation is completed by suturing the periosteum, where possible, and then closing the skin.

Multilateral Technique

The multilateral technique (Fig. 3) uses wires only in conjunction with two apparatuses attached on each side of the wire for lengthening by simultaneous distraction. This technique is indicated

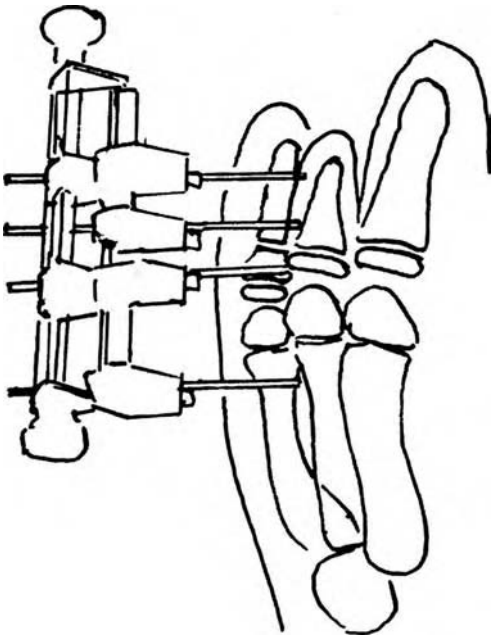

Figure 2 Apparatus fixation across the left little MP joint in a case of amniotic band syndrome. The pins for the proximal attachment are placed accross the MP joint. The proximal pin is placed in the metacarpal and the distal pin is placed in the proximal part of the osteotomized proximal phalanx. *Abbreviation*: MP, metacarpophalangeal.

for smaller bones that do not have adequate volume for accommodating screws. Therefore, it is often used for younger children. The osteotomy and wire positions are selected in the same way as in the monolateral method. However, direction of the wires differs from the monolateral method: wires are inserted in the coronal plane. This method enables lengthening of more than two metacarpals simultaneously in one lengthening treatment (Fig. 4). It should be noted that the thinner the wires, the more flexible they are and they can bend during distraction.

Postoperative Distraction and Treatment

After the operation, a splint is applied to protect the hand. In general, the method of distraction osteogenesis is used to lengthen the bone postoperatively (11–13). The knobs for distraction are kept outside the dressing to allow lengthening without disturbing the dressing. The dressing changes are done two or three times a week. Distraction starts three to five days after the operation with the rate of 0.5 to 0.75 mm per day. This is achieved by turning the knob two or three times a day. Subsequent radiographs are obtained each week throughout the distraction period to check appearance of the regenerate. The lengthening rates are adjusted according to the regenerate formation. The apparatus is removed only when there is radiological evidence of consolidation. The apparatus is detached from the pins one week prior to their removal.

COMPLICATIONS

Complications can occur at various stages of the treatment. At the time of surgery, potential problems are soft-tissue injuries (vessel, nerve, or tendon). Careful and sufficient exposure of the bone should be made during the operation. Potential complications that can occur during the lengthening period are pin loosening, superficial infection, angular deformity, premature consolidation, or insufficient regenerate formation.

Pin loosening is caused by various factors, including poorly positioned pins, loosening of the apparatus, and pin-tract infections. During surgery, the pin must be carefully inserted at the correct angle to ensure the proper application of force to the bone. The apparatus must be checked to ensure all bolts and knobs are correctly set and tightened.

Superficial pin-tract infection is successfully treated with prompt administration of oral antibiotics. Deep pin infection is treated with intravenous antibiotics, pin removal, or exchange.

Angular deformity of the regenerate can occur both during and after the distraction, and may be caused by imbalance and soft-tissue tension. This is best prevented with use of a stable

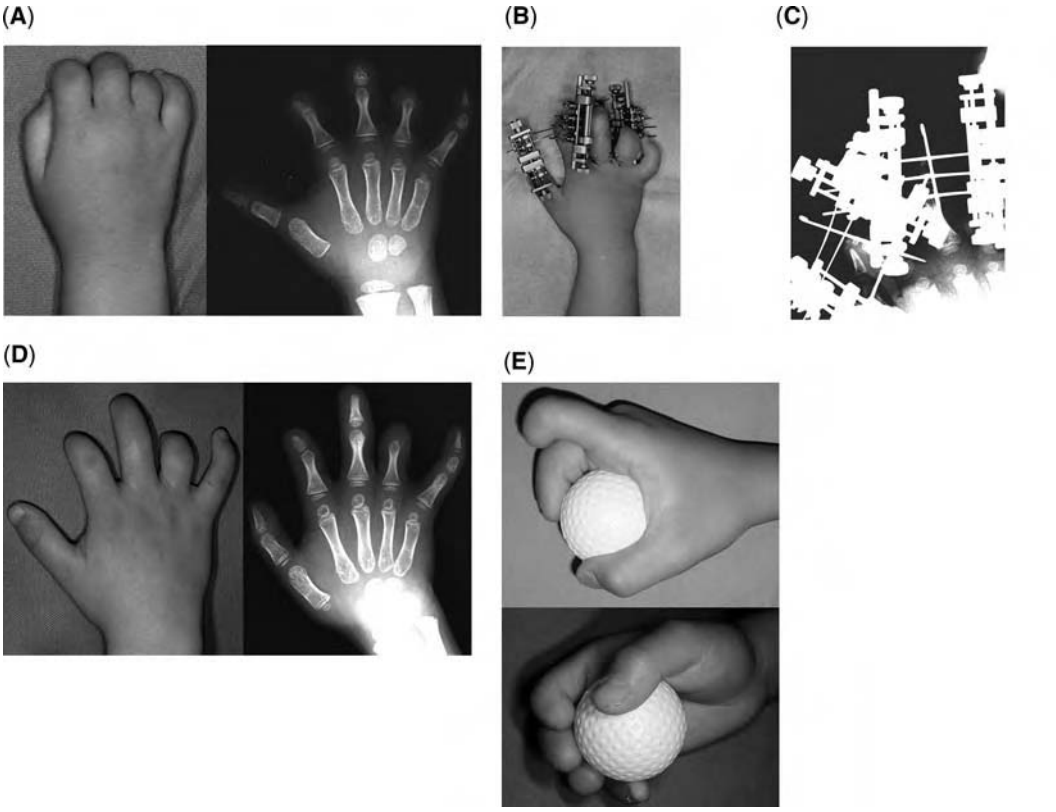

Figure 3 Multilateral technique for the lengthening of index and middle phalanges in a three-year-old boy with amniotic band syndrome. (**A**) A preoperative photograph and a radiograph of the right hand. The middle phalanges of index and middle were present but shorter than 10 mm. (**B**) Appearance after the operation. More care is needed handling the frames and taking care of the wound than in the monolateral method. (**C**) The procedure for the lengthening of the middle finger. The middle phalanx was cut center of it. A K-wire was penetrated in the distal osteomized fragment for the distal attachment, the other K-wires were penetrated in the proximal fragment of the middle phalanx and proximal phalanx for the proximal attachment. A longitudinal K-wire was inserted and fixed to prevent rotation of the distal fragment. Then the apparatures were attached to the K-wires on bilateral sides of the middle finger. (**D**) The results of lengthening shown on a photograph and a radiograph. (**E**) The patient reported that he could grasp larger objects such as a ball and throw it, after lengthening. *Source*: From Ref. 10.

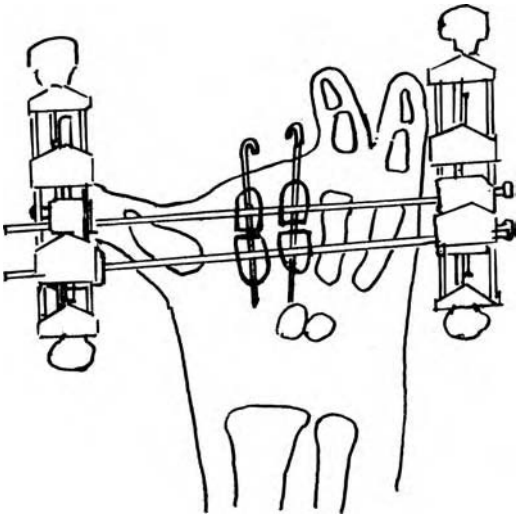

Figure 4 Simultaneous bone lengthening of the second and third metacarpals of the right hand in a case of symbrachydactyly. Bone lengthening for the two metacarpals was performed in one lengthening treatment.

frame, insertion of adequate number of pins, and release of soft-tissues as needed. If angular deformity does develop that would compromise function, correction osteotomy should be considered.

Premature or delayed consolidation can be unpredictable. Although children's fractures heal more readily than those of adults, the regenerate formation is different in each case. Careful radiological evaluation is required in order to adjust distraction rate accordingly during lengthening and to confirm a bony union at the time of apparatus removal.

After lengthening, regenerate fracture, angular deformity at an adjacent joint, and a bony prominence at the fingertip can occur.

A painful bony prominence at the fingertip is the most frequent complication in the lengthening of the terminal bones. Skin problems frequently occur as a result of insufficient skin stock to cover the newly lengthened digits. Although skin tension around the pins decreases during the consolidation phase, it often compromises the lengthening process. In such patients, the lengthening is often stopped early and the target length is not achieved.

FUTURE DIRECTIONS

There are a number of potentially interesting avenues of development in the area of congenital hand deformities, including enhancement of bony regenerate (14), tissue engineering, fetal surgery, and gene therapy.

Tissue engineering techniques may become effective methods for developing artificial biosynthetic fingers. Currently, craniofacial reconstruction is performed using collagen substitute and cultured chondrocyte graft (15). Fingers have more complex structures and function. At present, it is difficult to make organs composed of multiple tissues. In the future, technical innovations in relation to the culture and synthesis of tissues will probably enable us to reconstuct fingers using biosynthetic technologies.

Fetal surgery may develop as a treatment for congenital hand deformity. The early diagnosis of hand deformity in a young fetus became possible owing to the development of diagnostic instruments and techniques such as ultrasonography. An experimental tissue graft on a fetus has been tried (16). Further development of fetal surgery in combination with stem cell and embryonic research could be another way to approach hand reconstruction.

Many hand deformities are caused by genetic disorders. Through the process of genetic research, the etiologies of congenital hand deformities will be clarified gradually (17). Someday gene therapy could become a powerful tool in prevention of congenital anomalies of the hand.

REVIEW OF LITERATURE

Authors	Journal, Year	Title	Number of Patients	Results	Conclusions
Kessler	J Hand Surg (A), 1978	Experience with distraction lengthening of digital rays in congenital anomalies	11	All good and excellent	Lengthening of a digital ray or hypoplastic palm can improve function
Ogino	J hand Surg (B), 1994	Digital lengthening in congenital hand deformity	11	One symbrachydactyly case not improved	Metacarpal lengthening is a useful and reliable treatment in order to achieve functional and aesthetic improvement
Seitz	Orthopedics, 1995	Digital lengthening using the callotasis technique	14	All good and excellent	Adequate control of the distal segment, appropriate choice of location for corticotomy, and gentle handling can improve results

(*Continued*)

REVIEW OF LITERATURE (*Continued*)

Authors	Journal, Year	Title	Number of Patients	Results	Conclusions
Pensler	Plast Reconst Surg, 1999	Distraction osteogenesis in the hand	12	All good and excellent	Distraction osteogenesis has proven to be superior to the standard treatment modalities
Dhalla	J Hand Surg (A), 2001	A comparison of two techniques for digital distraction lengthening in skeletally immature patients	16	All good and excellent	The use of the single half-pin/K-wire technique allows substantial lengthening for bones shorter than 23 mm
Miyawaki	J Bone Joint Surg (A), 2002	Bone lengthening for symbrachydactyly of the hand with the technique of callus distraction	4	Pinch imrovement in all patient	Bone lengthening is a safe, simple, and cost effective method for Muller type-D symbrachydactyly
Matsuno	J Hand Surg (A), 2004	Bone lengthening for the congenital differences of the hands and digits in children	14	Two symbrachydactyly cases changed hand shape	Careful patient selection is needed in young symbrachydactyly patients

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Part XI STATURE LENGTHENING

43 Stature Lengthening: Skeletal Dysplasia

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INTRODUCTION

This chapter will discuss our latest methods of management of lower and upper limb deformities and short stature in patients with achondroplasia and hypochondroplasia. These represent the vast bulk of skeletal dysplasias. Other skeletal dysplasias are less common, and we tend to focus more on limb realignment rather than lengthening for those other conditions.

Children with achondroplasia typically present for orthopedic evaluation after already having been diagnosed by a geneticist. Achondroplasia accounts for about 75% of all dwarfism. However, it is nonetheless a rare condition, affecting 1/50,000 children. Therefore, most achondroplastic children will grow up in communities without any peers sharing the same diagnosis. The gene for achondroplasia is a point mutation on the fibroblast growth factor receptor-3 gene, located on the short arm of the fourth chromosome. Achondroplasia is transmitted as an autosomal dominant gene, with the double dose being lethal. Still, 80% of cases are spontaneous mutations, and these patients are born of normal stature parents. Hypochondroplasia is genetically similar to achondroplasia, with the same gene involved, but with a slightly different nucleotide change. Hypochondroplasia is phenotypically a much milder form of dwarfism. In this chapter, we focus on the more clinically challenging problem of achondroplasia because hypochondroplasia represents clinically less need for deformity correction and the total amount of lengthening is less.

In addition to the obvious limb deformities of short stature and bowing, children with achondroplasia also have frontal bossing, midface hypoplasia, thoracolumbar kyphosis, delayed developmental milestones, and spinal stenosis. They may suffer from foramen magnum stenosis and hydrocephalus. Therefore, prior to undergoing limb reconstruction surgery, patients with achondroplasia should be evaluated for possible spinal cord compression, hyperreflexia, clonus, sleep apnea, and thoracolumbar kyphosis.

Achondroplastic dwarfism presents with a rhizomelic disproportion, short stature, and several characteristic limb deformities. The primary defect is in endochondral ossification, which affects primarily the central region of the growth plate while sparing the peripheral ring of the growth plate. Therefore, the larger the proportion of central growth plate to peripheral growth plate, the greater the growth inhibition. According to the theory of Ponseti, the distal femur, which has the largest growth plate, is affected the most. This accounts for the rhizomelia of the lower limb. Similarly, the proximal humerus has the largest growth plate in the upper limb, so it is most affected, which leads to upper limb rhizomelia too. The fibula has a relatively small growth plate with a much higher ratio of peripheral to central growth cartilage compared with the adjacent tibia. The growth of the tibia is, therefore, more affected than that of the fibula, leading to relative fibular overgrowth. Overgrowth of the fibula causes developmental varus of the proximal and distal joint lines of the tibia, lateral collateral ligament laxity, and consequently dynamic varus with a lateral thrust during the single-leg stance of gait, and developmental internal tibial torsion. In the proximal femur, the femoral neck has a larger growth plate than the greater trochanteric apophysis. This leads to trochanteric overgrowth, coxa vara, and a short femoral neck.

In the upper extremity, flexion deformity of the elbow is common. This is partly due posterior bowing of the ulna, due to greater stunting of growth of the larger distal radius growth plate than the relatively small distal ulnar physis. In more severe cases, the radial head dislocates to accommodate the slower-growing ulna. The spine is relatively spared inhibition because the vertebrae grow in height from the apophyseal growth plate. This is a peripheral growth plate structure more than a central growth plate. The acetabulum is more anteriorly located in achondroplasts possibly due to differential growth of the three separate limbs of the triradiate cartilage. Because the pelvis is tilted forward and because the iliac side is more inhibited, the acetabulum becomes relatively posterior, shifting the center of the femoral head back. This leads to a flexion deformity of the hip, which is compensated by hyperlordosis of the lumbar spine.

CLINICAL EVALUATION

Prior to lengthening, children with achondroplasia should undergo a thorough orthopedic and neurologic evaluation with special emphasis on joint range of motion (ROM)/stability, including hip flexion contracture, ankle contracture, knee ROM, and elbow ROM. One should also assess gait and look for signs of neurologic involvement. Radiographs should include standing views of both legs (anteroposterior and lateral) and ones of the spine. The following table summarizes the clinical signs associated with these deformities:

Deformity	Clinical Manifestation
Varus distal tibia	Varus foot
Varus proximal tibia	Genu varum
Overgrowth of proximal fibula	Lateral thrust (lateral knee instability)
Internal tibial torsion	Intoeing gait
Overgrown trochanter	Waddling gait
Hip flexion deformity	Hyperlordosis
Bowing of ulna/dislocation radial head	Elbow flexion deformity
Flexion deformity of distal humerus	Elbow flexion deformity
Rhizomelia	Disproportionate short stature

STATURE IN ACHONDROPLASIA

The stature of achondroplastic dwarfs falls significantly below the standard growth charts for children. Therefore, special growth charts have been developed to follow the growth of these children. The average adult height for achondroplastic men is 131 cm and for women is 125 cm. The range (2 SD) of height for achondroplastic men at maturity is 117 to 144 cm and for women is 113 to 157 cm. In comparison, the low normal end of height at maturity for men without achondroplasia is 160 cm and for women is 151 cm. Tall parental height is associated with relatively taller height in achondroplastic offspring. The short stature of achondroplastic patients is not simply due to growth at a different rate than in normal children. These children have a completely different growth pattern. The height multipliers for achondroplastic children are completely different than for normal children (Tables 1 and 2).

GOALS OF LENGTHENING SURGERY FOR ACHONDROPLASIA

The goals of surgery are to correct upper and lower limb deformities while at the same time increasing stature to the low end of the normal height spectrum. This theoretically requires leg lengthening of about 30 cm (men) or 25 cm (women) for the average patient with achondroplasia. Lengthening of the lower extremities can be 10 to 15 cm per session, so the total number of leg-lengthening procedures required to achieve these goals is two to three leg-lengthening sessions. The humeri are lengthened separately. Deformity correction of the

Table 1 Height Multiplier for Achondroplastic Boys

Age (yr)	Average Multiplier
0	2.848
1	2.107
2	1.798
3	1.679
4	1.579
5	1.501
6	1.430
7	1.366
8	1.318
9	1.270
10	1.221
11	1.181
12	1.145
13	1.105
14	1.079
15	1.054
16	1.032
17	1.018
18	1.000

lower limbs for achondroplasia is a well-accepted procedure. Lengthening for stature is still somewhat controversial. This controversy will be discussed in detail below.

CLASSIFICATION AND TREATMENT STRATEGIES FOR LENGTHENING IN ACHONDROPLASIA

Strategy for Juveniles (Young Children)

First Lengthening Between Ages 7 and 10 Years

Simultaneously lengthen both femora and both tibiae a total of 10 cm each (5-6 cm in each femur and 4-5 cm in each tibia) (Fig. 1A–E).

Correct bowlegs and internal tibial torsion.

Average external fixation treatment time is five months.

Second, Third, and Fourth Lengthenings as in the Strategy for Adolescents

Total increase in lower limb length is 30 to 35 cm.

Total increase in upper limb length is 10 cm.

Table 2 Height Multiplier for Achondroplastic Girls

Age (yr)	Average Multiplier
0	2.655
1	1.943
2	1.744
3	1.632
4	1.535
5	1.461
6	1.400
7	1.343
8	1.274
9	1.222
10	1.170
11	1.130
12	1.098
13	1.066
14	1.042
15	1.023
16	1.009
17	1.000

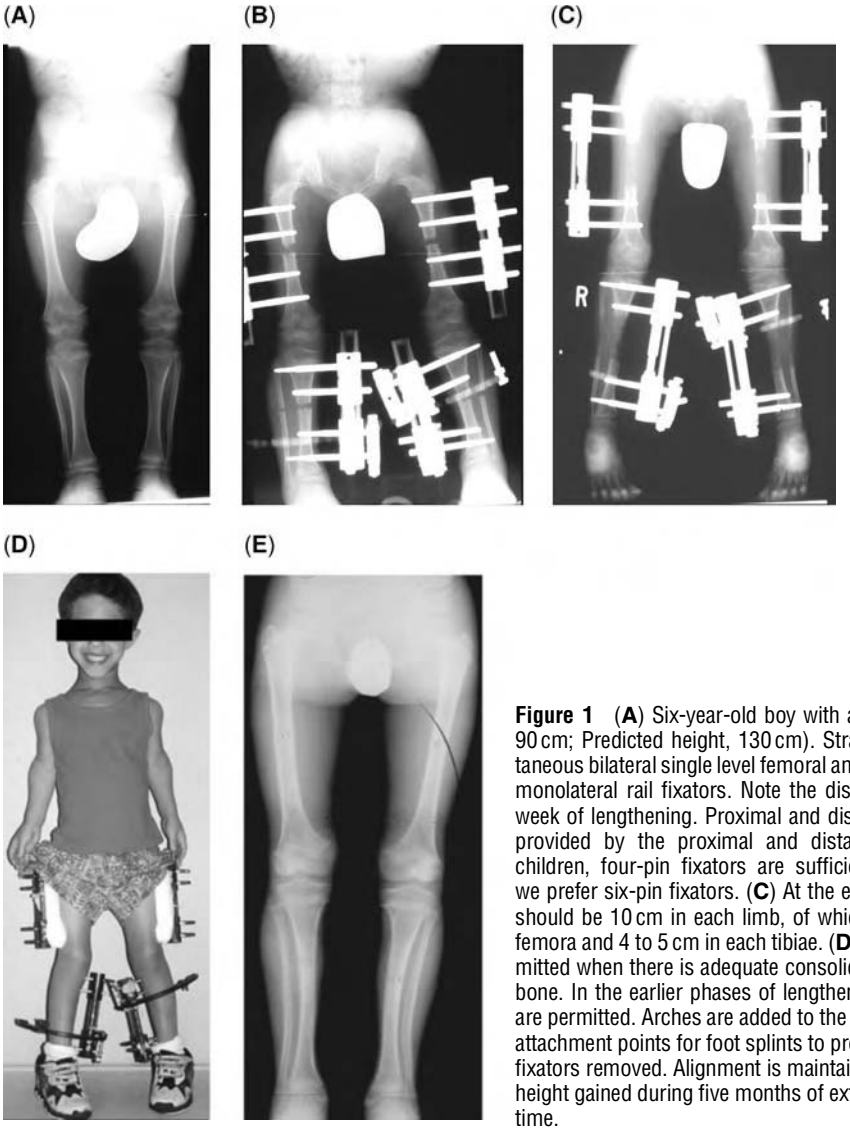

Figure 1 (**A**) Six-year-old boy with achondroplasia (Height, 90 cm; Predicted height, 130 cm). Straight limbs. (**B**) Simultaneous bilateral single level femoral and tibial lengthening with monolateral rail fixators. Note the distraction gaps after one week of lengthening. Proximal and distal fibular fixation were provided by the proximal and distal tibial pins. In small children, four-pin fixators are sufficient. In larger children, we prefer six-pin fixators. (**C**) At the end of lengthening. Goal should be 10 cm in each limb, of which 5 to 6 cm is in each femora and 4 to 5 cm in each tibiae. (**D**) Weight bearing is permitted when there is adequate consolidation of the regenerate bone. In the earlier phases of lengthening, standing transfers are permitted. Arches are added to the tibial fixators to provide attachment points for foot splints to prevent equinus. (**E**) After fixators removed. Alignment is maintained. Ten centimeters of height gained during five months of external fixation treatment time.

Strategy for Adolescents

First Lengthening at About 13 Years of Age

Option 1

Double-level tibial lengthening; if varus proximal tibia and varus distal tibia, then perform double-level bilateral tibial lengthening 10 to 15 cm with deformity correction (Fig. 2).

Typically, more length is achieved in the proximal osteotomy.

Correct the varus deformity of the proximal tibia through proximal osteotomy.

Correct the varus deformity of the distal tibia through the distal osteotomy.

Tighten the lateral collateral ligament during the final stages of the lengthening.

Average external fixation treatment time is six to nine months.

Option 2

Bilateral simultaneous femoral and tibial lengthening; if no significant distal tibial varus, then lengthen both femora and both tibiae 10 to 15 cm (5–8 cm in each femur and 5–7 cm in each tibia) (Fig. 3A–C).

Average external fixation treatment time is five to seven months.

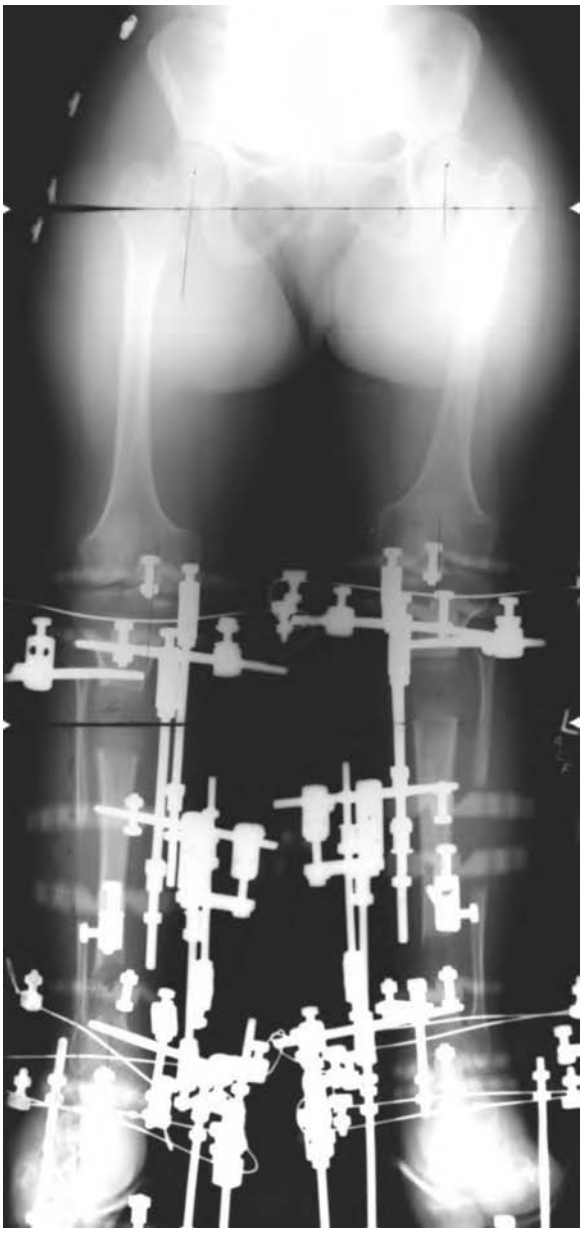

Figure 2 Sixteen-year-old achondroplastic dwarf underwent bilateral simultaneous double level tibial lengthening and straightening. At the end of lengthening, the patient had 6 cm of new bone in proximal osteotomy and 5 cm of new bone in distal osteotomy.

Second Lengthening at Age 14 Years

Lengthen both humeri 8 to 10 cm (Fig. 4A–E).
Correct flexion deformity of elbows if significant (>30°).
Average external fixation treatment time is six to eight months.

Third Lengthening Between Ages 15 and 16 Years

Option 1

Lengthen both femora 10 cm through proximal osteotomy (Fig. 5A–C).
Correct flexion deformity of both hips (this reduces the lumbar hyperlordosis).
Correct varus deformity of both hips.
Average external fixation treatment time is 10 to 12 months.

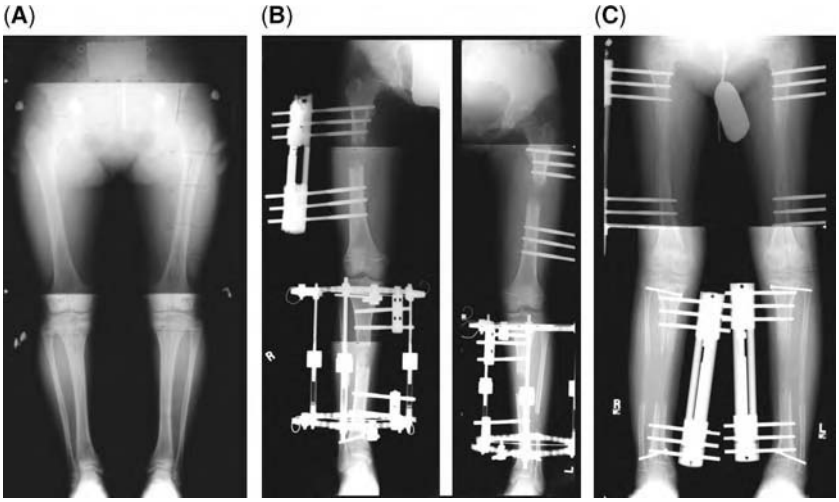

Figure 3 (**A**) Eleven-year-old hypochondroplastic girl underwent bilateral simultaneous femoral and tibial lengthening. No malalignment was observed preoperatively. (**B**) Six-pin monolateral rail fixators were applied to the femora and spatial ring fixators were applied to the tibiae. Initial lengthening in tibiae is with spatial rings and Ilizarov graduated telescopic rods. At the end of lengthening, the spatial struts may be inserted for correction of any residual axis deviation. (**C**) Eleven-year-old achondroplastic boy underwent an aggressive bilateral simultaneous femoral and tibial lengthening to achieve 7.5 cm in each tibia and 8 cm in each femur. Monolateral fixators were used on both femora and tibiae.

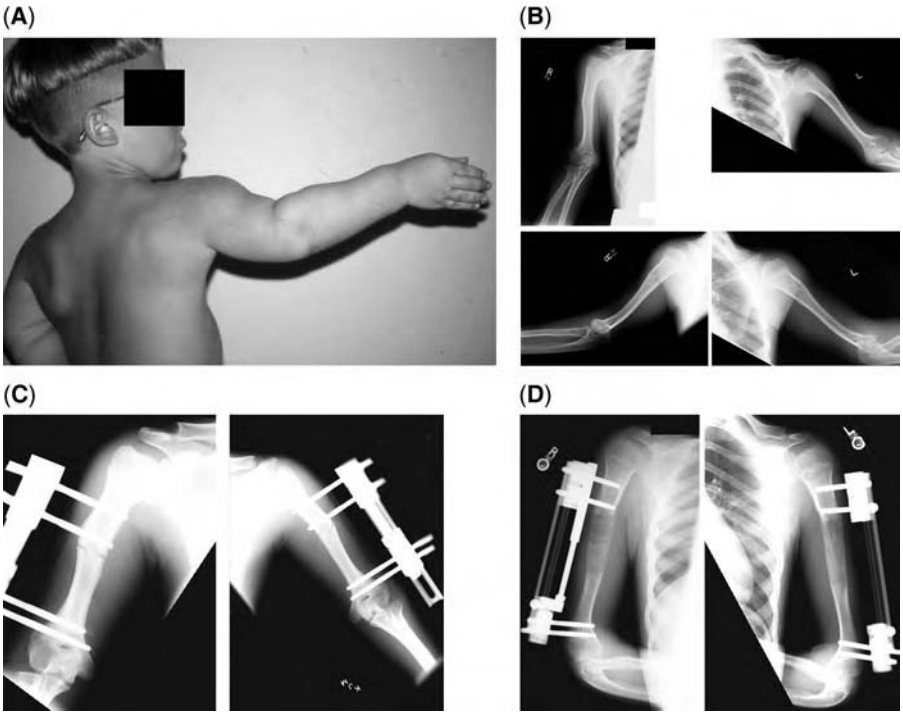

Figure 4 (**A**) Rhizomelic shortening is characteristic of achondroplasia. The humerus is disproportionately shorter than the forearm. The elbow lacks full extension. (**B**) Preoperative radiographs of a 16-year-old achondroplastic boy who underwent bilateral humeral lengthening. (C) Bilateral monolateral four-pin fixators applied. (**D**) Anteroposterior view of both humeri at the end of consolidation phase. About 8 cm of new bone has been achieved. (**E**) Fracture at the host-regenerate bone junction on the right side occurred from a fall one month postremoval of fixators. Intraoperative sequence of fracture reduction, reaming with flexible hand held reamer, and stabilization with Rush pin.

(E)

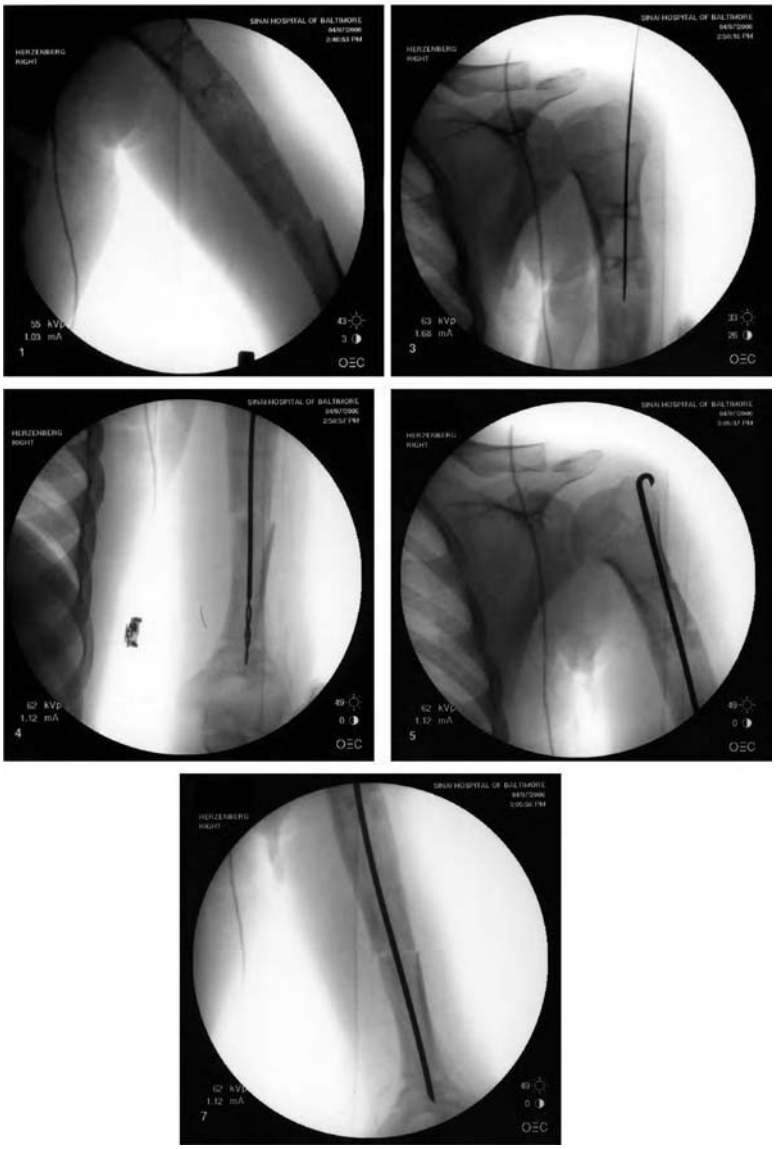

Figure 4 (*Continued*)

Option 2

Or again simultaneously lengthen both femora and tibiae for a total of 10 to 15 cm.
Correct flexion deformity of both hips to reduce lumbar hyperlordosis.
Correct any distal femoral varus or valgus.
Correct any residual tibial deformity.
Average external fixation treatment time is five to seven months.

Fourth Lengthening at Age 17 to 18 Years

If needed, final lengthening of the femora, preferably with intramedullary techniques (Fig. 6A–B).

By lengthening the arms during the period of time between the two lower-limb lengthenings, the legs get a well-needed break from lengthening. The minimal time between lengthenings is generally at least 6 to 12 months after the device is removed.

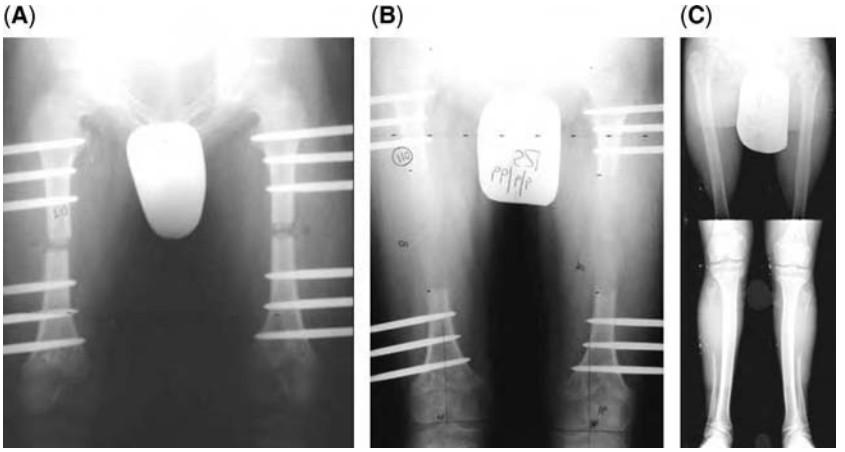

Figure 5 (**A**) Bilateral femoral lengthening with monolateral rail fixators. Six 6-mm pins are needed in adolescents to provide sufficient stability. (**B**) After 10 cm lengthening. Limited four-point gait is allowed in consolidation phase. (**C**) After external fixator removal. The femora are well aligned. Residual valgus is present in the tibiae, which was later corrected during a second tibial lengthening. Note the resected fibulae. This had been done as a youth to prevent the tibiae from growing into varus (Kopits procedure).

During bilateral tibial lengthenings, the patient is able to weight bear using a walker, crutches, or canes if using circular external fixators. Longer trips are by wheelchair. During bilateral simultaneous femoral plus tibial or bilateral femoral lengthenings, the patient is not permitted to walk on monolateral fixators during the lengthening phase. Weight bearing is allowed during the latter half of the consolidation phase. Standing is allowed for transfer only prior to that. Ambulation is by wheelchair only during the lengthening phase. During bilateral humeral lengthening, there is almost no restriction on activities.

The advantage of repeating simultaneous bilateral femoral and tibial lengthenings is that the total external fixation treatment time is reduced significantly. Furthermore, this method enables the maximum stature gain in the least time. For example, bilateral 10 cm femoral lengthenings take 10 months of external fixation time, while bilateral femoral and tibial lengthenings of 5 cm each for a total of 10 cm take only five months. The disadvantage of bilateral simultaneous femoral and tibial lengthenings is the increased nursing care and physical therapy involved.

SURGICAL TECHNIQUES

Bilateral Simultaneous Femoral and Tibial Lengthening

This is a lengthy procedure, and we often work with two teams sequentially. Start by performing the necessary soft-tissue releases under tourniquet control. For the first lengthening, there may be no need for soft-tissue releases. For the second lengthening, it may be advisable to lengthen the fascia lata, rectus femoris muscle, and gastrocnemius-soleus muscle (Fig. 7A–D).

Fixators

Use two LRS rail systems (Orthofix, McKinney, TX, USA) for the femora and two Taylor Spatial Frames (TSF) (Smith & Nephew, Memphis, TN, USA) or LRS rail system for the tibiae. Typically, the goal is length, so the femoral fixators may be applied parallel to the mechanical axis (Fig. 8). After placing the six pins, check knee ROM and consider fascia lata resection or sectioning, either percutaneously around the pin sites or through an open incision just anterior and distal to the distal pin cluster (Fig. 7D). For the tibial lengthening, either use a six-pin LRS rail system or a two-ring TSF. Either way, it is important to fix the fibula proximally and distally. Consider an anterior compartment fasciotomy. Pain can be difficult to judge postoperatively and fasciotomy eliminates the possibility of compartment syndrome. Always apply splints to the feet, suspended by Velcro straps attached to the tibial fixator.

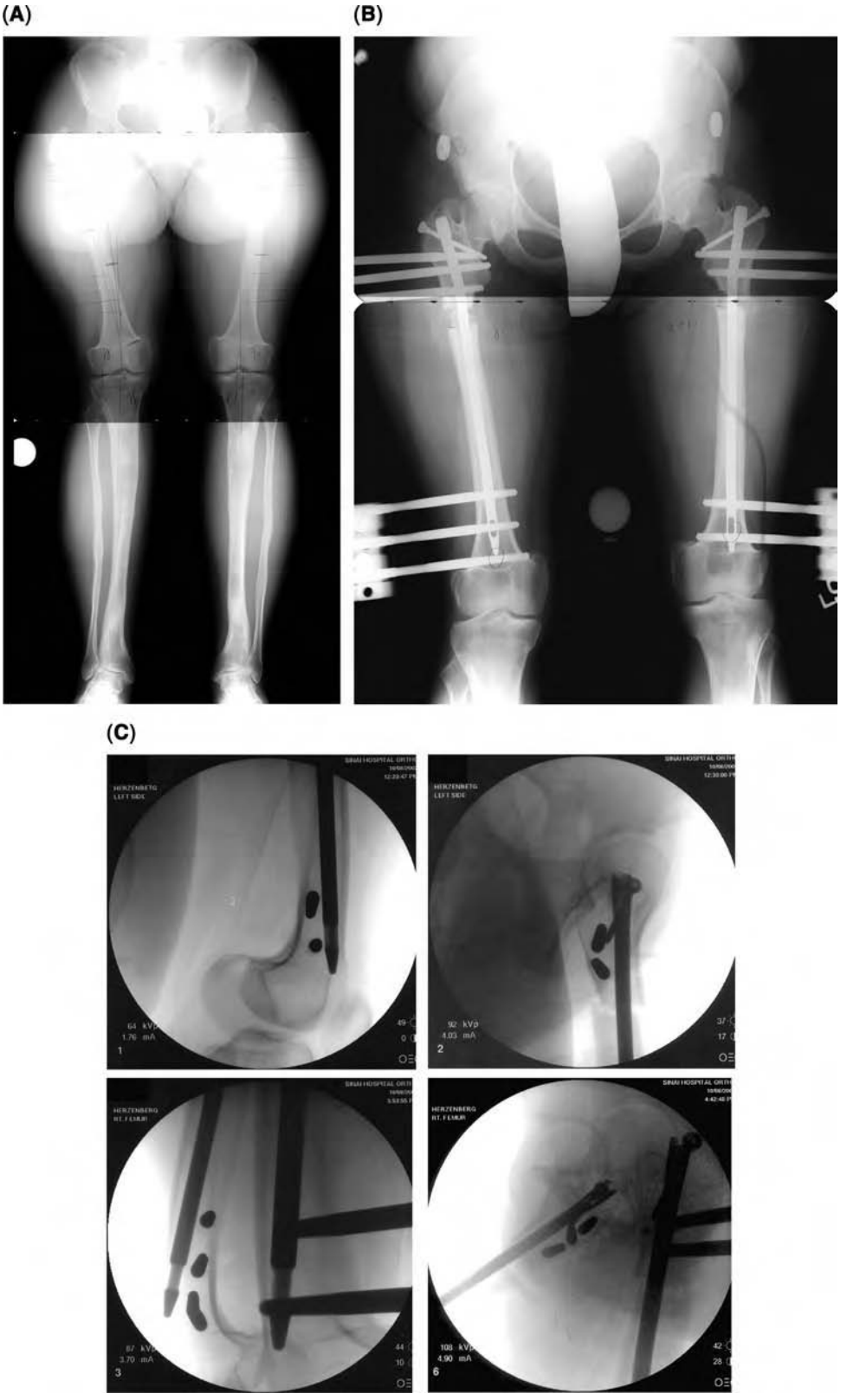

Figure 6 (*Caption on next page*)

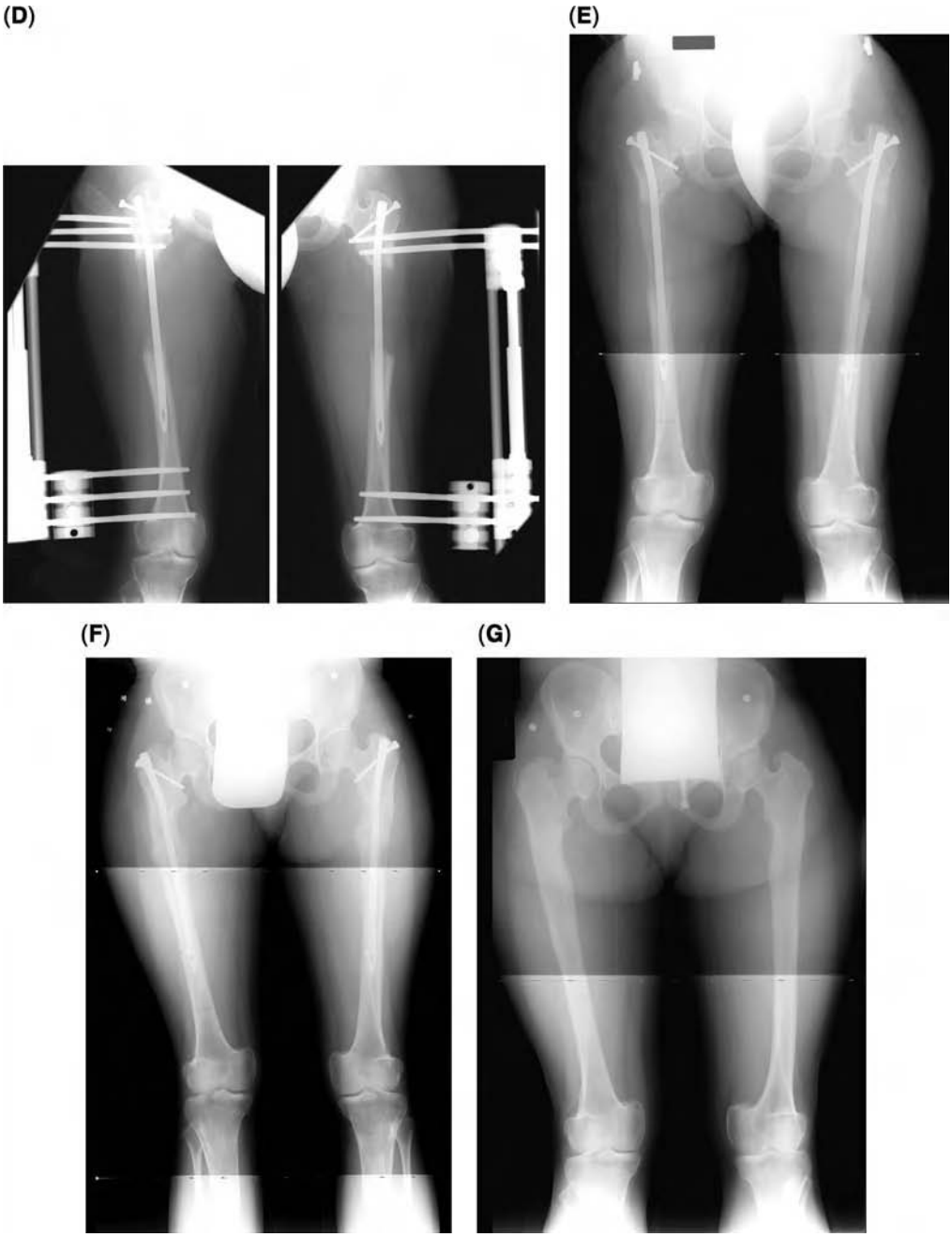

Figure 6 (*Continued from previous page*) (**A**) Young achondroplastic adult after healing of bilateral double level tibial lengthenings. (Same patient as in Fig. 2). (**B**) Bilateral femoral lengthenings over nails. Eight-millimeter humeral nails were used due to the small canal size. On the right side, a six-pin monorail device was used for distraction. The left side distracted well with only four pins. (**C**) Intraoperative fluoroscopic images showing the pin placement. The external fixation pins must not have contact with the intramedullary nail. (**D**) Anteroposterior (AP) view of right and left femur at the end of lengthening. Nine centimeters achieved. (**E**) Two weeks after insertion of distal locking screws and removal of fixators. The regenerate bone continues to heal under the protection of the intramedullary rods. (**F**) Three months after fixator removal, the regenerate bone is solid and hypertrophied. (**G**) AP view of both femora following removal of rods.

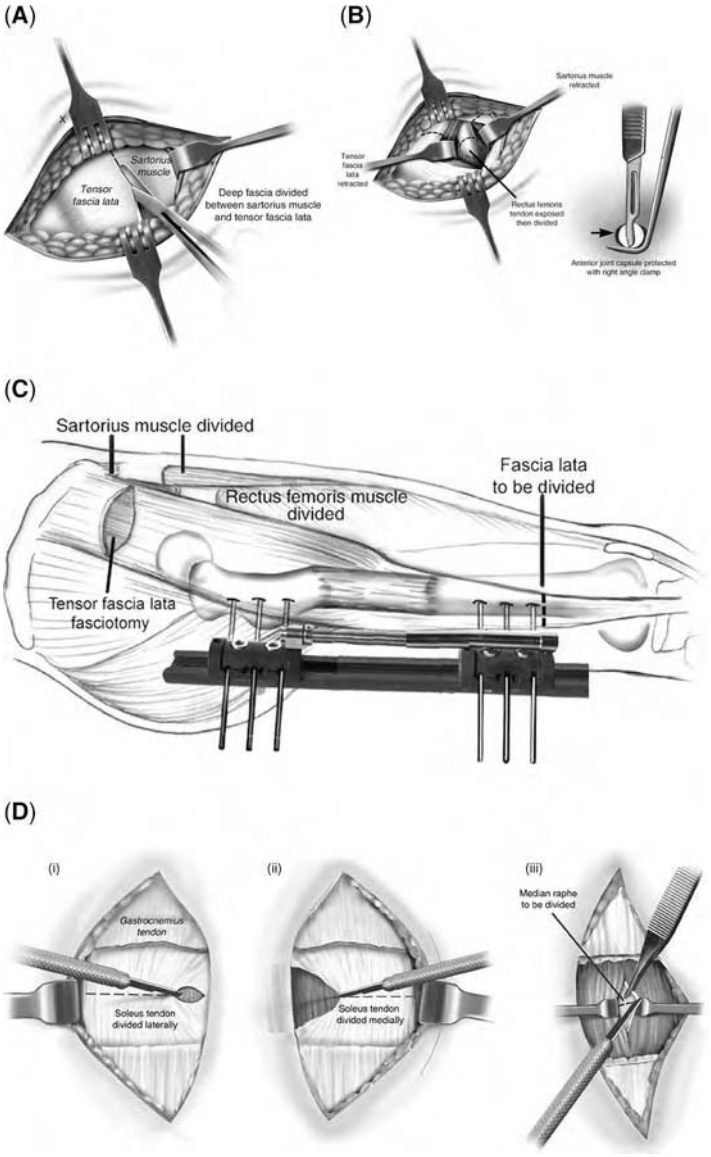

Figure 7 (**A**) Surgical approach for the proximal soft tissue release of the rectus femoris tendon. The fascia over the tensor fascia lata (TFL) and sartorius muscle should also be released. (**B**) Deep dissection for the proximal soft tissue release of the rectus femoris tendon. (**C**) In addition to releasing the proximal tendon muscle of the rectus femoris, the fascial membrane over the TFL and sartorius muscle is also incised. Distally, the fascia lata is divided through a separate incision near the distal pin cluster. (**D**) Vulpius recession of the gastrocnemius-soleus muscle (i) dividing the lateral edge of the tendon, (ii) dividing the medial edge of the tendon, and (iii) dividing the central portion and the median raphe. *Source*: Panel (D) reprinted from Lamm BM, Paley D, Herzenberg JE. Gastrocnemius soleus recession: a simpler, more limited approach. J Am Podiatr Med Assoc 2005; 95(1):18–25.

Bilateral Double-Level Tibial Lengthening

This is typically indicated when there is upper and lower tibial varus deformities. Begin by cutting the fibula and passing the Gigli saw at the proximal metaphyseal level. Distally, the osteotomy may be either diaphyseal or supramalleolar (metaphyseal). The Gigli saw is appropriate for metaphyseal levels but not diaphyseal levels. The very distal Gigli cut can be around both the tibia and fibula together. We recommend tibiofibular stabilization proximally and distally with a 4.5-mm solid bone screw (Fig. 9). This is accomplished by first drilling 1.5-mm wires from the head of the fibula into the tibia proximally and from the distal metaphysis of the fibula

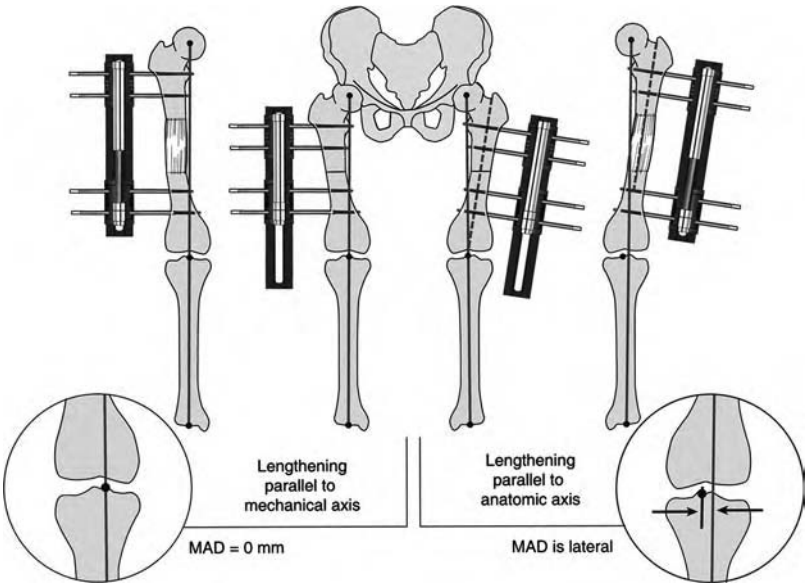

Figure 8 In the situation where there is no preoperative deformity, the monolateral fixator should be aligned with the mechanical axis, not the anatomic axis. Lengthening along the anatomic axis may induce valgus mechanical axis deviation.

distally. Check the position on lateral image intensification to prove that the fibula is captured and then ream over the wires with a 3.2-mm cannulated drill. The proximal screw may be deliberately left long, ''outside'' the skin medially, so that it can be easily removed toward the end of distraction. This allows the fibula to descend as the lengthening continues, thus tightening the lateral collateral ligament, if lax preoperatively. Only one fibular osteotomy is necessary. Use a three-ring external fixator such as the Ilizarov or TSF. The proximal ring is parallel to the knee, the distal ring is parallel to the ankle, and the middle ring is perpendicular to the diaphysis. There may not be room for TSF struts on a very short tibia. In such cases, it is possible to use TSF rings that are connected with regular Ilizarov hinges and rods. With some careful intraoperative planning, the half-pin fixation can be planned to allow later conversion to TSF struts in order to better correct rotational deformities and other unexpected axial deviations that

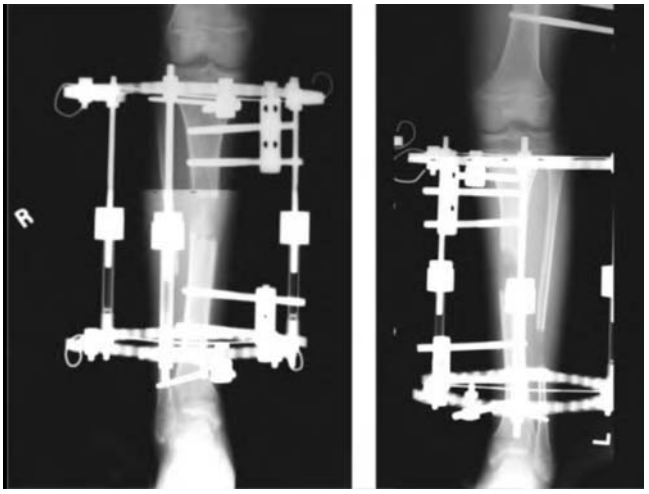

Figure 9 Hypochondroplastic dwarf undergoing bilateral tibial and femoral lengthenings. Tibiofibular stabilization with proximal and distal 4.5-mm solid bone screws. The distal screw should slant upwards to resist the upward pull of the fibula.

may occur during lengthening. Once the length has been achieved, there is more room between the rings to accommodate the TSF struts, so they can be added in the out-patient clinic setting to fine-tune the correction. Vulpius gastrocnemius-soleus recession is recommended for all double-level lengthenings. The hindfoot should be included in the frame by extending the distal ring to the foot for calcaneal fixation. The two tibial osteotomies are made with a Gigli saw if in the wide metaphysis and with an osteotome and drills if made through the diaphysis only. The osteotomies are lengthened at 0 to 75 mm per day at each level. Typically, bone formation is better proximally and so about 60% of length gained is from the proximal level. During the consolidation phase of treatment, the frame can be dynamized by "decommissioning" the half-pins on the intercalary segment. Pins are decommissioned by removing the cube to which they are attached but leaving the pin itself in situ. This is simpler than actually removing the pins.

Bilateral Femoral Lengthening with Correction of Hip Flexion Deformity and Coxa Vara

Correction of hip flexion contracture is done to help ease the strain on the lumbosacral spine, which is excessively lordotic. Typically, the amount of acute extension at the osteotomy site is about 15 to 20° (Fig. 10A–B). At the same time, it is useful to acutely valgusize the osteotomy in anticipation of some varus that will occur with a monolateral fixator under tension during lengthening (Fig. 11A–B). Precise pin placement is the secret to this procedure. We use the LRS rail system with straight clamps. The upper three pins are placed with the cannulated wire/drill technique at a slope to induce acute apex anterior correction. The middle pin is the pivot point for this correction. Typically, the one to two to four pin holes are used proximally and the one to three to five holes used distally. The distal three pins are placed in the typical pattern, perpendicular to the mechanical axis of the femur, which is not far from the anatomic axis in most patients with dwarfism. Cut the bone just below the lesser trochanter, and acutely translate the upper piece anteriorly and the lower piece posteriorly. Wait five days and begin distraction.

Bilateral Humeral Lengthening with and Without Correction of Flexion Deformity

This is discussed in Chapter 39 on Humeral Lengthening.

Lengthening Over Nail/Internal Lengthening Nail

In older adolescents and adults with achondroplasia, it is not unreasonable to consider lengthening over nail (LON) or internal lengthening nail (ILN) methods. These are discussed at length in other chapters in this book. Anatomically, a person with achondroplasia has a short femoral neck, making trochanteric entry safer and easier. This may necessitate the use of a tibial ILN in the femur or a humeral intramedullary nail for LON of the femur (Fig. 12A–C).

COMPLICATIONS OF LIMB LENGTHENING FOR ACHONDROPLASIA

Pin Site Infection

This is the most common minor complication of external fixation treatment. Most pin site infections are superficial presenting with pain and tenderness, redness, and drainage from the pin site. Systemic symptoms such as fever are uncommon. All patients are given a prescription for an oral antibiotic before discharge from the hospital with instructions on self-diagnosis and treatment of pin site infection. This minimizes delay of treatment even if it leads to over-treatment in some cases. If the signs and symptoms subside, the doctor is never notified. If symptoms do not clear, then the patient is instructed to call after 48 hours. At this point, a culture is taken and, if indicated, the antibiotic changed according to the specific sensitivities of the offending organism. If there is any necrotic tissue around the pin site, this is debrided in the office. Prevention of pin infection includes minimizing pin-skin motion especially in the proximal thigh, where there is more ample soft tissue. This can be achieved by dressings that apply direct pressure to the skin. Daily pin care consists of showering with soap daily and drying the pin site with a fresh clean towel. Any crust or scab at the pin site should be picked off with forceps. Cleaning of pins with swabs and saline or peroxide is avoided if the pin site is quiet. This avoids intentionally irritating a nonproblematic pin site. The key to

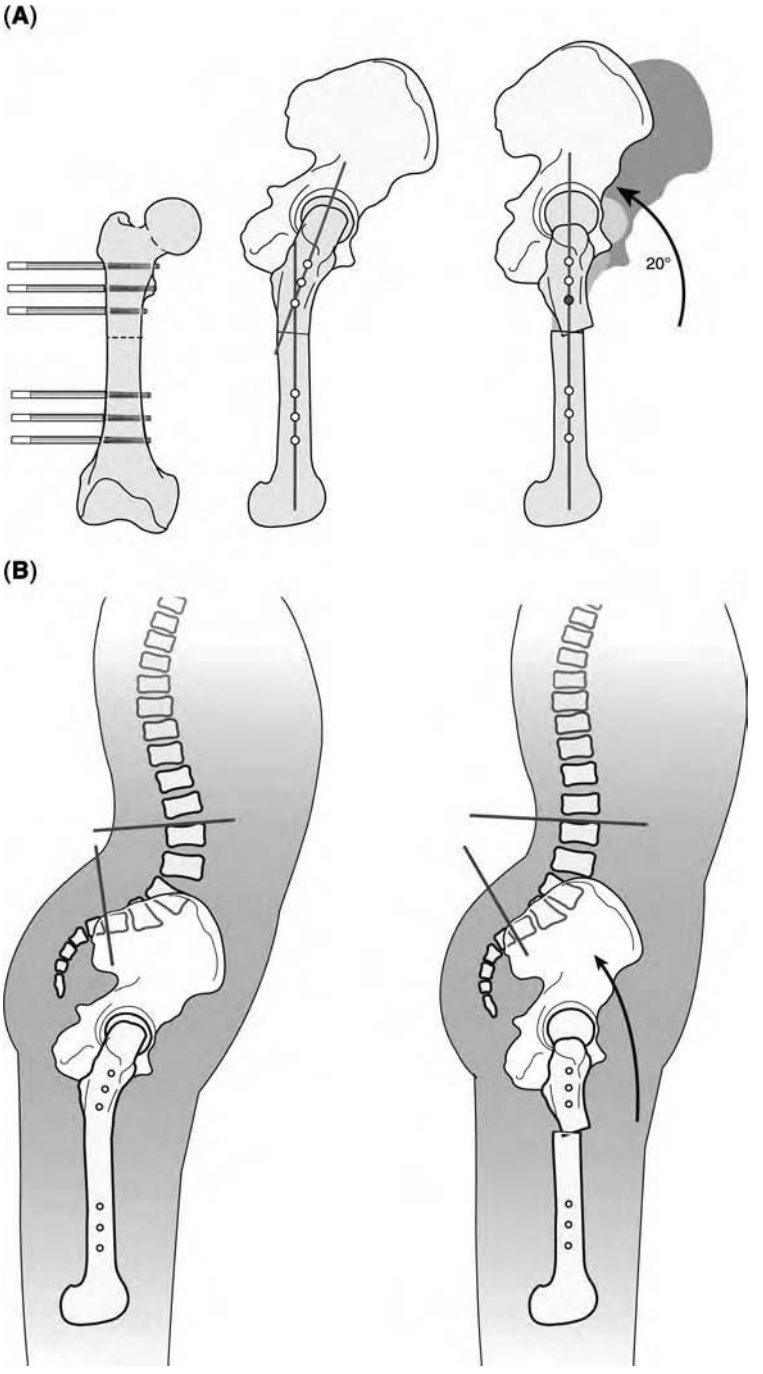

Figure 10 (**A**) The proximal three pins are inserted off axis in the sagittal plane. When the bone is cut and straightened to fit all six pins on the straight monolateral fixator, the proximal femur is extended to create a 20° apex anterior angular osteotomy. (**B**) The proximal extension osteotomy alleviates the hip flexion contracture and subsequently relieves the lumbar hyperlordosis. The hamstrings will further pull the pelvis into a straighter position during the lengthening, especially if the anterior soft tissues are released as described in Figure 7C.

cleaning the pin sites is as for any wound: removal of any not vital tissue around the pin site and washing the wound with water (shower). When fielding phone calls about pin site infection from patients with fixators, one should always ask pointed questions about the patient's well being. In particular, ask if the child is ill appearing, sickly, glassy eyed, pale, flushed, or

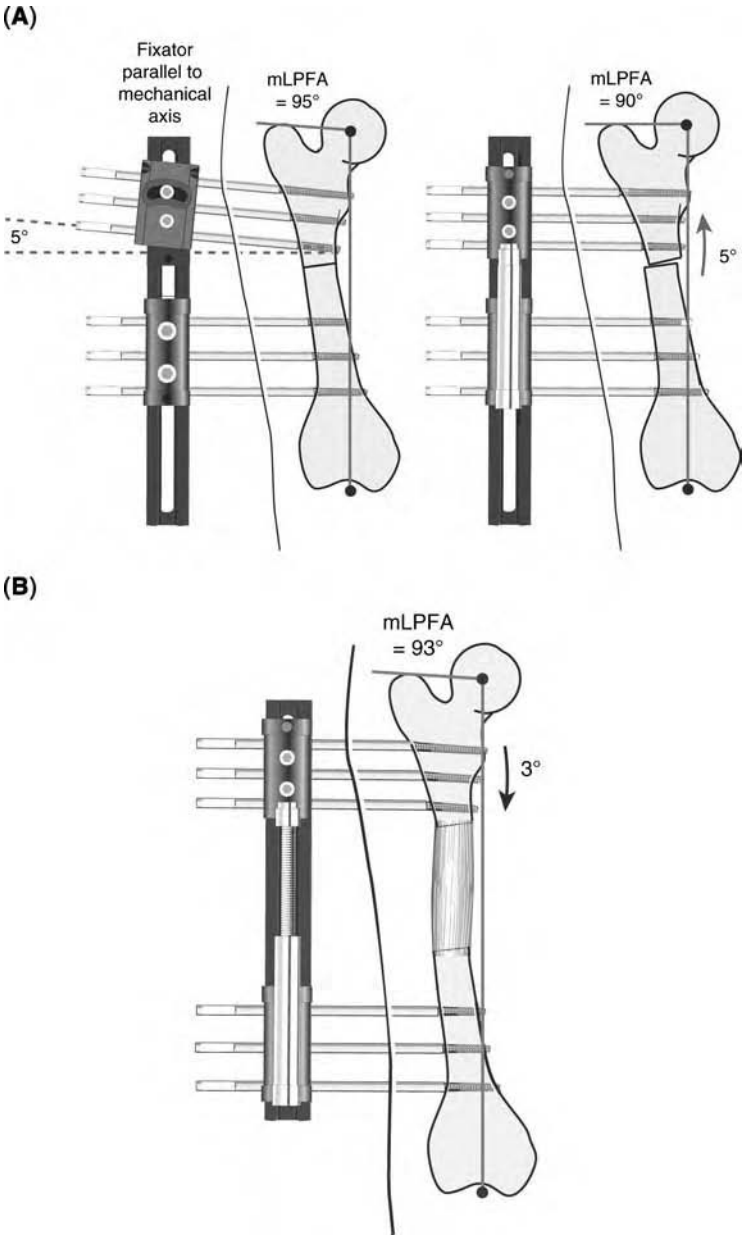

Figure 11 (**A**) The proximal three pins are deliberately inserted in about 5° of varus in the frontal plane. Once the bone is cut, the proximal pins are rotated to become parallel with the distal pins and inserted into the straight monolateral fixator. This causes an acute 5° of valgus at the osteotomy site. This will likely revert to neutral during lengthening from the force of the pull of the medial muscles. (**B**) During lengthening, the force of pull of the medial muscles causes the 5° of valgus to return back to the preoperative baseline alignment.

tachycardic. These could be symptoms of a severe, potentially life-threatening infection such as toxic shock or necrotizing fasciitis.

Nerve Complications

Nerve problems are common during bilateral double-level lengthening of the tibia and less common with single-level lengthening of the tibiae and femora or combined femora and tibiae. It is not clear why the incidence is increased in the double-level group because the nerve

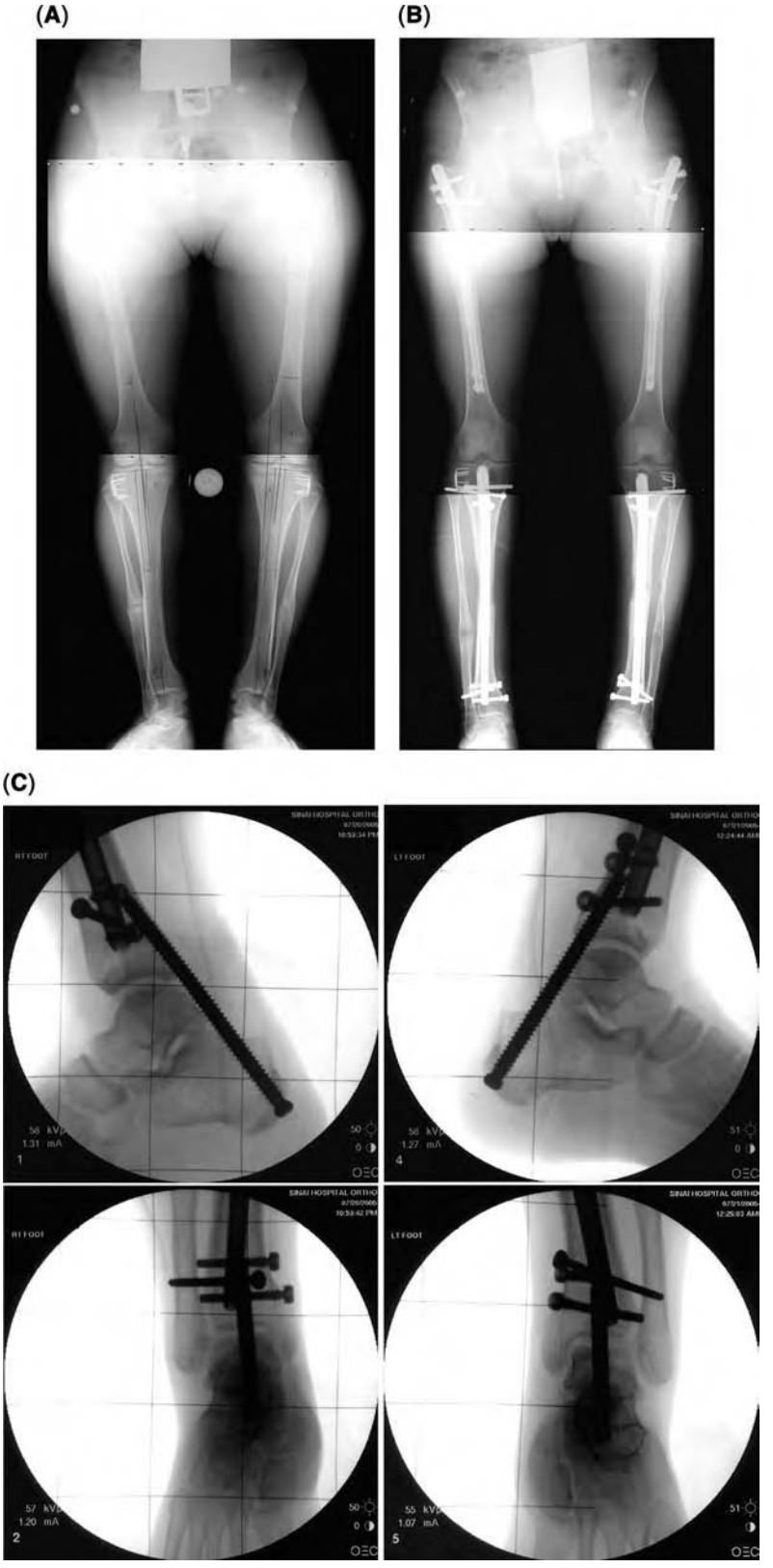

Figure 12 (*Caption on facing page*)

symptoms appear after an average of only 4 to 5 cm of lengthening. The predisposing factor is probably a small peroneal nerve tunnel. Previous fibulectomy is a predisposing factor. Peroneal nerve problems usually present as referred pain to the foot during tibial lengthening. If left untreated, they would produce a dropfoot. For this reason, it is essential to see the patient every two weeks during the distraction phase. If available, quantitative sensory nerve testing [pressure specified sensory device (PSSD) test] is performed at the time of the follow-up. This allows detection of impending nerve problems even before they are noticed clinically. If they are detected early and the rate of lengthening is slowed, the nerve symptoms may abate and the lengthening can be continued at a slower rate. If despite slowing the lengthening the nerve problem persists, the nerve needs to be surgically decompressed. Decompression should be performed at the level of the neck of the fibula decompressing the nerve at its entry into the common peroneal muscle fascia and the deep peroneal nerve as it passes beneath the intermuscular septum between the lateral and anterior compartments. After the decompression, lengthening is able to continue at the same rate. About 75% of patients undergoing double-level tibial lengthening require this additional procedure. Because it is so common, we consider the nerve decompression as a planned second-stage procedure. Because peroneal nerve entrapment is so common with double-level tibial lengthening, the question has arisen whether prophylactic decompression should be considered. Our preference to date has been to do this as a planned second-stage procedure rather than a prophylactic one. The reason for this is theoretical. We are concerned that if the nerve is decompressed before the onset of symptoms and if referred pain develop we have no remaining options except to stop the lengthening. The advantage of early therapeutic nerve decompression is that it permits the goal of lengthening to be achieved.

Spinal Cord Injury

Recently, we have become aware of an unusual but serious complication that can occur at the onset, during, or after the lengthening—paraparesis due to spinal cord compromise. We have had three cases of this complication of which two have completely resolved without residual problem and one still has weakness. All three patients had one thing in common—thoracolumbar kyphosis with unrecognized spinal stenosis. Only one had had any previous symptoms of spinal stenosis. In one patient, the problem occurred after the femoral lengthening was completed, in another it occurred during the lengthening, and in the third it occurred on the day of the initial surgery before any lengthening had been performed. We do not think this problem is related to the actual stretch of lengthening but rather to positional injury to the cord that occurred while sitting and sleeping in a wheelchair in a flexed posture for lengthy periods of time or while being transferred to or from the operating room table while under anesthesia. The latter occurred in one case at the time of removal of the fixator and the former at the time of application of the fixators. Both were treated by using a reclining wheelchair for sitting and prone position for sleeping. We now routinely obtain standing long spine radiographs in patients with clinical evidence of thoracolumbar kyphosis as well obtaining magnetic resonance imaging evaluation to assess for spinal stenosis. Patients at high risk in our opinion are sent for a preoperative consultation regarding their spine. Spinal decompression is a surgical option that is considered in a few cases to prevent or treat the spinal stenosis.

Premature Consolidation

Children with achondroplasia tend to heal regenerate bone faster than normal. Premature consolidation will arrest the lengthening process leading to bending of the pins without increase in length. Premature consolidation that is diagonsed radiographically is treated by lengthening at a faster rate (1.5 mm/day) for a few days. Sometimes this increased tension against resistance will lead to a sudden break through the regenerate bone or through a pin site. This is very painful and causes a sudden separation of the bone ends. If this occurs, it is imperative to diagnose it quickly and to undo any acute diastasis. If not addressed, delayed ossification may result.

Figure 12 (*Figure on facing page*) (**A**) Sixteen-year-old achondroplastic female before lengthening. No malalignment. (**B**) After bilateral lengthening with Intramedullary Skeletal Kinetic Distractor (ISKD) (Orthofix, Mckinney, TX, USA). Femora were lengthened first (5 cm) followed by tibiae (5 cm). (**C**) Temporary extra-articular screw ankle arthrodesis to prevent equinus during lengthening of tibiae with ISKD.

Fracture through a pin site is also suspected if the patient reports sudden bleeding at the pin site. If premature consolidation is diagnosed, the bone should be reosteotomized at a new level. It is preferable not to osteotomize through the regenerate because consolidation may become delayed.

Joint Contracture

During combined femoral and tibial lengthening, the emphasis is on knee and ankle ROM, respectively. Daily physical therapy is required. During bilateral double-level tibial lengthening, the feet are held in a 90° position by an extension of the external fixator to prevent contracture. Emphasis at therapy is on walking and knee-extension exercises. During bilateral humeral lengthening, the emphasis is on elbow ROM. During bilateral femoral lengthening, the emphasis is on knee ROM. In addition to formal physical therapy by a trained therapist, the patient is encouraged to exercise at home several times per day. Highly motivated patients recover faster.

Soft-tissue releases are occasionally required during the treatment. These include gastrocnemius-soleus recession when the foot is not included in the fixation. Achilles tendon lengthening should be avoided because it leads to permanent weakness of push off, while recession does not seem to weaken push off. The fascia lata may need to lengthen distally if the knee is developing valgus and flexion. The tensor fascia lata is released proximally to help correct the hip flexion deformity. More extensive release may be required if hip abduction contracture is developing. Recovery of hip, knee, and ankle ROM is predictable. The ankle ROM is the slowest to recover if the gastrocnemius-soleus unit is not lengthened and the foot is not immobilized for the duration of the lengthening to prevent equinus.

Fracture of Regenerate Bone

Fracture of the bone shortly after frame removal is a problem in lengthening for achondroplasia. Removing bilateral frames exposes both sides to high stress on transfers and weight bearing. In order to reduce the stress, we sometimes remove one side first while simultaneously dynamizing the opposite side (remove some pins), allowing it to strengthen, and then three weeks later remove the frame from the other side. Fractures can occur through the regenerate bone, through pin sites, and through the host-regenerate bone junction. Fractures may also be ''silent,'' i.e., bending but not catastrophic failure. Casting does not reliably prevent postremoval fractures. It is imperative to obtain postremoval radiographs on the day of removal, at about one-week postremoval, and one-month postremoval. Fractures after removal are generally best treated with intramedullary stabilization. In young children, a Rush rod typically is used, and in older adolescents and adults, a locked intramedullary nail is used (Figs. 13,4E). Nailing is challenging because the intramedullary canal is blocked by cortical tracks around old pin sites and the regenerate bone is not yet recannalized. We have found it helpful to use Ilizarov wires and cannulated drills to broach the canal, along with special T-handled reamers of small sizes (3–5 mm) that can be bent to help negotiate curves in the bone. Nailing allows prompt resumption of physiotherapy. The potential risk is infection, due to old pin sites. If the pin sites are not healthy enough to permit nailing, then reapplication of an external fixator is another, though less satisfactory, option.

Postoperative Management

Clinical follow-up is recommended every 10 to 14 days during the distraction phase. During these visits, radiographs are obtained and sensory nerve testing may be performed in addition to the clinical evaluation by the physician. During the consolidation phase, radiographs are obtained monthly. During one of these visits near the end of the consolidation phase, the external fixator is dynamized. Dynamization is the process of increasing the weight bearing taken by the bone and lessening the load taken by the external fixator. For double-level tibial lengthening, the middle pins are disconnected as part of the dynamization process. The rods or bar of the external fixator are unloaded to allow dissipation of the lengthening forces so that the bone takes more of the weight-bearing load. This helps the bone strengthen and thicken in preparation for removal of the external fixator.

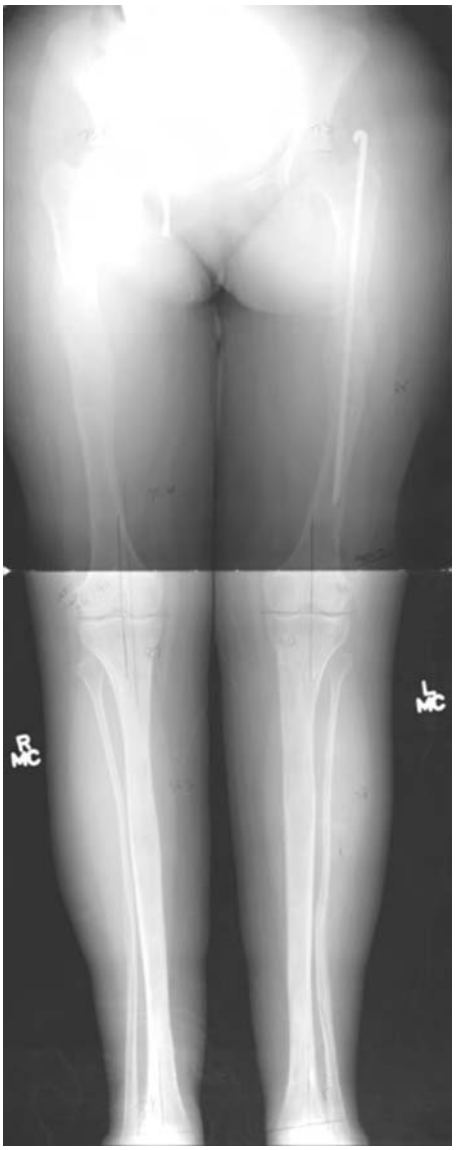

Figure 13 This female dwarf underwent bilateral simultaneous femoral lengthenings and sustained a fracture of the left femur following removal. This was realigned and held with a Rush Rod. Her tibiae had previously been lengthened and corrected with bilateral double-level lengthenings.

At the end of the distraction phase and before all adjustments are stopped, it is very important to ensure that the bone is straight. Measurements of joint orientation angles are performed on the radiographs to detect axial deviation. If present, the external fixator is adjusted to correct the angles to normal. If this step is skipped, the bone will heal malaligned. We routinely examine for this and correct the joint orientation angles to normal. This is especially important during the double-level tibial lengthening. The other important adjustment that is also made at this stage for tibial lengthening is derotation. An achondroplastic tibia is often internally rotated. Derotation can be facilitated by the TSF. Axial deviation in the femur should be corrected, although adjusting monolateral fixators can be more challenging than circular devices. The Yasui technique can be used to correct frontal plane malalignment provided the pins have been left long enough. Sagittal plane malalignment is somewhat easier to correct with the monolateral multi-axial correcting (MAC) (EBI, Parsippany, NJ, USA) device which can be grafted on top of the Orthofix device if necessary. For these reasons, we generally prefer circular devices for tibial lengthening because axial deviation and correction of deformities are critical (Ilizarov or TSF). In the femur, monolateral fixators are more convenient for the patient.

Bilateral femoral lengthening may be performed by two different methods. If correction of hyperlordosis is important, then formal lengthening is performed with bilateral monolateral

(straight bar) external fixators and extension osteotomy of the proximal femur. If hyperlordosis correction is not required, then LON or intramedullary skeletal kinetic distractor (ISKD) is preferred in the adolescent lengthening, provided the femoral canal can accommodate these devices. LON decreases the time in the fixator by over 50%. For achondroplasia, hyperlordosis and spinal stenosis are important issues. We, therefore, usually prefer to use the fixator-only method so that hyperlordosis is corrected and spinal stenosis is reduced. After six to eight weeks of lengthening, we perform soft-tissue releases of the flexor muscles at the hip to reduce flexion deformity. The external fixator is on for 10 to 12 months followed by a month of bracing following removal. There is usually a stature gain of 2 cm from reduction of lumbar lordosis. This is in addition to the length obtained from lengthening. For hypochondroplasia where hyperlordosis is usually not a problem, LON or, size permitting, fully implantable lengthening with the ISKD are options.

FUTURE DIRECTIONS AND CONTROVERSIES

Lengthening for stature for dwarfism has been termed extensive limb lengthening (ELL). ELL treatment is controversial in the United States and Canada, but it is a well-accepted part of the management of dwarfism in Europe, Asia, and South America. The interests of patients with dwarfism are represented by the Little People of America (LPA) and Little People of Canada organizations. They have rightfully been wary of such treatments that might lead to serious complications and injury to individuals with dwarfism. This concern dates back to the first experience with lengthening for stature in dwarfism using the Wagner method during the 1970s. Results were complication-ridden with the treatment often being worse than the condition. After several surgical treatments, patients achieved very limited increase in height. With the introduction of the Ilizarov and Orthofix devices to North America in mid-1980s, all this changed. ELL has been more reproducible in some hands. In Baltimore, we use the methods described above, stressing the goal of lengthening to increase the height to the short normal range—5 feet tall in girls and up to 5 feet 4 inches in boys. We also stress correction of deformities while lengthening to improve mechanical axis alignment and joint orientation, reduce the hyperlordosis of the spine, and in some cases, correct the flexion deformity of the elbows. Complications still occur, but they are almost all treatable without leaving permanent sequelae. Long-term results of the European experience and now up to 16-year follow-up on the North American experience have not shown deterioration in results, such as degeneration of joints, muscles, or nerves. In light of these experiences, we feel confident recommending ELL for patients with achondroplasia and hypochondroplasia. We feel that we can safely achieve the goals of treatment in nearly all cases with little to no permanent residual side effects, provided the treatment protocols are strictly followed.

We agree with the LPA that there is no ''need'' to be lengthened in order to live a productive, healthy life. Similarly, patients with limb-length discrepancy (LLD) do not ''need'' to undergo limb lengthening. In both situations, the affected individuals can live quite well with special devices to compensate for their disability, e.g., pedal extension so that patients with achondroplasia can use a car and a shoe lift for LLD. Therefore, what are the benefits of ELL that justify the risks and inconvenience of this difficult and lengthy treatment including its multiple planned and unplanned surgeries and extensive rehabilitation?

The benefits from ELL can be divided into functional, psychosocial, and therapeutic. The functional benefits derive from the increased stature and from longer reach. Society has designed the world based on a minimum height of five feet tall. Every day items in our homes, such as door knobs, coat racks, light switches, seat height and depth, toilet seat height, freezer doors, and shower controls, are designed based on a minimum height of 5 feet. The depth of the range and water faucet is based on a certain amount of arm reach. Office environments have copy machines, file drawers, and other surfaces that are out of reach of individuals with dwarfing conditions. Cars have their gas and break pedals out of reach of the short lower limbs. Air bags must be disabled to avoid hitting the short-armed achondroplastic person too strongly in the head because they are holding their head and torso so close to the steering column. Most public telephones are out of reach. By adding a foot of length and increasing height to over 5 feet tall, as well as increasing the reach by 4 inches, all of these items are made easily accessible. Compensatory devices such as stools and stepladders are no longer needed.

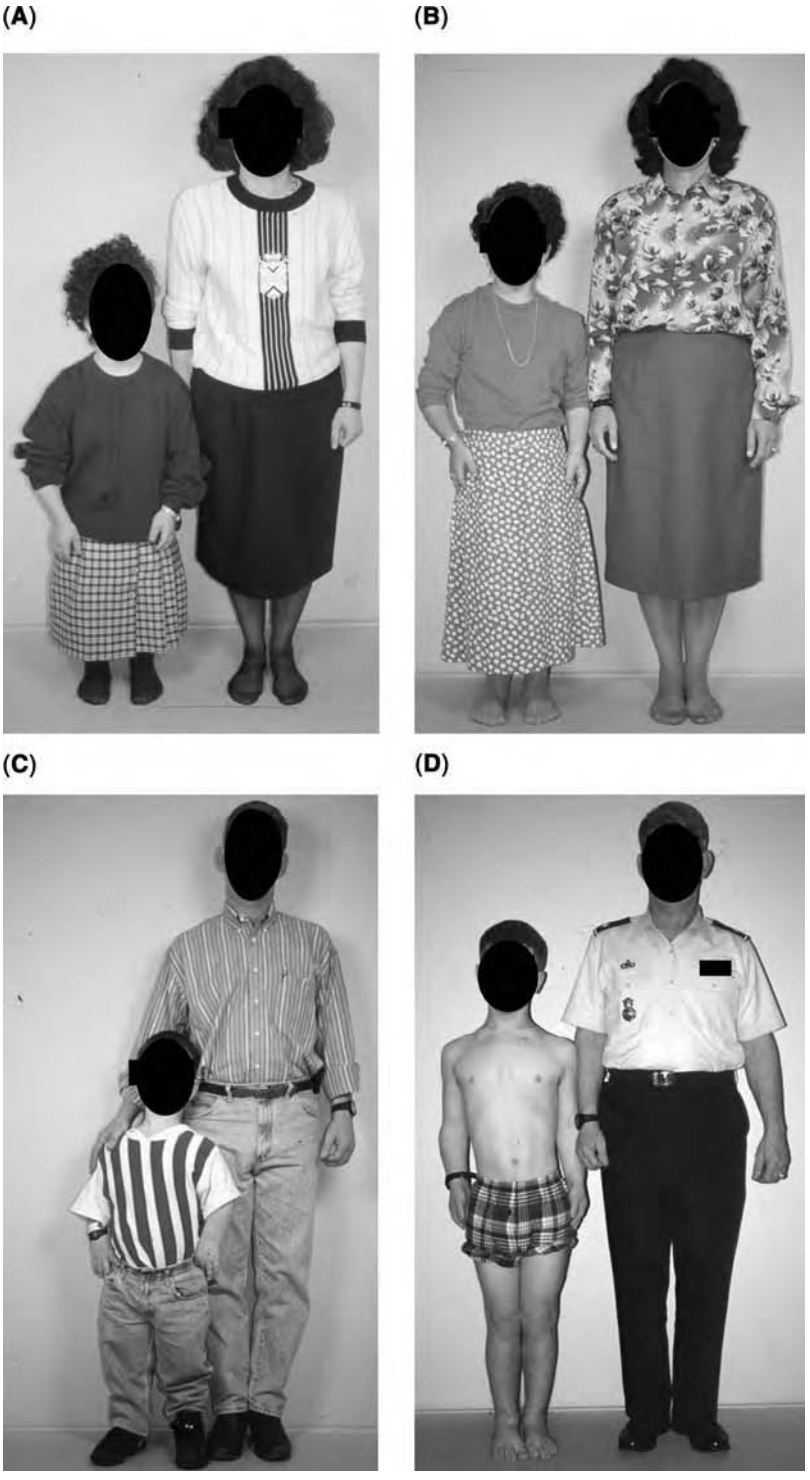

Figure 14 (**A**,**B**) Teenager standing next to her mother before and after bilateral tibial lengthening. The increase in height was judged to be satisfactory, and the patient went on to marry. (**C**,**D**) Achondroplastic dwarf standing next to his father before and after his first two lengthenings. He subsequently returned for a third lengthening to gain additional height.

Sitting can be easier after leg lengthening, because the feet rest comfortably on the floor rather than dangling.

One of the greatest benefits for achondroplastic patients is humeral lengthening, which has tremendous implications for personal hygiene. As the achondroplastic spine ages, it loses flexibility and it becomes more difficult or even impossible to reach the perineum and carry out personal hygiene.

Psychosocial benefits are also extensive. ELL leads to a complete change in body image. The trunk limb and rhizomelic disproportions are no less pronounced (Fig. 14A–D) . Increased height may lead to greater self-confidence and may increase dating options. Increased height may improve job marketability, making more types of employment available.

Finally, ELL has some prophylactic and therapeutic benefits. Reducing the hyperlordosis of the spine should theoretically lead to increased space available to the cord and cauda equina, theoretically reducing the spinal stenosis that is present in all of them. Realigning the knee should theoretically reduce the likelihood of arthritis, similarly for the ankle and hip. Tightening the lateral collateral ligament makes the knee more stable and eliminates the valgus thrust. Valgusizing the upper femur increases the abduction lever and tension arm leading to less lurch during gait. With the exception of one patient with spinal cord injury, all of our patients said they would undergo this surgery again and that the ordeal was worthwhile because the final result is such a dramatic improvement. Only one patient was made functionally worse with treatment, and now that we better understand the spinal cord problem, paraparesis should be completely avoidable and at least fully recoverable.

The decision to undergo ELL is a personal one and it is not for every family or child. However, we do feel that patients with achondroplasia or hypochondroplasia should be educated about this treatment modality. More studies are needed to document the long-term results and patient outcomes. More centers need to learn these techniques so that patients will not have to travel such long distances to the few centers that offer this treatment. As the technology of lengthening advances with more totally implantable intramedullary lengthening devices, we can expect to see an increased interest in lengthening for dwarfism.

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44 Stature Lengthening
Orthopedic Surgical Focus

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INTRODUCTION

Limb lengthening with the Ilizarov method has been used successfully in both the femur and the tibia (1). The most common indications for which this method is used are leg length discrepancies and deformities from either congenital or post-traumatic etiologies (2–5).

Bilateral lower extremity lengthening has been used to increase stature and correct deformity in patients with dysplasia (6–8). This is the subject of another chapter.

With the ability to safely and effectively perform these procedures comes an interest by many individuals to increase their stature (2,8–15). The procedure is too complicated, controversial (16–19), risky (20–22), and expensive to be used for any individual who just wants to be taller. However, there appears to be a subset of the population with short stature, who are very unhappy about their height. Patients with the psychological state of ''short stature dysphoria'' seem to feel an intense need to increase their height. With appropriate psychological screening, a select group of patients can be identified for treatment with stature lengthening.

Our experience treating such a group of patients has been very positive. Safe and successful stature lengthening of 2″ to 3″ can be accomplished. The result is an outcome of increased happiness and improved self-image. This chapter is divided into two parts—the first will deal mainly with orthopedic issues of limb lengthening and the second will address the significant psychological component.

CLINICAL EVALUATION

Lengthening for stature in normally proportioned individuals with short stature but without dysplasia (dwarfism) requires a different strategy for lengthening than in patients with disproportion and dysplasia. Most patients with constitutional short stature (CSS) or low normal stature (LNS) require only modest increase in stature compared to the extreme amount of lengthening required by patients with dwarfism. In fact, the starting stature of patients with LNS is the final goal of lengthening in patients with dwarfism.

When assessing distribution of height in the population, we consider the normal bell curve. We divide people by distribution around the mean (average), and normal height is considered ± 3 standard deviations from the mean. Short stature below 3 standard deviations from the mean in individuals without a medical condition such as dwarfism, growth hormone deficiency etc., is considered CSS. The lower limit of so-called normal stature for Caucasian men is 5′5″ and for women is 5′0″.

While patients with dwarfism are often treated as children (1,6–8,16), patients with CSS or LNS are not treated until they have completed growing. Patients with LNS and CSS also do not usually have deformities of the bones.

A thorough history is obtained including birth, pregnancy, childhood growth patterns, onset of puberty, and endocrine history. Early onset of puberty, low-growth hormone levels, childhood illness, and a variety of genetic conditions can contribute to CSS.

Family history is obtained. This includes the height of mother, father, and siblings. This helps put the patient's height into the context of his family. This perspective helps with an understanding of what is normal for this particular family.

Table 1 Clinical Management

Treatment Approach	Patient Factors	Complications/Pitfalls
Referral to endocrinologist for possible growth hormone treatment	Patient still growing	Medication side effects
No surgery	Not psychologically cleared; poor medical health; joint contractures or instability	Counseling
Bilateral tibia and fibula lengthening	Tibia: femur $<80\%$; abnormal knee	Equines contracture of ankle, may need gastrocnemius recession
Bilateral femur lengthening	Tibia: femur $>80\%$; abnormal ankle	Contracture of knee, may need quadricepsplasty
Traditional technique	Younger patients who are expected to have short consolidation time; patient does not want second surgery	Pin tract problems; extended time in frames
Technique		
Lengthening and then nailing	Use frames to control distraction and then remove frames after distraction phase	Risk of deep infection; second surgery for rod insertion
Internal lengthening nailing	Sufficiently sized intramedullary canals	Poor rate control; nonunion, premature consolidation, nail malfunction, iatrogenic LLD

Abbreviation: LLD, leg length discrepancy.

This often uncovers psychological pain, particularly if this patient is different from the family norm. Psychological testing is done in a very formal and thorough fashion by a psychologist, as is explained in a later section of this chapter. First encounter questions should try to uncover the degree of unhappiness caused by short stature, the frequency of this unhappiness, and overall psychological health. We have found that patients who are the best candidates for lengthening are those who have significant and frequent feelings of discontent about their short stature, yet in other areas of their lives are successful, happy, motivated, well adjusted, and most importantly, have realistic expectations of what surgical lengthening can do for them. This is not simply a cosmetic operation. This surgery is done for patients who feel the need to do this, yet are mentally healthy.

On physical examination, height is measured. Range of motion of hips, knees, and ankles are recorded. Neurologic and vascular examination is performed. This will uncover any contractures or deformities that may exist. Knee pathology or contracture may make femur lengthening a less attractive option because it may lead to joint stiffness. Ankle contracture may make tibia and fibula lengthening a less attractive option for the same reason. Deformities or LLD can potentially be corrected during stature lengthening (5).

Radiographs are done. These include a 51″ bipedal erect leg radiograph and lateral X-rays of one femur and one tibia. These are measured to uncover any deformity or LLD. Lower extremity lengths and both femora and tibiae are measured. Tibia to femur ratio is measured (normal is 0.8). Mechanical axis deviation and joint orientation angles are recorded (5). Intramedullary canal sizes of femur and tibia are measured on both anteroposterior and lateral X-rays.

CLASSIFICATION (TABLE 1)

Patients who are still growing are not usually considered for this surgery. Referral to an endocrinologist is made for possible growth hormone treatment.

If a patient has a deformity such as bowlegged alignment, one should consider correcting the deformity at the same time as stature lengthening. This requires osteotomy at the apex of the deformity (5).

Intramedullary canal size is an important factor. The use of fully implantable limb lengthening nails requires an adequately sized intramedullary canal. Similarly, combination techniques such as lengthening over a nail (LON) (23) and lengthening and then nailing (LATN) [Rozbruch SR, Kleinman D, Fragoknen A, Ilizavov S: Lengthening and the Nailing (LATN). Limb Lengthening and Reconstruction Society (ASAMI-NA) July 21–23, 2006] mandate a minimum sized intramedullary canal to accommodate the intramedullary (IM) nail.

Contractures, stiffness, or instability of the adjacent joints is an important factor. Abnormalities of the knee should discourage femur lengthenings. Abnormalities of the ankle should discourage tibia and fibula lengthenings.

The proportionality of leg to thigh may be a consideration. The normal tibia to femur length ratio is 80%. A lower ratio may encourage the surgeon to perform tibia lengthenings and a higher ratio may encourage femur lengthenings.

TREATMENT OPTIONS (TABLE 1)

Surgical site options include lengthening of bilateral femora or bilateral tibiae. Technique options include osteotomy and gradual lengthening with circular (15,24) or monolateral external fixation (1) (traditional technique), combination techniques of LATN [Rozbruch SR, Kleinman D, Fragoknen A, Ilizavov S: Lengthening and the Nailing (LATN). Limb Lengthening and Reconstruction Society (ASAMI-NA) July 21–23, 2006] (Figs. 1 and 2) or LON (23), and fully implantable lengthening nails.

The traditional technique uses the external fixator for gradual distraction as well as consolidation. This is well established and has been used for the longest time. The main disadvantage is excessive time in frames and problems with pin tract infection and soft-tissue tethering by pins. The main advantage is that the osteotomy can be done in the proximal tibia. This is the usual location of the apex of a congenital genu varum deformity. We use this technique for patients with open growth plates or when we aim to correct bilateral genu varum or valgum and add a modest length. One inch of length typically requires six months of time in frames (Fig. 3).

To minimize the treatment time for lengthening, we prefer the LATN technique (Fig. 1). This method minimizes the treatment time in the external fixator, limited to the time required to obtain the lengthening desired without having to wear the external device during the hardening or consolidation phase. This method also reduces the rehabilitation time and time to return to full weight bearing, work, school, and recreational activities. The LATN technique uses external fixation for controlled gradual lengthening during the distraction phase and IM nails to support the new regenerate during the consolidation phase. Advantages of this method include the following (Fig. 2):

1. Internal and external fixation are used in separate phases decreasing chance of infection,
2. The IM nail used for fixation during the consolidation phase can be as long and wide as possible to fill the tibia bone, and
3. Reaming through the regenerate seems to enhance bone healing.

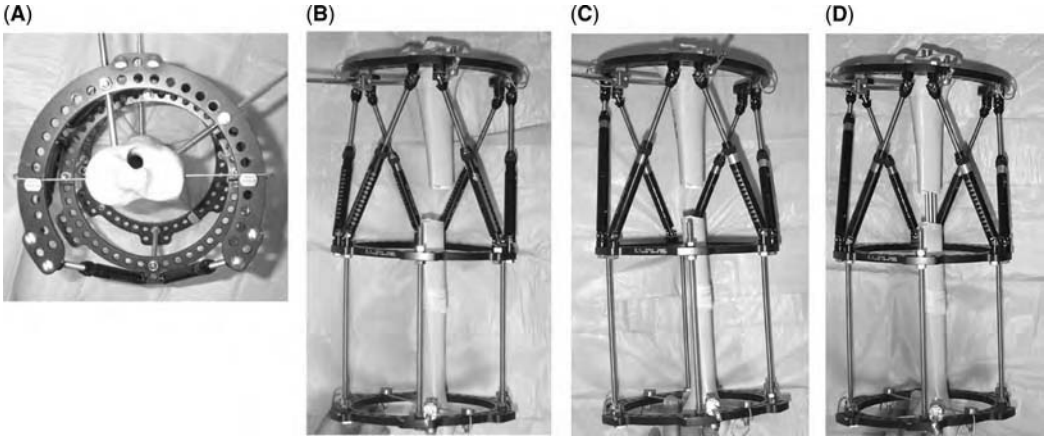

Figure 1 Sawbone demonstration of lengthening and then nailing of tibia (LATN). (**A**) Axial view at the proximal tibia showing configuration of pins and wires to miss the future intramedullary nail. (**B**) After distraction showing deformity. (**C**) Correction of residual deformity with the Taylor spatial frames. (**D**) After insertion of IM nail. *Abbreviation*: IM, intramedullary

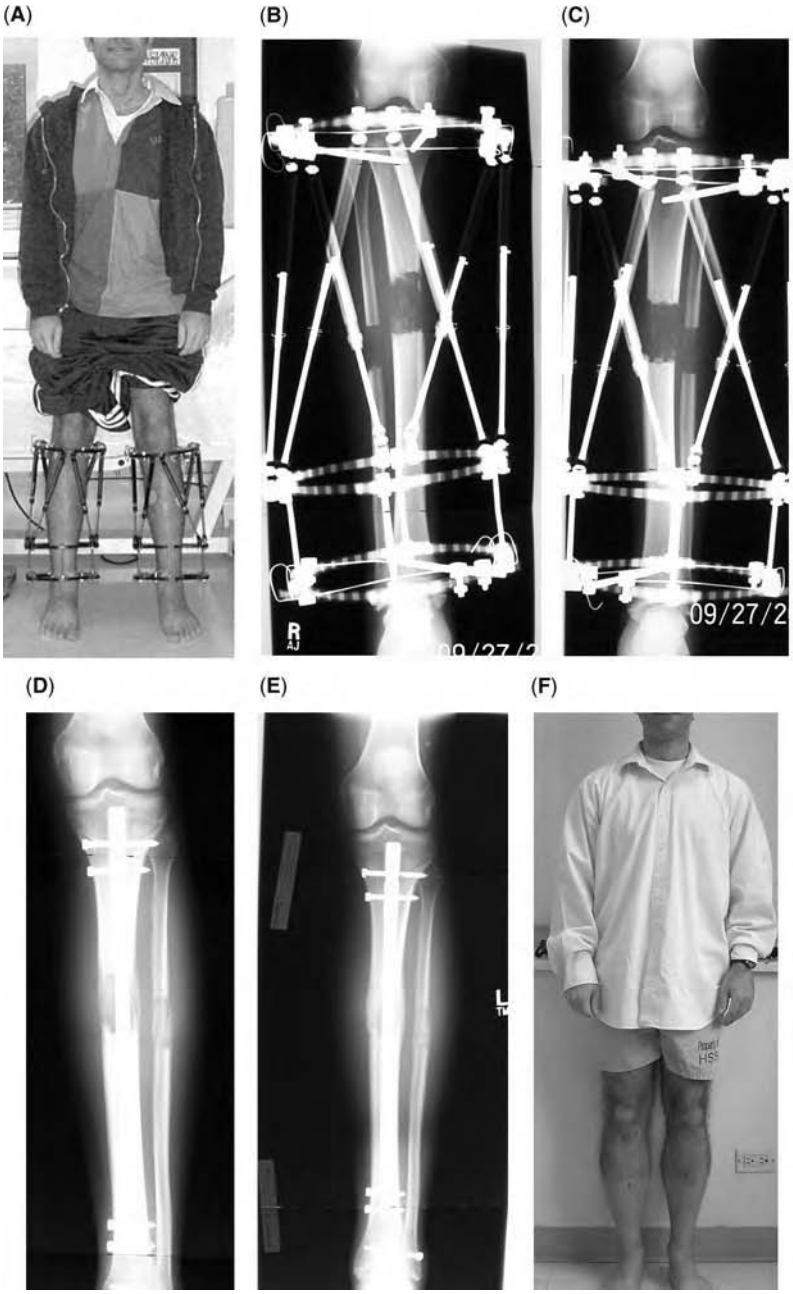

Figure 2 Case example of a 5 cm stature lengthening using lengthening and then nailing of the tibia. (**A**) Front standing view two months after surgery. (**B** and **C**) Anteroposterior (AP) radiographs of both legs showing 5 cm of distraction. (**D**) AP radiograph of left tibia two weeks after intramedullary (IM) nailing. Note oblique syndesmosis screw. (**E**) AP radiograph of left tibia two months after IM nailing. (**F**–**H**) Clinical photos four months after IM nailing and six months after initial surgery. (**I**–**L**) AP and lateral radiographs of both legs four months after IM nailing and six months after initial surgery. Note the absence of the syndesmosis screws.

A fully implanted lengthening nail is also available (Fig. 4). This approach does not require an external fixator at all. The only Food and Drug Administration approved devise in the United States is called the intramedulla skeletal kinetic distractor (ISKD) and is made by Orthofix Inc. The lack of precise rate control is a major disadvantage of this devise. Distraction that occurs at a rate of 1 mm/day leads to excellent bone formation. Slower distraction can

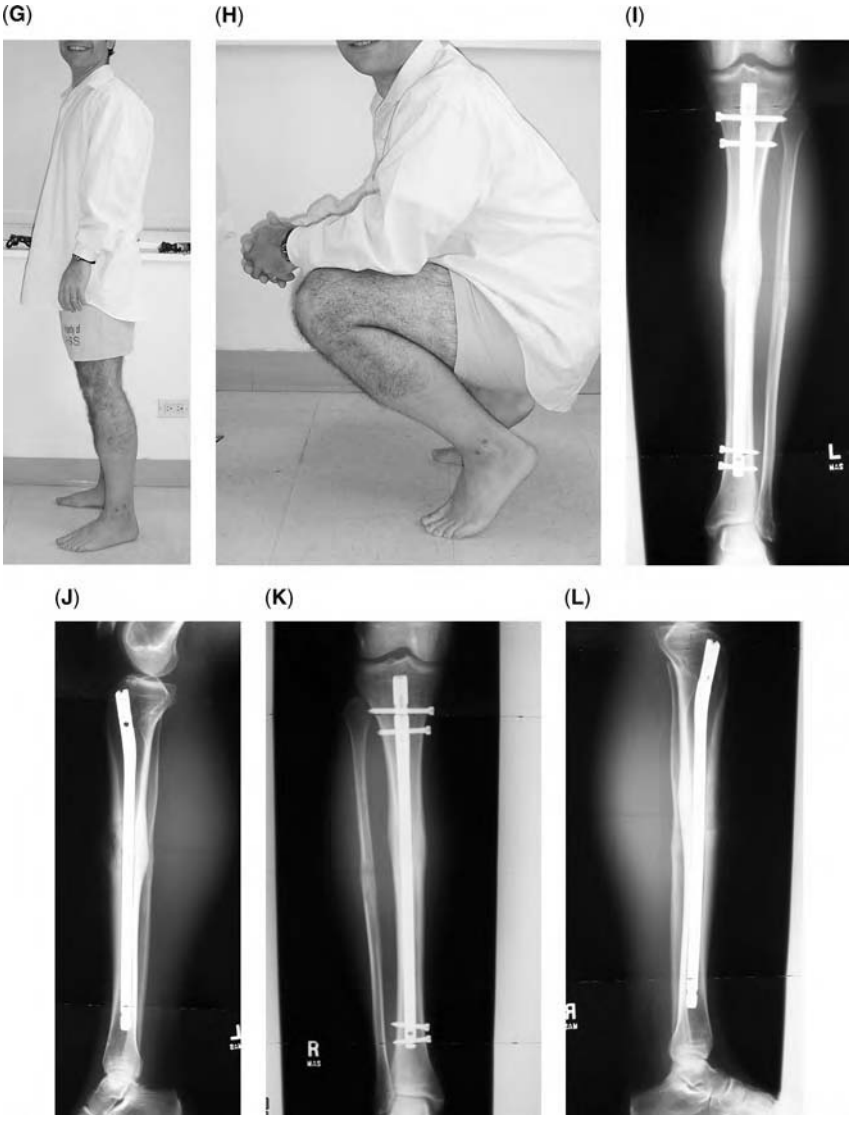

Figure 2 (*Continued*)

lead to premature consolidation; faster distraction can lead to poor bone formation, nonunion, joint contracture, and nerve dysfunction.

The usual goal of lengthening for stature for most patients with LNS is 2″ to 3″. More than that cannot be achieved safely in one pair of bones during one lengthening. For more lengthening, one needs to repeat the process in a second pair of bones.

Our usual strategy is to lengthen both tibias including the fibula by 2″ to 3″ using the LATN method or the internal lengthening nail. If a second lengthening is desired, then both femurs are lengthened by the same measurement. Most patients do not choose to have the femurs done because of the added time commitment and doubling of the expense. Furthermore, doing too much lengthening in the lower limbs in normally proportioned people leads to an obvious disproportion. Up to 3″ of lengthening produces no obvious disproportion.

Our preference for tibial lengthening over femoral lengthening is because of several good reasons. First of all, tibia lengthening makes one's legs look longer than femoral lengthening. The reason for this is that the level of the knees and feet are obvious but the level of the hips is not obvious. Depending on how we wear our belts, our hips seem to be higher or lower. When we see long lower legs, we extrapolate that the rest of the person has a normal

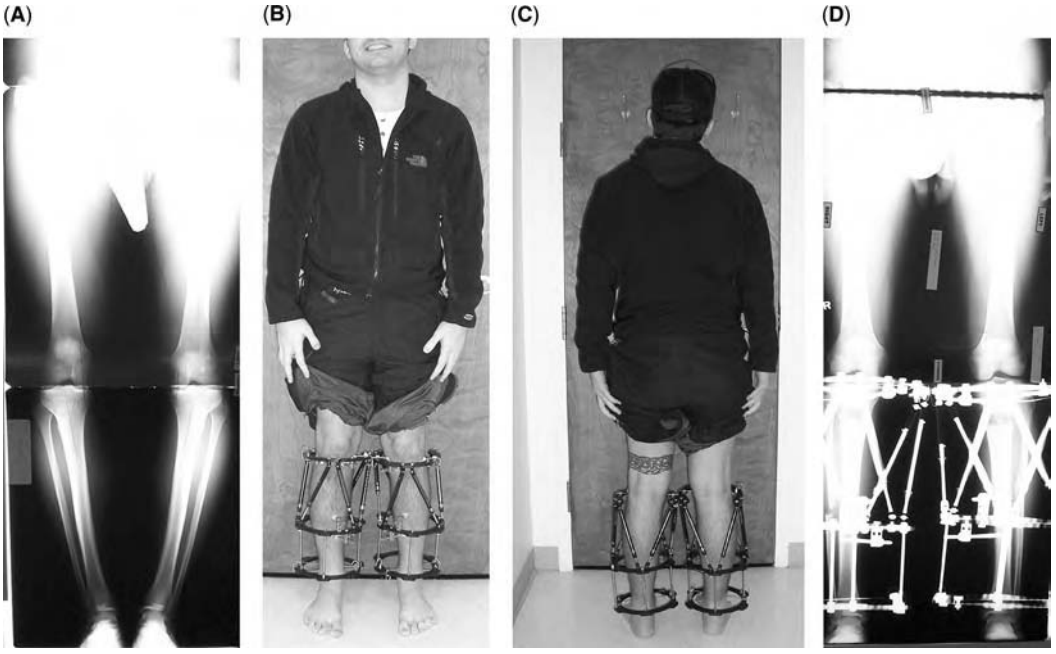

Figure 3 Case example of 1″ lengthening and correction of bilateral genu varum using the Ilizarov/Taylor spatial frames. (**A**) Preoperative 51″ radiograph showing bilateral genu varum. (**B**–**D**) At the end of distraction, showing correction of deformity and 1″ lengthening. (**E**) Anteroposterior radiograph four months after surgery of the right side showing progression of healing. (**F**) Clinical view at five months after distal rings have been removed for dynamization and to improve mobility. (**G**) Clinical view at six months showing a staged removal of three weeks between sides. (**H** and **I**) Front and side views one month after removal (seven months after initial surgery). (**J**) Standing 51″ radiograph four months after removal showing correction of deformity and 1″ lengthening.

proportion, which gives the impression that the individual is taller than they really are. Furthermore, wearing short skirts and dresses shows the length of the lower legs but not the thighs; therefore, one gets more mileage in apparent increased height from tibial than from femoral lengthening. The second reason we prefer tibia lengthening is that technically it can be performed in one operation in a reasonable period of time. Two femoral lengthenings require two separate setups during the same operation or two different operations. This greatly increases the operating time and cost. Furthermore, there is much less blood loss with tibial lengthening than with femoral lengthening. The third reason we prefer bilateral tibial rather than femoral lengthening is that it is much easier and better tolerated to have two external fixators on both lower legs than on both thighs.

SURGICAL TECHNIQUE

The LATN technique uses external fixation for controlled gradual lengthening during the distraction phase and IM nails to support the new regenerate during the consolidation phase (Figs. 1 and 2).

The first surgery is bilateral tibia and fibula osteotomies and application of Ilizarov/Taylor Spatial Frames (TSFs). Epidural anesthesia is typically used. The fluoroscopy unit is placed on the opposite side of the table. After procedure on the first leg is complete, the fluoroscopy unit is switched to the other side of the table. A tourniquet is used only for the fibula osteotomy. The fibula osteotomy performed is transverse and in the middle of the fibula. The fibula is approached through a 1″ lateral skin incision. The interval between the lateral and posterior compartment is used. A subperiosteal exposure of the fibula is maintained with mini-Homann retractors. A 1.8 mm Ilizarov wire is used to make drill holes in the fibula while cooling the

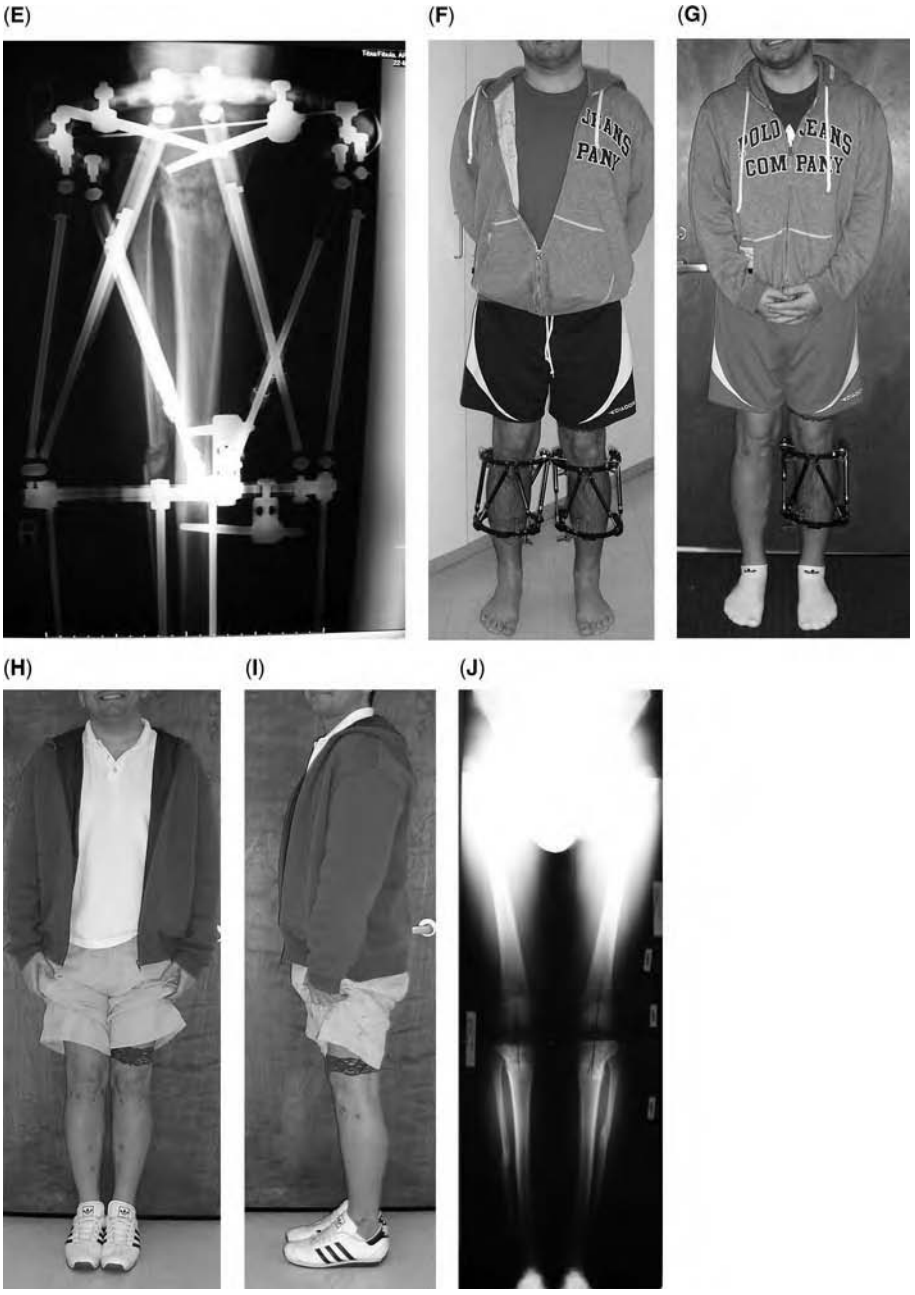

Figure 3 (*Continued*)

area with saline. The osteotomy is completed with an osteotome. The fascia is not closed. The tourniquet is released.

Next, the frame is mounted. A three-ring Ilizarov/TSF is used. The proximal is a 2/3 ring to allow knee flexion. The middle ring is empty. The distal is a full ring. Fixation from the proximal ring includes two tensioned wires and two half-pins. This fixation is placed so that the path of a future IM nail is not blocked (Fig. 1). The transverse wire is posterior. The tibiafibular wire is posterior. The anteromedial and anterolateral half-pins are peripheral. The proximal ring is placed close to the leg on the medial side to enable use of the targeting jig for interlocking screw insertion during future surgery.

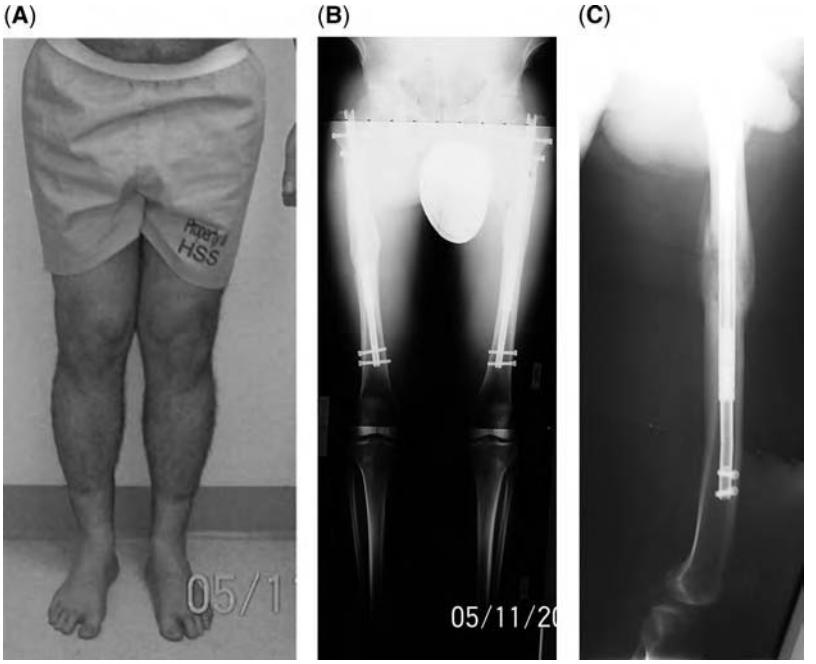

Figure 4 Case example of a 5 cm stature lengthening using ISKDs in bilateral femurs. (**A**) Clinical photo at the end of distraction. (**B**) Erect leg radiograph at the end of distraction. (**C**) Lateral radiograph of the right femur showing the progression of healing. *Abbreviation:* ISKD, intra medulla skeletal kinetic distractor.

The middle ring has no fixation. The distal ring is fixed to the bones with two wires and a half-pin. This fixation is just above the ankle and does not interfere with a future IM nail. One wire is placed transversely parallel to the ankle, one wire is used to fix the syndesmosis, and the half-pin is placed from the anteromedial direction just medial to the Tibialis anterior tendon.

A ''rings first method'' of TSF application (3,25,26) is used to apply the proximal ring, which will be the ''reference ring'' and the ring block consisting of the middle and distal rings. Strut connections are placed between the proximal and middle rings. The tibial osteotomy is performed about 10 cm from the proximal tibia joint line. The middle of the bone at the osteotomy site is considered the ''origin.'' This is a large enough proximal fragment to be stabilized with an IM nail. The tibial osteotomy is done with a multiple drill-hole osteotomy technique (5) after the struts are removed. The struts are then reapplied.

Distraction usually begins on day number 10 after surgery. Lengthening of 1 mm per day is started. Follow-up visits are every 10 to 14 days. The distraction rate is adjusted based on symptoms of pain, regenerate appearance on X ray, and joint mobility (especially ankle). On these visits, we perform nerve testing if needed, examine pin sites, and obtain X rays. Problems are identified and treated and medication prescriptions for pain or antibiotics are written. Occasionally adjustments to the external fixators are required in the office.

Once the complete length is achieved, deformity at the osteotomy site (especially translation) is corrected with the TSF ''residual deformity program.'' The goal is to be able to insert an IM nail across the distraction gap.

The second surgery entails insertion of IM nails, insertion of syndesmosis screws, and removal of frames. It is crucial that there are no infected pin sites. If necessary, an infected pin can be removed and surgery can be delayed. Pin sites are covered with Betadine soaked sponges. One leg is done at a time with the fluoroscopy unit across the table. A syndesmosis screw is placed in an oblique manner using a cannulated 4.5 mm screw system. Care is taken during this and all steps of the surgery to avoid contact between internal and external fixation. The entry site for the IM nail is planned with a percutaneously placed wire. A 1″ skin incision is made at the proximal tibia. The patella tendon is split. A 10 mm solid reamer is used over the guide wire to open the entry site into the proximal tibia. The guide wire is then inserted

across the distraction gap and into the distal tibia. This is uncomplicated if there is no deformity at the distraction site. The nail length is planned. Serial reaming is done (without tourniquet) and an IM nail 1 mm smaller than the biggest reamer used is inserted. The frames are still in place maintaining the length. Proximal and distal interlocking screws are placed. The frame is then removed. Surgical incisions are primarily closed; external fixation pin sites are left open to close in a secondary fashion.

If significant equines contracture has occurred, then this is addressed. Typically, the contracture is from tight gastrocnemius. This is confirmed by checking the contracture with the knee both in extension and in flexion. A gastrocnemius recession (5) is performed that enables ankle dorsiflexion to neutral. Soft dressings are applied.

Radiographs are checked six weeks after this surgery. Bone healing has usually progressed and patients are encouraged to begin walking as tolerated.

Most patients are in the hospital between two and four days after the procedure. Physical therapy to stretch the ankles begins in the hospital and continues after discharge as an outpatient. Patients need to have physical therapy for one to two hours everyday during the two to three months of lengthening. Diligent stretching of the ankle and knee joints both at home and at physical therapy is crucial. Therapy can be carried out at any therapy center. The patients from out of town can do their therapy in their hometown. Ankle range of motion is the limiting factor for lengthening.

One can assume approximately one month of distraction for every inch (2.5 cm of lengthening). Lengthening does not begin until approximately one week after surgery. Therefore, 2″ to 3″ of lengthening takes two to three months in the external fixator. Weight bearing is allowed within limits of pain. At the end of the lengthening, there is a second surgery to insert the IM nails, and the external fixators are removed. Because the rods only fix the tibias, an additional screw is needed for each fibula to protect the fibula from shifting from the ankle. This screw can be removed under local anesthesia one or two months later. After the external devices are removed, the follow-up is once a month for the first two to three months. Out-of-town patients can just send X-rays and physical therapy reports instead of coming in.

The internal lengthening nail method does not require the application or removal of external frames.

Patients remain non–weight bearing until the X rays show that the bone is healed enough to allow weight bearing. One or two intact cortices must be seen on X ray. This usually takes one to two months after nail insertion. Most patients can begin weight bearing two months after removal. Physical therapy continues mostly to regain full ankle motion and foot push-off strength. This can take several more months. Therapy is only three days a week after removal but daily home exercises are recommended. Removal of rods is recommended in the future. This is not critical and can be done at any time once the bones are fully healed. We generally perform this about one year after the lengthening is completed. Again, this is an outpatient or one-night stay surgery.

COMPLICATIONS

While there are many potential complications of lengthening, other than mild pin infections, they are uncommon. Ankle stiffness is a concern but a rare complication if intense therapy and home exercises are carried out and if the lengthening does not continue, if the foot position drops below 90° (equines contracture). Gastrocnemius recession is very helpful in preventing a permanent equines contracture. Deep infection with tibia lengthening over nail is rare. It is resolved by removal of rod. Nerve injury is also rare (21).

Delayed union and nonunion (25) are potential complications. We [Rozbruch SR, Kleinman D, Fragoknen A, Ilizavov S: Lengthening and the Nailing (LATN). Limb Lengthening and Reconstruction Society (ASAMI-NA) July 21–23, 2006] have not encountered this problem in 35 cases of LATN. In fact, bone healing seems to be faster than with routine lengthening. We think that reaming through the regenerate stimulates bony union with bone grafting and perhaps release of bone growth factors.

Finally, the most important issue to consider is that lengthening for stature in any individual over 5 ft. in height is primarily for psychological reasons and does not improve function. It does seem to improve body image in patients with what we call short person dysphoria. Nevertheless, the issue of function is of greatest concern to us. This procedure

can damage nerves, muscles, and joints. If such damage were to occur, it could become irreversible leading to long-term problems such as arthritis, limitation of joint motion, and pain. Rare cases can even develop reflex sympathetic dystrophy, which is a chronic pain condition and which may not be resolvable. Pulmonary embolism and deep vein thrombosis, which are common with other forms of orthopedic surgery, are rare with this surgery, but they can occur and could lead to sudden shortness of breath, chronic leg swelling, and even death. While loss of life and limb have never occurred to us with this procedure, one must still weigh the risks of undergoing a major surgical procedure versus the benefits of increasing ones stature by 2″ to 3″. Proceeding with the surgery is a very personal decision.

FUTURE DIRECTIONS

Improvements in the design of internal lengthening nails will expand the indications and practice of stature lengthening. The ability to control distraction rate with a simple method is needed.

The psychology of patients who are intensely unhappy with their stature needs further elucidation. Further study of this area should help with counseling and patient selection for surgery.

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Psychological Focus

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OVERVIEW

Each year, thousands of individuals undergo elective cosmetic surgery to alter their physical presentation. This is likely an underestimation of the actual number performed because cosmetic procedures are also performed by surgeons in other specialties (2–4). Ultimately, one elects to have cosmetic surgery to improve satisfaction with one's appearance and ultimately, self esteem. To a large degree, in many cases, cosmetic surgery can be considered in itself a psychological intervention with both physical and psychological consequences (4–6).

The physical body, literally and metaphorically, has long been considered fundamental to one's sense of self. Early psychoanalytic theorists as well as more recent writers note that the physical body is a representation and reflection of the self (4,6–8). As such, how one experiences his body is inextricably intertwined with his sense of self-esteem and self-worth. Developmental theories of body image disturbance focus on the contribution of childhood and adolescent experiences to adult body image (1,7). Most contemporary theorists believe that body image concerns are central to understanding the desire to undergo cosmetic surgery. Some investigators contend that many of these patients obtain much of their self-esteem from their appearance, and when their self-esteem declines, they seek surgical remediation of what is at least in part, a psychological difficulty (1,4,6,9).

While a great deal has been written about the psychological indications and sequelae of cosmetic surgery, there is little empirical data available with regard to patients who elect to undergo surgical lengthening for short stature. Available literature suggests that it is not in the same dimension as other cosmetic procedures because it is major surgery with a host of possible complications as well as a lengthy and arduous postoperative course (10). As such, surgeons who perform this procedure often recommend that patients undergo an intensive psychological evaluation to assess motivation as well as a variety of other factors outlined below.

CLINICAL ASSESSMENT

Patients who present for a psychological assessment as part of an overall evaluation for cosmetic limb-lengthening surgery often report chronic dissatisfaction with their height. Anecdotally, many of these patients are men, who cite concerns such as interpersonal anxiety with regard to romantic relationships, social anxiety, as well as negative perceptions of them at work due to their stature. Studies of cosmetic surgery patients have generally employed

clinical interviews as well as structured diagnostic tests to assess the psychological functioning of these patients [for a thorough review of assessment procedures of cosmetic surgery patients, Refs. (6,11)]. Assessment of patients prior to limb-lengthening surgery includes a structured clinical interview as well as a battery of standardized objective and projective measures. An evaluation that includes both objective, self-report measures, as well as projective, ambiguous tests is optimal because it captures both overt and unconscious levels of psychological functioning (4,12,13).

Objective measures such as the Symptom Checklist 90-Revised (SCL-90) (14) and the Minnesota Multiphasic Personality Inventory (MMPI) (15) assess primary psychiatric symptom dimensions including somatization, obsessive compulsivity, depression, psychoticism, and anxiety. The Beck Depression Inventory is also a useful component of the evaluation (16). The BDI is a 21 question multiple choice self report inventory that is a widely used measure for assessing the symptoms and severity of depression. Questions relate to the emotional manifestations of depression, such as hopelessness, irritability and feelings of guilt, as well as somatic symptoms such as fatigue and fluctuations in appetite. The BDI reflects the diagnostic criteria for depression described in the DSM. The Wechsler Adult Intelligence Scale (WAIS-III) is a widely accepted, empirically validated measure of overall intellectual and cognitive functioning. It assesses both verbal and nonverbal abilities, and can yield valuable information regarding a patient's ability to understand consequences of his actions, as well as the presence of any intellectual deficits. The WAIS can also be useful in elucidating difficulties in attention and concentration, as well as the impact of anxiety on intellectual functioning (17).

A thorough evaluation also incorporates projective measures such as the Thematic Apperception Test, Draw-A-Person and the Rorschach Inkblot Test (18–20). These measures are designed to assess a patient's unconscious attitudes and experiences that might not be accessible via clinical interview and self-report measures. The Thematic Apperception Test is a technique for the assessment of personality dynamics as they manifest in interpersonal situations. The Thematic Apperception Test taps into thoughts and feelings about oneself and others that might not be accessible to one's conscious mind. The TAT consists of a standard series of 31 provocative yet purposely ambiguous pictures about which the subject must tell a story. The subject is asked to include what might have happened before, during and after the image, as well as what the characters might be thinking and feeling. Recurrent themes reflected throughout the stories tend to mirror the subject's personality dynamics, as well as how the subject experiences himself in the world. The Draw-A-Person Test requires subjects to draw pictures including a house, tree, male and female human figures, and a family. These items are universally accepted and recognized. The subject is given complete freedom to draw these items, with no guidelines. He is then asked to tell stories about the drawings. Typically, drawings of the house can represent concerns about self and body image, while pictures of the tree often give the clinician insight into the subject's fantasy life (18). Pictures of human figures reflect real and idealized self-image, while the family drawing can offer clues as to the subject's subconscious experience of his own family dynamics. Of interest, patients evaluated by the author often drew houses in need of renovation, or with a recent addition.

The Rorschach Inkblot Test consists of a series of 10 inkblots. The subject of much empirical research, there are several empirically derived scoring systems that are used to interpret subjects' responses to the inkblots. Perhaps the most widely utilized is the Exner Comprehensive System, which assesses emotional, cognitive, and interpersonal functioning on a variety of dimensions (19).

EVALUATION OF PATIENTS FOR COSMETIC LIMB-LENGTHENING SURGERY

Patients who opt for this procedure present with a marked preoccupation with their height. Height dysphoria can be unrelated to actual physical height, and can sometimes be indicative of body dysmorphic disorder (BDD), which is a serious psychiatric condition that involves excessive preoccupation with a physical attribute (2,5). Patients with BDD repeatedly alter or obsess about the offending body characteristic to the point at which their global functioning is significantly impaired. Most patients with BDD show little to no psychological improvement post operatively, and often request further surgical procedures (6,21,22). Reports consistently suggest that BDD should contraindicate any elective cosmetic procedure (5,6,21–23). For some patients, surgery represents the only means by which positive self-esteem can be restored; for

others, surgery would provide remediation of a perceived physical deficit; however, these patients will often report an ability to lead a satisfying and fulfilling life should surgery not be an option. Overall, a psychological evaluation should assess a patient's internal and external motivation for and expectations of surgery, as well as psychological expectations and possible benefits derived from the procedure (4,6,22). Because limb-lengthening surgery involves a difficult postoperative course where the patient initially has limited ability to take care of himself without assistance, it is imperative that a patient's understanding of this process, as well as his psychological response to dependency and helplessness be assessed. Social support throughout the course of treatment should also be evaluated (4,23).

A detailed clinical interview generally focuses on understanding when a patient was first aware of his perceived physical deficit, the impact of the deficit on interpersonal and professional functioning, and what, if any attempts were made to compensate for the perceived deficit. Because early messages in childhood and adolescence appear to influence adult body image, an understanding of parental and peer attitudes toward the patient's stature should be assessed (4,5,7). Most developmental theories hold that one internalizes these attitudes and incorporates them into one's sense of self. Anecdotally, many patients evaluated by this author consistently reported parental anxieties and concerns about their height. Similarly, it is not uncommon for these patients to describe psychological experience in concrete, body-focused language. For example, patients will discuss ways in which they did not ''measure up'' to siblings, parents who made them ''feel small,'' or peers who ''looked down'' on them.

Structured measures designed to assess the presence of psychopathology can also yield useful information regarding a patient's motivation for and expectations of surgery (6). With the exception of BDD, the presence of a psychiatric condition might not necessarily preclude suitability for surgery especially if the patient is actively undergoing treatment for the condition (9,11,23). Studies suggest, however, that poor outcomes are associated with unrealistic expectations of the procedure; for example, a belief that surgical correction of the ''defect'' will be a panacea for all of life's woes; a history of multiple cosmetic procedures, and motivation for surgery based on relationship or other external motivators (4,6,8,22). As such, these patients are generally deemed unsuitable for the procedure.

Measures such as the Rorschach Inkblot test can be extremely useful in gauging a patient's underlying experience of his body. For example, in this author's sample of patients evaluated for limb-lengthening surgery, frequently seen percepts included decaying body parts, withered limbs, and an overall preoccupation with bodily functioning. Similarly, when asked to draw pictures of people, these patients often drew small bodies with oversized heads, regardless of artistic ability. It should be noted that while the interpretation of these clinical observations have not been empirically substantiated, they are generally consistent with studies outlining the psychological presentation of patients with a continuum of body image disturbances (18,19,24).

CASE EXAMPLES

Mr. A presented as a 48-year old, married man who reported generally positive relationships with peers, as well as a satisfying career. He reported that his stature had always made him feel inferior in relation to others, despite his personal and professional achievements. When asked how he might proceed if unable to have limb-lengthening surgery, he noted that he would get on with his life and find other ways to compensate for his perceived deficit. The clinical interview, as well as the MMPI, SCL-90, and BDI yielded normative responses, with no overt indication of psychopathology. Projective data, however, was suggestive of a marked preoccupation with his body, with images including decaying, truncated body parts, and helpless diminutive creatures. Although this finding did not contraindicate suitability for surgery, it was suggestive of the impact of Mr. A's height neurosis on his overall sense of self.

In response to a TAT picture, Mr. A provided the following narrative:

> ''The boy is remembering a surgery he had had or witnessed. He is wearing a suit, so you can see he's doing okay. Maybe he is thinking about the future, spending time with his family. I see a good outcome in this picture. He has a good life.''

The following narrative was provided in response to Mr. A's drawing of a house:

> "This house is in the suburbs. It's a single-family house with a two-car garage, two trees in front with a big backyard. The bedrooms are really warm and personal. The garage is a little disorganized but I think that eventually it gets cleaned out. It is on a block with other houses. In the future, I see an addition to the house—a bigger kitchen or another room, maybe."

These stories were reflective in theme and affective tenor, of the majority of stories provided in the battery of tests. They suggest a rich, positively charged internal life, as well as a sense of connectedness to others in his world. Mr. A eventually did have limb-lengthening surgery and reported feeling great relief from distress around his height.

Mr. B presented as a 25-year old, single man who reported feeling socially isolated with limited career options, both of which he attributed to his short stature.

The clinical interview yielded discussion of intense sibling rivalry that was covertly encouraged by his parents, which left Mr. B feeling as if he could never live up to their expectations of him. Mr. B noted during the interview that if he were taller, he might finally be able to have an impact on his family. He also indicated that if he were unable to have limb-lengthening surgery with the surgeon of his choice, he would consider searching elsewhere, despite financial and occupational constraints. Structured measures indicated the presence of mild depression. Projective measures revealed a preoccupation with his body integrity, with all negative thoughts and feelings focused on his body. In response to a TAT picture, Mr. B reported:

> "It is a soldier on the operating table—no one here is happy. There is no anesthesia. He never had much of a promising future to begin with. He eventually recuperates but he is regretful about the path he took. He was at the wrong place at the wrong time."

When asked to draw a picture of a house, Mr. B provided the following narrative:

> "A simple 'A' frame house. A pretty empty place to live, though no one lives there now. It's alone, on a rural road. It's not in good shape; it could be painted better. It could be bigger and prouder than it is. In the future, maybe the owner adds a garage, an extra bedroom, window boxes, maybe a garden. Then it'd be a mansion!"

These stories, taken together with themes woven throughout the majority of the test data, suggest some underlying depressive ideation, as well as a sense of himself as being at the mercy of his environment, rather than an active participant in his own life. In this case, it was recommended that Mr. A seek psychological remediation of his emotional difficulties, and revisit the prospect of surgery some time in the future.

AREAS OF FUTURE RESEARCH

Although empirical evidence supports, to some degree, the role of cosmetic surgery as a means of repairing disturbances in body image, there is little information about limb-lengthening surgery as a successful means of redressing concerns about stature. Further research should more clearly elucidate the relationship between body image dissatisfaction and limb-lengthening surgery. Additionally, research should more clearly define the characteristics of patients suffering from a circumscribed height neurosis, rather than more diffuse psychological difficulties. Research should also be undertaken to identify more clearly both patients for whom surgery may be beneficial, as well those for whom it is contraindicated. Studies should also be designed to assess the long-term health and general life satisfaction of those patients who elected to undergo elective limb-lengthening surgery. Finally, anecdotal reports suggest that a majority of patients who present with short stature dysphoria are men. It would be of interest to better understand how men and women differ psychologically, with regard to their interest in, and decision to pursue cosmetic limb-lengthening surgery.

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Part XII TECHNOLOGY

45 Taylor Spatial Frame

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INTRODUCTION

The Ilizarov system utilizes hinge and translation mechanisms, which are specifically oriented for a given case. Complex deformities are addressed by frames, which include hinge (rotation) and translation mechanisms in series or stages.

The Taylor Spatial Frame fixator consists of two rings or partial rings connected by six telescopic struts at special universal joints (Fig. 1). By adjusting only strut lengths, one ring can be repositioned with respect to the other. ''Simple'' or complex deformities are treated with the same frame. The multiple angles and translations of a given deformity are addressed by adjusting the lengths of struts only. The Taylor Spatial Frame fixator is capable of correcting a six-axes deformity.

PRIMARY METHODS OF TREATMENT

Acute Fracture Treatment with FastFx™ Struts

First a frame is mounted. (See illustration series below: Fig. 2A—foam bone; Fig. 2B—in surgery.)

With the struts in their sliding mode, the fracture is acutely reduced under direct vision or C-arm control and the strut slides locked. The frame is first fixed to the bone without worrying about reduction. The bone is then reduced without worrying about fixation. If the manual reduction is perfect, no further fine adjustment is necessary. In the postoperative period, any additional adjustment may be gradually performed without repeat anesthesia utilizing the Total Residual Deformity Correction Program.

CHRONIC DEFORMITY CORRECTION

Chronic deformity applies to congenital deformity, malunions, and stiff nonunions that can be measured fairly accurately by radiographs. The deformity should not change on a minute-to-minute basis, but allow accurate orthogonal radiographs to be taken. Based on these radiographs and a clinical exam, a Spatial Frame can be adjusted from its neutral or home position (at which all struts are equal length) to a deformed position that exactly matches the skeletal deformity. The Spatial Frame is then fixed to the skeleton. Because the frame is returned to its neutral or home position, the fragments are restored to their anatomic positions. This process is called Chronic Deformity Correction. A structure at risk and a safe velocity of correction may be decided and a daily adjustment schedule is generated by the computer program (Fig. 3A–C).

TOTAL RESIDUAL DEFORMITY CORRECTION

A more recently developed software program allows the surgeon to basically correct any skeletal deformity mounted to any arbitrarily adjusted Spatial Frame, sometimes referred to as the problem of a ''crooked bone in a differently crooked frame.'' As in the chronic deformity correction program, a structure at risk and a safe velocity may be selected to precisely control the correction (Figs. 4A–C, 5, and 6).

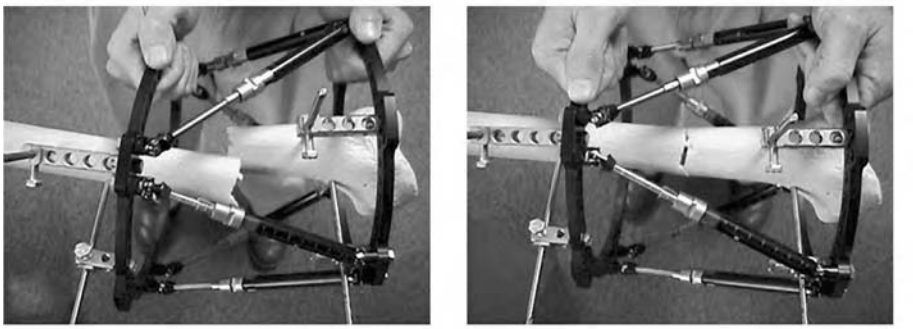
Figure 1 The Taylor Spatial Frame fixator consists of two rings or partial rings with six telescopic struts attached at special universal joints. The universal joints are passive and do not require clamping. Strut lengths are changed by rotating an adjustment knob. These strut lengths may be read directly off each strut.

CHOOSING A REFERENCE FRAGMENT AND THE ORIGIN

Orthopedic convention characterizes the deformity of the distal fragment with respect to the proximal fragment. (The proximal fragment is the reference fragment; the distal fragment is the moving fragment.)

Deformities could also be measured where the proximal fragment is characterized with respect to a reference distal fragment. This characterization of the deformity by describing abnormal position of the proximal fragment is especially useful in distal nonunions or malunions with a short distal fragment. The location of the attachment of the distal ring (using the joint surface as a landmark) will be more exactly determined in preoperative planning and in surgery than the level of attachment of the proximal ring on the longer proximal fragment. It also allows the surgeon to fully characterize the deformity even though the radiographs are too short to include the level of attachment of the proximal ring. (The distal fragment is the reference fragment; the proximal fragment is the moving fragment.)

(A)



(B)

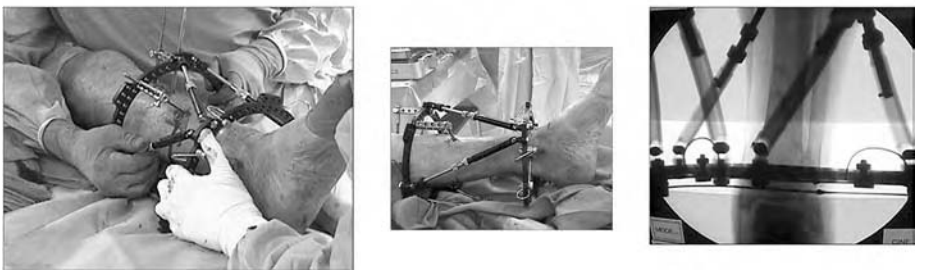


Figure 2 (**A**) Foam bone model showing frame mounted on bone with all struts unlocked. (**B**) After pin and wire fixation of each major fragment, with all struts unlocked, the fracture is reduced under C-arm control or direct vision for open fractures. Each strut is then locked, thereby securing the reduction.

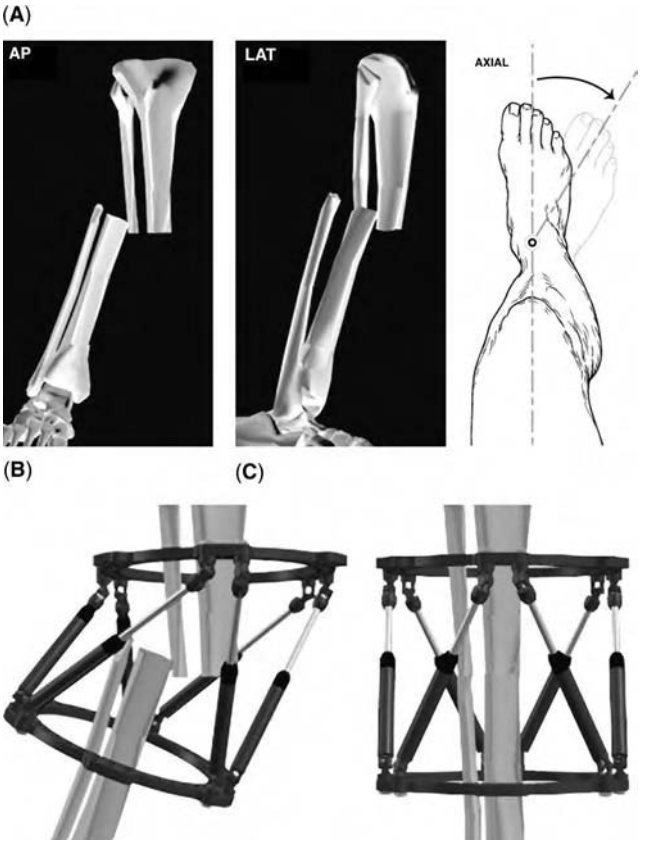


Figure 3 Chronic Deformity—For deformity correction, the surgeon measures AP and lateral radiographs and performs a clinical exam, which yield the six deformity parameters (**A**). Clinical exam determines which ring diameters to use and whether longer or shorter struts are required—the three frame parameters. The surgeon anticipates the position of the frame with respect to the origin (usually the interior end of the reference fragment), thus providing the four mounting parameters. These 13 parameters are input to a Chronic Deformity Correction Program which returns six specific strut lengths to adjust the Taylor Spatial Frame fixator to exactly mimic the deformity (**B**). The frame is then attached to the skeleton. The deformity is fully corrected as the struts are restored to their neutral length (**C**). *Abbreviation*: AP, anteroposterior.

Either fragment could be the reference fragment. Ideally, the reference fragment should satisfy two criteria:

1. The fragment whose anatomic planes most closely match the planes of the anteroposterior (AP) and lateral radiographs;
2. AP and lateral radiographs include the actual or anticipated level of attachment of a ring to the reference fragment.

The patella provides a prominent landmark for distal femoral or proximal tibial deformities. The foot provides a prominent landmark for distal tibial, ankle, and subtalar deformities. Frequently, the best choice for the reference fragment is the short fragment in conjunction with the prominent landmark. The X-ray technician is most likely to successfully align with the landmark (criteria 1) and if the joint line is included in the radiographs (as it should!), then the level of attachment of a ring to that fragment is also included (criteria 2).

Obviously, the actual deformity is the same whether the physician characterizes the distal fragment with respect to the proximal fragment or, alternatively, characterizes the proximal fragment with respect to the distal fragment. However, the working measurements of even an oblique plane angular deformity will be different depending upon which fragment is chosen for the reference fragment. Strangely enough, the final external fixation frames for these different deformity characterizations (based on alternative reference fragments) are identical and will effect the same complete correction. It is important that the same fragment be maintained as reference fragment for AP and lateral radiographs as well as clinical exam for malrotation.

Translation between fragments is measured from an origin on the reference fragment to its corresponding point on the moving fragment. The best choices for origin and corresponding point are points that are coincident in the anatomic (reduced) state. The tip of a spike on the reference fragment and the matching 'notch' on the moving fragment would be reasonable

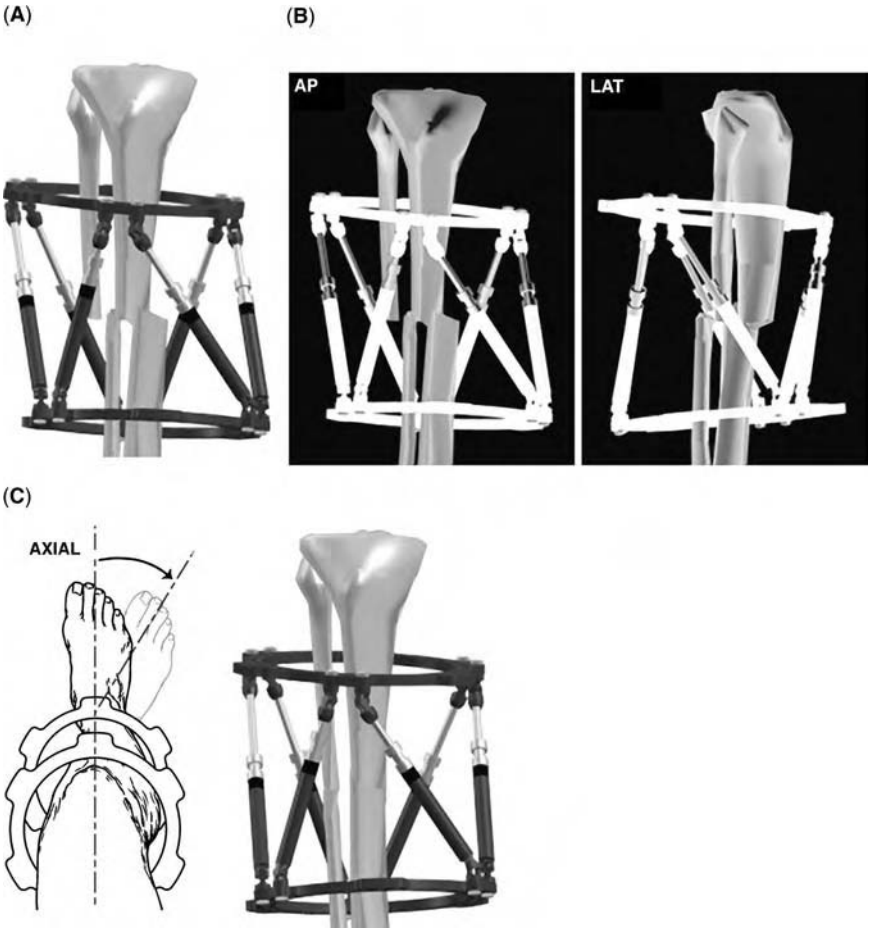


Figure 4 Total Residual Deformity Correction will fully correct any remaining skeletal deformity already mounted to a Spatial Frame. The Total Residual Deformity Correction can be undertaken at any time for fractures or during a chronic deformity correction. This method requires the surgeon to record each of the current 6 strut lengths for the computer program. At any time with a frame mounted to the bone (**A**), AP and lateral radiographs are obtained, six current strut settings are recorded, and a clinical exam is performed (**B**). From the radiographs, the six fracture deformity parameters and the four mounting parameters are measured. These 10 parameters, the two ring diameters, and the current strut settings are input to a Total Residual Deformity Correction Program, which returns six specific strut lengths to adjust the Spatial Frame to exactly correct the deformity. The deformity will be fully corrected when the struts are moved from their current settings to their specified lengths (**C**). *Abbreviation*: AP, anteroposterior.

Figure 5 Like the Ilizarov, the Taylor Spatial Frame fixator may be used to lengthen or shorten a limb by adjusting all struts by the same increment. Unlike the Ilizarov, there is no preload on the frame as adjustments to the struts are made. Because the universal joints are free to rotate, *any* combination of six strut lengths is a valid frame.

Figure 6 Regardless of the complexity of the case, essentially the same Taylor Spatial Frame fixator is used. In this case, of juxta-articular deformity, a Taylor Spatial Frame can accomplish the same rotation and translation as Ilizarov frames.

choices in post-traumatic deformity. These points are easily discerned on AP and lateral radiographs (Fig. 7).

However, the mechanical axis at the fracture site of the reference fragment and the mechanical axis at the fracture site on the moving fragment are the most commonly used choices for origin and corresponding point, respectively (Fig. 8). The implied coordinate system on which these translational and rotational measurements are made is the coordinate system of the reference fragment. Thus, imagine a grid aligned with the mechanical axes of the reference fragment. The AP view translation, lateral view translation, and axial translation are measured along these grid lines.

Choosing the origin and corresponding points for congenital and remodeled deformities will be presented in a later section.

PARAMETERS

By fully characterizing skeletal deformity, determining the appropriate frame size, and establishing the position of the frame on the limb, the surgeon can correct complex deformity and reduce fractures utilizing the Chronic Deformity Correction Method or the Total Residual Deformity Correction Method.

Skeletal deformity is completely characterized by measuring six deformity parameters: the three projected angles (rotations) and three projected translations between major fragments.

The frame parameters consist of proximal and distal ring diameters and neutral strut length or neutral frame height for chronic deformity correction. Total residual deformity correction requires all six current strut settings.

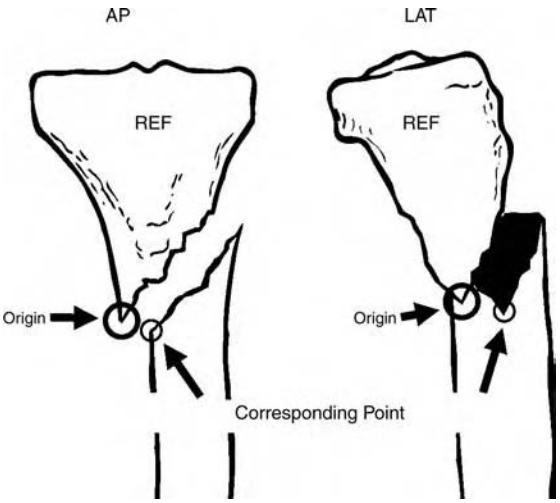


Figure 7 A spike and its matching notch, if discernible on AP and lateral films, is one possible choice for origin and corresponding point to measure translations at the fracture site. *Abbreviation*: AP, anteroposterior.

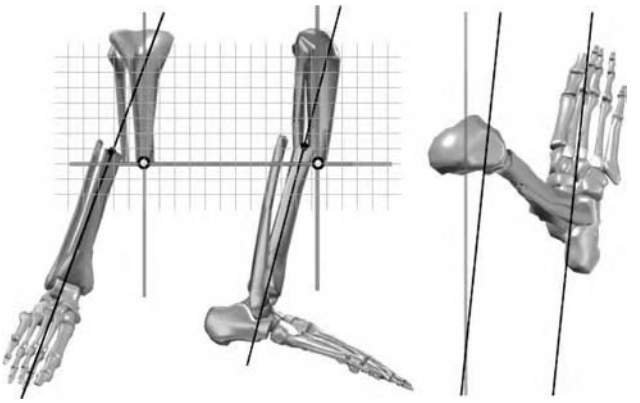


Figure 8 Measurements of translation and rotation are made along an imaginary grid aligned to the reference fragment.

Four mounting parameters are anticipated before surgery for chronic situations and measured radiographically and clinically after surgery for total residual deformity corrections. They are AP view, lateral view, axial, and rotary frame offsets.

These parameters are input in a computer program, which determines the strut lengths for the Taylor Spatial Frame Fixator.

Frame Parameters

The frame is completely described by the inner diameters of the proximal and distal rings and the six current strut settings.

Proximal and Distal Ring Internal Diameters

Complete rings range in size from 80 to 300 mm internal diameter in 25 mm increments. Two-third rings range from 80 to 300 mm. Special U-Plates, used about the ankle and foot are available in 105, 130, and 155 mm. Different size rings may be used on one frame. Tapered frames allow a lower profile mounting. A two-third ring permits more proximal fixation of the femur and humerus. Accessory rings and partial rings may be attached to extend the levels of fixation. Short and long footplates are available in 155 and 180 mm internal diameters. The internal diameter is printed on each ring, partial ring, or footplate (Fig. 9A–D)

Neutral Frame Height and Neutral Strut Length

Standard telescopic struts are available in x-short, short, medium, and long sizes ranging in functional length from 75 to 284 mm (Fig. 10). For a given size, the strut has a specific range from its shortest to longest length and a mid-position marked on each strut. Struts are marked with millimeter graduations with actual strut length printed every 10 mm. The strut length is read at the indicator (Fig. 11). FastFx^{TM} struts, likewise, are available in x-short, short, medium, and long sizes ranging in functional length from 91 to 311 mm.

Struts are attached to the rings in a specific pattern with special shoulder bolts. The holes in the rings for attaching the struts are apparent. Mounting holes in the footplates are highlighted by a shallow groove. When a strut runs out of travel or excursion, it can be exchanged for the next size strut. Before the exchange, simply apply an additional telescopic strut or threaded rod temporarily between rings. Any available holes may be used for this temporary seventh strut. After the exchange, remove the temporary strut. The short strut in its fully lengthened position overlaps the medium strut in its fully shortened position by 10 mm. Likewise, the medium strut in its fully lengthened position overlaps the long strut in its fully shortened position by 10 mm. Thus, the surgeon has some choice when the struts are exchanged. At the time of exchange, simply set the strut to be inserted at the same length as the strut being replaced (Fig. 12).

The neutral frame height is the distance from the center of one ring to the center of the other ring with all struts at their neutral length. The neutral frame height or the neutral strut length is chosen by the surgeon preoperatively for chronic deformity correction. This neutral frame height will be the target or final destination that will be achieved at full correction (Fig. 13).

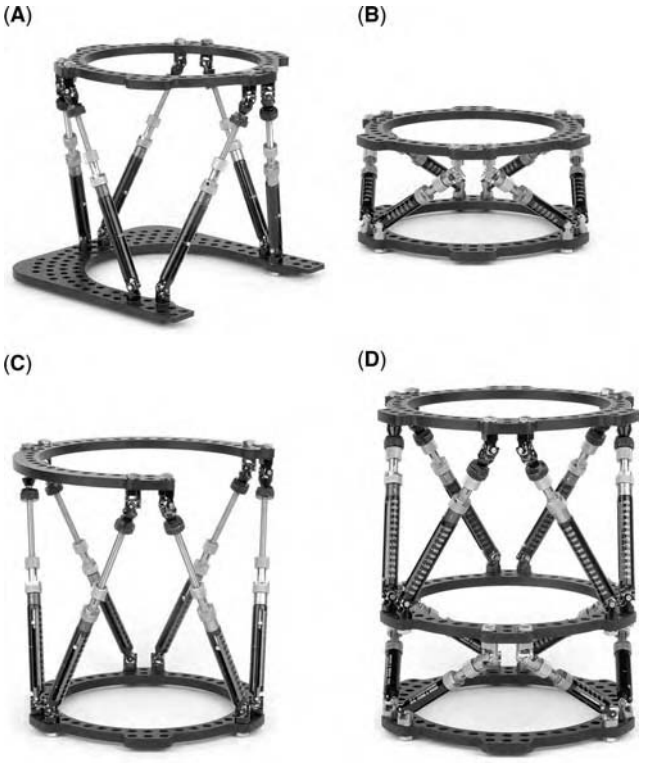


Figure 9 (**A**) Spatial Frame with short footplate and medium struts in mid-position. The short footplate would allow independent treatment of the forefoot. (**B**) Shorter struts allow more stable fixation when indicated. Accessory rings could be added to extend level of fixation. (**C**) The component system permits custom frames such as this tapered open section frame for distal femoral application. (**D**) Each ring has six tabs and can serve as the intermediate ring for a segmental application. Spatial Frames come preassembled or can be assembled from components.

The Taylor Spatial Frame software can use either neutral frame height or neutral strut length to calculate adjustments to the frame. Also, given one of these values, the software will immediately return the other, which may be useful in frame planning with the chronic method. When performing a chronic deformity correction with primarily angular and transverse plane translational deformity, choose the neutral strut length equal to the mid-length of one of the strut sizes. This permits some struts to shorten and some to lengthen without needing to exchange struts. If primarily performing a lengthening, choose a neutral strut length toward the lengthened end of the particular strut size. This better utilizes the available excursion and may decrease the need to exchange struts.

Deformity Parameters

The orthopedic surgeon must assess the patient for fixed pelvic obliquity. Long films (including hip, knee, and ankle) with a radiographic ruler are helpful to assess mechanical and anatomic axes and identify all deformities. Weight-bearing films identify ligamentous laxity, which may mimic skeletal deformity.

Imagine a limb segment in anatomic position as in Figure 14A and B. The two fragments adjoin at the origin. With fracture or deformity, the two fragments are angulated and translated. The translations are measured as the separation of the adjacent (or corresponding) point

Figure 10 Struts are available in xxshort, xshort, short, medium, and long bodies.

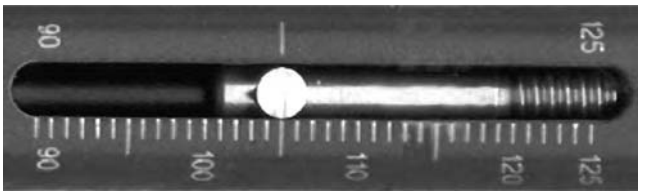


Figure 11 A close-up of a short strut shows the indicator at mid-position.

from the origin. Translations are measured along the coordinate axes of the reference fragment (actually the reference ring)(Fig. 14B).

Determine whether a conventional distal characterization or a proximal characterization is to be used. Angulations are determined by measuring the divergence of centerlines drawn in each fragment, as with traditional methods. Axial malrotation (internal or external rotation) is assessed clinically or with special films (Fig. 15).

Translation (displacement) is the perpendicular distance from the reference fragment to its corresponding point on the moving fragment.

To measure translations, determine where the corresponding point is with respect to the origin. *Note*: If a distal reference is chosen, the AP view and lateral view translations will generally be opposite those if a proximal reference is chosen.

Note: Angulation and rotation are determined by the reference fragment's view of the deformity in a traditional sense. For example, if a proximal reference is chosen and the AP view along the reference fragment shows a varus deformity, then it is a varus deformity.

If a distal reference is chosen and an axial view is taken along the axis of the distal fragment (usually clinical exam), which shows the distal fragment internally rotated with respect to the proximal fragment, it is an internal rotation deformity.

Mounting Parameters

Axial Frame Offset

Axial frame offset is the measurement of length parallel to the frame centerline from the origin to the reference ring (Fig. 16). This can generally be measured on AP or lateral films. This measurement in millimeters partially specifies the orientation of the frame with respect to the origin.

Anteroposterior View and Lateral View Frame Offset

In most tibial mountings with circular fixators, the tibia is located anterior to the geometric center of the ring. Measure the distance from the origin to the centerline of the frame. This distance in millimeters is lateral view frame offset. If the tibia is shifted from center on AP view, measure the distance from the origin to the centerline of the reference ring. This distance is AP view frame offset (Fig. 17).

The surgeon can choose the exact virtual center of rotation of the correction. This virtual center of rotation, the origin, can be placed at the convexity of the deformity rather than the center of the interior end of the moving fragment. Rotation at the convex cortex will be necessary especially for correction of congenital deformities, malunions, and stiff nonunions that require minimal or no lengthening. Otherwise, too much impaction and overconstraint at the convex cortex may result in excessive preload on pins and wires and undercorrection of the deformity.

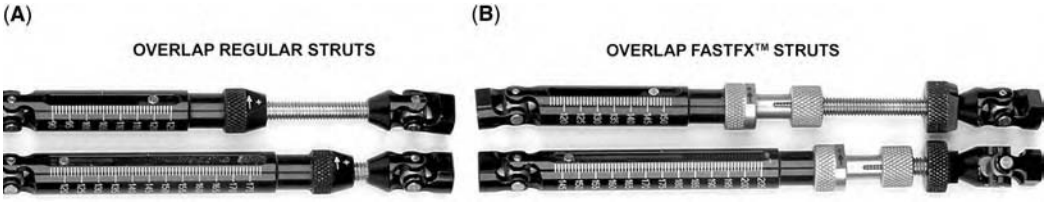


Figure 12 The achievable length of struts overlap by 5 to 10 mm from one body to the next in the series.

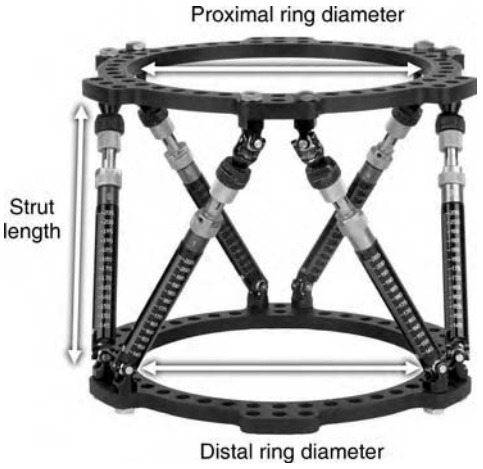


Figure 13 The Frame Parameters, the description of the frame, consists of the internal diameters of the ring, which are printed on the rings and the strut lengths, which are read at the indicator of each strut.

When treating stiff nonunions, malunions, and congenital deformities, the position of the frame with respect to the origin can be anticipated. Corresponding values for axial frame offset, lateral view frame offset, and AP view frame offset are entered into the Chronic Deformity Correction Program to determine exact strut lengths for the Taylor Spatial Frame to mimic the given deformity, maintaining the anticipated relative position of frame and bone.

When treating fresh fractures, axial frame offset, lateral view frame offset, and AP view frame offset are measured on postoperative films. These values are entered into the Total Residual Deformity Correction Program to determine exact strut lengths for the Spatial Frame to compensate for the residual deformity.

Rotary Frame Offset

The preferred (reference) rotational orientation of the Taylor Spatial Frame is with the proximal ring universal joints (master universal joints) for Strut 1 and Strut 2 located exactly anterior on the proximal fragment (Fig. 18).

When using two-third rings for mid-femur or humerus, the surgeon may place the master universal joints directly lateral by selecting 90° of external rotation for some femoral and humeral applications (Fig. 19A and B). When treating stiff nonunions, malunions, and congenital deformities, the position of the frame with respect to the bone can be anticipated, and corresponding values for rotary frame offset (Fig. 19C) are entered into the Chronic

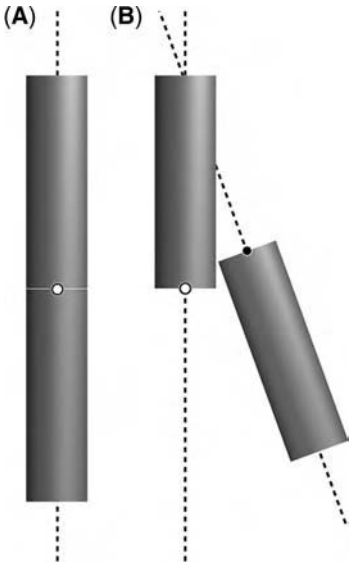


Figure 14 (**A**) When anatomically reduced, the origin and corresponding point are coincident and there is no angulation or rotation between the fragments. (**B**) In general, there could be a six-axis deformity after initial fracture reduction, consisting of three translations and three angulations.

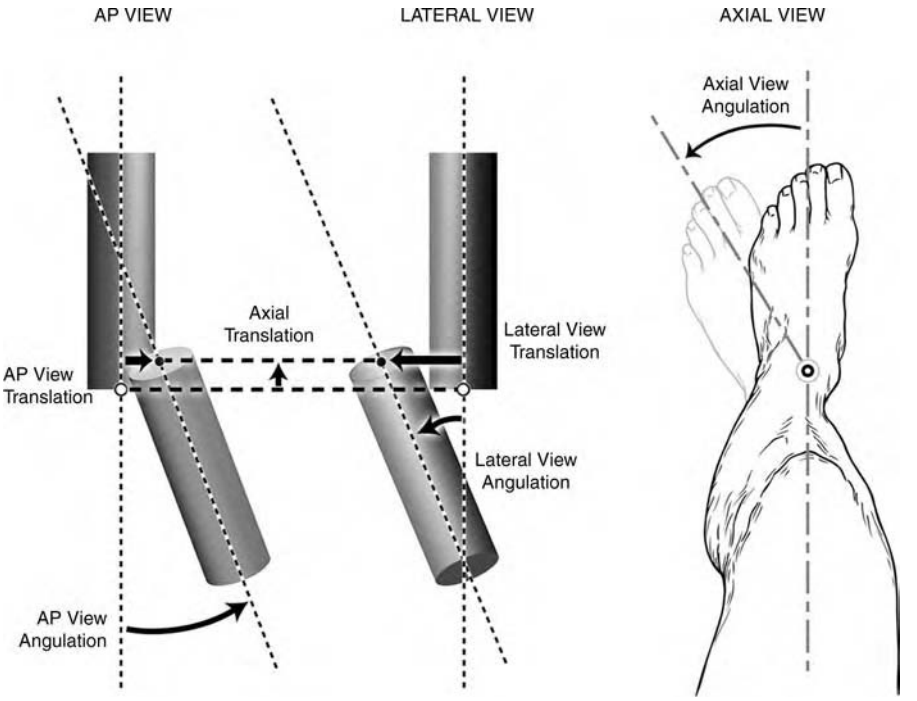


Figure 15 Translation (displacement) is the perpendicular distance from the reference fragment to its corresponding point on the moving fragment. To measure translations, determine where the corresponding point is with respect to the origin. (If a distal reference is chosen, the AP view and lateral view translations will generally be opposite those if a proximal reference is chosen.) *Note*: Angulation and rotation are determined by the reference fragment's view of the deformity in a traditional sense. For example, if a proximal reference is chosen and the AP view along the reference fragment shows a varus deformity, then it is a varus deformity. If a distal reference is chosen and an axial view is taken along the axis of the distal fragment (usually clinical exam) that shows the distal fragment internally rotated with respect to the proximal fragment, it is an internal rotation deformity. *Abbreviation*: AP, anteroposterior.

Deformity Correction Program to determine exact strut lengths for the Spatial Frame to mimic the given deformity maintaining the anticipated relative position of bone and frame.

When used for fractures, the frame may be inadvertently malrotated when applied. Simply enter the angular position of the sagittal plane of the reference ring with respect to the reference fragment in rotary frame offset.

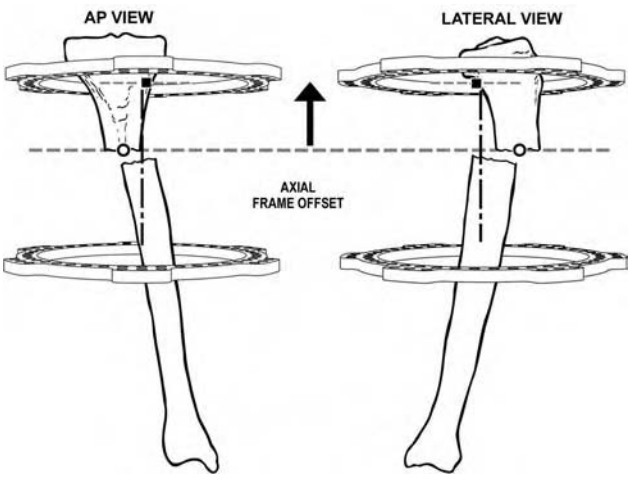


Figure 16 The mounting parameter, Axial Frame Offset, measures the axial distance from the origin to the reference ring.

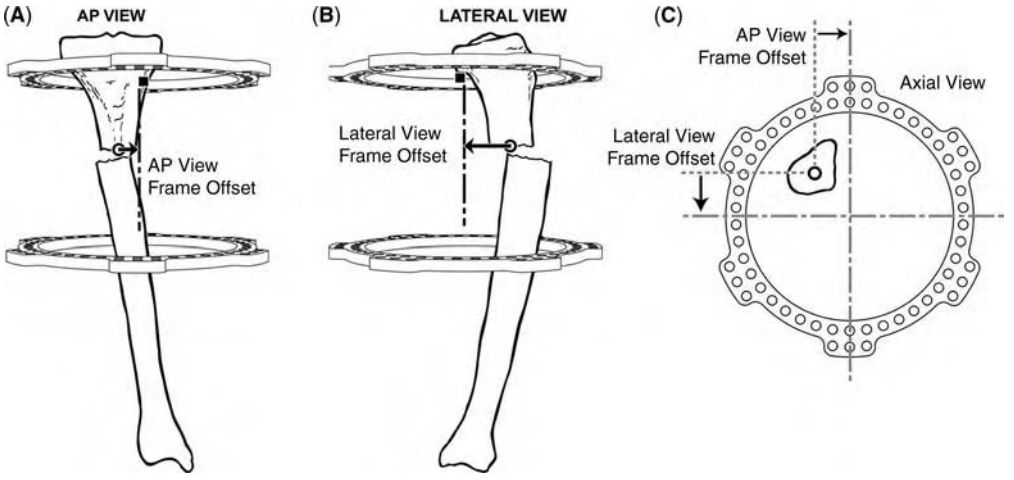


Figure 17 (**A**) AP View Frame Offset, one of the four mounting parameters, is measured from the origin to the centerline of the frame on AP view. (**B**) Lateral View Frame Offset, measured on lateral view, describes the distance from the origin to the centerline of the frame. (**C**) The AP and Lateral View Frame Offsets describe how the center of the reference ring is positioned with respect to the origin in a transverse plane. *Abbreviation*: AP, anteroposterior.

STRUCTURE AT RISK AND RATE OF CORRECTION

It is incumbent upon the surgeon to be aware of the structures at risk on the concavity of the deformity. When dealing with rotation about the longitudinal axis in addition to conventional angular correction, the risks may be less or greater depending on direction of axial rotation. For example, when correcting a flexion/valgus/external rotation deformity of the proximal tibia, the peroneal nerve is at increased risk. However, when correcting a flexion/valgus/internal rotation deformity, the axial rotation will tend to offset the stretch on the peroneal nerve created during the correction of flexion/valgus.

The chronic and total residual programs allow the surgeon to input the coordinates of the structure at risk with respect to the origin and the maximum daily displacement of the

Figure 18 The Spatial Frame computer program assumes the Master Tab is always directly anterior on the proximal ring.

structure at risk. The program creates a daily adjustment schedule moving the structure at risk by the prescribed amount each day until the deformity is eliminated.

CHRONIC DEFORMITY—A SECOND CHANCE FOR CORRECTION

Because of nonorthogonal initial radiographs, error in measuring radiographs, or excessive preload and bending of wires and pins, there may be residual skeletal deformity when the struts have reached their neutral lengths at the completion of a chronic deformity correction. Simply measure the radiographs to determine residual skeletal deformity parameters. Make a clinical exam for malrotation. Record the six strut lengths. The mounting parameters can be revised if necessary. Use the Total Residual Deformity Correction Program to determine new strut lengths to correct the residual deformity.

During any gradual correction of fractures or chronic deformity, a total residual deformity correction may be undertaken by measuring current parameters and noting current strut settings.

CHOOSING THE ORIGIN AND CORRESPONDING POINT FOR CONGENITAL AND REMODELED DEFORMITIES—ADDITIONAL CONSIDERATIONS

Congenital deformities, old remodeled malunions, and two bone deformities (tibia-fibula, radius-ulna) require additional considerations. The level of the origin (transverse plane) could be arbitrarily chosen provided the corresponding point could be determined. However, the most useful levels for establishing the origin has been the knee joint line for distal femoral deformities; proximal tibia-fibula joint for proximal tibial deformities; the ankle joint line for distal tibial deformities; and the subtalar joint for ankle and hindfoot deformities. The level of the anatomic center of rotation of the knee is a convenient level for the origin in correction of knee contracture.

Remodeled shaft deformities should have the level of the origin set at the intended osteotomy or a nearby anatomic landmark such as a boney prominence or broken screw. It is essential that the level of the origin can be accurately determined on both AP and lateral radiographs.

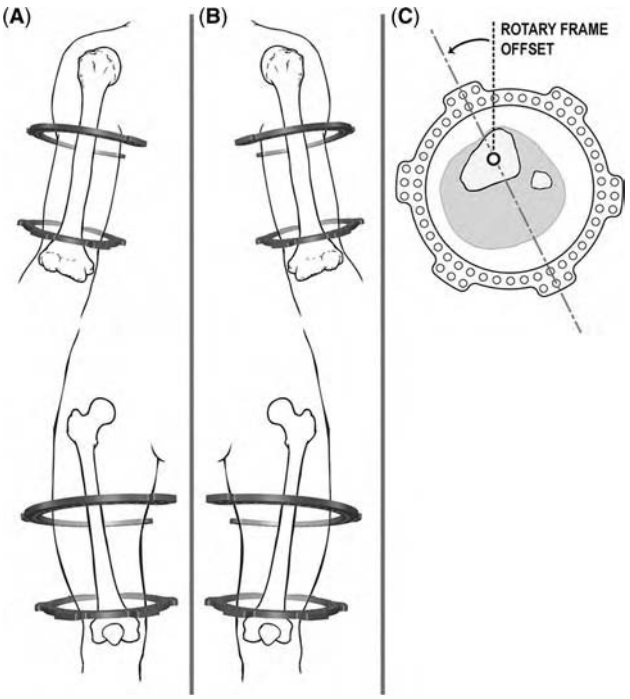


Figure 19 (**A**) To improve trunk and opposite thigh clearance, the Spatial Frame may be externally rotated 90° for right humeral or femoral applications. (**B**) To improve trunk and opposite thigh clearance, the Spatial Frame may be externally rotated 90° for left humeral or femoral applications. (**C**) Rotary Frame Offset, one of the four parameters, is measured clinically as rotation of the sagittal plane of the reference ring with respect to the sagittal plane of the reference fragment.

In general, the corresponding point will not reside at the level of the origin in the deformed position except for pure rotational or pure translational deformities. Even if the true corresponding point resided at the level of the origin in a pure remodeled varus tibial deformity, the best choice for determining the level of the corresponding point would reflect that the tibia is functionally short. Then, after the tibia is osteotomized and the angular deformity corrected, sufficient length is gained to prevent the remodeled bone on the convexity from impinging. This is equivalent to positioning the Ilizarov hinge axis to pass along the convexity of a deformity to prevent impaction. For each case of congenital deformity or remodeled malunion, a simple geometric local analysis is performed to determine the best choice of corresponding point to fully correct the deformity and prevent impaction at the osteotomy or to match the length of the other bone on the convexity in a two-bone system.

The mechanical axes are drawn for each of the fragments (Fig. 20). The origin is the intersection of the mechanical axis of the reference fragment and the specific level (transverse plane) that the surgeon has chosen. Generally, the surgeon wants to bring the mechanical axis of the deformed fragment to coincide with the mechanical axis of the reference fragment. The difficulty is determining which point along the mechanical axis of the deformed fragment corresponds to a particular point on the mechanical axis of the reference fragment, which has been chosen as origin.

Geometric Local Analysis of Extra Length to Prevent Impaction

Measure the distance from the origin to the convex cortex of the reference fragment ''*W*'' (Fig. 20). Reproduce a line segment ''T'' of length *W* with one end touching the plane passing through the origin and the other end touching and perpendicular to the mechanical axis of the deformed fragment. The point of contact of this line segment with the axis of the deformed fragment is the corresponding point (Fig. 21). When this corresponding point is reduced to the origin, the deformity will have been reduced with enough length gained to prevent impaction.

Similarly, in a two-bone system, draw the mechanical axes of each fragment. Determine the plane of the origin (Fig. 22). Measure the distance from the origin to the convex cortex of the second bone *W*. Reproduce a line segment T of length *W* with one end touching the plane passing through the origin and the other end touching and perpendicular to the mechanical axis of the deformed fragment. The point of contact of this line segment with the axis of the deformed fragment is the corresponding point (Fig. 23). When this corresponding point is reduced to the origin, the deformity will have been reduced with enough length gained to prevent impaction at the second bone at the convexity.

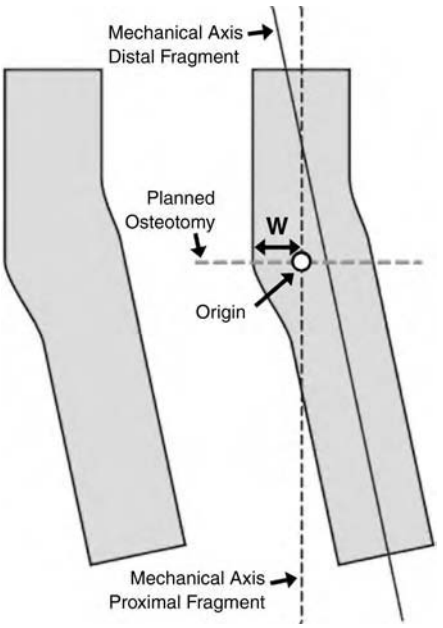


Figure 20 For congenital deformity or malunions, a good location for the origin is the level of the osteotomy. Start by drawing the mechanical axis of each fragment and locate the level of the osteotomy. Let the origin be the intersection of the plane of the osteotomy with the mechanical axis of the proximal fragment. Create a line from the origin to the convexity of the deformity, W.

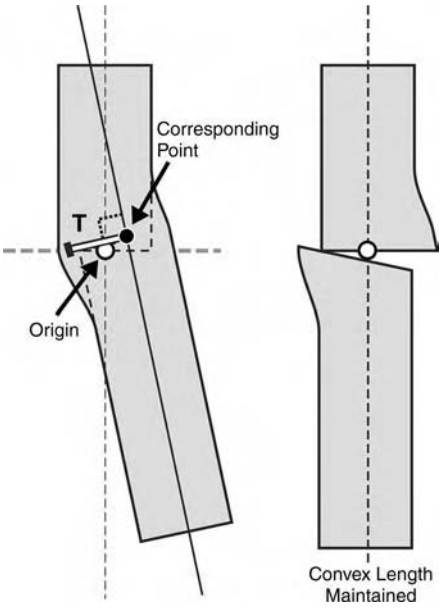


Figure 21 Create a line, T, equal in length to line W, with one end perpendicular to the mechanical axis of the deformed fragment and the other end resting on the plane of the origin. The point of intersection of line T with the mechanical axis of the deformed fragment is the corresponding point for the origin.

Trigonometric Local Analysis of Extra Length to Prevent Impaction

Similarly, a trigonometric analysis may be performed to find the most appropriate corresponding point so that when the angular deformity is corrected, the convex cortices will just clear one another. If W is the distance from the origin to the convex cortex and q is the angle between the mechanical axis of the reference fragment and the mechanical axis of the deformed fragment, then the amount of shortening "S" of the corresponding point is: $S = W \sin q$.

Thus, if a line is drawn distance S (shortened) from the origin, the corresponding point is located at the intersection of this new line and the mechanical axis of the deformed fragment (Fig. 24).

Sometimes it is more useful to utilize the anatomic axes for femoral cases rather than mechanical axes.

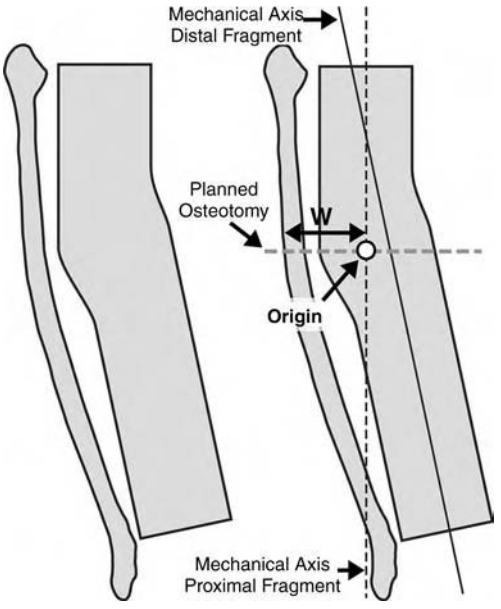


Figure 22 Likewise, for a two-bone system, draw the mechanical axes of the fragments and determine the level of osteotomy and thus the plane of the origin and the origin. Create a line W from the origin to the convexity of the second bone.

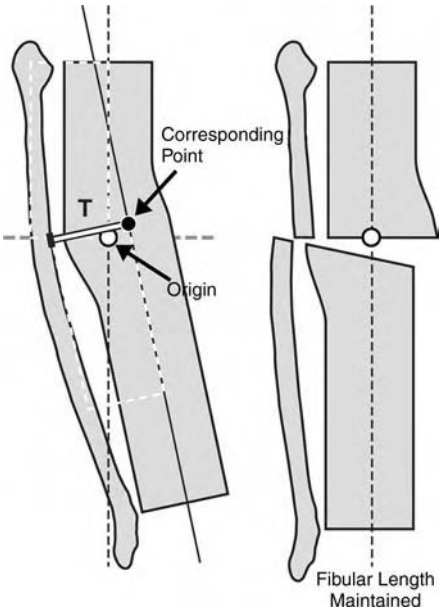


Figure 23 Create a line, T, equal in length to line W, with one end perpendicular to the mechanical axis of the deformed fragment and the other end resting on the plane of the origin. The point of intersection of line T with the mechanical axis of the deformed fragment is the corresponding point for the origin.

INTRINSIC VERSUS EXTRINSIC DEFORMITY

In the previous example, the origin and corresponding point chosen will characterize and thus serve to correct the inherent or intrinsic deformity. The origin selected is the intrinsic origin. When the corresponding point is restored to the intrinsic origin, the angular and translational deformity will be corrected with the corresponding point gaining just enough length through the local analysis to maintain a constant lateral cortical length.

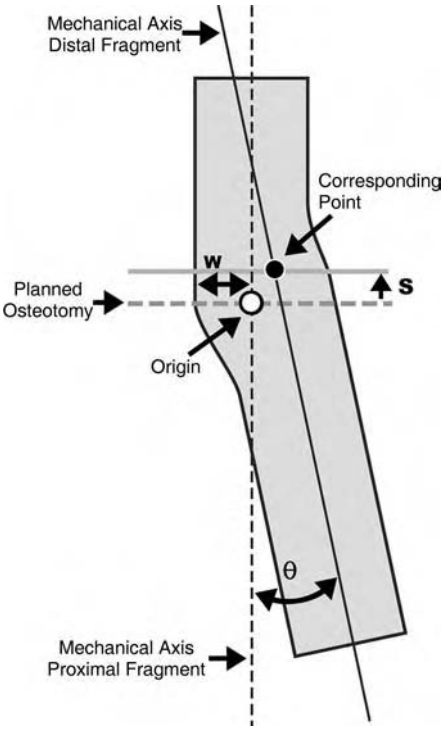


Figure 24 For the one-bone and two-bone system, the preceding analysis yields the AP view angulation theta, the AP view translation, and the amount of shortening *S*. This amount of shortening *S* is the key to unlocking the lateral radiograph. The corresponding point is located on the deformed fragment centerline a distance *S* shortened from the origin. *Abbreviation*: AP, anteroposterior.

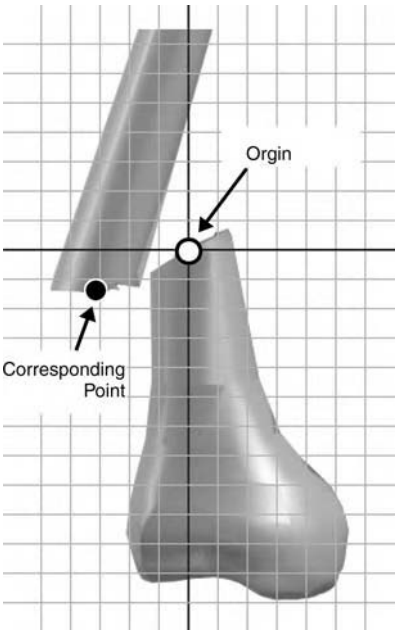


Figure 25 For post-traumatic nonunions of the distal femur, use the center of the distal fragment at the fracture for the origin and the center of the proximal fragment at the fracture as the corresponding point.

In other situations, the surgeon may have other additional or extrinsic information to take into account for a deformity correction. For instance, the physician may know by scanogram that the deformed limb is also 50 mm short or that based on growth charts, there will be a 50 mm limb length discrepancy. This additional shortening deformity is an extrinsic deformity. If the physician wants to correct the intrinsic plus extrinsic deformity, an extrinsic origin is selected 50 mm along the mechanical axis of the reference fragment from the intrinsic origin. The deformity parameters are measured from the extrinsic origin to the corresponding point. For the tibia, humerus, and forearm, the only change is in the axial shortening. The AP and lateral view translations and all rotation and angulations remain the same for an intrinsic or extrinsic correction. If there is a particular level of attachment of the reference ring that is to be used for intrinsic or extrinsic correction (as is usually the case), the surgeon must also add 50 mm to axial frame offset under the mounting parameters for the extrinsic correction.

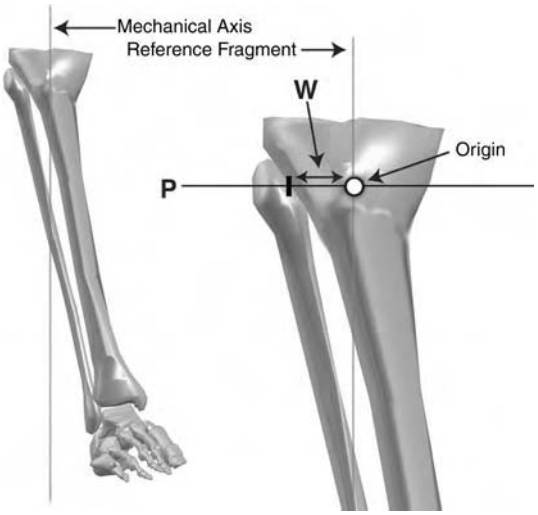


Figure 26 To analyze proximal tibial deformities, use either the level of the knee joint or more commonly the level of the proximal tibial fibula joint as the plane of the origin. The origin is located at the intersection of the plane of the origin and the mechanical axis of the proximal fragment. Create a line W from the origin to the proximal tibial fibula joint.

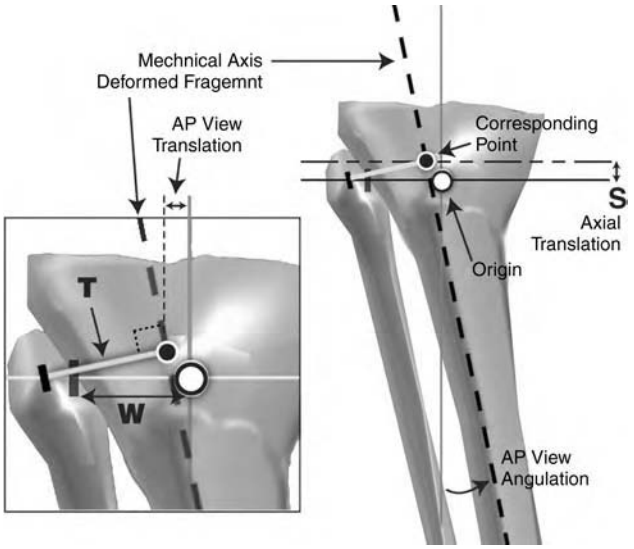


Figure 27 Draw the mechanical axis of the distal fragment. Create a line, T, equal in length to line W, with one end perpendicular to the mechanical axis of the deformed fragment and the other end resting on the plane of the origin. The point of intersection of line T with the mechanical axis of the deformed fragment is the corresponding point for the origin.

Performing an extrinsic correction is similar to performing an initial deformity correction followed by a residual deformity correction consisting of length only. The intrinsic origin is essentially a waypoint because the corresponding point moves to the extrinsic origin.

The following examples demonstrate typical choices for origin and corresponding point.

- Post-traumatic femoral shaft deformity: In this deformity, the bone ends are still discernable. The origin is established at the anatomic axis at the interior end of the reference fragment. The corresponding point is at the anatomic axis of the deformed fragment at its interior end. The points will be coincident when the deformity is corrected (Fig. 25).
- Remodeled or congenital femoral shaft deformity: In this deformity bone ends may not be discernable or new bone may be interposed. Set the level of the origin at the osteotomy site.

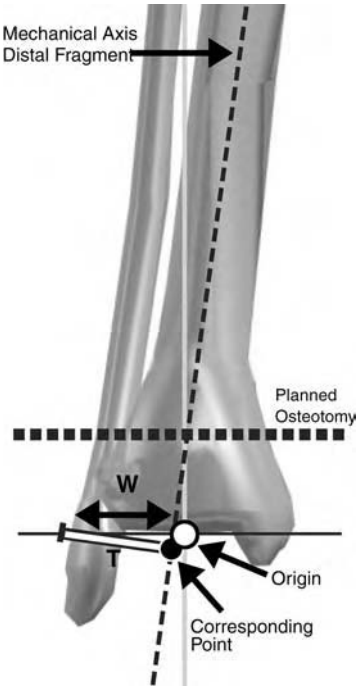


Figure 28 For distal tibial deformities, use either the level of osteotomy or the level of the joint as the plane of the origin. Use the same line W and line T construction to find the corresponding point.

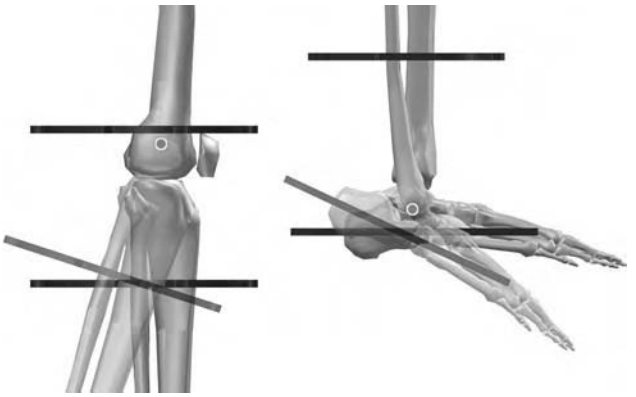


Figure 29 When correcting contractures of the knee or ankle, use the anatomic center of rotation at the origin and corresponding point. The contracture, therefore, is characterized as a Lateral View angulation only.

Local analysis is used to determine a level of corresponding point that will allow full correction of the angular deformity without creating impaction of the convex cortex.

- Proximal tibial varus: Set the level of the origin at the proximal tibia-fibula joint. The origin is the intersection of the mechanical axis of the proximal tibia with the plane passing through the tibia-fibula joint. The distance *W* from the mechanical axis to the tibia-fibula joint is measured and a second line T of length *W* is constructed so that one end lies on the proximal tibia-fibula joint plane and the other end rests on and is perpendicular to the mechanical axis of the distal fragment (Fig. 26). The corresponding point is the intersection of this constructed line segment with the mechanical axis of the distal fragment. This choice of origin and corresponding point will allow full correction of angular and translational deformity without changing fibular length. Fibular osteotomy is generally unnecessary unless there is significant malrotation (Fig. 27).
- Distal tibial varus: Set the level of the origin at the ankle joint. The origin is the intersection of the mechanical axis of the distal tibia at the ankle joint. The distance *W* from the mechanical axis to the tibia-fibula joint is measured and a second line of length *W* is constructed so that one end lies on the ankle joint plane and the other end rests on and is perpendicular to the mechanical axis of the proximal fragment. The corresponding point is the intersection of this constructed line segment with the mechanical axis of the proximal fragment. This choice of origin and corresponding point will allow full correction of angular and translational deformity without changing fibular length. Fibular osteotomy may only be necessary if there is significant distal fibular deformity and/or malrotation (Fig. 28).

When using the Spatial Frame to correct joint contracture (joint reduced), choose the anatomic center of joint rotation as the origin. By definition, the fragment centerlines intersect

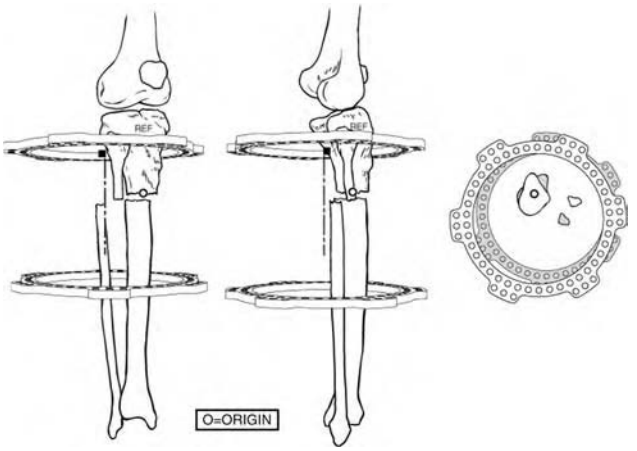


Figure 30 For rotational deformities, select the center of the interior end of the reference fragment to avoid concommitant translation as the moving fragment is rotated.

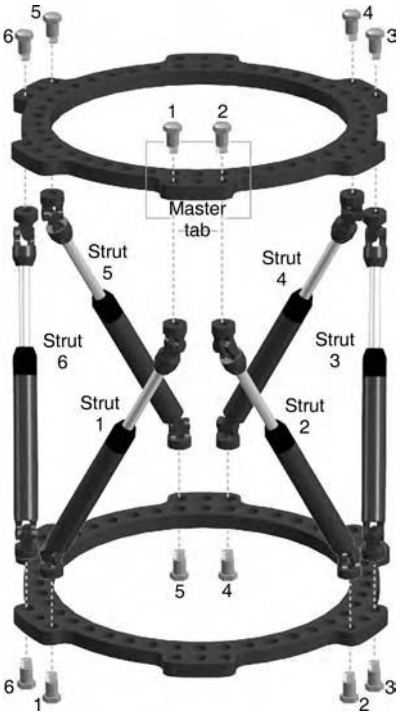


Figure 31 Each numbered/clip is applied to a strut beginning with strut 1 (which is attached to the designated Master Tab anteriorly) and progressing counterclockwise as viewed from the proximal end of the frame. The computer program assumes the universal joints connecting strut 1 and strut 2 to the proximal ring are aligned directly anterior with respect to the reference fragment. Different rotational alignments, especially for more proximal femoral and humeral applications, can be accommodated by changing rotary frame offset.

Figure 32 To begin, enter patient information, the side (left or right), and type of correction (long bone or foot).

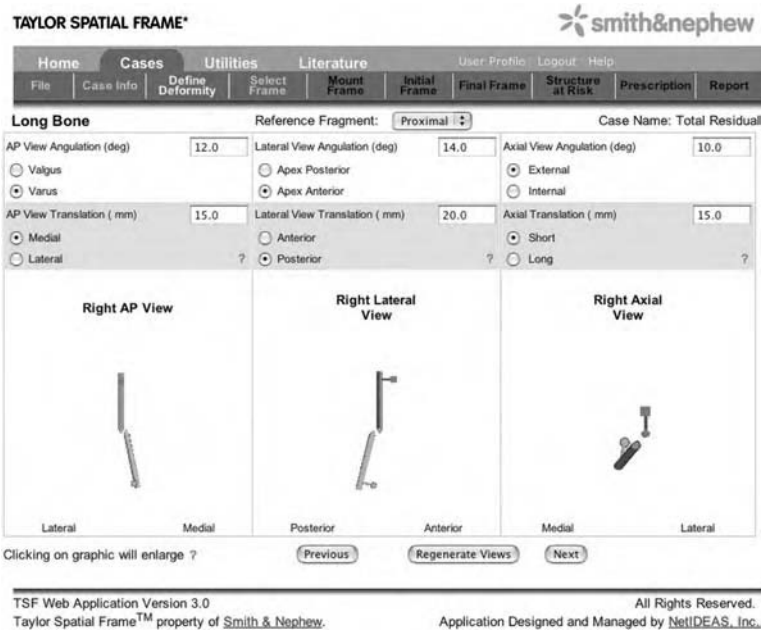


Figure 33 In the next window, select the proximal or distal reference fragment. Input the six skeletal deformity parameters. When regenerate views is then selected, the software provides the surgeon with updated AP, lateral, and axial views of the skeletal deformity. In this example, the pointed end of the top rod (with small cube attached) is the origin and the pointed end of the bottom rod (with small ball attached is the corresponding point). *Abbreviation*: AP, anteroposterior.

at this point; thus, there are no translations to measure. The deformity will consist only of rotational deformities (Fig. 29).

In deformities with significant malrotation, it is especially important to have the origin at the center of the reference fragment at the level of the interior end of the reference fragment (Fig. 30). In these cases, no additional translation is created by rotation about this center.

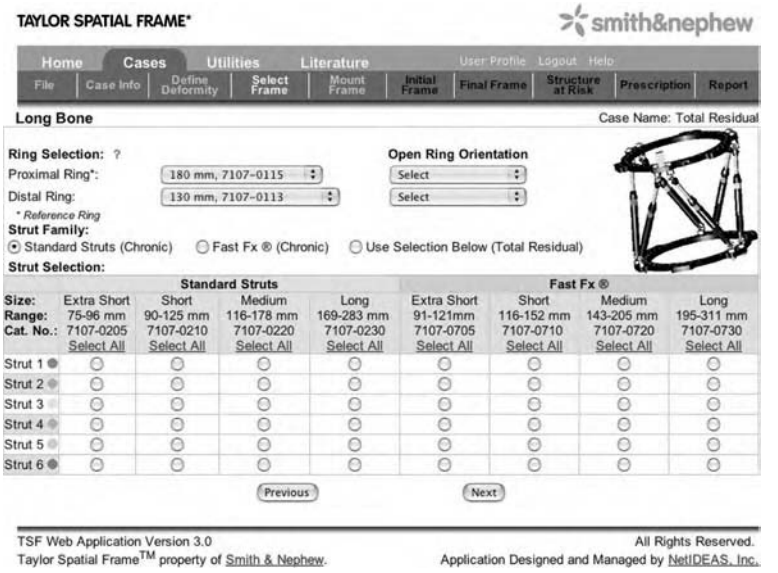


Figure 34 Using the pull down menu in the next window, select the proximal ring diameter, distal ring diameter, and the strut body type. Different sized rings may be used to create a tapered frame.

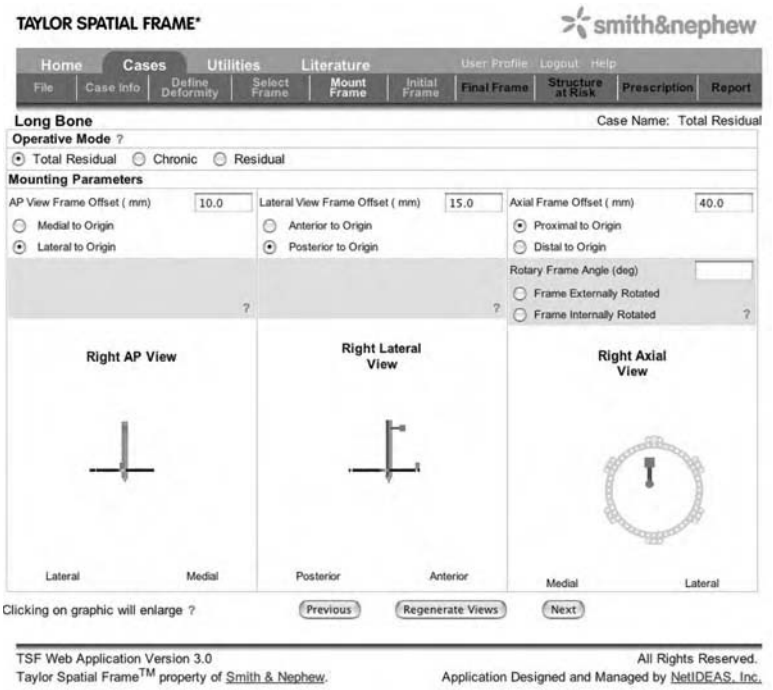


Figure 35 Next, select the operative mode, which in this case is the total residual button. The position of the reference ring with respect to the origin is input into the mounting parameters. The regenerated views confirm the relation of the reference ring on the reference fragment.

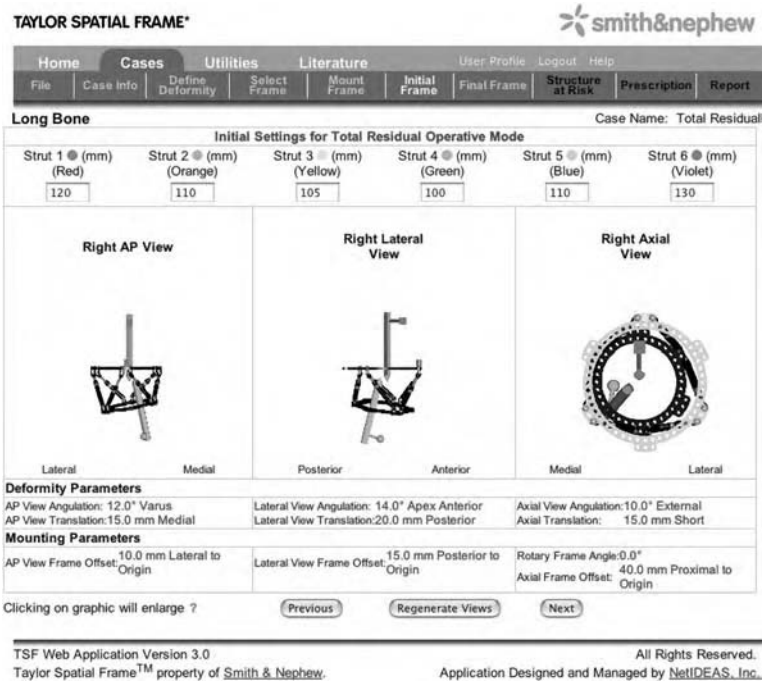


Figure 36 Each of the six strut lengths is entered in the next window. The computer then provides a graphical representation of the initial crooked frame on crooked bone.

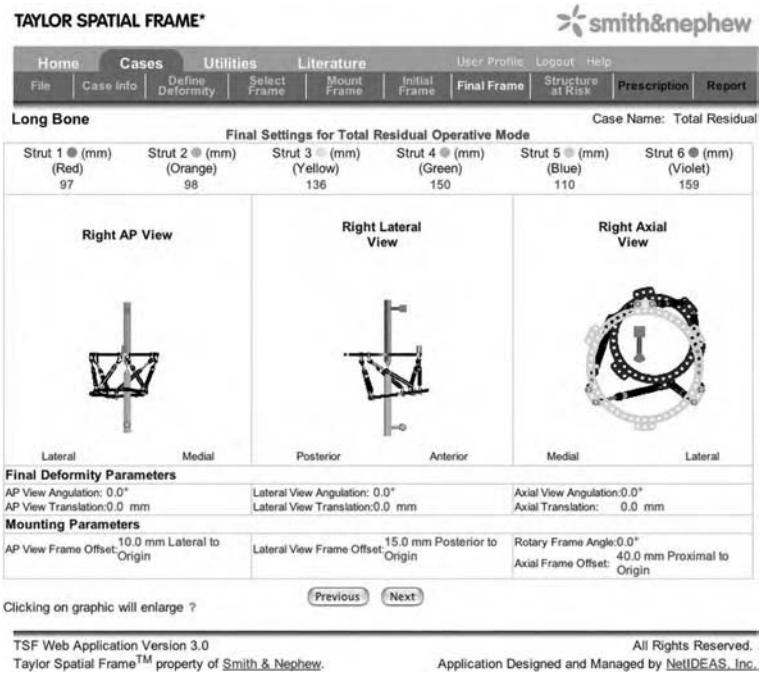


Figure 37 In the Final Frames window, the computer solves for the necessary strut lengths to correct the skeletal deformity and presents it graphically.

Thus, any translation measured at the level of the origin will be fully corrected and no additional translation will be created by correction of the malrotation.

FRAME ORIENTATION AND REVIEW OF COMPONENTS

The Taylor Spatial Frame fixator consists of two rings or partial rings connected by six telescopic struts at special universal joints. Telescopic struts are available in short, medium, and long sizes (Fig. 31). For a given size, the strut has a specific range from its shortest to longest length and a mid-position marked on each strut.

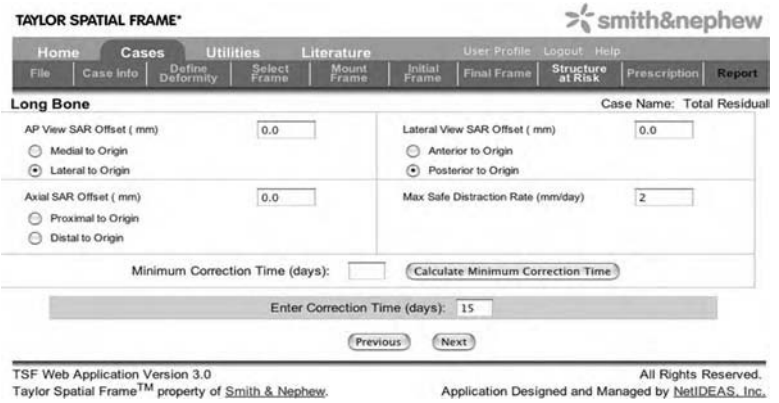
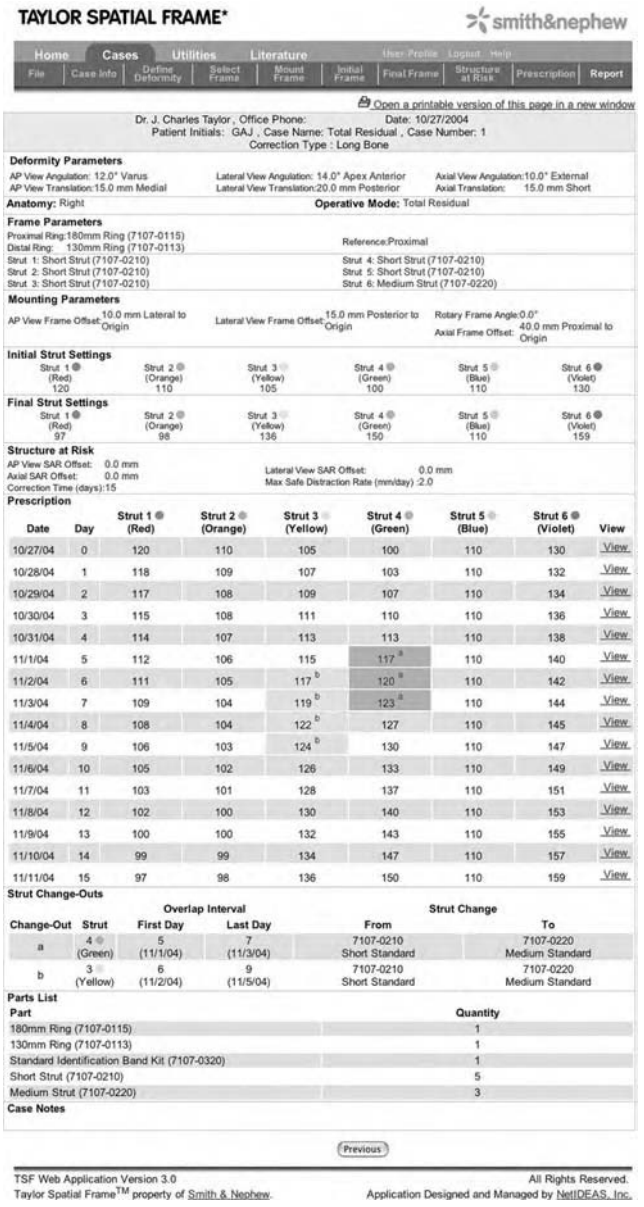


Figure 38 In the next window, the coordinates of the structure at risk and the maximum safe velocity are entered. The computer determines the number of days required for the correction. The surgeon may also override the number of days.



TAYLOR SPATIAL FRAME*

smith&nephew

Home | Cases | Utilities | Literature — User Profile Logout Help

File | Case Info | Define Deformity | Select Frame | Mount Frame | Initial Frame | Final Frame | Structure at Risk | Prescription | Report

Open a printable version of this page in a new window

Dr. J. Charles Taylor , Office Phone: Date: 10/27/2004
Patient Initials: GAJ , Case Name: Total Residual , Case Number: 1
Correction Type : Long Bone

Deformity Parameters

AP View Angulation: 12.0° Varus — Lateral View Angulation: 14.0° Apex Anterior — Axial View Angulation:10.0° External
AP View Translation:15.0 mm Medial — Lateral View Translation:20.0 mm Posterior — Axial Translation: 15.0 mm Short

Anatomy: Right **Operative Mode:** Total Residual

Frame Parameters

Proximal Ring:180mm Ring (7107-0115) Reference:Proximal
Distal Ring: 130mm Ring (7107-0113)
Strut 1: Short Strut (7107-0210) Strut 4: Short Strut (7107-0210)
Strut 2: Short Strut (7107-0210) Strut 5: Short Strut (7107-0210)
Strut 3: Short Strut (7107-0210) Strut 6: Medium Strut (7107-0220)

Mounting Parameters

AP View Frame Offset: 10.0 mm Lateral to Origin — Lateral View Frame Offset: 15.0 mm Posterior to Origin — Rotary Frame Angle:0.0° — Axial Frame Offset: 40.0 mm Proximal to Origin

Initial Strut Settings

Strut 1 (Red)	Strut 2 (Orange)	Strut 3 (Yellow)	Strut 4 (Green)	Strut 5 (Blue)	Strut 6 (Violet)
120	110	105	100	110	130

Final Strut Settings

Strut 1 (Red)	Strut 2 (Orange)	Strut 3 (Yellow)	Strut 4 (Green)	Strut 5 (Blue)	Strut 6 (Violet)
97	98	136	150	110	159

Structure at Risk

AP View SAR Offset: 0.0 mm — Lateral View SAR Offset: 0.0 mm
Axial SAR Offset: 0.0 mm — Max Safe Distraction Rate (mm/day) :2.0
Correction Time (days):15

Prescription

Date	Day	Strut 1 (Red)	Strut 2 (Orange)	Strut 3 (Yellow)	Strut 4 (Green)	Strut 5 (Blue)	Strut 6 (Violet)	View
10/27/04	0	120	110	105	100	110	130	View
10/28/04	1	118	109	107	103	110	132	View
10/29/04	2	117	108	109	107	110	134	View
10/30/04	3	115	108	111	110	110	136	View
10/31/04	4	114	107	113	113	110	138	View
11/1/04	5	112	106	115	117 [a]	110	140	View
11/2/04	6	111	105	117 [b]	120 [a]	110	142	View
11/3/04	7	109	104	119 [b]	123 [a]	110	144	View
11/4/04	8	108	104	122 [b]	127	110	145	View
11/5/04	9	106	103	124 [b]	130	110	147	View
11/6/04	10	105	102	126	133	110	149	View
11/7/04	11	103	101	128	137	110	151	View
11/8/04	12	102	100	130	140	110	153	View
11/9/04	13	100	100	132	143	110	155	View
11/10/04	14	99	99	134	147	110	157	View
11/11/04	15	97	98	136	150	110	159	View

Strut Change-Outs

Change-Out	Strut	Overlap Interval: First Day	Overlap Interval: Last Day	Strut Change: From	Strut Change: To
a	4 (Green)	5 (11/1/04)	7 (11/3/04)	7107-0210 Short Standard	7107-0220 Medium Standard
b	3 (Yellow)	6 (11/2/04)	9 (11/5/04)	7107-0210 Short Standard	7107-0220 Medium Standard

Parts List

Part	Quantity
180mm Ring (7107-0115)	1
130mm Ring (7107-0113)	1
Standard Identification Band Kit (7107-0320)	1
Short Strut (7107-0210)	5
Medium Strut (7107-0220)	3

Case Notes

Previous

TSF Web Application Version 3.0 — All Rights Reserved.
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Figure 39 In the report, window physician and patient information is reiterated. The mounting parameters, initial and final strut settings, the coordinates of the structure at risk, as well as the safe velocity are shown in table form. The daily adjustment schedule is also provided. Critical days when struts need to be swapped out are highlighted and color coded and identified with a letter. The strut exchanges are listed and explained in chronological order, and a list of necessary Spatial Frame parts is also given.

Six identifier clips, uniquely colored and numbered 1 through 6, are provided with each frame. Each numbered/colored clip is applied to a strut beginning with strut 1 (which is attached to the designated Master Tab anteriorly) and progressing counterclockwise as viewed from the proximal end of the frame.

The computer program assumes the universal joints connecting strut 1 and strut 2 to the proximal ring are aligned directly anterior with respect to the reference fragment. Different rotational alignments, especially for more proximal femoral and humeral applications, can be accommodated by changing rotary frame offset.

METHOD

Figures 32–40 illustrate the current web-based computer program for preparing an adjustment schedule for either the Chronic or Total Residual Method. Patient and file information as well

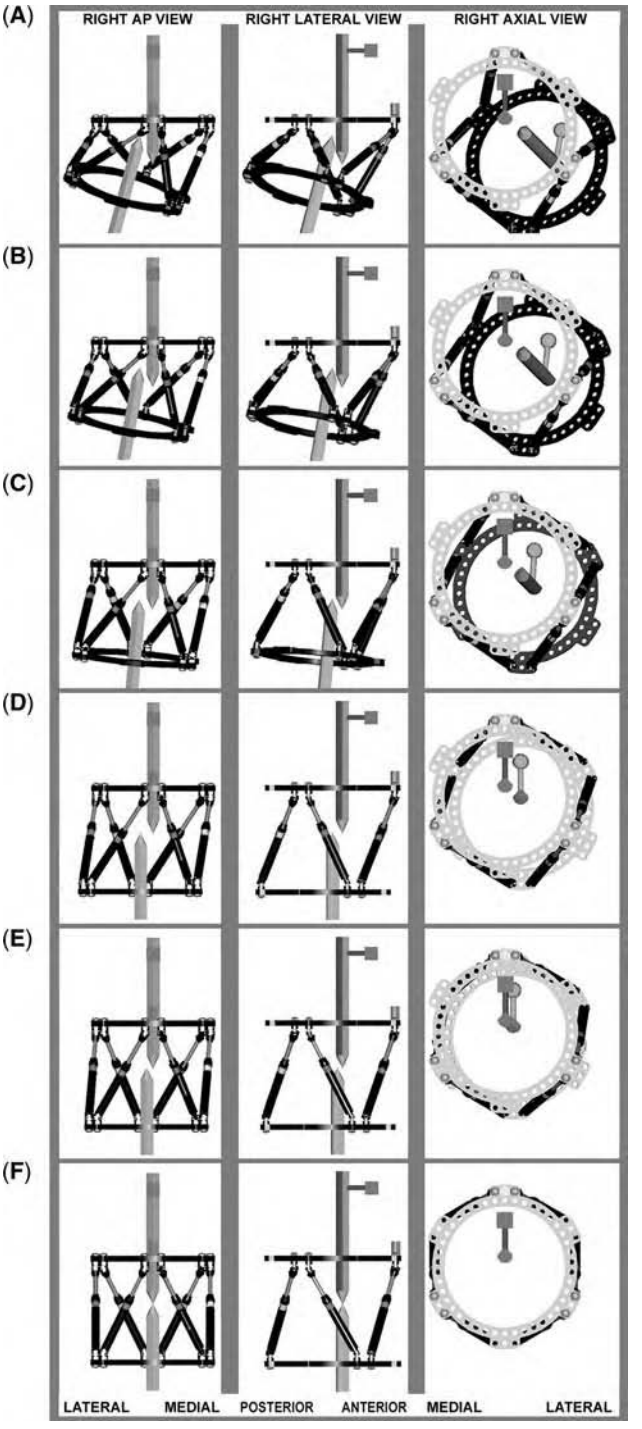

Figure 40 The Total Residual Correction Program can be used to prepare a daily schedule to correct a deformity via waypoints. In this case, correcting angulation, rotation, and length in the first step (rows **A-B-C**), and correcting translation in a second step (rows **D-E-F**).

as specific case notes can be saved to a secure web-based server. Saved cases can be reopened and modified, then saved again as a different case or scenario.

CLINICAL EXPERIENCE

Results with Spatial Frame compare favorably to the results with other external fixation, intramedullary nails, and plating of tibial fractures. During its development, the Spatial system was

thoroughly tested for theoretical accuracy of the computer program, combined mechanical accuracy in correcting severe six-axis deformities, and frame stiffness. The computer program is mathematically accurate to within a millionth of an inch and a ten thousandth of a degree. The combined mechanical accuracy of the program as applied to the frame by hand is 1 mm translation and 0.6° angulation. The Spatial Frame is as stiff as the Ilizarov in axial loading and twice as stiff in bending and torsion.

The Spatial Frame has been used since approximately 1996. Although initially reported for fractures and deformity correction (1), its early acceptance was for deformity correction and nonunions (2–7) and has gradually gained acceptance for primary treatment of fractures (4,8–13). Historically, malunion was the leading complication of external fixation reflecting the difficulty in making adjustments to other frames after the initial surgery and the reluctance to return to surgery. Not only does repeat surgery pose a risk to the patient, but there is also the real possibility that in an effort to improve one aspect of fracture reduction, another could be lost.

The ability to make prescribed gradual adjustments to the Spatial Frame in the postoperative period is the greatest strength of the system. The program provides a daily adjustment schedule for the struts, which are easy to adjust using the direct read indicator for each strut. MacFadyen and Atkins (9) demonstrated a 100% compliance of their patients adjusting the frame as prescribed. Near anatomic realignment and repositioning of the externally fixed major fragments has been routinely achieved. Binski and Hutchinson (8), MacFadyen and Atkins (9), and Whately (13) report a 95% to 100% rate of anatomic or near anatomic restoration of alignment and position. Likewise, these same authors report a 96% to 100% rate of primary union and remarkably low rates of reoperation, approximately 5%, for delayed union.

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46 Internal Lengthening Nail: Intramedullary Skeletal Kinetic Distractor

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INTRODUCTION

The recognition of the ability of bone to regenerate has stimulated the development of different procedures to lengthen or restore bone loss. External fixators, hybrid systems such as lengthening over an intramedullary (IM) nail, and lately, fully implantable lengthening devices, have been designed in an attempt to regain bone length.

HISTORICAL BACKGROUND

Ilizarov Technique

The pioneering work of Ilizarov (1,2) concerning biomechanical factors that affect osteogenesis has had a significant influence on the techniques currently used to perform bone lengthening and bone transport. Ilizarov's findings emphasized the importance of limited soft-tissue trauma, the significance of gradual distraction and preservation of IM circulation. The first two concepts are still accepted as pillars for both lengthening and bone regeneration. Violation of IM circulation was once widely accepted as having a negative effect on bone healing because it affected the degree of avascularity and revascularization of the cortex. In 1988, Kojimoto et al. (3) reevaluated the role of periosteum and endosteum on bone lengthening by callus distraction in rabbits. The results suggested that for bone lengthening by callus distraction to succeed, preservation of periosteum is essential. Likewise, periosteum is more important than careful corticotomy and preservation of the endosteal circulation.

Biological principles of the ''tension stress effect'' that Ilizarov described are generally accepted and should be followed during bone lengthening. His work added a great deal to the understanding of bone and tissue regeneration under ''tension stress.'' In many instances, external fixation is difficult for the patient and is associated with pin tract infections and tethering of the soft tissues at the pin sites. Some of the problems during lengthening that are directly associated with external fixation may be avoided with an internal lengthening device.

Lengthening-Over-Nail Technique

Combinations of internal/external fixation and distraction techniques have been reported. Bost and Larsen (4) first documented lengthening of an extremity over an IM rod in 1956. Lengthening-over-nail (LON) technique was developed to simplify the fixator construct and to minimize the amount of time that the external fixator was applied. The ultimate goals were to improve patient acceptance and decrease the complications associated with the traditional method, such as pin tract infection, scarring, and refracture. In the LON technique, the external fixator provides the ''tension stress effect'' necessary for osteogenesis to occur. The locked IM nail performs well as a stabilization device to guide the bone during lengthening or bone transport and provides definitive fixation at the completion of the lengthening or transport process, when locking is performed, allowing the early removal of the external device.

During the development of the internal lengthening nail, we evaluated lengthening and fixation methods that would provide a low profile and decrease the duration of the external fixation. From March 1988 through June 1992, 25 consecutive adult patients in our practice were treated for leg length discrepancies and segmental bone defects (5). Fourteen patients underwent lengthening and 11 underwent bone transport with the LON technique. Two of the lengthening patients and eight patients of the bone transport group had been treated

for osteomyelitis. Seven patients had nonunion and two had delayed union. Eight patients had previously sustained fractures with consequent bone loss. Seven of the patients suffered malunions and one patient had sustained a physeal injury as a child. The average length gained in the lengthening group was 4.3 cm while in the bone transport group, the average bone gain was 6.6 cm. The mean period of external fixation for lengthening was 20.2 days/cm of length gained and 23.6 days/cm of bone transported. Two patients in the lengthening group developed pin tract infections, while one patient responded to oral antibiotics and the other required an external fixator pin exchange. Three patients in the bone transport group developed pin tract infection, two patients responded to antibiotics, and the third patient required pin exchange. Two patients exhibited exacerbation of preexisting osteomyelitis; one of these responded to intravenous antibiotics while the second one required multiple surgical procedures including removal of distal screws, irrigation and debridement, antibiotic bead placement, and intravenous antibiotics. A 65-year-old patient developed delayed formation of regenerate. Ten of the 11 bone transport patients (91%) experienced complications related to docking of the transport fragment, and eight of these required bone graft at the nonunion of the docking site. Hardware failure occurred in one patient who experienced a breakage of the distal locking screw of the IM nail. One patient developed flexor hallucis longus and equinus contractures, ultimately requiring tendon releases.

Our experience with LON technique was not free of complications. Pin tract infection was still a frequent problem even though the external fixator time was significantly reduced through this technique compared with the traditional Ilizarov method. Infection proved to be one of the constant problems with the LON technique. Acute deep infection and exacerbation of latent osteomyelitis are two serious possibilities. The association of an external fixator with an IM nail in a patient with history of an open fracture or osteomyelitis is a major risk factor for development of deep infection.

Paley et al. (6) reported a matched-case comparison between femoral lengthening over IM nail with Ilizarov lengthening. Between March 1990 and November 1993, 32 femoral lengthenings (29 patients) were performed using the LON technique and 32 femoral lengthenings (31 patients) with the Ilizarov method. In this report, the time needed for radiographic consolidation was reduced significantly ($p < 0.001$) and the range of motion of the knee returned to normal a mean of 2.2 times faster in the LON as compared to the Ilizarov group. The rate of complications in the LON patients was 1.4 (one pin tract infection and one late infection) compared with 1.9 in the Ilizarov group (two deep pin tract infections).

Simpson (7) in 1999 reported on 20 bones lengthened by the LON technique. They did not encounter refracture or malalignment. There were three cases of deep infection, (two of which occurred in patients who had had previous open fractures of the bone, which was being lengthened); all were resolved with appropriate treatment. The conclusions drawn were that the method allows early rehabilitation, with a rapid return of knee movement. There is a lower rate of complications and, the time of external fixation is shorter than in other methods of leg lengthening. The high risk of infection calls for caution (three cases of deep infection—15% infection rate).

Silberg et al. (8) compared 10 patients who underwent femoral lengthening using the Ilizarov technique versus 10 patients with the LON technique and a monolateral external fixator. The infection rate was 5%. Kocaoglu et al. (9) in a recent report found complications in 16 (38%) of 42 bones (35 femora and 7 tibiae) in 35 patients who underwent lengthening over IM nail technique. The complications included premature consolidation, pin tract infection (one grade-2 and one grade-3), poor formation of regenerate bone (two patients), interlocking screw problems, Schanz screw cut-out, perioperative fracture, equinus contractures and nail impingement, and angulation at the osteotomy level. The complication rate was significantly higher if lengthening was greater than 21.5% of the original length or if more than 6 cm of total length was sought. The deep infection rate was 2.4% (one patient). Song et al. (10) reviewed the outcome in 22 patients who underwent femoral lengthening over IM nail. The complications included three patients with osteomyelitis (history of infection or open fracture), who required removal of the external fixator. Four patients developed knee complications (lengthening over 20% of the original bone length). In one patient, the lengthened segment collapsed. The conclusion drawn was that caution is required to prevent major complications when lengthening over an IM nail.

The current indications for LON in our practice include patients who need humeral lengthening, patients whose tibia or femur is too short, or whose canal is too narrow to allow

placement of the IM lengthener and patients who have current joint instability at the knee and ankle. This also pertains to those select patients in whom the distraction rate has to be strictly and precisely controlled.

Internal Bone Lengtheners

The patient's problems are the surgeon's source of inspiration. The concept of an implantable lengthening nail was developed in response to (i) the complications expected of the external fixator and its interference with the patient's lifestyle and daily activities, (ii) the opportunity to achieve restoration of limb length without pin site infections, (iii) the risk of chronic osteomyelitis and (iv) the sequelae from pins penetrating through soft tissue and muscle.

The early development of a fully implantable lengthening nail was plagued by multiple technical difficulties. The initial designs included an IM nail that was connected to an external component, increasing the risk of infection. The early experience of Golz and Schellmann (11) was reported in 1975. They used an IM nail that contained a hydraulic pressure system. Baumann and Harms (12) in 1977 published results of an animal study using a lengthening nail that contained a spindle mechanism. The first fully implantable distraction device was designed in 1977 by Witt and Jäger (13). It included an electronically controlled distractor that was regulated by transcutaneous radio control. It, too, had technical problems and to our knowledge, was not used further.

In 1997, the early results of a motorized IM nail for limb lengthening and bone transport (Fitbone) in 12 patients were reported by Baumgart et al. (14). The device uses a fully implantable, motorized programmable sliding mechanism. A subcutaneous reception antenna is connected to the motor in the proximal part of the nail. In 2002, Garcia-Cimbrelo et al. (15) analyzed the results of 24 femoral lengthenings and in 2003, Guichet et al. (16) reported that 32 patients underwent lengthening using the Albizzia Gradual Elongation Nail. The device has two telescoping cylinders connected by a ratchet system and lengthens by rotation movements of the limb segment. The Bliskunov nail was developed in Ukraine by Alexander Bliskunov. The device has two parts: the IM nail and a telescoping tube (the clicker). The telescoping tube is attached to the pelvis and to the nail. Lengthening occurs by rotatory motion between the nail and pelvic portions. Once the goal length has been reached, the pelvic portion is removed and the nail maintains alignment while the bone heals. The device is currently being used by Dr. Vladimir Dragan in Kiev.

INTRAMEDULLARY SKELETAL KINETIC DISTRACTOR

The Intramedullary Skeletal Kinetic Distractor (ISKD^{TM}, Orthofix®, McKinney, Texas, U.S.) (17) is a fully implantable lengthening nail designed to lengthen under physiological movement. The basic internal mechanism is shown in Figure 1. The ISKD lengthens as small oscillations between two telescoping sections are mechanically converted to one-way distraction. Because the patient rotationally oscillates the limb either manually or during walking, the device gradually distracts. Because the device is designed to lengthen under rotational displacement as small as 3°, nonphysiologic movement is not required to achieve distraction. However, rotational oscillations as large as nine degrees are allowed if greater rotation and fewer oscillations are desired. Thus, the rate of linear distraction depends on the frequency and intensity with which the patient oscillates the limb. The lengthening device is not as efficient when it is under load; hence the number of rotations to achieve length increase. The rate of distraction is monitored using an external hand-held sensing device or monitor.

Intramedullary Skeletal Kinetic Distractor Design

All the components of the ISKD have been manufactured from titanium alloy (Ti6A14V) in order to maximize strength and make the device biocompatible. The three main components of the ISKD are the telescoping sections, the drive mechanism, and the length indication feedback mechanism (Fig. 2). The proximal and distal sections are the only two components that have exposed surfaces. These sections have screw holes arranged in a pattern allowing for the device to be attached to the bone segments. The drive mechanism consists of two one-way roller clutches and a threaded rod. It is designed to convert rotary oscillations of the proximal and distal sections relative to one another into linear distraction. The length indication

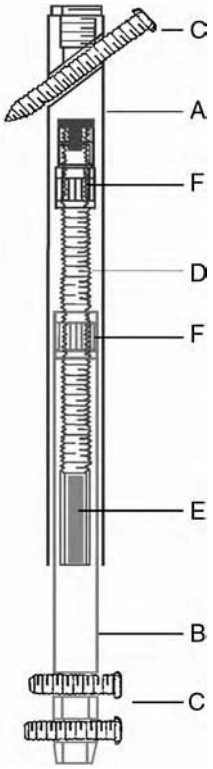

Figure 1 Basic internal mechanism. (**A**) proximal telescoping rod; (**B**) distal telescoping rod; (**C**) proximal and distal locking screws; (**D**) threaded rod; (**E**) magnet; (**F**) proximal and distal clutches. *Source*: Courtesy of Orthofix Inc.

mechanism consists of a magnet centered in the threaded rod. The rotation and relative position of the magnet can be detected by a hand-held external sensor and a minicomputer that calculates the amount of lengthening that has occurred based on frequent noninvasive measurements of the magnet position (Fig. 3).

Proximal and Distal Sections

The proximal section is the largest component of the ISKD. It is designed specifically for the bone to be lengthened. Consequently, the only difference between femoral and tibial ISKD is the design of the proximal section. The distal section telescopes inside the proximal section and has a diameter that is typically 15% smaller than that of the proximal section. Two parallel groves 180° apart are machined in the distal section and mate with a removable key ring that fits over the distal end of the proximal section. This keyed mechanism allows the distal section to slide within the proximal section with limited rotational displacement. The distal section and proximal section are matched in length but are capable of either 50 or 80 mm of distraction. Tibia lengtheners are available with proximal section diameters of 10.7 and 12.5 mm; femoral lengtheners in 12.5 mm. The 10.7 mm tibia design can be used in the femur using a greater trochanter entry or in the retrograde technique.

Drive Mechanism

The drive mechanism consists of a threaded rod, a threaded area within the distal section and two one-way roller clutches oriented in opposite directions to one another. The distal clutch is

Figure 2 Intramedullary Skeletal Kinetic Distractor sagittal view of the internal mechanism. *Source*: Courtesy of Orthofix Inc.

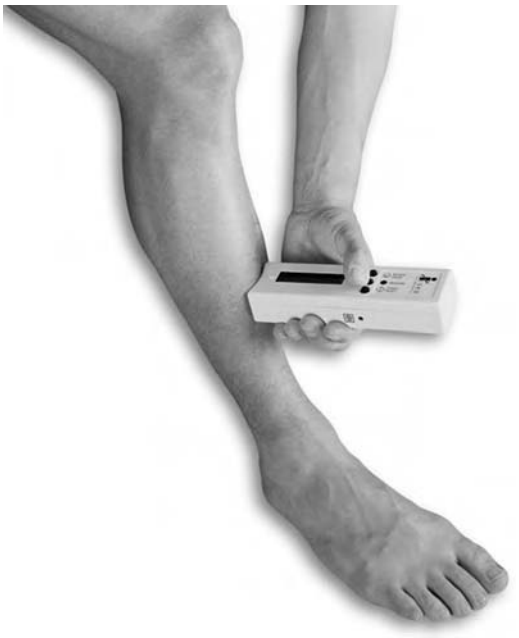

Figure 3 Measuring technique: the monitor should be in direct contact with the skin. *Source*: Courtesy of Orthofix Inc.

constrained by the movement of the distal section while the proximal clutch is constrained by the movement of the proximal section. As the distal section turns the distal clutch clockwise (with respect to the proximal section and proximal clutch), the threaded rod rotates with the clutch and ISKD does not distract. Because the distal clutch rotates counter-clockwise, the proximal clutch prevents the threaded rod from turning and the distal clutch rotates with respect to the threaded rod. Because the distal section rotates (with respect to the threaded rod), the low-friction female ACME threads in their threaded area traverse the threaded rod, forcing the proximal and distal sections to distract. Because the clutches only contact the perimeter of the threaded rod, compressive loads not involving rotation are distributed from the lower distal section to the upper proximal section without affecting the clutches; the clutches also act as bearings providing stability to the threaded rod.

Length Indication Feedback Mechanism

A natural magnet is encased within the tip of the threaded rod. The magnet is shaped in such a way that the north and south poles of the magnet are oriented perpendicular to the length of the nail. Because the threaded rod rotates during distraction, the magnet also rotates changing the orientation of the poles of the magnet relative to the vertical axis of the nail. An external magnetic sensing device is placed on the skin; it detects the strength of the magnetic field as well as the pole of the magnet that is facing the sensor (Fig. 4). A minicomputer within the sensing device records the time of measurement as well as the strength and position of the pole nearest to the sensor and compares that measurement with the last previous measurement. The computer then calculates the amount of rotation of the magnet in degrees and multiplies the angular change by the pitch of the threads of the threaded rod. Each pole change, 180° rotation of the threaded rod, equals 0.37 mm in length. The result of this calculation is the amount of distraction that has occurred since the last previous measurement. The result of each consecutive measurement is added to the sum of the previous measurements recorded by the device and is displayed for the patient on the LCD screen of the monitor. The patient adjusts his/her activity relative to the doctor's instructions concerning the rate of lengthening desired. The minicomputer within the sensor can download the full series of measurements to a PC to periodically permit analysis of the lengthening process and to check patient compliance. The device also has an alarm that tells the patient to take a measurement at regular intervals.

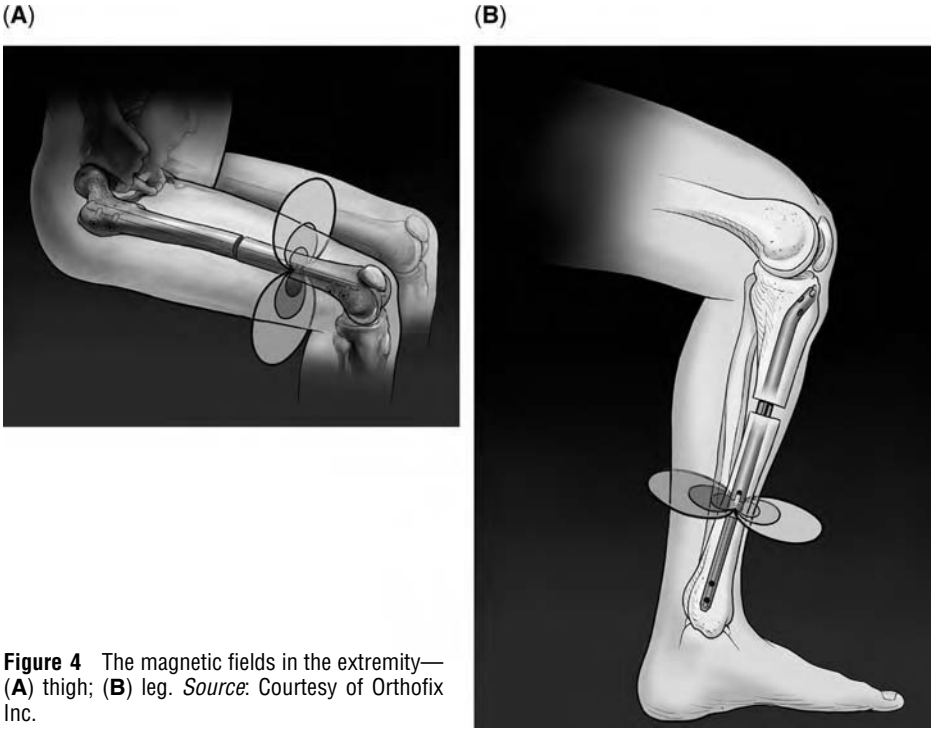

Figure 4 The magnetic fields in the extremity—(**A**) thigh; (**B**) leg. *Source*: Courtesy of Orthofix Inc.

Mechanical Evaluation

During the development of the ISKD, three types of tests were performed. A traditional four-point bend test demonstrated that the bending strength of the ISKD decreases significantly as the ISKD expands. Bending rigidity is an indication of an implant's inherent flexibility. Bending rigidity of the ISKD was found to decrease linearly as the ISKD expands. However, the rigidity of the 12 mm diameter ISKD remained higher than either the 8.0 mm commercially available Ti6A14V tibial nail or the solid 9.5 mm titanium alloy bar, and compared favorably with cannulated and slotted 316LVM implants tested by other authors.

A torsion-to-failure test revealed that the torsion values of 35 and 45 Nm required for torsional failure of the ISKD are within the range of values obtained by other researchers for conventional 11 mm diameter IM nails, which ranged from 15 to 147 Nm.

A test to measure performance of the nail during rotational oscillations under increasing compressive loads showed that the torque required to lengthen the ISKD increased proportionally as the applied compressive load increased. Due to the efficiency of the drive mechanism, even the highest loads of 1400 N (315 lb) required a relatively low torque of 2.2 Nm (19.5 in. lb) to distract the device.

CLINICAL RELEVANCE

Indications

As lengthening nails become a routine part of the armamentarium available to the orthopedic surgeon, strategies will most likely evolve to include their use in treatment of osteomyelitis, complex open fractures, and nonunions (Table 1). Bone resection surgery is at a stage of early development. Successful resection osteotomy technique, soft-tissue coverage schemes, and infection treatment will evolve to decrease complications and assure functional outcomes. Obviously, patients with osteomyelitis require additional treatment that may impact resection osteotomy and fixation methods depending on location of the infection.

In our practice, only a small percentage of the patients requiring lengthening have anatomically aligned extremities, the majority of patients have sequelae from osteomyelitis, open

Table 1 Intramedullary Skeletal Kinetic Distractor: Indications and Contraindications

Indications	Contraindications
Leg length discrepancy	Absolute
Congenital	Noncompliant patient
Developmental	Open physes
Fracture with segmental soft-tissue loss	Intramedullary (IM) canal diameter <9 mm
	IM canal length <215 mm
Osteomyelitis	Joint instability—knee subluxation
Nonunion	Significant contractures (bracing not an option)
Malunion with shortening	Pregnancy
Short stature	Relative (expect difficulties)
Congenital and social	Smoker
	Low pain tolerance
	Inability to follow up

Abbreviations: IM, Intramedullary nail.

fractures, nonunion, and deformities; therefore they require additional surgical procedures prior to lengthening. If the surgical plan includes using an ISKD, the different surgical steps need to be planned accordingly, (not to burn bridges) and the strategies directed at obtaining anatomical canal axes and an open IM canal. As a general rule, treatment of angular deformity, infection, and nonunion should be performed before attempting lengthening procedures.

Bone Resection

Bone resection should be done in a manner that reduces healing time by decreasing damage to the surrounding soft tissues and improves the torsion stability of the osteotomy fragments. An oblique osteotomy is not technically demanding and it provides inherent rotational stability along with an extended surface helpful for healing. The simple transverse osteotomy does not offer mechanical stability when compared to the oblique osteotomy; it is easy to perform, but it lacks rotational control even if fixed with a locked IM nail. In general, an oblique resection osteotomy should be used when possible, especially in the diaphysis; perhaps a guide system to optimize matched cuts will add to the stability of the fragments.

The following list includes the specific temporary fixation methods, by level of resection, recommended for the tibia.

1. Proximal epiphysis (intraarticular): temporary tibiofemoral rod (Fig. 5); temporary external fixation technique during soft-tissue coverage or infection treatment
2. Proximal metaphysis: posteromedial or posterolateral plate; locked IM nail and blocking screws; external fixation
3. Diaphysis: IM nail; antibiotic-impregnated cement nail (Fig. 6)
4. Distal metaphysis: screw fixation; external fixator; locked IM nail
5. Distal epiphysis (intraarticular): tibiotalar fusion nail (Fig. 7); external fixator

Soft-Tissue Coverage

Soft-tissue coverage schemes, perhaps, are the most challenging aspect of bone resection to the orthopedic surgeon. Intuitively bone loss with redundant soft tissues would allow easy coverage of the shortened limb segment; however ''puckering'' of the indurated traumatized soft tissue will make closure without a flap impossible in most situations. A z-incision (18) may be appropriate in the pretibial area when a healthy soft-tissue sleeve is present, but usually a flap or multiple flaps are required. In general, fasciocutaneous flaps are the workhorses of soft-tissue coverage of resection osteotomies. Planning must be performed at the time of debridement to assure that the best coverage opportunity is available. The incision for debridement will later become a border for the fasciocutaneous flap. Flaps must be based either medial or lateral over the septocutaneous vessels; the anterior pretibial area is not an acceptable base. Large flaps are possible either by staging or by increasing the base width. Debridement, therefore, should be performed in a manner that limits flap trauma and

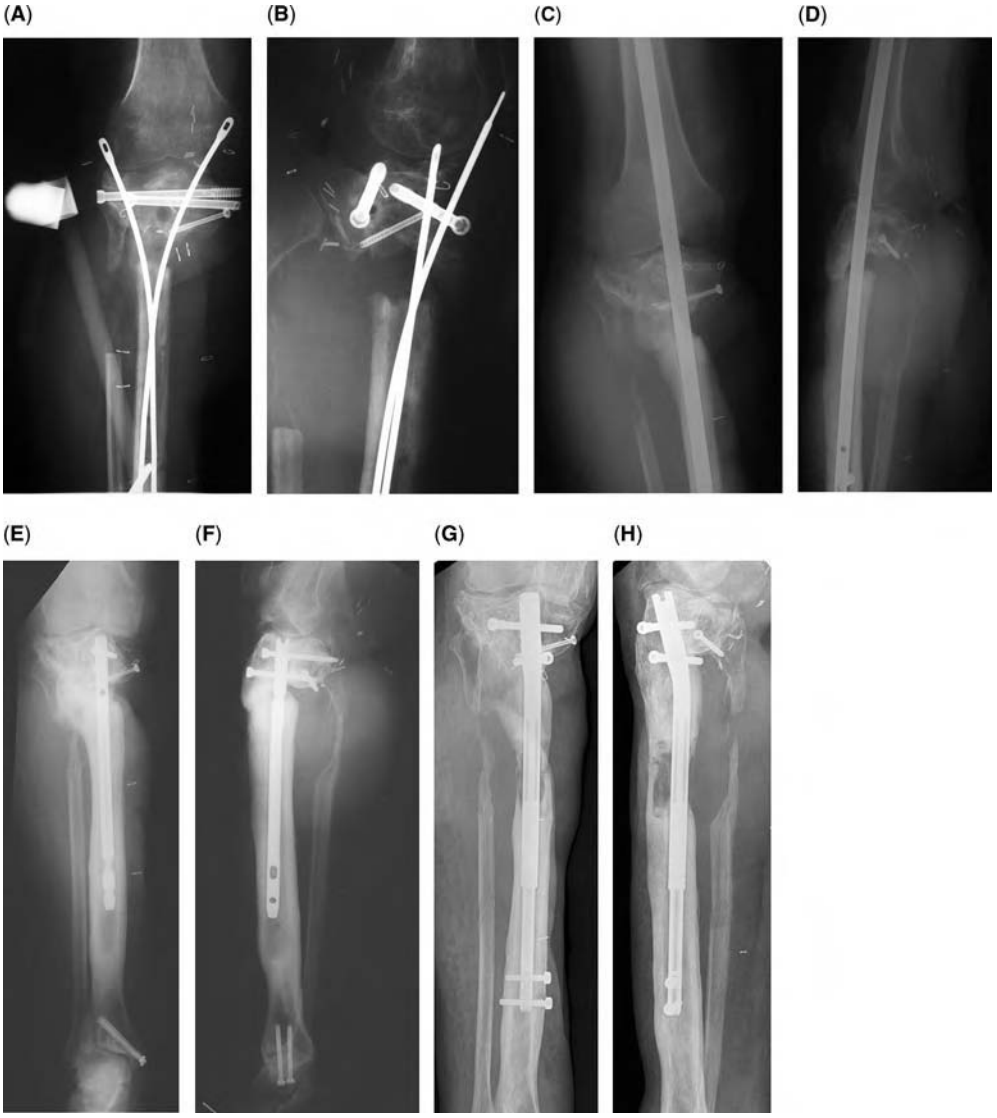

Figure 5 Proximal tibia periarticular fracture. (**A** and **B**) Radiographs of an open intraarticular proximal tibia fracture. (**C** and **D**) Temporary tibiofemoral fixation, provides stability to allow healing of proximal tibia fracture. (**E** and **F**) Tibia intramedullary nail to allow knee movement while bone healing progresses. (**G** and **H**) Successful lengthening of the tibia with Intramedullary Skeletal Kinetic Distractor.

ischemia. Flap completion prior to bone resection allows retraction of the flap to accomplish exposure without undue tension on the flap.

Osteomyelitis Treatment

The successful treatment of osteomyelitis with segmental resection is reliant on bone resection of necrotic cortices and canal contents. Choice of resection osteotomy location is therefore the primary concern. Surgical observation, MRI, and computed tomography scanning should lead to success. Use of cooled saw cut with a thin blade, decreasing thermal necrosis at the site, is very important. Canal debridement with careful reaming and brushing and fluid flow will be of benefit. Establishing a distal vent site theoretically will decrease the risk of leaving canal debris in the distal segment. Use of antibiotic-impregnated nail to fill the dead space and deliver high concentration of antibiotics in the local environment should decrease the risk of infection (19). Tensionless meticulous closure over the site with an appropriate flap is the

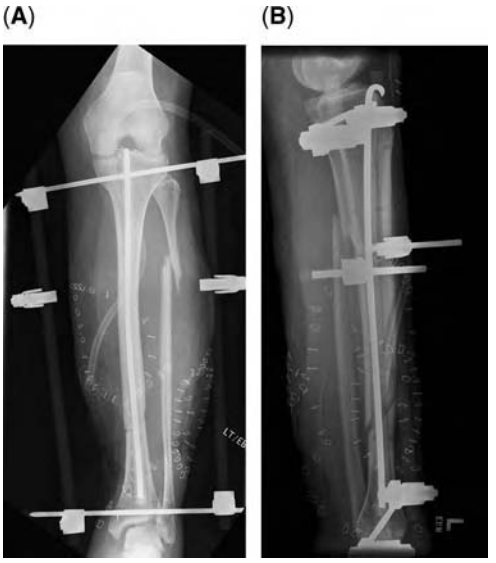

Figure 6 Antibiotic-impregnated cement rod for initial treatment of infected open tibia fracture.

key to success especially in today's hospital environment. Secondary contamination of exposed bone, even after debridement, is a concern that demands early sealing of the wound to decrease its risk.

Soft-Tissue Balancing

Another often-overlooked problem facing the surgeon treating patients with bone resection techniques is the soft-tissue balance of the limb. If an imbalance resulting from paresis, contraction or fibrosis is present in a shortened limb, it will most likely worsen with lengthening. Rates of distraction needed to avoid premature consolidation do result in increased soft-tissue tension and muscle fibrosis. Despite stretching exercises and bracing, contractures will increase during lengthening. Adequate joint mobility prior to lengthening should be assessed carefully. Coronal and sagittal plane motion of the hip joint, quadriceps, and hamstring mobility about the knee are important for femoral lengthening. Tibia lengthening requires careful assessment of coronal motion in the subtalar joint and sagittal plane balance in the ankle and knee joint.

Even forefoot contractures should be assessed and corrected prior to lengthening. Tendon transfers about the leg are necessary to balance motion lost by paresis; peroneal tendon or posterior tibial tendon transfers with heel cord lengthening or gastrocnemius recession are common.

Tendon transfers and joint contracture release with therapy preceding lengthening is necessary to avoid contractures. Calcaneal osteotomy to address a contracted varus heel after severe limb trauma is often necessary. If the foot does not remain plantigrade or the knee joint does not fully extend during gait with the lengthening procedure, a protracted complicated course will ensue. Less predictable outcome and long therapy requirements will result. Arthrofibrosis and destructive arthritis are possible if soft-tissue balancing and joint mobility issues are not given appropriate attention prior to lengthening.

A major disadvantage of the ISKD method compared to an external fixator is the lack of adjustability during lengthening. The rate of distraction as well as the inability to stop during distraction places a larger burden of appropriate preoperative planning on the surgeon. However, with careful patient selection limiting distraction limits and prelengthening soft-tissue balancing, successful outcomes are expected.

INTRAMEDULLARY SKELETAL KINETIC DISTRACTOR IMPLANTATION

Preoperative Planning

In patients with leg length discrepancy, determine the goal length using a reliable method as described by Paley (20). Candidates for cosmetic or stature lengthening must be evaluated for

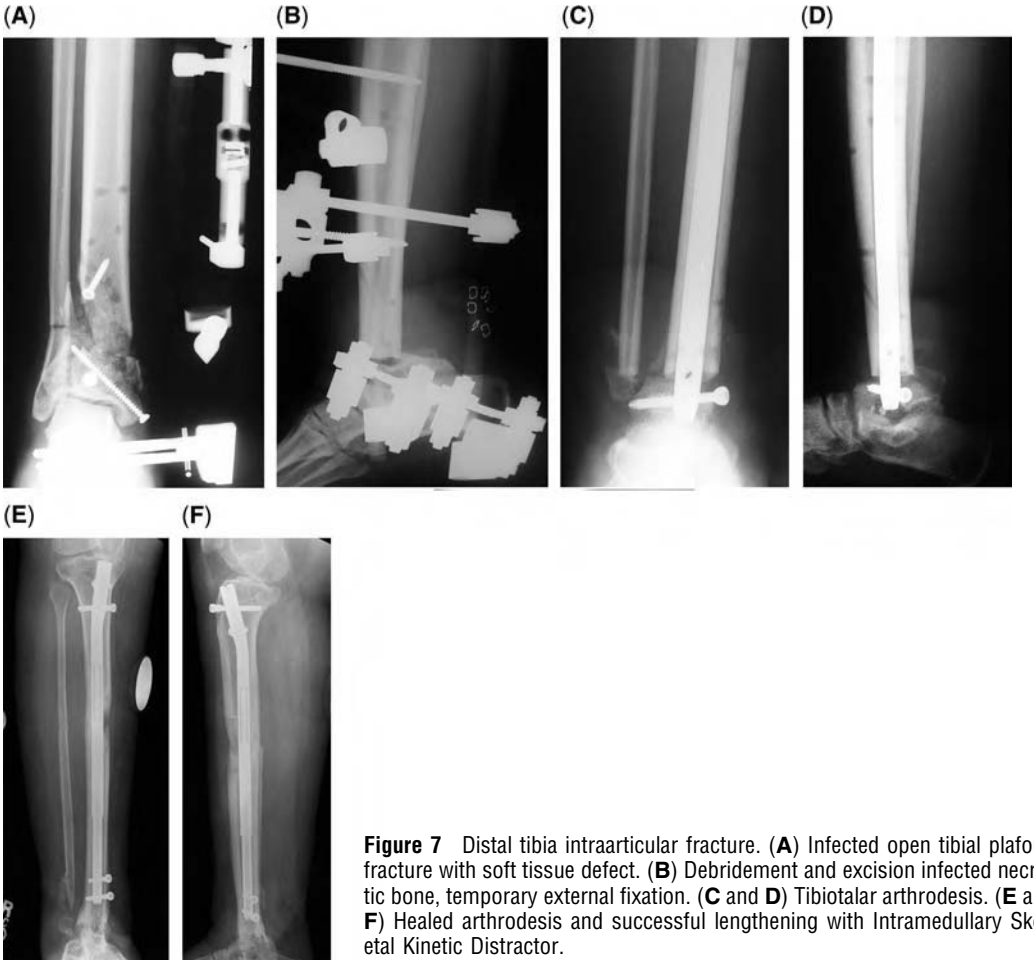

Figure 7 Distal tibia intraarticular fracture. (**A**) Infected open tibial plafond fracture with soft tissue defect. (**B**) Debridement and excision infected necrotic bone, temporary external fixation. (**C** and **D**) Tibiotalar arthrodesis. (**E** and **F**) Healed arthrodesis and successful lengthening with Intramedullary Skeletal Kinetic Distractor.

unapparent discrepancies, for determining which bones will be lengthened first and the goal length. To date, the smallest nail diameter available is limited to 10.7 mm; therefore, the IM canal must tolerate reaming to 12.5 mm. The ISKD is available in lengths of 50 and 80 mm; however, lengthening a tibia more than 50 mm carries a high risk of joint contractures and nonunion. The femur may tolerate lengthening greater than 50 mm, but the surgeon must carefully evaluate the elasticity of the soft tissues, the healing potential of bone, and the compliance of the patient. Once the goal length has been established, the appropriate nail size is determined. The length of the bone and the size of the canal will determine the range of sizes that may be used.

The optimal location of the osteotomy is also determined when choosing the nail. The distal end of the osteotomy (after lengthening) should be located at least 30 mm proximal to the distal point of the proximal rod, in the proximal diaphysis, to enhance the stability of the osteotomy by placement at the point where the nail has significant potential contact with the endosteal cortex. Ideally, the osteotomy is made away from the wide metaphyseal area to avoid varus or valgus malalignment during lengthening. When the surgical plan includes correcting a deformity simultaneously with the lengthening procedure, placing the osteotomy close to the metaphysis may be adequate. In this case, the blocking screw technique (Poller) screw is often necessary to control the alignment of the metaphyseal fragment (21).

When choosing a nail and an osteotomy site in the femur, attention should be directed at the location and the magnitude of the femoral bow. The apex of the femoral bow in the sagittal plane is an ideal place for the osteotomy; if a different site is chosen, the nail–endosteal contact will increase at the bow resulting in more torque necessary to achieve nail rotation and lengthening.

Retrograde femoral nailing is indicated in the presence of a proximal femur implant or deformity that prevents antegrade insertion of the ISKD. Other applications of retrograde femoral ISKD include patients who have previous retrograde femoral nails or in the selected patient with distal femoral deformity, that will be addressed at the same time as the length discrepancy. The choice between a femur and a tibia nail for retrograde insertion will depend on the distal femoral anatomy and the specific surgeon preferences. In retrograde femoral nailing, the shortest possible nail will prevent problems associated with passing a nail beyond femoral bow. If the ISKD is too long, the surgeon will encounter difficulties inserting the nail past the femoral bow. Increased cortical contact between the straight nail and the femoral cortices may increase friction during rotation. The direction that the magnet rotates is left to right from the patient's perspective in the retrograde inserted nail (opposite to the antegrade). This should be considered when monitoring the lengthening process.

Surgical Steps

In our practice, the first step of most ISKD surgeries is application of an external fixator. If used routinely, this step facilitates rotational control during insertion and locking of the nail. The external fixator is also useful to confirm that the osteotomy is complete and that distraction can be performed. The external fixator is used in the tibia and the femur. The pin placement and assembly of the frame is shown in Figure 8.

The position of the external fixator should not interfere with the placement of the instruments and the hardware. It should allow the surgeon to use dynamic stabilizers such as blocking screws or canal fillers to direct the bone segments when required. The external fixator is removed at the end of the surgical procedure.

Once the site for the osteotomy is selected, drill holes are performed percutaneously at the selected site. Closed osteotomy with an IM saw does not have advantages over percutaneous method; it increases the time and difficulty of this simple procedure. Drilling the cortex prior to reaming allows for 'venting' of the IM canal, perhaps decreasing the risk of fat embolism. This sequence also will deliver reaming debris to the osteotomy site.

The IM canal needs to be reamed 1.5 to 2 mm larger than the diameter of the proximal portion of the nail. Other than a 7° proximal bend in the tibial nail, both the femoral and tibial

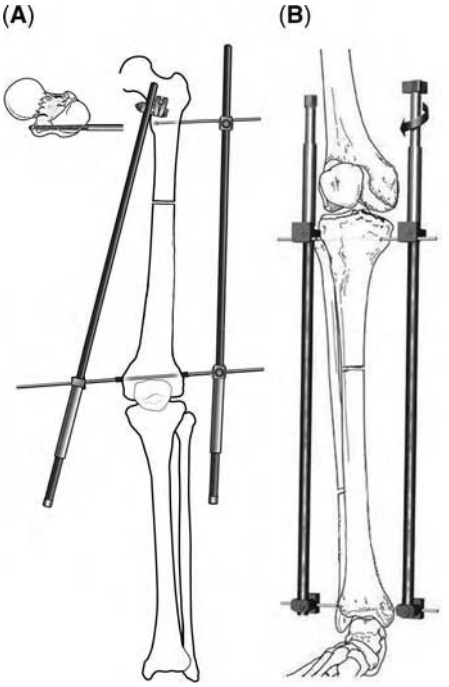

Figure 8 Fixator assisted nailing. (**A**) Femoral fixator–distractor, axial view on the left. (**B**) Tibial fixator–distractor.

ISKDs are straight. Once the full length of the canal has been reamed, the osteotomy is completed with an osteotome. The external fixator maintains the length and the rotation of the osteotomized bone for nail insertion. The ISKD is a solid nail; it does not allow for a guidewire to aid in nail insertion. When lengthening the tibia, the fibula needs to be osteotomized and usually fixed to the tibia.

Prior to nail insertion, the ISKD is tested for correct function by gently manipulating the proximal and distal sections and observing telescoping of the distal section out of the proximal section. Magnet function should also be tested using the magnetic sensor device wrapped in a sterile bag. When the goal length is smaller than the full potential length of the lengthener, the nail is manipulated or dialed out until the appropriate starting length is obtained. The nail insertion proceeds in the usual manner. All nails are locked both proximally and distally. Distal locking is performed with a radiolucent drill offset. Precise and accurate drilling of the bone for the locking screws is required. The nail's locking holes will only allow perpendicular screws due to a tighter tolerance than routine locking nails. After the nail is locked and the distraction of the nail has been confirmed, the proximal and distal tibia-fibula syndesmoses should be fixed with transverse cortical screws to prevent migration of the fibula during distraction (Fig. 9).

Postoperative Management

After the calculation of the goal length and the insertion of the ISKD, the surgeon does not have direct control over the lengthening process. Prior to the surgery, the physician and a qualified assistant must establish good communication with the patient and a caregiver. Patient education and close follow-up are vital to a successful lengthening procedure. The patients need to understand the ISKD's internal mechanism and the importance of their participation in the postoperative period. After the correct use of the measuring device or monitor is reviewed, the patient must demonstrate adequate measuring technique prior to discharge from the hospital. Starting the first postoperative day, the patient or a caregiver must measure every two to four hours while he/she is awake. Measuring during the night is recommended if the patient is awake and when using the restroom. During the first five days, the patient should try to have as minimal physical activity as possible and limit pole changes. Patients are evaluated on the fifth or sixth postoperative day and then at one- to two-weekly intervals until lengthening is complete. Patients must be instructed on the desired number of pole changes during each visit. The goal will be determined by the physician based on the

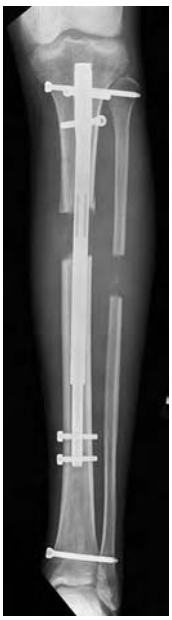

Figure 9 Distracted tibia with Intramedullary Skeletal Kinetic Distractor and proximal and distal syndesmosis fixation; discrete regenerate noted at the distraction site.

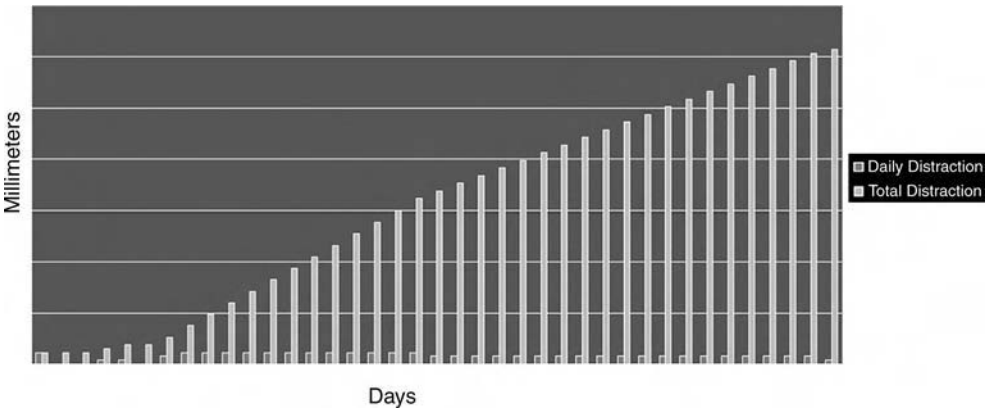

Figure 10 Total Distraction Chart. Five days after surgery: minimal distraction. Initial two weeks of distraction: three pole changes per day. Last phase of distraction: two pole changes. The rate was decreased based on the discrete regeneration noted on the radiographs.

healing potential and the characteristics of the regenerate on the radiographs. The recommended rate is two to three pole changes per day for the tibia and four to five for the femur. The monitor is downloaded at each visit, compliance with the number of measurements and the registered pole changes are evaluated (Fig. 10). Radiographs are taken at each visit to assess regenerate formation and accuracy of measurements.

Follow-up visits also include assessment of joint motion, ambulatory status, work status, and ability to perform activities of daily life such as driving. Patients are asked to avoid full weight bearing of the short limb during the lengthening phase: 20 to 50 lbs with the aid of crutches or a walker. After the goal length is reached, radiographs are obtained monthly to determine the amount and characteristics of the regenerate. When at least one cortex is seen in the distraction gap, weight bearing is advanced. The patient is encouraged to perform stretching exercises from the preoperative period and to continue stretching throughout the lengthening phase (Fig. 11). Once the goal length has been reached and weight bearing has increased, the patient starts progressive resistive and active strengthening exercises in addition to stretching.

Clinical Experience

From April 1995 to December 2005, 90 patients in our practice had 101 procedures with the ISKD: 44 femoral and 57 tibial lengthenings. The indications are listed in Table 2. One patient had four lengthening procedures to overcome a discrepancy of 167 mm. Eight patients have

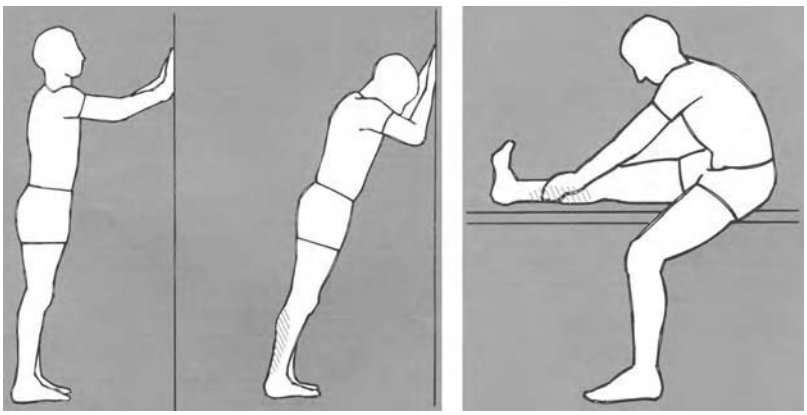

Figure 11 Stretching exercises before, during, and after lengthening. *Source*: Courtesy of Orthofix, Inc.

Table 2 Indications for Lengthening with Intramedullary Skeletal Kinetic Distractor in 90 Patients, April 1995 to December 2005

Indications	Patients
Malunion	28
Infection	27
Limb salvage	17
Deformity	11
Nonunion	7

required two procedures for discrepancies ranging from 70 to 100 mm. The average lengthening rate was 1.14 mm/day (range, 0.4–3 mm/day). The complications encountered include: six patients with premature consolidation, three hardware failures (two nails, one screw from the initial prototype), two patients with joint contractures, one patient with nonunion. Infection and malunion have not been associated with the use of the ISKD. The patients who developed complications have required additional surgery, and none had sequelae from lengthening.

FUTURE DIRECTIONS

Patient satisfaction drives medical care in the United States, perhaps at a cost of efficiency. Application of new and untried methods carries a high toll in costs and morbidity.

In the short time period of my peers' and mentors' orthopedic careers, we have participated in the transition to many patient-friendly surgical techniques including arthroscopy and minimally invasive approaches to trauma. One of these patient-friendly technologies developed during our careers is IM nail application to lower extremity long bone open or closed diaphyseal fractures. Locked IM nails have changed patient functional levels during treatment as well as the prognosis of their injuries. Application of nailing techniques to limb reconstruction is an obvious step toward improving patient satisfaction and patient lifestyle during treatment.

As we analyze and divide the steps involved in successful limb reconstruction, one may look at subjects such as bone growth, skeletal alignment, soft-tissue preservation, limb function, and patient lifestyle as separate fractions of the treatment. We should assess the value of a new limb reconstruction method by comparing it to past methods in terms of these divisions. The lengthening nail's only positive contribution in limb reconstruction to date is theoretical belief that IM nailing may improve patient lifestyle during treatment. Some skepticism is therefore warranted because bone growth, skeletal alignment, soft-tissue preservation, and limb function will not be improved by internal devices.

Overall, the future of limb reconstruction procedures is promising. As surgeons gain experience and the technologic advances are readily available, the process of managing a patient with severe injury, nonunion, and infection should improve. Limb lengthening is one of the final steps in obtaining a functional extremity. The development of newer technology will most likely include different mechanisms for internal lengthening devices. In the near future, however, the cost and limited market size will limit the application of devices that offer more patient and surgeon control.

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47 Emerging Intramedullary Bone-Lengthening Technologies

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INTRODUCTION

Limb lengthening has gained increasing acceptance in the past two decades across the globe. Progressives understanding of the principles of distraction osteogenesis and advancements in operative technique as well as in equipment (1,2) have allowed improved success rates of such procedures.

However, limb lengthening still entails a considerable amount of risk (3). Some of the problems arise from the application of the external fixator, such as soft-tissue infection, neurovascular injuries, and soft-tissue scaring (3,4). Other risks are associated with internal lengthening devices—namely rate control. The lack of control of the distraction mechanism makes it more difficult to prevent premature or delayed consolidation, nerve compromise, and joint subluxations.

In order to simplify postoperative care and to prevent the above-mentioned complications, we have designed a new internal lengthening nail.

Several telescopic intramedullary nails (IMNs) had been developed over the years. However, only few have matured into medically approved devices and remain in current use, namely the Bliskunov nail (Russia) (5), Albizzia (France) (6), ISKD (Orthofix, United States) (7), and the Fitbone (Wittenstein Intens, Germany) (8). Yet, the proposed designs need to address critical technical issues such as force generation, accurate control, and design efficacy (Table 1).

PREREQUISITES FOR TELESCOPIC INTRAMEDULLARY NAIL DESIGNS

Generation of Force

The telescopic IMN should be able to produce forces sufficient to overcome the resistance of the soft tissues including the deep fascia and the interosseous membrane as well as the regenerate. The peak resistance forces encountered during in-vivo bone lengthening by external fixators may range from 140 to 220 N in the tibia (9,10) and 420 to 680 N in the femur (11). The intramedullary location of the IMN is expected to require smaller forces during distraction osteogenesis in comparison to external fixators due to direct axial application of the force resulting in less loss of para-axial forces. However, no valid data is yet available regarding the distraction force that is necessary for bone lengthening by IMN.

Control, Accuracy, and Feasibility

One of the basic principles in bone lengthening is a timed and accurately controlled distraction rate in order to avoid premature consolidation of the distraction regenerate on one hand, and nerve injury, joint stiffness, joint dislocation, and delayed consolidation (1,2,12) on the other

Table 1 Comparison of Lengthening Intramedullary Nails

	The Ideal Intramedullary Nails (IMN)	ISKD	Fitbone	Orthogon (in development)	Albizzia	Bliskunov
Elongation principle	Axial	Foot rotation	Activated by charging an internal battery through an electric antenna	Magnetically activated	Foot rotation	Hip rotation
Surgical technique	Simple IMN	Simple IMN, overreaming of tibial cortex	Simple IMN	Simple IMN	Simple IMN	Use of external fixation, technically more demanding
Mechanical effect on callus	Axial distraction	Axial and rotational	Axial distraction	Axial distraction	Axial and rotational	Axial and rotational
Soft and neurovascular tissue injury	Minimal	Minimal	Minimal	Minimal	Medium	Medium
Number of cases performed	Established experience	Several hundreds	Less than hundreds	Experimental	Several hundreds	Several hundreds
Controllability	Excellent—accurate, controllable and repeatable	Poor	Excellent	Excellent	Poor	Poor
Nonradiological monitoring	Excellent	Mediocre	–	Good	None	None
Need for patient education	Minimal	High	Minimal	Minimal	High	Very high
Need for patient cooperation	Minimal	High	Minimal	Minimal	High	High
Mechanical fidelity	High	Good	High	High	Good	Good
Need for close follow up	Minimal	High	Minimal	Minimal	High	High
Possibility for ambulatory elongation	High	High	High	High	High	High

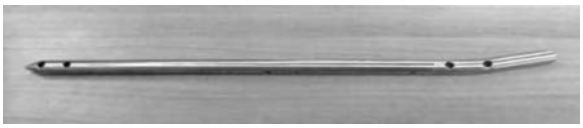

Figure 1 Orthogon's magnetically actuated tibial intramedullary telescopic nail.

hand. Moreover, because there is no access to the nail to correct mechanical failures once it is implanted, the device should be highly reliable.

Physical Properties and Biocompatibility

The telescopic IMN should be strong enough to withstand various mechanical forces operating on the limb until reliable union takes place, without compromising its distraction mechanism, especially during the distraction phase. The above prerequisites should be met within the confines of biocompatibility, the sterilization process, and size, i.e., diameter and length.

MAGNETICALLY ACTUATED NAIL

A new intramedullary magnetically actuated bone-lengthening system for the controlled lengthening of long bones (e.g., femur, tibia, and humerus) is currently under development. The system is composed of a telescopic IMN (Fig. 1) and a magnetic field generator. The telescopic IMN consists of two major sections, each fixed to the bone by two interlocking screws. The part that holds the magnetic and distraction mechanism is static. The second part is a telescopic element that moves out from the static part. This creates an axial distraction force on the regenerate. The unidirectional motion (lengthening) is mechanically constrained by a unique ratchet mechanism that enhances control over the distraction rate.

Placing the limb inside an electromagnetic coil (Fig. 2) activates the controlled motion of the telescopic nail. Electromagnetic pulses in the coil induce a low-level homogenous and directional magnetic field within the nail. These magnetic pulses have the shape and characteristics of a static magnetic field, causing a controlled lengthening of the telescopic nail. Each magnetic pulse in the coil induces a homogeneous and directional magnetic field inside the nail that distracts the nail by 0.02 mm. Each session for the distraction of 0.24 mm will require 12 pulses.

Orthogon's (Israel) coil system, integrated within a console, includes a power supply, amplifier, energy storage modules, and a coil. The system operates from a single-phase power outlet, thus enabling the patient or the physician to operate the system at home or in a clinic. The low-level magnetic field is only induced inside the inner diameter of the coil (around the limb), and there is no magnetic field outside the coil.

FUTURE DIRECTIONS

The use of the Orthogon (Israel) magnetically actuated intramedullary rod may have other clinical applications in addition to limb lengthening.

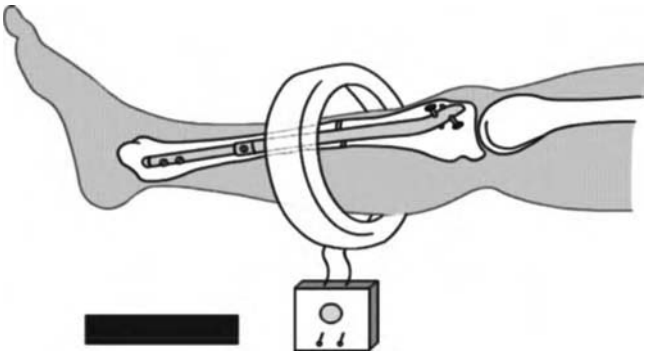

Figure 2 Schematic diagram of limb lengthening by a magnetic coil.

Nonunions

Interlocked intramedullary nailing is useful in the treatment of nonunions of long bones. The magnetically actuated nail can combine the advantages of IMN fixation with controlled compression. Technically, it could be achieved by using a nail with a reverse force generating mechanism. This nail would be magnetically actuated in a manner similar to the lengthening design, yet the motion of the telescopic part of the nail will be toward the static part. This would result in compression across the nonunion. The magnitude of the compression force and the frequency of its application can be predetermined and adjusted to the clinical requirements.

Fractures

During the past 30 years, IMNs have become the mainstay for treatment of lower extremity long bone fractures (13,14). However, other methods of fracture treatment (15–18), based on more dynamic fixation were developed in order to shorten the healing time. Kenwright and Goodship have demonstrated that controlled dynamization for 17 minutes a day by external fixators in humans, significantly shortened the healing time of tibial fractures and promoted callus formation and density in a sheep model (15–17). Choi et al. had similar results in a canine tibia model (19). Yet, the application of these principles to interlocked IMNs by dynamization resulted in 10.5% shortening and rotational instability, limiting its use (20).

A magnetically oscillating nail would combine the advantages of an interlocked IMN with that of dynamic external fixation when used in the treatment of primary long bone fractures. Moreover, the rate of oscillation as well as the magnitude of mechanical compression force would be externally controlled. Consequently, this could result in significantly shorter fracture healing times as observed in the use of external fixators by Kenwright and Goodship (16).

Table 2 Review of Literature

Authors	Journal, Year	Title	No. of Patients	Results	Conclusions
Guichet et al.	JBJS, 2003	Gradual femoral lengthening with the Albizzia intramedullary nail	31 (41 femur)	2–8.4 cm lengthening. Patients underwent on average three operations on each limb. Eleven surgeries for complications; 31 anesthesias for ratcheting. At follow up—no axial deviation of the limb secondary to lengthening	Reasonable alternative to external fixation, well tolerated by patients
Hankemeier et al.	Arch Orthop Trauma Surg, 2004	Improved comfort in lower limb lengthening with ISKD	4	Excellent functional result of the patients	High patient comfort during limb lengthening with the ISKD
Baumgart	Clin Orthop, 1997	A fully implantable motorized intramedullary nail for limb lengthening and bone transport	12	All of the bones healed and no fractures occurred in 2 yr margin	The Fitbone nail is a good alternative for limb lengthening

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48 Monolateral Frame Strategies: The Multiaxial Correcting Monolateral External Fixation System (EBI®)

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INTRODUCTION

While monolateral fixation has been available for most of the past century, its applications have been limited to acute fixation of fractures and osteotomies, as well as to limb lengthening. With the introduction of the Ilizarov fixation device in the mid-1980s outside the Soviet Union, external fixation took a giant leap into three-plane deformity correction. However, compared to monolateral fixation, the Ilizarov ring fixator was more cumbersome and less friendly to patient and physician. Since the 1980s, monolateral fixators have been redesigned to handle single plane angulation and translation. In the past two years, a Multiaxial Correcting (MAC) Monolateral External Fixation system (MAC system)(EBI®, Parsippany, New Jersey, U.S.) has been developed for the correction of linear, angular, translational, and rotational deformities of the limbs.

The rotational, angular, and linear application of these new monolateral external fixators is the subject of this chapter.

CLINICAL RELEVANCE

Gradual correction of deformity using external fixation can avoid or correct many problems of acute correction. Steel et al. (1) reported a 19% nerve and vessel complication rate in acute correction by proximal tibial osteotomies. Young et al. (2) demonstrated that six of six patients undergoing osteotomy for lengthening of the tibia showed electro-diagnostic abnormalities in the deep peroneal nerve or superficial peroneal nerve, as well as increased compartment pressures.

Even when performed by experienced orthopedic surgeons, acute corrections of limb deformities, fixed with internal fixation or static external fixation, have been reported to result in 10% to 26% inadequate corrections (3–6). However, these static devices do not permit gradual correction in the postoperative period. When treatment with internal fixation or traditional static external fixation results in inadequate correction, the only option is to return to the operating room to fix the problem.

While gradual correction with the multiplaner ring external fixation systems reduced the risks of these problems, they were bulky, uncomfortable for the patient, and difficult for the treating physician to alter the plane of correction (7,8). Early monolateral fixators, while user friendly for the patient and physician, could not accomplish the complex corrections of the ring fixators.

The key to all corrections of deformity using external fixation, as well as the development of a monolateral fixator that could correct multiple planes of deformity, lay in the application of geometric rules of correction organized by Paley (9–12). In summary, there are three elements to each deformity correction: the center of rotation of angulation (CORA), the angulation correction axis (ACA)(or the hinge of the fixator), and the osteotomy location. Quoting from Paley (12):

> "Osteotomy rule 1. When the osteotomy and the ACA pass through any of the CORAs, realignment occurs without translation. Osteotomy rule 2. When the ACA is through the CORA but the osteotomy is at a different level, the axis will realign by angulation and translation at the osteo-

tomy site. Osteotomy rule 3. When the osteotomy and ACA are at a level above or below the CORA, a translational deformity will result.''

A corollary to Rule 1 is that, when the ACA is along the bisector of the deformity, but not at the central CORA, lengthening or shortening must occur.

A practical problem continued to plague monolateral fixators in terms of multiaxial correction: only certain anatomic locations permitted safe placement of pins and screws. Whereas ring fixators permitted circumferential access to the fixator for safe placement of wires and bone screws, the nature of monolateral fixators did not. For example, a flexion deformity of the femoral diaphysis would require that the hinge of a traditional monolateral fixator be placed at the anterior thigh, risking injury to the femoral artery and nerve, as well as transfixing the quadriceps muscle. This meant that simple hinge fixators could be applied in a CORA centric or CORA perpendicular method only to specific anatomic locations, where the device could be applied at the CORA and the pins could be safely applied without injury to soft tissues.

In a CORA centric application, the hinge is directly over the central CORA and correction is obtained without lateral or axial translation (Fig. 1), such as with an anterior angular hinge fixator in the treatment of Blounts disease. In a CORA perpendicular application, the ACA is placed along the bisector on the concave or convex side of the deformity with the secondary alteration in length, such as in the treatment of distal femoral valgus with a laterally placed angulating rail (Fig. 2).

To resolve these problems of monolateral fixation, the MAC system provides two planes of angulation, two planes of translation, and lengthening and rotation components that allow correction at any location along the upper or lower extremities (Fig. 3).

HINGE FIXATORS AND ANGULATING RAILS

Two of the earliest monolateral fixators capable of gradual angular correction were the Garsches Clamp (Orthofix, Verona, Italy)(Fig. 4A) and the Anterior Angulating Hinge Fixator (EBI)(Fig. 4B). Placed on the anterior proximal tibia they could correct varus, valgus, or oblique plane deformity when applied in a CORA centric oblique plane fashion. These devices were well suited to the correction of proximal tibia vara (Blount's and degenerative diseases) or tibia valgus because they could be positioned on the CORA and had anatomically safe access for bone screw sites but not other sites. Additionally, by design, these single plane hinges could not correct for residual or secondary deformities perpendicular to the primary plane, nor could they correct rotation. Essentially, if they were not placed perfectly

Figure 1 Center of Rotation of Angulation (CORA) centric application of a Multiaxial Correcting External Fixation System. The Hinge of the device is centered on the CORA. *Abbreviation*: CORA, center of rotation of angulation.

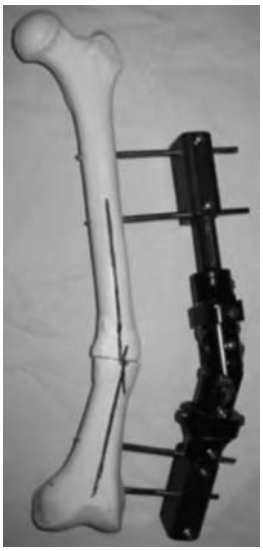

Figure 2 CORA Perpendicular application of a Multiaxial Correcting External Fixation System. The device is placed on the bisector of the deformity. *Abbreviation*: CORA, center of rotation of angulation.

on the CORA, the patient would have to live with the residual deformity or undergo additional surgery.

Angulating hinges were also applied to rails as CORA perpendicular fixators (Fig. 5A). These could be applied to the femur laterally to correct varus or valgus deformities or applied medially to the tibia to correct varus or valgus deformities. In some cases, they could also be applied to deformities of the humerus or the forearm.

When these angulating rails were placed on the convex side of a deformity lengthening would occur, but when placed on the concave side of a deformity shortening would occur. Clearly, when lengthening is desirable, when the distance from the deformity provides just the correct amount of length, and when the bone screws can safely be placed on the convex side of the deformity, such gradual correction would be beneficial. Still, calculating the length that would occur required knowledge of trigonometry and depended on the distance between the bone and the hinge (ACA). The further the hinge was from the bone, the greater the increase in length. However, such calculations were difficult.

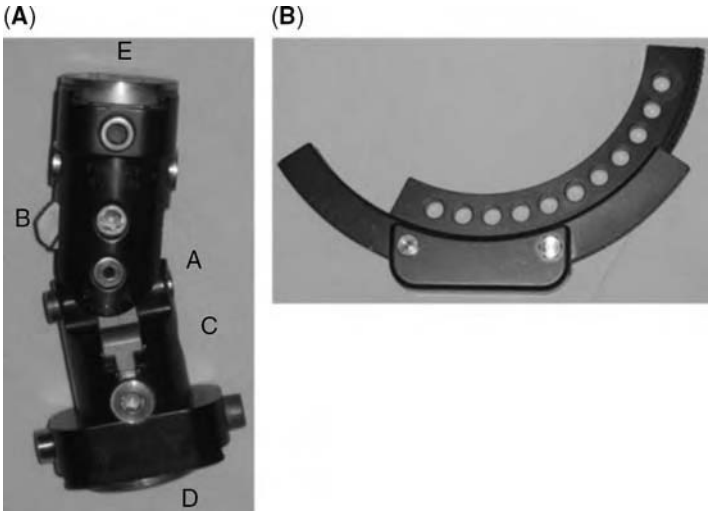

Figure 3 (**A**) The Multiaxial Correcting External Fixation System: A is the centering hole for the primary hinge, B is the primary hinge, C is the secondary hinge (90° to the primary hinge), D is a translation device, and E is a second translation device at 90° to D. The screw holes at either end allow attachment of rails, compression/distraction devices, arcs, rings, rotation arcs, etc. (**B**) The Multiaxial Correcting Rotation Arc.

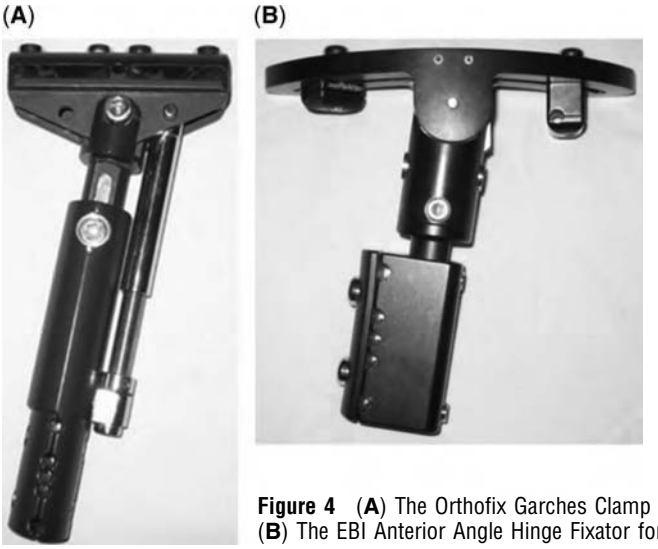

Figure 4 (**A**) The Orthofix Garches Clamp for single hinge/single plane deformity. (**B**) The EBI Anterior Angle Hinge Fixator for single hinge/single plane deformity.

When the hinge was positioned on the concave side of a deformity (such as with distal femoral valgus or a proximal tibia varus deformity), the shortening would have to be calculated first, then the bone lengthened, and then the angular correction would shorten the bone (Fig. 5). At 1 mm of lengthening a day, the process of angular correction without length change could take months. Often, consolidation of the gap would occur, interfering with correction of the angulation. Carefully calculated instructions and extremely compliant patients could often reduce the time by simultaneously lengthening and altering the angle (lengthen 1 mm per day, while shortening 1 mm per day through angulaltion or vice versa). Still, premature consolidation remained a common complication.

In spite of these shortcomings, the hinge and angulating rail monolateral fixators remain popular alternatives to the ring fixators because of their ease of use for both patient and physician.

Perhaps the most difficult issue in correcting deformity with any external fixation device (monolateral or ring) was the ability to apply the devices to the patient in exactly the correct

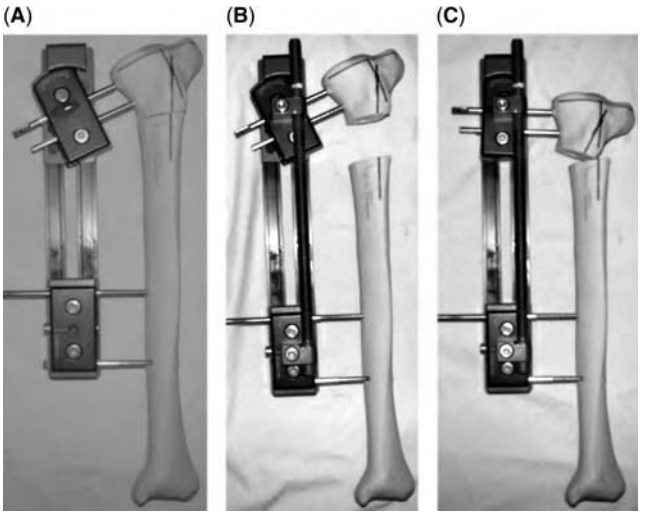

Figure 5 (**A**) The EBI angulating rail placed CORA perpendicularly on the bisector of a proximal tibia angulation deformity. (**B**) In order to correct this angular deformity, the bone must first be lengthened. (**C**) Then the deformity can be corrected using the angulation hinge, top. Translation in the plane of the deformity can be corrected by the translation clamp, bottom.

location. Most deformities are oblique to the frontal and sagittal planes. Precisely orthogonal (90° to each other) X-rays must be taken to assess the deformity. In the operating room, the surgeon must know how to apply the device exactly on the axis of the CORA or along the bisector. Errors occur, which lead to secondary deformities.

Misalignment in any plane leads to secondary (iatrogenic) translation, rotation and, angulation deformities. The traditional monolateral fixators lacked degrees of freedom to correct any deformity out of plane of the device. The Ilizarov frame (Smith and Nephew, Memphis, Tennessee, U.S.) required time-consuming and complicated adjustments in the clinic. The Taylor Spatial Frame (Smith and Nephew, Memphis, Tennessee, U.S.) required frequent multiple measurements and programming these into a computer. All of the ring fixators were cumbersome to the patients and complex for the physicians.

THE MULTIAXIAL CORRECTING MONOLATERAL EXTERNAL FIXATION SYSTEM: MULTIAXIAL CORRECTION OF LIMB DEFORMITIES

In spite of many advances in the past century, ring fixators remained cumbersome and complicated, while monolateral fixators had limited applications. An idealized monolateral external fixator would need to be applicable coaxially with the CORA or along the bisector, permit fixation through bone screws and wires at safe anatomic locations, and permit simple multiaxial correction including angulation, translation, lengthening (compression and distraction), and rotation. Additionally, the device should permit correction of secondary deformity from misapplication, bending of wires or screws, and soft-tissue resistance. The MAC device was designed to satisfy these principles.

The MAC system (Fig. 3) consists of a central component with a primary and secondary hinge capable of 80° and 35° to 55° of angulation, respectively, in two planes. There is a translation screw at either end mounted at 90° to each other and each is capable of 3 cm of translation. The MAC device can be mounted CORA centrically (Fig. 1) on a guide pin inserted into the axis of the CORA or along the bisector of the CORA in a CORA perpendicular fashion (Fig. 2).

Each end of the MAC device permits attachment of a variety of components, including compression/distraction bone screw blocks, rails, arcs, rings, or rotation arcs, to allow positioning of the device where needed and safe placement of fixation screws. This component system permits application virtually anywhere along either the upper or lower extremity.

Applications include acute or gradual correction of deformity or fixation of fractures at locations including: the proximal femur for correction of varus, valgus, rotation, or severe SCFE deformity; any long-bone diaphysis deformity; metaphyseal deformity of any long bone (tibia vara, Blount's, high tibial osteotomy, etc.); genus recurvatum; clubfoot at the midfoot or supramalleolar tibia; radial club hand.

Correction of deformity is attained through simple turning of the screws using an Allen wrench. Secondary (iatrogenic) deformities can be corrected just as easily by turning angulation or translation screws for the appropriate plane.

Assessment of deformity at each office visit requires only anteroposterior (AP) and lateral X-rays and the measurement of the angle. For CORA centric correction, most patients tolerate a correction rate of 1° four times per day, while for CORA perpendicular corrections lengthening bone and soft tissues one millimeter per day is tolerated.

Rotation can be corrected by choosing to apply a rotation arc to attach to either end of the MAC device. While ideally the arc should be attached with the bone centered in the arc to avoid secondary deformity through translation, the device can, within limits, correct such deformity.

A sawbone demonstration of adolescent Blount's disease of the proximal tibia is illustrated (Fig. 6). AP and lateral X-rays are obtained and demonstrate a deformity of 22° of metaphyseal deformity. The CORA is identified at about 1.5 cm distal to the physis. Tibial torsion of 30° is identified. An appropriate-sized guide pin is placed in the axis of the CORA with care to align the wire in the frontal and sagittal planes with the long axis of the leg. A two-finger breadth spacer of padding is placed on the skin at the guide pin. The primary guide hole of the MAC device is placed over the guide pin and the primary hinge is aligned so that the angle of the primary hinge matches the deformity. Two or three bone screws are inserted into the upper tibia from anteromedial to posterolateral and anterolateral to

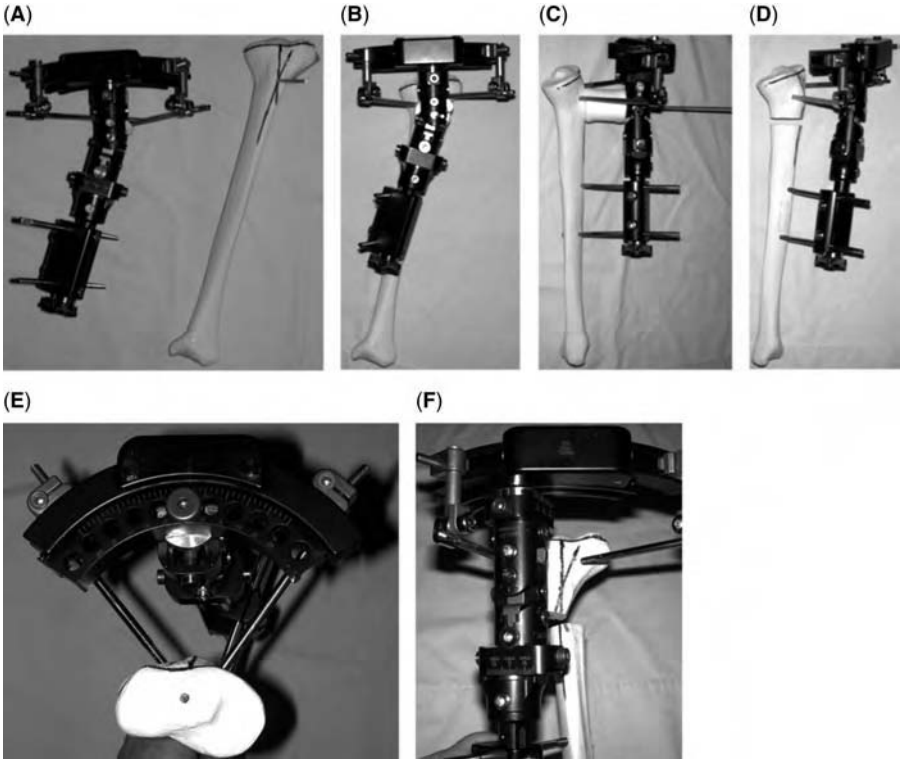

Figure 6 (**A**) The Multiaxial Correcting (MAC) External Fixation System assembled for an adolescent Blount's tibia application. Note the Sawbones® model with lines drawn to identify the CORA. (**B**) The MAC External Fixation System applied to the model. Note the arms of the device are aligned with the deformity. The primary hinge-centering hole is placed on a guide pin inserted into the CORA with a "two-finger breadth" spacer. (**C**) Lateral view shows that the MAC External Fixation System is applied with the arms parallel to the deformity in the sagittal plane. Note spacer to protect the skin. (**D**) Once applied and aligned to the deformity, the osteotomy is completed, gradually distracted, and correction is accomplished. (**E**) The rotation arc or the MAC External Fixation System is centered on the bone to minimize any secondary translation. (**F**) After correction, the lines demonstrate restoration of normal anatomy. Incorrect application of the device (not centering on the CORA) will cause secondary deformities, which can be corrected with the MAC External Fixation System. *Abbreviation*: CORA, center of rotation of angulation.

posteromedial, avoiding the joint (note, the proximal tibia slopes posteriorly) and the physis if it is still open. For proximal CORAs, the screws are simply aimed distally. The desired location for the osteotomy is at the CORA, but safe placement will be distal to the proximal tibial physis because of the tibial tubercle and attachment of the patellar ligament (the osteotomy must be placed distal to the attachment or the patella will be displaced distally). The safe placement of the proximal bone screws is in the metaphysis but distal to the joint and physis. If rotational correction is desired, then a rotation arc is chosen for the proximal component, otherwise a solid arc is chosen. Distally a compression/distraction bone screw block is chosen and assembled to the MAC device through a male adaptor. Two proximal screws are attached to the metaphysis, one anterolateral to posteromedial and the other anteromedial to posterolateral. Three distal screws are attached to the tibial diaphysis at the compression/distraction component. Care must be taken to assure that the distal part of the MAC device is parallel to the diaphysis in the frontal and sagittal planes, while the proximal part is aligned with the proximal tibia. The device is securely fixed to the bone screws. An osteotomy is performed either percutaneously with a Gigli saw with the device in place or with drill holes and an osteotomy removing the device. (Hint: measure the screw lengths extending anterior to the device prior to removing the fixator for the osteotomy and then replace it at the same position). After about one week, the compression/distraction device is used to lengthen $\frac{1}{4}$mm (a 90° turn four times each day) for one week. This creates a space allowing the flat osteotomy to slide along the arc defined by the MAC's hinge. Then the angle is corrected with the hinge. Once

the mechanical axis is restored, rotation can be corrected. If the MAC device was not properly aligned with the CORA, then secondary corrections can be made with the secondary hinge or translation screws without altering the device, returning to the operating room or using a computer to analyze the deformity or correction. Because the MAC device is based on a coordinate system, only AP and lateral X rays need be taken to define the alignment at each weekly visit (Fig. 6).

The rate of correction can be easily calculated using the principles of similar triangles (13) or from precalculated tables available from the manufacturers of hinged external fixators.

CONCLUSIONS

The principles of multiaxial deformity correction developed with ring fixation have served as a guide in the development of new strategies for the fixation and correction of deformity with monolateral fixators. The MAC monolateral fixation system can accomplish correction of these deformities with improved simplicity and comfort for the physician and patient. It can help to correct secondary deformity due to inadequate placement of the hinge and bending of screws under the loads of distraction. It works simply, using CORA principles without the need of computer technology.

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Part XIII POSTOPERATIVE CARE

49 Obstacles in Limb Lengthening
Nerve Problems

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Nerve injury is one of the most feared complications in limb lengthening. It could cause impairment of the limb function, compromising the results of limb lengthening or deformity correction. Reported incidence varies from 3 to 30% (1–11).

There is no direct correlation between the amount of lengthening and nerve injury, as well as no specific threshold in the number of centimeters for nerve injuries to occur. One possible explanation for this is anatomic variation. Lengths and circumferences of limbs vary from patient to patient depending on constitutional properties such as adipose mass, and ability of soft tissue to stretch. Moreover, the majority of nerve injuries in the leg occur only at 2 to 5 cm lengthening, suggesting anatomic predisposition to peroneal entrapment at the neck of the fibula (the most common nerve injury in the leg) rather than a stretch injury to the nerve, which would be expected with a greater amount of lengthening.

Early diagnosis and treatment of nerve injuries are very important in order to avoid permanent impairment. In recent years, newer techniques have been developed to detect impending problems with the nerves.

CLASSIFICATION

Nerve injuries can be classified into surgical and gradual stretch nerve injuries.

Surgical Nerve Injuries

They occur during the surgical procedure.

They are subdivided as follows:

1. ''Direct surgical injuries'' from half pins, osteotomes, drill bits, saws, or wires during surgery. This can usually be avoided with good surgical technique. In the operating room total muscle relaxation during anesthesia should be avoided. While inserting pins or wires in areas close to the nerve, the distal part of the limb should be observed for any movements or twitching. Soft tissue and nerves are protected with soft-tissue sleeves around drill bits. Wires should be tapped through the soft tissue to minimize nerve injury.
2. ''Intraoperative acute stretch injuries'' are due to acute corrections of a bone or joint deformity, a rotation osteoclasis maneuver, or an inadvertent acute distraction at the osteotomy site. It depends on the amount of corrected angulation. Acute corrections over 15° are at higher risk of nerve injuries (8,9,12,13). Osteoclasis must be performed gently, and in the preferential direction considering the nerve position (Fig. 1).
3. ''Nerve entrapment related to surgery''—the nerve gets entrapped by pins or wires located next to it during lengthening.

Gradual Stretch Nerve Injuries

They are the most common nerve injuries. They are related to microstructural changes and axonal flux disturbance (14–16).

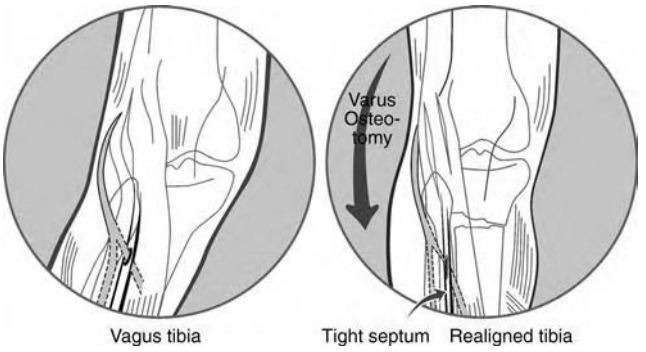

Figure 1 Nerve stretching in acute corrections.

They are subdivided into:

1. "Gradual stretch injuries"—they appear gradually, initially with referred pain in the nerve tract, followed by sensory impairment, and finally compromise of motor function (Fig. 2).
2. "Acute on gradual stretch injuries"—they appear with acute corrections during or at the end of lengthening, when the nerve is already under tension and the leg is acutely repositioned. These maneuvers should be avoided (Fig. 1).

ANATOMIC CONSIDERATIONS

Nerves are formed after leaving the spinal foramens and tract between different compartments in the limbs. Compartments are limited by fasciae and septae that do not stretch as the other connective tissues. They resist to lengthening and increase tension as they distract (Fig. 3).

Peroneal Nerve

The peroneal nerve injury is the most common in lower-limb lengthening and deformity correction. This could be explained by the anatomy of this nerve (17,18). The common peroneal nerve is one of the two main divisions of sciatic nerve in the popliteal fossa. It tracks lateral to the edge of the biceps muscle in the distal thigh, and enters the lateral compartment of the leg. It crosses the fibular neck, and enters behind the peroneus nerve muscle. This entrance is usually narrow and limited by the peroneus nerve fasciae. In the lateral compartment, the common peroneal nerve divides into two branches, the superficial peroneal nerve and the deep peroneal nerve. The superficial peroneal nerve innervates peroneal muscles and tracks down in the lateral compartment. The deep peroneal nerve passes to the anterior compartment of the leg crossing the anterior intermuscular septum through a tunnel with the same diameter as the nerve. With lengthening or deformity correction, the septum stretches and changes its orientation constricting the nerve.

It is important to remember that deformity correction could also affect the peroneal nerve during correction of the varus deformity. Intuitively, this direction should favor a nerve

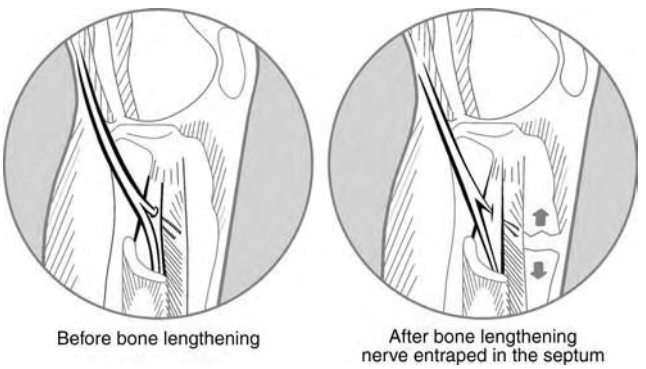

Figure 2 Nerve stretching during lengthening—nerve entrapment in the intermuscular septum.

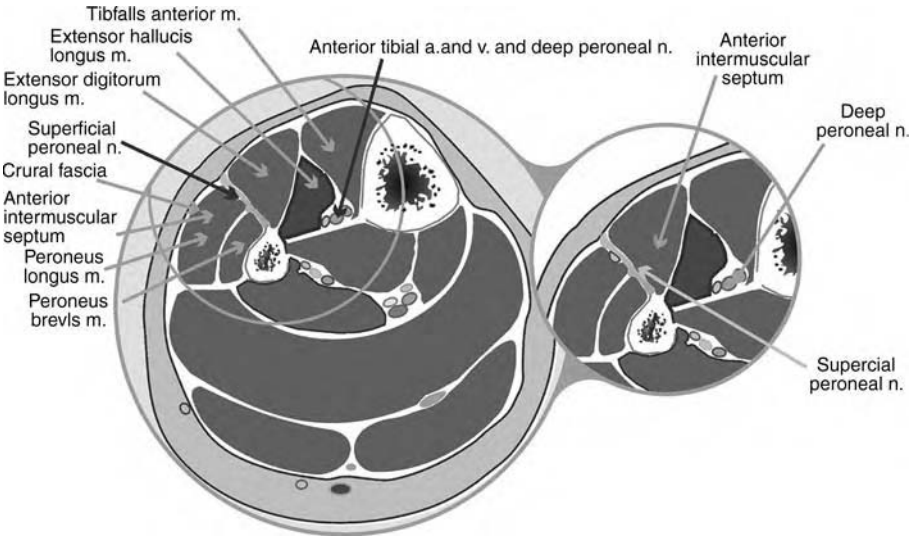

Figure 3 Compartments of the leg separated by fasciae and septae.

relaxation; however the nerve could get entrapped when it passes through the septum, which changes its tension and orientation.

Posterior Tibial Nerve

This nerve passes around the posterior aspect of the medial malleolus through the tarsal tunnel. It is situated between the posterior tibial artery, anteriorly and the flexor hallucis longus tendon, posteriorly. It enters the foot by diving under the abductor hallucis muscle. It gives off the calcaneal, lateral plantar, and medial plantar branches. Correction of a long-standing equinovarus deformity will lead to increased tension on the nerve and is a significant risk factor for a stretch injury to the nerve. Previous surgery and scar tissue on the medial aspect of the ankle further increases the risk even with gradual deformity correction.

DIAGNOSIS

Patients undergoing limb lengthening or deformity correction must be accessed for nerve function systematically with a detailed neurological exam with special attention to the area innervated by the nerve at risk for the procedure (19). The examination should include sensory exam, motor function, and reflexes. Tinel's sign could be helpful in defining the site of nerve entrapment. Patients should be warned about the early symptoms of nerve compromise and instructed to notify their surgeon immediately if they experience any of those symptoms. Pain is the first clinical symptom of nerve injury, followed by dysesthesia or loss of sensation. Motor-function impairment is usually a late sign of nerve-function impairment. Patients usually experience pain along the nerve and in its area of distribution. This pain could happen at rest, and must be differentiated from the pain in the inflamed pin sites or by muscle stretching during movement.

Nerve conduction studies have been used for the diagnosis of nerve injury, but they are invasive and most of the times painful for the patient. With a needle placed very close to the nerve, the so-called near nerve studies are highly sensitive to changes in the action potential when the injury occurs (20). Reduction of the amplitude is considered to be the most important electrophysiologic sign of nerve function impairment (16). Electromyography is usually not recommended, because of the common changes in muscle function that happen with lengthening and muscle stretching, like fibrillation potentials. Conduction studies and animal experiments suggest that the alterations in nerve function by stretching can be explained by axonal damage (14,15,21,22).

Nerve compromise can also be detected earlier by quantitative testing of sensory function. The pressure specified sensory device (NK Biotechnical Corporation, Minneapolis, Minnesota, U.S.A.) is able to identify changes such as reduced density of nerve terminations, as well as function of the conducting fibers (23). The device consists of a two-pronged pressure transducer connected to a computer. Patients can be tested by having the examiner touch the area in the nerve distribution with the transducer (with a recorded pressure) and patients press a button when they feel the transducer, or when they can differentiate two different points. The data are integrated by the computer, and graphs are plotted comparing the obtained results with normal values. Patients are tested before the surgery, and then every two weeks during lengthening or deformity correction. If any changes in those values are detected, patients are told to reduce the rate of lengthening, and recovery is expected in the following week. If this does not occur, a nerve decompression is indicated.

TREATMENT

In limb lengthening and gradual deformity correction, the nerve injury must be detected and treated aggressively in order to continue lengthening. The nerve will not recover spontaneously with observation only if the causal factor, which is lengthening, is still active (Fig. 4).

Treatment must be based on the type of nerve injury, surgical or gradual, and on the clinical presentation and results of diagnostic tests.

Surgical lesions must be treated by surgical decompression, because swelling of the nerve will cause a double-crush phenomenon. This phenomenon can be explained by the fact that the nerve was not entrapped at the moment of injury, but as soon as the inflammatory process takes place, the nerve swells, and gets entrapped. This is the basis of recent guidelines to decompress chronic nerve injuries such as diabetic neuropathies.

In gradual stretch injuries, the first measure is to stop or reduce the rate of distraction—the patient is reevaluated after two weeks and if he/she recovers, the lengthening rate could be resumed.

If the patient does not improve with reduction of lengthening rate, nerve decompression is indicated (24–26). Delay in nerve decompression surgery is not recommended, because the recovery is better and faster if the nerve is decompressed just after diagnosis. Patients decompressed within two weeks of diagnosis have the best prognosis of nerve recovery, usually about one month after decompression.

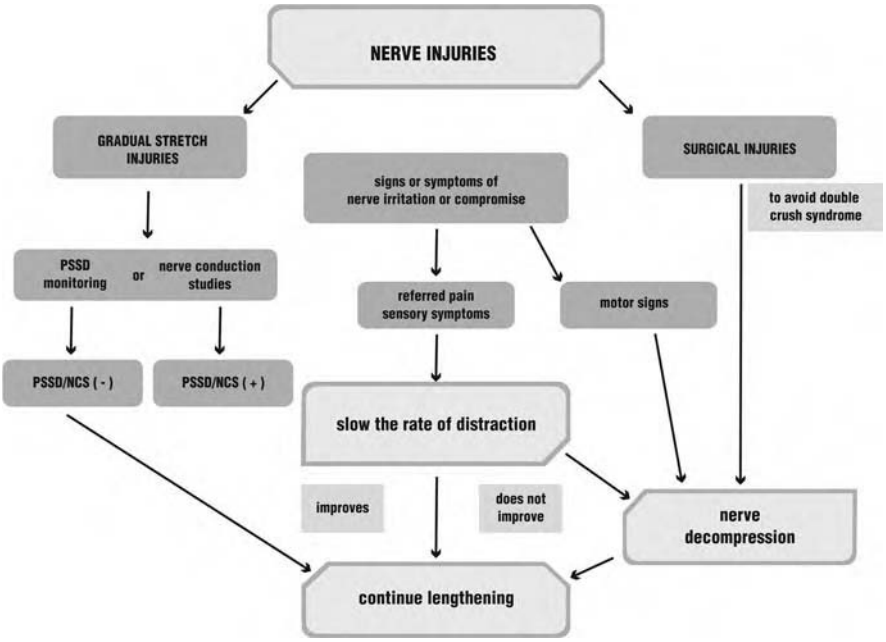

Figure 4 Strategies in nerve injury treatment.

When patients present with motor impairment, this usually is a late clinical sign of nerve injury, and decompression is advised.

Prophylactic decompression is indicated in patients at higher risk of nerve injuries, such as those undergoing double-level lengthening, or big angular deformities. However, in patients with skeletal dysplasia, especially achondroplasia, when a large amount of lengthening is desired, it is preferable to wait and watch for nerve entrapment to occur, and then decompress the nerve as needed.

Nerve Decompression

Nerve decompression must address the specific site of compression of each particular nerve.

For the upper extremity, the radial nerve can be entrapped in the lateral intermuscular septum, between the triceps and brachialis muscles, and in the supinator's fascia. The median nerve can be compressed by Struthers' ligament, lacertus fibrosus, pronator teres, flexor digitorum superficialis, and distally in the carpal tunnel. The ulnar nerve can be entrapped in the cubital tunnel and Guyon's canal.

In the lower limb, the femoral nerve is entrapped in the inguinal tunnel; under the inguinal ligament. The sciatic nerve decompression must address the superficial and deep piriformis fasciae.

Posterior tibial nerve decompression is performed by opening the tarsal tunnel. The nerve is followed in the neurovascular bundle behind the medial maleolus, the fascia of the flexor retinaculum is cut, as well as the superficial and deep fasciae of the abductor hallux (site of division into plantar medial and plantar lateral branches). The release of the calcaneal branches posteriorly must be important to treat pain in the posterior plantar aspect of the foot.

Classically, peroneal nerve decompression was done through a long incision, from the edge of biceps muscle on the lateral aspect of the thigh, to the peroneus nerve muscle (27). However, with better understanding of particular anatomical aspects (28,29), decompression should be focused on two most common sites of peroneal nerve entrapment: the superficial and deep portions of peroneal muscle fascia, and the anterior intermuscular septum. This modified decompression was initially performed and described by Paley, who reported successful decompression of 51 nerve injuries in patients undergoing limb lengthening and deformity correction (8,25).

The skin incision is made about 1 cm under the fibular head in the oblique direction following the nerve and is usually 3 to 5 cm long. The nerve is identified entering under the peroneal muscles. The superficial fascia is cut horizontally, and the peroneus nerve muscle is retracted medially exposing the deep peroneal longus fascia, which should be cut in the direction of the nerve (Fig. 5).

The second site of nerve entrapment is the anterior intermuscular septum. It is found by retracting the peroneal muscles laterally, in the division between the peroneal muscles and the

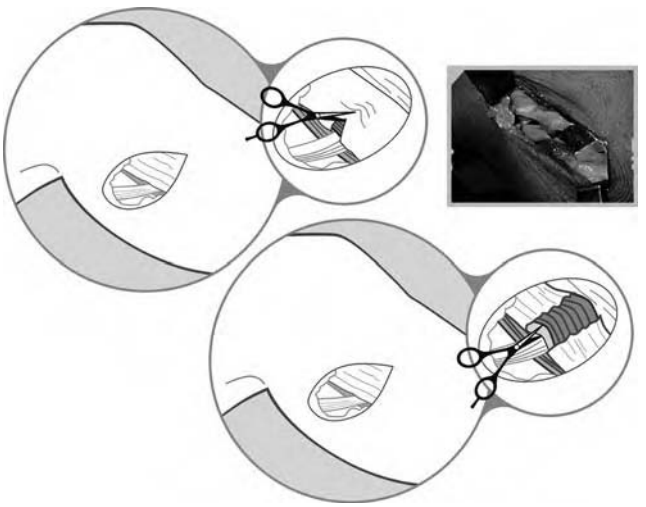

Figure 5 First site of peroneal nerve decompression: superficial and deep fasciae of the peroneus longus muscle.

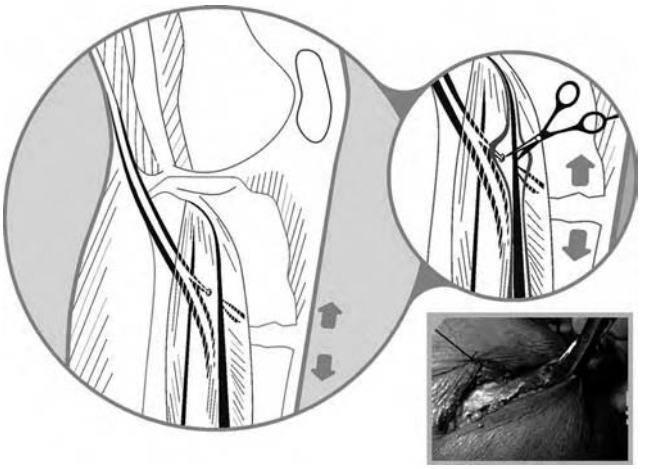

Figure 6 Second site of peroneal nerve decompression: anterior intermuscular septum.

extensor digitorum longus muscles. The septum is isolated and transected from superficial to deep in the leg. It is important to avoid injuring the deep peroneal nerve passing through the septum (Fig. 6).

FUTURE DIRECTIONS

Early detection, by means of noninvasive techniques before clinical signs and symptoms have developed, could improve the lengthening process, patient comfort, and decrease risk of permanent injuries.

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Fractures

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Fractures can lead to residual deformity, increased treatment time, and failure to achieve treatment goals. These patients often require further surgery. By identifying the incidence of fractures, patients can be more informed of the relative risk of this complication. Identification of fracture patterns and risk factors can help assist the surgeon in avoiding this complication.

Fractures can occur through the regenerate bone or remote from the lengthening site during or after lengthening. General complication rates of limb-lengthening procedures have been reported in many publications (1–3) but few have concentrated on fractures (4). Simpson and Kenwright (5) published a series of 180 lengthening segments in 173 patients leading to 17 fractures thus reporting an overall fracture rate of 9.4% per lengthening segment.

O'Carrigan, Paley, and Herzenberg (6) presented a series of 986 lengthening segments in 650 patients treated in the Maryland Center for Limb Lengthening and Reconstruction over a 10-year period. There were 80 fractures in 70 patients for an overall fracture rate of 8% per lengthening segment.

There were higher fracture rates in lengthening of the femur (11%) and the humerus (14%). This reflects the larger lengthenings that were done in the humerus (average lengthening of 90 mm or 50% increase on original length) and the higher deforming soft-tissue forces in the femur often with a stiff knee that increased the lever arm forces (9).

There was a much higher fracture rate for congenital cases, particularly congenital femoral shortening with a 34% fracture rate per lengthening segment. This reflects the relatively large lengthening amounts, relatively small bone diameter, and the significant soft-tissue deforming forces and lever arm forces.

CLINICAL EVALUATION

Some fractures occurred with the frame in situ (15%) (6) and they are usually associated with a fall. The diagnosis is usually quite obvious on history and examination and can be easily confirmed with X ray.

The vast majority of fractures (81%) occur after frame removal and 69% of those within six weeks (6). The presentation can be more subtle in this situation. There can be the typical fracture signs and symptoms or a gradual deformity progression with weight bearing. This occurs through the regenerate and is not always associated with pain. Therefore, careful monitoring of progress with radiographs is important following frame removal particularly during the first six weeks.

The process of frame removal is associated with risk of fracture. Five percent of fractures in our series (6) occurred at the time of frame removal. The risk factors are depth of anesthesia (which may contribute to pain induced muscle spasm), osteoporosis, and joint stiffness. Junior staff are sometimes involved in frame removal and they should be cautioned about the risk of fracture by the supervising surgeon. If applied, casts should have minimal padding and be well molded.

CLASSIFICATION

Simpson and Kenwright (5) presented a fracture classification based on the location of the fracture (Fig. 1).

They identified that the patterns of fracture occurred through the regenerate (Type 1a and 1b) and could be of the greenstick pattern in a regenerate that was still malleable or a complete fracture. The Type 2 was at the host-regenerate junction because this area acted as a stress riser due to differential tissue stiffness. Type 3 was remote from the regenerate and was

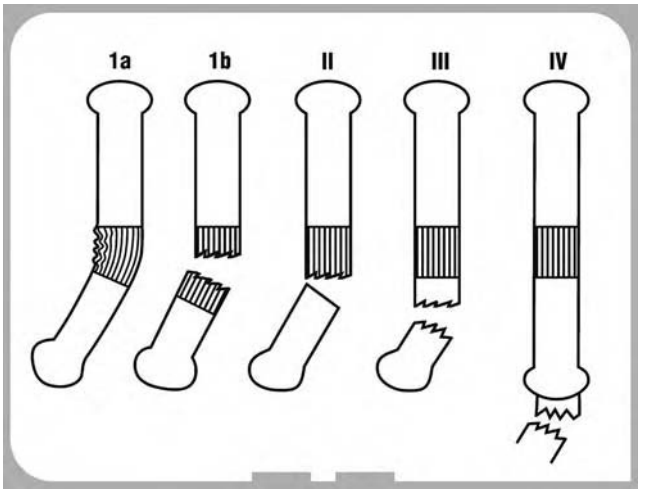

Figure 1 Classification of fractures in lengthening by Simpson and Kenwright.

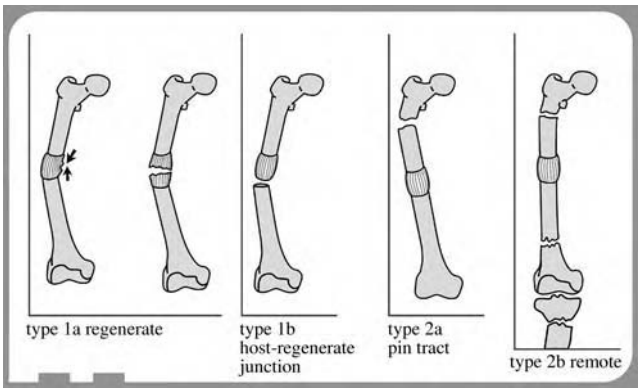

Figure 2 Classification of fractures in lengthening by O'Carrigan, Paley, and Herzenberg.

usually through a pin site and Type 4 were remote from the lengthened limb segment and disuse osteoporosis was implicated as a factor.

Paley and O'Carrigan (6) simplified that classification with type 1 signifying regenerate fractures and type 2 remote fractures (Fig. 2).

LOCATION

In our series (6), 72.5% of fractures occurred in the regenerate and 27.5% remotely.

TREATMENT OPTIONS

Treatment is aimed at achieving fracture healing without deformity or other complications such as infection. There are a number of treatment options available and the individual choice is dependent on the fracture type and location, the patient's preference, and the surgeon's experience.

Whatever technique is chosen, the important principle is that soft-tissue deforming forces and the lever arm effect of joint stiffness above and below the fracture site is neutralized.

Casting

The patient and family are often reluctant to have further invasive surgery and a cast can be attractive to the patient and surgeon because of its relative simplicity. However, this treatment was associated with the highest rate of deformity, defined as greater than 5° of angulation or 1 cm of shortening.

External Fixation

This allows control of soft-tissue deforming forces and joint stiffness and is associated with a lower rate of residual deformity, but, coming after what is often a long and difficult treatment process, it is often the least popular treatment choice for the patient and family.

Intramedullary Nail

This can take the form of flexible small diameter nails or locked intramedullary (IM) nails.

The flexible nails control angulation well but not length.

Locked intramedullary nails control length and alignment well but, if used in the Femur, they are associated with a risk of avascular necrosis in the pediatric population.

The main concern with any form of intramedullary fixation is the risk of infection. None of our seven patients (6) treated with IM nails were complicated by infection but all three patients treated with an IM nail in the Simpson and Kenwright series developed infection (4) and it was felt to be a significant risk. The most likely source of infection is the pin tracts.

Plate

Only one patient in our series and four in the Simpson and Kenwright series had open reduction and internal fixation. It is a possible solution and should be considered in the light

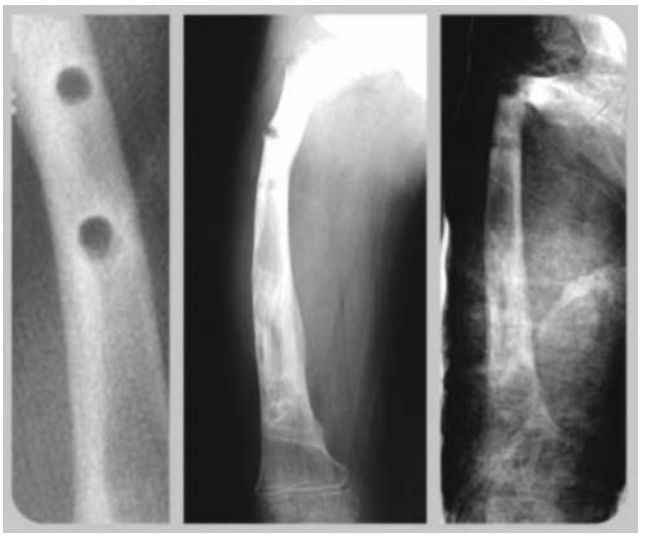

Figure 3 Fracture through eccentric pin hole.

of previously discussed factors. The new generation of minimal incision locked plates with a targeting devise may be helpful, particularly in the distal femur or proximal tibia.

STRATEGIES/PRECAUTIONS

Surgical Techniques

Eccentric pinholes should be avoided if possible. In our series, 60% of the fractures occurring at a pinhole were associated with eccentric pins. Use of cannulated drills after the placement of the wire is confirmed on fluoroscopy can help avoid eccentric placement of pins (Fig. 3).

Pin placement should span the length of the limb segment to maximize control and frame stability. One should avoid subtrochanteric pin placement in the femur because this is a significant stress riser and was associated with fracture on more than one occasion (Fig. 4).

Stiff Joints

Joint stiffness to some degree is almost invariably present at the end of limb lengthening. In the lower limb, knee stiffness increases the lever arm forces and this increases the risk of fracture.

All efforts should be made to minimize joint stiffness during the lengthening and consolidation phase. If it is present, then this should be taken into account when considering

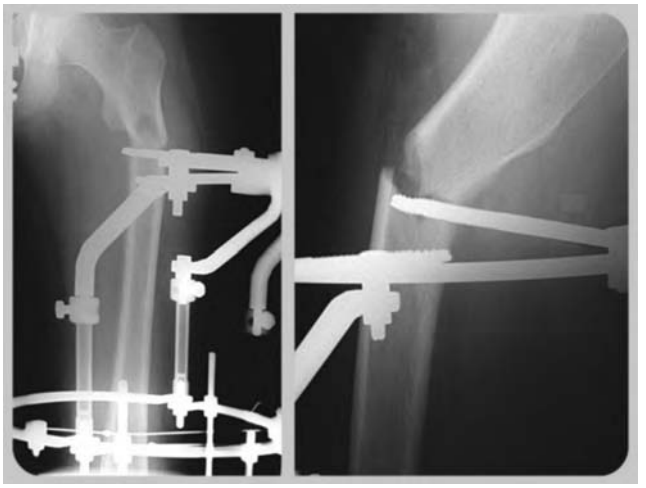

Figure 4 Fracture in the proximal half pin.

postremoval bracing or casting. Selected soft-tissue releases can facilitate the restoration of joint range of motion.

Staged Removal

It is always wise to maximize the strength of bone before frame removal. One of the ways to do this is dynamizing the frame by taking out parts and connections of the frame, reducing rigidity of construction, and reducing the risk of fractures after frame removal.

Prophylactic Internal Fixation

Prophylactic internal fixation should be considered if there is a high risk of fracture (e.g., hypotrophic regenerate or marked joint stiffness). This is particularly true of congenital lower-limb lengthening with fracture rates of 34% per lengthening segment (6) in congenital shortening of the femur patients.

Shortening and angular deformities are common sequelae secondary to fractures, due to fracture regenerate malformation or fracture malunion. The psychological distress experienced by the patient, their family, and the treating surgeon should not be underestimated and the best treatment is prevention if possible.

FUTURE DIRECTIONS

There may be a role for bisphosphonates with limb lengthening and this is thoroughly discussed by Dr. David Little in a previous chapter.

The role of prophylactic internal fixation has yet to be clarified, with the main concern being infection.

Hydroxyapatite (HA)-coated pins (7) reduce the incidence of loosening and pin-tract infection and, therefore, allow functional weight bearing earlier thereby promoting regenerate maturation.

Intramedullary lengthening devices such as the ISKD (Orthofix) (8) may have a role in reducing joint stiffness traditionally associated with lengthening and the risk of fracture because the implant is not removed until full healing has been achieved. The results at this stage are only preliminary.

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Muscle Contractures

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Muscle contractures are often responsible for poor functional results during and after limb lengthening, and should be prevented and aggressively treated to preserve adequate range of motion and normal gait (1–3).

If not detected, they can also cause subluxation and dislocation of joints with severe compromise of the functional outcome. When not treated, the muscle contractures can lead to joint stiffness and degeneration.

CAUSES OF SOFT-TISSUE CONTRACTURES

Contractures occur when elastic tissues and contractile elements cannot accommodate changes in length. Because myofascial tissue resists elongation more then other tissues, it is most prone to contractures. In such cases, a relative shortening of a muscle that bypasses two joints is often the cause. Muscle contractures tend to occur in the overpowering muscle group, due to the imbalance of strength between flexors and extensors. In addition, some muscles, as the gastrocnemius, develop passive tension much more rapidly in response to limb lengthening's passive stretching (Fig. 1).

These muscles are strong and have more muscle mass compared to the anterior muscles of the leg. They will tend to flex the knee and plantarflex the ankle rather then lengthen if they are left unopposed (4). In lengthening of the femur, the hamstrings are the largest and bulkiest muscle group, and for this reason, posterior subluxation of the knee can occur. Ligament instability, common in patients with congenital shortening, is also a predisposing factor to subluxation during lengthening (Fig. 2).

The quadriceps muscle also crosses two joints, and that is why lengthening of the femur can cause difficulties with knee flexion leading to knee extension contractures (Fig. 3).

Animal studies show that in spite of muscle fiber adaptation by sarcomere addition (5,6), the fiber length increases more than the sarcomere length, concluding that this insufficient sarcomere production, especially in the posterior muscles, is related to the development of equinus contractures (6–8).

Biomechanical studies show that even with those intrinsic changes, the muscle fibers can maintain many of their biomechanical parameters, and indicate that those isolated changes were not alone responsible for contractures (6,9–11).

There are three factors responsible for weakness of the lengthened muscles:

1. Transitory muscle denervation (as shown in eletrophysiologic studies)
2. Decreased fiber cross section due to the increase of fiber length after limb lengthening
3. Pain, and decreased motion of the muscle

CONNECTIVE TISSUE ROLE IN CONTRACTURES

Connective tissue does not stretch or lengthen as well as the contractile elements of muscle fibers. The fasciae and septae do not lengthen well and often are an obstacle to lengthening. It was shown by the histology of stretched muscle that distracted muscles have increased deposition of collagen type III, and presence of a dense perimysial weave surrounding the distracted muscle fibers. Prolonged stretch may lead to damage to the perimysial and endomysial network, with subsequent fibrosis and loss of muscle compliance (8,12,13). Those changes in the connective tissue could account for the increased stiffness demonstrated by physiological measurements.

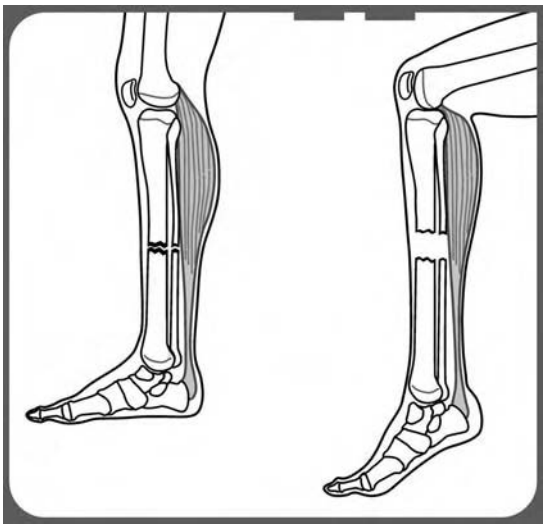

Figure 1 Usual contractures in leg lengthening: knee flexion and equinus of the ankle.

PROPHYLAXIS AND TREATMENT

Soft-tissue contractures during lengthening are often related to pain, weakness, and problems in rehabilitation. They could be difficult to treat and it is important to avoid them, by following certain technical aspects in external fixator application, and prophylactic therapy (4,14).

Application of External Fixation

When applying an external fixator using half pins and wires, muscular planes should be avoided. For example, in the distal femur, in order to preserve the quadriceps muscle function, half pins should be inserted in the intermuscular planes. The best configuration is delta with a medial half pin and a lateral half pin from posterior to anterior, 60° in the horizontal plane of the knee (15) (Fig. 4).

Physical Therapy

Physical therapy in a customized and intense schedule has an important role to prevent development of contractures (Fig. 5). Hydrotherapy is very beneficial in facilitating joint motion using muscle relaxation and exercises with reduced weight bearing.

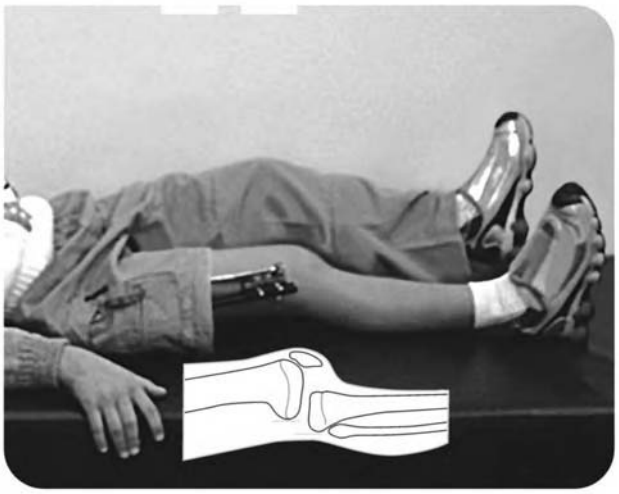

Figure 2 Posterior knee subluxation.

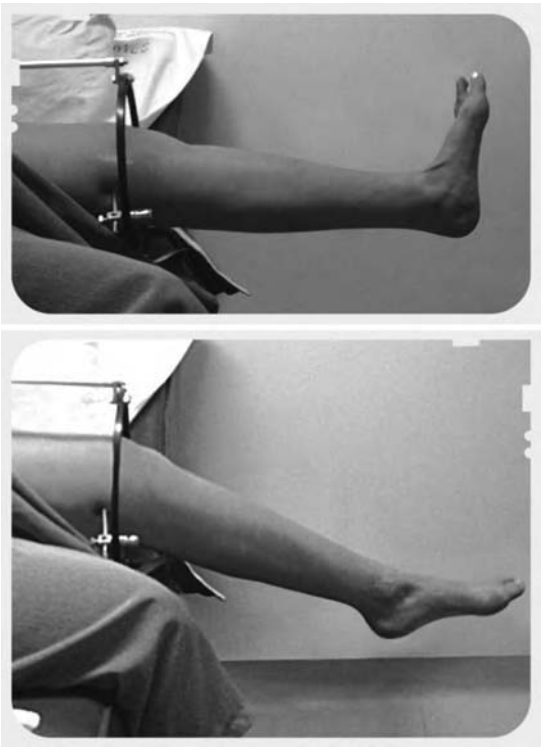

Figure 3 Extension contracture in femur lengthening.

Splints

Splinting is also a very good approach to prevent and treat muscle contractures. It helps to maintain position gained in therapy sessions. After surgery, if the feet are not included in the fixation, they must be splinted in neutral position to avoid equinus contractures (Fig. 7). If mild contractures are detected, the rate of lengthening can be decreased, and the therapy protocol must be adjusted to emphasize gaining back lost degrees of range of motion.

The principle of splinting is to place the muscle under tension for as many hours as possible (9,12,16).

Dynamic splints are very useful by keeping the desired position of the joint and allowing passive stretching of the contracted muscles as well as active movement of joints and more comfort. They can be applied in intermittent time intervals, depending on the severity of contracture and patient tolerance (17–19).

Figure 4 Delta configuration in distal femur fixation.

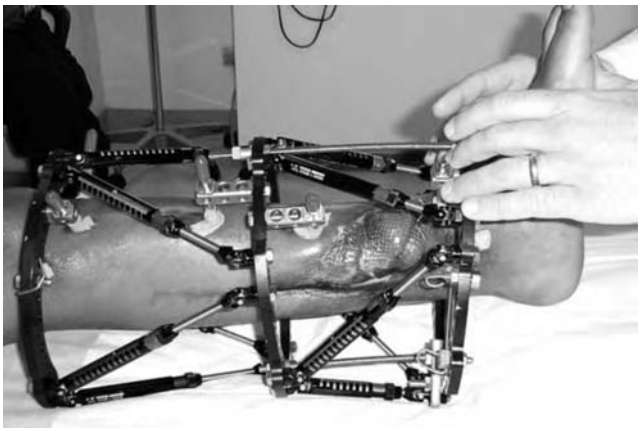

Figure 5 Physical therapy showing knee extension and passive dorsiflexion exercises of the ankle to prevent equines contracture.

Hinges could be applied and connected to an external attachment to the thigh and leg (with or without external fixators, by casts or external fixators parts), for the lower limb, and upper and lower arm in elbow contractures. Hinges can be spring-loaded, or passive. Passive hinges depend on some other attachments (for example, elastic bands) to give the mechanical tension in flexion or extension. Hinges can also be placed strategically anterior to reduce a posterior subluxation of the knee. The Taylor Spatial Frame (Smith and Nephew Inc., Memphis, Tennessee, U.S.A.) is particularly useful to help correct subluxation of the knee.

Another example of dynamic splinting is used to avoid flexion contracture of toes during tibial lengthening. Elastic bandages are applied to the toes and connected to a dorsal rod. They allow active motion of the toes preventing claw deformities (4) (Fig. 6).

Splints can be used either with external or internal lengthening devices.

SURGICAL TREATMENT OF CONTRACTURES

Muscles and fasciae can be surgically released, if the regular therapy protocol and splinting are not able to treat the contracture successfully. An equines contracture of the ankle that develops during tibial lengthening is usually related to contracture of the gastrocnemius. This is confirmed when the equinus corrects with flexion of the knee. Gastrocnemius recession surgery will correct the contracture. This is accomplished by releasing the gastrocnemius fascia at the junction of the middle and distal third of the leg through a posterior approach.

Extension contracture of the knee that occurs during femoral lengthening can be corrected through a limited Quadricepsplasty (20). A 5 cm anterolateral incision at the junction of the middle and distal third of the thigh is used. The iliotibial band and anterior fascia of the thigh is transversely incised. The vastus lateralis muscle is longitudinally split and the vastus intermedius is exposed. This is noted to be predominantly fascia. This vastus intermedius fascia is incised transversely. A gentle manipulation under anesthesia leads to significant increase in knee flexion. This has led to quick return of knee flexion without loss of strength and without extensor lag.

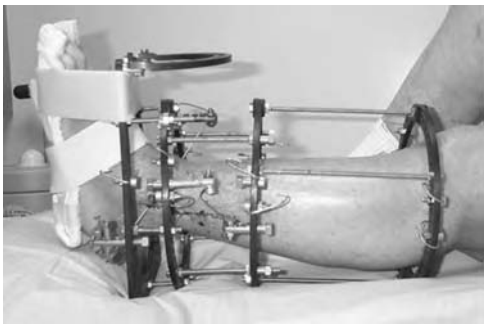

Figure 6 Avoiding equinus contracture of the ankle.

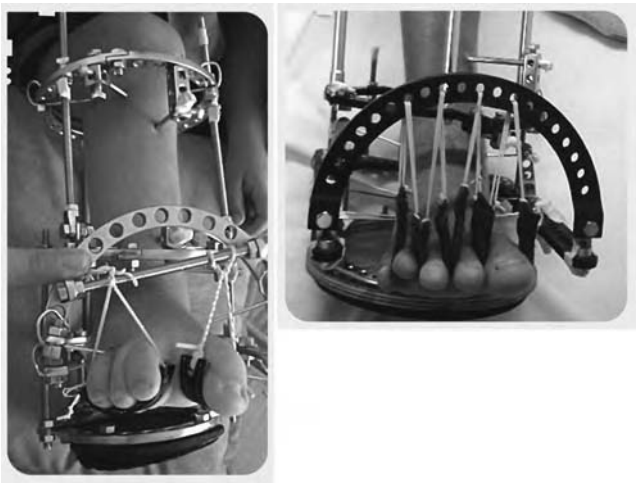

Figure 7 Elastic bandages applied to the toes and connected to a dorsal rod.

Another surgical solution is to bypass the joint with the external fixator, and distract it, correcting the contracture. Major contractures can be treated with muscle lengthening and capsule releases (4).

Internal lengthening devices could require extending the fixation across the joints as well. For example, this can be done by placing a screw through the calcaneous to the posterior tibia, to avoid equinus contracture of the ankle during tibial lengthening.

FUTURE DIRECTIONS

Development of muscle contractures could be better avoided in the future by a better understanding of soft-tissue structure, physiology, and behavior of those tissues during lengthening.

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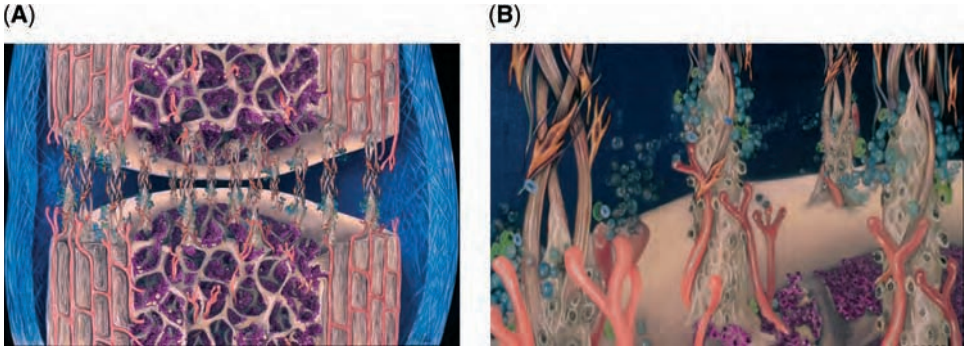


Figure 2.8 (**A**) Artist rendition of histological findings demonstrates multiple bone forming units (BFUs) bridging the entire cross-section of the two distraction surfaces. Note the central collagenous zone of each BFU, where strain energy is absorbed following each distraction. This drawing demonstrates why the overall distraction force (load) is proportional to the number of BFUs (cross-section of bone formation) and indirectly related to the length of unmineralized collagen. (**B**) Closer examination of the BFUs reveals massive zones of proliferating precursor cells, which migrate from local bone marrow and periosteum, concentrating on the tips of each new microcolumn at the zone of primary matrix formation. *Source*: Courtesy of Ron Tribbel. *(See p. 39)*

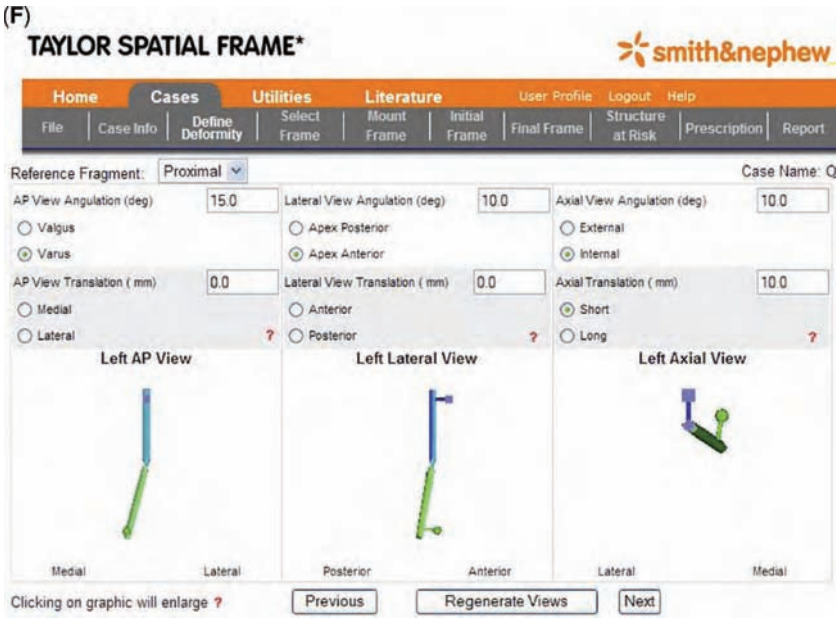
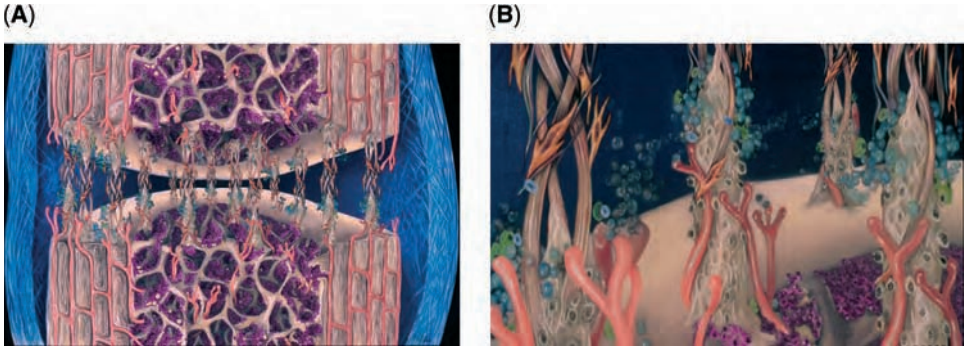


Figure 23.1 Case example of 37-year-old male with medial compartment arthritis after a medial plateau fracture. (**F**) The deformity data is entered into the web-based computer program and an image is generated recreating the deformity. (With reference to the stick diagrams, the blue box represents the knees and the green circle the foot.) *(See p. 316)*



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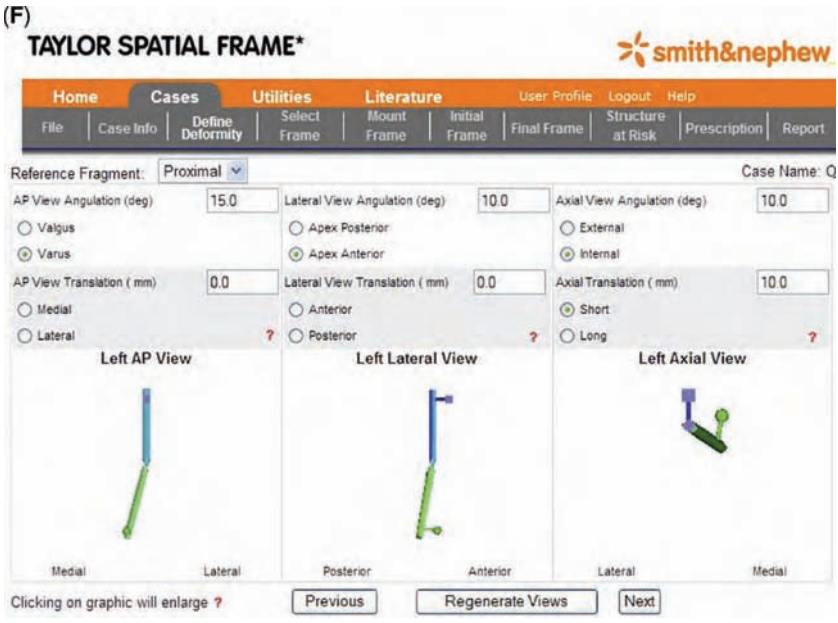

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Orthopedics

about the book . . .

An illustrative and in-depth overview of the many available applications and techniques for limb lengthening and reconstruction, this guide provides step-by-step details on the latest surgical procedures for the correction of limb deformities due to congenital defects, growth disturbances, infection, and trauma in both children and adults. Supplying effective surgical approaches, technical tips, methods to manage complications, and clinical case studies in each chapter, this guide will be a constant companion for all orthopedic, reconstructive, pediatric, foot, ankle, upper extremity, tumor, and trauma surgeons.

Covering the gamut of new technologies and advancements related to limb lengthening and reconstruction, this source offers authoritative contributions from key opinion leaders on acute trauma; post-traumatic reconstruction; tumors; stature lengthening; and post-operative care...provides expert surgical approaches for the hip, knee, foot, and ankle...stands as a clear compendium of treatment guidelines for practitioners specializing in limb lengthening, reconstruction, and deformity correction...includes tables outlining the authors' classification and treatment recommendations...contains real-world technical tips and methods for complication management and avoidance, as well as an abundance of clinical case examples...and organizes each chapter to include principles, a practical approach to each subject, and a review of the recent literature.

about the editors . . .

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Printed in the United States of America

informa
healthcare
www.informa.com

270 Madison Avenue
New York, NY 10016

2 Park Square, Milton Park
Abingdon, Oxon OX14 4RN, UK

DK051X
ISBN 0-8493-4051-9
90034
9 780849 340512