Plastic and Reconstructive Surgery

Wei Wang
Jianmin Yao
Editors

Congenital Deformities of the Hand and Upper Limb

Springer

Plastic and Reconstructive Surgery

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Congenital Deformities of the Hand and Upper Limb

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Profile of Chief Editors of *Congenital Deformities of Hands and Upper Limbs*

Wei Wang Wang Wei is a tenured professor of plastic surgery and one of the founders of the Chinese Plastic and Reconstructive Surgery Association and Plastic Surgery Branch of the Chinese Medical Doctor Association as well as an international editorial board member of *Plastic and Reconstructive Surgery*. He has successively served as chairman, vice chairman, member of standing committee, associate editor, and editorial board member of over 30 associations and journals at home and abroad.

He started his career in plastic surgery with a master's degree in 1961. When he was a senior in college, he was elected as a physician of the Cardiology Department of Shanghai Ruijin Hospital and has been in charge of 26 sickbeds for half a year. From1981 to 1982, he was a visiting scholar and visiting professor at Baylor College of Medicine in the United States. Since 1967, he has served as the discipline leader, deputy director (executive) and director, and leader of Shanghai's key medical disciplines as well as an academic leader of the "211 Project" for the Ministry of Education.

He reported and published over 40 leading achievements at home and abroad. For example, in 1965, he reported *An Experimental Study on Free Replantation of Skin Tissues*. In 1975, he applied the dorsal island flap grafting and free grafting. In 1977, he performed cervical esophageal reconstruction with a free jejunum flap and a jejunum-esophagus grafting as well as cervical and thoracic esophageal reconstruction with a proximal jejunal pedicle and a distal anastomosis grafting. In 1985, he was hailed as the "world's most experienced doctor in esophageal reconstruction with intestinal grafting" by American scholars. In 1990, he created cervical esophageal reconstruction with the pectoral musculoskin flap grafting. In 1991, he created cervical esophageal reconstruction with the tubular latissimus dorsi flap. In 1977, he expanded this technique to second-toe transplantation. In 1979, he created a treatment for postburn claw hand with the superficial temporal fascia flap and skin grafting. That year, he also performed island skin flap grafting in the medial plantar and of the metatarsophalangeal joint flap grafting with a 0.8 cm × 5.0 cm area of skin, nerves, and blood vessels, which were used for hand or temporomandibular joint reconstruction. In 1980, he and Yang Guofan reported a free forearm flap grafting. In 1980, he proposed in *Medical Encyclopedia* that the free forearm flap grafting be used for cervical esophageal or penile reconstruction, creating the grafting of the reversed forearm island flap. In 1982, he reported the etiology and classification of lymphedema. In the same year, he created the treatment of traumatic anal sphincter reconstruction with the greatest gluteal muscle flap grafting and subsequently used it for anal sphincter reconstruction to treat colorectal cancer in situ. In 1985, he created Y-shaped microvascular anastomosis method. In 1989, he reported the treatment for advanced facial nerve paralysis with the grafting of free latissimus dorsi muscle (Phase I). In 1995, he performed treatment for advanced facial nerve paralysis (Phase I) with the grafting of the latissimus dorsi muscle free flap. He has many innovative achievements in classification of thumb dysplasia, aesthetic reconstruction, modern

abdominoplasty, augmentation mammoplasty with prosthetics, facial contour beautification, eyelid and nasal surgery, and other fields. Since 1984, he has been engaged in the practice of "plastic surgery" and "cosmetic surgery."

He prepared the *Rules for Training Chinese Orthopedic Surgeons* and *Rules for the Scope and Classification of Plastic Surgeries* for the competent department. The majority of surgeons under his guidance have become academic leaders of provinces and municipalities. He also trained over 20 professors and surgeons for the United States, the United Kingdom, Italy, and other countries.

He has won the National Invention Award for over 20 times. He is listed in the *History of Microsurgery* and *Who's Who* as a medical celebrity.

Jianmin Yao Yao Jianmin, a chief physician, was awarded the title "Top 10 Young Doctors" in Hangzhou in 1991. In 1982, he graduated from the Hangzhou Campus of Zhejiang Medical University. Currently, he is an academic leader in Hangzhou's medical hand surgery key specialties, a standing committee member of the Hand Surgery Branch for Zhejiang Medical Association, a member of the Plastic Surgery Branch of Zhejiang Medical Association, vice chairman of the Limb Function Reconstruction Professional Committee for Zhejiang Provincial Rehabilitation Medicine Association, and the seventh and eighth special editorial member of the *Chinese Journal of Microsurgery*.

He has published 49 academic papers, including 11 SCI papers. In 1997, he published the "Treatment of Simple Syndactyly by Reverse Insertion Method of Webbed Fascia Pedicle Skin Flap" in the journal *Plastic and Reconstructive Surgery*. Currently, he has published 22 papers in first-class Chinese journals and12 papers that won awards. He has won first, second, and third prizes via his excellent papers at municipal, provincial, and national meetings.

His research "Breast Splitters for Mammoplasty" won a national patent. He completed nine research projects independently or by cooperating with others, which have reached a leading level at home and abroad. In addition, he has won the first, second, and third prizes of science and technology awards at the municipal level, at the provincial level, and of the Ministry of Health, respectively. In 1995, the project *Repair of Acute Thumb Avulsion with Bi-lobed Island Flap of the Second Web* won the third prize of the Zhejiang Provincial Science and Technology Progress Award. In 1997, the project *Microanatomy Research and Clinical Applications of Hand and Forearm Island Flaps* won the third prize of the Ministry of Health and the first prize of Hangzhou Science and Technology Progress Award. In 2006, he compiled the *Atlas of Clinical Surgery for Flap Repair of Hand and Foot Wounds.* In 1997, he studied at the Beckman Laser Institute & Medical Clinic of the University of California in the United States. In September 2007, he went to Luisenhospital Aachen in Germany for an exchange program. In 1997, he was listed as a third-level talent in Hangzhou Talent Education Training.

Editorial Board of *Congenital Deformities of Hands and Upper Limbs*

Preface

Congenital anomaly of the hand and upper limb is a common disease. Patients with such a disease can pay a visit to a department of plastic surgery, department of pediatric surgery, department of hand surgery, department of orthopedic surgery, or department of general surgery. Therefore, it is necessary for healthcare providers to know and be familiar with this type of disease as well as its treatment.

In 1986, in the first National Symposium and Workshop of Hand Surgery held in Qingdao, the author presented the report *Classification of Congenital Hand and Upper Limb Deformities*. Thereafter, lectures on congenital deformities of the hand and upper limbs were delivered in Shanghai, Guangzhou, Beijing, Wuhan, and other places. In *Huang Jiasi Surgery* and *Plastic Surgery*, of which the author is the chief editor (1999), relevant chapters on congenital deformities of the hand and upper limbs were written. In 2004, with the assistance of the author, Professor Hong Guangxiang compiled and published *Congenital Deformities of Hands.* The book *Congenital Deformities of Hands and Upper Limbs* has been written and published based on them.

The author had the honor to invite professors and scholars in plastic surgery, hand surgery, and orthopedics from North and South China as well as in the Hand Surgery Center at University of Louisville Hospital to participate in the composition and compilation of *Congenital Deformities of the Hand and Upper Limb.* It took more than 2 years to finish the book.

During the compilation process, the authors summarized their decades of experiences and reviewed hundreds of thousands of pictures of hand deformities. In addition, they referred to over a thousand articles and books on congenital deformities of hands and upper limbs published at home and abroad.

In clinical practice, there appears to be many malpractice cases in the treatment of congenital deformities of hands and upper limbs, most of which are caused by a lack of understanding of the morphological, structural, and functional defects of hand deformities as well as inappropriate selection of treatment measures. Therefore, only when the morphological, structural, and functional defects of hand deformities are fully understood can the best therapeutic regimen be generated. To this end, hand examination method and hand function assessment method have been added to this book. Professor Tsai from the Hand Surgery Center of University of Louisville Hospital in the United States actively participated in the compilation of this book, for which the author is deeply grateful.

Although every effort has been made to compose and compile this book, shortcomings are inevitable, so any feedback is welcome.

Shanghai, China
October 18, 2014

Wei Wang

Acknowledgments

During the compilation of *Congenital Deformities of Hands and Upper Limbs*, the editors have contacted professors from famous hand surgery centers in the United States for many times, and Professor Tsai and Professor Tien from US Kleinert Kutz Hand Surgery Center have taken an active part in our compilation and made amendments and supplements to relevant chapters. We are very glad to have American peers participate in the compilation of this book, and they will be introduced as follows.

It has been over 50 years since US Kleinert Kutz Hand Surgery Center was founded, and it has a large number of famous surgeons and scholars in hand surgery. This center is currently one of the world's largest training bases of hand surgeons. From 1960 to now, a total of over 1200 surgeons from 58 countries have received professional training in the field of hand surgery here. This center owns medical bases in Louisville and Lexington in Kentucky and New Albany in Indiana, and it provides comprehensive medical services in upper limbs for patients from all over the world. During the past over five decades, Kleinert Kutz Hand Surgery Center has performed seven allogenic hand transplantations and the world's first batch of hand replantations. It has been the cutting edge in the field of free tissue transplantation and reconstruction; its research in peripheral nerve blood supply has won national honors; it is the first to report finger arterial repair, bilateral upper arm replantation, bilateral forearm replantation, and vascularized epiphysis transplantation; and it has successfully created a set of techniques in flexor tendon repair and rehabilitation.

Tsu-Min Tsai (M.D.) is a renowned specialist in hand surgery and microsurgery. Dr. Tsai once performed replantation of amputated finger for Armstrong, the one who first landed on the moon. Dr. Tsai graduated from National Taiwan University in 1961 and received the training of National Taiwan University and University of Louisville Hospital. Dr. Tsai acted as chief of surgery in Taipei Peace Hospital from 1970 to 1975. In 1976, Dr. Tsai received training at Kleinert Kutz Hand Surgery Center. Currently, Dr. Tsai is the clinical professor in orthopedics in University of Louisville Hospital, a member of the American Society for Reconstructive Microsurgery, a member of the World Society for Reconstructive Microsurgery, and an official member of the American Association for Hand Surgery. Dr. Tsai has been devoted to the clinical and research work in the field of hand surgery and microsurgery and has published a total of over 130 articles.

Huey Y. Tien (M.D.) is a renowned specialist in hand surgery and microsurgery and has treated patients with injuries in wrist joint and brachial plexus from all over the world. Dr. Tien graduated from Chung Shan Medical College in 1986. He completed the training for resident orthopedist in Taoyuan Armed Forces General Hospital and the training for resident surgeon in University of Louisville Hospital. In the last year, Dr. Tien acted as the chief resident and subsequently received specialized training in hand surgery in Kleinert Kutz Hand Surgery Center. Currently, Dr. Tien is clinical associate professor of University of Louisville Hospital, official member of the American Association for Hand Surgery and official member of the Association for the Study of Internal Fixation, and also director of Surgeon Training Department of Kleinert Kutz Hand Surgery Center.

Special thanks should go to Dr. Han Dong from the Department of Plastic Surgery of Shanghai Ninth People's Hospital, Shanghai Jiaotong University School of Medicine, who has made great efforts in the compilation of this book during his study in Kleinert Kutz Hand Surgery Center. Thanks also go to Di Jing, associate professor of Anatomy Teaching and Research Office of China Medical University for the charting in relevant chapters.

Jinghong Xu

July 2014

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1 Embryonic Auxanology, Etiology, and Pathology of Congenital Deformities of the Hands and Upper Limbs

Jinghong Xu, Jialiang Chen, Wei Wang, Bin Wang, Yijia Yu, Bo Chen, and Jianmin Yao

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1.1 Embryonic Development of the Hands and Upper Limbs

Jinghong Xu, Jialiang Chen, Wei Wang, and Bin Wang

Embryonic development period is a period starting from fertilization of eggs to the formation of main structure of the body, as for human beings, the 8 weeks from postfertilization to embryogenesis. The development period of limbs is basically the same as that of other human organs, and its duration is from the fourth to eighth week of embryogenesis.

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1.1.1 Formation and Differentiation of the Body Axis

Limb development is a three-dimensional process that occurs in the proximal-distal axis, front-rear axis, and dorsal-ventral axis. During the early period of embryonic development, homeobox (HOX) transcription factor mediates the skull-tail axis to start the differentiation of somites [1–3]. Around the fourth week, the upper limb germinal area is established; the expressions of T-box (TBX5), wingless-type MMTV (WNT), and fibroblast growth factor (FGF) are launched; and the body begins to grow, and the upper limb bud covered with the surface layer of dermal layer bulges from lateral plate mesoderm. The failure of limb bud induction (tetra-amelia syndrome, limb absence) is correlated with WNT3 and FGF10 mutations [4–8]. TBX4 and TBX5 are correlated with development of lower limbs and upper limbs, respectively. TBX5 mutation (Holt-Oram syndrome, hand-heart malformation syndrome) will lead to a series of upper limb deformities. With the formation of the limb bud, its development is along the three axis directions: proximal-distal axis, front-rear (radius-ulna) axis, and the dorsal-ventral axis (Fig. 1.1).

1.1.1.1 Proximal-Distal Axis

The development and differentiation of each axis is controlled by a cluster of cells, which send development message to local tissues and cells that are called "signal center." The FGF10 in mesoblast is connected with radical fringe gene (RFNG) in ectoderm at the apex of the dorsal-ventral border, which enables ectoderm to get thickened to form a signal center of proximal-distal axis which is called "apical ectodermal ridge (AER)" [9–12]. AER can produce WNT3 and some FGFs (FGF4, FGF8, FGF9, and FGF17) to maintain the expression of FGF10 in mesoblast. The FGF10 may promote the cell proliferation in sub-AER areas, and these areas are called progress zones. The mesoderm cells are regulated by the signal center to determine the final differentiation of limb bud. The interaction between FGF and WNT of ectoderm and mesoderm maintains the growth and development of the proximal-distal axis. In humans, there are four HOX gene clusters, labeled as A, B, C, and D, respectively; in rodents, similar genes are replaced by lowercase letters (hoxa 11). For the developing limbs, HOX gene family is expressed sequentially in a complex overlapping manner, that is, expression from near to far in the natural order. Expression of HOX9 starts when the humerus appears, and HOX11 expression is in the forearm region, HOX12 expression in the wrist area, and HOX13 expression in hand. In mice, hoxa 11 or hoxd 11 gene knockout has no significant impact on the development of limbs, and in this way, some excessive structures exist in the gene system; however, hoxa 11 or hoxd 11 gene knockout in a mouse strain will cause the absence of the radius and ulna. The cascade expression of

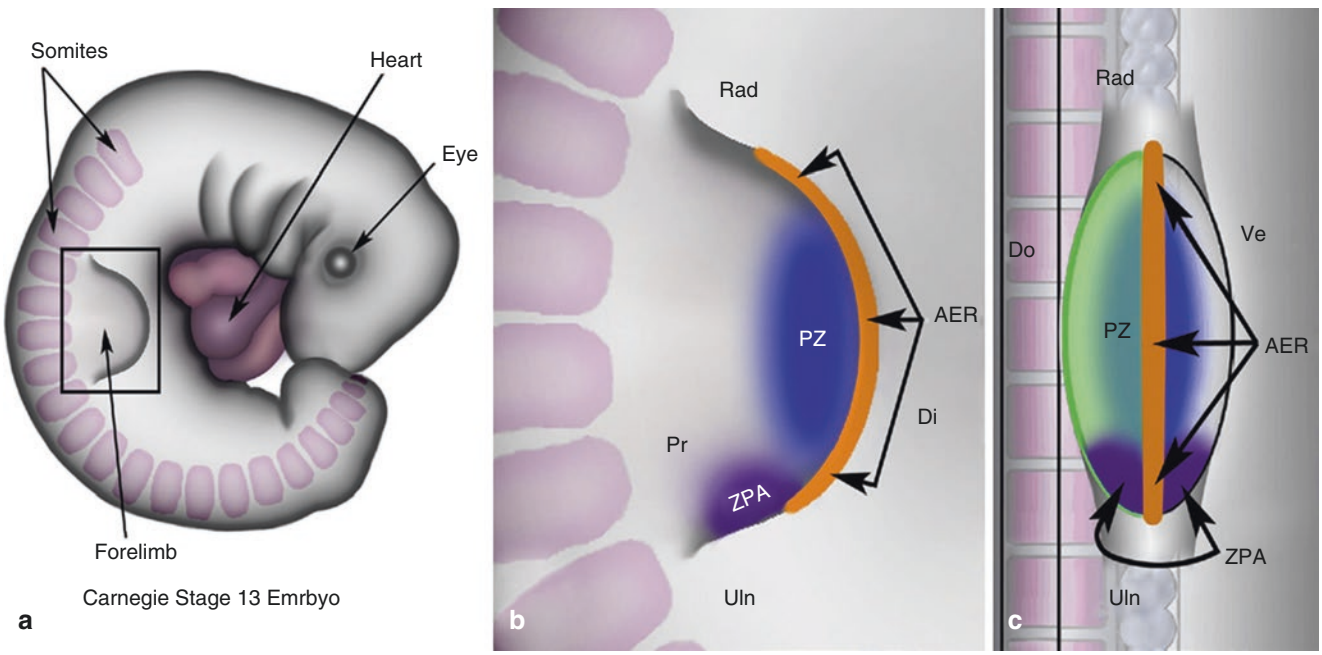

Fig. 1.1 Limb bud regulation center and body axis (Originate from Sekine K, Ohuchi H, Fujiwara M, Yamasaki M, Yoshizawa T, Sato T, et al. Fgf10 is essential for limb and lung formation. Nat Genet 1999;21:138–141) (Reprint with permission from NATURE PUBLISHING GROUP LICENSE TERMS AND CONDITIONS)

HOX gene is partially regulated by FGF secreted by the apex ectodermal ridge and also affected by sonic hedgehog (SHH) signal path [13, 14].

1.1.1.2 Front-Rear Axis

Studies have shown that the development position of hand plate will change from front axis to rear axis when a small area of the rear axis of limb buds is transplanted to the front axis, so that mirror hand is formed. Under the influence of the rear-axis tissue polarization, the front axis of limb bud becomes rear-axis zone of polarizing activity (ZPA). The development and differentiation of the front-rear (radial-ulnar) axis is controlled by the ZPA behind the mesoderm. ZPA can increase the width of the limbs, making it develop backward (ulna). One of the early active factors in ZPA is possibly the vitamin A acid, because the transplantation of vitamin A acid can replicate the effect of ZPA transplantation; however, subsequent studies have shown that vitamin A acid cannot play a biological role in the normal development of limb. ZPA activity is achieved by morphogen SHH [15]. AER is closely linked with ZPA through a feedback loop to maintain the expression of SHH in the rear (ulna) border of distal AER in the growth progress (Fig. 1.2).

1.1.1.3 Dorsal-Ventral Axis

Early limb bud is clearly divided into two parts: dorsal part and ventral part. There is no cell that crosses dorsal and ventral border during development process. The ectoderm covering the limb bud has a certain influence on the interface of dorsal-ventral axis, because cutting and separating of the ectoderm and the dorsal-ventral transposition will lead to reversion of dorsal and ventral parts during limb bud development. Engrailed 1 (En1) protein expression is strictly confined to the ventral chamber. En1 is controlled by one or more bone morphogenetic proteins (BMP), which produce the biological effects through En1. En1 inhibits the activity of WNT7a in ventral chamber. WNT7a makes the mesoderm of the lower limb grow toward the dorsal side by inducing Lim homeobox isogenous transcription factor LMX1B; therefore, the WNT7a deficiency will cause the growth and development disorder of the limbs at the ulnar side. It also suggests another important role of WNT7a of maintaining the production of SHH associated with ZPA [16, 17].

Signal center is also able to regulate the generation of downstream target tissues such as bone, blood vessels, muscles, and nerves through conventional and special, asymmetrical molecular pathways. For example, the generation of bones requires several factors to play regulatory roles at the right time and location, including high mobility group protein9 (SOX9) associated with the Y in sex-determining region, which makes the bone primordial concentrate. WNTs and growth differentiation factor5 (GDF5) regulate the development of joint; parathyroid hormone (PTHLH), Indian hedgehog (IHH), insulin-like growth factor (IGF), bone morphogenetic proteins (BMP), WNTs, FGF, and osteoblast-specific transcription factor2 (RUNX2) will promote the growth of bone primordial and the subsequent endochondral ossification. In addition, short homeobox gene2 (SHOX2) gets upregulated in the cartilaginous membrane of proximal side, promoting the extension of the humerus; at the same time, the forearm cartilaginous membrane induces SHOX to regulate the growth of radius and ulna. Therefore, the correct induction of downstream path is essential to the full differentiation in each axial direction.

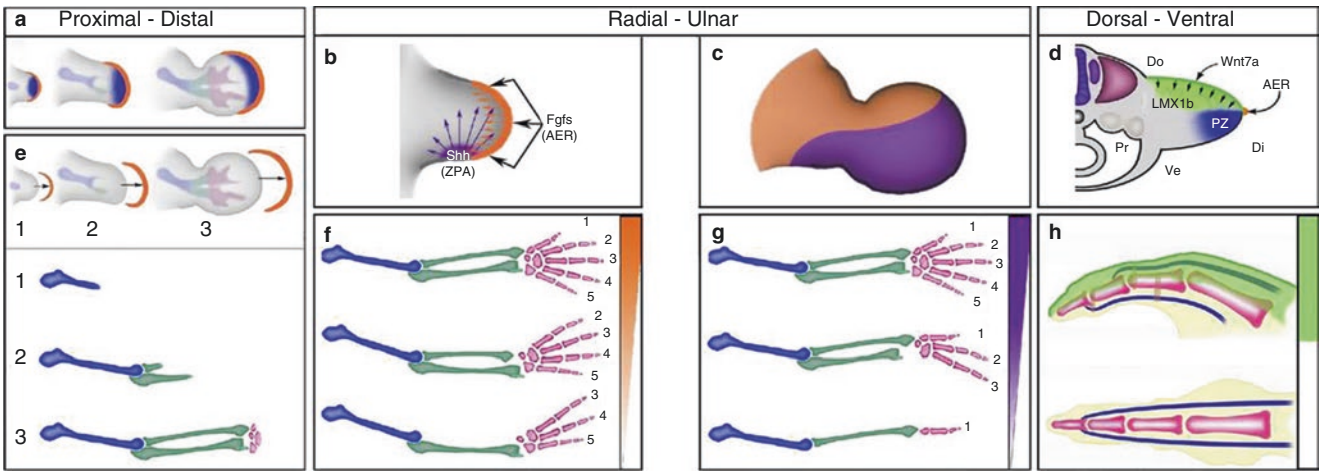

Fig. 1.2 Axial differentiation and developmental disorder (Originate from Saunders JW. The proximo-distal sequence of origin of the parts of the chick wing and the role of ectoderm. J Exp Zool 1948: 108: 363–403) (Reprint with permission from JOHN WILEY AND SONS LICENSE TERMS AND CONDITIONS)

1.1.2 Limb Bud Formation

At 26 days of development, segment, corresponding to the first cervical vertebra at 5–8, a mesodermal ridge appears at the ventrolateral wall of both embryonic sides and near the root of the neck segment, and it is covered with the ectoderm. This is just the upper limb buds, which is the famous Wolff top (Fig. 1.3); at 28 days, upper limb buds are clearly visible (Fig. 1.4).

The limb bud consists of mesoblastic mesenchymal tissues and one ectodermic layer on their surface. In the early stage when the limb bud appears, the apical ectodermal ridge is formed by the ectoderm at the top of limb buds. Many studies have shown that apical ectodermal ridge directly influences and controls the development and differentiation of limbs. Some earliest limb deformities are caused by incomplete differentiation or injuries of apical ectodermal ridge.

At 28–30 days of development, the upper limb bud becomes thickened and bends toward the body side (Fig. 1.6); At 31–32 days, the cylindrical proximal portion and the flat distal portion can be distinguished in upper limb buds, the latter of which is called hand plate; at 33 days, the upper arm, forearm, and hand plate can be distinguished in upper limb buds, and even the segmented structure of hand, i.e., wrist, hand, and finger plate, can be respectively seen, but no finger-dividing sign is seen (Fig. 1.7).

At 35 days of development, finger-dividing signs appear on the hand plate of upper limb buds, and muscles and bone tissue are visible in the limb buds in mesodermal tissues, but at this stage it is unable to distinguish between the bones and muscle tissues (Fig. 1.8). At 37 days of development, the upper limb bud development goes through the fin shape and the slurry shape, and then the fingers with traces are formed with webbed appearance, which are called webbed fingers, and elbows emerge (Fig. 1.9).

1.1.3 Limb Development

Limb develops in the order from the proximal end to the distal end. At 5–8 weeks of development, the generation of upper limbs of each segment is shown in Fig. 1.5.

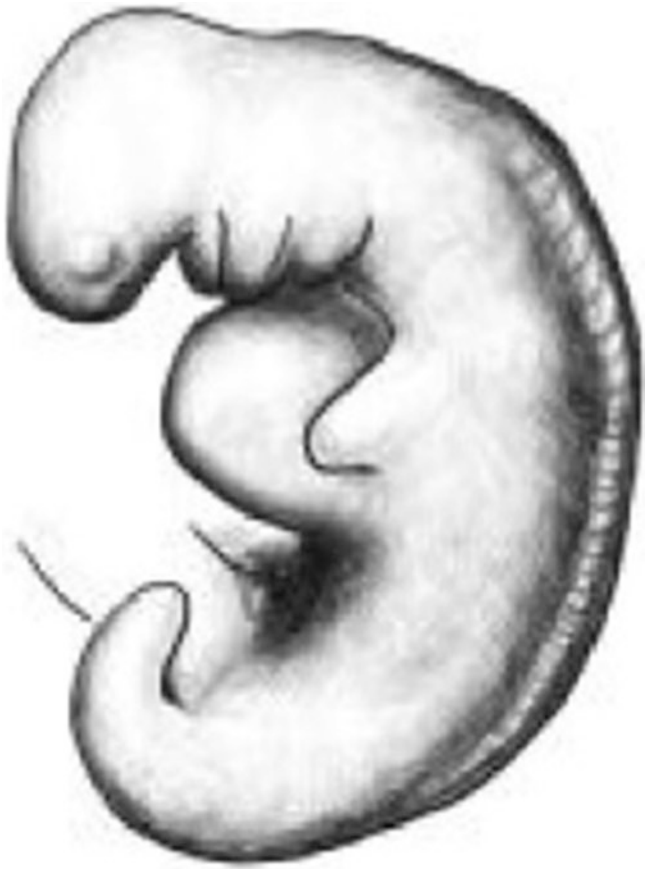

Fig. 1.3 At 26 days of development, with a length of 3.5 mm, the trace of upper limb bud appears at ventral side

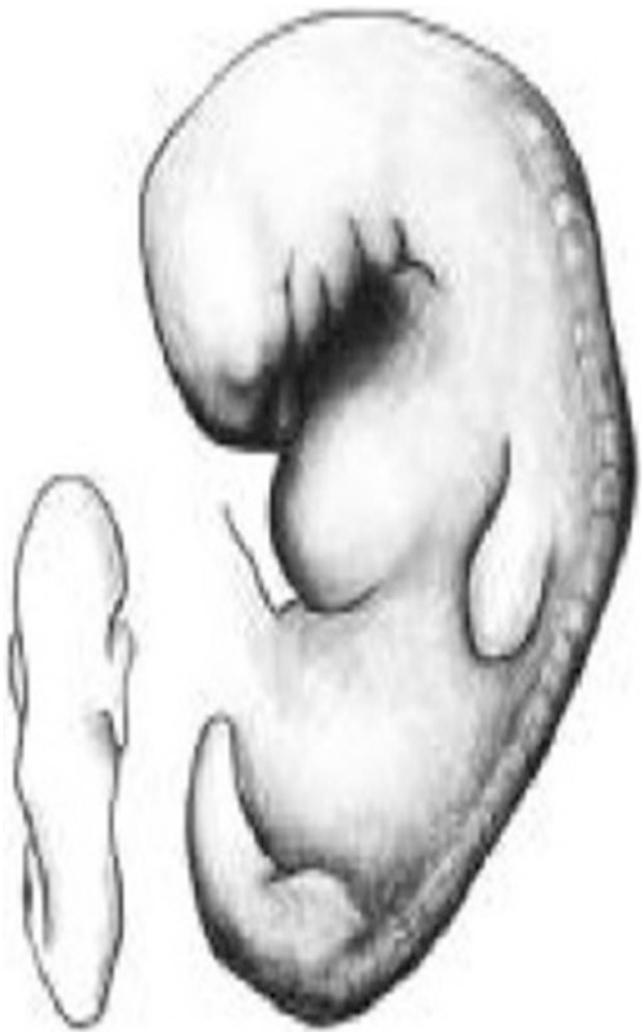

Fig. 1.4 At 28 days of development, with a length of 4–5 mm, upper limb buds are clearly visible

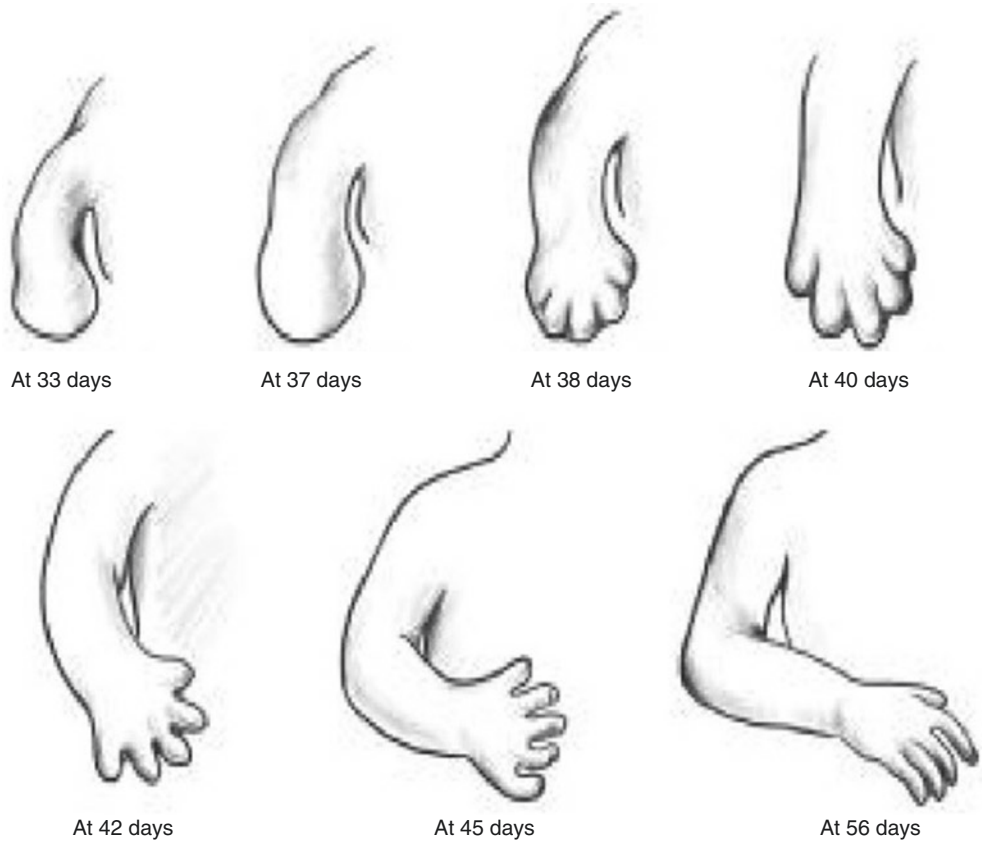

Fig. 1.5 At 5–8 weeks of development, the segments of upper limb develop smoothly

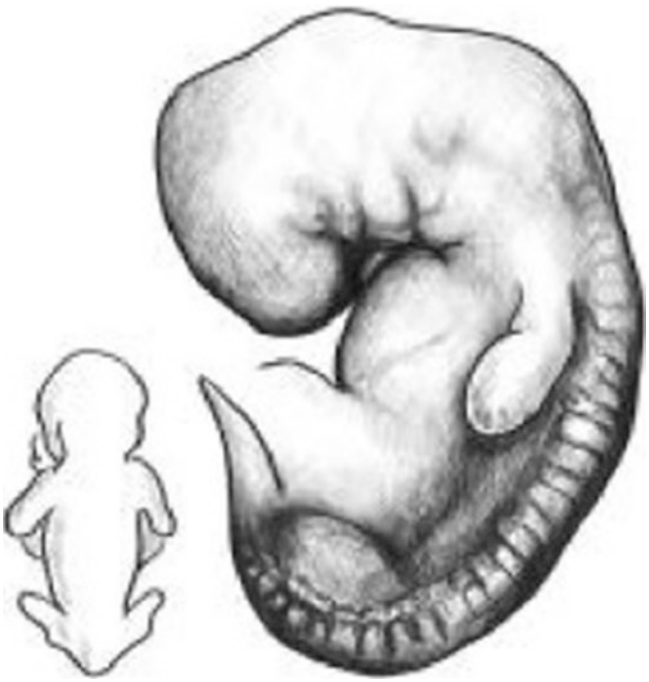

Fig. 1.6 At 28–30 days of development, with a length of 6 ~ 7 mm, upper limb bud already has segmentation

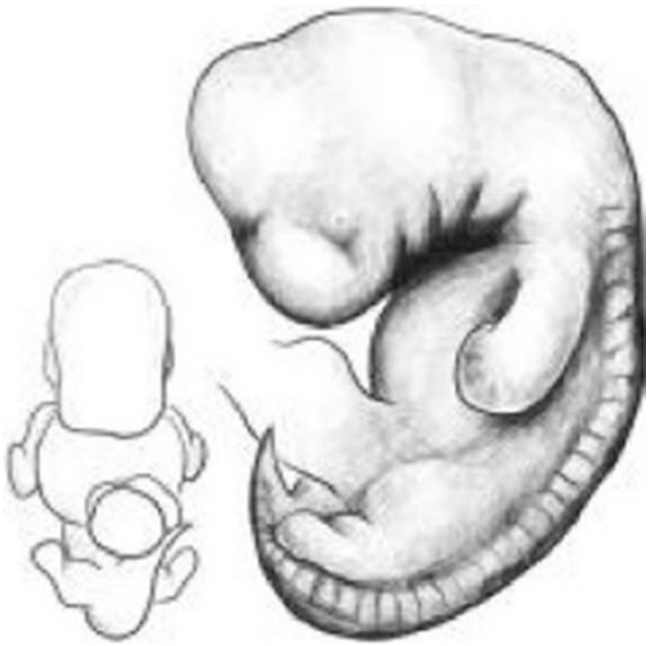

Fig. 1.7 At 33 days of development, with a length of 8 ~ 11 mm, upper arm, forearm, and hand plate can be distinguished in upper limb buds, and lower limb buds are also segmented structurally

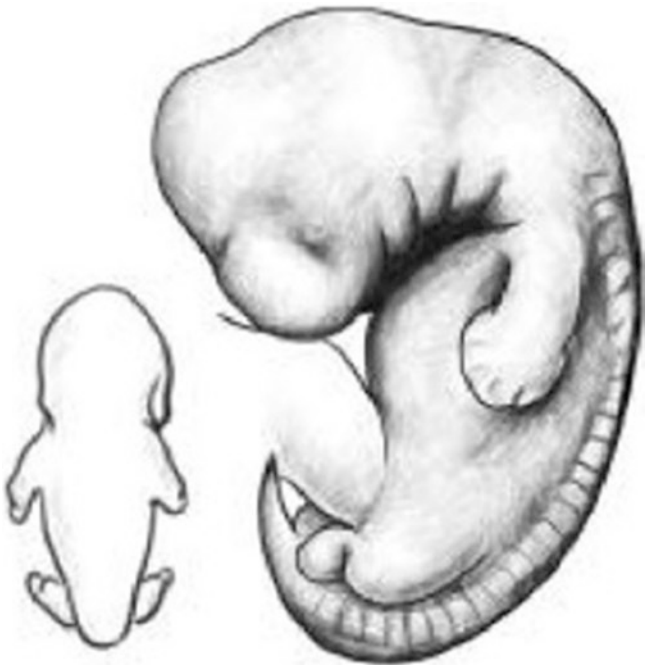

Fig. 1.8 At 35 days of development, with a length of 11–14 mm, finger-dividing signs appear on the hand plate of upper limb buds

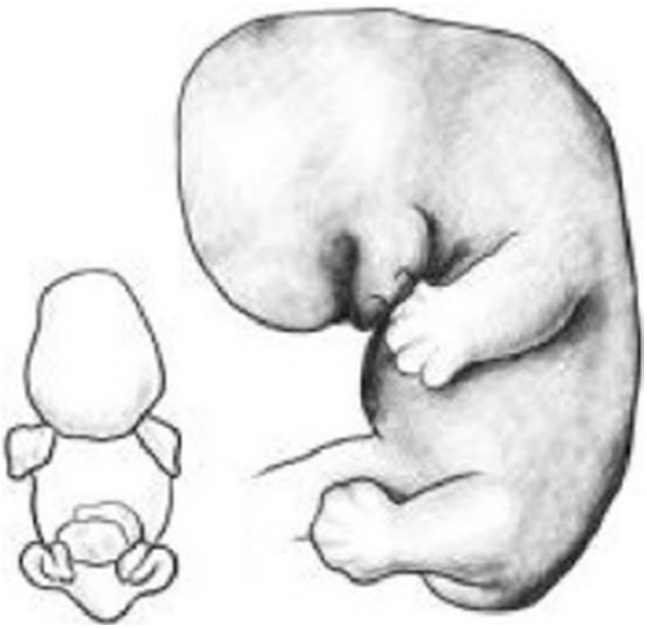

Fig. 1.10 At 39 days of development, with a length of 17–20 mm, palms face each other

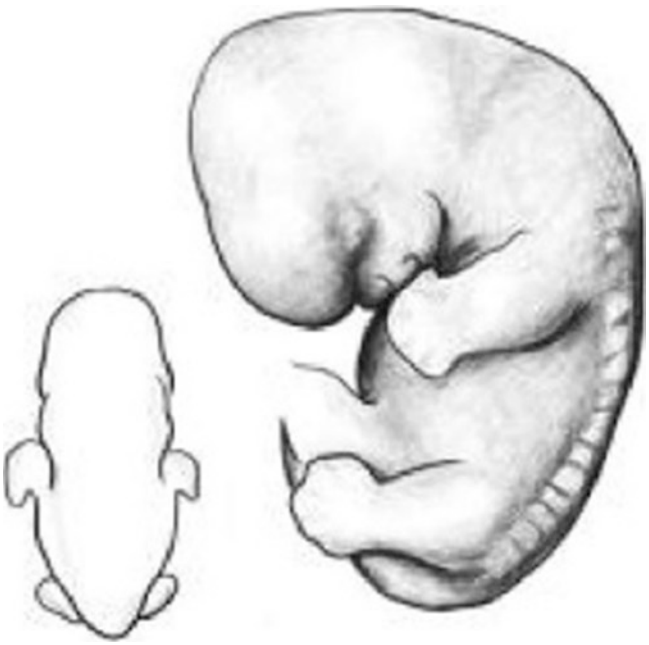

Fig. 1.9 At 37 days of development, the hand plate of upper limb bud has clearly divided fingers, which are webbed

Fig. 1.11 At 40–45 days of development, with a length of 21–23 mm, the fingers have divided completely

At 39 days of development, the palms face each other (Fig. 1.10); at 40–45 days, the finger begins to differentiate, and it becomes bigger and takes shape. At this time the nerves come into the mesenchymal tissues of limb buds from the spinal cord, and muscle composition is clearly visible. And the rest of mesenchymal tissues evolve into the original shape of cartilage and then become ossified to the bones (Fig. 1.11); at 56 days, upper limb takes shape (Fig. 1.12).

Fig. 1.12 At 56 days of development, with a length of 25–27 mm, upper and lower limbs have taken shape

With the increase in limb length, bones are gradually formed, and the myoblasts gather and then differentiate into limb muscle group. These muscle groups are divided into extensor group at dorsal part and flexor group at ventral part. At 7 weeks of development, most of the limb structures have been formed, and joints emerge. Muscles and unique individual muscles can be differentiated. The limbs rotate from the original ventral direction to the opposite direction. Initially, its flexor surface turns toward the ventral side, and then the extensor surface turns toward the dorsal side. Whether it is the upper or lower limb bud, there is a difference between cephalic side and caudal side. The former refers to the side next to the embryonic head, which is located in the anterior edge of the long axis of the limbs, called axis anterior edge; the latter refers to the side close to the rear embryonic side, which is located in the posterior edge of the long axis of the limbs, called axis posterior edge. Axis anterior edge is facing the head, while the axis posterior edge is facing the end (Fig. 1.13).

1.1.4 Skeletal Development

In the early stage of limb morphogenesis, the mesenchyme of the limb bud becomes dense. At around 6 weeks of development, chondrocytes appear and are embedded in a matrix of basophils, gradually showing the characteristics of hyaline cartilage, and further generating bone tissues in the way of entochondrostosis.

Limb bones include shoulder girdle, pelvic girdle, and free bones of upper and lower limbs. The time when ossification centers appear and the corresponding number vary. Clavicle bone is the first bone that experiences ossification, which appears at 7 weeks of development and contains two ossification centers. Entochondrostosis occurs at both ends,

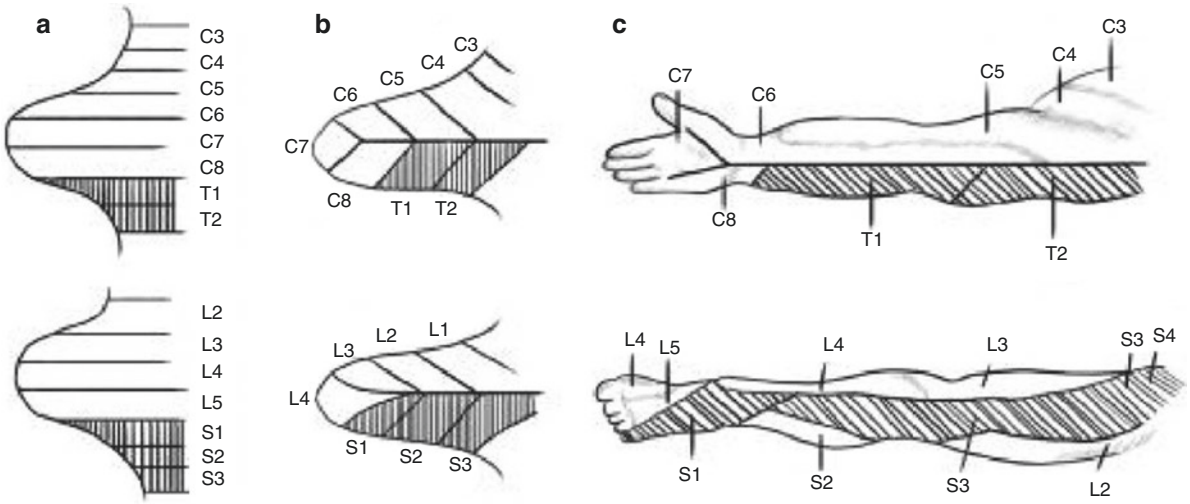

Fig. 1.13 Relationship between distribution of adult upper and lower limb cutaneous nerves and neural development process of embryonic stage

while intramembranous ossification occurs in the middle. The shoulder peak and the mesoscapula of the shoulder girdle bone have an ossification center, and there is an ossification center at the coracoid. Iliac ossification center of pelvic girdle bone appears earlier than pubis and ischium, but it is not completely ossified until adolescence, and when people are 14–16 years old, they become mutually healed to become hip bone.

The upper limb ossification centers appear in such order: the humeri (8) → radii (3) → ulnae (3) → distal knuckle bones (2) → metacarpi (2) → proximal knuckle bones (2) → middle knuckle bones (2) → carpale (1). Generally, if a comparison is made in terms of the growth rate of fetal long bones of four limbs, the growth rate of the forearm bone is the same as that of focile, but the growth rate of the distal segment is slightly faster than that of the proximal segment; the growth rate of lower segment of lower limbs is faster than that of the corresponding segment of the upper limb. Women's ossification centers appear earlier than men's.

1.1.5 Muscular Development

According to embryology and tracing observation of adult innervation, it is generally considered that limb muscles are formed by the myotome of neck and lumbar region extending into the ventral limb bud; however, the source of limb muscles' somites cannot be traced for mammalian embryos, so most scholars now believe that mammalian limb muscle does not originate from the somites. With the elongation of limb bud and formation of limb bud bone, at 7 weeks of development, the myoblasts evolving from the corresponding section of mesoblastic mesenchymal cell layer become aggregated locally and become differentiated to myoblasts, which are gradually transformed into muscle cells. With the bone as the vertical axis, it can be divided into postaxial muscles at the back of limbs (extensor group) and preaxial muscle at the ventral surface of the limbs (flexor group). The upper limb muscles appear a little earlier than the lower limb muscles, the proximal muscles of limbs appear earlier than the distal muscles, and the extensors appear earlier than the flexors. At the beginning of the seventh embryonic week, limbs become extended to the ventral side until the late stage of development, and the upper limbs rotate outward by 90° along their long axis, causing the future elbow to protrude toward the dorsal side and the extensor group toward the dorsal-lateral side. The lower limbs rotate by nearly 90° toward the interiors of their long axis, making the future knee protrude forward, that is, the ventrolateral side, the extensor group turning to the ventral side and the flexor group turning to the dorsal side. Usually at the 8 weeks of development, the main muscles of the limbs nearly take shape.

1.1.6 Vascular Development

Limb arteries originate from intersegmental artery of the corresponding body segments of limb buds; the lateral branch of intersegmental arteries constitute vascular plexus and grow along the long axis of the limbs to form the limb axial artery and its branches. Sources of the upper limb arteries on both sides are slightly different. The left side originates from the left artery in section 7, and the right side originates from the right artery in section 7 and the fourth arterial arch. Subclavian artery is formed by artery axis, and it extends downward into the upper limbs, becomes close to the descending median nerve, and reaches the front of the forearm interosseous membrane. In its advancement, vessels develop into axillary artery, brachial artery, palmar interosseous artery, and deep palmar arterial arch, and ulnar artery and radial artery occur relatively late.

Superficial vein of limb is formed by the edge vein of the upper limb bud. As the fingers of the upper limb bud become differentiated into finger arrangement, the edge vein of the upper limb preaxial margin (i.e., upper limb cephalic vein) is developed into cephalic vein, and the edge vein of the postaxial margin (i.e., upper limb caudal vein) is developed into basilic vein. In adults, the venous location proves the existence of the rotating stage of limbs during embryonic development.

1.1.7 Nerve Development

With the development of embryonic body wall (ectoderm and somatic mesoderm), spinal nerves also enter the limbs. The feeling of adult limb skin is strictly divided according to the source of spinal nerve, i.e., according to dermatomere distribution. The boundary of each dermatomere is determined by the distribution of its corresponding spinal nerves and sympathetic nerves.

The nerves of limbs are from the corresponding nerve plexus. For example, the brachial plexuses entering the upper limbs consist of the anterior branches of spinal nerves from the lower segment of neck and the upper segment of chest. These nerves are re-splitted and recombined during the limb advancement process and make up different nerve trunks and nerve bundles. They are both anatomical unit and functional unit. For example, the medial and lateral cords of the brachial plexus are dominated by dominating flexor groups, and the rear cord is dominated by dominating extensor group. But there are exceptions. For example, the innervation of brachial muscles is both from the musculocutaneous nerves of dominating flexor group and the radial nerves of dominating extensor group.

1.2 Function Development of the Hands and Upper Limbs

Jinghong Xu, Yijia Yu, Bo Chen, and Jianmin Yao

The hands and upper limbs are the main human organs that are engaged in precise operation and daily activities. They not only have motor function and can complete a variety of intricate movements accurately and powerfully but also serve sensing function, particularly physical and sensory functions, which is very important for flexible movement of hands. Maria Montessori (1909) [18], an Italian educator, called our hands "the wise tool" in his book. Application of hand in all aspects including movement, society, language, and cognition starts when life begins. The long-term hand development itself shows the complexity of hand function. Because of this, people call hands the "almighty hand." The hand functional development follows the law of overall development of motion, that is, from top to bottom, from near to far, from generalization to concentration, from positive to negative, and from roughness to precision.

1.2.1 Factors Affecting the Development of Hand and Upper Limb Functions

1.2.1.1 Relationship Between the Nervous System and Hand Functional Development

The evolution of the human cerebral cortex allows hands to possess high flexibility and skill; therefore, hand is the executor and recipient of the brain. Single finger movement depends on the original motor cortex and pyramidal tract and also depends on the sensory feedback reaching the original sensory cortex. For example, touch can adjust grip strength, which is maintained through continuous monitoring; in the integration of sensory information (visual, tactile, and temperature sensation) of complex action (such as tying shoestrings), the posterior cerebral parietal cortex plays an important role [19]. Hand feeling has important effects on sports, that is, it offers various kinds of environmental information, and affects and adjusts hand movement. On the occasion of hand movement, the sensory information continues to be imported to the cerebral cortex, so as to guide and coordinate its motion. The cerebral cortex can integrate information from the tactile neurons, which allows the cooperative work of agonistic muscle and antagonistic muscle to control and coordinate hand movements.

The visual and tactile (sensory and motor) functions of infants are independent from each other within the several months after their birth. However, as they grow, they will coordinate their eyes and hands and become gradually able to fiddle with objects out of their visual range. Sensory nerves and motor nerves of the forearm, wrist, and hand are equally important, which can detect the size, weight, temperature, texture, and other information of objects through touch, pain and temperature sensation, and other body surface receptors, and can transmit stimulation to the brain cortex by nerve impulses. The deep receptors at the nerve endings within the muscle and tendon provide proprioception, kinesthesia, and perception of pressure and joint sites [20–22].

1.2.1.2 Relationship Between Movability and Stability and Hand Functional Developmental Development

Paillard (1970) believed that stability of posture is essential to complete grip movement, and this movement can be divided into three major phases: ① eye-head positioning, which determines the correct position of the hand and arm; ② torso stability, which ensures effective upper limb movement; and ③ control over motion and stability of each joint of hands and arms, to complete precise grip. This order is consistent with the order of normal development [23–26].

1.2.1.3 Relationship Between Anatomic Structure and Hand Functional Development

The arrangement of 27 metacarpal bones and phalanges of human body forms an internal structural basis for hand function. The good combination of bone and supporting soft tissue, such as ligaments, tendons, and muscles, provides stability and mobility for various activities of hands including all joints. Throughout the entire childhood stage, the hand functional development continuously changes proportionally in the same way of the changes in the size of the hand. Hand functional development process of children aged 6–11 shows that the size of hands is correlated with changes in physical strength, which enables surgeons to predict the effects of hand surgery on children in a development phase through the height, age, sex, dominant hand, or other parameters of children [27–32].

1.2.1.4 Relationship Between Environment and Hand Functional Development

Various functional activities of hand require relatively stable external environment or body position. The relatively stable shoulder, elbow, and wrist joints can help effectively control and adjust the direction of motion and strength of the hand, and five-finger separation movements dominated by palm bow and the intrinsic muscles also depend on the stability of wrist joint. Gravity plays a key role in the final development

and improvement of the body, and mechanical stimulation generated by gravity is necessary to bone growth and establishment of the integrity of soft tissues surrounding the joint [30, 31].

1.2.2 The Development Process of the Hands and Upper Limbs

Hand functional development includes the whole process, from unconscious reflex activity to conscious posture maintaining and precision movement, such as initial perception and use of tools, development of dominant hand, and development of object gripping capabilities [18, 32].

1.2.2.1 Development of Grip Ability

Grip is the primary and most fundamental precision movement of individuals. Continuous development of grip ability enables human to make complex, smart, and accurate movements and enables hands to use tools.

At the age of around 3 months, with the disappearance of grasp reflex, a baby begins to unconsciously grip objects, which marks the beginning of its hand functional development. At the age of around 6 months, a baby begins to casually grip objects. During this process, its hand-eye coordination gradually develops. When gripping an object, it often first uses the ulnar side of the palm and then the entire palm. As it is growing, it gradually uses its radial side to grip an object or even nips up an object with its fingers (Figs. 1.14, 1.15, and 1.16) [33].

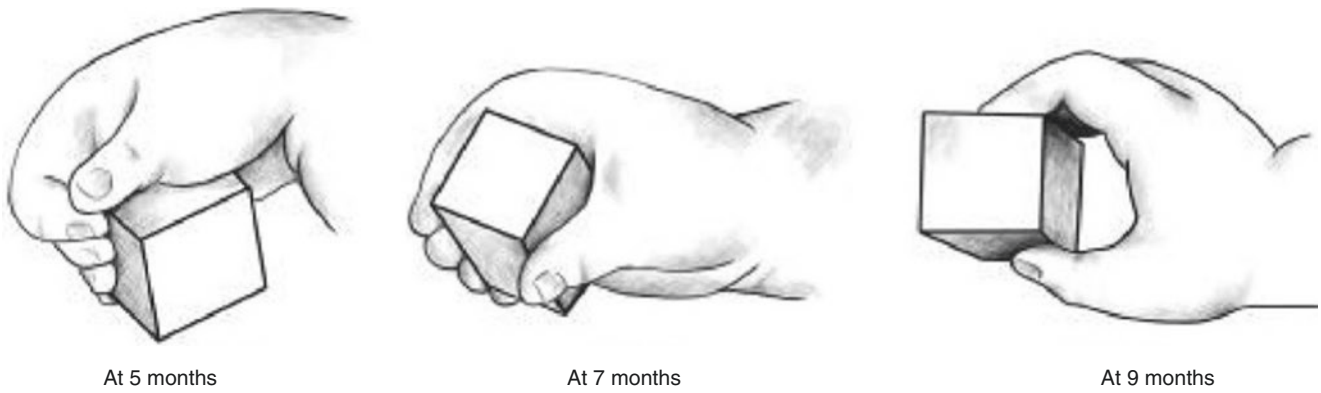

Fig. 1.14 Development process of grip function

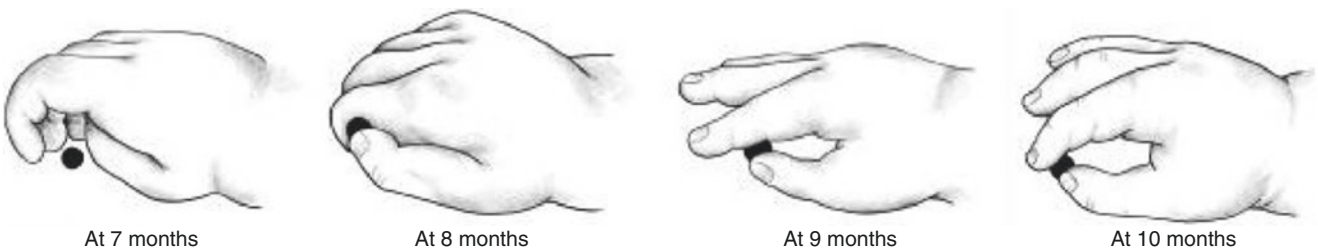

Fig. 1.15 Development process of pellet gripping action

Fig. 1.16 Development process of cylindrical object gripping action (Redrawn from Stephen J. Mathes. MD. plastic surgery second edition, 2007., volume 8, the hand and upper limb, part 2:43)

1.2.2.2 Tools Can Guide and Promote the Hand Functional Development

The morphological characteristics of tools used for eating for a baby can affect the function and efficiency of food intake. For example, the use of spoon with a recess handle enables a baby to more easily grasp it and get food. People usually experience a tool-using process from clumsy grip to precise and delicate pinch and holding; the contact with the object often shifts from the proximal end to the distal end, and the process is that flexion of wrist joint is gradually developed into dorsiflexion.

Pencil-pinching action for children aged 1–6 has shown organic combination of stability and movement (Fig. 1.17). Neuromuscular development proceeds from proximal part to distal part. Take writing and painting, for example. The shoulder joint first moves, and then the elbow joint, and then the wrist joint, and finally the metacarpophalangeal joint. Large joints at the proximal side maintain stability, while the more distant joint shows flexibility. All actions follow a similar pattern: every successful action depends on the stability and proper movement of the upper limbs. The palmar supinated grip requires the coordination of shoulder joint motion, while the pronated grip requires the coordination of elbow and forearm movement and even requires the shoulder joint to remain stable; static tripod grip is often accompanied by the movement beyond from the wrist joint, while mature dynamic tripod grip requires relatively fixed shoulder joint, elbow joint, wrist joint, metacarpophalangeal joint, etc., so that fingers can make subtle and precise movements of flexion and extension.

1.2.2.3 Development of Dominant Hands

Children under 3 show no obvious dominant hand. One does not develop obviously dominant hand until he/she becomes 8–9 years old. Researches reveal that infants under the age of 1 year often go through many stages of hand development before they can use one hand and both hands to perform one action more skillfully. According to the demand of functional development, children need to develop one dominant hand. The hands are used to different extents according to the actual need. For example, when pushing a heavy cart, one needs to exert one's strength and hence use both hands equally; when peeling bananas, one uses both hands but mainly uses one of them; when tying shoes, one uses both hands alternately. Work division of both hands and development of dominant hand are of vital importance to the hand functional developments, especially to the performance of complex actions [31].

1.2.2.4 Development of In-Hand Manipulation Capabilities

Napier (1967) observed and analyzed the static grip of hand and classified hand grip into two types: power grip and precision grip [34]. According to Exner (1992), in-hand manipulation refers to a process when a single hand is utilized to adjust the object inside it before grip is loosened so that the object can be in a more effective position [33]. For example, when grasping several coins, one moves them to the fingertips within the hand and then drops them into a vending machine; when writing, one clenches one end of the pen with the thumb and other fingers. Organic integration of stability and movement is the key to in-hand manipulation. When one is doing up a button, doing up a zip, tying up shoelaces, using scissors, or performing a more complex finger action, usually his/her fingers at the ulnar side play the role of stabilizing the object in the hand. A child at the age of around 2 years has basically fully developed his/her grip function of hands and gradually developed his/her in-hand manipulation capabilities. Of course, the in-hand manipulation capabilities need to be improved with the joint use of both hands.

1.2.3 Law of Hand Functional Developments

In the process of a baby's hand development, the ulnar side first develops, and then the radial side, and then the fingers.

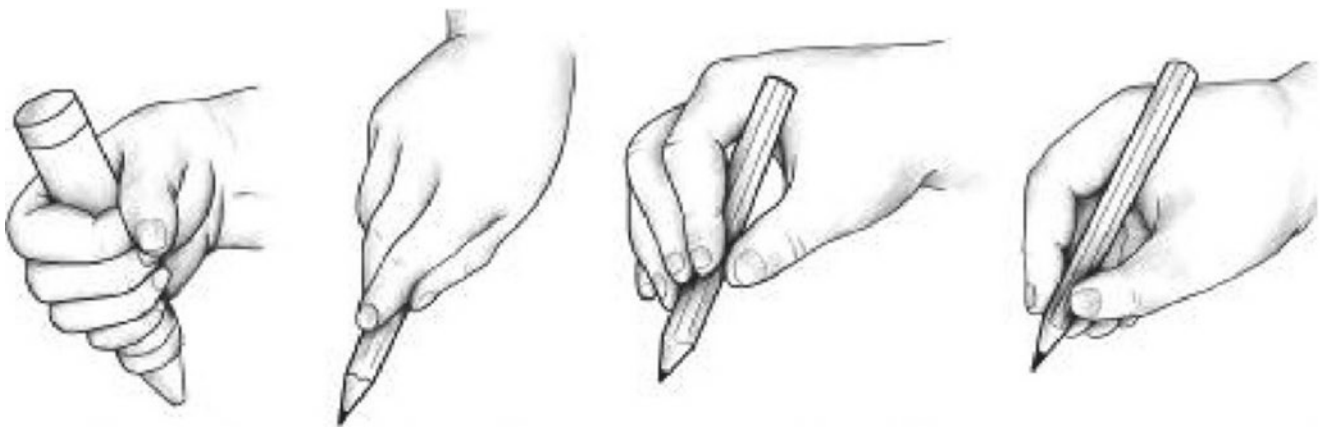

Fig. 1.17 Development process of pencil gripping from near to far

During the early development of hand, a baby gets an object by pressing its four fingers against the center of its palm. With the development of its hand, it tries to grab an object with its thumb and index finger. At an early age, a baby grabs an object without letting it go. It can only lose hold of an object when getting older. Generally, a baby begins to unconsciously touch something at the age of 3 months and becomes able to grab something consciously at the age of 5 months. The baby does not use all its five fingers to cooperatively do different parts of work until it turns 6 months [33, 35].

The process of hand functional developments is described briefly as follows [30]:

1. Neonate. The infant reflectively clenches its fist due to grasping reflex. The thumbs are often adducted and it clenches its fist more tightly when forced to be opened.
2. Month 2. The two hands are opened occasionally, and the time of opening becomes lengthened gradually.
3. Month 3. The two hands are completely opened, and the infant can grasp the object and put it onto the hands; the infant occasionally drag sits own clothes with difficulty.
4. Month 4. The infant stares at the hands, which can be put together; the infant starts the development of centric position direction (the two hands can be reached to the middle of the body) and uses hands or mouth to touch objects, and the infant can also put hands into the mouth.
5. Month 5. The infant can put any object in its hand into the mouth and can use two hands to do various actions; the infant begins to make conscious grasping action with the ulnar side (the little finger side). When lying in the supine position, the infant will stretch out the hands to touch the above toys.
6. Month 6. It is a transitional stage of infantile hand functional development. The infant can reach for a toy, can grip the toy with the entire hands, can knock the toy on the desk, can pat its image in the mirror, and can pass the toy from one hand to another.
7. Month 7. The infant starts to grip objects with radial side (the thumb side), can grip the objects with three fingers, i.e., thumb, index finger, and middle finger, and can pick up fallen objects.
8. Month 8. The infant still grasps objects with the radial side and can pick a raisin-sized object through thumb and index finger and with the remaining three fingers in extension position; the infant can occasionally use two hands to firmly grip a toy.
9. Month 9. The infant can use the ventral side of the last segment of the thumb and index finger to pinch an object and freely let go of the object gripped in the hand and can use two hands or one hand to grip an object; the hands can cross the body midline, that is, stretch out diagonally.
10. Month 10. The two hands coordinates, and the infant can use two hands to grasp one object each and knock it; when supporting the body with its forearm on a desk, it can nip objects by pressing the thumb against the index finger.
11. Month 11. The infant can use the tips of thumb and index finger to pinch objects, but the hand still needs to be left on the desk as the infant cannot pick the object up.
12. Month 12. The infant can pinch an object with thumb and index finger like pinching an object with a forceps; after pinching, the hands can be elevated and leave the desk.
13. Month 15. The infant can put a small object into a cup or bottle and can also pour the object from the cup or the bottle.
14. Month 18. The infant can build 2–3 layers of toy bricks and can pour the water in one cup into another cup.
15. Month 21. The infant can build 4–6 layers of toy bricks and can scribble on the paper with a pencil.
16. At the age of 2 years. The infant can arrange the 2–3 toy bricks in a horizontal line, can screw open the screwed bottle cap and remove it, can turn the pages one by one, and can pass the string through the small holes of beads.
17. At the age of 2.5 years. The infant can use scissors to cut paper and cloth.
18. At the age of 3 years. The infant can use the toy bricks to make them into a door or tunnel shape and keep the hands away from the desk when building the toy bricks; the infant can stretch the upper limbs to grasp a ball.
19. At the age of 4 years. The infant can grasp a large ball when the upper limbs are at the status of flexion and can throw the ball outward from the top of the head.
20. At the age of 5 years. The infant can use scissors to cut various objects.
21. At the age of 6 years. The infant can sustain an object with one hand and do things with the other and can throw a ball and pat a ball. At this stage, the infant can hold a pen basically well like an adult.
22. At the age of 7 years. The infant can knock in a nail with hammer and can throw and hit a ball.
23. At the age of 8 years. The infant can use one hand to grasp a ball and skillfully use scissors.

1.2.4 Evaluation Method of Hand Functional Developments

The fine movement age evaluation scale[36, 37] (Table 1.1) can be used to evaluate the fine movement capabilities of infants aged from 4 months to 6 years, and this scale includes 42 examination items with a total score of 72 points. The less the score is, the lower the developmental level of the fine movement is.

Table 1.1 Fine movement age evaluation scale

Age (month)	Item of examination	Score	Evaluation
4	Gently clench one's fist (1 hand)	4	
7	Grasp a dice with an edge length of 2.5 cm with 1 hand	1	
	Grasp a dice with an edge length of 2.5 cm with thumb and other fingers	1	
	Grasp a dice with an edge length of 2.5 cm with 1 hand and pass it to the other hand	1	
10	Correctly pinch a bead with a diameter of 0.6 cm with the thumb and other fingers	3	
12	Pinch 1 bead and put it into a bottle with a diameter of 5 cm	1	
	Stack 2 cubes with an edge length of 3.7 cm	1	
18	Stack 3 cubes with an edge length of 3.7 cm	6	
21	Stack 5 cubes with an edge length of 3.7 cm	3	
24	Stack 6 cubes with an edge length of 3.7 cm	1	
	Use hands to turn pages (turn 4 pages in 6 pages)	1	
	Pass a string through the small hole of a bead with a diameter of 1.2 cm	1	
30	Stack 8 cubes with an edge length of 3.7 cm	3	
	Grip a crayon to write	3	
36	Stack 9 cubes with an edge length of 3.7 cm	3	
	Put a bead into 1 bottle (10 beads, completed within 30 s)	3	
48	Put a bead into 1 bottle (10 beads, completed within 25 s)	3	
	Draw a circle with a pen	3	
	Use a healthy hand to press 3 buttons (9 times completed within 10 s)	1.5	
	Use an affected hand to press 3 buttons (8 times completed within 10 s)	1.5	
	Lift 45 small rods (completed within 180 s)	3	
60	Draw a quadrangle with a pen	6	
	Put a bead into one bottle (10 beads, completed within 20 s)	6	
66	Wind a coil (completed within 30 s)	0.6	
	Lift 45 nails (completed within 140 s)	0.7	
	Lift 5 nails with a forceps (completed within 60 s)	0.7	
	Use a healthy hand to press 3 electric buttons (10 times completed within 10 s)	0.7	
	Use an affected hand to press 3 electric buttons (9 times completed within 10 s)	0.7	
	Press 2 electric buttons at the horizontal level (6 times completed within 10 s)	0.7	
	Press 2 electric buttons at the vertical level (6 times completed within 10 s)	0.7	
	Use a healthy hand to turn the screw (completed within 55 s)	0.6	
	Use an affected hand to turn the screw (completed within 55 s)	0.6	
72	Draw a 5-pointed star with a pen	0.6	
	Wind a coil (completed within 15 s)	0.6	
	Lift 5 nails with a forceps (completed within 35 s)	0.6	
	Lift 45 nails (completed within 130 s)	0.6	
	Use a healthy hand to press 3 electric buttons (11 times completed within 10 s)	0.6	
	Use an affected hand to press 3 electric buttons (10 times completed within 10 s)	0.6	
	Press 2 electric buttons at the horizontal level (8 times completed within 10 s)	0.6	
	Press 2 electric buttons at the vertical level (7 times completed within 10 s)	0.6	
	Use a healthy hand to turn the screw (completed within 50 s)	0.6	
	Use an affected hand to turn the screw (completed within 55 s)	0.6	

1.3 Incidence, Etiology, and Pathogenesis of Congenital Deformities of the Hands and Upper Limbs

Wei Wang and Jianmin Yao

1.3.1 Incidence

Congenital deformities of the hands and upper limbs are a common disease. The Society of Congenital Anomaly of International Federation of Societies for Surgery of the Hand has made a statistical analysis on the report results of seven research centers in the UK, Japan, and the USA in1982 and found that its incidence was 1.1‰. The author et al. (1982) once investigated the birth records of 350,000 neonates in Shanghai urban area. According to them, the incidence of deformities of the hands and upper limbs was 0.85‰. As the neonate birth records in some hospitals were incomplete, the actual incidence might be higher than this figure. A European report indicated that the incidence of the congenital deformities of the limbs was 59.1/10,000 for live newborns. Among all kinds of human congenital deformities, limb deformity accounted for 26%. Lamb (1982) reported that the incidence of congenital deformities of the hands and upper limbs was 1.8‰; as the same neonate might suffer two kinds of deformities of the hands and upper limbs, the actual incidence was 1.09‰. Ivy reported that the incidence of congenital deformities of the upper limbs was as high as 8‰. Woolf et al. reported that the incidence of congenital deformities of the upper limbs was 1/1064 in live newborns in Salt Lake City, USA, from 1951 to 1967.

Congenital deformities of the hands and upper limbs may exist in the form of either one anomaly or multiple deformities, or it can be the manifestation of multiple syndromes. As is reported by Froster (1993), among the 1,213,913 live neonates, a total of 659 suffered from limb defects, 24 of whom were induced by amniotic band. Giele (2001) reported the 11-year incidence of congenital clubhand in Western Australia was 1/506, 46% of the cases were accompanied by other deformities, 51% of the cases were deformities of two hands, and 17% of the cases were multiple hand deformities; the commonest was dysdifferentiation (35%), followed by twin deformity (33%) and dyspoiesis (15%); there was no difference between white and black people.

However, few studies have been done on the incidence of each type of deformity. In 1987, Cheng et al. classified the different kinds of deformity suffered by the 1673 infants with congenital deformities of the hands and upper limbs in eight clinics. Some statistical results obtained by them can be used for reference (Table 1.2).

Flatt A. (1994) [38] recorded the incidence of 2758 cases of various congenital deformities of the hands and upper limbs in his book *Treatment of Congenital Clubhand*, which was of significance (Table 1.3).

Table 1.2 Different kinds of deformity suffered by the 1673 infants with congenital deformities of the hands and upper limbs in eight clinics

Type of deformities	Number of cases (fingers)	Percentage (%)	Range of proportion (%)
Transverse deletion	128	5	0.7–32.5
Longitudinal deletion	410	16	6.6–37.3
Dysplasia	291	11.3	4.3–19.3
Ring constriction	120	4.7	0.9–6.5
Neoplasm	626	24.3	2.4–35.9
Megalomelia and macrodactylia	22	0.9	0.5–2.1
Dysdifferentiation	442	17.1	8.3–26.4
Translocation	166	6.6	2.5–16
Soft tissue deletion	366	14.2	1.2–21.3

Table 1.3 Incidence of various congenital deformities of the hands and upper limbs

Type of deformities	Number of case (fingers)	Percentage (%)
Syndactylia	501	18.2
Polydactylia (total)	403	14.6
Radial polydactylia	184	6.7
Ulnar polydactylia	142	5.1
Central polydactylia	77	2.8
Anomaly of finger flexion	189	6.9
Broken limbs (all)	186	6.7
Broken limbs, broken hands, and broken fingers	80	2.9
Broken limbs, broken arms, and broken forearms	76	2.8
Broken limbs and broken wrists	30	1.1
Anomaly of finger lateral flexion	151	5.5
Anomaly of brachydactylia	143	5.2
Radial clubhand	127	4.6
Central ray deletion (cleft hand)	106	3.8
Dysplasia of thumb	97	3.5
Crossing syndactylia	92	3.3
Snapping finger	63	2.3
Apert's syndrome	62	2.3
Poland's syndrome	60	2.2
Ring constriction	56	2.0
Deletion of muscles and tendons	49	1.8
Deformities of muscles and tendons	45	1.6
Deletion of thumb	39	1.4
Dysplasia of ulnar (bone)	34	1.2
Deletion of ulnar fingers and metacarpal bones	32	1.2
Radial-ulnar bone fusion	30	1.1
Ulnar clubhand	28	1.0
Dysplasia of whole hands	27	1.0
Macrodactyly and megalomelia	26	0.9
Triphalangeal thumb	24	0.9
Phocomelia	19	0.7
Deletion of thumb	19	0.7
Dysplasia of radial (bone)	18	0.6
Phalangeal bone fusion	15	0.5
Others	117	4.2

Congenital deformities of the hands and upper limbs can be accompanied by cardiovascular deformity, hematopoietic system diseases, digestive tract deformity, facial deformity, cranial deformity, genitourinary apparatus deformity, and lower limb deformity.

About 5% of congenital deformities of the hands and upper limbs are one of the manifestations of syndrome. With the development of genetics, people find that the relation between hand deformity and syndrome is as follows: 48 kinds of syndromes show the symptom of aschistodactylia, 36 kinds of syndromes show the symptom of finger lateral flexion, 20 kinds of syndromes show the symptom of finger flexion deformity, and 18 kinds of syndromes show the symptom of brachydactylia. The author finds that the actual figure in clinical practice is bigger than the statistical figure. Take finger flexion deformity for example. Half of the cases are one of the symptoms of different syndromes.

McGuirk (2001) analyzed 161,252 live births and dead births in Boston, USA, from 1972 to 1974 and from 1979 to 1994 and found that the incidence of limb deformity was 0.69‰. The reasons were as follows: ① pure gene mutation, family genetic factors, and known syndrome (24%); ② chromosomal aberration (6%); ③ administration of teratogenic agent (4%); ④ vascular impairment (35%); and ⑤ unknown reasons (31%).

1.3.2 Etiology

The etiologic factors of congenital deformities are very complex, and the exact teratogenic reasons and mechanism are still very unclear at present. The teratogenic reasons can be generally divided into two types: one is intrinsic factor, namely, the genetic factor; the other is external cause, namely, the external factors during the embryonic period.

1.3.2.1 Genetic Factors

The genetic factors include the following: ① chromosomal abnormality, namely, abnormality in chromosome number or structure, and as miscarriage and dead births are induced in most cases, clinical cases are not very common; ② gene mutation, 10–15% of congenital deformities are induced by gene mutation, but most gene mutations will not induce congenital deformities [39–43]. Gene mutations are classified into polygenic mutation and monogenic mutation. The former can result in multiple congenital deformities, and the latter can sometimes result in multiple defects such as acrosphenosyndactylia induced by one single dominant gene.

Gene mutation and congenital deformities are closely associated. P63 gene is one member of the p53 gene family and can be structurally classified into transactivation domain (TAD), DNA binding domain (DBD), oligomerization domain (OD), and SAM structural domain (SAM) (Fig. 1.18). The coding products are multiple isomers with different activities and can be classified into two types: ① the isomers which start transcription from exon 1 and have transactivation domains are called TA isomers; ② the isomers which start transcription from the site between exon 3 and exon 4 and do not have transactivation domain are called Δ*N* isomers; at the same time, due to the differences of 3′ end shearing methods, isomers with the following different C-ends (α, β, and γ) are produced. It has been verified at present that p63 gene plays an important role in the development, differentiation, and morphogenesis of various epithelial tissues, formation of embryo, and development and differentiation of ectoderm. p63 gene is widely expressed in human tissues, such as the esophagus, lungs, skin, muscles, breast glands, spleen, lymphocytes, nervous tissues, digestive system and urogenital system, etc., but its composition in such histiocytes and subcellular localization varies. P63 gene is expressed in AER of limb buds, and the mutation of p63 gene renders the AER unable to complete differentiation, so its structural integrity is maintained. At present, it is found that the mutation of p63 gene is present in ectrodactyly-ectodermal dysplasia-cleft lip/palate syndrome (EEC syndrome), split-hand/split-foot malformation (SHFM), and ankyloblepharon-ectodermal dysplasia-cleft lip/palate syndrome (AEC syndrome). The heterozygous mutation of p63 gene is associated with ectodermal dysplasia, orofacial cleft, and deformity at the limb ends. Till now, 31 mutation sites have been found in patients with EEC syndrome, including five mutational hot spots (R204, R227, R279, R280, and R304), which are located at the DBD of p63 gene and influence the binding of p63 gene and DNA, resulting in the decrease in the transcription activity (Fig. 1.19). The SAM structural domain of p63 gene participates in the interactions between proteins during tissue development and differentiation; therefore, it is speculated that the mutation that occurs to this structure will inhibit the interactions of specific proteins [44–46].

The primary cause of congenital deformities is the inheritance of the genetic factors in cell chromosome to the next generation. Genetic factors play an important role in the occurrence of congenital deformities; according to statistics, about 5% of the hand deformities are genetically induced. Due to blood relationship, among the family members with a family history of deformities, the incidence of deformities is 25 times that of normal population. The majority of hand deformities are induced by monogenic inheritance. The mode of inheritance is autosomal dominant or recessive inheritance and sex-linked inheritance, among which the common type is autosomal dominant inheritance [40, 47, 48]. The genetic regularity is as follows:

The thin arrowhead represents the mutational amino acid residue (K194 and R280) in SHFM, and the thick arrowhead represents the mutational amino acid residue (R279 and R304) in EEC.

1. The morbigenous dominant gene is located at one pair among the 1–22 autosomes. Inheritance is unrelated with

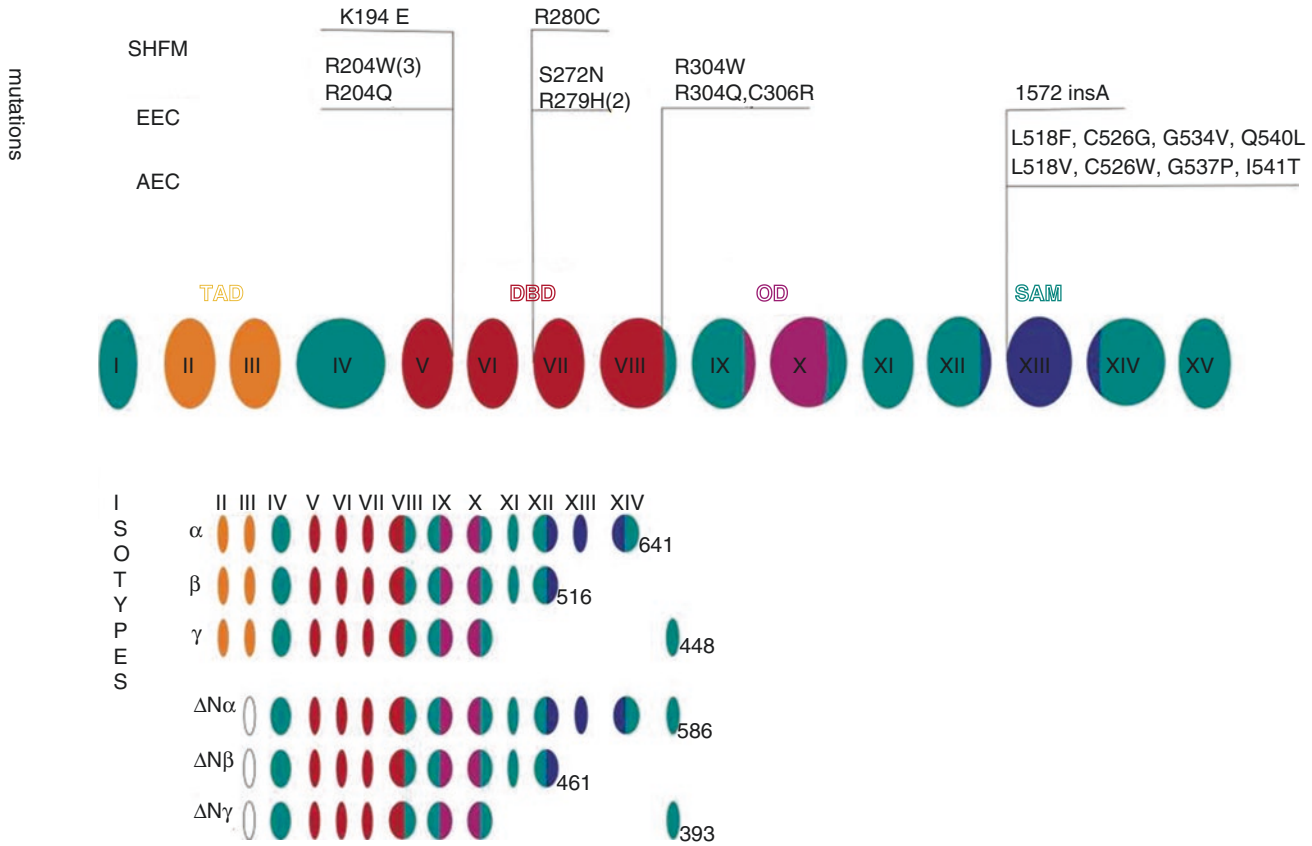

Fig. 1.18 SHFM, EEC, and AEC mutational hot spot of p63 gene (Redrawn from Sifakis S, Basel D, Ianakiev P, Kilpatrick M, Tsipouras P. Clin Genet. Distal limb malformations: underlying mechanisms and clinical associations. 2001;60(3):165–72)

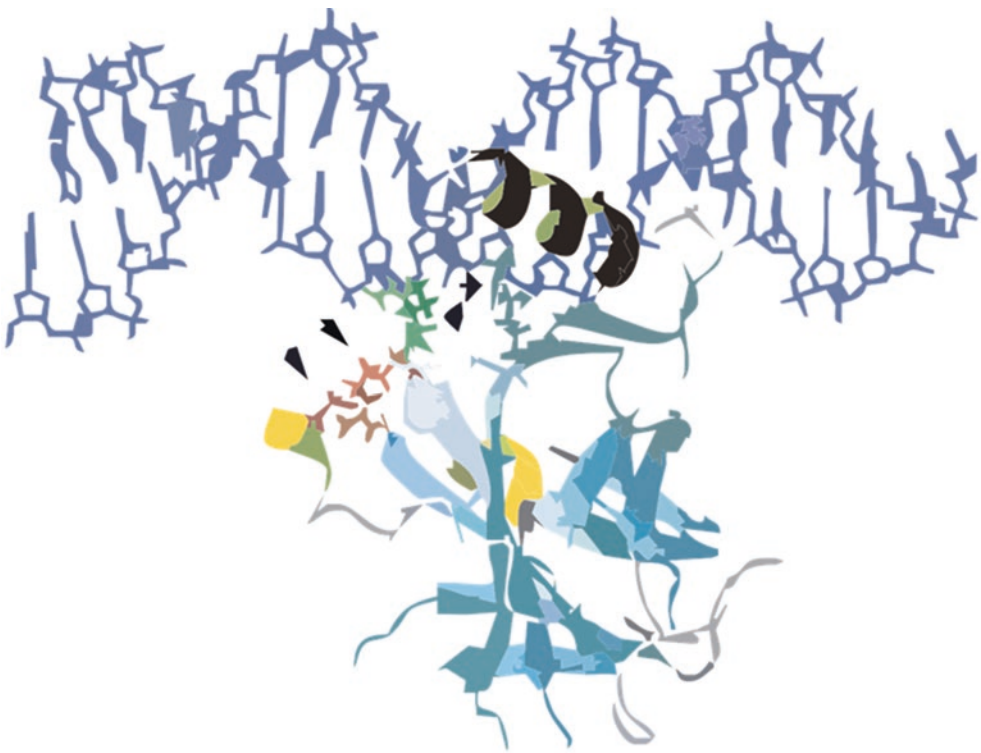

Fig. 1.19 p63 gene DNA binding domain ribbon diagram (Redrawn from Ianakiev P, Kilpatrick MW, Toudjarska I, Basel D, Beighton P, Tsipouras P. Split-hand/split-foot malformation is caused by mutations in the p63 gene on 3q27. AmJ Hum Genet 2000: 67: 59–66) (Reprint with permission from ELSEVIER LICENSE TERMS AND CONDITIONS)

gender, and the pathogenic probability among men and women in the family is the same.
2. Patients can be found in each generation, and the disease may occur commonly within continuous generations in the family.
3. If a patient gets married with a healthy person, the incidence of deformities in their children is 50%; if both are patients, the incidence of deformities in their children is as high as 75%.
4. Different expressivities are present, which means that despite the onset in different individuals with the same genotype, the severity of lesions varies.

The common deformities induced by dominant inheritance include symphalangia, brachydactylia, cleft hand, hyperdactylia, etc. Patients with recessive inheritance usually have no obvious family history. The manifestations of the sex-linked recessive inheritance are men developing the disease and women being the carrier of the pathogenic factor.

Consanguineous marriage is the primary cause of congenital deformities. In case of non-consanguineous marriage, the incidence of deformities in the children is 1‰; in case of consanguineous marriage, the incidence of deformities in the children is 25% to 50%, which is 250 to 500 times as high as that of normal population.

1.3.2.2 External Factors

Under the influence of external factors during the embryonic stage, especially the teratogenic factors in the embryo, hand deformities can also occur. The occurrence of deformities does not involve the genetic factors in chromosomes, so such deformities will not appear in the future generations. The key stage of occurrence of embryonic deformities is the first 3 months after gestation, which might be associated with the following factors:

1. Nutritional factor. Animal experiment studies indicated that, in case of lack of vitamin C in diets for mother mice, the embryonic mice might suffer deformities in flexural limb after birth. Warkany and Nelson fed the white mice with vitamin B_2 deficient feeds. Among the 484 mice born afterward, 189 (39.05%) developed various congenital deformities; half of them developed deformity of forepaw. The deficiency of vitamin A can affect the development of embryonic soft tissues and can induce the deformities in the heart, eyes, diaphragm, and genitourinary apparatus.

   In human kind, there are few cases where the mothers are undernourished, but some placental lesions will affect fetal nutritional supply, further influencing embryonic development and inducing the occurrence of hand deformities.
2. Drug factor. Many drugs have a teratogenic effect, including sedatives such as thalidomide, anticancer drugs, and oral contraceptives. Animal experiments have verified that cortical hormone, trypan blue, mustard nitrogen, etc. can induce limb deformities in animal embryos. Kosenow and Pfeiffer (1960) reported the relation between occurrence of phocomelia anomaly (short upper limbs, and the hand is connected with the shoulder) and administration of thalidomide during the early pregnancy, and the incidence is up to over 20%. Thalidomide once caused thousands of children in Europe to develop deformities of amelia and micromelia from 1957 to 1961, and this event spurred scholars to carry out the studies on the cause and pathology of congenital deformities. In addition, organomercury, insecticides, etc. are also very important teratogenic factors. For the same drug, the differences in its dosage, route of use, absorption, and metabolism will induce different types of deformities.
3. Radioactive factors. Radioactive rays have a decisive influence on embryonic genetic characteristics and can even cause developmental arrest. After X-ray radiation is performed on white mice, water blister, blood blister, and hematomas can be found in the paws of the embryonic mice. Multiple deformities such as lipomeria, adactylia, cleft limbs, hyperdactylia, and syndactylia are formed after birth, and in the meantime, ocular and renal deformities also occur. After the Second World War, a random examination was carried out on 205 children who were exposed to the atomic bomb explosion during the first half stage of embryo and found 28 developed deformities, which accounted for over 13%, indicating that their incidence was much higher than that of the common population.
4. Endocrine factor. Daraiswami injected a small amount of insulin into the shell of eggs whose incubation was traced so that the hatched chickens would develop multiple deformities, but the occurrence of deformities could be prevented if both niacinamide and vitamin B2 were injected into it. Clinically, the incidence of deformities of future generation of diabetes patients is five to seven times higher than that of healthy population.
5. Disease factor. The exposure of mothers to the infection of some pathogens during the first 3 months such as rubella virus, cytomegalovirus, toxoplasm, herpes simplex virus, Asian influenza virus, epidemic parotitis virus, and *Treponema pallidum* can induce fetal deformities. Gregg (1941) found, in case the mothers developed rubella during the first 2 months after gestation, their fetuses could suffer various kinds of congenital deformities, such as cataract, decreased hearing, heart deformities, and dysostosis, and the reason might be that the virus directly influenced embryonic development through placenta. Other scholars hold that the poor maternal health status might be one supplementary factor that causes

the fetuses with a certain genetic factor for anomaly to develop deformities. In addition, maternal diabetes, chronic ethylism, etc. could also induce fetal deformities. Some reports indicated that thalassemia can induce digital arterial embolism, which results in the deformity of congenital dactylolysis.

6. Trauma factor. Some scholars consider that, during the early embryonic stage, the hematomas in the embryo can inhibit the development of some embryonic parts, further resulting in deformities. During the later stage of pregnancy, the fetus grows rapidly, and the amniotic fluid decreases gradually; in the meantime, the abdominal cavity and pelvic cavity are under increasing pressure. Cotwin or uterine deformities, hysteromyoma, etc. would especially cause the fetal limbs to be subject to compression and their movement to be confined so that the fetal limbs will become flexed and deformities will occur. In addition, the fetal constraint by amniotic band or fibrous ring will also result in intrauterine amputated extremities (fingers).
7. Environmental factor. According to Jones (1973), fetal alcohol syndrome is the syndrome of abnormalities in the head, face, four limbs, heart, and external genitals accompanied by generalized scoliosis and listlessness that are found in infants delivered by pregnant women with ethylism. Hundreds of cases have been reported after his definition of such syndrome. Therefore, pregnant women have been warned that moderate drinking during pregnancy is harmful to the fetuses. In addition, pregnant women who smoke will deliver low birthweight infants, and their mean weight can decrease by 150–250 g. As is reported by other articles, smoking could lead to an increase in miscarriage rate and perinatal mortality rate, and the risk of deformity occurrence in the children delivered by smoking pregnant women is two to three times higher than that in the children delivered by nonsmoking pregnant women.

1.3.3 Pathogenesis

At present, the real pathogenesis of congenital deformities of the upper limbs is little known. Regarding this, two views are currently prevailing: one is that the development process is programmed by gene from the beginning; the other is that development is the result of biological, chemical, and physical effects of sequences and is subject to the influence of four-dimensional spacetime. The two viewpoints lead to the theory of genetic determinism and the theory of environmental determinism. However, more materials indicate that the majority of deformities result from the joint effect of the two factors, but the influence of environmental factors is more significant. The author believes that some congenital deformities of the hands and upper limbs are gene programmed from the beginning, such as deformity of camptodactilia, deformity of finger lateral flexion, cleft hand, and some deformities of syndactylia, and the patients often have an obvious family genetic predisposition; in addition, environmental factors can induce gene mutation or change the normal expression of gene, such as radial clubhand, and the gene defect can somewhat induce deformities only under the influence of some environmental factors.

From the children treated by the author, it is known that the family of one patient has obvious medical history of syndactylia inheritance (Fig. 1.20). There are 38 direct relatives in the five generations of ancestors in their family, 21 of whom suffer congenital aschistodactylia with the incidence of 55.3%. In terms of the shape of deformities, the manifestations are syndactylia of middle and ring fingers, most of which were complete syndactylia, fingernail amalgamation, and ungual phalanx fusion. Among them, there were 19 cases of syndactylia of middle and ring fingers of both hands, a proportion of 90.5%, and two cases of syndactylia of middle and ring fingers of one single hand, a proportion of 9.5%. In terms of gender, nine are men and account for 42.9%, while 12 are women and account for 57.1%.

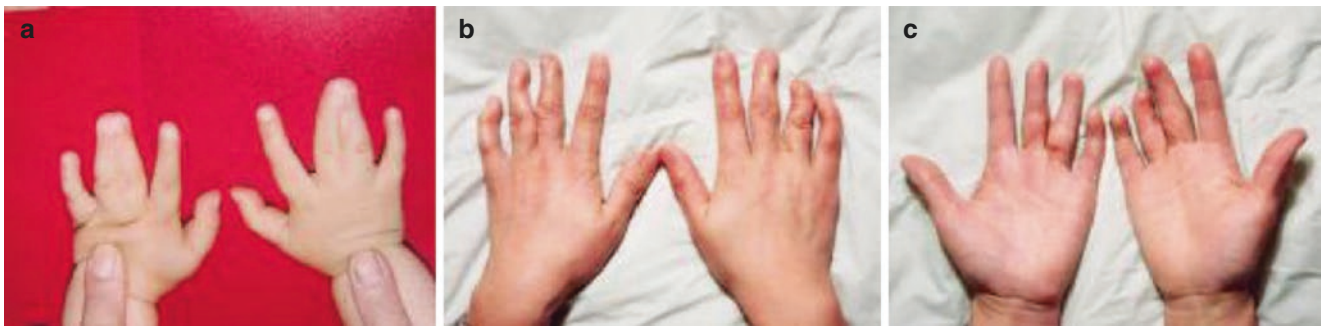

Fig. 1.20 A girl aged 2 years and a half, complete symphalangia of middle and ring fingers, with a family history (**a**). Hand appearance of the patient (**b**). (**c**) Post-surgery for complete syndactylia of both hands of the patient's mother

According to a large quantity of experimental materials, Wilson theoretically classified the mechanism of teratogenic action into nine categories: gene mutation, chromosome aberration, interference with mitochysis, changes in nucleic acid functions and synthesis process, substance insufficiency before the synthesis of protein and enzyme, blocked energy supply, inhibited enzymatic activity, self-stabilization function disorder, and changes in cell membrane characteristics. From the clinical perspective, Beckman and Brent classified the mechanism of action of human teratogenic sources into the following categories: cell death, delayed mitochysis, prolonged cell cycle, delayed differentiation, forced position, insufficient vascular blood supply, histogenesis disorder, and inhibited cell migration.

1.4 Pathology and Genetics of Congenital Deformities of the Hands and Upper Limbs

Jinghong Xu, Jialiang Chen, and Wei Wang

1.4.1 Pathology

The entire process of embryonic development is expressed through gene regulation. The occurrence of all histiocytes is mutually constrained in differentiation and development according to certain genetic information; the primordia of all organs are formed through different mechanisms of histiocyte proliferation, differentiation, local growth, degeneration, and absorption. It should be emphasized that the development zone is a region or a group of cells in embryo and can give a response to the endogenous or the exogenous stimuli as a whole. The defects in the development zone are correlated with the function disorder of primitive cells and the mutual reaction of multiple tissues; for instance, the rostral mesoblastic development disorder can induce head and facial multiple deformities, and the disorder of hypothalamus or vascular tissues can induce deformities in the reproductive organs and heart. During the process of embryonic development, the interference and disorder at any level can induce various kinds of dysplasia and functional disorder, leading to various kinds of congenital deformities and abnormalities, and even embryonic development termination and death. The abnormal phenotypes appearing at all levels of development include:

1. Metabolism disorder: it can be manifested as autosomal recessive or dominant genetic disease.
2. Histogenesis disorder: if two to three embryonic layers and the derivant tissue structures are influenced, the severity is relatively serious; otherwise, the clinical manifestation is relatively mild, and the mode of inheritance can be dominant or recessive.
3. Organ formation disorder: namely, the defects in the structures and functions in organs; various congenital deformities may appear and account for 2–7% of neonate deformities, and 1% of newborns suffer multiple deformities.
4. Deformation disorder: it often occurs in the fetal period 3 months after the conception. The manifestations are obvious changes in the shape and structure of relevant body sites, the majority of which is local involvement. In fetal period, both enzygotic twins and dizygotic twins may suffer deformation due to fetal compression, and such deformations account for 2% of the newborn deformities and can be corrected after birth.

According to the law of embryonic development, different development modes can result in various kinds of deformities, and Patten once proposed six methods: ① hypogrowth, ② hypoabsorption, ③ hyperabsorption, ④ absorption at the wrong sites, ⑤ growth at the abnormal positions, and ⑥ overgrowth of tissues or structures. Arey proposed nine similar methods: ① aplasia, ② hypoplasia, ③ impaired development, ④ adhesion of adjacent primordia, ⑤ overgrowth, ⑥ dislocation, ⑦ incorrect migration, ⑧ atypical, and ⑨ atavism. The modes of occurrence of deformities listed by Cohen (1981) are shown in Table 1.4.

Table 1.4 The congenital deformities induced by abnormalities in embryonic shape

Type	Clinical manifestations
Morphological hypoplasia	
Absence of development	Abrachia, arhinia, renal aplasia, aproctia
Hypoplasia	Achondroplasia, head deformity, and micrognathia deformity
Incompetence	Cleft palate, cheilognathus, and iridocoloboma
Incomplete separation	Ankylodactyly, persistent truncus arteriosus
Incomplete migration	Eversion of cloaca (incomplete orchiocatabasis)
Incomplete rotation	Incomplete intestinal rotation (situs inversus)
Incomplete subsidence	Choanal atresia and Meckel diverticulum
Retention of early positions	Low-set ears and cryptorchidism
Excessive morphogenesia	Polydactylia, dactylomegaly, and big earlobe
Morphogenesia aberration	Struma endothoracica, paradidymal tumor

1.4.2 Genetics

1.4.2.1 Monogenic Inheritance

The alleles arranged in pairs on the homologous chromosomes according to the linear sequence can be divided into dominant type and recessive type. The dominant type is usually expressed with A and the recessive type is expressed with a. On the homologous chromosomes, if the nature of each pair of alleles is the same and it is dominant (AA) or recessive (aa), it is a homozygote; on the contrary, if the nature of each pair of alleles is different and it is Aa, it is a heterozygote. The gene in one pair of alleles which presents the heterozygote (Aa) and manifests the hereditary character or hereditary diseases is called a dominant gene; the gene in one pair of alleles which presents the homozygote (aa) and manifests the hereditary character or hereditary diseases is called a recessive gene. In case the recessive disease-causing gene presents the heterozygote (Aa), due to the presence of normal dominant gene A, the effect of the recessive disease-causing gene a cannot be expressed, and such heterozygotes will not develop a disease. It means the phenotype is normal but the disease-causing gene a will be passed to the next generation. The patients with such genes are called carriers of disease-causing genes. Because of this, the single-gene inheritance diseases can be classified into two types: dominant inheritance and recessive inheritance. Because the single gene can be located on autosome or sex chromosome, the diseases can also be classified into autosomal dominant type and recessive inheritance type and sex chromosome dominant inheritance type and recessive inheritance type.

1. Autosomal dominant inheritance. One hereditary character or disease-causing gene is located on the 1–22 pairs of autosomes. In case their alleles present heterozygote (Aa) and manifest the hereditary characters or hereditary diseases, this is called autosomal dominant inheritance. The modes of transmission are: ① the diseases can occur if the genotype is a heterozygote (Aa), and the conditions are more serious if it is a homozygote (AA); ② the incidence for each generation is 1/2; ③ the chance of onset between men and women is half and half; ④ there are patients that develop the diseases in each generation; ⑤ if the parents are disease-free but their children develop disease, this may be caused by gene mutation or insufficient penetrance. Among the deformities of congenital cleft hands (feet), SHFM1 (7q21.2~22.1, 61, 62), SHFM3 (10q24~25, 37~40), and SHFM4 (3q27) are autosomal dominant inheritance and induced by p63 gene mutation.

   The autosomal dominant inherited diseases associated with congenital deformities include syndactylia, hyperdactylia, osteogenesis hypoplasia, cartilaginous developmental disorder (insufficiency), multiple epiphyseal dysplasia, Marfan's syndrome, and neurofibroma.
2. Autosomal recessive inheritance. One hereditary character or disease-causing gene is located on the 1–22 pairs of chromosomes. In case their alleles present homozygote (aa) and manifest the hereditary characters or hereditary diseases, this is called autosomal recessive inheritance. The modes of transmission are: ① the diseases can attack if the genotype is a homozygote (aa); ② if the filial generation has disease developers, the parents are carriers with normal heterozygote (Aa) phenotypes; ③ in each generation, 1/4 of the filial generation develops the disease, 2/4 are carriers, and 1/4 is normal; ④ both men and women may develop disease; ⑤ generally the onset is scattered; ⑥ in case of consanguineous mating, the incidence for filial generation is significantly increased.
3. X chromosome inheritance (sex-linked inheritance). It means that one kind of hereditary character or disease-causing gene is located on the sex chromosomes, most of which are located on X chromosomes, so it is also called X-linked inheritance. For instance, SHFM2 (Xq26) is an X chromosome genetic locus. The disease-causing genes on the X chromosomes are classified into dominant type and recessive type. As for the dominant type, in case the gene combination presents heterozygote (Aa), the disease can occur, which is called X-linked dominant inheritance; as for the recessive type, in case the gene combination presents the homozygote (aa), disease can occur and those with a heterozygote (Aa) are carriers, which is called X-linked recessive inheritance. The male sex chromosomes only have one X chromosome, the size of Y chromosome is relatively small, and it has no corresponding alleles with X chromosome. There is only one gene present in the paired alleles, and this is called hemizygote. Therefore, men cannot have homozygote. Although the disease-causing gene is recessive, men can develop the disease as long as one disease-causing gene presents hemizygote, so most patients with sex chromosomal genetic diseases are men, and women are mostly carriers. If one kind of hereditary character or disease-causing gene is located on Y chromosome and there is no corresponding allele on X chromosome, this gene will be transmitted along the Y chromosome, namely, from father to son and from son to grandson. As women do not have Y chromosome, they cannot transmit relevant genes. The number of genes that can be located on the Y chromosome is relatively small, and the following are the most important factors: ① testis-determining factor, located on Yp (Y short arm 1 region 1 band 3 subband); ② deleted in azoospermia gene, located on Yp (Y long arm 1 region 1

band); ③ STA, located on Yq (Y long arm 1 region 2 band); and ④ zigzag gene, located on Yq [40, 41].

1.4.2.2 Polygenic Inheritance

The diagnosis of polygenic inheritance is different from that of single-gene inheritance diseases except that it is based on the clinical phenotypes of all diseases.

For many hereditary characters including quantitative characters such as height, body shape, intelligence, blood pressure, and life span as well as multiple hereditary diseases, the hereditary basis that controls them is not one pair of genes but over two pairs of polygenes, which is called polygenic inheritance. To confirm whether a certain disease has a polygenic hereditary basis, an investigation must be carried out on the incidence among the population and the incidence in the first-degree relatives. The presence of significant difference indicates a hereditary basis and can serve as one of the basis for diagnosis of the disease of multifactorial inheritance.

The causes of multiple diseases of multifactorial inheritance are relatively complex, and the location of predisposing genes and genetic analysis are the new high-profile spots in current studies. Domestic and foreign investigators have carried out studies from the perspective of modifying experimental technologies and analysis methods, including linkage analysis, involved sibling and involved family member analysis, correlation study, and polygenic analysis of animal models. Currently some progress has been made, enabling the predisposing genes of diseases of multifactorial inheritance to be located and cloned and making possible genetic diagnosis and treatment of diseases of multifactorial inheritance.

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Functional Anatomy, Examination, and Functional Assessment of Hands

2

Dong Han, Hong An, Wei Wang, Bo Chen, Jianmin Yao, Fengjing Zhao, Bin Wang, Feng Ni, and De Shi

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2.1 Functional Anatomy of Hands

Hong An, Dong Han, and Bo Chen

Human hands have very accurate and complex functions, and their relevant anatomic structures include brachial plexus and various structures from shoulder girdles to fingertips [1]. According to the features of this specialty, this section describes the functional anatomy of wrists and hands.

2.1.1 Hand Functions

2.1.1.1 Hand Posture

To perform complex actions, hands can make various gestures, the completion of which not only needs muscular contraction to generate sufficient strength but also needs muscular coordination to make very accurate movements. The hand skin, especially volar skin, has a large number of sensory receptors, which can perceive the texture and appearance of objects as well as small changes in them; there is a large number of proprioceptors in the muscles and joints, which can feel the small changes in hand gestures, and such sensors can guarantee that hand actions are accurate and smooth. There are the following two kinds of hand gestures:

1. Resting position. During sleep or rest, the muscles of forearms and hands become relaxed; at this time, the hands are under a special natural status, which is called resting position. This resting position is maintained by tissues such as ligaments with the following manifestations: the wrist joint extends in a dorsal direction by 10°–15° with a mild ulnar deviation; the thumb tip is close to the radial side of the middle segment of index finger, the remaining four fingers take the semiflexion shape, and the degree of flexion increases from the index finger to the little finger and the fingertip points toward the tubercle of scaphoid bone, indicating that the strength of hand flexor aspect exceeds that of the extensor aspect (Fig. 2.1) [2]. During passive flexion and extension of the wrist joints, all fingers will do the corresponding flexion and extension actions. The correct understanding of the resting position of hands and their changes are important basis for the diagnosis and treatment of hand injuries and diseases. In case of rupture of the flexor tendon of a certain finger, the finger does not take the semiflexion shape but is straightened in the resting position; when tendon transplantation is adopted to repair flexor and extensor tendons, they can be adjusted to the resting position for anastomosis, and at this time the tension is optimal [3].
2. Functional position. The body position that can give full play to hand functions is called functional position. At this time, the wrist extends in a dorsal direction by 25°–30°, the degree of ulnar deviation is about 10°, the metacarpophalangeal joint flexes by 30°–45°, the proximal interphalangeal joint flexes by 60°–80°, the distal interphalangeal joint flexes by 10°–15°, the carpometacarpal joints of thumbs are fully abducted, the thumbs take the opposing position, and the rest four fingers are separate. For instance, gripping one tennis ball into the hand can basically reflex the status of hand functional position (Fig. 2.2). In clinical work, hands should be placed in the functional position in hand fracture reduction or finger joint fusion [1–3].

2.1.1.2 Motor Functions of Hands

1. Pinching and holding. It is a fine action of fingers, including pinching and holding with finger pulps, pinching and holding with fingertips, pinching and holding with the lateral aspects of thumbs and index fingers, and the actions of gripping, pinching, revolving and twisting with multiple fingers.
2. Gripping. It is a result of forceful flexion of fingers and palms and used for doing heavy work.
3. Lifting. It is an action that fingers flex to raise objects.

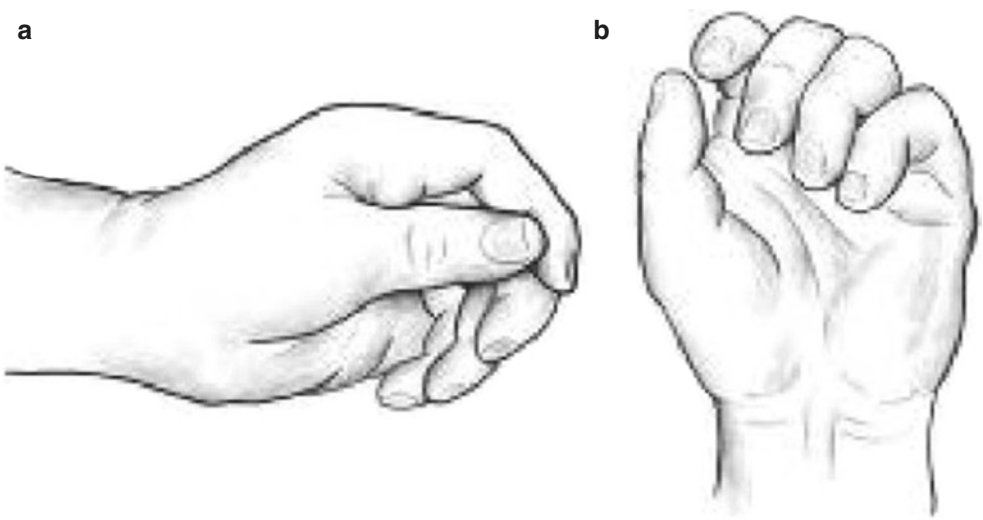


Fig. 2.1 Resting position of hands

4. Clamping. It is an action that two fingers approach each other to gently hold fine objects. If objects are clamped through extension of finger joints, volar interosseous muscles contract; if objects are clamped through flexion of finger joints, both the volar interosseous muscles and finger flexors contract.
5. Pushing. It is an action that wrist joint extends in the dorsal direction and palms touch objects under the joint action of the muscles of forearms and upper arms.
6. Plucking and poking. It is an action of sudden and rapid flexion and extension of fingertips. The action can be performed by either two fingers or one finger. This action is mostly performed when playing some musical instrument or in daily life and largely depends on the presence of fingernails.

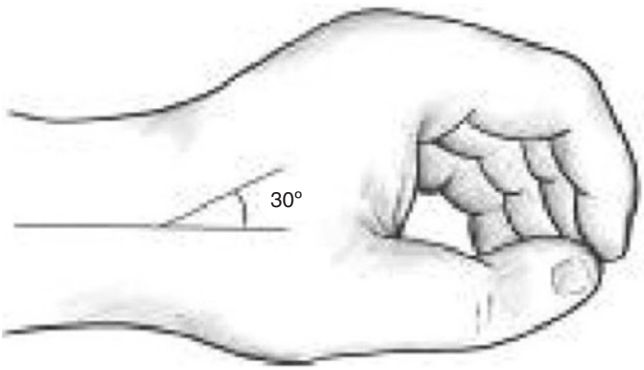


Fig. 2.2 Functional position of hands

Understanding the hand maneuvers (Fig. 2.3) is of vital importance to physical examination in case of hand injuries and diseases and to training on hand rehabilitation.

2.1.1.3 Sensory Function of Hands

The most sensitive site of human hands is the palmar side, especially the fingertip, and it is of vital importance to the completion of various complex actions. The importance of recovering the motor functions after nerve injuries was overemphasized previously, while not enough emphasis was laid on the recovery of sensor functions. Actually, any small sensor injury in hands will influence the motor functions of hands. In addition to the sensitive algesthesia, thalposis, thigmesthesia, and piesesthesia, hands have stereognosis, which is unique to hands. Stereognosis refers to a special function possessed by hands after the brain's comprehensive analysis and accumulation of the above various sensations and life experience. It enables humans to, without using the eyes, identify the category of objects merely by relying on hand touch and make some fine actions. The skin grafting used to treat the finger pulp skin defects may recover gross sensations, but stereognosis cannot be recovered. The

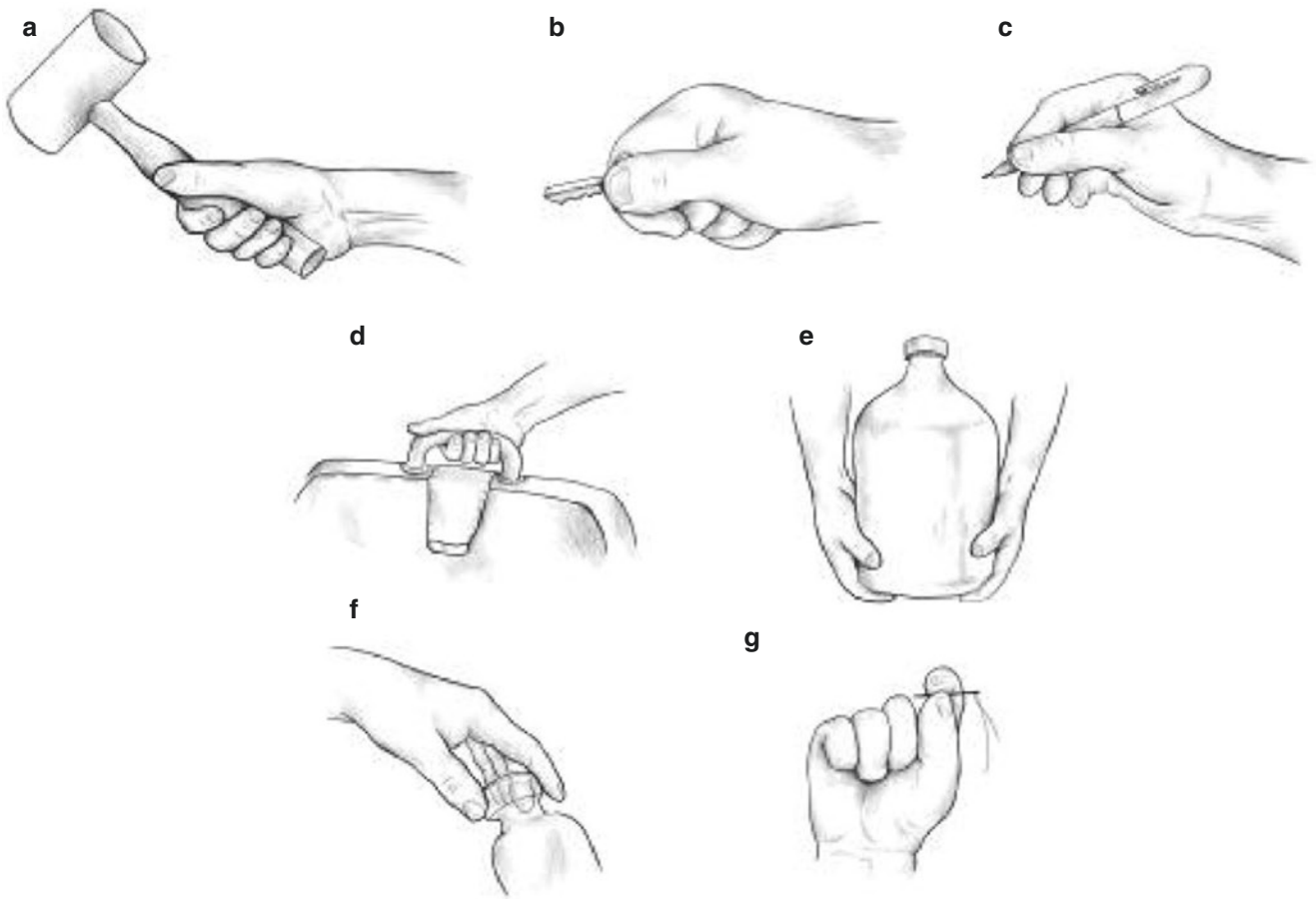


Fig. 2.3 Various actions of hands. (**a**) Gripping. (**b**) Revolving. (**c**) Holding. (**d**) Lifting. (**e**) Moving. (**f**) Twisting. (**g**) Pinching

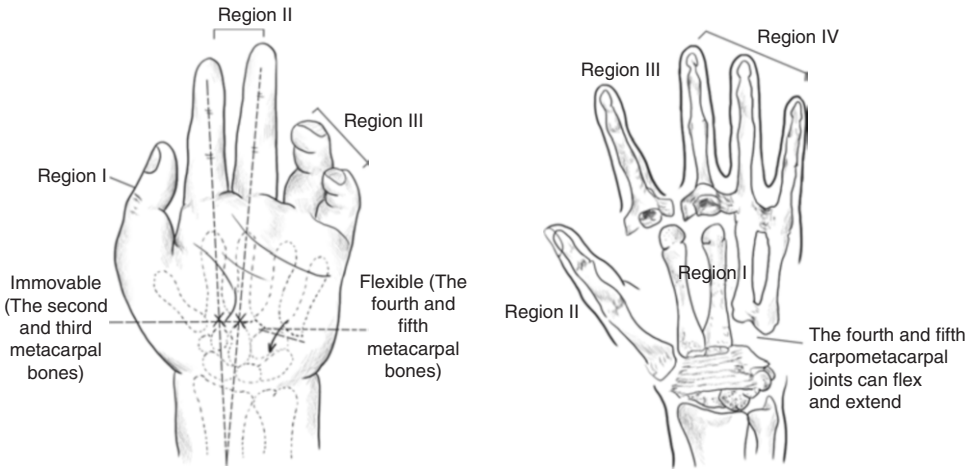


Fig. 2.4 Zonation of hand functions

emphasis on hand sensory functions is to emphasize the repair of sensory nerves during the repairing surgery after hand injuries [4, 5].

2.1.1.4 Zonation of Hand Functions

The hands can be divided into three zones according to structure and function (Fig. 2.4a) [1, 3]:

1. Zone I. It includes the thumbs and thenar area, which mainly perform palmar opposition and finger-to-finger functions.
2. Zone II. It includes the index fingers and middle fingers as well as the second and third metacarpal bones and can coordinate the finger-to-finger functions of thumbs; as the second and third metacarpal bones are relatively fixed, their longitudinal axis and radial longitudinal axis are nearly overlapped to produce the effects of transmitting force.
3. Zone III. It includes the ring fingers, little fingers, hypothenar, as well as the fourth and fifth metacarpal bones, and its main function is to consolidate grip strength and form transverse arch.

Hands are also divided into four zones (Fig. 2.4b).

2.1.1.5 Specificity of Hand Functions [3, 6, 7]

1. The difference between dominant hand and non-dominant hand is relatively large, and it is of certain significance to hand function reconstruction and medical identification.
2. The hands have a strong compensation function and can compensate for some functional disorders. Some functional disorders occurring during childhood are always surprisingly compensated for when one enters adulthood. Therefore, complicated hand functional reconstruction surgery is not recommended for children.
3. The rotation function of forearms will directly affect the normal functions of hands, so the rotation function should be well protected during treatment of forearm injuries.
4. Currently there is no ideal artificial hand with the functions of hand recovery when a comparison is made with the effect of artificial lower limbs.
5. Shortening of upper limbs (especially shortening of upper arm) has a small influence on the hand function, which is different from the big influence exerted by shortening of lower limbs on the foot functions.
6. There are more studies and applications using muscle tendons, muscles, nerve translocation or grafting to reconstruct hand functions than those on lower limbs, and the studies are profounder.

2.1.2 Surface Anatomy of Hands

1. Fingerprints. The dermatoglyphics of finger pulps of hands are different and can strengthen the sensations of finger pulps. Fingerprint is an important evidence in criminal investigation, but with the wide application of skin grafting or skin flaps in fingertip repair, various changes occur to the fingerprints, which is a problem worth attention in fingerprint identification.
2. Palm prints. The dermal ridges at the proximal and distal interphalangeal joints of fingers are basically parallel to the joint lines; the dermal ridges at the root of fingers are at the base of proximal phalanx and away from the metacarpophalangeal joints. The levels of distal palmar transverse striation and metacarpophalangeal joint are close, and the middle palmar transverse striation is the highest vaulted point of the vessels of palmar transverse striation. The intersection between the big thenar dermal

ridges and the middle finger ulnar longitudinal axis is equivalent to the ramification between the distal end of the vessels of deep palmar arch and the thenar muscles of median nerve. The proximal transverse crease of the wrist is parallel to the radial carpal articular surface (Fig. 2.5). Without subcutaneous tissues, the deep surface of palm prints is directly connected with flexor tendon sheath, so the incised wounds at this site will always involve the tendon sheath, flexor tendon, joint capsule, etc. [3, 8].

3. Nasopharyngeal fossa wrist joint is located at the neutral position, and the thumbs flex in the dorsal direction and become mildly adducted. One triangular concave from the tip to the distal side is visible at the dorsal radial side, and it is called nasopharyngeal fossa. The ulnar margin is the tendon of long extensor muscle of the thumb, the radial margin is the short extensor muscle of thumb, the distal end is the root of the first metacarpal bone, the proximal end is the styloid process of radius, and the bottom is the trapezium bone and the dorsal side of the tubercle of scaphoid bone. Cephalic vein, deep branch of radial artery, and superficial branch of radial nerve pass this fossa (Fig. 2.6). In case of rupture of the above tendons, the contour of the nasopharyngeal fossa becomes unclear; in case of fracture of scaphoid bone, the nasopharyngeal fossa becomes welling with the presence of tenderness [1, 3].
4. Palmar wrist muscle tendons. The wrist joint is located at the neutral position. If one's fist is clenched with force, several string-shaped elevated muscle tendons are visible or palpable in the transverse crease of the wrist at this time. The most obvious in the middle is the long palmar muscle tendon, the radial margin is the radial wrist flexor tendon, and the ulnar margin is the ulnar wrist flexor tendon. The tendons of superficial flexor muscle of the middle and ring fingers are present between the ulnar wrist flexor tendon and long palmar muscle tendon. The median nerve is located between the long palmar muscle tendon and radial wrist flexor tendon and slightly deviates to the side of long palmar muscle tendon. The position is relatively superficial, so attention should be paid when procedures are performed to block the median nerve of the wrist. Ulnar nerve, ulnar artery, and ulnar vein are located at the radial side of the ulnar wrist flexor tendon, and radial artery is located at the radial side of the radial wrist flexor tendon (Fig. 2.7) [2].
5. Muscular markers. The bulge at the palmar radial eminence is the muscle of thenar, the ulnar side is the muscle of hypothenar, and what are among the metacarpal bones of back of hands are the dorsal interosseous muscles. After median nerve or ulnar nerve injuries, these muscles will experience atrophy as a result [1, 3, 8].
6. Bone markers. Lentiform bone is located at the intersection between the distal side of the wrist transverse striation and the ulnar longitudinal axis of the little finger. The distal hamate bone jointly forms one of the attachment

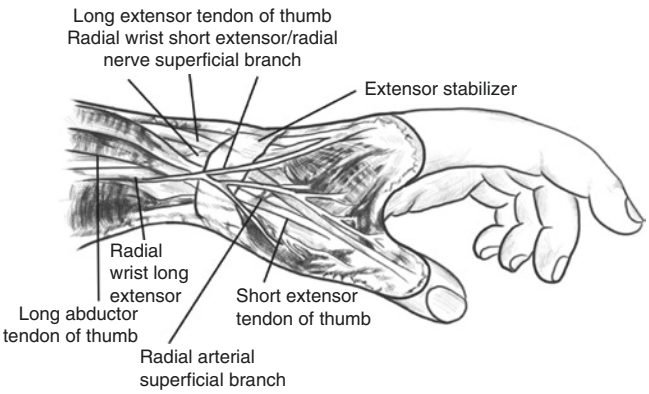


Fig. 2.6 Structure of nasopharyngeal fossa

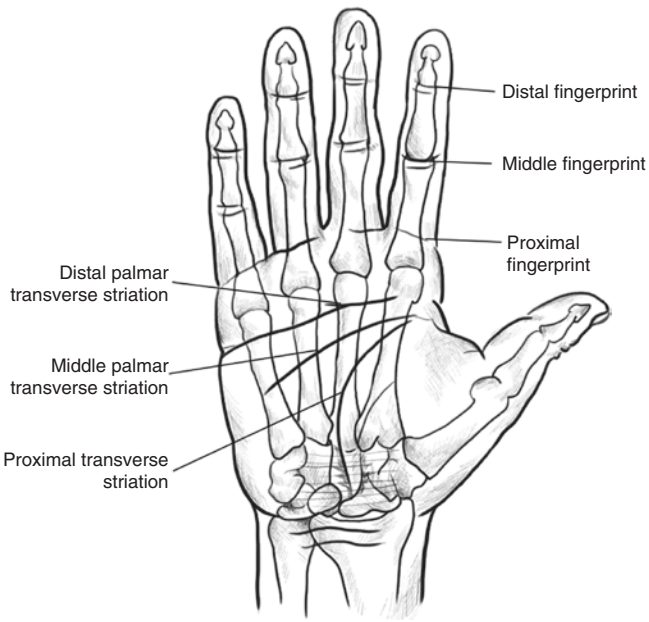


Fig. 2.5 Palmar dermal ridges

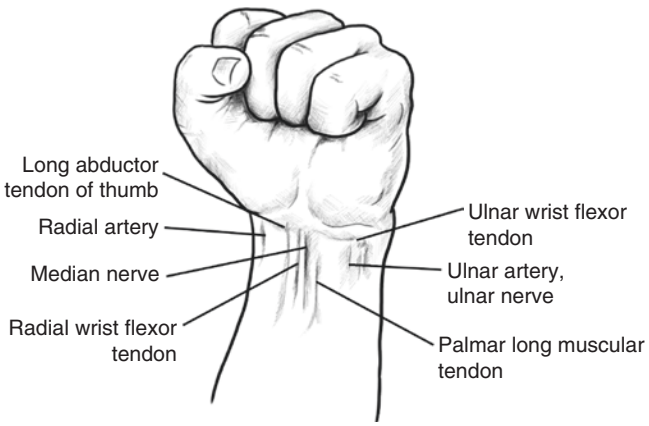


Fig. 2.7 Wrist palmar surface anatomy

points of wrist transverse ligament, and the ulnar nerves and the ulnar vessels are adjacent to the radial side. The tubercle of scaphoid bone is located at the intersection between the distal side of the transverse crease of the wrist and the radial longitudinal axis of middle fingers, it is the longitudinal axis intersection when the index, middle, ring and little fingers flex naturally, and it is also an important marker used to assess the presence of rotation displacement in case of metacarpal and phalangeal fracture reduction. The styloid process of radius and the styloid process of ulna are apt to be visible and palpable, and they are the markers from which we can know whether there is displacement fracture and deformities and whether the radial wrist joint and sub-ulnar and sub-radial joints are stable. In addition, the tenderness at the styloid process of radius can indicate the tenosynovitis of thumb short extensor tendon and thumb long abductor tendon, and the distal tenderness of the styloid process of the ulna indicates the possible presence of injuries of triangular fibrocartilage or ligament [8, 9].

7. Thumb web. The soft tissue space between the first and second metacarpal bones is called thumb web, equivalent to the fingerweb between the thumb and index finger. The first dorsal interosseous muscle is at the dorsal side under the skin, and the adductor muscle of thumb is at the palmar side. Under normal circumstances, thumb web can become abducted by 90°, and its role is to facilitate the palmar opposition of thumb, improve finger functions, and strengthen grip strength. In case of thumb web stenosis induced by adductor contracture or skin cicatricial contracture, hand functions will be obviously affected. The sensation of the dorsal skin of thumb web is the single dominance region of radial nerve, and it is clinically considered as one of the criteria used to judge radial nerve injuries [10–12].

2.1.3 Functional Anatomy of Hands

2.1.3.1 Skin Characteristics

1. Palmar skin
   (a) Sensitive skin: There are dense nerve endings structures and complex nerve net inside the palm skin and in the subcutaneous regions. Their density is higher than that of any site in the body, so they are very sensitive. The Meissner's corpuscle inside the finger pulp epiderm is mildly sensitive to tactile sensation, and the Merkel disks below it can perceive general tactile sensations. The Ruffini's corpuscle inside the dermis is correlated with thermal sensation, the Krause's corpuscle is correlated with cold sensation, the lamellar corpuscle at the deep layer of dermis is correlated with pressure sensation, and the pain sensation is transmitted via the nonmyelinated nerve fiber of the endings.
   (b) Relatively stable holding of object: There is no hair or sebaceous gland in the palm, so it can hold objects stably. Therefore, the palmar mass cannot be diagnosed as sebaceous cyst.
   (c) Thick horny layer: The horny layer of the palm is especially thick, so it can increase the resistance to physical injuries.
   (d) Relatively small mobility, which facilitates object holding: The palmar dermis and deep fascia or periost (fingertip or joint) are connected through a kind of complex fibroareolar fascia structure, forming fine and small septa which are filled with adipose tissues (the structure connected with periost is also called the bone skin ligament); therefore, the mobility of the palmar skin is relatively small, which facilitates object holding. In case of a lack of such structures after the skin flaps of other sites or the skin grafting is used to repair the palmar skin defect, the fine small or round object is apt to roll and fall when pinched.
   (e) Facilitating the flexion of fingers: The transverse striation of palms and fingers facilitates the flexion of fingers. The lack of transverse striation or abnormal distribution is common in patients with inborn dementia, and the palm print of "simian line" is common in patients with trisomy 21 syndrome [13].
2. Skin at the back of the hands
   (a) Thin and flaccid: The horny layer of the skin at the back of the hands as well as the stratum lucidum are thin, the quantity of subcutaneous tissues is small, the connective tissue is flaccid, and the number of veins and lymphatic vessels is large, so trauma is easily induced, and diffusion is apt to occur and swelling is obvious in case of infection.
   (b) Good elasticity: The skin at the back of the hands has good elasticity and high mobility, which facilitates the clenching of fists and full flexion of fingers.

2.1.3.2 Fingernails

The nail is a special structure at the end of fingers which includes nail plate, nail bed, onychostroma, eponychium, hyponychium, lunula, and medial nail groove [14, 15]. Nail plate is the derivative of stratum corneum, and the granular layer, stratum spinosum, and basal lamina of the skin are transformed into onychostroma and nail bed. The mechanism of lunula formation has not been clear, and the possible explanation is that it is formed after the cells in the stratum corneum experience light scattering. Eponychium is connected with the epidermis at the dorsal side of nail root; hyponych-

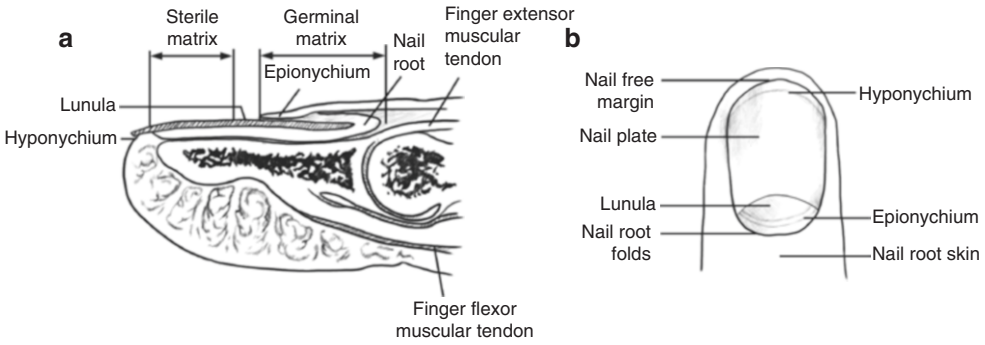


Fig. 2.8 Structure of fingernails. (**a**) Section. (**b**) Surface

ium refers to the extended part of nail bed at the finger free margin and is connected with the skin at the fingertip (Fig. 2.8). The functions of fingernails include the following: ① nail bed is closely connected with the phalangeal periost, and nail plate is hard, so they are the good protective layer of distal phalanx and also a kind of natural external fixture after the fracture of distal phalanx; ② the fingernails can prevent the finger pulp soft tissues from rotating toward the dorsal direction, making the fingertip to have a good pinching function; ③ the pressure suffered by the finger pulp is blocked by the fingernails, producing one counteracting force and making the finger pulp more obviously compressed, so that the finger pulp sensation can be enhanced; ④ the fingernails can help perform some special actions in daily life, such as peeling the fruit skin, scratching, untying the knots, and poking small objects; ⑤ the fingernails are also an important part for a person's overall beauty. Without nail, a finger appears to be weird. The time of nail regeneration varies from person to person and it usually needs 3–6 months [16–18].

2.1.3.3 Palmar Aponeurosis

Palmar aponeurosis is one layer of solid aponeurosis on the deep surface of palmar subcutaneous tissues, and its proximal aspect is connected with the palmar long muscle tendon and wrist transverse ligament; the distal aspect is divided into four bundles and ends at the index, middle, ring, and little finger tendon sheath and metacarpophalangeal joint accessory ligaments; the center takes the triangular shape and is relatively thick; the bilateral sides become thin and become extended into big thenar and small thenar fascia (Fig. 2.9). There is a connection of longitudinal fibers between the palmar aponeurosis and the skin, playing the role of fixing the palmar skin and enhancing the object holding function of palm [19, 20]. The tenacity of palmar aponeurosis makes it difficult for the palmar deep infection to penetrate in the palmar side, so in case of palmar infection the manifestations of the back of the hand are more obvious. The connection between palmar aponeurosis and flexor tendon sheath can make the metacarpophalangeal joint and interphalangeal joint flex but not extend when contracture occurs [21, 22].

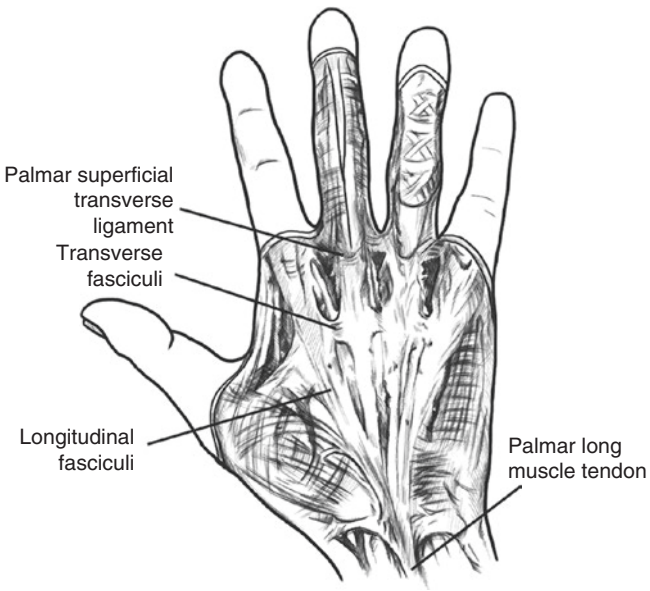


Fig. 2.9 Palmar aponeurosis

2.1.3.4 Bone, Joint, and Ligament

1. Bone
   (a) Phalanx: Except that the thumb has proximal and distal phalanx, the other four fingers consist of proximal, middle, and distal phalanx. From the perspective of the longitudinal plane of phalanx, the dorsal side is one straight line, the palmar side presents mild arch-shaped excavation, which may be because the flexor tendon is relatively thick; the cross section presents a dual bent line that protrudes in the dorsal side.
   (b) Metacarpal bone: It has five metacarpal bones; the shape of their longitudinal section is similar to that of the phalanx, and the cross section is similar to the triangle with the base extending to the dorsal side; such a structure makes the subcutaneous metacarpal section of the back of the hands relatively smooth, causing less friction and injuries of surface tissues. The palmar surface triangular space can hold the

volar interosseous muscles and maximally reduce palmar thickness to make it easy to hold objects.

(c) Carpale: Proximal row and distal row. The proximal row of carpale includes scaphoid bone, lunar bone, triquetrum and lentiform bone. The proximal sides of the lunar bone and some scaphoid bones correspond to the lower end of radial bone, forming the radiocarpal joint. The distal row of carpale includes trapezium, trapezoid, capitate bone, and hamate bone, and the former three work with the first, second, and third metacarpal bones, and the latter works with the fourth and fifth metacarpal bones to form carpometacarpal joints. Clinically, the problems in scaphoid bone, lunar bone, and trapezium are relatively common, which is somehow correlated with their anatomic structure. Most of the scaphoid bones are joint cartilage surfaces, and only the vessels at the rear and in the front of lumbar region enter the bones via the radiocarpal ligaments, so in case of fracture at the lumbar region or distal end of scaphoid bone, the ischemic necrosis of the proximal bones is apt to occur. The lateral surface of lumbar region of scaphoid bone can be directly subjected to the impact of radial styloid process in case of the radial deviation of the wrist joint, which is the anatomic basis for the occurrence of fracture. There are two or more ossification centers in the scaphoid bones. In case of the presence of two centers, dual scaphoid bone variation occurs, and the lumbar region is divided into two parts, and the symptom is usually misdiagnosed as old fracture [9]. The ulnar, radial, distal, and proximal sides of lunar bones are joint cartilage surfaces. Their blood supply is from the small vessels of palmar and dorsal ligaments. In case of complete dislocation and ligament rupture or chronic injuries and vascular occlusion, ischemic necrosis can occur. Lunar bone is formed by the fusion of two ossification centers. In case of nonfusion, the smaller one is called supra-lunar ossicle. Lunar bone is occasionally fused with triquetrum, but there is one pseudofracture line between the two, which is worth attention. Trapezium has three articular surfaces, of which the most important is the saddle-shaped joint formed with the first metacarpal bone. It is the joint with the largest range of motion among all the metacarpophalangeal joints and can directly influence the functions of the thumb. The trapezium has one sulcus at the slightly medial side of the palmar surface, and the radial wrist flexor tendon passes it; the dorsal side has two bulges, and the thumb long extends or tendon passes it there.

(d) Sesamoid bone: There are two small sesamoid bones at the palmar side of thumb metacarpophalangeal joints, and the thumb long flexor tendon is located between the two sesamoid bones. In case of the abnormal position of sesamoid bones, the incidence of congenital tenosynovitis increases. In addition, sesamoid bones can be occasionally seen at the ring and little finger metacarpophalangeal joints and the thumb interphalangeal joint and the index and middle finger metacarpophalangeal joint palmar sides.

(e) Accessory bones: All the hand accessory bones are near the wrist bones. This is frequently reported in literatures, but they are not common in clinical work. Understanding of accessory bones that may occur is of certain significance to the differential diagnosis of the injuries on the carpale (Fig. 2.10).

(f) Other rare circumstances: The density of phalangeal epiphysis of some children is very high. Such epiphysis is called ivory epiphysis, which will not affect

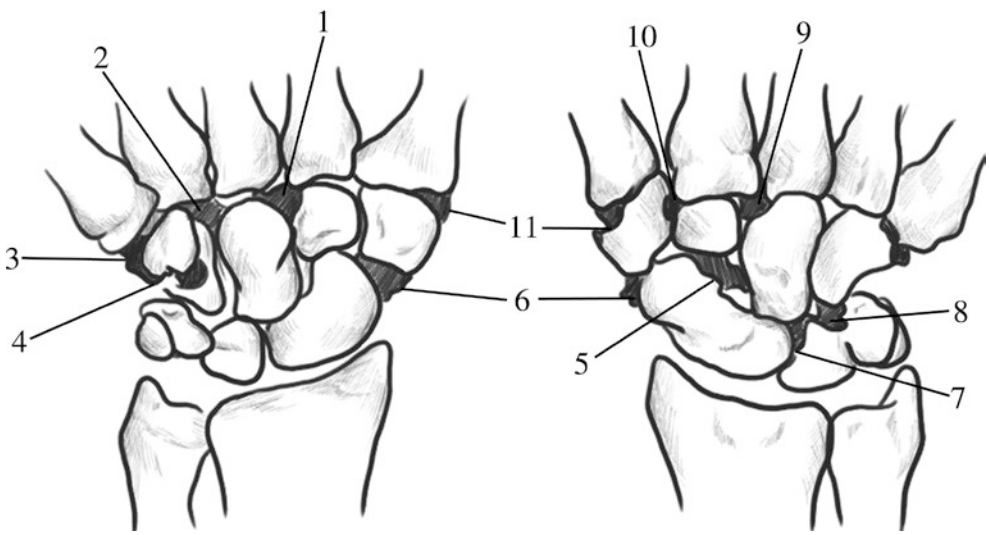


Fig. 2.10 Accessory bones of carpale

phalangeal development, but cannot be judged as epiphysitis without careful examination. Conic shape of the phalangeal epiphysis of children might be caused by insufficient central blood supply of epiphysis. It is generally considered that single finger conic epiphysis is a kind of congenital variation and multi-finger conic epiphysis falls into the category of pathological changes.

2. Joint and ligament
   (a) Interphalangeal joint: It is a kind of uniaxial joint. The thumb has only one uniaxial joint, and all the other four fingers have the proximal and distal interphalangeal joints. The joint capsule of interphalangeal joint is relatively weak and relies on the surrounding ligaments and muscle tendons to consolidate its stability. There is enhancement in collateral ligaments at the joint lateral side from anterior-inferior side of the phalangeal side superior part to the basal part of another phalanx. The accessory ligaments of metacarpophalangeal joints can be divided into sarciniform portion and fan-shaped portion, and the latter is also called collateral accessory ligament. The sarciniform portion of the collateral accessory ligament becomes flaccid in extension position and becomes tense in flexion position, so if the interphalangeal joint is fixed for too long in the extension position and the flaccid sarciniform portion contracts, it is difficult for the fingers to flex. On the contrary, the collateral accessory ligament is relatively tense in extension position and is flaccid in flexion position. However, as it is relatively weak, the braking position has a small influence on joint movement, so in case of contracture of collateral ligaments, the deformity can be corrected only by removing the sarciniform portion (Fig. 2.11). The joint palmar surface has palmar accessory ligament, namely, volar plate. The middle and distal sides of this ligament consist of fibrous cartilage plates, and the distal end is connected with the phalangeal periost; the proximal part is the fibrous tissues and connected with the proximal phalangeal periost; the two sides are connected with the collateral accessory ligament. In case of flexion of interphalangeal joint, the proximal fibrous membrane experiences folds and the cartilage plates will not become deformed, so that the finger flexor tendon can normally glide on the surface of cartilage plate. Because the distal end of the palmar accessory ligament is the junction between cartilage plates and periost, the difference between the two in terms of tensile strength is very large, so the chance of the occurrence of laceration considerably exceeds that of the junction between the proximal fibrous membrane and periost [1, 3].
   (b) Metacarpophalangeal joint: It is multi-axis spheroid joint; in addition to flexion and extension, it may have lateral movements to some extents at the extension position; thumb metacarpophalangeal joint also has the action of mild rotation. The ligament structure of metacarpophalangeal joints is basically the same as that of interphalangeal joint, but the role of the collateral ligament becomes more prominent, while the intensity of the palmar cartilage plate is weaker than that of the interphalangeal joint. The volar plate of the thumb metacarpal joint board contains two sesamoid bones, which is different from the metacarpophalangeal joint of other fingers. Sesamoid bones are often wrapped in the connection between the collateral accessory ligament and cartilage plates; if it is necessary to remove the sesamoid bones, the connection between the collateral accessory ligament and the cartilage plates should be repaired in the meantime [23].
   (c) Transverse ligament of head of metacarpal bone: There are three metacarpal transverse ligaments from the second to fifth metacarpal bone which are about 1 cm wide and are connected with the adjacent palmar cartilage plates. The main role of the ligaments is to control metacarpal so that overdispersion can be avoided, thereby strengthening the grip. There are digital proper neurovascular bundle in the volar ligaments, and there is interosseous muscular aponeurosis at the dorsal side. Between the first and second metacarpal bones, there is no such structure which facilitates thumb web's opening and thumb's actions [23].

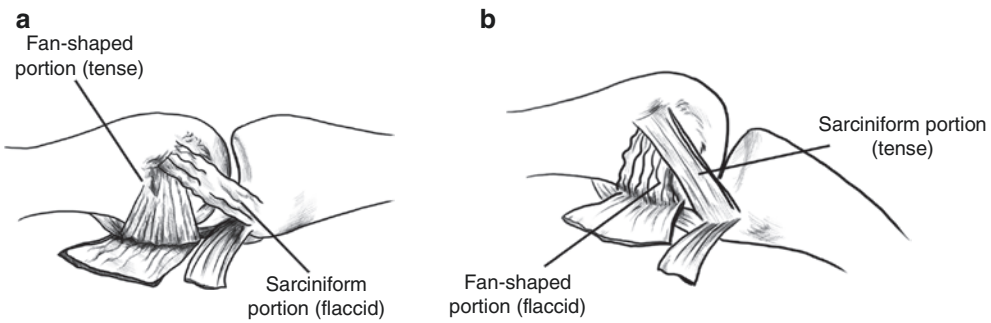


Fig. 2.11 Interphalangeal joint collateral ligaments. (**a**) Interphalangeal joint extension position. (**b**) Interphalangeal joint flexion position

(d) Intermetacarpal joints: There are three intermetacarpal joints at the lateral side of the second to fifth metacarpal bases, and the joint volar and dorsal ligaments are reinforced. This is a sagittal amphiarthrodial joint, whose joint capsule is connected with carpometacarpal joint.

(e) Carpometacarpal joint: It consists of metacarpal base and the distal row of carpal bones, and their composition and relation have been previously described. In carpometacarpal joints, the second and third carpometacarpal joint are the most stable and performs fewest action; the ring finger carpometacarpal joint flexes and extends by 15°; the little finger carpometacarpal joint flexes and extends by 30°. Carpometacarpal joint of the thumb is of particular importance and performs such actions as flexion, extension, adduction, abduction, and some rotational movements; once this joint suffers rigidity in nonfunctional position, most of the thumb functions will be lost [24].

(f) Intercarpal joint: It includes the proximal and distal row of intercarpal joints (midcarpal joints) and the joints between the carpal bones. Because the carpal positions of the midcarpal joint proximal and distal rows are not on the same plane, the joint line takes a U shape with a large opening. Its ulnar side is smooth and radial side is straight. When the lumbar region of the scaphoid bone experiences fracture, the active power line will pass the fracture line, which is one of the reasons that influence fracture healing (Fig. 2.12). There are many intercarpal joints, which are connected with interosseous ligament and palm and dorsal ligaments, and the blood supplied finds its way to each carpal bone. A gap is observed between the carpal volar radiocarpal ligament (including radial head ligament, radiolunate ligament, and radioscaphoid ligaments) and the capitohamate ligament, which happens to the distal port of the lunar bone and a weak area of the volar ligament structure. This gap becomes larger when the wrist extends in the dorsal direction and becomes smaller when the palm flexes. Therefore, if excessive force is exerted at the dorsiextension position, the lunar bone is apt to become dislocated toward the volar side (Fig. 2.13).

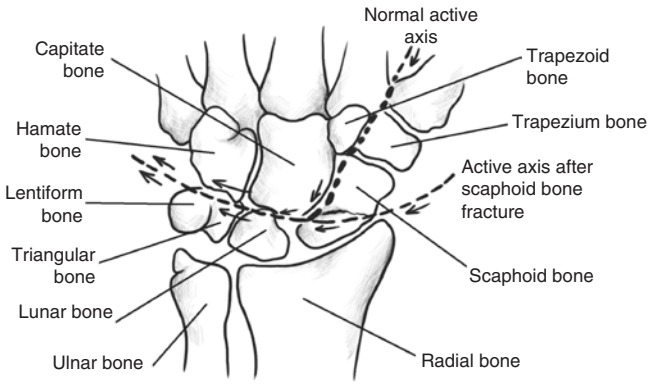


Fig. 2.12 Changes in force line of midcarpal joint and the force line after scaphoid fracture

(g) Radiocarpal joint: It consists of the lower end of the radius bone, scaphoid bone, and lunar bone. The physiological angles of the ulnar tilt by 20° and the volar angle by 15° are present on the articular surface of the lower end of radial bone, and it is not only an important sign for reduction of radial fractures at the lower end but also correlated with the normal movement range of the wrist. The flexion and extension movement of the wrist is jointly performed by the radiocarpal joint and the midcarpal joint; due to the different scopes of activities, the degrees of activity of the two joints are different (Fig. 2.14). After radiocarpal joint fixation is performed, the wrist still retains some flexion and extension functions [24].

(h) Inferior radioulnar joint: Inferior radioulnar articulation (distal radioulnar joint) has two articular surfaces: one is the rotary joint surface consisting of ulnar head circumferentia and the radial ulnar notch, and the other is flexion and extension articular surface between the distal side of ulna and the triquetrous cartilage. The two articular surfaces are connected to each other, making the articular cavity take an L shape. In radiocarpal arthrography, if the contrast agent finds its way to the

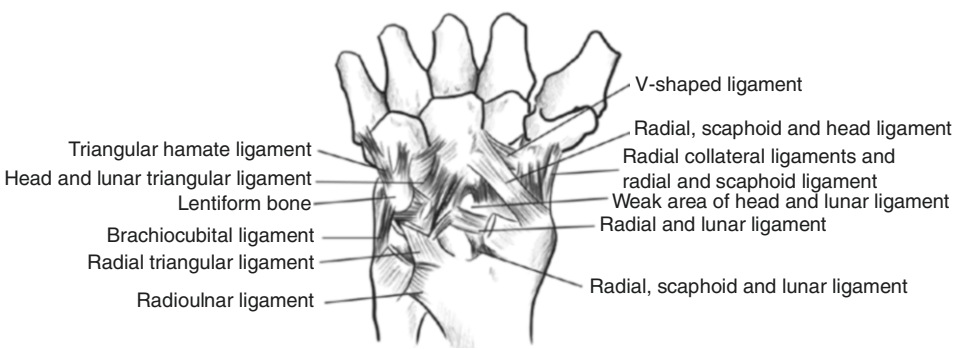


Fig. 2.13 Anatomic basis of lunar volar dislocation

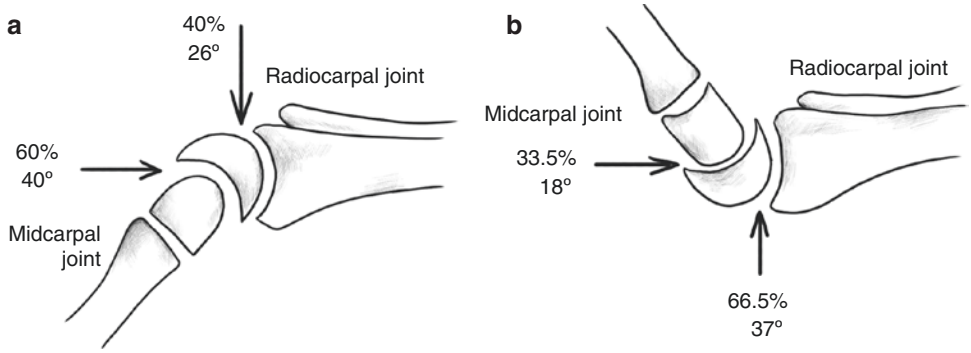


Fig. 2.14 The range of motion of radiocarpal joint and midcarpal joint during flexion and extension of the wrist joint. (**a**) In palmar flexion. (**b**) In dorsiextension

inferior radioulnar joint, it indicates the breakage of triquetrous cartilage. The inferior radioulnar joint and the superior radioulnar joint control the rotation of the forearm together; during forearm rotation, whether it is the superior joint or inferior joint, the radial bone rotates by surrounding the ulnar bone, and the ulnar bone is fixed and remains unchanged. As far as the anatomical characteristics are concerned, in case of double fracture of the ulnar and radial bones, both bones need rigid fixation, so that the rotational functions are trained during the early stage without the union of fracture being affected. In addition, when the forearm cannot rotate normally, if resection of ulnar head is performed, its rotational functions will not be improved. What is worse, new problems such as carpal instability will be caused.

2.1.3.5 Muscles and Tendons

During movement of limbs, dynamic muscles, synergistic muscles, and antagonistic muscles work. If the dynamic muscles are the extensor digitorum muscles, the synergistic muscles will be the extensor muscle of the wrist. Since the hand functions are complex, in order to fully perform hand functions or repair hand dysfunction, attention should be paid to the role of different muscle groups [25].

1. Rotator muscle of forearm. The action of forearm supination is mainly generated by the supinator muscle dominated by the median nerve. When the elbow position is flexed, the biceps muscle of the arm is synergistic muscle; when the elbow position is extended, the thumb long and short extensors and the thumb long abductor play the roles of synergistic muscles. The action of forearm pronation is controlled by the round pronator muscles and the quadrate pronator muscles dominated by the median nerve, and the synergistic action of other muscles is not obvious. When the forearm muscle suffers ischemic contracture, the pronator muscle is likely to be involved, resulting in the deformity of pronative position fixation.
2. Flexor muscle of the wrist. The dynamic muscle is the radiocarpus and long palmar muscle dominated by the median nerve and the ulnar flexor muscle of the wrist dominated by the ulnar nerves, and all finger flexor muscles are synergistic muscles. The palmar long muscle tendon is thin and about 14 cm long. It is the first-choice material for autogenous tendon transplantation and also one of the optional transposition tendons to reconstruct thumb opposition and abduction functions and the first interosseous dorsal muscular function and to treat unstable wrist joint. After the radial nerves are injured, translocating the wrist flexor tendon to the dorsal side of hands can reconstruct the functions of total extensor muscles of fingers [26].
3. Extensor muscle of the wrist. The dynamic muscles include the radial wrist long and short extensors and the ulnar extensor muscle of the wrist, which are all dominated by radial nerves. Extensors of fingers are their synergistic muscles. Translocating the radial wrist long extensor tendons can reconstruct the thumb opposition functions and the ring and little finger interphalangeal joint extension functions, and translocating the ulnar extensor muscle tendon of the wrist can reconstruct the thumb adduction and flexion functions.
4. Digital flexor. The digital flexors of the forearm are classified into profound and superficial layers. The superficial layer is the superficial digital flexor dominated by median nerve, and plays the role of proximal interphalangeal joint. Its anatomical features are as follows: the four superficial digital flexors are completely independent, the muscle belly can be substantially separated, and particularly the finger ring superficial digital flexor has the independent sarcolemma wrapping. Thus, the index, middle, ring and little finger flex the proximal interphalangeal joints, respectively. According to this feature, the superficial digital flexor tendon (especially the ring finger) is used for reconstruction of thumb opposing function, and the thumb can be independently used to perform abduction opposition function without the need to flex other fingers. The deep layer has profound digital flexor and thumb long flexor. The radial side of profound digital flexor is dominated by the median nerve, and the ulnar side is

dominated by the ulnar nerves and can flex the proximal and distal interphalangeal joint. The profound digital flexors of index fingers are independent, so index fingers can flex alone; the profound digital flexors of the middle, ring, and little fingers are connected together at the beginning part, so the three fingers cannot flex independently. Thumb long flexor is a completely independent single pinnate muscle which is dominated by the median nerve, and no lumbrical attachment is found on their chordae; after palm fracture, there are many proximal retractions, which can be found through dilation of wounds. Because of this, after finger rupture, the extensibility of the lower forearm tendon can be taken advantage of to directly move the proximal broken end to the tip of the thumb tip to rebuild its functions without the need of performing tendon grafting. In addition, the thumb flexor tendon passes the first metacarpophalangeal joint and enters the tendon sheath at the proximal phalanx, the process is very tough, and a greater strength can be generated during thumb movement. Here stenosis tenosynovitis is often caused, which is commonly known as "snapping finger" [27].

5. Intrinsic muscles. Though the hand intrinsic muscles are short, their starting and ending points are at the medial side of hands, so they are very efficient. Intrinsic muscles are usually classified into four groups [1, 3, 26].
   (a) Thenar muscle: It includes thumb short abductor muscle, thumb short flexor muscle, thumb opposing muscle, and thumb adductor muscle. Thumb short abductor muscle is dominated by the median nerve; it not only allows thumb abduction but also can assist thumb extension because some fibers end at the thumb extensor aponeurosis. During reconstruction of thumb abduction function, the displacement tendon insertion should pass the digital dorsum through the radial side of the root of thumb proximal phalanx and become fixed at the ulnar side to ensure that the thumb force will not be reduced. The superficial head of flexor pollicis brevis muscle is dominated by median nerve, and the thumb adductor muscle is dominated by ulnar nerves. In addition, the fibers of thumb short abductor muscle, thumb short flexor, thumb opposing muscle, and oblique head of adductor pollicis run relatively vertically with thumb web, and only the fibers of the thumb oblique head of adductor pollicis and the first dorsal interosseous muscles run in parallel to thumb web. Therefore, during the thumb web dilating orthopedics, the tendon insertion of the two posterior parts of muscles should mainly be cut. In case of excessive stripping of muscular insertions, the thumb adduction capabilities will be weakened, and the reconstructed thumb web takes the concave V-shape after the operation, which is inaesthetic.
   (b) Hypothenar muscle: It includes palmar short muscle and little finger abductor at superficial level and little finger short flexor and little finger opposing muscle at deep level, which are all dominated by ulnar nerves. The contraction of palmar short muscle can cause inferior fovea of hypothenar skin and slightly abduct the little finger. The little finger abductor muscle can not only abduct the little fingers but also help flex metacarpophalangeal joint, extend interphalangeal joint, and play the role of lumbrical muscle, because some fibers end at the aponeurosis of the digital dorsum of the little fingers. Little finger short flexor ends at the palmar ulnar side of proximal phalanx, so it can both flex the metacarpophalangeal joint and abduct little fingers. The little fingers act on palmaris so that the little finger and the thumb are mutually pinched.
   (c) Interosseous muscle: It is classified into volar interosseous muscle and dorsal interosseous muscle, both of which are dominated by ulnar nerves. There are three volar interosseous muscles, all of which are single pennate muscles. Their contraction makes the index, ring, and little fingers approach the middle fingers. There are four dorsal interosseous muscles, all of which are bipennate muscles. Their contraction keeps the index and ring fingers away from the middle and makes all metacarpal bones get close to each other. The abduction of little fingers and thumbs are controlled by their corresponding abductors (Fig. 2.15).
   (d) Lumbrical muscle: There are four lumbrical muscles which begin at the fibrous connective tissues of epitendineum of palmar profound digital flexor. The first and second lumbrical muscles are single pennate muscles, which start at the radial side of profound digital flexor tendon of index and middle fingers. Their chordae pass by the radial side of metacarpophalangeal joint and form the lateral funiculus of extensor aponeurosis jointly with the chordae of interosseous muscles, which are dominated by the median nerve; the third and fourth lumbrical muscles are bipennate muscles, which start at the two adjacent side of the middle-ring finger and ring-little finger profound digital flexor tendon. Their chordae also participate in the formation of the lateral funiculus of the finger extensor aponeurosis, which is dominated by ulnar nerves (Fig. 2.16). The single action of lumbrical muscles is to flex the metacarpophalangeal joint and extend the interphalangeal joint, but in daily activities, it acts with interosseous muscles, especially the volar interosseous muscles, to jointly work. It is due to this coordinating effect that the fingers can make a number of fine movements. In case of paralysis of lumbrical muscle and interosseous muscle, the current function reconstruction methods cannot restore fine finger functions satisfactorily. In case of ulnar nerve injuries, the interosseous muscles and the third and fourth lumbrical muscles will become paralyzed, the dynamic balance of finger joint will be lost,

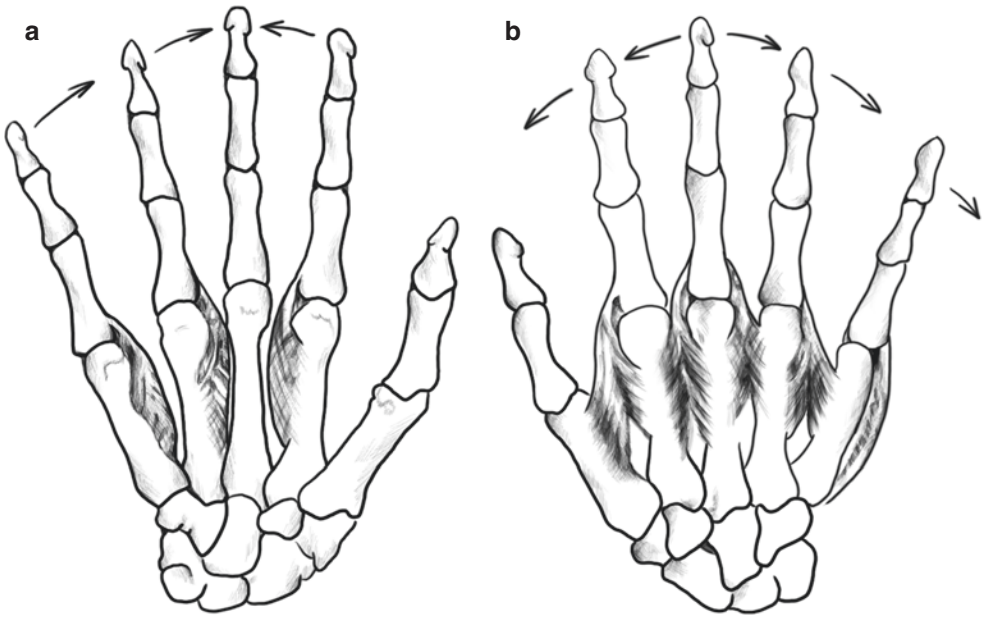


Fig. 2.15 Interosseous muscles. (**a**) Volar interosseous muscles. (**b**) Dorsal interosseous muscles

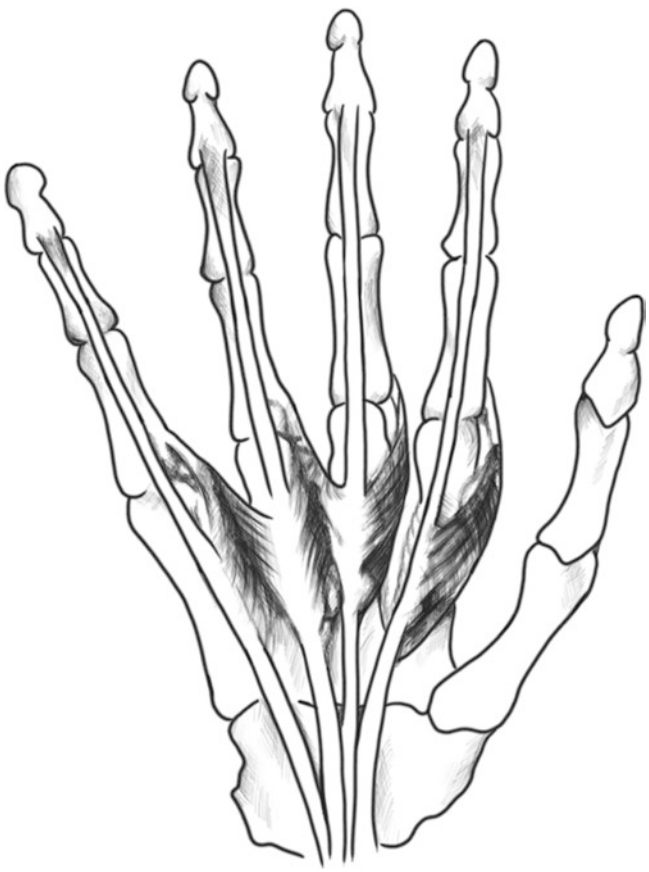


Fig. 2.16 Lumbrical muscles

causing the metacarpophalangeal joints of the ring and little fingers to experience hyperextension as well as deformity of claw hand in interphalangeal joint flexion. In case the median nerve is impaired, although the first and second lumbrical muscles become paralyzed, the interosseous muscles are normal, a relative balance between the flexion and extension strength of fingers can still be achieved, and the deformity of claw hand will not occur basically.

6. Finger flexor tendon. There are nine finger flexor tendons. Except that the thumb has one, all the other four fingers have one profound and one superficial flexor tendon, respectively. The ends of superficial flexor tendon are divided into two, which end on both sides of the palmar side of the middle phalanx; the profound digital flexor passes below and ends at the palmar side of the root of the distal phalanx (Fig. 2.17). The tendon fiber surface of finger flexor tendon has endotenon, and a number of fibers synthesize chordates. The surface is covered with perimysium, and the neurovascular bundle of tendons is distributed between the tendon posterior 1/2 endometrial membrane and perineurial membrane. Therefore, when tendon anastomosis is performed, the tendon should be sutured through the middle part of tendons and knotted in the front in order to reduce the impact on the blood supply of tendons. The outside of the entire tendon is covered with tendon outer membrane; there is a layer of synovium on the surface of epitendineum that has tendon sheath, and the synovia it secretes can nourish the muscle tendons. There is a thin layer of loose connective tissues around the epitendineum that has no tendon sheath. When tendon free grafting is performed, grafting tendon and this layer of loose connective tissue membrane should be removed together so that the grafting tendon is not prone to be directly adherent to new tendon beds, which facilitates the gliding movement of tendons [26].

Hand flexor tendon can be divided into five areas according to anatomical characteristics, which is of certain significance to clinical work. Area I is the tendon end area, extending from the insertion of superficial digital flexor tendon of middle phalanx to the insertion of profound digital flexor tendon. This area has only profound digital flexor tendon, and the effects of injury repair are good. Area II is the sheathing canal, previously known as no man's land. It extends from the distal transverse striation to the insertion of superficial digital flexor of middle phalanx. The tendon sheath in this area is narrow and thick, the deep and superficial flexor tendons overlap each other, the adhesion chance after injury repair is high, and the effect is poor. Area III is the palm area, extending from the distal end of wrist transverse ligament to the insertion of flexor tendon sheath, and the lumbrical muscles are attached to the surface of the profound digital flexor tendons in this area. As there are many loose connective tissues around the tendon and the tissues are covered by muscles, the effect after tendon repair in this area is good. Area IV is the carpal canal area, and nine flexor tendons are closely wrapped inside the solid carpal canals. The deep and superficial flexor tendons are wrapped by ulnar bursa, and the thumb long flexor tendon is wrapped by radial bursa. Nine tendons are arranged in three layers: the superficial layer consists of the middle and ring finger superficial digital flexors, the middle layer consists of index and little finger superficial digital flexors, and the deep layer consists of thumb long flexor tendon and four profound digital flexor tendons. In this area, if multiple tendons rupture on the same plane, the chance of mutual adhesion after repair is great, and the finger fine movements will be affected. Area V is the forearm area which extends from the juncture between muscles and tendons to the proximal side of carpal canal. In this area, they are many loose connective tissues. Even if multiple tendons on the same plane rupture, it is easy to use paratenons or fascial flaps to separate the stoma, thus reducing mutual adhesion. The zonation of thumb long flexor tendon is basically the same as the above zonation, and merely due to the absence of middle phalanx in thumb, area II is made relatively short. However, the thumb long flexor tendon of thumb metacarpophalangeal joint has one sesamoid bone on both sides to form a small bone fiber pipe, and the repaired tendons are apt to adhere at this site.

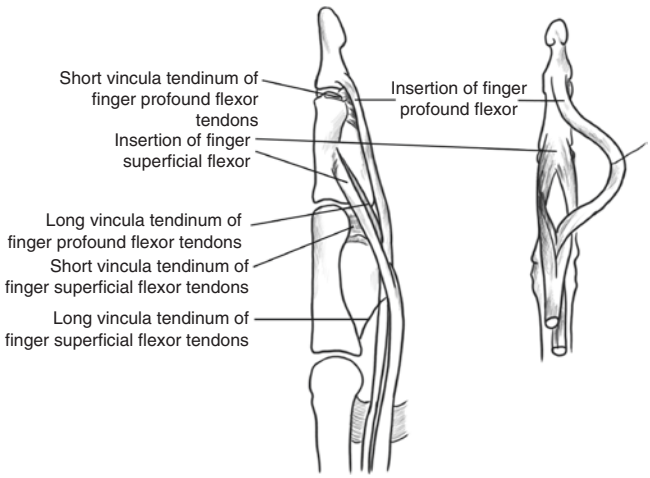


Fig. 2.17 Finger profound and superficial flexor tendons

Previous literatures stressed that it was not suitable to perform phase I repair after the injuries of flexor tendons in area II, but with the development of microsurgical techniques, the improvement of tendon suturing methods, the popularization of rehabilitation and physiotherapy as well as the adoption of various anti-adhesion measures, phase I repair can be performed on the flexor tendon injuries at any site, including the repair of the deep and superficial flexor tendons and tendon sheath. The superficial digital flexor tendons are of great importance to hand force and movement accuracy.

There is a certain difference in the gliding distance of all hand joints of finger flexor tendons, and understanding this is of clinical guiding significance to tendon repair or transplantation (Table 2.1).

Table 2.1 The gliding distance of all hand joints of finger flexor tendons (mm)

Finger flexor tendon		Distal interphalangeal joint	Proximal interphalangeal joint	Metacarpophalangeal joint	Wrist joint	Carpometacarpal joint	Total
Index finger	Superficial digital flexor	0	16	16	16		48
	Profound digital flexor	5	20	15	16		56
Middle finger	Superficial digital flexor	0	16	26	46		88
	Profound digital flexor	5	17	23	38		83
Ring finger	Superficial digital flexor	0	11	21	40		72
	Profound digital flexor	5	12	15	45		77
Little finger	Superficial digital flexor	0	8	17	40		65
	Profound digital flexor	3.5	11	15	45		74.5
Thumb			12	8	23	20	63

7. Finger flexor tendon sheath and bursa mucosa [28]
   (a) Finger flexor tendon sheath: The medial layer is the synovial tendon sheath and the outer layer is the fibrous tendon sheath. The synovial tendon sheath has two layers: the visceral layer synovium that directly covers the tendon surface and the wall layer synovium located at the medial surface of fibrous tendon sheath. The movement of tendons is the intersynovial movement. Synovial tendon sheath extends from the proximal side of metacarpophalangeal joint to the distal insertion of tendons. The thumb synovial tendon sheath is connected with the radial bursa mucosa, and the little finger synovial tendon sheath is connected with the ulnar bursa mucosa (Fig. 2.18). Fibrous tendon sheath is not a complete canal and presents irregular segmental distribution, and the shapes of joints and phalanxes are different. As the fibrous tendon sheath is connected with phalangeal periost and they form the osseofibrous canal, the flexor tendon may not necessarily leave the bone surface during gliding movement so that the fingers can be fully flexed. Synovia can be squeezed into the tendon fiber to nourish it. Usually, fibrous tendon sheath is called trochlea, and the second to fifth finger trochleas are ring-shaped (5) and cruciform-shaped (3); there are only three thumb trochleas, two of which are ring-shaped and one of which is oblique-shaped. As the functions of trochleas are special, it is necessary to perform selective repair of 1–2 trochleas when tendon transplantation is being performed. The integrity of annular ligaments 2 and 4 (A2 and A4) is very important, and can make the flexor tendons glide along the proximal and middle phalanxes and not render the appearance of bowstring-like bouncing to guarantee the performance of the above functions (Fig. 2.19).
   (b) Mesotendons and vincula tendinum: Both are the connections with deep tissues after the synovial tendon sheath wraps the tendons, so both of them are at the dossal side of the tendons. This structure takes the long patchy shape in palmar and wrist parts and is called mesotendon; it presents segmental distribution in the finger tendon sheath and is called vincula tendinum. Mesotendons and vincula tendinum contain the vessels, nerves, and lymphatic vessels that enter the tendons. Near the insertion of finger deep and superficial flexor tendons, the vincula tendinum is triangle-shaped and is relatively constant, and this is called short vincula tendinum; there are still long vincula tendinums which are thin and long at the proximal side of finger deep and superficial flexor tendons, whose variation in position, shape, and number is great, and number is usually 1–3 (Fig. 2.20). As the

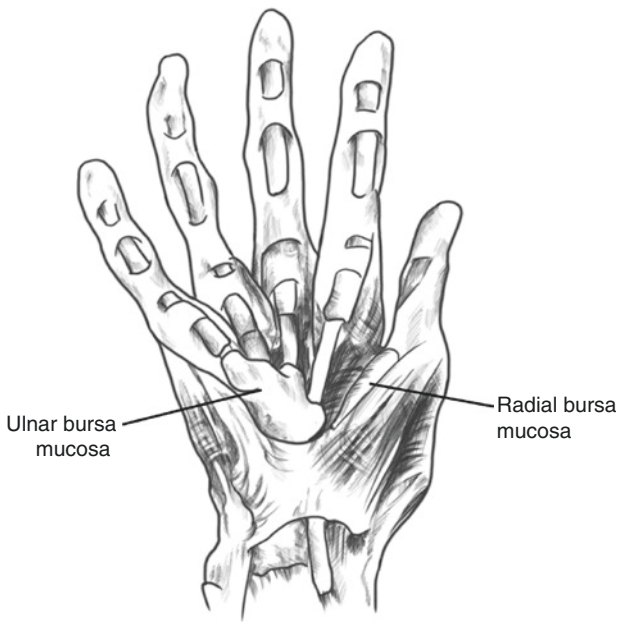


Fig. 2.18 Finger flexor tendon sheath and bursa mucosa

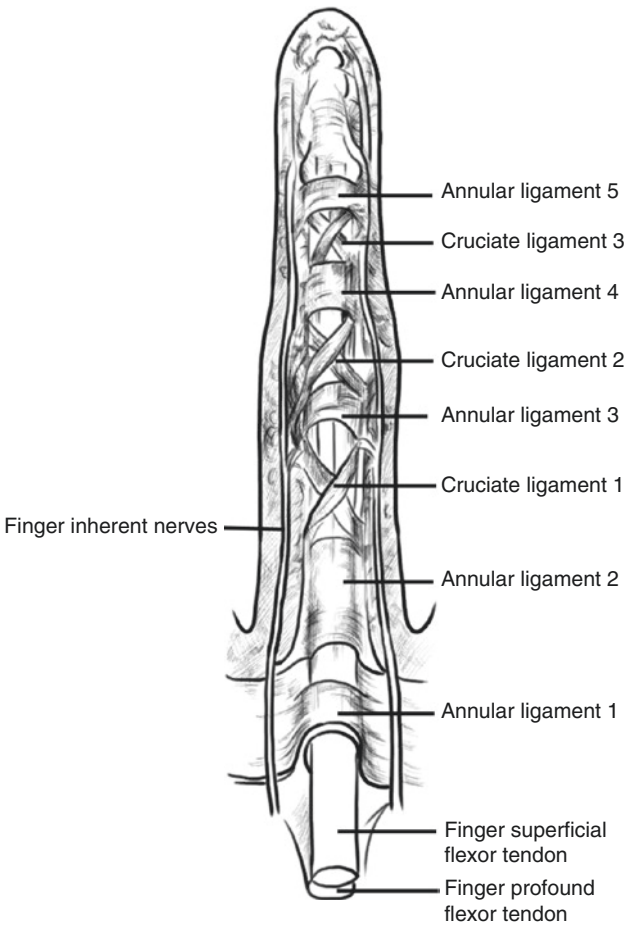


Fig. 2.19 Finger trochleas

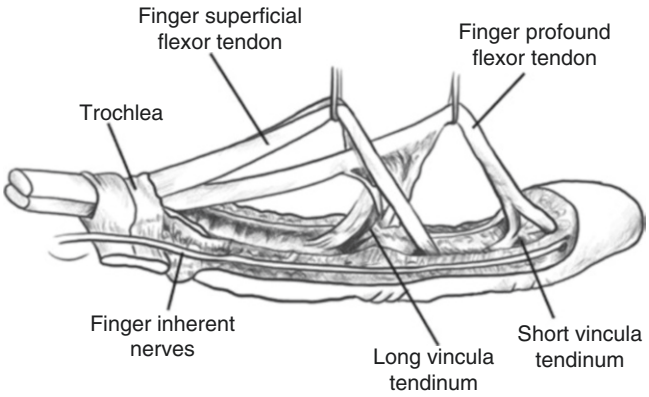


Fig. 2.20 Vincula tendinum of finger flexor tendon

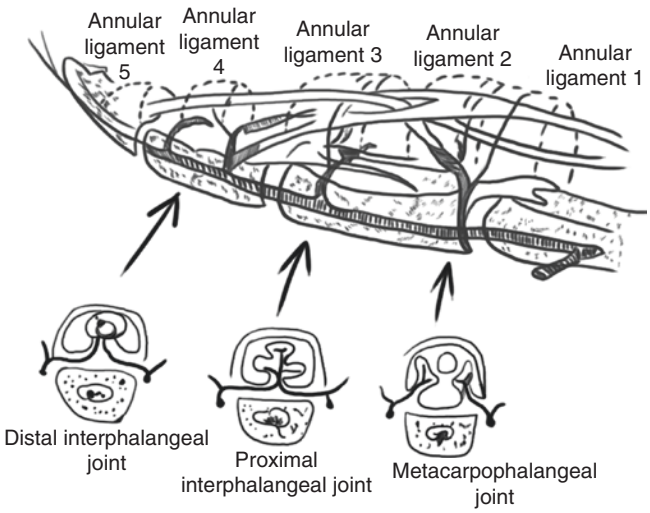


Fig. 2.21 Blood supply of finger flexor tendons

flexor tendon vincula in the tendon sheath is the blood-supplying channel, the blood supply of flexor tendons is also segmental, which is of vital importance to the simple resection of superficial digital flexor. So, attention should be paid to protect the long vincula tendinum of profound digital flexor tendons. What is more, the short vincula tendinum of superficial digital flexors should be preserved because the long vincula tendinums of most profound digital flexor tendons originate from the short vincula tendinums of the superficial digital flexor tendons.

(c) Synovia: Synovia can not only lubricate tendons and reduce physical injuries but also provide nutrients to the vessel-free areas of the anterior 1/2 part of the finger flexor tendons. With the development of microsurgical techniques, if the tendons are injured, it can be used to repair the tendon sheath while repairing tendons, greatly helping to heal tendon and reduce adhesion.

(d) Relation of tendon sheath with finger vessels and nerves: As the structures of finger flexor tendons at metacarpophalangeal joint and interphalangeal joint are different, the ways in which vessels enter the tendons via the vincula tendinum vary. As there are only long vincula tendinums of superficial digital flexor near the metacarpophalangeal joints, finger vascular vessels enter the finger superficial flexor tendons via this site, but no vessel enters into profound digital flexor; the inherent vascular branches of the proximal interphalangeal joint enter the tendons via the short vincula tendinum of finger superficial flexor tendons and the long vincula tendinum of finger profound flexor tendons as well as supply blood to the finger deep and superficial flexor tendons; there is no finger superficial flexor tendon in the distal interphalangeal joint, and the vessels enter the finger profound flexor tendons via the short vincula tendinum of finger profound flexor tendons (Fig. 2.21). When we inject methylene blue into the tendon sheath, the transverse fine pathways are connected with the finger neurovascular bundles near all interphalangeal joints, the methylene blue flows out via this channel and segmentally stains the finger neurovascular bundles, which can explain the mechanism of anesthesia in the finger flexor tendon sheath.

8. Finger extensor tendon. Interosseous muscle and lumbrical muscle composite device can help extend interphalangeal joints; the insertion of thumb short abductor joins the thumb long extensor and has the effect of extending thumb. In addition, the remaining extensor tendons are hand extrinsic muscles, dominated by radial nerves.

(a) Hand dorsal finger extensor tendon: The thumb has one thumb long extensor tendon and short extensor tendon. The former ends at the dorsal side of the basal portion of the distal phalanx and the latter ends at the dorsal side of the root of the proximal phalanx. Thumb short extensor tendon is of vital importance to the stabilization of thumb metacarpophalangeal joint. Even if the thumb long extensor tendon is normal, the thumb metacarpophalangeal joint cannot be extended completely after rupture of the thumb short extensor tendons, so the two tendons should be repaired. This can be easily neglected by non-hand surgeons. Finger total extensor tendons are distributed in the fan shape in the second and fifth fingers at the back of the wrist, and they are mutually connected with oblique fibers among the hand dorsal finger extensor tendons. This structural feature can be manifested as follows: after the rupture of one certain extensor tendon, the metacarpophalangeal joint can still retain a certain extending function under the traction of adjacent extensor tendons. In addition,

both index finger and little finger has one independent inherent extensor tendon and both of them are located at the ulnar side of the finger total extensor tendon. After the rupture or paralysis of finger total extensor tendons, the index finger and little finger can still extend, which is called paralysis syndrome (Horner's sign). In case of old injuries in thumb long extensor tendons, the index finger inherent extensor tendon is the commonest grafting tendon [1, 3].

(b) Digital dorsal extensor tendon: As the digital dorsal extensor tendon is thin, it is called digital dorsal aponeurosis and is a relatively complex digital dorsal anatomic structure. Digital dorsal aponeurosis consists of finger total extensor tendon, interosseous muscle tendon, and lumbrical muscle tendon. The extended part of finger total extensor tendon at the dorsal side of proximal phalanx is the digital dorsal aponeurosis central tendon, which ends at the proximal side of middle phalanx and mainly serves to extend metacarpophalangeal joint and proximal interphalangeal joint. Interosseous muscle tendon and lumbrical muscle run toward the distal side via the dorsal side and volar side of transverse ligament of the head of metacarpal bone, the two join together to form the lateral tendon of the digital dorsal aponeurosis at the lateral side of proximal phalanx, and then one medial cord is separated to join the central tendon. At the similar level, the lateral cord diverges from the central tendon and joins the lateral tendon, and the lateral tendon moves to the dorsal side of middle phalanx by passing by the lateral side of proximal interphalangeal joint. Finally, the lateral tendons at the ulnar and radial sides join together and end at the fundus of the dorsal side of the distal phalanx, and mainly serve to extend the distal interphalangeal joint and help extend the proximal interphalangeal joints. At the insertion of proximal lateral tendon, there is connection of transverse triangular ligaments between the two lateral tendons; the superficial surfaces at both sides of proximal interphalangeal joints have one layer of relatively thin fibrous tissues, they join together near the insertion of central tendon by passing by the lateral tendon, the oblique part is called Cleland ligament, the transverse part is called Grayson's ligament, and all the above three kinds of ligaments have the functions of preventing the over-translocation of lateral tendons and coordinating the proximal and distal interphalangeal joints (Fig. 2.22). The execution of the digital dorsal aponeurosis functions relies on the balance of finger flexor muscle strength and the balance between the finger total extensor and the muscular strength of interosseous muscles and lumbrical muscle, and the latter is particularly important (Table 2.2). When the muscular strength of interosseous muscles and lumbrical muscles is weakened, deformity of claw hand will occur (hyperextension of metacarpophalangeal joint and flexion of interphalangeal joint). When the muscular strength of the two is too high, swan-neck deformity occurs (flexion of metacarpophalangeal joint and distal interphalangeal joint and hyperextension of proximal interphalangeal joint). In case of rupture of insertion of central tendon

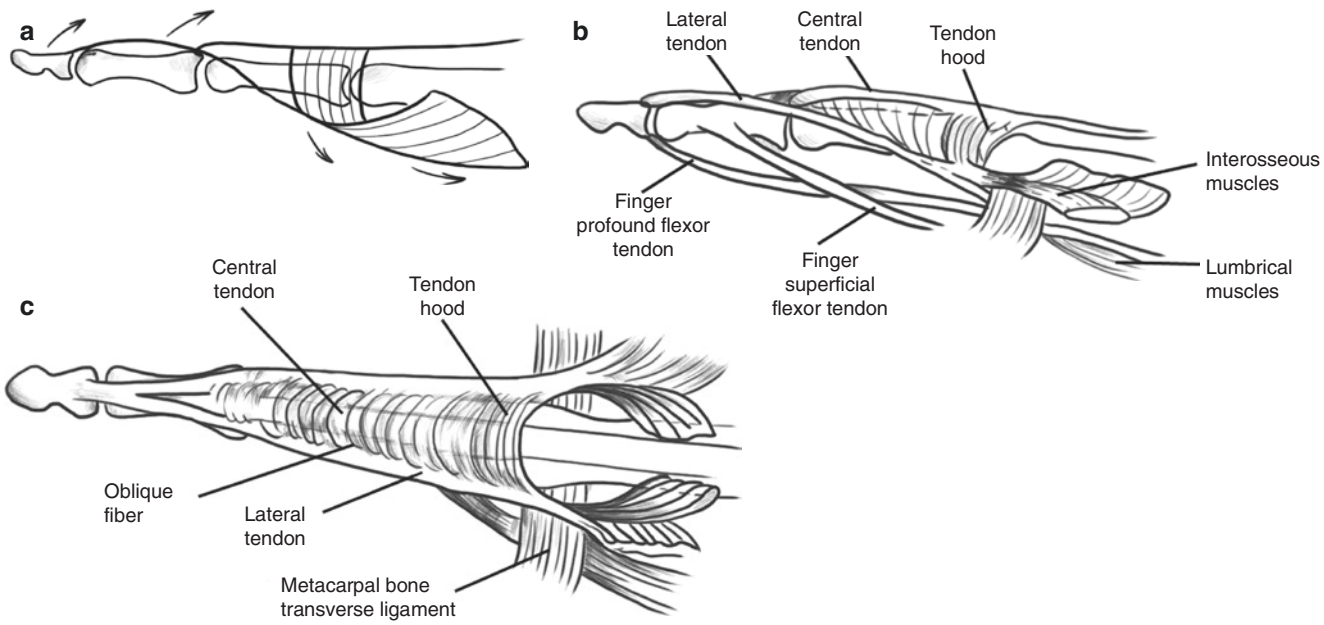


Fig. 2.22 Digital dorsal aponeurosis

or the rupture of medical cord, triangular ligament and Cleland ligament, boutonniere deformity may occur; in case of rupture of insertion of digital dorsal aponeurosis, mallet finger deformity will occur (Fig. 2.23) [1, 3].

Thumb digital dorsal aponeurosis has the thumb long extensor tendon as the main body, thumb short abductor and thumb short flexor chordae are added to the radial side, thumb adductor chordae are added to the ulnar side, so the ulnar surface is more rough and robust. The main function of thumb short extensor tendon is to stabilize the metacarpophalangeal joints, making it easy for the thumb long extensor to give full play to the thumb extending function. It should be noted in clinical work that at the dorsal side of thumb proximal phalanx, the above chordae mutually present the membrane-shaped connection, and only the main tendon becomes slightly thickened. In case of rupture of the thumb long extensor tendon at this site, those who do not understand this usually seek the broken ends of "recovery" at the proximal side. In fact, as the chordae and adjacent tendons are connected, the thumb long extensor tendon will not retract obviously when it ruptures at this site, and the thickened aponeurosis at the proximal side of incision is the proximal broken end.

Table 2.2 Factors of muscle strength balance of finger joints

Joints	Extension strength and flexion strength
Metacarpophalangeal joint	Finger total extensor, interosseous muscle, lumbrical muscle, profound digital flexor, superficial digital flexor
Proximal interphalangeal joint	Finger total extensor, interosseous muscle, lumbrical muscle/profound digital flexor, superficial digital flexor
Distal interphalangeal joint	Interosseous muscle, lumbrical muscle/profound digital flexor

2.1.3.6 Osseofibrous Canal

1. Carpal canal. It is the main osseofibrous canal of the wrist. Its top is the wrist transverse ligament, the posterior wall is the radiocarpal joint and midcarpal joint, the radial wall is the tubercle of scaphoid bone and tubercle of trapezium, and the ulnar wall is the lentiform bone and hook of hamate bone. Wrist canal has four digital profound flexor tendons, four superficial flexor tendons, one thumb long flexor tendon, and medial nerves. Finger flexor tendon is wrapped by ulnar bursa, thumb long flexor tendon is wrapped by radial bursa, and the medial nerves are located outside the bursa mucosa, and the position is the most superficial (Fig. 2.24). If the carpal canal cavity narrows (e.g., displacement fracture), or the volume of the content enlarges (e.g., bursitis), or new content appears (e.g., space-occupying mass and variant muscle bellies), the intracavity pressure increases. During long-term flexion and extension, the median nerve is first subject to the changes in this pressure. Once corresponding nerve dysfunction occurs, carpal canal entrapment syndrome occurs.
2. Ulnar canal. It is located at the wrist ulnar side: the anterior wall is the wrist palmar ligaments and short palmar muscle, the posterior wall is the wrist transverse ligament and pisohamate ligament, the ulnar wall is the wrist ulnar flexor muscle tendon, lentiform bone, and little finger abductor, and the radial wall is the hook of hamate bone. The ulnar canal is divided into proximal, middle, and distal segments: the proximal segment is called Guyon canal, equal to the bifurcation between ulnar nerve deep and superficial branches; the middle segment is called lentiform-hook canal, which is one segment of oblique space between lentiform bone and hook of hamate bone and covered by little finger short flexor; the distal segment is called opposing muscle canal and located deep in the little finger opposing muscle. In the ulnar canal, the ulnar nerve, ulnar artery, and two ulnar veins pass by; in case of compression of ulnar nerve at Guyon canal, abnormality occurs to the sensation and movement of the

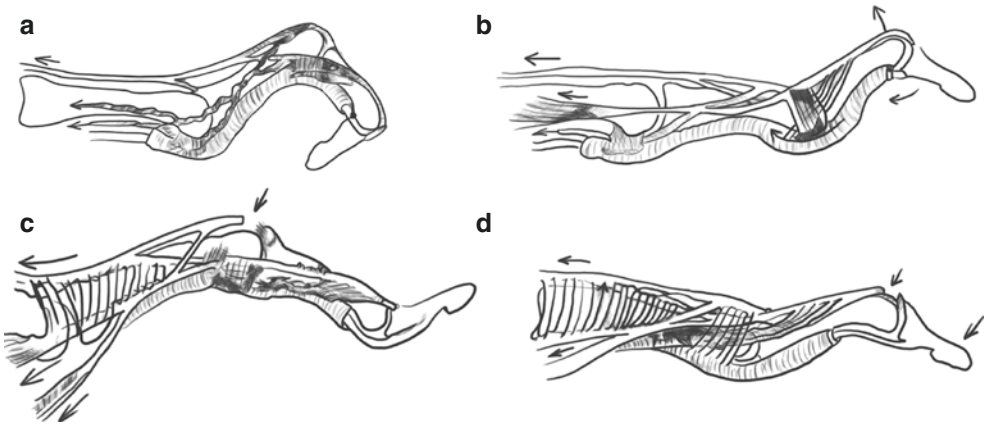
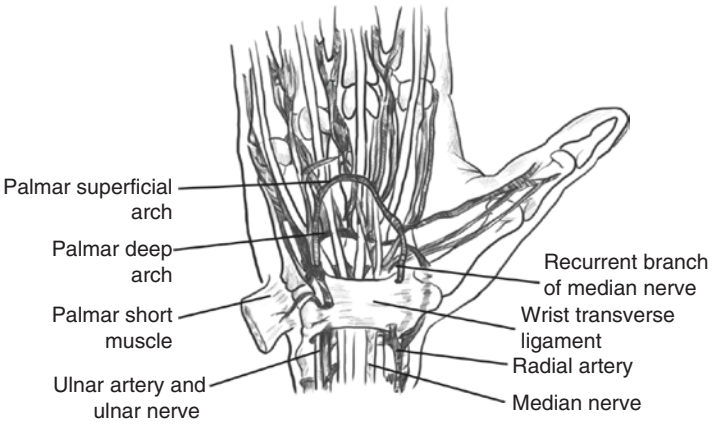


Fig. 2.23 Deformities induced by digital dorsal aponeurosis. (**a**) Claw hand deformity. (**b**) Swan-neck deformity. (**c**) Boutonniere deformity. (**d**) Hammer finger deformity



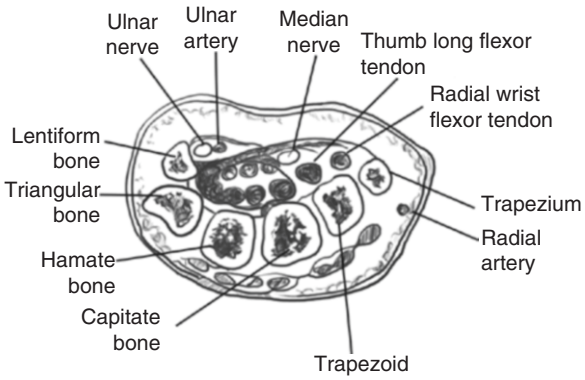
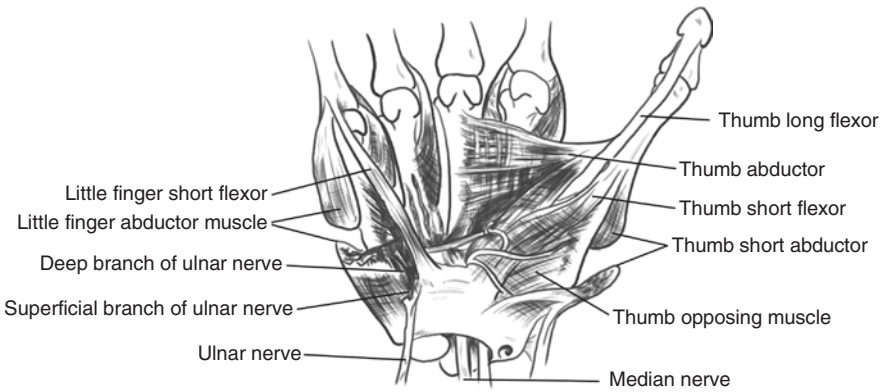


Fig. 2.24 Carpal canal

Fig. 2.25 Ulnar canal



hand-dominated areas; in case of compression of lentiform-hook canal, only inherent muscular dysfunction occurs without abnormal sensation; in case of compression at the palmaris canal, no disorder in addition to sensory disturbance is observed in little finger abduction function (Fig. 2.25).

2.1.3.7 Vessels

1. Palmar arch. The terminal branches of ulnar and radial arteries form deep and superficial arches at the palm to guarantee the blood supply of the hand (Fig. 2.26). The blood of about 60% of the superficial palmar arch is supplied by the terminal branch of ulnar artery, the blood of about 30% of ulnar and radial arteries is supplied by the terminal branch of ulnar and radial arteries in a balanced way, and various rare variations are seen in other 10%. The deep palmar arch is formed mainly by the terminal branches of radial artery, and about 95% is anastomotic with the terminal branches of ulnar artery, forming a complete deep arch; the remaining 5% is not anastomotic with the terminal branches of ulnar artery. It is worth noting that in case variations are present in superficial palmar

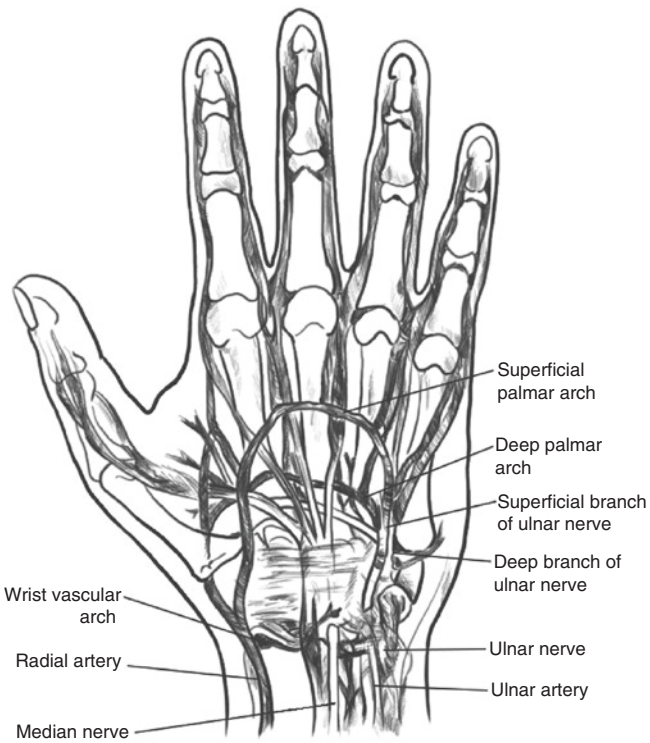


Fig. 2.26 Vessels of palmar arch

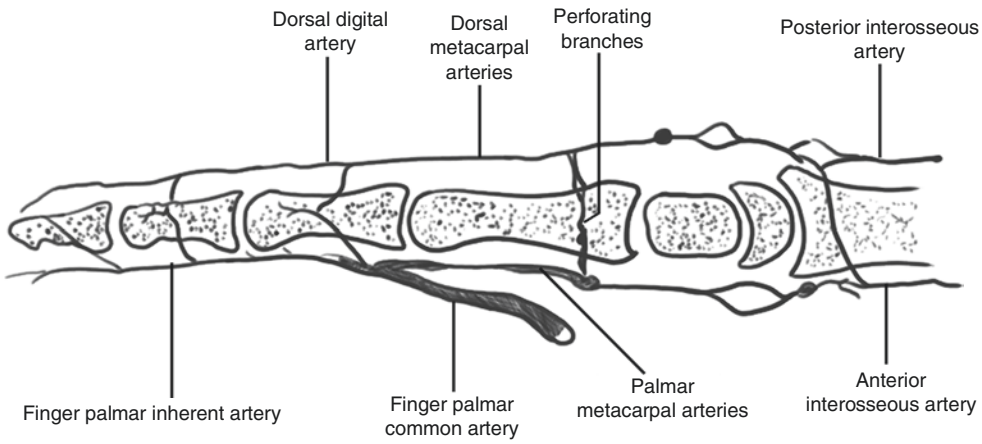


Fig. 2.27 Finger arteries

arch, the deep palmar arch is usually not complete; if the ulnar or arterial artery suddenly cannot supply blood, the corresponding hemi-side of hands will experience ischemic necrosis. So when ulnar and radial arterial ligation is performed or the ulnar and radial arterial flaps are removed, conventional advanced examinations should be carried out to examine whether the palmar arch is complete [29].

2. Digital arteries. They include inherent artery and dorsal digital artery; the former is the main source of finger blood supply. About 80% of thumb inherent arteries stem from the main thumb artery originating from the radial artery, and most of thumb dorsal digital arteries stem from the deep branch of radial artery and the first dorsal metacarpal arteries of their branches. The index finger radial inherent arteries mostly originate from the deep palmar arch or deep and superficial palmar arches, and the ulnar inherent arteries originate from the digital common arteries; the middle and ring finger inherent arteries originate from the digital common arteries; the little finger radial inherent arteries originate from the digital common arteries, and the ulnar inherent arteries originate from the deep and superficial palmar arches. The dorsal digital arteries of four fingers originate from the dorsal metacarpal arteries which are usually thin; the bilateral communications take the network shape, and they are connected with inherent arteries and mainly supply blood to the hand proximal and middle dorsal structures (Fig. 2.27). The ulnar and radial inherent arteries of each finger are mutually communicated with the joint adjacent and fingertip sites, where the anatomical basis of finger lateral retrograde is land flap.
3. Dorsal metacarpal arteries. There are four dorsal metacarpal arteries. The first dorsal metacarpal artery originates from the deep branch of radial artery, and the other ones from the second to the fourth are formed by the anastomosis between the perforators of deep palmar arch and the dorsal carpal arterial net. Each dorsal metacarpal artery has communicating branches at fingerweb and digital common arteries; antegrade or retrograde flaps are formed at the back of the hand with one certain dorsal metacarpal artery as the axis. As the position of the first dorsal metacarpal artery is relatively constant, the terminal branches reach the dorsal side of index finger, so it serves as the axis to make an island skin flap with vessel pedicles 4–5 cm long and it can be used to repair the skin defect of thumb web and the dorsal side of thumb [30, 31].
4. Dorsal interosseous artery. It is the dorsal branch of dorsal interosseous artery and the vessels that nourish the forearm extensor group. The upper segment has many cutaneous branches to supply the blood to the forearm dorsal skin, and the terminal branches, ulnar and radial arterial wrist dorsal branches, and the deep palmar arches form the wrist dorsal vascular network. Therefore, the proximal skin of the dorsal side of forearm can be utilized, and the distal side of dorsal interosseous artery serves as the pedicles to make retrograde island skin flaps to repair the skin defect of the hand back, especially to repair the soft tissue defects of the skin at thumb web. The appearance is beautiful; the trauma in this way is smaller when compared with the utilization of ulnar and radial artery as the pedicles without having to sacrifice the main vessels.
5. Finger veins. Finger palmar veins are the accompanying veins of the inherent arteries. The branches are few and fine, and the number and position are not constant. The vessel wall of dorsal digital veins is thicker than that of the palmar ones. The number of their communicating branches is large, and their distribution is always constant. The valve of the finger veins is open from the palmar side to the dorsal side, which decides that the dorsal veins are the main entrance of finger blood backflow. Therefore, during reconstruction of finger blood supply, whether the dorsal digital veins are smooth is a problem of vital importance.

2.1.3.8 Nerves

1. Ulnar nerves
   (a) The only muscle dominated independently by ulnar nerves at forearm is ulnar flexor muscle of the wrist, which has a high muscle strength, and the ulnar nerve muscular branches are mainly located at the proximal end of the muscle belly, so clinically the insertion of this muscle at lentiform bones can be cut to perform separation until the vascular nerve bundle enters the muscle belly and to move tendon ends upward to the upper arm, which is a good method to reconstruct the function of bending the elbows.
   (b) Ulnar nerves emit the volar cutaneous branches from the middle lower segment of forearm and dominate the sensation of palmar ulnar skin; the nerves emit the cutaneous branch at the back of the hand at the site 5–7 cm away from the proximal side of wrist joint and dominate the sensation at the ulnar side of the back of hand and the dorsal side of the proximal segment of little finger. Therefore, in case of ulnar nerve injuries at the dorsal part of the wrist, the sensation at the above sites is still present.
   (c) In the wrist ulnar canals, the ulnar nerves are divided into deep and superficial branches: the deep branch dominates hypothenar muscle, interosseous muscles, the third and fourth lumbrical muscles, thumb adductor muscle, and thumb short flexor deep head; the superficial branch dominates the sensation of volar ulnar proximal metacarpophalangeal joint and the skin of little and ring fingers (mostly at the ulnar sides).
   (d) Ulnar nerves play a dominant role in hand function. In case of occurrence of forearm defects, the median nerve can be sacrificed to repair the ulnar nerves, and the opposing dysfunction induced by median nerve impairment can be corrected by tendon grafting [32].
2. Median nerve
   (a) Forearm median nerve muscular branches dominate all flexors except the ulnar head of ulnar flexor muscle of the wrist and profound flexor muscle of fingers as well as round pronator muscles and quadrate pronator muscles.
   (b) Hand median nerve muscular branches dominate muscles of thenar as well as the first and second lumbrical muscles. The thenar muscular branches are mostly emitted behind the carpal canal and prone to be injured in case of palmar puncture wounds. The lumbrical muscular branches are fine with rare independent injuries.
   (c) Median nerve cutaneous branches dominate the sensation of three and a half fingers at the radial sides and ulnar sides of the palms. The cutaneous branches of big thenar and the palmar center are emitted from the proximal side of cutaneous branches; the cutaneous branches of the radial skin of thumb, index, middle and ring fingers first form the common digital nerves at the palm and then emit digital inherent nerves to reach corresponding fingers.
3. Radial nerves
   (a) Forearm radial nerve trunk only includes the segment from the external condyle of humerus to the upper margin of supinator muscle without main branches. Ninety-five percent of the brachioradialis muscles and the radial wrist long extensor muscular branches extend from the upper region of external condyle of humerus, so most of the wrist extensor functions can be maintained after the forearm radial nerves are injured.
   (b) Radial nerve forearm muscular branch is also called posterior interosseous nerve which extends from radial nerve trunk and then enters the supinator muscle canals. The upper margin of supinator muscle usually consists of muscular and tendon tissues, and only 1/5 are completely tendinous (called Frohse arch); the lower margin of supinator muscle is mostly tendinous tissues. The potential space near the supinator muscle passed through by posterior interosseous nerves is called supinator muscle canal (Fig. 2.28). The nerves are prone to be compressed on the upper and lower margins of this canal, inducing paralysis at the finger extensor, ulnar extensor muscle of the wrist, and supinator muscle [32].
   (c) The superficial branches of radial nerves are emitted by the radial nerves at the lower margin of the elbow joint, then they descend and reach the dorsal side of the wrist and the radial side of the back of the hand, thumb web, the dorsal side of the proximal segment of thumb, index and middle fingers dominate the skin sensation of corresponding regions.
4. Sensory nerves of forearm skin. The sensory nerves of forearm skin are the cutaneous branches emitted by upper arm nerve trunks. It is worth noting that the skin sensations at the palmar surface (near the radial side) of the forearm are not dominated by median nerve and radial nerves but dominated by one terminal branch of the musculocutaneous nerves (forearm lateral cutaneous nerves); likewise, the skin sensations of the palmar surface (near the ulnar side) of the forearm are dominated by the medial cutaneous nerves of the forearm.
5. Communicating branches of forearm. There are different degrees and types of communicating branches between the median nerve and ulnar nerves of the forearm and hand, and this is important in electrophysiological examination. Clinically, if only one single nerve is injured, the results of electrophysiological examination indicate that

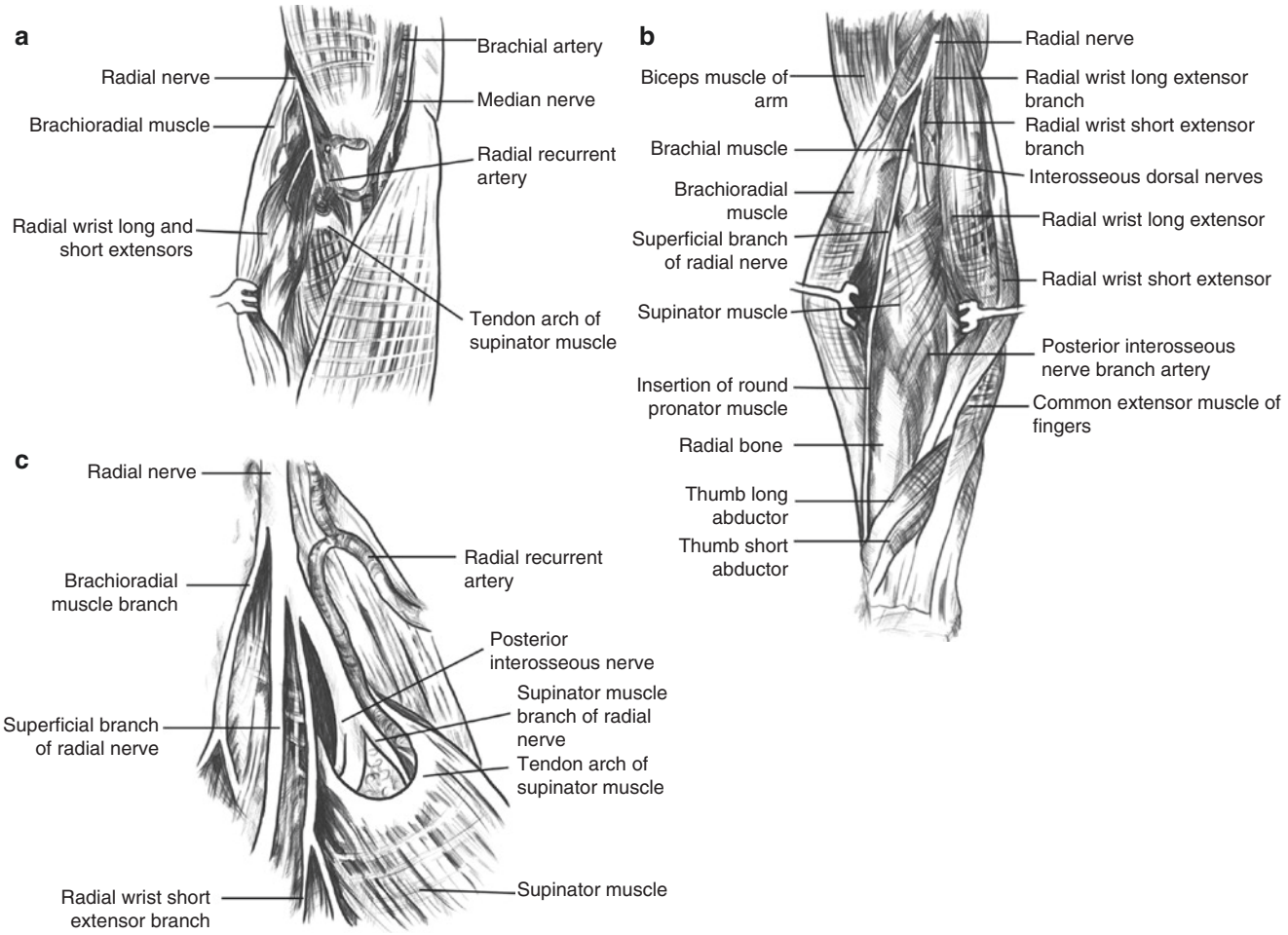


Fig. 2.28 Supinator muscle canals and relevant structures

not only is this nerve injured but also the other nerve has partially functional disorder. At this time, factors such as edema, inflammation, and disuse after trauma of the limbs should be considered, the problem of branch communication between the two nerves should be noted, and this is of significance to preoperative diagnosis and medicolegal identification.

2.2 Hand Examination and Diagnosis

Hong An, Dong Han, and Bo Chen

Like those for other human parts, examination and diagnosis of the hand include inquiry about medical history, physical examination, special examinations, etc. The contents of some examinations and diagnosis have been mentioned in the section of functional anatomy, and the following will focus on the questions closely correlated with hand diseases.

2.2.1 Medical History

Surgeons and doctors from rehabilitation department should have a clear understanding of the entire medical history of the patients, especially the influence of hand trauma or diseases on the patient's physical, mental, and economic conditions so that the entire treatment can proceed smoothly.

2.2.1.1 Deformities

First, a distinction should be made between congenital deformities and acquired deformities. In most cases, it is easy to distinguish between the two, but some cases are very confusing. As for birth injury induced neck fifth and sixth nerve root injuries (injuries of upper brachial plexus nerves, Erb paralysis) or neck eighth and chest first nerve root injuries (injuries of lower brachial plexus nerves, Klumpke paralysis), though they can be found at birth, they fall into the category of acquired injuries of birth injury; as for trigger finger induced by congenital thumb tendon sheath narrowing, although they cannot be easily found at birth and cannot be noted until the children become 1–2 years old, they should

not be considered to be induced by acquired factors. In addition, the duration from the appearance of deformities to the doctor visit time has a direct influence on the difficulty and effect of treatment. The diagnosis of the congenital deformities should be made not only based on surface phenomenon (e.g., joint flexion contracture) but also on the problems in internal structures (e.g., vessels and nerves), to avoid vascular and nerve injuries induced by excessive orthopedic surgery. In addition, changes in nails and skin are always associated with diseases in the kidneys, liver, and respiratory system [14, 15, 34].

2.2.1.2 Diseases

Joint inflammation and benign masses are common hand diseases, and different ages, genders, and disease courses are of some value in differentiation of diseases. For example, repeated and sudden occurrence of arthritis in men's thumb metacarpophalangeal joints may be one of the manifestations of gout; the symmetric inflammation of proximal interphalangeal joint and metacarpophalangeal joint in old women is mostly rheumatoid; the chronic inflammation and Heberden's nodes of distal interphalangeal joint in old people are the characteristics of osteoarthropathy. The common symptom of hand benign mass is tendon sheath cyst, but the tendon sheath cyst at the finger volar side is often small and hard and is often misdiagnosed as fibrous tubercles; the painful nodules near the nails or below the nails may be glomus tumor; the mass that grows along the tendons may be giant cell tumor of tendon sheath or tuberculous synovitis; the common symptom of phalangeal expanded destruction is enchondroma.

2.2.1.3 Trauma

Trauma is the commonest and most complicated issue in hand damages. Its diagnosis is relatively easy, but its treatment and functional reconstruction are difficult. In medical history, it is necessary for the doctors to learn the trauma-inducing reasons. It should be noted that due attention should be paid to high-pressure injury and animal bite, especially human bite and meat chopper injuries. If such small wounds are not properly treated, infections and tissue necrosis that are difficult to control are very prone to occur. In addition, the trauma-inducing environment should be learned, including contamination degree, contaminant nature, early treatment, and the duration from injuries are sustained to when treatment is accepted; in addition, whether there are factors that influence tissue healing or whether chronic diseases such as diabetes, arteriosclerosis, rheumatoid disease, TB, and chronic liver diseases are suffered should be learned. The author once compared the postoperative infection rate in patients with hand trauma and found that the postoperative infection rate of patients with systemic diseases was as high as 25%.

2.2.2 General Examination

2.2.2.1 Abnormalities of Rest Position

Rest position is the position where the strength of hand intrinsic muscles and extrinsic muscles are relatively balanced. Abnormalities of rest position mean a disequilibrium, which usually occurs during rupture of a certain tendon, muscular tissue paralysis, ankylosis, or injuries of peripheral nerves [6, 7].

2.2.2.2 Deformities

1. Anomaly of claw hand. Claw hand caused by ulnar nerve injuries are confined to the ring and little fingers, and its passive activities are normal and accompanied by hand ulnar paresthesia; claw hand induced by forearm ischemic contracture can involve all fingers, including the metacarpophalangeal and even joint wrist, and the passive activities will not improve deformity; as for the claw hand caused by burns, the scar on the palm surface is very obvious and the patient has a clear medical history [35].
2. Anomaly of spade hand. After the median nerve and ulnar nerve are damaged, both the thenar and hypothenar muscles shrink, the metacarpophalangeal joint extends, the interphalangeal joint flexes, the palmar arch disappears, and the palm becomes flat. Such deformities share similar manifestations with muscle atrophy and joint extension position rigidity induced by too long cast fixation of hand extension position, but the causes of the two are different. The spade hand induced by congenital hand dysplasia and the spade hand caused by trauma can be easily distinguished.
3. Gooseneck anomaly. It is the deformity of proximal interphalangeal joint hyperextension and distal interphalangeal joint flexion. Gooseneck anomaly is the typical manifestation of hand intrinsic muscle contracture, which occurs usually during metacarpophalangeal joint's deviation toward the ulnar side and passive stretching of interosseous muscles in case of osteofascial compartment syndrome, cerebral palsy, or rheumatoid arthritis; it may be the increase of digital common extensor muscle induced by a variety of causes or the joint disequilibrium induced by the rupture of superficial flexor tendon and proximal interphalangeal joint volar plate. Thumb gooseneck anomaly is mostly induced by rupture of thumb long extensor tendon and excessive traction of thumb short extensor tendon. Burn of back of hand is also the common cause of gooseneck anomaly [36].
4. Anomaly of boutonniere finger. It is the deformity of proximal interphalangeal joint flexion and distal interphalangeal joint hyperextension. The rupture of central tendon of dorsal aponeurosis or the olisthe of lateral tendon toward the volar side will cause the anomaly of boutonniere finger. Trauma and burns are common etiological

factors: sometime since rheumatoid arthritis involves finger extending structures and makes them reflex, such deformities will arise [36].

5. Anomaly of hammer finger. It is the distal interphalangeal joint flexion deformity induced by the rupture of distal insertion of dorsal aponeurosis or the avulsion fracture at the lateral basilar part of distal phalanx. It is commonly accompanied by hyperextension of proximal interphalangeal joint.
6. Anomaly of thumb adduction. Thumb web cicatrical contracture, median nerve injuries, thumb abductor paralysis, etc. can all induce anomaly of thumb adduction. In the past, muscular adductor ischemic contracture was induced most commonly by injection of drugs into Hegu acupoint; in severe cases, the first dorsal interosseous muscles will be atrophic, and the index finger can deviate toward the radial side. At present, anomaly of thumb adduction induced by injection is rare.
7. Anomaly of ape hand. Low median nerve damages can induce thenar muscular atrophy and disappearance of the opposing function of the thumb, in which case the thumb can only flex resembling the appearance and actions of ape hand may be abnormal.

2.2.2.3 Swelling

In case of acute infection of the palm, swelling is significant, and palmar side is hard with severe tenderness; swelling of back of hand is severer than that of the hand palm and presents pitting edema, but tenderness is slighter. Tendon sheath infection presents band-shaped tenderness, and the thumb and little finger can, respectively, lead to the radial synovial bursa and ulnar synovial bursa and can extend to the wrist. When compared with infectious welling, the majority of hand ganglionic cysts have a relatively hard texture and are small and localized; especially at the dorsal wrist to wrist, they may sometimes be misdiagnosed as osseous lump. Wrist volar ganglionic cysts or tuberculous synovitis can present calabash-shaped swelling after passing the carpal canal, palm center or forearm, and when the latter is compressed, the gliding of irregular particles can be felt inside the mass. The thenar angiocavernoma bounces when compressed, and light blue or dull red traces are visible on the skin. Rheumatoid nodules are mostly found in the subcutaneous regions of digital dorsum which are hard and flexible. The hand implantation cysts are mostly adherent to the skin with a moderate texture, and pigmentation is often present in the original injuries.

2.2.2.4 Joint Activities

The range of joint activities of fingers and wrists are correlated with the working nature of different individuals; those engaged in fine work have a relatively large range of activities; those engaged in heavy labor have a relatively small range of activities; therefore, the normal range of activities of hand joints recorded by different books varies. For each patient, when compared with the same joint at the healthy side, the optimal criteria of evaluating the disorder degree of joint activities can be based on the normal mean value in case of injuries of both hands (Table 2.3) [1].

The range of activities determination method proposed by Eaton in 1975 is the main method used to judge the joint active movement after tendon repair and is also a good method used to judge joint functions. The previous method of measuring the distance from the fingertip to the center of palm is simple. However, in case of the presence of the anomaly of finger flexion and contracture, the fingertip is very near the center of palm, but the functions of the two joints are very poor. The main method completely rules out this possibility [37, 38]. The content of the main method includes:

1. TAF (total active flexion). It means that the range of active flexion is equal to the total sum of active flexion degrees of metacarpophalangeal joint and proximal and distal interphalangeal joints.
2. TEL (total extension lack). It means that the degree of loss of active extension is equal to the total sum of extension deficit degrees of metacarpophalangeal joint and proximal and distal interphalangeal joints.
3. TAM (total active movement). It means the range of active movement, which is TAF-TEL.
4. TAM %. It means the range of active movement and is calculated using the following formula: TAM of the affected side ÷ TAM of the healthy side ×100%. Results: 95~100% is superior, 75~94% is good, 50~74% is fair, and less than 50% is inferior.

Table 2.3 Mean value of range of activities of all hand joints

Joints	Flexion	Extension	Adduction	Abduction
Wrist joint	70°~80°	60°~70°	20°~40°	10°~20°
Proximal interphalangeal joint	80°~90°	0°	0°	0°
Distal interphalangeal joint	70°~80°	0°	0°	0°
Metacarpophalangeal joint	80°~90°	0°~20°	30°	30°

Note: ① The middle finger metacarpophalangeal joint has no adduction or abduction function; ② the judgment of wrist adduction and abduction should be made when the upper limbs are at the anatomical position; ③ the judgment of metacarpophalangeal joint adduction and abduction should be made when the joint is approaching or leaving the middle finger

2.2.3 Examination of Vascular Nerve Functions

2.2.3.1 Allen's Test

This is a method used to examine whether the wrist ulnar and radial arteries as well as the palmar arch formed by them suffer obstruction or congenital defects. First, the patient is asked to clench the fist to expel hand blood, and then the examiner uses the two thumbs to compress the ulnar and radial arteries of the wrist. At this time, the patient is asked to extend the fingers and it is evident that the hand becomes pale. If the examiner relaxes the ulnar artery or radial artery, the palm becomes red again completely, which means that the ulnar artery or radial artery trunk and the palmar arch are smooth. If the examiner relaxes the compression on the radial artery and the palm does not become rosy, it means that the wrist radial artery is blocked; if only radial side becomes rosy, it means that the palmar arch is blocked or defective. The method of only relaxing the compression on the ulnar artery is to examine whether the ulnar artery and palmar arch are smooth (Fig. 2.29). Allen's test can also be used to detect single fingers through the abovementioned method, but it is only necessary to compress and relax digital inherent arteries.

2.2.3.2 Finkelstein's Test

This test is also called test of ulnar deviation in clenching fist. During the examination, the patient is asked to clench the fist after thumb flexion and perform ulnar deviation of the wrist joint; if pain is felt at radial styloid process, the result is positive, indicating that the thumb short extensor tendon and thumb long abductor tendon develop tendovaginitis at the radial styloid process (Fig. 2.30). In order to determine whether thumb long abductor tendon and thumb short extensor tendon develop tendovaginitis at the same time, some scholars have modified this method. Specifically, the wrist joint is first maintained at the ulnar position; the passive flexion of the first carpometacarpal joint makes the thumb long abductor tendon nervous. If sharp pain is felt at the radial styloid process, it means that the thumb long extensor tendon develops tendovaginitis, and then the first carpometacarpal joint is maintained at the flexion position and the thumb metacarpophalangeal joint is passively flexed; if sharp pain is felt at the radial styloid process, it means that the thumb short extensor tendon develops tendovaginitis.

Fig. 2.30 Finkelstein's test

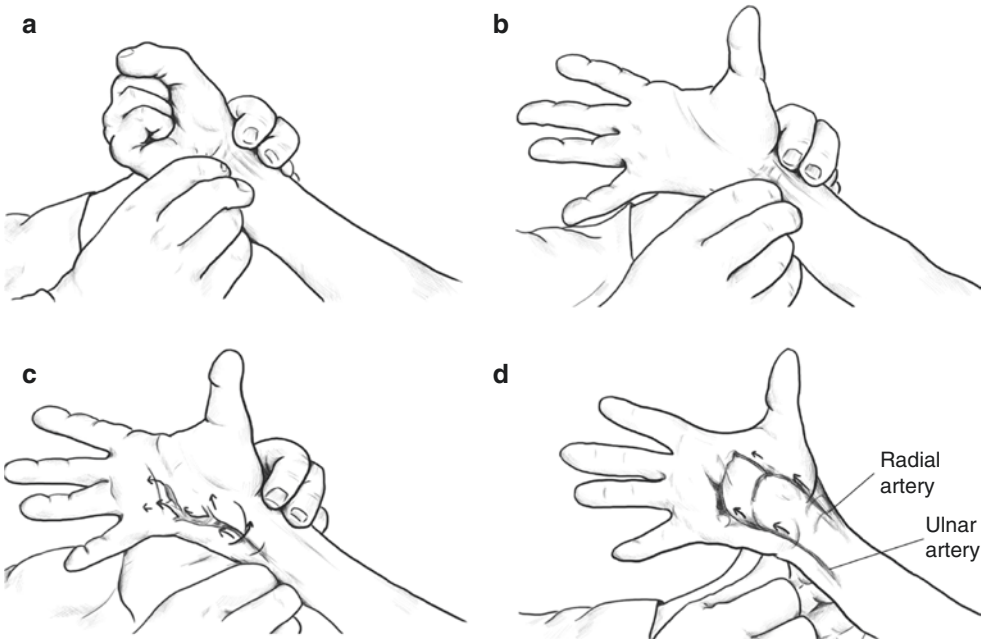


Fig. 2.29 Allen's test. Make a fist (**a**). Open fist (**b**). Release side artery (**c**). Release bilateral artery (**d**)

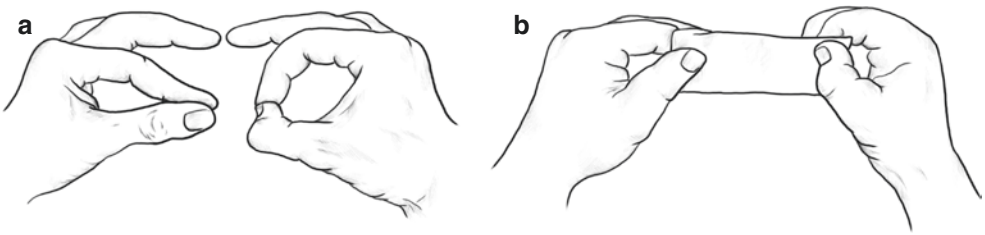


Fig. 2.31 Froment's test. (**a**) Pinching using the lateral side of thumb and index finger, positive right hand, normal left hand. (**b**) Pinching paper using thumb and index finger, positive right hand, and normal left hand

2.2.3.3 Froment's Test

Ulnar injuries induce thumb myoparalysis, in which situation the patient uses thumb flexor contraction to compensate for partial adduction function; the partial paralysis of thumb short flexor and thumb short abductor makes the thumb metacarpophalangeal joint unstable and thus be under hyperextension status, and when the patient is asked to use the lateral side of thumb and index finger or use the finger pulp for pinching one small object, the phenomena of interphalangeal joint flexion and metacarpophalangeal joint hyperextension of the affected side of thumb often occur (Fig. 2.31).

2.2.3.4 Wartenberg's Sign

When fingers are straightened, the little finger is in the abduction position and cannot adduct, which means the Wartenberg's sign is positive. The mechanism of generation is as follows: after ulnar nerves are injured, the hand intrinsic muscles become paralytic, the little finger loses active adduction and abduction functions, and the insertion of the inherent extensor tendon of little finger deviates toward the ulnar side of the little finger. Therefore, in case of extension of the little finger, anomaly of passive abduction will occur. This positive sign indicates that lesions occur to the motor branches of ulnar nerves, but the specific position of ulnar nerve injuries cannot be located.

2.2.3.5 Phalen's Test

This is a reliable method used to examine carpal tunnel syndrome. The patient is asked to symmetrically flex both wrists. In case of carpal tunnel syndrome at one side, the volume of carpal canal becomes small, further compressing median nerve. Within 2 min, anesthesia or pricking-like paresthesia (Fig. 2.32) immediately occurs to the hand median nerve innervation area, especially at the middle finger site. In case of no occurrence of paresthesia within one minute at this position, the median nerve is considered to be normal.

2.2.3.6 Tinel's Sign

When the nerve injure point is percussed, the occurrence of radiating pain toward this nerve innervation area means that the Tinel's sign is positive. Its significance lies in that: (1) Judge the site reached by neuranagenesis and the regeneration degree, (2) understand nerve tumor. It is especially important to examine Tinel's sign within 4 months after injuries. If the Tinel's sign at the injury level is persistently positive and it is negative beyond the injury level, it means that the injured nerves are not regenerated toward the distal end. If the Tinel's sign beyond the injury level is weakly positive, it means that the regeneration quality of nerves is poor. Only if the Tinel's sign is persistently positive and continuously advances to the distal end can prove that the neuranagenesis be satisfying.

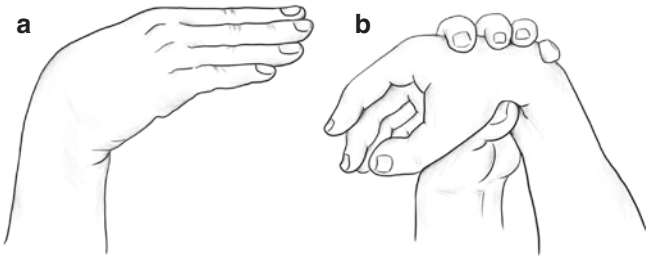


Fig. 2.32 Phalen's test

2.2.3.7 Palmar Short Muscle Radiation Test

The forceful compression of the radial side of the wrist lentiform bone and the stimulation of ulnar nerves to cause palm short muscle contraction can induce the appearance of scattered pitting on the hypothenar skin. In case the ulnar nerves are injured at the proximal side of the wrist, no such phenomenon occurs.

2.2.3.8 Two-Point Discrimination Test

It is a method used to learn skin epicritic sensibility and can be classified into static two-point discrimination and dynamic two-point discrimination. It can reflect the distribution density of sensation corpuscles and is of important reference value to the evaluation of nerve recovery degree after they are injured. The finger is one of the most sensitive sites of human skin, and its ability to discriminate the distance between the two stimulation points is the strongest. Its minimal discrimination distance is only 2 mm. Disk-Criminator is used in examination, and it can be substituted by two-footed compasses (dull ends). The two ends gently touch the skin, and the connecting line between the two stimulation lines should ideally be parallel to the finger longitudinal axis (to avoid the influence caused by nerve crossing). As two-point discrimination varies greatly due to different individuals and different occupations, the patient's healthy and affected fingers should be examined, and a comparison should be made between them. After nerves are injured, the two-point discrimination ability will improve as nerves are recovered.

2.2.3.9 Ninhydrin Test

This test aims to examine whether the hand skin is sweating for the purpose of reflecting autonomic nerve function. First, the examined hand is radiated under the intense incandescent lamp for several minutes, and then the finger pulp is pressed on the test paper sprayed with ninhydrin liquid. In case of occurrence of clear fingerprint, it means that the finger sweat glands are normal, that is, the autonomic nerve functions are normal. Also, the examiner can be sprayed with iodine tincture. A layer of flour is spread on the surface after it becomes dry, and then the finger is placed under the intense light for radiation and heating. In case of sweating, the flour will turn blue; otherwise, it will remain unchanged. This method is simple and feasible, and it is most practical for grass-root units that lack corresponding devices.

2.2.4 Special Examinations

2.2.4.1 X-ray, CT, and MRI Examination

1. X-ray examination. X-ray plain film is a basic method used to examine the wrist, each bone of hands, and joint morphology, structure and location of them, which is described in detail in relevant books. What should be noted here is that attention should be paid to the projection position of X-ray. The fingers can be fully displayed through anteroposterior and lateral film; metacarpal bones are frequently overlapped on the lateral film, and can only be displayed through volar and dorsal lateral oblique radiography. The scaphoid axial view film is a good method used to get a full view. The wrist dorsiextension position 45° projection can display hook of hamate bone, and this projection place is the only place for diagnosis of fracture of hook of hamate bone.
2. CT examination. CT can also be used to check the wrist, plays an important role in understanding hidden wrist fracture, displacement of bone fragments, fracture healing, and bone nonunion, and is especially helpful to the evaluation of dislocation and semi-dislocation of distal radioulnar joints.
3. MRI examination. MRI is the gold standard for the observation and confirmation of soft tissue injuries. It is especially helpful to the diagnosis of carpal ligament, triangular fibrocartilage disk, tendon injury, and tenosynovitis. However, MRI examination cannot be performed on patients using heart pacemaker and artificial cochlea [39–44].

2.2.4.2 Arthroscopy

A 1.5–3 mm arthroscope can be used to examine the wrist joint. Arthroscopy can be used to observe the articular cartilage and learn whether any triangular cartilage is ruptured, can be used to identify the cause of carpal joint instability and to know whether any intercarpal interosseous ligament is injured, and can be used to perform observation and biopsy of synovial bursa. Additionally, arthroscopy can be used to perform open decompression of cubital tunnel and carpal tunnel as well as the lysis of adhesion of tendovaginitis, and it can be combined with drug injection to prevent adhesion; it can also be used for the treatments under direct vision such as reduction of fracture fragments within the joints at the lower end of descending radial bone and fixation by per cutem. However, since the carpal joint cavity is relatively small, arthroscope operation is difficult.

2.2.4.3 Ultrasound Examination

Ultrasound is a noninvasive blood test method. With the improvement of device fabrication, the patency degree and the blood flow speed of digital blood vessels can be examined, and arteries and veins can be differentiated [45].

B-mode ultrasound with a high-frequency probe can identify the physical properties and the adjacent relation of the hand mass and contribute to the differential diagnosis and preparation of preoperative surgical plan; it can also display the morphology and travel arrangement of vessels and perform diagnostic puncture on mass under the ultrasound monitoring. Since the method has no damages to the patient and is easy to repeat, it is practical and popular. The 3-D color ultrasound technology that has emerged during recent years can also be used for the stereotactic positioning of the caliber and travel arrangement of vessels, and is helpful to the design of each kind of flap [46].

2.2.4.4 Emission Computed Tomography (ECT)

Emission computed tomography (ECT) can early detect the presence of lesions in the hand bones and can diagnose the disease 3 months or so earlier than X-ray examination. In addition, ECT can facilitate the diagnosis of hand hemangioma and lymphangioma. The utilization of nuclide for angiography is relatively effective for the ulnar and radial artery, and the display of digital small vessels remains unclear.

2.2.4.5 Electrophysiologic Examination

1. EMG. It can identify the types of muscular contraction functional disturbances: neurogenic, myogenic, neuromuscular junction, or psychogenic, and it can also serve as an important method used to assess the efficacy after nervous lesion treatment. The judgment of EMG results and the examiner's anatomy knowledge are closely correlated, and whether patients can be fully cooperative during the examination is also an important factor. If necessary, clinicians should also participate in the examination and describe in detail the clinical physical examinations to the examiner so that they can cooperate with each other and reach the accurate conclusion.
2. Nerve conduction velocity test. It includes examination of motor nerve conduction velocity (MNCV) and sensory nerve conduction velocity (SNCV). The MNCV is calculated as follows: (proximal latency period − distal latency period)/the distance between the two stimulation points; the sensory nerve conduction velocity is calculated as follows: the latency period from the stimulation point to recording point/the distance from the stimulation point to the recording

point. For the same nerve, the conduction velocity of the proximal end is faster than that of the distal end, and the conduction velocity of the adults is faster than that of children and the elderly. After nerve injury, the conduction velocity becomes slow, which is a specific pathological manifestation, and is not subject to the influence of subjective factors. In case of nerve adhesions or compression, MNCV can sometimes be within the normal range, but the sensory nerve conduction velocity has become slow, the latency period after stimulation is prolonged, the wave amplitude becomes low, and the diagnostic significance is greater.

3. Somatosensory evoked potential test. Somatosensory evoked potentials (SEP) are bioelectric reactions that can be detected at any site on the specific pathway in this system after one point in the somatic sensation system is stimulated. After peripheral nerves are injured, SEP will show some characteristic manifestations: ① After nerve rupture, the sensory nerve action potentials cannot be measured. As for incomplete injury, even if a small number of neurites stays in touch with the nerve center, grade one somatosensory cortex primary electric potential can be recorded. ② When most of the injured nerves are recovered, there may be basically normal nerve conduction velocity and EMG, but at this time, making the SEP that crosses the original injury point can also help find the disappearance of sensory nerve action potential. ③ During the recovery period of nerve, the sensory nerve action potentials that cross the injury points cannot be detected, but the corresponding grade one somatosensory cortex primary potentials can be recorded. This is evidence that the nerve fiber has been regenerated, which could be used to judge nerve regeneration and used as a method to learn the nerve regeneration speed. ④ When the nerves suffer from compression damage, in case the corresponding nerve innervation area is stimulated beyond the compression point, it is evident that the wave amplitude of grade one somatosensory cortex primary potential decreases, the latency period is prolonged, and the time course is widened; when the proximal side of the compression side is stimulated, SEP is within the normal range.

2.3 Assessment of Hand Functions

De Shi, Dong Han, Wei Wang, Bo Chen, Jianmin Yao, and Fengjing Zhao

In recent years, rapid development has been seen in China's hand surgery. Various surgeries such as replantation of amputated finger, thumb reconstruction, as well as reconstruction of tendons, muscles, nerves, bones and joints have been extensively applied and have reached a high level, but comments on the efficacy of various surgeries vary. For a long time, there has been a lack of a unified standard for hand function assessment in China, and the hand function assessment methods of the United States Swanson and the American Medical Association (AMA) is not yet fully applicable to China. In 1989, the Society of Hand Surgery of Chinese Medical Association held hand function evaluation standard workshops in Guangzhou; in addition, the department where the author worked was designated to carry out the measurement of normal hands of Chinese people in 1990. According to the hand shape and the range of activities of the finger and hand wrist joints of Chinese people, we proposed a normal reference value for the assessment of hand functions of Chinese people. The Society of Hand Surgery of Chinese Medical Association proposed the standard for assessment of upper limb functions in 2000 in Wuxi City, and in September of that year, it published the standard in the "Chinese Journal of Hand Surgery" Vol. 16, No. 3, suggesting its trial application nationwide. However, no matter what assessment methods are used, it is required that the data obtained should be accurate and reliable; the device should be easy to use with a high repeatability so that the detection purpose can be served [47–50].

Each assessment method has its own limitations; for example, it is difficult for the factor of age to be separated independently from complex factors, but it does affect the final result; patient's compliance and cognitive ability can also affect the accuracy of results. Pain is a difficult-to-accurately-assess factor, especially for those with related diseases (such as nerve diseases), and the assessment method cannot address the problem.

Now, our measurement results and the hand function assessment method of United States Swanson, AMA, and DASH (disability of arm, shoulder, and hand) evaluation methods are described as follows.

2.3.1 Examination Method

Assessment of upper limb function should include hand anatomy, appearance, function, etc., in order to accurately reflect the functions of patient hands. Assessment of anatomical lesions is based on the medical history and careful examination of the patient; assessment of appearance is about the reactions of patients and the society to injuries; assessment of function contains many aspects and is the most important, and it reflects the functions of upper limbs and their performance in daily life [3].

For every patient, it is important to keep a full and complete preoperative and postoperative follow-up examination logs. The former is the basis for selection of treatment program, while the latter is the basis for assessment of the efficacy and also the indispensable valuable materials for medical workers to summarize experience and lessons. To make functional assessment standard uniform, during preparation of standards, it is necessary to have a unified format for the examination records concerning diseases and trauma so that they can gradually be standardized and normalized. Tables 2.4 and 2.5 briefly summarize patient's general conditions, diagnosis, medical history, laboratory tests and treatment, list

Table 2.4 Records of preoperative or postoperative examinations of hand diseases

Name: Gender: □ Male □ Female Date: Date of Birth
Address: Occupation: Dominant hand: □ Left □ Right Hospital: Examiner:
Diagnosis:
Treatment plan or surgical method: Surgery date:
Date of onset:
Site of onset:
Check whether the followings are complete: □ X-ray □ Radiography □ Cinematograph □ Radiological inematograph image
[Range of motion (ROM) the neutral position 0 ° is used]
(Codes 1 to 25 represents the Deformitisobserved and measured)
(Severity is represented by a, b and c, meaning mild, moderate and severe, and then codes 1 to 25 are used to represent the c ategories)

Thumb	For thumb, the following codes are used: 7, 2, 3, 9, 14, 19 and 22		Abduction (the angle between the first and second metacarpal bone) adduction (the distance from the digital last segment to the cross striation of the fifth metacarpophalangeal joint) opposition (the distance from the digital last segment cross striation of the third metacarpophalangeal joint)			
	Code			ROM		
	Right	Left	Joint	Right	Left	
			Abduction			
			Adduction			
			Opposition			
			MP			
			IP			

Codes for clinical abnormalities: 13. Synovial hypertrophy
1. Thumb swan-neck deformity 14. Snapping in motion
2. Thumb anomaly of boutonniere finger. 15. Extensor tendon subluxation
3. Suto the bluxation and luxation 16. Introversion angel
4. Digital swan-neck deformity 17. Ectropion angel
5. Digital anomaly of boutonniere finger 18. Rotating deformity
6. Hand internal muscular tension 19. Erosion
7. Ulnar deviation 20. Joint space narrowing
8. Radial deviation 21. Subchondral sclerosis
9. Arthrokleisis 22. Pain in joint activities
10. Instability 23. Nerve compression
11. Tendon rupture 24. Vasculitis
12. Constrictive tenosynovitis 25. Nodules

Codes for hand use: 3 ~ 15, 19, 22 ~ 25 ROM					
Index finger			MP		
			PIP		
			DIP		
	Distance from DIP flexion crease to palm transverse striation				
Middle finger			MP		
			PIP		
			DIP		
	Distance from DIP flexion crease to palm transverse striation				
Ring finger			MP		
			PIP		
			DIP		
	Distance from DIP flexion crease to palm transverse striation				
Little finger			MP		
			PIP		
			DIP		
	Distance from DIP flexion crease to palm transverse striation				

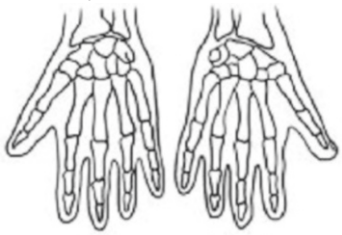
Grasp mode: examine the presence of abilities

Grip		Right	Left
Cylinder	2.5cm		
	5cm		
	7.5cm		
	10cm		
Spherical body	5cm		
	7.5cm		
	10cm		
	12.5cm		
Strength: □ lb (pound) □ kg □ kPa		Right	Left
Pinching with finger pulp	Index finger		
	Middle finger		
	Ring finger		
	Little finger		
Lateral pinch or key pinch			
Clip			

Wrist	The following codes are used: 3, 7~14, 19, 20, 22 and 23				
			Flexion		
			Extension		
			Ulnar deviation		
			Radial deviation		

Musclar strength determination:
Radial nerve
Brachioradial muscle _ Radial wrist long and short extensors_ Supinator muscle_
Digital common exensor -Ulnar wrist extensor_ Thumb long abductor _
Thumb short extensor_ Thumb long extensor_ Index finger inherent extensor_
Little finger inherent extensor_
Median nerve
Round pronator muscle_Quadrate pronator muscle_ Radial wrist flexor_
Palm long muscle _ Superficial digital flexor _Thumb long flexor _
Profound digital flexor_ Thumb short abductor_ Thumb opposing muscle_

Ulnar nerves
Thumb short flexor _ 3rd and 4th lumbrical muscle_
Ulnar wrist flexor_Finger profound flexor _ Thumb adductor_
Litter finger abductor_ Small little finger short muscle _ Little finger opposing muscle_
Thumb short flexor deep head _ Lumbrical muscle_ Volar interosseous muscle_
Dorsal interosseous muscle_
Sensory impairment or amputated finger plane



Palmar surface right Palmar surface left

Note: *MP* means metacarpophalangeal joint, *IP* means interphalangeal joint, *PIP* means proximal interphalangeal joint, *DIP* means distal interphalangeal joint

Table 2.5 Records of preoperative or postoperative hand function of hand injuries

Name: Age: Date: Dominant hand:
Occupation: X-ray film: Photo: Medical history:

Shoulder joint: left right	Wrist joint: left right	Perimeter: left right
Forward	Dorsiflexion	Biceps muscle of arm
Backward	Palmar flexion	Forearm
Abduction	Radial deviation	Forearm: Pronation
Adduction	Ulnar deviation	Supination
Intorsion	Elbow joint: Flexion	Grip strength: Left
Extorsion	Extension	Right

		MP	IP		Functional lesions (%)	
Thumb	Flexion			Abduction (the angle between the first and second metacarpal bone)		
	Extension			Adduction (the distance from the digital last segment to the fifth metacarpophalangeal joint)		
	Arthrocleisis			Opposition (the distance from the digital last segment to the third metacarpophalangeal joint)		

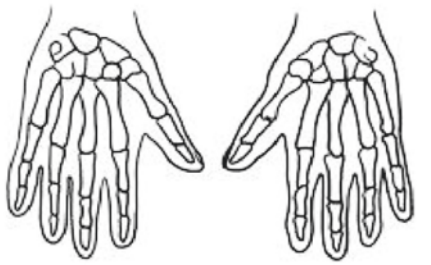
		MP	PIP	DIP	From finger pulp flexion crease to middle transverse striation	
Index finger	Flexion					
	Extension					
	Arthrocleisis					
Middle finger	Flexion					
	Extension					
	Arthrocleisis					
Ring finger	Flexion					
	Extension					
	Arthrocleisis					
Little finger	Flexion					
	Extension					
	Arthrocleisis					

Codes for clinical abnormalities:
1. Amputated extremity
2. Scar
3. Skin - subcutaneous tissue defects
4. Nail bed damage
5. Main nerve defects: R.M.U.
6. Nerve bundle defect
7. Neuroma
8. Pain and tendon
9. Bone injury
10. Joint injury
11. Flexor tendon defect
12. Extensor tendon defect
13. Ligament injury
14. Sensation - Object picking test

Note: the movement degree is recorded according to left and right

Total (%)

Two point discrimination
Ninhydrin test (sweat test)
15. Grasp
Grip
Pinch: Finger pulp
Finger tip
Lateral side of fingers
Hook: Distal end
Proximal end
Joggle
16. Maximal improvement
17. Need in rehabilitation
18. Further treatment
19. Classification



Dorsal surface of right hand Palmar surface of left hand

Dorsal surface of left hand or Palmar surface of right hand

a variety of tests and measurement items, and draw the rough sketch of hand volar and dorsal injuries. Entry in the table records the range of motion and strength of each joint, fist shape, motor ability of daily living and activity status, and common clinical abnormalities, and list the name and mark as the codes for remarks and indexes. Wherein, the records on left-handedness and right-handedness are of great significance to functional assessment and rehabilitation.

A set of standard photos should be taken, including each hand view of flexion, extension, gripping, and pinching of fingers. Continuous pictures or various function tests help assess patient's adaptability for daily living functions.

Standard X-ray examination is a part of records, involving the dorsaventral position, lateral position, and oblique position of the hand and wrist joint, and these photos must be taken within 3 months. In order to display the degree of anomaly, anatomical macrophotograph is usually preferred but is not compulsory. X-ray cinematography helps to display the range of motion of the finger and wrist joints [40].

2.3.1.1 Anatomical Examination

Anatomical examination should involve examination of the entire body and all its structures, including the skin, nail bed, nerves, blood vessels, muscles, tendons, bones and joints, as well as measurement of the circumferentia of two sides of limbs. Finger examinations include examination of thumbs, index fingers, middle fingers, ring fingers, and little fingers, to observe the condition of every joint and to see whether there is any synovitis, bone and joint instability, subluxation, stiffness, contractures, or lateral deviation deformity, and their degree if any.

2.3.1.2 Measurement of Range of Joint Motion

During the measurement of range of joint motion, the examiner should stick to the principle that the neutral position of joint activities is 0°, which means that, 0° is set as the starting position for measurement of all joint activities. Measuring the angle of the joint activity helps to determine the motion range of joint. The active movement is achieved by the muscle strength of all flexors or extensors, and measurement of passive movement needs to overcome the resistance of normal soft tissues to movement (which is about 0.5 kg on the finger joints).

Extension refers to the movement of the fingers inversely moving to the flexion toward the 0° starting position, and it can be seen in the extending movement of fingers, elbows, and knee joints. If the extension exceeds the 0° starting position, it is called hyperextension and plus is used to represent the angle of hyperextension; if a flexion position cannot completely extend to the 0° starting position, it is called deficiency of extending movement, and minus is used to represent the angle. For example, if one finger has a 15°~45°flexion contracture, the motion is marked −15°~45°. If the finger joint has a 15° hyperextension to 45° flexion, the motion is marked +15°~45°.

The measured movement of all fingers should be recorded in the form of tables, and angles are used to represent the range of motion. In case of measurement of distal joint, the proximal joint should be placed in the neutral position or extension position. The finger distance and the corresponding strength should also be measured.

The measurement of thumb movement also includes radial abduction, adduction, flexion, extension, opposition, antelocation, and retrocession.

The measurement of wrist joint movement includes the range of dorsal extension, palmar extension, radial deviation and ulnar deviation, and some assessment methods can also include the range of pronation and supination.

The measurement of elbow and shoulder joint movement includes the range of flexion, extension, pronation, supination, adduction, abduction, anteflexion, backward extension, and rotation (Figs. 2.33, 2.34, 2.35, 2.36, 2.37, and 2.38).

2.3.1.3 Examination of Grasp Strength

Grasp strength can be examined through force measuring instrument, and the method of comparing the examiner and the examinee can be adopted for the examination of forearm and hand strength. However, a hemomanometer may be used for a hand with weak strength: first, roll the cuff of the hemomanometer into a cylinder with a diameter of 5 cm, and then aerify it until atmospheric pressure of 6.67 kPa (50 mmHg) is obtained; when the cuff is gripped, the value which exceeds 6.67 kPa is recorded as the grasp strength. Age, nutritional status, pain, fatigue, different time points in the day, the

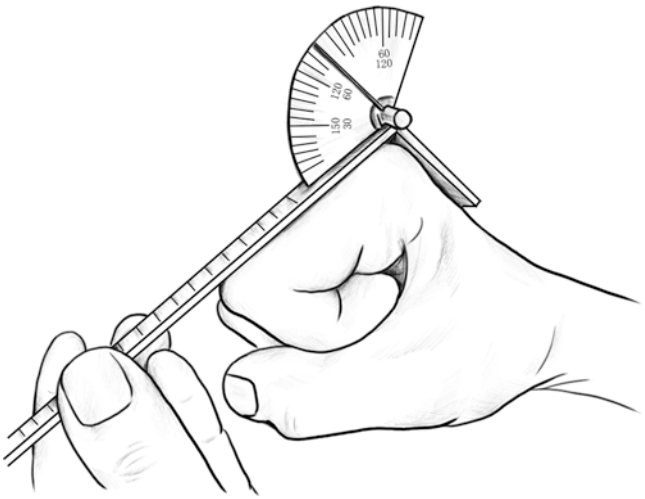


Fig. 2.33 Using joint angle measuring scale to measure the range of flexion of metacarpophalangeal joint when the fist is clenched

Fig. 2.34 Using joint angle measuring scale to measure the range of flexion of proximal interphalangeal joint when the fist is clenched

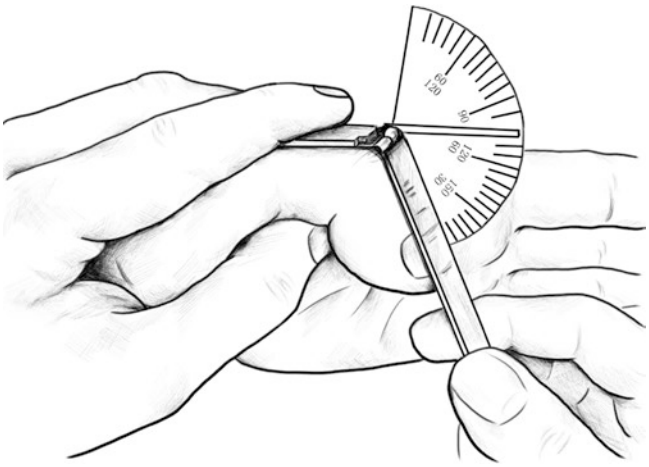


Fig. 2.35 Using joint angle measuring scale to measure the range of movement of thumb interphalangeal joint

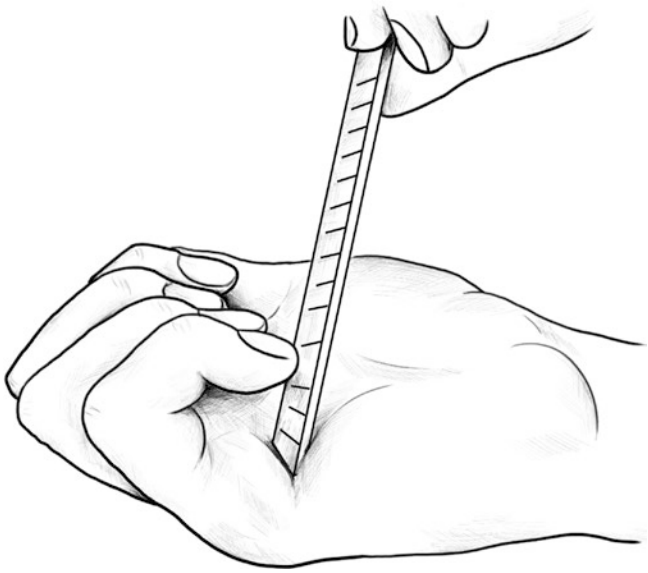


Fig. 2.36 Using measuring scale to measure the distance between finger pulp and the distal palmar transverse striation during the active and passive activities

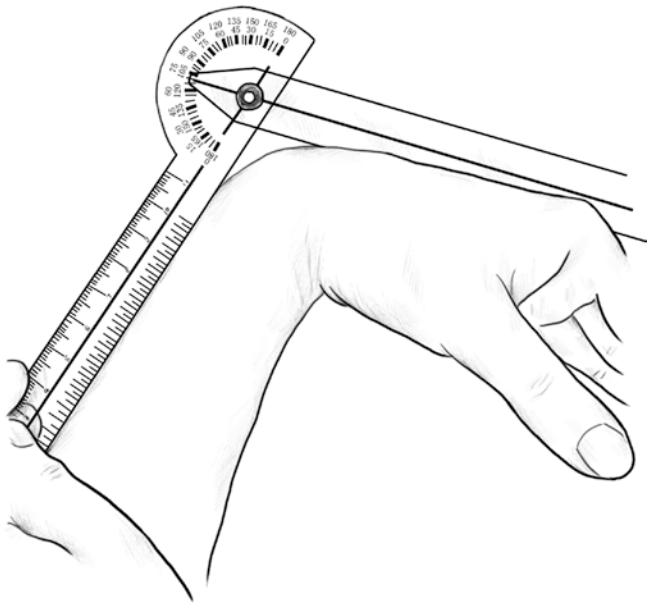


Fig. 2.37 Using joint angle measuring scale to measure the range of flexion of the wrist joint

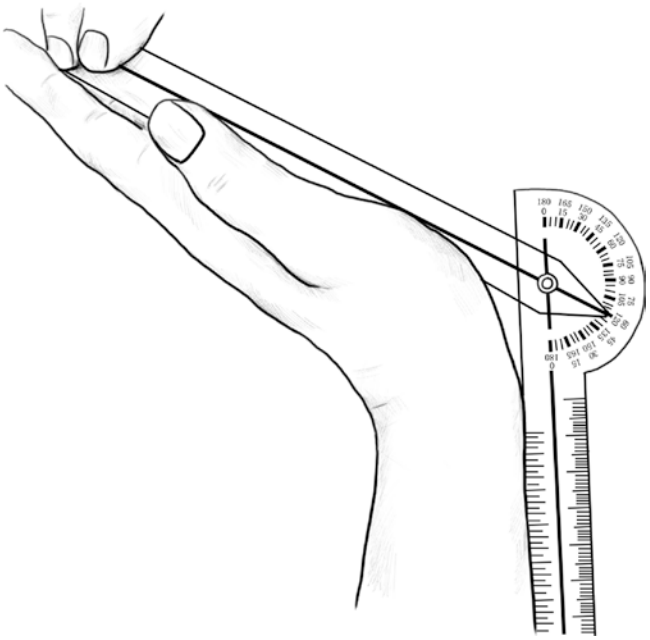


Fig. 2.38 Using joint angle measuring scale to measure the range of dorsal extension of the wrist joint

patient's cooperation, etc., are likely to be factors that affect grasp strength [51].

The detection methods in Figs. 2.39, 2.40, 2.41, and 2.42 are very important to the assessment of the strength of hand, and these examinations are closely correlated with various actions in daily life.

2.3.1.4 Muscle Strength Examination

The rehabilitation of patient's limbs always needs 1–2 years, and the results of muscle strength examination can reflect the final status.

If the patient's limbs suffer restricted movement, pain and partial amputation, muscle strength testing cannot be performed, because the result obtained from test under such condition is often inaccurate. Doctors carry out muscle strength test for the purpose of figuring out whether there is any decline in muscle strength or to find reasons for the decline if so [33].

In 1912, Lovett, according to how much the muscles can offset resistance during contraction, divided muscle strength into six grades, expressed as a percentage: ① 100%, grade 5, normal strength; ② 75%, grade 4; the muscles can offset some of the resistance and drive joint movement; ③ 50%,

Fig. 2.39 The ergometer is used to measure the grip strength of the hands, and the results are accurate and reliable

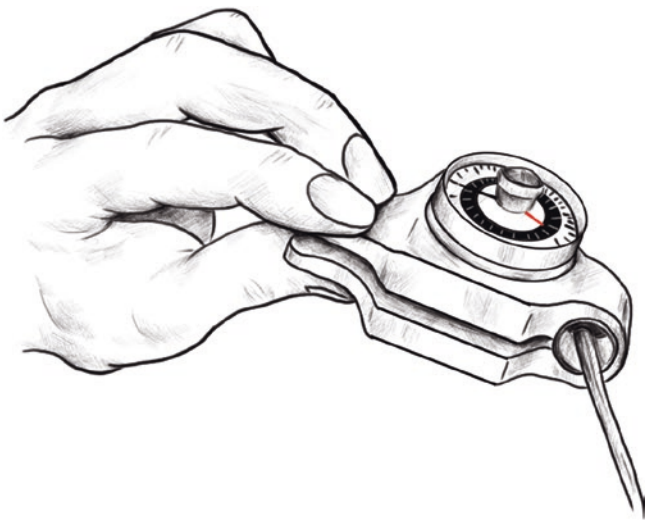


Fig. 2.40 Pinch strength of three fingers

grade 3; muscle can offset gravity, but not resistance; ④ 25%, grade 2, when gravity is excluded, muscles can drive joint movement; ⑤ 10%, grade 1; muscles can only contract very slightly, without causing joint movement; ⑥ 0, grade 0; the muscles do no contract at all.

Fig. 2.41 Measurement of lateral pinch strength of thumb and index finger

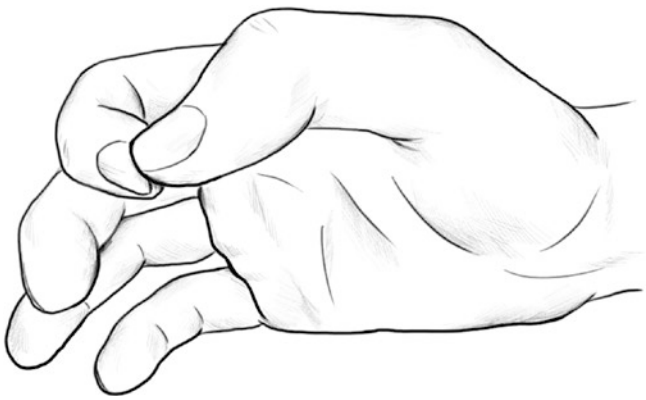


Fig. 2.42 Measurement of pinch strength of fingertips

Table 2.6 Relation between grip strength and occupation (Swanson)

	Grip strength (kg)			
	Male		Female	
Occupation	Forehand	Backhand	Forehand	Backhand
Technical job	47.0	45.4	26.8	24.4
Sedentary job	47.2	44.1	23.1	21.1
Manual job	48.5	44.6	24.2	22.0
Average	47.6	45.0	24.6	22.4

The motor functions are classified into six grades, i.e., M5, M4, M3, M2, M1, and M0, and they are the commonest methods used to assess the degree of recovery of motor function.

The mean values of hand grip strength and pinch strength measured by foreign scholars are of reference value to the assessment of hand functions (Tables 2.6, 2.7, 2.8, 2.9 and 2.10) [52].

2.3.1.5 Sensory Examination

Hand sensation is an extremely important hand function; without hand sensation, people may feel eyeless. Comprehensive sensations of hands can help identify objects and spread emotion, and blind persons can also use hands for reading, developing literacy and so on.

Table 2.7 Relation between grip strength and age (Swanson)

	Grip strength (kg)			
	Male		Female	
Age	Forehand	Backhand	Forehand	Backhand
20 Years	45.2	42.6	23.8	22.8
20–30 Years	48.5	46.2	24.6	22.7
30–40 Years	49.2	44.5	30.8	28.0
40–50 Years	49.0	47.3	23.4	21.5
50–60 Years	45.9	43.5	22.3	18.2

Table 2.8 Relation between grip strength of three fingers and occupation (Swanson)

	Grip strength of three fingers (kg)			
	Male		Female	
Occupation	Forehand	Backhand	Forehand	Backhand
Technical job	7.3	7.2	5.4	4.6
Sedentary job	8.4	7.3	4.2	4.0
Manual job	8.5	7.6	6.1	5.6
Average	7.9	7.5	5.2	4.9

Table 2.9 Pinch strength of finger pulp of different fingers (Swanson)

	Pinch strength of finger pulp (kg)			
	Male		Female	
Fingers	Forehand	Backhand	Forehand	Backhand
Index finger	5.3	4.8	3.6	3.3
Middle finger	5.6	5.7	3.8	3.4
Ring finger	3.8	3.6	2.5	2.4
Little finger	2.3	2.2	1.7	1.6

Table 2.10 Relation between digital lateral pinch strength and occupation (Swanson)

	Digital lateral pinch strength (kg)			
	Male		Female	
Occupation	Forehand	Backhand	Forehand	Backhand
Technical job	6.6	6.4	4.4	4.3
Sedentary job	6.3	6.1	4.1	3.9
Manual job	8.5	7.7	6.0	5.5
Average	7.5	7.1	4.9	4.7

Sympathetic and sensory nerves have similar courses in the hands; in case of nerve injury, the regions with sensory loss and the regions with sympathetic dysfunction are similar; therefore, the functional examination of sympathetic nerves and functional examination of sensory nerves are always combined clinically as the results can be more accurate and reliable. Under the condition that nerves are partially damaged or neuranagenesis is incomplete, the functions of sympathetic nerve can be recovered, but the dysfunction of sensory nerves can be persistent. Clinicians can observe during the examinations the plumpness of fingers and the presence of finger sweat; presence of local depression and dryness are the manifestations of nerve injuries.

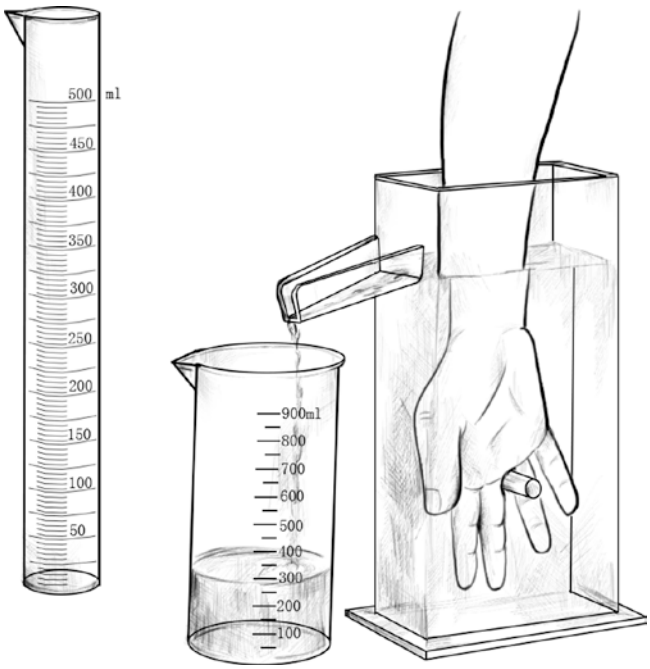


Fig. 2.43 Water-elimination test

1. Water-elimination test. It is a test to measure the extent of hand fullness after repair of hand nerve rupture (Fig. 2.43). The volume measurement of last segment of fingers is carried out through water-elimination test with the flexion side and the transverse striation of the interphalangeal joint of the last segment of fingers as the borders. This method is very difficult to carry out and inaccurate, and now it is rarely applied.
2. Sweat secretion test. This is a method to check whether nerves are injured and it is also early evidence indicating whether nerves are rehabilitated. There are two methods of sweat secretion tests: one is iodamyl test; the other is observation of sweat secretion of hands through microscope.
3. Static and dynamic two-point discrimination determination. Two-point discrimination, especially the dynamic two-point discrimination determination, is a simple and effective method for examination of hand sensation and is also a pure sensation examination method; other tests such as object picking test with eyes closed and activity time determination method are also methods for examination of sensation, but they are based on the prerequisite that the motor functions are good.

   The static two-point discrimination can be performed with Disk-Criminator or compasses (Fig. 2.44), and the dynamic two-point discrimination can be performed using paper clip. During dynamic two-point discrimination, first bend the paper clip into 2 ft, flatten through

Fig. 2.44 Use of Disk-Criminator to detect two-point discrimination

grinding the part with rough edges, draw the 2 ft of the paper clip close to each other, glide it along one side of the finger from the proximal part to the distal part, and ask the patient whether one point or two points are moving; then separate the distance between the 2 ft by 5–8 mm, move it from the proximal part to the distal part, and then ask the patient how he/she feels. When the patient is aware of this examination method, then officially start the examination from another finger: first start with the distance of 5–8 mm and then slowly narrow the distance until 2 mm. Sometimes, if the patient cannot clearly answer whether one point or two points are moving, the test can be repeated for several times until most answers are similar [53].

Li Diren carried out two-point discrimination determination on the 39 hands of 32 healthy persons aged between 4 and 83 years old. The dynamic two-point discrimination of thumb pulp is about 2 mm, there is no difference between the left hand and the right hand, the ulnar side and the radial side of fingers are similar, and there is no obvious relation with gender and age. This test is often used for the examination of effect after nerve repair and can serve as the basis for diagnosis of nerve compression syndrome [53].

4. Other examinations. They are touch, pain, cold and heat sensation examinations and tuning fork test.

The degree of the nerve injuries can be determined according to the movement and sensory paralysis. Determination of nerve sensations covers tenderness, allergies, thermalgia, etc. Ninhydrin test (sweat secretion test) is an important method to detect nerve functions. Two-point discrimination is an effective method to detect the sensitivity of tactile sensation, and the two-point discrimination of normal thumbs is 1–4 mm; when the two point discrimination is over 20 mm, the tactile sensation might disappear completely. Object picking test with the eyes closed is a method to detect the comprehensive sensory functions of fingers. During examination, the patient is asked to close the eyes and identify objects with hands. The examiner observes whether the patient can identify objects such as paper clip, pin, plastic product, screw, and nut; if the two-point discrimination is within 12 mm, it means the patient has the ability to identify objects (Tables 2.11, 2.12, 2.13, 2.14, and 2.15) [54].

Table 2.11 Test standard of two-point discrimination of AMA finger sensory functions [55]

Two point discrimination	Nerve damage
<6 mm	0
7~15 mm	50%
>15 mm	100%

Table 2.12 Normal value of two-point discrimination of finger palmar surfaces (mm) [56, 57]

Author	Last segment of fingers	Fingers with callosity	Middle segment of fingers	Proximal segment of fingers
Moberg	2~4	4~6		4~6
Parry	0.5~4		1~6	4~6
Gellis, Pool	2~4			
Millesi, Rinderer	1.5~6			

Table 2.13 Normal value of two-point discrimination of finger dorsal surfaces (mm) [56, 57]

Author	Last segment of fingers	Middle segment of fingers	Proximal segment of fingers
Parry	1~6	1~8	1~12
Gellis, Pool		2~7	

Table 2.14 Normal value of two-point discrimination of hands (mm) [56, 57]

Author	October	Thumb web of back of hand	Palmar thenar area	Central region of palm	Palmar hypothenar area
Moberg	8~11				
Parry			4~11	4~15	5~9
Gellis, Pool		5~15	4~8	4~15	4~8

Table 2.15 Mean time of using object picking test with closed eyes to identify objects (Parry) [56, 57]

Objects	Mean identification time (s)	Objects	Mean identification time (s)
Small coin	2	Key	2
Large coin	3	Paper clip	2
Plastic piece	4	Soft cork stopper	2
Small wooden block	5	Matchsticks	2
Abrasive paper	2	Screw	2
Rubber band	1	Safety needle	2

Sensory function of the hand is an important hand function. During the Second World War, the sensation was classified into five grades, i.e., grade 0 to grade 5. In 1954, the British Medical Research Council revised and improved the method and developed a 0–4 rating method, which has been adopted by the majority of scholars and has become a common sensation assessing method at present [56, 57].

Level 0–4 grading: S_0, sensory loss; S_1, deep sensory loss; S_2, partial shallow pain sensation and tactile sensation recovery, to protect injured fingers from damages; S_2^+, the same as S_2, but there is a hypersensitivity phenomenon; S_3, shallow pain sensation and tactile sensation recovery; there is no skin hypersensitivity phenomena; S_3^+, the same as S_3, with good positioning capability; two-point discrimination is close to normal; and S_4, normal sensation.

2.3.1.6 Evaluation of Pain

The evaluation of chronic pain is very difficult. Pain can be defined as "an uncomfortable feeling induced by individual afferent nerve stimulation accompanied by emotional state and modified by their past experience, induction, and mental state" which is based on a composite of many components. Through examination, the examiner can know whether pain is correlated with anatomical abnormality or nerve dysfunction or is pretended. Persistent functional lesions caused by pain can be determined only after the most appropriate physical adjustments and the best medical recovery [58]. The pain correlated with the proximal spinal nerve lesions can be classified into four grades according to their interference on the actions completed: ① weak (0~25%), namely, whether there is any discomfort; ② slight (26%~50%), namely, whether activities are interfered; ③ moderate (51%~75%), namely, whether activities are hindered; and ④ severe (76%~100%), namely, whether activities are hindered and vexation is induced. The fsunctional impairment induced by pain or discomfort can be similarly graded like the functional impairment that is used to assess the anesthesia or dactylolysis at this site and is expressed as a percentage (e.g., 100% of the patients with a severe predisposing cause may lose the practical value of limbs). AMA classified the damage criteria of complex regional pain syndrome (CRPS) (Table 2.16) [55], which is of important reference and significant value to the diagnosis and treatment in clinical work.

2.3.1.7 Examination of Hand Appearance

Hand appearance can be divided into passive hand appearance and active hand appearance. Passive hand appearance is the position of the hand when it is being rested, which is a common form of artificial hand imitation. Active hand appearance is the position of the hand when it is moving. Surgeons assess the appearance of the hand according to its position when it is moving or not moving. Scar, stiffness, joint imbalance, rotating deformity, etc. are included in the examination of hand appearance.

2.4 Judgment Criteria

2.4.1 Assessment of Amputated Extremities

The gradation and definition (Table 2.17) of limb damage degree by AMA are helpful to the understanding and interpretation of other assessment criteria.

Amputation of the entire upper limb or 100% of upper limb loss is defined as 60% loss of entire human functions. The amputation at the level of distal insertion of the elbow biceps means 95% loss of the functions at one upper limb; the amputation at the proximal level of metacarpophalangeal joint means 90% loss of the functions at one upper limb.

Dactylolysis of fingers and thumb means 100% loss of the functions of one hand or 90% loss of the functions of one upper limb. As the loss of one upper limb is equal to 60% loss of the entire human function, 90% loss of the function of one upper limb is equal to 54% loss of the entire human function. By means of this method, the relation between the defects of each part of hands and the functional lesions of the

Table 2.17 Gradation and definition of limb damage degree by AMA

Classification	Existing problems	Damage degree	
		Percentage of the entire upper limb	Percentage of the entire body
Grade 0	No	0	0
Grade 1	Slight	1~13%	1~8%
Grade 2	Moderate	14~25%	9~15%
Grade 3	Severe	26~49%	16~29%
Grade 4	Very severe	50~100%	30~60%

Table 2.16 Classification of damage criteria of complex regional pain syndrome by AMA [55]

Diagnosis classification	Grade 0	Grade 1					Grade 2					Grade 3					Grade 4				
Range of injuries (upper limb)	0	1%~13%					14%~25%					26%~49%					50%~100%				
Number of trigger points		≥4					≥6					≥8					≥8				
Severity		Slight					Moderate					Severe					Very severe				
Classification	0	A	B	C	D	E	A	B	C	D	E	A	B	C	D	E	A	B	C	D	E
	Negative diagnosis	1	3	7	1	1	1	1	2	2	2	2	3	3	4	4	5	0	7	8	9
					1	3	4	7	0	3	5	6	2	8	4	9	0	6	0	0	0

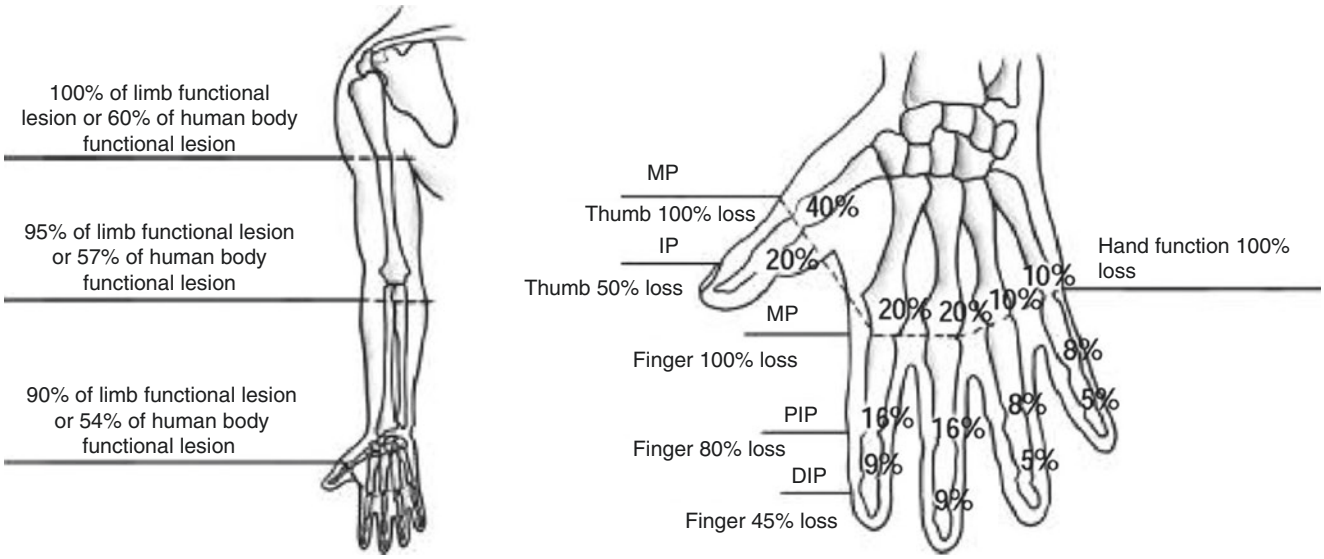


Fig. 2.45 Proportion of functional lesions of the corresponding fingers, hands, lateral upper limb and upper limbs on different levels of amputation

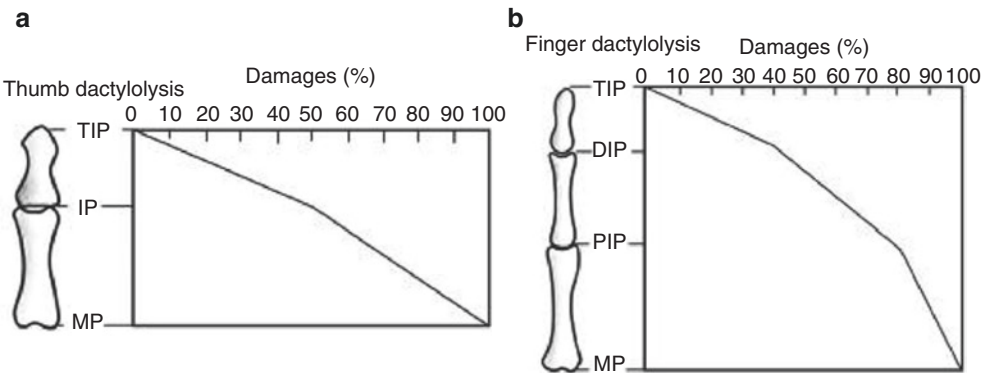


Fig. 2.46 Percentage of dactylolysis damages of the thumb and other fingers

upper limb and even the entire human body can be figured out (Fig. 2.45).

Among the entire hand functions, the thumb accounts for 40%, index finger and middle finger account for 20%, and ring finger and little finger account for 10%.

The proportion of the functions of each site of fingers in the entire finger functions is as follows: the proximal segment and distal segment of the thumb account for 50% of the thumb function; the distal segment and the middle segment of other fingers account for 40% of the functions of the finger, and the proximal segment accounts for 20% of the functions of the finger (Fig. 2.46).

According to the proportion of the functions of each site of fingers among the entire finger functions, the relation between the defects of each site of fingers and the functional lesions of the entire hand, the upper limb, and even the entire body can be figured out. For instance, the complete defects of index finger mean 20% loss of the entire hand functions; the PIP dactylolysis of index finger means 80% loss of index finger functions, equal to 16% loss of the functions of the entire hand (80% × 20%). In case of loss of multiple fingers, the percentage of the functional loss of each site is added to get the total sum, which can reflect the degree of the functional loss of the entire hand. For instance, complete dactylolysis of the thumb is equal to 40% loss of the functions of the entire hand; the accompanying dactylolysis at the level of DIP of index finger means 40% loss of the functions of index finger; because the functions of index finger account for 20% of the hand functions, it means 8% loss of the functions of the entire hand (40% × 20%). Therefore, complete dactylolysis of the thumb accompanied by dactylolysis at the level of DIP of index finger can result in 48% loss of the functions of the entire hand.

AMA formulated a series of tables for the assessment of amputation damages, and the degree of functional loss of hands, upper limbs, and the entire body can be found directly in the tables. Table 2.18 is the diagnosis criteria for amputation damages [55].

Table 2.18 Diagnosis criteria of AMA amputation damages

Diagnostic criteria	Grade 0	Grade 1					Grade 2					Grade 3					Grade 4				
Damage range	0	1%~13% (percentage in upper limb)					14%~25% (percentage in upper limb)					26%~49% (percentage in upper limb)					50%~100% (percentage in upper limb)				
Classification		A	B	C	D	E	A	B	C	D	E	A	B	C	D	E	A	B	C	D	E
Thumb							18%	18%	18%	20%	22%	36%	36%	36%	38%	40%					
							Interphalangeal joint					Metacarpophalangeal joint									
												37%	37%	37%	39%	41%					
												1/2 metacarpus									
												38%	38%	38%	40%	42%					
												Carpometacarpal joint									
Index finger or Middle finger		8%	8%	8%	9%	10%	14%	14%	14%	16%	18%										
		Distal interphalangeal joint					Proximal interphalangeal joint														
							18%	18%	18%	20%	22%										
							Metacarpophalangeal joint														
							19%	19%	19%	21%	23%										
							1/2 metacarpus														
							20%	20%	20%	22%	24%										
							Carpometacarpal joint														
Ring finger or Little finger		5%	5%	5%	6%	7%															
		Distal interphalangeal joint																			
		7%	7%	7%	8%	9%															
		Proximal interphalangeal joint																			
		9%	9%	9%	10%	11%															
		Metacarpophalangeal joint																			
		11%	11%	11%	12%	13%															
		1/2 metacarpus																			
		12%	12%	12%	13%	13%															
		Carpometacarpal joint																			
Hand																	54%	54%	54%	58%	58%
																	Metacarpophalangeal joints of all fingers except thumb				
																	90%	90%	90%	92%	94%
																	Metacarpophalangeal joints of all fingers				
																	92%	92%	92%	94%	96%
																	From insertion of biceps muscle to metacarpophalangeal joint				
Arm																	92%	92%	92%	94%	96%
																	From insertion of triangular muscle to insertion of biceps muscle				
																	100%	100%	100%	100%	100%
																	Beyond insertion of triangular muscle				
Shoulder																	100%	100%	100%	100%	100%
																	At shoulder joint				

2.4.2 Assessment of Sensory Damage

Any loss of function induced by sensory disorder, pain, and discomfort must be plain and continuous. The loss of function at the dorsal side of fingers is not disability-causing. Only the loss of function at the palmar side of fingers has a disability-causing effect on finger functions.

1. Complete loss of function. The complete loss of function at the palmar side is considered to be 50% loss of hand functions. For example, the loss of function of peripheral nerves at both sides of the thumb means 50% loss of finger functions (Fig. 2.47). As the thumb function occupies 40% of the hand functions, the complete loss of thumb sensation can result in 20% loss of hand functions (50% × 40%). In a similar fashion, the complete loss of functions of the index finger and middle finger can result in 10% loss of hand functions; the complete loss of functions of the ring finger and little finger can result in 5% loss of hand functions [52, 58].

   Swanson formulated the criteria for assessment of the functions after nerve injuries of each finger (Tables 2.19 and 2.20) [52], and the assessment results can be directly found in the table according to the injuries.

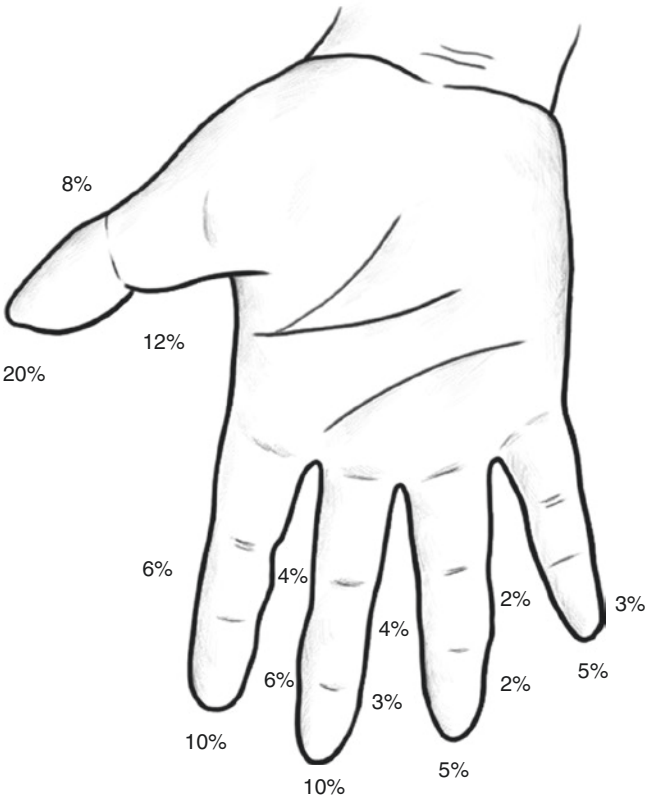


Fig. 2.47 Damages caused by complete loss of finger sensations to the entire hand functions, calculated according to 50% of dactylolysis damages

2. Transverse (segmental) sensory deprivation. Transverse (segmental) sensory deprivation is calculated by the percentage of the functions of each part of fingers in the functions of the hand. For example, the sensory deprivation at the level of thumb IP is equal to the dactylolysis from the IP level, namely, 25% (50% × 50%) loss of thumb sensation, or equal to 10% loss of the entire hand functions (25% × 40%).
3. Longitudinal sensory deprivation. Due to the different importances of sensory functions on both sides of fingers, the damages of sensory deprivation to hand functions vary (see Fig. 2.47). The sensory deprivation at the radial side of the thumb can result in 40% of the functional damages of the thumb, and the sensory deprivation at the ulnar side can result in 60% of the functional damages of the thumb. The sensory deprivation at the ulnar side of the remaining fingers can result in 40% of the functional

Table 2.19 Criteria for functional assessment after nerve injuries of thumb and little finger (Swanson)

Percentage in finger length (%)	Percentage of hand injuries					
	Transverse loss		Longitudinal loss			
	Nerves of bilateral fingers		Nerves of ulnar fingers		Nerves of radial fingers	
	All (%)	Partial (%)	All (%)	Partial (%)	All (%)	Partial (%)
100	50	25	30	15	20	10
90	45	23	27	14	18	9
80	40	20	24	12	16	8
70	35	18	21	11	14	7
60	30	15	18	9	12	6
50	25	13	15	8	10	5
40	20	10	12	6	8	4
30	15	8	9	5	6	3
20	10	5	6	3	4	2
10	5	3	3	2	2	1

Table 2.20 Criteria for functional assessment after nerve injuries of the index, middle, and ring fingers (Swanson)

Percentage in finger length (%)	Percentage of hand injuries					
	Transverse loss		Longitudinal loss			
	Nerves of bilateral fingers		Nerves of ulnar fingers		Nerves of radial fingers	
	All (%)	Partial (%)	All (%)	Partial (%)	All (%)	Partial (%)
100	50	25	20	10	30	15
90	45	23	18	9	27	14
80	40	20	16	8	24	12
70	35	18	14	7	21	11
60	30	15	12	6	18	9
50	25	13	10	5	15	8
40	20	10	8	4	12	6
30	15	8	6	3	9	5
20	10	5	4	2	6	3
10	5	3	2	1	3	2

damages of this finger, but the little finger is an exception because the sensation of the ulnar side of the little finger is more important. Then the functional damages of each finger are converted into the functional damages of the entire hand. For example, the complete loss of thumb sensation is equal to 20% of the damages of the entire hand, and the longitudinal sensory deprivation at the ulnar side of thumb is equal to 60% of the functional damages of the thumb, namely, 12% of the functional damages of the entire hand (60% × 50% × 40%).

2.4.3 Assessment of Motor Damage

The motor functional loss of hands is the final clinical manifestations of various hand diseases mostly induced by lesions in joints, tendons, muscles, nerves, and vessels. Clinically, it is difficult and impossible to evaluate the indexes of the above factors one by one. Therefore, there are many factors that contribute to motor functional loss, and numerous scholars have been exploring the clinical methods for comprehensive assessment [52, 54, 55, 59].

During many years, Boyes, Litchmon, Paslay, Vant, Heiple, White, Tubiana et al. have proposed different assessment methods in terms of the motor functional loss of the hand and consider that Swanson's assessment method on the basis of AMA "assessment of continuous injuries of four limbs, lumbus and back" and in combination with Boyes' linear measurement method formula "A% + B% × (100% − A%) = Composite Value of A% + B%" is relatively systematic and practical and is recognized by the International Hand Surgery Societies. This method is recommended in China.

According to AMA guidelines, the calculation of damage value of arthrokleisis and flexion is based on the assumption that the normal dorsal extension at MP and IP joints is 0°. Swanson proposes a compensation method used to assess the dorsal extension ability and he makes a special compensation for the hyperextension of normal upward movement by 20° of MP joints [54, 60].

The movement amplitude of one joint is the sum of the angles constituted by the maximal dorsal extension to the maximal flexion movement. In determining the movement amplitude, the measure is expressed by V: V_{flexion} (V_{flex}) = attainable maximal flexion degree and $V_{\text{extension}}$ (V_{ext}) = attainable maximal dorsal extension degree.

1. Assessment of movement amplitude (V) of metacarpophalangeal joint The normal movement amplitude of MR joint is 0°~90°. V_{flex} is 90° and V_{ext} is 0°, meaning no presence of movement damages. The hyperextension of normal MP joint will be discussed in the following section.
   (a) Range of flexion of joint loss (F): when the range of flexion of the joints is calculated, the lost range of flexion is expressed by F, equal to the value obtained by the theoretically maximal V_{flex} minus the measured V_{flex} (Fig. 2.48). For a MP joint with V_{ext} = 0° and V_{flex} = 60°, the lost range of flexion can be expressed as follows: F = 90°(maximal V_{flex}) − 60°(measured V_{flex}) = 30°.
   (b) The range of dorsal extension of joint loss (E): if a joint has a loss of dorsal extension 20°, it is expressed as V_{flex} = 20° (Fig. 2.49). The range of dorsal extension of joint loss can be expressed with E, equal to the value obtained by the measured V_{ext} value minus the minimal V_{ext} value in theory. For a loss of dorsal

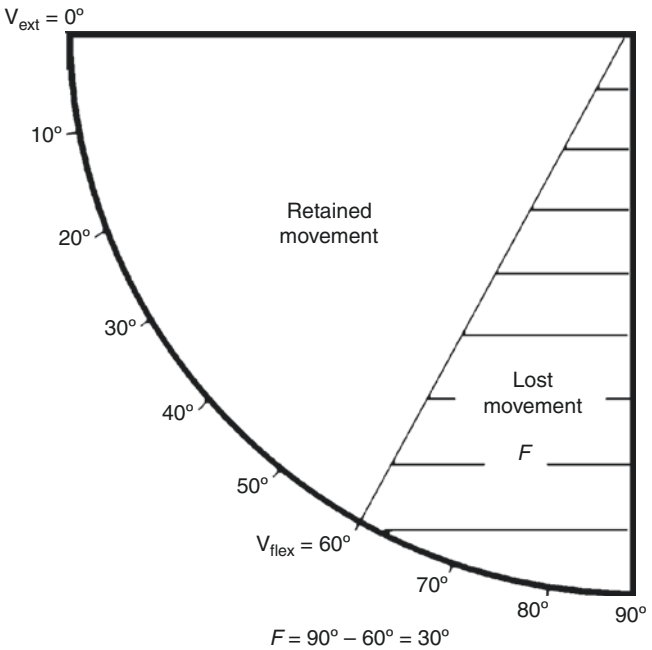


Fig. 2.48 Calculation of range of flexion of joint loss

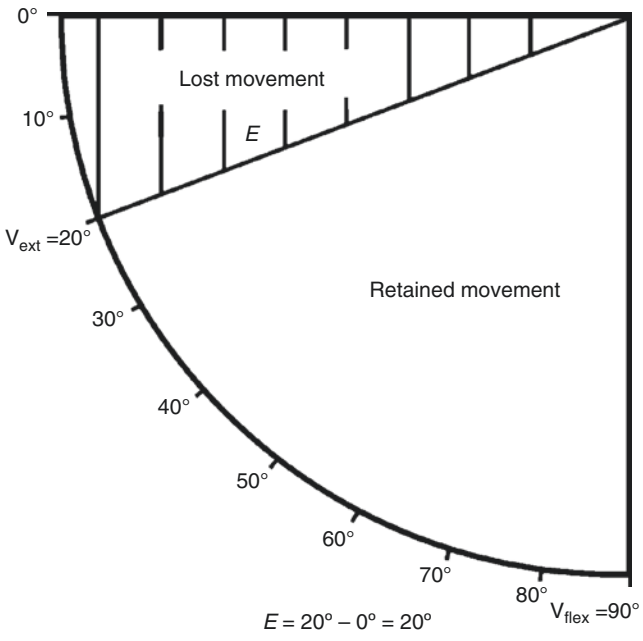


Fig. 2.49 Calculation of range of dorsal extension of joint loss

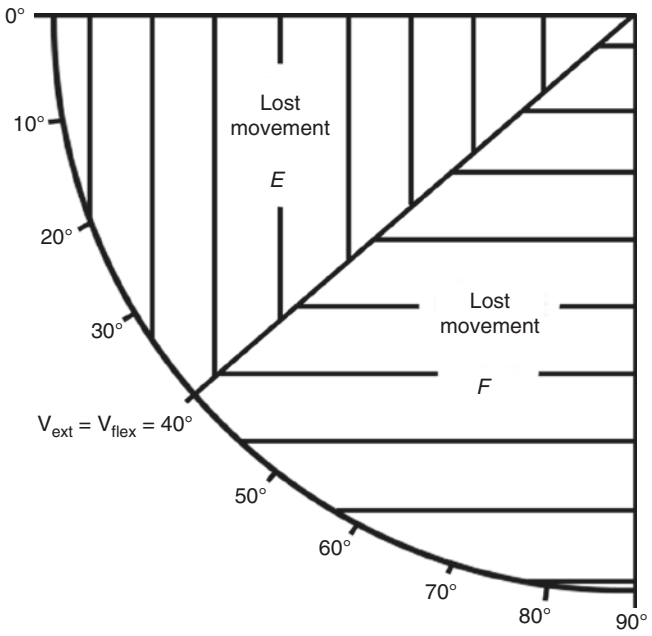


Fig. 2.50 Complete loss of movement when ankylosis occurs (*A*) is equivalent to the loss of dorsal extension (*E*) 40° + flexion loss (*F*) 50°, that is, *A* = 90°

extension of the MP joint 20°, it can be expressed as *E* = 20° (measured V_{ext}) −0° (minimal V_{ext}) = 20°.

(c) Arthrocleisis (*A*): in case of a decrease in range of flexion, V_{flex} decreases; in case of a decrease in range of dorsiflexion, V_{ext} increases. The two values finally stay at the same angle, namely, $V_{ext} = V_{flex}$, and at this time this presents arthrocleisis (Fig. 2.50). Complete loss of joint motor function is represented by *A*, which does not mean that ankylosis occurs at this angle, but it refers to the sum of decrease in range of dorsal extension induced by rigidity (*E*) and the decrease in range of flexion (*F*). Complete loss of joint movement can be expressed as *A* = *E* + *F*. If ankylosis occurs at 40°, then $V_{ext} = V_{flex} = 40°$, *E* = 40°, *F* = 90° − 40° = 50°, *A* = 40° + 50° = 90°.

It should be noted that, *A* value reflects the total loss of joint movement and is always equal to the normal movement amplitude of the joint. As for the MP joint, no matter where the rigidity occurs to the arc, as long as $V_{flex} = V_{ext}$, then *A* = 90°. When rigidity occurs at 30°, *A* = 30° (*E*) + 60° (*F*) = 90°; When rigidity occurs at 80°, *A* = 80° (*E*) + 10° (*F*) = 90°.

Finger functional injury may be induced by dorsal extension loss (*E*), loss of flexion (*F*) or arthrocleisis (*A*). Therefore, the percentage of finger functional lesions can be expressed using I_E, I_F and I_A, i.e. I_E for dorsal extension damage, I_F for flexion damage, I_A for rigidity damage (these can be measured in examinations). In a more professional sense, the percentage of damage can be expressed as: I_E is V_{ext} (measured minimum dorsal extension angle) function; when the V_{ext} reaches its theoretical minimum value (when MP joint is at 0°), I_E is 0; I_F is V_{flex} (measured maximum flexion angle) function; when the V_{flex} reaches its theoretical maximum value (when MP joint is at 90°), I_F is 0; when $V_{flex} = V_{ext}$, $I_A = I_E + I_F$.

Table 2.21 Rate of finger functional lesions induced by functional loss of different flexion positions of MP joint

Joint flexion angle	Range of flexion of loss (*F*)	Finger functional lesion rate (I_F) (%)
0°	90°	55
10°	80°	49
20°	70°	43
30°	60°	37
40°	50°	31
50°	40°	24
60°	30°	18
70°	20°	12
80°	10°	6
90°	0°	0

Table 2.22 Rate of finger functional lesions induced by different rigidity positions of MP joint

Arthrocleisis angle	Finger functional lesion rate (I_A) (%)
0°	55
10°	52
20°	48
30°	45
40°	54
50°	63
60°	72
70°	82
80°	91
90°	100

Functional lesions are expressed as a percentage, and the loss of function affected is reflected in the 100% graduation scale. AMA guidelines provide the percentage of finger functional lesions from 0° to 90° induced by flexion function loss (*F*) and arthrocleisis (*A*) (Tables 2.21 and 2.22), and this percentage can also be expressed with figures (Figs. 2.51 and 2.52). According to the formula *A* = *E* + *F* (or *E* = *A* − *F*), we can obtain the dorsal extension functional lesions of a certain angle (IE) according to the formula $I_E = I_A - I_F$. For example, when the arthrocleisis is at the 30° position, according to AMA table, I_A = 45%, while I_F = 37%, so you can get the I_E value: I_E = 45% (I_A) −37% (I_F) = 8%. This step can also be used for each angle on the 0° ~ 90° motion arc, to obtain their I_E values (Fig. 2.53). In case of joint dorsal extension injury 40°, I_E = 54% (I_A) −31% (I_F) = 23%. I_E is the function of V_{ext}, and when V_{ext} = 0° or *E* = 0°, its value is 0. But the AMA guideline does not consider the value of MP joint hyperextension, so we will somewhat modify the I_F value of AMA guideline (Table 2.21), to compute the value of hyperextension 20° of normal MP which is also considered normal (Fig. 2.54); so when arthrocleisis is at

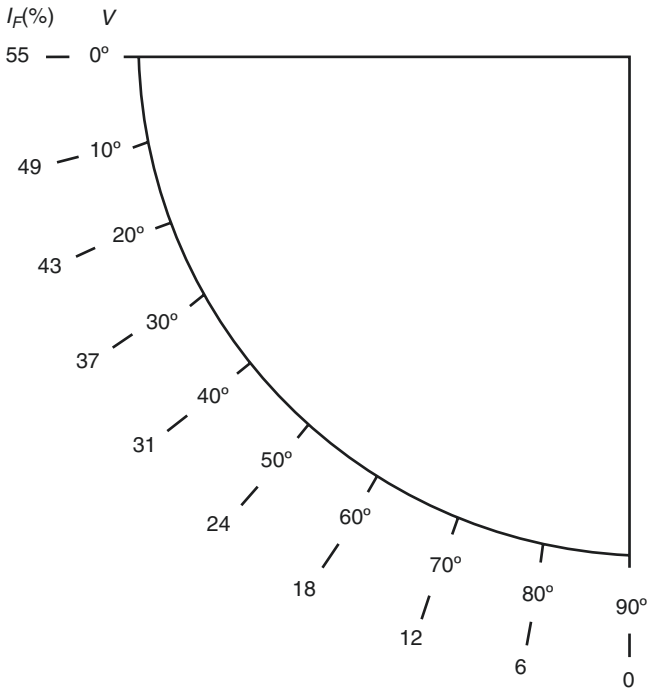


Fig. 2.51 Rate of finger functional lesions (I_F) induced by flexion function loss (F) converts the AMA's I_F value (Table 2.21) into arc motion

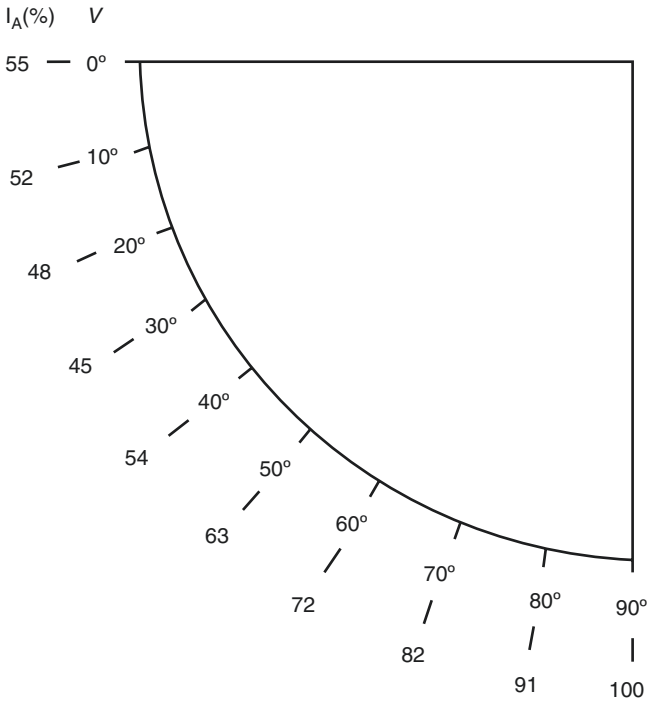


Fig. 2.52 Rate of finger functional lesions (I_A) induced by arthrocleisis (A) converts the AMA's I_A value (Table 2.22) into arc motion

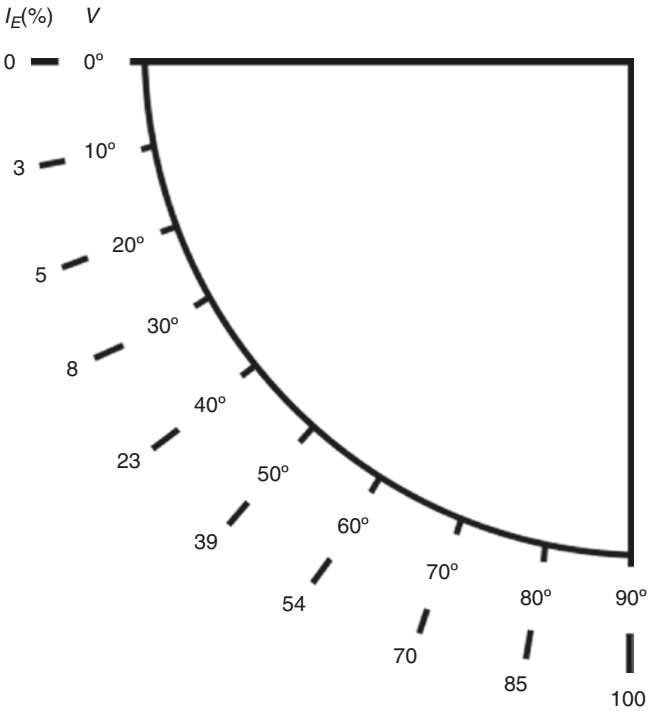


Fig. 2.53 According to the formula $I_A = I_E + I_F$ or $I_E = I_A - I_F$, obtain the corresponding I_E value

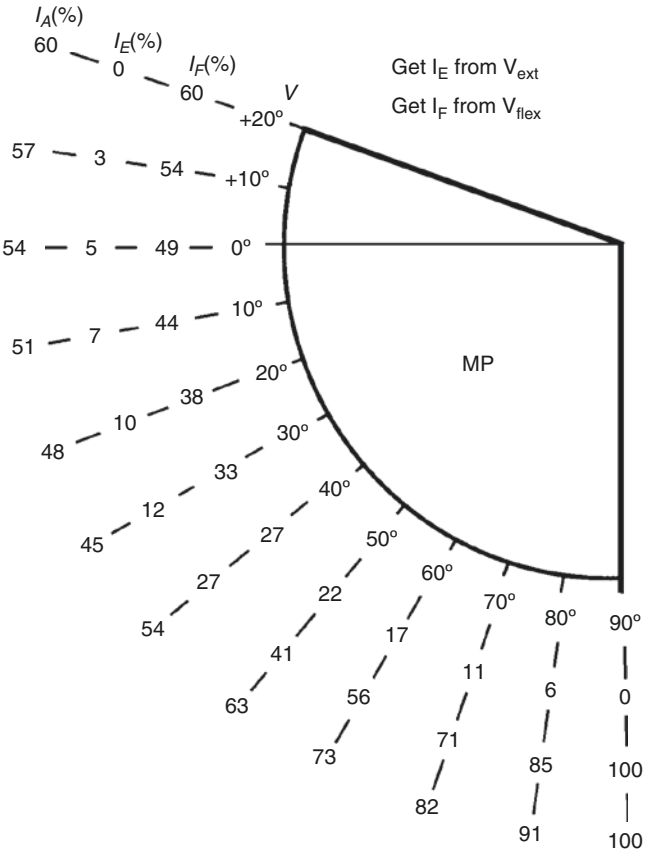


Fig. 2.54 MP joint functional lesion rate I_A is minimal at 30° or at other functional positions (45%). The figure includes the functional lesion rate of hyperextension deformity

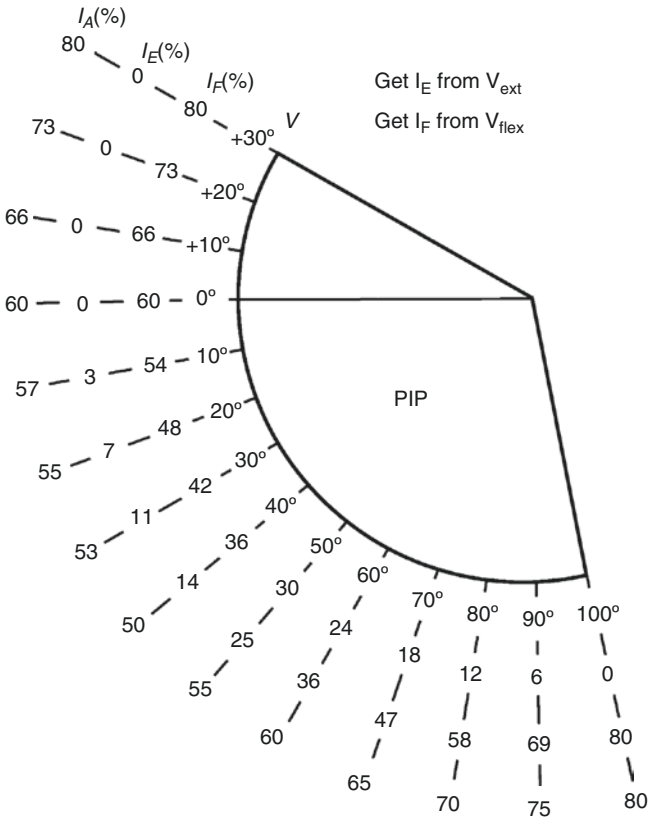


Fig. 2.55 As for the assessment of PIP joint damage rate, the function position of PIP joint is at 40°, and the IA at this angle is the lowest (50%)

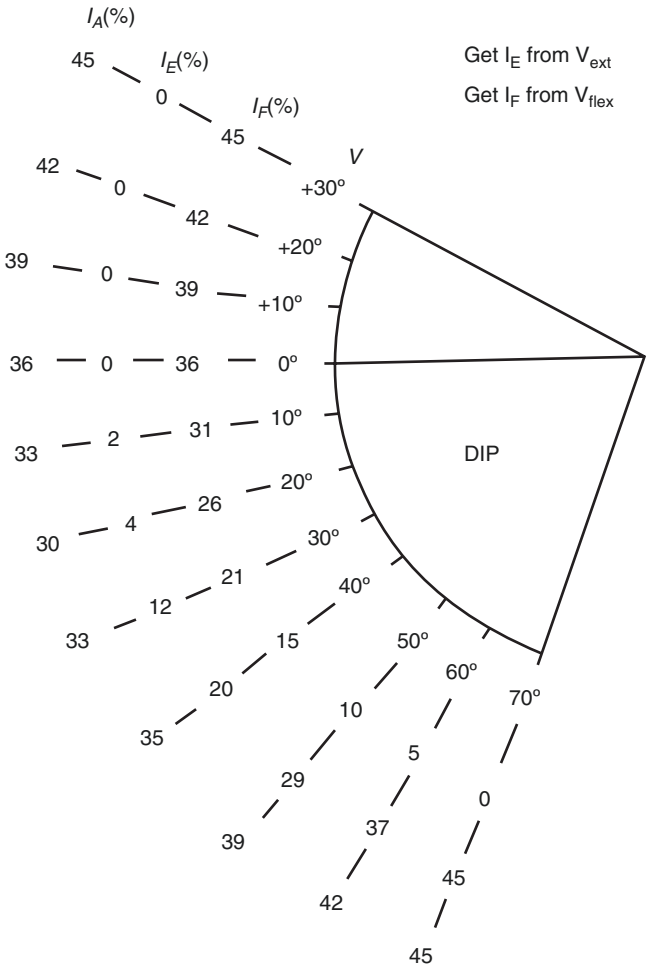


Fig. 2.56 As for the assessment of DIP joint (including hyperextension position) damage rate, the function position of DIP joint is at 20°, and the IA at this angle is the lowest (30%)

30° position, I_F 37% in Table 2.21 is replaced by 33%. According to the above formula, I_E = 45% (I_A) – 33% (I_F) = 12% [54, 59–61].

IE derivation is very important. It provides the values of I_E and I_F in order to correctly estimate the percentage of functional lesions. This not only reflects the degree of loss of movement but also and more importantly reflects the position with functional loss in the finger movement arch.

For example, a MP joint has a movement amplitude of 30°. In case of movement from extension −10° to flexion 40°, the functional lesion is not as severe as the movement from −50° to 80°. As for the MP joint moving from extension −10° to flexion 40°, I_E = 7%, I_F = 27%, and the total damage rate is 34%. As for the MP joint moving from extension −50° to flexion 80°, I_E = 41%, I_F = 6%, and the total damage rate is 47%. If the MP arthrocleisis is at the 30° position (i.e., functional sites), I_A = 12% (I_E) + 33% (I_F) = 45%, and the value is low. If the MP arthrocleisis is at the 80° position, the degree of damage is much more serious, i.e., I_A = 85% (I_E) + 6% (I_F) = 91%.

The tables of functional lesions of the finger, thumb, wrist, elbow, and shoulder joint are derived from the abovementioned basic formula.

2. Assessment of functional lesions of the index, middle, ring, and little fingers. Figs. 2.55 and 2.56 show the three different functional lesions (I_A, I_F, I_E) of each finger joint (MP, PIP, DIP), and the functional sites of each joint are based on AMA guideline with materials of hypertension attached. For a normal hand, a MP joint can realize hyperextension by 20°; in case of loss of this hyperextension function, only a very small damage rate is marked. When the MP joint is in dorsal extension 0° position, I_E = 5%, PIP and DIP joint normal dorsal extension is 0°, so when these joints are at dorsiflexion 0° position, I_E = 0. However, if the hyperextension angle of these joints is taken into account, when ankylosis occurs to the hyperextension position, the damage rate of flexion function can be determined. For example, when the PIP arthrocleisis is at the 30° position, 80% of functional lesion can be considered.

As for the functional decrease and lesions induced by the rigidity of each joint (I_A), the lowest value is reached in the functional position; for example the MP joint I_A is

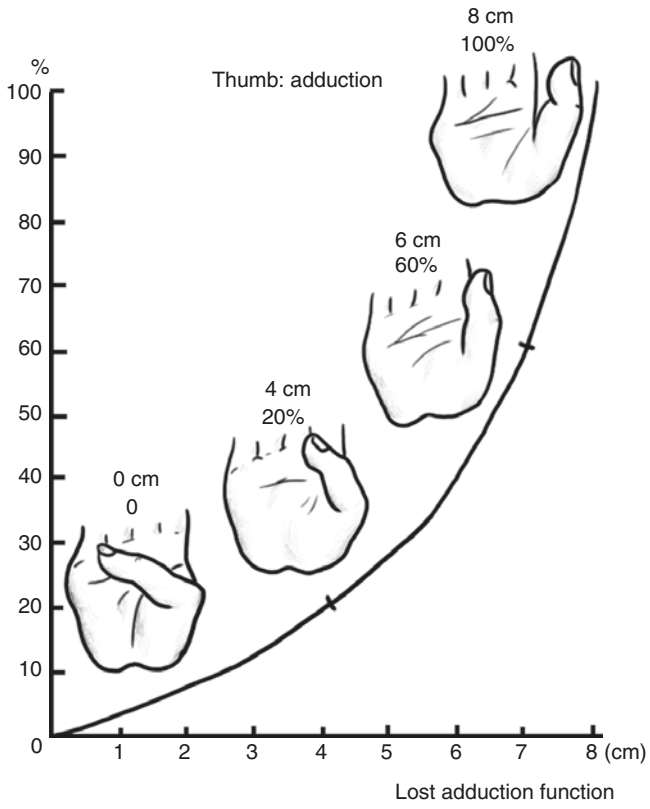


Fig. 2.57 Calculation of lesion rate of thumb adduction function

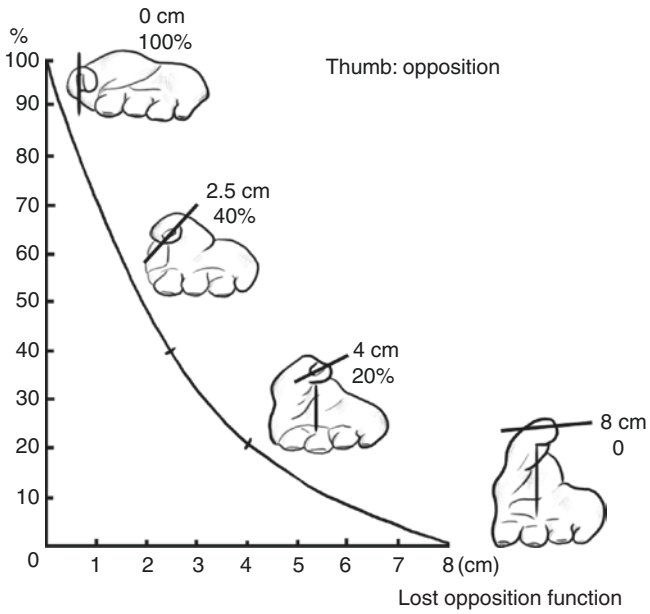


Fig. 2.58 Calculation of lesion rate of thumb opposition function

Table 2.23 Percentage of lesion rates of thumb adduction and opposition function in the lesion rate of the entire thumb function (based on the concept that the adduction function and opposition function, respectively, occupy 20% and 60% of the thumb function)

Distance (cm)	Lesion rate of thumb adduction function (%)	Lesion rate of thumb opposition function (%)
0	0	60
1	0	42
2	1	29
3	3	19
4	4	12
5	6	7
6	8	3
7	13	2
8	20	0

45% at 30°, PIP joints I_A is 50% at 40°, and DIP joint I_A is 30% at 20°.

Figure 2.54 can be used to measure the movement amplitude and assess the functional lesions. Take the MP joint activities from −20° to 60° for example. The dorsal extension damage of this angle should be checked in the row labeled with I_E, that is, I_E is 10% of dorsal extension injury. Flexion damage should be viewed in the row labeled with I_F, that is, I_F is 17%. The total functional lesions of this patient is 10% + 17% = 27%.

3. Assessment of thumb functional lesions. The thumb function occupies 40% of the entire hand function and consists of three kinds of functions: ① Flexion and extension functions of MP and IP; ② adduction and abduction functions; ③ opposition function. The flexion and extension functions of MP and IP joints occupy 20% of the thumb motor function, the adduction function occupies 20% of the thumb motor function, and the opposition function occupies 60% of the thumb motor function.
   (a) Adduction and opposition functional lesions: as is shown in Figs. 2.57 and 2.58, the functional lesions of the entire thumb are the sum of the value of each function of the thumb. The thumb adduction function is measured by the straight-line distance between the palmar—distal palmar-cross striation of thumb IP joint and the MP joint of little finger. In Fig. 2.57, the curve indicates that the lesion rate of thumb adduction function is correlated with the adduction function loss instead of the entire thumb function. As the adduction function occupies 20% of the entire thumb function, the lesion rate of the entire thumb function should be 20% of the lesion rate in the figure, as shown in Table 2.23. The thumb opposition function is measured by the maximal distance between the palmar—distal palmar-cross striation of thumb IP joint and the MP joint of middle finger. In Fig. 2.58, the curve indicates that the lesion rate of thumb opposition function is correlated with the opposition function loss instead of the entire thumb function. As the opposition function occupies 60% of the entire thumb function, the lesion rate of the entire thumb function

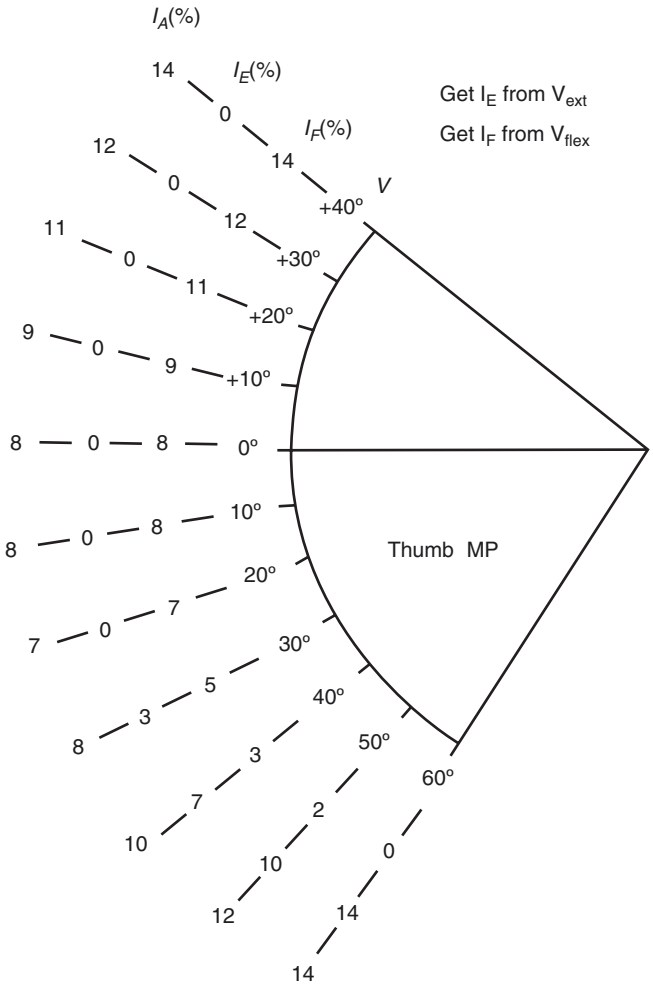


Fig. 2.59 Rate of lesions of hand functions caused by loss of flexion and extension function or arthrocleisis of thumb MP joint; the functional position is 20°

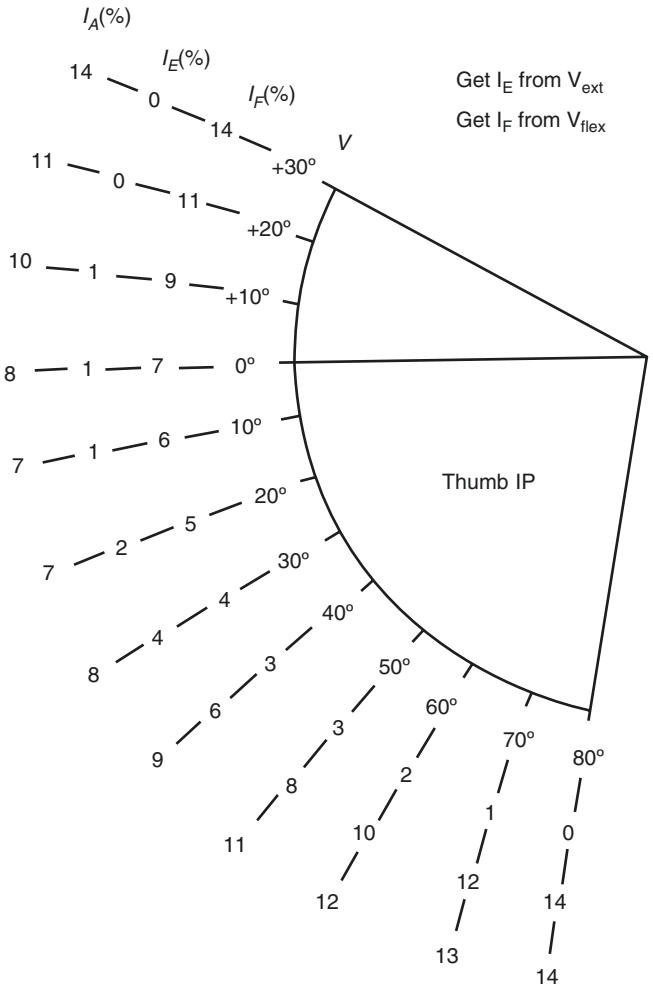


Fig. 2.60 Rate of lesions of hand functions caused by loss of flexion and extension function or arthrocleisis of thumb IP joint; the functional position is 20°

should be 60% of the lesion rate in the figure, as shown in Table 2.23.

(b) Flexion and extension functional lesions and arthrocleisis: Figs. 2.59 and 2.60, respectively, show the rate of lesions of hand functions caused by loss of flexion and extension functions or arthrocleisis of MP and IP joints. The flexion and extension functions occupy 20% of the entire thumb functions. The functional position of thumb MP and IP joints is flexion 20°, so the I_A value is the lowest at this angle, and it is the 7% lesion of rigidity.

In actual use, the function of each part can be assessed according to Figs. 2.57, 2.58, 2.59, 2.60 and Table 2.23, for the purpose of determining the influence of each direct lesion value on the entire thumb function. Then these values are directly added to get the entire functional lesions of the thumb without using the composition table. Table 2.24 shows the criteria established by AMA to assess the motor function lesions in the elbow and wrist, which can be directly queried [55, 59–61].

2.4.4 DASH Questionnaire

In 1996, Hudak et al. [47] first used DASH questionnaire to assess the patient's shoulder, arm, and hand function defect. Since then, increasing importance has been attached to DASH questionnaire by clinicians all over the world. DASH questionnaire has a total of 34 items, mainly used to assess functions closely associated with daily activities and the resulting symptoms. A 5-point scoring system is adopted for each item. The higher the score is, the severer the functional defect is. For athletes and musicians, additional items are needed for assessment. Although DASH questionnaire is not perfect, it is simple and easy to use with a good reliability and repeatability as well as a wide scope of application. It has been translated into multiple languages and widely applied in numerous countries.

DASH questionnaire is divided into three parts: A, B, and C. Part A (Table 2.25) is used to learn the functional

Table 2.24 AMA guiding criteria of motor function lesions [55]

Motor functions of each site	<10% of motor function lesions	>10% of motor function lesions
Elbow joint		
Flexion	≥120°	<120°
Extension	≥ − 40°	<−40°
Wrist joint		
Supination	≥20°	<20°
Pronation	≥45°	<45°
Flexion	≥45°	<45°
Extension	≥45°	<45°
Radial deviation	≥10°	<10°
Ulnar deviation	≥20°	<20°
Thumb		
Abduction	≥40°	<40°
Adduction	≤2.5 cm	>2.5 cm
Index finger		
Metacarpophalangeal joint flexion	≥70°	<70°
Metacarpophalangeal joint extension	≥ − 10°	<−10°

Table 2.25 DASH questionnaire (A)

Item		Mobility				
		No difficulty	Low difficulty	Some difficulty	Great difficulty	Unable
1	Open a tightly sealed or new glass bottle	1	2	3	4	5
2	Write	1	2	3	4	5
3	Open the door	1	2	3	4	5
4	Prepare a meal	1	2	3	4	5
5	Push open a gate	1	2	3	4	5
6	Place items into the space of a cabinet over the head	1	2	3	4	5
7	Do heavy household chores (e.g., wash wall, wash floors)	1	2	3	4	5
8	Do garden or yard work (e.g., cleaning, scarification, mowing, and trimming trees and flowers)	1	2	3	4	5
9	Make a bed	1	2	3	4	5
10	Carry a shopping bag or briefcase	1	2	3	4	5
11	Carry a heavy object (weighing more than 5 kg)	1	2	3	4	5
12	Change a lightbulb overhead	1	2	3	4	5
13	Wash or blow dry hair	1	2	3	4	5
14	Wash the back	1	2	3	4	5
15	Put on a sweater	1	2	3	4	5
16	Cut food with a knife	1	2	3	4	5
17	Join relaxing recreational activities (e.g., cardplaying, knitting)	1	2	3	4	5
18	Take strenuous exercise (e.g., golf playing, hammering, tennis playing)	1	2	3	4	5
19	Join recreational activities in which arms are moved freely (e.g., badminton playing, squash playing, and frisbee playing)	1	2	3	4	5
20	Drive or take means of transport	1	2	3	4	5
21	Sexual function	1	2	3	4	5
22	Effect on social interaction with family members, friends, neighbors or other groups	1	2	3	4	5
23	Effect on work or other daily activities	1	2	3	4	5

Note: Please evaluate your performance of the abovementioned actions last week and circle the number of the corresponding grade

activities of upper limb, Part B (Table 2.26) is used to investigate the symptoms of upper limb, and Part C (Table 2.27) is used to investigate the upper limb functions of athletes and musicians. Part A and Part B are already enough for most patients. In calculation, add all the selected numbers of Part A and Part B and then refer to the following formula:

DASH functional disorder (symptom) scores = [(the mean of scores *for n* answer) − 1] × 25, where *n* represents the number of questions that have been answered.

Table 2.26 DASH questionnaire (B)

	Item	Severity of symptoms				
		Not at all	Slightly	Moderately	Severely	Extremely
1	Arm, shoulder, or hand pain during break	1	2	3	4	5
2	Arm, shoulder, or hand pain during movement	1	2	3	4	5
3	Numbness or needle-like pain of shoulder, arm, or hand	1	2	3	4	5
4	Weakness of arm, shoulder, or hand	1	2	3	4	5
5	Stiffness of arm, shoulder, or hand	1	2	3	4	5
6	Effect of arm, shoulder, or hand pain on sleep	1	2	3	4	5
7	Poorer ability or lack of confidence due to functional disorder of arm, shoulder or hand	1	2	3	4	5

Note: ① Please evaluate the severity of the above symptoms last week and circle the number of the corresponding grade; ② DASH will not be evaluated if there are more than three missing items

Table 2.27 DASH questionnaire (C)

Item		Mobility				
		No difficulty	Low difficulty	Some difficulty	Great difficulty	Unable
1	Play the musical instrument or do sports as well as before	1	2	3	4	5
2	Play the musical instrument or do sports not as well as before due to arm, shoulder or hand pain	1	2	3	4	5
3	Be able to play the musical instrument or do sports as required	1	2	3	4	5
4	Be able to play the musical instrument or do sports for long	1	2	3	4	5

Note: ① Investigate the effects of shoulder, arm, and hand functional disorder on your musical or sports activities; ② if you play multiple musical instruments or do multiple sports activities, please write the most important musical instrument or sports event; ③ please circle the number of the corresponding grade according to the action performance last week

When DASH value is 0, it means the upper limb is completely normal; when DASH value is 100, it means that the upper limb suffers severe functional defect.

DASH questionnaire is used to know the symptoms and daily performance of upper limb of the patient. The patient should circle the number of corresponding grade in the table according to the actions it performs during the last week and must answer every question. If the patient does not perform the action last week, he/she can be asked to judge which item most conforms to his/her upper limb, and circle the corresponding number. No matter which hand performs the action and no matter how it is performed, the patient just needs to answer the question according to the actual condition.

Scoring Optional Modules: Add up assigned values for each item, divide by 4 (number of items), subtract 1, and multiply by 25. The score for an optional module may not be calculated if there are any missing items.

If you do not provide answer to over 10% of the questions (e.g., over three questions), DASH functional disorder (symptom) score will not be calculated according to this rule. Likewise, in the working module of high performance sports (or performance arts), as there are only four questions, every question should be answered. The rule of this missing item of this table is applicable to original version and modification version of scoring method.

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Classification of Congenital Deformities of Hands and Upper Limbs and Selection of Surgery Timing

3

Bin Wang, Wei Wang, and Feng Ni

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3.1 Classification of Congenital Deformities of Hands and Upper Limbs

Wei Wang

The congenital deformities of the upper limb are complicated, so establishment of a complete classification system for congenital deformities of the upper limbs is conducive to the understanding of anomaly formation, the design of clinical treatment planning, the study on clinical treatment and etiological factors, and the dissemination and academic exchange of knowledge on hand and upper limb congenital deformities.

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The classification of congenital deformities of hands and upper limbs includes the overall classification of congenital deformities of hands and upper limbs, each of which can be divided into different types. Up to now, there has been no complete method used to classify congenital deformities of hands and upper limbs, and the reasons are as follows: firstly, congenital deformities of hands and upper limbs vary greatly in shape, and their occurrence and development have no limits; secondly, congenital deformities of hands and upper limbs display various features in terms of functional defects; thirdly, there are differences in the anatomical structure and developmental defects in different patients; and in the process of growth and development, deformity occurrence and development will change, and some have not been known to people; fourthly, congenital deformities of hands and upper limbs are often accompanied by other deformities, which are also varied; although many authors continue to find new ones, there are still new complex deformities constantly discovered; and fifthly, many problems in the etiological factors of congenital deformities of hands and upper limbs are to be studied. All these show that the classification of congenital deformities of hands and upper limbs is a complex task, and the occurrence and development of congenital deformities of hands and upper limbs are changing. Therefore, there is no complete classification system for congenital deformities of hands and upper limb in the field of hand surgery all over the world. Currently the hand surgeons and plastic surgeons are using the congenital deformities of hands and upper limbs classification system proposed by Swanson in 1964 [1], and this system was accepted by the International Federation of Societies for Surgery of the Hand (IFSSH) in 1974 [2, 3]. This classification system is a relatively improved classification method for congenital deformities of hands and upper limbs. In spite of no coverage of every aspect, it takes into consideration the embryological, etiological, and anatomical features of congenital deformities of hands and upper limbs and classifies the deformities according to the morphological and structural features. Therefore, it is relatively systematic and detailed. Now, the Swanson's congenital deformities of hands and upper limbs classification system (IFSSH classification) is introduced as follows.

3.1.1 Type I: Limb Formation Disorder

3.1.1.1 Transverse Limb Defects

1. Shoulder level. Without limb deformity
   (a) The deformity of missing upper limbs below the shoulder (congenital amelia)
   (b) The deformity of missing upper limbs below the clavicles
2. Congenital broken upper arms at the level of the upper arm and deformity of missing upper limbs below the arms
   (1) High limb deficiency above the elbow
   (2) Low limb deficiency above the elbow
3. Congenital broken elbow at the level of the elbow and deformity of missing upper limbs below the elbow
4. Congenital broken forearm at the level of the forearm and deformity of missing upper limbs below the forearm (Fig. 3.1)
   (a) High limb deficiency below the elbow
   (b) Low limb deficiency below the elbow
5. Congenital broken wrist at the level of radiocarpal joint and deformity of missing limb blow the wrist (achiria)
6. Congenital broken wrist at the level of the carpal bone and deformity of missing hand below the carpal bone (Fig. 3.2)

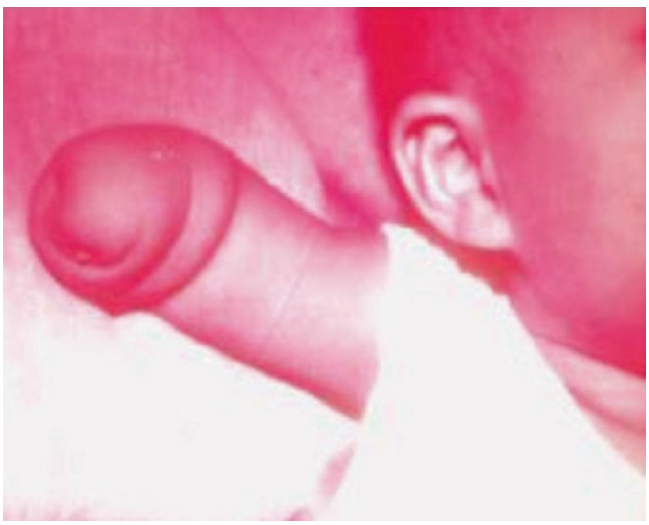

Fig. 3.1 Deformity of congenital broken forearm

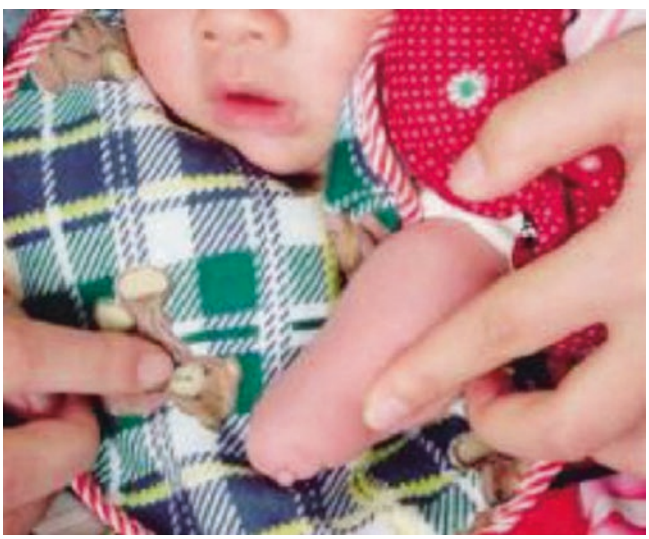

Fig. 3.2 Deformity of congenital broken wrist

(a) Deformity of missing hand below the proximal row of the carpal bone
(b) Deformity of missing hand below the distal row of the carpal bone

7. Congenital broken palm at the level of metacarpal bone and deformity of adactylia (Fig. 3.3)
8. Level of phalanx congenital broken fingers and deformity of adactylia
   (a) Ectodactylism at the level of proximal segment
   (b) Ectodactylism at the level of middle segment
   (c) Ectodactylism at the level of distal segment

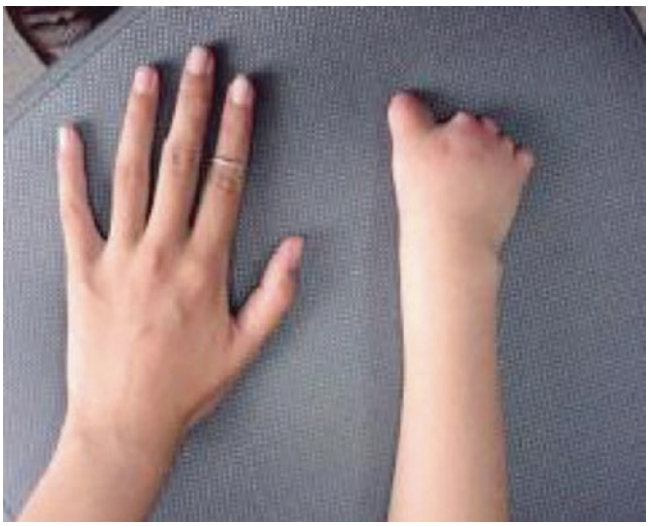

Fig. 3.3 Deformity of congenital broken palm

3.1.1.2 Longitudinal Limb Defects

1. Radial ray deficiency (preaxial) and radial ray defect deformity (radial club hands) (Fig. 3.4)
   (a) Normal radial type: ① thumb dysplasia (functional type); ② thumb dysplasia (nonfunctional type); ③ thumb missing
   (b) Radial dysplasia (fine but complete radial bone): ① thumb dysplasia (functional type); ② thumb dysplasia (nonfunctional type); ③ missing thumb; ④ Madelung deformity; ⑤ others
   (c) Partial radial absence: ① thumb dysplasia (functional type); ② thumb dysplasia (nonfunctional type); ③ thumb missing
   (d) Complete radial absence: ① thumb dysplasia (functional type); ② thumb dysplasia (nonfunctional type); ③ thumb missing
   (e) Others: ① poor or missing big thenar development; ② extensor dysplasia or absence; ③ flexor dysplasia or absence
2. Ulnar ray deficiency (postaxial) and ulnar ray defect deformity (ulnar club hand)
   (a) Normal ulnar types: ① poor metacarpal and phalangeal development; ② poor metacarpal development and phalangeal absence; ③ absence of both metacarpal and phalangeal bones
   (b) Ulnar dysplasia (ulna is fine and small, but complete): ① poor metacarpal and phalangeal development; ② poor metacarpal development and phalangeal absence; ③ absence of both metacarpal and phalangeal bones

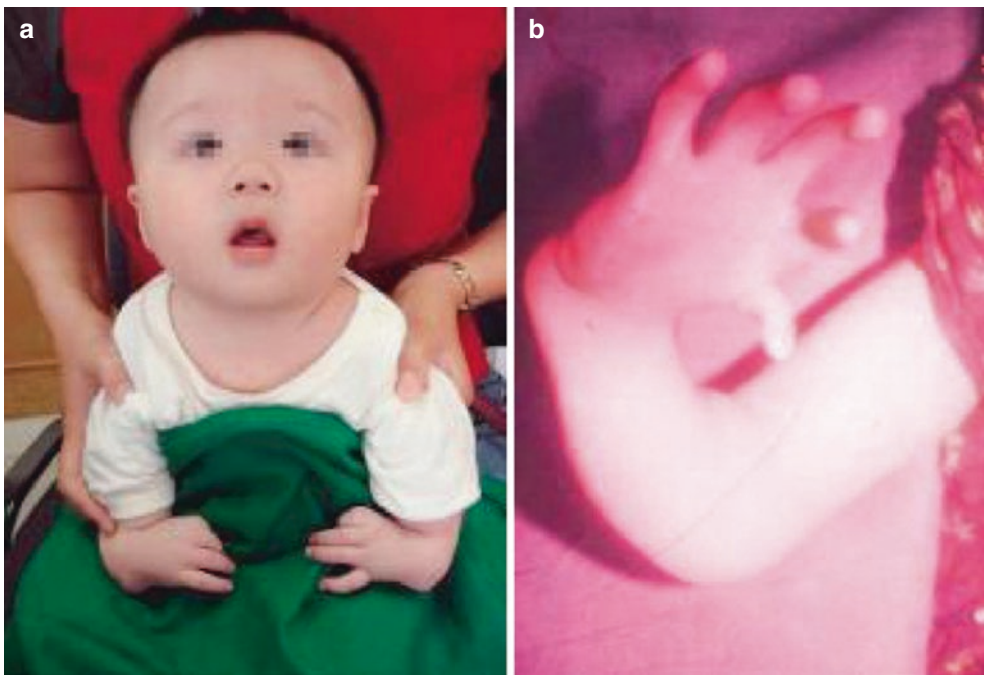

Fig. 3.4 Radial ray defect deformity (radial club hand). (**a**) Bilateral. (**b**) Unilateral

(c) Partial ulnar absence: ① poor metacarpal and phalangeal development; ② poor metacarpal development and phalangeal absence; ③ absence of both metacarpal and phalangeal bones
(d) Complete ulnar absence: ① poor metacarpal and phalangeal development; ② poor metacarpal development and phalangeal absence; ③ absence of both metacarpal and phalangeal bones
(e) Ulnar absence complicated by humeroradial joint fusion
(f) Absence or dysplasia of hypothenar muscles
(g) Absence or dysplasia of extensor
(h) Absence or dysplasia of flexor

3. Central ray deficiency and central ray deficiency deformity (cleft hand) (Figs. 3.5, 3.6, and 3.7)
   (a) Typical cleft hand: ① poor metacarpal and phalangeal development; ② poor metacarpal development and phalangeal absence; ③ absence of both metacarpal and phalangeal bones
   (b) Atypical cleft hand: ① syndactylous cleft hand; ② hyperphalangeal cleft hand; ③ monodactylous cleft hand; ④ others

3.1.1.3 Segmental Limb Deficiency (Hollow Developmental Disorder)

1. Seal hand (Fig. 3.8)
   (a) Proximal type: upper arm absence
   (b) Distal type: forearm absence
   (c) Complete type: absence of both forearm and upper arm
2. Others

3.1.2 Type II: Limb Dysdifferentiation

3.1.2.1 Suffering Soft Tissue

1. Sporadic at multiple sites and multiple joint flexion (including congenital multiple joint flexion deformity) (Fig. 3.9)

   The shape of patient's hand deformity seems to be the deformity of bilateral radial ray defect (radial club hand), but not the radial club hand, and the manifestations are as follows: ① poor development of multiple extensor muscles, dysplasia of deltoids, triceps, thumb extensor, and finger extensor; ② multiple joint flexion deformity with

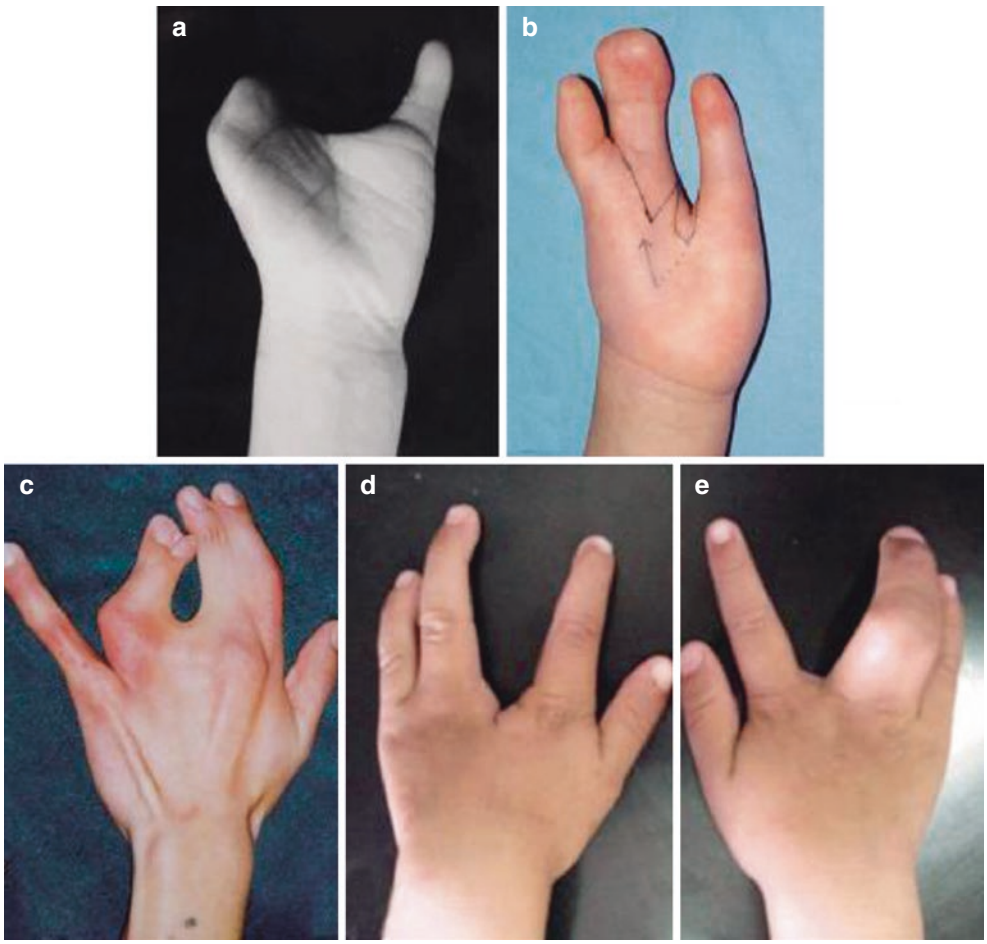

Fig. 3.5 Cleft hand. (**a**) Two-finger cleft hand; (**b**) three-finger cleft hand; (**c**) four-finger cleft hand; (**d**) five-finger cleft hand; (**e**) six-finger cleft hand with aschistodactylia

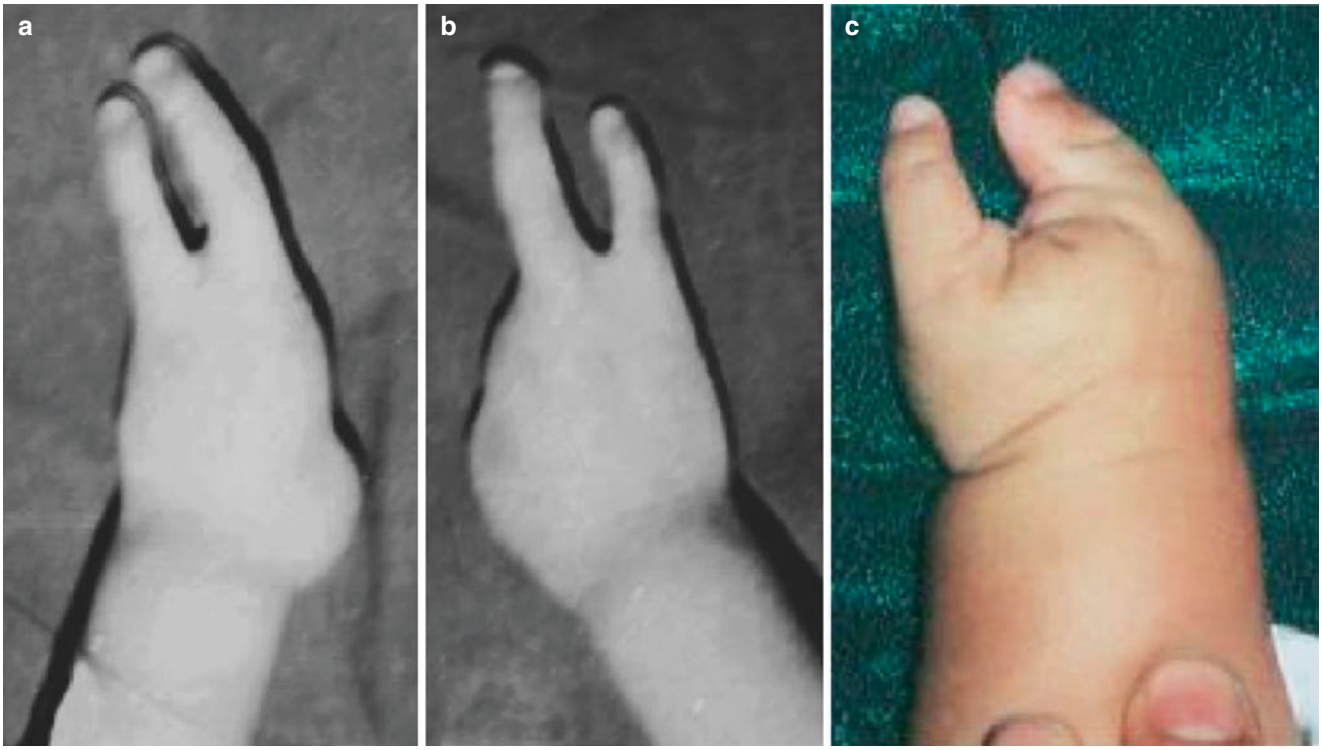

Fig. 3.6 Two-finger cleft hand; (**a**, **b**) the left and right hands of the same child. (**c**) The left hand of another child

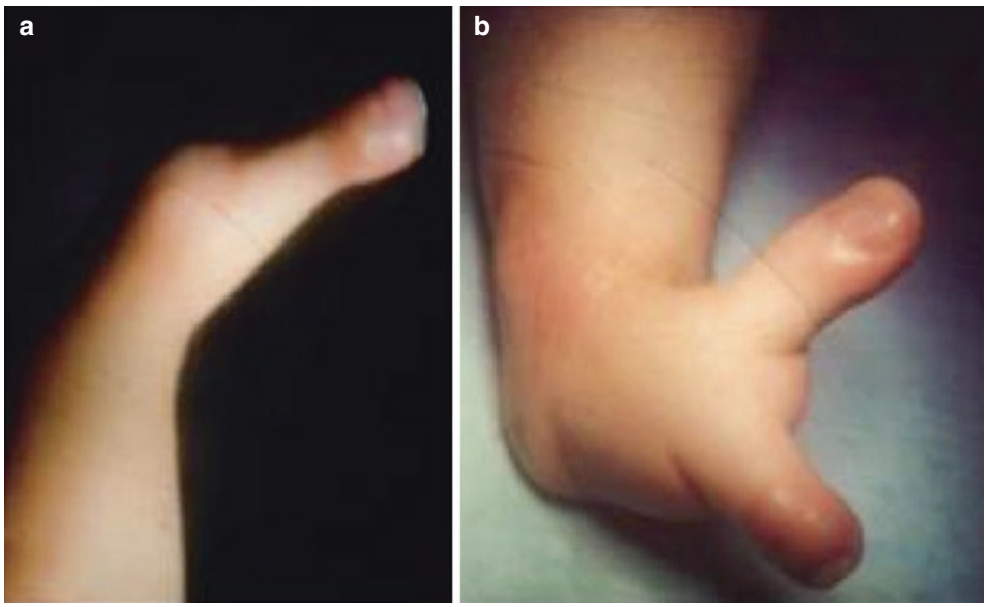

Fig. 3.7 Cleft hand and cleft foot (the same patient). (**a**) One-finger cleft hand. (**b**) Cleft foot

the axillary web wrist and flexion deformity of the wrist joint, thumb, and all the fingers; the hand shape seems to be ulnar deviation of the hand (windblown hand), but the development of thenar and hypothenar muscles is good.

(a) Severe
(b) Moderate
(c) Mild

2. Shoulder level
   (a) Deformity of shoulder shrugging (incomplete shoulder descending, Sprengel shoulder) (Fig. 3.10)
   (b) Absence of pectoral muscles (including Poland syndrome) (Figs. 3.11 and 3.12): ① absence of major pectoral muscle; ② absence of major and minor pectoral muscles; ③ others

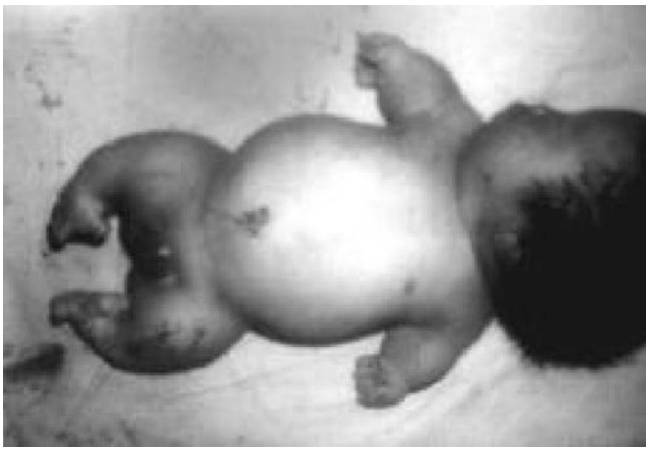

Fig. 3.8 Seal hand (foot)

3. Levels of the elbow and forearm
   (a) Hand long flexor deformity (Fig. 3.13)
   (b) Hand long extensor deformity
   (c) Hand internal muscle deformity
   (d) Others
4. Levels of wrist and hand
   (a) Syndactyly (complete, incomplete): ① radial type, the first finger web; ② central type, the second and third finger web; ③ ulnar type, the third finger web; ④ joint type, ① + ② or ③
   (b) Congenital finger flexion deformity: ① little finger (Fig. 3.14); ② otssher fingers
   (c) Palm center and thumb deformity
   (d) Non-osseous finger deviation deformity (arthrochalasis induced by muscles, ligaments, and joint capsule

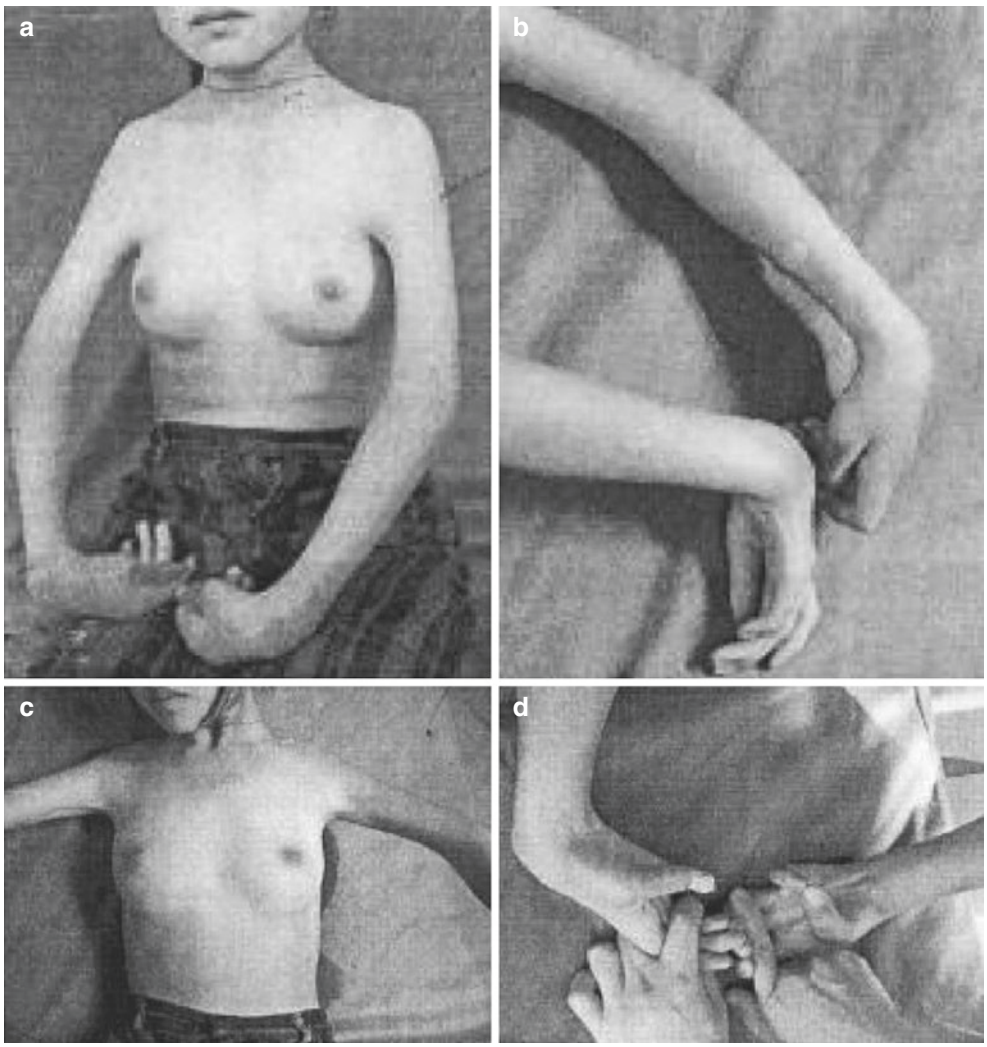

Fig. 3.9 Congenital multiple extensor dysplasia and multiple joint flexion deformity

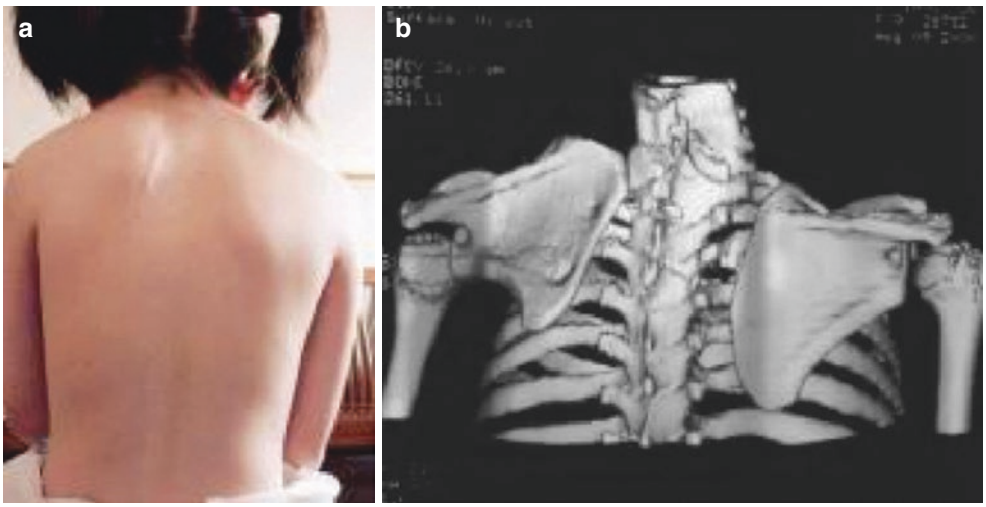

Fig. 3.10 Deformity of shoulder shrugging (incomplete shoulder descending). (**a**) Dorsolateral view. (**b**) 3-D image

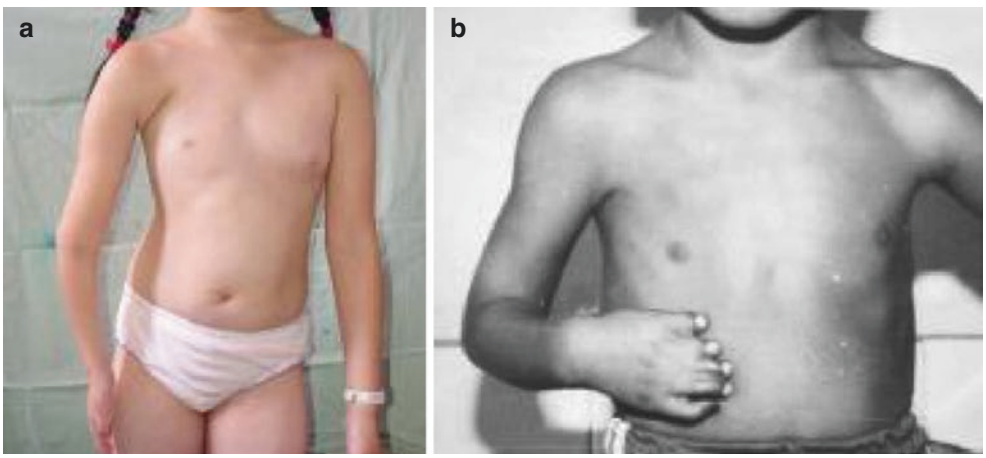

Fig. 3.11 Two pediatric female patients, absence of the right pectoral muscle with right micromelia, brachydactylia and syndactyly deformities (Poland syndrome), breast dysplasia

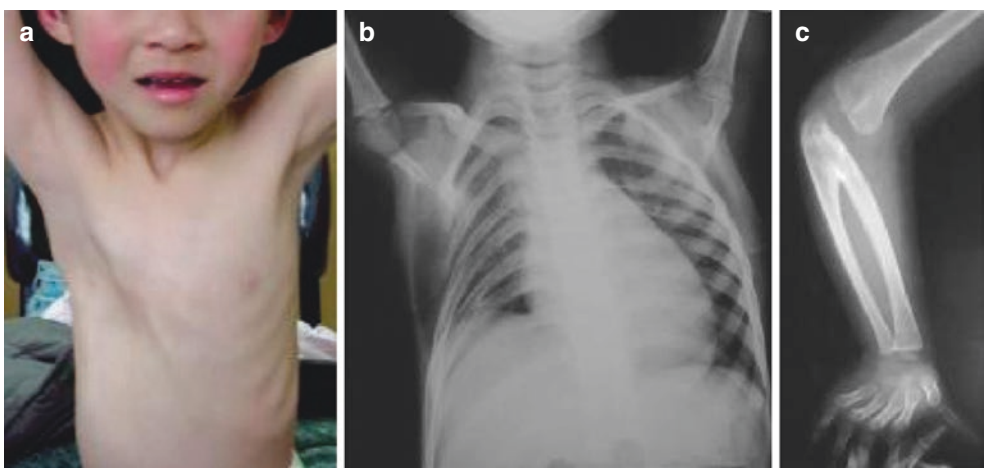

Fig. 3.12 A pediatric male patient, congenital dysplasia of right thorax, right thorax deformity, absence of right pectoral muscle, partial absence of the second to sixth ribs, accompanied by multiple bone joint deformities, radial-ulnar fusion, dysplasia of the carpal bone, and phalangeal deformities

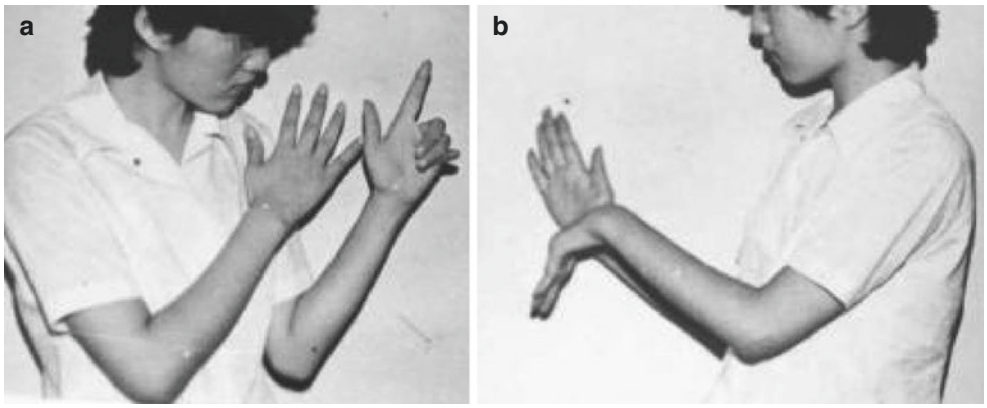

Fig. 3.13 Female patient, left hand long flexor deformity. (**a**) The middle, ring, and little fingers of the left hand cannot extend. (**b**) Only in case of flexion of left wrist joint, the middle, ring, and little fingers can extend

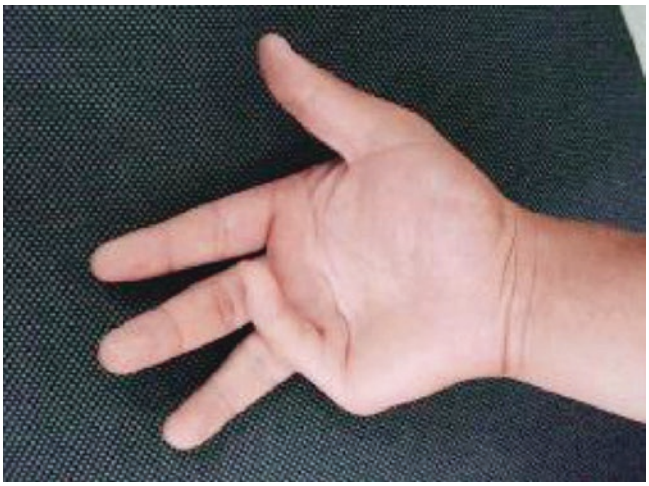

Fig. 3.14 Congenital little finger flexion deformity

dysplasia): ① independent finger deviation; ② ulnar deviation deformity of hand (windblown hand) (Fig. 3.15); ③ others
   (e) Congenital deafness trigger finger
   (f) Others
5. Skin and appendages
   (a) Wing-like axillary webbed elbow and webbed deformity (wing webbed syndrome)
   (b) Skin dysplasia
   (c) Drumstick finger deformity (Fig. 3.16)
   (d) Long fingernail deformity
   (e) Others

3.1.2.2 Involving Bones (Including Palmar Nail and Koilonychia Deformity)

1. Shoulder level
   (a) Congenital humerus varus
   (b) Others
2. Elbow joint fusion at the elbow level (Fig. 3.17)
   (a) Humeral and radial bone fusion
   (b) Humeral and ulnar bone fusion
   (c) Complete elbow bone fusion
3. Level of forearm
   (a) Proximal radial and ulnar joint bone fusion: ① presence of radial head dislocation; ② absence of radial head dislocation
   (b) Distal radial and ulnar bone fusion
4. Wrist and hand
   (a) Carpal bone fusion: ① lunate-triquetrum fusion; ② capitate-hamate fusion; ③ scaphoid-lunate fusion; ④ others
   (b) Metacarpal bone fusion: ① ring finger-little finger metacarpal bone fusion; ② others
   (c) Phalangeal bone fusion: ① radial type, the first and second finger bone fusion; ② central type, the second and third finger or the third and fourth finger bone fusion; ③ ulnar type, the fourth and fifth finger bone fusion; ④ spade hand (including Apert syndrome) (Fig. 3.18); ⑤ others
   (d) Interphalangeal joint bone fusion: ① distal interphalangeal joint bone fusion; ② others
   (e) Finger lateral flexion deformity (Fig. 3.19): ① little finger (including triangle phalanges); ② thumb (including triangle phalanges); ③ other fingers
   (f) Multi-phalanx malformations: ① three-segment phalanx thumb deformity (Fig. 3.20); ② others

3.1.2.3 Congenital Tumor-Induced Deformities

1. Vascular
   (a) Hemangioma
   (b) Vascular deformity: ① capillary deformity (including port wine stains); ② venous deformity; ③ venous lymphatic vascular deformity; ④ arterial deformity (including arteriovenous fistula); ⑤ lymphatic vascular deformity; ⑥ others
2. Neurogenic
   (a) Neurofibroma
   (b) Neuroblastoma
   (c) Others
3. Connective tissue derived
   (a) Juvenile aponeurotic fibroma
   (b) Others

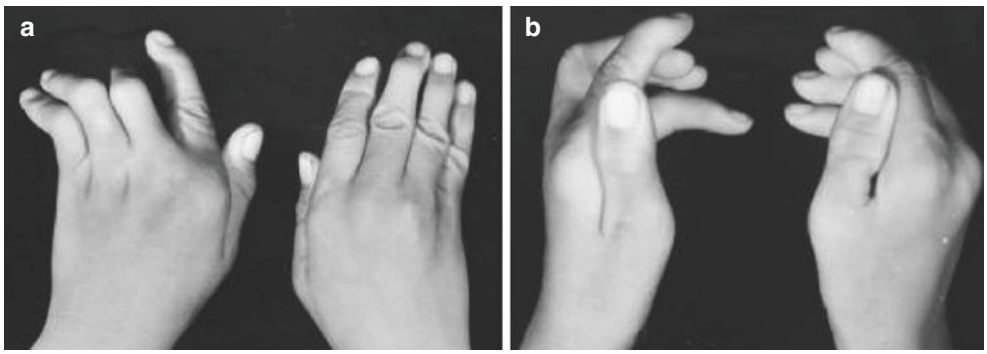

Fig. 3.15 Windblown hand deformity

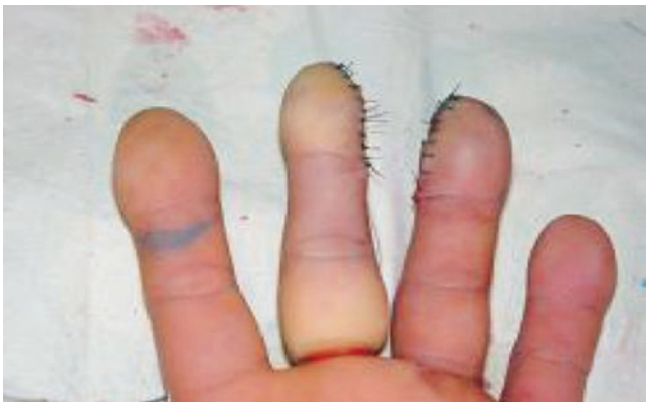

Fig. 3.16 Congenital drumstick finger deformity (plastic operation has been performed on the middle and ring fingers, and no operation is performed on index and little fingers)

4. Osseous (excluding overgrowth syndrome)
   (a) Osteochondromatosis (including multiple hereditary exostoses)
   (b) Enchondromatosis
   (c) Fibrous structural dysplasia
   (d) Epiphyseal abnormalities
   (e) Others

3.1.3 Type III: Twin Deformity

1. Whole body; neoplastic hand
2. Two humeral bones
3. Two radial bones
4. Two ulnar bones
   (a) Mirror hand deformity (Fig. 3.21)
   (b) Others
5. Polydactyly and polydactyly deformity
   (a) Radial ray (preaxial): thumb polydactylia, deformity of the duplication of the thumb
   (b) Central ray
   (c) Ulnar ray (postaxial): little finger polydactylia
   (d) Complexity: the simultaneous presence of any of the above two
6. Epiphyseal repetition (supernumerary epiphyseal deformity)
   (a) Thumb longitudinal ray
   (b) Index finger longitudinal ray
   (c) Others

3.1.4 Type IV: Overgrowth

1. Whole body
   (a) Overgrowth of half body (Fig. 3.22)
   (b) With vascular deformity
   (c) Others
2. Partial limbs
   (a) With vascular deformity
   (b) Others
3. Macrodactylia deformity (Figs. 3.23 and 3.24)
   (a) Accompanied by vascular deformity
   (b) Accompanied by neurofibroma
   (c) Accompanied by chondrophyte

3.1.5 Type V: Low Development

1. Whole body
2. Forearm
3. Hand dysplasia of hands
   (a) Dysplasia of whole hands
   (b) Dysplasia of partial hands
4. Metacarpal bone. Short and small metacarpal bone deformity
   (a) Short and small fifth metacarpal bone deformity
   (b) Others
5. Fingers. Short and small finger deformity
   (a) Brachydactyly and syndactyly: ① accompanied by absence of pectoral muscle (Poland syndrome) (Fig. 3.25); ② not accompanied by absence of pectoral muscle
   (b) Polydactyly: ① absence of middle phalanx; ② absence of two-segment or multi-segment phalanx; ③ absence of proximal or distal phalanx; ④ others

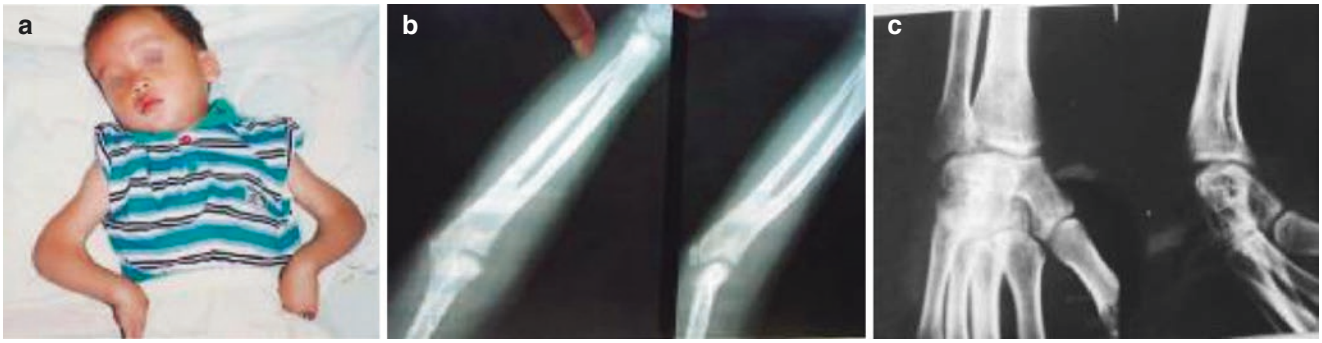

Fig. 3.17 A pediatric male patient, congenital elbow joint and wrist joint bone fusion, bilateral elbow joint and wrist joint flexion deformity. (**a**) Deformed appearance. (**b**, **c**) X-ray film indicates the proximal ulnoradial joint bone fusion and wrist joint bone fusion

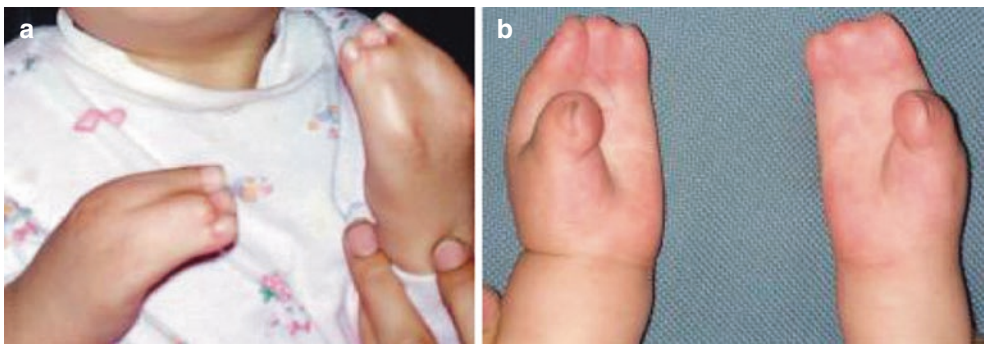

Fig. 3.18 Spade hand of Apert syndrome

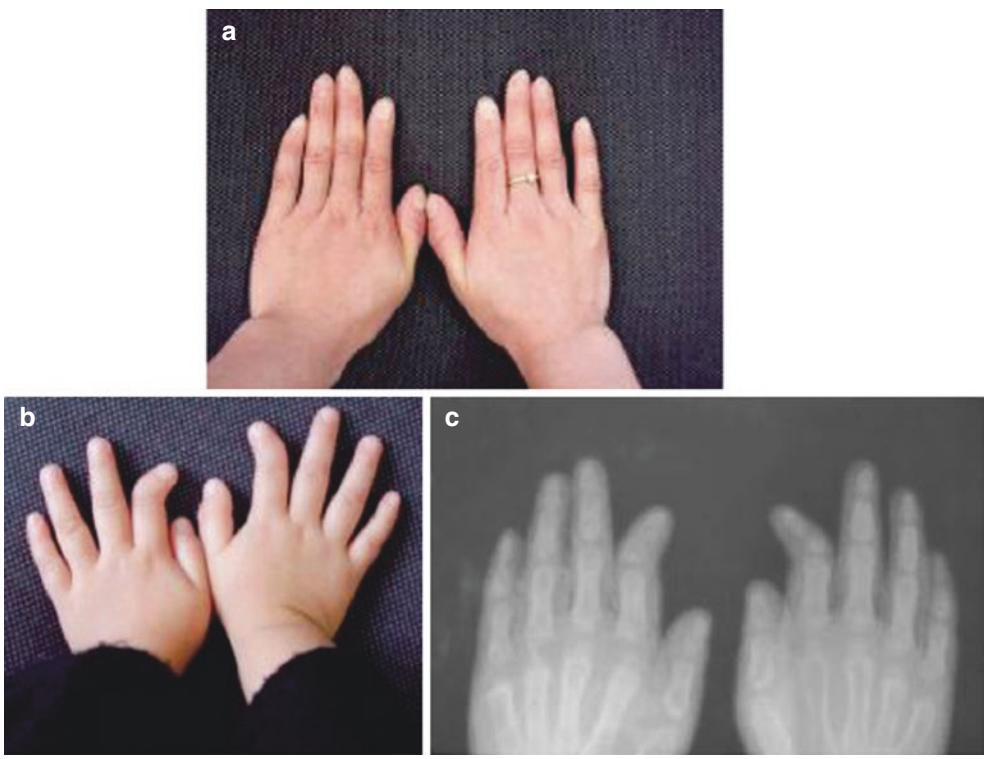

Fig. 3.19 Congenital finger lateroflexion deformity. (**a**) Index finger and little finger lateriflection deformity (*left*). (**b**, **c**) Index finger lateriflection deformity of both hands; X-ray shows the triangle phalanges of index finger. (**d**, **e**) Index finger lateriflection deformity of right hand, with deformities in bones, joints, muscles, fascia, and ligaments

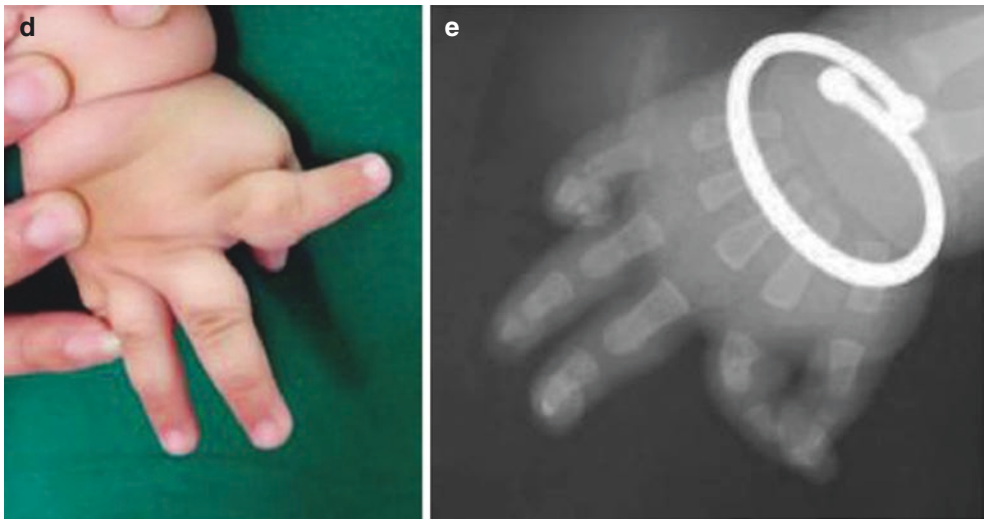

Fig. 3.19 (continued)

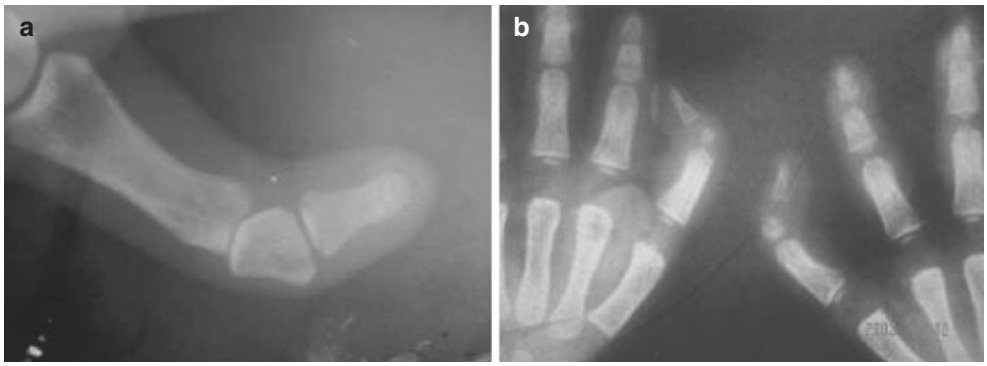

Fig. 3.20 Three-segment phalanx thumb deformity

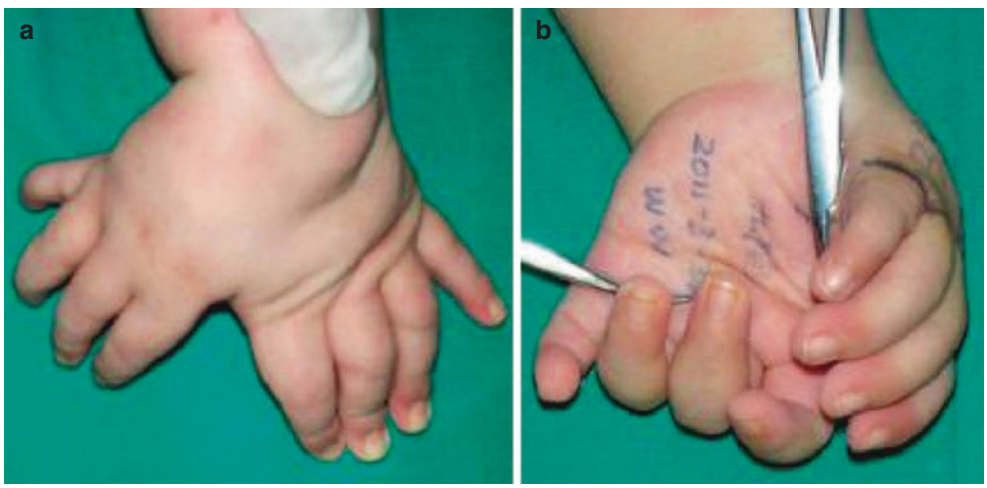

Fig. 3.21 Two ulnar bones. Mirror hand deformity

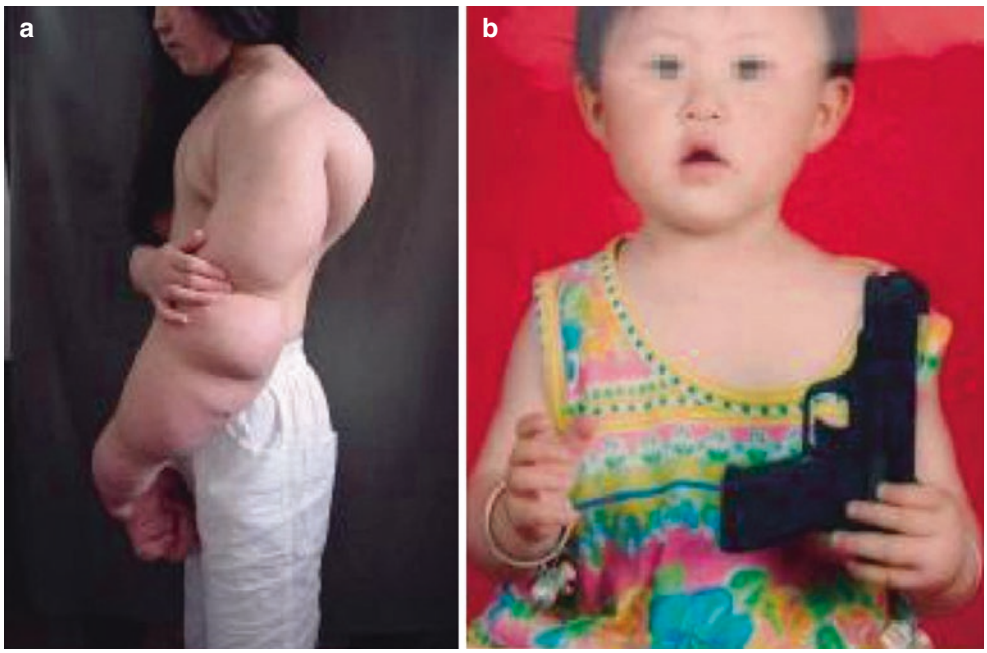

Fig 3.22 Overgrowth of half body. (**a**) A 6-year-old girl; the entire left upper limb, including shoulder, scapula, and inter-thoracic soft tissue, suffers disorderly growth. (**b**) Five years ago, when the patient was 1 years old, the difference between the left and right limb was not great

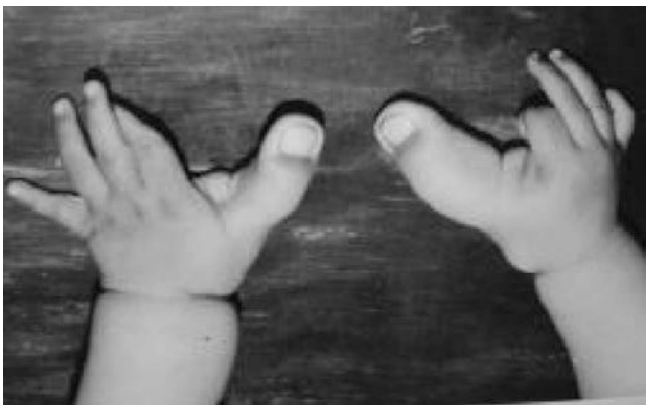

Fig. 3.23 A 1-year-old boy, macrodactylia deformity of both thumbs

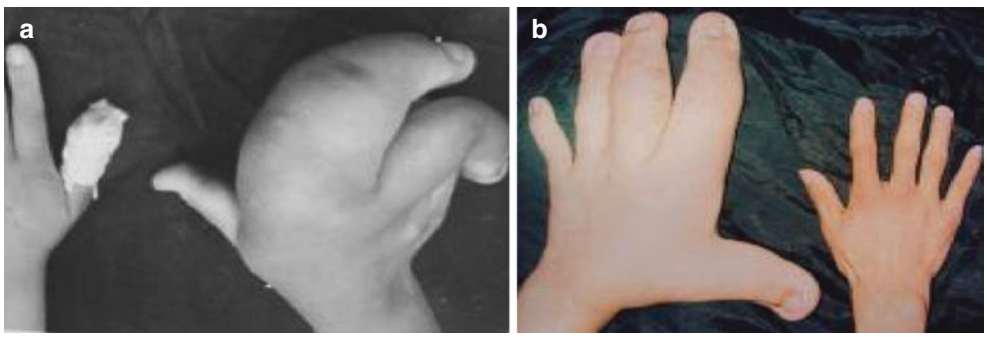

Fig. 3.24 Deformity of overgrowing macrodactylia. (**a**) Index finger and middle finger macrodactylia deformity of right hand. (**b**) Thumb, index finger, middle finger, and ring finger macrodactylia deformity of left hand

3.1.6 Type VI: Ring Constriction Syndrome

1. Focal necrosis
   (a) Constrictive band (partial or a week): ① with lymphedema; ② no lymphedema
   (b) Syndactylia at the fingertip
2. Intrauterine amputation (finger) (Fig. 3.26): ① wrist; ② palm; ③ phalanx; ④ joint type, ① + ② or ② + ③; ⑤ others

3.1.7 Type VII: Systemic Skeletal Deformities and Syndromes

1. Holt-Oram syndrome
2. Whistling face syndrome
3. Cranio-facial-corporal syndrome
4. Oculo-dento-digital syndrome
5. Oro-facial-digital syndrome
6. Aarskog syndrome

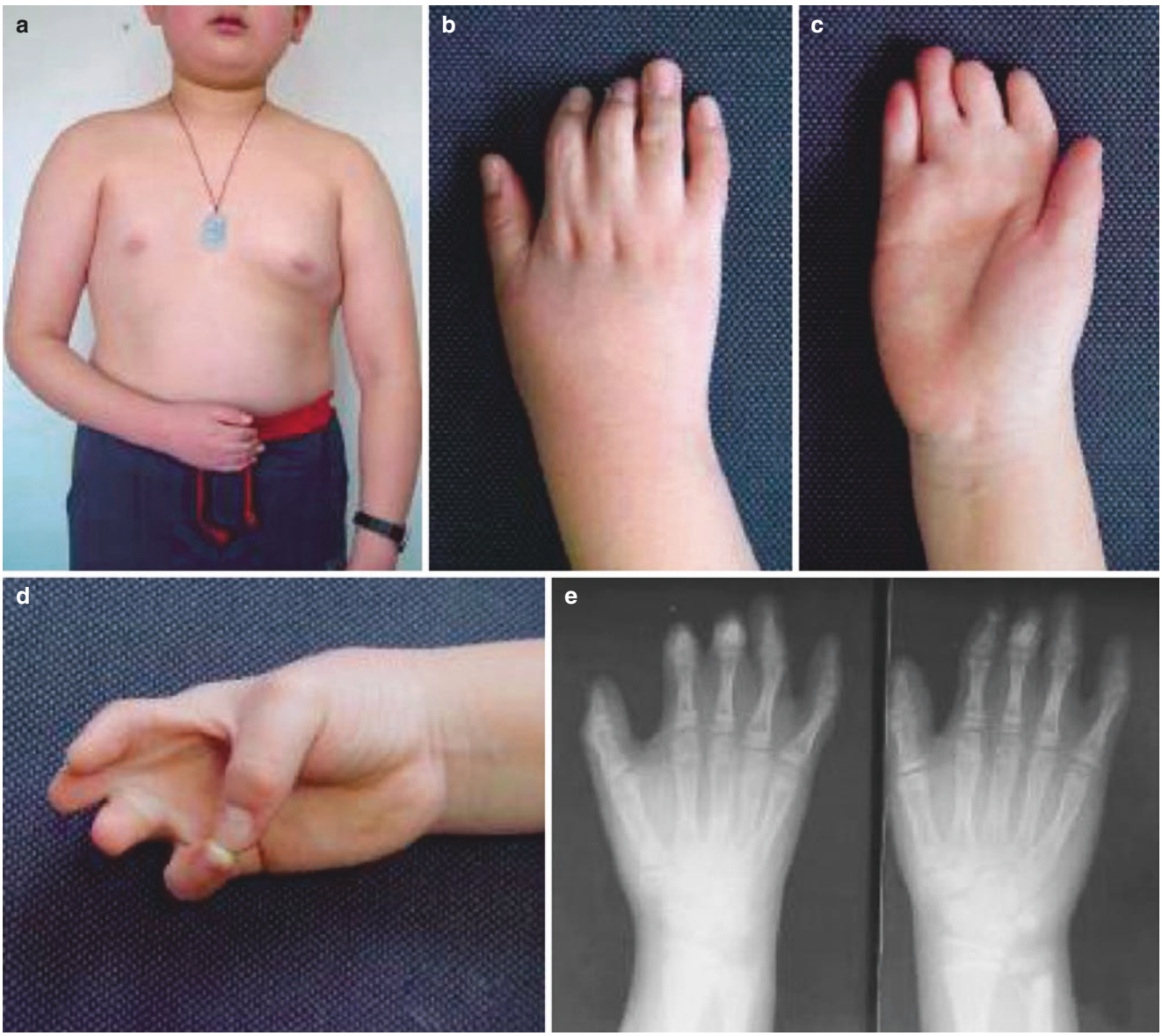

Fig. 3.25 Brachydactyly and syndactyly accompanied by absence of pectoral muscle (Poland syndrome). (**a**) Physical appearance. (**b–d**) Hand appearance. (**e**) Hand X-ray findings

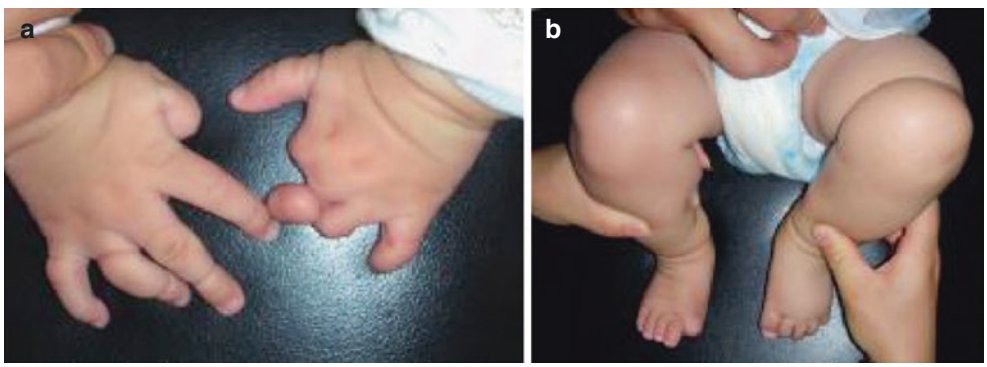

Fig. 3.26 Intrauterine amputation (finger). (**a**) Ring constriction of two hands accompanied by left hand amputation. (**b**) Ring constriction of the left thigh

7. Teebi-Shaltout syndrome
8. Robinow syndrome
9. Coloboma of the eye, heart defects, atresia of the choanae, retardation of growth, genital abnormalities, ear abnormalities syndrome
10. Gordon syndrome
11. Ectrodactyly-ectodermal dysplasia-cleft lip/palate syndrome, EEC syndrome
12. Shprintzen-Goldberg syndrome
13. Fryns syndrome
14. Klippel-Trenaunay syndrome

Swanson's classification system of congenital deformities of hands and upper limbs (IFSSH classification) is a practical classification method at present, but it is not perfect. In 1997, De Smet L. et al. [4] analyzed 925 hands of 650 patients with congenital upper limb deformities, involving 1013 hand deformities, among which 26.7% also had other deformities. Among the above deformities, those that could be included for classification accounted for 86%, those that could barely be classified accounted for 6.6%, and those that could not be classified accounted for 7.8%. Nonetheless, IFSSH classification is still the classification method most recommended by plastic surgeons, hand surgeons, and orthopedic surgeons today.

3.2 Developmental Biology and Classification of Congenital Deformities of Hands and Upper Limbs

Bin Wang and Feng Ni

One of every about 600 newborns suffers upper limb deformities [5]; therefore, the study on the molecular mechanism coordinating four limb development has been the research focus during the several decades. Recent studies have found that there is a correlation between the discontinuation of different molecular pathways and the upper limb deformities.

Utilization of one accurate method to describe and classify these deformities not only facilitates clinical diagnosis and treatment but also benefits the discussion and exchange by clinicians and researchers on the classification protocol and deformity occurrence and development. Currently, the commonest classification method is based on the protocol proposed by Swanson in 1964 [1], which was adopted by IFSSH in 1974 [2]. The classification method combines the morphological knowledge of four limb development and the clinical surgical viewpoints, and it is characterized by easy use. It is almost adopted by hand surgeons all over the world. However, the classification of the etiological factors in this protocol includes the descriptive and predictive contents, and its simplicity can also make it impossible to classify some development-related deformities and is apt to lead to misdiagnosis or multiple diagnoses. With the constant deepening in the understanding of development biology, these descriptive terms can no longer accurately summarize the features of diseases; therefore, this classification has been subject to objections [6–11].

Manske and Oberg [8] have recently modified the original classification method on the basis of clinical experience and developmental biology. The article aims to outline developmental biology, propose the latest concept in pathological genetics of upper limb deformities to facilitate the modification of deformity classification method, and make it more accurate and applicable.

3.2.1 Limb Development: Formation and Differentiation of Body Axes

During the early stage of embryonic development, homeobox (HOX) transcription factor mediates the cranial rear axis to initiate the differentiation of body segments. About 4 weeks after the initiation of embryonic development, the germinal area of upper limb is established; the expression of T-box (TBX5), wingless-type MMTV (WNT), and fibroblast growth factor (FGF) is initiated; the limb growth starts; and the upper limb bud covered by the thin ectodermal surface layer bulges from the lateral plates (refer to Fig. 1.1). Limb bud induction failure (tetra-amelia symptom, absence of four limbs) is correlated with WNT3 and FGF10 [12, 13], TBX4 and TBX5 are respectively related to the development of lower limb and upper limb, and the TBX5 mutation (e.g., Holt-Oram syndrome and hand-heart deformity syndrome [14]) will lead to a series of upper limb deformities.

With the formation of limb buds, the development proceeds at three axial directions: proximal-distal, anterior-posterior (radial-ulnar), and dorsal-ventral; the development and differentiation of each axial direction are controlled by a group of cells; they will send the development information to the local cells and tissues, and we call this group of cells the information center. Mesodermal FGF10 and ectoblast radical fringe gene (R-FNG) are linked at the top of dorsal and ventral border to make the ectoderm become thickened to form the information center of proximal-distal axis, which is called apical ectodermal ridge (AER) [15–17]. AER can produce WNT3 and some FGFs (FGF 4, 8, 9, and 17) to maintain the expression of mesodermal FGF10; FGF10 can promote the cell proliferation in AER lower areas, which are called progress zone [18]. The mesodermal cells in the progress center are subject to the regulation of information center to decide the final evolution. The interactions between ectodermic and mesodermal FGF and WNT maintain the development and growth of proximal-distal axis [19, 20].

The development and differentiation of front-rear (radial-ulnar) axis is controlled by the mesodermal posterior zone of polarizing activity (ZPA). ZPA increases the width of limbs and makes them develop toward the rear (ulnar) direction, and it produces effects through generating morphogen sonic hedgehog (SHH). AER and ZPA are closely associated through the feedback loops and maintain the expression of SHH at the border area of AER distal rear (ulnar) direction during the growth process (refer to Fig. 1.2) [21–23].

The WNT7a generated at the dorsal side of ectoderm regulates the development of limb dorsal-ventral axis. WNT7a [17, 24, 25] makes the mesoderm of the lower layer limb grow toward the dorsal side through inducing the Lim homeobox transcription factor LMX1B, so the deficiency of WNT7a will lead to the limb ulnar growth and development disorder, indicating that another important role of WNT7a is to maintain the production of ZPA-related SHH. It can be said SHH [26] plays an important role in limb development and is associated with the development of the proximal-distal axis, front-rear (radial-ulnar) axis, and the dorsal-ventral axis [27].

The information center can also regulate the occurrence of downstream target tissues such as bones, vessels, muscles, and nerves through conventional, special, and asymmetric molecular pathways. For example, skeletal occurrence needs several factors to play the regulating roles at the proper timing and sites, including high-mobility group protein 9 (SOX9) correlated with the sex-determining region (Y), which makes the bone primordia concentrate; WNTs and growth differentiation factor 5 (GDF5) regulate joint development; parathyroid hormone-like hormone (PTHLH), Indian hedgehog (IHH), insulin-like growth factor (IGF), bone morphogenic protein (BMP), WNTs, FGF, and runt-related transcription factor 2 (RUNX2) promote bone primordia growth and subsequent endochondral ossification. In addition [28], upregulated expression of short stature homeobox-containing gene 2 (SHOX2) is found at the proximal cartilaginous membrane, promoting the extension of humerus; in the meantime, the forearm cartilaginous membrane induces SHOX to regulate the growth of radio-ulna. In addition, the correction induction of downstream pathway is of vital importance to the complete differentiation of all axial directions.

3.2.1.1 Defects of Proximal-Distal Axis (Transverse Defect)

The FGF produced by AER promotes the growth of ZPA-related limbs. The thorough removal of AER [31] or blockage of FGF receptor [32, 33] will at the same time discontinue the development of limbs at the proximal-distal axis, and the clinical manifestation is blockage of transverse growth. Animal experiments indicate that the blockage degree of transverse growth is correlated with the time of destruction of this axis [34].

Recently, Winkel et al. [35] have found that WNT in brachydactylia B1 (BDB1) and tyrosine-protein kinase receptor (ROR2) often experience mutation. Under the regulation of AER-related FGF, WNTs can promote the extension of the entire limbs through ROR2.Therefore, AER functional disorder often leads to brachydactylia, which also falls into the category of developmental disorder of proximal-distal axis.

Limb absence or phocomelia is generally not purely development problem, and the majority of them are often accompanied by severe longitudinal or radial-ulnar axis defects. However, recent studies on the mechanism and potential of SHOX2 and SHOX during the growth of proximal-distal axis have indicated that they may be correlated with the rare deformities of limb absence [29, 30, 36].

3.2.1.2 Radial-Ulnar Axis Defect (Longitudinal Defect)

AER-related FGF function loss often results in transverse absence, and lack of FGF function may result in longitudinal absence. FGF hypofunction can induce slowing in limb growth and diminution of shape. Although the ulnar growth and proliferation under the action of ZPA is still ongoing, the development results are manifested as the radial longitudinal ray absence in the classification of the same deformities. Malformation syndromes of FGF receptor 2 mutations [37, 38], such as Apert syndrome, Pfeiffer syndrome, and Saethre-Chotzen syndrome, are characterized by the radial (front) joint abnormality and the formation of forearm synostosis [39].

SHH induces the formation of upper limb ulna and hand ulnar phalanx; in addition, SHH is correlated with the growth of posterior (ulnar) limb. During limb development, decrease in SHH expression or discontinuation of target signals can slow down the growth of limbs and diminish their size. The results of SHH absence [40, 41] are manifested as ulnar longitudinal ray absence, and the clinical manifestations vary with time points, degree, and duration of SHH absence. Additionally, SHH absence can reflectively induce decrease in FGF expression; therefore, in addition to decrease in limb length, size, and FGF expression, radial structure, especially thumb development, is potentially subject to influence, and the clinical manifestation is thumb and ulnar ray absence.

In the past, the longitudinal absence of several kinds of proximal limb absence falls into the category of segmental absence. After many patients are carefully examined, the defect at the distal side is consistent with that of the radial ray or ulnar ray, indicating that the distal deformity is the extension of the longitudinal ray deformity [42, 43].

3.2.1.3 Dorsal and Ventral Defects (Dorsal Defects)

The decrease or absence of WNT7a at the dorsal side of ectoderm or LMX1B at the dorsal side of mesoderm can affect dorsal development [44, 45] [dorsal side refers to the

posterior side (extending side) of the upper limb and the anterior side (extending side) of the lower limb before and after limb rotation at the embryonic phase]. In rat models, the single dose is insufficient to produce phenotypes; in humans, the defect of LMX1B single allelomorphic gene can induce dorsal hypoplasia, such as nail-patella syndrome, namely, development abnormality of the elbow and fingernails [45].

3.2.2 Formation and Differentiation of Hand Plates

At 5 weeks of embryonic development, the hand plates are visible. The interaction between HOX transcription factor (especially HOXD9~13 and HOXA9~13) determines the number and category of fingers. SHH [46, 48] can also induce the generation of BMP gradient from the rear to the front (ulnar side to radial side), which plays at least two roles in the development and differentiation of fingers: first, BMP can induce interdigital apoptosis or programmed cell death, which can be partially achieved by inhibiting the FGF expression of AER; second, BMP [49–51] participates in finger differentiation through the phalanx formation area, which, located at the distal bone primordia of fingers, regulates the chondrification through the upregulated expression of SOX9, maintains the FGF expression, and promotes the continuous growth of fingers. However, currently, it is still unclear how BMP family members (such as BMP2 to 7 and GDG5 and 6) periodically shift the functions during apoptosis and chondrosis (Fig. 3.27) [52].

3.2.2.1 Interdigital Area Formation and Differentiation Defect (Soft Tissue Defect)

Web can be found in the interdigits of some animals, such as ducks and bats. In some animal models [50, 53], BMP antagonist Gremlin (GREM) is expressed in the interdigital area, further limiting cell apoptosis. The ectopia or overexpression of BMP antagonist in the interdigital area [52] and the continuation of FGF function of AER [54] or mesoderm inhibit the cell apoptosis of interdigital area and lead to aschistodactylia. The mutation of BMP antagonist Noggin (NOG) is associated with joint bone fusion, syndactylia, and hyperdactylia, indicating the role of BMP in it.

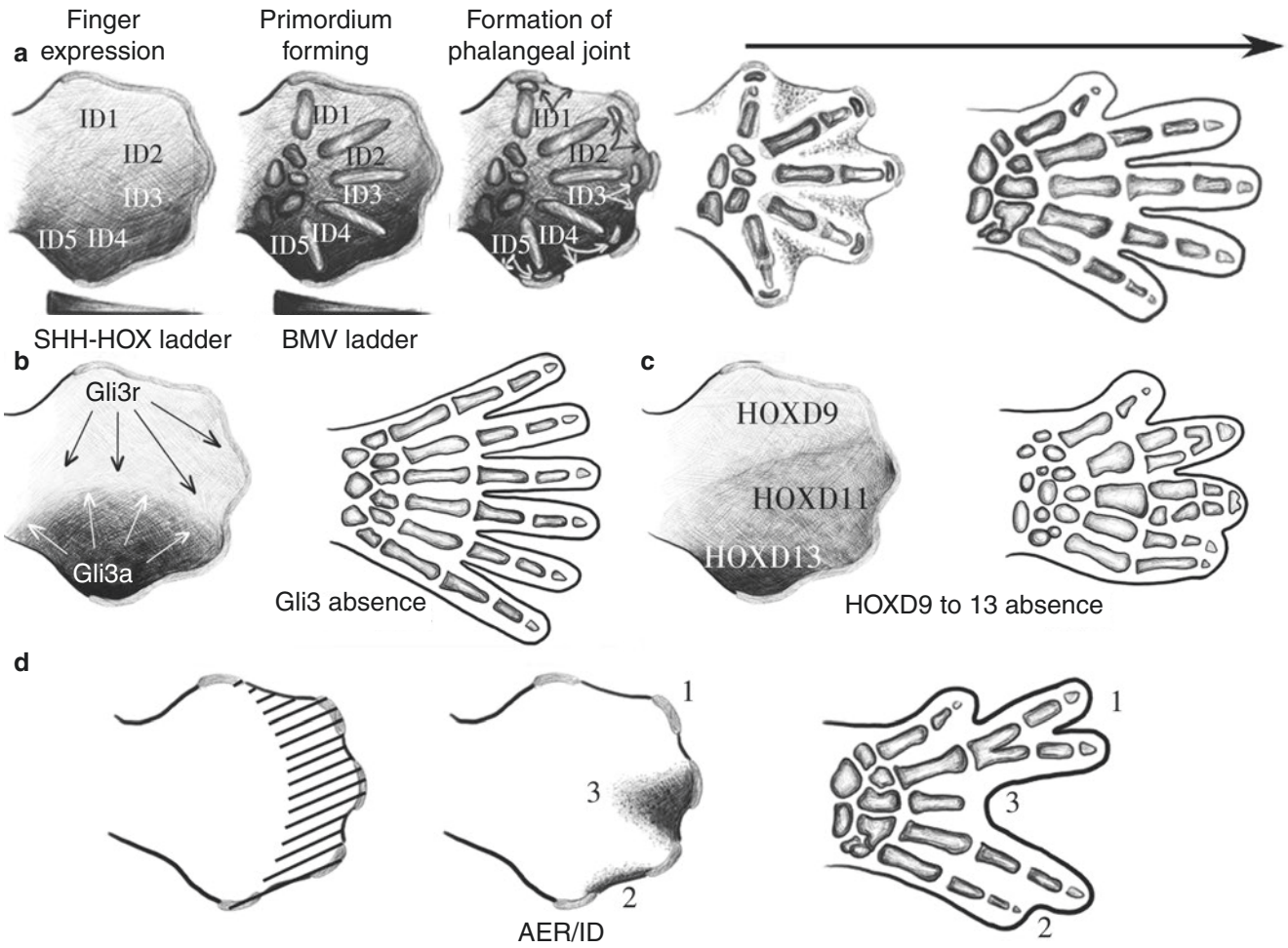

Fig. 3.27 Formation of hand plates and differentiation disorder

The syndactylia of Apert syndrome is induced by the mutation in FGF receptor 2 (FGFR2), which leads to increased activity of binding of FGF and glycosaminoglycans with the receptors [39, 47], making the body under the state of continuous activation. The constant effect of FGF functions covers the signal mechanism of BMP and further causes complicated aschistodactylia.

The formation and connection mechanisms of ligaments or tendons of the dorsal and ventral side and all fingers still have not been well explained. Candidate gene may be correlated with flexion contracture deformity. For example, the camptodactyly may be derived from the interactions of nerve muscles and (or) extracellular matrix structures (e.g., PRG4). Profound studies still need to be done on the etiological basis of relevant soft tissue deformities of these hands.

3.2.2.2 Finger Formation and Differentiation Defect (Bone Defect)

As described above, deficiency of SHH will induce the absence of ulnar fingers and the defect of radial-ulnar axis; however, the abnormality in SHH signal system will also lead to polydactylism. In the transmission of SHH signals, the transcription factor Gli3 reflects the role of SHH gradient on the radial-ulnar axis. On the radial side of limb bud, Gli3, after transcription, is expressed into the form of short-acting inhibitor, which inhibits the SHH function. In ZPA zone, SHH blocks the conversion of Gli3 and becomes the full-range form of activator. The mutation of such dual functional transcription factors [55–59] can induce the combined hyperdactylia of ulnar hyperdactylia (also called postaxial hyperdactylia type A and type B [56, 57]) and radial hyperdactylia (also called preaxial hyperdactylia type IV). In rat models, the complete absence of Gli3 makes the characteristics of absence of each toe similar to the deformity of five-fingered hand or the deformity of triphalangeal thumb in humans. In addition, the variation [59] in limb SHH regulatory region may lead to abnormal expression at the front side (radial side) and can also lead to deformity of triphalangeal thumb [60, 61].

HOXD13 defect [62] or the absence of the entire HOXD9 ~ 13 regions may lead to various deformities of syndactylia and polydactyly, and disorder may occur to the finger and interdigital structure and morphology. The absence of HOXA13 functions is associated with the hand-foot-genital syndrome, with the manifestation of middle phalanx and finger deviation of hypoplasia [63, 64].

The formation, segmentation, and cartilage development of fingers involve a number of factors and channels, so their mutation or defect may cause brachydactylia. HOXD13 [65] variation may lead to type D or type E1 brachydactylia. As for the factors in cartilage development, including IHH (A1 type), BMPR1B (A1 type and A2 type), GDF5 (A2 type and C type), ROR2 (B1 type), NOG (B2 type), and PTHLH (E2 type), their variation can also lead to brachydactylia.

In the process of development and differentiation of phalanx, BMP regulates the activation of AER upper SOX9 and maintains the transition between FGFs. The complete absence of SOX9 [66] can cause limb degeneration, and it cannot induce cartilage formation. The haploinsufficiency of SOX9 can result in bone flexion dysplasia, manifested as cartilage reduction, bow shape of long bones, and brachydactylia.

Ogino [67] used the alkyl agent busulfan and established the models of central absence (cleft hand). During the formation of hand plates, he observed that the teratogenic factor made cell apoptosis in the distal progression area and AER increase, leading to central fissure, central syndactylia, and central hyperdactylia. The increase in hand plate cell apoptosis can also lead to the AER-related FGF expression deficiency, making the BMP secretion of AER and the mesoderm below it decrease and be in disorder and also making the ZPA-related SHH expression decrease. These constitute [68] the general mechanism of central fissure, syndactylia, and hyperdactylia during the formation of hand plates in spite of the fact that the molecular mechanism is very complicated.

3.2.3 Relation Between Hand Developmental Biology and Classification

The characteristics of hand developmental biology are inconsistent with the IFSSH/Swanson classification method. The molecular channel regulates the unique asymmetric development process of upper limbs, including proximal-distal axis, anterior-posterior axis (radial-ulnar axis), and dorsal-ventral axis. The destruction of such molecular pathways will affect the development of the entire body in each axial direction. According to the IFSSH/Swanson classification method, IFSSH type 1 is the formation disorder type, which mainly affects the relevancy of the proximal body axis and may extend to the hand deformities. Correspondingly, it is very reasonable to classify it into the types that can reflect the etiology and the overall influence. We call this classification the axial formation (differentiation) defect, complete upper limb type, and the subcategory is determined based on the specific axis.

IFSSH type 2 is the differentiation disorder type, which mainly describes the defect in structures and differentiation of hand plates, and it is not purely a hand plate differentiation defect. The polydactylism that falls into the category of the IFSSH type 3 also involves the anterior-posterior (radial-ulnar) axis formation and differentiation disorder; therefore, it is not logic to exclude hyperdactylia from type 1 and type 2. These deformities induced by hand plate defects can be subdivided according to axial formation and (or) differentiation disorder. For example, polydactylism is generally determined as anterior-posterior (radial-ulnar) axis deformation. Also it can be classified into subcategories according to hand plate formation and (or) differentiation disorder, such as aschistodactylia. In addition, none of the IFSS grouping

can properly classify cleft hand, which has various deformities; therefore, Japanese Society for Surgery of the Hand has added one group in the IFSSH/Swanson classification—digit ray formation disorder [3].

IFSSH type 4 is the overgrowth type, a descriptive term which does not include nosogenesis. Likewise, IFSSH type 5 is the growth failure type without etiologic information and characteristics, and it seems arbitrary if they are classified. Growth failure (e.g., brachydactylia, symbrachydactyly), like transverse defect or longitudinal defect, also involves the disturbance in molecular channel, and this type should be excluded from this type.

IFSSH type 7 is generalized skeletal deformity and syndrome, often including upper limb deformity and various skeletal abnormalities, and it is generally a genetic syndrome. When compared with description of deformity shape, more detailed classifications are less significant [6–11].

With constantly deepening understanding in limb morphogenesis and deformity production, we find some problems in IFSSH classification, which will spur surgeons, pathologists, and geneticists to make joint efforts in the field of congenital deformity of limbs and question this classification method.

Teratology provides a framework for the etiological study of congenital deformities of limbs. Deformities refer to the abnormal formation of some body part or the complex tissues; deformations, different from deformities, are induced by injuries after normal formation of tissues. Dysplasia refers to abnormalities in size, shape, and components of cells in tissues. Teratology also describes phase 4, namely, disruption. Because there are changes in the deformed tissues during this process, it is classified into the category of deformation in order to complete the classification.

Although the nosogenesis of some special deformities still remains unclear, it is reasonable to classify congenital upper limb deformities into deformity, deformation, and dysplasia. Therefore, in order to combine IFSSH classification and our understanding of limb development at the molecular level, we propose the following categories—deformity, deformation, and dysplasia. Then deformities are subclassified according to the main axis of the defects and the site of the defects (the entire limb or the hand). The following classification methods are made in combination with these principles.

3.2.3.1 Deformity

1. Axis deformation or differentiation disorder—the entire upper limb. In order to more accurately reflect the nosogenesis of multiple deformities in IFSSH type 1, we classify them into three subcategories according to axial defect. We will include the brachydactylia (symbrachydactyly and brachydactylia type B1) and transverse defects, segments, and defects that affect the entire limbs into the category of proximal-distal axial defect. The radial-ulnar defect not only includes the radial and ulnar longitudinal defects but also includes the repeated deformities of radial and ulnar structure, such as the ulnar dimelia and radial bone fusion (previously fall into the category of the twin deformity type in the IFSSH type 3 and the differentiation disorder in the IFSSH type 2). We also add the type of dorsal-ventral axial defect, such as nail-patella syndrome.
2. Axial formation or differentiation disorder—hand plates. The axial defects limited to the hand plates are classified into the second subcategory. In the past, polydactylism was classified into the range of twin deformity; however, it is typical axial signal channel disorder. For example, the functional disorder of transcription factor Gli3 in the radial-ulnar axis will lead to the radial (preaxial) supernumerary fingers (type IV or synpolydactyly [69]) and ulnar (postaxial) supernumerary fingers (A1 type) [58].

   We also include triphalangeal thumb into this category. Recently the genetic study on the radial polydactylism (preaxial hyperdactylia type II) has found that point mutation or paired mutation is present in SHH accommodation region. In the animal models, similar point mutation will lead to radial SHH ectopic expression and preaxial supernumerary fingers [70–75].

   The defect of dorsal-ventral axis can also be limited to hand plates, such as dorsal dimelia, so it also falls into this subcategory.
3. Hand plate formation and differentiation defect—nonspecific axis. This deformity is included into type 2 in the IFSSH classification, namely, the type of differentiation disorder, which mainly refers to hand plate development defect but is not unique. We include the deformities limited to the hand plates but without axial defects into this category, such as the deformities correlated with the molecular channels that regulate finger web formation and phalanx differentiation. We can also include the deformities that might involve multiple pathways into this category, such as synpolydactyly and cleft hand. Ogino and his colleagues demonstrated that injuries to the hand plates during development would lead to syndactylia, central symphalangia, and cleft hand, indicating that there are some connections between the factors, but one of them is not necessarily connected with others [67, 68, 76].

3.2.3.2 Deformation

According to the nomenclature of teratology, we have determined the second major category and defined the deformation and destruction of each limb sites that have taken shape. Spastic constriction ring is also included in this category, and it is one manifestation of the syndrome and can be correlated with amniotic band. Arthrogryposis or congenital contracture can occur alone or involve multiple joints, and the etiological factors can include nervous, muscular, or connective tissue factors. Arthrogryposis [77, 78] is also included into this

category because the formation time of contracture is about the second trimester of pregnancy, namely, after osteoarticular development and formation. Trigger finger [79] also falls into this category, and its incidence in childhood is higher than that of the fetal period. The deformation or destruction induced by virus infection, vascular injury, or mechanical stimulation does not follow a fixed pattern. But in order to facilitate classification, we include it into subcategory D—nothing special.

3.2.3.3 Dysplasia

This category includes special deformities correlated with the appearance, cell heterotype, or tumor; for example, hypertrophy of limbs is often correlated with oncogenesis, and macrodactylia is currently still correlated with still unclear cell dysplasia. In the past, these deformities were classified as IFSSH type 2—differentiation disorder (tumor factor)—or classified as IFSSH type 4 as the descriptive term, overgrowth. There is a controversy on whether these are deformities or deformations, but further deep study on development biology will help us to understand the essentials of deformity and deformation.

A repeatable and consistent nomenclature is needed for congenital deformities of hands and upper limbs, and only a universal language all over the world can allow discussion of complicated clinical examples, selection of treatment indications, and comparison of efficacy. As wse get a basic understanding of the molecular theory of morphogenesis and deformity generation, classification will be made more detailed. This classification method may need to be modified in the future, but we believe that teratological nomenclature will provide a suitable framework to satisfy future demand (Table 3.1).

Table 3.1 Comparison between IFSSH classification and new classification

IFSSH classification	New classification
1. Formation disorder (a) Transverse loss (b) Longitudinal loss • Radial longitudinal ray defect • Ulnar longitudinal ray defect • Central longitudinal ray defect (c) Segment defects 2. Dysdifferentiation (a) Soft tissue absence (b) Bone absence 3. Twin deformity (a) Neoplastic hand (b) Two humeral bones (c) Two radial bones (d) Two ulnar bones (mirror hand deformity) (e) Polydactylism • Radial polydactylia • Ulnar polydactylia 4. Overgrowth 5. Poor development (dysplasia) 6. Congenital constriction band syndrome 7. Systemic skeletal deformities and syndromes	1. Deformities (a) Axial deformation or dysdifferentiation—the entire upper limbs • Proximal-distal axis defect Anomaly of brachydactylia Transverse loss Segmental loss • Radial-ulnar axis defect Radial longitudinal ray defect Ulnar longitudinal ray defect Ulnar dimelia Radial bone fusion Humeral-ulnar bone fusion • Dorsal-ventral axial defect Nail-patella syndrome (b) Axial deformation or dysdifferentiation—hand plates • Radial-ulnar axis defect Radial polydactylia Ulnar polydactylia Triphalangeal thumb • Dorsal-ventral axial defect Dorsal dorsum (volar finger nail) Insufficient hypoplasia (3) Formation of hand plates and differentiation—nonspecific axis • Soft tissue abnormalities Syndactylia Phalange flexion • Bone abnormality Brachydactylia Oblique finger Kirner's deformity Volar carpal bone fusion • Composite type Cleft hand Hyperdactylia and syndactylia Apert hand 2. Deformity (a) Ring constriction (b) Joint contracture (c) Trigger finger (d) Others 3. Dysplasia (a) Megalomelia (macrodactylia) (b) Tumor

3.3 Selection of Surgery Timing of Congenital Deformities of Hands and Upper Limbs

Wei Wang

3.3.1 Principles for Selection of Surgery Timing

When is the best time to do a surgery for congenital deformities in hand and upper extremity is a problem that often puzzles domestic and foreign scholars. Early surgical correction of deformities is an important principle that benefits the patient's physical and mental development and the parents' physical and mental health. However, during early surgery, consideration should be given not only to the possibility and safety of surgical techniques and the possibility of obtaining the optimal efficacy and prediction of long-term efficacy but also to the influence of the development of the patient's immune system and other important organs on surgery. Based on the above various influencing factors, the author [80] believes that the following principles should be followed when the surgery time of congenital deformities of hands and upper limbs is selected.

3.3.1.1 Anaplasty Is Carried Out During the Infantile Period

Attempts should be made to carry out anaplasty on the congenital deformities of hands and upper limbs during the infantile period, which facilitates the development and reconstruction of anatomical structures and functions of repaired hands and upper limbs, is good for the patient's psychological development and the parents' physical and mental health, and helps reduce the residue of the postoperative local scars.

3.3.1.2 The Safety of Surgery and Anesthesia Should Be Guaranteed

The children with congenital deformities of hands and upper limbs often concomitantly suffer from cardiovascular, digestive tract, urinary system, and respiratory system deformities, which will affect the safety of anesthesia and surgery, so the deformities of the main organs should be first corrected when the surgery is carried out during the infantile period. On the other hand, during the infantile period, the surgery of multiple deformities of hands and upper limbs should be carried out in different stages to guarantee safety and achieve the efficacy of good shape and functions.

3.3.1.3 Decide Whether to Perform Surgery According to the Actual Circumstances

During the infantile period, the structure of hand and upper limb tissues is fine and small, often making it difficult to carry out deformity repairing surgery; therefore, surgeons should decide whether to carry out the surgery during the infantile period according to the hospital equipment conditions and their operative skills. Currently, with the help of precise hand surgery and microsurgery instrument and devices, plastic surgery can be performed on any fine tissue structures. Especially among the numerous domestic plastic surgeons and hand surgeons, most of them have received training in microsurgery, which obviously makes it easy for them to carry out surgeries during the infantile period.

3.3.1.4 Weigh the Advantages and Disadvantages to Decide Whether Early Surgery Should Be Performed

Whether orthopedic procedure performed during the infantile period will affect the normal development of hands and upper limbs after operation is often a problem that puzzles hand surgeons and plastic surgeons. Different surgery timing should be selected according to the differenced in the disease categories. In the meantime, the degree of the influence of various deformities on the patient's hand functions and physical and mental development should be considered to decide whether to perform early surgery.

3.3.1.5 It Is Unnecessary to Delay the Surgery Due to Hypogenesis of Immune System

Netscher (1990) summarized the discussion of over 60 authors on the selection of surgery timing for congenital deformities of hands and upper limbs and proposed that it is necessary to delay the surgery due to hypogenesis of infantile immune system. According to the author's experiences in the plastic surgeries on chapped lips, cleft palate, and many congenital deformities of hands and upper limbs, if patients are surgically corrected within several hours to several months after birth, the hypogenesis of immune system will not affect the surgical effects. Therefore, the authors believe that, as long as the conditions permit, it is unnecessary to delay the corrective procedure during the infantile period due to the hypogenesis of immune system.

3.3.1.6 For Special Patients, Surgical Correction Can Be Postponed

The author [80] thinks that surgical correction can be performed during the infantile unless the deformities can be relieved as the child grows, such as congenital snap finger,

under which condition the surgical correction can be postponed. Most limb formation disorder, finger contracture deformity, tumorous finger deformity, ring constriction and some deformities of dysplasia can be surgically corrected around 6 months after the birth. For some patients, it is necessary to perform preoperative rehabilitative therapies in the first place; for example, for the pediatric patients with syndactyly, the finger webs should be rubbed for 3–6 months before the surgery to increase the width and length of inter-syndactylous skin. The author of Campbell's Operative Orthopedics suggests that the surgery of syndactylia be scheduled before the preschool period, and the purpose is to increase the flabbiness of inter-syndactylous skin. The author believes that this is unacceptable to some Chinese parents because they hope that the deformed hands of their children can be corrected before they meet strangers. Especially for the pediatric patients with synpolydactyly of the entire hand, the manifestation is often web-shaped deformities of both hands. The author suggests that their parents should rub and tract their hands after their birth. When they become able to perform the grabbing action 3 months later, they can have finger separating and thumb reconstructing surgery in different times.

Auxiliary physical therapies such as fixation with cleat and brackets should first be performed for some contracture deformities. Pediatric patients with such contracture deformities should be given surgical correction at the age of 2–4, and the delay of surgery will affect the effects of correction. Pediatric patients with ulnar deviating hand (namely, windblown hand deformity) should be given bracket traction therapy before the surgery, and then surgery is performed during the infantile period; otherwise, the delay of surgery will lead to poor postoperative effect (Figs. 3.28 and 3.29).

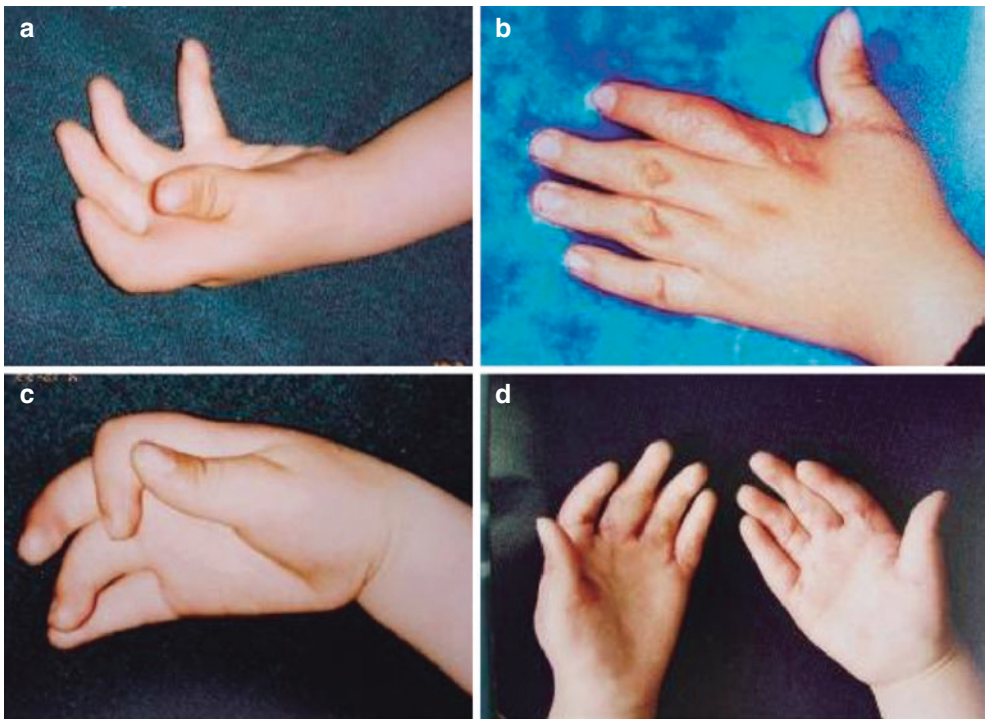

Fig. 3.28 A 4-year-old girl, before and after surgery on windblown hand deformity. (**a**) Before the surgery of left hand. (**b**) After the surgery of left hand. (**c**) Before the surgery of right hand. (**d**) Good postoperative effect

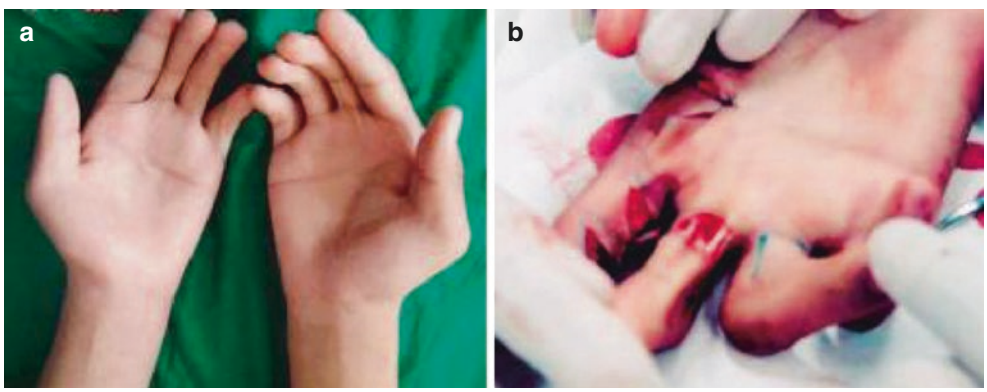

Fig. 3.29 A 14-year-old boy, surgical treatment of windblown hand deformity. (**a**) Before operation. (**b**) During the surgery, the middle finger vascular nerve tract can be found very tense, making it difficult for the finger flexion to be corrected

3.3.2 Selection of Surgery Timing

3.3.2.1 Plastic Surgery on Congenital Deformities of Hands and Upper Limbs of Patients Under 1

The indications of plastic surgery carried out on congenital deformities of hands and upper limbs of patients under 1 include the following:

1. As for the pediatric patients with serious congenital deformities of hands and upper limbs that affect the hand and upper limb functions or endanger limb survival, such as serious limb ring constriction, surgical correction should be given as early as possible; otherwise, serious lymphedema at the distal end of stenosis of the affected limb will arise, and even gangrene will appear due to complications.
2. The conditions of congenital deformities of hands and upper limbs are very mild, and simple surgeries are only needed to correct the deformities and improve functions, such as the type 7 deformity of thumb duplication, and radial or ulnar hyperdactylia without other deformities in hand joint.
3. Hand deformities that can be corrected within 2–4 h, such as cleft hand, neoplastic hands with partial displacement, simple syndactylia, dysplasia of partial hands, dysplasia of thumbs, etc.

As for pediatric patients who need complex surgeries such as thumb reconstruction for mirror hand, the author suggests that corrective surgery should be performed at 7 months after birth when they become able to perform gripping, nipping, grasping, and holding actions.

3.3.2.2 Plastic Surgery on Congenital Deformities of Hands and Upper Limbs of Patients Aged Within 2

It is best for a pediatric patient to undergo plastic surgery for congenital hand and upper limb deformities before he/she becomes 2 years old. Any pediatric patient with congenital deformities should receive the first plastic surgery or have the entire deformity corrected during this period, including repair of thumb dysplasia, thumb reconstruction, tendon grafting, repair and correction of thumb adduction deformity, repair of complicated thumb duplication deformity, surgical treatment of complicated syndactylia, polydactylism and mirror hand, plastic surgery of complicated cleft hand, plastic surgery of mild limb ring constriction, and plastic surgery of some windblown hand deformities.

It is worth noting that, in case of surgery that may result in epiphysis destruction or affect blood supply, plastic surgery should be delayed.

3.3.2.3 Plastic Surgery on Congenital Deformities of Hands and Upper Limbs of Patients Aged Over 2 Years

The indications of plastic surgery on congenital deformities of hands and upper limbs of patients aged over 2 include bone joint fusion, macrodactylia (megalomelia) deformity, windblown deformity, finger flexion or lateral flexion deformity, complicated neoplastic hand deformity, and brachydactylia (micromelia) deformity. Plastic surgery on congenital thecostegnosis should be performed after the patients become 2 years old because many pediatric patients with congenital micromelia may recover at the age of 2–3. In addition, because the plastic surgery of radial or ulnar club hands needs cleats and brackets to correct the deformity of deviation and the cleats and brackets should be replaced as the patient gets older, corrective procedure can be performed when the patient is between the age of 2 and school age [81].

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Disorders of Formation of Upper Limbs

4

Bin Wang, Wei Wang, Ping Yao, Xu Gong, Laijin Lu, Bo Chen, Jianmin Yao, Longchun Zhang, Feng Ni, and Fei Tian

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4.1 Introduction

The upper limb disorder during the embryonic period will result in transverse or longitudinal deformities of upper limbs. The manifestation of the transverse one is lipomeria or adactylism, and the manifestation of the longitudinal deformities is cleft limb or cleft foot. Partial segmental or longitudinal dysplasia of limbs will produce various deformities of limb (finger) shortness, and the manifestations can become various and complicated.

4.1.1 Etiological Factors

So far, the cause of most limb formation disorders has still been unclear, and the related factors include the following three kinds:

4.1.1.1 Genetic Factors

The material basis of heredity is genes, which are arranged on the chromosomes in the linear shape and have their own specific positions on the chromosomes. Genes may determine the morphological characteristics and physiological and biochemical characteristics of human body. Likewise, various congenital malformations can be passed to the next generation through the genes in the cell chromosomes. The various congenital deformities of hands are no exception, and some may even be passed to the next few generations in the same family. Some believe that the deformity of abrachia is a long-distance inheritance, namely, atavism.

4.1.1.2 Embryological Factors

During embryonic development, the basic components of upper limbs are mainly formed at 3–7 weeks of embryonic development. During this period, if the limb bud development of embryo is affected by various factors, congenital deformities may result. Different deformities will be formed when the teratogenic factors work in different periods: if they work at 3 weeks, such deformities as abrachia, micromelia, and lipomeria will be formed; if they work at 6 weeks, such deformities as adactylia, brachydactylia, and syndactylia will be formed. One teratogenic factor may affect not only one limb bud of embryo but also the differentiation and development of other sites. Therefore, clinically the patients with congenital hand deformities often concomitantly suffer from physical deformities of other sites. Particular attention should be paid and missed diagnosis should be avoided.

4.1.1.3 External Factor During the Embryonic Period

During the embryonic period, some external factors will also lead to some deformities, which are irrelevant to the genetic inheritance in the chromosomes, so there is no inheritance phenomenon. There are many external factors that induce embryonic deformities. Currently, the external factors that have been verified by animal experiments and clinical observation include the following:

1. Nutritional factors. Experiments have proved that when the diets of white mother rats are deficient in vitamin A or vitamin C, bent limbs can be found in infantile rats after their birth, and soft tissue development will be affected; when their diets are deficient in vitamin B2, a variety of deformities can occur to the infantile rats, and the deformity of forepaw accounts for 50%.
2. Drug factors. Experiments have proven that drugs such as adrenocortical steroid, caryolysine, and trypan blue will lead to limb deformities. If a small amount of insulin is injected into the egg shells, cleft foot can be found in the newly born chickens; if some vitamin B_2 is injected together with insulin, this deformity will not appear.
3. Disease factor. The embryonic development will also be affected if its mother contracts a disease during pregnancy. For example, if the mother is infected by rubella virus within the 4 weeks after she becomes pregnant, the fetus may suffer multiple congenital deformities.
4. X-ray factor. If the mother rat has even been exposed to X-ray, the infantile rat it gives birth to may suffer such deformities as ectrodactyly, syndactyly, polydactyly, and lipomeria and also concomitantly deformities in the eyes and kidneys. Experiments have proven that X-ray can inhibit or stop embryonic development so as to further induce various deformities.

4.1.2 Pathological Basis

In case of transverse or longitudinal dysplasia in limbs, the involved bones, joints, muscles, vessels, and nerves will suffer damage or absence, further resulting in various deficits in upper limbs and hands or the deformity of cleft, accompanied by corresponding functional disorder.

4.1.3 Classification

According to the plane, direction, and axial line of the upper limb absence, limb formation disorders can be classified into two categories, namely, transverse formation disorder and longitudinal formation disorder. The manifestation of the former is defective or amputated extremity on different planes from shoulder to fingers; the manifestation of the latter is radial ray deficiency (the lower limb is the tibial side), ulnar ray deficiency (the lower limb is the fibular side), and central ray deficiency. The central ray deficiency is often manifested as deformity of cleft hand (cleft foot), namely, cleft limb or cleft finger.

4.2 Upper Limb Transverse Formation Disorder

Congenital limb (finger) transverse defect is called congenital ectrodactyly. The planes of defect are from shoulder level, upper arm, elbow level, and forearm to the wrist, palm, and fingers, among which the most common one is below the elbow joint or the distal end of the forearm. Disease almost occurs to one side. In terms of embryonic development, the earlier the duration of pregnancy of defect is, the higher the plane of defect is. In case of involvement at 3 weeks of pregnancy, the deformed plane will be the arm; in case of involvement at 6 weeks of pregnancy, the deformed plane will be the fingers. Clinical findings indicate that the higher the involved plane is, the more likely a patient is to concomitantly suffer from syndromes.

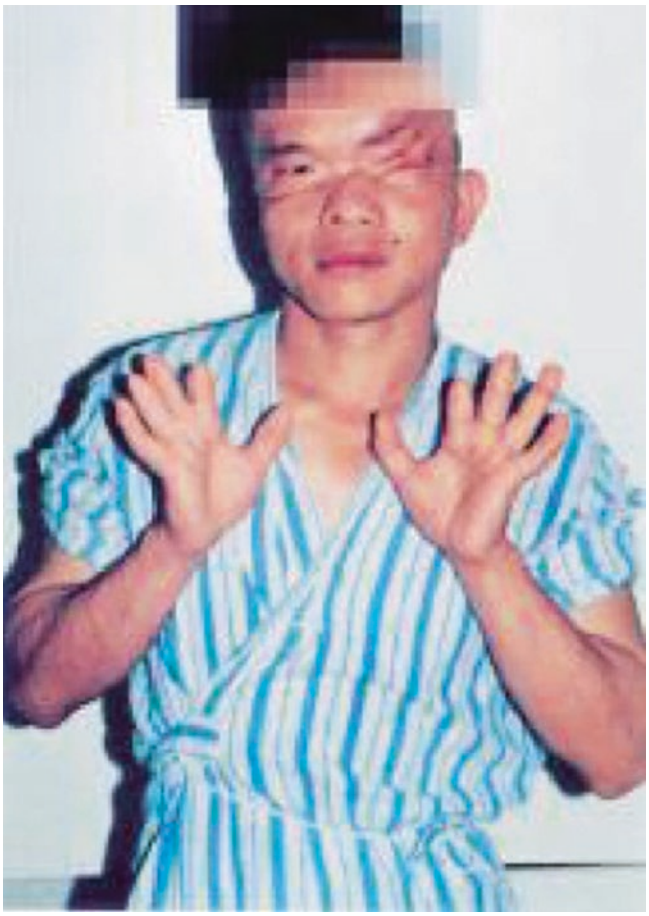


Fig. 4.1 A patient with congenital severed finger and the ring constriction of his head

4.2.1 Ectromelia

4.2.1.1 Etiological Factor and Epidemiology

Congenital limb (finger) transverse defect is sporadic. The etiological theory is the vascular damage of limb bud development or the injury of apical ectodermal ridge, and the risk factors include the use of misoprostol [1], ethanol, tobacco, and cocaine during pregnancy.

4.2.1.2 Clinical Manifestations

Congenital lipomeria (adactylism) can occur at any level from congenital amelia to congenital severed finger. The level of amputation (severed finger) is often similar to that of limb ring constriction, but the causes of the two are different. Congenital lipomeria (adactylism) can be classified into congenital broken arms, forearm inferior, elbow inferior, wrist inferior and palm inferior lipomeria, and adactylism deformities (Figs. 4.1, 4.2, and 4.3).

Patients with congenital lipomeria (adactylism) often concomitantly suffer from hydrocephalus, schistorachis, cerebral spinal meningocele, equinus, ulnar-radial bone fusion, and radial head dislocation. Because patients with congenital

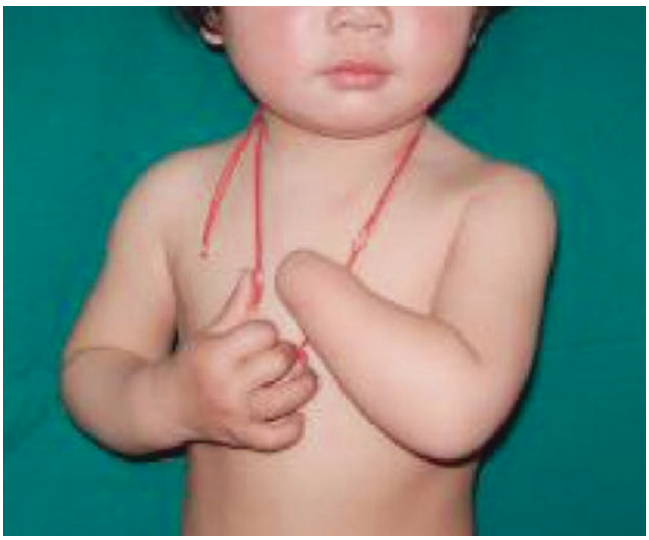


Fig. 4.2 Absence of left hand and congenital absence of the wrist plane

lipomeria (adactylism) often concomitantly suffer from bone and muscular absence and deformities, other deformities should be considered when a therapeutic regimen is formulated.

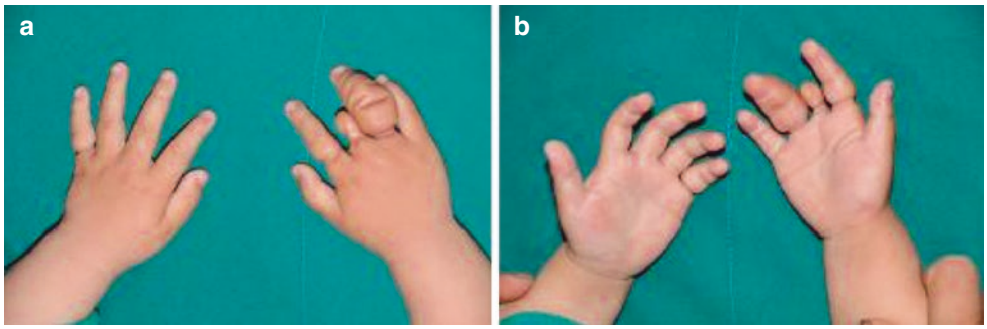


Fig. 4.3 Congenital severed right middle finger accompanied by ring constriction

4.2.1.3 Treatment

The treatment on patients with amelia (aphalangia) is relatively difficult.

Artificial limbs (fingers) can functionally compensated for some congenital broken arms and lipomeria and adactylia below the forearm and wrist. In addition, they can improve the appearances of the patients suffering from such diseases. It is usually suggested that artificial limbs (fingers) be used by infants who are not less than 6 months old or can independently get up and sit down, and at the beginning passive artificial limbs (fingers) can be used. When the child is 2–3 years old, an assessment should be carried out to guide the use of active artificial limbs (fingers). According to the differences of control source, active artificial limbs (fingers) can be classified into myoelectrical type and body-driven type. Crandall R.C. et al. [2] carried out a follow-up visit for 14 years on the average of 34 patients and found that 15 patients chose cosmetic artificial limbs (fingers) (passive type), 14 chose the body-driven type artificial limbs (fingers), and only 5 chose myoelectrical type artificial limbs (fingers).

For the patients with bilateral low congenital broken arms and lipomeria and adactylia below the wrist, especially for those whose both eyes are blind, forearm splitting operation (Krukenberg reconstruction) is indeed a good option. The typical surgery procedures are shown in Fig. 4.4.

As for the lipomeria and adactylia below the congenital broken palm, if conditions permit, the patients can receive surgeries such as toe-to-hand grafting and expanded toe-to-hand grafting to reconstruct the thumb or other fingers.

Limb bone extension can also be adopted: first choose the proper bone lengthener to extend the limb by 1–2 mm every day, which is of great help to the lipomeria (adactylism) at multiple levels. This surgery can serve as a therapeutic surgery as well as a preparatory surgery of other surgeries (e.g., toe-to-hand grafting), but attention should be paid to its relatively high incidence of complications. Alekberov C. et al. [3] applied Ilizarov [4] technology (distraction osteogenesis) to perform bone extension on six patients with lipomeria at elbow joints (who have been fitted with and used artificial limbs). The follow-up visit lasted for 39–48 months, and 5.6 cm (3.4–8.4 cm) was extended on average.

4.2.2 Defect at the Middle Segment of Upper Limbs: Phocomelia Deformity

According to the description of Goldfiab, the proximal dysplasia at the radial side and the ulnar side can lead to proximal continuous defects rather than middle defects. The deformed hands which are formed are shaped like seal limbs, so it is called phocomelia.

4.2.2.1 Clinical Epidemiology

Phocomelia is a rare kind of congenital deformity. Kallen et al. [5] studied 1,368,000 newborns, and its incidence among them was only 4.2/100,000. Among the 48 patients studied by him, 29.2% suffer the deformity of the right limb, 22.9% suffer the deformity of the left limb, and 47.9% suffer the deformity of both limbs. In the same study, those with involved upper limbs accounted for 68.8%, those with involved lower limbs accounted for 29.2%, and those with involved both upper and lower limbs only accounted for 2.1%. So far, no one has published a study on the risk factor of phocomelia, but thalidomide has a clear relation with the occurrence of phocomelia. Brent and Holmes [6] believed that 24–53 days after fertilization was the sensitive period of involvement of upper limbs, and the 28–33 days after fertilization was the sensitive period of involvement of lower limbs.

4.2.2.2 Clinical Manifestations

Phocomelia can occur to both lateral side and bilateral sides, and similar deformities can occur to the upper limb and the lower limb at the same time (Fig. 4.5).

According to the anatomical type, Frantz and O'Rahilly classified phocomelia into three types: type I, the hand was directly connected with the shoulder, without humeral bone or forearm; type II, humeral bone suffered absence or serious hypoplasia, and forearm and hands were connected to the trunk; and type III, the hands were connected to the humeral bone, without forearm (Fig. 4.6).

According to Tytherleigh-Strong and Hooper [7], although Frantz and O'Rahilly classification was direct and straightforward, it has some defects in clinical practice. They reviewed 44 cases of confirmed phocomelia and found that only 11 conformed to Frantz and O'Rahilly [8] classification

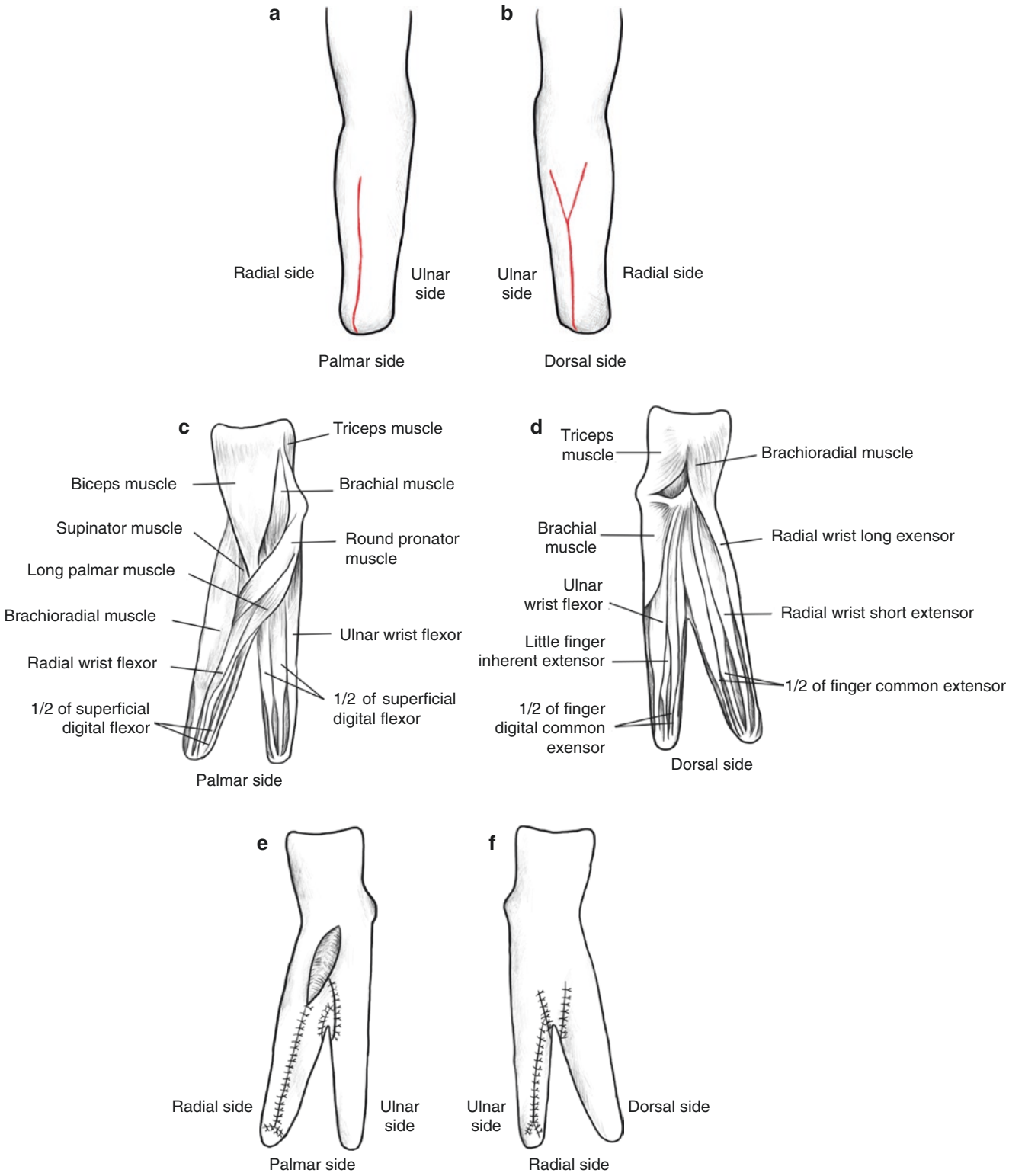


Fig. 4.4 Forearm splitting operation (Krukenberg reconstruction) of congenital lipomeria. (**a**, **b**) Design of forearm incision. (**c**, **d**) Divide forearm muscles into two groups. (**e**) Close palmar incision (the *oval-shaped region* is the skin defect area, where the dermatoplasty with skin graft with an intermediate thickness can be performed). (**f**) Close dorsal incision (Reprint with permission from Wolters Kluwer Health, Inc. License Terms and Conditions) (originate from Swangson AB:The Krukenberg procedure in the juvenile amputee. J Bone Joint Surg 46A; 1540, 1964)

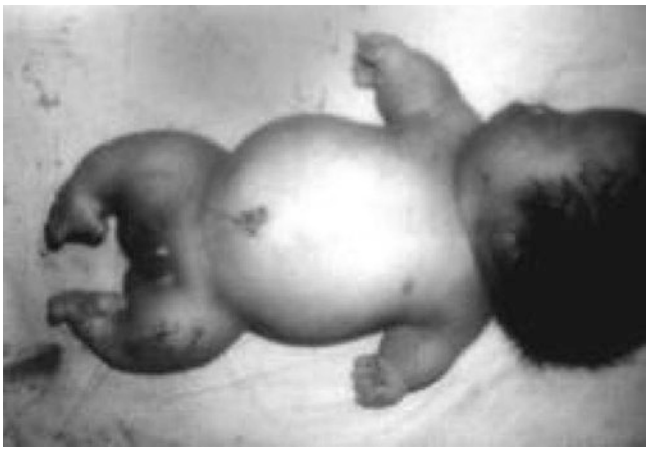


Fig. 4.5 Bilateral incomplete phocomelia (foot), dead infant

(nine cases of type I, two cases of type III, and no case of type II). They classified the remaining 33 cases into three types: type A, abnormal humeral bone was connected with abnormal radial bone or ulnar bone; type B, abnormal humeral bone was connected with abnormal radial bone and ulnar bone; and type C, abnormal humeral bone was fused with radial bone and ulnar bone (Fig. 4.7).

4.2.2.3 Treatment

There are few reports on the surgical indications of phocomelia. The main reason is that there are a small number of such cases and the pathological study and judgment on the affected limbs are very superficial. To solve the problems of unstable shoulder joint or thumb opposition function disorder, clavicles downward shift and thumb opposition function

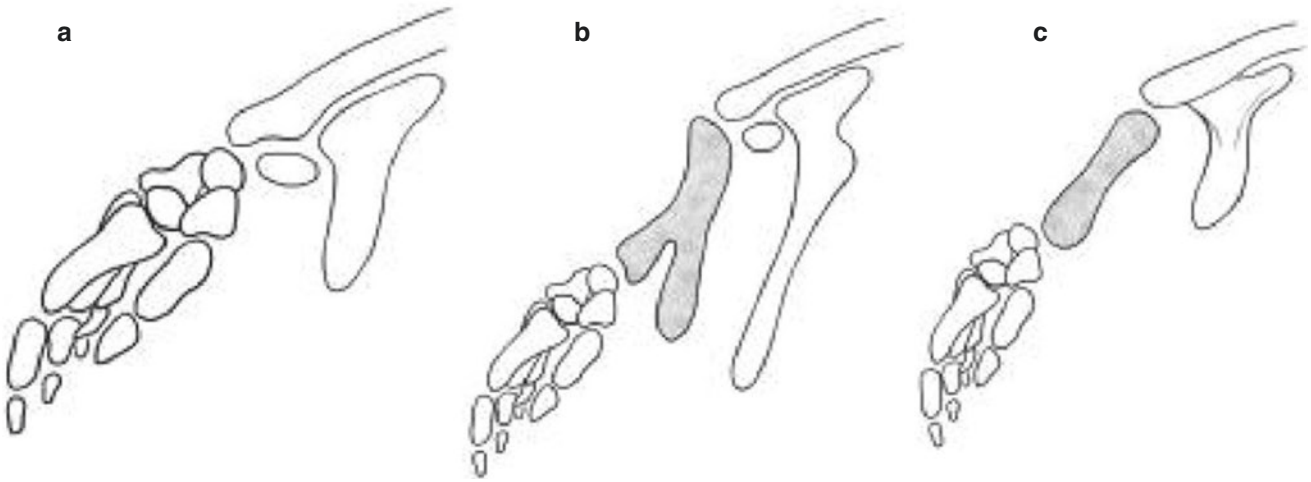


Fig. 4.6 Frantz and O'Rahilly classification of phocomelia. (**a**) Type I. (**b**) Type II. (**c**) Type III (Redrawn from Frantz CH, O'Rahilly R; Congenital skeletal limb deficiencies, J Bone Joint Surg 43A: 1202, 1961)

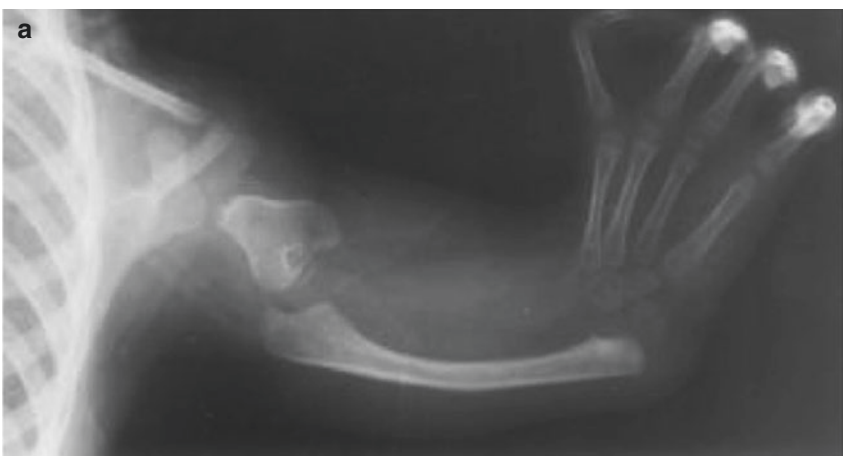


Fig. 4.7 Unclassifiable phocomelia. (**a**) Type A. (**b**) Type B. (**c**) Type C

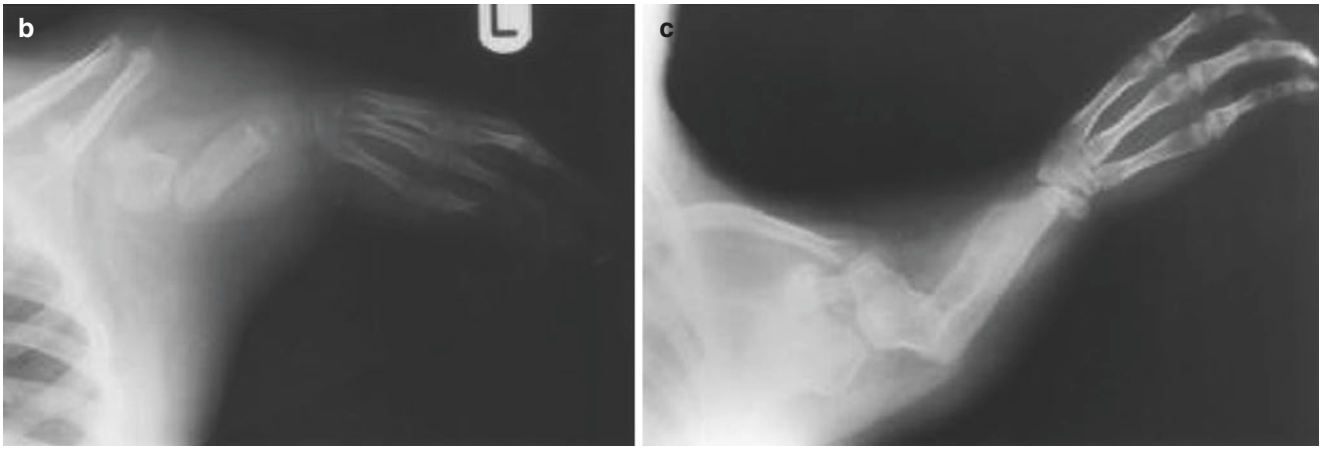


Fig. 4.7 (continued)

reconstruction can be performed. In case of presence of some limbs and relatively thick tubular bones, Ilizarov technology (distraction osteogenesis) can be performed to increase the length of limbs. In order to increase the stability and length of phocomelia, surgeries of clavicular grafting or fibular grafting can be adopted. In case of deformities in the lower limbs and apraxic presence, the residual bone tissues can be considered as the donor area.

4.3 Incomplete Shoulder Descending

Incomplete shoulder descending is also called congenital high scapula and falls into the category of limb differentiation disorder in taxology. Congenital high scapula is a rare kind of congenital shoulder deformity, which was first described by Eulenburg [9] in 1863. Sprengel [10] reported four cases in 1891; from then on, this deformity has been called Sprengel's deformity.

4.3.1 Etiological Factors

At the end of the first 3 months of embryonic development, the bilateral shoulder girdles should descend from the neck to the upper region of thorax, and congenital high scapula is the result of incomplete shoulder girdle descending. Excessively high intrauterine pressure and polyhydramnios or hypamnion during the pregnancy, the abnormal embryonic scapula and interspinal girdle, cartilage connection, amyoplasia, etc. can prevent scapula from descending to the normal position. There are studies revealing that the generation of upper and lower limbs can be expressed by HOXA and HOXD genes, and the mutation of such genes can induce congenital high scapula.

4.3.2 Pathological Changes

Khairouni A. et al. [11] summarized 19 pediatric patients (23 shoulders, four patients had diseases in bilateral shoulders) and proposed four kinds of abnormal anatomical phenomena:

4.3.2.1 Dysplasia of Scapula

The changes in thoracic girdle blastema and the changes in the early growth of the epiphyses of vertebral margin of scapular bones and spines lead to the dysplasia of scapula, whose manifestations are: the scapular angle is very smooth, the medial margin is very straight, and the height of the bilateral scapular bones is obviously different.

4.3.2.2 Omovertebral Bone Bridge

Fifty-four percent of the pediatric patients have omovertebral bone bridge, which is an important reason that induces deformities. Omovertebral bone bridge consists of bones, cartilages, fibers, or several tissues, is located at the medial margin or the upper medial corner of the scapula, and is connected with the cervical vertebral spinous processes or transverse processes.

4.3.2.3 Muscular Abnormality

In case of amyoplasia around the scapula, the lower part of the inferior part can be absent, and the dysplasia of the rhomboid muscle and the levator scapulae muscle can present partial fibrosis. They also found one pediatric patient with absence of sternomastoid muscle.

4.3.2.4 Simultaneous Presence of Other Congenital Deformities

The patient may concomitantly suffer from other congenital deformities such as cervical and thoracic vertebral hemivertebrae, cervical vertebral lateral curvature, spina bifida, rib absence, and rib fusion.

4.3.3 Clinical Manifestations and Classification

Incomplete shoulder descending mainly affects the appearance and the motor function of the shoulder joint of the pediatric patients. The manifestation of the pediatric patients is the asymmetry of bilateral shoulders. The superior angle of the affected scapula can upward reach the level of the fourth cervical vertebra and can downward reach the level of the second thoracic vertebra, and the abduction of the affected shoulder can be limited (Fig. 4.8).

Currently, the Cavendish classification method [12] is mainly adopted to classify incomplete shoulder descending into four grades: grade I, light deformity, the height of bilateral shoulders is almost the same after clothes are put on; grade II, light deformity, the heights of the two shoulders are almost the same, and the superior medial angle of affected scapula is visible when no clothes is worn; grade III, the height of the affected scapula increases by 2–5 cm; and grade IV, severe deformity, the superior medial angle of the affected scapula almost reaches the level of occipital bone; the shoulder has skin web and presents the deformity of short neck. However, Khairouni A. et al. [11] believe that Cavendish classification is a morphological and aesthetic classification and the classification is too subjective and not accurate enough, and they recommend Rigault's classification [13] (radiological classification): grade I, the superior angle of scapula reaches the level of the first thoracic vertebra (T1) and below; grade II, the superior angle of scapula reaches the level between the first thoracic vertebra (T1) and the fifth cervical vertebra (C5); and grade III, the superior angle of scapula reaches the level of the fifth cervical vertebra (C5) and above (Fig. 4.9).

The application of 3D CT to the diagnosis and treatment of incomplete shoulder descending has been increasingly wide. According to the results of spiral CT scan, Liu Tao et al. [14] measured the height difference of the bilateral scapulae with the medial margin of the scapular bone and scapular spine as the reference point and classified them into four degrees according to Cavendish classification method: degree I, the height difference of bilateral scapular bones is less than 1 cm; degree II, the height difference of bilateral scapular bones is 1–2 cm; degree III, the height difference of bilateral scapular bones is 2–5 cm; and degree IV, the height difference of bilateral scapular bones is more than 5 cm.

4.3.4 Treatment

In the treatment of this disease, consideration should be given to the age, deformity degree, and general conditions of the patient. As to the age at which a patient should receive treatment of this disease, different reports have different answers. But most consider that the age between 3 and 7 is the best treatment time. As for patients under 3 years or with relatively mild deformity, pediatric patients can be asked to make active movements and passive movements to enhance the amplitude of upper limb abduction. However, age is not the decisive factor for surgery. Khairouni A. et al. [11] consider that the age of the pediatric patients does not affect the postoperative effects, but the concurrence of cervical vertebral deformity has adverse effects on prognosis. Generally,

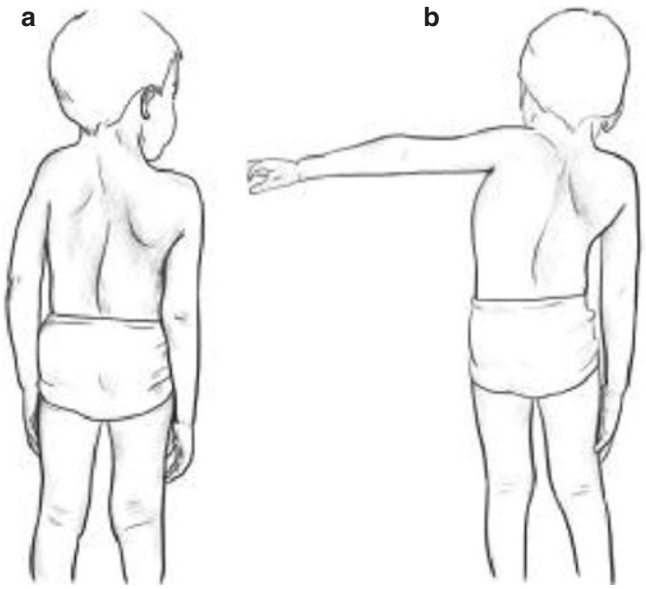


Fig. 4.8 Incomplete shoulder descending. (**a**) Shoulder appearance. (**b**) Limited abduction of the affected shoulder

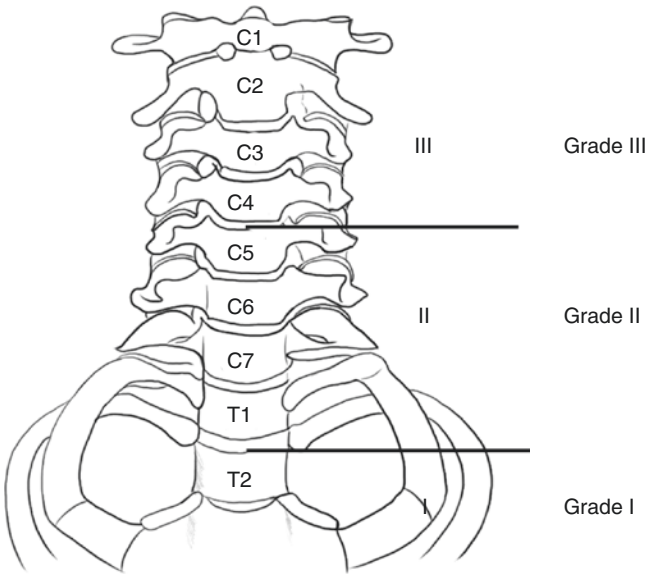


Fig. 4.9 Rigault's classification method (Redrawn from Sprengel OK. Die angeborene verschiebung des schulterblattes nach oben. Berlin: Archiv Fur Klinische Chirurgie; 1891;42:545–549)

surgical correction is inapplicable to Cavendish grade I deformity; as for grade II deformity, the consent of the patient's parents should be won as to whether surgery should be performed; as for grade III and IV deformities, surgery must be performed. The classic surgical methods mainly include Woodward's operation [15] and Green's operation [16], and many modified operations have developed from these two kinds of operations.

4.3.4.1 Woodward's Operation

Woodward's operation refers to the exposure of midline section, the remission of trapezius muscles and rhomboid muscles, the resection of omovertebral bone, and the reduction of scapular bones. Dendane A.M. et al. [17] reported the application of Woodward's operation to six pediatric patients (four girls and two boys), whose surgical age ranged from 4 to 15 (10 on average) and the follow-up visit duration was 8 months to 10 years (4 years and 2 months on average). The mean improvement of postoperative shoulder joint abduction was 40° (30°–75°), and the results of functional and appearance assessment were excellent for three cases, good for two cases, and fair for one case. The author considers that it is best for one to receive scapular bone displacement before he/she is 4 years old. Khairound A. et al. [11] applied the modified Woodward's operation [15] (through correction of scapular inclination) and treated 19 cases of incomplete shoulder descending, among whom four suffered from bilateral diseases and 79% showed improvement in appearance and functions. It was believed that the age and omovertebral bone of the pediatric patients did not affect the postoperative effects, and the prognosis of the patients concomitantly with cervical vertebral deformities was poor.

4.3.4.2 Green's Operation

Green's operation [16] refers to the making of the reversed L-shaped incision from the superior region of the scapular spine to the medial margin of scapula; the remission of trapezius muscle, levator muscle of scapula, and greater and lesser rhomboid muscle; separation of omovertebral bone chorda; rotation of the scapula; downward shift of the scapula to the normal position, and suturing it into the latissimus dorsi bag. Steel silk is used to tract the scapula to the iliac bone, the scapular correction position is maintained, and after operation the gypsum is used for external fixation. As this operation model causes serious injuries, many authors have modified it. Bellemans M. et al. [18] applied the modified Green's operation [16] (the serratus anterior muscle is not incised, and the patient immediately moves after operation) to treat seven pediatric patients with incomplete shoulder descending, and the results indicated that postoperative abduction function was improved. They believed that this modified operation mode has advantages and can improve postoperative functions.

4.3.4.3 Modified Operation

Some scholars believed that conventional surgeries just focused on solving aesthetic problems of incomplete shoulder descending and paid scant attention to improvement of functions. They used other methods for surgical treatment and have achieved good clinical effects. Meals [19] reported treatment of eight patients with congenital elevated scapula through partial resection of the scapula and remission of long head of triceps. If omovertebral bone was found during the operation, it was also removed, nearly 50% of the scapular body was cut, and acromion, coracoid process, and glenoid cavity were kept from injuries. The mean postoperative follow-up visit duration was 5.5 years, and the shoulder joint abduction was recovered to 150°. X-ray film indicated that nearly 50% of scapular body was amputated during the operation, and the postoperative follow-up visits could basically return to the preoperative level. Masquijo J.J. et al. [20] pointed out that the use of Meal's method to treat incomplete shoulder descending could significantly expand the range of motion of shoulder joint and significantly improve the appearance, and complications were rare. They treated a total of 14 patients, the mean postoperative follow-up visit duration was 45 months (from 12 to 74 months), the shoulder joint flexion increased from the preoperative mean level of 83.9° (50°~100°) to the mean level of 152.1°(110°~180°), the abduction increased from the preoperative mean level of 81° (50°~120°) to the mean level of 145° (100°~180°), no patients experienced muscle strength decline or brachial plexus nerve injuries, and the mean appearance grade increased by two grades (Cavendish classification). Zhang Z.M. et al. [21] treated the deformity of elevated scapula through resection of the superior part of scapular spine; omovertebral bone was also removed if any. They adopted the modified reversed L-shaped incision and relieved the contractural tissues at the medial side of the scapula. They treated a total of 26 patients (28 shoulders), the follow-up visit duration was 10 months to 7 years (3.9 years on average), and the preoperative abduction of 18 shoulders was less than 120° with a mean improvement of 52°; the preoperative abduction of ten shoulders was over 12° with a mean improvement of 19°; improvement to varying extents was achieved in 23 shoulders. During and after the operation, no nerve complication occurred, and no patient complained of the problem of scar.

The application of 3D CT to the treatment of incomplete shoulder descending has been increasingly wide. Liu Tao et al. [14] believed that CT plain scan could display the planar structural relationship and clearly display the horizontal plane connection between the cervical vertebrae and the shoulder vertebral connector. Spiral CT scan and three-dimensional reconstruction imaging can show the three-dimensional image of the lesion site, and before the operation they can directly indicate the lesion degree of incomplete shoulder descending and the relation between the cervical, thoracic,

and shoulder vertebral connector and the affected scapula. In case of concomitant deformities, the difference between the affected scapula and the healthy scapula in appearance can be accurately classified through measuring the height difference of the scapula to facilitate the development of surgical regimen. To some extent, this shortens the surgical duration, avoids the blindness of surgical procedures, and reduces the occurrence of iatrogenic complications.

4.3.5 Complications

The postoperative complications of incomplete shoulder descending vary according to surgical mode and mainly include brachial plexus nerve injuries, keloid, exostosis, and superficial wound infections. Brachial plexus nerve injury is a serious postoperative complication. Wang Hongqiang et al. [22] reported one 10-year-old pediatric patient that received Woodward's operation. After the operation, the patient experienced brachial plexus nerve injuries. But the nerve functions were recovered, and the movement function of shoulder joints was satisfying. In order to prevent brachial plexus nerve injuries, they recommended the pediatric patients over 8 years old and with deformities of grade III–IV to receive scapular downward transposition and also clavicular osteotomy, so as to avoid the occurrence of brachial plexus nerve injuries. Masquijo J.J. et al. [20] reported two patients having residual exostosis in the area of omovertebral bone resection and needing second-stage surgical resection. They believed that this was because the periosts that surrounded the omovertebral bone were not completely removed.

To sum up, treatment of incomplete shoulder descending should be individualized. Surgical regimen should be designed before the operation, operation should be done gently, and timely guidance should be offered after the operation so that good surgical effects can be achieved.

4.4 Dysplasia of Upper Limb Girdle

Ping Yao, Longchun Zhang, Jianmin Yao, Wei Wang, and Bo Chen

Dysplasia of upper limb girdle is considered to be the upper limb deformity induced by the combined action of gene programming error and disorder or environmental biological, physical, and chemical factors on the development process.

4.4.1 Anatomy

Upper limb girdle mainly consists of upper limb girdle bones, upper limb girdle muscles, and upper limb girdle bone connections. The upper limb girdle bones include clavicles and scapula. The upper limb girdle muscles mainly include deltoid, supraspinatus, infraspinatus, teres minor, teres major, and subscapularis muscle, which are all distributed around the shoulder joint, starting from the upper limb girdle bones and ending at the humerus, and they can move the shoulder joint and enhance the stability of the joint. The upper limb bone connections include sternoclavicular joint, acromioclavicular joint, and coracoacromial ligament.

4.4.2 Etiological Factors and Pathological Basis

The real etiological factors and pathogenesis of congenital deformities of upper limb girdles are little known, but two views are currently prevailing: one is that the development process is programmed by genes from the beginning; the other one is that development is the result of biological, chemical, and physical effects of sequences and is affected by four-dimensional space-time. The two viewpoints lead to the theory of genetic determinism and the theory of environmental determinism. However, more data indicate that the majority of deformities result from the joint effect of the two factors, but the influence of environmental factors is more significant. During embryonic development, malfunction of upper limb girdles may happen if any link is influenced. The development process of the upper limb girdles is as follows.

4.4.2.1 Embryonic Development

About 26 days after fertilization, the upper limb bud begins to take shape at the 9th–12th body segment of body wall. The limb buds that begin to appear consist of the mesodermal lateral plates covering ectoderm. Thereafter, the growth and differentiation of the limb buds proceed in the directions of three coordinate axes: proximal-distal, dorsal-ventral, and anterior-posterior, and the growth in each direction is regulated by independent signal centers. According to the hypothesis of Saunders (1957) and Zwilling (1960), the embryo proliferates and extends at the somatopleure from the heart to the cloaca and gradually forms a cylindrical region called Wolff's crest; it proliferates along the two sides of the crest to form limb buds. The external layer of limb bud is encapsulated by ectoderm, and the internal layer stems from the undifferentiated mesenchymal cells at the mesodermal lateral plates. These undifferentiated mesenchymal cells have been divided into two groups at this time with two different fates: one group forms the muscle mass and the other group produces cartilaginous primordium, tendons, and synovium. During the early stage, muscles and chondroblast cell groups are already present in the limb buds.

4.4.2.2 Fetal Development

The growth and development of skeletal musculature needs the growth and differentiation of cells and tissues in the accurate order to form the bone and joint structures. During this complicated process, occurrence of any disorder may cause serious deformity. During the evolution of human, the upper limb bones and soft tissues underwent a series of changes for being adapted to the posture of upright walking. First, the shoulder and neck could be identified at the glenoid cavity, the dorsal shoulder blade and ventral coracoid process were visible, and the gap between the two developed into shoulder joint. The separation of shoulder and neck enabled the upper limbs to move more freely, and there was a transitional stage before complete separation. In case of any problem arising during this stage, the deformity of scapular non-descending to varying extents could occur, or between the shoulder and neck, the high scapular fiber bundle was connected with the neck. The formation of shoulder joint can be generally divided into four stages: ① when intermediate zone appeared between the blastemas, ② when cartilage shapes appeared at the two ends of intermediate zones, ③ when intermediate zone fissures occurred, ④ and when synovial layers appeared around the articular cavity and were surrounded by joint capsules.

During the evolution of human, the components of strong bones that originally supported the anterior limbs disappeared, the neck and shoulders were completely separated, and the attached muscles had relatively big shift, making the upper limbs move more flexibly. The rotation of the upper limbs created conditions for the motion of the anterior side of the body. Descending from the neck, the scapula is located at the posterior upper chest wall and serves as the fundus of upper limbs. The inferior part of scapular spine becomes enlarged, while the superior part of the spine shrinks. In the meantime, the infraspinous muscularity is better than the supraspinous muscularity, the supraspinous muscles are no longer pulled in the anterior and posterior directions, and the mechanical effects become less significant. The importance of shoulder muscles is that they not only provide strength required for joint movement but also accurately control joint movement. Many upper limb muscles still maintain the initial attachment points at skulls and vertebral columns, making it possible to place effective control and produce lever effects for the purpose of making various kinds of complicated movements. As for the distribution of the upper limb muscles, it is first required that there should be a solidly attached fundus, followed by the development of upper arm muscles, and finally the muscles that control the distal side of the lever.

4.4.3 Classification

Currently there has been no uniform standard that classifies dysplasia of upper limb girdles. This article roughly classifies dysplasia of upper limb girdles into three types according to the components of upper limb girdles: ① dysplasia of upper limb girdle bones, ② dysplasia of upper limb girdle muscles, and ③ dysplasia of upper limb girdle bone connections. During the process of growth and development, from the beginning of limb bud formation to the maturity of shoulder joint, any factor that affects shoulder joint growth will lead to dysplasia of upper limb girdles and cause different clinical manifestations. The following will give a brief introduction to the clinical manifestations and treatment of dysplasia of upper limb girdles.

4.4.3.1 Dysplasia of Upper Limb Girdle Bones

1. Congenital clavicular pseudoarthrosis. This disease is rare, the etiology is unknown, and the main manifestations are obvious bulge of the middle clavicles and no pain and sensation of flabby movement as indicated by palpation. Different from the excessive bony calluses of newborn fracture and also different from the other concomitant tissue changes of hypoplasia of clavicles and skulls, X-ray only displays the hypertrophy and sclerosis at the two ends of false joints. The primary purpose of treatment is to improve appearance, and the optimal age for surgery is 3–5 years.
2. Dysplasia of clavicles and skulls. It can be manifested as clavicular development defect or complete aplasia, the bilateral clavicles can present asymmetric dysplasia often without acromial end, but absence of sternal end is rare, or it can be manifested as defect of middle clavicles, formation of false joint, unstable shoulder, or enlarged range of motion. Due to abnormal development of clavicles, the patients can suffer drop shoulder, dolichoderus, and shortened distance of inter-shoulder distance, and they can draw the two shoulders together at the anterior side. When the patients are born, their frontal bones and occipital bones are small sized, inducing delayed closure of fontanel. Most of the patients with this disease have a family history and the disease can occur sporadically. Dysplasia of clavicles and skulls can have multiple clinical manifestations and X-ray abnormalities, such as hypoplasia of bilateral clavicles, delayed closure of anterior fontanel and cranial sutures, and ossification defects of ulnar bones and ischial bones. In addition, the patients may also suffer facial osteodysplasty, dentition disorder, abnormal lengthening of the second metacarpal bone, blunting of the last segment of the phalanx, and coxa vara.
3. Dysplasia of the scapula. In addition to the abovementioned deformity of scapular non-descending, other deformities can also occur, such as abnormal scapular margins manifested as ossification of superior transverse ligament of the scapula and hook-shaped upper margin;

abnormal acromion, such as disunion of acromial epiphysis, bilateral acromions, and lengthened acromion; abnormal coracoid process; and abnormal inferior angle of the scapula.

4.4.3.2 Dysplasia of Upper Limb Girdle Muscles

1. Dysplasia of major pectoral muscle. The sternal head and the clavicular head of major pectoral muscles can be absent. Even the entire part of one side of major pectoral muscle can be completely absent, which can be displayed when the two hands press the seat handrail downward, but the movement is always not affected. The latissimus dorsi muscles can present compensatory hypertrophy.
2. Dysplasia of smaller pectoral muscle. The smaller pectoral muscles can start from the first rib and ends with the sixth rib. The smaller pectoral muscles can have the attached insertions or run over the coracoid process and over the coracoacromial ligament toward the posterior and lateral side and stop at the shoulder joint capsule, greater tubercle, and the middle one third site of the glenoid cavity or expand to the medial and the lateral sides of the coracoid process; the former extends to the costocoracoid ligament in most cases, and the latter extends to the coracobrachial muscle in most cases. Sometimes one longitudinal sternal muscle occurs on the breast bones, as if the chest rectus of some animals was on a line. The sternal muscles start from the anterior aponeurosis in front of the manubrium sterni, in the superior region become connected with the tendon fiber of the sternomastoid muscle breast bone head, and in the inferior region become attached to the sixth to seventh cartilages and the anterior layer of the sheath of rectus abdominis.
3. Dysplasia of sternal muscle. The incidence of sternal muscles in China is about 13%. It has a higher rate in newborns and a lower rate in adults. There are different explanations as to the source of sternal muscles: some believe that they are formed by the upward extension of musculus rectus abdominis; some believe that they are formed by the downward extension of sternomastoid muscles; and others believe that they are formed by the separation of major pectoral muscle. In terms of phylogenesis, the interpretation which considers sternal muscle as the residue of mammalian major cutaneous muscles is relatively reasonable. Sternal muscles can have many variations and can take myocutaneous shape or have abdominal muscles; they can also take digastric shape, bicipital shape, or the shape of multiple abdominal muscles. Sternal muscle can be unilateral or bilateral; sternal muscles at one side can run over the midline to the opposite side. Sternal muscles are dominated by anterior thoracic nerves or intercostal nerves.
4. Dysplasia of subscapular muscles. Subscapular muscles have a tendency of being separated into several muscle bundles, but complete separation has never been seen. Subscapular muscles have sesamoid bones inside, and most of them have cartilaginous nuclei at bilateral sides during the fetal period and are located in the tendons or muscles. Sesamoid bones may have cartilage surfaces and humeral head-related segments.
5. Dysplasia of supraspinous muscles, infraspinous muscles, teres major muscles, and teres minor muscles. The insertions of the supraspinous muscles are often fused with some of or all the smaller pectoral muscle tendons. Infraspinous muscles can be classified into upper, middle, and lower parts. The upper part starts from below the shoulder blade, and some start from the medial surface of the sub-spine fascia. It has a tendency of separation in the middle, which is called small infraspinous muscle, accounting for about 4%. Union can still be found between infraspinous muscles and teres minor muscles; partial union accounts for 8%, complete union accounts for 4%, and the latter can be misdiagnosed as absence of teres minor muscle. Partial union between teres minor muscle and triceps muscle of the arm can be observed, and separation often occurs. The part of teres minor muscle attached to the surgical neck of humerus may sometimes be separated more or less, to form independent muscle called teres minor muscle. The insertion of teres major muscle is often fused with the insertion of the broadest muscle of the back, and the proportion is about 62%; there can also be some additional muscle bundles, fused with rhomboid muscles or long head of triceps.

4.4.3.3 Dysplasia of Upper Limb Girdle Bone Connection

1. Abnormal glenoid cavity. In case of normal development, glenoid cavity should present the biconcave shape in the longitudinal and transverse directions, but there can be some variations in the depth. Some are nearly flat, slightly dented, or deeply dented. Glenoid joint may excessively incline, causing instability in the rear of the shoulder. If the secondary ossification center in the middle part of the glenoid cavity still does not appear after the age of 15, the glenoid fossa can become flat and stop developing. Coupled with excessive posterior inclination of the glenoid cavity, obvious deformity of unstable development can be induced. As for such patients, wedge-shaped osteotomy can be performed at the rear of the scapular neck, iliac bones or acromions are used for bone grafting and filling, and osteotomy site is at least 1 cm away from the medial side of the posterior margin of glenoid cavity to ensure survival. The measured glenohumeral index is the

maximal transverse diameter of the glenoid cavity or the maximal transverse diameter of the humeral head. If it is 56.6 ± 5.6, it indicates dysplasia of glenoid cavity.

2. Congenital shoulder dislocation. Congenital shoulder dislocation means abnormality in the relationship between humeral head and glenoid cavity caused by scapular-humeral dysplasia, which is clinically rare. There are two types of glenoid cavity dysplasia: (1) the humeral head is at the normal position, but the size and proportion are inappropriate, like the congenital hip joint dislocation and acetabular bone aplasia, and (2) the abnormality of glenoid cavity. The infantile glenoid cavity is in the front. Although the humeral head develops normally, it shows the tendency of upward and backward development. Sometimes, the coracoid process extends forward and outward, causing the glenoid cavity and humeral head to form joint allotopia backward.

The upper region of glenoid cavity often suffers deviation, while the lower region is normal, because of the lower scapular development without independent epiphysis, the abnormal scapular shape, the elongation of scapular spine, the irregularity in some acromions, the deviation in glenoid cavity, and the loss of normal oval shape. The upper region presents a convex surface and is closely correlated with coracoid process; the lower region presents a concave surface, which basically remains normal. Under this circumstance, only the lower region of the humeral head is connected with the hypoplastic glenoid cavity, other parts are deformed, the level of coracoid process is outward, and there is a longitudinal and horizontal part. Due to fundus twist, the upper region of scapular spine faces forward and the lower region faces backward. The acromion is also deformed and does not take oblique shape but presents the horizontal position. The scapular body is flat, straight, and deformed, and it is not angulated. This kind of congenital shoulder joint dislocation needs to be differentiated from the dislocation induced by joint spasm, and the latter is bilateral and often accompanied by other deformities.

Another phenomenon is the congenital posterior semiluxation of shoulder joint. Its pathology is joint posterior flabbiness, aplasia of glenoid cavity, and posterior inclination of humeral head, and it is not found until the patient becomes 5–7 years old. Initially, the joint only has the sensation of being hindered, but gradually posterior protrusion is observed. Flexion can make the humeral head protrude backward. In case of extorsion and abduction, this does not happen. In case of intorsion, the relation between the humeral head and glenoid cavity can be changed. The congenital semiluxation is mostly bilateral, and it is more apt to occur at the dominant side. It has been reported that different surgeries are performed to correct this deformity, such as posterior arthrocapsuloplasty or bone block procedure. The efficacy of rotary ostectomy is relatively good. Performing transverse osteotomy at the upper end of the humerus makes the distal end rotate outward by 20°, and steel plate screws are used for fixation.

4.5 Pathogenesis of Upper Limb Longitudinal Formation Disorder

Bin Wang and Feng Ni

In most cases, upper limb longitudinal formation disorder is induced by abnormal development of embryonic limb bud, with the manifestation of hand and upper limb longitudinal development deformity. The limb buds develop along three axes. The development of proximal-distal axis is regulated by apical ectodermal ridge (AER) and mesoderm through growth factors. AER can produce WNT3 and some FGFs (FGF 4, FGF 8, FGF 9, and FGF 17) to maintain the expression of mesodermal FGF10. FGF10 can promote cell proliferation in AER lower areas, which are called progress zone. The mesodermal cells in the progress center are subject to the regulation of information center to decide the final differentiation. The interactions between ectosermic and mesodermal FGF/WNT maintain the development and growth of proximal-distal axis. The development and differentiation of front-rear (radial-ulnar) axis are controlled by the mesodermal posterior zone of polarizing activity (ZPA). ZPA can increase the width of limbs and make them develop toward the rear (ulnar) direction, and it produces effects through generating morphogen-sonic hedgehog (SHH). AER and ZPA are closely associated through the feedback loops and maintain the expression of SHH at the border area of AER distal rear (ulnar) direction during the growth process. The WNT7a generated at the dorsal side of ectoderm regulates the development of limb dorsal-ventral axis. WNT7a makes the mesoderm of the lower layer limb to grow toward the dorsal side through inducing the Lim homeobox transcription factor LMX1B. The deficiency of WNT7a will also lead to disorder of limb ulnar growth and development, indicating that another important role of WNT7a is to maintain the production of ZPA-related SHH. It can be said that SHH plays an important role in limb development and is associated with the development of the proximal-distal axis, front-rear (radial-ulnar) axis, and the dorsal-ventral axis.

AER-related FGF function loss often results in transverse absence, and lack of FGF function may result in longitudinal absence. FGF hypofunction can cause the growth of limbs to slow down and their size to diminish. Although the ulnar growth and proliferation under the action of ZPA are ongoing, the development results are manifested as the radial longitudinal ray absence in the classification of the same deformities. Malformation syndromes of FGF receptor 2 mutations, such as Apert syndrome, Pfeiffer syndrome, or Saethre-Chotzen

syndrome, are characterized by front (radial) joint abnormality and formation of forearm synostosis.

SHH induces the formation of upper limb ulna and hand ulnar phalanx; in addition, SHH is also correlated with the growth of posterior (ulnar) limb. During the development of limbs, the decrease in SHH expression or discontinuation of target signals can slow down the growth of limbs and diminish their size. The development results of SHH absence are manifested as ulnar longitudinal ray absence, and its clinical manifestations vary with the time point, degree, and duration of SHH absence. Additionally, SHH absence can reflectively induce the decrease in FGF expression. Therefore, not only are the length, size, and FGF expression of limbs reduced, but the development of the radial structure especially the thumb is potentially affected, and its clinical manifestation is thumb and ulnar ray absence. The influence of both abnormal gene expression and teratogenic factors on AER and ZPA may induce upper limb longitudinal formation disorder.

4.6 Radial Longitudinal Ray Deficiency

Wei Wang and Bin Wang

4.6.1 Overview

Radial longitudinal deficiency (RLD) refers to the congenital developmental defect covering the radial bones, muscles, tendons, nerves, vessels, etc. of the upper limbs, and it is clinically classified according to the degree of thumb and radial defects. Radial longitudinal ray deficiency is often bilateral and asymmetric, and patients with mild symptoms even have no obvious clinical manifestations [23]. Radial longitudinal ray deficiency often occurs to some congenital malformation syndromes (Table 4.1). Therefore, it is necessary for patients to undergo comprehensive physical examinations and clinical genetic diagnoses.

Good hand surgery and genetic diagnosis are necessary. The pediatric patients with bilateral and serious radial longitudinal ray deficiency will suffer serious limb function disorders due to thumb dysplasia or absence, wrist joint deviation and instability, and upper limb crispation, and it is very difficult for them to independently perform daily actions such as buttoning up, zipping, and keeping personal hygiene. The hand functions of the pediatric patients with radial longitudinal ray deficiency should be detected objectively for many times as early as possible. X-ray examination is very necessary for classification and can objectively display the involvement degree of radial bones, thumbs, and carpal bones (over 8 years). Lumbar vertebral X-ray, renal ultrasound, and cardiac ultrasound examination are of great importance to the diagnosis of the accompanying deformities. The patients with radial longitudinal ray deficiency mostly concomitantly suffer deformities in skeletal musculature, which aggravated the severity of the disease. For patients with Fanconi's anemia, it is necessary to perform genetic detection, and bone marrow transplantation can be performed for the life-threatening pancytopenia. The other accompanying upper limb abnormalities of the patients with radial longitudinal ray deficiency include humeral dysplasia, ulnar-radial proximal bone union, congenital radial capitulum dislocation, and finger rigidity, and rare deformities include metacarpal fusion, symphysodactyly, contralateral radial hyperdactylia, and phocomelia.

Bayne and Klug classified the radial longitudinal ray deficiency into four categories according to the X-ray findings, and James included the patients with normally long radial bones but with thumb and carpal defects in this classification (Table 4.2 and Fig. 4.10).

4.6.2 Incidence

Congenital absence and hypoplasia of radius are often manifested as shortening of hands and forearms, partial or complete deficiency of radial bones, deviation of ulnar bone to

Table 4.1 Malformation syndromes correlated with radial longitudinal ray deficiency

Syndrome	Concomitant deformities	Genetic characteristics
VACTERL syndrome	Vertebral columns, anus, heart, trachea, esophagus, and kidney deformities, radial and limb deformities	Sporadic
Holt-Oram syndrome	Interventricular septal defect, other upper limb deformities	Autosomal dominant inheritance
Radial defects accompanied by thrombocytopenia, TAR	Thrombocytopenia, anemia, radial defect, but thumb existence	Autosomal recessive inheritance
Fanconi's anemia	Pancytopenia, multiple congenital deformities	Autosomal recessive inheritance
Trisomies 13 and 18 (chromosome aberrations)	Multiple deformities	Sporadic
Nager syndrome, Rothmund-Thomson syndrome, IVIC syndrome	Craniofacial deformities	Varying according to syndromes

Originate from Joseph Upton, MD. Management of transverse and longitudinal deficiencies (failure of formation). Plastic Surgery Second Edition. Edited by Stephen J. Mathes. MD. Philadelphia. Vol. 8. 2006:51–137

the radial side, dysplasia or absence of scaphoid bones and trapezium, soft tissue dysplasia, thumb absence or dysplasia, index finger dysplasia, syndactylia or shortening of index fingers and middle fingers, deviation of the hand and wrist to the radial side, and bending of forearm to the radial side. The hand suffering from it is shaped like a hockey stick and is therefore called hockey stick hand or radial club hand (Fig. 4.11). As the hand deviates toward the radial side, the active movements of the entire forearm, hand, thumb, and fingers are seriously limited, and the hand shape and functions are very poor. Radial club hand is a kind of serious composite congenital deformity of upper limbs which is induced by dysplasia of hand and forearm radial longitudinal rays and pathologically manifested as invasion into upper limb preaxial skeletons, muscles, tendons, aponeurosis tissues, joints, ligaments, nerves, and vessels. It almost involves hands, wrists, forearms, shoulder, or other organs, accompanied by deformities in the heart and digestive organs. It falls into the category of radial defect in formation disorder in the classification of congenital deformities of hands and upper limbs. Therefore, it is also called hand with radial deficiency or radial dysplasia.

Table 4.2 Modified Bayne's classification of radial longitudinal ray deficiency

Type	Thumb	Carpal bones	Distal segment of radial bone	Proximal segment of radial bone
Type N	Dysplasia or absence	Normal	Normal	Normal
Type 0	Dysplasia or absence	Dysplasia, absence or fusion	Normal	Normal, ulnar-radial fusion, congenital radial head dislocation
Type 1	Dysplasia or absence	Dysplasia, absence or fusion	Above 2 mm shortening compared with ulnar bones	Normal, ulnar-radial fusion, congenital radial head dislocation
Type 2	Dysplasia or absence	Dysplasia, absence or fusion	Dysplasia	Dysplasia
Type 3	Dysplasia or absence	Dysplasia, absence or fusion	Epiphysis defect	Serious dysplasia
Type 4	Dysplasia or absence	Dysplasia, absence or fusion	Deficiency	Deficiency

Petit reported the world's first case of hand and forearm radial deficiency in 1733. Petit described one newborn case with deformities of upper limbs and established the detailed autopsy materials in hand and forearm radial deficiency, where the records were almost as detailed as the present ones. The pictures and written records on deformities of bilateral upper limbs described by Petit can be found in the book *The Growing Hand* compiled by Gupta (2000). Gruber (1865) reported 14 cases after literature review, Antonelli (1905) reported 114 cases after literature review, and Kato [24] (1924) reported 253 cases after literature review. Radial club hand is a rare kind of congenital upper deformity, and its incidence is 1/100,000 to 1/30,000 of the neonates. Birch-Jensen (1950) made a statistical analysis on four million people and learned that the incidence was 1/55,000 of the live births. Temtamy and McKusick (USA) reported that the incidence of radial club hand among the neonates was 0.03‰, Bod (Hungary) et al. reported that the incidence of

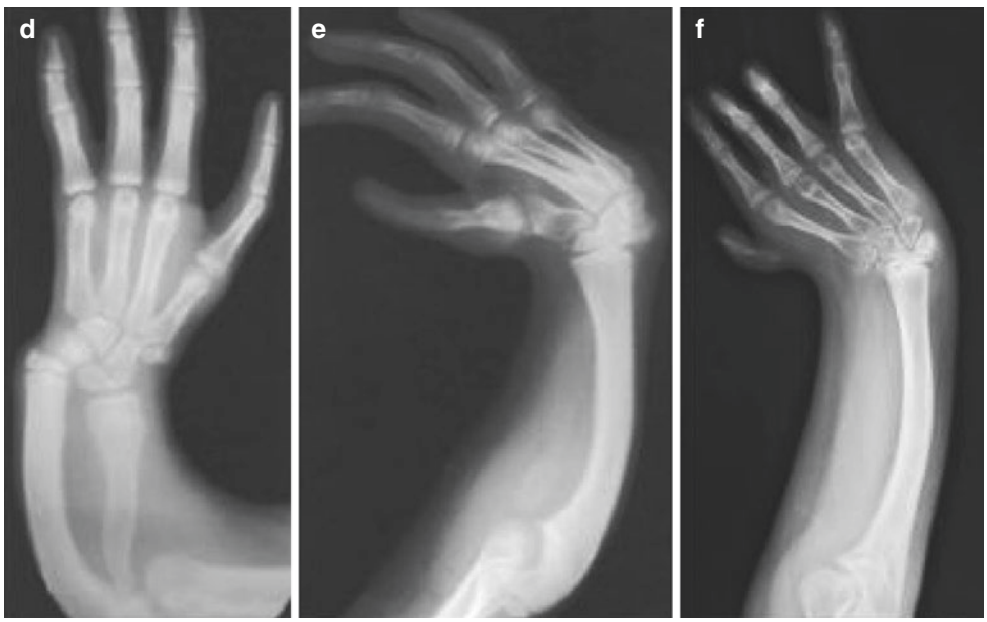


Fig. 4.10 Classification of radial longitudinal ray deficiency. (**a**) Type N, separate thumb dysplasia, normally long radial bones. (**b**) Type 0, radial carpal absence or dysplasia, normally long radial bones, radial tissue tension. (**c**) Type 1, the radial bone is over 2 cm shorter than the ulnar bone, radial deviation of hands. (**d**) Type 2, radial dysplasia, significant shortening. (**e**) Type 3, radial distal deficiency. (**f**) Type 4, complete radial absence

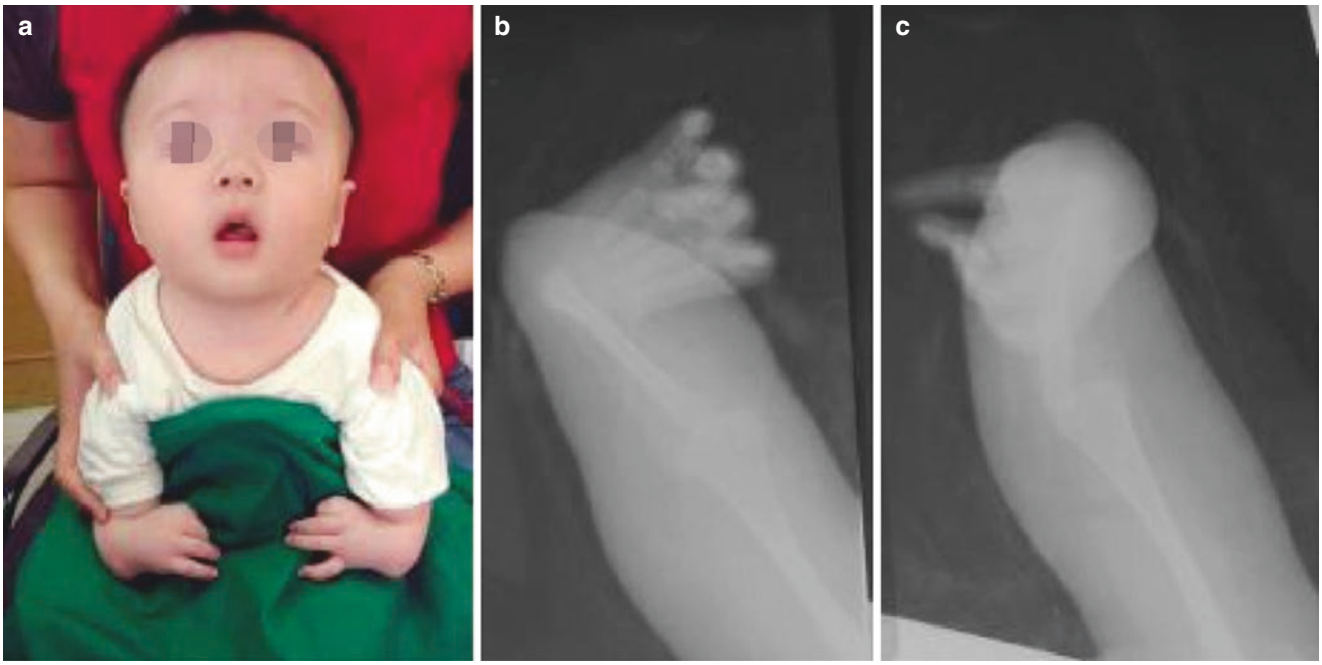


Fig. 4.11 Bilateral radial club hands. (**a**) Hand appearance. (**b**, **c**) X-ray findings

radial club hand was 0.09‰, and Kallen and Tamerek (Sweden) reported that the incidence of radial club hand was 0.08‰. As is reported, male patients with this disease outnumber female patients, and Caucasians have the highest incidence rate. The authors only saw 15–20 cases of radial club hand since 1998, indicating that this disease is very rare in China (no statistics are available before 1998).

4.6.3 Etiological Factors

Etiological factors of radial longitudinal ray dysplasia or radial club hand have been discussed for over 100 years, but still remain unclear until now. Genetic factors have been reported, but still remain unclear, and genetic defect may be an important cause; radiation injury, virus infection, chemical factors, drug, malnutrition, etc. can all cause such deformities. Radial club hand induced by thalidomide has been reported for many times. Saunders (1950) found, during the study, that the ectosermic ridge crest of the chicken's embryonic limb buds will result in the radial dysplasia of chicken wings. As is recorded in the book *The Growing Hand* compiled by Gupta, the administration of busulfan (Myleran), a drug that is used to treat chronic myeloid leukemia, to the mother mice could cause the baby mice to suffer deformities similar to radial longitudinal dysplasia. Gupta (2000) recorded that, during the limb development period at 4–7 weeks of pregnancy, the adverse factors from surroundings acted on the ectosermic ridge crest, which could induce fetal deformities.

4.6.4 Pathological Anatomy

Radial longitudinal dysplasia or radial club hand is pathologically manifested as absence or dysplasia to varying extents of radial bones, radial carpal bones, and thumbs, and the corresponding muscles, tendons, aponeurosis, vessels, nerves, skin, and subcutaneous tissues located at the radial side are manifested as deficits and distortion. The D'Arcangelo's detailed description in the book *The Growing Hand* compiled by Gupta concerning the pathological anatomy of radial club hand is of very high reference value.

4.6.4.1 Bone and Joint Deformities

1. Upper. It may be shorter than the normal one and can often be manifested as superficial deformities in humeral bone, trochlea, and olecranon fossa as well as absence of intertubercular groove of the humerus, coronary sulcus, internal epicondyle, or the end of humeral bones.
2. Elbow. The range of active movement of elbow becomes narrower, and the range of elbow flexion becomes even narrower than that of elbow extension. According to the records of Heikel (1959), the pronation and supination motions of the forearm are absent. Before a child becomes 2 years old, its elbow joint is often rigid in the extension position. After it grows up, the active or passive flexion of its elbow joint can gradually increase to 90°, which may be correlated with the dysplasia of elbow joint structures and the insertions of elbow-flexing muscles.

3. Forearm. The forearm is short, and the length of ulnar bones is only 60% of the healthy side. The end of epiphysis occurs late but closes early. Due to the absence of radial bones, the hand and wrist become displaced from the end of epiphysis of the ulnar bones and glide to the radial margin of the ulnar ends. The backbone of the ulnar bones bends to the radial side, which is correlated with dysplasia or absence of the radial bones. Lamb [25] (1977) once reported 117 cases of radial club hand, only four of which had ulnar bending at birth.

   Fiber primordium is a kind of fibrous tissues and also the remaining structure of radial bone dysplasia or radial partial absence, which is the reason that induces bending between the forearm and the wrist. The incidence of trapezoid, lunar bone, and lentiform bone is relatively low (10%), and the lentiform bone, hamate bone, and trapezoid of most patients are normal.
4. Wrist and carpal bone. Among the patients with complete radial absence or partial absence, 80–100% concomitantly suffer from the absence of scaphoid bone and trapezium. The forearm is short and small, the hand and wrist are located at the radial margin of the end of the ulnar bones, the ulnar and carpal bones constitute the ulnar dislocated ulna-wrist joint, and the backbone of the ulnar bones bends to the radial side (Fig. 4.12).
5. The deformity of metacarpals, thumb, or fingers is the most common among the radial longitudinal ray dysplasia, and it is 100% present. According to the statistics of Heikel (1959), among the complete radial absence, thumb absence accounted for 60%; among the radial dysplasia, thumb absence accounted for 30%, and others were thumb dysplasia or residue. Lamb observed that 86% of his cases suffered from thumb absence. In addition to the absence of the first metacarpal, scaphoid bone and trapezium are also missing in the case of thumb absence or residual thumbs.

Usually, other fingers are normal, the metacarpophalangeal joints of fingers are normal, but the index finger and the proximal end of the second metacarpal may suffer dysplasia, and the index finger takes the shape of pronation or rotation. Among the authors' cases, the pediatric patients once suffered syndactylia of index finger and middle finger (Fig. 4.13).

4.6.4.2 Muscular Deformity

1. Upper arm and major pectoral muscles. The clavicles or ribs of the major pectoral muscles may be absent, and the smaller pectoral muscles are generally normal. The triangular muscles may be normal, but may be fused with brachial muscles or triceps muscles of the arm. The origin of coracobrachial muscles is often fused with the coracoradialis. The biceps muscle of the arm is often abnormal; in case of complete absence of radial bones, the long head of the biceps muscle of the arm is often absent and only ends at the fiber bundles; the short head may be present or fused with the coracobrachial muscles. The brachial muscles can be normal and can present low development or absence; usually their origin is fused with the biceps muscle of the arm without an independent insertion, which emerges with the insertion of the common extensor muscle of fingers. The triceps muscle of the arm is often normal, but can be fused with the surrounding muscles.

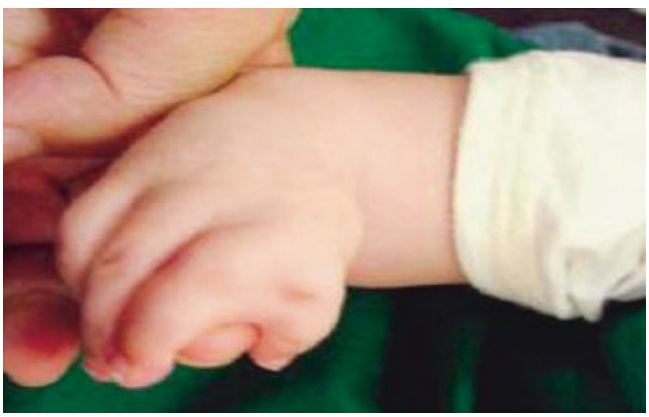


Fig. 4.13 Deformity of radial longitudinal ray dysplasia (thumb and index finger dysplasia and syndactylia of index finger and middle finger)

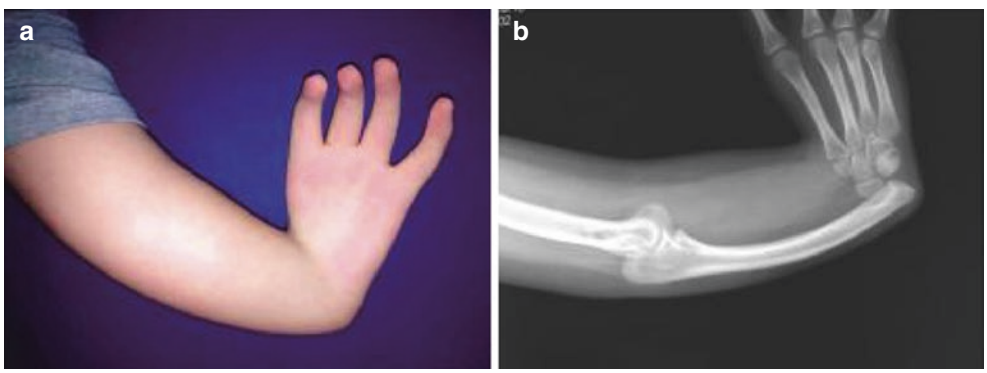


Fig. 4.12 Right radial club hand. (**a**) Hand appearance. (**b**) X-ray findings

2. Forearm muscles. The round pronator muscles are often absent and can be fused with biceps muscle of arm, brachial muscles, radial wrist flexors, or long palmar muscles. In the cases with complete radial absence, the brachioradial muscle is often absent; in case of presence of the brachioradial muscles, dysplasia may occur, or fusion with radial wrist long extensor and radial wrist short extensor can be observed, or the muscular insertions are abnormal, ending at the carpal bones or ulnar bones.
3. Hand-wrist muscles. The long palmar muscles are often absent; in case of presence, their fusion with superficial digital flexor or other flexors can be observed. For the patients with partial or complete absence, radial wrist flexors may be absent. In case of presence of the radial wrist flexors, almost all of them suffer from deformity or are fused with other muscles. Superficial digital flexors are often present, but may be fused with profound digital flexors or suffer atrophy or defects. The origin of the radial bones can be absent, and the tendons to the index fingers are absent. The profound digital flexors are normal, but the finger profound flexor tendons of index finger are often absent, and some reports indicate that the tendon insertions are at the basal part of the middle phalanx. The ulnar flexor muscles of the wrist are almost normal. The quadrate pronator muscle and round pronator muscle are often absent and can only be present at the proximal end of radial bones. The radial wrist long and short extensors can be absent or can be fused with the common extensor muscle of fingers or brachioradial muscles. The common extensor muscle of fingers is often present, but often fused with the radial wrist long extensors and little finger extensors. Some reports indicate that the insertions of the common extensor muscle of fingers are abnormal and end at the basal part of the proximal phalanx. The index finger inherent extensors are absent; in case of presence, the insertions are often abnormal. The ulnar-wrist extensors are often present and normal; in case of abnormality, the insertions are often abnormal or fused with the common extensor muscle of fingers and ulnar-wrist flexors. The lumbrical muscles and interosseous muscles are generally normal, but the first lumbrical muscle and the first dorsal interosseous muscle are often absent. Hypothenar muscles are normal in most cases, and the hand outer muscles including thumb long flexor, thumb long abductor, thumb long extensor, and thumb short extensor are often absent; in case of presence, serious dysplasia can be observed. The hand internal muscles of thumb including thumb short abductor, thumb adductor, thumb short flexor, and thumb opposing muscle are often absent; in case of presence, serious dysplasia can be found. The origin and insertion can be abnormal, and the metacarpal bones of the index finger and middle finger are the abnormal insertions.

The comparison between the cross-sectional structure of normal arms and the cross-sectional structure of arms with radial dysplasia and radial defects is shown in Figs. 4.14, 4.15, 4.16, and 4.17 (thank the foreign peers for plotting the sectional drawing).

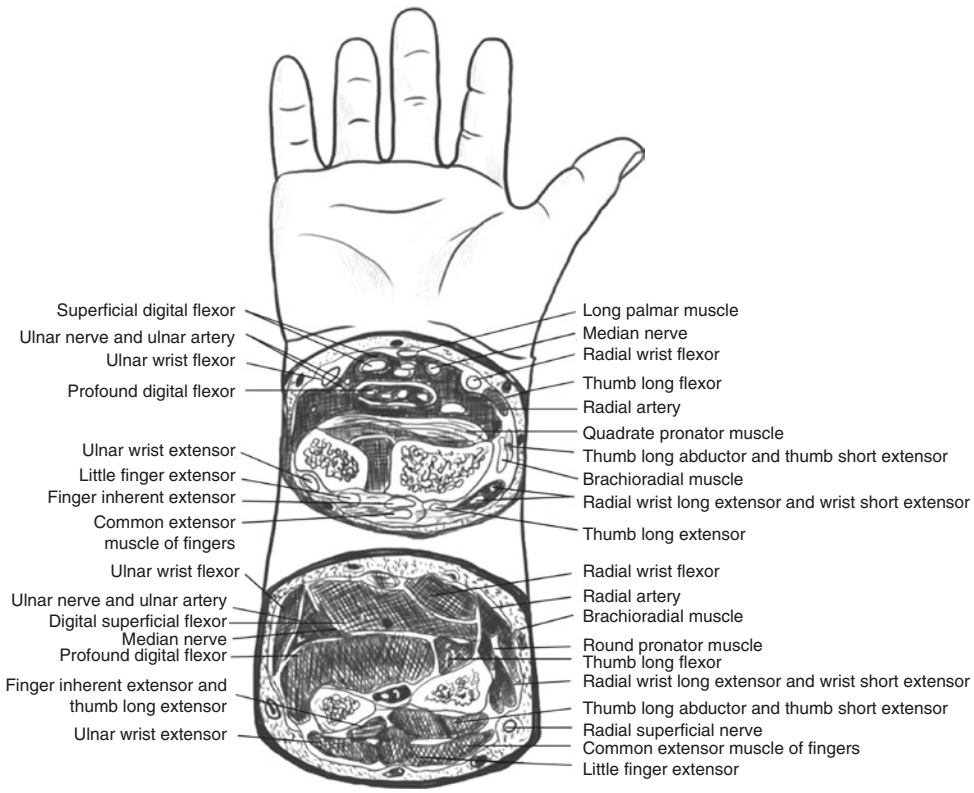


Fig. 4.14 Cross-sectional structure of normal arms

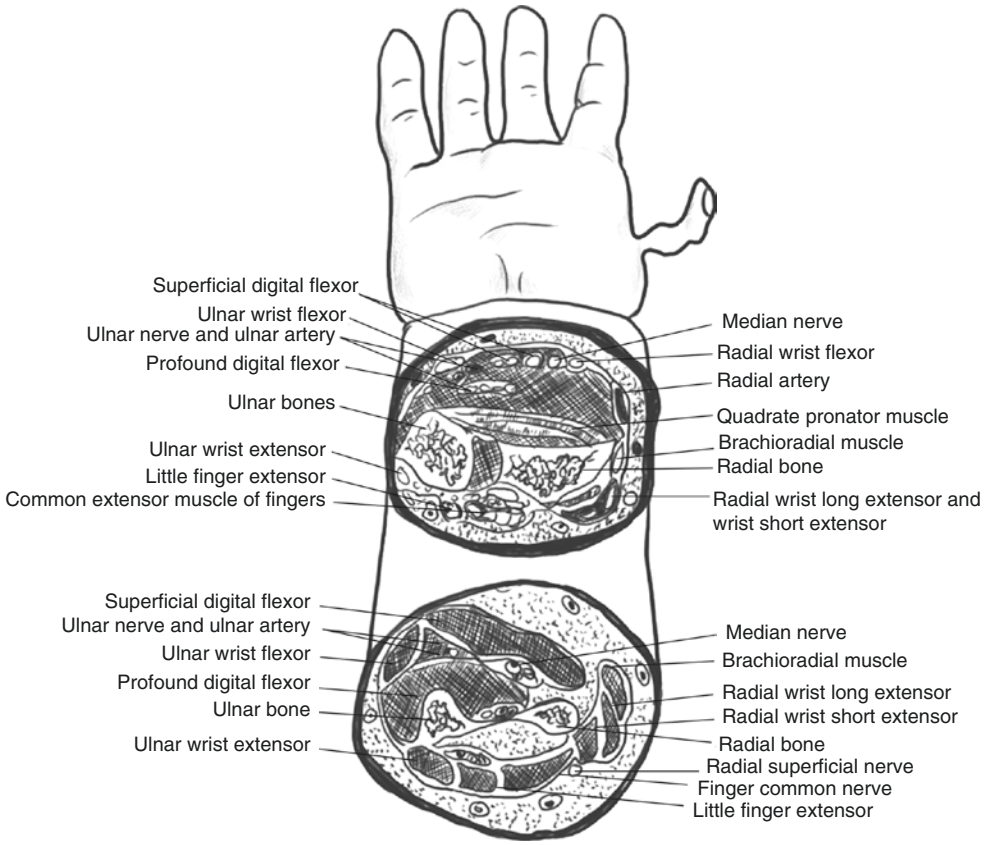


Fig. 4.15 Cross-sectional structure of arms with radial dysplasia

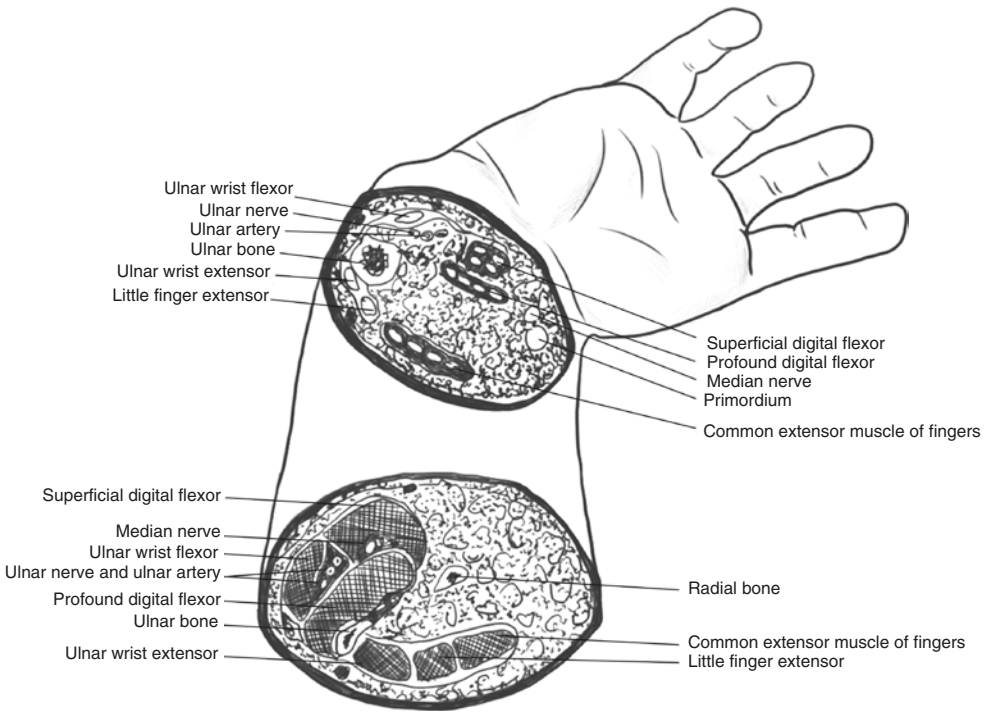


Fig. 4.16 Cross-sectional structure of arms with partial radial defects

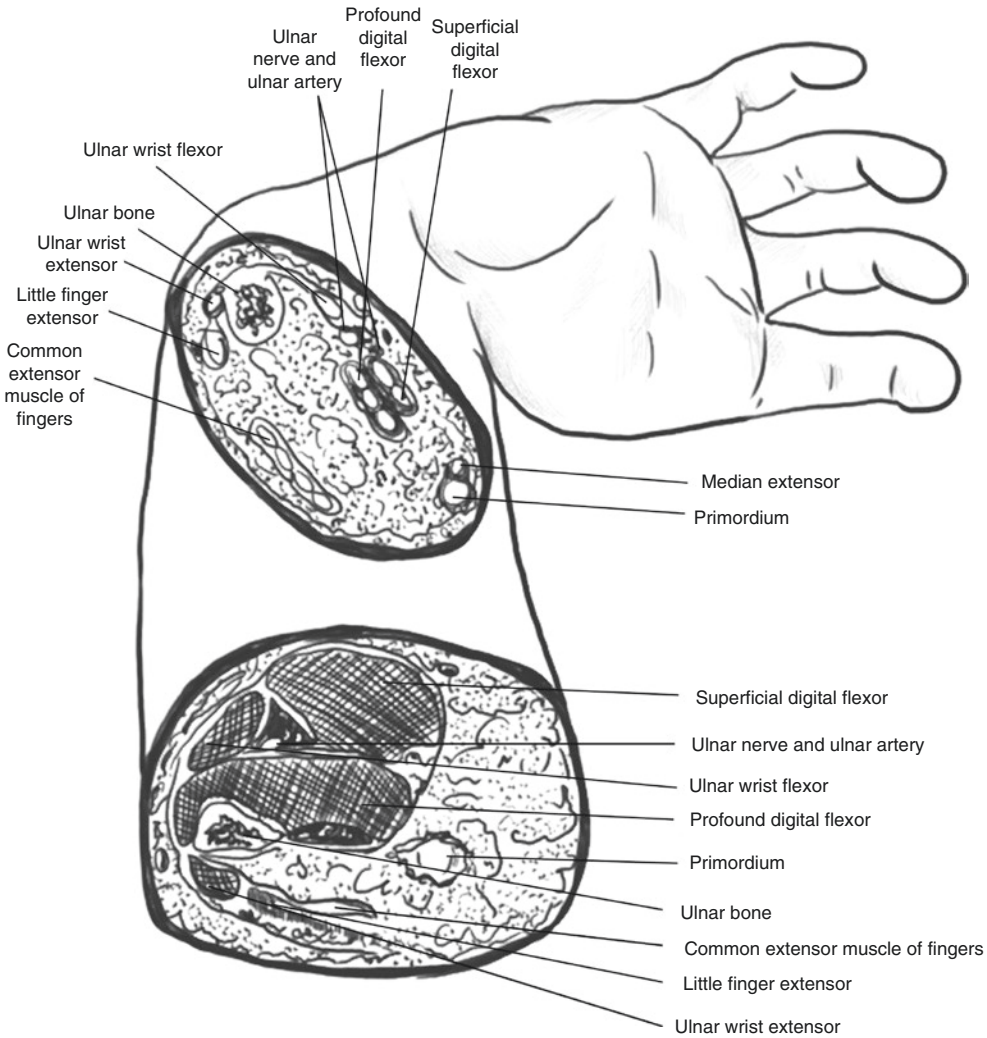


Fig. 4.17 Cross-sectional structure of arms with complete radial defects

4.6.4.3 Nerve Deformities

The deformities of main nerves are normal. The brachial plexuses of most patients described in the literature are normal, and the ulnar nerves are also normal. The musculocutaneous nerves of most patients are absent. The median nerve dominates the front muscles of the upper arm. The radial nerves become the terminal branch at the elbow joint after dominating the triceps muscle of arm, and the superficial branches are absent. The forearm radial and hand sensations are dominated by the median nerve, which coincides with the sensory nerves of the ulnar nerves, called radial-medial nerves. This nerve is thicker than normal one because this contains the radial nerve sensory branch and median nerve, which are found at the end of forearms and the wrist superficial regions.

4.6.4.4 Vascular Deformities

The abnormality in artery system, especially the radial artery, is common. Brachial artery is often present and can be divided into two branches at the upper part of upper arm. Ulnar artery is often normal and may also deviate to the radial side. Radial artery is often absent. Even if it is present, its bore is very small and the pulsation is very weak. Overgrowth of interosseous artery is called median artery, which can replace radial artery and ulnar artery. Deep palmar arch and the radial digital artery radiating to the index finger are often absent. Some performed an angiographic study and found 6 cases of radial artery absence (46%), 5 cases of dysplasia (38%), 10 cases of presence of median artery (77%), 2 cases of superficial palmar arch absence (15%), 12 cases of small deep palmar arch or absence (92%), 5 cases of absence of thumb radial digital artery (38%), and 1 case of absence of index finger radial digital artery (8%).

4.6.5 Clinical Manifestations

4.6.5.1 Forearm Deformities

The clinical manifestations of radial club hand are very characteristic, and especially for the serious radial club hand, its

clinical manifestations are fairly self-explanatory: the affected forearm is short and bent with partial or complete absence of radial bones, the ulnar bone bends to the radial side, the distal end of the ulnar bone is protruding, the carpal joint is completely dislocated toward the ulnar-radial side, and angulation deformity is observed in the longitudinal axis of fingers and ulnar bone. The serious complete radial defect is manifested as arch-form bending of ulnar bones, hand bending toward radial side and almost being parallel to the forearm, and the fingertips pointing to the cubital fossa, taking typical hockey stick shape (Fig. 4.18).

All forearm muscles starting from external epicondyle of the humerus, radial bones, and interosseous membrane as well as the muscles starting from thumb metacarpal bone and phalanx have abnormalities. The round pronator muscles are often absent or are fused with biceps muscle of arm, brachial muscles, radial wrist flexors, or long palmar muscles. However, some reports indicate that round pronator muscle, ulnar-wrist flexor, and radial wrist flexor usually have a good differentiation, but the origin and insertion of radial muscles are abnormal, and these muscles make the hand incline to the radial side. Most of the muscles starting from external epicondyle of humerus have variations. Although brachioradial muscles are present, most of the radial wrist flexors, wrist long extensors, and wrist short extensors are abnormal; therefore, flexion and extension are weak. Thenar muscles are dysplastic or absent.

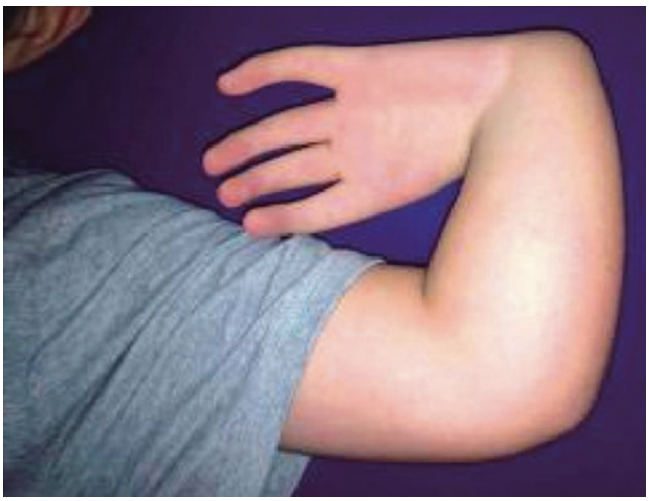


Fig. 4.18 Left radial club hand (the hands take typical hockey stick shape)

The radial skin of the forearm takes web shape. Skin, subcutaneous tissues, fascia, aponeurosis, and tendons are obviously short with anlage tissue, the tension of radial tissues of the hand and forearm is obviously higher than that of the ulnar side, the hand inclines toward the radial side, and the wrist becomes dislocated toward the radial side. The radial club hand is often bilateral, whose incidence is 38–58%. If the deformity is bilateral club hands, the lesion degree and deformity types of bilateral sides are often different.

4.6.5.2 Finger Deformity

The finger deformities of radial club hand are mainly manifested as thumb and index finger deformities. Its corresponding phalanx, metacarpal bones, carpal bones, muscles, and tendons all have deformities, inducing the abnormal shape of the thumb and index finger. Thumb deformity is one major feature of radial club hand. The thumb dysplasia in shape and structure almost covers all types of thumb dysplasia, including fine thumb, short thumb, adducted thumb, abducted thumb, floating thumb, and complete thumb absence. Index finger deformity can be manifested as short index finger and dysplasia.

Radial club hand can be accompanied by syndactylia and polydactylism, but similar reports are rarely seen in literature. The author once encountered a boy with radial club hand. One side was the typical radial club hand; the other side was the duplication of the thumb, accompanied by the radial dysplasia of the left hand, manifested as short radial bone, metacarpal bone, and phalanx neoplasm, with seven metacarpal bones and seven fingers, similar to mirror hand, which is very rare (Fig. 4.19). In addition to preaxial polydactylia, the author once encountered radial club hands with multiple radial deformities.

4.6.5.3 Concomitant Deformities

Forty percent of unilateral radial club hands and seventy-seven percent of bilateral radial club hands are accompanied

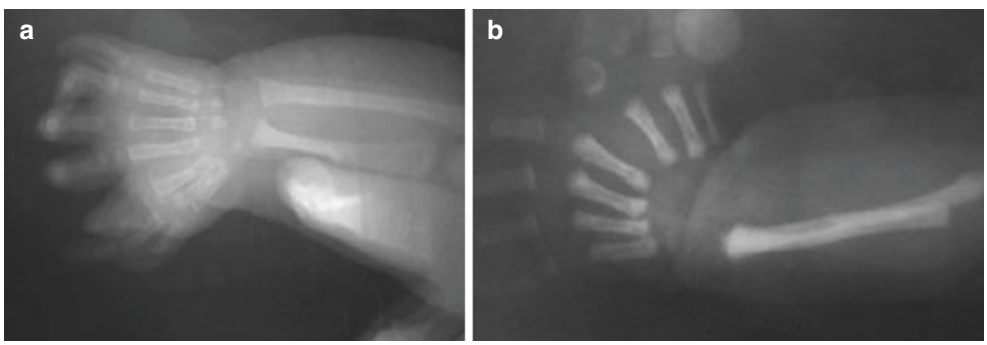


Fig. 4.19 X-ray findings of left radial club hand

by deformities in other organs, such as interventricular septal defect, pulmonary stenosis, gastrointestinal tract malformation, tracheoesophageal fistula, hedratresia, inguinal hernia, and still with microcephaly, decreased intelligence, hydrocephalus, thrombopenia, and other muscular and skeletal deformities. Radial club hand can also be one of the symptoms of congenital malformation syndrome, such as VATER association (e.g., deformity of spinal column, hedratresia, tracheoesophageal fistula, and radial deformities), Holt-Oram syndrome (a kind of chromosomal disorder, manifested as interventricular septal defect and radial defect; and the chromosomal aberration is located at 12 q), and Fanconi's anemia (manifested as pancytopenia).

4.6.6 Classification

The clinical characteristics and lesion degrees of radial longitudinal ray deficiency vary greatly, and some scholars have classified the deformity according to the thumb and radial defect degrees.

4.6.6.1 Michelle's Classification

Michelle [26] (1999) analyzed the 196 limbs with radial longitudinal ray deficiency of 119 patients from 1923 to 1996 and classified the 181 limbs of 104 patients:

1. Type N, normal radial bones, normal carpal bones, thumb dysplasia
2. Type 0, normal radial bones, radial carpal deformity
3. Type I, radial defect for over 2 mm, accompanied by carpal dysplasia
4. Type II, radial dysplasia
5. Type III, radial dysplasia, distal defect
6. Type IV, complete radial deficiency

Eighty-two percent of the patients suffer thumb dysplasia, fusion of proximal ulna-radial joints, and dislocation of radial bones, and 44% are type I.

4.6.6.2 Classification in This Book

This book classifies radial longitudinal ray (preaxial) dysplasia, namely, radial longitudinal ray defect deformity (radial club hand) into five categories:

1. Normal radial bones
   (a) Dysplasia of thumb (functional type)
   (b) Dysplasia of thumb (nonfunctional type)
   (c) Absence of thumb
2. Radial dysplasia (fine but complete radial bones)
   (a) Dysplasia of thumb (functional type)
   (b) Dysplasia of thumb (nonfunctional type)
   (c) Absence of thumb
   (d) Madelung's deformity
   (e) Others
3. Partial radial (distal) absence
   (a) Dysplasia of thumb (functional type)
   (b) Dysplasia of thumb (nonfunctional type)
   (c) Absence of thumb
4. Complete radial absence
   (a) Dysplasia of thumb (functional type)
   (b) Dysplasia of thumb (nonfunctional type)
   (c) Absence of thumb
5. Others
   (a) Dysplasia or absence of thenar eminence
   (b) Dysplasia or absence of extensor
   (c) Dysplasia or absence of flexor

4.6.6.3 Other Classifications

Most scholars classify radial longitudinal ray deficiency into four categories according to the degrees of radial defects (Fig. 4.20).

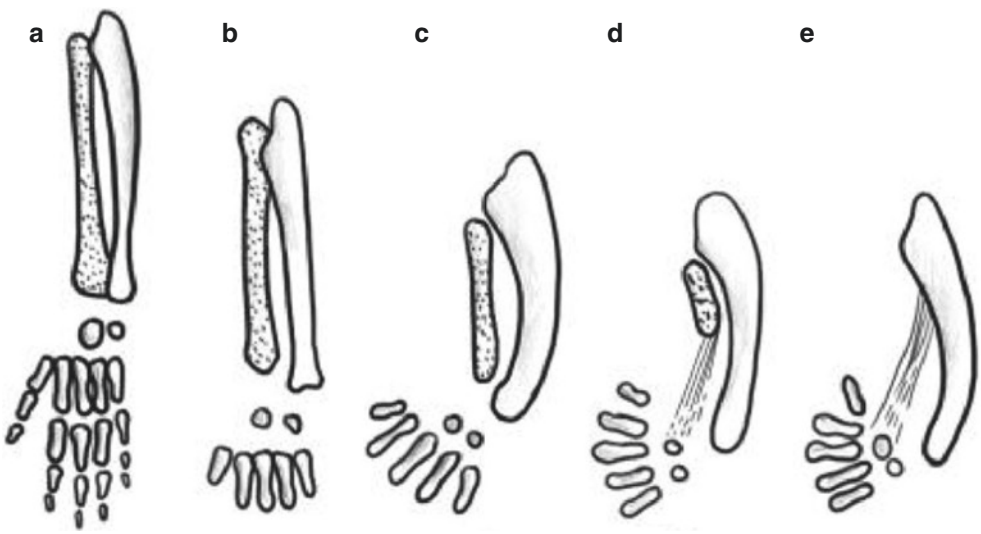


Fig. 4.20 Development status of radial bones with radial longitudinal ray deficiency and its classification. (**a**) Normal. (**b**) Type I. (**c**) Type II. (**d**) Type III. (**e**) Type IV

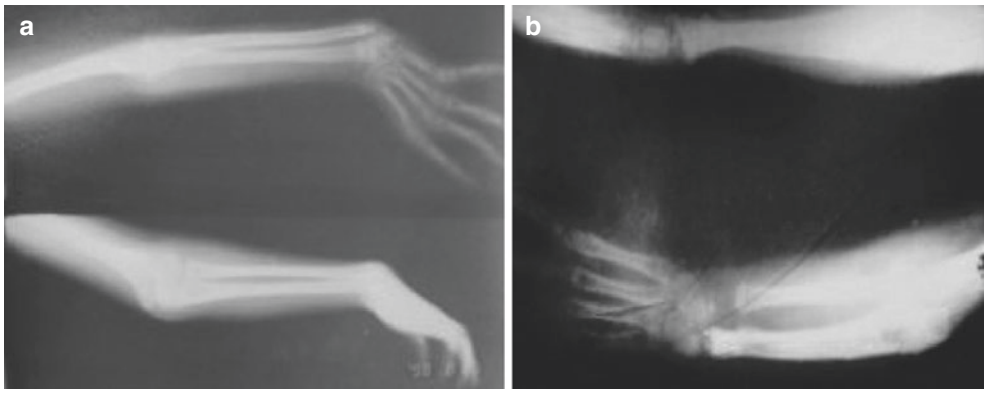


Fig. 4.21 X-ray findings of type I radial club hand

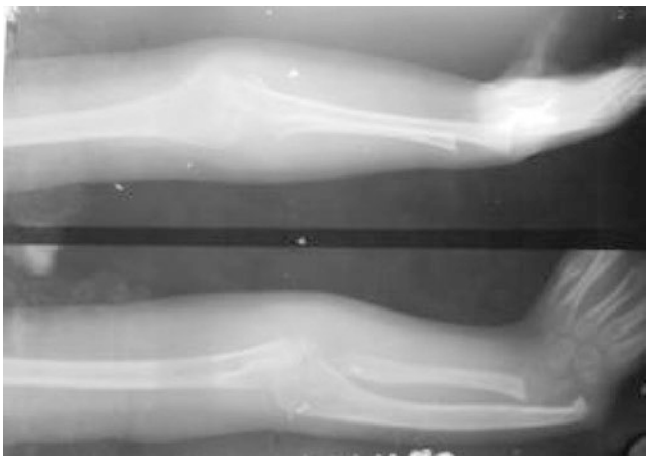


Fig. 4.22 X-ray findings of type II radial club hand

1. Type I (shortening of radial distal end). The appearance of radial distal end of epiphysis is delayed; the growth is deficient, resulting in the shortening of the radial distal end. The radial deformity is relatively small sized; the patient has sufficient support at the wrist joints, so there is no deformity of radial bending. The distal proximal end is normal, the activity of elbow joint is normal, and there are also reports on the dysplasia of radial proximal end or distal end. The thumb has dysplasia to varying extents and the radial carpal bones have dysplasia (Fig. 4.21). Such patients visit a doctor mostly due to thumb deformities.
2. Type II (radial dysplasia). The forearm is short, the epiphysis of radial distal and proximal end is present, but they all suffer dysplasia. The radial bone is bent and fine, and the radial carpal bone and thumb suffer dysplasia to varying extents. The wrist joint becomes dislocated toward the radial side and takes the club shape. The ulnar bone becomes thickened and bends toward the radial side (Fig. 4.22). Such patients should adopt plaster or plastic scaffolds to correct the deformities.
3. Type III (partial absence of radial bone). Partial absence of radial bone can occur to the distal end, middle segment, or distal end of the radial bones, and the one third absence at the distal end or the middle segment is common. Due to the presence of the proximal end of radial bone, the stability of elbow joint is ensured, the distal radial absence leads to the dislocation of hand toward the radial side, and the ulnar bone becomes thickened, bends toward the radial side, and presents typical club hand deformity. Among the eight patients treated by the author, five fall into this category. Such patients often suffer thumb and radial carpal absence or serious dysplasia.
4. Type IV (complete radial absence). Bayne considered that such a deformity was the most common, manifested as short forearm, obvious deviation of the hand toward the radial side, complete loss of support, presence of false joint at the distal end of radial carpal bone and ulnar bone, thickening of ulnar bone and bending toward the radial side, contracture of radial soft tissues of forearm and presentation of web-shaped deformity, and thumb and radial carpal bone absence or serious dysplasia (Fig. 4.23).

   In addition to the above classification, the following deformities should also be included into the classification, and the author classifies them into type V and type VI.
5. Type V The radial bone is close to normal; thumb suffers dysplasia (Fig. 4.24).
6. Type VI Radial longitudinal ray dysplasia, accompanied by preaxial hyperdactylia, similar to the mirror hand or the radial defect hand of polyostotic radial bones.

4.6.7 Treatment

Corresponding therapies are adopted for the radial club hand according to the severity of lesions and the status of deformities, including surgical treatment and nonsurgical treatment, and the entire treatment is called serial treatment or systematic treatment.

Nonsurgical treatment of the radial club hand can be performed immediately after birth and can be postponed to at 2–3 months after birth. Brace is applied to correct hockey stick like deformity of the forearm to reduce ulnar bending and soft tissue contracture.

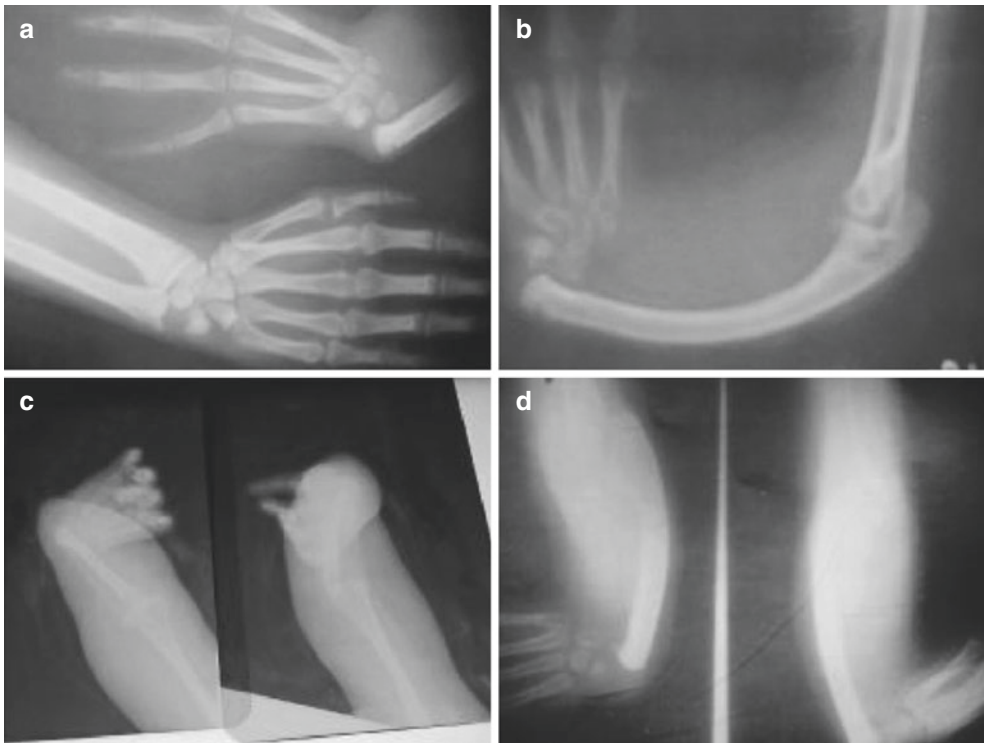


Fig. 4.23 X-ray findings of type IV radial club hand. (**a**). Complete absence of radial bone, short forearm, obvious deviation of the hand toward the radial side, complete loss of support; (**b**) presence of false joint at the distal end of radial carpal bone and ulnar bone, thickening of ulnar bone and bending toward the radial side; (**c**, **d**) contracture of radial soft tissues of forearm and hand presentation of web-shaped deformity, and thumb and radial carpal bone absence

Fig. 4.24 X-ray findings of type V radial club hand (the radial bone is close to normal; thumb suffers dysplasia)

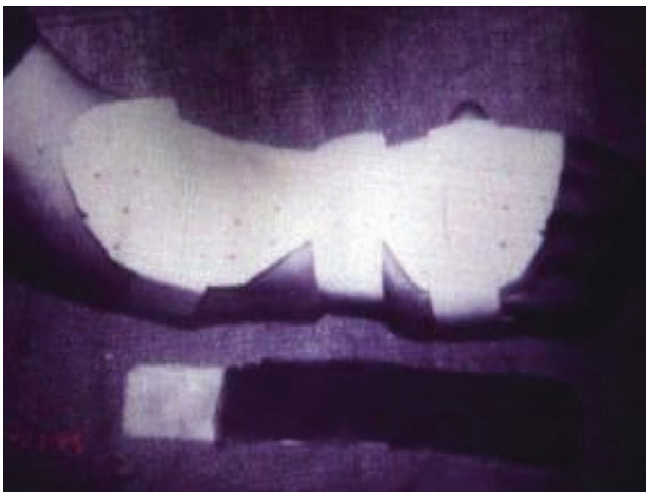


Fig. 4.25 Application of forearm brace to radial club hand

The systematic treatment of radial club hand should be given in different stages: use the brace to correct deformities during the early stage; perform centralized surgery of wrist joint, thumb reconstruction, repair of wrist soft tissue absence, and reconstruction of dynamic tendon at the age of about 2 years old; and subsequently correct ulnar bending.

4.6.7.1 Application of Forearm Brace

The forearm brace is applicable to type II, III, and IV deformities. The early application of brace can not only help correct forearm radial bending and dislocation of wrist joint to the radial side but also elongate the shortened soft tissues of the radial side for preoperative preparations. Forearm brace is also an important postoperative measure to maintain joint stability. The use of corrective brace should last until bone maturation, and after operation the patient can move the affected hand during the daytime and use braces during the night. With the development of bones and the correction of deformities, the braces should be constantly adjusted and replaced (Fig. 4.25).

4.6.7.2 Surgical Treatment

1. Surgical purpose: The deformities in the anatomic structure and shape of radial club hands are serious; therefore, the main purpose of surgical treatment is to improve the functions of hands and forearms. It will be better if surgical treatment can improve both their functions and appearance, but improvement of appearance should not be the primary purpose. Although the radial club hand of adults has an ugly appearance, it has the ability required for daily life and work, so operation is unnecessary [23, 27, 28].
2. Content of surgery. The contents of surgical treatment include correction of wrist joint radial dislocation, correction of the deformity of ulnar bending, correction of radial soft tissue contracture, reconstruction of radial deficiency, reconstruction of thumb defects, or repair of thumb functional defect.
   (a) Type N: For the treatment principles, refer to Chap. 5 "Congenital Thumb Dysplasia."
   (b) Type 0: Only when the angulation is over 20° and the active extension of the wrist to the neutral position is limited can surgery be considered. The surgery should focus on soft tissue release and tendon transposition. During the surgery, it is necessary to release the radial, volar, and dorsal joint capsules and the tense wrist long and short extensor tendons so as to make the wrist joint reach the neutral position. Anastomosis of radial wrist extensor tendon grafting and ulnar-wrist extensor tendon distal segment is utilized to consolidate the active ulnar deviation strength, and the method of suturing between the proximal segment of ulnar-wrist tendon and the dorsal joint capsule of the third metacarpal bone is utilized to consolidate the wrist joint dorsal extension strength. As is reported in the literature, this process can improve the mean ulnar deviation of 46° and dorsal extension of 54°.
   (c) Type I: Except the shortening of radial bone, striction can be found in both radial carpal extending device and the joint capsule. The joint release and tendon transposition are the same as those of type 0. In addition, it is necessary to use a bone lengthener to extend the radial bone so that it can be as long as the ulnar bone.
   (d) Type II: As the radial bone is short and takes arch shape, bone lengthener can be used to lengthen the ulnar bone. Some literatures also report that radial extension can improve functions.
   (e) Type III and type IV: Usually surgery is needed which should proceed from two aspects: first, relieve the striction of radial soft tissues, and place the hand and carpal bone at the distal end of the ulnar bone (centralized surgery); second, apply metatarsophalangeal joint transplantation to reconstruction of the radial bony support of the wrist joint.

   The centralized process includes three main links: ① pre-centralization of soft tissue extension, ② stabilization of carpal bones at the distal end of ulnar bones after rearrangement of them, and ③ tendon transposition and correction of ulnar bending.

   Usually, from the infancy, braces are used to perform pre-centralization of soft tissue extension to correct ulnar angulation and make the carpal joint located at the distal end of the ulnar bones. Brace correction starts after birth and lasts before the centralized surgery. During the recent years, the application of external fixed traction apparatus before centralized surgery for traction for 6–8 weeks becomes an effective measure to improve the striction of radial soft tissues. The pre-centralization of soft tissue traction can not only improve radial deviation and angulation but also improve hand crispation. Recent reports indicate that this can produce correction effect of a radial deviation of 80° and a hand ulnar deviation of 29 mm so that performance of centralized surgery is made easier.

   Correction of thumb dysplastic is usually performed after the centralized surgery.
3. Selection of surgery timing. Both centralized surgery of wrist joint and index finger pollicization for thumb defects can be performed at 6 months after birth.
4. Surgical methods.
   (a) Pre-centralized treatment: This is an important step to correct soft tissue contracture before the centralization or radial formation surgery. First it is necessary to determine the rotation center of deformed wrist joint, and generally capitate bone is considered as the reference standard (Fig. 4.26a). Adjust the track of external bracket to become adapted to the connection of volar-dorsal side and radial-ulnar side (Fig. 4.26b). Make a longitudinal incision at the fifth metacarpal bone, expose the bone cortex, and protect the neurovascular bundle. Reserve sufficient space for traction or pressurization between the screw holders. If the fifth metacarpal bone is excessively short, insert a screw or steel needle into the fourth metacarpal bone for the placement of the distal track (Fig. 4.26c). Insert the first screw or steel needle, with the lateral anterior side perpendicular to the backbone; its diameter should not exceed 30% of that of backbone (Fig. 4.26d). Place brackets, and insert the second screw or steel needle; the length of penetrated contralateral cortex should be shorter than 2 mm to prevent injuries to soft tissues. Screw the distal screw holders, and install the proximal screw holders at the ulnar

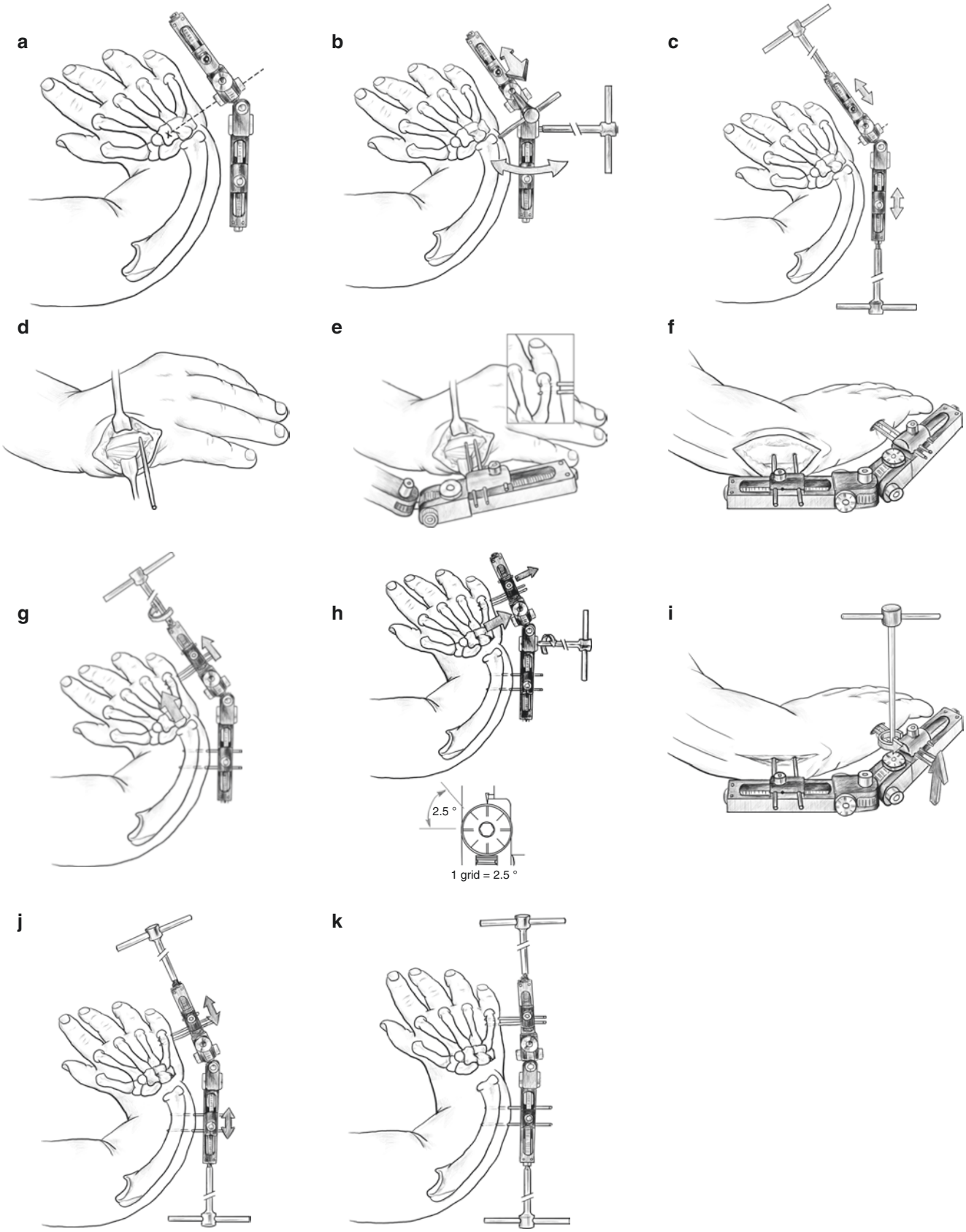


Fig. 4.26 Process of pre-centralized treatment

side (Fig. 4.26e). Make an ulnar longitudinal incision to expose the bone cortex. Insert two screws or steel needles through the same method, and screw the proximal screw holders (Fig. 4.26f).

Traction is started on the next the day after operation. The patient and his/her family members must be fully informed of the pressurization or traction methods as well as the dorsal and radial-ulnar correction plans and adjustment methods, and the patient should receive rechecks once to twice a week. The traction speed is 1 mm a day, and one fourth of the screw diameter is rotated each time and four times a day (Fig. 4.26g).

The radial-ulnar gear is used for ulnar correction, and there is an increment of 1°–3° every day (completed in three to four times). The speed of correction depends on the rigidity and individual difference of the tissues. To prevent ulnar impaction, 2 ~ 4 mm or more traction is required. The center of rotation will vary with ulnar correction; therefore, it is necessary to timely change the pressurization or traction strength to keep it correct (Fig. 4.26h). Volar and dorsal gears can be used for correction of the volar and dorsal side, and there is a daily increment of 1°–3° (Fig. 4.26i). Initially, the scaffold should be located at the radial deviation position to become adapted to deformities; with the increase of ulnar deviation, the distance from the ulnar distal end to the carpal center will change, and the tracks can be adjusted to become accommodated to the changes in the rotation centers (Fig. 4.26j). Continue the correction until the wrist joint experiences an excessive traction of 2–4 mm, and the radial-ulnar arrangement reaches the targets (Fig. 4.26k).

(b) Wrist joint ulnar centralized surgery: This is an important surgery for the correction of club hand deformity, and the purpose is to correct wrist joint dislocation and ulnar-wrist false joint deformity:

- Incision: Some scholars abroad selected the dorsal side of wrist joint to make a horizontal wedge-shaped incision at the prominent part of ulnar bone, the exposure area of this incision is wide, and excessive skin at the ulnar head can be resected at the same time. The author selected the dorsal side of wrist joint to make an S-shaped longitudinal incision, the wrist joint exposure of this incision is good, it is unnecessary to resect the excessive skin at the ulnar head, and adjustments can be made after the operation. The adoption of this incision facilitates repair of radial skin deficit, can make it possible to perform index finger pollicization, reduces vascular injuries, and does not affect the blood supply of the distal grafted skin flap.
- Exposure and translocation of ulnar head: Incise the skin, expose the ulnar extensor muscle of wrist and ulnar flexor muscle of the wrist, protect the cephalic vein and the dorsal carpal branch of ulnar nerves, and expose the distal end of carpale and ulnar head as well as the radial joint capsule. As the ulnar head goes beyond the wrist joint, shape the web-shaped skin with the radial contracture at the forearm into a Z form, or make a traverse or wedge-shaped incision at different levels of skin and forearm deep fascia layers, and perform V-Y reshaping at the radial skin defect areas. After the skin deficit is addressed, incise the joint capsule and expose the ulnar head.
- Ulnar centralization: Cut the ulnar head into the anteroposterior wedge shape, resect partial or all lunar bones, form wedge-shaped space in the center of wrist joint, insert the ulnar bones into the site of lunar bone, and fix them with two Kirschner wire, one of which passes through the third metacarpal bone and capitate bone into the ulnar bones and the other obliquely passes through the carpal bone and ulnar bone. Some scholars abroad also repair the ligaments around the joints. Six to eight weeks after operation, remove the Kirschner silk and perform functional training. After the correction of wrist joint dislocation, use a brace for maintenance in order to prevent the recurrence of club hand deformity until epiphysis growth is completed. Steel plate screws can also be fixed at the functional position of wrist joints. Watson (1984) did not remove any carpal bone when performing wrist joint centralized surgery. Our experience is, for patients of types III and IV, if the carpal bone is not resected and the ulnar bone is not shortened, it is not easy to perform ulnar centralized surgery because the ulnar bone is relatively long. Only in case of repair of radial skin deficit and the extension of the radial tendons, the carpal bone may not be resected.

(c) Ulnar radialization: Buck-Gramcko [29] proposed the concept of radialization in 1990. The method is to dissociate carpale and ulnar bones, correct the dislocation of wrist joint, make the ulnar bones located at the proximal end of carpal bone, and pass the Kirschner silk through the second metacarpal bone for fixation to make the dislocation of ulna-wrist joint hypercorrected; in order to make them more stable, shorten the flaccid ulnar extensor muscle of the wrist or make its insertion move forward. Radial extensor muscle tendon of the wrist and radial flexor muscle tendon of the wrist are also grafted to the insertion of ulnar extensor muscle tendon of the wrist to strengthen the dorsal extension and correct the strength of radial deviation to balance the muscular strength after

deformity. After follow-up visits of 23 patients (the longest duration was 23 months), the efficacy on all of them turns out good.

(d) Correction of soft tissue contracture: As for the mild cases, it is only necessary to perform Z forming and tendon extension; as for the severe cases, local or distal skin flap grafting or skin grafting for repair can be considered. However, considering that the surgery range of such patients is wide, the operation is relatively complicated. Currently, free skin flaps are rarely used for the grafting and repair of skin deficit to reduce surgery time.

(e) Reconstruction of radial supporting tissues: As for type II and III deformities, due to radial defect, fibular free grafting can be adopted, and fibular bones can carry one piece of skin flap to repair the radial skin defect while repairing the radial bone defect. This is a relatively complicated surgery. Before the operation, the position and shape of grafted bones and fibular skin flaps, their mutual relation, the vessel pedicle position, and the vascular anastomosis position of the recipient site should be elaborately designed so that the effect of both repair of radial bone defect and repair of radial skin deficit can be produced. After ulnar reconstruction and correction of dislocation of ulna-wrist joint, sometimes Kirschner silk cannot well serve fixation functions. In this case, it is necessary to mount the wrist joints to the functional position through steel plate screws.

The author once used fibular skin flaps to perform free grafting to repair the radial skin defect and at the same time performed radial bone reconstruction without resecting the carpal bones to perform ulna-wrist joint reduction and thumb reconstruction via index finger pollicization. The above surgeries were all completed in one phase (Fig. 4.27). However, long-term follow-up visits are lacked.

As for type II and IV deformities, metatarsophalangeal joint grafting can be utilized to reconstruct wrist joint radial bone support (Fig. 4.28). This method was first proposed by Vilkki, namely, fusion between the proximal end of the phalanx and the basal part of the second metacarpal bone, fusion between the proximal end of metatarsal bone and the distal end of ulnar bone, and performance of free metatarsophalangeal joint grafting during soft tissue traction. Although the early results are good, there is no follow-up visit for the long-term results.

(f) Correction of ulnar bending: It can be finished jointly with the wrist joint ulnar centralization and can also be performed in different stages. Make a longitudinal S-shaped incision at the ulnar side and ulnar middle segment bending areas of the dorsal surface of the

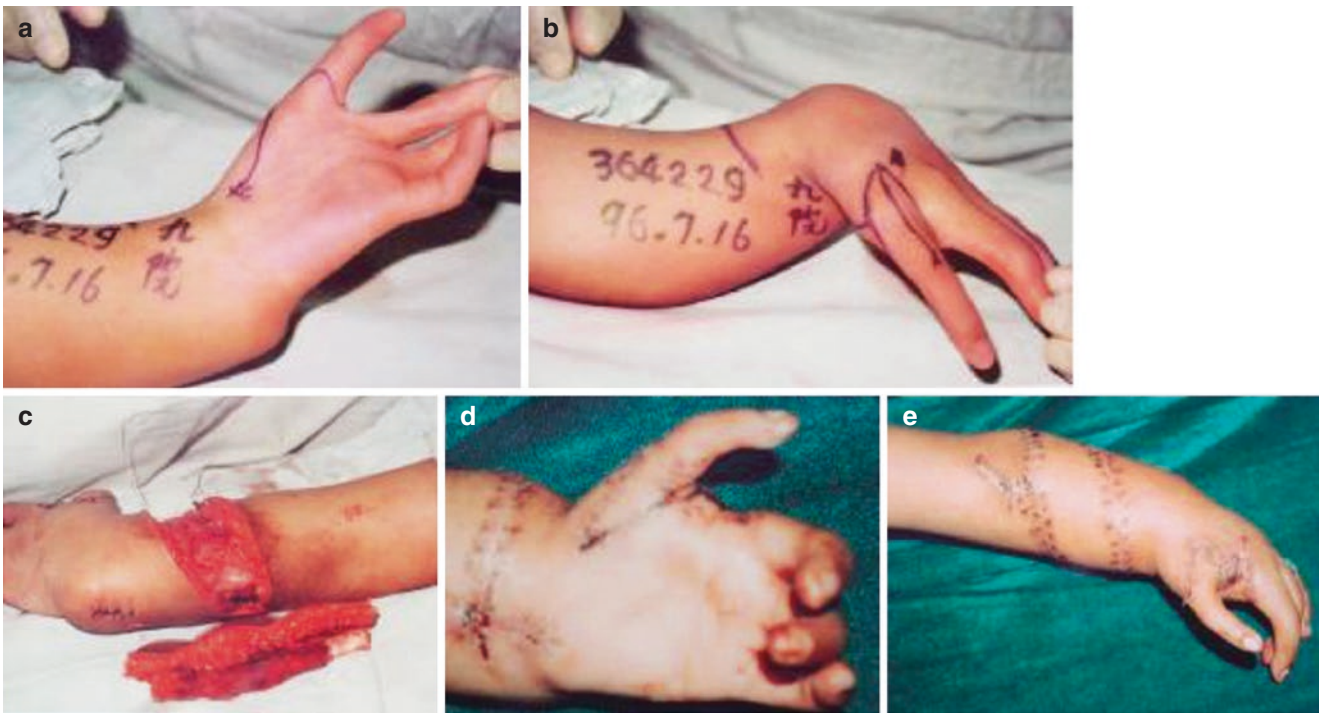


Fig. 4.27 Utilization of fibular skin flaps to perform free grafting for the treatment of radial club hand. (**a**, **b**) Design of incisions for index finger pollicization (**a** is for type I and **b** is for type IV). (**c**) Preparation of vascular anastomosis during the free grafting using fibular skin flaps for radial bone reconstruction (**d**). (**e**) 2 weeks after operation

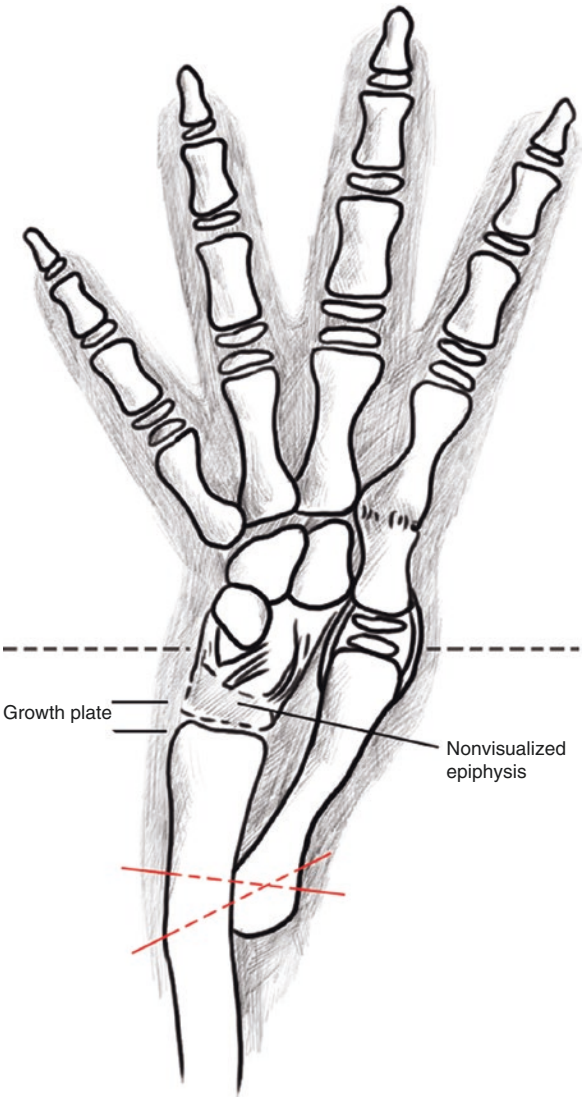


Fig. 4.28 Metatarsophalangeal joint grafting can be utilized to reconstruct wrist joint radial bone support

forearm, expose the dorsal surface of the ulnar bones, and perform the wedge-shaped osteotomy to correct the deformity of ulnar bending (Fig. 4.29). This is a surgery which turns out effective in correcting bending deformity.

Typical cases: A 4 year old boy, type III right radial club hand, IV degree absence of the right thumb; in September 1988, the first ulnar centralized surgery was performed, and 4 months thereafter, index finger pollicization was performed.

(g) Bone lengthening: For the type II and III radial dysplasia, radial bone lengthening can be adopted, that is, self-made lengthener can be used to correct deformities (Fig. 4.30). This is also an optional method to correct radial dysplasia and shortening.

(h) Repair and reconstruction of thumb defect and deformity: After the wrist and forearm deformity is corrected, or during the correction, reconstruction of thumb defect or correction of thumb deformity can be performed. The first choice for thumb reconstruction is index finger pollicization, and the details of the method are shown in Chap. 5 "Congenital Thumb Dysplasia."

(i) Surgical design of the author: The surgery performed by the author for the treatment of radial club hand was mainly ulnar centralized surgery, thumb reconstruction, and treatment of radial bone shortening, and if necessary ulnar bone bending should be corrected.

- Ulnar centralized surgery is the main surgery that corrects the radial dislocation of wrist joint in the radial club hand deformity. Adopt the S-shaped incision at the dorsal side of the wrist at the lower end of ulnar bone, and the wide exposure can facilitate the design of grafted skin flaps at the dorsal side of the wrist. Incise the skin, expose the ulnar extensor muscle of the wrist and the ulnar flexor muscle of the wrist, and protect the dorsal carpal branch of the ulnar nerves. Expose the distal end of the ulnar head of the wrist joint, and cut the ulnar head into the posteroanterior wedge shape. Cut the lunar bone, insert the ulnar bones into the site of lunar bone, and fix them with two Kirschner silk, one of which passes through the third metacarpal bone and enters into the ulnar bones via capitate bone and the other obliquely passes through the metacarpal bone and enters into ulnar bone via carpal bone. Six to eight weeks after the operation, pull off the Kirschner silk, and use external scaffolds for braking during the night. After the ulnar centralized surgery, the wrist joint can still have partial activities. If steel plate screws are used for fixation, the wrist joint should be mounted to the functional positions.
- All the author's patients have undergone the pollicization of the index finger or the fingers with a relatively good radial development, and the details are shown in Chap. 5.
- One pediatric patient in this group used the free fibular skin flaps for grafting. The fibular bone was 8 cm long with skin flaps; fibular end scaffold wrist joint. The free proximal end was placed on the ulna-humeral joint plane, joint capsule remission at the ulna-wrist joint was performed, the ulna-wrist joint dislocation was corrected, the skeletal flaps were used to repair the radial skin deficit areas of the wrist joint, and two Kirschner silks were used to fix the ulna-wrist joint and reconstructed radiocarpal joint for 6 weeks.
- If necessary, correct the ulnar bending.

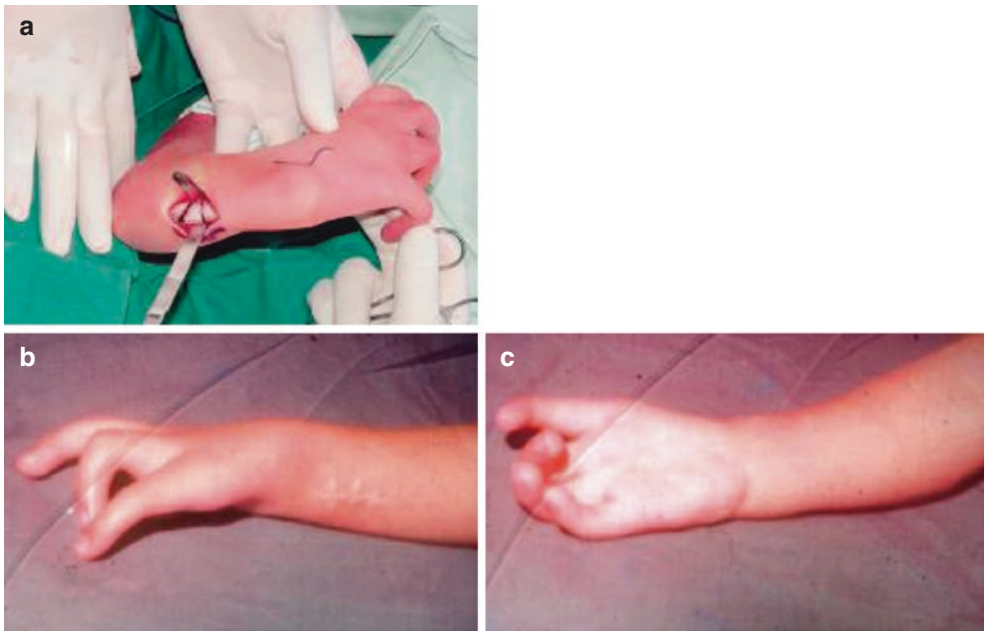


Fig. 4.29 Correction of ulnar bending of radial club hand. (**a**) Ulnar middle segment osteotomy. (**b**) Correction of ulna-wrist dislocation. (**c**) After the completion of the first-phase surgery

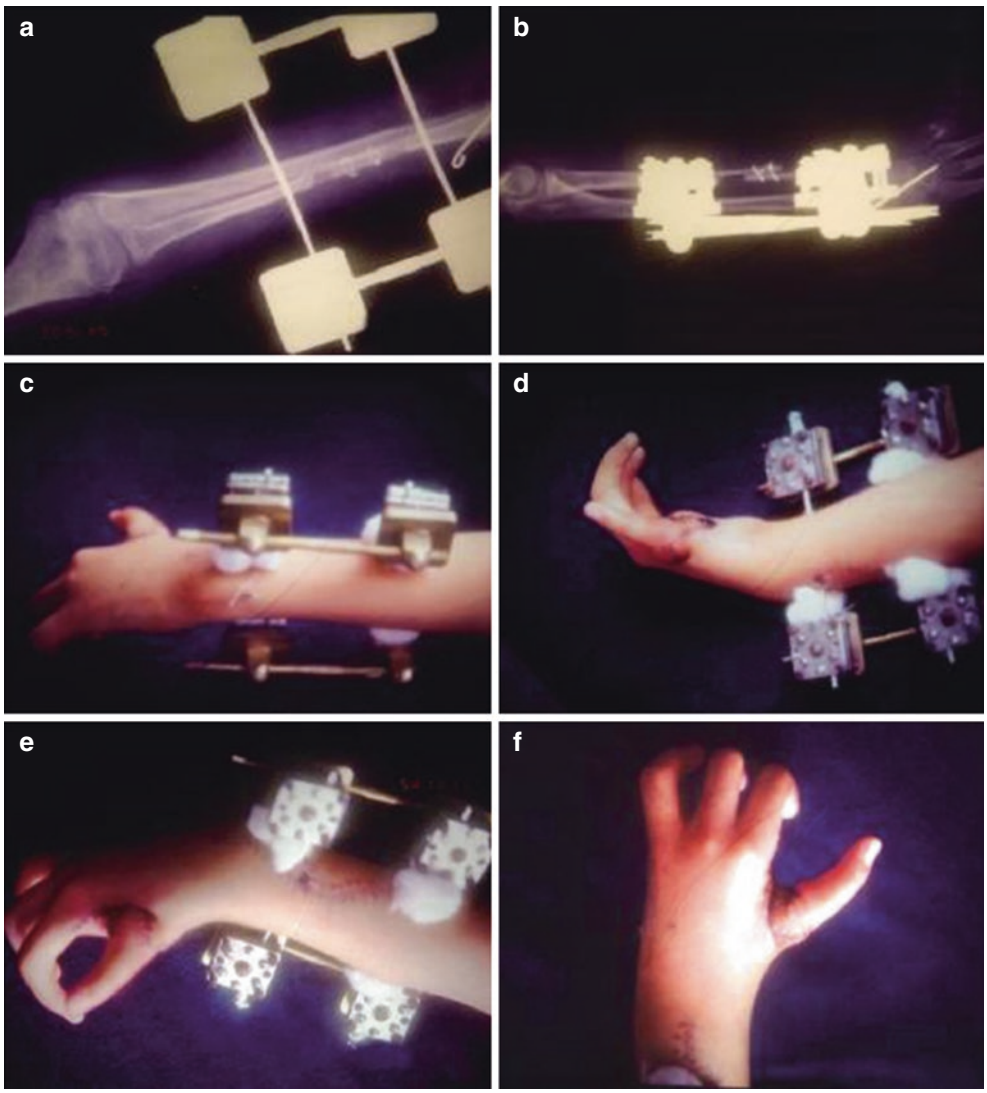


Fig. 4.30 Treatment of left type II radial club hand and thumb absence. (**a**–**e**) First mount bone lengtheners to the radial bones to lengthen the radial bones. (**f**) Then perform index finger pollicization for thumb reconstruction

4.7 Central Longitudinal Ray Deficiency

Laijin Lu, Xu Gong, Fei Tian, and Bo Chen

Hand central longitudinal ray deficiency, namely, cleft hand, refers to the presence of the dysplasia or absence of central longitudinal ray fingers, inducing obvious separation of bilateral fingers and palms. The characteristic manifestations are: the center of the hand presents V-shaped defect, the entire hand is divided into the radial side and ulnar side, also one-finger or multi-finger congenital absence exists, and in terms of naming, the use of the offensive words such as "crab-shaped hand" and "lobster claw" should be avoided. Cleft hand is relatively rare clinically. Rogala et al. reported that its incidence rate was 0.4/10,000 live births, while Birch-Jensen reported that it was only 0.14/10,000; Flat reported that cleft hand only accounted for 3.9% among the 2758 cases of upper limb congenital deformities. About 70% of the central longitudinal ray deficiency is autosomal dominant inheritance, and its etiology is considered to be wedge-shaped defect of ectosermic top of limb buds. The disease can occur to one side or two sides and can also involve the feet. Barsky classified the cleft hand into typical and atypical cleft hands. Typical cleft hand is manifested as congenital ectrodactyly, and atypical cleft hand is manifested as symbrachydactyly [30, 31]. However, Buck-Gramcko et al. consider that atypical cleft hand falls into the category of transverse ray deficiency was a type of hypoplasia rather than central longitudinal ray deficiency.

4.7.1 Clinical Manifestations

Due to the differences in the structures and degrees of finger and palm defects induced by central longitudinal ray deficiency, the forms of their clinical manifestations are different. For the in less severe cases patients, the manifestation is only the defect at the third and fourth metacarpal bone transverse ligament, widening the space, and the deepened third fingerweb possibly concomitantly with dysplasia of middle finger; for the relatively severe patients, the manifestation is absence of middle finger and partial or full absence of the third metacarpal bone; for the severer patients, due to the absence of index finger and middle finger, the manifestation is three-finger cleft hand, or the manifestation can also be normal thumb but absence of the index finger and middle finger with dysplasia of ring finger and little finger [31–33].

Most of central longitudinal ray deficiency is accompanied by other deformities, such as partial and even full syndactylia, flexion contracture and declination of proximal interphalangeal joint, and fusion of phalanx and metacarpal bone. The central longitudinal ray deficiency can also involve two feet, which is called cleft foot. As is reported, it can also be accompanied by congenital heart disease, hedratresia, anonychia, cataract, etc. As is reported by Flatt, the main musculoskeletal deformities accompanying hand deformity include clavicle dysplasia or false joint, absence of major pectoral muscle, shortening of the humerus, elbow joint fusion, forearm shortening, ulnar absence, ulna-radial fusion, hip joint dislocation, femur shortening, whirbone dysplasia, tibia absence, deformed foot, calcaneus splayfoot, and talipes varus [34, 35].

The X-ray of hand central longitudinal ray deficiency varies greatly in different patients, the common one is appearance of transverse bone, and triangular phalanx occasionally appears. It is also possible that two metacarpal bones support one phalanx or one furcal metacarpal bone supports two phalanx. In relatively older children, carpal fusion may occur [33, 34].

Frequent hand movements of patients with typical central longitudinal ray deficiency are normal; even patients with atypical deficiency have certain gripping and grasping capabilities. Pediatric patients often hide hands into the pockets to avoid drawing attention, but as they grow older, they will do this less and less [35].

4.7.2 Classification

Barsky classifies the disease into typical and atypical ones according to the clinical manifestation of the central longitudinal ray deficiency [31]. The central part of the hands of typical patients presents V-shaped cleft, which extends to the palm. The middle finger and the ring finger are often accompanied by dysplasia to varying degrees, and deformity of two-finger syndactylia can be commonly found at the ulnar and radial sides. Transverse phalanx is common, making the bilateral fingers separated so that they cannot be drawn close to each other in a passive way. In most cases, both hands suffer deformities, both feet can be involved, and the patients often have a family history (Fig. 4.31). If the V-shaped cleft at the center of palm is present at birth, the middle fingers are often completely absent, the interdigital space at bilateral sides of the palmar cleft has weds to varying extents, often inducing thumb adduction and contracture, and similar foot deformities often appear.

The hand of the atypical patients takes U shape, the index finger, middle finger, and ring finger are absent, the metacarpal bones are partially or fully absent, and the hand only has thumb and little finger which possibly suffers dysplasia; for the most severe patients, the manifestation can be complete absence of all other fingers than the little finger (Fig. 4.32). This deformity is often single without the occurrence of foot deformity, and it is non-hereditary.

Experts such as Flatt and Buck-Gramcko consider that the typical and atypical central longitudinal ray deficiencies are different in nature not only in appearance but also in

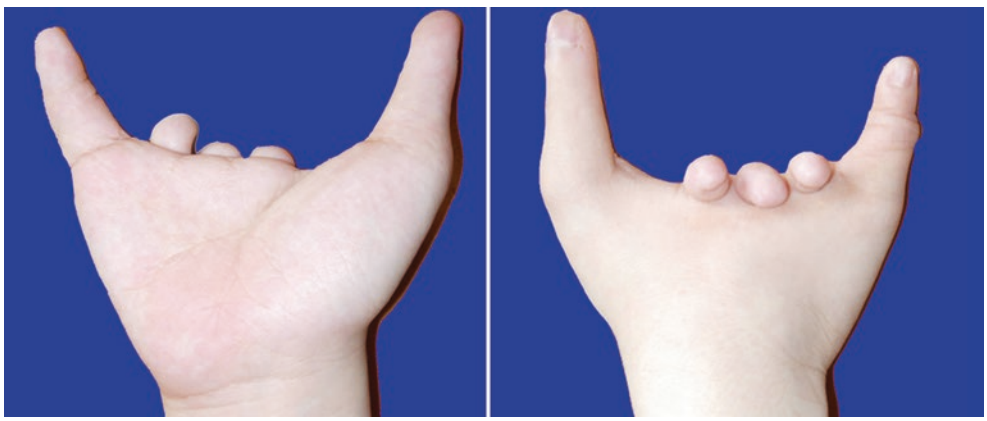
Fig. 4.31 Typical hand central longitudinal ray deficiency and the cleft hand takes V shape

Table 4.3 Manske and Halikis (1995) classification of cleft hand

Type	Characteristics
Type I	Normal thumb web: middle finger or central ray defect, presence of slit in the palm
Type IIA	Mildly stenotic thumb web: central ray defect, presence of slit in the palm
Type IIB	Severely stenotic thumb web: the spaces between the first and second interosseous metacarpals become narrow or have bony fusion
Type III	Thumb web absence: syndactylia of thumb and index finger, middle finger defect
Type IV	Fusion of thumb web and hand slit: absence of index finger and fusion of thumb web into the hand slit
Type V	Thumb web absence: absence of thumb, index finger, and middle finger, and only the ulnar ray fingers remain

Originate from Manske, P.R. and Halikis, M.N. (1995) Surgical Classification of Central Deficiency According to the Thumb Web. Journal of Hand Surgery, 20, 687–697

Fig. 4.32 The atypical hand central longitudinal ray deficiency; the cleft hand takes U shape, and the index finger, middle finger, and ring finger are absent



formation mechanism, so the patients with central longitudinal ray deficiency should refer to the typical ones. The atypical deficiency falls into the category of transverse ray defect, which is a kind of hypoplasia rather than central longitudinal ray deficiency [31, 32].

Clinically, the visits by the patients with cleft hand outnumber the visits by the patients pursuing good hand appearance, and the hand functions are not affected in most cases; for the patients with severe conditions, the main reason for the occurrence of functional disorder is the stenotic or the absent thumb web. In 1995, Manske and Halikis classified the disease into five types according to the thumb web shape of typical cleft hands (Table 4.3) which is used to offer guidance for clinical treatment [33].

1. Type I. Normal thumb web; the middle finger or central ray suffers defects to varying extents, and there exist slits in the palm (Fig. 4.33).
2. Type IIA. Mildly stenotic thumb web, present of defects to varying extents in central ray, and presence of slit to varying extents in the palm (Fig. 4.34).
3. Type IIB. The thumb web is severely stenotic. The X-ray indicates that the first and second interosseous metacarpal spaces become narrow or the bony fusion appears, but the

Fig. 4.33 Type I cleft hand

bony structures of index finger and thumb are not fused (Fig. 4.35).

4. Type III. The thumb and index finger form the syndactylia accompanied by middle finger defect, the thumb web disappears completely, and slits are formed in the center of the palm (Fig. 4.36).
5. Type IV. Absence of index finger, fusion between thumb web and palm slit, and formation of an expanded thumb web (Fig. 4.37).
6. Type V. The thumb, index finger, and middle finger are all absent, only the ulnar ray fingers remain, and the thumb web is completely absent (Fig. 4.38).

4.7.3 Pathogenesis

The etiological factor of hand central longitudinal ray deficiency has not been clear yet, and the majority are sporadic cases. The typical ones are inhibited as autosomal dominant trait, but the penetrance is often incomplete. Maisels [36] once proposed the centripetal inhibition theory, that is, the deformity is simple slit in case of mild inhibition, without obvious tissue defect; in case of aggravation of inhibition degree, absence of middle finger first appears, and subsequently absence of radial fingers appears; in the most severe cases, all fingers are absent (Fig. 4.39). According to Muller [37], cleft hand and web-shaped finger have different

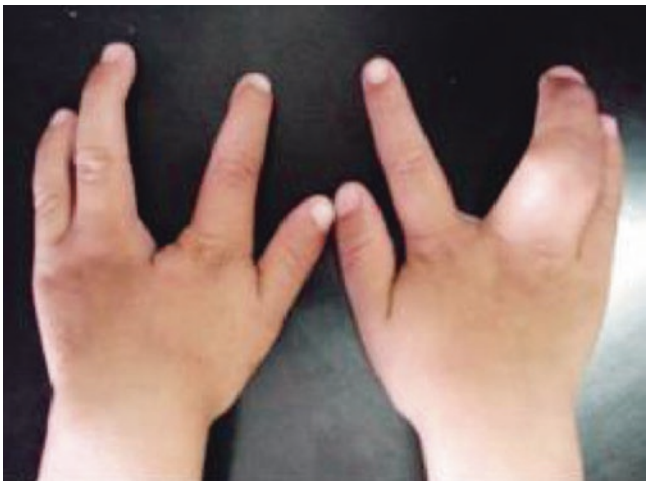


Fig. 4.34 Type IIA cleft hand

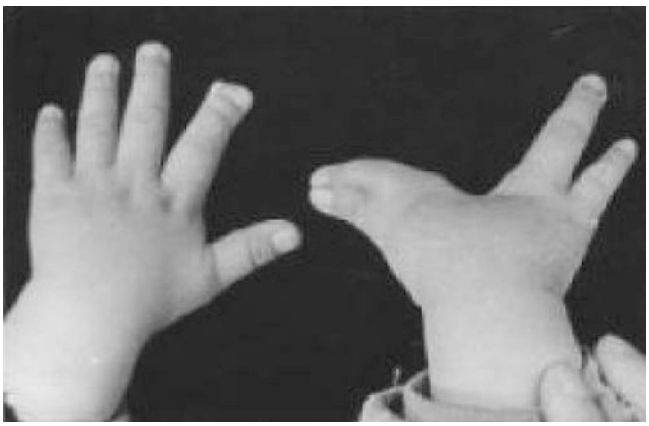


Fig. 4.36 Type III cleft hand

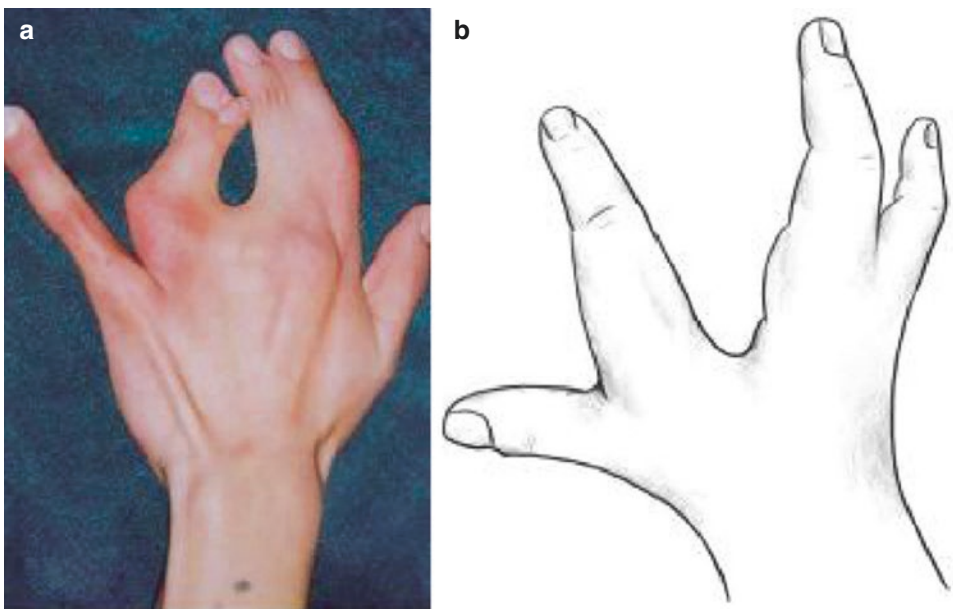


Fig. 4.35 Type IIB cleft hand

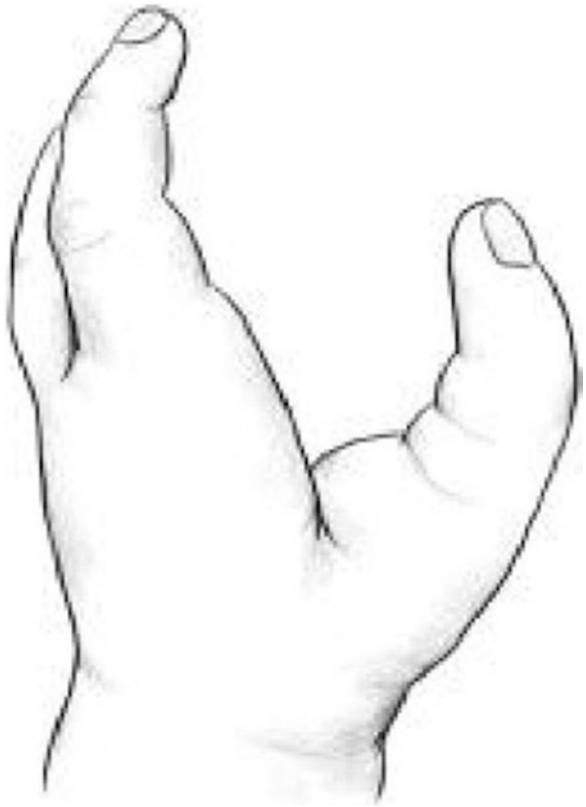


Fig. 4.37 Type IV cleft hand

etiological factors: cleft hand may originate from the early development disorder of ectosermic ridge, while web-shaped finger may originate from the deep surface skeletogeny defect during the early stage, which can explain the absence of finger end vestige in case of simple central longitudinal ray deficiency. Central longitudinal ray deficiency and central hyperdactylia can coexist, increasing the complexity of deformities. The patients with cleft hand have central absence between the little finger and the index finger or the central absence between the little finger and the middle finger. Ring finger absence and little finger dysplasia may also exist.

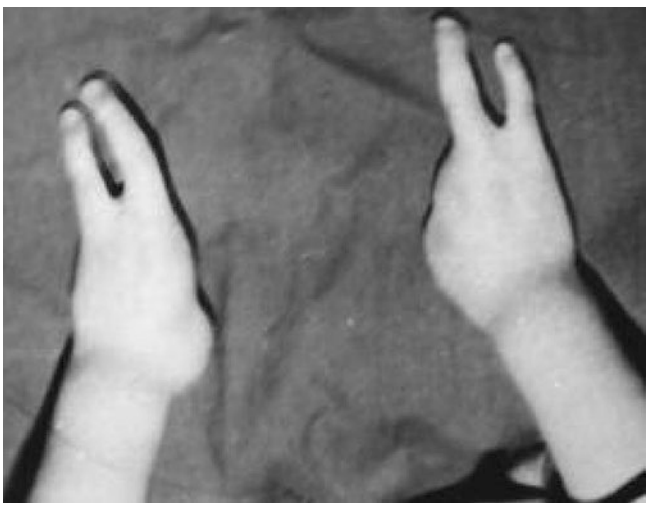


Fig. 4.38 Type V cleft hand

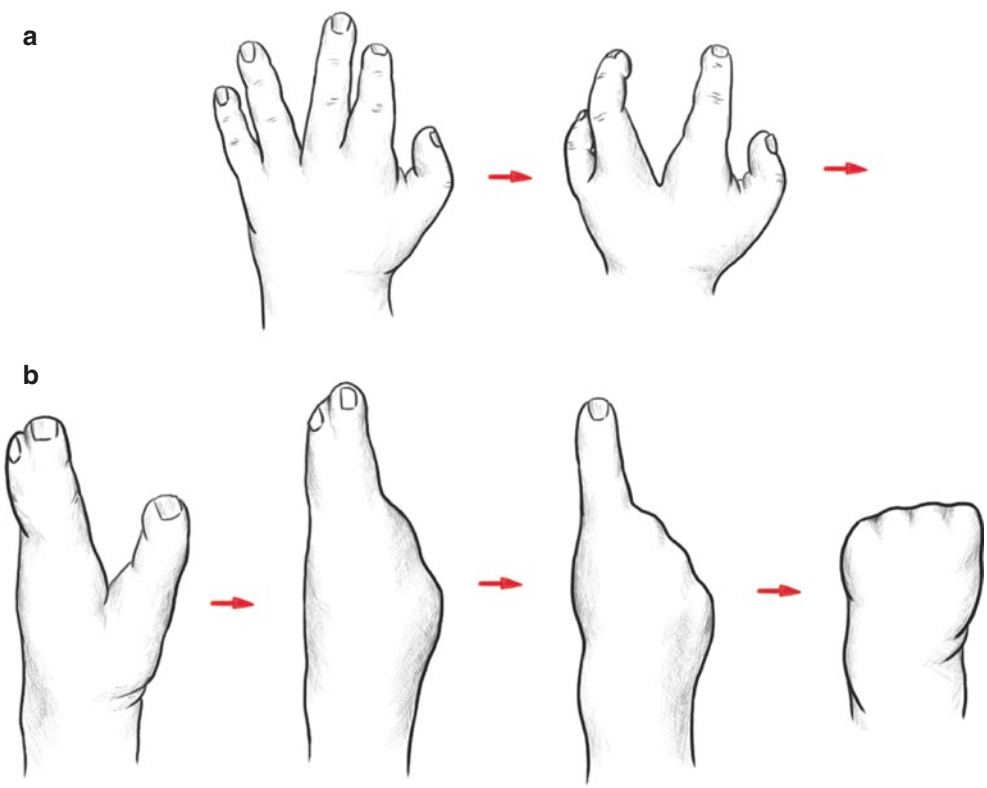


Fig. 4.39 Maisels's centripetal inhibition theory (Redrawn from Maisele DO: Lobster-claw deformities of the hand. Br J Plast surg, 1970, 23:269–281)

4.7.4 Treatment

This chapter only describes the treatment of typical cleft hand, and the treatment of atypical cleft hand will be introduced in other chapters. Besides surgical intervention, so far there has been no nonsurgical treatment method for such deformities. The specific deformity and anatomic structure of each patient should be considered for surgical treatment. The surgical purpose is to improve the functions and appearance of the hand. Surgical treatment should follow the general principles of hand surgery. That is, priority should be given to the pinching and grasping functions of the hand, and then consideration should be given to cosmetic appearance. However, it is difficult for the current medical technology to completely solve the problem of hand appearance of cleft hand. Therefore, for patients who have hand surgery for the purpose of improving hand appearance, a considerable degree of caution should be taken in the selection of the surgical treatment methods in order to avoid the postsurgical complications and hand functional disorder.

The problems that need to be solved by surgical operation include ① slit in the middle of the closed palm, ② stenotic or absent thumb web, ③ transverse ligament of head of metacarpal bone and avoidance of deformity recurrence, and ④ other defects existing in hands, such as instability caused by dysplasia of accessory ligament at the side of thumb metacarpophalangeal joint and flexion deformity or aschistodactylia of index finger, little finger, and ring finger.

Surgical treatment regimen of cleft hand should be formulated after the careful examination and preoperative X-ray of the hands. The surgeons should examine the skeletons and joints of thumb ray, index finger ray, ring finger ray, and little finger ray one by one, observe the grasping, clamping, and pinching functions of the pediatric patients, and formulate individualized therapeutic regimens. When deciding the sequence and time of surgery, the surgeons should carefully follow Flatt's suggestions:

1. As for the syndactylia, remission should be performed according to normal time sequence, the remission of marginal syndactylia should be performed 6 months later, and the remission of central syndactylia should be performed 18 months later.
2. The closure of the central cleft should be preferred after 6-month rehabilitation from the remission.
3. The thumb deformity of adduction contracture should be corrected at the same time, but usually it is unnecessary to correct mild adduction contracture.
4. The bony factors impeding the closure of clefts should be resected as less as possible because the absence of central metacarpal bone will make the palm weak and make the cleft apt to reoccur.
5. Functional fingers with proximal phalanx cannot be injured during surgery because these fingers can obviously improve the gripping and grasping capabilities.
6. Triangular phalanx, especially those concomitantly with little finger ulnar deviation or thumb radial deviation, should be corrected at the age of about 3 [35].

As the cleft hand is relatively rare, currently there is no clear therapeutic regimen that is applicable to each infantile patient. Manske and Halikis recommend the surgical methods according to the classification [33]. See Table 4.4.

In addition to this, the following surgeries can be selected according to the degree of hand deformities of pediatric patient.

1. Closed palm slits. Barsky [31] (1964) surgery can be adopted. Barsky's skip flaps can be used to close the palm slits of type I and type IIA cleft hands of Manske and Halikis classification. The surgical methods are as follows:

   The patient takes a supine position, and tourniquets are fixed at the middle segment of upper arm. Flaps are designed at the lateral side of one finger of the slit, the flap pedicle is located at the distal side, and the shape is

Table 4.4 Treatment suggestions according to the Manske and Halikis's surgical classification

Type	Reconstruction of thumb fingerweb	Central absence and closure
Type I	Treatment is unnecessary	Resecting the intermetacarpal tissues to close the cleft, graft the surrounding tendons, attaching the local tissues to the metacarpal bone (metacarpal translocation from index finger to middle finger), and if necessary resecting the excessive phalanx
Type IIA	Local pediculated skin flap grafting (Z-plasty flap transposition)	Same as described above
Type IIB	Pediculated skin flap grafting at the dorsal side (volar side) of the cleft	Same as described above, skin grafting if necessary
Type III	Syndactylia remission and skin grafting, pediculated skin grafting at the dorsal side (volar side) of the cleft, or resection of the index finger phalanx	Same as above, or resect the bony components of the index finger
Type IV	Treatment is unnecessary, or resect the tissues to reconstruct stable metacarpophalangeal joint	Treatment is unnecessary (the cleft is the web space)
Type V	Toe-finger grafting or carpometacarpal lengthening	Treatment is unnecessary

pentagon. The length and width ratio of the flaps are designed according to random flap design principle, and the proportion should be less than 1.5: 1. The flap position is at the relative position of dorsal side, and a slope inclining from the dorsal side to the volar side can be formed after suturing. Incise the skin and subcutaneous tissues, form flaps, and resect the excessive subcutaneous adipose tissues at the dermal-subcutaneous vascular plexus superficial surfaces. Incise the skin of the relative margin of slit along the bilateral free margins of the slit, expose the metacarpal bones, resect the soft tissues or bone tissues that impede the closure of adjacent metacarpal bones, close adjacent metacarpal bones, and reconstruct the transverse ligaments of the head of metacarpal bone. The reconstructing methods of the transverse ligaments of the head of metacarpal bone include: ① use the dysplastic tendons that remain in the palm as the grafts, and perform circular suture at the neck of metacarpal bone; ② incise the A1 trochlea of the adjacent flexor digital tendons in the opposite direction, and suture and reconstruct the transverse ligaments; ③ utilize the residual soft tissues of adjacent interdigits to reconstruct transverse ligaments; and ④ drill a hole in the neck of the metacarpal bones, and use silk to suture and close the metacarpal bones. After reconstruction of transverse ligaments of the head of metacarpal bone, suture the interslit skin from the near to the distant. Utilize the Z-plasty principle during suturing, resect excessive skin, make the suture lines form a Z shape at the palm cross striation, and avoid the occurrence of contracture. Adopt the perpendicular or horizontal mode to suture the palm skin, and adopt intracutaneous suture of the skin at the back of the hand. After operation, perform pressure dressing, and do functional exercise 3–4 weeks after fixation with plaster support (Fig. 4.40).

2. Thumb web reconstruction and index finger translocation. Snow-Littler surgery can be adopted. Snow-Littler surgery means that by means of slit skin, form local flaps with the volar side or the dorsal side as the pedicle, reconstruct the stenotic thumb web skin, translocate the index finger ray, distract the stenotic thumb web, and close the palm slit [38]. This method is applicable to type IIB and III cleft hand, but the formed local skin flap tip is apt to suffer necrosis due to the insufficient blood circulation, so attention should be paid during the surgery. The surgical methods are as follows:

(a) Use slit skin to form regional flaps, and reconstruct thumb web skin: make an arc incision along the slit margins at the dorsal sides of index finger and ring finger of the bilateral sides of slit and away from the head of metacarpal bone. The proximal sides of the two incisions intersect at the proximal side of the head of metacarpal bone, and the distal side is extended to the volar side after passing the head of metacarpal bone. Extend the incisions along the volar median line of the index finger and ring finger until the cross point level of the dorsal incisions is reached. Incise the skin and cutaneous tissues, respectively, and raise the skin flaps from the dorsal side to palmar side to form the regional flaps with the volar side as the pedicle.

(b) Remission of thumb web gap: make a longitudinal incision at the dorsal side of thumb web, make it parallel to the index finger until the free margin of thumb web is reached, and lengthen it toward the volar side. After the skin and cutaneous tissues are incised, relieve the soft tissues of the first and second interosseous metacarpal spaces layer by layer. Peel the first dorsal interosseous muscle at the relative margin of the first and second metacarpal bones, and partially break the adductor muscle belly of thumb. During the operation, be sure to protect the deep branches of the radial artery that passes the fundus gap of the first and second metacarpal bones.

(c) Index finger translocation: perform osteotomy at the relative level of the second metacarpal bone accord-

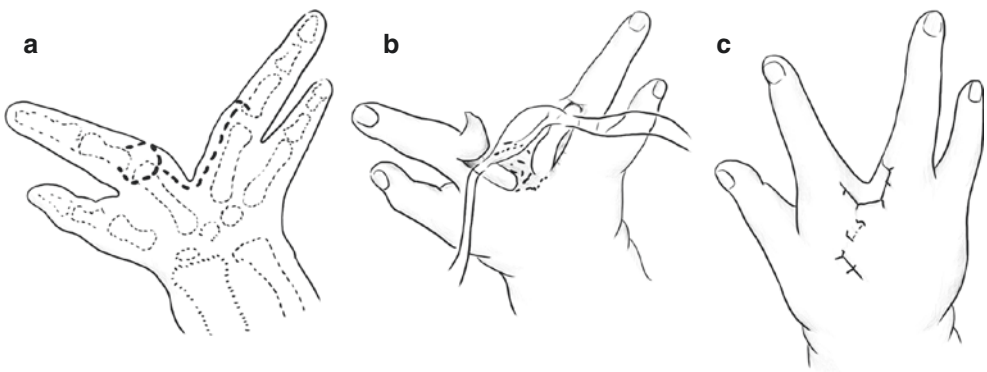


Fig. 4.40 Barsky's surgery (Reprint with permission from WOLTERS KLUWER HEALTH, INC. LICENSE TERMS AND CONDITIONS) (Redrawn from Barsky AJ: Cleft hand: Classification, incidence, and treatment. J Bone Joint Surg, 1964, 46A: 1707–1719)

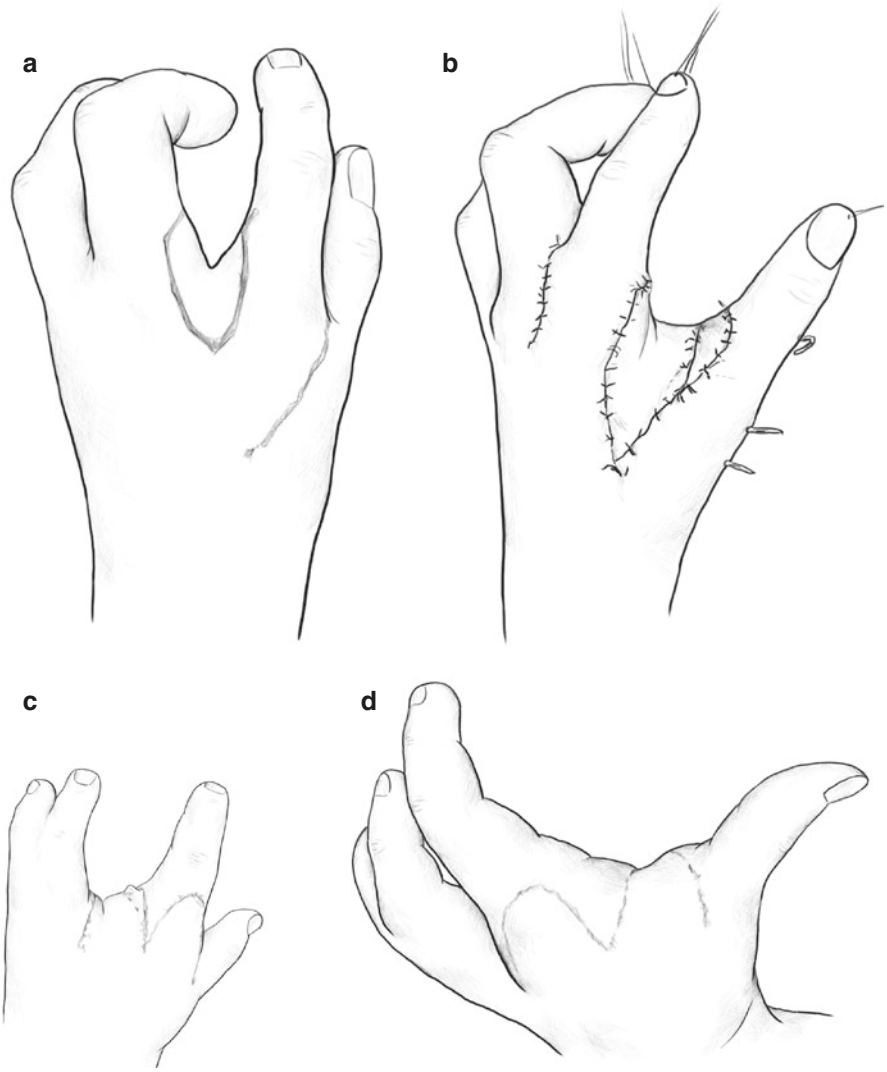


Fig. 4.41 Snow-Littler surgery. (**a**, **b**) Reconstruction of thumb web with the volar side as the pedicle. (**c**, **d**) Reconstruction of thumb web with the dorsal side as the pedicle

ing to the length of the residual third metacarpal bone. If the residual third metacarpal bone is very short, then perform osteotomy at the fundus of the second metacarpal bone. Perform peeling under the periosteum, expose the shaft of metacarpal bone, and complete osteotomy. Before fixation, it is necessary to examine whether the translocated index finger suffers any deformity of rotation.

(d) Closing the palm slit: make a relatively dorsal longitudinal incision at the ulnar side of index finger, and suture it with the radial incision of the ring index to form fingerweb. Graft the volar pedicle flap to the newly established thumb web. During operation, avoid tension in suturation. In case of skin defects, use the hypothenar eminence of the same hand to take the full-thickness skin graft for suturing.

(e) Relax the tourniquet: after examining the blood circulation of skin flaps, perform bandaging. Use plaster support for external fixation at the hand functional position, and expose the fingertip. After 6 weeks, pull out the Kirschner wire, and perform functional exercise (Fig. 4.41).

4.8 Ulnar Longitudinal Ray Deficiency

Laijin Lu and Xu Gong

Ulnar longitudinal ray deficiency refers to the congenital dysplasia or defect in the ulnar structure of upper limbs, and it can involve the ulnar bony and (or) soft tissue structures of elbow joint, forearm, and hand. Ulnar longitudinal ray deficiency is quite rare clinically, and its incidence is

1/100,000 to 7.4/100,000, lower than that of the radial ray defect and central ray defect. Ulnar longitudinal ray deficiency is often sporadic usually concomitantly with other musculoskeletal defects.

4.8.1 Clinical Manifestations and Classification

Ulnar longitudinal ray deficiency often has the following manifestations: ① short upper limbs, ② occurrence of elbow joint motor disorder due to the dislocation of radial capitulum or radial-humeral fusion, ③ presence of forearm radial bones and the formation of stable joints with the wrist bones, and ④ ulnar defects or dysplasia and occurrence of pronation or forward prominent arch form, making the hand present supination or backward deformity position. Sixty-eight percent to one hundred percent of the ulnar longitudinal ray deficiencies are accompanied by hand development abnormalities. The deformities include adactylia, syndactylia, musculoskeletal dysplasia, etc., and both the ulnar side and the radial side can be involved. Clinically, the ulnar longitudinal ray deficiency can be classified according to forearm and elbow joint deformities and can also be classified according to hand deformities.

4.8.1.1 Classification According to the Forearm and Elbow Joint Deformities

Currently there are six kinds of similar classifications according to the degree of forearm and elbow joint deformities. The Bayne classification is widely applied as it is similar to the classification of radial longitudinal ray deficiency. Its feature is that it is classified in accordance with the dysplasia of ulnar bone and elbow joint, and the coverage is from ulnar dysplasia (ulnar minus variant) to complete ulnar defect accompanied with radial-humeral bony fusion. As hand deformities are not covered in the current classification method, in 2005 Havenhill et al. [39] included the deformities involving the hand and wrist but not involving the forearm and elbow joint in the Bayne classification and were named type 0. In addition, phocomelia is listed as radial ray defect, and the proximal manifestation of ulnar ray defect is included as type V:

1. Type 0. The length of ulnar bone relative to the radial distal end is normal, and the defect is limited to the hand (Fig. 4.42).
2. Type I. The ulnar bone is dysplastic with shortening, but the proximal and the distal epiphysis of the ulnar bone is present (Fig. 4.43).
3. Type II The distal end of ulnar bone is not developed, namely, ulnar distal defect (Fig. 4.44).

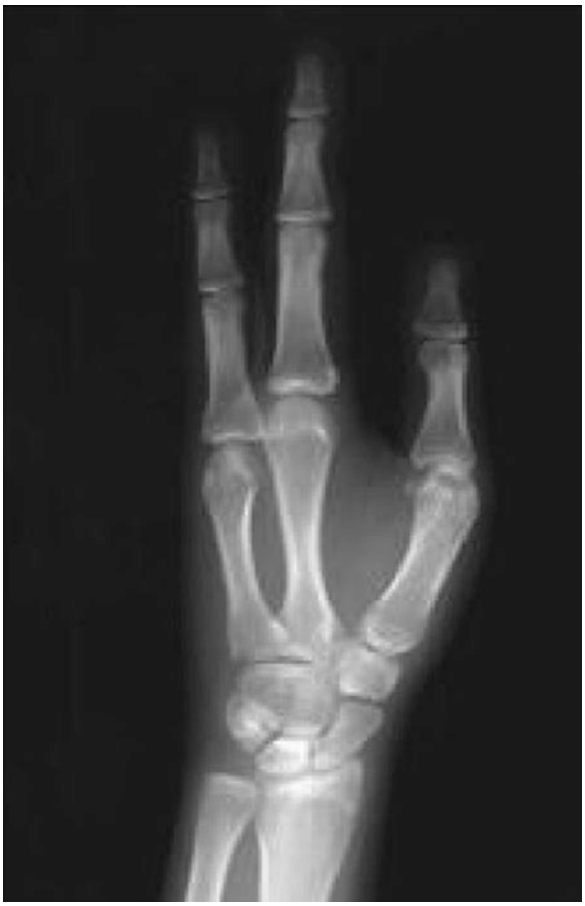


Fig. 4.42 Type 0 of ulnar longitudinal ray deficiency

4. Type III. The ulnar bone has complete defects, and the forearm only has the radial bone left (Fig. 4.45).
5. Type IV. The ulnar bone has complete defects concomitantly with radial-humeral fusion, namely, elbow joint defect (Fig. 4.46).
6. Type IV. Not only do the hand, wrist joint, forearm, and elbow joint suffer dysplasia, but also the proximal end of the upper limb has dysplastic glenoid cavity and upper limb osteocomma.

4.8.1.2 Classification According to Hand Deformities

1. Ogino classification. According to the number of hand ray deficiency, Ogino et al. classified the hand deformities of ulnar longitudinal ray deficiency from the ulnar side to the radial side into five types:
   (a) Type A: little finger defect
   (b) Type B: the fifth ray defect
   (c) Type C: the fourth and fifth ray defect
   (d) Type D: the third to fifth ray defect
   (e) Type E: the second to fifth ray defect

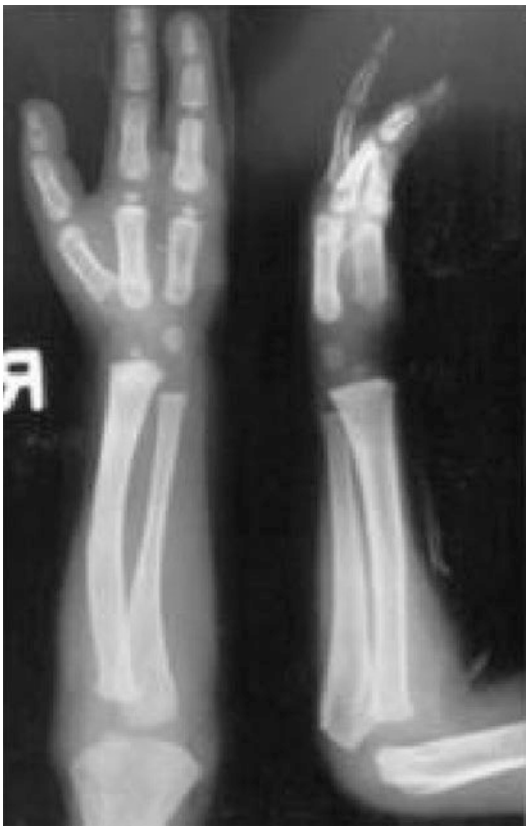


Fig. 4.43 Type I of ulnar longitudinal ray deficiency

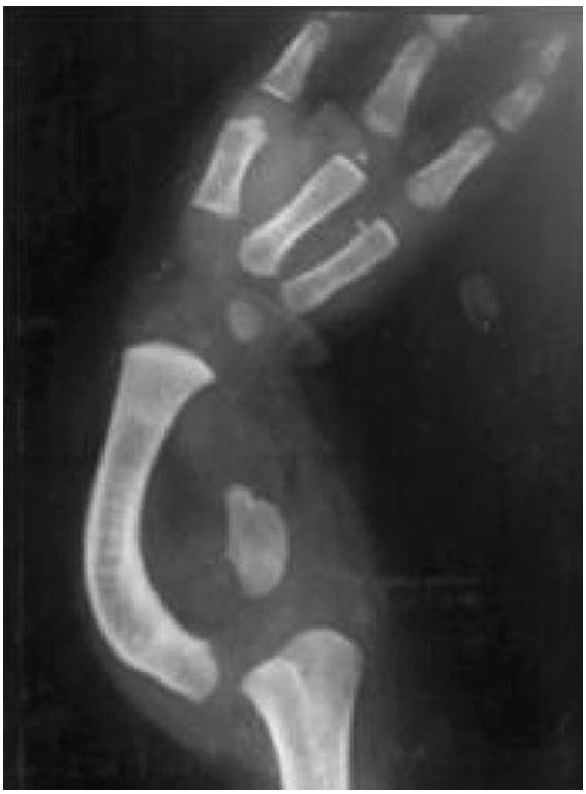


Fig. 4.44 Type II of ulnar longitudinal ray deficiency

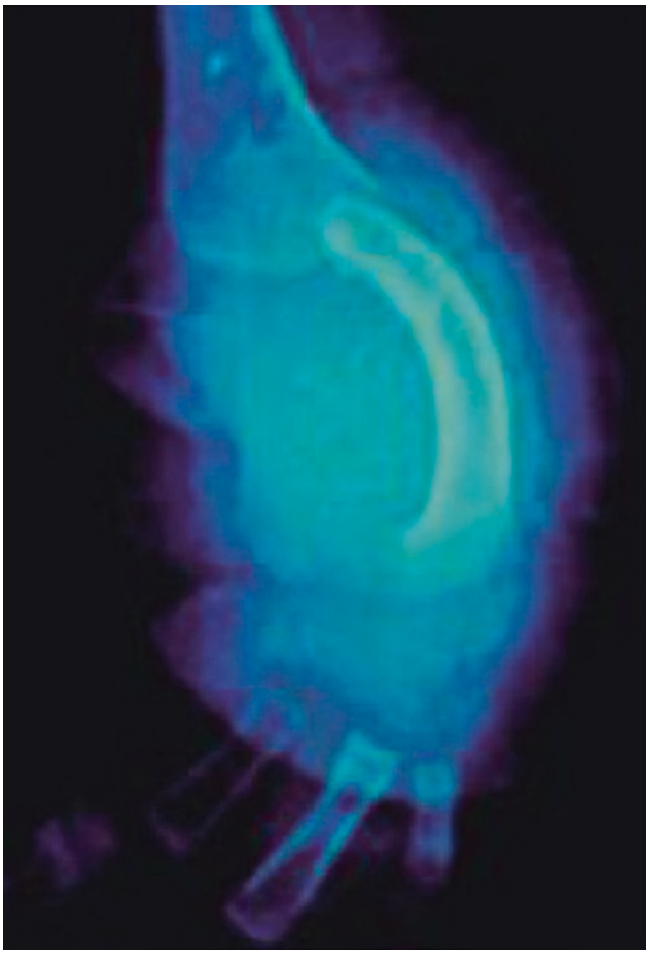


Fig. 4.45 Type III of ulnar longitudinal ray deficiency

2. Cole classification. As Ogino classification does not cover thumb dysplasia, Cole et al. propose the following classification according to the characteristics of thumb dysplasia:
   (a) Type A: thumb and thumb web are normal.
   (b) Type B: thumb web and thumb have mild defects.
   (c) Type C: thumb web has mild and severe defects, including thumb and index finger syndactylia, thumb rotation deformity, thenar muscle dysplasia, hand extrinsic muscle defects, etc.
   (d) Type D: thumb defect.

4.8.2 Treatment

4.8.2.1 Correction of Forearm Deformities

Although ulnar longitudinal ray deficiency has defects in appearance, the upper limbs function well and seldom suffer functional disorder. As there are fibrocartilage vestiges among the radial distal end, carpal bone, and poorly developed ulnar distal end, it was believed in the past that the constraint effects

Fig. 4.46 Type IV of ulnar longitudinal ray deficiency

of the vestiges can lead to further deformities of forearm and wrist joint, such as retarded growth of radial distal end, deformity of ulnar deviation of wrist joint, deformity of radial arch shape, and dislocation of radial capitulum. However, present studies hold that deformity of ulnar longitudinal ray deficiency is relatively static; therefore, the fibrocartilage vestige is not of clinical significance. Surgical resection can only be performed under the condition of deformity of progressive wrist joint ulnar deviation, deformity of radial arch shape, and dislocation of radial capitulum.

The most common forearm surgery is radial osteotomy, which can be used to correct bowing deformity of forward radial bones or serious pronation deformity. During radial osteotomy, the forearm is placed in the neutral position so that the affected limb can coordinate with the contralateral upper limb. For the dislocation of radial capitulum concomitantly with pain or obvious deformity, the radial capitulum can be resected, but it should be noted that the resection of radial capitulum can make the previously stable and functional elbow joint lose the functions and stability. For the patients with unstable forearm, the forearm reconstruction for osteocomma can be performed, but unstable forearm is rarely found in such patients [40].

4.8.2.2 Correction of Thumb and Thumb Web Deformities

The most common surgery adopted for ulnar longitudinal ray deficiency is correction of thumb and thumb web deformities, and the surgical methods include thumb web deepening, finger separation of thumb and index finger syndactylia, thumb ray rotation-valgus osteotomy, and utilization of pedicled skin flap to reconstruct thumb web. For other deformities, pertinent modes of operation can be adopted. For example, thumb reconstruction can be adopted for thumb defect, and little finger abductor muscle translocation can be adopted to reconstruct the opposition functions as for thenar muscle defects.

4.8.2.3 Correction of Other Hand Deformities

Except for the deformities in thumb and thumb web, corresponding modes of operation can be adopted to correct other hand deformities such as syndactylia and flexion contracture.

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Congenital Thumb Dysplasia

5

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5.1 Introduction

5.1.1 Concept of Congenital Thumb Dysplasia

Thumb dysplasia (aplasia) and congenital thumb deformities are the commonest congenital deformities of hands and upper limbs. Many authors discuss them by including them into radial defects; the author describes it in this chapter because it holds an important position in the congenital deformities of hands and upper limbs.

The author believes that thumb dysplasia (aplasia) is a major disease among congenital deformities of hands and upper limbs, and it is necessary to have a profound understanding of thumb dysplasia (aplasia) from the depth and width of the congenital deformities of hands and upper limbs. Duplication of the thumb also falls into the category of thumb dysplasia, but due to its special nature, a chapter will describe it in detail.

During the first to third month of a life, thumb is in the position of flexion and adduction into the center of the palm; during the sixth to seventh months, the infants become able to grasp and pinch an object by pressing the thumb against other finger(s); during the ninth month, the thumb can move freely and independently from the center of the palm; at the age of 1, the thumb can perform most functions of hands.

Congenital abnormality in thumb shape, structure, position, number and range, amplitude and strength of movement as well as abnormality in the position and depth of the first fingerweb all fall into the category of congenital thumb deformities or dysplasia.

Congenital thumb deformities include congenital thumb dysplasia, defect or deformity, and generally refer to congenital deformities of hands and upper limbs accompanied by thumb deformities.

When treating the pediatric patients with congenital thumb dysplasia, the surgeons should know the following well:

1. Clinical manifestations, hereditary character, and etiological factor of thumb dysplasia as well as the hard-to-notice manifestations of generalized deformities
2. Classification of thumb dysplasia
3. Evaluation of the shape of thumb dysplasia, scaffold structure, stable structure, functional defect, and lesion degree
4. Principles and methods of correcting the shape of thumb dysplasia, scaffold structure, stable structure, and functional defect
5. Treatment steps, content, methods, staging, and perioperative treatment of thumb dysplasia
6. The treatment stage of thumb dysplasia and evaluation of final effects, the consistency and difference of doctors and patients in the understanding of the treatment results, and the common understanding of the final evaluation of treatment effects

5.1.2 Denomination of Congenital Thumb Deformity

There are many nouns for the clinical description of congenital thumb deformities, which is due to the diversity of the deformities and constant changes in the understanding and study of them. The representations of congenital thumb deformities include congenital thumb deformities, congenital pollical maldevelopment, congenital aplastic thumb, short thumb, triphalangeal thumb, radial polydactyly, thumb duplication, congenital adducted thumb, congenital abducted thumb, thumb syndactyly, delta phalanx of thumb, congenital trigger thumb, thumb-in-palm deformity, and triplication thumb, and the duplication of the thumb include three finger nails, two metacarpal bones, seven phalangeal bones, etc. Among the above thumb deformities, some are dyspoiesis, such as congenital thumb dysplasia; some are dysdifferentiation, such as syndactylia of thumb; some are twin deformity, such as triphalangeal thumb and radial polydactyly; some are low development, such as short thumb and the thumb deformity; and dysplasia in syndromes such as the Poland syndrome and the Apert syndrome.

Congenital thumb dysplasia is the main part of congenital thumb deformities and is referred to as congenital hand deformity mainly characterized by thumb dysplasia or aplasia.

5.1.3 Incidence of Congenital Thumb Dysplasia and Congenital Deformities of Hands and Upper Limbs

The author et al. investigated the birth records of 350,000 neonates in Shanghai urban areas. According to the investigation, the incidence of congenital deformities of hands and upper limbs was 0.85‰. Congenital thumb dysplasia is manifested as congenital thumb deformity, and it is the

commonest kind of congenital deformities of hands and upper limbs. Entin (1959) reported that, among the congenital deformities of hands and upper limbs in Canada, the incidence of thumb deformities was 16% [1, 2]. According to the report made by Tay S.C. et al. (2006), in recent years, the incidence of congenital deformities of hands and upper limbs accounted for 3.4/10,000 [3–5] of the new births, and thumb deformities accounted for 16% of congenital deformities of hands and upper limbs. Among the etiologic factors of congenital hand deformities, 40–50% were unclear. Judging from the author's many years of clinical practice, the author feels that the incidence of congenital thumb deformities and their proportion in hand deformities are much higher than the statistics obtained by Entin et al., and the reasons are as follows: ① every category in the classification of congenital upper limb deformities can be manifested as congenital thumb deformity; ② most congenital hand deformities are accompanied by thumb deformities; ③ in the treatment of congenital deformed hands, in case of concomitant thumb deformities, repair and reconstruction of thumb functions are the first link in hand function reconstruction; ④ among the patients with multiple syndromes admitted into department of plastic surgery, the patients concomitantly suffering congenital dysplasia are very common. Upton J. reported that the incidence of thumb deformities accounted for 37% of that of upper limb deformities, which is similar to the author's clinical findings, because the multiple syndromes are accompanied by thumb dysplasia [6].

5.1.4 Concept of Congenital Thumb Dysplasia and Diversity of Deformities

As for the congenital thumb dysplasia, there are always some problems difficult to solve whether in exploration of etiology, description of morphological diversity, demonstration of deformity classification method, or discussion of treatment method. Having treated over 1000 patients with congenital thumb deformities and consulted a lot of domestic and foreign materials, the author deeply believes that congenital thumb deformities are caused by many reasons and take a variety of forms, and their treatment means are a challenge for surgeons for their limited medical knowledge and difficulty in aesthetic reconstruction. At present, understanding of congenital thumb deformities is being constantly deepened. During clinical practice, the more in-depth and detailed the understanding and evaluation of the thumb deformity factors is, the more effective treatment decisions can be made by surgeons in plastic surgery, microscopic reconstruction, and hand surgery.

Every category in the classification of congenital upper limb deformities can be manifested as congenital thumb deformity.

5.1.4.1 Limb Formation Disorder

Limb formation disorder includes the following kinds:

1. Transverse arrest. Congenital broken hand or congenital broken thumb deformity can occur.
2. Longitudinal arrest. Both radial longitudinal ray deficiency (preaxial) (radial clubhand) and ulnar longitudinal ray deficiency (postaxial) (ulnar clubhand) can be manifested as thumb dysplasia or disorder of thumb functions to varying extents accompanied by thumb dysplasia.
3. Central ray deficiency. Among the central ray deficiency deformities (cleft hand), thumb deformity is common and can be accompanied by thumb dysplasia.

5.1.4.2 Failure of Differentiation of Parts

Failure of differentiation of parts includes the following kinds:

1. Multiple arthrogryposis of soft tissues. Multiple arthrogryposis of soft tissues (congenital multi-joint flexion deformity) can occur to patients with congenital elevated scapula (incomplete shoulder descending), pectoral muscle absence (including the Poland syndrome), hand long flexor distortion, hand long extensor distortion, and hand internal muscle distortion induced by elbow and forearm horizontal muscle deformity; thumb deformities may also occur. It can also occur to patients with cutaneous syndactylia (complete and incomplete), congenital flexion deformity (congenital finger flexion deformity), thumb-in-palm deformity, non-bony finger deviation deformity (arthrochalasis induced by dysplasia of muscles, ligaments, and joint capsule), independent finger deviation, and ulnar deviating hand (windblow hand), and thumb deformities can almost occur including congenital trigger finger.
2. Bony deformities. Bony deformities include carpal bone fusion, metacarpal bone fusion, phalangeal bone fusion, knucklebone fusion, congenital finger lateral flexion deformity, and multi-segment phalange deformity, to which thumb deformities can occur.
3. Congenital tumor-induced deformities. Among the patients with congenital tumor-induced deformities, vascular, neurogenic, connective tissue-derived and skeletal osteochondroma (including multiple hereditary exostosis), enchondroma and epiphysis abnormalities might all occur to the thumb, with the manifestation of thumb deformity or dysplasia.

5.1.4.3 Duplication

Duplication can be manifested as follows: ① abnormal superfluous growth of the entire limb, ② mirror hand deformity in double ulnar bones, ③ radial (preaxial) thumb hyperdactylia or duplication of the thumb, and ④ thumb longitudinal ray epiphyseal repetition (deformity of excessive epiphysis) deformity, etc. All the above deformities can be accompanied by thumb deformities.

5.1.4.4 Overgrowth

Overgrowth can occur to the entire upper limb or partial limb or is manifested as finger overgrowth, which is common in thumbs.

5.1.4.5 Undergrowth

Undergrowth is often accompanied by thumb deformities.

5.1.4.6 Constriction Ring Syndrome

Patients with constriction ring syndrome can suffer constriction ring deformity of thumb or thumb dysplasia.

5.1.4.7 Systemic Skeletal Deformities and Syndromes

Systemic skeletal deformities and syndromes can be accompanied by thumb deformities.

Simple congenital thumb deformities refer to isolated congenital thumb deformities, including congenital thumb absence, thumb dysplasia, and various kinds of thumb adduction deformity, thumb-in-palm deformity, triangle triphalangeal thumb, triphalangeal thumb, duplication of the thumb, congenital thumb thecostegnosis, congenital thenar muscle absence, thumb constriction band deformity, etc.

Thumb deformities are common in upper limb preaxial longitudinal formation disorder, such as thumb deformity of radial clubhand in the radial longitudinal ray deficiency, which is induced by the first longitudinal ray dysplasia of hands with the manifestation of thumb dysplasia.

Multiple-hand congenital deformities can be accompanied by thumb deformities, such as spade hand, cleft hand, syndactylia, mirror hand, and ulnar-deviating hand (including windblow hand) in hand dysplasia. Thumb dysplasia and deformities to varying extents can be found.

Some generalized deformities, such as cardiovascular abnormalities, genitourinary malformations, musculoskeletal abnormalities, and vertebral deformity, can also be accompanied by thumb deformities. The patients with some syndromes, such as Holt-Oram's syndrome characterized by slenderness and dysplasia of distal segment of fingers; Fanconi's anemia with hemopoietic system diseases complicated by hand deformities; the Apert syndrome characterized by short, flat, and broad distal finger segments and acrocephalosyndactylia; and the Poland syndrome, with brachydactylia and syndactylia deformities accompanied by the absence of major pectoral muscle, can concomitantly suffer from thumb deformities.

5.2 Etiological Factors of Congenital Thumb Dysplasia

Thumb dysplasia is one type of congenital thumb deformities. Both short thumb deformity and complete absence of thumb fall into the category of thumb dysplasia. Thumb dysplasia can exist independently and is also one of the manifestations of congenital hand deformities or syndromes.

Thumb dysplasia shares the same etiologic factors with other congenital deformities of hands and upper limbs, and the etiologic factors of it remain unclear. Thumb dysplasia may be caused by the mother's contraction of diseases, trauma, and administration of teratogenic drugs during early pregnancy. It is also often correlated with genetic factors.

Some people have noticed that thumb dysplasia is induced by embryonic limb bud deficiency during embryonic development, and its pathogenesis is similar to that of radial clubhand; it may be induced by limb bud dysdifferentiation. Several decades ago, thalidomide was sold on the market as a kind of sedative. Pregnant women taking this drug delivered a considerable number of infants with congenital deformities of hands and upper limbs. Inheritance was also found to be another important factor. The author once admitted and treated several pediatric patients with congenital thumb dysplasia, all of whom had a history of familial inheritance. Case one is a patient with six fingers without thumb, both his/her maternal grandmother and mother have five fingers but no thumb, and six among the four generations in the family suffer thumb dysplasia, four of whom are females and two are males. Case two is a patient suffering congenital thumb absence, his/her father suffer congenital absence of left thumb with triphalangeal deformity in the right thumb. Case three is a boy with seven fingers accompanied by brachydactylia, syndactylia, and thumb dysplasia, and his father and paternal grandfather suffer similar deformities. Case four is a girl with five fingers but no thumb in the left hand as well as six fingers but no thumb in the right hand; both her father and paternal grandmother suffer polydactyly in both hands with the absence of thumb; among the generation of her paternal grandmother (four females and two males), the generation of his father (five females and eight males), and the generation of the pediatric patient (five females and six males), only the child's father and paternal grandmother suffer deformities in two hands (Fig. 5.1).

5.3 Classification of Congenital Thumb Dysplasia

The pathology, etiologic factors, and clinical symptoms of congenital thumb deformities and thumb dysplasia are complicated. Knowing the classification of thumb dysplasia and deformities is the basis for treatment.

Classification is the basis for understanding of diseases and selection of treatment methods. Both short thumb deformity and complete absence of thumb fall into the category of thumb dysplasia. Thumb dysplasia is diverse in terms of morphology, anatomical structure, and etiology. In order to distinguish different kinds of congenital thumb deformities and facilitate the selection of treatment methods, a number of authors have classified thumb dysplasia. The work done by such authors is of certain reference value to clinical practice.

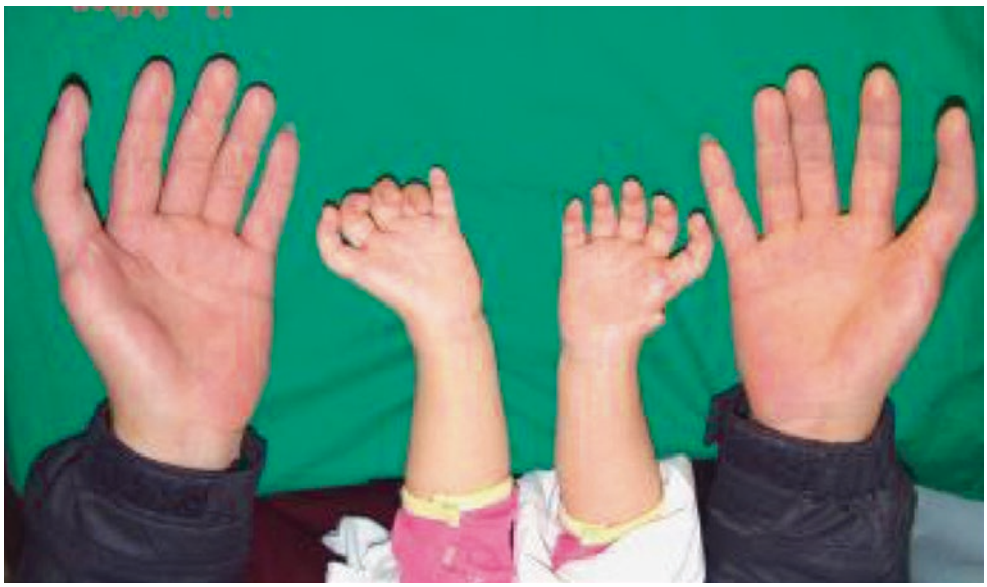


Fig. 5.1 The case is a female infantile patient; her father suffers polydactylism with no thumb

5.3.1 Blauth's Classification of Thumb Dysplasia

Blauth (1967) [7] classified it into five types according to the degree of thumb dysplasia, and a number of authors have made some supplement. This is a classification method adopted by many fellows, but it is not comprehensive enough. At present, many authors adopt Blauth's classification method of thumb dysplasia [8–12] (Table 5.1).

Table 5.1 Blauth's classification of thumb dysplasia

Type	Clinical manifestation
Type I	The thumb is small, short, and narrow; all structures of thumb are present and accompanied by mild dysplasia of hand internal muscles; and the thumb functions are basically normal
Type II	The thumb suffers obvious dysplasia, the shape and functions are not comprehensive, the thenar muscle suffers dysplasia, the thumb is adducted, the thumb web is stenotic, the collateral ligaments of metacarpophalangeal joint are flabby with poor stability, and the thumb bony structures are present but relatively small
Type III	The thenar muscles are absent, the hand external muscles suffer dysplasia, and the disease is classified into type a and type b according to the development degree of the first metacarpal bone
Type IIIa	Dysplasia of type II thumb + dysplasia of the first metacarpal, dysplasia of thumb extensor tendon, and stable carpometacarpal joint
Type IIIb	Dysplasia of type II thumb + obvious dysplasia of the first metacarpal, dysplasia of thumb extensor tendon, and unstable carpometacarpal joint
Type IV	Thumb is floating and thumb-end bones are present, but dysplasia is serious; metacarpal bones are absent, the thumb as soft as silkworms are connected with hands by very small skin bridges, nerves and vessels are present inside the skin bridges, and the functions of floating functions are completely absent
Type V	Complete absence of thumb
Five-fingered hand	The hand has five fingers, which become extended on one plane at the volar side, and the appearance and functions of thumb are absent

5.3.2 Bayne's Classification of Thumb Dysplasia

Bayne L G. (1982) classified thumb dysplasia into 5° (Table 5.2).

5.3.3 Manske's Classification of Thumb Dysplasia

Manske's [11, 12] classification (1995) of thumb dysplasia is a supplement to the previous classification methods, and it is brief (Table 5.3).

5.3.4 Author's Classification of Thumb Dysplasia

The new classification method proposed by the author is based on the deformed shape of congenital thumb dysplasia, the absence or abnormality of anatomical structure, the deficiency degree of basic functions of thumb (flexion, extension, abduction, adduction, rotation, and opposition), the selection of treatment methods, and the indications, and it corresponds to the etiologic factors, symptoms, and treatment methods of thumb dysplasia.

Bases used by the author for classification of congenital thumb dysplasia:

Table 5.2 Bayne's classification of thumb dysplasia

Type	Clinical manifestation
Degree I	Deformity of short thumb
Degree II	Deformity of adducted thumb
Degree III	Deformity of abducted thumb
Degree IV	Floating thumb
Degree V	Complete absence of thumb

Originate from Joseph Upton, MD. Management of transverse and longitudinal deficiencies (failure of formation). Plastic Surgery Second Edition. Edited by Stephen J. Mathes. MD. Philadelphia. Vol. 8. 2006:51–137

Table 5.3 Manske's classification of thumb dysplasia

Type	Clinical manifestation
First category	The thumb is small, short, and fine
Second category	The thumb web is stenotic, the thenar muscles suffer dysplasia, and the metacarpophalangeal joint is unstable
Third category	In addition to the manifestations of the second category, it also shows manifestations as follows: ① hand external muscles and tendons are abnormal, partial metacarpal bones suffer dysplasia, and carpometacarpal joint is stable; ② the hand external muscles and tendons are abnormal, the metacarpal bones suffer dysplasia, and the carpometacarpal joint is unstable
Fourth category	Floating thumb
Five category	Deletion of thumb

Originate from PR Manske, Halikis MN. Surgical classification of central deficiency according to the thumb web. J Hand Surg Am 1995 Jul;20 (4): 687–697

1. Morphological characteristics. The location and morphological characteristics of congenital thumb dysplasia.
2. Anatomical structure. The development deficiency or abnormalities of thumb anatomical structure, including supporting structures of bones and joints; the stable structures between ligaments, bones, and joints; and development status of dynamical structures of muscles and tendons.
3. Functional status. The basic functions of thumb, including flexion, extension, abduction, adduction, rotation, and damage of opposition functions and absence degree, to assess the development of the above anatomical structures.
4. Relation between thumb deformity and hand deformity. The importance of thumb dysplasia in congenital hand deformities.
5. Relation between thumb deformity and condition of the whole body. Whether thumb deformity is accompanied by deformities in other organs.
6. Selection of treatment methods. The treatment methods that should be selected are sometimes inconsistent with

Table 5.4 Upton J.'s classification method of thumb dysplasia and absence [6]

Dysplasia of thumb	Supplementation for thumb dysplasia
Type I: mild dysplasia of thumb	Type VI: central longitudinal ray dysplasia accompanied by thumb dysplasia
Type II: moderate dysplasia of thumb	Type VII: congenital constriction band accompanied by thumb dysplasia
Type III: severe dysplasia of thumb	Type VIII: thumb dysplasia in five-fingered hands
Type IV: floating thumb	Type IX: radial polydactylism accompanied by thumb dysplasia
Type V: thumb aplasia	Type X: syndrome of thumb dysplasia of short thumb

Originate from Joseph Upton, MD. Management of transverse and longitudinal deficiencies (failure of formation). Plastic Surgery Second Edition. Edited by Stephen J. Mathes. MD. Philadelphia. Vol. 8. 2006:51–137

the classification of congenital hand deformities because most thumb dysplasia is limb formation disorder, such as complete thumb absence and radial clubhand of radial longitudinal ray deficiency. Thumb dysplasia concomitantly with syndactylia and pectoral muscle absence falls into the category of limb dysdifferentiation; five- and six-fingered congenital deformities and thumb deficiency fall into the category of polydactylism in taxology, namely, duplication; as for congenital constriction band accompanied by deformities of thumb dysplasia or broken thumbs in hands and thumbs, their treatment methods vary. The author's classification method also refers to the classification methods of other authors, including Upton J.'s (2006) classification method (Table 5.4) of thumb dysplasia and absence, and it is a practical classification method.

The new classification method proposed by the author is developed according to the supporting structure of deformities (osteoarticular developmental status), dynamical structures (muscle and tendon development status), stable structures (ligament and aponeurosis developmental status), and shape and structure (the proportion of position, length, thickness, diameter and transverse diameter, the size of nails, the development of nail folds, etc.). The objective of classification is to help know the nature of deformities and performance characteristics to better understand disease, select treatment methods, and disseminate knowledge (Tables 5.5 and 5.6). Although various deformities of duplication of the thumb are accompanied by obvious thumb dysplasia, they are not included in the classification of radial polydactylism in thumb dysplasia, and they will be discussed in detail in a chapter.

Table 5.5 New classification methods of congenital thumb dysplasia (Wang Wei)

Type	Clinical manifestation
Type I (mild dysplasia of the thumb)	The thumb is short, small, and narrow, accompanied by mild adduction or abduction; functional lesions are mild; and the supporting structures, dynamical structure, and stable structures of the thumb are nearly normal. In spite of dysplasia to varying extents, the flexion, extension, abduction, adduction, rotation, and opposition functions of thumb are basically normal; the muscular strength is over grade 4; and it is deformity of short thumb with functional insufficiency
Type II (moderate dysplasia of the thumb)	The thumb suffers obvious dysplasia accompanied by thenar muscles or extensor and flexor dysplasia in the thumb. The thumb is short with deformity of adduction, and moderate impairment is observed in shape, structures, and functions. Due to the differences in deformity degree, it can be classified into four subtypes: a, b, c, and d
Type IIa	Thumb adduction, thumb web stenosis, and obvious dysplasia in thenar muscles
Type IIb	Thumb adduction, thumb web stenosis, dysplasia in thumb extensor, and hand external muscles, with/without dysplasia in thenar muscles and possibly accompanied by mild supporting structures such as metacarpal bone and phalanx dysplasia
Type IIc	Thumb adduction, thumb web stenosis, dysplasia in thumb flexor, with/without dysplasia in thenar muscles, dysplasia in thumb flexor tendon, abnormal insertion position, possibly accompanied by mild supporting structures such as dysplasia in metacarpal bone and phalange
Type IId	Mild damage in thumb phalange and/or the first metacarpal bone; moderate abnormalities in shape and structure; relatively small phalange or metacarpal bone; ability of flexion, extension, abduction, adduction, rotation, and opposition of thumb; and hypoplasia of muscular strength to varying extents
Type III (severe dysplasia of the thumb)	Obvious dysplasia in thumb dynamical structure, supporting structure, stable structure, and shape, manifested as thumb adduction, dysplasia in thenar muscles, dysplasia in hand external muscles, obvious dysplasia in thumb phalange and the first metacarpal bone, and dysplasia in part or most of the interphalangeal joint, metacarpophalangeal joint, and/or carpometacarpal joint. In spite of the presence of the thumb at the opposition position, the first metacarpal bone is sometimes finger-type metacarpal bone and phalange, and the first metacarpal bone suffers serious dysplasia. Due to the differences in deformity degree, it can be classified into three subtypes: a, b, and c
Type IIIa	Type II thumb dysplasia + severe dysplasia of the first metacarpal bone, dysplasia of the first carpometacarpal joint, and severe impairment in thumb supporting and dynamical functions
Type IIIb	Type II thumb dysplasia + obvious dysplasia or partial absence of the first metacarpal bone, severe dysplasia or the absence of the first carpometacarpal joint, and severe impairment in thumb supporting and dynamical functions
Type IIIc	Type II thumb dysplasia + phalange dysplasia; severe dysplasia; abnormal position of the first metacarpal bone, metacarpophalangeal joint, and carpometacarpal joint; deformity of thumb abduction; and severe impairment in thumb position, shape, and function
Type IV (floating thumb)	Thumb-end bones and soft tissues are present, but the dysplasia is serious, metacarpal bones are absent, the thumb as soft as silkworms are connected with hands by very small skin bridges, nerves and vessels are present inside the skin bridges, and the functions of floating functions are completely absent
Type V (dysplasia of thumb absence type)	Four-fingered hand, the absence of the thumb at the opposition position, fair development of four-fingered hand, good or mildly poor finger appearance and functions
Type VI (hyperphalangeal thumb dysplasia)	Hyperphalangeal thumb defect, including five fingers or six, seven, and eight fingers; all fingers being on the same plane, the absence of the thumb at the opposition position, good shape and functions of fingers, or partial dysplasia (radial hyperdactylia includes various kinds of thumb deformities; in spite of the accompanying obvious thumb dysplasia, they are not listed as thumb dysplasia of hyperdactylia; likewise, the thumb dysplasia of mirror hand does not fall into the category of thumb dysplasia of hyperdactylia)
Type VII (deformity of web-shaped hand)	Syndactylia and/or hyperdactylia of the entire hand, the absence of the thumb at the opposition position and the first fingerweb, possibly manifested as five-fingered, six-fingered, and seven-fingered syndactylia, or deformity of finger flexion and thumb hypogenesis
Type VIII (dysplastic thumb dysplasia of the entire hand)	Dysplasia of the entire hand, thumb dysplasia, deformity of short thumb, including multiple types of syndromic thumb dysplasia and short thumb; and the common ones include the Apert syndrome, the Poland syndrome, and spade hand deformity
Type IX (thumb dysplasia of cleft hand)	Central longitudinal ray deficiency, thumb dysplasia, thumb complete absence or deformity, including thumb dysplasia of four-fingered cleft hand, thumb dysplasia of three-fingered cleft hand, thumb dysplasia of two-fingered cleft hand, and thumb dysplasia of one-fingered cleft hand
Type X (thumb dysplasia of ring constriction syndrome)	The limb, body, or skull suffers deformity of ring constriction syndrome accompanied by short thumb, distal defects, ring constriction, congenital thumb deformities, etc.

Table 5.6 Brief classification of congenital thumb dysplasia (10 types and21 classes)

Type I thumb dysplasia: mild dysplasia of the thumb (the thumb is slightly short but functions well)
Type II thumb dysplasia: moderate thumb dysplasia
Type IIa thumb dysplasia: thumb dysplasia + dysplasia of thenar muscles
Type IIb thumb dysplasia: thumb dysplasia + dysplasia of thumb extensor
Type IIc thumb dysplasia: thumb dysplasia + dysplasia of thumb flexor
Type IId thumb dysplasia: moderate dysplasia of thumb phalange and metacarpal bone, with/without amyoplasia
Type III thumb dysplasia: severe thumb dysplasia
Type IIIa thumb dysplasia: type II thumb dysplasia + severe dysplasia of the first metacarpal bone
Type IIIb thumb dysplasia: type II thumb dysplasia + severe dysplasia of the first metacarpal bone and the first carpometacarpal joint
Type IIIc thumb dysplasia: abduction-type thumb dysplasia
Type IV thumb dysplasia: floating thumb
Type V thumb dysplasia: dysplasia of thumb absence
Type VI thumb dysplasia: hyperphalangeal thumb dysplasia
Type VIa thumb dysplasia: five-fingered thumb dysplasia
Type VIaa thumb dysplasia: five-fingered thumb dysplasia (good finger development type)
Type VIab thumb dysplasia: five-fingered thumb dysplasia (short finger type)
Type VIb thumb dysplasia: six, seven and eight-fingered thumb dysplasia
Type VII thumb dysplasia: web-shaped hand deformity
Type VIIa thumb dysplasia: five-fingered web-shaped hand deformity
Type VIIb thumb dysplasia: hyperphalangeal web-shaped hand deformity
Type VIII: dysplastic thumb dysplasia of the entire hand
Type IX thumb dysplasia: thumb dysplasia of cleft hand
Type IXa thumb dysplasia: thumb dysplasia of four-fingered cleft hand
Type IXb thumb dysplasia: thumb dysplasia of three-fingered cleft hand
Type IXc thumb dysplasia: thumb dysplasia of two-fingered cleft hand
Type IXd thumb dysplasia: thumb dysplasia of one-fingered cleft hand
Type X thumb dysplasia: thumb dysplasia of ring constriction syndrome

5.4 Manifestations of Congenital Thumb Dysplasia

5.4.1 Type I Thumb Dysplasia (Mild Thumb Dysplasia)

Type I thumb dysplasia, also called mild thumb dysplasia, has the following manifestations: the thumb has opposition position but is short, small, and narrow compared with the normal side, possibly accompanied by deformity of mild adduction or abduction; function impairment is mild, and the supporting structure and dynamical structure exist. In spite of dysplasia to varying extents, the thumb still has the functions of flexion, extension, abduction, adduction, rotation, and opposition. In addition, the muscular strength may become declined. It falls into the category of deformity of short thumb with functional insufficiency. Type I thumb dysplasia takes various shapes and shows various functional defects. The thumb suffers flexion and extension disorder to varying extents and is different from normal thumbs in terms of the muscular strength of thenar muscles, but the affected hand is capable of performing actions such as gripping, grasping, pinching with two fingers, pinching with three fingers, digital lateral pinching, and opposition. In spite of insufficiency, it is difficult to improve it though a proper surgery, so it is often unnecessary to treat such deformities. The deformity of short thumb in this section only includes the deformity of short thumb in mild thumb dysplasia (Figs. 5.2 and 5.3).

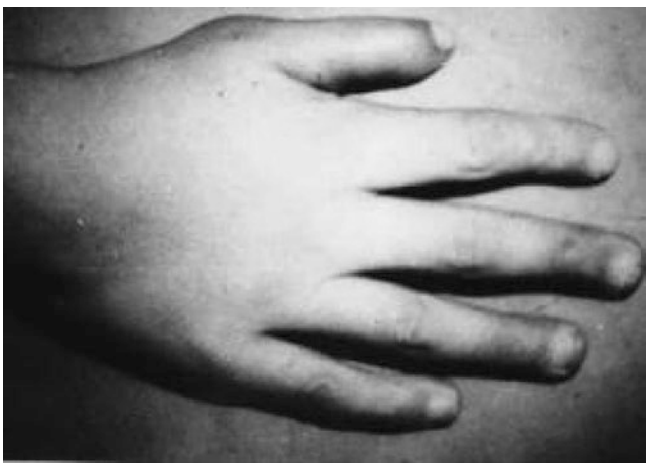


Fig. 5.2 Type I thumb dysplasia of the right hand (mild dysplasia of thumb): the right thumb is slightly short and small but functions well

In some cases, type I thumb dysplasia is only manifested as deformity of lateral deviation at the tip of the thumb; some cases only have the manifestation of too short thumb (Fig. 5.4). Since such deformity is only slightly harmful to the thumb functions, most patients with such deformity give up treatment. As for this type of dysplasia, surgical treatment can be arranged if the patients want to have it corrected. For example, one patient with the angulation deformity at the tip of the thumb and interphalangeal joint suffers rare deformity of lateriflection of thumb interphalangeal joint (Fig. 5.5). Adult patients who pursue better thumb appearance through surgery may wish to have such deformity corrected, and in this case surgery can be performed.

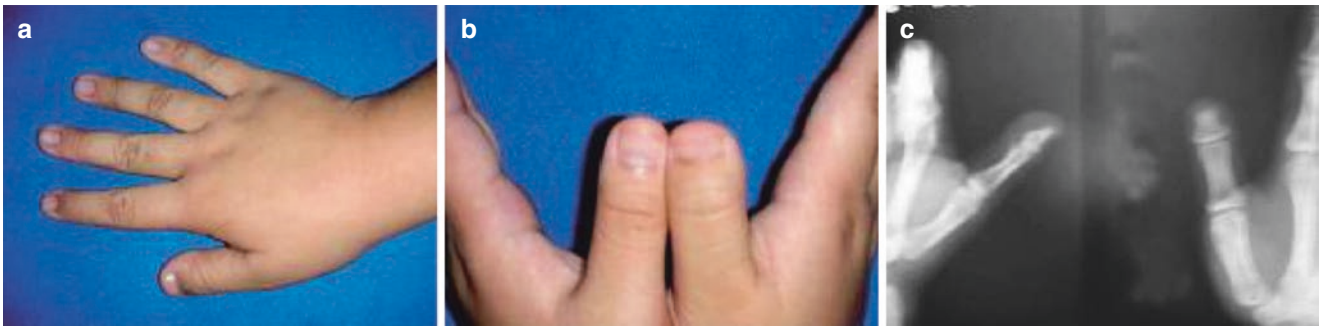


Fig. 5.3 Type I thumb dysplasia of the right hand: mild dysplasia of the right thumb and the right thumb has the deformity of being slightly short and small. (**a**, **b**) The right thumb functions well, and its strength is close to normal, but it is a bit unsightly. (**c**) X-ray film indicates dysplasia of the distal phalanx of the right thumb

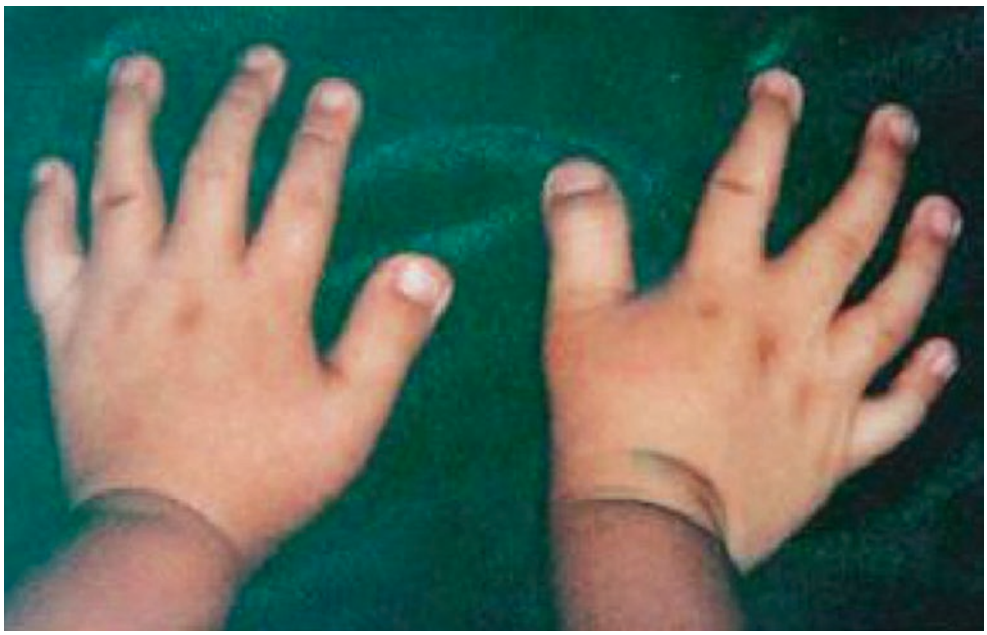


Fig. 5.4 Type I thumb dysplasia: the thumb is relatively short and small, the thickness is close to normal, other fingers are also small, the thumb and hand functions are almost close to normal, and treatment may not be necessary

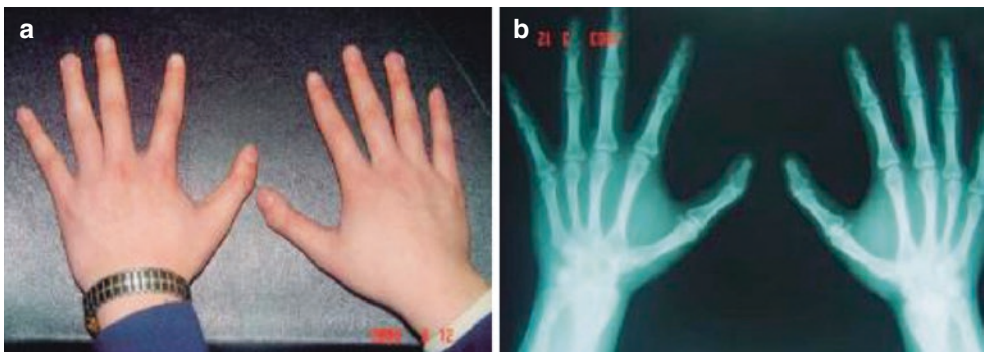


Fig. 5.5 Type I thumb dysplasia: the thumb functions well, and the tip of the thumb and the interphalangeal joint suffer angulation deformity. (**a**) Hand appearance. (**b**) X-ray findings

5.4.2 Deformity of Short Thumb

Type I thumb dysplasia is sometimes called short thumb. In fact, clinically, the concept of short thumb deformity is broader. In the author's classification of thumb dysplasia, short thumb deformity refers only to mild thumb dysplasia, but in clinical practice, it does not refer only to mild thumb dysplasia but also in some cases involves more functional lesions. Short thumb deformity is not rare in hand congenital deformities and is often one symptom of some syndromes. Therefore, a section is purposefully devoted to describing it.

Short thumb deformity is widely present in congenital deformities of hands and upper limbs. Too short thumbs are also often found in type II and type III thumb dysplasia. The severity and manifestations of short thumb deformity vary greatly, and lots of patients with syndrome can concomitantly suffer from the deformity of short and small thumb.

The feature of the short thumb deformity is that the length of the thumb cannot reach the level of interphalangeal joint of the index finger, and the main etiologic factor is dysplasia of the bony supporting structure which leads to the loss of thumb functions to varying extents. This deformity is often accompanied by other congenital deformities.

The X-ray film of the short thumb deformity indicates that, when metacarpal bone is short and fine, the patient may concomitantly suffer from deformities in cardiovascular system, gastrointestinal tract, or spinal column. As for the patients with Fanconi's anemia, short thumb deformity may be accompanied by hemopoietic system diseases. When the thumb is short and the metacarpal bone is short, flat, and broad, the patient may concomitantly suffer from dystrophic dwarf, progressive ossifying myositis, or hand-foot-uterus syndrome. When short thumb deformity is only manifested as short, flat, and broad proximal phalanx, the middle phalanx of each finger is often thick and short, presenting the deformity of brachydactylia and excessively shallow fingerweb. Short thumb deformity characterized by short and flat distal phalanx can exist independently and can be one of the symptoms of certain syndrome, such as the Apert syndrome and the Poland syndrome.

The manifestations and complications of the short thumb deformity are various. Bayne (1982) classified it into some subtypes according to the shape and size of phalange and metacarpal bones, and this method is a classification method of some reference value (Table 5.7).

Patients with short thumb deformities also suffer other deformities in addition to short and small metacarpal bone and phalange. Manske (1995) included short thumb deformities into the category of type IIIa thumb dysplasia (Table 5.8).

Note: The author classifies such deformities into the category of type IIIa thumb dysplasia.

In the above literature review, the author gives a detailed description of the functional defects of short thumb deformities, including thumb web deformity, metacarpophalangeal joint collateral ligament relaxation, thenar muscle dysplasia, and hand external muscle dysplasia (e.g., extensor tendon absence, flexor tendon absence, flexor tendon abnormalities, and mutual combination of tendons), which fall into the category of mild thumb dysplasia (type I), moderate thumb dysplasia (type II), and severe thumb dysplasia (type III).

In the author's clinical practice, in addition to the extensor tendon absence, flexor tendon absence, flexor tendon abnormalities, and mutual combination of tendons that occurs in thumb web deformities, collateral ligament relaxation, thenar muscle dysplasia, and hand external muscle dysplasia that accompany the short thumb deformities of 54 patients reported by other authors as summarized by Manske (1995), the dysplasia of the first carpometacarpal joint and trapezium bones, dysplasia or abnormalities of the first metacarpal bone, and abnormal development of thumb proximal or distal phalanx are often observed. In addition to the metacarpophalangeal joint ulnar accessory ligament relaxation reported in the literature, the crispation of metacarpophalangeal joint accessory ligament can also be observed. Among the 13 patients with short thumb deformities summarized by Manske (1995) in Table 5.8, two patients also concomitantly suffer from mutual combination of hand external tendons. In the author's experience, the incidence of mutual combination of hand external tendons is very high, and it is manifested as

Table 5.7 Subtypes of short thumb deformity

Metacarpal bone—short metacarpal bone and fine metacarpal bone	Metacarpal bone—short metacarpal bone and flat metacarpal bone
Exist independently to varying extents	Cornelia de Lange's syndrome, hand-foot-uterus syndrome
Accompanied by deformities in the spinal column, heart, and digestive system	Dystrophic dwarfism
Accompanied by hemopoietic systemic diseases (e.g., Fanconi's anemia), with the manifestation of pancytopenia and radial defect (TA syndrome)	Progressive ossifying myositis
Holt-Oram syndrome: the hand suffers obvious deformities, accompanied by deformities in the hemopoietic system, urogenital system, heart, or musculoskeletal system	Short, flat, and broad proximal phalanx
Juberg-Hayward syndrome is a rare genetic syndrome of endocrine development abnormalities, manifested as chilopalatognathus, microcephaly, limb abnormality, thumb shortness and absence	Brachydactyly
	Short, flat, and broad distal phalanx
	Rubinstein-Taybi syndrome, including short and flat thumb and toes, facial deformities, and dysgnosia
	The Apert syndrome and acrosphenosyndactylia
	Carpenter's syndrome, brachydactylia, and syndactylia deformities and craniofacial deformity

not only mutual combination of thumb extensor and flexor tendons but also mutual combination of the tendons of thumb and other fingers and abnormalities in tendon insertions.

The patients with dysplasia of the entire hand often suffer short thumb deformities, such as congenital hand dysplasia, the Poland syndrome, and the Apert syndrome (Figs. 5.6 and 5.7).

Table 5.8 Clinical manifestations of congenital short thumb deformities (literature review)

Author (year)	Number of cases	Thumb web deformity	Ulnar accessory ligament relaxation	Thenar muscle dysplasia	Dysplasia of hand external muscles			
					Extensor tendon absence	Flexor tendon absence	Flexor tendon abnormalities	Mutual combination of tendons
Tupper (1969)	4	Presence	Presence	Presence	Presence	Presence	Presence	
Strauch (1976)	7	Presence	Presence	Presence	Presence	Presence	Presence	
Nevaiser (1979)	10			Presence	Presence	Presence	Presence	
Blair (1981)	1	Presence	Presence		Presence	Presence		
Blair (1983)	1	Presence	Presence	Presence	Presence	Presence	Presence	
Fitch (1984)	3	Presence	Presence	Presence			Presence	
Rayan (1984)	4	Presence	Presence	Presence	1	3	2	
Lister (1991)	11	Presence	Presence	Presence			Presence	
Manske (1995)	13	Presence	Presence	Presence	8	6	6	2

Originate from Joseph Upton, MD. Management of transverse and longitudinal deficiencies (failure of formation). Plastic Surgery Second Edition. Edited by Stephen J. Mathes. MD. Philadelphia. Vol. 8. 2006:51–137

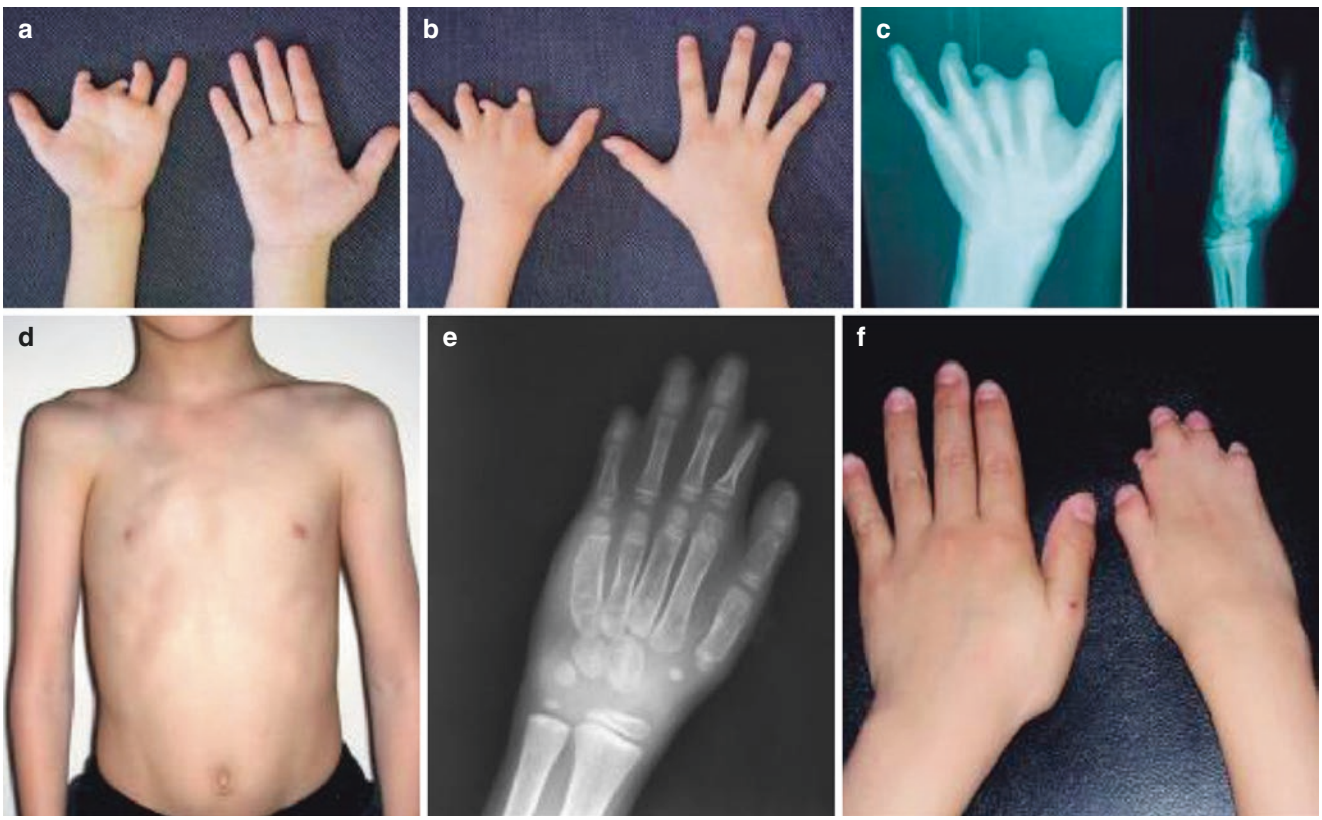


Fig. 5.6 Congenital hand dysplasia (brachydactylia deformity). (**a**–**c**) The Poland syndrome, the left thumb is shorter and smaller than the right thumb, accompanied by congenital brachydactylia of the index, middle, and ring fingers. (**d**–**f**) The Poland syndrome, the right thumb is short and small, accompanied by congenital brachydactylia of the index, middle, and ring fingers and the dysplasia of the right major and minor pectoral muscles

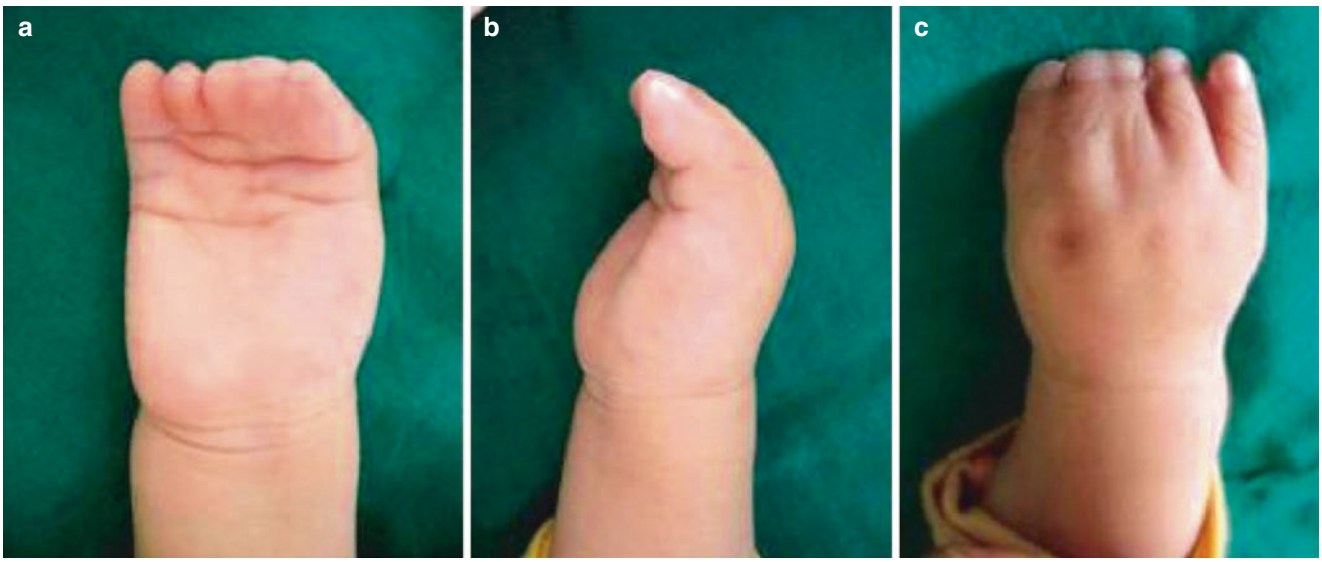


Fig. 5.7 The Apert syndrome (congenital right hand dysplasia and spade hand deformity)

5.4.3 Type II Dysplasia of Thumb (Moderate Dysplasia of Thumb)

5.4.3.1 Clinical Manifestation

Type II dysplasia of thumb, also called moderate dysplasia of thumb, is characterized by thumb dysplasia, accompanied by dysplasia of thenar muscles or thumb flexor and extensor, which obviously damages the dynamical functions of the thumb, mild to moderate dysplasia of the supporting structures of phalange and metacarpal bone, and not obviously damage in the functions of the thumb-supporting structures. The specific manifestations are as follows:

1. Abnormal shape. The thumb is short and small or is manifested as thumb adduction and thumb web stenosis.
2. Dysplasia of dynamical structure It includes dysplasia of thenar muscles, thumb flexor, or thumb extensor to varying extents.
3. Impaired supporting and stable structures. The supporting structures include mild to moderate dysplasia in phalange, metacarpal bone, metacarpophalangeal joint, and carpometacarpal joint, and the stable structures might suffer damages to varying extents, including relaxation of metacarpophalangeal joint accessory ligament, poor stability of the carpometacarpal joint, and existence but mild to moderate deformities of the phalange and metacarpal bone of the thumb.
4. Impaired functions. Dysplasia in thumb flexion, extension, abduction, adduction, rotation, and opposition functions and muscular strength as well as significant drop in the finger-to-finger pinch strength, digital lateral pinch strength, three-digit pinch strength, and grip strength.

5.4.3.2 Classification

Due to the differences in deformity degree, type II thumb dysplasia can be reclassified into the following subtypes:

1. Type IIa thumb dysplasia: moderate thumb dysplasia + dysplasia of thenar muscles:
   (a) Clinical manifestations: Fine and small thumb with adduction, thumb web stenosis, obvious dysplasia of thenar muscles, flat palmar surface and radial side of the first metacarpal bone, absence or serious dysplasia of thumb thenar muscles, thumb function defect, palm-opposition and finger-opposition hypofunction, and obvious drop in grip strength, finger-to-finger pinch strength, digital lateral pinch strength, and three-finger pinch strength. In spite of dysplasia in the supporting structures of thumb phalange, metacarpal bone, metacarpophalangeal joint, and carpometacarpal joint, there is no serious impairment and the joint stability is fair (Fig. 5.8).
   (b) Typical case: Case one, a 2-year-old male, with radial hyperdactylia in the right hand and dysplastic and fine thumb of the left hand, but the position is good, the nail plane and other four fingers present the opposition position accompanied by the dysplasia of thenar muscles, thumb adduction, lack of strength in pinching and grasping, limited palm-opposition and finger-opposition functions, and stenosis in the first fingerweb local examination: the absence of palm thenar muscles, subcutaneously palpable metacarpal bones, obviously fine thumb, relative small range of motion of interphalangeal joint, limited thumb abduction, and incapability of palm opposition. The patient suffers functional disorder in pinching, grasping, and gripping, and the deformed

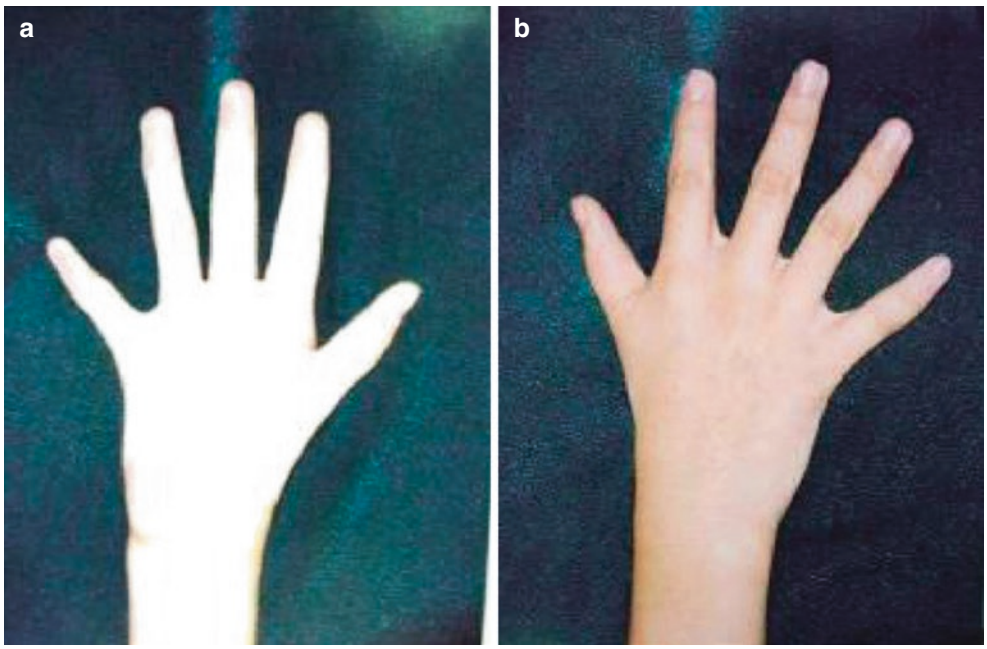


Fig. 5.8 Type IIa thumb dysplasia: adduction-type thumb dysplasia

thumb depends on the ulnar surface and the index finger or the index and middle fingers to grip objects. With the increase in age, both the deformity of thumb adduction and the stenosis and contracture of the first fingerweb became aggravated. X-ray film indicates that the phalange and the first metacarpal bone of the left thumb are finer and smaller than the right thumb and suffer dysplasia, and the distal phalanx of the left thumb suffers mild ulnar deviation. During the reconstruction of thumb opposition functions, the thumb short abductor, thumb opposing muscle, and thumb short flexor are found absent, and the thumb adductor is contractural. Then the thumb web is expanded to reconstruct the thumb opposition functions. The tendon of the superficial digital flexor of the ring finger is translocated and fixed at the volar insertion of the thumb abductor, and after the operation, the thumb is fixed at the abduction and opposition positions (Fig. 5.9).

2. Type IIb thumb dysplasia: moderate thumb dysplasia + dysplasia of thumb long extensor:
   (a) Clinical manifestations: Short, small, and fine thumb; stenotic thumb web; thumb adduction; dysplasia of thumb extensor and hand external muscles, with/without dysplasia of thenar muscles; flexion of thumb distal segment or metacarpophalangeal joint, possibly accompanied by mild or moderate dysplasia of the supporting structures of phalange and metacarpal bone; and relatively small phalange and the first metacarpal bone. Dysplasia of metacarpophalangeal joint and carpometacarpal joint, abnormal shape, relaxation of metacarpophalangeal joint accessory ligament, poor stability but existence of basic structures. The absence or obvious weakening of thumb extension functions, with abduction, adduction, rotation, and palm-opposition functional disorder and muscular strength hypoplasia when accompanied by thenar muscle dysplasia. Obvious drop in thumb extension strength, finger-to-finger pinch strength, digit lateral pinch strength, three-finger pinch strength, and grip strength and loss of palm-opposition function of the thumb when accompanied by dysplasia of thenar muscles. As for this category of dysplasia, different patients have different manifestations.
   (b) Typical cases:
   - Case two: Deformity of adduction of two thumbs, thumb dysplasia, dysplasia of thumb long extensor tendons, accompanied by thenar muscular dysplasia, stenosis of the first fingerweb, angulation deformity of thumb interphalangeal joint, moderate dysplasia of phalange and metacarpal bone, and the presence of metacarpophalangeal joint and carpometacarpal joint (Fig. 5.10)
   - Case three: Adduction deformity of the right thumb; dysplasia of thumb long extensor tendon, accompanied by thenar muscular dysplasia, with the manifestations of fineness and adduction of the right thumb; stenosis of thumb web; flexion of the interphalangeal joint; subcutaneous emptiness of the first metacarpal area; dysplasia of phalange and the first metacarpal bone of the thumb; the presence of metacarpophalangeal joint and carpometacarpal joint with poor stability; and moderate dysplasia of supporting structures and stable structures of phalange and metacarpal bone (Fig. 5.11).
   - Case four: Adduction of two thumbs; dysplasia of thumb extensors and hand external muscles, accompanied by thenar muscular dysplasia; thumb web steno-

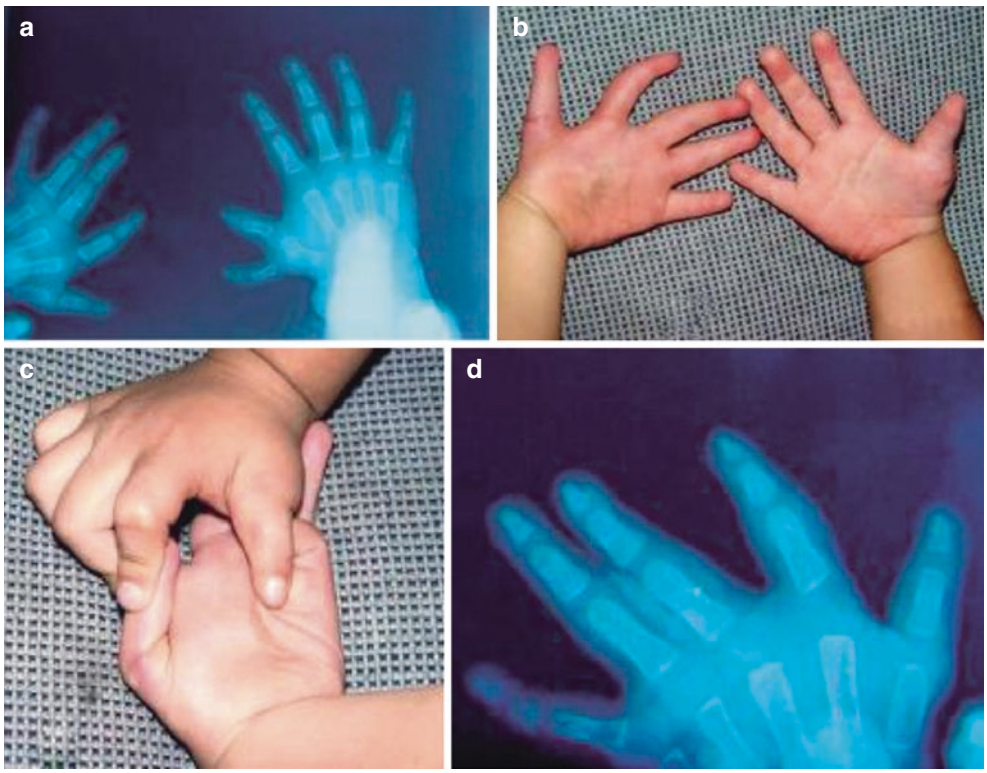


Fig. 5.9 Case one: type IIa thumb dysplasia of the left hand. (**a**) X-ray indicates radial hyperdactylia of the right hand (deformity of duplication of the thumb), type IIa left thumb dysplasia. (**b**) At 3 months after surgical resection of radial hyperdactylia of the right hand. (**c**) The left thumb cannot perform finger-opposition and palm-opposition actions, and the assistance of the right hand is needed. (**d**) X-ray film indicates mild dysplasia of the phalange and metacarpal bone of the left thumb

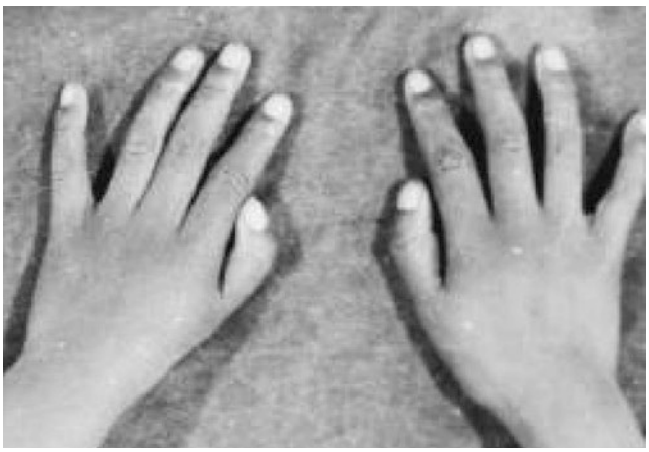


Fig. 5.10 Case Two: type IIb thumb dysplasia of two hands (adduction-type thumb dysplasia)

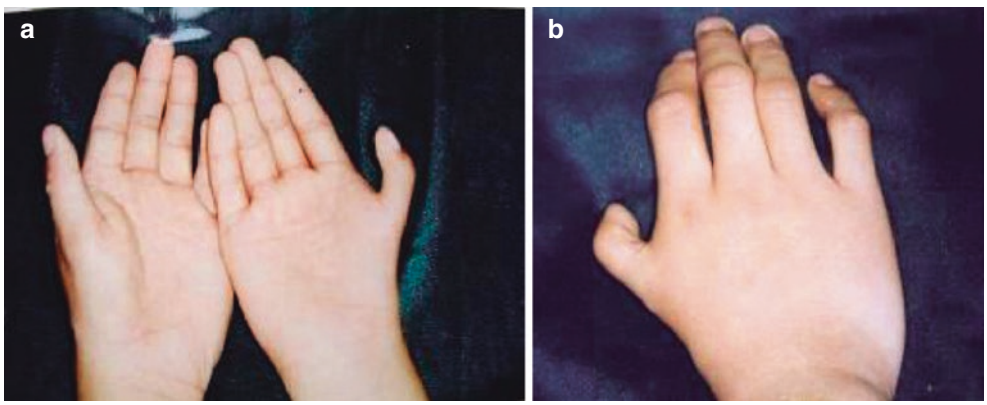


Fig. 5.11 Case three: type IIb thumb dysplasia (adduction-type thumb dysplasia)

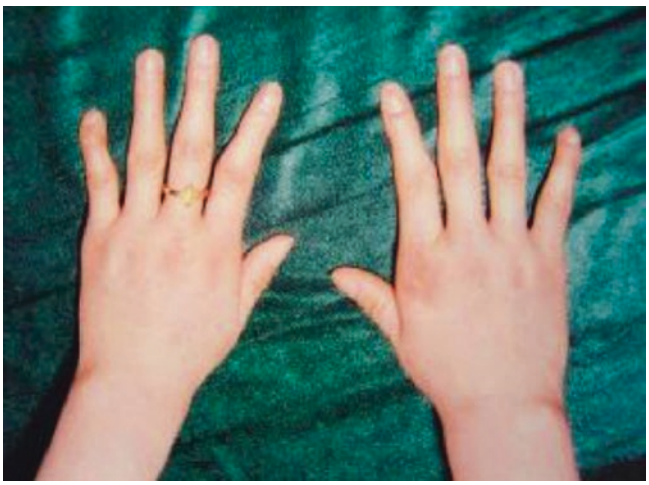


Fig. 5.12 Case four: type IIb thumb dysplasia (adduction-type thumb dysplasia)

sis; deformity of lateral curvature of bilateral index fingers and little fingers; still fair shape and anatomical structures of the thumb; the presence of phalange and the first metacarpal bone of the thumb; moderate dysplasia of supporting structures and stable structures; dysplasia of flexion, extension, abduction, adduction, rotation, and palm-opposition functions of thumb; and hypoplasia of muscular strength (Fig. 5.12).

3. Type IIc thumb dysplasia: moderate thumb dysplasia + dysplasia of thumb long flexor. Type IIc thumb dysplasia is characterized by congenital dysplasia of thumb long flexor tendon accompanied by thumb short flexor dysplasia.
   (a) Clinical manifestations: Shortness and adduction of thumb, stenosis of thumb web, and thumb flexor dysplasia, with/without thenar muscular dysplasia. The presence of thumb extensor functions to varying extents, dysplasia of thumb flexor tendon, and/or abnormal insertion position. The presence of bone support structures of the thumb, which are small; abnormal shape of the phalange, metacarpal bone, metacarpophalangeal joint, and carpometacarpal joint; and relaxation of metacarpophalangeal joint accessory ligament, with relatively poor stability. Limited or no thumb flexion function; insufficiency in thumb lateral flexion, extension, abduction, adduction, rotation, and opposition functions and muscular strength; and obvious drop in grip strength, finger-to-finger pinch strength, digital lateral pinch strength, and three-digit pinch strength.
   (b) Typical case: Sun Yufang et al. (2006) [13] reported one 3-year-old girl suffering congenital thumb long flexor dysplasia with limited functions. Case five: The right thumb suffers dysplasia, the little finger is under abduction status, the interphalangeal joint cannot actively flex, the thenar muscles suffer dysplasia, and the appearance and motion of the remaining four fingers are normal. X-ray film indicates that the thumb skeletons are relatively small and diagnosed as congenital thumb long flexor tendon abnormality (Fig. 5.13). It is found during operation that the thumb long flexor tendon is drawn to the radial lateral side by one aponeurosis-shaped structure, then it runs along the thumb radial side and toward the distal end and becomes bifurcated into two sides at the interphalangeal joint, the palmar head ends at the volar distal phalanx base (the normal insertion is near the radial side), and the dorsal head takes patchy shape and ends at the radial lateral distal phalanx base (Fig. 5.14). Do the following during operation: remission of aponeurosis-shaped structures, dissociation of thumb long flexor tendon, cutting of dorsal head from the bifurcation position, cutting of the abnormal insertion, and removal of this tendon for future use. Expose the thumb short abductor at the metacarpophalangeal joint, break the tendinous part, move the thumb long flexor until it is under the thumb short abductor, and intermittently suture the thumb short abductor. Use an alternate tendon to form a trochlea at the volar side of the thumb long flexor tendon at the distal end of metacarpophalangeal joint (at the site of interphalangeal

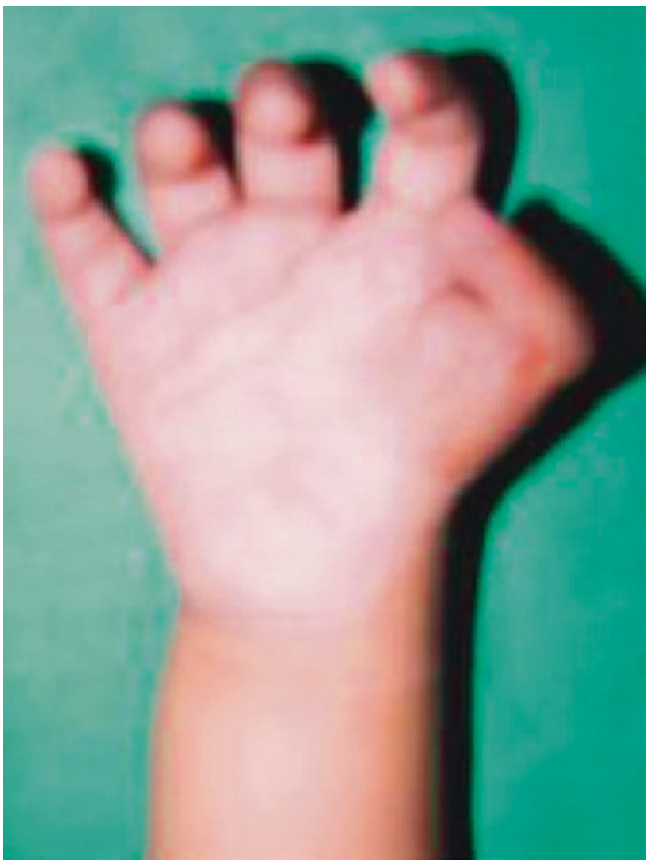


Fig. 5.13 Case five: type IIc thumb dysplasia (congenital thumb long flexor tendon abnormalities)

joint) and maintain the position of the thumb long flexor tendon in the volar median aspect. Examine whether the flexor tendons slide freely, passively tract thumb long flexor tendon, and subsequently the thumb is able to flex. The possible manifestations may be thumb brevis flexor dysplasia and thumb adduction deformities, and there are similar cases in Lister's (1985) [14] report and the author's clinical practice.

4. Type IId thumb dysplasia. The frequent thumb long extensor dysplasia in the thumb dynamical structure + moderate dysplasia in the thumb-supporting structures. No literature has included this deformity into the category of type II thumb dysplasia, but there are clinical cases with this type of thumb dysplasia.
   (a) Characteristics: Shortness and adduction of thumb, induced by dysplasia of hand internal muscles of the thumb or accompanied by dysplasia of hand external muscles. The thumb phalange and metacarpal bone suffer dysplasia, and this type of bone scaffold structures does not cause serious functional impairment to the deformed thumb and only manifests moderate impairment.
   (b) Clinical manifestations: The dynamical structure of thumb—dysplasia of hand internal muscles or hand external muscles, accompanied by moderate dysplasia in the phalange, metacarpal bone, metacarpophalangeal joint, and carpometacarpal joint of the thumb. Mild abnormality in the shape and structure of the thumb, and insufficient flexion, extension, abduction, adduction, rotation, and palm-opposition functions.
   (c) Typical case: Case six, a 5-year-old female, with type IId thumb dysplasia, short and small thumb and little finger, deformity of thumb adduction, thenar and hypothenar muscles, and thumb long extensor dysplasia, accompanied by deformities of the proximal phalanx, the first metacarpal bone, and metacarpophalangeal joint. The thumb has extension, flexion, adduction, and abduction functions, but the palm-opposition function is insufficient. Her deformed thumb cannot touch the little finger, but can touch the ring finger, and at the same time, the finger-to-finger pinch strength of the thumb is too low. In spite of functional impairment, the impairment is not serious. X-ray film indicates that her right thumb is short and small, the phalange and the first metacarpal bone of the thumb are relatively fine and are finger-type metacarpal bones, and the epiphysis is at the distal end of metacarpal bone. The metacarpophalangeal joint suffers mild deformities, and the two segments of phalange and proximal phalanx suffer deformities. The surgery is likely to improve the functions of deformed thumbs. After the patient was informed of the state of illness and the therapeutic regimen, the patient chose nonsurgical treatment (Fig. 5.15).

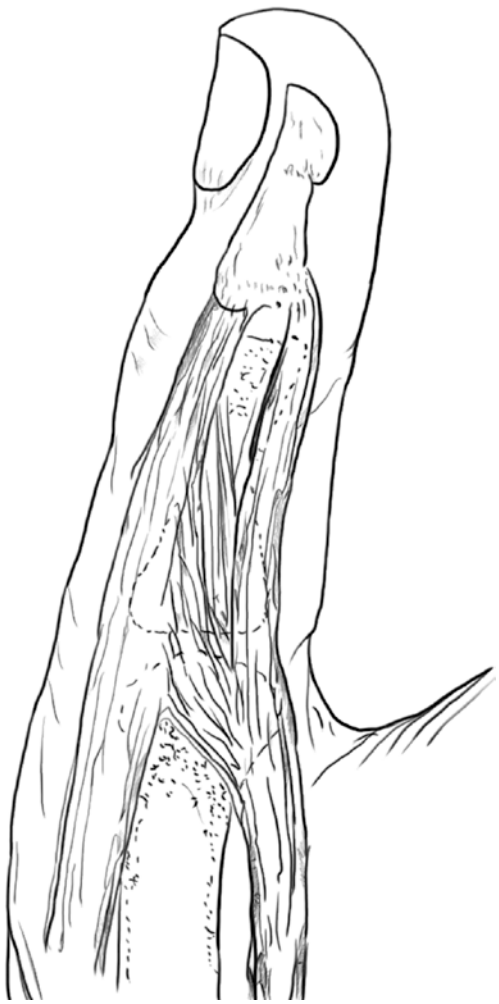


Fig. 5.14 Type IIc thumb dysplasia (the thumb long flexor tendons are divided into two parts, which end at the flexor aspect and extensor aspect of the thumb)

5.4.4 Type III Hypoplasia of Thumb (Severe Hypoplasia of Thumb)

Type III hypoplasia of thumb, also called severe dysplasia of thumb, refers to the type II thumb hypoplasia + severe dysplasia in the skeletal supporting structures (the first metacarpal bone and metacarpophalangeal joint), dynamical structures, and stable structures of the thumb.

5.4.4.1 Clinical Manifestation

Severe thumb dysplasia refers to severe dysplasia in the dynamical structures and supporting structures of the thumb, manifested as thumb adduction; dysplasia in thenar muscles; dysplasia in hand external muscles, especially dysplasia in

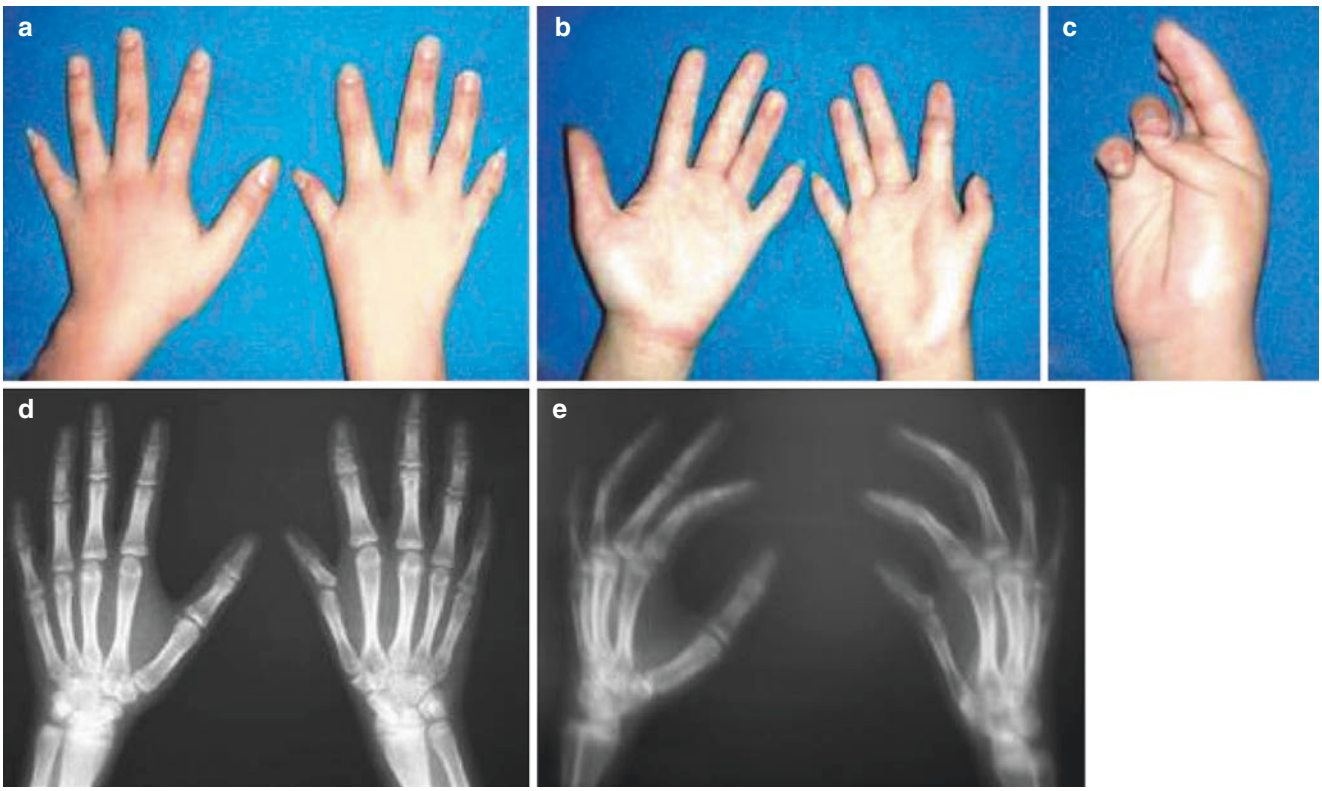


Fig. 5.15 Case six: type IId thumb dysplasia of the right hand. (**a**–**c**) Appearance of the two hands. (**d**, **e**) X-ray findings

the thumb long extensor; severe dysplasia in thumb phalange and metacarpal bone; and dysplasia in part or most of the metacarpophalangeal joint and/or carpometacarpal joint. The thumb is in the palm-opposition position, the first metacarpal bone is sometimes the finger-type metacarpal bone, the epiphysis is located at the distal end of metacarpal bone, and triphalangeal and triangular phalanx deformities can be simultaneously found. Type III thumb dysplasia often leads to severe pollical functional insufficiency.

5.4.4.2 Classification

According to the degree of dysplasia, type III thumb dysplasia can be reclassified into the following three subtypes:

1. Type IIIa thumb dysplasia. Also called type IIIa severe adduction-type thumb dysplasia, it refers to type II moderate thumb dysplasia + severe dysplasia of the first metacarpal bone. Its manifestations include short and small thumbs and adduction deformity, accompanied by thenar muscular dysplasia, thumb extensor dysplasia, severe dysplasia of thumb phalange and metacarpal bone but the presence of carpometacarpal joint. There may also be deformities of triphalangeal, triangular middle phalanx, and interphalangeal joint deformities, but they are different from the five-fingered thumb absence deformity, in which the dysplastic thumb and other fingers are in the palm-opposition position. The flexion, extension, adduction, abduction, rotation, and palm-opposition functions of the thumb are obviously limited, and the muscular strength drops significantly. The difference between it and type IIIb thumb dysplasia is that type IIIa suffers no severe dysplasia of the first carpometacarpal joint.
2. Type IIIb thumb dysplasia. Also called type IIIb severe thumb dysplasia, it refers to type II moderate thumb dysplasia + dysplasia of the first metacarpal bone + severe dysplasia of the first carpometacarpal joint:
   (a) Clinical manifestations: Deformities of short, fine, narrow, and adducted thumb, dysplasia in the supporting structures and dynamical structures of the thumb, obvious dysplasia or partial absence of the first metacarpal bone, dysplasia or the absence of carpometacarpal joint, unstable metacarpocarpal joint, dyplasia of extensor tendon, and severe impairment in supporting and dynamical functions of the thumb.
   (b) Typical cases:
   - Case one: The author once admitted one pediatric patient with congenital severe thumb dysplasia in both hands. The right hand of the patient suffered type IIIa thumb dysplasia, and the left hand suffered type IIIb thumb dysplasia, manifested as deformity of

adducted thumb, thenar muscular dysplasia, thumb extensor dysplasia, serious dysplasia of thumb phalange and metacarpal bone, triphalangeal deformities accompanied by triangular phalanx deformities, and interphalangeal joint deformities, but not five-fingered thumb absence deformity. The dysplastic thumb and other fingers were in the palm-opposition position; the thumb flexion, extension, adduction, abduction, rotation, and palm-opposition functions were obviously poor; the muscular strength dropped significantly; the X-ray film indicated that the two thumbs suffer the deformity of adduction; and the right hand suffered type IIIa thumb dysplasia and the left hand suffered type IIIb thumb dysplasia, which were the short thumb deformity of severe functional disorder type (Fig. 5.16).

Very coincidentally, Kaissi (2007) reported two pediatric patients who were identical twins suffering congenital thumb dysplasia of right hand and left hand, respectively. Their hand deformities were similar to those of the author's above cases, that is, the deformities of the two hands of the pediatric patients admitted by the author occurred to the left hand and right hand of the two pediatric patients (who were identical twins) reported by Kaissi [15].

- Case two: Type IIIb thumb dysplasia of the right hand, small, fine, short, and soft right thumb, accompanied by thenar muscular dysplasia. Due to the severe dysplasia of the carpometacarpal joint, there was no tension for the support, and the residual functions of the thumb were similar to those of the floating thumb (Fig. 5.17).
- Case three: Type IIIb thumb dysplasia of the right hand, fine and small thumb, severe dysplasia in the phalange and the first metacarpal bone, and the absence of carpometacarpal joint, which was nonfunctioning thumb deformity (Fig. 5.18).

3. Type IIIc thumb dysplasia. Most literature has no classified description of type IIIc thumb dysplasia but lists this type of thumb dysplasia as abduction-type thumb dysplasia. According to the author, as such thumb dysplasia is often accompanied by abnormalities in the position, shape, structure, and dynamical functions of the carpometacarpal joint, hand functions are seriously affected, it is difficult to incorporate them into type IIIa or IIIb thumb dysplasia, and it is relatively suitable to include it into type IIIc thumb dysplasia. The author once treated a large number of pediatric patients with type IIIc thumb dysplasia during clinical practice.

(a) Clinical manifestations: Phalangeal dysplasia of the thumb, dysplasia of the first metacarpal bone, dysplasia of the carpometacarpal joint, and abnormal position in the first metacarpocarpal joint. The features are serious dysplasia of the thumb, abduction deformity, abnormal presence of osteoarticular scaffold, and effects on the normal functions of hands.

Such deformity is not rare clinically and changes greatly, with the manifestations of dysplasia and anamorphosis. In the case of type IIIc thumb dysplasia, despite the existence of the thumb, the position is abnormal; the carpometacarpal joint is located at the site of the proximal row of carpal bone accompanied by thenar muscular dysplasia, which is the deformity of nonfunctioning thumb and seriously incompetent thumb. The presence of deformed thumb affects the functions of other fingers to some extents.

The manifestations of the shape, structure, and functions of abduction-type deformed thumb are various. If such deformity is mild, it is just enough to repair the shape and dynamical functions of the thumb of the patient. If such deformity is severe, complete resection and thumb reconstruction should be performed.

(b) Typical cases:

- Case four: Type IIIc thumb dysplasia of the left hand, thenar muscular dysplasia, limited adduction function of the thumb, finger extensor dysplasia, and limited flexion of the metacarpophalangeal joint of four fingers of the left hand, with the manifestations of

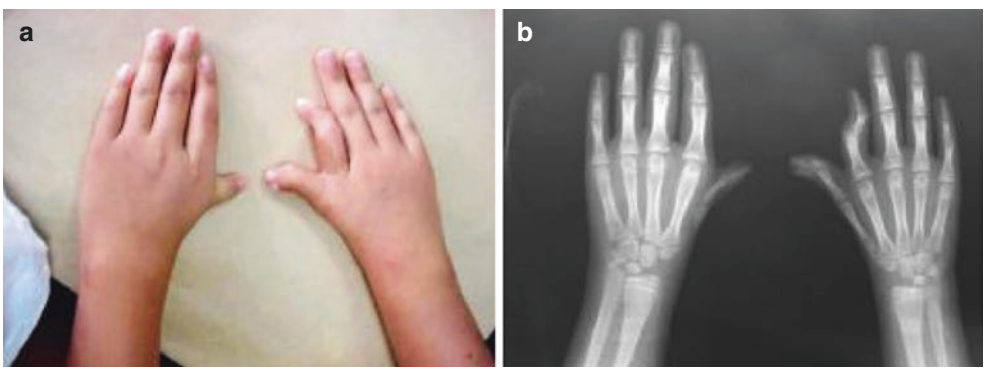


Fig. 5.16 Case one: type IIIa thumb dysplasia of the right hand, type IIIb thumb dysplasia of the left hand. (**a**) Hand appearance. (**b**) X-ray findings

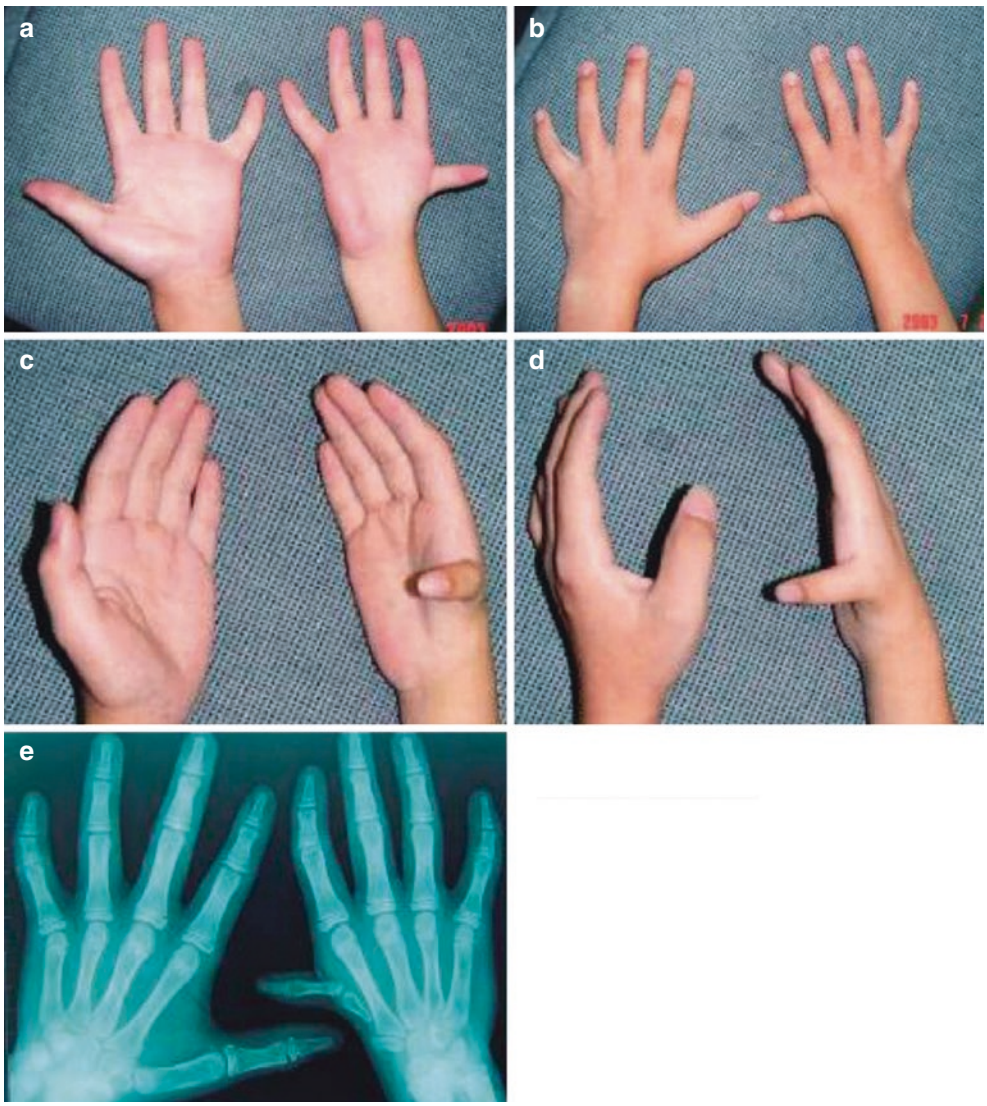


Fig. 5.17 Case two: type IIIb thumb dysplasia of the right hand. (**a**–**d**) Hand appearance. (**e**) X-ray findings

type II thumb dysplasia, phalangeal dysplasia, dysplasia of the first metacarpal bone, abnormal development of the first carpometacarpal joint, and deformity of abducted thumb (Fig. 5.19).

- Case five: Type IIIc thumb dysplasia of the right hand; fine, small, short, and flat thumb; serious dysplasia of the phalange and metacarpal bone; and dysplasia of thenar muscles and hand external muscles; although the thumb is in the abduction position and palm-opposition position, it basically has no function (Fig. 5.20).
- Case six: Type IIIc thumb dysplasia of both hands, dysplasia in the phalange and metacarpal bone, abnormal development of the first metacarpophalangeal joint and carpometacarpal joint, and deformity of abducted thumb and thenar muscular dysplasia, which is the deformity of nonfunctioning thumb and the seriously incompetent thumb (Fig. 5.21).
- Case seven: Type IIIc thumb dysplasia of the right hand, the first carpometacarpal joint at the site of the proximal row of carpal bone, deformity of abducted thumb, thenar muscular dysplasia, and complete loss of thumb functions. The existence of deformed thumb can affect hand functions to some extents (Fig. 5.22).
- Case eight: Type IIIc thumb dysplasia of both hands, dysplasia in phalange and metacarpal bone, abnormal development of the first metacarpophalangeal joint and carpometacarpal joint, and deformity of abducted thumb. In spite of the presence of the thumb, the thenar muscles and the thumb flexor are dysplastic, the finger skin contracts, and it is the deformity of nonfunctioning thumb and seriously incompetent thumb.

Such thumb deformity is often accompanied by thenar muscular dysplasia, deformities in the phalange and

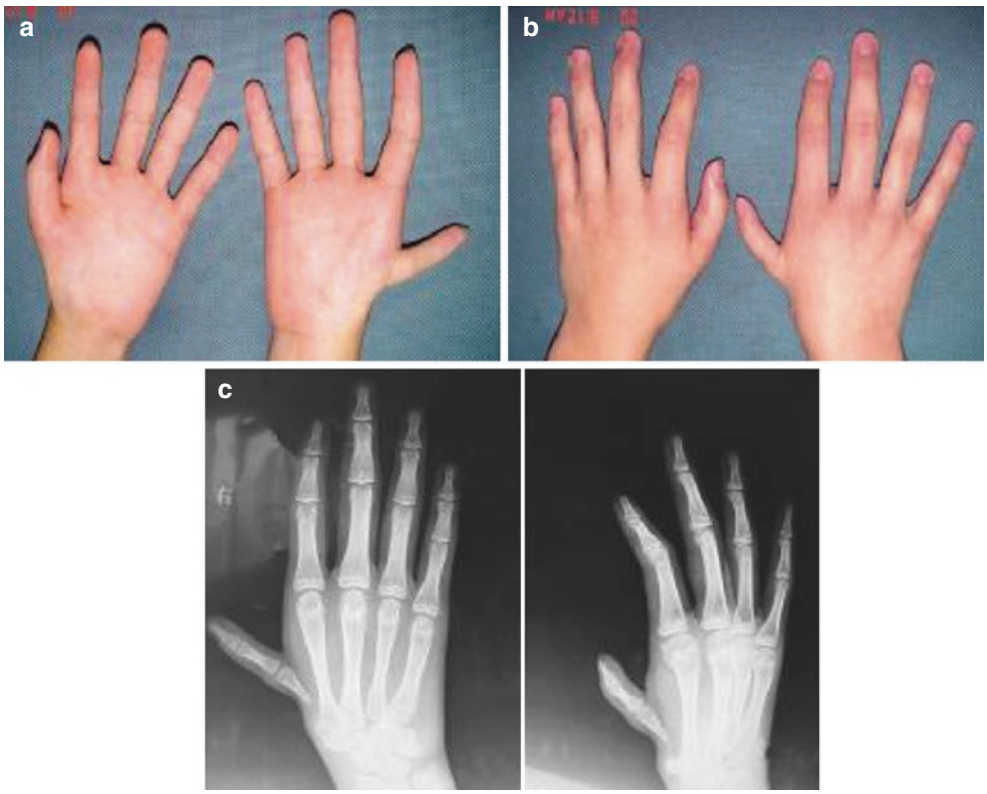


Fig. 5.18 Case three: type IIIb thumb dysplasia of the right hand. (**a**, **b**) Hand appearance. (**c**) X-ray findings

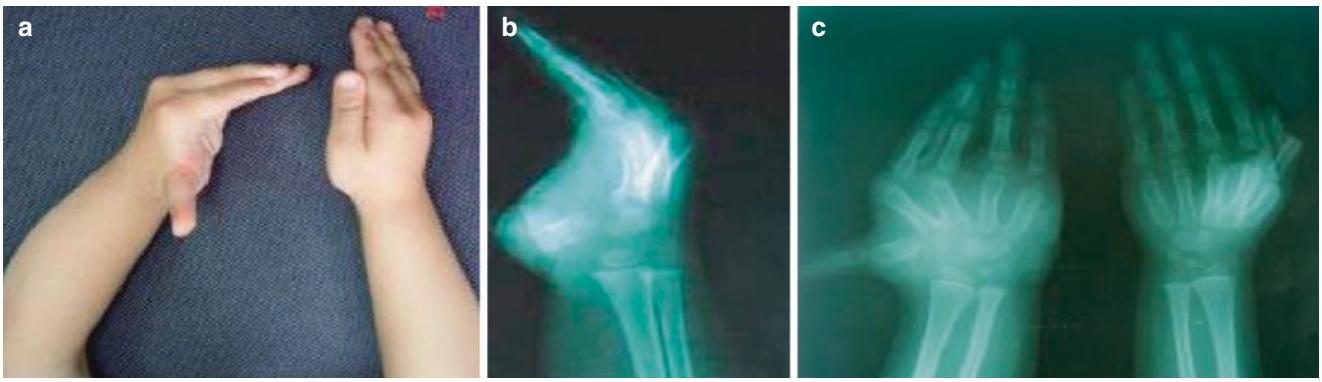


Fig. 5.19 Case four: type IIIc thumb dysplasia of the left hand. (**a**) Hand appearance. (**b**, **c**) X-ray findings

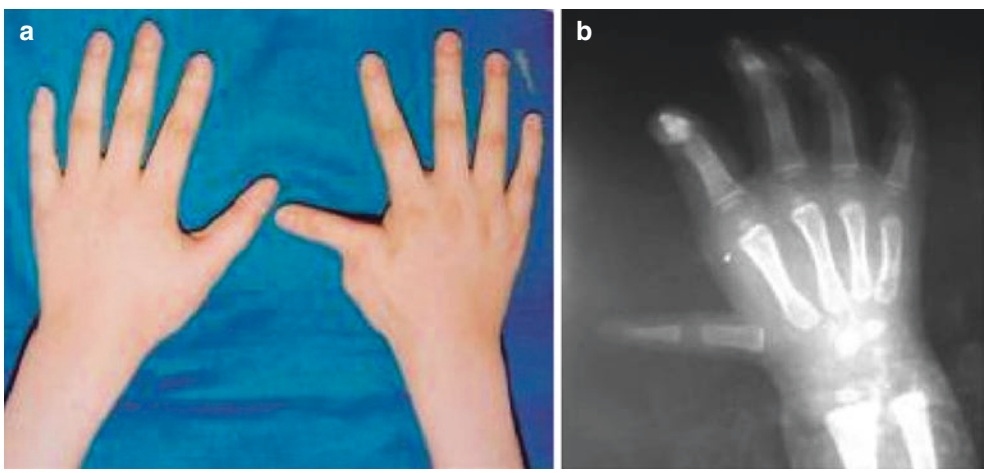


Fig. 5.20 Case five: type IIIc thumb dysplasia of the right hand. (**a**) Hand appearance. (**b**) X-ray findings

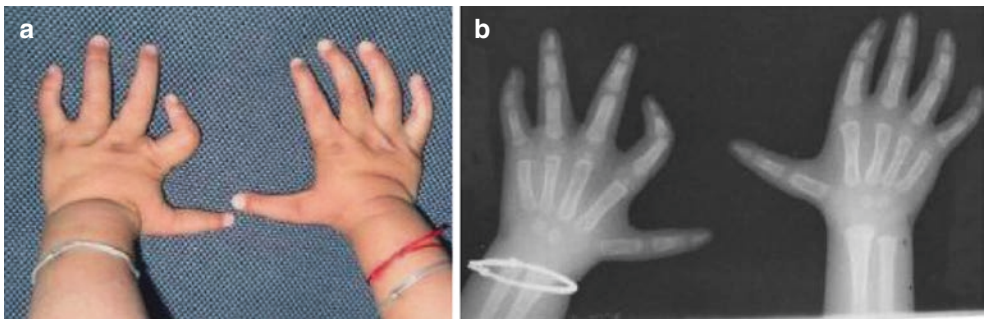


Fig. 5.21 Case six: type IIIc thumb dysplasia of both hands. (**a**) Hand appearance. (**b**) X-ray findings

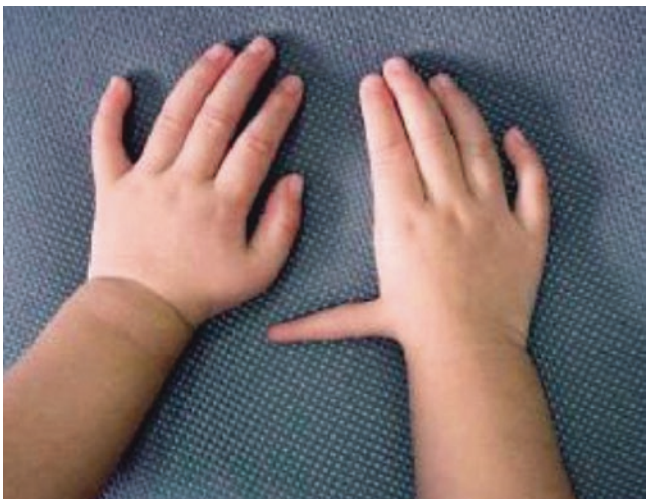


Fig. 5.22 Case seven: type IIIc thumb dysplasia of the right hand

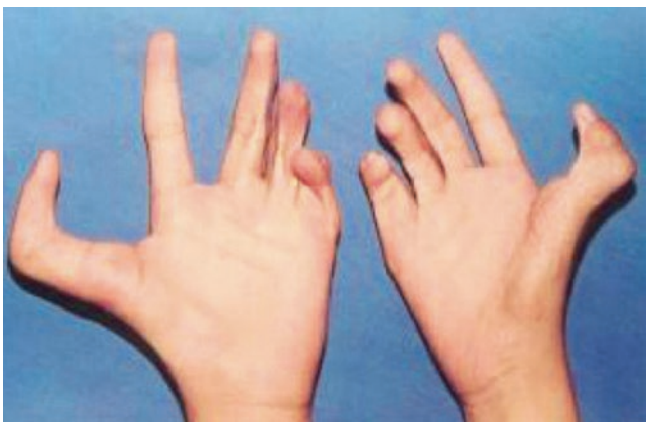


Fig. 5.23 Case eight: type IIIc thumb dysplasia of both hands

metacarpal bone, and deformities in the metacarpophalangeal joint, interphalangeal joint, and carpometacarpal joint. The thumb with the thumb weblike first fingerweb is in the palm-opposition position, the skin on the volar surface of the fingers contracts, and the deformity is not the simple thumb deformity or dysplasia, but type IIIc thumb dysplasia mainly characterized by deformity of abducted thumb (Fig. 5.23).

5.4.5 Type IV Thumb Dysplasia (Floating Thumb)

5.4.5.1 Clinical Manifestations

Type IV thumb dysplasia, also called floating thumb, is manifested as the presence of thumb as soft as silkworm at the radial side of the hand. The phalange at the tip of thumb and proximal phalanx may be present but are very fine and small with serious dysplasia. The first metacarpal bone is absent or seriously dysplastic, and the first metacarpophalangeal joint and carpometacarpal joint are absent. The floating thumb can be connected with hands through many skin bridges as there are nerves and vessels inside the skin bridges; or it can be connected with hands through few skin bridges. The pedicles of the skin bridges are mostly located near the carpometacarpal joint, and they can also be located on the plane of proximal phalanx of the index finger or the proximal part of the metacarpocarpal joint (Fig. 5.24). The floating thumb completely has no function. Type IIIb and IIIc thumb dysplasia can be considered as the mild floating thumb.

5.4.5.2 Typical Cases

An infantile patient; the left hand suffers type IV thumb dysplasia, close to the floating thumb, and serious dysplasia of the phalange, metacarpal bone, carpometacarpal joint, and metacarpophalangeal joint; and the right hand suffers type VII deformity of duplication of the thumb (Fig. 5.25).

5.4.6 Type V Thumb Dysplasia (Thumb Aplasia-Absent Thumb)

Type V thumb dysplasia, also called thumb aplasia-absent thumb, is manifested as four-fingered hand and lack of the thumb in the palm-opposition position. The four fingers develop well, and the appearance, anatomic structure, supporting structure, and dynamical structure and functions of the fingers are often normal. It is the most typical kind of deformity in congenital thumb dysplasia and manifested as congenital complete defect of thumb both in structure and shape, including the absence of thumb proximal and distal phalanx, the first metacarpal bone, metacarpophalangeal joint, carpometacarpal joint and thenar muscle.

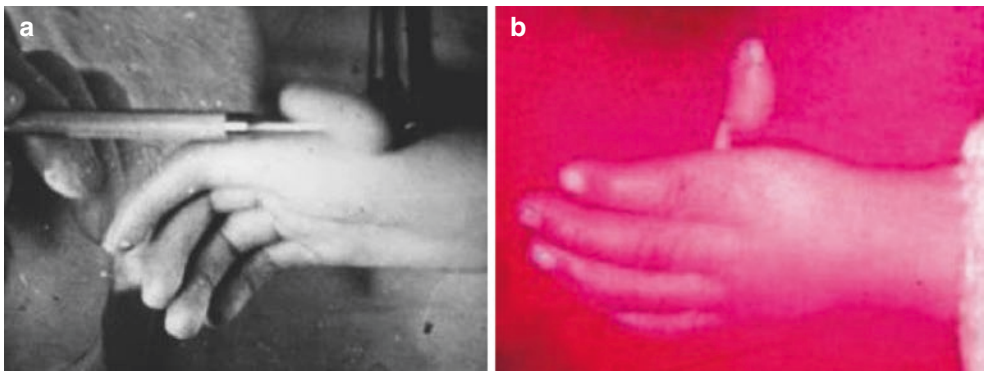


Fig. 5.24 Skin bridge parts of floating thumb. (**a**) The pedicles of skin bridges are located near the proximal row of carpometacarpal joint. (**b**) The pedicles of skin bridges are at the base of the proximal phalanx of the index finger

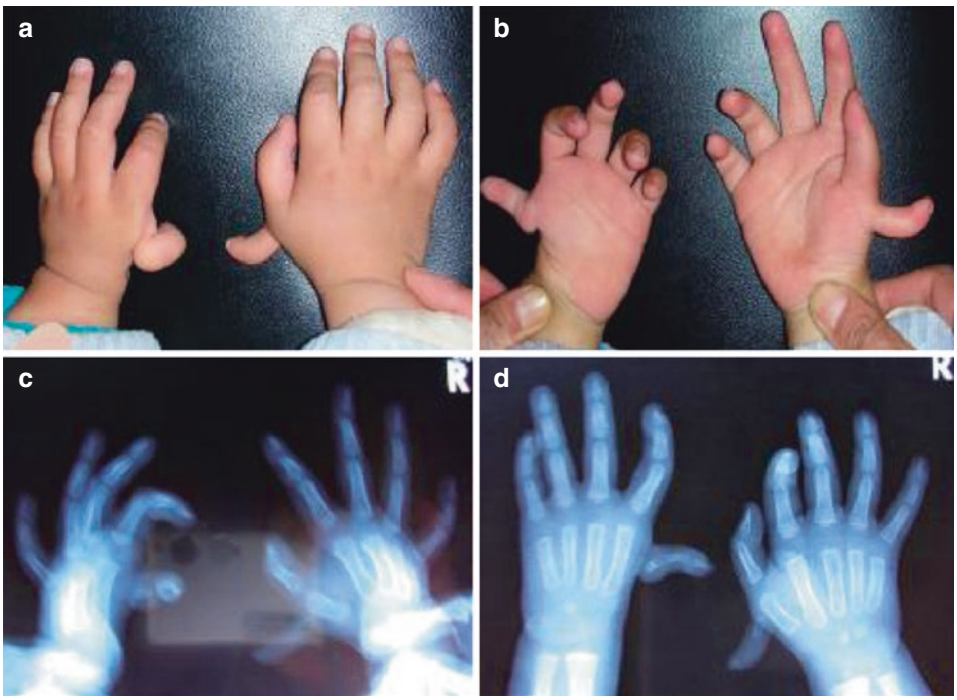


Fig. 5.25 Type IV thumb dysplasia (floating thumb). (**a**, **b**) The left hand suffers serious dysplasia of thumb, close to the floating thumb, and the right hand type suffers VII deformity of duplication of the thumb (**c**, **d**). The X-ray film displays the right hand suffers type VII duplication of the thumb

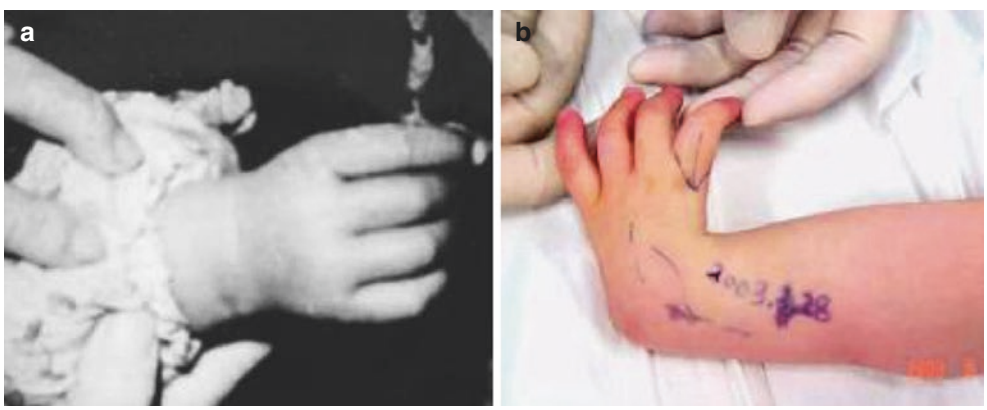


Fig. 5.26 Type V thumb dysplasia. (**a**) Four-fingered hand, thumb absence type dysplasia, good finger development, and complete thumb defect. (**b**) Radial clubhand, with the manifestation of four-fingered thumb absence

In the past, when classifying the congenital thumb dysplasia of the thumb, many authors classified the four-fingered thumb absence as type V thumb dysplasia, namely, serious thumb dysplasia, and the classification ends with this type. In fact, type V thumb dysplasia not only includes congenital thumb dysplasia. Abdel-Ghani H. et al. (2004) [9] also include in the report the four-fingered radial clubhand in type V thumb dysplasia. Although both the four-fingered radial clubhand accompanied by radial absence and the simple four-fingered thumb absence originate from the radial longitudinal ray dysplasia, many authors describe them in different chapters (Fig. 5.26).

5.4.7 Type VI Thumb Dysplasia (Polydactyly with Thumb Aplasia)

Type VI thumb dysplasia, also known as polydactyly with thumb aplasia, was also called five-fingered hand in the previous literature, and the thumb absence of more-than-five-fingered hand is included into another category, but this classification is not so accurate.

Polydactyly with thumb aplasia cannot be simply named five-fingered hand because six-fingered, seven-fingered, and eight-fingered thumb dysplasia also fall into this category. In the report of Abdel-Ghani and Amro (2004) [9], the five-fingered hand is listed as type VI thumb dysplasia, and there is no description of the thumb absence of more-than-five-fingered hand. In the second edition of *Plastic Surgery Science* (2006) edited by Mathes S.J. and Upton J.'s [6] classification of thumb dysplasia, five-fingered hand is classified as type VIII thumb dysplasia, and the thumb absence of more-than-five-fingered hand is classified as type IX thumb dysplasia, called radial hyperdactylia [6], which is not so accurate. The author believes that, whether the hand is five-fingered, six-fingered, seven-fingered, or eight-fingered, such thumb deformity and dysplasia should be called polydactyly with thumb aplasia. The reasons are:

1. Their etiologic factors and treatment principles are the same. The author clinically treated pediatric patients with five-fingered thumb absence, as well as the pediatric patients with six-fingered and seven-fingered thumb absence. These deformities can occur independently and also occur to many persons from the same family.
2. Radial hyperdactylia includes the radial hyperdactylia present in the first metacarpal bone, classified as deformity of duplication of thumb, and the radial hyperdactylia of the first metacarpal dysplasia falls into the category of polydactyly with thumb aplasia.
3. For the same infantile patient, one hand may suffer five-fingered thumb dysplasia, and the other hand suffers six-fingered thumb dysplasia; the manifestation can also be that the pediatric patient suffers six-fingered thumb dysplasia and his/her father suffers five-fingered thumb dysplasia. Therefore, the hyperphalangeal thumb dysplasia of more-than-five-fingered hand should be named type VI thumb dysplasia.

5.4.7.1 Clinical Manifestation

Polydactyly with thumb aplasia includes thumb dysplasia of five-fingered or more-than-five-fingered hand. A normal thumb should have the following five elements: ① the thumb is located in the palm-opposition position; ② the thumb has two segments of phalanx and is shorter than the index finger, and its length is level with the interphalangeal joint level of the index finger; ③ the first fingerweb is broad (thumb web); ④ the first metacarpal bone region has muscle of thenar; and ⑤ the epiphysis of the first metacarpal bone is located at the proximal end of the metacarpal bone.

The patients with five-fingered hyperphalangeal thumb dysplasia have no thumb, but the shapes and functions of other fingers are often normal; the patients with more-than-six-fingered hyperphalangeal thumb dysplasia may suffer various deformities of finger development. Type VI thumb dysplasia has the following manifestations: the hand has more than five fingers, all fingers grow on the same plane from the ulnar side to the radial side, there is no thumb with two dactylopodites in the palm-opposition position, the excessive radial metacarpal bone is finger metacarpal bone, and the epiphysis is located at the distal end of metacarpal bone. As the main feature of such deformity is thumb absence and its treatment mainly aims to reconstruct the function and shape of thumb, the author has classified these deformities as thumb dysplasia since the 1980s, which is the difference from the six-fingered or seven-fingered hand of the deformity of duplication of the thumb. In 1984, the author treated one 5-year-old boy with six-fingered deformity in both hands and thumb absence. The shape, structure, and function of his ulnar four fingers developed well, he has no typical thumb, and plastic therapy is required to be performed on him. The patient's mother and maternal grandmother were five-fingered in both hands with thumb dysplasia; obviously such deformity has a familial genetic predisposition (Fig. 5.27).

In the past, many authors classified hyperphalangeal thumb dysplasia as deformity of hyperdactylia during classification of congenital thumb dysplasia. Actually, the characteristic of polydactyly with thumb aplasia is thumb defect accompanied by deformity of hyperdactylia, and the key to treatment is resection of extra finger and reconstruction of thumb. The deformity of radial hyperdactylia includes various kinds of deformities of duplication of thumb, but this kind of hyperdactylia has fingers and thumb web with the characteristics of thumb. In spite of the presence of obvious thumb dysplasia, it is difficult to classify it as type VI thumb dysplasia. As the deformity of the duplication of the thumb has many unique features not enjoyed by other kinds of deformity, this book describes it in a particular chapter; likewise, mirror-hand hyperphalangeal thumb dysplasia is not listed as thumb dysplasia.

5.4.7.2 Classification

Type VI thumb dysplasia can be reclassified into many subtypes. For example, five-fingered thumb dysplasia is type VIa thumb dysplasia, and six-fingered thumb dysplasia is type VIb thumb dysplasia. It is found during clinical practice that these deformities often have familial genetic predisposition. It should be noted that, as for type VIb thumb dysplasia, one hand can have more than seven fingers accompanied by complete thumb absence.

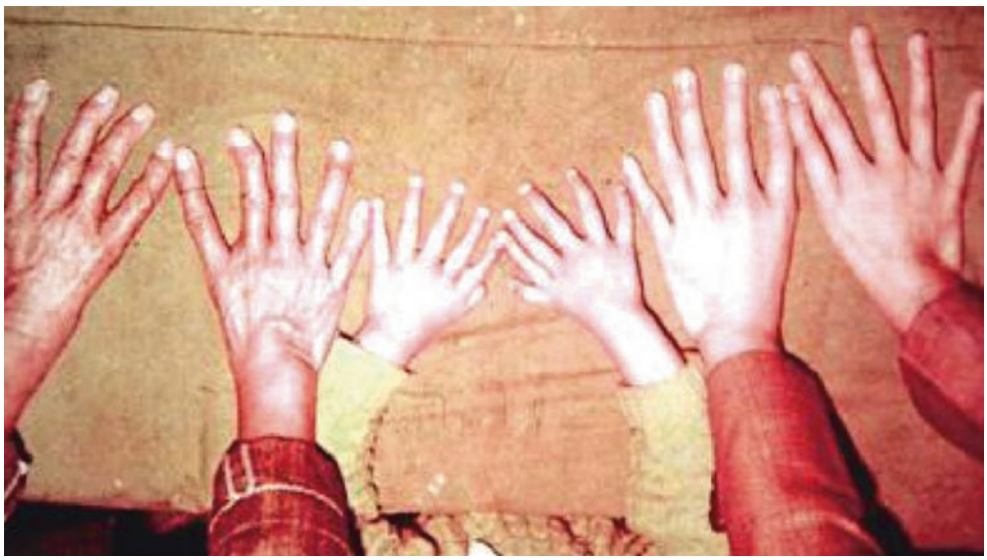


Fig. 5.27 Type VI thumb dysplasia: the one in the middle is a 5-year-old boy with the manifestations of six-fingered hyperphalangeal thumb dysplasia; the ones at two sides are, respectively, the patient's mother and maternal grandmother, who suffer five-fingered hyperphalangeal thumb dysplasia

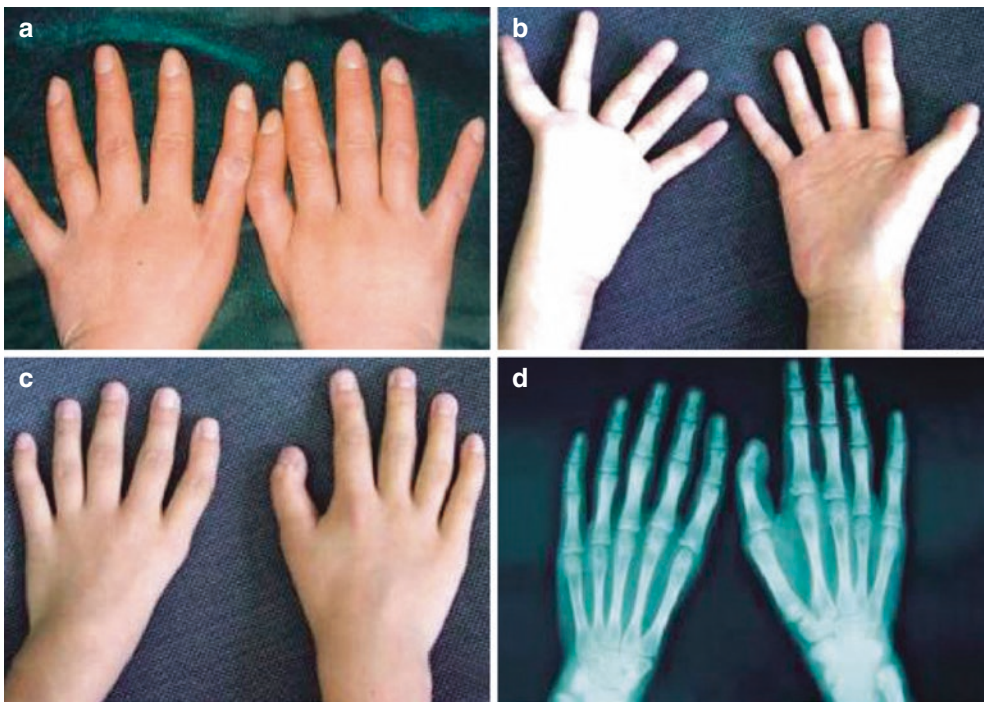


Fig. 5.28 Type VIaa thumb hypoplasia. (**a**) Type VIaa thumb hypoplasia of both hands. (**b**, **c**) Type VIaa thumb hypoplasia of the left hand and moderate hypoplasia of the right thumb. (**d**) X-ray film indicates that the epiphysis of the first metacarpal bone of the left hand is located at the distal end of metacarpal bone, the right thumb suffers moderate dysplasia, the epiphysis of the first metacarpal bone of the left hand is located at the distal end of metacarpal bone, it is the finger-type metacarpal bone, and thenar muscular dysplasia suffers dysplasia

1. Type VIa thumb dysplasia. Five-fingered thumb absence, thumb dysplasia; the category of five-fingered thumb dysplasia can be reclassified into two subtypes:
   (a) Type VIaa (well-developed finger-type thumb dysplasia): The manifestations are the presence of five-fingered hand and thumb dysplasia, but good development or basically good development of ulnar four fingers (Fig. 5.28).
   (b) Type VIab (finger dysplasia-type thumb dysplasia): The manifestations are the presence of five fingers and short and small entire hand or ulnar finger, which are common in syndromic short thumb deformity (Fig. 5.29).

Some include type VIaa thumb dysplasia into the category of polydactylism, but there is no increase in the number of fingers as for this type of deformed hand. The patients have five fingers, and the main manifestation is thumb absence. As for the treatment, simple resection of hyperdactylia cannot be performed. Instead, reconstruction of defected thumb must be conducted. Type VIab dysplasia is characterized by dysplasia of the entire hand, with the manifestations of dysplasia in hands and fingers, brachydactylia, and five-fingered thumb absence. The first step in treatment of these hand deformities is to reconstruct the thumb function, so it is

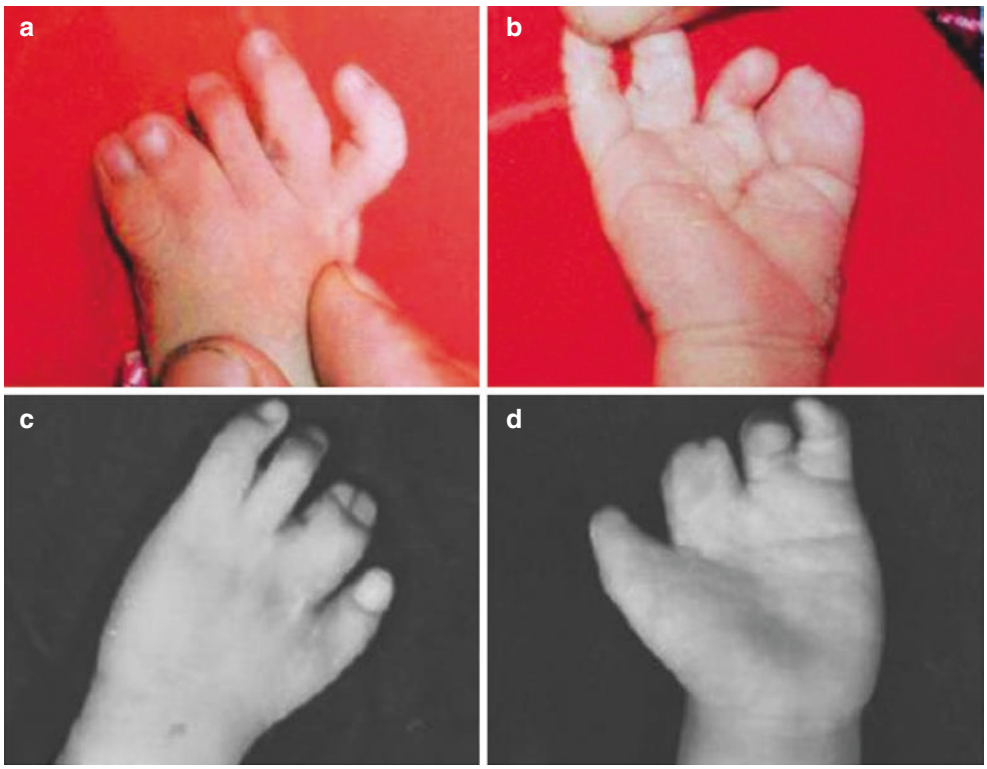


Fig. 5.29 Type VIab thumb dysplasia. (**a**, **b**) The presence of five fingers, thumb absence or dysplasia, and serious dysplasia in other fingers. (**c**, **d**) Five-fingered dysplasia, the presence of thumb, but short and small thumb, dysplasia, and the presence of thenar muscle

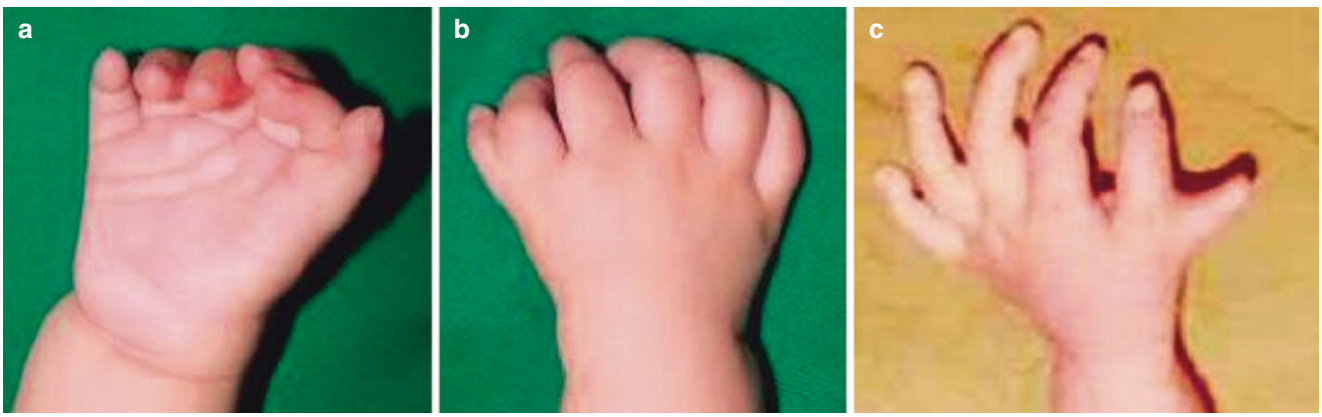


Fig. 5.30 Type VIb thumb dysplasia and deformity of duplication of thumb. (**a**, **b**) Hyperphalangeal thumb dysplasia, no presence of thumb in the palm-opposition position, no thumb web, and serious dysplasia of thenar muscles. (**c**) Deformity of duplication of the thumb, whose second finger at the radial side has the characteristics of thumb

relatively suitable to classify such deformity as congenital thumb dysplasia.

2. Type VIb thumb dysplasia More-than-six-fingered thumb absence, thumb dysplasia

As for the more-than-six-fingered thumb absence or the more-than-six-fingered thumb dysplasia, there is a great difference in shape and structure, but the manifestation is the complete absence of thumb and is different from deformity of duplication of the thumb (Fig. 5.30). This type of thumb dysplasia is similar to type VIa thumb dysplasia. Sometimes, the manifestation is dominant inheritance; if the mother suffers from type VIa thumb dysplasia, her son suffers from type VIb thumb dysplasia.

Type VIb thumb dysplasia (viz., more-than-six-fingered hyperphalangeal thumb dysplasia) is different from the thumb dysplasia with the deformity of duplication of the thumb. The latter one has one relatively short thumb, which is located at the palm-opposition position, with the presence of the first metacarpal bone, broad thumb web, or with the presence of thenar muscles, etc., but the type VIb thumb dysplasia and the deformity of duplication of

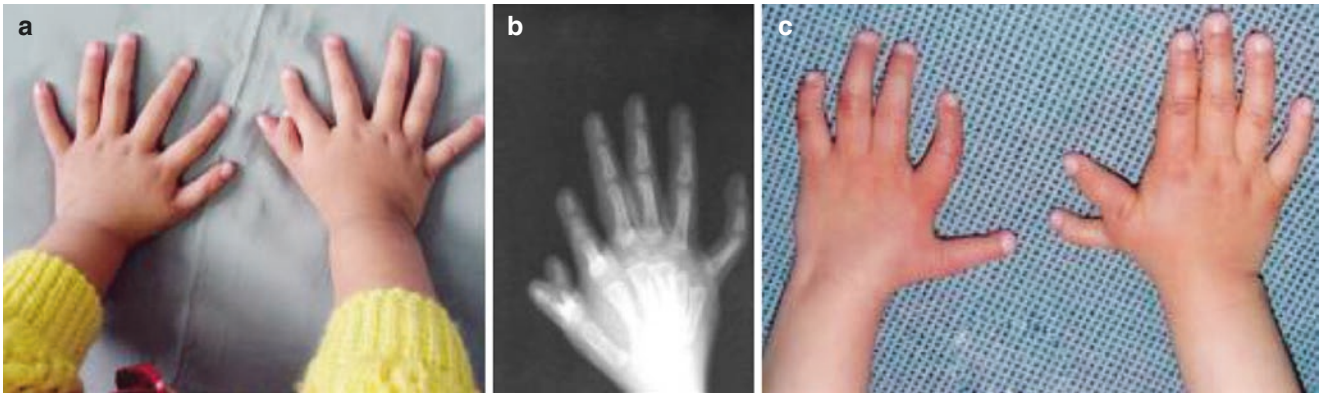


Fig. 5.31 Type VIb thumb dysplasia and the deformity of duplication of the thumb. (**a**, **b**) Patient I, the left hand suffers type VIb thumb dysplasia, the right hand suffers seven-fingered deformity of duplication of the thumb, X-ray film indicates the right thumb suffers dysplasia, and the quasi-first-metacarpal-bone is present. (**c**) Patient II, the left hand has five fingers, which is type IIIc (abduction type) thumb dysplasia; the right hand suffers radial hyperdactylia, which is the deformity of duplication of the thumb in the hyperphalangeal thumb dysplasia

the thumb can also be present in the left and right hands of the same pediatric patient (Fig. 5.31). Therefore, although the author does not describe the deformity of duplication of the thumb into this chapter, the deformity of duplication of the thumb falls into the category of thumb dysplasia in nature.

5.4.8 Type VII Thumb Dysplasia (Hypoplasia of Thumb with Polydactyly and Syndactyly)

Type VII thumb dysplasia is also called hypoplasia of thumb with polydactyly and syndactyly. This type of thumb dysplasia is classified as syndactylia in the literature and is one type of deformity suffering the most serious functional lesions in thumb. The first element in treatment is reconstruction of thumb function, structure, and shape. This type is similar to type VI thumb dysplasia in terms of deformity structure, functional disorder, and treatment methods.

This type of deformity is manifested as each hand with five to eight fingers, syndactylia and/or hyperdactylia, deformity of hand flexion, thumb absence, and the shape of the entire hand like a spider; therefore, it is called web-shaped hand deformity.

The author once treated a 15-year-old girl in 1966 with more than eight fingers and toes in hands and feet.

According to the degree of deformity of finger flexion, type VII thumb dysplasia can be reclassified into two types: type VIIa (general flexion type), with the manifestation of thumb absence, deformity of syndactylia and hyperdactylia, the number of fingers ranging from 5 to 8, accompanied by phalangeal and metacarpal dysplasia, and deformity of mild flexion in interphalangeal joint and type VIIb (serious flexion type), with the manifestation of thumb absence, deformity of hyperdactylia and syndactylia, and serious flexion of interphalangeal joint (90°) (Figs. 5.32 and 5.33).

5.4.9 Type VIII Thumb Dysplasia (Entire-Hand-Dysplasia-Type Thumb Dysplasia)

Type VIII thumb dysplasia, also called entire-hand-dysplasia-type thumb dysplasia, has the manifestations of dysplasia of the entire hand and thumb dysplasia, including multiple syndromic thumb dysplasia, such as syndromic short thumb deformity, the Apert syndrome, and the Poland syndrome (Figs. 5.34, 5.35, and 5.36).

5.4.10 Type IX Thumb Dysplasia (Thumb Dysplasia of Cleft Hand)

5.4.10.1 Clinical Manifestation

Type IX thumb dysplasia, also called thumb dysplasia of cleft hand, falls into the category of thumb dysplasia of central longitudinal ray deficiency, which is manifested as complete absence or deformity of the thumb and cleft-shaped deformity of hand.

5.4.10.2 Classification

This type of thumb dysplasia can be reclassified into the following several subtypes:

1. Type IXa thumb dysplasia. It is also called four-fingered cleft hand-type thumb dysplasia with the manifestation of four-fingered hand accompanied by thumb absence, thumb dysplasia, and syndactylia (Fig. 5.37).

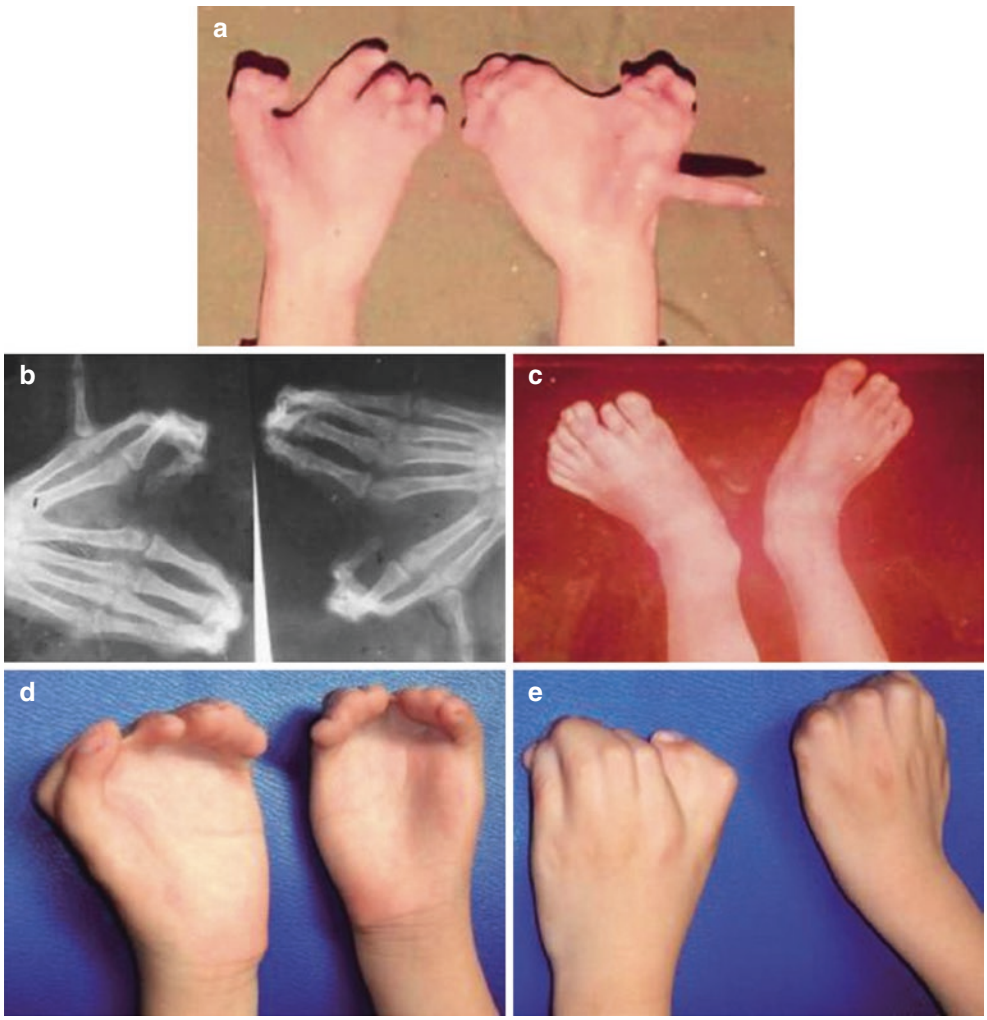


Fig. 5.32 Type VIIa thumb dysplasia (general flexion type). (**a**–**c**) Case one. (**d**, **e**) Case two

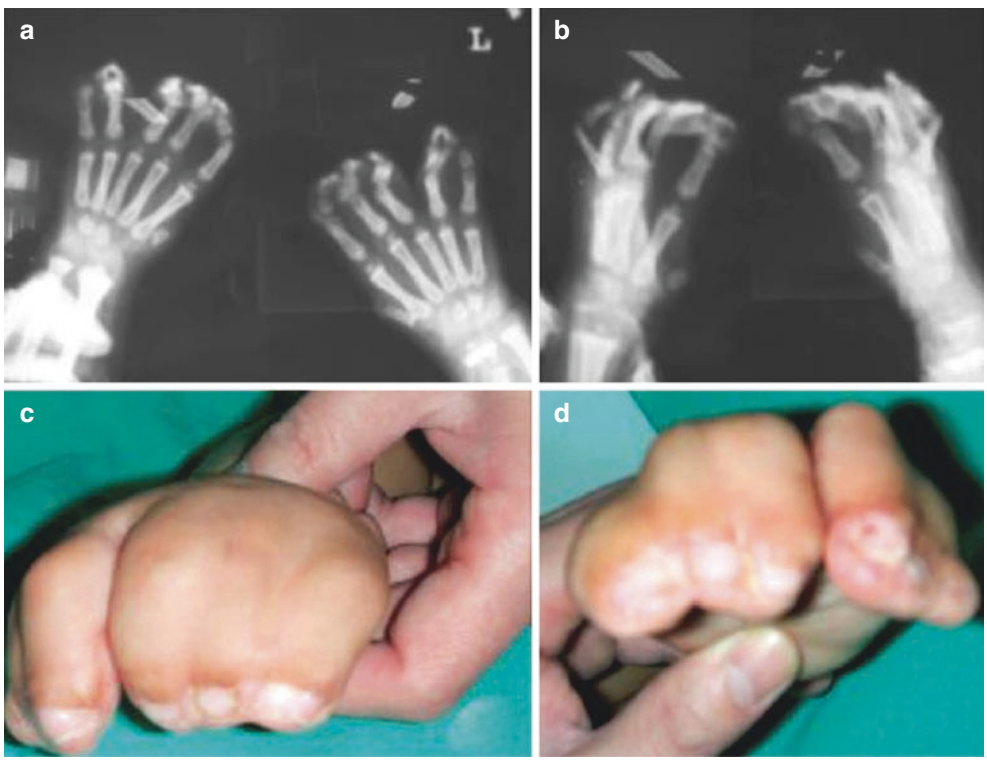
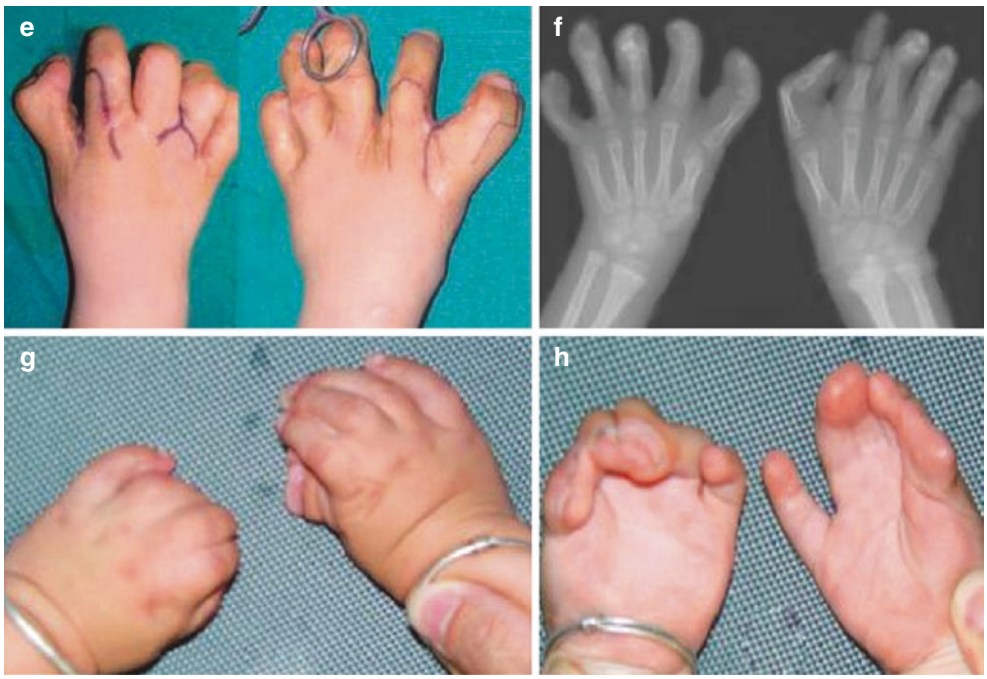


Fig. 5.33 Type VIIb thumb dysplasia (serious flexion type). (**a**–**d**) Case one. (**e**, **f**) Case two. (**g**, **h**) Case three

Fig. 5.33 (continued)



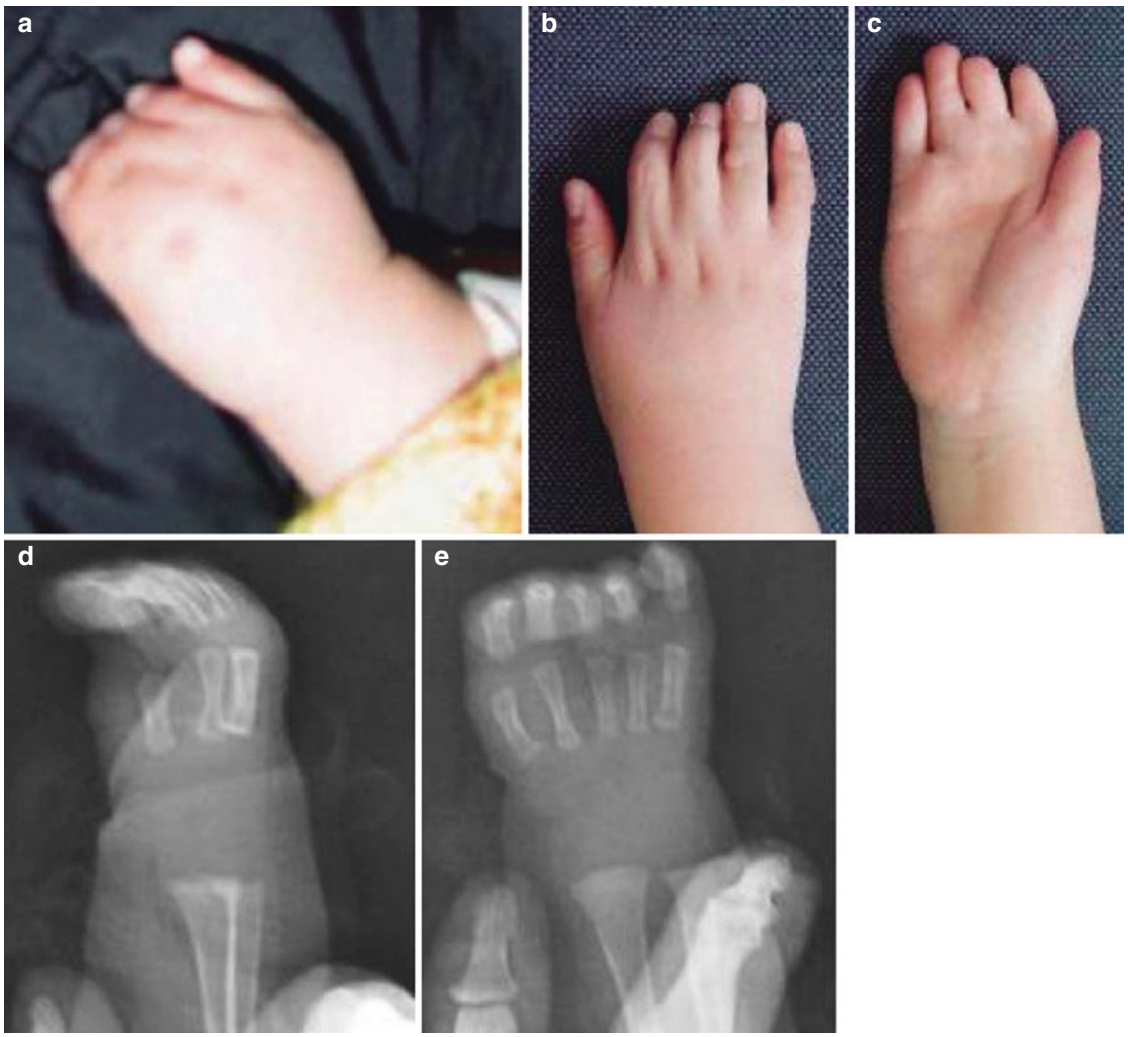


Fig. 5.34 Type VIII thumb dysplasia, the Apert syndrome. (**a**–**c**) Hand appearance. (**d**, **e**) X-ray findings

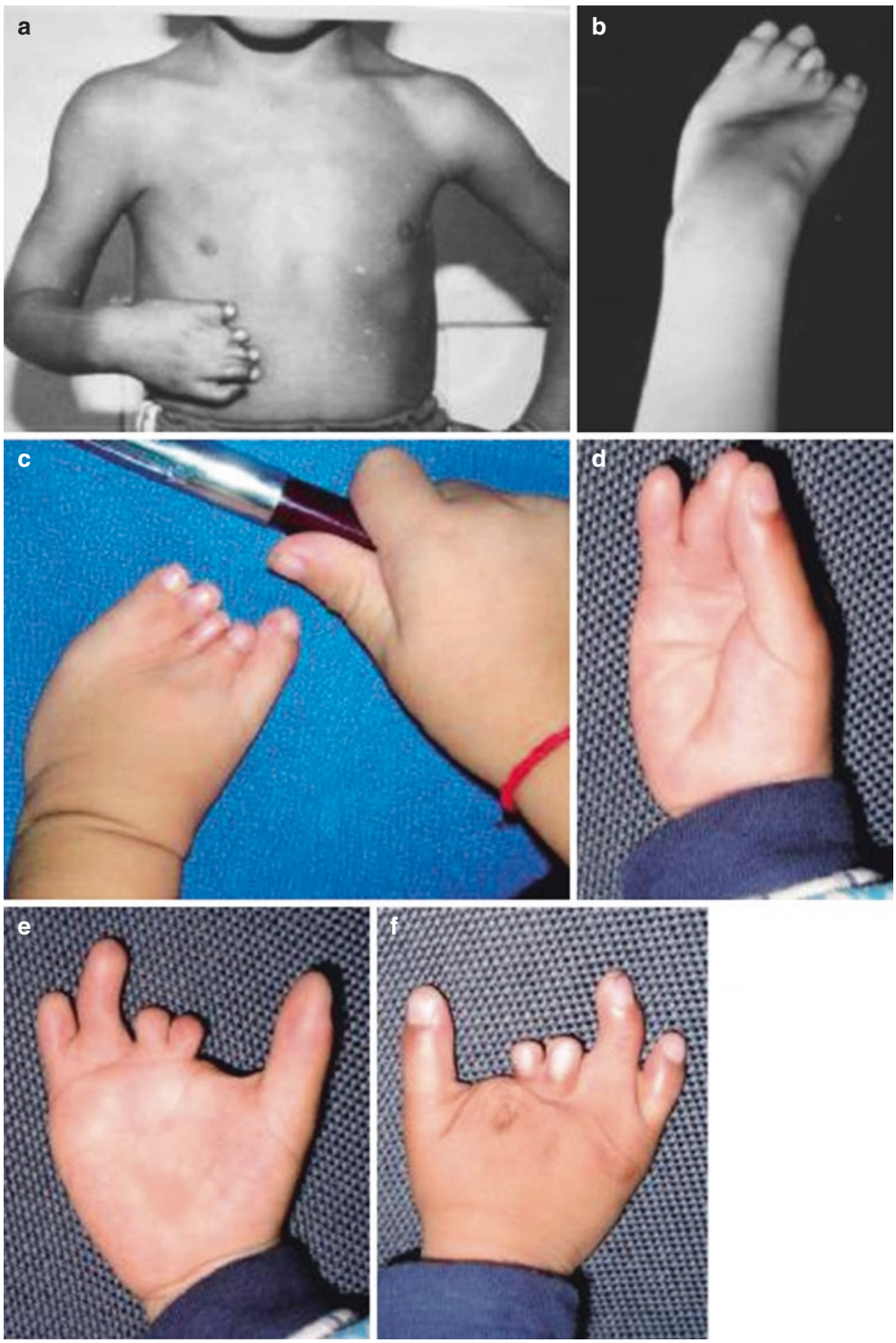


Fig. 5.35 Type VIII thumb dysplasia, the Poland syndrome. (**a**, **b**) Case one. (**c**, **d**) Case two. (**e**, **f**) Case three

2. Type IXb thumb dysplasia, also called thumb dysplasia of three-fingered cleft hand (Fig. 5.38).
3. Type IXc thumb dysplasia. It is also called two-fingered cleft hand-type thumb dysplasia with the manifestation of two-fingered hand accompanied by thumb absence and thumb dysplasia and syndactylia (Fig. 5.39), and the surgical treatment should focus on thumb reconstruction.
4. Type IXd thumb dysplasia. It is also called one-fingered cleft hand-type thumb dysplasia with the manifestation of the complete absence of thumb, finger dysplasia, and one finger in the entire hand (Fig. 5.40), and the surgical treatment should focus on thumb reconstruction. This type of thumb dysplasia is mostly bilateral, often accompanied by bilateral one-toed cleft

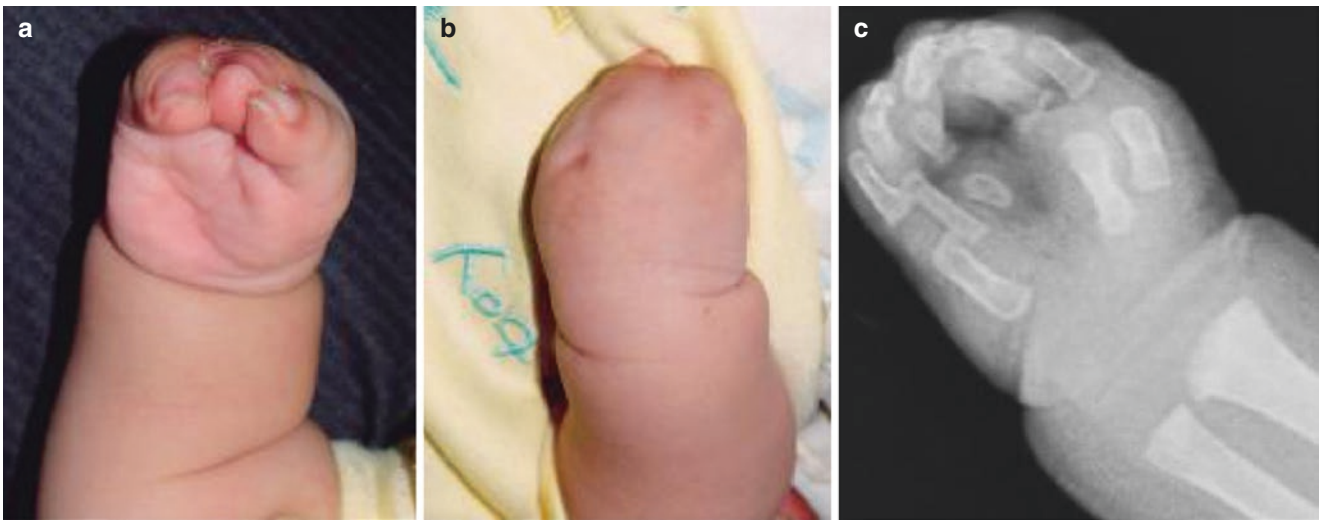


Fig. 5.36 Spade hand deformity. (**a**, **b**) Hand appearance. (**c**) X-ray findings

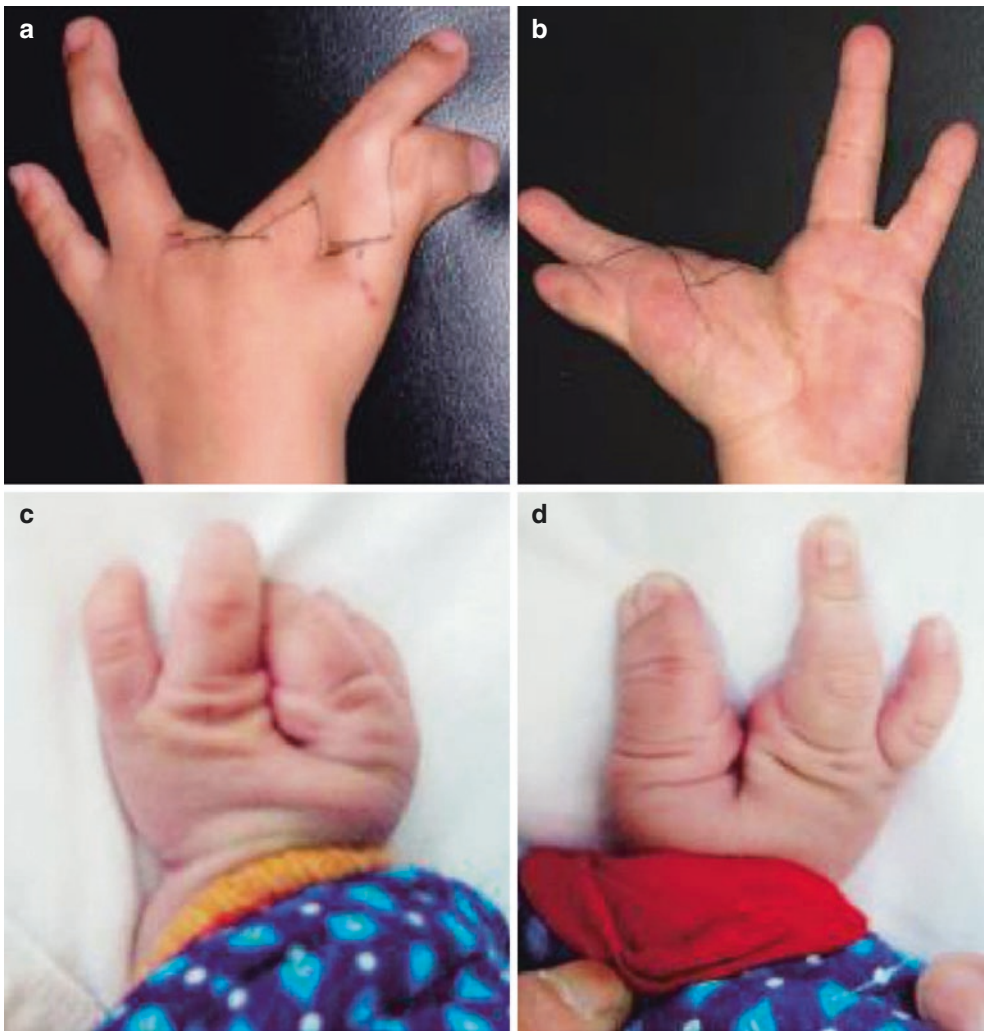


Fig. 5.37 Type IXa thumb dysplasia (four-fingered cleft hand). (**a**, **b**) Syndactylia of the first and second fingers, four-fingered cleft hand. (**c**, **d**) Syndactylia of the first and second fingers, thumb dysplasia

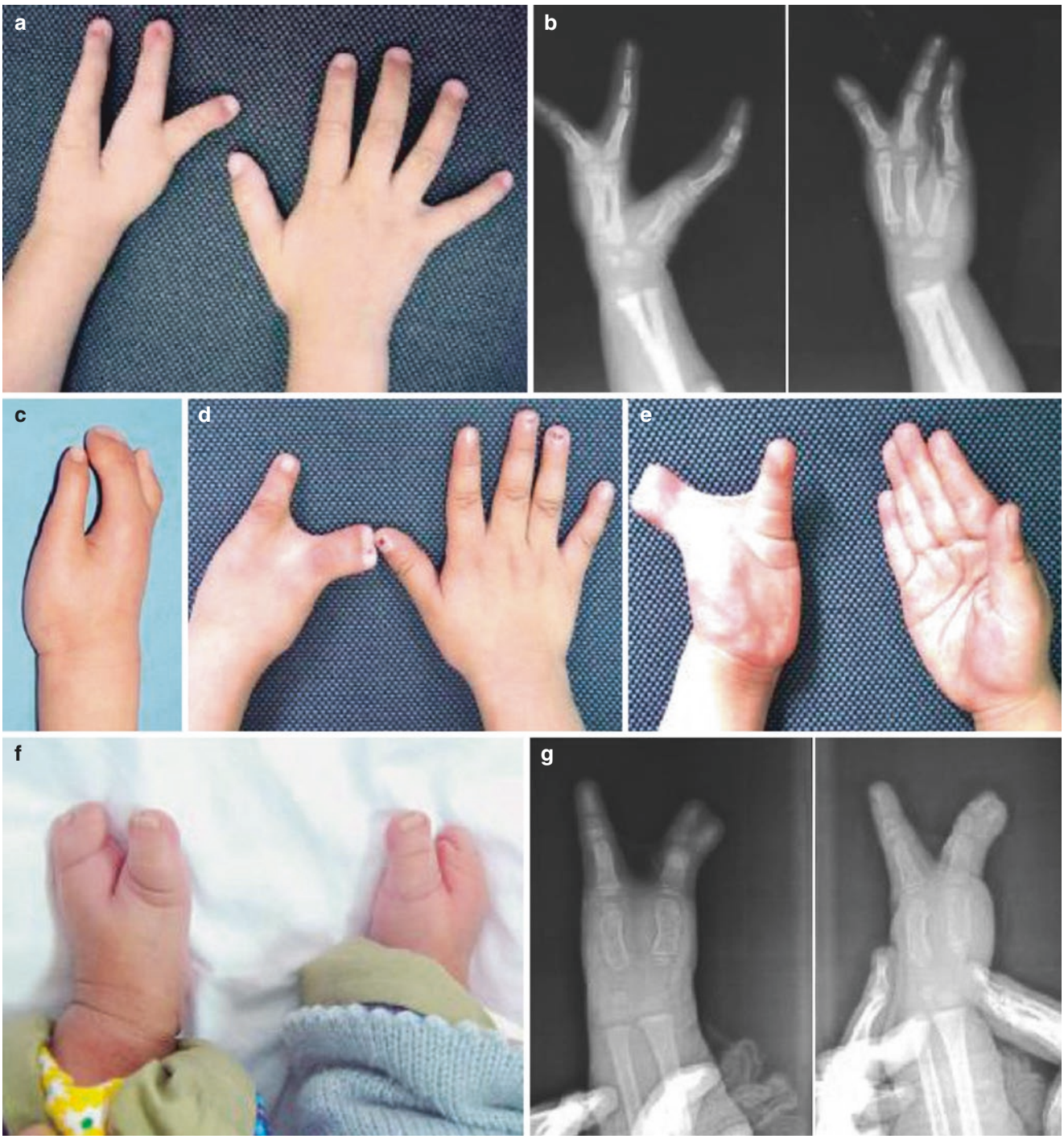


Fig. 5.38 Different types of type IXb thumb dysplasia (thumb dysplasia of three-fingered cleft hand). (**a**, **b**) Hand appearance and X-ray findings of three-fingered cleft hand of the left hand. (**c**–**e**) Complete syndactylia of the second and third fingers of the left hand. (**f**, **g**) Hand appearance and X-ray findings of wide and flat fingers

foot. The author once encountered two pediatric patients with one-fingered cleft hand and dysplasia of foot vessels; therefore, before the toe-to-hand grafting for thumb reconstruction, examinations should be performed on the vascular development to judge whether it is possible to perform successful toe-to-hand grafting for thumb reconstruction.

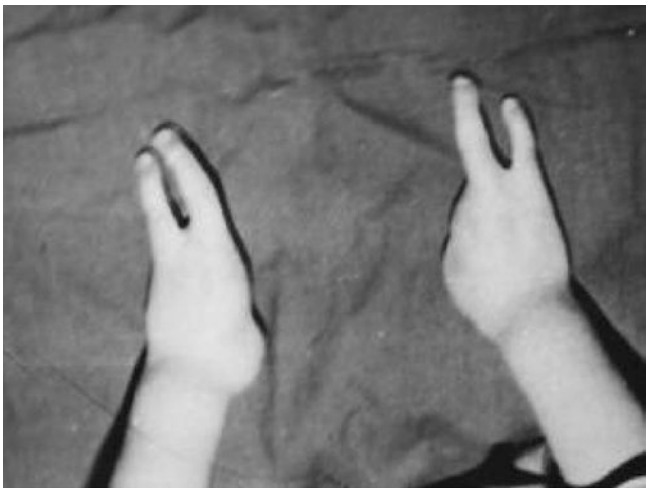


Fig. 5.39 Type IXc thumb dysplasia (thumb dysplasia of two-fingered cleft hand)

5.4.11 Type X Thumb Dysplasia (Constriction Ring Syndrome with Hypoplasia of Thumb Dysplasia)

5.4.11.1 Clinical Manifestation

Type X thumb dysplasia, also called constriction ring syndrome with hypoplasia of thumb, has the manifestation of short and small thumb and distal defect, accompanied by deformity of ring constriction (Fig. 5.41).

Congenital ring constriction of the limb accompanied by thumb dysplasia is often diagnosed as limb congenital ring constriction deformity, and thumb dysplasia is considered as one of symptomatic manifestations. X-ray thumb dysplasia can be manifested as thumb ring constriction, or congenital deformity of severed thumb, and possibly accompanied by other deformities. Therefore, in congenital ring constriction deformity, only congenital severed thumb falls into the category of congenital thumb deformity, and most of the other simple thumb ring constriction deformities are the symptomatic manifestations of ring constriction band syndrome.

5.4.11.2 Typical Case

1. Case one. The pediatric patient suffers ring constriction deformity in the left thumb and the little finger, the thumb suffers mild dysplasia, the digital middle and distal phalanx

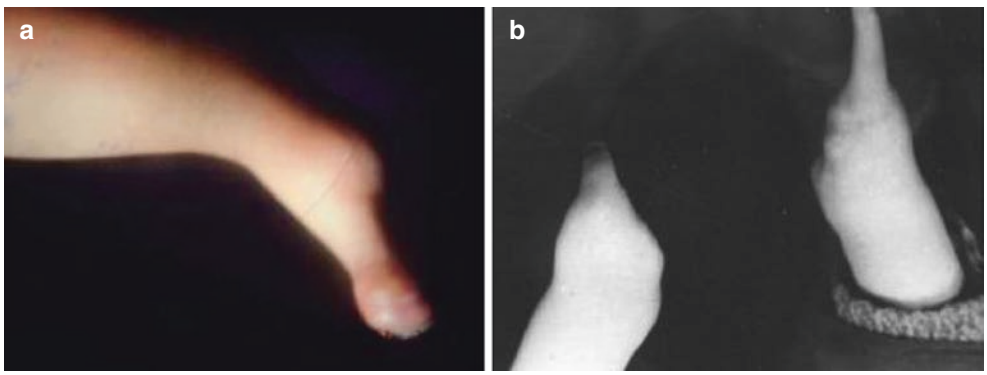


Fig. 5.40 Type IXd thumb dysplasia (thumb dysplasia of one-fingered cleft hand). (**a**) Only one finger in the entire hand, thumb absence. (**b**) The one-fingered cleft hand accompanied by one-toed cleft foot

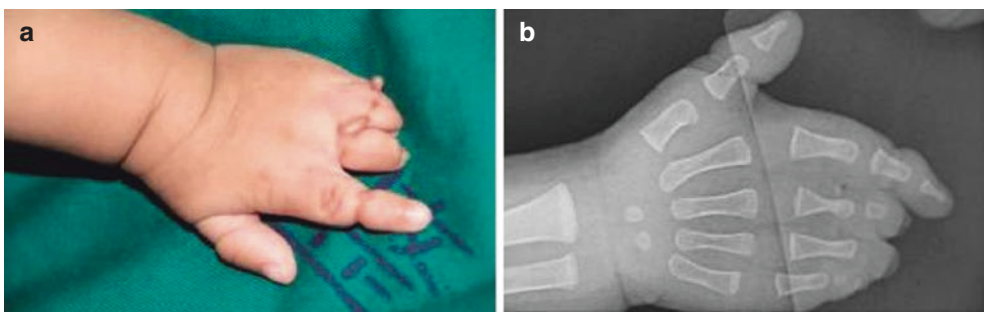


Fig. 5.41 Type X thumb dysplasia. (**a**) Hand appearance. (**b**) X-ray findings

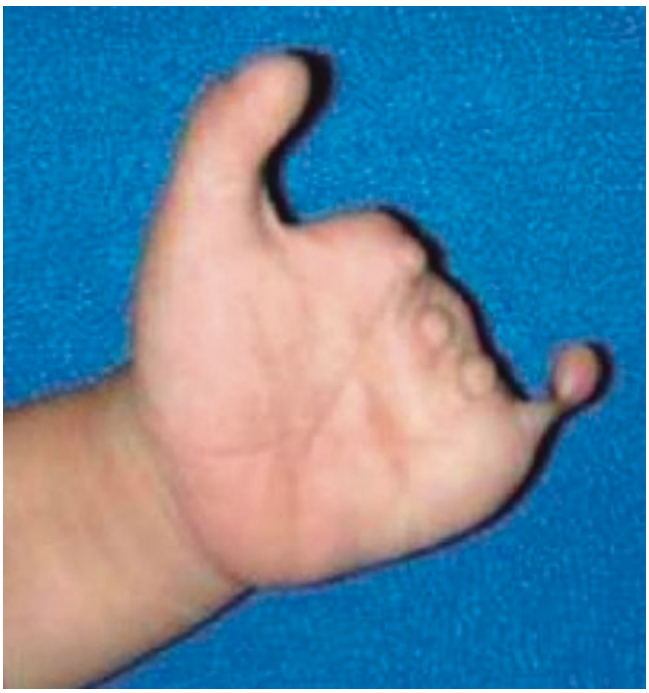


Fig. 5.42 Case one

suffer obvious absence and dysplasia, and the index, middle, and ring fingers suffer complete deficiency. The patient was diagnosed with thumb ring constriction deformity (Fig. 5.42). This type can be also manifested as mild thumb dysplasia, other fingers suffer ring constriction or congenital severed finger; or many parts of the body suffer ring constriction deformity accompanied by mild thumb dysplasia, etc.

2. Case two. In the type X thumb dysplasia-type constriction ring syndrome, dysplasia can also be seen in the entire hand and forearm accompanied by serious thumb dysplasia. The pediatric patient suffers serious dysplasia in the right forearm, right hand, and right thumb, the index finger is absent, most part of the middle finger is absent with ring constriction at the tip, the ring and little fingers suffer syndactylia, and dysplasia is observed (Fig. 5.43).

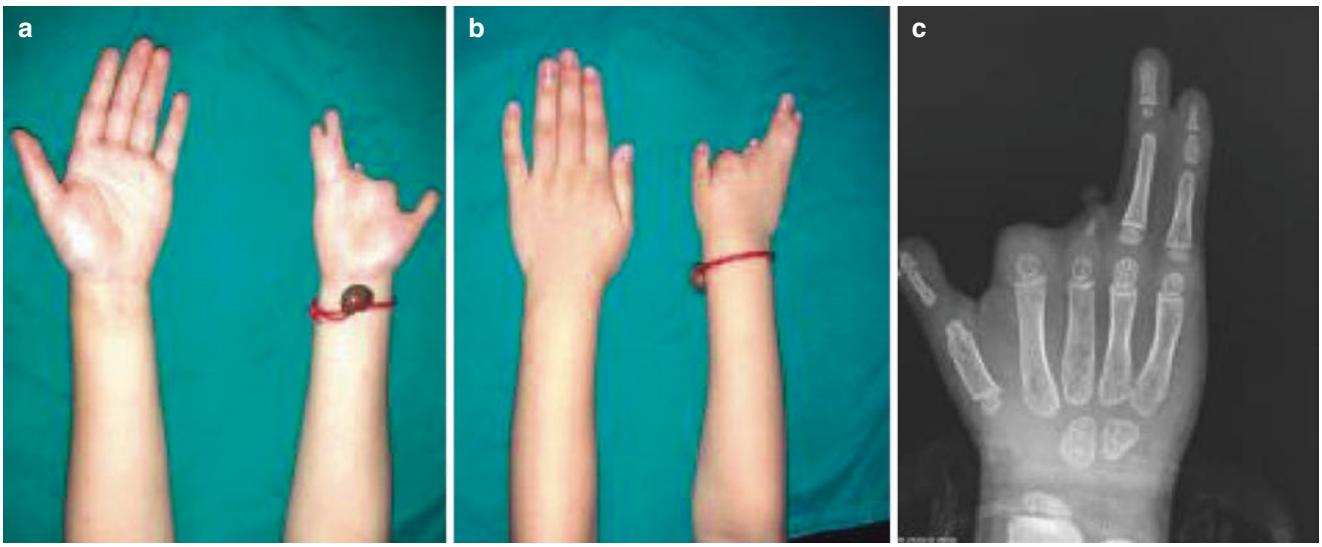


Fig. 5.43 Case two. (**a**, **b**) Hand appearance. (**c**) X-ray findings

5.5 Treatment of Congenital Thumb Dysplasia

5.5.1 Basis of Thumb Function Reconstruction

The objective of congenital thumb dysplasia is to reconstruct the thumb functions and appearance, and the treatment methods include ① reconstruction of the first fingerweb, namely, deepening of the first fingerweb and subsequent thumb web reconstruction; ② thumb lengthening; ③ local reshaping of the first metacarpal bone (pollicization of metacarpal bone) and reconstruction of thumb shape; ④ repair and reconstruction of thumbnail; ⑤ tendon transplantation for the reconstruction of dynamical functions; ⑥ repair and construction of metacarpophalangeal joint and carpometacarpal joint; ⑦ pollicization of index finger or radial finger for thumb reconstruction; and ⑧ toe-to-hand grafting for thumb reconstruction, or other skin flaps, osteoplastic thumb reconstruction, etc.

5.5.1.1 Classification of Thumb Defects and Selection of Surgical Methods

The thumb defect includes congenital and acquired thumb defect; as for the thumb repair and reconstruction of congenital thumb dysplasia, the fundamental principle is similar to related thumb reconstruction methods.

The author classified thumb defect into six categories in 1980 (Fig. 5.44): type I thumb defect, the defect of distal thumb dysplasia; type II thumb defect, the defect of the distal end of interphalangeal joint of thumb; type III thumb defect, the defect of the proximal phalanx area of the thumb; type IV thumb defect, the defect of the distal end of the metacarpophalangeal joint of the thumb, the complete defect of the thumb; type V thumb defect, including the thumb defect with partial defect in the first metacarpal bone; and type VI thumb defect, the thumb defect on the plane of carpometacarpal joint.

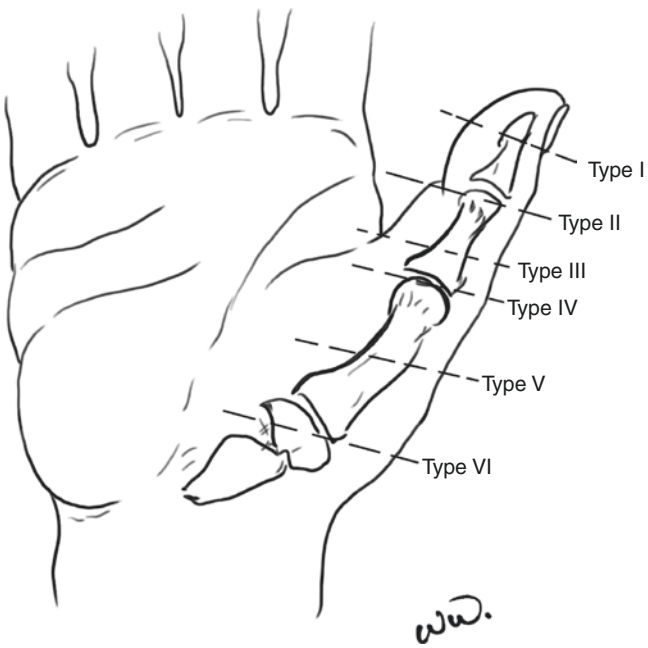


Fig. 5.44 Classification of six types of thumb defect (Wang Wei's classification)

The surgical methods for the reconstruction of various kinds of thumb defect are shown in Table 5.9, and the method can be selected according to the type of thumb defect in congenital thumb dysplasia.

Table 5.9 Selection of surgical methods of thumb defect reconstruction

Defect type	Surgical methods
Type I and II thumb defect	1. Thumb prolonging, including pollicization of metacarpal bone and phalanx prolonging 2. Thumb web deepening or metacarpal bone prolonging + thumb web deepening 3. Nail bed transplantation 4. Grafting of partial phalanges of foot 5. Grafting of toe nail flap or second toe nail flap 6. Toe V-shaped toe web cutaneous-free transplantation
Type III thumb defect	1. Thumb prolonging, including pollicization of metacarpal bone, thumb degloving, bone grafting and prolonging, phalanx prolonging, etc. 2. Thumb back and fingerweb skin flap over and out transplantation, bone grafting, and thumb reconstruction 3. Toe transplantation, bone grafting, and thumb reconstruction 4. Transplantation, bone grafting, and thumb reconstruction of toe nail flap or second toe nail flap 5. Toe V-shaped toe web skin flap transplantation, bone grafting, and thumb reconstruction 6. Skin tube + bone grafting and thumb reconstruction
Type IV thumb defect	1. Grafting of second phalange of foot 2. Grafting of second phalange of foot for expansion 3. Thumb back and fingerweb skin flap over and out transplantation, bone grafting, and thumb reconstruction 4. Transplantation, bone grafting, and thumb reconstruction of toe nail flap or second toe nail flap 5. Toe V-shaped toe web skin flap transplantation, bone grafting, and thumb reconstruction 6. Finger transposition and thumb reconstruction 7. Cutaneous-free grafting + bone grafting 8. Skin tube + bone grafting
V. Type VI thumb defect	1. Grafting of second phalange of foot for expansion 2. Toe transplantation for expansion 3. Finger transposition and thumb reconstruction 4. Transplantation, bone grafting, and thumb reconstruction of toe nail flap or second toe nail flap for expansion 5. Cutaneous-free grafting + bone grafting 6. Skin tube + bone grafting

5.5.1.2 Fundamental Principle of Thumb Reshaping and Aesthetical Reconstruction

As for the thumb reconstruction of congenital thumb dysplasia, as the pediatric patients are young, the hands may concomitantly suffer from dysplasia and deformities in multiple sites and tissues such as vessels, nerves, bones, joints, muscles, tendons, ligaments, fasciae, and aponeurosis; therefore, aesthetical reconstruction of thumb functions and shape should be focused on during selection of surgical methods, and finger pollicization is a preferred surgical regimen. The surgery of finger pollicization is designed to include index finger pollicization, radial finger pollicization, pollicization of radial metacarpal bone, thumb web deepening for thumb prolonging, etc. Toe-to-hand free grafting is also an optional surgical regimen; the follow-up results after the free toe-to-hand grafting for thumb reconstruction turn out satisfying, indicating that the functions are normal, and the grafted toes will grow with increase in age. However, free toe-to-hand grafting for thumb reconstruction should be regarded as the preferred indication for congenital thumb absence and can only be used as an optional surgical method. In case of no alternative local donor site, the free toe-to-hand grafting can be used for thumb reconstruction, or free grafting of disused finger can be used for thumb reconstruction. Aesthetical reconstruction of the thumb includes reconstruction on the length and thickness of thumb, dactylopodite shape, finger pulp, finger nail shape, dynamical functions, shape, etc., which should be carefully designed one by one.

The patients with congenital thumb dysplasia suffer not only generalized development deformities but also deformities and dysplasia of hand skin, subcutaneous tissues, muscles, tendons, bones, joints, ligament, fasciae, nerves, and vessels. The surgeons must carry out careful preoperative examinations; make all-round and accurate evaluation on the morphological, structural, and functional status of the affected hands; and have some understanding of the generalized development status. Only in this way can thumb reconstruction on congenital thumb dysplasia be performed successfully.

5.5.1.3 Treatment Principles of Various Kinds of Congenital Thumb Dysplasia

1. Type I thumb dysplasia (Fig. 5.45):
   (a) Clinical manifestations. Although the thumb is short, small, and narrow and the patients can concomitantly suffer from mild adduction or abduction deformity, the supporting and dynamical structures of thumb are present, and the thumb can basically function.
   (b) Treatment principles. Treatment is generally unnecessary, and thumb aesthetical reshaping and treatment, thumb prolonging, correction of thumb angulation deformity, and correction of finger nail deformity can be selected. Deformity of short thumb is one kind of thumb deformities, and its treatment is not described in this section.

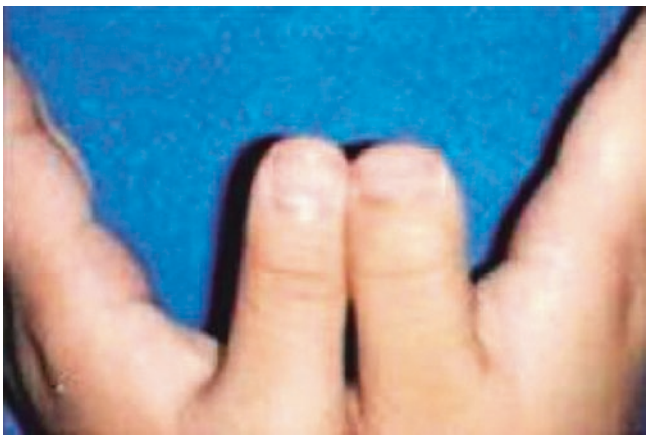


Fig. 5.45 Type I thumb dysplasia

2. Type II thumb dysplasia (Fig. 5.46):
   (a) Clinical manifestations. Obvious thumb dysplasia and obvious defects in shape and function.
   (b) Treatment principle:
   - Correction of thumb adduction deformity and expansion of stenotic thumb web.
   - The treatment of thenar muscular dysplasia includes reconstruction of thumb palm-opposition functions, usually transposition grafting of finger superficial flexor tendon of the ring or little fingers can be adopted, and wrist flexor and brachioradial muscle can also serve as the muscular donor sites for dynamical reconstruction.
   - Correction of anomaly of thumb flexion.
   - The radial wrist long extensor tendon grafting plus volar long tendon grafting and inherent extensor grafting of the index finger are is adopted for thumb function reconstruction.
   - Before correction of thumb interphalangeal joint flexion deformity, the reasons must be identified. If it is caused by thumb long flexor insertion and thumb long extensor tension fusion, thumb long flexor insertion reconstruction and repair or thumb interphalangeal joint fusion can be performed.
   - The repair and reconstruction of the supporting structure of the thumb should cover the striction of the metacarpophalangeal joint accessory ligaments to improve the stability of metacarpophalangeal joints. As for the patients with interphalangeal joint deformities induced by deformities in bone supporting structures, corresponding correction should be performed.

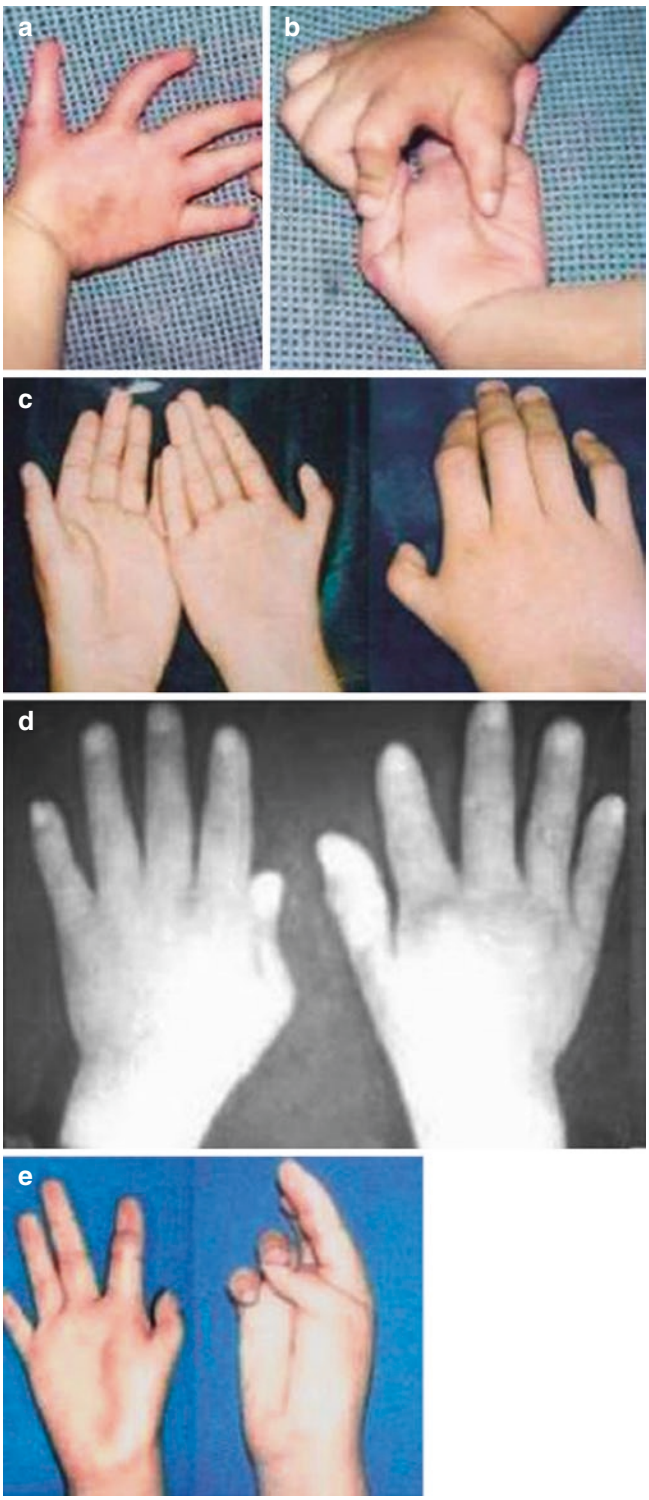


Fig. 5.46 Type II thumb dysplasia

3. Type III thumb dysplasia (Fig. 5.47):
   (a) Clinical manifestations: severe dysplasia of the thumb.
   (b) Treatment principle:
   - As for type IIIa thumb dysplasia, thumb web should be expanded, and functional reconstruction should be performed for corresponding dynamical defects.
   - As for type IIIb thumb dysplasia, it is necessary to consider thumb reconstruction due to the basic loss of thumb functions. In case of simple carpometacarpal joint dysplasia, microsurgical metatarsophalangeal grafting for the reconstruction of carpometacarpal joint reconstruction can be performed.
   - Type IIIC thumb dysplasia is abduction-type thumb dysplasia, and it is necessary to perform correction of abduction deformity and functional reconstruction of corresponding joints; or abducted fingers are removed, and pollicization treatment and thumb reconstruction are performed, etc.
4. Type IV thumb dysplasia (Fig. 5.48):
   (a) Clinical manifestations. The floating thumb.
   (b) Treatment principles: As for the resection of floating thumb, treatment of index finger pollicization should be the first option.
5. Type V thumb dysplasia (Fig. 5.49):
   (a) Clinical manifestations. Thumb absence and thumb dysplasia.
   (b) Treatment principles: The treatment of index finger pollicization should be the best option.
6. Type VI thumb dysplasia (Fig. 5.50):
   (a) Clinical manifestations. Hyperphalangeal thumb dysplasia.
   (b) Treatment principle:
   - Thumb grafting for reconstruction of shortened radial fingers
   - Resection of hyperdactylia, thumb grafting for reconstruction of shortened radial fingers
   - Resection of hyperdactylia, deepened thumb web fingerweb, and thumb grafting for reconstruction of shortened radial fingers
   - Island skin flap grafting of resected fingers, to perform the volume enlargement of reconstructed thumb and re-shaping
7. Type VII thumb dysplasia (Fig. 5.51):
   (a) Clinical manifestations. Syndactylia and hyperdactylia, thumb dysplasia, the number of fingers being more than five, which is six-fingered, seven-fingered, and eight-fingered synpolydactyly
   (b) Treatment principles. Thumb reconstruction and separation of syndactylia, correction of flexed finger deformity, and dynamical reconstruction of finger flexion functions
8. Type VIII thumb dysplasia (Fig. 5.52):
   (a) Clinical manifestations. Dysplasia of the entire hand accompanied by thumb dysplasia and web-shaped hand deformity.

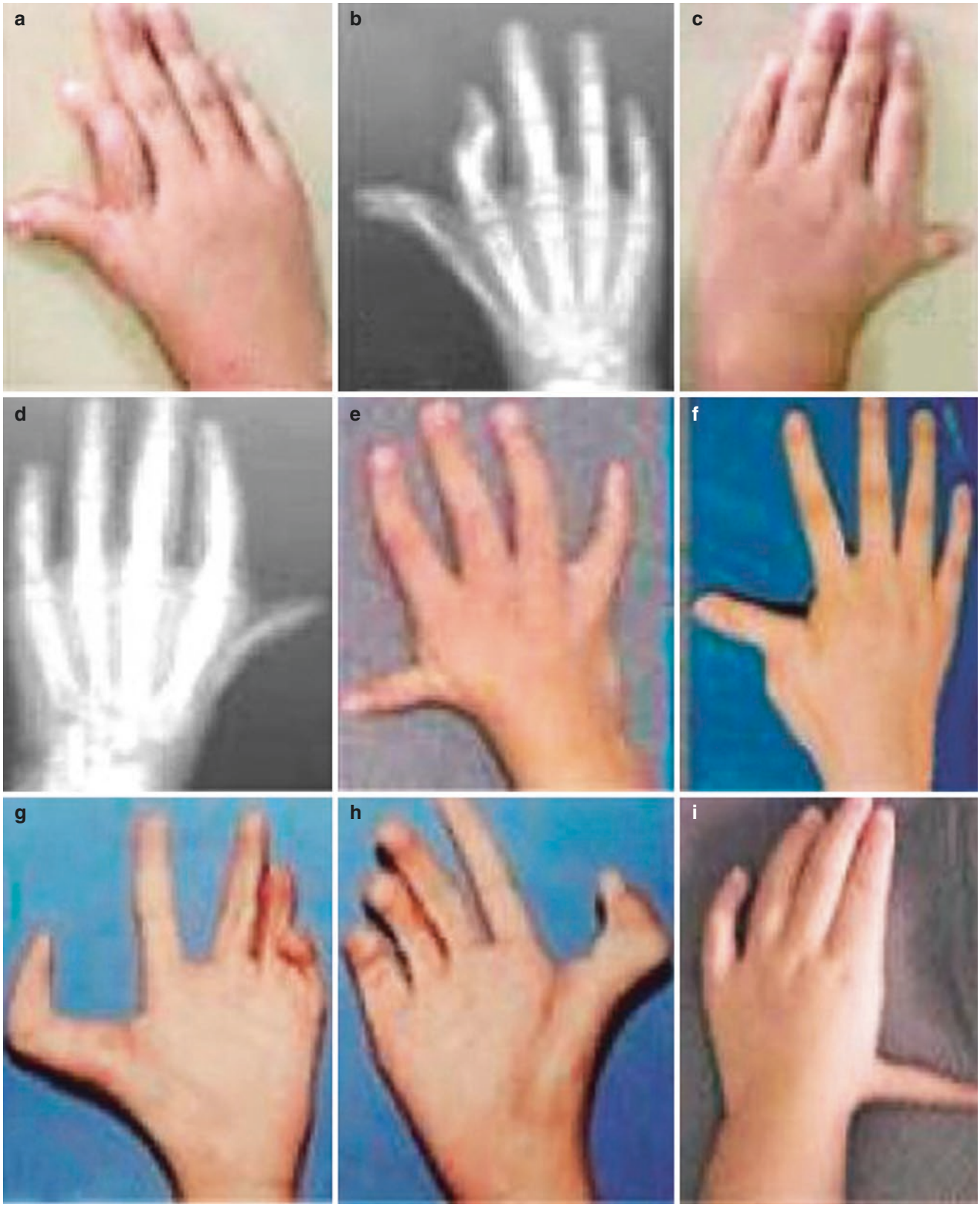


Fig. 5.47 Type III thumb dysplasia. (**a**, **b**) Type IIIa. (**c**–**e**) Type IIIb. (**f**–**i**) Type IIIc

Fig. 5.48 Type IV thumb dysplasia

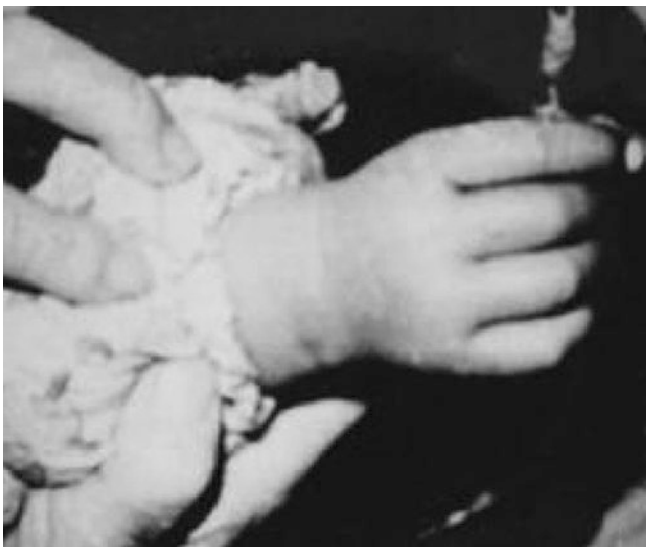


Fig. 5.49 Type V thumb dysplasia

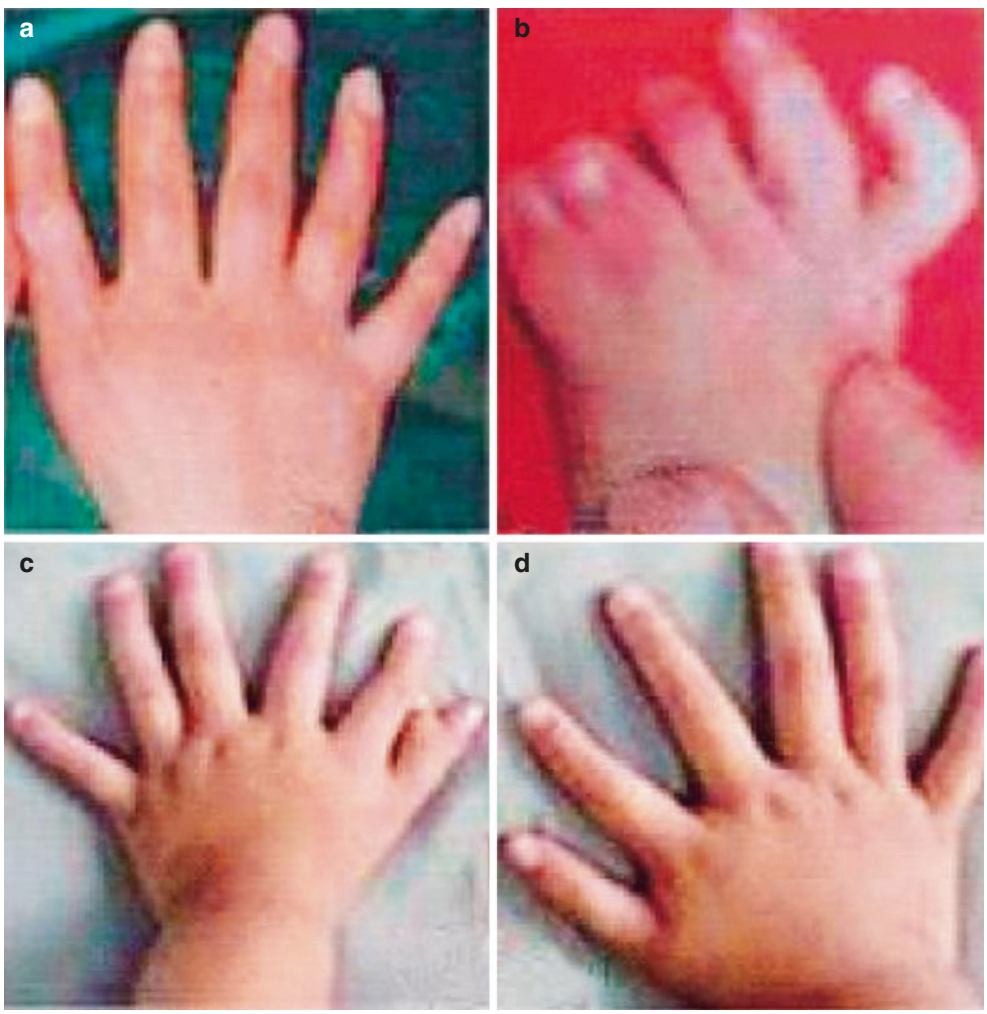


Fig. 5.50 Type VI thumb dysplasia. (**a**) Type VIaa. (**b**) Type VIab. (**c**, **d**) Type VIb

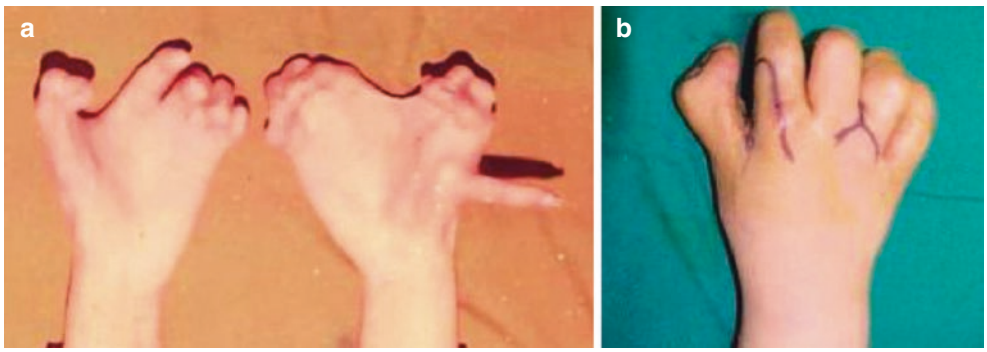


Fig. 5.51 Type VII thumb dysplasia. (**a**) Type VIIa. (**b**) Type VIIb

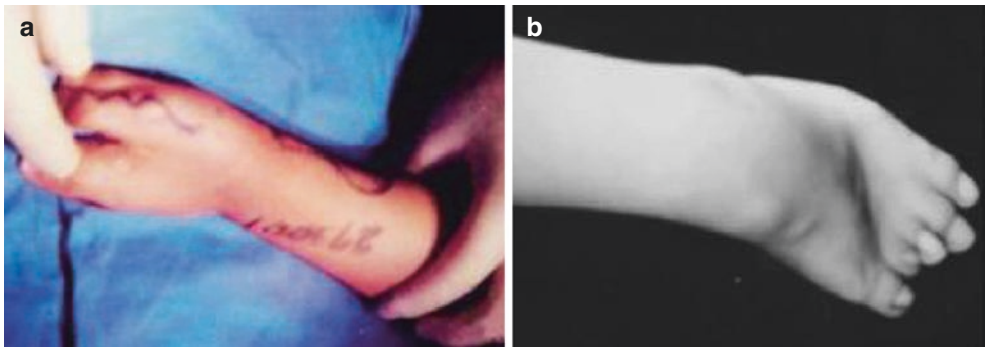


Fig. 5.52 Type VIII thumb dysplasia

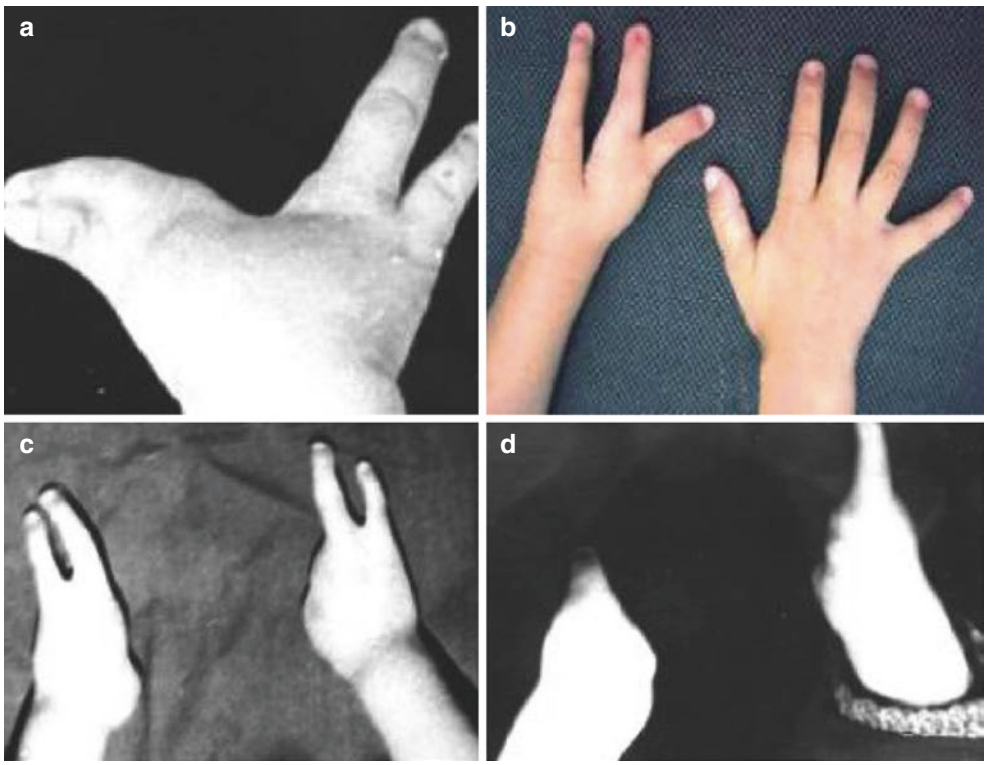


Fig. 5.53 Type IX thumb dysplasia

(b) Treatment principles. The separation of syndactylia and the radial finger grafting for thumb reconstruction are adopted, including radial finger metacarpal bone osteotomy, rotation, and palm-opposition position fixation, deepening and reconstruction of thumb web, etc.

9. Type IX thumb dysplasia (Fig. 5.53):
   (a) Clinical manifestations. Central longitudinal ray deficiency and deformity of cleft hand
   (b) Treatment principles. Correction of deformity of cleft hand and thumb reconstruction

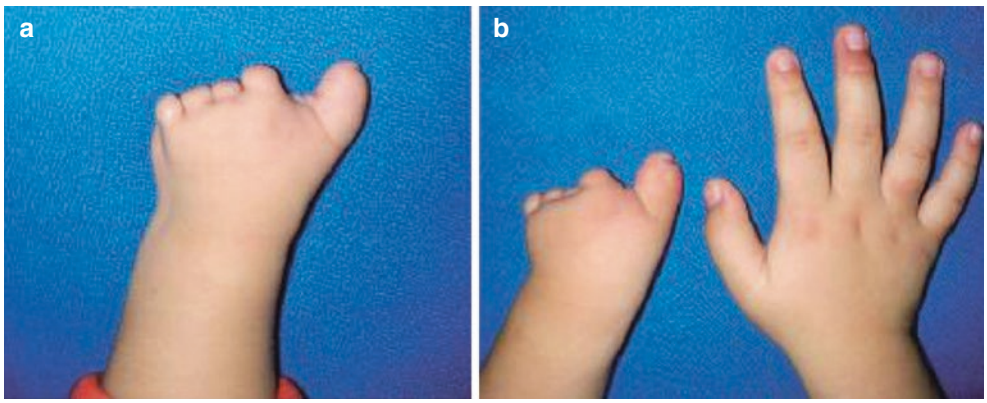


Fig. 5.54 Type X thumb dysplasia

10. Type X thumb dysplasia (Fig. 5.54):
    (a) Clinical manifestations. Constriction ring syndrome plus congenital thumb dysplasia
    (b) Treatment principles. Correction of ring constriction, thumb repair, and reconstruction and finger reconstruction according to the deformity status.

5.5.2 Surgical Timing and Preoperative Preparations

The surgery for thumb dysplasia includes thumb reconstruction, thumb web reconstruction, thumb dynamical reconstruction, beautification and reconstruction of thumb shape, etc.

5.5.2.1 Surgery Timing

Early surgical treatment is a choice for thumb dysplasia, and this treatment involves physiological and psychological factors. The thumb palm-opposition function of a 6-to-7-month-old infant develops gradually, and it is reasonable to perform thumb reconstruction before the full development of grasping and holding functions of thumb within the age of 1. If the general conditions of the pediatric patient are good, early surgery is advisable. It is generally believed that thumb reconstruction surgery can be performed at the age between 1 and 3. Buck-Gramcko once reported an infant at the age of only 11 weeks who underwent this surgery. If the condition for safe anesthesia is met, it is advisable to perform surgical correction during the early stage of infancy, but generally the author choose to perform thumb reconstruction when the infant is 6–7 months ago, because the palm-opposition development of the thumb at this time is relatively full (Fig. 5.55).

During the infancy, finger pollicization for thumb reconstruction is adopted in most cases; if microsurgical toe-to-hand grafting is used for thumb reconstruction, the surgery will be safer after the age of 2. In the selection of surgical timing, top priority should be given to safety. The special requirements for the plastic operation of congenital hand deformity are minimal invasiveness, accuracy, best tissue transplantation, and morphological and functional reconstruction, application of microsurgery technology is demanded, and surgeons should pay due attention.

5.5.2.2 Preoperative Preparations

As the pediatric patient may concomitantly suffer from abnormalities in other organs, especially the cardiovascular system deformities, digestive system deformities, blood system dysplasia, etc., a full physical examination should be performed before operation to confirm the diagnosis and rule out the surgical contraindications.

The abnormal development of limb vessels is also a common complication, and careful examination and diagnosis are needed to be made. In congenital hand deformities, radial arterial absence is common in thumb absence of radial club hand and inherent vascular dysplasia in fingers and also in thumb absence of one-fingered cleft hand and thumb absence of two-fingered cleft hand. The diagnosis of these diseases should be confirmed before the operation; otherwise, any intraoperative negligence may cause accident to the grafted fingers.

The patients with hand radial arterial absence often have relatively thick medial artery (volar interosseous artery). Necrosis of the grafted fingers will be not induced as long as the vessels of the grafted fingers are protected from injuries during operation. Before preparation for finger grafting, the blood supply of the grafted fingers should be carefully tested.

When the toe-to-hand grafting is selected for thumb reconstruction, no matter whether the supply site is at the foot or the hand, the patients may concomitantly suffer from vascular dysplasia; therefore, the vascular status of the supply site and donor site should be learned before surgery, and accidences that may occur during the surgery should be made known to the patient's family members.

The pediatric patients receiving congenital thumb absence surgery are mostly only 1–3 years old, whose tendons, vessels, and nerves are still very delicate. Only by means of operating magnifier or operating microscope can reconstruction of thumb and its dynamical functions be performed

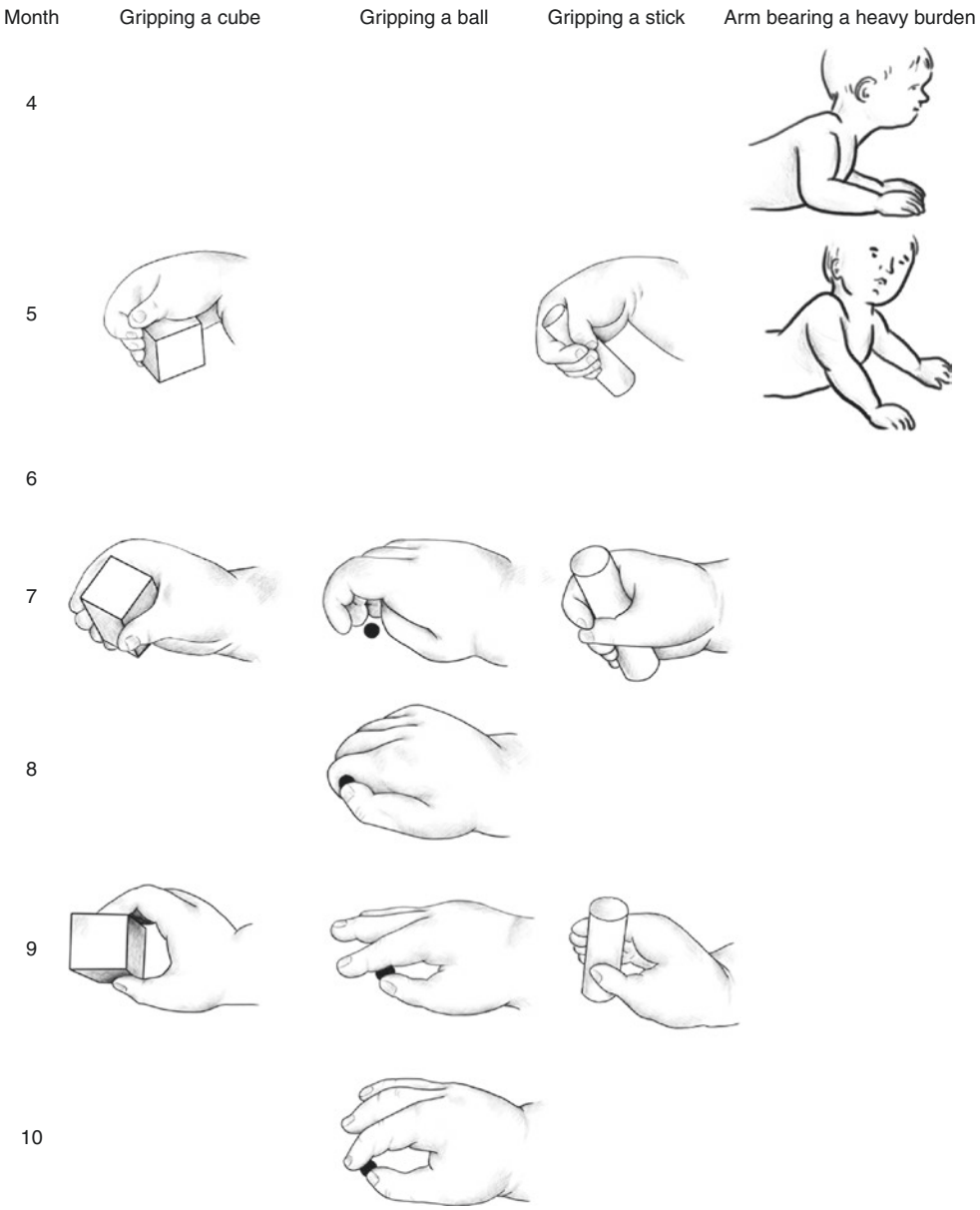


Fig. 5.55 Hand functional development (Redrawn from Upton J: Hypoplastic or absent Thumb in Henth VR ed. The Hand and Upper Limb, Part 2, P 43 Editor Mathes SJ ed. Plastic Surgery Vol. 8. Philadelphia WB Sanders, 2006)

accurately without causing any mistake. The reconstruction of radial clubhand in older children is a reshaping and aesthetical reconstructive surgery and should follow the operation principles of microsurgery—minimally invasive operation and aesthetical shaping no matter what is the design of skin flap or the processing, incision, and suturing of bones and joints, so that the surgery can produce good effects in appearances and functions.

5.5.3 Common Surgical Methods

5.5.3.1 Treatment of Type II Thumb Dysplasia and Deformity of Short Thumb

The manifestations of type II thumb dysplasia and deformity of short thumb include ① short and small thumb; ② thumb web stenosis; ③ hand internal muscle dysplasia; ④ hand external muscle dysplasia; and ⑤ unstable metacarpophalangeal joint, often manifested as accessory ligament relaxation. As for these problems, the following surgeries can be selected:

1. Phalangeal and metacarpal prolonging of the thumb. The deformity of short thumb can sometimes be manifested as short and small thumb, but the hand internal and external muscles develop well with one broad thumb web; the short thumb has flexion, extension, adduction, abduction, and palm-opposition functions; and phalangeal and metacarpal prolonging of the thumb can be performed. Currently, proximal phalanx, metacarpal extension osteotomy, or bone grafting and prolonging are often adopted. The surgery is performed under the condition of general anesthesia or brachial plexus anesthesia. The phalange or metacarpal middle segment ladder is adopted for osteotomy, oblique or traverse

osteotomy can also be performed, the arms of the bone lengtheners are put at the two ends of osteotomy and inserted into external lengtheners, and it is lengthened by 1 mm every 1–2 days until the needed length is obtained. After the bone is lengthened to the target value, the external clamp stand continues to be used for 3 weeks until the osteotomy regions become healed and stable; iliac bone flap grafting can also be performed after osteotomy and steel silk or Kirschner silk can be used for fixation for 6 weeks, and the Kirschner silk is pulled out after bones are healed. However, after the phalange or metacarpal osteotomy, the insufficient space often makes it difficult to perform lilac bone grafting.

2. Reconstruction of thumb dynamical functions. Reconstruction of thumb dynamical functions is often adopted for type IIa, IIb, and IIc thumb dysplasia, and good effects can be achieved. Among all cases of moderate thumb dysplasia, thenar muscular dysplasia is the commonest. Its treatment includes reconstruction of thumb palm-opposition function, and some patients still need to receive thumb web expanding surgery. The main objective of thenar muscular dynamical reconstruction is to reconstruct the palm-opposition function of the thumb, the usual practice is grafting of superficial digital flexor of the ring finger, and other tendons can also be selected for grafting as the motive force. In the reconstruction of palm-opposition function, fabrication of trochlea is an important step, and using the ulnar flexor muscle tendon of wrist as the trochlea is a typical surgical method.
   (a) Surgical method (grafting of superficial digital flexor of the ring finger for opponensplasty): The traditional method is to make a median incision at the ulnar side of the interphalangeal joint of the proximal segment of the ring finger and expose the bifurcation of the superficial digital flexor. Although the incision has a good exposure, the trauma at the linear trauma is big. The author is more willing to choose to make a traverse incision at the basal transverse striation of the proximal segment of the ring finger to cause the ring finger to flex, and by means of the skin retractor, the distal insertion of the superficial digital flexor of the ring finger can be exposed. Tunnels are made at the parts from the ring finger to the wrist and from the wrist to the metacarpophalangeal joint so that the palm-opposition dynamical tendons can pass. Make a traverse incision at the wrist transverse striation, expose the ulnar flexor muscle tendon of wrist, cut half of the tendon at the radial side by about 3 cm, and roll it into a button hole shape and make it into trochlea. Let the superficial flexor tendon of the ring finger pass the button hole-shaped trochlea and end at the distal end of metacarpophalangeal joint of the thumb, and perform suturing for fixation. Before the tensions are sutured, make the thumb maintain the tension in the palm-opposition position.

      The author sometimes uses the superficial flexor tendon of the ring finger as the dynamic tendon and palmar fascia as the trochlea because the surgical trauma is relatively small and the efficacy is good. The dynamical reconstructing surgeries of type IIb and IIc thumb dysplasia are similar to this.
   (b) Typical case: An infantile patient, a 1-year-old boy, suffered type IIa thumb dysplasia of the left hand and thenar muscular dysplasia of the thumb. The superficial digital flexor of the ring finger is taken for grafting to reconstruct the thumb palm-opposition functions. Under the condition of general anesthesia, make an incision at the transverse striation of the proximal segment of the left ring finger and then make another incision at the transverse striation of the distal palm. Take out the superficial flexor tendon of the ring finger from the small incision of transverse striation of the distal palm, form palmar aponeurosis fabricated trochlea via the subcutaneous pathway of superficial layer of palmar aponeurosis, surround it from the radial side to the ulnar side for one cycle after the thumb metacarpophalangeal joint is reached, and end it at the distal level of the horizontal line of the metacarpophalangeal joint of the thumb. Fix the tendon insertion via the incision, reconstruct the palm-opposition functions of the thumb, and make sure that the thumb is in the palm-opposition position after the surgery (Fig. 5.56).
3. Thumb web expansion and correction of corresponding deformities. Including expansion of thumb web, remission of corresponding ligament and contracted muscles, and repair and reconstruction of skin defects. There are multiple options for repair and reconstruction of skin defects, including deepening of the first fingerweb, Z-plasty, free skin grafting, or skin flap grafting. As for skin flap grafting, in addition to the retrograde skin flaps at the interosseous dorsal side, the forearm radial or ulnar retrograde skin flaps, groin flaps, upper arm medial or lateral flaps, and pedal dorsal flaps can be adopted.
4. Correction of deformity of adducted thumb. Correction of the deformity of adducted thumb includes thumb web expansion, correction of deformity of flexed thumb, and dynamical reconstruction of thumb extensor tendons and thumb abductor tendon. In the correction of deformity of adducted thumb, thumb web expansion is the common and preferred basic technology and the specific surgical methods are as follows:
   (a) Dorsal skin flap rotation and grafting of the index finger or thumb for thumb web expansion: As for serious thumb web stenosis of congenital hand deformities, it is difficult for local Z-plasty to serve the purpose of correction. The dorsal skin flaps of the index finger or the thumb can be adopted for rotation, grafting, and repair, and this is also a clinically preferred surgical method. Although distal skin flap

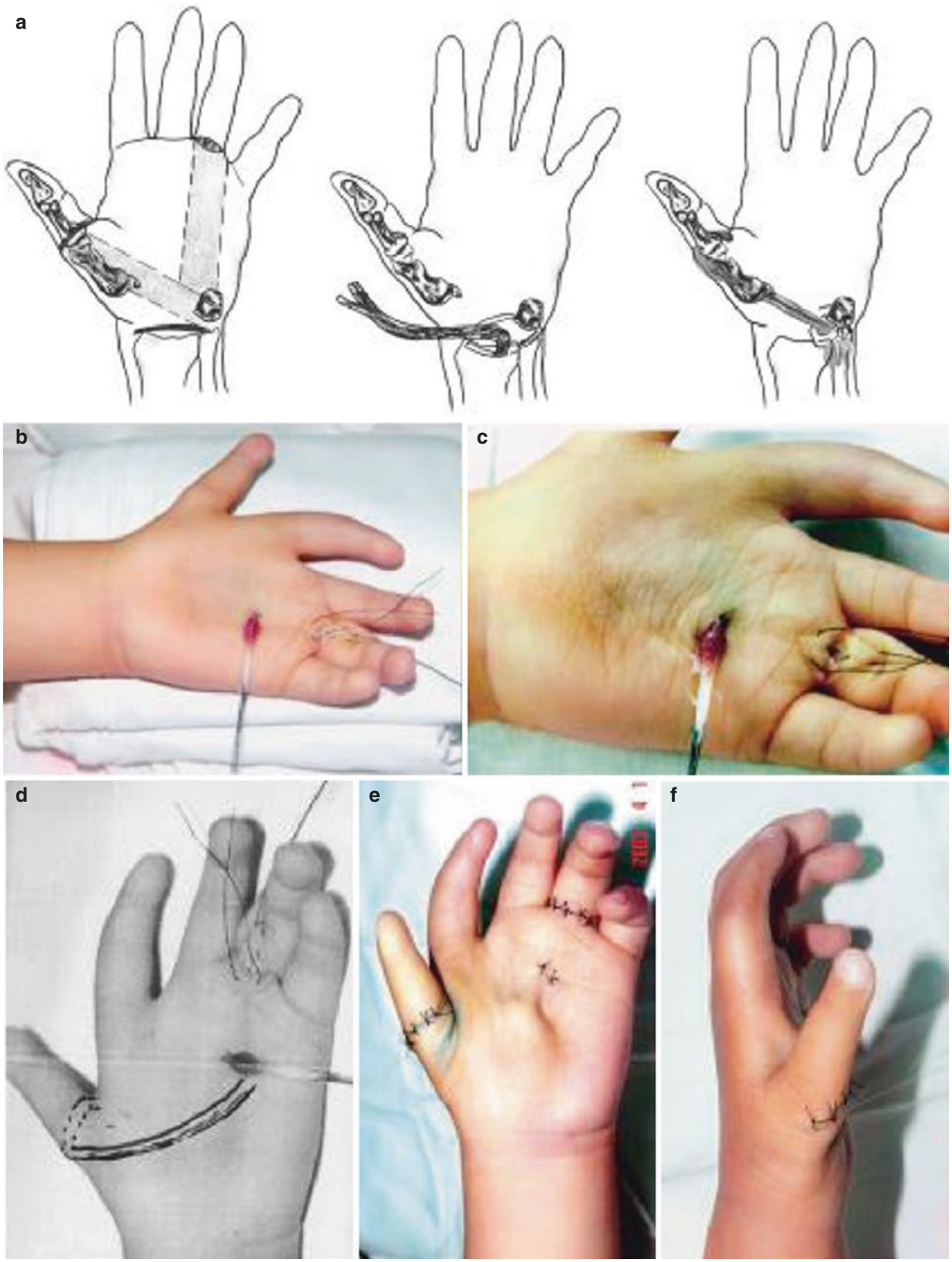


Fig. 5.56 Dynamical reconstruction of opposition functions of superficial flexor tendon of the ring finger. (**a**) Schematic diagram of three surgical incisions and resection, grafting and fixation of ring finger tendons. (**b**–**d**) Removal of superficial flexor tendon of ring finger for dynamical reconstruction of opposition functions. (**e**, **f**) Postoperative hand appearance

pedicles can be adopted for grafting or free grafting, both morphological and functional reconstruction are complicated; therefore, it is less frequently used.

Preparation of dorsal rotation flaps of the index finger or the thumb: The ratio of length and width of the designed skin flaps at the dorsal side of the index finger or the thumb is 2.1, 2.5: 1, or 3: 1. Incise the skin along the designed line of the incision until the surface of the extensor tendon aponeurosis is reached to retain the cutaneous and subcutaneous veins inside the skin flaps. Design incisions at the volar side of hand, and rotate the prepared digital dorsal skin flaps to the thumb web and palm to serve the purpose of thumb web expansion (Fig. 5.57).

(b) Z-plasty coupled-triangular skin flap grafting for thumb web expansion: Usually called Z-plasty thumb web expanding surgery. No. 1 and No. 2 triangular skin flaps are designed, the axial position is located at the junction between the palm and hand dorsum of the thumb and the index finger, and the angles of the skin flaps should range from 45° to 60°. When the skin flaps are incised, the subcutaneous deep fascia should be reached, and the finger nerves and vessels should be protected from injuries, and dual cross-transplantation is performed to serve the purpose of thumb web expansion (Fig. 5.58).

(c) Four-flap method thumb web expansion: Four-flap thumb web expansion is the extension of Z-plasty coupled-triangular skin flap grafting for thumb web expansion. There are two design plans for the four-flap method repair of thumb web stenosis, namely, dual Z-plasty skin flap coupled-grafting and four-cutaneous-flap cross-grafting transplantation. Incise the skin and subcutaneous tissues according to the design, release aponeurosis, perform No. 1 and No. 2 triangular skin flap coupled cross grafting, and perform No. 3 and No. 4 triangular skin flap coupled cross grafting; or perform No. 1 and No. 3 triangular skin flap coupled cross grafting, and perform No. 2 and No. 4 triangular skin flap coupled cross grafting. The skin flap angle should range from 30° to 45° or a little larger; but when the flap-included angle is too large, it is apt to produce varying tension in rotation, grafting, and suturing (Fig. 5.59).

(d) Five-flap method thumb web expansion method, namely, V-Y skin flap reshaping plus dual Z-plasty coupled-triangular skin flap grafting, in which the skin flap angles should be within the range of 45° to 60° (Fig. 5.60).

5. Correction of the deformity of thumb flexion. In type II thumb dysplasia, the deformity of thumb adduction and the deformity of flexion often coexist. The deformity of thumb flexion is associated with brevis flexor dysplasia and contracture of the thumb and can also be correlated with dysplasia of thumb extensor. Due to dysplasia and contracture of the thumb adductor or the thumb brevis

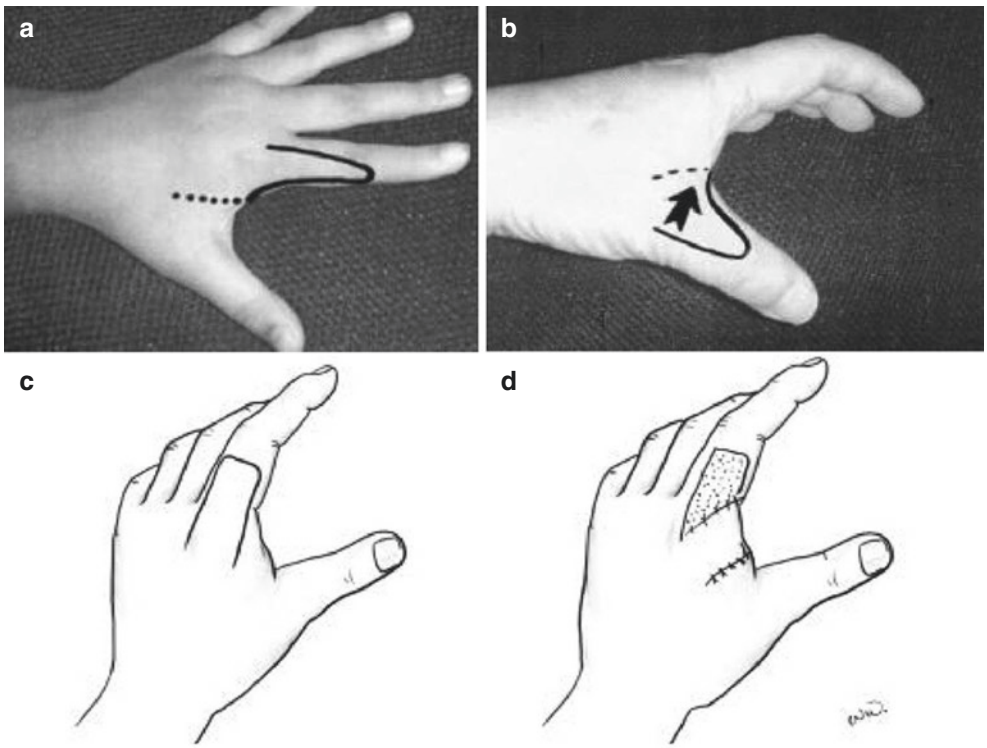


Fig. 5.57 Rotation of digital dorsal skin flaps of the index finger or the thumb for thumb web expansion. (**a**, **b**) Design incisions at the back of hand (indicated by the *dash line*). (**c**, **d**) Repair of thumb web stenosis

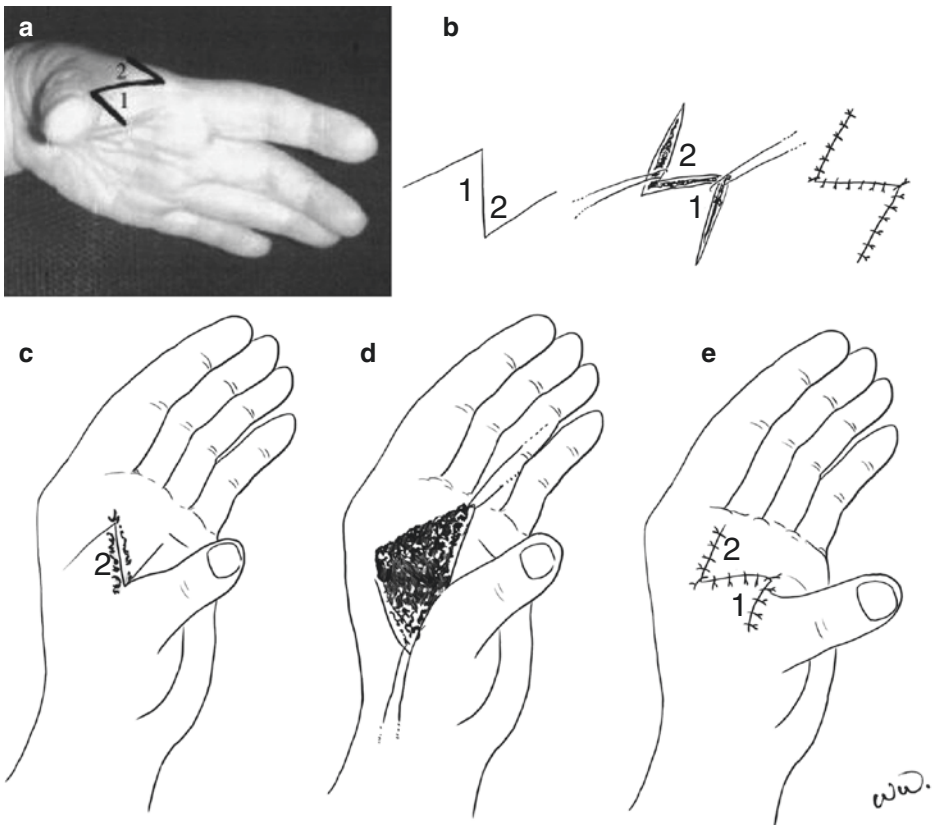


Fig. 5.58 Z-plasty coupled-triangular skin flap grafting for thumb web expansion. (**a**, **b**) Design of Z-plasty skin flaps. (**c**–**e**) Repair of thumb web stenosis

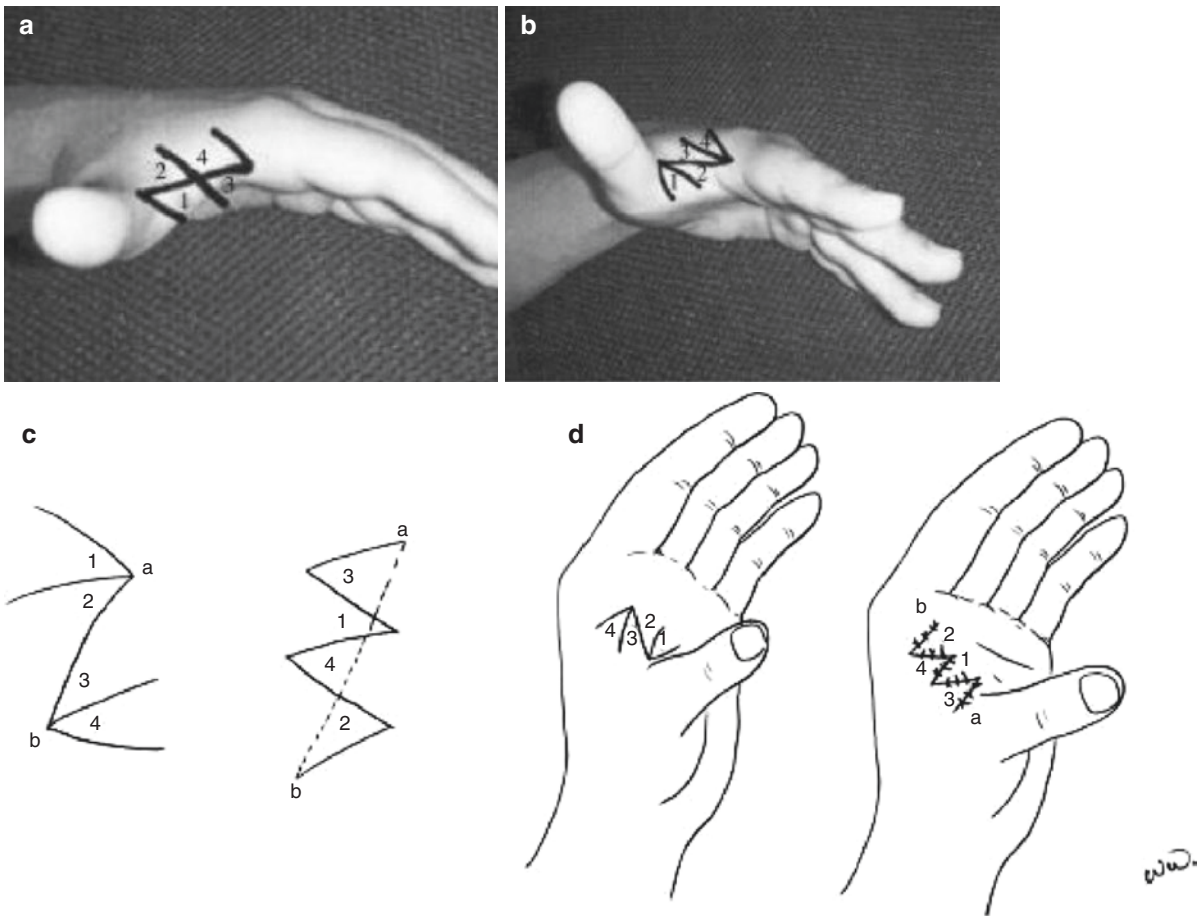


Fig. 5.59 Four-flap method thumb web expansion

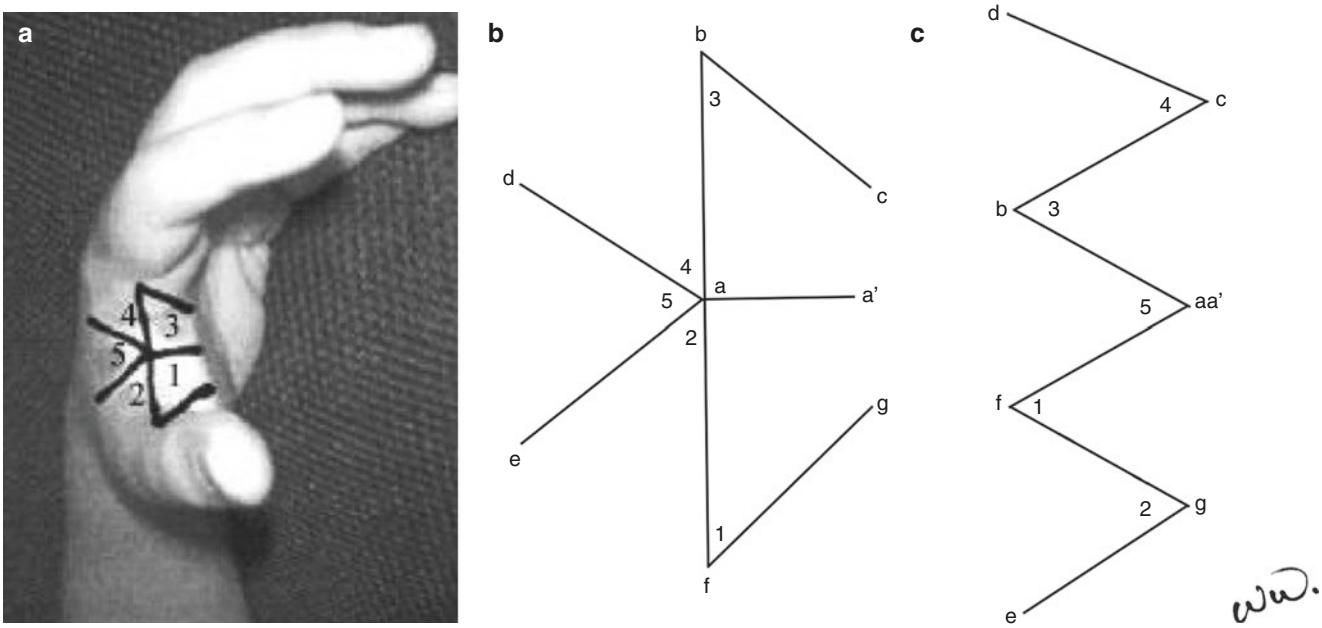


Fig. 5.60 Five-flap method thumb web expansion

flexor, the procedures of release, lengthening, or disjunction should be performed, and in severe cases, antedisplacement of volar plate can be considered. As for those with thumb long extensor dysplasia, it is suitable to perform dynamical reconstruction of the thumb long extensor, during which index finger inherent extensor grafting, radial wrist long extensor grafting, and volar long tendon grafting can be adopted to perform dynamical reconstruction of the thumb long extensor.

6. Treatment of hand external muscle dysplasia. Hand external muscle dysplasia includes thumb extensor dysplasia and thumb flexor dysplasia. As for thumb extensor dysplasia, inherent extensor tendon grafting is adopted in most cases, and radial wrist long extensor can also be used as dynamic tendon. In the case of the simple tendinous junction abnormality, the manifestation is abnormal junction between the thumb long flexor tendon and the thumb long extensor tendon apparatus; the abnormal tendinous junctions can be resected or disjuncted; if the strength of thumb flexion and extension is insufficient, tendon grafting can also be performed simultaneously. In case of thumb flexor dysplasia, the ring finger superficial flexor tendon grafting is adopted for the repair in most cases, and as for thumb extensor dysplasia, the index finger inherent extensor tendon grafting can be adopted for repair.
7. Correction of metacarpophalangeal joint accessory ligament relaxation. The accessory ligaments can be antedisplaced for fixation, and tendon transplantation can also be adopted for the replacement. After the accessory ligament reconstruction, the metacarpophalangeal joint is in the extension position. After fixation with Kirschner silk for 3 weeks, the patient can carry out functional activities during the daytime after the Kirschner silk is pulled out, and brace continues to be used during the night for braking for 3 weeks.

5.5.3.2 Treatment of Type III Thumb Dysplasia

The treatment methods of this type of deformity vary greatly, so the regimens can only be established after thorough consideration and can only be implemented after the profound consent from both doctors and patients is obtained.

Type III thumb dysplasia is the type with the greatest changes in congenital thumb dysplasia, and the degree of its functional lesion ranges from dysplasia of fingers, thumb, and the first metacarpal bone, to dysplasia of the metacarpophalangeal joint, carpometacarpal joint, hand internal muscles, and hand external muscles, to deformities of thumb growth direction and position. However, the patients with this type of deformity always have one finger which is morphologically and structurally similar to the thumb. According to the severity of deformity, type III thumb dysplasia can be reclassified into type IIIa, IIIb, and IIIc. Different deformities have different influences on hand functions. Thumb functional insufficiency, thumb functional absence, and the presence of diseased thumb can all affect the functions of the entire hand. If type IIIc is congenital abduction-type severe thumb dysplasia, in spite of the presence of the thumb, it serves no function. There is no uniform surgical method for this type of thumb dysplasia. Only designs have been made according to the status of hand function absence, and in most

cases, the deformed abducted thumb is resected and the thumb is reconstructed.

The treatment policy of type III thumb dysplasia is based on the results of an all-around evaluation on the shape, position, size, thickness, muscular strength, and thumb web of the fingers and thumbs of the deformed hands, as well as various kinds of supporting structures and dynamic apparatus.

1. As for the thumb with structural deformities and the absence, repair, and reconstruction can be performed according to the osteoarticular structures of the thumb and the deformities in hand internal and external muscles, the method of replacement can also be adopted, or free grafting of microsurgical tissue and organ can be performed.
2. As for the patients suffering nonfunctional thumb or the abduction-type thumb or serious dysplasia of the first metacarpophalangeal and carpometacarpal joints which cannot be repaired, the nonfunctional thumb can be totally or partially resected with the first radial finger transposition for pollicization plus reconstruction of the first fingerweb, and at the same time, the dynamic function of the thumb can be reconstructed. Partial resection of the abduction type morphologically unpleasant and nonfunctional thumb is to utilize the tissues at the retained parts as the tissue donor site for aesthetical remodeling and functional reconstruction.
3. As for the thumb with serious dysplasia or abduction-type thumb, in case the first metacarpal bone or the first metacarpophalangeal joint is usable, toe grafting, expanded toe grafting, or partial toe grafting for thumb reconstruction can be considered; metatarsophalangeal joint grafting for the reconstruction of metacarpophalangeal joint or carpometacarpal joint can be adopted.
4. As for the thumb with serious dysplasia or abduction-type thumb, in case phalange and metacarpal bone are usable, the original phalange and metacarpal bone can be used to correct deformities, and the structurally and functionally similar tissues can be selected for reconstruction of suitable dynamic devices and also as the donor sites for tissue transplantation for the morphological and functional reconstruction of the thumb.

   Typical case: A 16-year-old women, with type IIIc thumb dysplasia, namely, abduction-type thumb dysplasia; as for the patients with serious functional defect-type thumb deformities, right thumb adduction, palmar flexion deformity, thumb web stenosis, thenar and hypothenar dysplasia, and serious disorder in thumb flexion, extension, adduction, abduction, and palm-opposition functions, the X-ray film indicates that the metacarpophalangeal joint develops relatively well. According to the preoperative assessment, the deformity of thumb abduction is mainly induced by thumb radial contracture; therefore, the surgery should focus on correction of skin, tendon, joint and ligament deformities. The surgery is performed under the condition of brachial plexus anesthesia. First correct the thumb radial deviation deformity; design linguiform skin flaps at the dorsal side of thumb metacarpophalangeal joint; set the pedicle at the distal end, grafting the linguiform skin flap pedicle to the interior of the radial longitudinal incision of the metacarpophalangeal joint of the thumb; and correct the radial skin defect of the thumb. Perform the reshaping of the metacarpophalangeal joint, incise the skin and subcutaneous tissues, expose the extensor tendon hood of the thumb, incise the thumb exposure tendon hood, shorten the thumb long extensor tendon, cut off the metacarpophalangeal joint accessory ligament, prolong the radial metacarpophalangeal joint accessory ligament, and fix the metacarpophalangeal joint with a Kirschner silk (Fig. 5.61).
5. As for the deformity of nonfunctional or seriously functionally insufficient thumb (including the adduction-type thumb dysplasia), the following methods can be adopted:
   (a) The nonfunctional adduction-type thumb can be totally or partially resected with the first radial finger transposition for pollicization plus the reconstruction of the first fingerweb (viz., thumb web reconstruction), and in the meantime, the dynamic functions of the thumb can be reconstructed; or the total or partial resection of the nonfunctional adduction-type thumb can provide the tissue transplantation donor site for the aesthetical reconstruction of the thumb.
   (b) If the first metacarpal bone or the first carpometacarpal joint of the dysplastic adduction-type thumb is usable, toe grafting or expanded toe grafting can be considered for thumb reconstruction.
   (c) The index finger grafting or the grafting of the first finger at the radial side is the basic surgical method of the congenital thumb dysplasia and applicable to the reconstruction of various kinds of thumb dysplasia or the thumb absence, including the reconstruction of type IV thumb dysplasia (floating thumb), type V thumb dysplasia-type dysplastic complete thumb absence, and type VI thumb dysplasia.

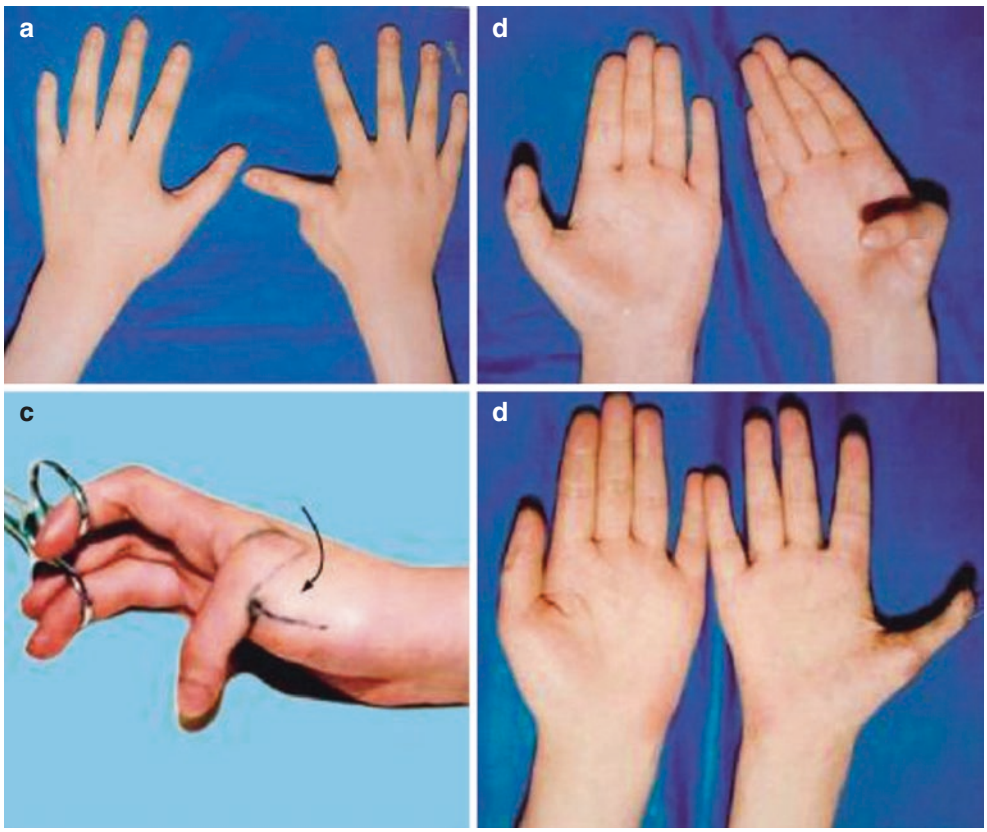


Fig. 5.61 Treatment of type IIIc thumb dysplasia. (**a**, **b**) Type IIIc thumb dysplasia of the right hand, appearance of hand dorsum and palm. (**c**) Surgical design: abduction-type thumb transposition; design one linguiform skin flap at the dorsal side of the thumb, repair thumb flexion and abduction after rotational transplantation, and perform repair and reconstruction of the corresponding tendons, joints, and ligaments. (**d**) Postoperative hand appearance

5.5.3.3 Treatment of Type IV Thumb Dysplasia (Floating Thumb)

The treatment method of this type of deformity is resection of the abandoned floating thumb and reconstruction of thumb in the rotating position of the index finger. For the surgical design and surgical methods, see the transposition of the second radial finger of the type V thumb dysplasia for thumb reconstruction (Fig. 5.62).

5.5.3.4 Treatment of Type V Thumb Dysplasia

Type V thumb dysplasia is a typical category of thumb dysplasia, manifested as complete thumb absence, the absence of all thumb structures including the first carpometacarpal joint but normal shape, structure, and functions of the remaining four fingers. The thumb reconstructing surgeries include the reconstruction of the first carpometacarpal joint, the reconstruction of the metacarpophalangeal joint, the reconstruction of the first metacarpophalangeal joint, and the dynamic reconstruction of the hand internal muscles and hand external muscles. Pollicization of the index finger is the optimal surgical regimen.

1. Surgical method. The surgical methods for reconstruction of thumb with congenital dysplasia are based on Buck-Gramcko's [16, 17] design of index finger pollicization worldwide. The author makes some modifications on this basis and calls it transposition of the radial finger for thumb reconstruction. The specific procedures are as follows:
   (a) Skin flap design: The surgical requirements are transposing the radial hand for thumb reconstruction, shortening the radial hand, and rotating it to the palm-opposition position. The main points include the following: ① the reconstructed thumb has two dactylopodites, and their length, thickness, ratio between the transverse diameter and the anteroposterior diameter, and the finger nail shape are close to those of normal thumb; ② the reconstructed thumb is located in the palm-opposition position and has a broad thumb web; ③ it has the stable first carpometacarpal joint, metacarpophalangeal joint, and interphalangeal joint; ④ it can effectively perform actions of abduction, adduction, flexion, and extension and has

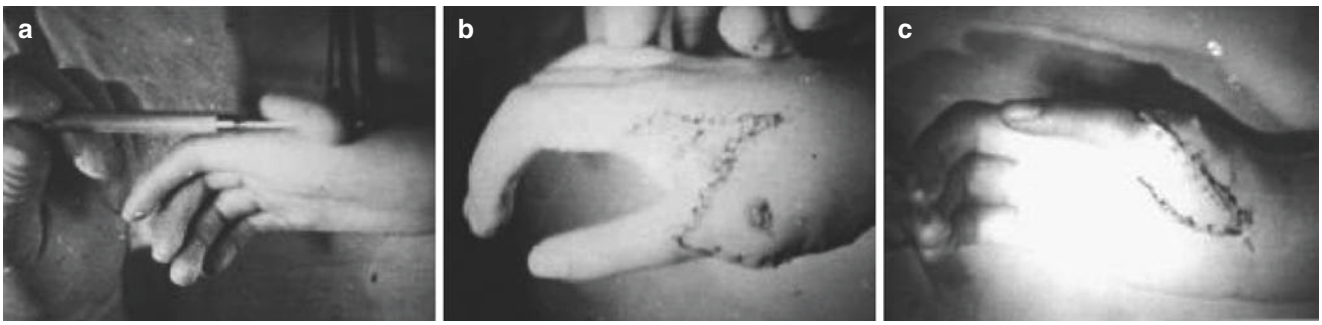


Fig. 5.62 Resection of the abandoned floating thumb and reconstruction of thumb in the rotating position of the index finger

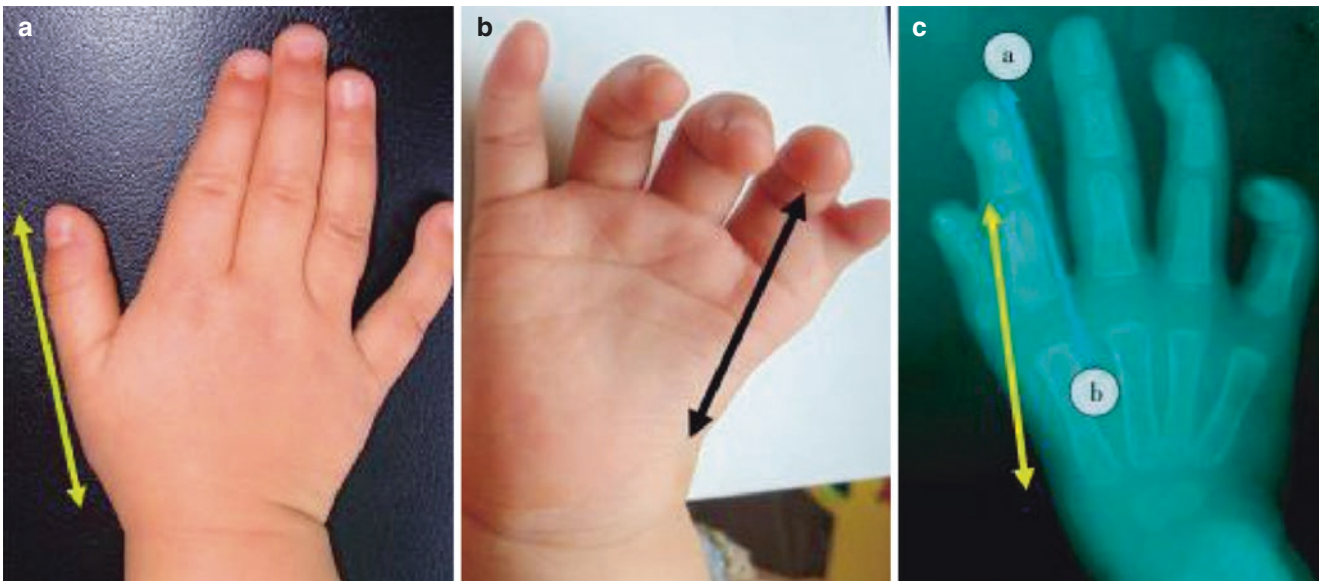


Fig. 5.63 Design of the length of the reconstructed thumb. (**a**) Measure the distance between the thumb tip and the thumb carpometacarpal joint; (**b**) measure the distance between the center of the carpometacarpal joint of the index finger and the volar transverse striation of the proximal interphalangeal joint of the index finger; (**c**) the selected length of the reconstructed thumb, namely, the line between *a* and *b*

rotation functions; and ⑤ the muscular strength of extension, flexion, abduction, adduction, and rotation functions are up to grade +4 and are aimed to reach grade 5.

In order to achieve the above effects, the specific procedures are designed as follows:

- Design of the length of the reconstructed thumb: There are two methods (Fig. 5.63): ① design the length of the reconstructed thumb according to the length of the healthy side of the thumb and measure the distance between the thumb tip and the carpometacarpal joint of thumb and ②measure the distance between the center of the carpometacarpal joint of the index finger and the volar transverse striation of the proximal interphalangeal joint, which can serve as the reference value for the length of the reconstructed thumb. The length of the thumb of children aged between 1 and 4 is 5.5–8 cm.
- Localization of the first carpometacarpal joint of the reconstructed thumb: At the radial margin of the palm, which is equivalent to the site of the first carpometacarpal joint, design the fixed-point c at the carpometacarpal joint of the reconstructed thumb as the basilar part of the reconstructed first carpometacarpal joint.
- Design of rotation flap in the palm-opposition position of the index finger: At the proximal interphalangeal joint at the dorsal side of the grafted finger (index finger), design skin flaps 1 and 2, which are retrograde flaps, with transverse striation part of the proximal interphalangeal joint of the digital dorsum as the pedicle, and flap 1 is slightly wider than flap 2. The width of flaps 1 and 2 at the dorsal side of the index finger can control the rotation angle of the grafted finger; if flap 1 is wider, the angle of rotation of the thumb to the palm is larger. Flaps 1 and 2 are overridden on flap 3,

the transposed flap 2 constitutes the thenar skin of the reconstructed thumb, and the transposed flap 1 constitutes the dorsum of the reconstructed thumb.

- Design of the basal flap of the reconstructed first metacarpophalangeal joint: At the radial margin of the palm, design flap 3, which is an isosceles triangular flap; the middle point of the bottom edge is the radial middle point of the metacarpophalangeal joint of the reconstructed thumb and also the basal part of the radial margin of the reconstructed thumb. The size of flap 3 should be decided according to the size of the reconstructed thumb, the length of the bottom edge is about 1/2 of the circumference of the reconstructed thumb, and the side length is the total length of flaps 1 and 2. The cusp of flap 3 is inserted between flaps 1 and 2.
- Flap design for thumb web reconstruction: At the volar side of the finger, design flap 5, which is used for thumb web reconstruction. The flap 5 should be made larger in design so that the thumb web of the reconstructed thumb can be wide enough. The size of flaps 3 and 5 can be adjusted according to the tissue quantity of the hand skin.
- Then at the volar transverse striation proximal side of interphalangeal joint of the grafted index finger, design flap 4, whose distal margin is 0.5–1 cm below the transverse striation of the proximal interphalangeal joint of the grafted finger. This flap can be involved in the composition of the first fingerweb, namely, the basal part of the volar margin of the proximal end of the reconstructed thumb, and it can move toward the thumb web skin (Fig. 5.64).

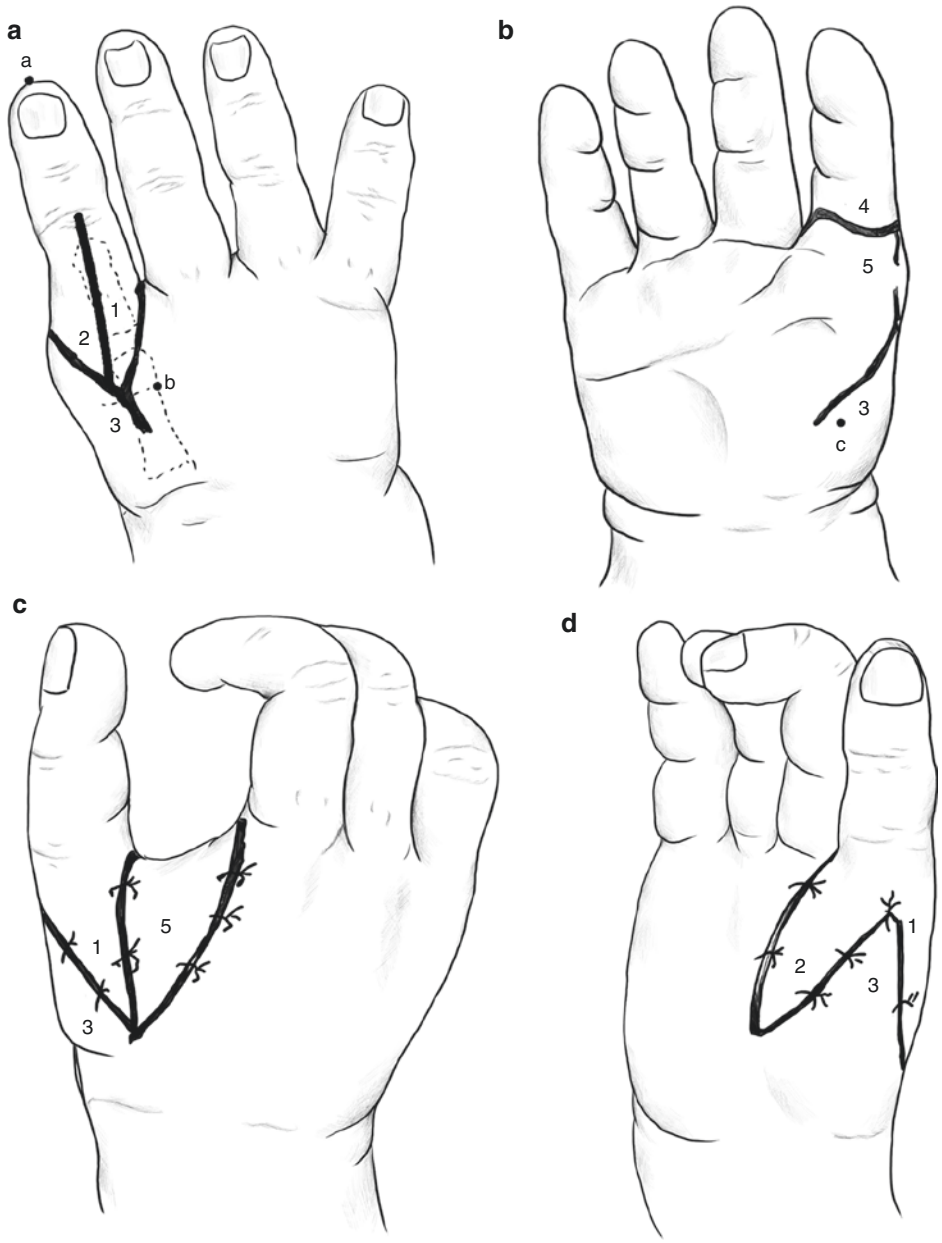


Fig. 5.64 Design of radial finger grafting for thumb reconstruction for patients with type V thumb dysplasia. (**a**) Set point *a* at the index finger tip, and design point *b* at the body surface projection of the neck of metacarpal bone of the radial finger; *ab* is the length of the reconstructed thumb; design flaps 1 and 2 of the grafted fingers, which, respectively, constitute the basal part of the reconstructed thumb. (**b**) At the margins of the palm and the dorsum, design flap 3, which constitutes the radial bottom edge; design flaps 4 and 5, which form thumb web. (**c**, **d**) Post-flap transposition for thumb reconstruction

(b) Reconstruction and early fixation of the first carpometacarpal joint: Shorten the metacarpal bone of the index finger, and use the metacarpophalangeal joint of the index finger to reconstruct the metacarpophalangeal joint of the thumb. The author has made improvements in terms of flap design, thumb web reconstruction, application of abandoned fingers and tissues for thumb enlargement and reconstruction, as well as reconstruction and fixation of carpometacarpal joint and completed the reconstruction of various skin flap thumb dysplasia in over 100 patients with thumb absence. The main point of the surgery is to shorten the metacarpal bone, transform the metacarpophalangeal joint with a large range of motion into the stable first metacarpophalangeal joint, transform the proximal phalanx of the index finger into the first metacarpal bone, and transform the proximal interphalangeal joint of the index finger into the metacarpophalangeal joint of the reconstructed thumb. The surgical methods are as follows:

- Reconstruction of the first metacarpal bone: Reconstruct the proximal phalanx of the grafted finger into the first metacarpal bone. Partially amputate the first metacarpal bone, raise flaps 1 and 2 under the incisions of the hand dorsal skin, cut down the common extensor muscle of finger and mark them, and retain them until the late surgical stage and perform anastomotic repair to shorten the disjuncted extensor tendons. Expose the shaft of the first metacarpal bone; protect the hand dorsal veins from injuries; at the site about 8 mm away from the proximal end of the metacarpophalangeal joint of the first metacarpal bone (viz., the neck of the metacarpal bone), use 4–6 mm hand surgery bone chisel or miniature reciprocating saw to cut off the first metacarpal bone; and only retain the head of the first metacarpal bone. Reamputate the residual metacarpal bone part at the proximal end of the shaft of the first metacarpal bone, and resect periosteum simultaneously, until the carpometacarpal joint is reached to form a space so that the carpometacarpal joint of the reconstructed thumb and the reconstructed first metacarpal bone can be contained.
- Reconstruction of the first metacarpophalangeal joint and establishment of the palm-opposition position of the reconstructed thumb: Transform the metacarpophalangeal joint into the carpometacarpal joint, and transform the metacarpophalangeal joint of the grafted index finger into a trapezial shape, which may aim to lengthen the reconstructed first metacarpal bone. Because the volar plate of the first metacarpophalangeal joint is relatively relaxed, its range of flexion and extension can be up to 115°–120°. In order to correct the relaxed joint capsule, rotate the head of the first metacarpal bone to the dorsal side by 90°, and use a 3-0 nylon thread to suture, and fix the joint capsule at the broken end of metacarpal bone and dorsal base of proximal phalanx of index finger to correct the relaxed volar plates and tightened accessory ligaments of joints. Rotate the carpometacarpal joint of the reconstructed thumb to the palm-opposition position and mount it to the wrist (Fig. 5.65).
- Localization and fixation of the carpometacarpal joint of the reconstructed thumb: In the position where the skin at the hand dorsum is incised, namely, the basal part of flap 3, use a vein retractor to rip the hand dorsal vein, expose point c of the palm, use a thin steel silk or 3-0 nylon thread to suture, and fix the head of metacarpal bone into the trapezial position. At the end of surgery, use the tracted fine steel silk or 3-0 nylon thread outlay as minus-tension pad and tie them for fixation, and take them out after 3 to 4 weeks. The minus-tension fixation should not be too tight to prevent the skin compression in the minus-tension region from inducing ischemic necrosis (Fig. 5.66).

(c) Reconstruction of palm-opposition position of pollicized fingers: After thumb reconstruction, it is necessary to make the reconstructed thumb be in the palm-opposition position. Abduct the reconstructed thumb by 80°–90°, make the coronal plane of the finger pulp pronate so that it can form an included angle of 45°–60° with the coronal plane of the index finger, and make the thumb present the palm-opposition position so that it can be opposite to the remaining four fingers. The reconstructed thumb from the index finger spins to the palm-opposition position per se, namely, a rotation in the axial direction by 135°–140°, and it is fixed in this direction (Fig. 5.67).

(d) Dynamical reconstruction after finger pollicization: This is an important link in thumb reconstruction. After the grafting of the flexor tendon of the transposed index finger (or other finger), the thumb long flexor and thumb brevis flexor are formed, and after the transposition of the original finger flexor tendon, it is shortened in a natural way. However, after the transposition of the finger extensor tendon, it is necessary to shorten and suture it after disjunction and perform the functions of the thumb long extensor; after changing the directions, fix the inherent extensor of the index finger at the bottom of the proximal phalanx to perform the functions of the thumb long abductor. Suture the dorsal interosseous muscles with the radial bundle of the extensor tendon of the transposed finger to perform the functions of thumb short abductor; suture the volar interosseous muscles with the ulnar bundle of the extensor tendon of the transposed finger to perform the functions of the thumb adductor. The surgical methods are as follows: separate the dilating

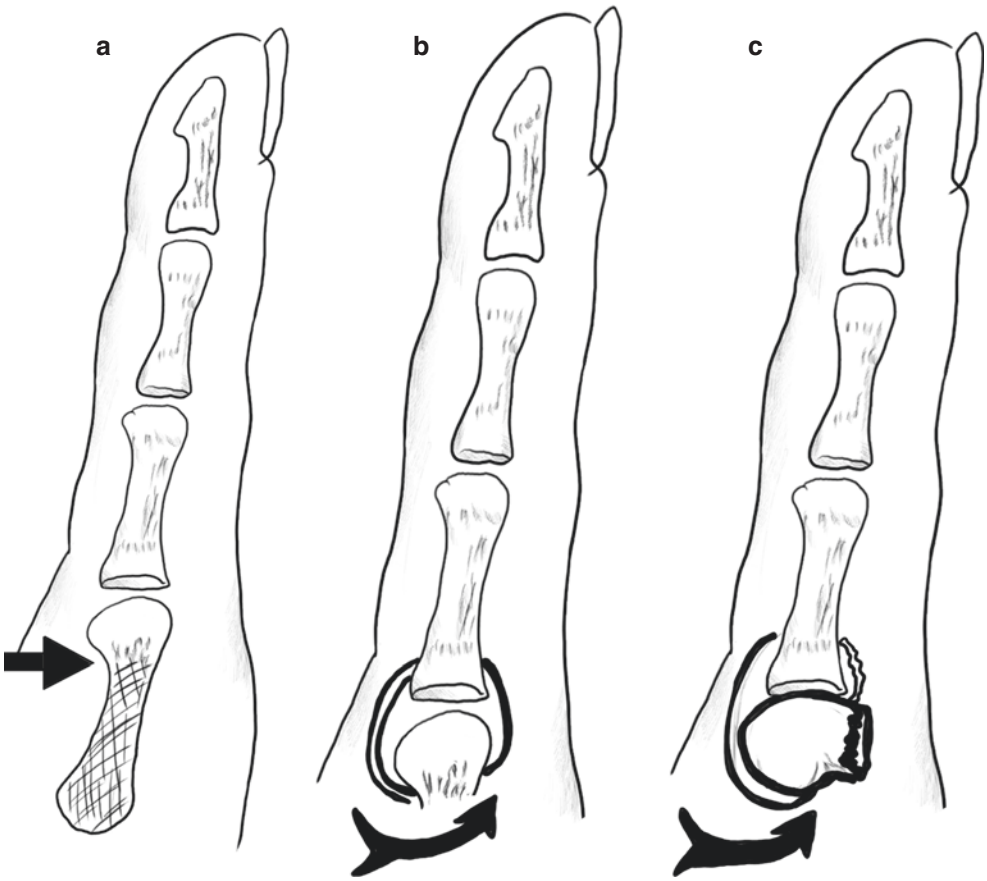


Fig. 5.65 Schematic diagram of reconstruction of the first carpometacarpal joint. (**a**) Partially resect the first metacarpal bone at the radial side of the reconstructed thumb, and amputate the neck of the metacarpal bone; (**b**) Rotate the first head of metacarpal bone to the dorsal side by 90°, and use a 3-0 nylon thread for suturing and fixation. (**c**) Tighten the relaxed metacarpophalangeal joint

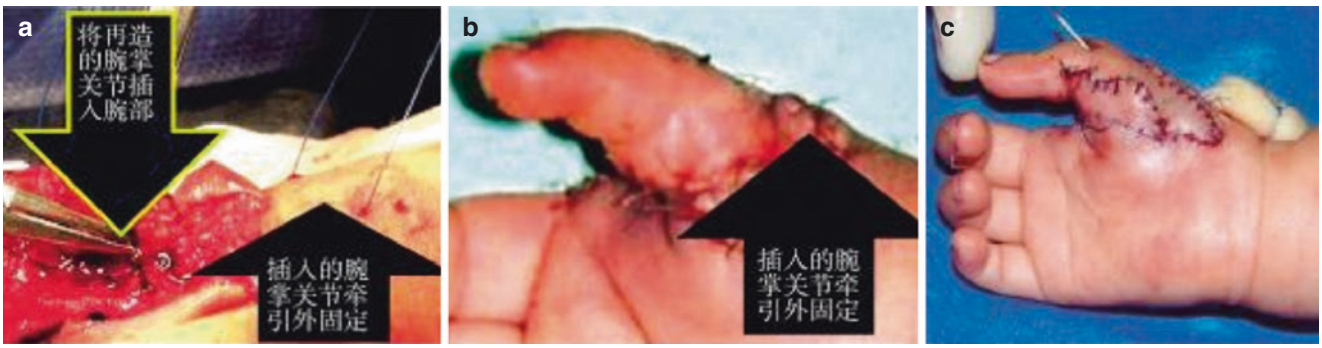


Fig. 5.66 Localization and fixation of the carpometacarpal joint of the reconstructed thumb. (**a**) Insert the reconstructed first carpometacarpal joint into the basal part of flap c. (**b**) Use a thin steel silk to tract the first carpometacarpal joint inserted into the wrist and fix it on the wrist skin. (**c**) Immediately after thumb reconstruction

part of the index finger extensor tendon from the digital dorsum, and cut the proximal part of the proximal phalanx. Divide the distal ends of the cut tendons into three bundles, suture the middle bundle with the proximal end of the common extensor tendon of the index finger, and suture the internal and external bundles with the reconstructed thumb adductor tendons and the thumb short abductor tendons. After changing the directions of the inherent extensor of the index finger, fix it at the bottom of the proximal phalanx, and reconstruct it into the thumb long abductor. Microsurgical minimally invasive techniques are adopted for the dynamical reconstruction of pollicized fingers, whether it is retaining, disjunction, or suturing of the tendons, and the operation is carried out under a magnifier or operating microscope. The tendon suturing method is to use 6-0~7-0 nylon threads to perform penetrating weaving and

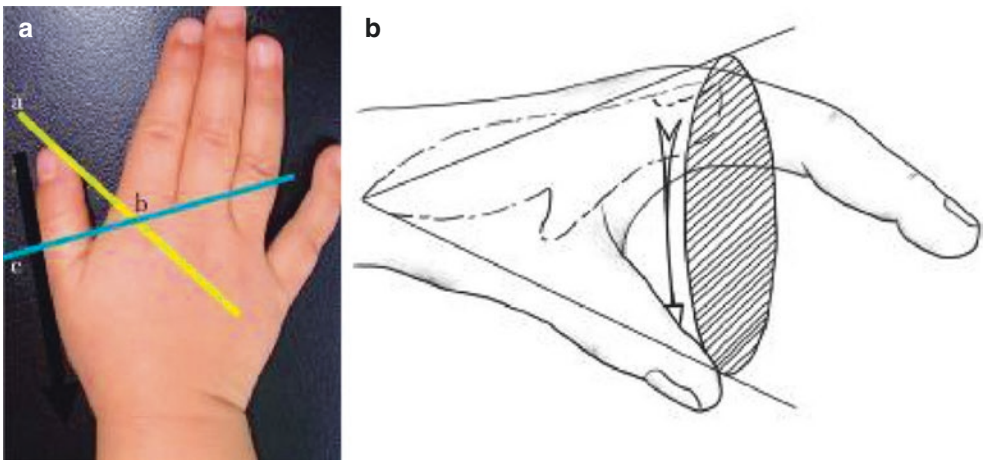


Fig. 5.67 Reconstruction of palm-opposition position of pollicized fingers. (**a**) Take a normal hand as an example. Rotate the thumb by 45°, and make it at the palm-opposition position. *ab* extending line (*yellow line*) is the coronal position horizontal line of the thumb pulp. *cb* extending line (*blue line*) is the coronal position horizontal line of the index finger pulp, and *abc* included angle is 45° and is the rotation amplitude of the index finger (or the finger). (**b**) The index finger of the original hand is rotated to the volar side by 90°, the thumb reconstructed from the index finger spins per se, namely, an axial rotation of 135°–140°, and the palm-opposition position of other fingers are formed (the *dash lin*e is the reconstructed thumb designed to be rotated)

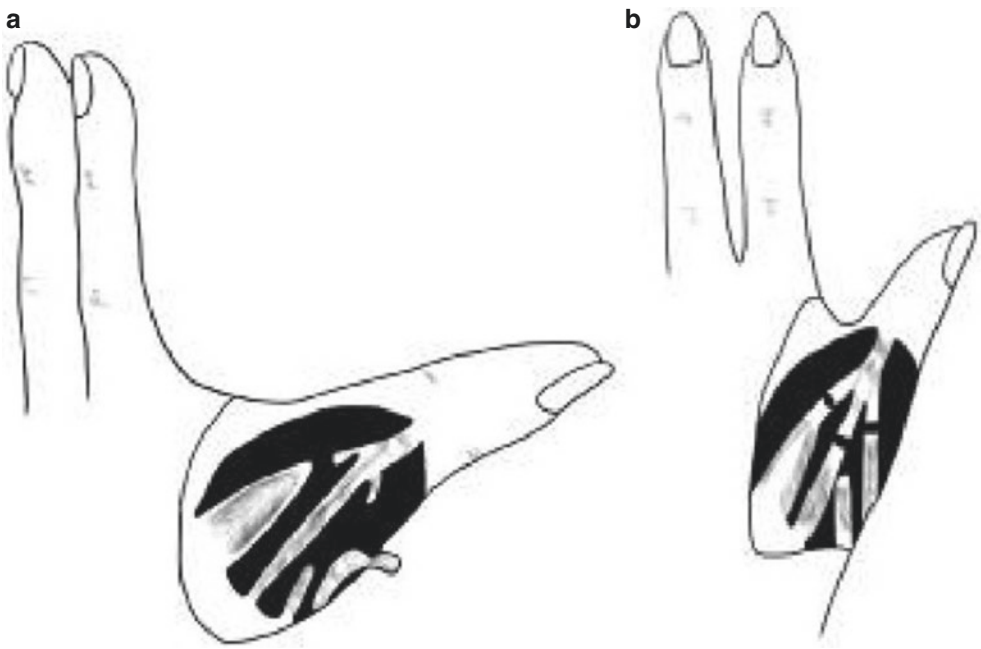


Fig. 5.68 Dynamical reconstruction after finger pollicization

reinforced suturing as well as accurate aponeurosis microsurgical suturing by surrounding the tendons (Fig. 5.68). Only with good dynamical devices can thumb reconstruction serve optimal and complete fine movement functions.

(e) Application of brace and functional training after surgery: Fix brace in thumb palm-opposition position. Remove the fixation sutures of the first carpometacarpal joint 2–3 weeks after operation. As the reconstructed first carpometacarpal joint is not stable enough, and healing needs a period of time after tendon grafting, it is advisable to use a static brace after surgery to keep the thumb palm-opposition position and maintain it for 5–6 weeks and perform functional training of the reconstructed thumb afterwards.

(f) Typical case: A 2-year-old boy with type V thumb dysplasia, four-fingered thumb absence, and thumb dysplasia; the patient underwent the transposition of the first finger for thumb reconstruction.

Surgical design:

- Design points a and b at the back of hand; their tie-line is the length of the reconstructed thumb (from the thumb tip to the margin of the proximal end of the first carpometacarpal joint).
- Design point c, which is approximately located at the proximal end of the middle of the distal and proximal rows of the carpal bones and is the midpoint of reconstructed first carpometacarpal joint. The length from point c to the cross striation of the proximal segment of the second finger at the radial side is equal to ab line, namely, the length of the reconstructed thumb, and the point c of the palm is lower than the point b of the hand dorsum.
- Design flaps 1 and 2 at the root of radial fingers. In order to control the rotation angle of palm-opposition position of the reconstructed thumb, the width of flap 1 should be equal to or larger than that of flap 2, and flap 1 constitutes the basal part of the dorsal side of the reconstructed thumb; flap 2 constitutes the basal part of the volar side of the reconstructed thumb.
- Design flap 3, which constitutes the radial skin covering edge of the carpometacarpal joint of the reconstructed thumb.
- Design flap 4 at the basilar part of the radial finger of the palm, which constitutes the thumb web edge of the root of the reconstructed thumb.
- Design flap 5, insert the ulnar side of flap 1, and reconstruct thumb web.
- The top (viz., point d) of the midline segmentation line of flaps 1 and 2 is located at the proximal end of the transverse striation of the proximal phalanx of the digital dorsum of the reconstructed thumb, and during the operation, point d and the top of flap 3 are sutured (Figs. 5.64 and 5.69).

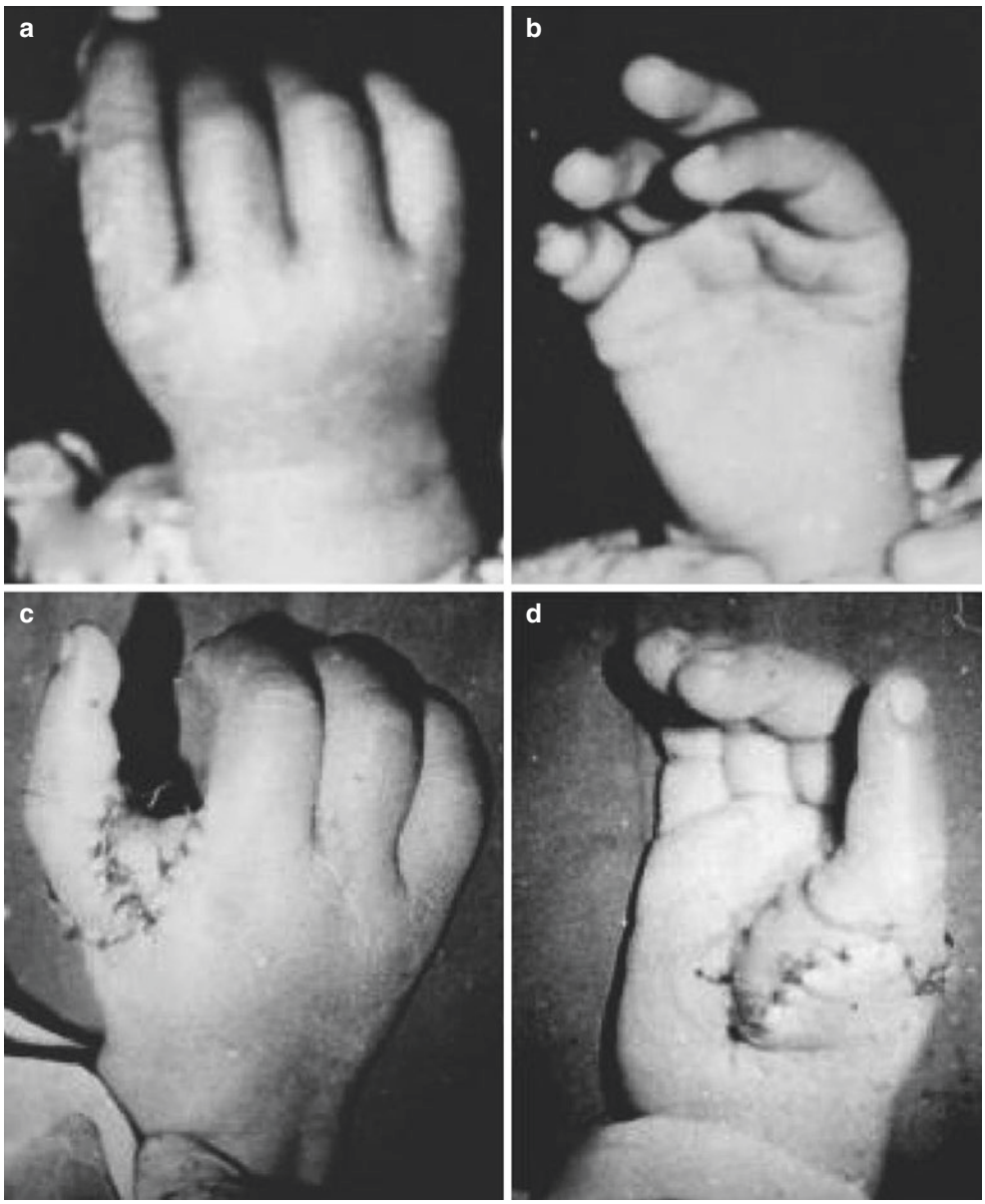


Fig. 5.69 Reconstruction of the thumb with type V thumb dysplasia. (**a**, **b**) Before operation. (**c**, **d**) After operation

5.5.3.5 Treatment of Type VI Thumb Dysplasia

Type VI thumb dysplasia includes hyperphalangeal thumb dysplasia and brachydactylic thumb dysplasia (Fig. 5.70). As for the hyperphalangeal thumb dysplasia, the radial side is selected for hyperdactylia removal, and finger pollicization is performed for treatment; the treatment of brachydactylic thumb dysplasia however is discussed in another section.

The anatomical features of type VI thumb dysplasia include ① the absence or serious dysplasia of the first carpometacarpal joint; ② no sellaeform first carpometacarpal joint able to move in three axial directions; ③ thumb phalange absence or serious dysplasia; ④ no broad first fingerweb; ⑤ thenar muscular absence or serious dysplasia; ⑥ the absence or serious dysplasia of thumb extensor, thumb flexor, and thumb long abductor; ⑦ often concomitantly with vascular and nervous dysplasia in the entire hand and forearm; and ⑧ sharp and fine index finger and poor shape and functions. As the patient has the above anatomical defects, replacement and correction should be thoroughly considered in the design of thumb reconstruction.

1. Treatment principle:
   (a) As for the patients with five-fingered thumb absence and good finger development, shorten the radial fingers, shorten the first carpometacarpal joint, reconstruct the first carpometacarpal joint, reconstruct the dynamical functions of hand internal and external muscles, and reconstruct the thumb; the surgical methods are similar to the thumb reconstruction in patients with type V thumb dysplasia. If the deformed hand has five fingers, the shape and function of the reconstructed thumb and hands are good.
   (b) As for the patients with brachydactylic thumb absence or dysplasia, shorten the radial fingers, correct and transplant deformities, shorten the first metacarpal bone, transpose the palm-opposition position, recon-

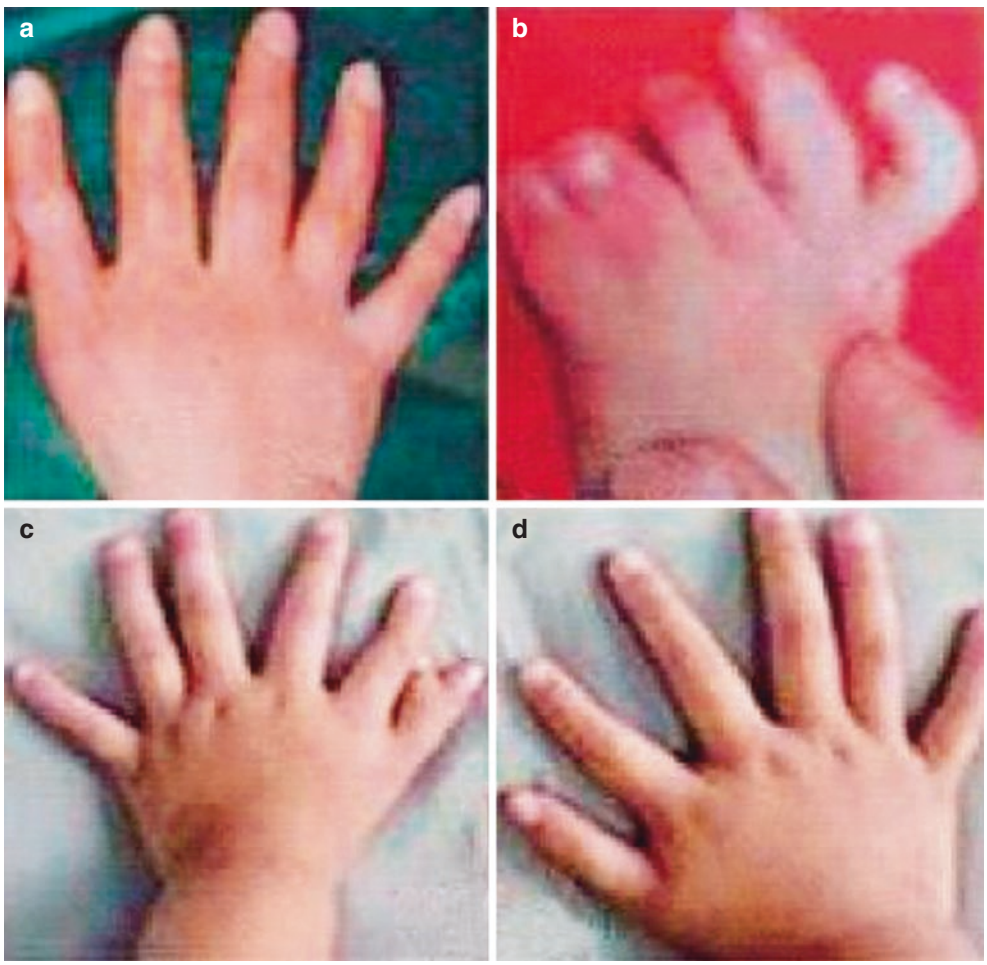


Fig. 5.70 Type VI thumb dysplasia. (**a**) Type VIaa: five-fingered hyperphalangeal thumb dysplasia. (**b**) Type VIab: five-fingered brachydactylic thumb dysplasia. (**c**, **d**) Type VIb: six-fingered and seven-fingered hyperphalangeal thumb dysplasia

struct the first carpometacarpal joint, reconstruct thumb web, reconstruct the dynamical functions of hand internal and external muscles, and reconstruct thumb.

(c) As for the patients with more-than-five-fingered hyperphalangeal thumb dysplasia, adopt the partial resection of hyperdactylia, fabricate the hyperphalangeal island skin flaps to enlarge and reconstruct the thumb, shorten the radial fingers, perform palm-opposition transposition and transplantation, shorten the first metacarpal bone to reconstruct the first metacarpocarpal joint, reconstruct the functions of hand internal and external muscles, and reconstruct thumb.

2. Skin flap design. Taking type VIb thumb dysplasia (six-fingered thumb dysplasia) as an example, perform the design of thumb reconstruction similar to type V thumb dysplasia.

Design 5 to 6 flaps for grafting at the hand dorsum and the palm; the procedures are as follows:

(a) Design of length of reconstructed thumb: Measure the distance between the tip and the carpometacarpal joint of the normal thumb (viz., the distance between the proximal end of interphalangeal joint and the carpometacarpal joint of index finger) to decide the length of the reconstructed thumb.

(b) Point fixation of the carpometacarpal joint of the reconstructed thumb: At the radial margin of the palm, design the fixed-point c at the carpometacarpal joint of the thumb as the part of the reconstructed first carpometacarpal joint.

(c) Design of the medial and lateral flaps of the constructed thumb: Design flaps 1 and 2 at the proximal end of the interphalangeal joint at the dorsal side of the grafted finger of the reconstructed thumb, flap 1 is larger than flap 2 (both of which are retrograde flaps), and end at point d with the transverse striation part of the proximal interphalangeal joint as the pedicle.

(d) Design of basal part of carpometacarpal joint of the reconstructed thumb: Design flap 3 at the carpometacarpal joint of radial margin of the palm to serve as the radial margin of the carpometacarpal joint of the reconstructed thumb; its size is up to the size of the reconstructed thumb. Flap 3 is a triangular flap, and its bottom edge is 1/2 of the diameter of the reconstructed thumb. This flap is inserted between flaps 1 and 2.

(e) Design of thumb web of reconstructed thumb: Design flap 5 at the margin of the palmar surface of the finger; its distal end is about 0.5 cm away from the transverse striation of the interphalangeal joint as the basal part of the volar side of the reconstructed thumb web. Flap 5 should be larger and will constitute the lower margin of the volar side of the thumb web of the constructed thumb. In the patients with hyperphalangeal thumb absence, the skin to be removed due to hyperdactylia can be fabricated into the tissue flaps connected with flap 6, which can serve as the skin source to enlarge and beautify the reconstructed thumb or expand thumb web.

(f) Design of thumb web margin of reconstructed thumb: Design flap 4; its proximal end is 0.5–1 cm away from the volar transverse striation of the interphalangeal joint. This flap can be involved in the composition of the first fingerweb (thumb web), namely, the basal migration part of the ulnar margin of the reconstructed thumb.

Incise the skin along the flap of the palm, raise flap 5, incise the skin at the proximal part of flap 5, and dissect the digital vascular nerve bundle and protect it. Use an arterial vascular clamp to block the ulnar side of the reconstructed thumb, the radial digital common artery of the adjacent fingers, the ulnar inherent arteries of the reconstructed thumb, and the radial inherent arteries of the adjacent fingers. After these arteries are blocked, under the condition that finger blood supply is not affected, the radial inherent arteries of the adjacent fingers can be cut and disjuncted safely so that the grafted reconstructed thumb has the blood supply from ulnar and radial inherent arteries.

(g) Design of aesthetical morphology of the reconstructed thumb: Use the island vascular neural flap of resected finger as flap 6 to increase the fullness of the reconstructed thumb as well as beautify finger shape and improve finger functions. This flap design is applicable to thumb reconstruction of six-or-more-fingered thumb defects (Fig. 5.71).

3. Typical case: A 5-year-old boy with type VIb thumb dysplasia, six-fingered thumb absence, and dysplasia. The patient has an obvious history of familial genetic diseases. His maternal grandmother and mother suffer from five-fingered thumb dysplasia, namely, type VIa thumb dysplasia (Fig. 5.27). As for the surgery, remove the extra fingers, perform the transposition of the radial second or third finger for thumb reconstruction, prepare the partial finger pulp of the resected fingers into vascular nerve island shape flaps for the enlargement of the reconstructed thumb, and assist in the morphological and functional reconstruction of the reconstructed thumb. There is no difference between the postoperative reconstructed thumb and the normal thumb in terms of shape and function (Fig. 5.72).

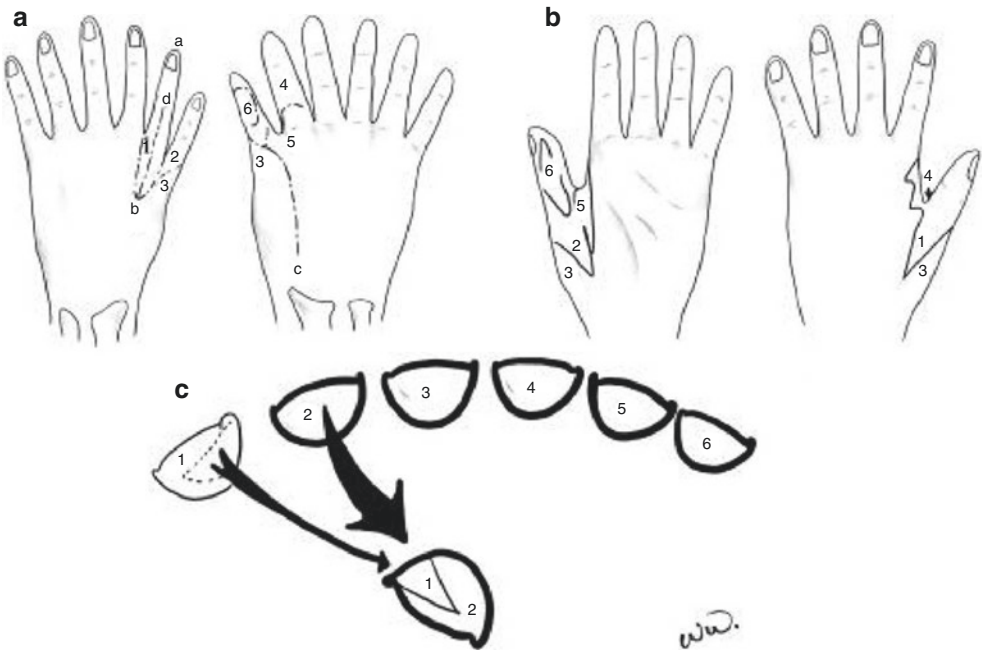


Fig. 5.71 Design of flap surgery for reconstruction of thumb with hyperphalangeal thumb dysplasia. (**a**) Flap design for reconstruction of six-fingered hand. (**b**) Flap position after thumb reconstruction. (**c**) Thumb reconstruction for flap combination of the radial first and second fingers

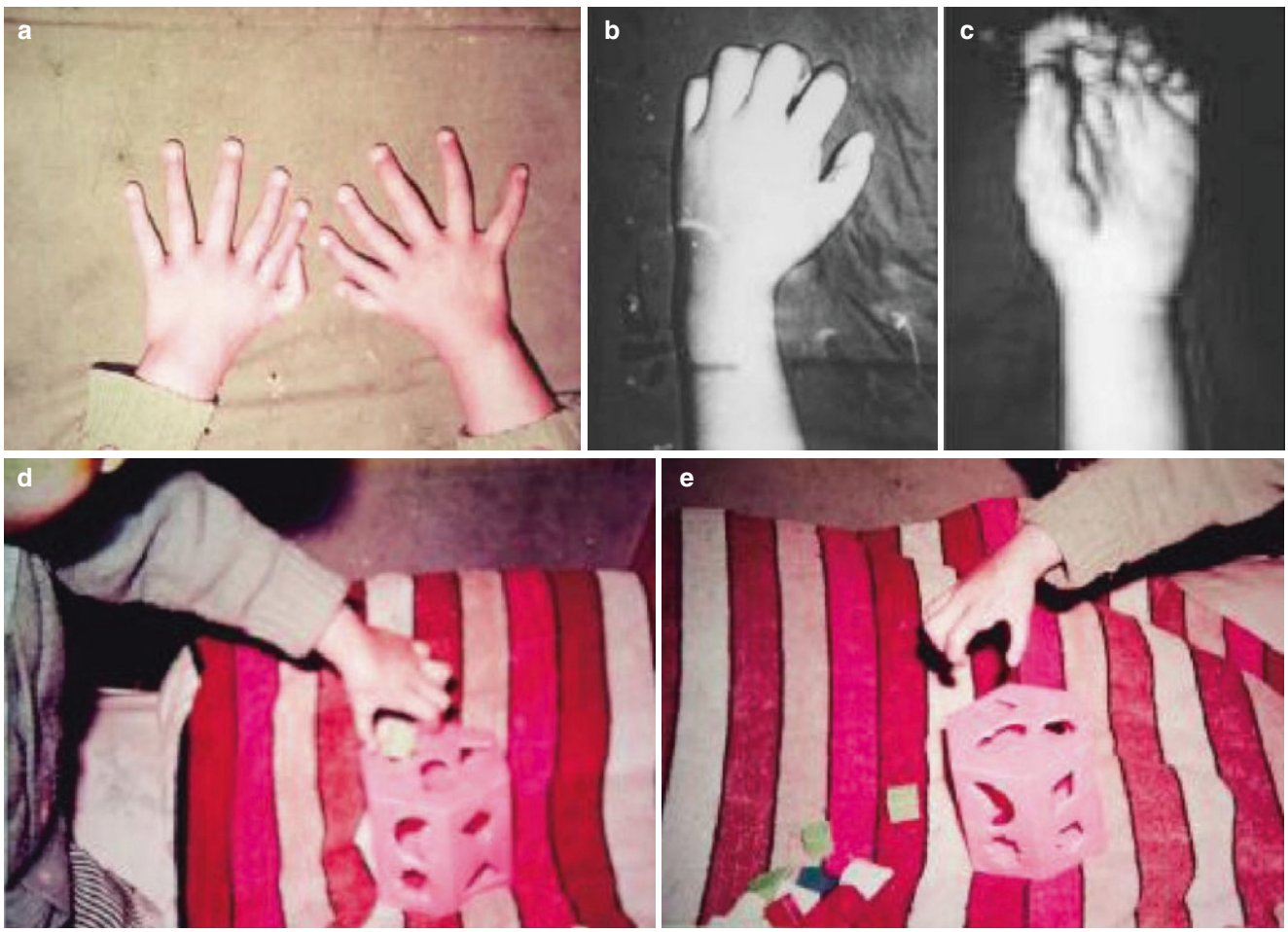


Fig. 5.72 Treatment of type VIb thumb dysplasia. (**a**–**c**) Before operation. (**d**, **e**). After operation

5.5.3.6 Treatment of Atypical Type VIa (Five-Fingered) Thumb Dysplasia

Atypical type VIa (five-fingered) thumb dysplasia refers to five-fingered hand, but the serious dysplasia of the radial first and second fingers is often manifested as syndactylia of the first and second fingers. This deformity is relatively common clinically.

Typical case: A girl at the age of 2 years and 9 months, with type VIa thumb dysplasia in both hands, atypical type VIa thumb dysplasia in the left hand, five-fingered thumb dysplasia, syndactylia of the radial first and second fingers, and serious dysplasia of the radial first finger, underwent the pollicization of the radial second finger of both hands for thumb reconstruction. The surgical design and procedures include design of thumb length, resection of the radial first finger, and preparation of the first syndactylia into the flaps for the enlargement of the reconstructed thumb (Fig. 5.73).

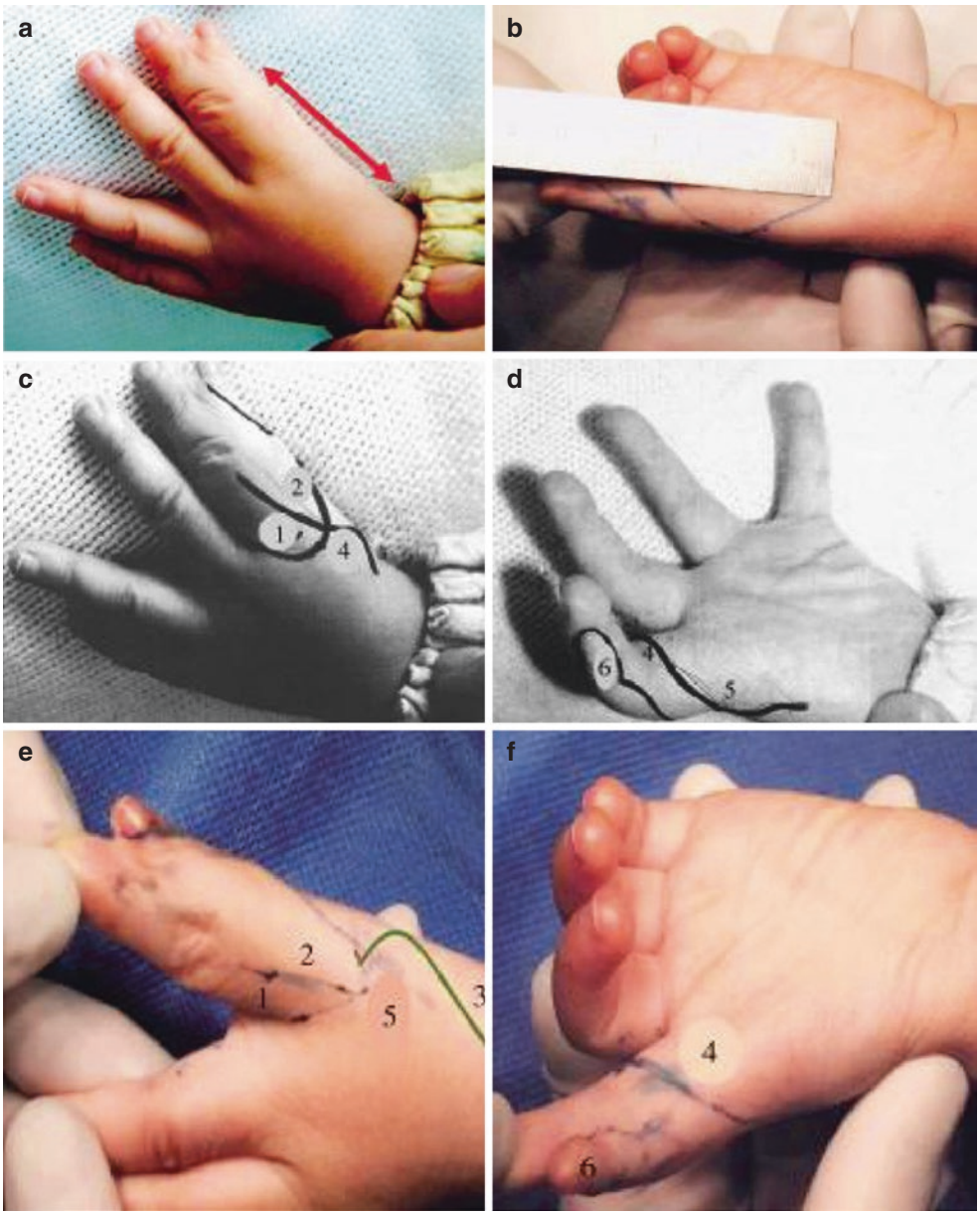
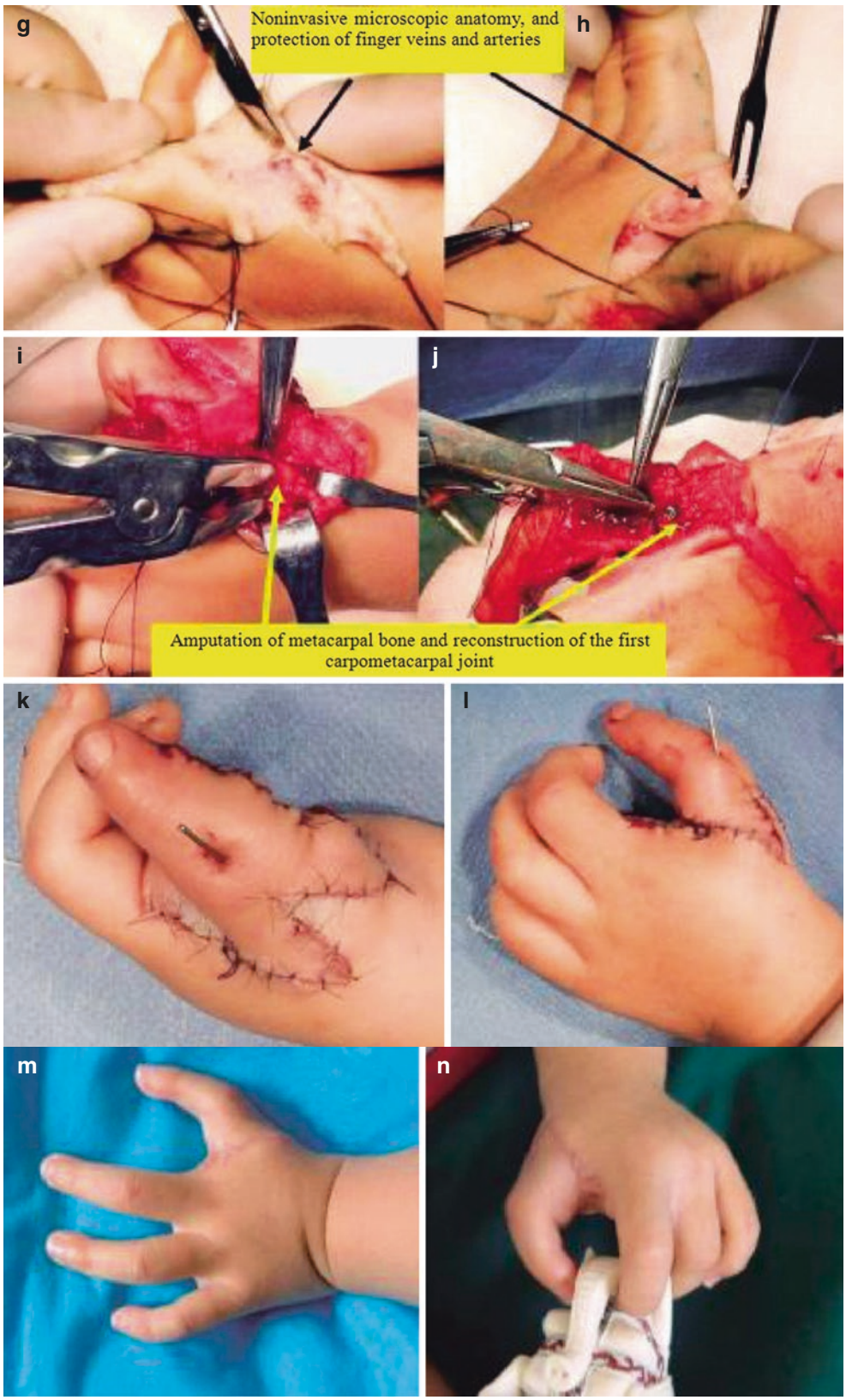


Fig. 5.73 Treatment of type VIa thumb dysplasia. (**a**, **b**) The designed length of reconstructed thumb is 5.5 cm. (**c**–**f**) Flap design of thumb reconstruction. (**g**, **h**) Shorten the radial second finger, perform palm-opposition transposition and grafting for thumb reconstruction, protect the dorsal digital vein and digital artery during operation, and prepare flap for thumb reconstruction. (**i**, **j**) Reconstruction and fixation of the first carpometacarpal joint. (**k**–**n**) After surgery, the shape, size, position, and length of the reconstructed thumb are similar to those of normal thumb, the width of thumb web is normal, and the palm-opposition function and palm-opposition muscular strength are good

Fig. 5.73 (continued)



5.5.3.7 Traction and Fixation of Carpometacarpal Joint Are Important Steps in Thumb Reconstruction

Traction and fixation of thumb carpometacarpal joint are important steps in thumb reconstruction, but the operation is not easy. Generally, the reconstructed metacarpophalangeal joint is sutured to the distal row of carpal bone, but as the position is relatively deep, it is sometimes difficult to finish the operation. The author used steel silk to suture the reconstructed carpometacarpal joint and led it to the outside of the wrist skin through the injection needle; the gauze minus-fixation method can produce good effects.

Typical case: Take the thumb reconstruction of the atypical type VIa thumb dysplasia (five-fingered thumb dysplasia) as an example. Nine-year-old girl of 10 months, with type VIa thumb dysplasia in both hands, atypical type VIa thumb dysplasia in the right hand (five-fingered), syndactylia of the radial first and second fingers, and serious dysplasia of the radial first finger, similar to the floating thumb, and with the left hand suffering typical type VIa thumb dysplasia, underwent finger pollicization for thumb reconstruction in the two hands at the same time. In the right hand, the seriously dysplastic radial first finger is resected, the radial second finger is transposed and shortened for pollicization, and the dynamical reconstruction of hand internal and external muscles are reconstructed; in the left hand, the radial first finger is pollicized for thumb reconstruction, the designed length of the reconstructed thumb is 8 cm, and the surgical methods and techniques are the same as those for the right hand (Fig. 5.74).

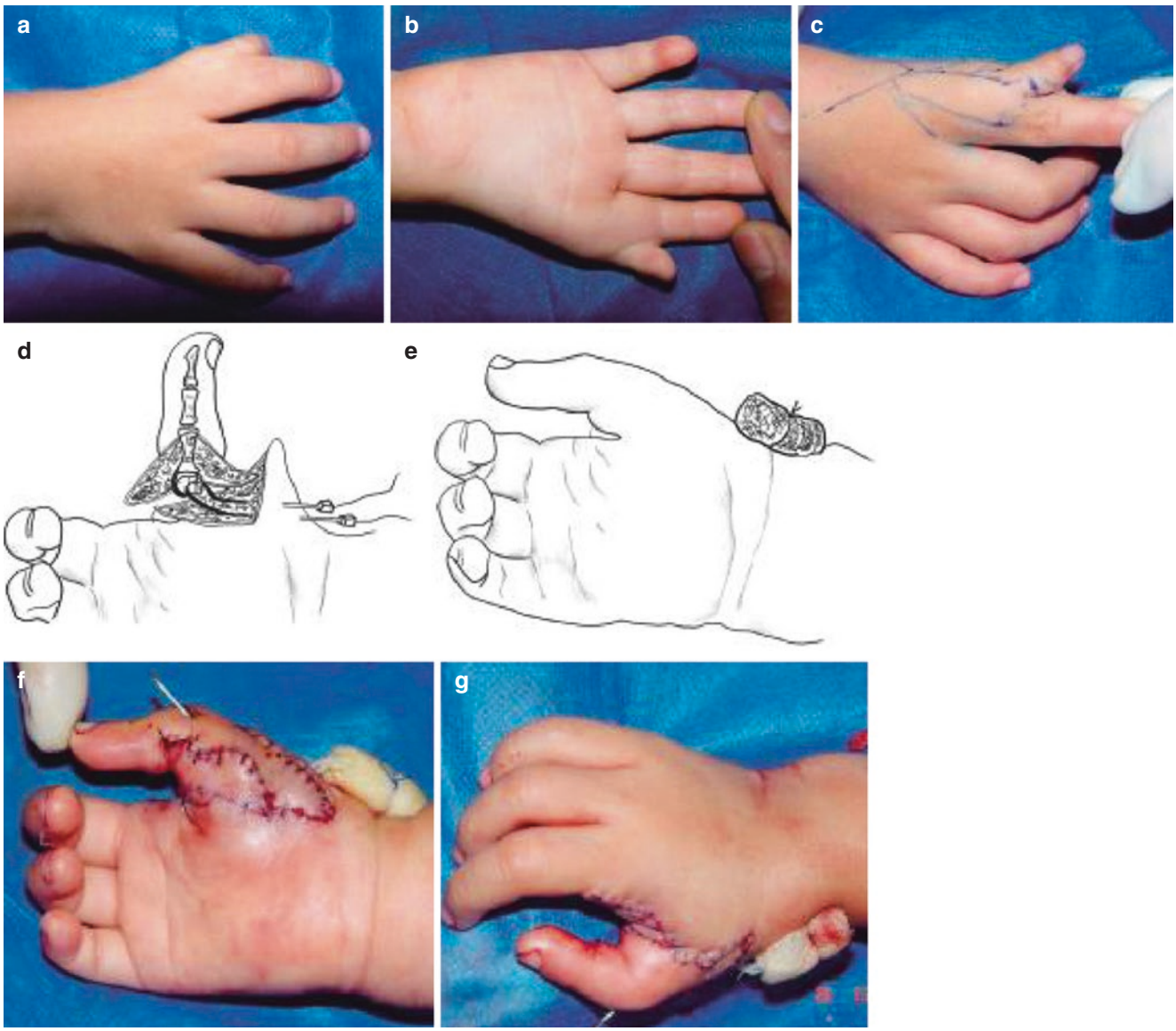


Fig. 5.74 Treatment of atypical type VIa thumb dysplasia. (**a**–**c**) Preoperative design. (**d**, **e**) Surgery demonstration: transpose and shorten the radial second finger for thumb pollicization, and tract the carpometacarpal joint of the reconstructed thumb with a fine steel silk and fix it at the wrist part. (**f**, **g**) After operation, the shape and position of the reconstructed thumb are good

5.5.3.8 Treatment of Hand External Muscle Tendons of Thumb and Functional Reconstruction of Hand Internal Muscles

As for thumb reconstruction for the congenital thumb dysplasia, dynamical reconstruction is also important in addition to the above flap preparation, thumb transposition and reconstruction, metacarpal shortening, reconstruction of the first carpometacarpal joint, as well as the island flap prepared by finger resection to enlarge the thumb. The main points in surgery are the following: ① protect the finger flexor tendons from injuries, and they will become shortened naturally after operation; ② divide the finger extensor tendons into three parts at the proximal end of the proximal interphalangeal joint to make anastomosed with the thumb adductor, thumb short adductor, and thumb long extensor tendons; and ③ as for the thumb long abductors, take the inherent extensors of the index finger for reconstruction. Reconstruction of hand dynamical functions by using this method can produce relatively good surgical effects (Fig. 5.75).

Typical case: A pediatric patient, with hyperphalangeal (six-fingered) thumb dysplasia in both hands, type VIb thumb dysplasia in the right hand, and thumb absence. During the first admission, thumb reconstruction of the left hand was completed; during the second admission, the thumb reconstruction of the right hand was completed (Fig. 5.76).

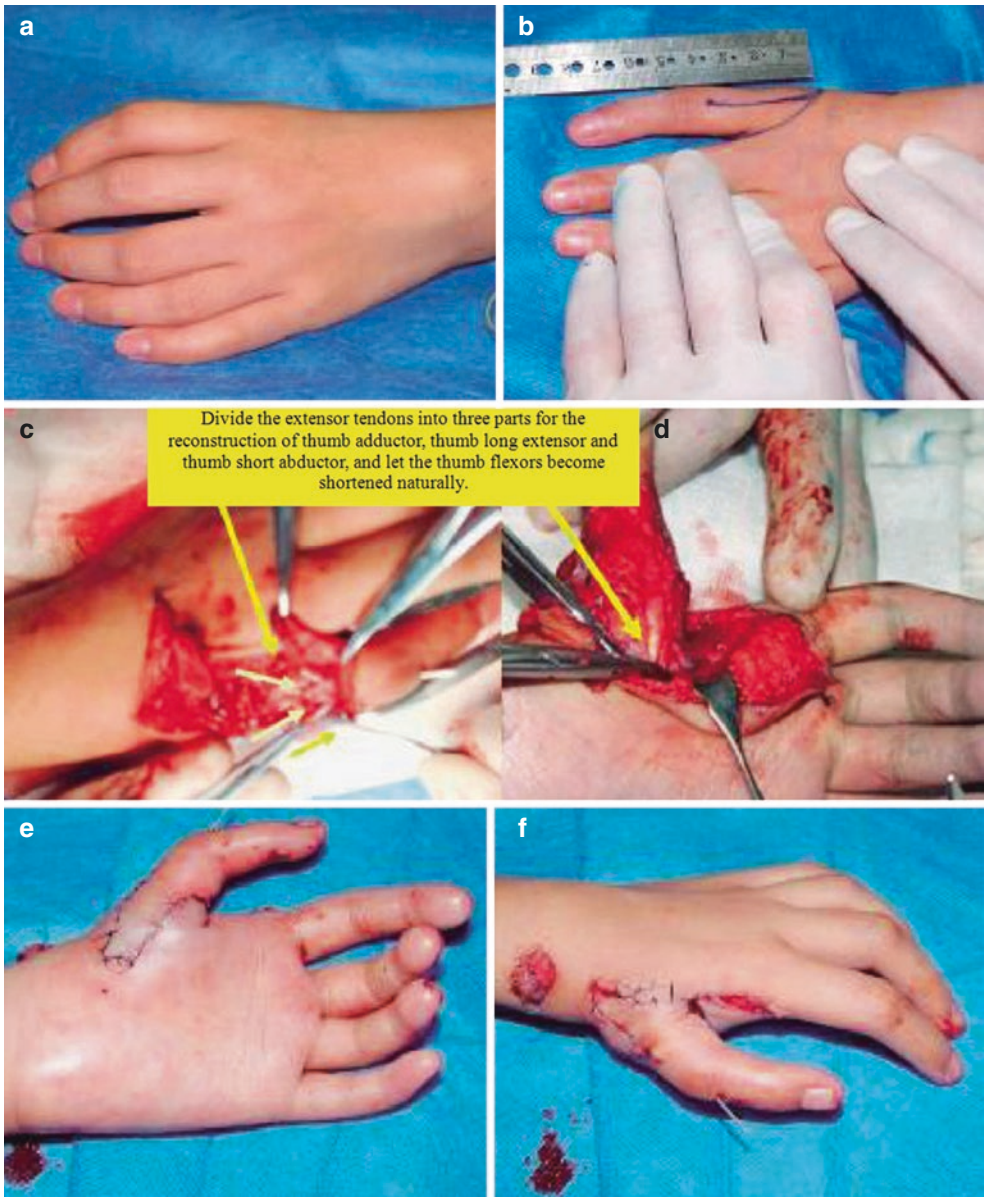


Fig. 5.75 Treatment of hand external muscle tendons of thumb and functional reconstruction of hand internal muscles. (**a**, **b**) Preoperative design. (**c**, **d**) Surgery demonstration. (**e**, **f**) Postoperative hand appearance

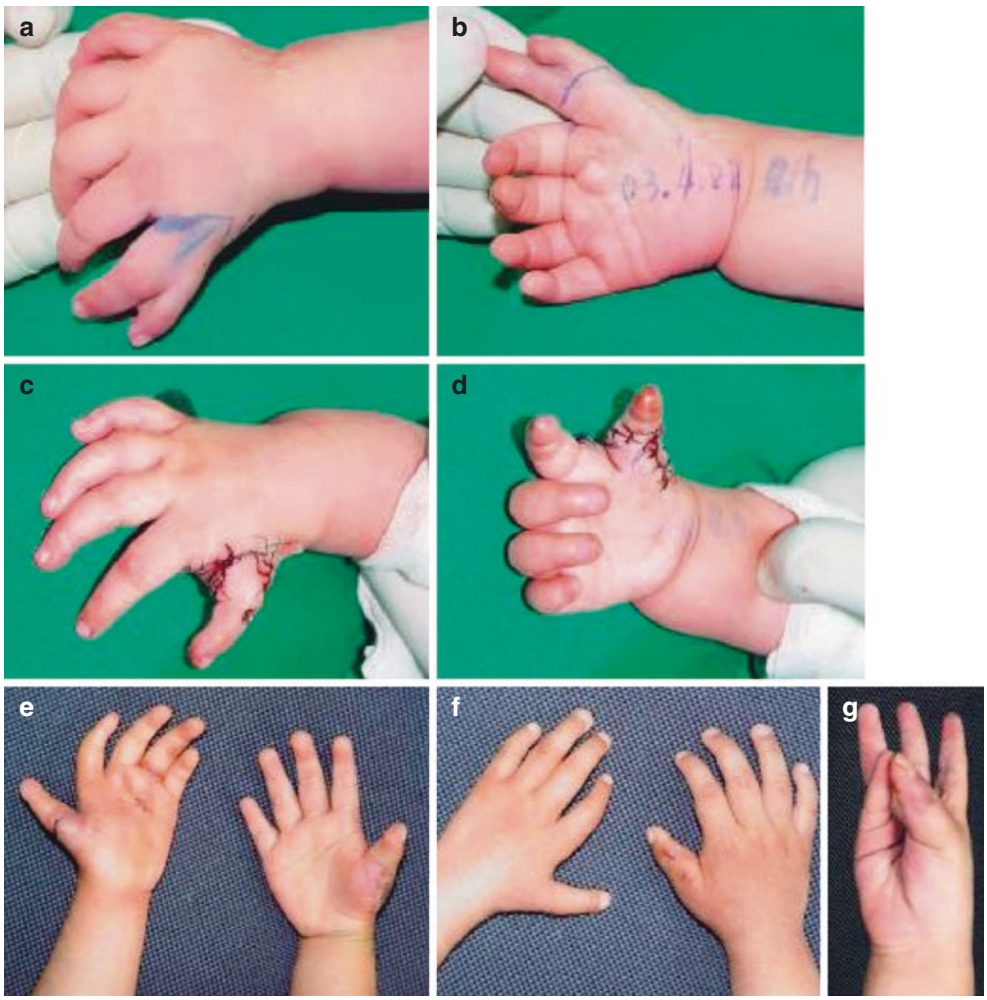


Fig. 5.76 Treatment of type VIb thumb dysplasia in the right hand. (**a**, **b**) Before thumb reconstruction. (**c**–**g**) After the surgery, the reconstructed thumb has complete palm-opposition and adduction functions

5.5.3.9 Thumb Reconstruction for Infants 6–12 Months Old with Congenital Thumb Dysplasia

As for type IV to VI congenital thumb dysplasia, the author usually performs thumb reconstruction on the pediatric patients before they become older than 6 months because early surgical operation is easier. But minimally invasive microsurgery techniques must be adopted for tissue anatomy, grafting, repair, and reconstruction. In order to reduce postoperative need of stitch removal, the author chooses to use the sutures absorbable within a short period of time to suture the skin.

Typical case: A 1-year-old boy with type VIb thumb dysplasia in the right hand, six-fingered thumb absence, and radial hyperphalangeal thumb dysplasia. Due to the shape and size, the index finger is suitable to serve as the donor site of reconstructed thumb. As the radial second finger develops well, it is just sufficient to perform index finger transposition for thumb reconstruction as well as remove hyperdactylia; it is unnecessary to adopt the skin and tissues of the extra fingers at the radial side to enlarge the reconstructed thumb (Fig. 5.77).

5.5.3.10 Treatment of Atypical Type VIb (Six-Fingered) Thumb Dysplasia

Atypical type VIb thumb dysplasia is six-fingered thumb dysplasia and the syndactylia of the first and second fingers. Its treatment is similar to type VIa five-fingered thumb dysplasia.

In the patients with atypical type VIb (six-fingered) thumb dysplasia, the possibility of the radial first finger is very low, and only embryo-bud like neoplasm can be observed. The surgical regimen of thumb reconstruction of this deformity is stated as before: resect the neoplastic dysplastic thumb

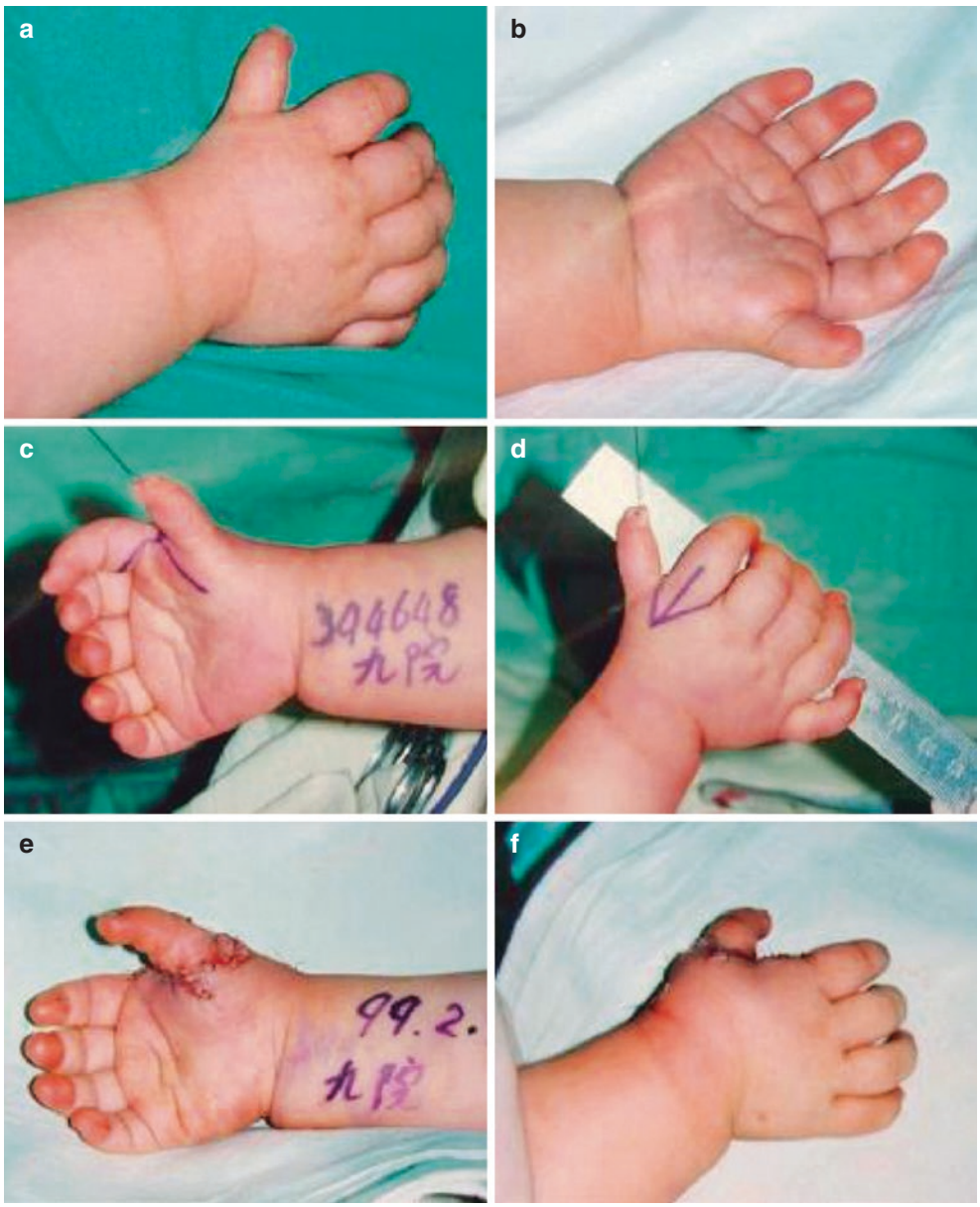


Fig. 5.77 Treatment of type VIb thumb dysplasia in the right hand. (**a**, **b**) Preoperative hand appearance. (**c**, **d**) Surgery design. (**e**, **f**) Postoperative hand appearance

residual end, and utilize the transposition of the radial second finger to design several skin flaps and fabricate one thumb in the palm-opposition position. The typical cases are as follows:

1. Case one: A boy at the age of two and a half, with atypical type VIb thumb dysplasia in the right hand, six-fingered thumb absence, syndactylia of the first and second fingers, thin radial first finger, and serious dysplasia such as floating thumb and type VIa thumb dysplasia in the left hand and five-fingered thumb absence. The surgery is performed in two times: during the first time, shorten and transpose the radial first finger of the left hand, and perform dynamical reconstruction of the first metacarpal bone and carpometacarpal joint; perform the second surgery at 3 months after the operation, namely, thumb reconstruction of type VIb thumb dysplasia of the right hand (Fig. 5.78).
2. Case two: A boy, with atypical type VIb thumb dysplasia in the right hand, six-fingered thumb absence, brachydactylia in the radial first finger and type VIa thumb dysplasia in the left hand. His father and paternal grandmother suffer from type VIa thumb dysplasia in both hands. His paternal great grandparents have six children (four daughters and two sons), and only his paternal grandmother suffers from thumb dysplasia in

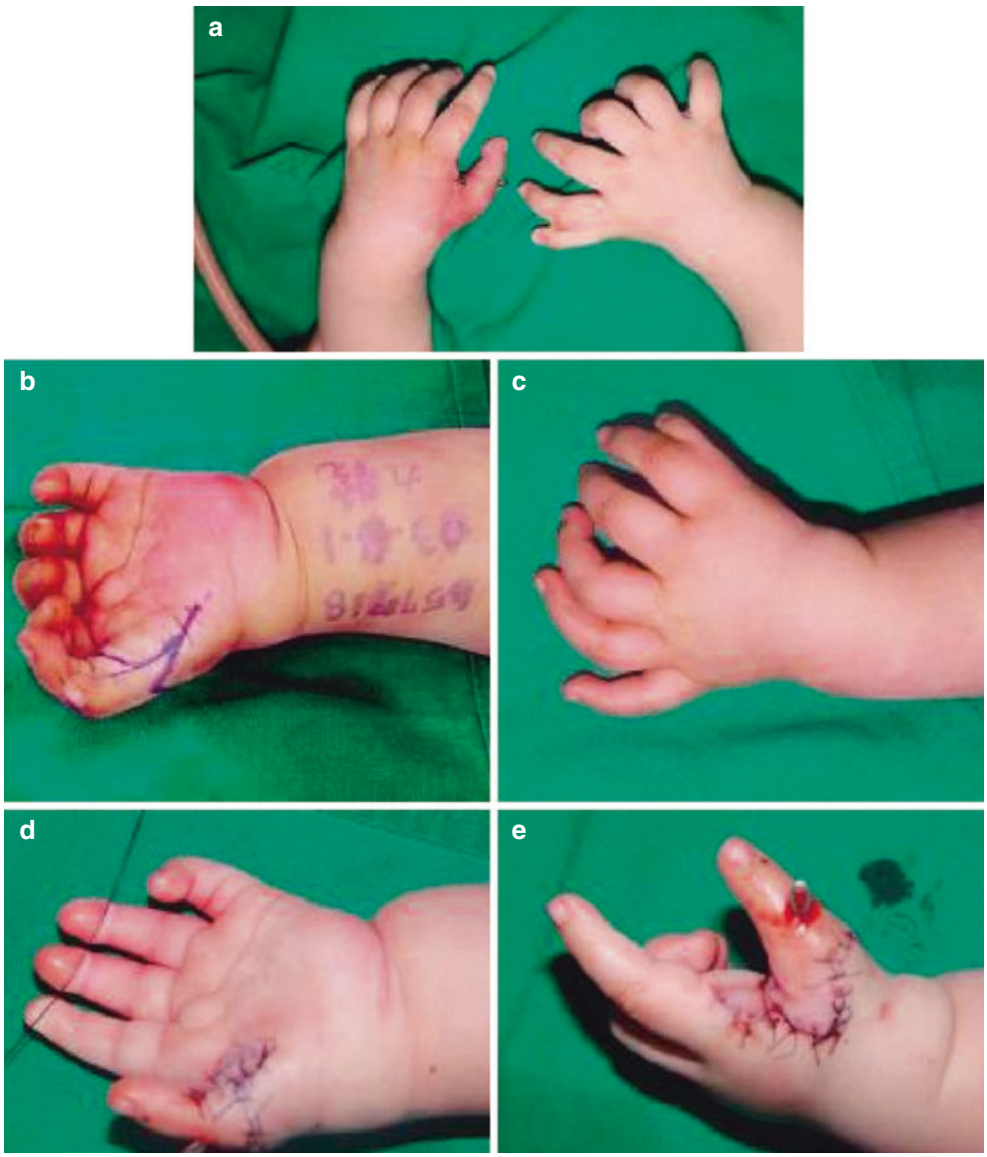


Fig. 5.78 Case one. (**a**) At 3 months after the first surgery of the left hand. (**b**, **c**) Incision design of the second surgery in the left hand. (**d**, **e**) After thumb reconstruction

both hands. His father has 12 cousins (seven males including and five females) who are all normal. Among the 11 cousins of the infantile patient, no one suffers from similar hand deformities.

During the operation, the thumb extensor tendons of the reconstructed thumb are shortened, and the insertions of the hand internal muscles are fixed. In the reconstruction of the first carpometacarpal joint, the author adopts the method of pulling the first carpometacarpal joint of the wrist and fixing the Kirschner silk, and sometimes use of the Kirschner silk can also produce good effects. This case is just a good example (Fig. 5.79).

5.5.3.11 Treatment of Type VII Thumb Dysplasia (Web-Shaped Hand Deformity)

The treatment of type VII thumb dysplasia (web-shaped hand deformity) is as follows: ① resection of radial hyperdactylia, separation of syndactylia, and reconstruction of thumb; ② separation of syndactylous other fingers; ③ as the nourishing blood vessels of the fingers may suffer deformities, it is recommended that separation of the adjacent fingerwebs be performed in different times unless the vessels are proven to develop well; ④ release of flexed and contracted fingers and dynamical reconstruction of finger extensor tendons; and ⑤ dynamical reconstruction of hand internal muscles.

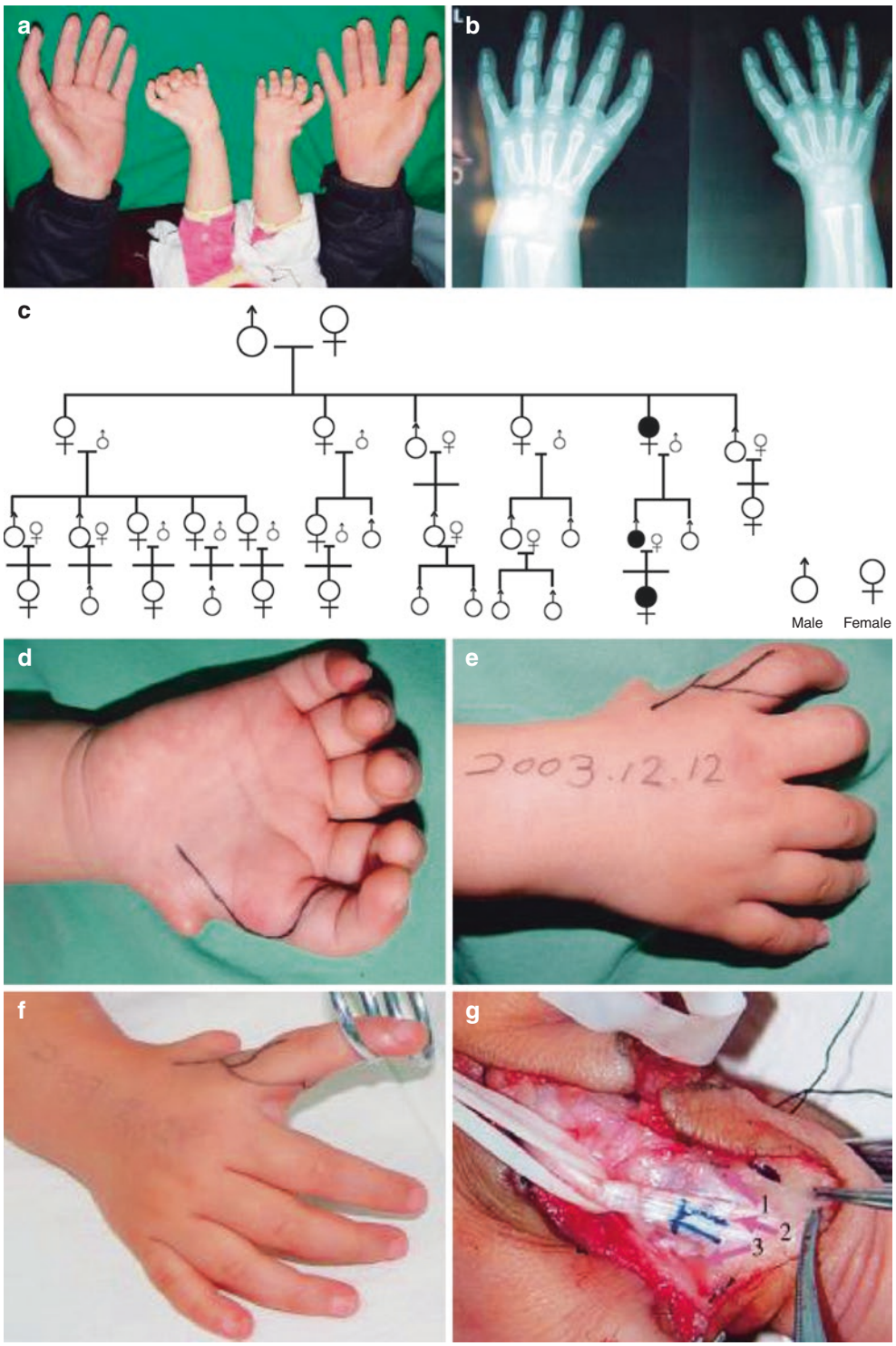


Fig. 5.79 Case two. (**a**) A boy, with type VIb thumb dysplasia in the right hand and type VIa thumb dysplasia in the left hand; his father suffers from type VIa thumb dysplasia in both hands. (**b**) X-ray findings of the pediatric patient. (**c**) Genetic genealogy of the pediatric patient. (**d–f**) Flap design of thumb reconstruction. (**g**) Dynamical reconstruction of thumb extensor. (**h–j**) The Kirschner silk is adopted to fix the carpometacarpal joint

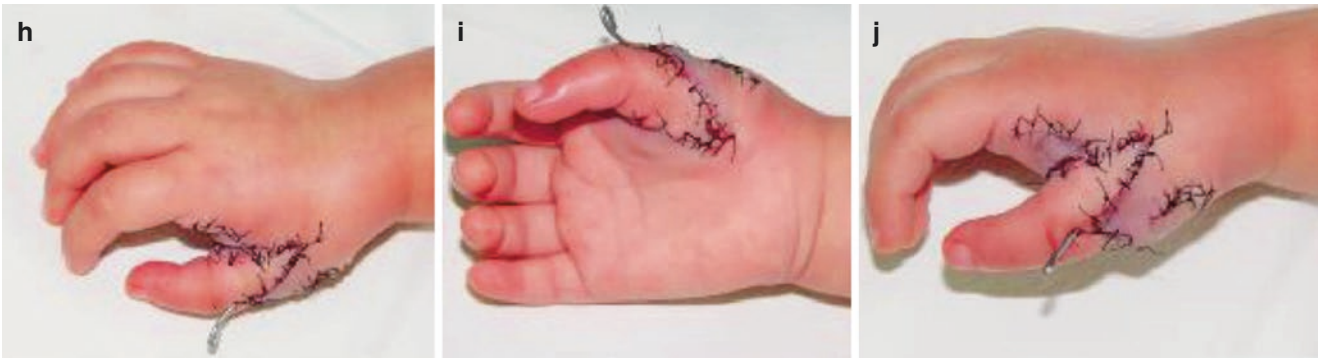


Fig. 5.79 (continued)

The web-shaped hand deformity can be classified into two types: ① type VIIa synpolydactyly-type thumb dysplasia, namely, deformity of general flexion synpolydactyly, and ② type VIIb synpolydactyly-type thumb dysplasia, namely, deformity of serious flexion synpolydactyly. Most of the patients of the second type need extensor dynamical repair. The typical cases are as follows:

1. Case one. A 4-year-old male infantile patient, with congenital thumb dysplasia in both hands, seven-fingered deformity in the left hand and six-fingered deformity in the right hand; thumb absence; partial separation of the third and fourth fingers of the left hand; syndactylia in the remaining fingers; thumb web absence in both hands; synonychia of multiple nails; abnormal size, position, and shape; deformity of flexed fingers; dysplasia of digital extensor; dysplasia of thenar and hypothenar muscles; no obvious disorder in finger sensation; no functional disorder detected in wrist joint; and concomitantly deformity of polydactyly and syndactyly in both feet. After admission, reconstruct the left thumb, separate the syndactylia of the right hand, and repair the reconstructed thumb (Fig. 5.80).
2. Case two. A 2-year-old boy, with thumb dysplasia in bilateral hands and deformity of synpolydactyly. Five-fingered hand in the right hand, thumb absence, incomplete syndactylia, namely, syndactylia of the first, second, and third fingers, and syndactylia of the fourth and fifth fingers. The patient was admitted to hospital at the age of one and a half and underwent reconstruction of thumb absence of the right hand, during which radial first finger was transposed for thumb reconstruction; the surgical methods were similar to thumb reconstruction of hyperphalangeal thumb dysplasia, including separating correction of syndactylia of the first and second fingers. In case of thumb reconstruction and thumb web reconstruction during the separation due to the syndactylia of the first and second fingers, it is necessary to perform skin grafting or local flap grafting for local skin defects. The interval between the second surgery and the first surgery was half a year. During the second surgery, the left thumb was reconstructed, the thumb web was reconstructed, the syndactylia of the fourth and fifth fingers was separated, the right thumb was corrected, and the syndactylia was corrected (Fig. 5.81).

5.5.3.12 Treatment of Type VIII Thumb Dysplasia

Type VIII dysplastic type thumb dysplasia of the entire hand is manifested as dysplasia in the thumb, fingers, and palm of the entire hand, syndactylia and brachydactylia, thumb absence, and spade hand. It is common in thumb dysplasia of the Apert syndrome.

1. Surgical methods. The osteotomy and transposition of the metacarpal bone of the radial first finger were adopted for pollicization, retrograde flap grafting was performed on the forearm, and the thumb web of the first fingerweb was reconstructed.

   As this type of thumb dysplasia is often concomitantly accompanied by dysplasia in other fingers, thumb web stenosis and syndactylia, there are some differences between radial finger transposition for pollicization and index finger pollicization. The main manifestation is that reconstruction of the first fingerweb is difficult because locally this

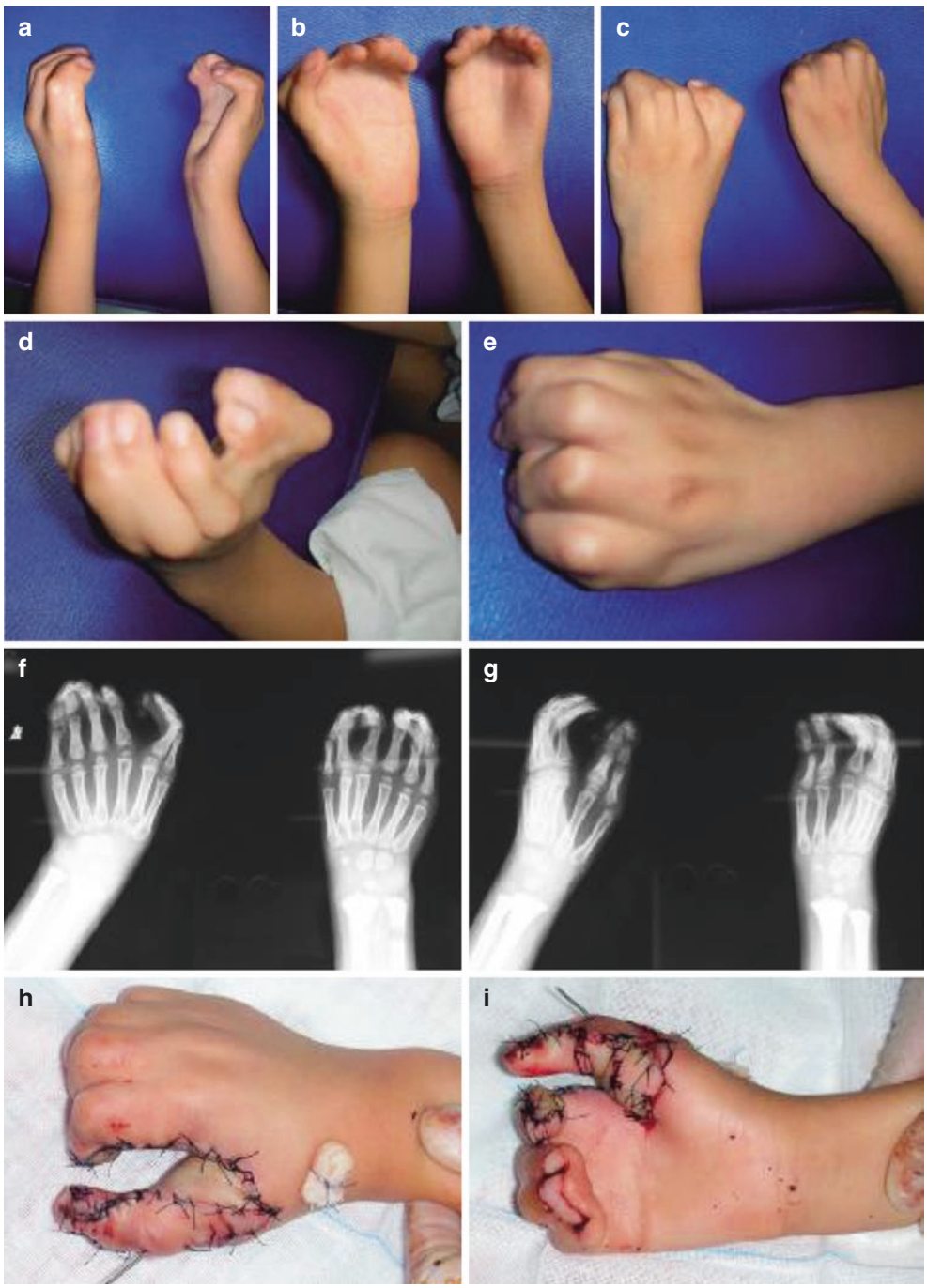


Fig. 5.80 Case one. (**a**–**e**). Preoperative hand appearance. (**f**, **g**) Preoperative X-ray findings. (**h**, **i**) Postoperative hand appearance

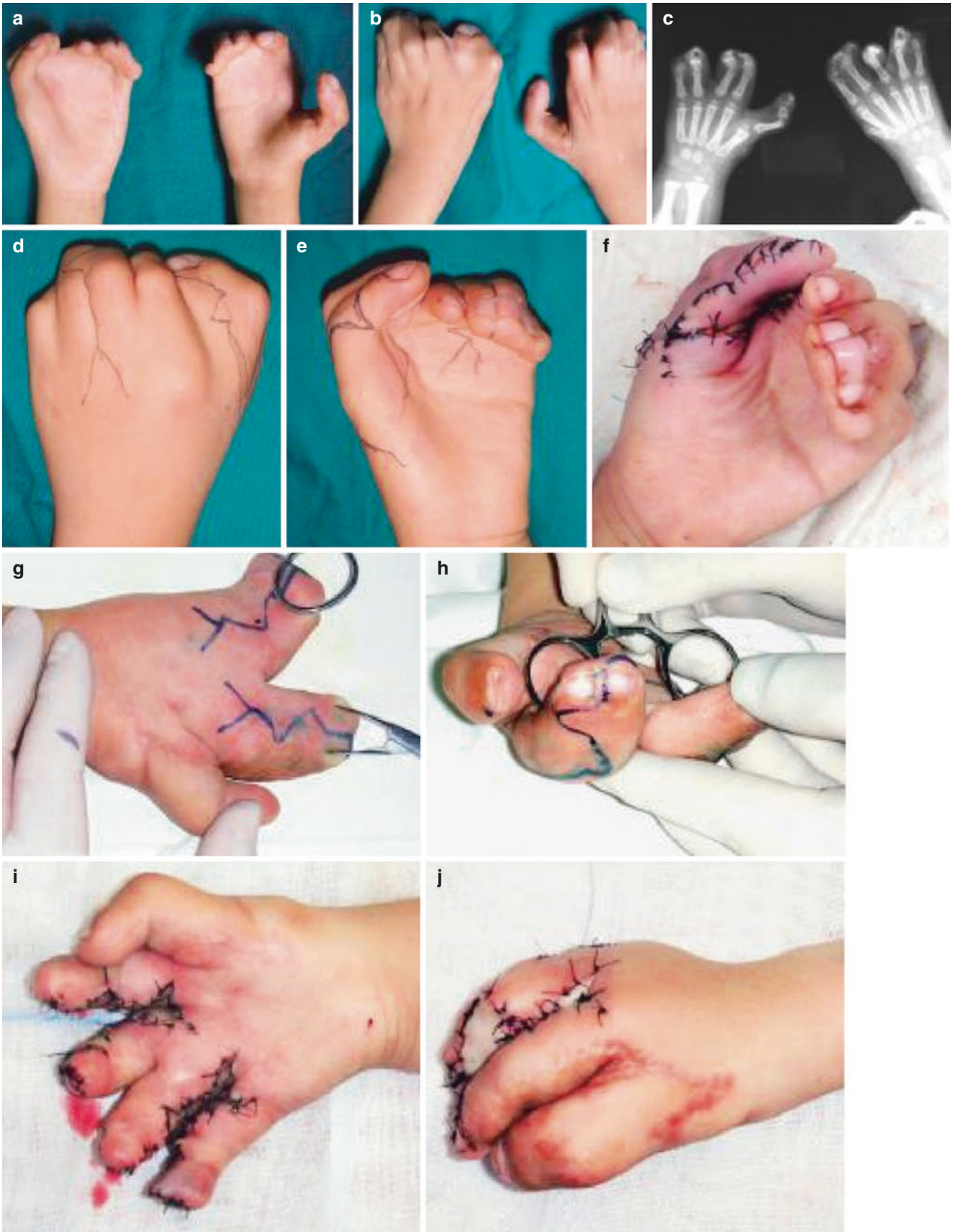


Fig. 5.81 Case two. (**a**–**c**) Before the first surgery. (**d**, **e**) Design of the first surgery. (**f**) Immediately after the first surgery. (**g**, **h**) Design of the second surgery. (**i**, **j**) Immediately after the second surgery

deformed hand does not have sufficient skin and subcutaneous tissues as the donor site of the tissues for the reconstruction of the first fingerweb, and the free grafting of skin flap is apt to induce local contracture and the efficacy is poor, so the flap grafting at the distal site must be considered. In addition, this type of thumb dysplasia is often accompanied by short and small finger; therefore, the length of the original fingers can be retained when radial finger transposition for pollicization is performed, and only osteotomy and transposition of the first metacarpal bone is performed for thumb web reconstruction.

2. Typical case
   (a) Case one: A 5-year-old girl, with the Apert syndromic type VIII thumb dysplasia, spade hand, three-fingered thumb absence, and finger dysplasia, growth of fingers on the same plane, syndactylia of the second and third fingers, and concomitantly with craniofacial deformity.

   On Nov. 26, 1984, the patient received surgery. Surgical design: transposition of radial fingers for pollicization, osteotomy of the first metacarpal bone and rotation of it to the palm-opposition position, and grafting of retrograde interosseous dorsal flaps for thumb web reconstruction. This may be the case with the earliest clinical application of grafting of retrograde interosseous dorsal flaps. Before the operation, Doppler examination is used to examine the reliable pathway of hand dorsal and wrist dorsal arteries. Arterial passage is palpable in the middle of the wrist dorsum of the affected hand. With this site as the pedicle, retrograde interosseous dorsal flaps are fabricated at the dorsal side of the forearm, and grafting is performed with fascia as the pedicle. The surgery is performed under the condition of general anesthesia. First, incise and deepen the first fingerweb, then perform osteotomy in the middle of the first metacarpal bone, rotate it to the palm-opposition position and fix it, and graft the retrograde interosseous dorsal flaps for thumb web reconstruction. In addition to the retrograde interosseous dorsal flaps, retrograde flaps at the radial side of forearm, retrograde flaps at the ulnar side of the forearm, groin-free skin flaps, free flaps at the median or lateral side of upper arm, free flaps at the foot dorsum, etc. can be grafted for thumb web reconstruction (Fig. 5.82).

   (b) Case two: A 3-year-old female patient, with type VIII entire-hand-type thumb dysplasia, spade hand, four-fingered thumb absence, and finger dysplasia and syndactylia of the first and second fingers as well as the third and fourth fingers.

   The surgical design includes the following: ① separate syndactylia of the first and second fingers; ② perform osteotomy of the first metacarpal bone, rotate it to the palm-opposition position and fix it, and reconstruct the thumb; ③ graft the forearm retrograde

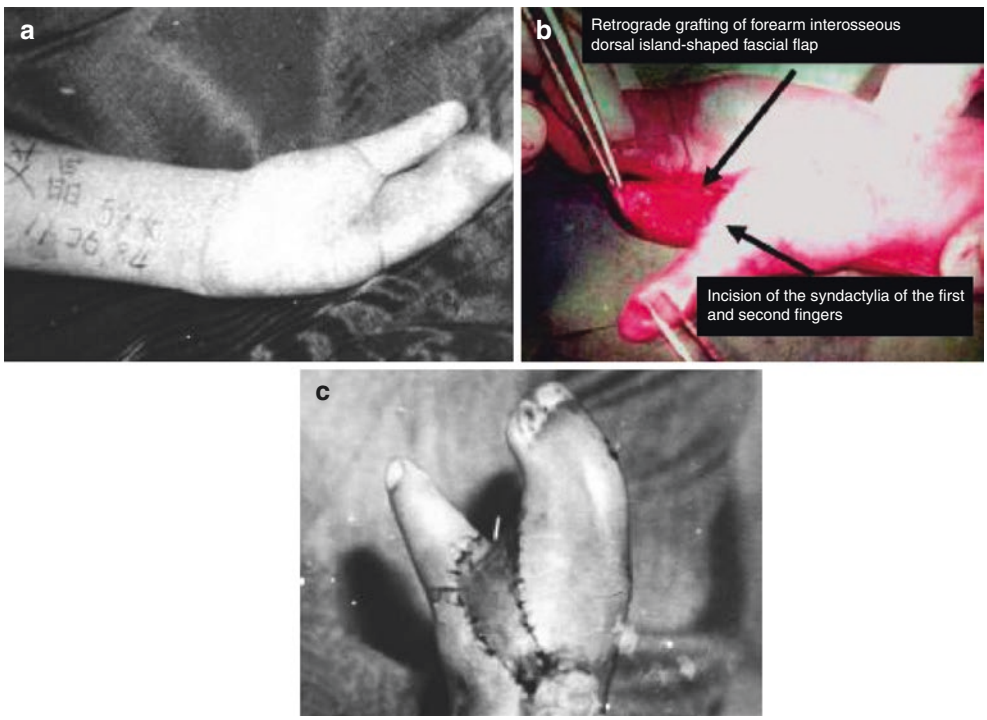


Fig. 5.82 Case one. (**a**) Preoperative, the Apert syndrome, spade hand. (**b**) Intraoperative. (**c**) Postoperative

fascial flap, and reconstruct the first fingerweb, namely, thumb web; and ④ separate syndactylia of the third and fourth fingers (Fig. 5.83).

(c) Case three: During the treatment of brachydactylia- and syndactylia-type thumb dysplasia, the author was willing to perform the separation of the first and second fingers at the radial side during the first surgery, osteotomy was performed on the first metacarpal bone, then it was rotated to palm-opposition position and fixed, and the forearm retrograde flaps were grafted for thumb web reconstruction. To prevent poor blood supply at the distal ends of fingers during the separation of syndactylia, the conventional practice is to correct it in different surgeries (Fig. 5.84).

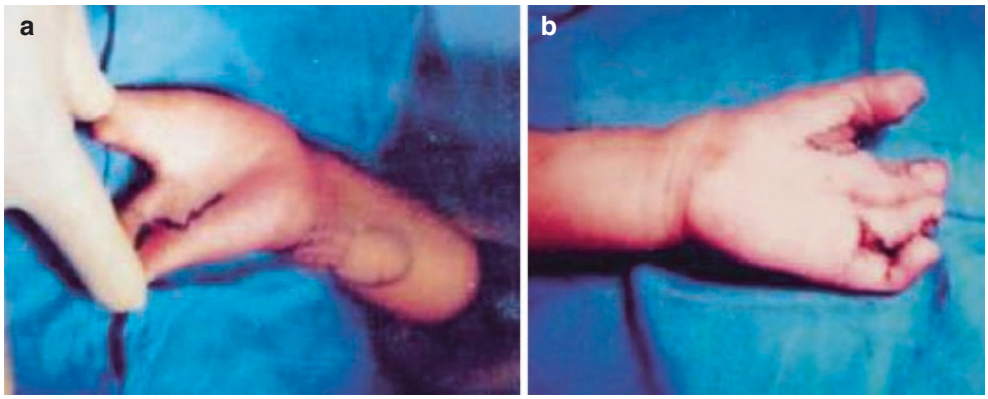


Fig. 5.83 Case two. (**a**) Before operation. (**b**) After operation

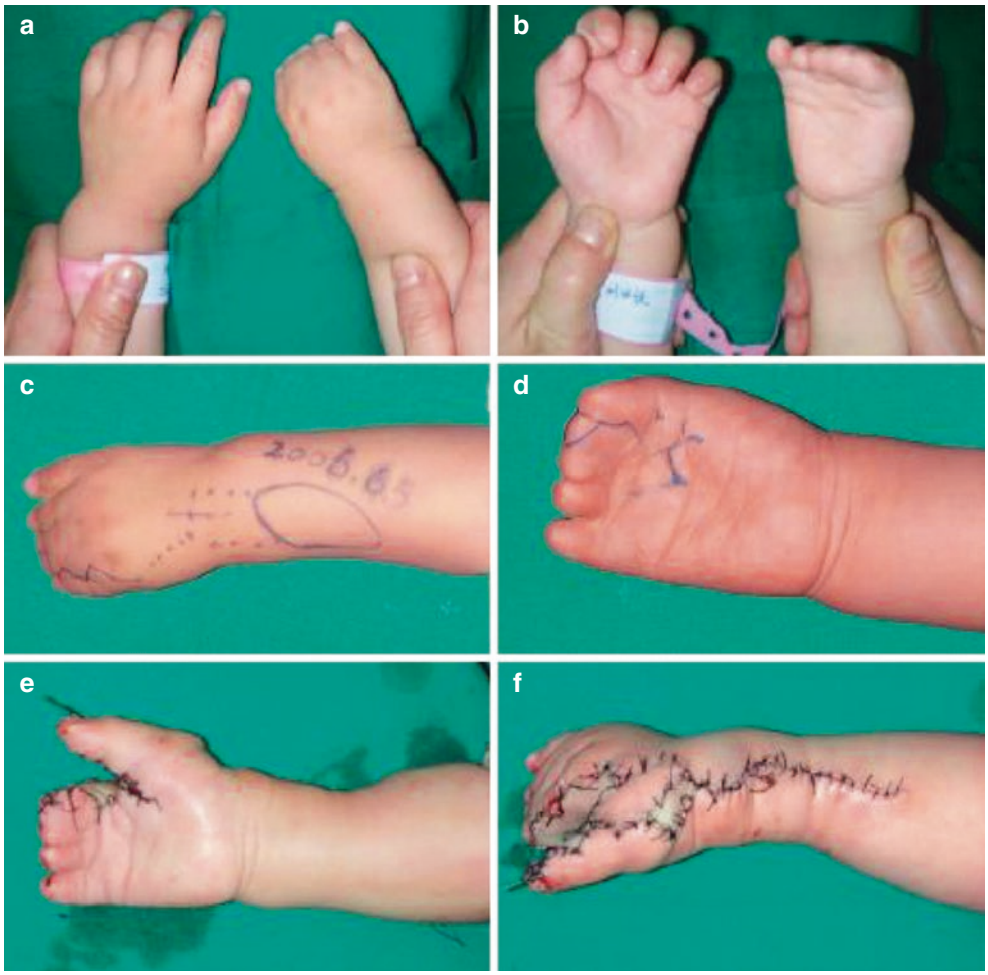


Fig. 5.84 Case three. (**a**, **b**) Before operation. (**c**, **d**) Surgery design. (**e**, **f**) After operation

5.5.3.13 Treatment of Type IX Thumb Dysplasia (Deformity of Cleft Hand) and Application of Toe-to-Hand Free Grafting to the Thumb Dysplasia

Microsurgical grafting of the second toe, grafting of the metatarsophalangeal joint, and grafting of partial toes or expansion of the second toe for transplantation can be regarded as the good choices for the functional reconstruction of type IIIa, IIIb, and IIIc serious thumb dysplasia as well as the good options for the functional reconstruction of metatarsophalangeal joint and carpometacarpal joint. Toe-to-hand grafting or other methods for thumb reconstruction can be used for the treatment of thumb dysplasia, but as for the patients capable of receiving the finger transposition for thumb reconstruction, the author never utilizes toe-to-hand grafting, metatarsophalangeal joint grafting, or partial toe-to-hand grafting. Although the thumb reconstructed after toe-to-hand free grafting can grow with the increase in age, the thumb reconstructed after toe-to-hand grafting is not as good as the reconstructed thumb after finger transposition both in terms of shape and function, because in most cases, toe-to-hand grafting is not the preferred method for thumb reconstruction due to congenital thumb defects, and it can only serve as a supplementary method for the congenital thumb dysplasia for thumb reconstruction.

As for type IX thumb dysplasia, namely, cleft hand deformity, there is no proper finger donor site for the treatment of complete thumb absence, especially the one-fingered cleft hand, two-fingered cleft hand, and three-fingered cleft hand. So sometimes the surgeons have to choose free toe-to-hand grafting for thumb reconstruction.

As for the surgical methods, the grafting of the second phalange of foot for expansion for thumb reconstruction or the partial toe-to-hand grafting for thumb reconstruction can be selected. The palm-opposing fingers can be reconstructed; after one finger was cut, it was transposed to the palm-opposition position, for example, the first finger at the radial side was transposed for pollicization, and then the first finger-web, namely, thumb web, was reconstructed. Although the reconstructed thumb and the hands take the morphologically poor lobster tong shape, the functions are normal; finger pollicization can also be performed. As for one-fingered cleft hand, toe-to-hand grafting can be considered for thumb reconstruction or the reconstruction of palm-opposing fingers, but it should be noted that cleft hand is often accompanied by cleft foot and dorsal pedal arteries and toe arteries also suffer deformities, so examinations must be conducted before the surgery. Only when blood supply in toes is good can toe-to-hand free grafting be adopted. In order to reconstruct hand functions, single finger resection can be performed, and forearm bifurcation is also a regimen that can be considered. In addition, the present artificial limb production is very advanced; the installation of artificial limbs can be a good choice due to such advantages: (1) good shape and (2) certain functions.

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Limb Dysdifferentiation

6

Wei Wang, Bin Wang, Jinghong Xu, Zhenyu Bian, Jianmin Yao, Xu Gong, Yunfei Zhang, Hongxing Zhang, Shengbo Zhou, Yongkang Jiang, Biwei Zeng, Jialiang Chen, Wangxiang Yao, Longchun Zhang, Liulong Zhu, Yizhen Chen, Feng Ni, Sheng Ding, and Laijin Lu

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6.1 Syndactylia

Wei Wang, Bin Wang, Yunfei Zhang, Hongxing Zhang, and Shengbo Zhou

Syndactylia is the fusion between the adjacent interdigital soft tissues and skeletons to varying extents, which is induced by the failure in a certain stage during the normal finger (toe) separation and formation of fingerweb. During normal development [1], fingers are formed during the mesoderm differentiation inside the terminal hand plates of the upper limbs during the embryonic phase. The formation of finger interdigital space is an apoptosis-regulating process; the direction is from far to near until the normal fingerweb is reached [2]. This process depends on the molecular signal of apical ectodermal ridge and multiple cytokines, including bone morphogenetic protein, transforming growth factor β, fibroblast growth factor, and tretinoin [3–7]. The normal second, third, and fourth fingerwebs have an inclination of 45° from the dorsal side to the volar side, a sand clocklike structure from the head of metacarpal bone to the middle point level of proximal phalange, added to the proximal digital transverse striation (Fig. 6.1). The second and the fourth fingerwebs are wider than the third fingerweb, which makes the index finger and the little finger have a larger degree of abduction. The first fingerweb is a piece of broad rhombic skin, consisting of volar hairless skin and the skin with a large range of motion at the dorsal side which is relatively thin.

6.1.1 Epidemiology

Syndactylia is a common congenital hand deformity with an incidence of about 0.5‰. Fifty percent of them is bilateral syndactylia. 10%~40% of the pediatric patients have a family history with the manifestation of autosomal dominant inheritance (Fig. 6.2). The variable expressivity and the incomplete penetrance can induce a high incidence in males (the male/female incidence ratio is about 2:1), and the phenotypes are various in the same family. As part of the congenital hand deformities in children, syndactylia can independently occur or appear in many syndromes, concomitantly with multiple deformities, such as polydactylism, finger flexion deformity,

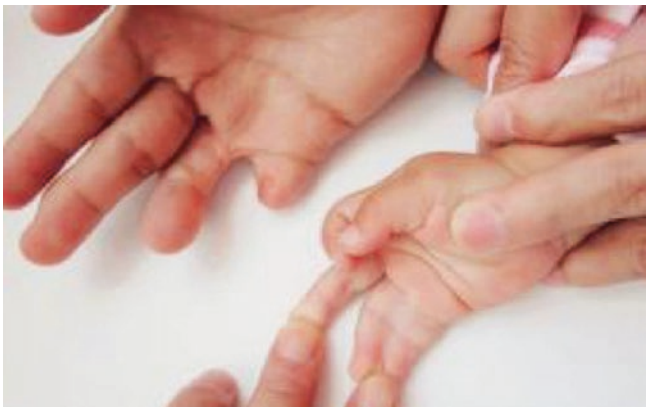


Fig. 6.2 The father and his daughter suffer the deformity of syndactylia of the right ring and little fingers (the father once underwent surgery)

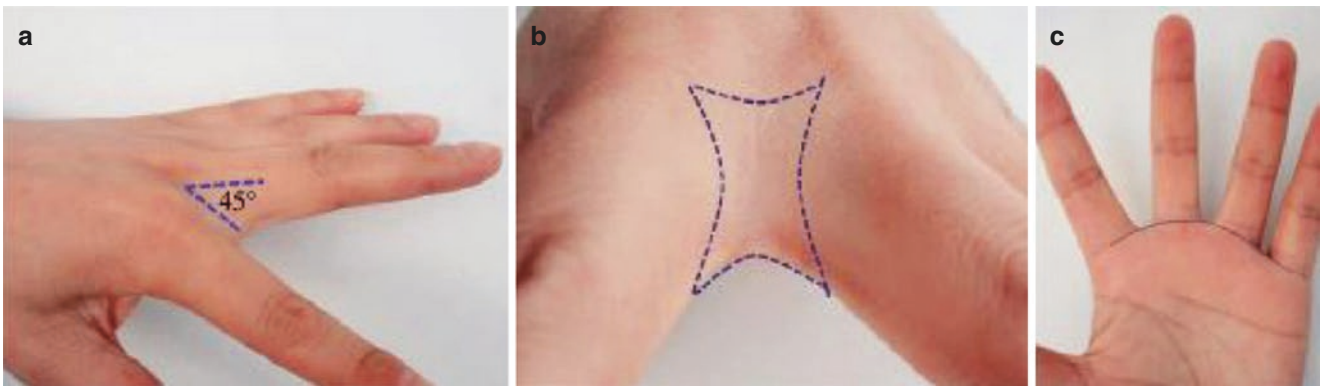


Fig. 6.1 The normal second, third, and fourth fingerwebs have an inclination of 45° from the dorsal side to the volar side, a sand clocklike structure from the head of metacarpal bone to the middle point level of proximal phalange

brachydactylia, congenital interphalangeal joint fusion, and synostosis. Among the patients with syndactylia that occurs independently, the syndactylia of the middle and ring fingers is the commonest (57%), followed by the syndactylia of the ring and little fingers (27%), and the syndactylia of the thumb and index finger as well as index and middle fingers is relatively rare. Among the patients with syndromes, the syndactylia of the thumb and index finger as well as index and middle fingers is relatively common [2, 4].

6.1.2 Pathological Classification

The fingers that are connected together can present deformities in fingernails, digital nerve vascular bundles, skeletons, and tendons. The outer layer of the syndactylous skin is insufficient to cover the corresponding margins after finger separation, and the subcutaneous abnormal fascia consists of continuous and thickened structures that run through the syndactylia. Complete syndactylia refers to complete connection between adjacent fingers from the background to the tip; incomplete syndactylia refers to the partial connection between adjacent fingers, and the fingerweb takes shape in any position between the normal place and fingertips (Fig. 6.3). Simple syndactylia only has the connection of skin or soft tissues of adjacent fingers, but the joints are normal in most cases and the finger flexion tensor tendons can move independently. Although the finger structural bifurcations may be closer to the terminal when compared with the normal level, the anatomic structures of the finger nerve

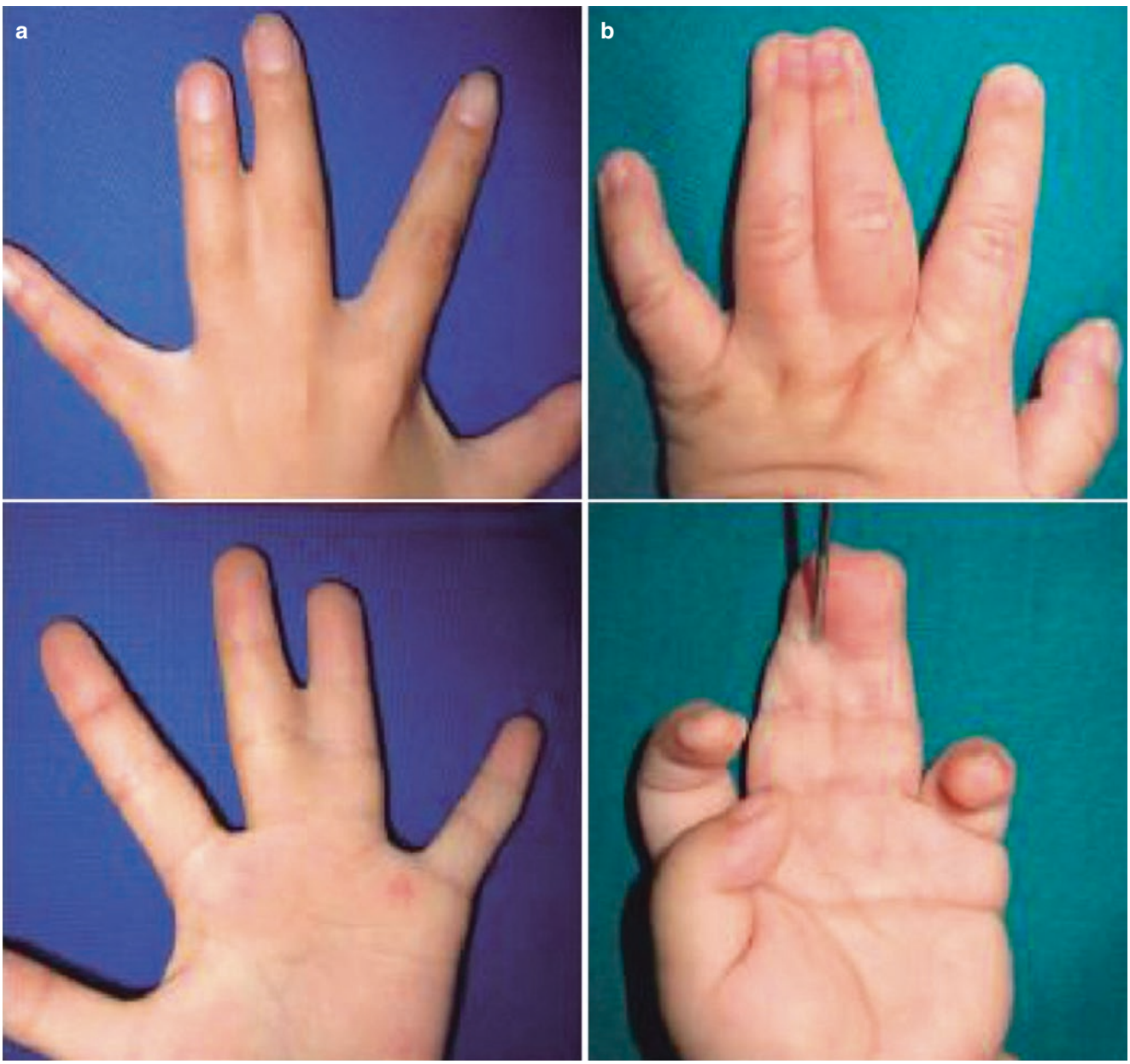


Fig. 6.3 Deformity of syndactylia of middle and ring fingers. (**a**) Incomplete. (**b**) Complete

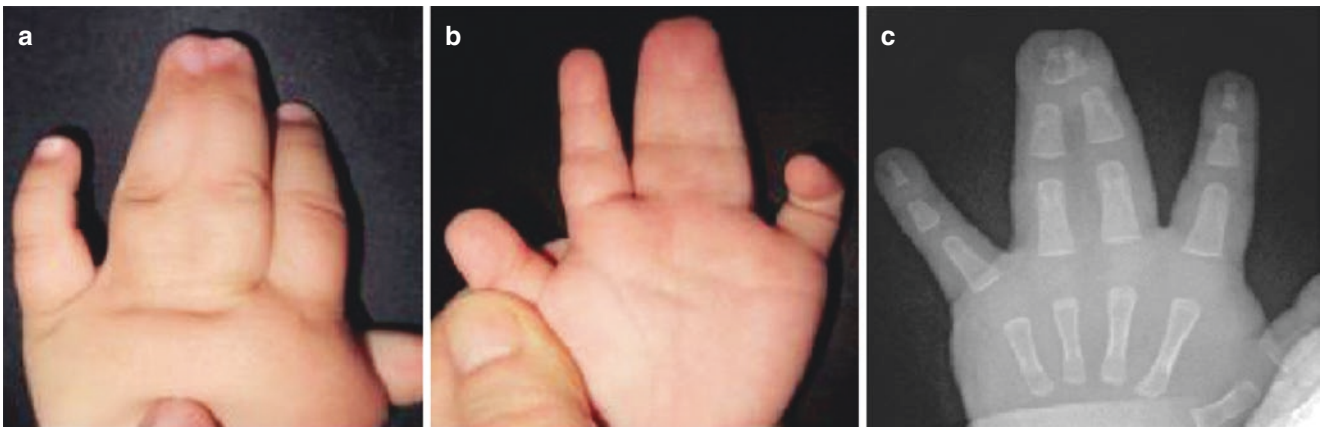


Fig. 6.4 Combined syndactylia: lateral fusion between distal phalanges. (**a**, **b**) Hand appearance. (**c**) X-ray findings

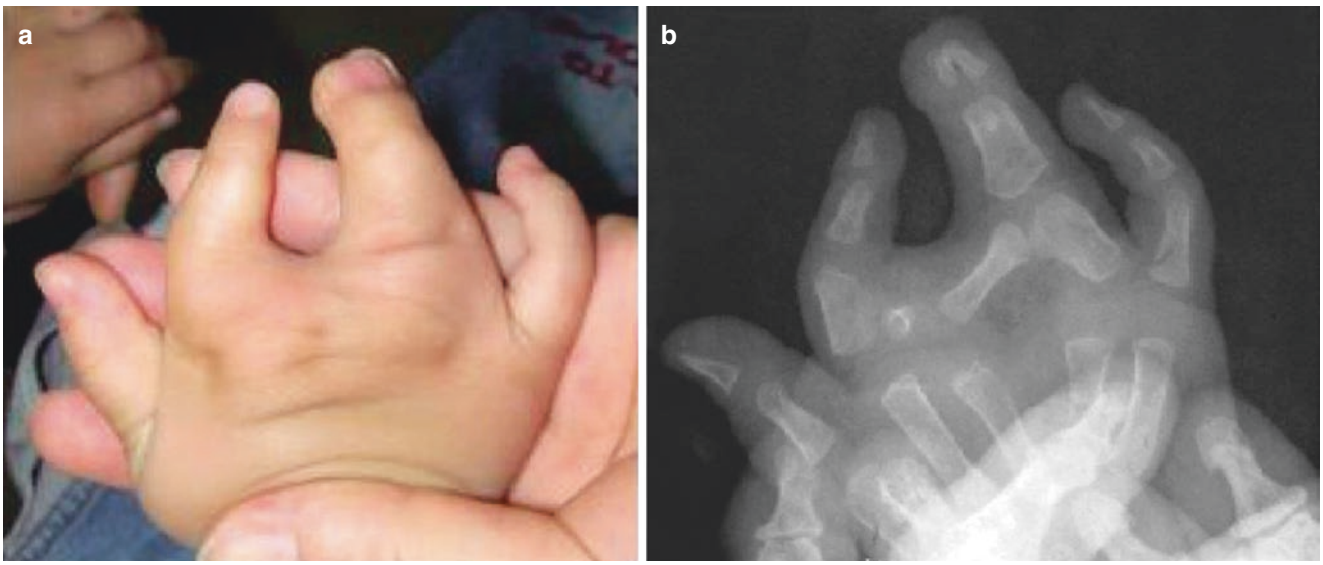


Fig. 6.5 Combined syndactylia: the phalange or the finger is inserted between the abnormal fingerwebs. (**a**) Hand appearance. (**b**) X-ray findings

vessels are normal. Combined syndactylia is characterized by skeletal abnormalities. The commonest abnormality is the lateral fusion between distal phalanges, the distal synostosis is often manifested as synonychia, and the concomitant symptoms are nail folds decrease and the onychostromas of two fingers that cross bone blocks become flat (Fig. 6.4). Combined syndactylia means that the phalange or the finger is inserted between the abnormal fingerwebs, and the incidence of tendon and nerve vascular deformities is positively correlated with the complexity of syndactylia (Fig. 6.5) [1].

6.1.3 Classification and Rating of the Damage Degree of Deformities

All congenital deformities of hands and upper limbs have damages in appearance and functions to varying extents, and how to measure the damage degree of deformities is a common concern of doctors in plastic surgery and hand surgery. The classification of Eaton and Lister (1990) on the damage degree of congenital syndactylia is a valuable attempt [8].

The classification of damage degree of deformities includes three parts, namely, grading of fingerweb adhesion, grading of bone structure deformities and range of motion, and grading of morphological damages. The author believes that the development of an ideal method to grade the damage degree of syndactylia should be based on the hand function assessing methods. The range of total active motion (TPM) and the range of total passive motion of each site of hands should be measured together with the defect degree. But it is objectively not easy for a 1–2-year-old pediatric patient to get access to these data. Therefore, Eaton and Lister's method to grade congenital syndactylia is an easy and feasible method in current clinical practice because it can not only facilitate the selection of surgical methods but also be used as the basis for assessment of surgical effects.

6.1.3.1 Grading of Fingerweb Adhesion Degree

Measure the relatively long finger: when the finger is completely extended or abducted, measure the distance between the head of metacarpal bone and the fingertip and calculate the degree of fingerweb adhesion. The criteria are as follows:

1. Degree I. Syndactylia range ≤ 1/8 of the distance between the head of metacarpal bone and the fingertip
2. Degree II. Syndactylia range ≤ 1/8 to 1/4 of the distance between the head of metacarpal bone and the fingertip
3. Degree III. Syndactylia range 1/4 to 3/8 of the distance between the head of metacarpal bone and the fingertip
4. Degree IV. Syndactylia range > 3/8 of the distance between the head of metacarpal bone and the fingertip

6.1.3.2 Grading of Range of Active Abduction

1. Degree I. Thumb-index finger abduction ≥ 60°, finger abduction ≥ 30°
2. Degree II. Thumb-index finger abduction 45°–60°, finger abduction 20°–30°
3. Degree III. Thumb-index finger abduction 30°–45°, finger abduction 10°–20°
4. Degree IV. Thumb-index finger abduction <30°, finger abduction <10°

6.1.3.3 Grading of Degree of Active Digital Extension or Flexion Damages

Measure the finger extension and flexion insufficiency by number of centimeter; as for the thumb, measure the abduction functional loss by number of centimeter. The criteria are as follows:

1. Degree I. The finger extension or flexion range decreases by <0.5 cm.
2. Degree II. The finger extension or flexion range decreases by 0.5–1 cm.
3. Degree III. The finger extension or flexion range decreases by 1–2 cm.
4. Degree IV. The finger extension or flexion range decreases by >2 cm.

6.1.3.4 Grading of Morphological Damages

1. Degree I. Normal appearance
2. Degree II. Close to normal appearance
3. Degree III. Obvious deformity
4. Degree IV. Serious deformity or no change in preoperative and postoperative morphology

6.1.4 Syndactylia Accompanied by Syndrome

Syndactylia accompanied by various syndromes has been described in the deformity of combined syndactylia.

Syndactylia can not only appear independently but also is one of the symptoms of other deformities. Among the deformities induced by multiple kinds of hand dysplasia, syndactylia is one of the important manifestations. In the deformity of the cleft hand, syndactylia is very common, and there are also hyperdactylia and syndactylia, brachydactylia and syndactylia, fingertip crossing and syndactylia, limb ring syndrome accompanied by syndactylia, and spade hand with syndactylia [9]. Among the multiple syndromes, syndactylia is one of the symptoms, such as Apert syndrome and Poland syndrome (Table 6.1). As is recorded by literature, the clinical manifestations of 48 syndromes include syndactylia.

6.1.4.1 Poland Syndrome

Poland syndrome (PS) is a rare type of congenital deformity that was first reported long ago, including unilateral sternal rib dysplasia and unilateral major pectoral muscle, minor pectoral muscle, and ipsilateral upper limb dysplasia; it often occurs to the right side, and girls often concomitantly suffer from breast dysplasia. The manifestation of hand dysplasia is short and small hand, syndactylia and brachydactylia. The etiologic factors are often considered to be correlated with the deformities in subclavian arterial series (Fig. 6.6).

In literature, the patients with Poland syndrome accompanied by multiple bone deformities are very rare. Professor Wang Wei found one 6-year-old patient with chest and hand dysplasia, with right major pectoral muscle and minor pectoral muscle absence accompanied by multiple bony deformities; the thorax was deformed, the second to sixth ribs suffered partial absence, thoracic bulging and excavation deformities were found in respiration, clavicles suffered dysplasia, ulnar and radial bones had fusion, carpal bones suffered dysplasia, and phalanges suffered deformities, but the shape of the affected hand was close to normal, and the diagnosis was Polandoid syndrome (Fig. 6.7). Polandoid syndrome is different from Poland syndrome, which generally has deformities in chest soft tissues; therefore, this case has its particularity [10].

6.1.4.2 Apert Syndrome

Apert syndrome is also called acrocephalosyndactyly and was first reported by a French neurologist Apert in 1906. It is a rare syndrome with an incidence of about 1/80,000 and characterized by craniosynostosis, exophthalmos, midfacial hypoplasia, and symmetric syndactylia. It is induced by the mutation in the fibroblast growth factor receptor 2 gene (FGFR2), and the gene is located at chromosome 10q, which falls into the category of autosomal dominant inheritance [11]. The cranial face shape of Apert syndrome is similar to that of Crouzon syndrome, but some characteristics are different: the head is flat at the anterior side and high at the posterior side, the bregmatic fontanel is protruding, the superior border of eye socket is sinking, the maxillary bone suffers hypodevelopment, and the palatal arch is high and narrow and often accompanied by cleft palate with the presence of abocclusion of anterior teeth. The patients are

Table 6.1 Syndromes accompanied by syndactylia

Syndrome	Clinical manifestations	Genetic characteristics
Poland syndrome	One-sided brachydactylia and syndactylia, major pectoral muscle, minor pectoral muscle, sternal head dysplasia, breast dysplasia, axillary web	Undecided
Apert syndrome	Narrow skull, ocular hypertelorism, exophthalmos, maxillary hypoplasia, mental retardation, and combined syndactylia at the fingertip	Autosomal dominant inheritance
Saethre-Chotzen syndrome	Narrow skull, ocular hypertelorism, exophthalmos, maxillary hypoplasia, and incomplete simple syndactylia	Autosomal dominant inheritance
Waardenburg syndrome	Acrocephalia, asymmetrical face and mouth, cleft palate, ear and nose deformities, simple brachydactylia and syndactylia, and occasional cleft of distal phalange	Autosomal dominant inheritance
Pfeiffer syndrome	Brachycephalia, wide and short thumb and big toes, accompanied by three-segment phalangeal simple syndactylia	Autosomal dominant inheritance
Summit syndrome	Acrocephalia, various types of deformities in feet and hands	Autosomal dominant inheritance
Noack syndrome	Acrocephalia, huge thumb, big toes and syndactylia, and syndactylia	Autosomal dominant inheritance
Carpenter syndrome	Acrocephalia, mandibular dysplasia, flat nose, mental retardation, and simple middle finger and ring finger syndactylia	Autosomal dominant inheritance
Oculodentodigital syndrome (Meyer-Schwickerath disease)	Microphthalmia and microcornea, glaucoma, microrhinia and small nasal wing, microtooth and adamantine hypoplasia, ring finger and middle finger syndactylia	Autosomal dominant inheritance
Oro-facial-digital syndrome I (orofaciodigital syndrome I)	Lingual frenum dysplasia, split tongue, cleft palate, middle fissure of lips, mandibular sulcus and groove, alveolar processes, abnormal teeth, maxillary hypoplasia, simple syndactylia; male patients are apt to die	X-linked dominant inheritance
Oro-facial-digital syndrome II	Slit tongue, middle fissure of lips, alveolar cleft, mandibular dysplasia, and syndactylia	Autosomal inheritance
Acropectorol-vertebral syndrome	Syndactyly, small toes and polydactyly, metacarpal bone or phalange fusion, sternal protrusion, cryptorachischisis, mental retardation, and thumb and index finger syndactyly	Autosomal inheritance

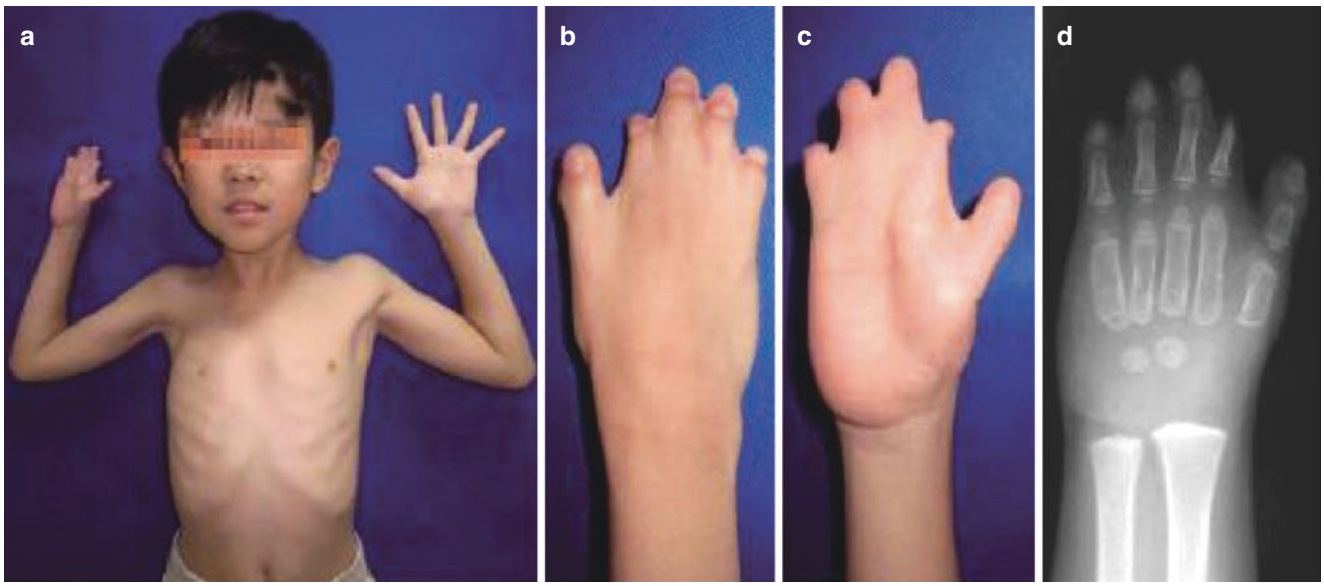


Fig. 6.6 A 9-year-old boy with Poland syndrome. (**a**) Right major pectoral muscle and minor pectoral muscle dysplasia. (**b**, **c**) Right-hand dysplasia, brachydactylia, and syndactylia. (**d**) X-ray indicates osteodysplasty of the second to the fifth finger and thumb web stenosis

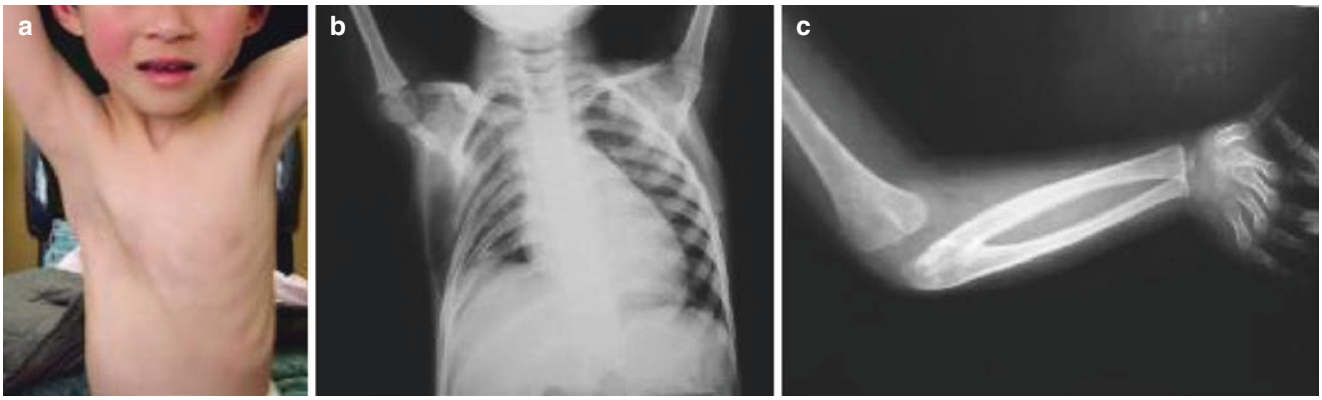


Fig. 6.7 A 6-year-old boy with Polandoid syndrome. (**a**) Right pectoral muscle absence, partial defect of the second to sixth ribs, and presence of thoracic bulging or excavation deformity in respiration. (**b**, **c**) Concomitantly with multiple bone deformities, ulnar and radial bone fusion, carpal bone dysplasia, and phalange deformity

apt to concomitantly suffer acne, oculomotor paralysis, ptosis, skinfold at the frontal part, big ear lobe, etc. [12].

The syndactylia that accompanies Apert syndrome is serious and complicated. Although many other Apert syndromes are defined, the child's hand deformities were not as complicated as Apert syndrome. In addition to the characteristic hand deformities, the manifestations also include shoulder and elbow deformities and tuberosity overgrowth and glenoid cavity dysplasia induced by heterauxesis of glenohumeral joint. Along with the growth, the limitation in the activity of the shoulder joints is increasingly serious. The commonest involvement of elbow deformities lies in humeroradial joint.

Hand deformities include the combined syndactylia of index, middle, and ring fingers as well as the simple syndactylia of ring and little fingers. The syndactylia of the thumb and index finger to varying extents impedes the effective gripping functions, and this impediment becomes aggravated due to the lateral curvature of the radial side of the thumb. The middle finger is short, and the interphalangeal joint is rigid (Fig. 6.8) [13, 14].

In the most serious cases, all distal phalanges are mutually fused. With the development of the finger, the shape becomes flowerlike or sarciniform, and due to the mutual restriction of each finger, a deep hole is formed at the palm. Due to overlapping and the inward growth of the adjacent nail plates, nail fold infections are often induced. Head and hook synostosis and the bony synostosis between the metacarpal bones of the ring and little fingers are common. This finding is very common clinically; the reason is that it prevents the fifth finger from transforming to the limited thumb. The degree of hand deformities and the degree of deformities in cranial face are inversely correlated. The classification of hand deformities includes the presence of involvement in the first fingerweb and the conditions of central digital plates (Table 6.2).

The diagnosis is mainly based on clinical examination. The diagnosis can be confirmed in case of the presence of family history (often sporadic) with the assistance of skull X-ray and CT films, and the hand and foot X-ray films can confirm the diagnosis of bony deformities in the hand and foot [15].

6.1.4.3 Bardet-Biedl Syndrome

Bardet-Biedl syndrome (BBS), also called Bardet-Biedl disease, is a kind of autosomal recessive inherited disease [16]. This is a syndrome that is not included in Table 6.1, with the manifestations of abdominal obesity, mental retardation, and limb deformity, including syndactylia, brachydactylia, or polydactylism, accompanied by retinal dystrophy, pigmentary retinopathy, sexual hypofunction, or hypoplasia of gonads. Lannello S. (2002) reported three cases with Bardet-Biedl syndrome in a family, and the patients were two females and one male (siblings) at the age of 66, 64, and 54, respectively. Bardet-Biedl syndrome may be accompanied by syndactylia, but the symptoms are various [17].

6.1.4.4 Neurogenic Fat Fibroplastic Proliferation

Progressive macrodactylia and syndactylia induced by neurogenic fat fibroplastic proliferation are rare types of deformity, and correction of both syndactylia and macrodactylia should be considered in the treatment of this deformity [18].

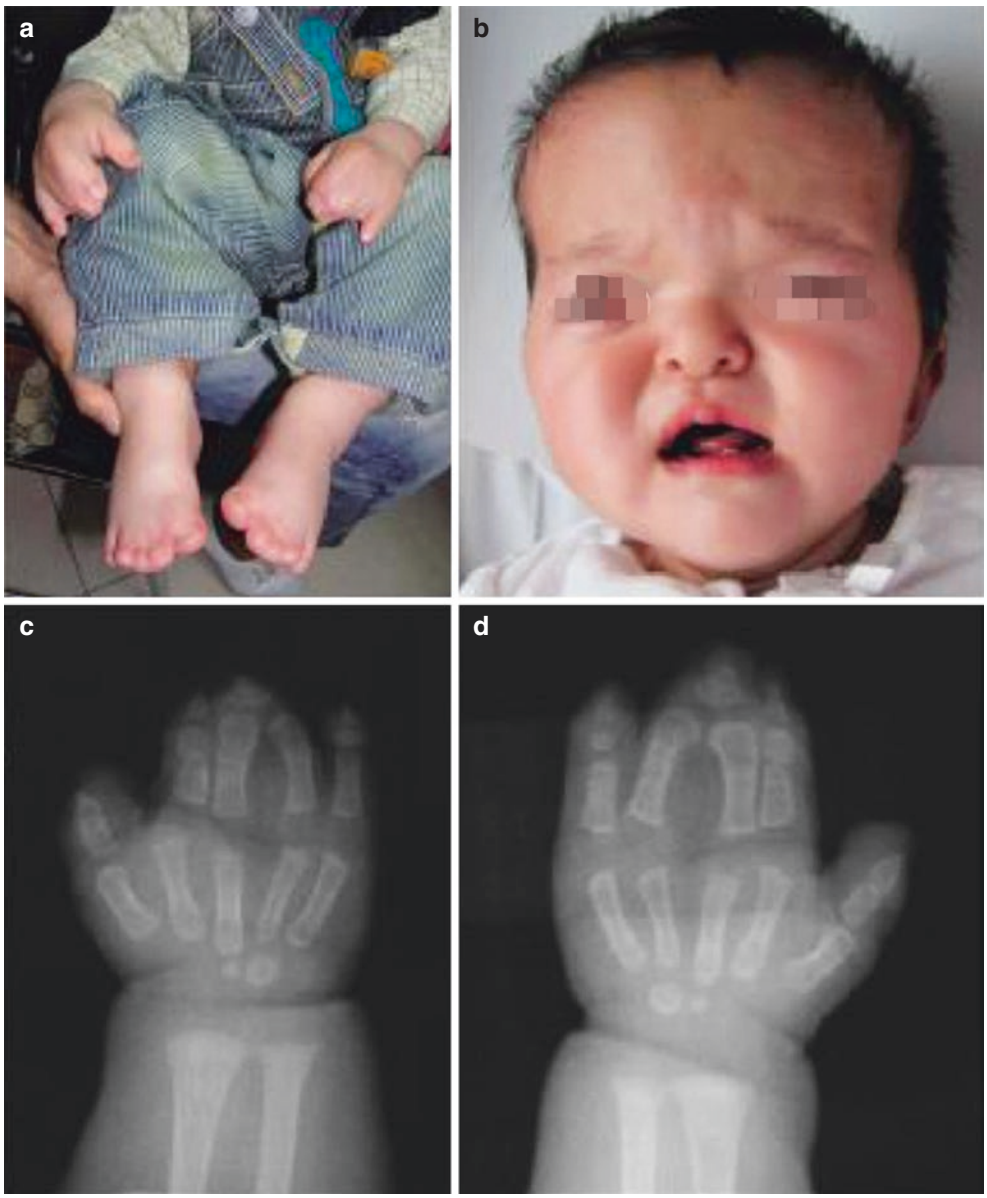


Fig. 6.8 Apert syndrome. (**a**) Combined syndactylia and hyperdactylia. (**b**) Facial appearance. (**c**, **d**) Hand X-ray findings

Table 6.2 Classification of hand deformities in Apert syndrome

Type	The first fingerweb (hukou)	Central digital plates	The fourth (ring and small fingers) fingerweb
Type I: spade hand	Incomplete simple syndactylia	The volar surface of the digital plates is smooth, the metacarpophalangeal joint is normal, and the interphalangeal joint has fusion to varying degrees	Incomplete simple syndactylia
Type II: spoon-shaped hand	Complete simple syndactylia	Volar excavation of digital plates, outward abduction of the proximal end of metacarpal bones, fingertip fusion, and synonychia	Complete simple syndactylia
Type III: hoof-shaped hand	Complete combined syndactylia	The thumb is involved and forms a cup-shaped structure together with the digital plates. In addition to little fingers, all fingers have synonychia, skeletal deformities in index finger ray, nail groove infection, and immersion-like changes of the volar skin	Simple syndactylia, commonly with the bony synostosis of the fourth and fifth metacarpal bones

6.1.5 Preoperative Evaluation

When the patients are preoperatively evaluated, the important factors that should be considered include the number of involved fingerwebs, range of syndactylia, conditions of nail involvement, and the presence of combination with other deformities. The absence of interdigital differential motion can indicate the presence of bone fusion and/or the hiding of one extra finger into the adjacent finger. Physical examination should cover the entire upper limb, contralateral hand, chest wall, and feet [19]. Radiological examination can help find whether bone fusion is present and whether recessive hyperdactylia (syndactylia and hyperdactylia) or other bone and joint deformities are suffered. Further ultrasound or magnetic resonance examination helps to judge whether the flexor tendon and vascular anatomy of combined syndactylia are abnormal.

For a child in the middle of growing, syndactyly may have effects on many aspects including its appearance, functions, and development. The hand appearance of patients is different from that of healthy people, especially those with complete combined syndactylia. The syndactylia of the thumb and index finger will impede the development of hand gripping and pinching functions; the syndactylia between the remaining fingers will inhibit the independent movement of each finger, especially their abduction; therefore, the hand transverse span will be decreased. The digital syndactylia of varying lengths will cause the longer finger to be restrained so that it will laterally bend to the shorter finger. With further growth, flexion contracture can be induced at the proximal interphalangeal joint [20] (Figs. 6.9 and 6.10).

6.1.6 Treatment

Surgical treatment is applicable to most cases, but the contraindications include mild incomplete syndactylia without functional disorder, health conditions to which surgery is inapplicable, or the presence of combined syndactylia in which case finger separation can induce further functional disorder (Figs. 6.11 and 6.12) [20]. Sometimes, the quantity of tissues is not enough to reconstruct independent, stable, and movable fingers; this is common in central brachydactylia and syndactylia deformity or synpolydactyly deformity, which may induce functional impairment after finger separation.

The main points in the surgical treatment of syndactylia mainly include selection of surgical timing, finger separation in different stages of syndactylia and hyperdactylia, fingerweb reconstruction, finger separation, skin coverage, and postoperative bandaging and care.

6.1.6.1 Surgery Timing

Dactylolysis can be performed in neonatal period, infancy, or childhood. The long-term follow-up visits of Flatt and Ger [20, 21] found that, although early surgery should be performed due to the influence of deviation and deformities of skeletons, better effects can be produced if dactylolysis is performed after 18 months. The treatment objective is to complete all finger-separating surgeries before the school age. The syndactylia of multiple fingers needs to be surgically treated in different stages, and during each stage, only one side of the affected finger can be separated in order to avoid injuring the skin flap or the digital vessels. If the syndactylia involves all fingers, the treatment often needs to be

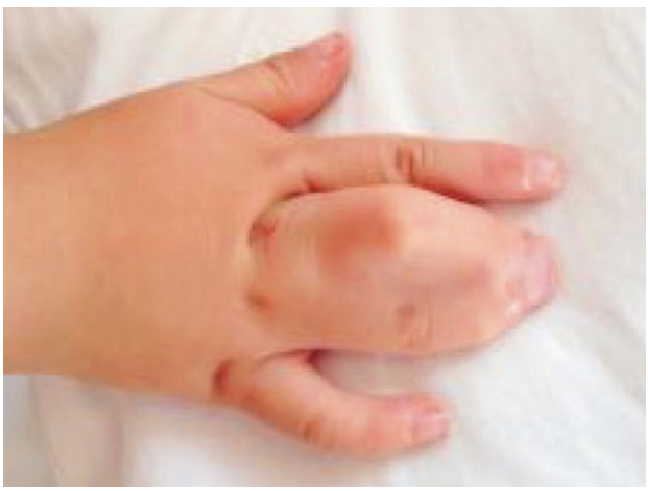


Fig. 6.9 Syndactylia affects the development of the finger and induces flexion contracture of interphalangeal joint

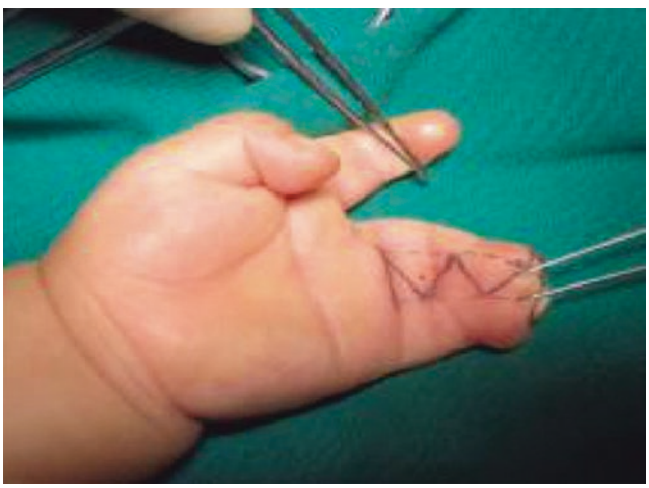


Fig. 6.10 Syndactylia causes the middle, ring, and little fingers to become angulated toward the ulnar side

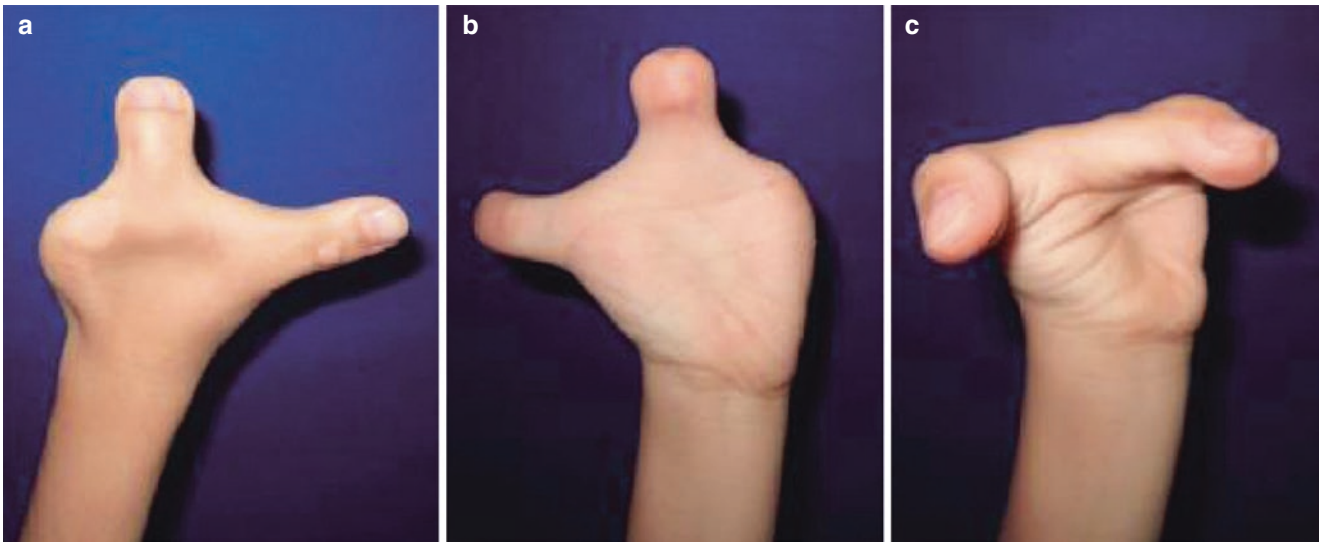


Fig. 6.11 Combined syndactylia

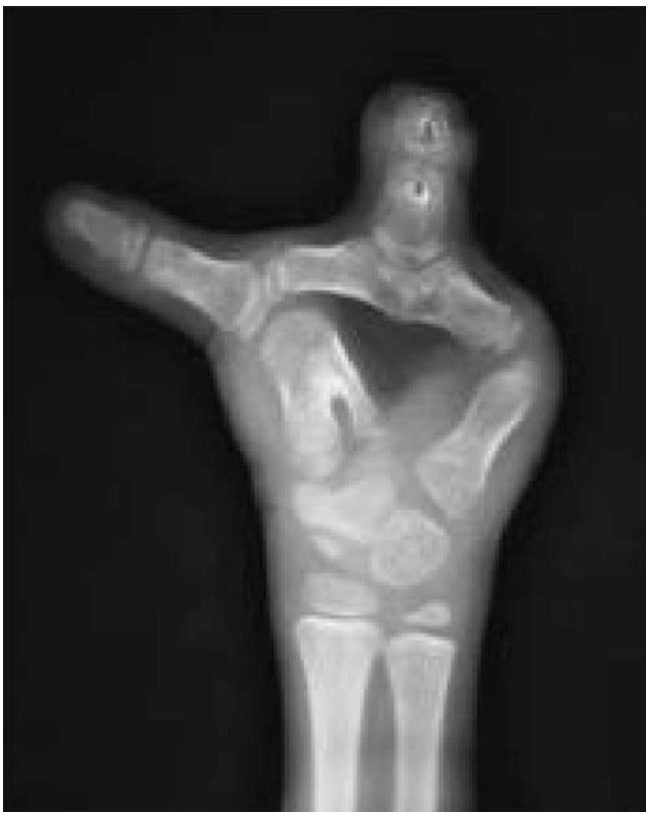


Fig. 6.12 X-ray findings of combined syndactylia with complete phalange fusion to which surgical separation is inapplicable

finished in two stages: during the first stage, the thumb and index finger as well as the middle and ring fingers are separated; the second-stage surgery is performed 3 months later to separate the index and middle fingers as well as the ring and little fingers. In addition, during the first stage, the separation of fingertips of all fingers and the separation of the distal phalange fusion can be performed at the same time, which can lay a solid foundation for the second stage of surgery.

6.1.6.2 Reconstruction of Fingerweb

The key of dactylolysis lies in the reconstruction of fingerwebs with good functions and shapes, and the commonest method is to make a rectangle flap at the proximal end at the dorsal side of syndactylia as well as many variational forms, such as dorsal ladder flap and dorsal flap with lateral wing. There are many alternatives available for the skin at junctures. The skin at the back of the hand is designed into island skin flap and is pushed into the fingerweb space according to V-Y. As for the volar surface of independent syndactylia (or with the dorsal surface), mutual insertion of triangular flaps can be utilized to reconstruct the fingerweb. As for the incomplete syndactylia limited to the proximal segments of fingers, the simple Z-plasty, four-flap Z-plasty, or butterfly flaps can be used to deepen or lengthen the existing fingerwebs to achieve the effects of reposition (Fig. 6.13), and other methods include mutual combination of local skin flaps, such as three-flap fingerweb plasty or V-M plasty [22]. Incomplete syndactylia often induces the pseudomorph that there is sufficient skin locally. However, when the juncture is reconstructed and the local flaps are grafted, skin defect often appears to the fundus of the fingerweb, and superfluous skin is present at the distal end of fingerwebs. Brennan and Fogarty introduce a technology, namely, advancing distal skin through the island flaps to the proximal end and combing with triangular flaps to reconstruct the junctures [23].

Z-plasty is the most frequently used and effective surgical method in syndactylia treatment, but due to the high

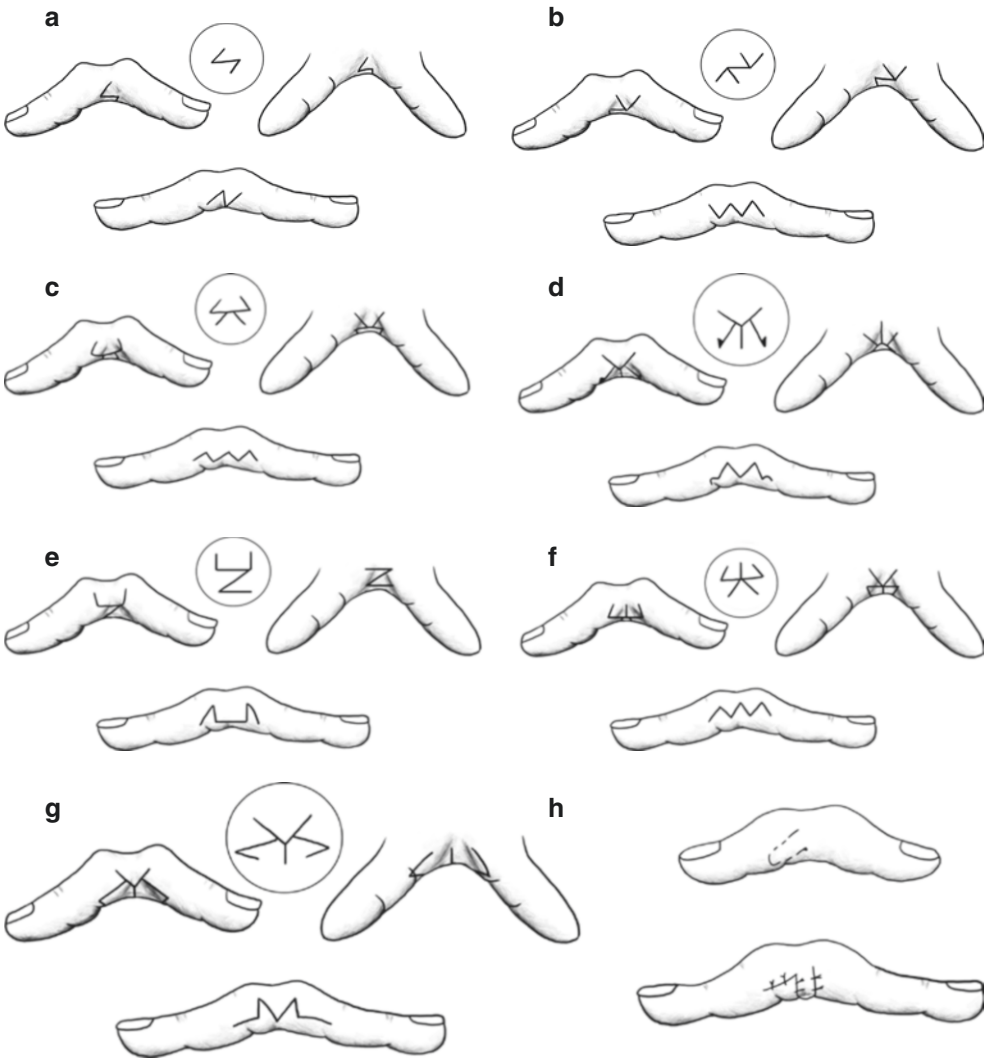


Fig. 6.13 Fingerweb plastic technique for syndactylia. (**a**) Single Z-plasty. (**b**) Overlapping four-flap method, negative direction dual Z-plasty. (**c**) Mirror image-type Z-plasty with two opposite ones. (**d**) Y-V plasty and V-Y plasty. (**e**) Rectangular flap advancement with Z-plasty. (**f**) Y-V plasty + dual Z-plasty with pedicle in the palm (five-flap plasty). (**g**) V-Y plasty + sea gull flap dual Z-plasty with pedicle on the back of the hand (five-flap plasty). (**h**) Digital linguiform flap grafting (Redrawn from Eaton, CJ: Syndactyly Hand Clinics 1990;6: 555)

flexibility of Z-plasty, there may be various changes, and proficient mastery of it needs long-term practice.

Single Z-plasty, also called coupled-triangular flap plasty or overlapping triangular flap plasty (Fig. 6.13a), is applicable to degree I syndactylia, namely, syndactylia range ≤ 1/8 of the distance between the head of metacarpal bone and the fingertip.

Surgical design of fingerweb Z-plasty: use the syndactylia fingerweb margin as the Z-plasty axis, and make an oblique incision at two sides, called arm; the axis and two arms form two triangular flaps at the opposite directions. After the skin is incised, fabricate two coupled-triangular flaps, make the two triangular flaps exchange positions, and suture them, so that the distance of the axial line can be lengthened, that is, tension is eased, and the purpose of relieving the syndactylia deformity is achieved. The best angle between the two flaps is 60°, which means the lengthening distance after translocation is the longest and can be up to 75%; in case of 45°, the distance can be lengthened by 50%; in case of 30°, the distance can be lengthened by 25%; in case of larger than 90°, the mutual transposition of the coupled flaps is very difficult. The angles of the two triangular flaps in Z-plasty transposition flaps can be equal or unequal (namely, one is larger, the other is smaller), and the surgery is called asymmetrical Z-plasty. This is the most frequently used surgical design. Based on this, there are many evolutions including double Z-plasty, continuous Z-plasty, and four-flap, five-flap, and six-flap plasty [24, 25].

The two-arm length of Z-plasty transposition flaps can be 0.5 cm, 1 cm, 1.5 cm, and 2 cm. It should be noted that the incisions of two arms of Z-plasty do not necessarily have to be made into a straight line and can be made into an arch or streamline shape according to the changes in cleavage lines.

Dual Z-plasty, commonly known as four-flap method, is better than single Z-plasty because the axial distance is lengthened. Figure 6.13b is an overlapping four-flap method; Fig. 6.13c is a mirror image-type Z-plasty with two opposite ones, which is another four-flap method and commonly used

for the reshaping of degree I and degree II syndactylia, but it is relatively common in degree I syndactylia.

Y-V plasty or V-Y plasty (Fig. 6.13d) is often applied to the correction of syndactylia. Y-V plasty is the skin Y-shaped incision and V-shaped suturing to increase the transverse length of the skin to serve the purpose of syndactylia correction. V-Y plasty is the skin V-shaped incision to release the triangular skin tissues and return them to the necessary position, and Y-shaped suturing can realize tissue reduction. Multiple Y-V plasties can greatly increase the transverse length of the skin to serve the purpose of syndactylia correction.

Rectangular flap advancement + Z-plasty (Fig. 6.13e) is designing one rectangular advancement flap at the back of the hand and designing one single Z-plasty at the volar side of the fingerweb to increase the depth of syndactylia correction, and it is applicable to the reshaping of degree I and II syndactylia.

Y-V plasty + dual-Z-plasty forms the five-flap plasty. Design Y-shaped incision of the skin in the middle of the fingerweb, perform V-shaped suturing, and then design one single V-plasty at the two sides of Y-V plasty to increase the transverse length of the skin to serve the purpose of syndactylia correction. Figure 6.13f shows the Y-V plasty + dual Z-plasty with pedicle in the palm, and it forms the five-flap plasty. Figure 6.13g shows the Y-V plasty + sea gull flap dual Z-plasty with pedicle on the back of the hand, which is also five-flap plasty [26].

The digital lateral linguiform flap grafting (Fig. 6.13h) can deepen the fingerweb, which is the surgical design commonly adopted by the author to correct the incomplete syndactylia induced by burning and which can also be used for the correction of congenital syndactylia. This surgical design is easy to perform and can be classified as one of Z-plasty surgeries.

Yao Jianming [27] and Xu Jinghong [28] established the fascia pedicle fingerweb flap setback surgery to treat simple syndactylia, and the main points are shown in Fig. 6.14. Design the fingerweb skin flap at the distal end of the fingerweb of the simple syndactylous fingers, and use the proximal end of the longitudinal fascia between the syndactylous fingers as the pedicle. Design a V-shaped incision at the skin of the digital volar and dorsal fingertips; according to the normal fingerweb proportion, the length of the volar surface is twice that of the palm surface. Extend the sawtooth-shaped incision toward the proximal end until the pedicle is reached. After the skin of the fascia pedicle at the distal end of the fingerweb is dissociated and grafted, explore the deep part of

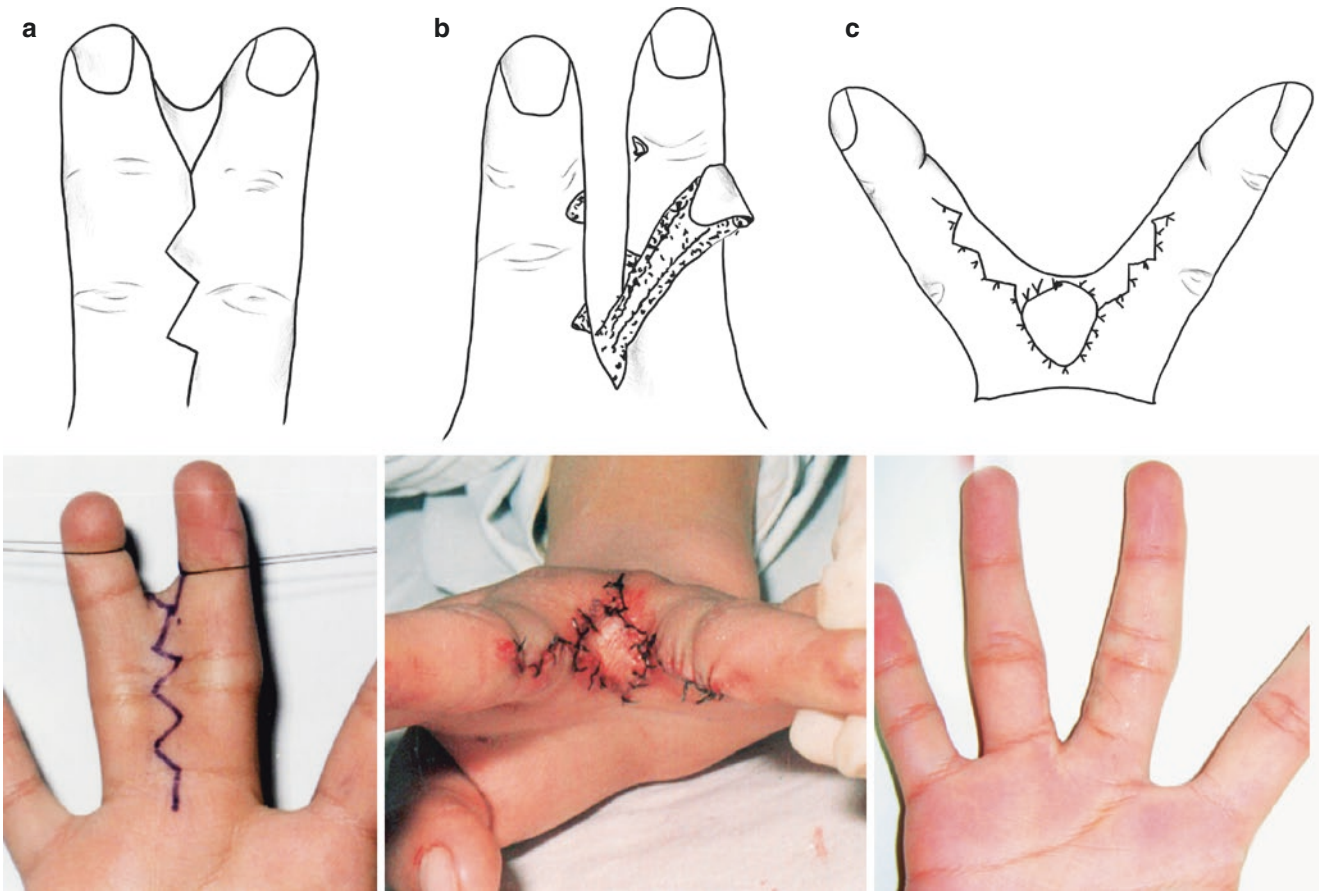


Fig. 6.14 Fascia pedicle fingerweb flap setback surgery. (**a**) Flap design. (**b**) Intraoperative findings. (**c**) Hand appearance 7 years after operation

the fingerweb. Carefully separate the subcutaneous tissues, form a skin pedicle, avoid injuries of digital arteries and fascia pedicle, and perform multiple Z-plasty suturing on the skin at two sides between the fingers. This surgical mode is applicable to the simple syndactylia with rich skin at the fingerweb, but cannot be used for the combined syndactylia and complete syndactylia with very fine fingertips.

Di Sheng [29] designs interdigital proximal and distal fascia pedicle flaps to reconstruct the skin defects induced by syndactylia separation, and the main points of this surgery are to design rhomboid flaps in the interdigital middle part of the skin, which is divided into two parts after transection. The anterograde pedicle point is located at the root of the proximal phalange, while the retrograde pedicle point is flexibly designed at the distal end of the axial line. The flap anterograde part is used to reconstruct the fingerweb, while the retrograde part is used to repair the distal defects of the finger (Fig. 6.15).

The syndactylia of the first fingerweb is common in patients with syndrome; for example, the patients with Apert syndrome are often accompanied by thumb deformities, which exert a greater influence on hand functions compared with other syndactylia deformities. The mild to moderate syndactylia of the first fingerweb can be treated through regional flaps [30], such as four-flap Z-plasty; other options include local flap grafting at the index finger or combined application of local flap grafting at the radial side of the index finger and the ulnar side of the thumb or the V-Y flap advancing at the central fingerweb. In case of serious syndactylia accompanied by significant stenosis of fingerweb at the thumb and index finger, it is necessary to provide more skin than what is provided by regional flaps. Under this situation, the skin can be harvested through the dilation of tissues at the back of the hand, or rotation is used to advance the skin; the distal pedicle skin flap or the axis-shaped flaps, such as inguinal, interosseous dorsal, or arm medial flaps, can also be used (Fig. 6.16). Free skin flaps can provide more coverage for the hands of the patients with serious skin defect and syndactylous syndrome.

6.1.6.3 Finger Separation and Skin Coverage

It is necessary to carefully design the incisions in separation of syndactylous finger so that the available skin can be optimized. When the surgery exposes the structure of the separated fingers, the incisions must be designed in such a way that the cicatricial contraction will not lead to interarticular and inter-fingerweb contractures. At present, it has evolved into numerous incisional designs, including the triangular flaps and rectangular flaps at the lateral fundus. Cronin technique [31] has always been the commonest technique in separation of syndactylia. Through multiple sawtooth-shaped incisions, triangular flaps at the volar side and dorsal sides of syndactylous fingers are formed to avoid skin coverage of contracture (Fig. 6.17a). There are many modifications of this method, and the aim is to redistribute the available skin so that the grafting of the skin at two sides of the fingerweb can be avoided (Fig. 6.17b–e). Sawabe [32] published a series of papers on separation of syndactylia. He adopted the skin grafting method with lateral central linear incisions to close the wounds. After operation, braces were used to avoid contracture, and subsequently, one acceptable lateral central scar was retained after removal. Although this method goes

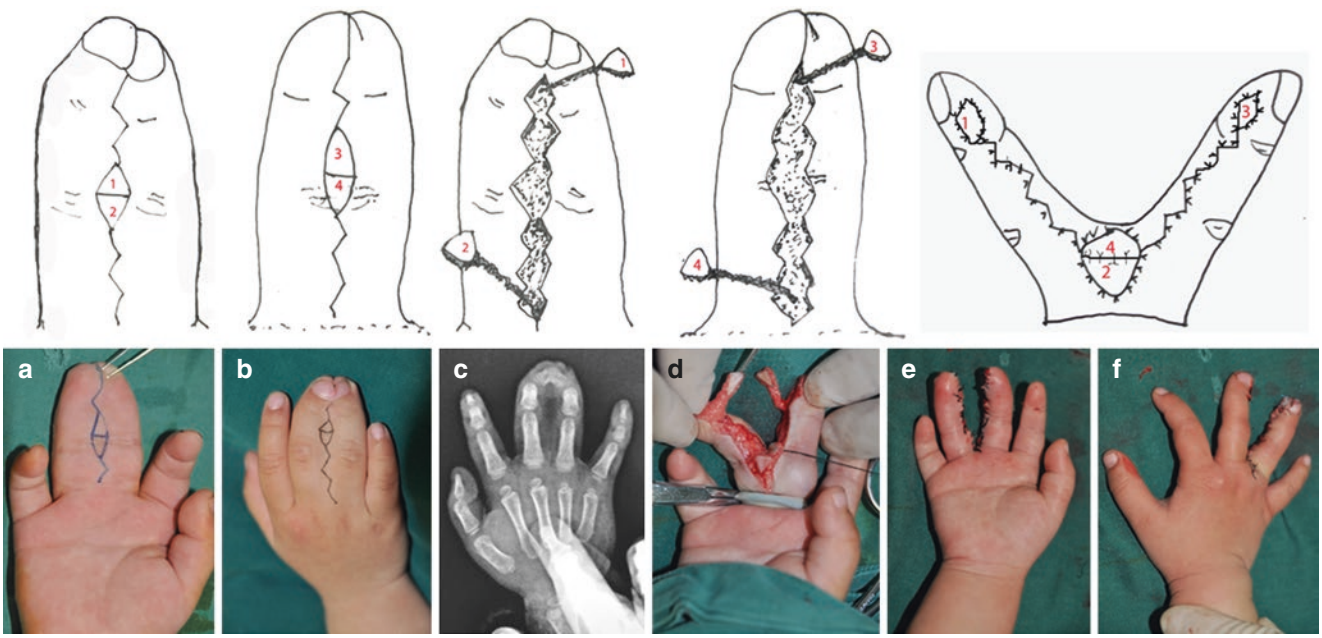


Fig. 6.15 Design main points of interdigital proximal and distal fascia pedicle flaps

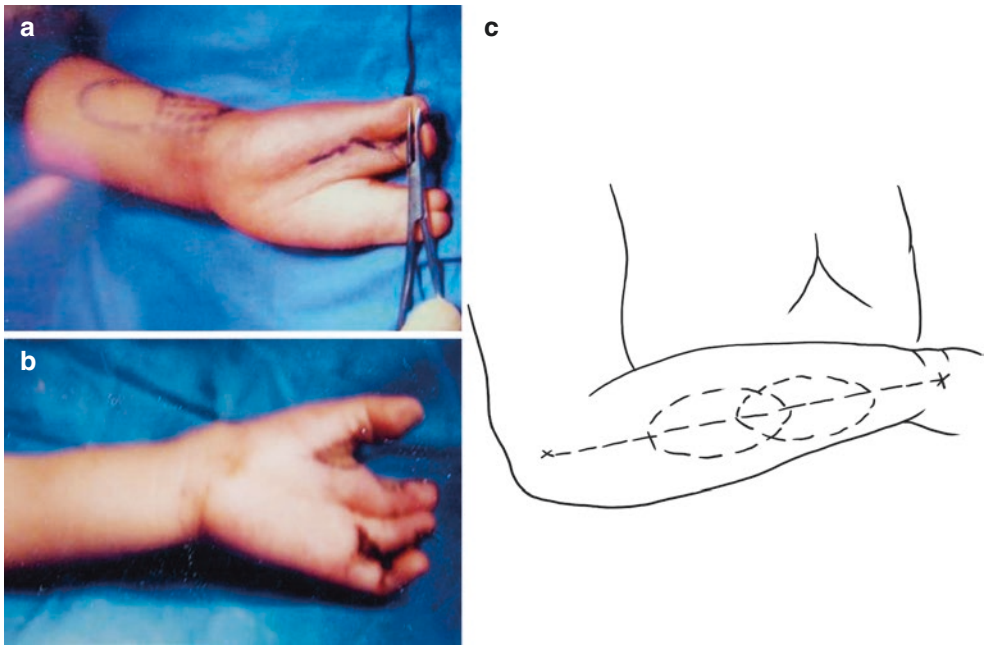


Fig. 6.16 Utilization of interosseous dorsal island flaps to reconstruct the thumb web stenosis induced by syndactylous finger separation (operator by Wang Wei)

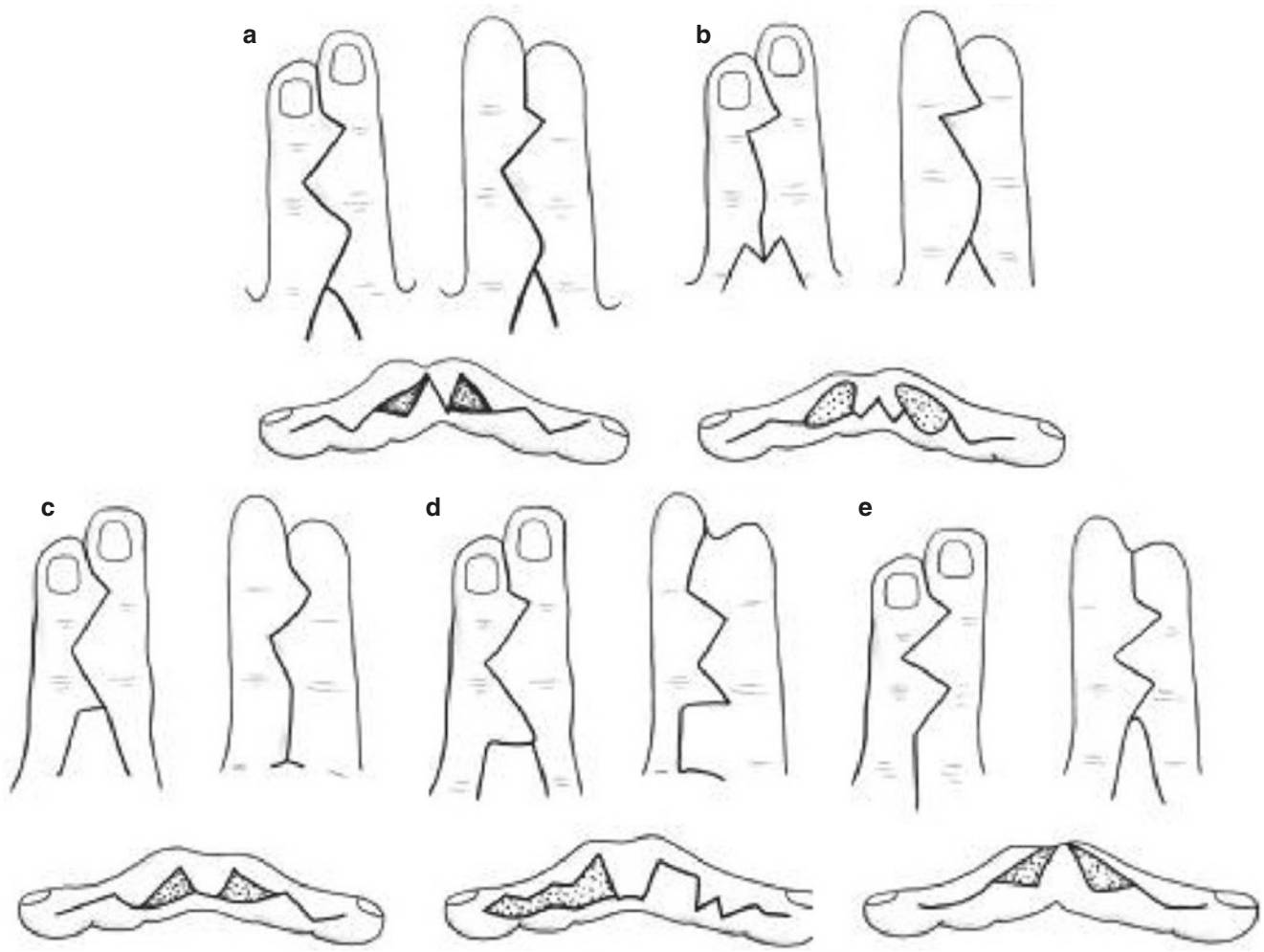


Fig. 6.17 Separation of syndactylous finger. (**a**) Separation of two fingers, dual triangular flap method (Cronin technology). (**b**) V-M flaps for fingerweb reconstruction. (**c**) Rectangular flaps at the back of the hand. (**d**) Palm transverse rectangular flaps. (**e**) Volar triangular flaps, V-Y plasty (Redrawn from Bauer TB, Tondra JM, Trusler HM: Technical modification in repair of syndactylism, Plast Reconstr Surg 17:385, 1956)

against the traditional technology, it is effective when the skin and scar experiences pigmentation or proliferation. Another method is Sommerlad's digital exclusion technique, namely, no treatment of residual skin defects for healing by second intention; however, most surgeons are worried that this will lead to the formation of the second phase scar [33].

During the separation, it is necessary to cut and remove the fascia between the two fingers. Not only should the neurovascular bundles of each finger be identified and protected, but also the fingerweb should have sufficient venous regurgitation (Fig. 6.18). The bifurcation of the digital nerves and arteries may be at a farther position compared with the designed fingerweb. In this case, if no surgery is performed on another side of this finger or the postoperative digital artery is intact, the digital artery can be ligated; otherwise, the level of fingerweb is limited to the level of arterial bifurcation, or the veins can be grafted to lengthen the arteries (this is applicable to very rare situations). When multiple fingers need to be separated, at least one digital artery should be retained for each finger; therefore, the surgical design must be accurate. The processing of the distal bifurcation of the digital fingers can be interfascicular cutting and separation of proximal end.

The skin coverage of the fingers depends on the grafting of the volar and dorsal flaps at the syndactylous fingers assisted by skin grafting. The full thick skin graft is superior to moderate thick skin graft, and it can reduce contracture [34]. In most cases, the selected donor sites for skin grafting are at the inguinal regions, and other donor sites include the medial side of the upper arm, antecubital fossa, hypothenar eminence, wrist, or accessory fingers. Although the foreskin may have the disadvantages of insufficient skin and mismatched color, it was once used. No matter what site is used as the donor site, it should be explained to the patients in detail and consent should be obtained from them because this will produce scars.

In order to improve the overall matching degree of the skin and avoid the occurrence of contracture after skin grafting, the dactylolysis without skin grafting starts to be applied [35]. In implementation of this technique, the vessels and nerves should be protected while the subcutaneous fat should be removed so that the diameter of the fingers can be reduced, so fine operation is needed (Fig. 6.19). Another method to avoid skin grafting is to harvest the skin from the back of the hand and/or the adjacent fingers; in case more skin is needed, it can be harvested through tissue dilation. Some scholars place skeletal traction scaffolds at the distal end of syndactylous finger as the transverse traction so as to dilate the skin at the distal end of the syndactylous finger so that sufficient graftable skin can be harvested at the distal end of the complete syndactylia. Though the application of this technology to syndactylia is limited, it provides new measures for separation of combined syndactylia.

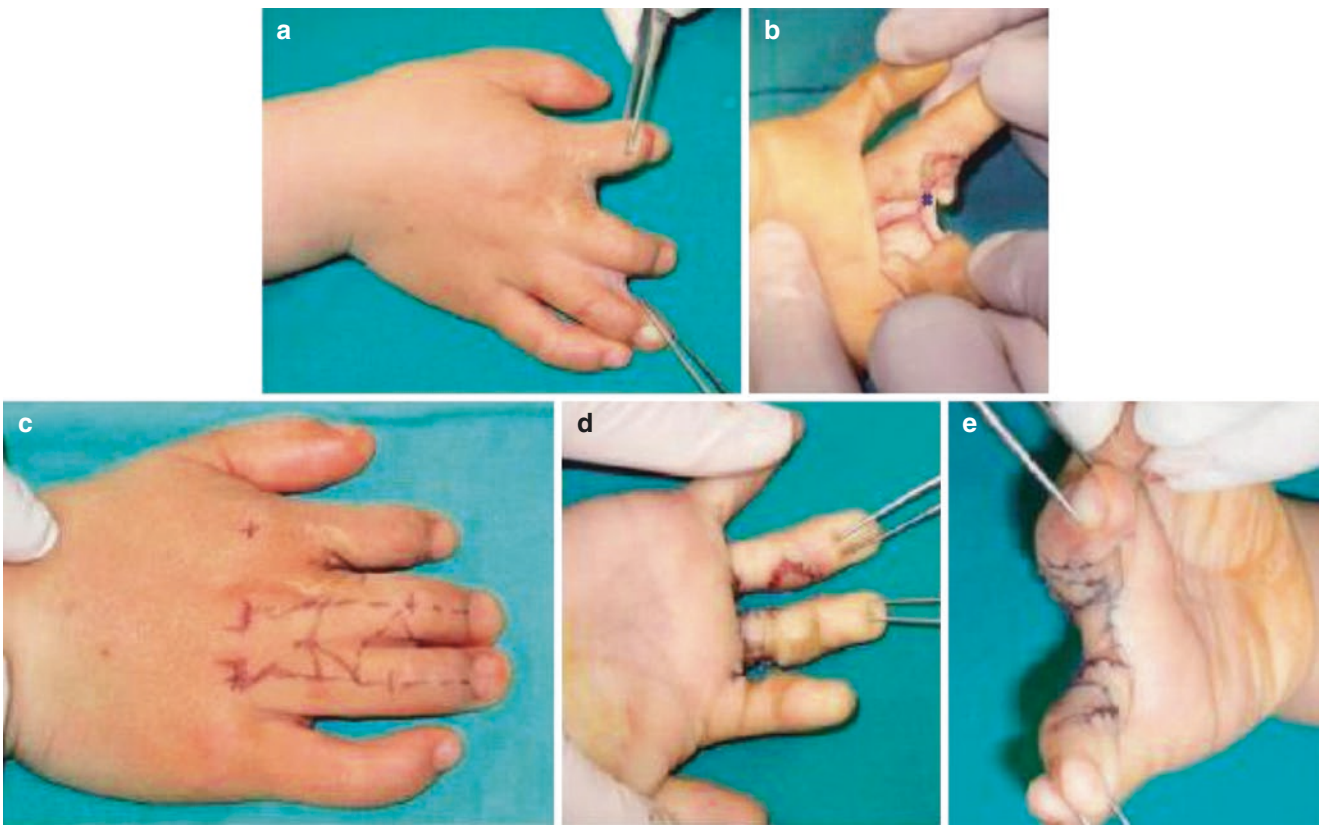


Fig. 6.18 Identify the bifurcations of the nerve vessels, and avoid the bilateral blood supply of single fingers from being affected

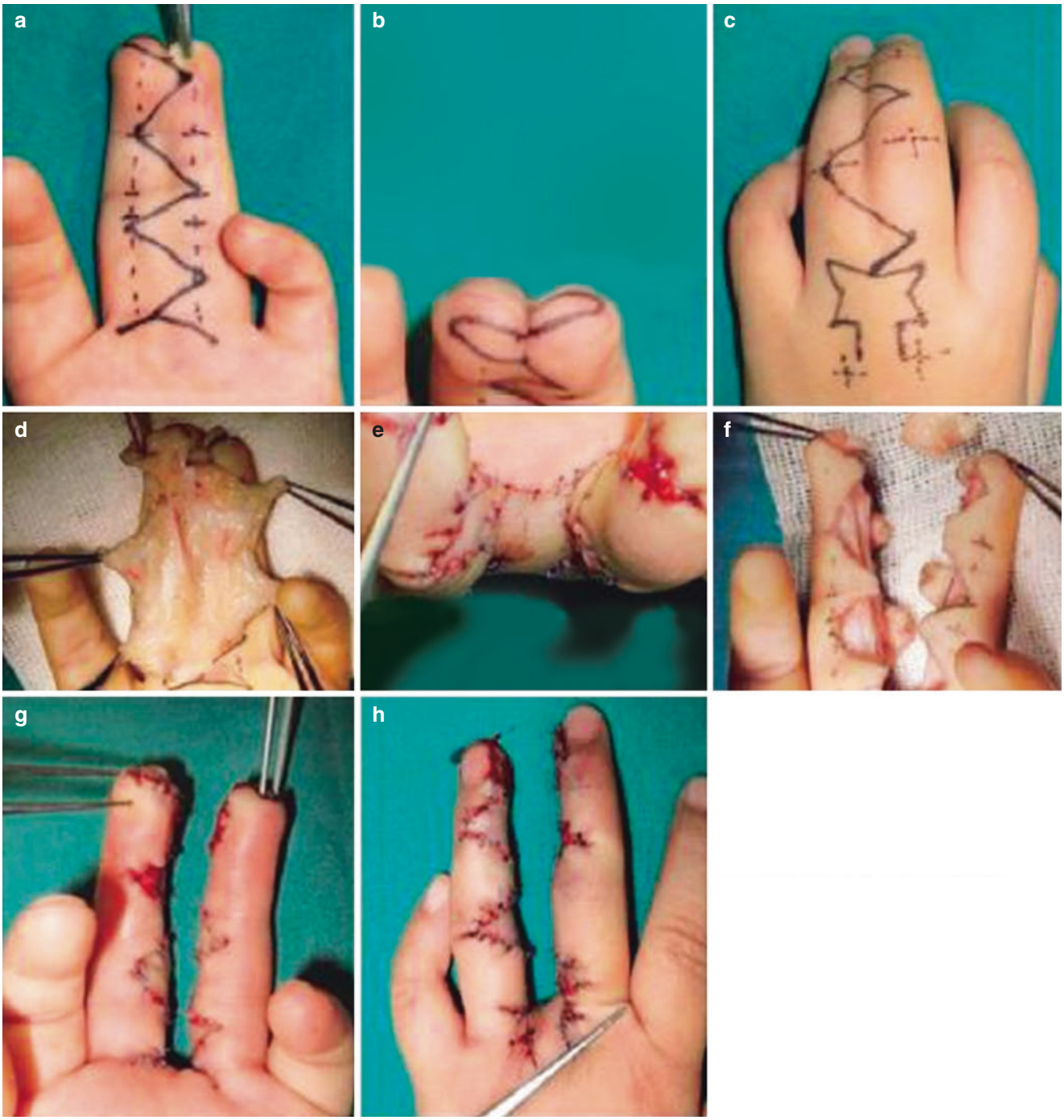


Fig. 6.19 Dactylolysis without skin grafting. (**a**–**c**) Preoperative design. (**d**–**f**) Fat-reducing technology with retained vascular nerves. (**g**, **h**) After accurate suturing

6.1.6.4 Nail Fold Formation

Nail fold reconstruction is needed for complete separation of syndactylia especially combined distal phalange fusion. The distal phalange can be treated by adopting Buck-Gramcko [9] technology: design linguiform rotational flaps at the distal end of the syndactylous finger, and fold them to reconstruct the bilateral nail folds; or design the digital dorsum linguiform rotational flaps + fingertip linguiform flaps to reconstruct the nail folds (Figs. 6.20 and 6.21); one flap can also be made at the combined finger pulps to reconstruct the nail folds of one finger, and the subcutaneous fat flap + skin grafting at this site can be used to reconstruct the nail folds of another finger; pedicle skin flaps such as thenar flaps can be applied, and the grafting of skin and subcutaneous tissues from the toes can be performed to reconstruct the nail folds.

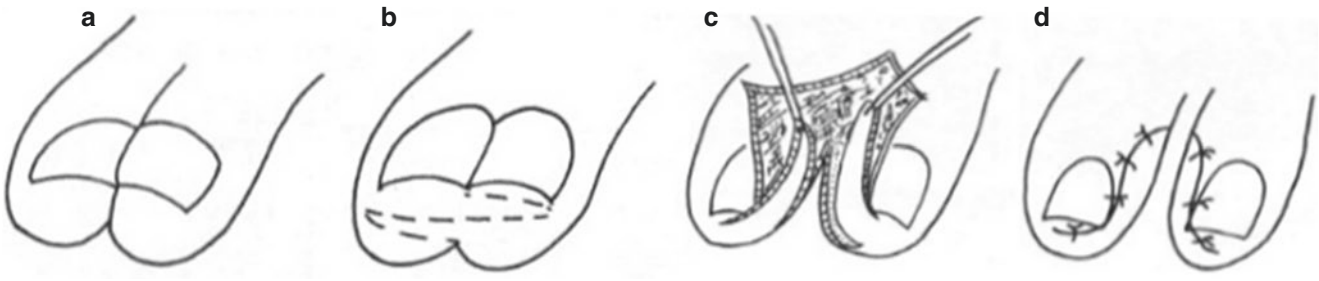


Fig. 6.20 Fingertip linguiform rotational flaps to repair fingertip defects and reconstruct nail folds

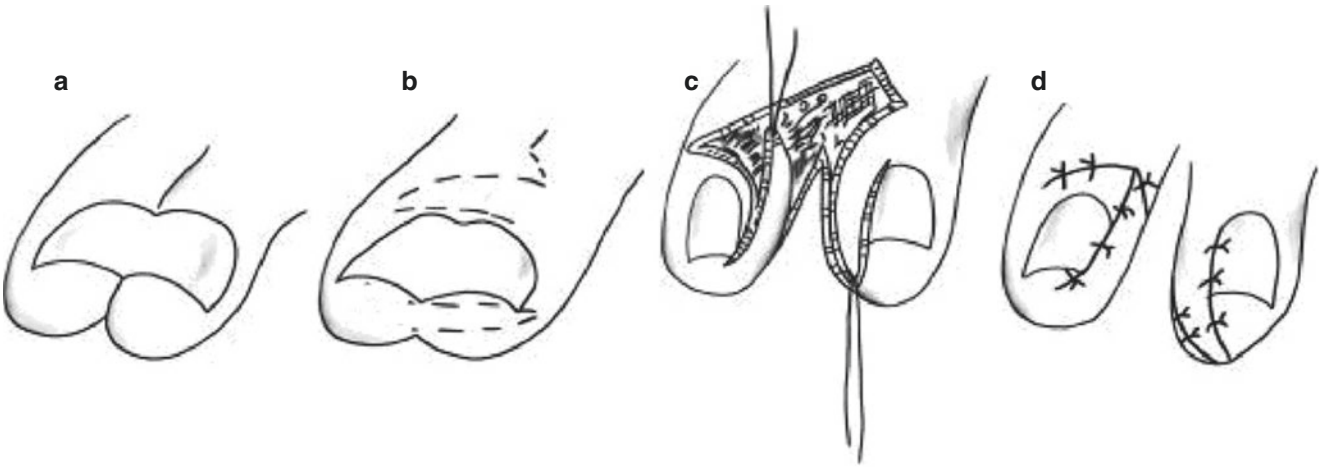


Fig. 6.21 Digital dorsal linguiform rotational flaps + fingertip linguiform flaps to repair the defects at the fingertip and reconstruct the nail folds

6.1.7 Complication

Early complications include vascular injuries, infections, wound dehiscence, and grafted skin necrosis. The intraoperative fine separation can avoid vascular injuries. Trimming and cleaning of nails before operation can substantially reduce the occurrence of infection, stainless suturing can help to avoid wound dehiscence, and the skin grafting on the tissue bed with a good blood supply can reduce the necrosis rate.

Late complications include:

1. Loss of fingerweb depth (Fig. 6.22): the reasons are poor design of flaps and formation of longitudinal scars at the fundus of fingers; it is also correlated with necrosis of the grafted skin and dehiscence of fingerweb flaps.
2. Joint contracture (Fig. 6.23): it is induced by volar cicatricial contracture of interphalangeal joint. It is usually necessary to resect the cicatricial tissues and further perform skin grafting; in case of sufficient skin in the region, Z-plasty can be performed to lengthen the scars.
3. Deformity of onychogryphosis and nail plate inclination: often induced by insufficient soft tissue amount at the fingertip and finger pulp.
4. Unstable joint: mostly induced by posterior collateral ligament defects after separation of combined syndactylia.
5. Keloid formation (Fig. 6.24): it is mostly correlated with the constitution, and it is often necessary to perform keloid resection and re-grafting of the skin or perform an all-round treatment on scar.

6.1.8 Author's Method

Under the condition of general anesthesia, the patient receives dactylolysis after tourniquets are applied.

First, design the dorsolateral flaps to reconstruct the fingerweb. The flaps start at the head of metacarpal bone, the shape is similar to the sand clocklike structure of the fingerweb, and then it extends to about two-thirds of the length of the proximal phalange. Mark the transverse striation level of the proximal finger that needs to be reached at the distal end of the flaps, and subsequently make sawtooth-shaped incisions at the volar and dorsal sides of the fingerweb flaps and proximal digital transverse striations. The apex of the incisions reaches the midline of the finger so that the triangular flaps have a relatively large degree of excursion. This design attempts to reduce the possibility of postoperative finger flexion contracture, and at the same time, maximize the coverage area. The flaps undergo a sharp separation, and bipolar electrocoagulation is used for hemostasis. First, separate the dorsal flaps,

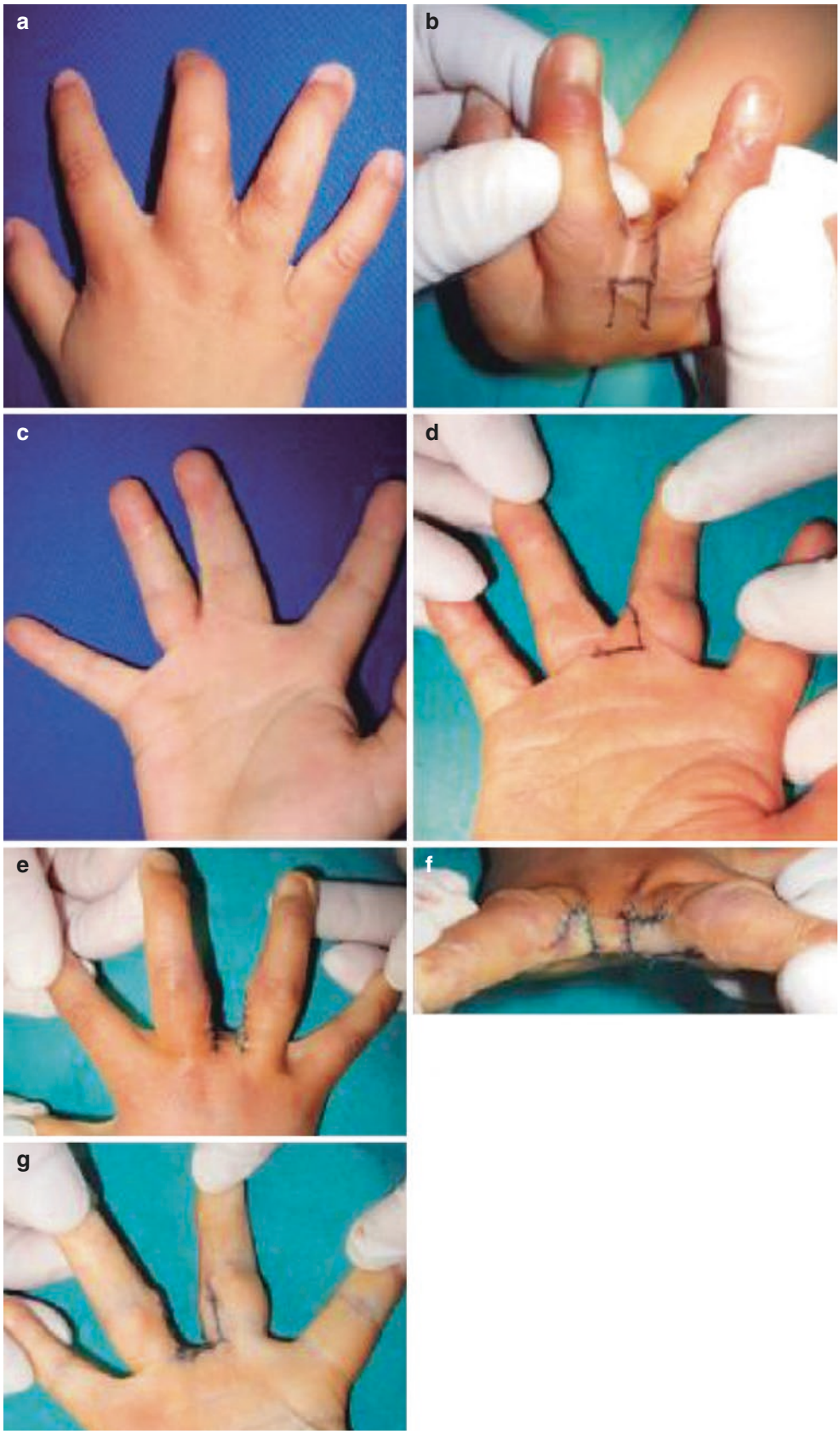


Fig. 6.22 As for the loss of fingerweb depth induced by longitudinal cicatricial contracture, rectangular flaps can be used to re-deepen the fingerweb and reconstruct the subunit structures of fingers. (**a**) Complications after syndactylia surgery. (**b**–**d**) Re-design of surgical incision. (**e**–**g**) Postoperative fingerweb appearance

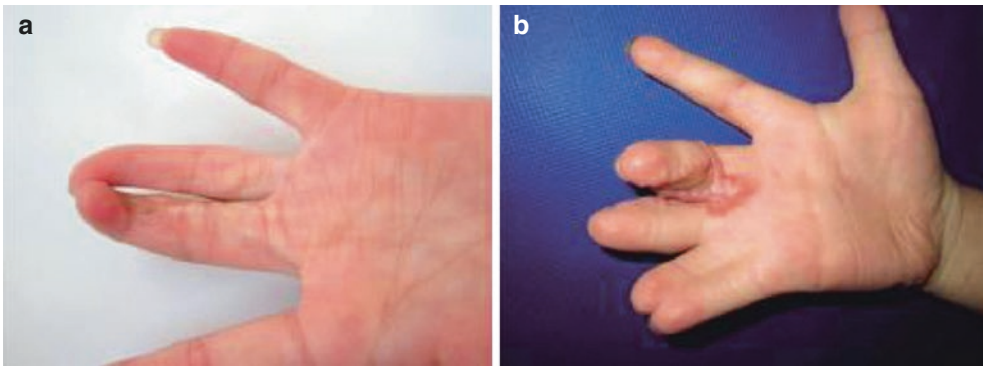


Fig. 6.23 Joint contracture deformity induced by volar cicatrix of interphalangeal joint

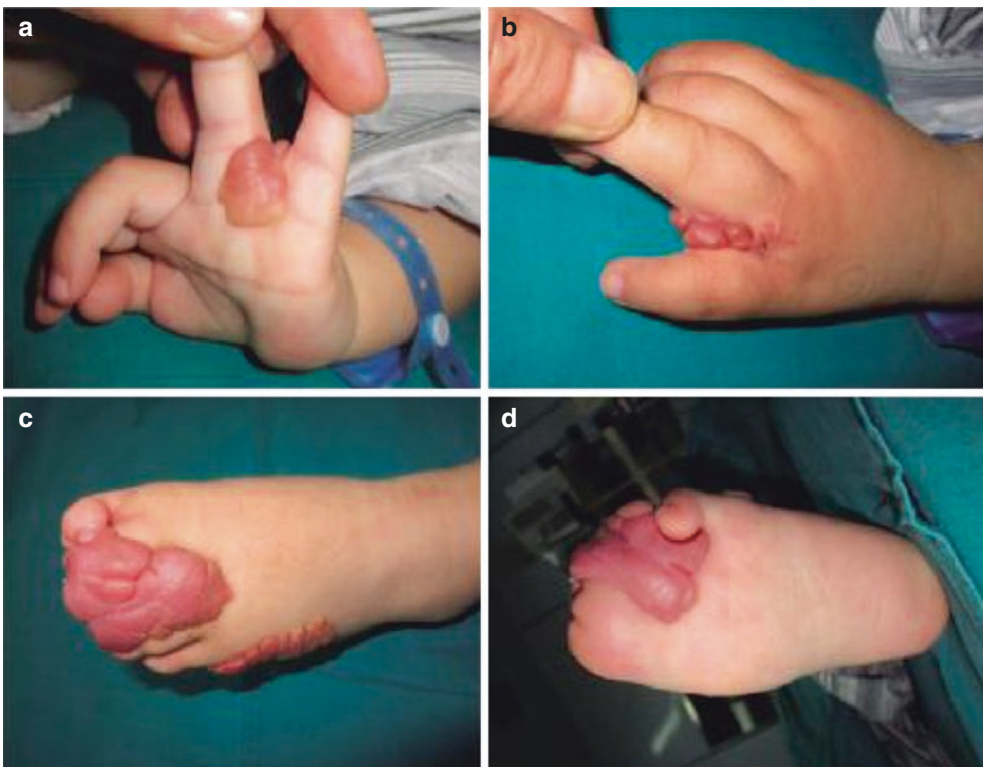


Fig. 6.24 Formation of keloid after dactylolysis

protect the paratenons of the extensor tendons, and then separate the volar flaps and the nerve vascular bundles below them. Separate the syndactylous fingers from far to near while protecting the nerve vascular bundles. Pull out the two fingers. Processing the tissues under the condition that the tensile strength is maintained helps to distinguish the vascular nerve bundles. In proximal anatomy, mark the bifurcations of the vascular nerves. The finger nerves at the distal end can be easily separated when microscope is used. When the arterial bifurcation is far away from the position of reconstructed fingerweb, the processing method is stated as before.

Sometimes, it is necessary to ligate the digital fingers, and the selection of the arteries to be ligated is dependent on whether the adjacent digital arteries are successfully separated. If the bilateral digital arteries of the two fingers are intact, small digital arteries are usually ligated; however, if another surgery needs to be performed on a certain finger (e.g., phase II separation of syndactylous fingers), sometimes it is necessary to ligate large arteries. If the contralateral digital arteries are unclear, the vascular clamps can be used to clamp the digital fingers before the tourniquets are loosened to confirm the blood supply of each finger is reliable. Before the flaps are inserted, fat should be reduced at the adjacent site of the two fingers to reduce the flap tension and improve the overall appearance of the fingers [36]. First, suture the fingerweb flaps, make sure that the distal end joins the proximal finger transverse striation, maintain a 45° tilt angle of the fingerweb, and form a sand clocklike structure [35]. Then use a 5-0 or 6-0 suture line to perform suturing of interdigital triangular flaps, and avoid excessive tension (Fig. 6.25).

Proper ligation is an important part of surgery. In applying dressings, pressure should be provided to the skin grafting

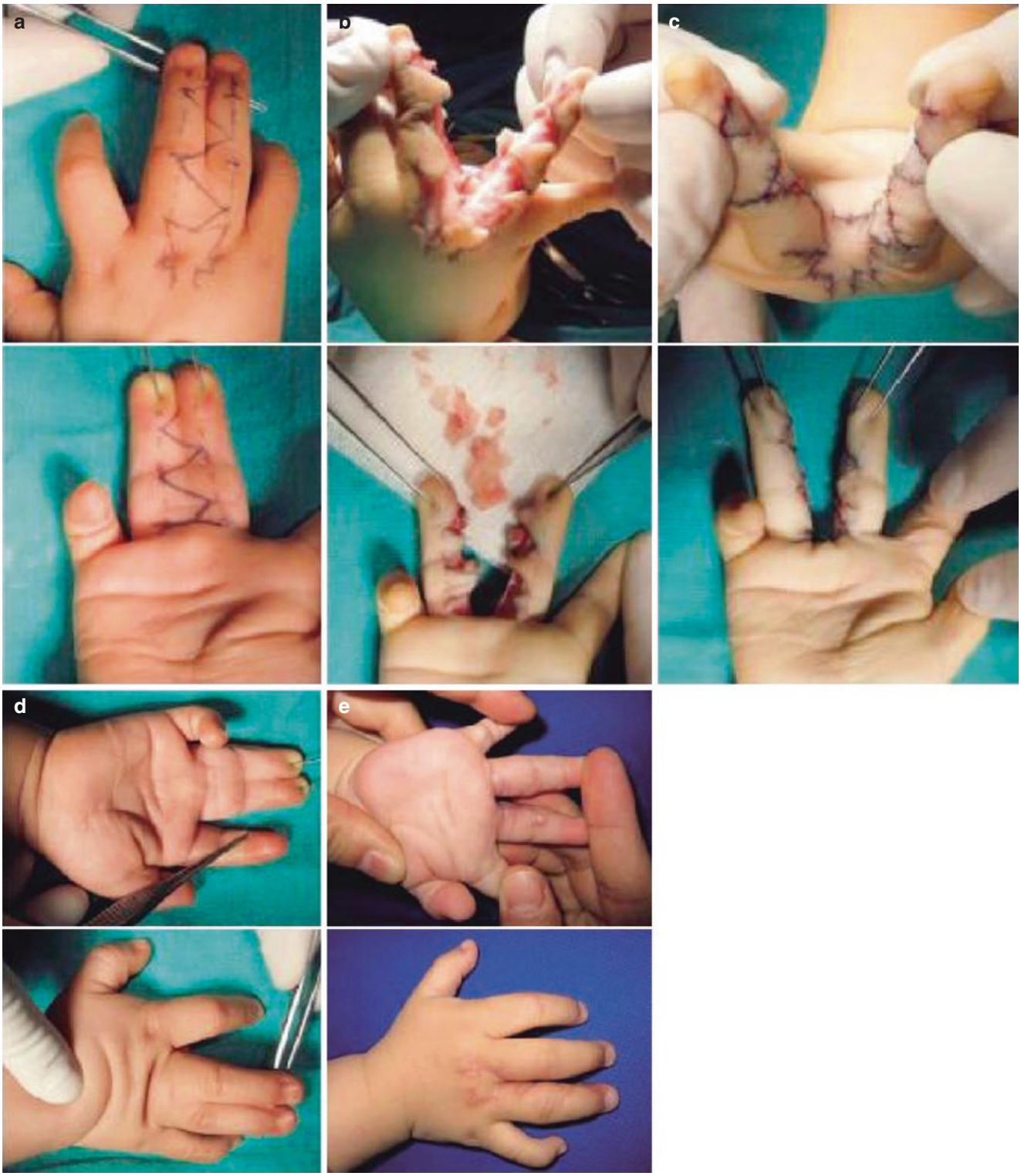


Fig. 6.25 Utilization of dorsolateral flaps to reconstruct fingerweb. (**a**) Surgical design. (**b**) Fine fat reduction with nerve vascular bundles retained. (**c**) Formation of sand clocklike structure of the fingerweb, reconstruction of flexed finger transverse striation, and accurate apposition of triangular flaps. (**d**) Preoperative hand shape. (**e**) Hand shape at 6 months after operation

region, and at the same time, the separated fingers should be protected. The anti-adhesion dressings can be placed to the fingerweb, and large gauze should then be used for ligation and fixation. For young children, the braces that pass the elbow are used for external fixation after pressure dressing to prevent unintentional translocation. Three weeks after operation, remove dressings, change dressings, care for the wounds, and protect them before the wounds become dry and healed. After the dressings are removed, the hands can be normally used. After the wounds become healed, elastic sleeves can be

used for 3 months after wound healing to control the cicatrization. Application of silica gel and injection of anti-adhesion drugs (e.g., triamcinolone acetonide) into the cicatrix can treat the local hyperplastic scar.

6.1.9 Treatment of Special Patients

6.1.9.1 Acrosyndactyly

Acrosyndactyly refers to the fusion of the terminals and the connection of proximal ends between the two fingers, which is one of the characteristics of constriction ring syndrome (amniotic band syndrome) [37]. Fifty percent of the patients suffer a bilateral onset and 50% of the patients are accompanied by hypodactylia. Syndactylia can be manifested as simple syndactylia and can also be manifested as the fusion of multiple fingers at the distal end and combined syndactylia with fingers forming a mass. The interdigital gaps can be large or small (range from the pinhole size to the broad channel), and in most cases they are beyond the fingerwebs (Fig. 6.26). The finger body beyond constriction ring can suffer edema or atrophy.

The treatment method is up to the degree of distal deformities and position and size of the sinus tracts. The mild deformities with well-preserved distal fingers can be separated according to conventional procedures, and the specific methods are stated as before. The sinus tract can be covered or resected in epidermis reconstruction. In case of more complicated deformities, staging operation is suggested. After the fingertip is separated, delay the reconstruction of the juncture. During the staging operation, the fingers can grow without restraint. In case of serious deformities, the first option is removal of the atrophic fingertip because the possibility of its successful splicing with a normal finger is very low. In releasing the syndactylia, the constriction ring can be resected at the same time, and V-Y plasty can be performed.

6.1.9.2 Apert Syndrome

The treatment of hand deformities of Apert syndrome patients must coordinate with the treatment of cranial face deformity and other relevant deformities. Actually, this is a complex surgery, and careful coordination between hand surgery and cranial face surgery is needed. The surgical objective is to complete separation and correction of thumb deformities before the patients become 2 years old so that the hand can develop normally. If the little finger is functional, the metacarpal bone fusion of the ring and little fingers can be surgically released to make the little finger movable. Few surgeries are performed on the shoulder and elbow.

The first task of surgery is to reconstruct sufficiently large thumb web. During the phase I surgery, sequentially release the skin and fascia, lengthen the hand internal muscles, and incise the carpometacarpal joint capsule so that the thumb array is in the abduction 45° position. Regional flaps such as four-flap Z-plasty can be used for mild thumb web stenosis; in case of serious stenosis accompanied by insufficient skin amount, it is necessary to rotationally advance the flaps at the dorsal side or pre-dilate the tissues at the back of the hand. In case of incomplete syndactylia of the thumb and index finger, we tend to make a transposition flap from the radial side of the index finger; the index finger flap can expand the thumb web and is sutured at the thenar patterns to correct any related flexion and adduction contracture deformities. As for the more serious thumb web contracture, Buck-Gramcko [38] once reported the case of application of one large dorsal rotational flap to release contracture. The inguinal and arm medial flaps can serve as the free tissue flaps for transplantation. And contrary to popular belief, the thumb web vessels are enough for microvascular anastomosis; however, this kind of surgery has high requirements on anesthesia, especially when the surgery is simultaneously performed on two hands. Thumb bending deformity must be corrected by phalange osteotomy. Open wedge osteotomy and bone grafting can extend the shortened thumb. It is recommended that the

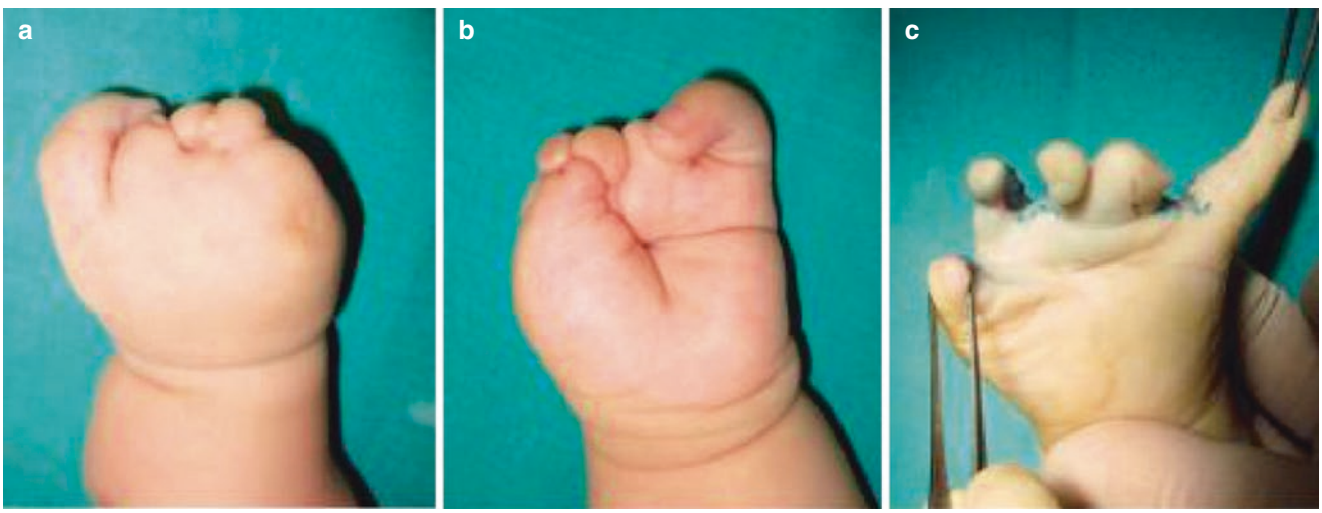


Fig. 6.26 Syndactylia in patients with congenital constriction ring syndrome

osteotomy be completed during the first stage of finger separation, and the surgeries that can be simultaneously performed include thumb web expansion and fixation needed for bone grafting. The bone graft can be taken from separation surgery of the bony synostosis of the fourth and fifth metacarpal bones. Under normal circumstances, the radial side of the thumb will have the problem of insufficient skin, which can be addressed through Z-plasty (Fig. 6.27).

Dactylolysis is generally performed in different stages, and the surgical plan is dependent on thumb web reconstruction and index finger status. Although we wish to retain all fingers, fingers with serious deformity can be given up in case of poor prognosis. Three-dimensional CT can help acquire more information on skeletal deformities, and it can be simultaneously performed in cranial surface imaging.

After the thumb is released, release the distal synostosis to make hoofed hand into a special type of hand such as type I spade hand. This also helps to prevent repetitive nail infection (the commonest hand infection during the early infancy). The subsequent separation of each finger should be performed in different stages, to avoid performing surgery on the two sides of one finger.

As for serious hand deformities, we recommend release in different stages. First, separate the bone fusion at the distal end, make a longitudinal incision at the dorsal side between the two fingers, and separate the nail bed. This process can transform combined syndactylia into simple syndactylia and relieve the fingers from bony constraint. The interdigital defects can be covered by full thick skin graft, and there may be some degrees of motion between the two fingers. Grafting can produce sufficient finger pulp skin, and when finger separation is performed during the late stage, the nail folds can be reconstructed. The fingers of patients with Apert syndrome suffer rigid extension, so linear incisions can be adopted for the separation of the proximal end of the syndactylous fingers because there is no risk of contracture induced by longitudinal incisions made at the interphalangeal joint in other patients. The skin defects are covered by full thick skin graft; due to a huge demand of skin grafting, it is necessary to harvest skin from the lower abdominal region instead of the inguinal region. It should be emphasized that the purpose of fingerweb reconstruction is to make the metacarpophalangeal joint able to move independently and freely. Dorsal skin flaps are commonly used.

In case the little finger is functional, surgical release is needed. The release of bony synostosis of metacarpal bones of the ring and little fingers needs to include fascia or fat implantation to avoid recurrence. The cut bones can be used as the transplanted bone during the correction of thumb bending deformity. Upton suggests that this procedure be performed after the patient becomes 5 years old, which can reduce the recurrence. The position of the fifth digital ray can be improved through releasing the carpometacarpal joint and allowing the metacarpal bone flexion.

6.1.9.3 Symbrachydactyly

The word "symbrachydactyly," originating from Greek, is characterized by shallow fingerweb and short and small finger body and is common in Poland syndrome. In most cases, it has a unilateral onset. The severity ranges from almost entire adactylia to relatively complete brachydactylia. When the finger body is intact, surgical treatment is needed [31]. The surgical mode is cutting superficial transverse metacarpal ligament to increase the length and the range of motion of each finger. The fingerweb should not be reconstructed in the position between the heads of metacarpal bones at the proximal side as V-shaped fingerweb can be shaped.

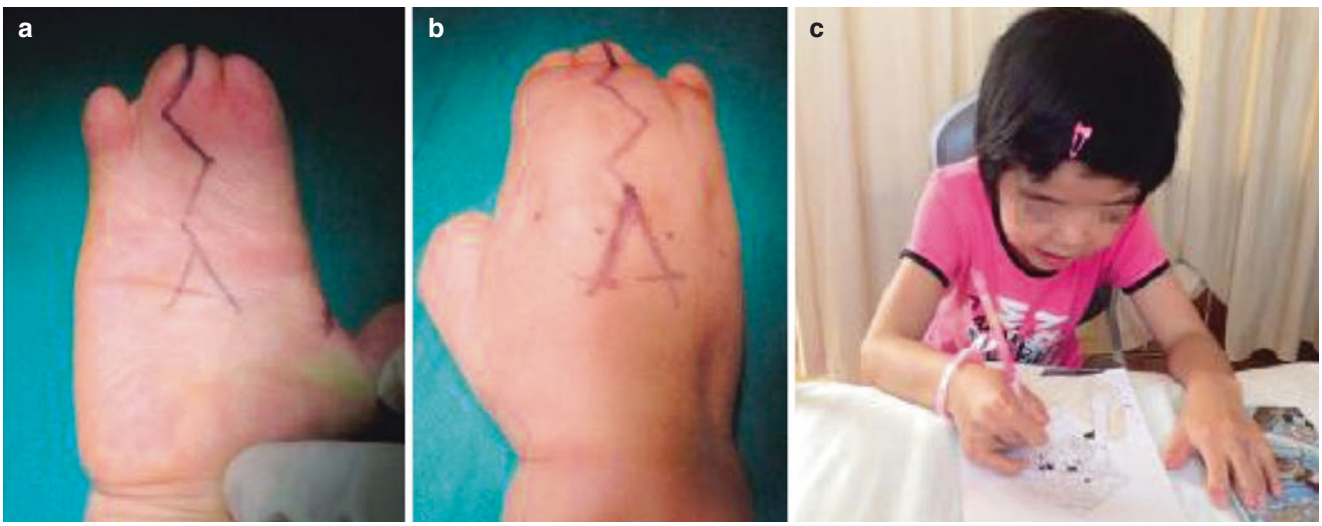


Fig. 6.27 Treatment of hand deformities in Apert syndrome. (**a**, **b**) Preoperative design. (**c**) Postoperative function

The hand deformities of Poland syndrome are various; the commonest involvement lies in the several fingers in the center [39]. As the middle phalange is short, each finger is also short. The syndactylia is simple in most cases and can be complete or incomplete. The aforementioned timing and technique are adopted for the surgical separation. It is combined with the treatment of chest wall malformations (ipsilateral muscle displacement) and the reconstruction of female breast (Fig. 6.28).

6.1.9.4 Dystrophic Epidermolysis Bullosa

The syndactylia of the patients with dystrophic epidermolysis bullosa (EB) does not refer to the congenital deformities in a real sense, and it is induced by the cicatrice on the squamous epithelial surface. EB is a rare congenital blistering disorder in a heterogeneous group, with the structural destruction induced by the loss of adhesiveness between different skin layers. According to the extent and causes, the blistering can be classified: simple bullous epidermolysis bullosa is induced by blistering of the basal keratinocyte layer, junctional epidermolysis bullosa is induced by the blistering of the basement membrane transparent layer, and dystrophic epidermolysis bullosa is caused by a defect of type VII collagen protein in the dermic layer of mastoid process. Dystrophic epidermolysis bullosa is often accompanied by repeated dermic injuries, and the inevitable cicatrix causes syndactylia and contracture. The dystrophic epidermolysis bullosa induced by autosomal dominant inheritance has been publicly known. The dystrophic epidermolysis bullosa induced by autosomal recessive inheritance is more serious. The typical hand deformities evolve into flexion and contracture of the fingertip, making the fingers become adherent into a mass, and the thumbs can become adherent to take a silkworm cocoon shape. Hand problem is only part of the complicated symptoms, so the diagnosis of this disease needs the combined consultations and treatment of multidisciplinary experts, and the specialties involved include dermatologic department, gastroenterologic department, ophthalmologic department, stomatologic department, oncologic department, psychologic department, and anesthesiologic department. The hand surgical operation is based on the separation of syndactylous fingers, release of contracture, and subsequent skin reconstruction. The syndactylia in this situation (called pseudosyndactyly) is generally induced by interdigital adhesion, and dull separation can be performed after the envelop of the scar surface is removed [40].

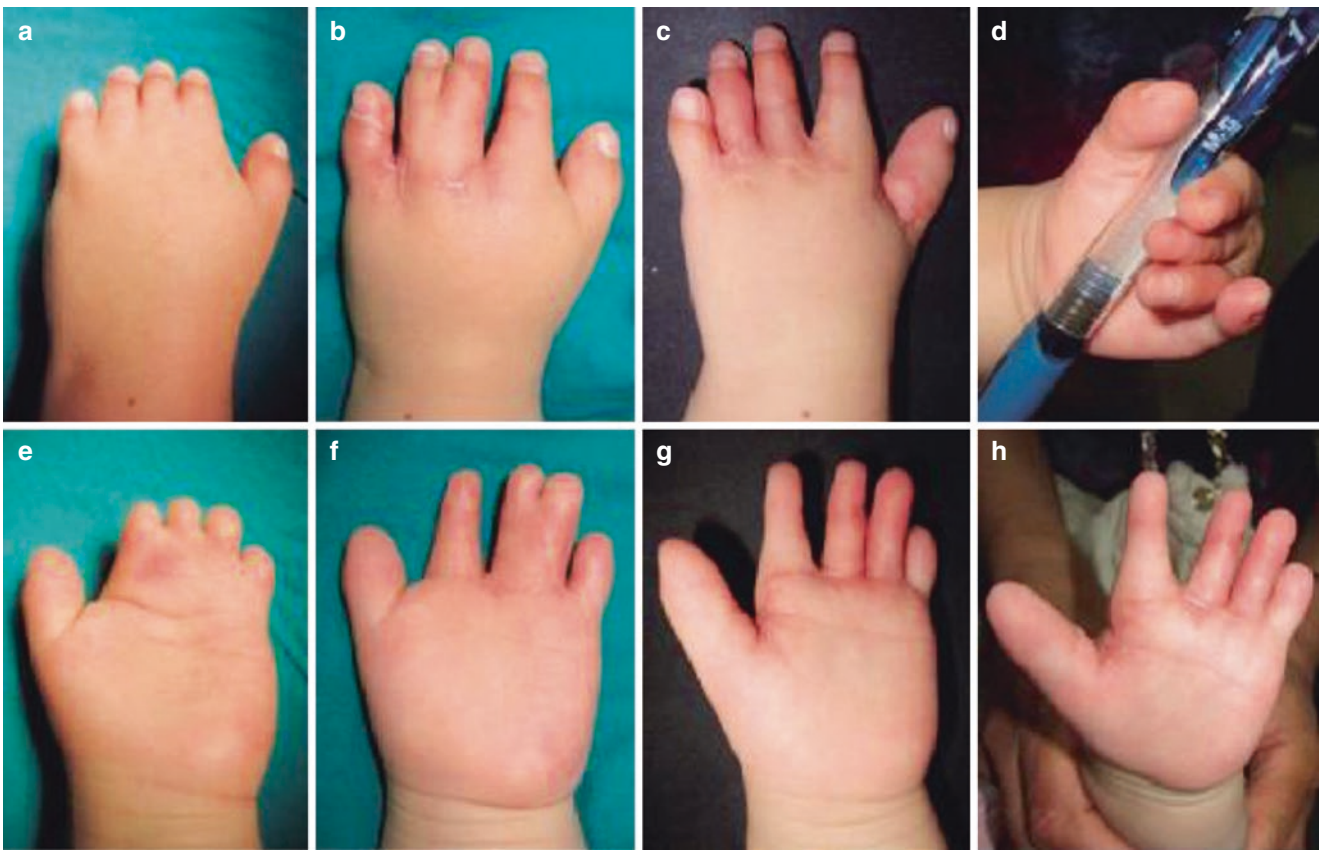


Fig. 6.28 Poland syndrome patients with brachydactylia and syndactylia have the flexed finger transverse striations and fingerweb structures reconstructed through finger separation in different times; as a result, the finger body has a good development basis, and its functions and appearance have significantly improved

Syndactylia will generally lead to finger flexion contracture or thumb web stenosis. We believe that retaining thumb web is the key for the patients to perform hand functions, and skin grafts can be transplanted from the cicatrice-free site (e.g., inguen). Regional flaps are basically unusable because the hands with serious deformities have too many cicatrices. Skin defects may be healed after two phases, by using the skin on the adherent finger body, or by the skin graft of the cut fingers [41]. Although we have successfully used the inguinal free flaps for the reconstruction of vital thumb web, we still want to use the intermediate split-thickness skin graft. The postoperative effects are still satisfying, but the deformity is frequently recurrent, and the probability of re-surgery exceeds 50%.

6.2 Congenital Finger Flexion Deformity

Wei Wang, Bin Wang, and Yunfei Zhang

Congenital finger flexion deformity is a common noninvasive congenital hand deformity characterized by progressive development of flexion contracture of the proximal interphalangeal joint and mostly frequently involves little finger. Tamplin first described this deformity in 1846. When morphologically observed, all finger flexion deformities fall into the category of congenital finger flexion deformity. They can be induced by the primary abnormality in soft tissue structures or distribution and can also be induced by the primary abnormality in bone and joint structures [42].

During recent years, when its relation with inheritance is described in literature, some reports are about the autosomal dominant inheritance, some are about the autosomal recessive inheritance, some are about the chromosome gap absence and chromosome translocation, and some are about the deformities induced by consanguineous mating. The author finds during clinical observation that there is a case in which the patient's maternal grandmother, mother, and daughter suffer from deformities in the final segment of the ring finger or the little finger and there is another case in which the patient's older sister and younger brother concomitantly suffer from oculodentodigital syndrome and finger flexion deformity. Increasingly in-depth genetic study is being conducted on this disease.

6.2.1 Clinical Manifestations and Classification

Congenital finger flexion deformities are manifested as finger flexion and extension functional absence or insufficiency, which frequently occur to the little, middle, and ring fingers. It can independently occur to one finger, is common in the proximal interphalangeal joint, and can also occur to the distal interphalangeal joint.

Clinically, congenital finger flexion deformity is classified into three types (Benson's classification) [43]:

1. Type I (finger flexion deformity during the infancy). The lesions are often limited to one side or two sides of the little finger, and the ring finger and middle finger can also be involved. The incidence in both men and women is similar, which is generally induced by autosomal dominant inheritance of incomplete penetrance (Fig. 6.29).
2. Type II (finger flexion deformity during the adolescence). It is often found during the adolescence. The child's parent may tell the surgeon that the finger flexion deformity is due to the child's fall or due to other "traumas." It is common in women and often occurs to the right hand (Fig. 6.30).
3. Type III (serious finger flexion deformity). It usually involves multiple fingers, often accompanied by other congenital deformities, such as multiple chromosomal diseases, cranial face deformities, and osteoarticular syndrome (Fig. 6.31).

Figuera (1993) reviewed 59 cases of congenital finger flexion deformities. Among them, the number of the patients with type I (finger flexion deformity during the infancy) was 24 and occupied 40.7%; the number of type II (finger flexion deformity during the adolescence) was 5 and occupied 8.5%; the number of type III (serious finger flexion deformity) was 30 and occupied 50.8% [44]. Clinically, the incidence of type III finger flexion deformity may be higher because many syndromic patients concomitantly suffer from finger flexion

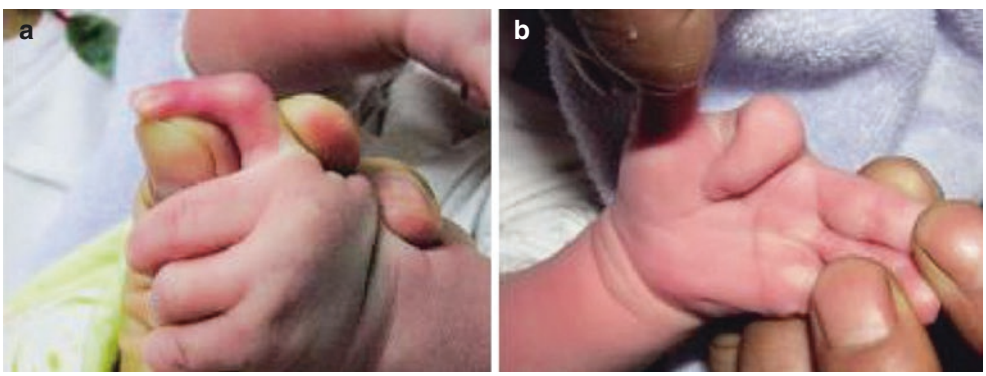


Fig. 6.29 Type I finger flexion deformity (finger flexion deformity during the infancy)

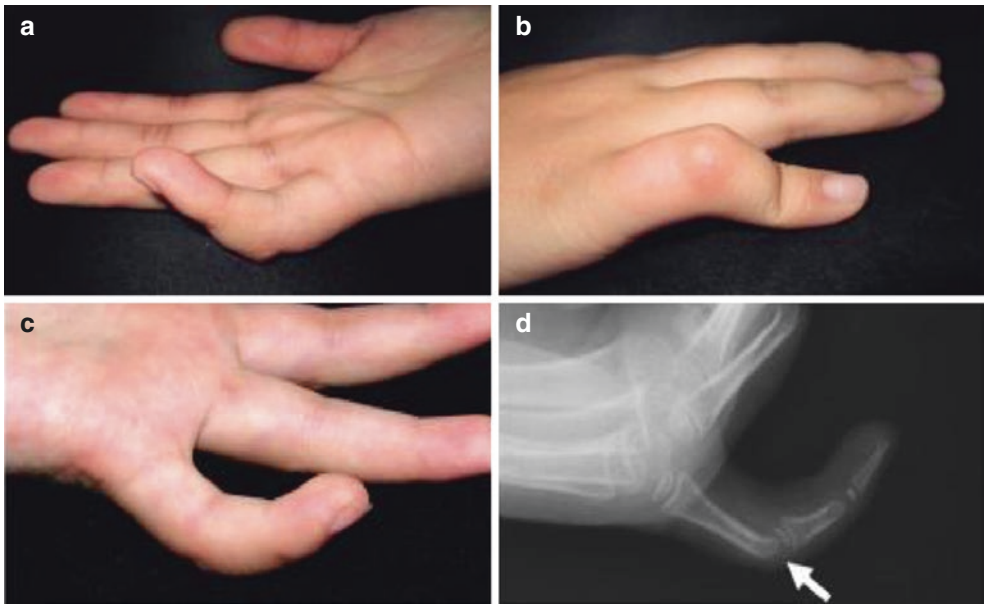


Fig. 6.30 Type II finger flexion deformity (finger flexion deformity during the adolescence). (**a**–**c**) Hand appearance. (**d**) X-ray lateral film indicates that the proximal phalange head of the little finger suffers dysplasia

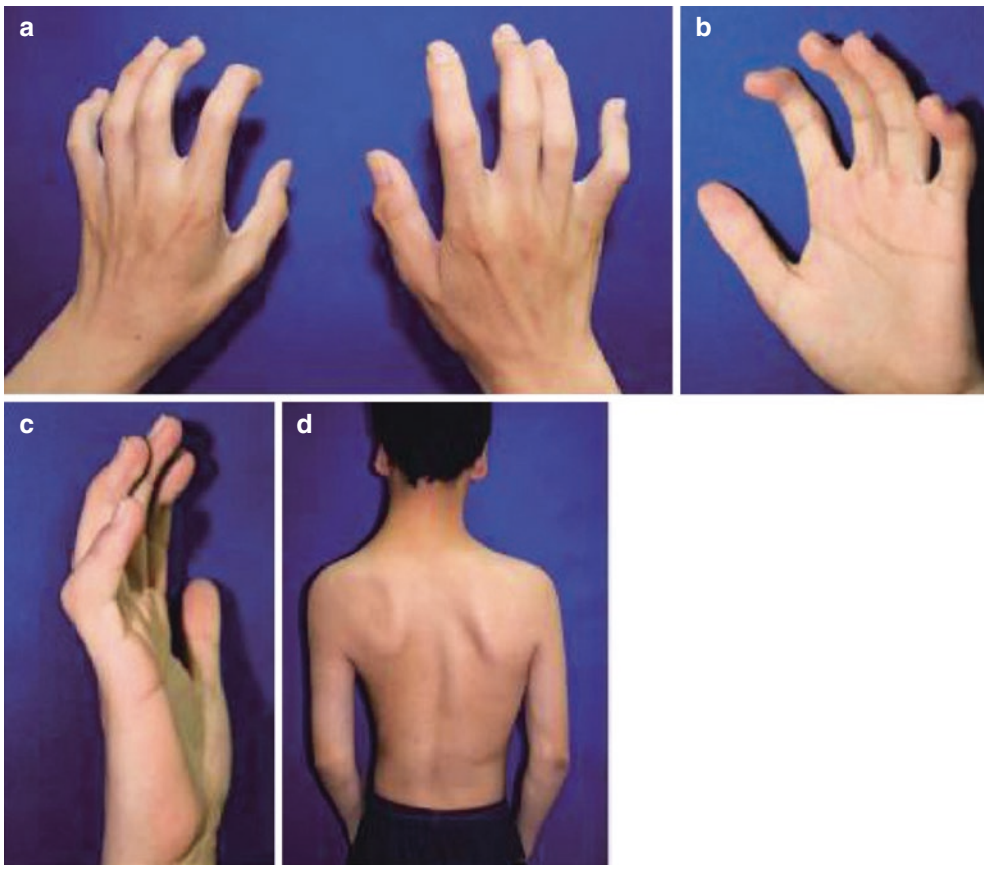


Fig. 6.31 Type III finger flexion deformity (serious finger flexion deformity). (**a**) Multiple finger flexion deformities in bilateral index, middle, ring, and little fingers. (**b**, **c**) The final segment of interphalangeal joint of the left index and little fingers and the proximal interphalangeal joint of the right little finger are serious. (**d**) The patient concomitantly suffers from spinal lateral curvature deformity and auricular dysplasia

deformities. When they visit a doctor, the finger flexion deformities are often covered due to the protruding symptoms of the syndrome.

Many syndromic patients concomitantly suffer from finger flexion deformities, such as orofaciodigital syndrome, with the manifestations of syndactylia and finger flexion deformities, dental deformities, dental dysplasia, enamel dysplasia, etc.; Aarskog's syndrome has the manifestations of finger flexion deformity, microcheiria, ocular hypertelorism, nostril facing forward, wide upper lip, and scrota wrapping the penis like a scarf; the patients with Guadalajara finger flexion syndrome have the history of slow intrauterine growth with the manifestations of dwarf, abnormal facial contour, and finger flexion and skeletal deformities; the patients with Teebi-Shaltout syndrome have the manifestations of cranial surface deformities, hair abnormalities,

finger flexion deformity, unilateral microphthalmos, and renal deformities; cerebrohepatorenal syndrome, a new syndrome reported by Hug A.H. (1997), is accompanied by bone deformity, finger flexion deformity, facial deformity, difficulty in food intake and breath, etc. [45]; the Tel-Hashomer finger flexion syndrome is accompanied by finger flexion deformity, atrial septal defect, and inguinal hernia. Crisponi (1996) reported the 17 neonate cases of bilateral finger flexion within 25 years among 12 family trees of Nansan Santing, and they were accompanied by gnathosasmus, hypersecretion of saliva, facial deformities, and facial muscular spasms. In case of stimulation or crying, the symptoms are obvious; when the infants are quiet, the symptoms gradually disappear [46]. This syndrome has not been named yet, and most pediatric patients die due to high fever within several weeks to several months after birth. Among the 17 patients reported by Crisponi, only 2 survived, 1 of whom was a 14-year-old girl with dysgnosia. In addition, Robinow syndrome is manifested as facial deformity, hollow phocomelia, brachydactylia, finger flexion, duplication of the thumb, clitoral dysplasia, and foot deformity; the CHARGE syndrome reported by Prasad (1997) is often accompanied by finger flexion and lateriflection, tibial excavation, semi-tibial dysplasia, and club foot [47]; Gordon syndrome includes finger flexion, finger lateriflection, cleft palate, disappearance of bilateral hearing, cyrtosis fold, epicanthus, saddle nose, microstomia, enorchia, etc.; ectodermal adactylia cheiloschisis syndrome is also accompanied by finger flexion; the familial fibrous pleural and serosal syndrome reported by Verma (1996) has the manifestations of serious finger and toe flexion deformities; Shprintzen-Goldberg syndrome is manifested as craniostenosis and finger flexion deformity; the Fryns syndrome reported by Stratton (1993) has the deformities of bilateral diaphragmatic muscle defects, duodenal atresia, tracheomalacia, and finger flexion deformities [48]; Schrander-Stumpel (1992) reported one 16-year-old girl with mandibular dysplasia with progressive finger and toe flexion; Van Maldergem (1992) reported the eye-nose-face deformity with finger flexion and lateriflection; Klippel-Trenaunay syndrome is also accompanied by finger flexion deformity and finger deviation deformity, and it is a congenital finger deformity with an obvious genetic predisposition.

In addition to the above syndromes, many patients with congenital hand deformities are also accompanied by finger flexion deformities, such as syndactylia, polydactylism, duplication of the thumb, cleft hand, hand and finger dysplasia, macrodactylia, and windblow hand.

6.2.2 Pathology

The pathological anatomy of congenital finger flexion deformities often includes the following situations:

1. Dysplasia of finger flexor and extensor tendons, which might be induced by congenital shortening in superficial digital flexor tendon and/or profound digital flexor tendon, insertion abnormality, or dysplasia; the finger flexion deformity of such patients disappears in case of flexion of carpal joint; this may be the secondary contracture induced by imbalanced finger flexor tendon development.
2. Abnormal lumbrical muscle, including abnormal lumbrical muscle initial point, for example, the lumbrical muscles become attached to the superficial digital flexor or transversal carpal ligaments; lumbrical muscle atrophy; and abnormal lumbrical muscle insertion, for example, the lumbrical muscle ends at the lateral side of the lumbrical muscle canal.
3. Shortening and abnormality of volar plate.
4. Skin and subcutaneous proliferative ligaments and abnormalities in connective tissues and structures.
5. Basal dysplasia of the head of the proximal phalange or middle phalange. In fact, nearly all relevant structures of proximal interphalangeal joints can be involved, but it is very difficult to ascertain the causes of the abnormality in some structures.

6.2.3 Treatment

Many mild patients only suffer deformities of flexion of the final segment of the finger. In addition to slight defects in appearance, the hand function is almost not affected, and the patient can even normally play an instrument or operate a computer without having to receive any treatment. There are surgical indications only when the flexion contracture of the proximal interphalangeal joint exceeds 60°. Currently the content of the surgical treatment and nonsurgical treatment is summarized as follows:

1. Extension of finger flexion tendon. As for the finger flexion deformity induced by shortened finger deep or superficial flexor tendons, Z-plasty extension can be performed for the digital deep or finger superficial flexor tendons of the forearm; or the functions of the finger superficial flexor tendon can only be retained (only one of the two tendons is retained) to extend the tendons. As for the simple finger superficial flexor tendon (FDS) contracture, the different planes of FDS bifurcate tendon bundles are disjuncted for re-suturing to produce lengthening effects (Fig. 6.32).
2. Resection of abnormal finger superficial flexor tendon. If the superficial digital flexor is abnormally attached to the palmar aponeurosis, flexed finger tendon sheath, or transverse metacarpal ligament, the abnormal points of attachment should be resected to correct the deformities.
3. Displacement of initial point of lumbrical muscle. In case of the finger flexion deformity induced by the lumbrical muscles originating from the superficial digital flexor, the

muscles can be displaced to the finger profound flexor tendon.

4. Grafting of finger superficial flexor tendon of the ring and little fingers. If the flexion deformity of the ring and little fingers is induced by lumbrical muscle dysplasia, the grafting of finger superficial flexor tendon can be adopted to replace the functions of lumbrical muscles.
5. Volar plate release or antedisplacement. Volar plates can be released or antedisplaced to correct finger flexion deformity.
6. Wedge-shaped osteotomy for correction of deformities. In case of the patients with proximal interphalangeal joint flexion who are unable to receive dynamic tendon repair or grafting, wedge osteotomy of proximal phalange can be performed to correct finger flexion deformity.
7. Regional flap grafting after the release of soft tissue contracted skin. Due to the different degrees of interphalangeal joint contracture, the palmar skin often has varying degrees of defects after releasing surgery, and the author utilizes the finger lateral flap grafting method and properly addresses this problem (Fig. 6.33).

Clinical evaluation of postoperative functions is mainly performed on the congenital finger flexion deformity according to the range of motion of the proximal interphalangeal joint (PIP) (Siegert method [49]) (Table 6.3).

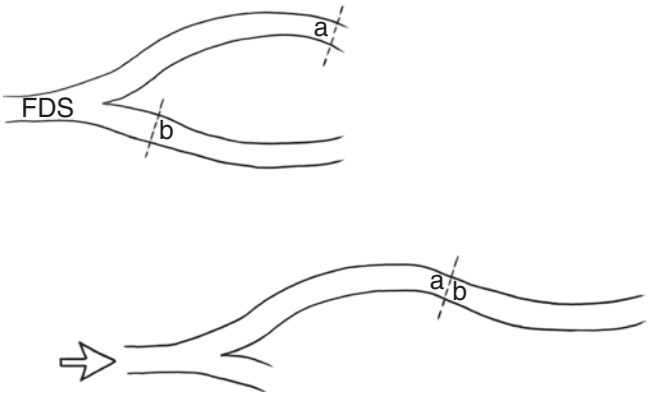


Fig. 6.32 In case of contracture of finger superficial flexor tendon, the different planes of FDS bifurcate tendon bundles are disjuncted for re-suturing to produce lengthening effects

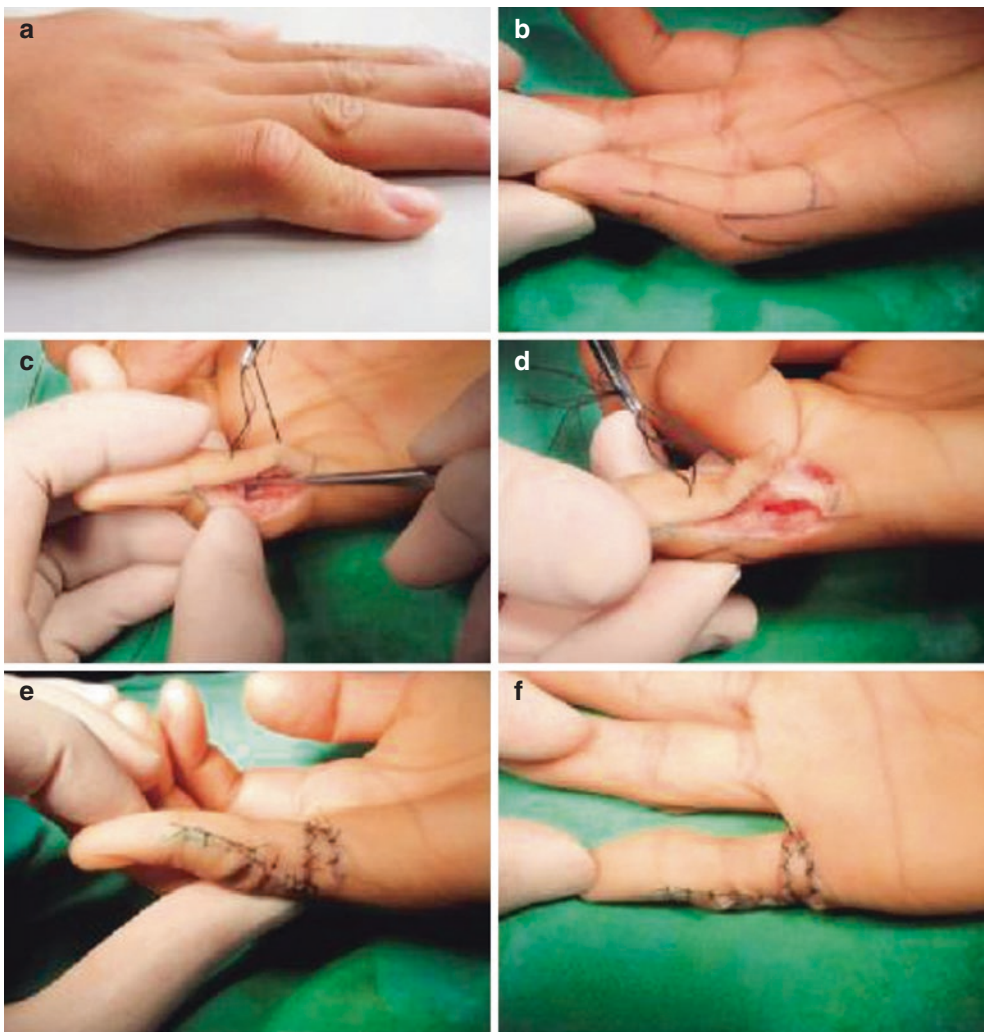


Fig. 6.33 The surgical methods for finger flexion deformity recommended by the author. (**a**) Hand appearance of type II finger flexion deformity. (**b**) Design linguiform regional flap at the affected side of fingers. (**c**) Intraoperative release of volar plates. (**d**) Appearance of soft tissue defects after joint reduction. (**e**) Rotate the linguiform flaps by 90° and then insert them into the soft tissue defects. (**f**) Postoperative appearance of the volar side of the affected finger

Table 6.3 Postoperative functional evaluation of congenital finger flexion deformity

Result	Manifestations
Excellent	No pandiculation of PIP, loss of range of flexion <15°
Good	Pandiculation of PIP <20°, improvement of range of flexion >40°, and loss of range of flexion <30°
Medium	Pandiculation of PIP <40°, improvement of range of flexion >20°, and loss of range of flexion <45°
Poor	Improvement of range of extension of PIP >20°, total range of motion <40°

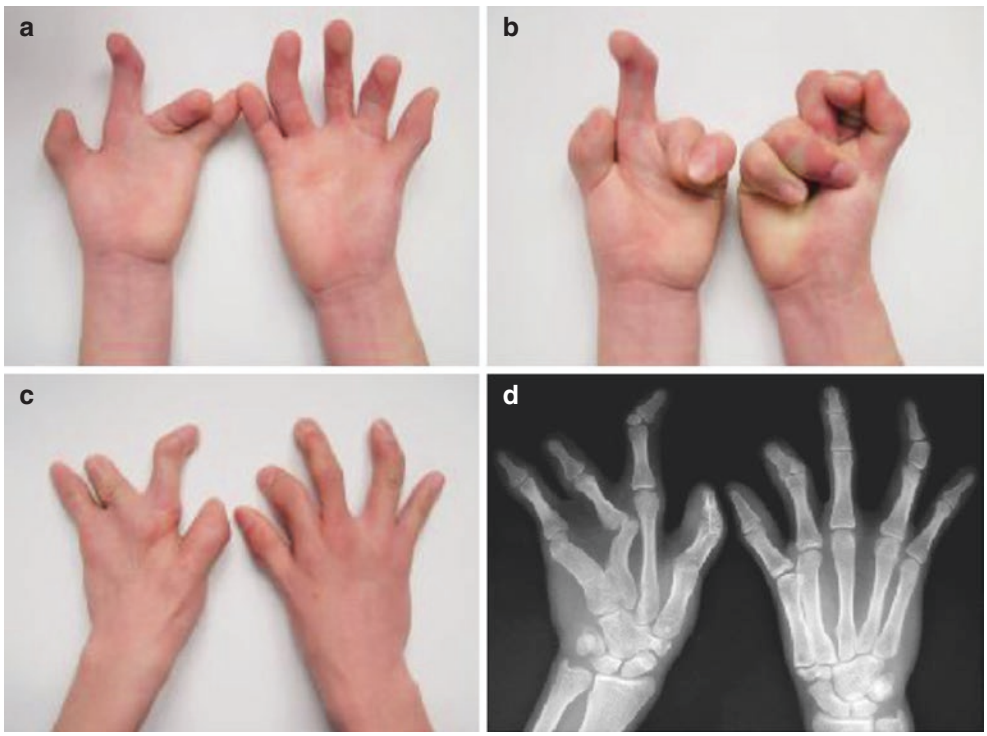


Fig. 6.34 Bilateral multiple deformities of finger deviation. (**a**–**c**) Hand appearance. (**d**) X-ray findings

8. Nonsurgical treatment. It is also an option. Some report that physical treatment can correct 20% of the congenital finger flexion deformity. Therefore, traction, dynamic or static splint, and finger physiotherapy are still the preferred treatment methods for non-tendon-shortening finger flexion deformity.

6.3 Deformity of Finger Deviation

Bin Wang and Yongkang Jiang

Deformity of finger deviation refers to the angulation deformity of finger and radial-ulnar planes. Compared with the normal shape, slight angulation deformity is common, especially in little fingers. The obvious deformity of finger deviation refers to the deformity with a deviation angle of over 10° mostly because the shape of one or more phalanges takes triangular or polygonal shape so that the corresponding interphalangeal joint seriations deviate from the normal axial line of the fingers. This dissymmetric longitudinal growth leads to the non-normal shape of fingers (Fig. 6.34) [50].

6.3.1 Epidemiology and Clinical Manifestations

The reports on the incidence of deformity of finger deviation are various, and the rate of different fingers ranges from 1% to 19.5%. This deformity is often manifested as finger deviation because the middle phalange takes triangular or polygonal shape. The deformity of finger deviation is mostly induced by autosomal dominant inheritance and often has a bilateral onset, common in males (Fig. 6.35). Deformity of finger deviation can be part of many syndromes and compound hand deformities.

Serious finger deviation can be found in neonate period and infancy, and in most cases it can be found when the angulation deformity becomes more obvious with growth and development. Because compensatory abduction of fingers will seriously impair their flexion function, surgical intervention is usually required. The deviation deformity of the thumb or middle finger can lead to functional impairment such as changes in finger flexion or limited pinching functions.

The radiological manifestations are different according to patient age and degree of bone involvement. Among the

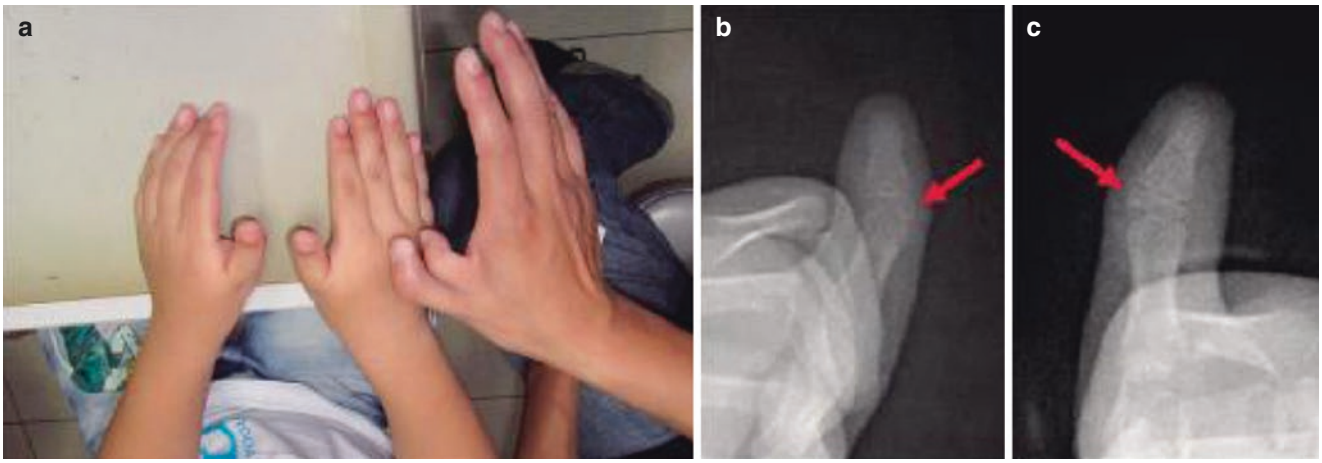


Fig. 6.35 Bilateral deformities of thumb deviation. (**a**) The child's father suffers thumb deviation and polydactylism. (**b**, **c**) X-ray film indicates heterauxesis of the thumb delta phalange and epiphyseal plates

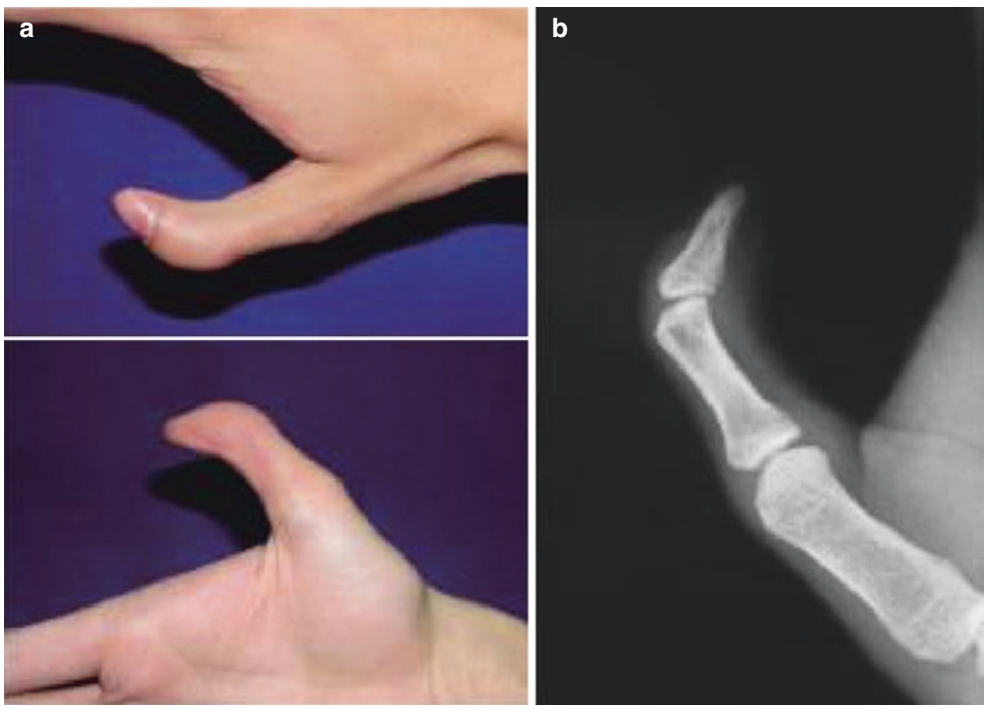


Fig. 6.36 Deformity of right thumb deviation (phalangette articular surface inclines obviously and the articular surface is located at the ulnar side)

patients with complete skeletal development, obvious triangular or polygonal bones can be found, and in some cases, the manifestation can be the deviation of interphalangeal articular surface (Fig. 6.36); among the patients with incomplete skeletal development, morphological abnormality of secondary ossification center can be seen, and it becomes overlapped with the basal part of the phalange and can run along the shortened margin of bones. The most serious shape is the parentheses-shaped epiphyseal plates formed by longitudinal C-shaped epiphysis, which connects the proximal and distal growth centers (Fig. 6.37).

The deformity of finger deviation can be classified according to etiology or pathology, and the most practical one is the classification method proposed by Cooney (Table 6.4) [51].

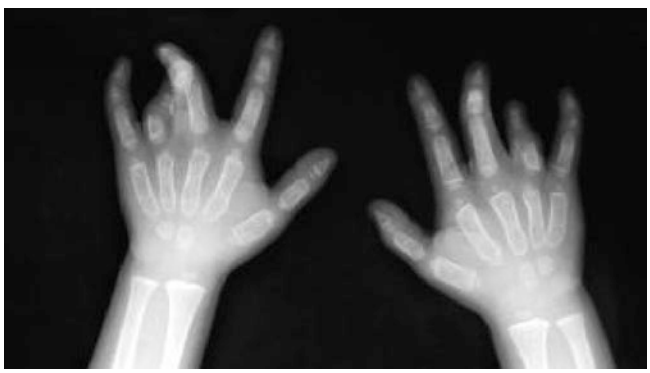


Fig. 6.37 Bilateral deformities of ring finger deviation (the proximal phalange presents bracket-shaped epiphyseal plates)

Table 6.4 Cooney classification of finger deviation deformity

Type	Deformity site	Angularity	Relevant deformities
Single type	Middle phalange	<45°	
Single complex type	Middle phalange	>45°	
Compound type	Bones and soft tissues	<45°	Syndactylia
Compound and complicated type	Bones and soft tissues	>45°	Polydactylism or macrodactylia

Originate from Cooney WP: Camptodactyly and clinodactyly. In Carter P (ed): Reconstruction of the Child's Hand. Philadelphia, Lea & Febiger, 1991

6.3.2 Pathology

The longitudinal growth of tubular bones such as phalange originates from the epiphyseal growth plate, and their axial arrangement relies on the endogenous and exogenous factors to act on the growth plates for maintenance. The growth plate abnormalities secondary to congenital deformities or induced by postnatal injuries will cause further deformations with the growth and development. The growth plate abnormality correlated with finger deviation deformity is the parentheses-like structures formed by the C-shaped epiphysis extending along one side of the bones so that the longitudinal growth at this side of bones is limited. Jones [52] first described the pathology of this disease, namely, the presence of triangular or delta phalange in serious finger deviation deformity. The longitudinal parentheses-shaped epiphysis, severity, and classification system are pathologically described. When the arch-shaped epiphysis is completely ossified during the early stage, longitudinal growth can completely disappear, and short triangular bones and delta phalange can be produced; if the parentheses-shaped epiphysis is incomplete or cartilaginous, there can be partial longitudinal growth, and the phalange is polygonal. The parentheses-shaped epiphysis often affects the middle phalange because it is the phalange that is finally ossified. The parentheses-shaped epiphysis can also appear in the extra phalange among the proximal phalange or the polyphalangeal bones. There may be more complicated deformities, e.g., the dual delta phalanges in specific malformation syndromes such as Rubinstein-Taybi syndrome and Cenani-Lenz syndrome [53]. The acquired etiologic factors of finger deviation deformity include trauma, cold injury, and inflammation, and the concomitant dissymmetric epiphyseal arrest, etc. can lead to injuries in growth plates. Bone tumor can also induce secondary angulation deformity. Although this deformity is often correlated with asymmetric deformity of articular surface induced by juxta-articular tumor, it does not affect the growth plates.

Although skeletal deformity induces the angulation of fingers, the potential soft tissues will also be affected. Serious angulation deformity can lead to shortening of finger concavity soft tissues and possible displacement of extensor tendon structures. In surgery, the factors of this soft tissue deformation need to be considered.

6.3.3 Treatment

Most patients with simple finger deviation deformity visit the hospital for aesthetical need instead of function improvement. For these patients, surgery should be avoided because any improvement in the appearance will subject the patients to the risk of scars and ankylosis. The indication of surgical correction is serious finger deviation accompanied by shortening and angulation, especially when the radial finger or the thumb is involved and the angulating deformity affects the pinching functions.

Application of splint therapy to finger deviation deformity is meaningless. The correction method of skeletal deformity is either rearrangement of fingers through osteotomy or resection of longitudinal parentheses-shaped epiphysis to release the longitudinal growth potential of the shortened side of the fingers. As for the rearrangement of finger axial lines, enclosed wedge osteotomy, open wedge osteotomy, or the combination of the two can be applied, and reverse wedge osteotomy can also be adopted. Enclosed wedge osteotomy (Fig. 6.38) is easy and safe. However, in case of shortening of fingers, further shortening of the length may be unacceptable. In this case, open wedge osteotomy or reverse wedge osteotomy can be used to increase or maintain the finger length [54] (Fig. 6.39). Bone grafting is needed for open wedge osteotomy, which is somewhat difficult for children due to the limitation in donor site. Reverse wedge osteotomy is technologically challenging because the phalange of children is fine and small and usually the two sides of the phalange are involved.

Corrective osteotomy is applicable to patients with moderate deviation and functional impairment of the finger. Surgery should be performed after the bones become mature. Otherwise, too much or too little osteotomy is apt to occur, leading to epiphysis injuries. During surgery, the problem of small amount of defects in the skin can be solved through concave Z-plasty. As for serious angulation deformity, merely Z-plasty is insufficient. Flap advancing or flap rotation is needed, and in the meantime extensor tendons can be adjusted to maintain the balance of finger force [55]. As for the three segments of triphalangeal thumb and multiphalangeal fingers, the deformed phalange can be resected, and ligament reconstruction and joint fusion can be performed simultaneously to achieve mechanical stability.

Another method is resection of parentheses-shaped epiphysis and fat transplantation (Fig. 6.40). This surgery is based on the following assumption: within 1–2 years after resection of parentheses-shaped epiphysis, normal epiphysis growth plates will produce the effects of "pursuing" the longitudinal growth

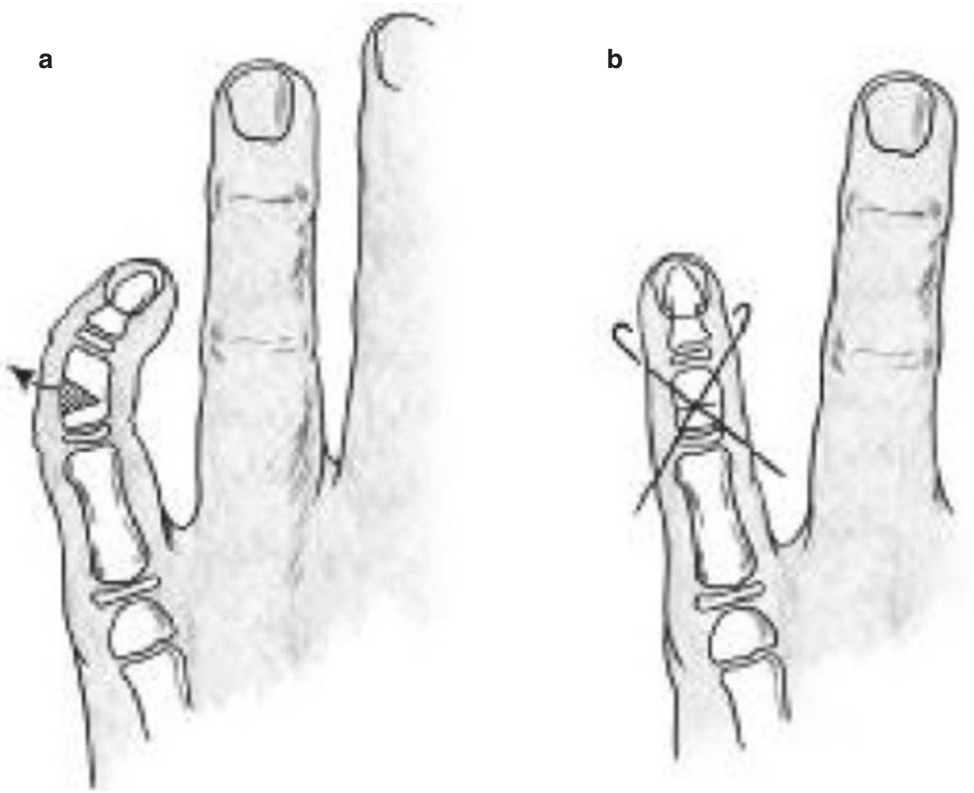


Fig. 6.38 The enclosed wedge osteotomy at the abnormal middle phalange completed at the finger convexity. (**a**) Before operation. (**b**) After operation (Redrawn from Flatt AE: The Care of Congenital Hand Anomalies, St. Louis, CV Mosby, 1977:154–163, with permission. Redrawn by Elizabeth Martin)

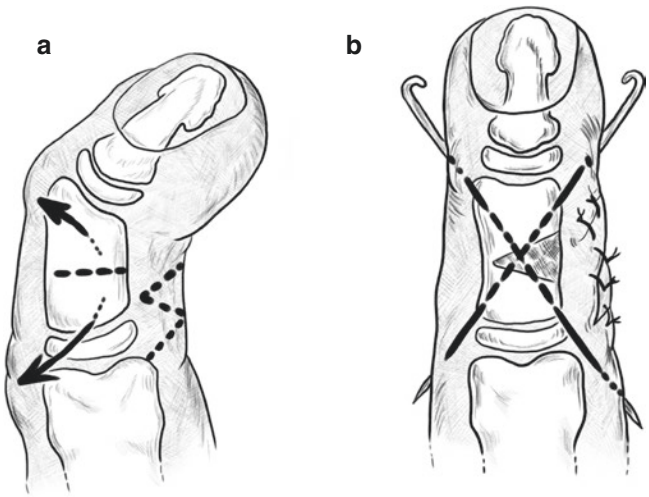


Fig. 6.39 Open wedge osteotomy can help obtain longer fingers, but soft tissue release such as Z-plasty is required. (**a**) Before operation. (**b**) After operation (Redrawn from Flatt AE: The Care of Congenital Anomalies.pp.147–154. CV Mosby, St Louis, 1977)

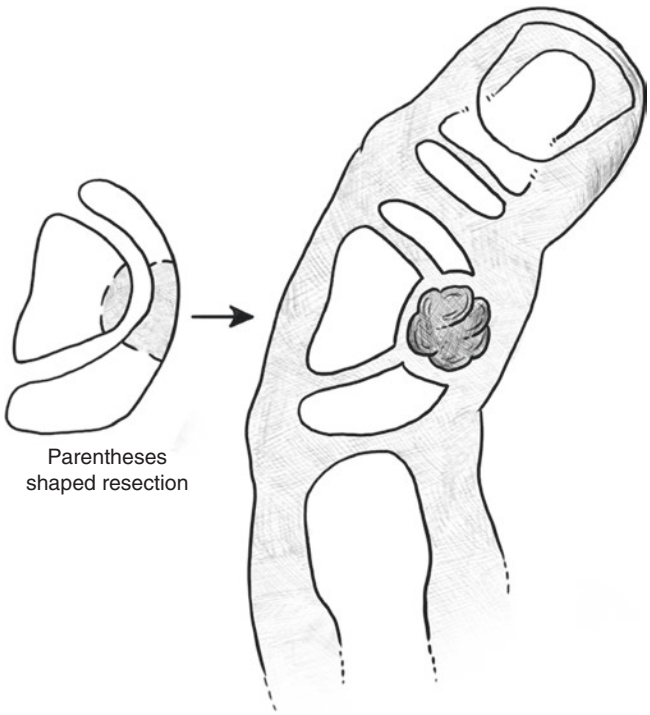


Fig. 6.40 Resection of parentheses-shaped epiphysis and fat transplantation

at the shortened side. The surgery is performed through the lateral median incision at the shortened side of the phalange to resect the cartilaginous or bony parentheses until the bone metaphysis is exposed from the dorsal side to the volar side, and growth plates are clearly exposed. The resulting defects are filled by the fat from the forearm, the ineffective cavity is enclosed, and the trans-growth plate fusion is prevented. This surgery is only limited to the patients with open growth plates, and early surgery is advocated. Vickers performed this surgery

on pediatric patients with remaining 1–2 years of epiphysis growth and obtained good effects [56]. In the report about this surgery [50], some people performed surgery on the 35 fingers of 23 children. After a 3.2-year follow-up visit on average, it was found that a correction of 11° on average was achieved. Further analysis indicates that performing of surgery before the patient becomes 6 years old can achieve better effects. In addition, when a comparison is made between the finger with an angulation of over 40° and the finger with an angulation of less than 40°, the former can make greater improvement (the former makes an improvement of 20°, and the latter makes an improvement of 7.5°). The resection of parentheses-shaped epiphysis does not impede the application of corrective osteotomy on the remaining angulations, and in the meantime, small splints need to be used for fixation or postoperative braking.

6.3.4 The Author's Technology

As for mild to moderate finger deviation, we suggest that surgery be avoided; especially for ordinary mild deformity of little finger, surgery should be abandoned. When the angulation is high and the phalange takes the delta shape, resection of parentheses-shaped epiphysis and fat transplantation can be performed during the early stage. When the patient's angulation is serious and the bones have become mature, the selection of surgical methods can be based on the phalangeal shape. The polygonal phalange can tolerate the shortening induced by enclosed wedge osteotomy, and the triangular phalange is more applicable to open wedge osteotomy. After osteotomy is used to correct the deformity, the axial direction Kirschner silk is used for fixation. The soft tissue defects that correspond to serious finger deviation deformity can be repaired through the dual pedicle skin flaps described by Evans and James [55]. As for the fingers with small triangular phalange, the technical requirements on reverse wedge osteotomy are high; therefore, simple osteotomy with bone transplantation can be selected. As for thumb deviation, thorough osteotomy is needed for correction to form thumb seriations with good alignment and gradual correction after parentheses-shaped epiphysis resection cannot be relied on (Fig. 6.41).

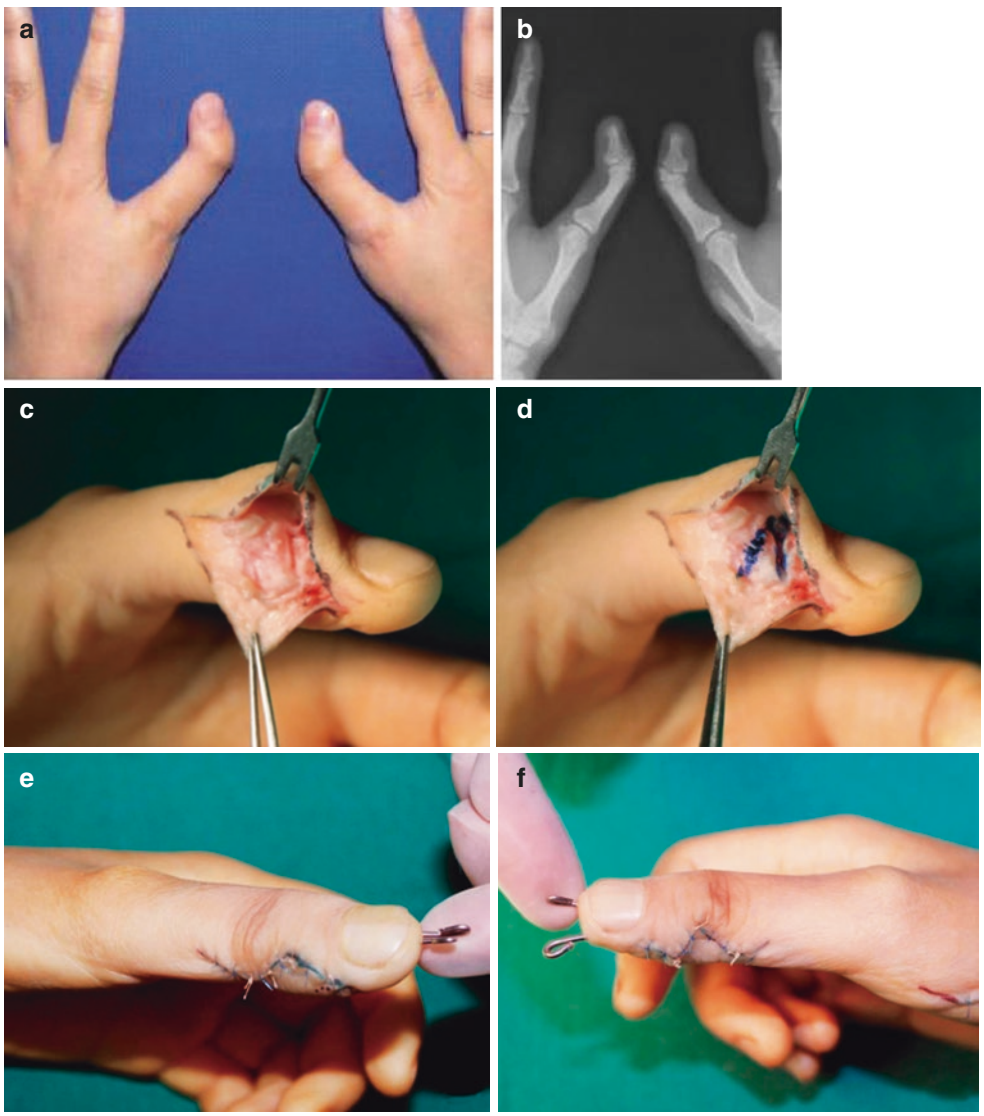
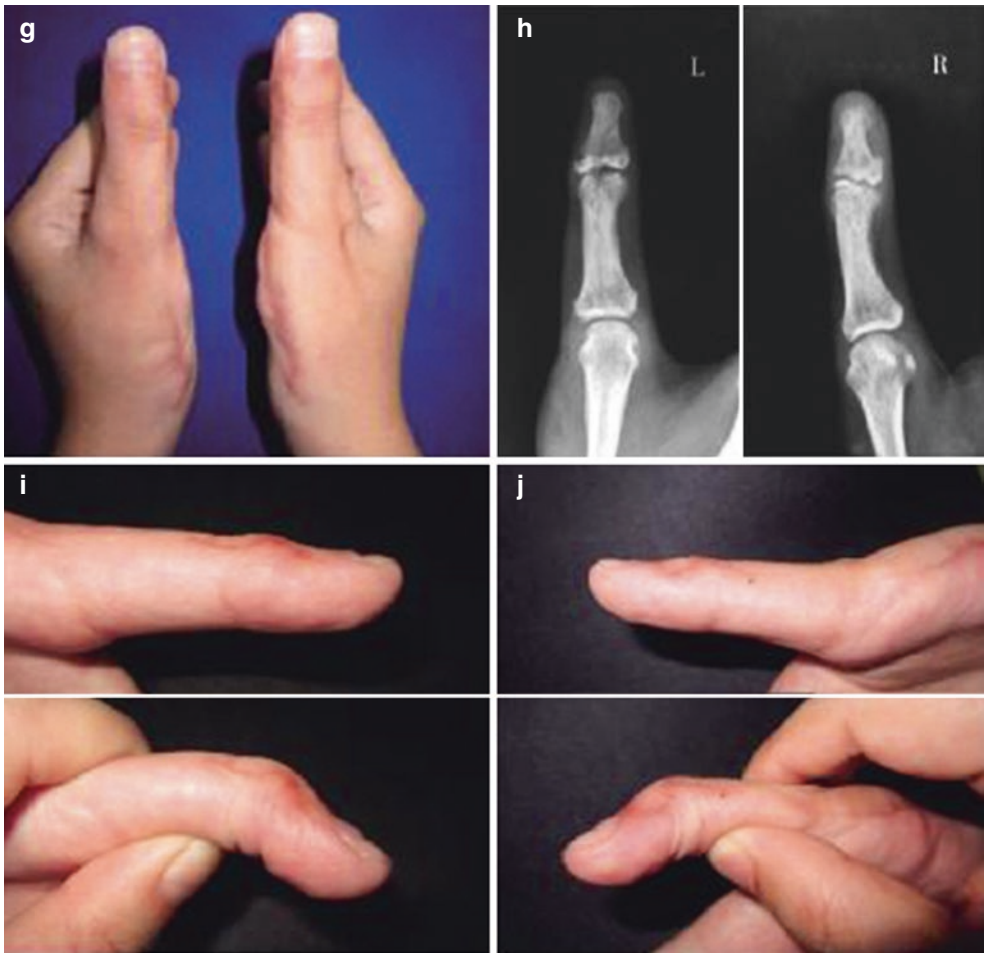


Fig. 6.41 Open wedge osteotomy treatment for bilateral deformities of thumb deviation. (**a**) Preoperative hand appearance. (**b**) X-ray indicates the presence of delta phalange. (**c**, **d**) Perform the sawtooth-shaped incision at the convex side, retain the normal interphalangeal articular surface, and perform wedge osteotomy to resect the delta phalange and phalangette articular surface. (**e**, **f**) Postoperative use of Kirschner silk for fixation. (**g**) Hand appearance at 4 months after operation. (**h**) X-ray indicates that the delta phalangeal and distal phalangeal articular surfaces have been fused. (**i**, **j**) The bilateral thumb interphalangeal joints have good flexion and extension functions.

Fig. 6.41 (continued)



6.4 Joint Contracture

Jinghong Xu, Biwei Zeng, and Jialiang Chen

Arthrogryposis multiplex congenita (AMC), also called congenital arthrokleisis or congenital amyoplasia, refers to the clinical syndrome in which multiple joints suffer contracture and muscular atrophy after birth and different positions suffer joint rigidity due to little movement of the fetus. It can invade partial joints of the four limbs and can also involve all the joints, but it rarely invades the vertebrae [57]. This disease is clinically rare, and the domestic and foreign data collected in 1985 indicated that its incidence was about 1/3000. During 5 weeks and a half of the embryonic period, the intermediate lobes start to develop into joints; at 7 weeks, many articular cavities appear; at 8 weeks, the limbs can move; therefore, the development of joints and the adjacent tissues is very important to the early joint development and movement.

6.4.1 Anatomy

6.4.1.1 Metacarpophalangeal Joint

There are a total of five metacarpophalangeal joints, which are the ball socket-shaped joints composed of capitulum ossis metacarpalis and proximal phalange and can make the movements of flexion and extension, adduction and abduction, and rotation, but the thumb metacarpophalangeal joint is a hinge joint and can only make flexion and extension movements. The volar ligaments and accessory ligaments around the joint capsules become enhanced. Volar ligament, also called volar plate, is a fibrous chondral plate whose connection with the metacarpal bones is loose but whose connection with the first phalange is tight. In movement, the baffle plates are located in front of the head of metacarpal bones. The second to fifth heads of metacarpal bones are connected via the volar deep transverse ligaments (Fig. 6.42).

6.4.1.2 Interphalangeal Joint

The interphalangeal joint consists of proximal phalange head and middle phalange fundus and middle phalange head and distal phalange fundus. There are a total of nine joints, and they fall into the category of axial trochlear joints and can only make the movements of flexion and extension. The joint cavity is broad, the joint capsule is flaccid and weak, and volar ligaments (volar plates) and accessory ligaments around the joint capsules become enhanced (Fig. 6.43).

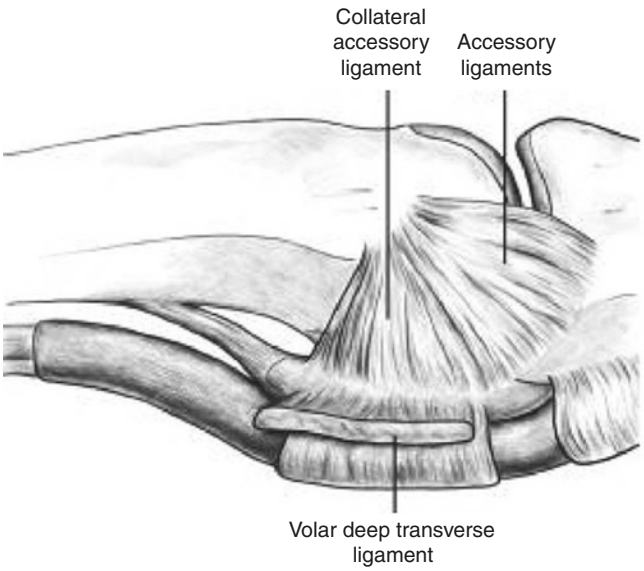


Fig. 6.42 Metacarpophalangeal joint (lateral view)

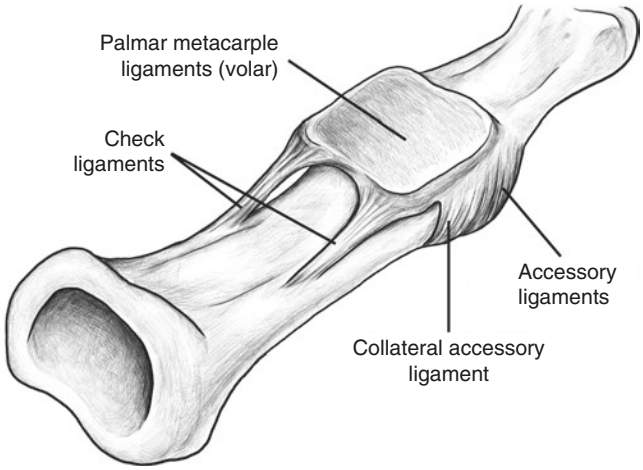


Fig. 6.43 Interphalangeal joint (appearance of palmar surface)

6.4.1.3 Palmar Metacarpal Ligaments (Volar Plates)

The lateral side of the proximal volar plates has a close connection with the proximal phalange, and it is adjacent to the A2 and C1 trochleas to constitute the check ligament. Under the pathological status, volar plates can impede the extension of the interphalangeal joints, which is induced by the thickening of the check ligaments. The thickened check ligament, called checkrein ligament, will impede the deviation of the volar plates and further limit joint movement (Fig. 6.44).

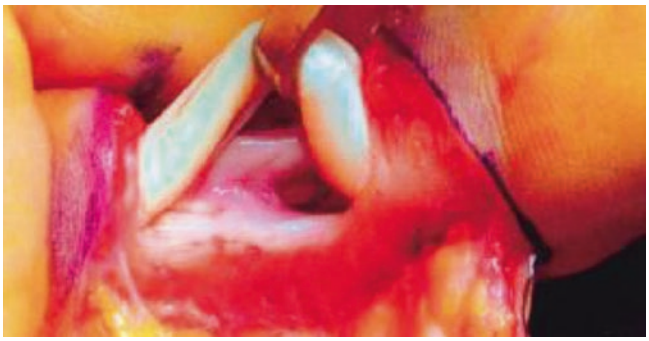


Fig. 6.44 Checkrein ligament secondary to the flexion deformity of the proximal interphalangeal joints

6.4.2 Etiologic Factor

The exact etiologic factors of this disease are unknown. Generally, the etiologic factors of congenital joint contracture include the following:

6.4.2.1 Spinal Nerve Abnormality

This is the main reason for joint contracture, such as meninx bulge, dysplasia of spinal cord anterior horn cells, prenatal spasm, and cerebral tissue defects (e.g., anencephalia, hydrocephalus, and holoprosencephaly).

6.4.2.2 Muscular Abnormality

Muscular abnormalities are mostly muscular hypoplasia, rarely fetal myopathy, and occasionally muscle tension malnutrition.

6.4.2.3 Joint and Adjacent Tissue Abnormalities

The main joint and adjacent tissue abnormalities include bone fusion, joint dysplasia, and joint peripheral soft tissue contracture.

6.4.2.4 Trauma

Trauma is common in fetal crowding and compression and soft tissue injuries, such as injuries in volar plate, accessory ligaments, flexor and extensor tendon sheath, skin, nerves, and arteries.

6.4.2.5 Viral Infection

Animal experiments show that, 2 days after chicken embryos are given ourari, the chickens can suffer similar multi-joint contracture with humans. In addition, after the chicken embryos are infected with coxsackie virus and Newcastle disease virus, multi-joint contracture can be formed, which

indicates that the viruses have certain effects on the occurrence of this disease.

6.4.2.6 Genetic Factor

This disease is sporadic, and there are individual cases on autosomal dominant and recessive inheritance as well as sex-linked inheritance.

6.4.3 Pathology

6.4.3.1 Pathological Classification

It is pathologically classified into two types, namely, muscular type and nervous type. The muscular type is manifested as muscular paleness, hard texture and fibrosis, normal spinal cords, anterior horns, and nerve roots, and this type accounts for 7%. The spinal cords, anterior horns, and nerve roots in the nerve type are abnormal, and this type accounts for 93%.

6.4.3.2 Pathological Characteristic

1. Muscular tissues. They take the shape of a mass of fibro-fatty tissues, with fatty or degenerative muscle fibers messily distributed in the middle, and the transverse diameter of the muscular fiber decreases. This change is similar to infantile muscular dystrophy, so it is also called fetal muscular dystrophy.
2. Nerve tissues. The main manifestations are degeneration of spinal cord anterior horn cells of the central nervous system, cell shrinkage, and decrease in cell number, especially drop in the number of nerve roots in front of cervical and lumbar vertebrae, but the nerve roots at the rear are normal. There are also reports on the degeneration of spinal cord posterior horn cells. There are manifestations of pyramidal tract and motor nerve root demyelination, and the number of perineural axons decreases.
3. Soft tissues. The gross appearance of joints is relatively normal, but the joint capsules become thickened and fibrous, the joints lack plica, and the articular cartilage becomes degenerative. The skeletons become thin and deformed; the connection of the skin, subcutaneous tissues, and surface of the bone is tight; and there is adhesion between the tendons and tendon sheath.

6.4.4 Clinical Manifestation

6.4.4.1 Basic Manifestation

There is little fetal movement during the maternal pregnancy, and the position of fetus is improper. The patients with typical symptoms can be diagnosed immediately after birth. The common postures after birth are extension of the knee joint, horse's hoof shape of feet, flexion of the elbow joint and wrist joint, and atrophy to varying extents of both upper and lower limbs. The deformity of feet is the most serious, and their rigidity is more serious than congenital equinovarus. Due to muscular atrophy, athrepsy is often induced. The skin has no normal folds and is nervous without luster, but when the joint is fixed in the flexion position, obvious skin and subcutaneous web-shaped deformities can appear. The joints suffer quasi-fibrous rigidity, the skin at the flexion side is shortened, the normal skin striae disappear, amyoplasia is noted, the active activities of the joint decrease, and there is a small quantity of passive activities without pain. The sensation is normal, but the deep tendon reflex becomes declined or disappears, and the electrical muscle stimulation reactivity is low. The intelligence is normal. Accompanying deformities such as club foot, hip joint dislocation, and knee joint dislocation are common [58–60] (Fig. 6.45).

Fig. 6.45 Congenital multiple arthrogryposis

The patients with typical symptoms can be diagnosed according to the presence of joint deformity found immediately after birth, and the diagnosis cannot be confirmed only if there is special deformity in single joint, congenital hip joint rigidity and dislocation, and difficult-to-correct spinal column lateral curvature. X-ray examinations can show that muscular tissues at the soft tissue level decrease and the subcutaneous fat is relatively thickened, some patients can experience carpal bone and radial-ulnar and talocalcaneal fusion, the joint shadow density becomes thickened, and the development of the femoral head is poor. The child's electrical muscle stimulation reactivity is very low, and electromyogram does not indicate degenerative changes.

6.4.4.2 Clinical Classification

1. Extension contracture. It can involve all of the four limbs or partial joints. The upper limb deformity is manifested as shoulder adduction, elbow extension, forearm pronation, and wrist flexion; the lower limb deformity is manifested as hip extension, abduction and extortion, and deformity of the back knee; deformities such as equinovarus, hip joint dislocation, or whirlbone absence can also be suffered.
2. Flexion contracture. Any joint can present symmetric contracture; the hip joint suffers deformities of abduction, extortion, and flexion; the knee suffers serious flexion contracture; and in severe cases, the heel stick can prop up the buttock.
3. Mixed contracture. This type is the commonest, manifested as elbow, wrist, and hip flexion deformities and knee extension deformity.

6.4.5 Clinical Examination

6.4.5.1 Joint Passive (Active) Flexion and Extension Test

Joint passive (active) flexion and extension test is the most direct and important examination (Fig. 6.46). If the passive movement of the finger is limited in a certain direction, the active movement is also limited in this direction.

6.4.5.2 Internal Muscle Tautness Test

Internal muscle tautness test is used to evaluate the effects of metacarpophalangeal joint movement on the proximal interphalangeal joint (Fig. 6.47). In metacarpophalangeal joint flexion or extension, measure the passive flexion range of the proximal interphalangeal joint; the enlargement of passive flexion range when the metacarpophalangeal joint becomes flexed indicates the tautness of finger internal muscles (internal muscular positive sign).

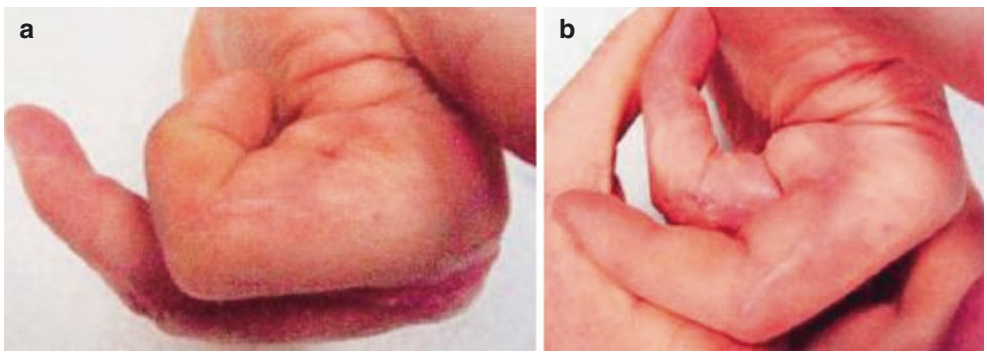


Fig. 6.46 Joint passive (active) flexion and extension test. (**a**) Finger active flexion. (**b**) Finger passive flexion

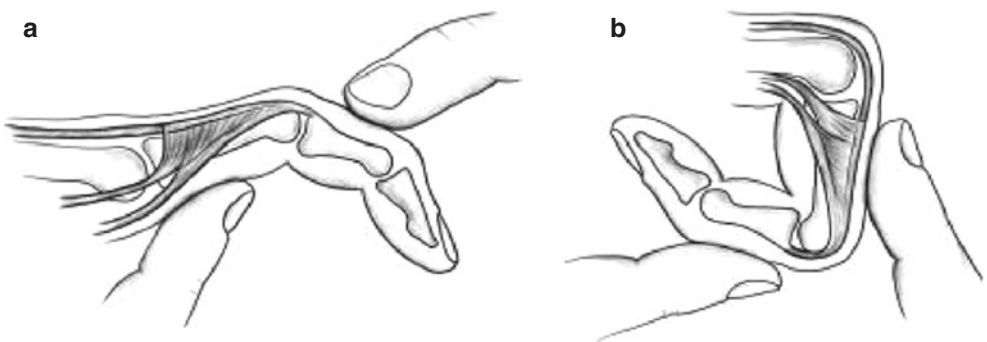


Fig. 6.47 Internal muscle tautness test

6.4.5.3 Examinations of Skin and Subcutaneous Tissues at Diseased Joints

Obvious soft tissue defects can limit the passive movement of fingers, and the solution of the skin problems of involved sites is the prerequisite for reconstruction of the diseased joints.

6.4.5.4 Imaging Examination

Imaging examinations can confirm whether the fracture is healed and whether the involved joint paraposition is accurate, and the examinations rule out osteoarticular lesions; they can also be used to rule out whether there is any bone block that affects joint movement. Exostosis can directly impede joint movement or change tendon movement to further affect joint functions.

6.4.6 Treatment

6.4.6.1 Nonsurgical Therapy

The early stage should focus on nonsurgical therapy, and the treatment objective is to increase joint activities and stabilize joints. Nonsurgical therapies include adoption of traction, splint, plaster, etc. to correct joint deformities. In the meantime, physiotherapy, massage, and medical gymnastics should be combined, and the specific methods vary according to the degree of joint contracture. If the deformity is serious and the muscles have been deformed, then the prognosis is poor.

6.4.6.2 Surgical Treatment

1. Surgical indications: ① Failure in nonsurgical treatment, ② no special surgical contraindications, and ③ patients with skin and joint capsule contracture who need to have function and appearance improved.
2. Basic principles of surgery:
   (a) The preferred treatment of joint contracture deformity induced by hand trauma is to recover normal anatomic structure, stabilize joint structure and fracture, and subsequently repair the ligaments, vessels, and nerves, without tension skin grafting. Before removal and reconstruction of the soft tissue, the anatomic structure of the normal ligaments and vessels must be protected.
   (b) In most cases, surgery should be avoided from being performed on bilateral sides of the fingers simultaneously. In most cases, the surgery is performed at the volar side to avoid injuries at the dorsal side; in this way, postoperative pain and edema can be effectively reduced. The extensor system of some patients has been involved. After the surgery is performed at the volar side, it is still necessary to remove the dorsal ligaments, which, though bring large injuries, will facilitate the recovery of motor functions.
   (c) Long-term flexion contracture will induce finger nerve and vascular contracture. After contracture release, the fingers should be fixed in the largest extension position to make sure that the fingertip has sufficient blood supply.
3. Surgical method:
   (a) Palmar proximal interphalangeal joint release: use the lateral median line of the metacarpophalangeal joint, proximal interphalangeal joint, and distal interphalangeal joint as the tangent line; mark the central part of the tangent line; connect it with the proximal interphalangeal joint and the distal interphalangeal joint, respectively, with a line; and form an angled palmar surface; in this way, a mixed incision is established, which is apt to dilate the flap area (Fig. 6.48).

   Incise the sheath between the A2 and A4 trochleas; remove the C1, A3, and C2 trochleas; retain the A2 and A4 trochleas; and expose the volar plates (Fig. 6.49). With stepped deep penetration, recognize the vascular commuter zone and protect it properly. Recognize the checkrein ligaments at the proximal end of volar plates and perform separation, and release contracture.
   (b) Z-plasty, joint capsule release, and free skin grafting: In the release of contracted skin, try not to expose the tendons in order to be able to receive free skin grafting. After the contracted joint capsule is released, Kirschner silk must be used to fix the joint in the extension position to reduce the postoperative recurrent contracture in joint capsule and free skin grafting area.
   (c) Dorsal proximal interphalangeal joint release: Dorsal contracture is rarer than volar contracture. If the contracture is secondary to the fracture, then the extensor

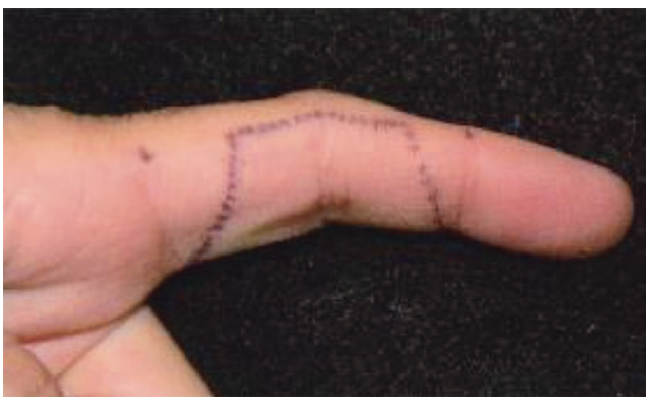


Fig. 6.48 Design of palmar surface incision of the proximal interphalangeal joint

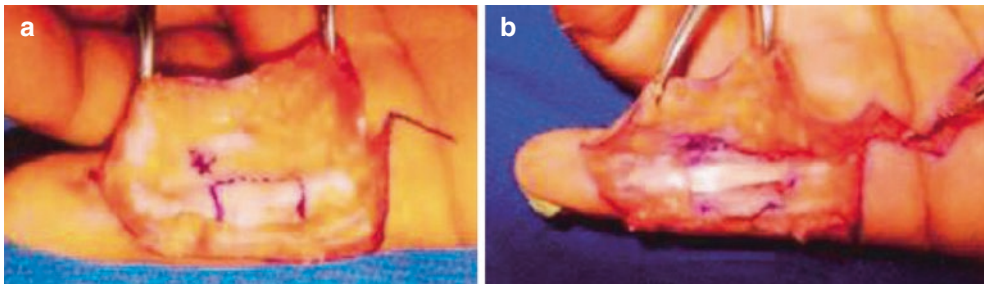


Fig. 6.49 Remove C1, A3, and C2 trochleas, and retain A2 and A4 trochleas

tendon system will experience regional adhesion. In case of the presence of spot-shaped adhesion at the proximal phalange, linear ligament removal can be performed. If passive flexion is still limited, the volar plate capsule should be separated on the basis that the central chordae are properly protected.

(d) Dorsal metacarpophalangeal joint release: The rigidity of metacarpophalangeal joint is common in extension status; the dorsal longitudinal incision is usually used to expose the extensor tendon system of the metacarpophalangeal joint. To fully expose the dorsal capsule, it is usually necessary to move the extensor tendon system away. It is possible to separate the tendons into sagittal stripes or disjunct it (generally this method is adopted on the fingers with two tendons such as the index finger or little finger, but it can also be used in other fingers), and after operation, these tendons can be properly repaired.

(e) Removal of dorsal ligaments and volar ligaments: If necessary, the removal of dorsal ligaments and volar ligaments can be performed.

(f) The use of compass-type hinged external fixator: The compass-type hinged external fixator is used to perform phased correction of proximal interphalangeal joint contracture, and at the same time, the distal interphalangeal joint can also be fixed. After the use of this external fixator, the joint will be pulled out gradually until the formation of the vascular communicating branch. Once the joint is pulled out to the proper length, the fixator can be removed and passive functional exercise can be done.

4. Postoperative care: The key of postoperative care is edema control, pain processing, activities (gentle passive activities), splint fixation (3–5 months), and delayed clenching enhancement. What is the most important in postoperative care is the early hand physiotherapy. If the finger is fixed with splint during the surgery, the physiotherapy can be initiated 3–5 days after operation [61, 62].

Hand physiotherapy is important in two aspects. First, physiotherapy helps to reduce the postoperative edema; second, the use of fixing splint and active and passive recovery training will help to maintain postoperative movement effects.

6.4.7 Prognosis

The complications after the release and ligament resection are not very common, including infection, tendon injuries, and nerve injuries. Careful and gentle skin and soft tissue operation can lower the possibility of infection and flap necrosis to the minimum, and attentive separation and careful recognition of the anatomical variation can reduce the probability of injuries in ligaments, nerves, and vessels.

Excessive correction on the joint flexion contracture is improper because this may sprain the nerve bundles, so the correction should be made in accordance with the patient's age, contracture degree, and the previous presence of nerve sprain.

6.5 Congenital Trigger Finger Deformity

Wei Wang, Wangxiang Yao, Jianmin Yao, and Sheng Ding

Congenital trigger finger deformity (congenital trigger digit), also called congenital thumb or finger thecostegnosis, was first described by Notta (1850). Among all types of congenital trigger finger deformities, trigger thumb deformity is the commonest. Due to the congenital stenosis in thumb flexor tendon or the finger long flexor tendon fibrous sheath wall, namely, the thickening of A1 trochlea and tendon sheath stenosis, the thumb long flexor tendon or the finger long flexor tendon is limited when gliding inside the stenotic tendon sheath, and there is a trigger-like blocking sensation in case of the extension of the thumb or finger metacarpophalangeal joint and interphalangeal joint, so it is called trigger finger. After a long time, the proximal end of the flexor tendons with limited gliding presents the nodular-like structure.

Congenital trigger finger is a relatively common congenital deformity. According to the statistics of Flatt (1977) and Ger (1991), its incidence was 0.5‰. Its incidence reported by China is 0.05–0.3‰, accounting for about 2.2% [63] of all fetal upper limb deformities. In most cases, congenital trigger finger can occur independently; in some cases, the disease is multiple, with both trigger thumb and trigger finger. Among the 41 patients who suffered from congenital trigger finger deformity as reported by Steenwerckx (1996),

33 suffered from trigger thumb (10 suffered bilateral deformity, 10 suffered deformity in the right thumb, and 13 suffered deformity in the left thumb), 7 suffered trigger finger, and 1 suffered multiple trigger fingers. Among the patients reported by Rodgers (1994), 73 children had 89 trigger thumbs, among whom 5 children had 11 trigger fingers, 1 had the trigger thumb in one side and in 3 fingers, and 1 had multiple trigger fingers. Among the author's over ten pediatric patients, they almost suffered the trigger thumb deformity in one hand or two hands, and they rarely suffered trigger finger deformity.

6.5.1 Etiology and Pathology

Many scholars believe that the etiologic factors of congenital tendon sheath stegnosis are different from those of the acquired thecostegnosis. In terms of nosogenesis, it is mainly induced by the pathological changes in the thumb long flexor tendon and the tendon sheath with the main manifestations as follows: ① the thumb long flexor tendons present the nodular-like thickening, also called Notta nodules [64]; ② tendon sheath stegnosis at the level of A1 trochlea; ③ both of them are present; and ④ the first metacarpal bone head is relatively thick or the tendon sheath orifice is stenotic. When the tendons are across the joints, in case the grafting angle or the gliding amplitude is large, the solid tendon sheath will be confined to the periost; therefore, the solid tendon and bones form the osseofibrous canal with little elasticity. The situation of the thumb is more special. There is a pair of sesamoid bones at the metacarpophalangeal joint, the thumb long flexor tendons pass through the two sesamoid bones, the thumb short abductor and the thumb breviflexor tendon end on the radial sesamoid bones, and the thumb adductor tendon ends on the ulnar sesamoid bones. There are more bony components of the osseofibrous canal at this place, and the three sides are the hard bones, so it is more stenotic. Errol [65] proposed that the original pathological changes in children's thumb long flexor tendon are collagen degeneration; the tendons often glide at the A1 trochlea to induce the rupture of synovium and produce stenosis; the thumb long flexor tendons present annular thickening at the metacarpophalangeal joint, inducing stenosis and compression; the thumb long flexor tendon shows excavation and grooves with enlargement of the two ends; and the free gliding of tendons within the tendon sheath is limited. Some scholars believe that the etiologic factor is as follows: the tendon nodules are formed at the metacarpophalangeal joint of the thumb long flexor tendon, and they are bound to be blocked when passing the relatively stenotic tendon sheath at the metacarpophalangeal joint; once the passage is successful, recovery is difficult, so two dents are formed at bilateral sides of the nodules.

The lesions of the acquired trigger finger and the congenital trigger finger are similar, with tendon sheath stegnosis as the main etiologic factor.

Pathological sectioning examination indicates that the tendon nodules and hypertrophic tendon sheaths present disorder in collagen and fibrous tissues with mild glass-like degeneration and the volar plate ligaments become thickened with mild inflammatory cell infiltration, which may be associated with repeated wear and tear. The thumb long flexor tendon sheaths are stenotic and thickened, and occasionally the tendon sheaths are located at the proximal end of the first trochlea. Chronic inflammation is also common; in case it subsides before the patient becomes 1 year old, there will no fixed contracture. With the increase in age and the extension of time of thumb interphalangeal joint locking, the thumb interphalangeal joint will suffer secondary contracture to varying extents in skin and joint capsule, even making the final segment of the thumb suffer deformity of ulnar deviation.

6.5.2 Clinical Manifestations

In flexion and extension of the thumb or fingers, there is a sensation of escape at the A1 trochlea (e.g., when the examiner uses fingers to palpate the A1 trochlea, there is a sensation of sudden passage of the obstacle). In severe cases, the thumb or the fingers cannot actively extend. In passive stretching of the thumb or fingers, there is nodular gliding and sensation of escape, or it is difficult for the flexed thumb to be extended, and even there is accompanying adduction deformity.

The time of congenital trigger finger found by parents varies. Some newborns are found with trigger finger deformity within just over 10 days after birth, but some pediatric patients' trigger finger is not found until they become 1 year old, and most parents take the children to the hospital because the children's thumb or fingers cannot extend. The trigger thumb mostly occurs to one side, and in rare cases it is seen in bilateral sides. It is rarely accompanied by other trigger fingers. The manifestations are as follows: the thumb flexion and extension are limited, the interphalangeal joint takes flexion shape, pain occurs and local skin becomes pale in passive extension, some cannot be passively extended, or flexion becomes impossible after extension, and nodules are palpable at the hyperextension position with inconspicuous tenderness. Therefore, in case the infant suffers deformity of thumb flexion and nodules are palpable at the metacarpophalangeal joint without obvious tenderness, the infant can be diagnosed with congenital cavity sheath stenosis. The long-term flexion contracture makes the metacarpophalangeal joint present hyperextension and semiluxation and may affect the development of the thumb; in addition, vicious

cycle may appear due to repeated wear and tear, so timely treatment should be given after the diagnosis is confirmed; otherwise, the phalangeal and articular development of the pediatric patients will be affected and the hand appearance and functions will be affected. Sometimes, the patient has a familial history of genetic diseases.

6.5.3 Auxiliary Examination

The diagnosis of congenital trigger finger is mainly based on the medical history and physical examination. Unless in severe cases, ordinary X-ray film cannot detect abnormalities, while ultrasound examination can clearly display the thickening of thumb long flexor tendons and trochlea and can be used for the auxiliary diagnosis of infantile trigger thumb. With the constant improvement of ultrasound probe frequency, the discrimination on superficial tissues has also been improved, and ultrasound examination has become the main examination method for superficial tissue small joint and muscle tendon lesions. The pathological changes of infantile trigger thumb are mainly the local thickening of the thumb long flexor tendon and trochlear thickening, and ultrasound examination can clearly display the changes in the trochlea at the metacarpophalangeal joint and flexor tendons. The ultrasound examination performed by domestic scholars [66] on 71 cases of infantile trigger thumb displays that the thumb long flexor tendon has obvious local thickening at the metacarpophalangeal joint; for the pediatric patients in different age groups, the section area of the thumb long flexor tendons at the metacarpophalangeal joint at the affected side is significantly larger than that at the healthy side, and the intergroup difference is of statistical significance; there is no significant difference between the two sides in terms of the section area of the thumb long flexor tendon at the middle level of thenar eminence, which is consistent with the fact that the thumb long flexor tendon presents nodular thickening at the head of metacarpal bone. In addition, the trochlea with thickening at the level of metacarpophalangeal joint of the trigger thumb is also the characteristic ultrasound manifestation, which may be correlated with the trochlear edema induced by the thickening of the thumb long flexor tendon.

6.5.4 Differential Diagnosis

This disease should be differentiated with congenital finger flexion deformity. The manifestation of congenital trigger finger is mostly manifested as limited finger extension, the interphalangeal joint is fixed in the flexion position, a snap sound is produced during extension of the interphalangeal joint or the interphalangeal joint is locked in the flexion position, and when the metacarpophalangeal joint is in the hyperextension position, nodules are palpable on the volar surface of the metacarpophalangeal joint, and the tenderness is not obvious. Congenital finger flexion deformity is a clinically rare hand deformity with the main manifestations of flexion contracture of the proximal interphalangeal joint, it has the onset within 1 year after birth, and it commonly involves the little, ring, and middle fingers, followed by the index finger, and the thumb is seldom involved and the involvement in bilateral sides is common. A differential diagnosis should be made when the congenital trigger finger is locked in the flexion position; metacarpophalangeal joint is in the hyperextension position in case of congenital trigger finger locking, and nodules are palpable on the palmar surface, which is of help to the differentiation.

6.5.5 Treatment

The key to treatment of congenital trigger finger is early detection and early treatment. Osteopaths especially hand surgeons need to be aware of this. It is difficult to confirm the diagnosis during the early stage of infancy. The patient's parents must be informed that rechecks should be performed every 3–6 months, and the patient should be advised not to rub the local region.

6.5.5.1 Nonsurgical Therapy

Thirty percent of the pediatric patients that suffer obvious symptoms before they are 1 year old can get better without intervention. The methods of observation and gentle manipulation therapy should be adopted, and braces or splints [67] can be used for fixation when the interphalangeal joint is in the extension position. If there is no remission of the symptom, intrathecal injection of adrenocortical steroids can first be given, but usually the efficacy is poor, and severe drug reactions or embrittlement and rupture of tendons may occur. If the symptoms persist when the patient becomes 3 years old, there will be thumb osteoarticular dysplasia, which affects the hand function [68]. Timely releasing treatment should be given. After the operation, the thumb flexion and extension functions may return to normal, and the effects of the thumb osteoarticular dysplasia are satisfying.

6.5.5.2 Surgical Treatment

As for the patients at the age of over 2 with disorder in finger flexion and extension, active surgical treatment should be given. The surgery can be performed during the infancy to prevent the occurrence of aplasia of the affected finger. The surgery should be performed under the condition of general anesthesia. The drape is disinfected according to the routine procedures, tourniquets are applied, and a transverse incision about 1 cm is made at the volar fold of the thumb metacarpophalangeal joint. The bilateral digital nerves should be

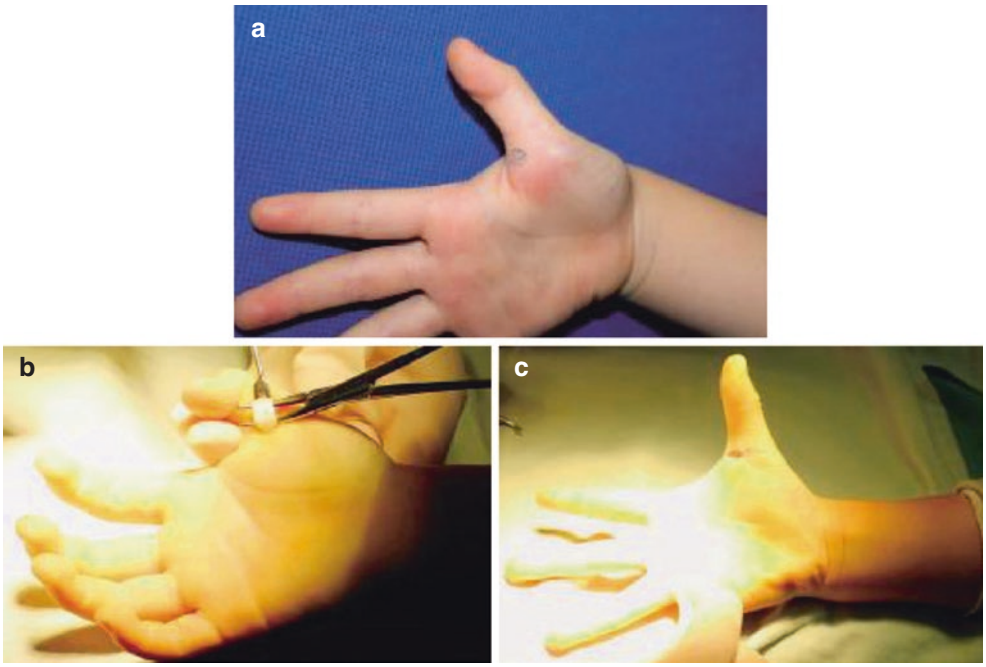


Fig. 6.50 Surgical treatment of congenital trigger finger. (**a**) Before the operation, the deformity of the trigger thumb is displayed, and Notta nodules are palpable at the proximal side of the A1 trochlea. (**b**) The thumb long flexor tendon and tendon sheath are exposed during the operation. (**c**) The annuliform trochlea is released and the thumb extending motions are recovered

protected. The thumb long flexor tendon and the tendon sheath are exposed, the tendon sheath becomes thickened, the tendons are continuously present but become thin, and there is adhesion of fibrous connective tissues around. Look for the proximal margin of the first trochlea, point the sticker cutting edge toward the distal end under the direct vision, perpendicularly cut the tendon sheath from the proximal end to the distal end, separate some of the tendon sheaths that become adherent to the fibrous connective tissues, and then move the affected finger; the free flexion and extension and the disappearance of the bouncing sensation indicate that the stenotic tendon sheaths have been longitudinally incised, and the inflated tendons can pass them freely. After the surgery, the entire layer of the incisions is sutured, and sterile dressings are used for ligation and fixation (Fig. 6.50). The main surgical point is to thoroughly release the stenotic sites of the thumb tendon sheath to avoid injuring the vascular nerves. After operation, it is necessary to strengthen the training of flexion and extension functions of the affected finger to avoid the recurrence of the adhesion.

6.6 Congenital Thumb Adduction and Flexion Deformities

Wei Wang

Congenital thumb adduction and flexion deformities are two kinds of concepts in terms of the property of disease, but the congenital thumb adduction deformity is often accompanied by flexion deformity; therefore, in this section, they are described as congenital thumb adduction and flexion deformities and called congenital thumb adduction deformity for short. Congenital thumb adduction deformity mainly features stenotic or too shallow fingerweb between the thumb and the index finger; morphologically, thumb web stenosis is displayed, and the thumb adduction, flexion, abduction, and palm opposition functions suffer disorder to varying extents. Up to now, the etiologic factors have been unknown. Clinically, the thumb adduction deformity has the following several manifestations, including thumb extensor tendon dysplasia, thumb flexor tendon contracture, thumb adductor contracture, contracture of the first dorsal interosseous muscle, and contracture of the thumb A2 trochlea. The pathomechanism is that the secondary lesions induced by the dysplasia or abnormality of some hand internal or hand external muscles result in the adduction and flexion of the thumb as well as the functional and appearance defects of the first fingerweb, and it may be the dysplasia of the entire thumb with the characteristics of thumb adduction deformity.

The congenital stenosis of the thumb is characterized by thumb adduction and flexion deformities, but in passive extension, adduction and flexion deformities are corrected.

Bayne (1982) classified the thumb adduction deformity into the category of thumb dysplasia. The thumb adduction deformity described by Bayne is often accompanied by thumb dysplasia or accompanied by thenar muscular dysplasia. Some call this deformity thumb-clutched hand or deformities of the palm center and thumb. Actually this is one type of congenital thumb dysplasia, so the author classifies this deformity into the category of congenital thumb dysplasia.

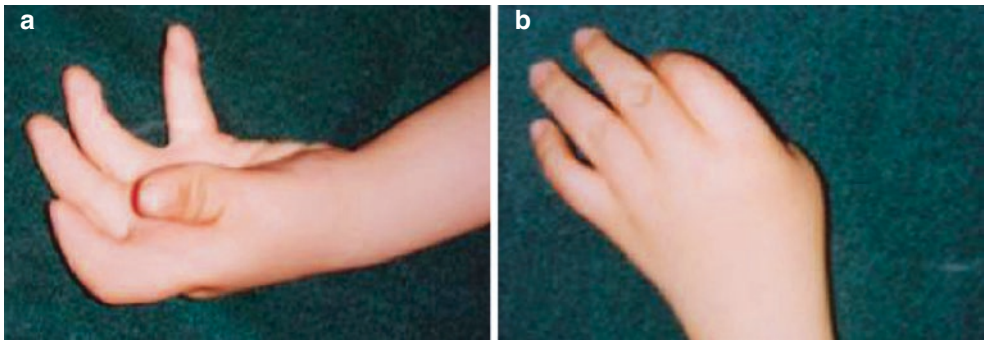


Fig. 6.51 Congenital thumb flexion deformity. (**a**) Windblow hand deformity. (**b**) Wicker hand deformity

There is also congenital thumb flexion deformity accompanied by deformity of flexion in the remaining four fingers. It is not suitable to classify this deformity into the category of congenital thumb adduction deformity. The author calls it windblow hand or wicker hand (Fig. 6.51), and this deformity will be described in another chapter.

6.6.1 Clinical Manifestation

It is manifested as thumb adduction to varying extents after the birth of the newborns, e.g., the thumb is gripped within the palm, which takes the shape of thumb clutching, but it will disappear after 6–12 months.

The congenital thumb adduction deformity is manifested as thumb web stenosis. In the static status, the thumb takes flexion and adduction shapes; the abduction abilities of the thumb decrease or disappear; the gap between the first and second interosseous space of metacarpus becomes narrow; the inter-thumb web skin, subcutaneous tissues, and fascia become tightened; there is no margin of abduction and relaxation; concomitantly dysplasia or labyrinth is seen in the hand internal muscle or hand external muscle of the thumb; and sometimes the deformity may be accompanied by dysplasia in thumb phalange, metacarpal bone, metacarpophalangeal joint, interphalangeal joint, and carpometacarpal joint as well as articular ligament relaxation. According to pathological anatomy and clinical manifestation, the author classifies congenital thumb adduction and flexion deformities into the following five categories: ① thumb extensor tendon dysplastic thumb adduction deformity, ② thumb breviflexor contractural thumb adduction deformity, ③ the first interosseous dorsal muscle contractural thumb adduction deformity (the author's case, no literature report), ④ pollical dysplastic thumb adduction deformity, and ⑤ congenital trigger thumb deformity.

6.6.1.1 Thumb Extensor Tendon Dysplastic Thumb Adduction Deformity

This deformity is induced by thumb long extensor and/or thumb short extensor dysplasia, and due to the different degrees, there are some differences in the deformity status.

Fig. 6.52 Thumb extensor tendon dysplastic thumb adduction deformity, accompanied by finger flexion deformity induced by extensor tendon dysplasia

The thumb long and short extensor tendons can be completely absent, but they are often manifested as dysplasia. Tendons are a thin layer of membrane-shaped tissues so that the extensors are weak, which is one kind of thumb deformities; they can also be manifested as thumb adduction and metacarpophalangeal joint flexion deformities, the gap between the first and second metacarpal bones are stenotic, and the thumb adduction deformity described in foreign literature is mainly this type. Although the author once treated multiple patients with this type of thumb adduction deformity, the relatively common cases were the congenital thumb adduction deformity induced by thumb breviflexor contracture. In addition to the thumb adduction deformity induced by thumb long extensor and/or thumb short extensor dysplasia, the deformity can also be accompanied by finger flexion deformity induced by extensor tendon dysplasia (Fig. 6.52).

6.6.1.2 Thumb Breviflexor Contractural Thumb Adduction Deformity

This deformity is manifested as stenotic and too shallow first fingerweb, the thumb metacarpophalangeal joint is flexed, and the thumb is located on the palmar surface of the hand; it can also be manifested as thumb-clutched hand deformity,

and there will be great resistance when the adducted thumb is pulled to the abduction position. The thumb long and short extensors often develop well and can be weak. The active and passive abduction functions of the thumb are limited, but the thenar muscles develop well. This deformity is mainly pathologically manifested as thumb breviflexor contracture, most of which is the contracture of the thumb breviflexor deep head, and sometimes contracture is in both deep and superficial heads; the deformity can also be contracture of the thumb adductor, and the latter is secondary in most cases (Fig. 6.53).

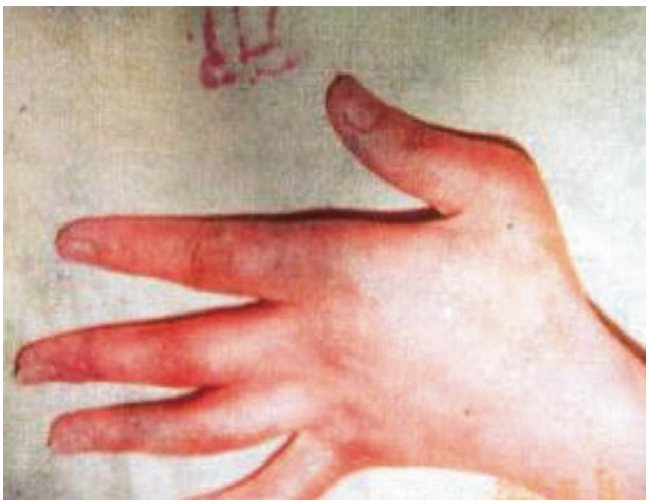


Fig. 6.53 Thumb breviflexor contractural thumb adduction deformity

6.6.1.3 The First Interosseous Dorsal Muscle Contractural Thumb Adduction Deformity

This is a rare type of congenital thumb adduction deformity, and there is no literature report about it. Among the acquired thumb adduction deformities, the relative common etiologic factor in China is necrotic and cicatricial contracture of the first dorsal interosseous muscle induced by the drug injection at the Hegu acupoint or trauma. The author witnessed two cases of congenital the first dorsal interosseous muscle contractural adduction deformity, who suffered from thumb adduction deformity immediately after birth without the medical history of the first dorsal interosseous muscle injury or the history of drug injection at the first fingerweb. The characteristics are as follows: the first fingerweb is stenotic and too shallow, the gap between the first and second metacarpal bones is stenotic, the index finger is abducted and located at the radial deviation position, an obvious tension sensation is felt during the adduction, the thumb metacarpophalangeal joint is slightly flexed, and the thumb extensor, flexor, and thenar muscles are normal (Fig. 6.54). It is found during the operation that the muscle belly of the first dorsal interosseous muscle is soft, the color is rosy, the luster is good, and there is no sign of cicatricial necrosis, but the tendons are thick and short, and the passive adduction makes it meet with obvious resistance in the normal neutral position.

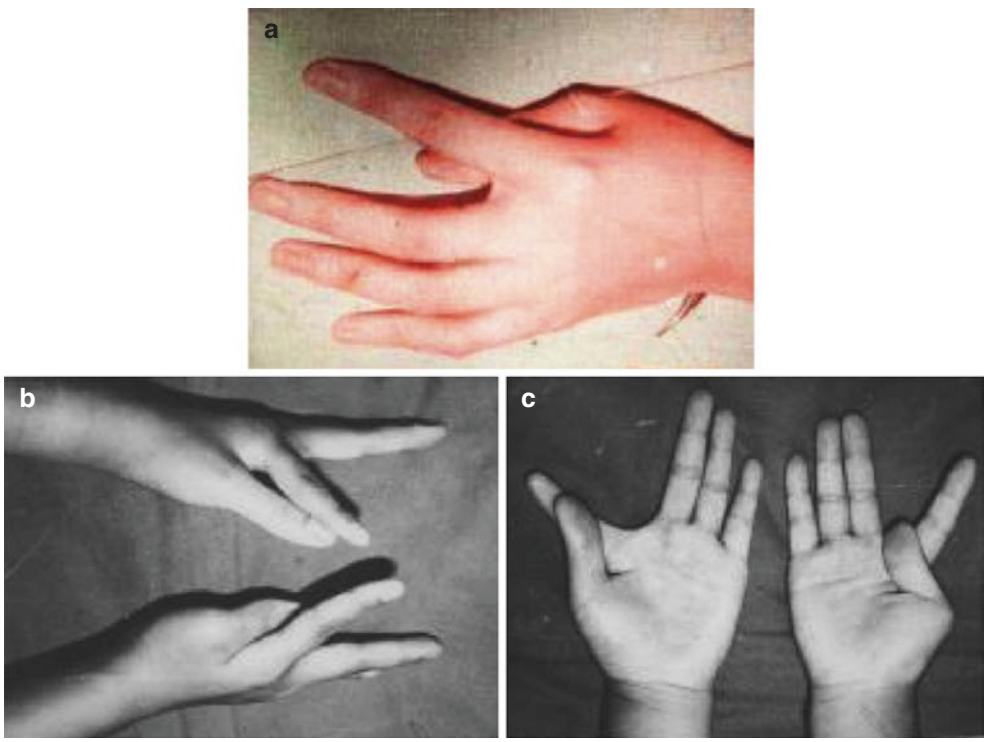


Fig. 6.54 The first interosseous dorsal muscle contractural thumb adduction deformity

6.6.1.4 Pollical Dysplastic Thumb Adduction Deformity

The congenital thumb adduction deformity is accompanied by short thumb deformity with functional insufficiency, with the presence of the first metacarpal bone, thumb proximal and distal phalanges, and metacarpophalangeal joint. The main manifestation is stenotic or too shallow first fingerweb, often accompanied by the short, small, and fine thumb, dysplasia in the first metacarpal bone and the thumb phalange, thenar muscular dysplasia, unstable metacarpophalangeal joint, and variation in insertions of thumb long flexor tendons, resulting in incomplete flexion or inability of flexion of the thumb interphalangeal joint, and in most cases the thumb extensor tendons are normal (Fig. 6.55).

6.6.1.5 Congenital Trigger Thumb Deformity

This deformity is manifested as temporary thumb adduction deformity and is another kind of thumb deformity.

6.6.2 Treatment

6.6.2.1 Treatment Timing

The surgery on the thumb adduction deformity induced by congenital thumb maldevleopment should be performed during the infancy; as for other congenital thumb adduction deformities, the surgery should be performed after the patient becomes 1 year old in order to avoid misdiagnosis, because thumb adduction of the infants under 1 is a normal phenomenon. The thumb adduction and flexion deformities during the infancy are often manifested as flexor contracture, and splint braces can be adopted for its treatment. Before surgery, it should be made clear whether it is real congenital thumb adduction deformity. The congenital trigger thumb can also be manifested as thumb flexion and mild adduction deformity and presents thumb-clutched hand, so a differentiation should be made because the congenital trigger thumb has the possibility of spontaneous cure.

6.6.2.2 Treatment of Thumb Web Contracture in Thumb Adduction Deformity

The treatment of thumb web contracture in congenital thumb adduction deformity refers to the repair of thumb soft tissue functions, including the following two aspects: one is expansion of thumb web, including repair of skin and subcutaneous tissue defects; the other is correction of muscle, tendon, fascia, aponeurosis, and articular ligament contracture, as well as reconstruction of dynamical functions:

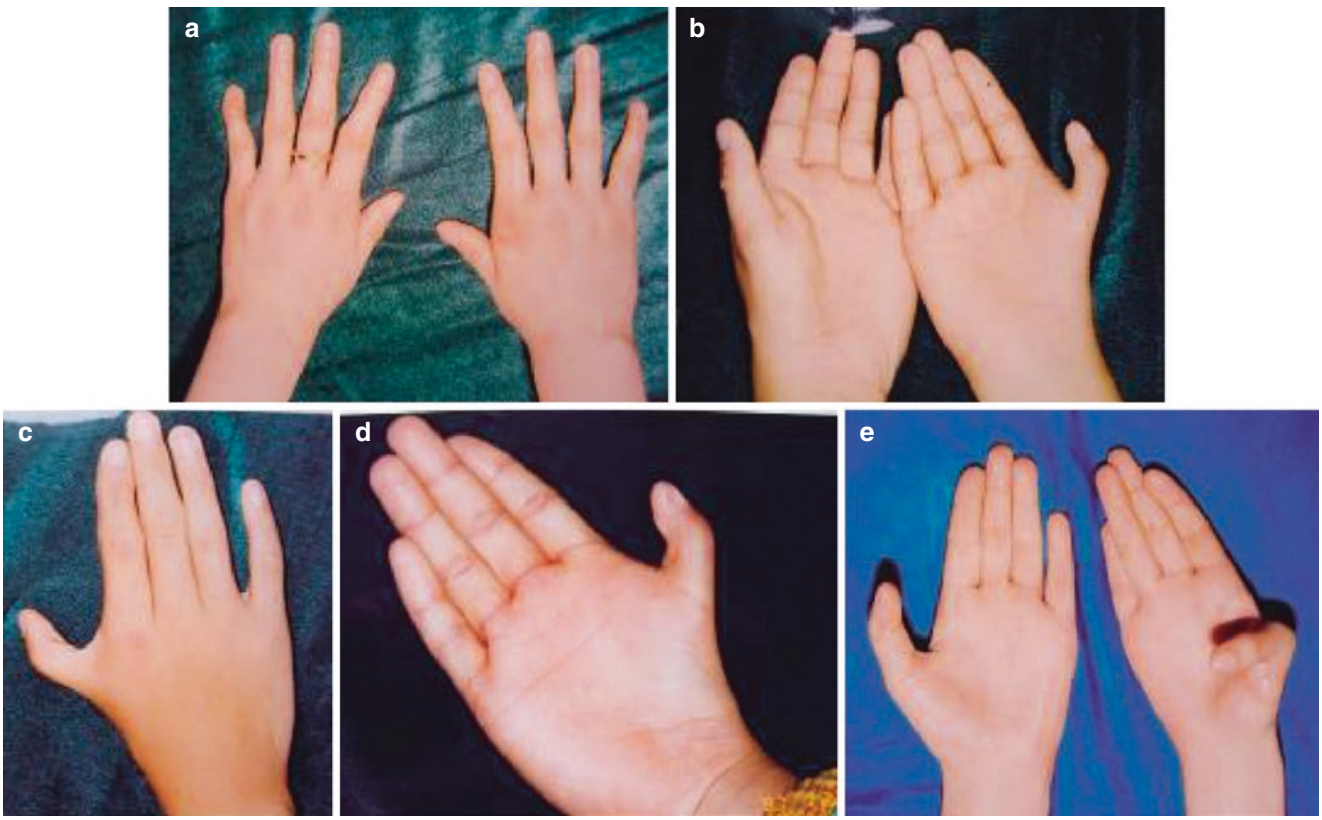


Fig. 6.55 Pollical dysplastic thumb adduction deformity

1. Thumb web expansion and repair of skin and subcutaneous tissue defects. The preferred method is the dual Z-plasty in the first fingerweb, namely, the coupled-triangular flap grafting (Fig. 6.56); this method is easy and effective. The second method is index finger dorsolateral flap grafting, namely, the rotational flap with the pedicle designed at the proximal end of the proximal digital dorsum of the index finger, to repair thumb web skin defects (Fig. 6.57), which is a good option. Selection of dorsal interosseous artery island skin flap is another good method to repair the skin defects at the thumb web area. Other pedicled skin flaps and free flaps for transplantation can also be adopted.
2. Correction of contractural muscles and reconstruction of dynamical functions. In order to deepen the thumb web, sometimes it is necessary to disjunct the transverse head of the thumb adductor; in case of inability of being corrected, partial oblique head of the thumb adductor can be disjuncted. In case of dysplasia of thumb long flexor or thumb long extensor, the ring finger superficial flexor tendons or the index finger inherent extensor tendon grafting can be adopted. For the patients with unstable or contractural metacarpophalangeal joint, the metacarpophalangeal joint accessory ligaments can be repaired or reconstructed (the details are shown in Chap. 5 "Congenital Thumb Dysplasia").

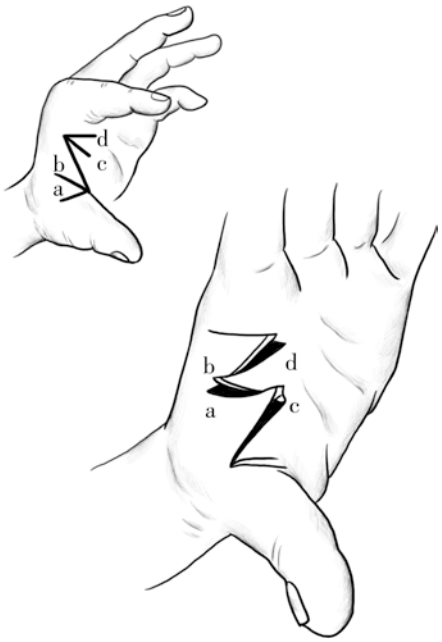


Fig. 6.56 Thumb web expansion, the dual Z-plasty in the first fingerweb area

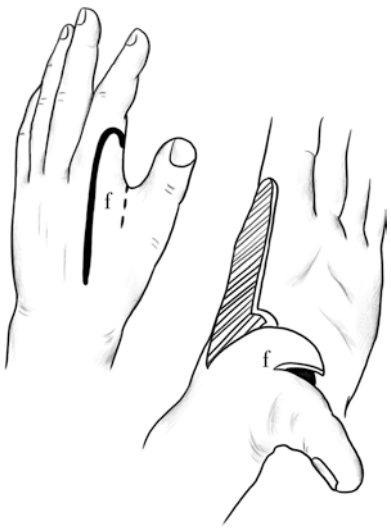


Fig. 6.57 Thumb web expansion, index finger dorsolateral flap grafting

6.6.2.3 Treatment of Thumb Extensor Tendon Dysplastic Thumb Adduction Deformity

The method is expansion of thumb web, repair of skin and subcutaneous tissue defects, and dynamical functional reconstruction of thumb extension. Inherent extensor tendon transplantation is commonly adopted:

1. The methods of thumb web expansion and repair of the skin and subcutaneous tissue defects are the same as above.
2. Dynamical functional reconstruction of thumb extension. The methods such as index finger inherent extensor tendon grafting, brachioradial muscle grafting + free tendon transplantation, and radial wrist long extensor grafting can be adopted.

6.6.2.4 Treatment of Thumb Breviflexor Contractural Thumb Adduction Deformity

1. Nonsurgical treatment. The thumb adduction and flexion deformities during the infancy are manifested as flexor contracture, and splint braces can be used for treatment. The thumb of the infant under 6 months presents flexion and adduction status, namely, thumb-clutched status; if the thumb still presents thumb-clutched status at the age of 1, the splint braces can be adopted for correction to make the first metacarpophalangeal joint be in the abduction and extension position to dilate the first fingerweb. The splint braces should be adjusted according to the correction status of the deformities. If the symptoms improve after the application of splint braces, they are exchanged every 6 weeks for 3–6 months; if no improvement is seen in the symptoms, surgery can be adopted for correction.

2. Surgical treatment. The main method is to disjunct or lengthen the contractural thumb breviflexor tendon; in mild cases, it is just necessary to disjunct the deep head of the thumb breviflexor to correct deformities; in severe cases, it is necessary to disjunct or lengthen the deep and superficial heads of the thumb breviflexor, or at the same time, some insertions of the contractural thumb adductor are disjuncted to dilate the first finger-web (Fig. 6.58). Both methods can produce satisfactory effects.

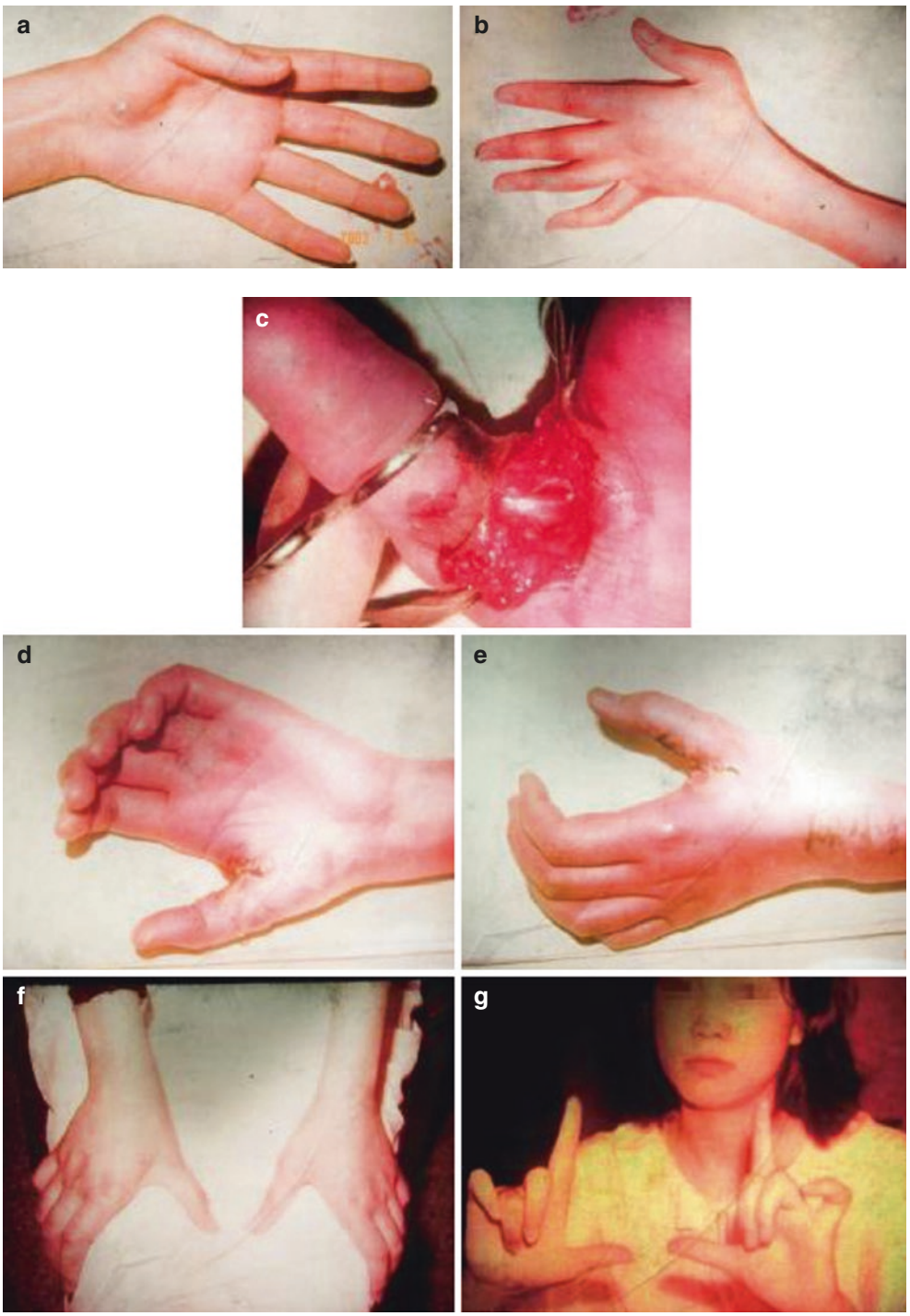


Fig. 6.58 Treatment of thumb breviflexor contractural thumb adduction deformity. (**a**, **b**) Hand appearance before operation. (**c**) Disjunction of the deep head of the thumb breviflexor during operation. (**d**, **e**) Hand appearance after operation. (**f**, **g**) Good recovery of functions after operation

6.6.2.5 Treatment of the First Interosseous Dorsal Muscle Contractural Thumb Adduction Deformity

This deformity is relatively rare, the preoperative accurate diagnosis is the prerequisite, and the main treatment content is relief of contracture.

Make an S-shaped incision at the radial side of the metacarpophalangeal joint of the index finger, and the contractural first dorsal interosseous muscle is exposed. Make a Z-plasty incision on the tendons, lengthen the tendons, and correct the radial correction deformity of the index finger. Under general circumstances, after the first dorsal interosseous muscle is lengthened, the thumb adduction deformity is corrected. In case the radial deviation of the index finger is still present after the tendon is lengthened, the accessory ligaments of metacarpophalangeal joint of the index finger can be released. After the operation, the Kirschner silk can be used to temporarily fix the metacarpophalangeal joint for 2–3 weeks. As for the patients with thumb web stenosis induced by skin and subcutaneous defects, corresponding Z-plasty or skin transplantation should be performed (Fig. 6.59).

6.6.2.6 Preoperative and Postoperative Measures for Thumb Adduction Deformity Treatment

In most cases, the thumb adduction deformity is induced by hand internal muscle contracture or hand external muscle dysplasia, and there is a tendency of retraction of the first fingerweb after surgical correction. In order to prevent the recurrence of deformity, Kirschner silk can be used to hold the seriously adducted thumb between the first and second metacarpal bones for 3–4 weeks, and then braces are used to expand the thumb web for 3 weeks. However, under general conditions, the splint braces are used to maintain the effects of surgical treatment; e.g., braces are adopted for 3 weeks; 3 weeks later, if the effects are poor, the patient can move during the daytime and braces are adopted for maintenance during the night for a total of about 3 months. Splint braces are not only the tools that prevent postoperative recurrence of deformity but also can serve as the preoperative preparation to dilate the first fingerweb and to improve surgical effects.

6.7 Congenital Ulnar Deviation Hand Deformity

Wei Wang, Longchun Zhang, and Jianmin Yao

6.7.1 Overview

Congenital ulnar clubhand, also called congenital windblow hand deformity and congenital digital and palmar contracture deformity, is manifested as flexion and ulnar deviation deformities of the entire hand, and it is a kind of hand congenital deformity syndrome induced by hand and upper limb dysdifferentiation.

The congenital windblow hand deformity is manifested as thumb adduction and flexion, thumb web stenosis, palmar

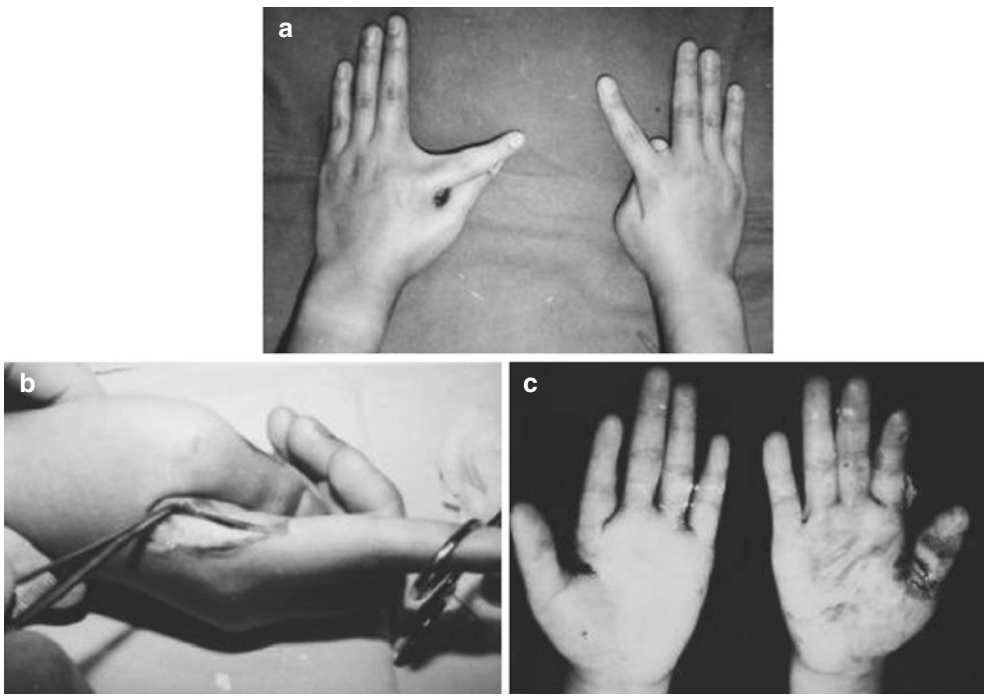


Fig. 6.59 Treatment of the first interosseous dorsal muscle contractural thumb adduction deformity. (**a**) Hand appearance before operation. (**b**) During the operation, the contractural first dorsal interosseous muscle tendon is exposed for lengthening. (**c**) After thumb web expansion, flap grafting, and skin transplantation

contracture, finger metacarpophalangeal or interphalangeal joint flexion, disorder in thumb extension, thumb adduction and finger extension functions, and thumb and finger dysplasia. It is characterized by obvious finger metacarpophalangeal joint flexion and ulnar deviation, and the supination function of the forearm is often limited. The etiologic factor of this congenital hand deformity is unknown, and the patient often has a family history. As the functional defects of this deformity are diverse and can affect each other, there is no uniform rule for the naming of such deformity. The author believes that it is suitable to call it congenital windblow hand deformity, congenital ulnar deviation hand deformity, or congenital windblow hand deformity syndrome.

Congenital windblow hand deformity is a type of congenital thumb and finger flexion deformity accompanied by the ulnar deviation of the metacarpophalangeal joint and fingers. As early as 1897, Emile Biox [69] described the characteristics of windblow fingers, but some scholars believe that this name cannot reflect the direction of finger deviation. In 1976, Powers et al. [70] started to use the name of congenital hand ulnar deviation or congenital finger contracture to describe this deformity, which has been gradually accepted by the academic circles. In 1984, Zancolli [71] named this deformity congenital ulnar deviation hand deformity. This congenital hand deformity can be one of the symptoms of some syndromes, the most famous of which is Freeman-Sheldon syndrome [72], and these syndromes have the main manifestations of cranial, hand, and foot deformity. In 1963, Burian [73] described whistling face syndrome, and its symptoms also include congenital windblow hand deformity. The patients with whistling face syndrome have long and protruding lips with rigid facial expression, which looks as if the patient were whistling, hence its name. The hand deformities of all of the above syndromes include finger ulnar deviation, metacarpophalangeal joint flexion, thumb adduction deformity, and the first fingerweb contracture. At present, it is common to use the name of congenital windblow hand or congenital ulnar deviation hand deformity, because these two names vividly reflect the morphology and characteristics of this deformity.

This deformity can be included into the same category of finger flexion deformity, finger lateriflection deformity, thumb-clutched hand deformity, thumb-in-palm deformity, etc., and they have the characteristics of contracture deformity.

6.7.2 Etiologic Factor

The occurrence of this disease is correlated with hereditary defect, and it is often autosomal dominant inheritance. Some once carried out a genetic study on the congenital palmar contracture deformity among the races in South Africa and found that autosomal genetic genopathy was correlated with this disease [74]. Among the dozens of patients with congenital windblow hand deformity treated by the author, some had the familial onset tendency with the obvious hereditary feature, but it is not universal. Among them, there was a 4-year-old girl with flexion contracture in both hands, thumb adduction and flexion deformities, thumb web stenosis, palmar contracture, metacarpophalangeal joint flexion in four fingers, and the degree of ulnar deviation of over 30°, and her mother and maternal grandmother suffered similar hand deformity (Fig. 6.60).

6.7.3 Pathology

There are many theories on the pathological basis of the congenital windblow hand deformity. Fisk (1974) [75] believed that too strong hand flexor tendons resulted in hand flexion and ulnar deviation deformity; some patients concomitantly suffered from atrophy of upper limb muscles with unilateral onset often, which is another pathological cause of windblow hand deformity. Malkawi (1983) [74], after detecting the specimens of patients with windblow hand deformity, found that there was fatty infiltration and fibrosis inside the muscular tissues with the presence of diffuse muscular atrophy. The main mainstream theories among the multiple theories on the pathological basis of windblow hand deformity are as follows:

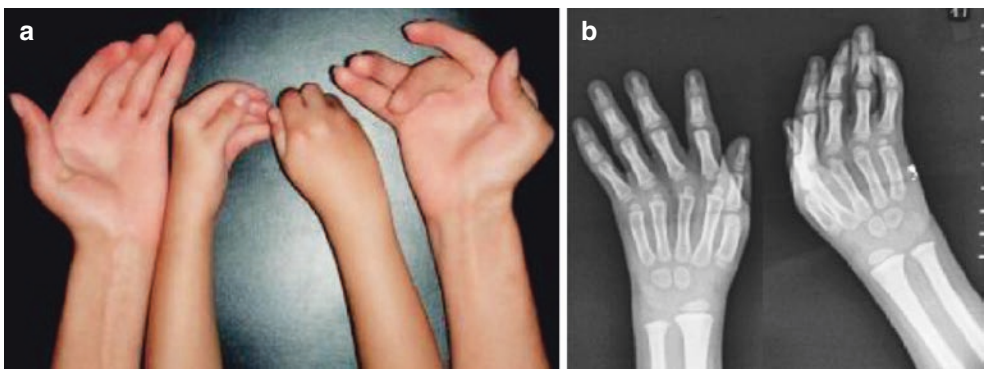


Fig. 6.60 Congenital windblow hand deformity in both hands. (**a**) The child's two hands are the interior ones, and the two hands of the child's mother are exterior ones. (**b**) X-ray film of the child's deformed hands

6.7.3.1 Soft Tissue Contracture Theory

Zancolli et al. [76] believed that windblow hand deformity was the secondary change induced by soft tissue dysdifferentiation. The dysdifferentiation in the hand, skin, or midpalmar fascia makes the palm and interdigital regions form thick and irregular subcutaneous contracture cords, and these abnormal subcutaneous cords and the shortened fingers and palm skin will result in secondary changes in tendons, joints, and bones and become the main cause of metacarpophalangeal flexion deformity and finger ulnar deviation.

6.7.3.2 Theory of Muscular Abnormality

According to Lanz and Teoh L.C. et al. [77, 78], the action of abnormal or proliferative muscles on the ulnar side of the proximal phalange was the pathological basis that resulted in windblow hand deformity, and it was necessary to remove these abnormal muscles during operation. In 2004, Grunert J. [79] proposed a bolder presumption that the pathological basis of congenital ulnar deviation hand deformity was a type of atavism, which means that incomplete degeneration of some muscles would temporarily occur during embryonic development.

Congenital windblow hand deformity can be divided into two types according to these two kinds of pathological basis: one is bilateral onset with the main pathological basis of soft tissue contracture inducing the joint contracture; the other is unilateral onset, in which muscle hypertrophy is usually observed in hands or upper limbs, and abnormally proliferative muscles end at the phalangeal ulnar side.

6.7.3.3 Theory of Poor Differentiation of Hand-Supporting and Dynamical Structures

The author believes that congenital windblow hand deformity is induced by the dysdifferentiation of supporting structures (bones, joints, ligaments, fascia, and aponeurosis), dynamical structures (muscles and tendons), as well as the corresponding vessels, nerves, and skin coverage to varying degrees and in different combinations, and the clinical characteristics are palmar aponeurosis contracture, ulnar deviation of finger extensor and thumb extensor tendons, and finger and thumb dysplasia.

Due to this, the patient's morphological and functional defects in hands and upper limbs are many-sided and comprehensive, with both dysdifferentiation in the skin or palmar fascia and abnormal development to varying extents of thenar muscles, interosseous muscles, palmar fascia, finger extensors, thumb extensors, bones, joints, and ligaments, manifested as short and small thumb and fingers, thumb web stenosis, and short and superficial fingerweb; and the deformities cover the areas in the thumb, all fingers, palm, and forearm; therefore, it is suitable to call this deformity "congenital windblow hand syndrome."

The theory of soft tissue contracture proposed by Zancolli et al. [76] holds that windblow hand deformity is the secondary change induced by soft tissue dysdifferentiation, but this conclusion cannot easily explain whether the windblow deformity involves the abnormal development in multi-layers, full structures, full fingers, and palms. If the deformities in muscles, tendons, bones, and joints are secondary according to the above theory, postnatal use of scaffolds can help correct the deformity of ulnar deviation. In fact, preoperative use of scaffolds cannot reduce or prevent the deformities and aberrations in muscles and tendons and cannot treat bone and joint deformities. This is the consensus of many clinicians in practice.

It can also be seen from the author's many cases that such deformity is often accompanied by short and small thumb and fingers, thumb web stenosis, relatively superficial fingerweb, and concomitant presence of supination disorder to varying extents in the forearm. When the wrist is in the flexion position, the finger ulnar deviation is somewhat relieved, which can also explain why it is difficult for the simple soft contracture theory to summarize the mechanism of the production of multiple windblow hand deformities. When treating serious windblow hand deformity, the author often adopts finger extensor tendon hood correcting surgery that is intended for the military salute hand and can effectively correct the ulnar deviation deformity of metacarpophalangeal joint, indicating that this windblow hand deformity suffers defects in hand internal muscle development, which is one of the main pathological and etiologic factors for the dynamical defects of hand ulnar deviation. The windblow hand deformity often has the bilateral manifestations with the presence of thumb extensor and thumb long abductor dysplasia, indicating that this deformity is induced by the multi-structural dysplasia in hands and forearms.

6.7.4 Clinical Manifestation

The hand ulnar deviation, palmar contracture, and finger deviation deformities of congenital windblow hand deformity can appear immediately after birth. With the increase in age, the deformity becomes more obvious, with the manifestations of thumb adduction and flexion deformities and thumb web stenosis. In severe cases, the thumb is in the center of the thumb, the thumb extension strength decreases or disappears, there is resisting tension to varying extents when the thumb is passively extended, and the second to the fifth fingers also suffer flexion and ulnar deviation deformities to varying extents, with metacarpophalangeal joint flexion and ulnar deviation as the main features. Some patients can suffer military salute-like hand deformity. When the fingers are passively extended, the palm skin and the structures below them have an obvious tension. All fingerwebs are too superficial and web shaped, the thumb and the fingers are shorter than those of healthy persons, and the metacarpophalangeal joints of the second to fifth fingers incline to the ulnar side, become flexed, and present mild pronation deformity.

The finger extensor muscle strength is normal or decreases, and the muscular strength of the thumb extensor and finger extensor often become reduced. Among finger deformities, pathological changes involve the metacarpophalangeal joint, proximal interphalangeal joint, and distal interphalangeal joint with the manifestations of flexion deformity and limited extension, but the symptoms in the proximal interphalangeal joint are severer with few manifestations of boutonniere deformity in the interphalangeal joint. In addition to hand deformities, the patient may concomitantly suffer from amyoplasia of the forearm. Foot deformities may be accompanied by hand deformities, with the manifestations of hockey club foot, crank handle foot, and pedal digital and plantar contracture. The patients also suffer from witless facial expression (taking masklike shape), microstomia deformity (the appearance is like whistling), asymmetric chest and shoulders, and scoliosis.

6.7.5 Classification

Zancolli et al. classified congenital windblow hand deformity into three categories according to the severity of the lesions: ① contracture of hand skin and subcutaneous tissues; ② presence of finger-involving tendon contracture, in addition to skin and subcutaneous tissue contracture; and ③ contracture of joint ligaments and joint capsules as well as bone deformities in addition to the above lesions.

The author believes that it is suitable to classify windblow hand deformity into three types: ① mild windblow deformity, mainly contracture in the skin, fascia, and palmar aponeurosis; ② moderate windblow deformity, dysplasia of hand internal muscles and hand external muscles in addition to contracture in the skin, fascia, and palmar aponeurosis; and ③ severe windblow deformity, serious deformities of bones and joints of hands, which need to be treated through osteoarticular correction.

6.7.5.1 Mild Windblow Hand Deformity

Mild windblow hand deformity mainly features palmar and finger flexion contracture. In case of wrist flexion, the flexion deformity of the finger metacarpophalangeal joint and interphalangeal joint is obviously relieved, they can be passively extended under the low tension, the finger metacarpophalangeal joint has mild ulnar deviation (less than 20°), the thumb web has mild stenosis, the thumb suffers mild deformity of adduction and flexion, it can be passively abducted under the low tension, and the muscular strength of finger extensor, thumb extensor, and thumb abductor is of over grade 4 (Fig. 6.61).

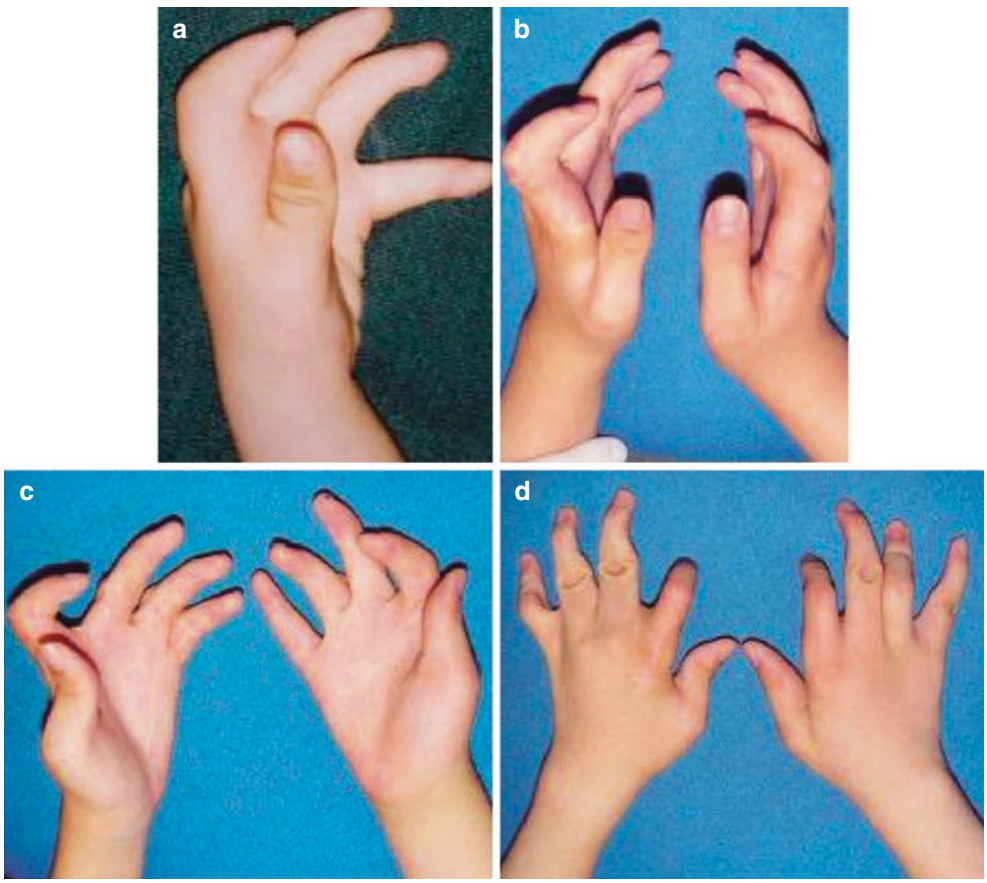


Fig. 6.61 Mild windblow hand deformity mainly features palmar and finger flexion contracture

6.7.5.2 Moderate Windblow Hand Deformity

Moderate windblow hand deformity is manifested as serious flexion contracture and ulnar deviation in palms and fingers, with the main manifestation of metacarpophalangeal flexion and military salute hand deformity. When the carpal flexed finger is passively extended, the tension is highly resistant, the thumb suffers serious adduction and flexion deformity, it is often adducted to the palm center, and the thumb web has serious stenosis. When the thumb is passively abducted, the resistance is high, the finger metacarpophalangeal joint has serious ulnar deviation (over 20°), the thumb extensor and/or finger extensor suffers dysplasia, and the muscular strength of thumb extensors and thumb abductors is of less than grade 4 (Fig. 6.62).

6.7.5.3 Severe Windblow Hand Deformity

Severe windblow hand deformity is manifested as serious hand osteoarticular deformity, which needs to be treated through osteoarticular correction.

6.7.6 Differential Diagnosis

The manifestations of windblow hand deformity are flexion and ulnar deviation deformities in the entire hand, and the single-digital and multi-digital flexion deformities are not within the range of windblow deformity (Figs. 6.63 and 6.64).

6.7.7 Treatment

The author believes that the disease should be treated as early as possible, surgical treatment can be given before the patient becomes 2 years old, splint braces can be used before operation to correct finger flexion and ulnar deviation deformities, and they should become conventional preparations before treatment. The reasons are as follows: (1) as the pediatric patient does not cooperate well in treatment, it is difficult to obtain good effects; (2) the use of splint braces for the pretreatment preparations can reduce the tension in correction of finger flexion contracture, which is of help to the correction of metacarpophalangeal joint deformities. As for the patients on which brace correction produces good curative effects, it is suggested that the surgery be performed when the patient is 2–4 years old, and the purpose is to correct and reduce the tissue tension arising from flexion deformity. In addition, the children at the age of over 2 can cooperate well during the preoperative physical examinations, allowing the surgeons to accurately judge the functional defects of hand deformities and especially know the functional defects of hand muscles, so that they can propose rational treatment policies. Most of the windblow hand deformities treated by the author are in both hands, but no one is cured due to adoption of physical therapy such as scaffolds and splints. According to the reports of Kaliainen L. [80], among the 18 patients, only 1 patient avoided surgery due to the use of brace for treatment. This pediatric patient started to wear

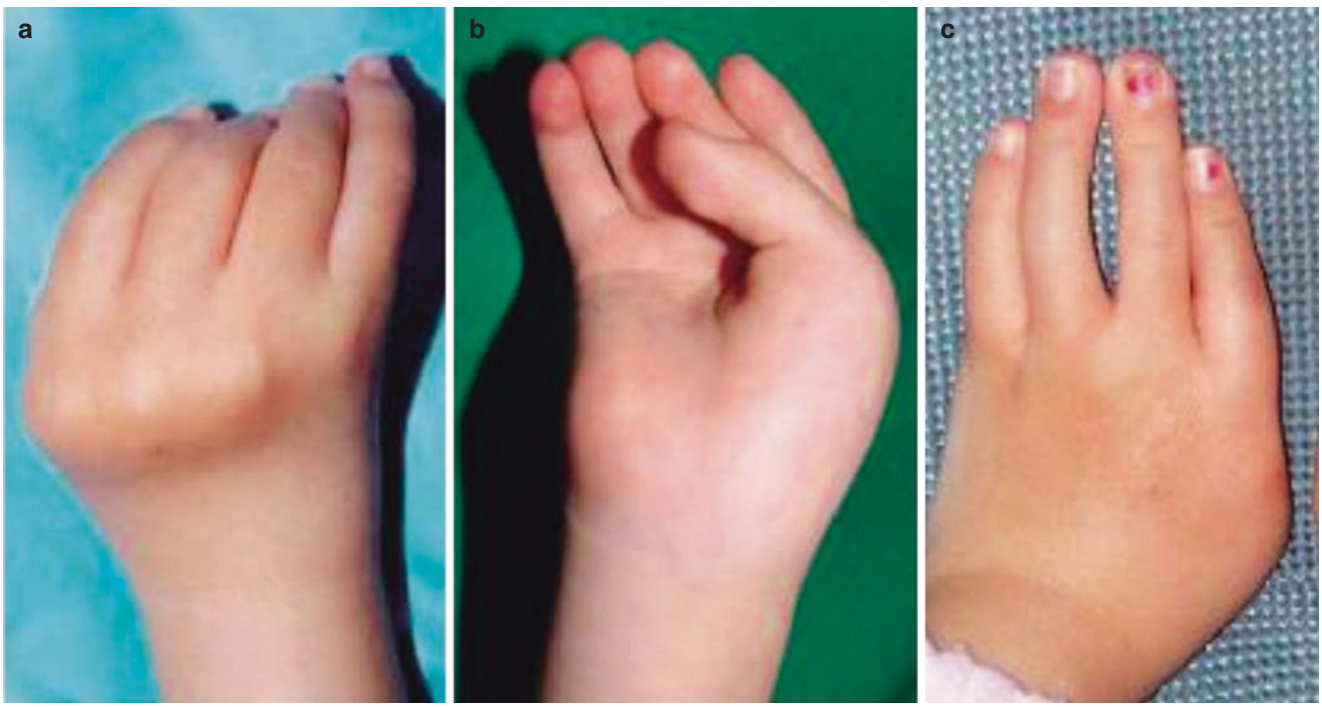


Fig. 6.62 Mild windblow hand deformity: the palms and fingers suffer serious flexion contracture and ulnar deviation deformities with the presence of military salute hand deformity. (**a**, **b**) Right hand. (**c**) Left hand

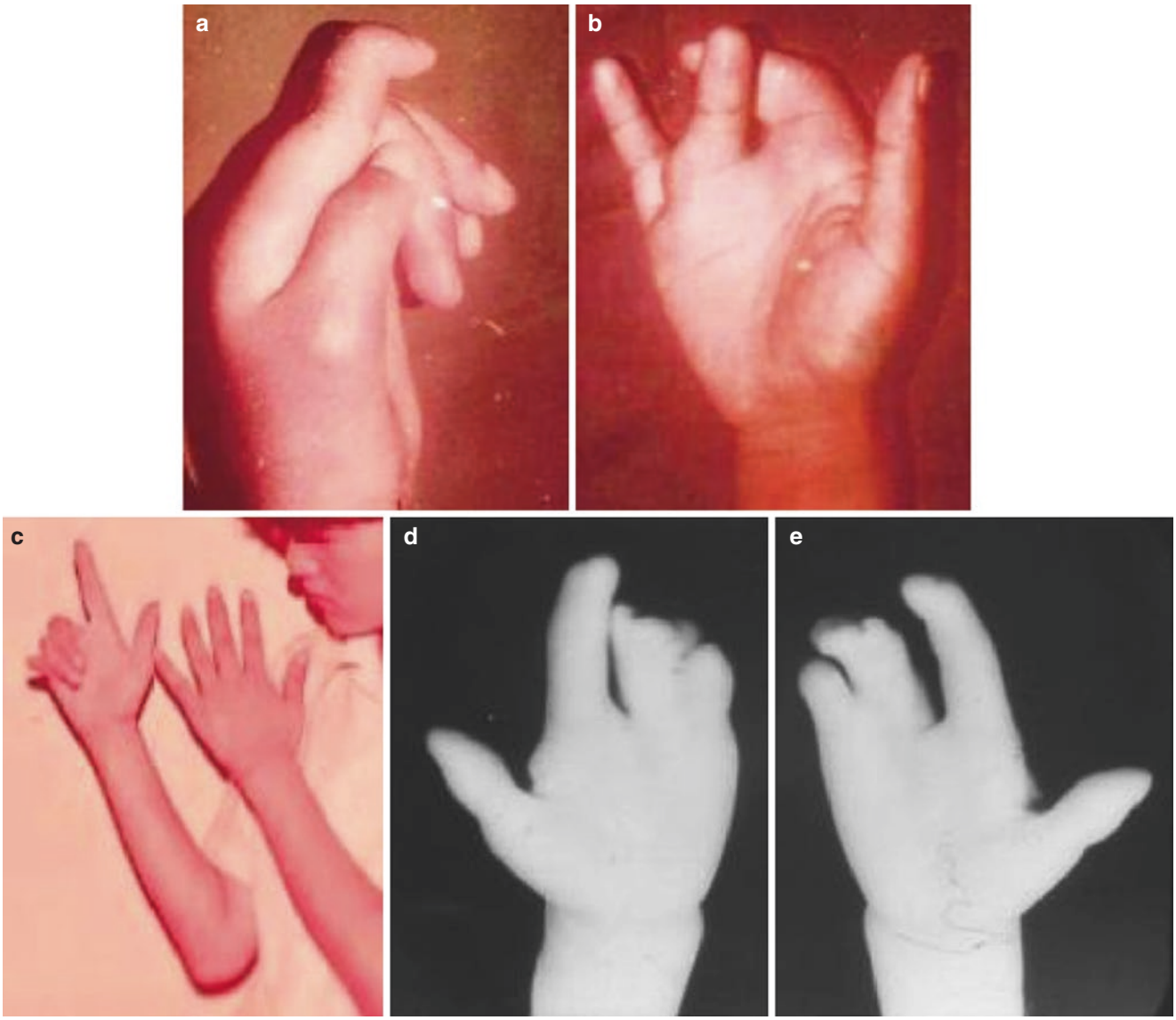


Fig. 6.63 Various types of finger flexion deformities. (**a**) Single-digital congenital flexion contracture. (**b**) Finger flexion deformity of the cleft hand. (**c**) Flexion deformity of middle, ring, and little fingers induced by superficial digital flexor dysplasia of the third to fifth fingers. (**d**, **e**) Multi-digital flexion deformities induced by congenital hand dysplasia

braces at 5 months after birth and discontinued their use until it became 2 years old. The surgical treatment during the infancy focuses on soft tissue correction of the deformity of shortening, and the youngsters and adult patients can obtain good efficacy only when they are cooperative during the osteotomy for correction. The patients treated by the author were children, and no patient needed to be corrected through metacarpal bone osteotomy.

6.7.7.1 Correction of Thumb Flexion and Adduction Deformities

First, incise the skin with flexion contracture, perform Z-plasty or regional flap transplantation + free skin grafting, correct thumb flexion deformities, and expand thumb web to make the thumb reach the radial abduction position. In case of mild thumb web contracture, two flaps can be used to perform Z-plasty or four flaps and five flaps are used for reshaping; in case of serious thumb web stenosis deformity accompanied by hand internal muscle dysplasia, if the method of local Z-plasty or four-flap and five-flap does not work or the efficacy is not as good as expected, the index finger dorsal flaps or the thumb dorsal flaps can be adopted for rotation and transplantation to repair thumb web, and in this way, good surgical effects can be obtained. Some scholars adopt interosseous dorsal island-shaped skin flap grafting, free flap grafting, distal pedicle flap grafting, etc., which, according to the author, are also considered an option, but they are not the best regimen for pediatric

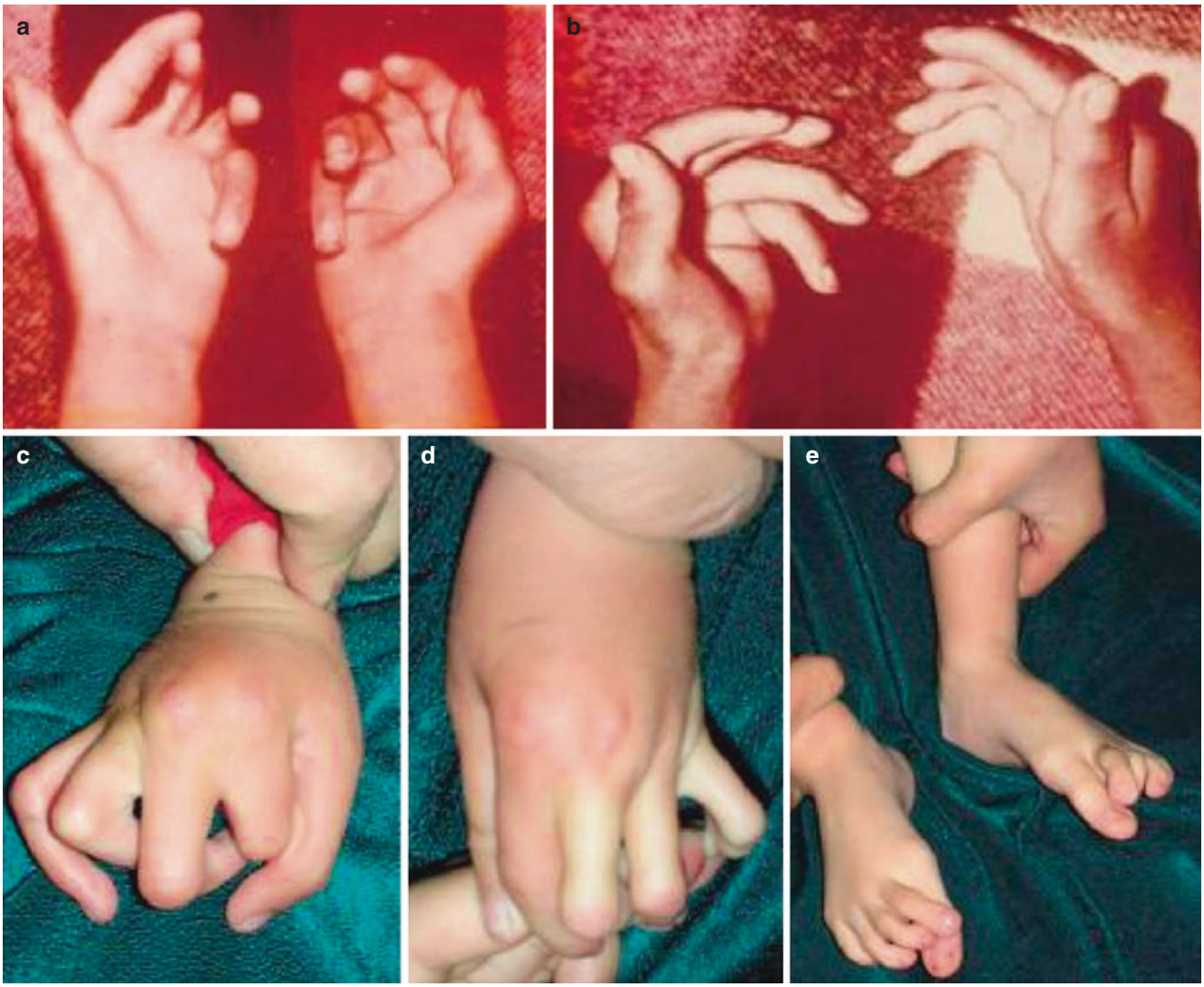


Fig. 6.64 Windblow hand deformity. (**a**, **b**) Congenital windblow hand deformity in both hands, thumb adduction and flexion deformities, and finger metacarpophalangeal joint flexion accompanied by ulnar deviation deformity. (**c**–**e**) Congenital windblow hand deformity in both hands, accompanied by flexion, adduction, external rotation, and other deformities in toes of feet

patients because regional flap grafting often can only serve the purpose of expanding thumb web.

6.7.7.2 Correction of Finger Flexion Deformity and Web-Shaped Deformity with Too Shallow Fingerweb

Finger flexion deformity and too shallow fingerweb are associated with volar skin defects and shortened vascular nerve tracts of the palms and fingers, finger volar Z-plasty or + V-Y reshaping can be adopted, and skin grafts can be adopted to repair skin defect areas. The reshaping of web-shaped deformity is similar to correction of mild syndactylia, and the details are shown in Sect. 7.1 Syndactylia in this chapter.

There are two kinds of finger flexion deformity: One is ulnar deviation of the finger metacarpophalangeal joint, accompanied by flexion deformity of the proximal interphalangeal joint; the pathogenesis of interphalangeal flexion deformity is similar to that of boutonniere deformity. This deformity is not common in typical windblow hand deformities. The author mainly adopts apposition suture at the bilateral lateral bundles of the finger extensor tendons at the interphalangeal joint to make them draw close to the center, which has a certain effect on correction of the interphalangeal joint deformity. The second one is metacarpophalangeal joint flexion, accompanied by extension deformity of proximal interphalangeal joint, which is similar to positive hand internal muscles. The author adopts the removal of mesh-shaped ligaments of the dorsal extensor tendon device of the metacarpophalangeal joint, which facilitates deformity correction.

According to the pathogenesis of such deformity, antedisplacement of hand internal muscles and lengthening of

interosseous muscle tendons are also the surgical regimens that can be considered. The author uses these methods for the treatment of severe windblow hand deformity and obtains good efficacy.

Even after the above surgery is completed, it is still difficult for fingers to be passively extended because the flexor tendons still have a high tension. So for adult patients, shortening of metacarpal bone can not only correct finger flexion deformity but also facilitate correction of ulnar deviation and internal rotation deformities of metacarpophalangeal joint. It is the easiest surgery.

6.7.7.3 Correction of Ulnar Deviation and Internal Rotation Deformities of Metacarpophalangeal Joint

Correction of ulnar deviation and internal rotation deformities of metacarpophalangeal joint has always been the key to treatment. The author believes that it is necessary to correct finger ulnar deviation and flexion deformity to reconstruct the balance of the muscular strength of hand internal muscles [81], because imbalance of muscular strength of hand internal muscles is one of the main reasons for finger ulnar deviation. If hand internal muscles tighten the ulnar structure, such as the third and fourth dorsal interosseous muscles and little finger abductors, then these structures should be lengthened or cut off through surgery. In case the metacarpophalangeal joint capsule or the accessory ligaments are tense when the finger is passively in the neutral position (Fig. 6.65), some joint capsules can be removed, and the accessory ligaments can be lengthened during surgery. If the superficial digital flexor is short and tense and finger ulnar deviation is severe, it is necessary to transpose the superficial digital flexor and then suture it onto the radial hand internal muscles of the adjacent finger, but this method is rarely adopted by the author.

As for the correction of metacarpophalangeal joint ulnar deviation, for the mild symptoms, the reshaping of extensor tendon device can be adopted, e.g., centralize the extensor tendon hood, reposition the extensor tendons between the human metacarpal bone heads that are dislocated, and fix them to the median line of the metacarpophalangeal joint. The author adopts the dynamical correction method of ulnar deviation of extensor tendon hoods, e.g., performing longitudinal incision at the ulnar side of the extensor tendon hoods, to release the tension arising from the ulnar deviation of the tendon hoods; in case of relaxation of the radial side of the tendon hood, perform longitudinal incision and plicating suture to correct the deformity of axial ulnar deviation of the extensor tendons, and in the meantime release the metacarpophalangeal joint accessory ligaments and resect the tendon hood triangular ligaments. In order to guarantee the stability after the correcting surgery, it is necessary to use Kirschner silk for fixation of the metacarpophalangeal joints for 3 weeks to maintain the tension balance of metacarpophalangeal joint. Some adopt the method of removing one of the finger extensor tendons and suturing it to the finger extensor tendon after circling it around the phalange for 1 week; some use the method of drilling a hole on the dorsal side of the proximal phalange and using 2-0 silk to suture and fix the tendons, but the author does not recommend it. In order to guarantee the effects of correcting surgery, Kirschner silk can be used for fixation for 3 weeks in the metacarpophalangeal joint extension position, the use of scaffolds is continued after the steel needles are pulled out, and the patients can voluntarily move but wear the scaffolds during the nights. During these operations, attempts should be made to avoid injuring the epiphysis. After these surgeries, most of the deformities can be obviously corrected. Even so, sometimes, deformity correction is not perfect, or as for the patients that miss the surgical opportunity, osteotomic correction needs to be done. If the deformity falls into the category of severe windblow hand deformity, it is necessary to perform osteotomic correction of bones and joints.

Osteotomy under the head of metacarpal bone can produce better effects than phalangeal osteotomy. Use a longitudinal incision at the hand dorsal regions between the index finger and middle finger or between the ring finger and little finger, use an electric saw to perform wedge osteotomy on the metacarpal bones after the tendons are pulled out, use two Kirschner silk or small steel plates or miniature steel plates to fix the metacarpal bones, and after operation use cast-form plaster or scaffolds of short forearms for fixation

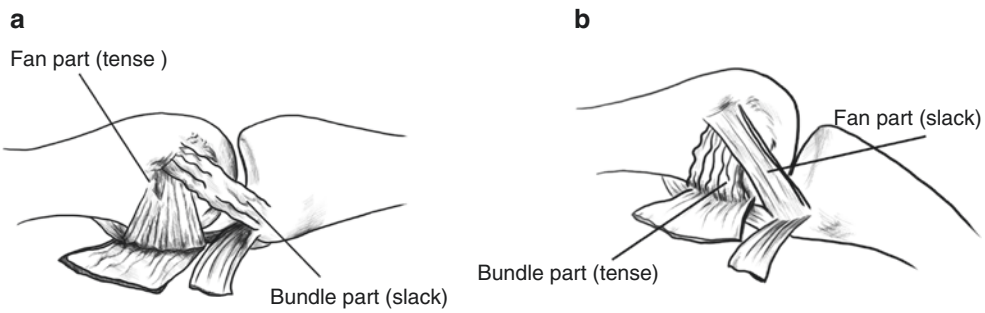


Fig. 6.65 Tension of accessory ligaments of metacarpophalangeal joint capsules. (**a**) Volar interphalangeal joint extension position. (**b**) Volar interphalangeal joint flexion position

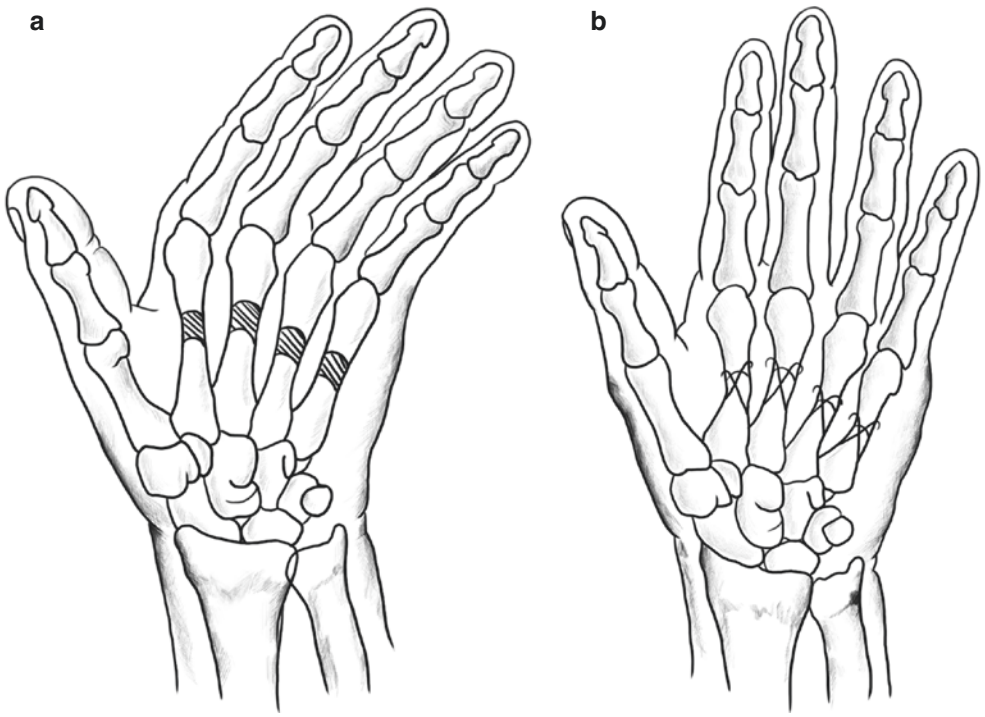


Fig. 6.66 Osteotomy below the head of the metacarpal bone to correct windblow hand deformity. (**a**) Before operation. (**b**) After operation (Redrawn from David P. Green. Operative hand surgery.1988,Second edition volume 1: 421)

for 6 weeks (Fig. 6.66). In the author's cases, they were given correcting treatment during the childhood or adolescence, and no one needed to be treated through osteotomy below the head of metacarpal bone.

6.7.7.4 Treatment of Young and Adult Patients

Ulkur E. et al. [82] reported the treatment of two adult patients with windblow hand deformities. These patients are often accompanied by the deformities in osteoarticular ligaments; therefore, the use of splints for fixation almost produces no curative effect, and surgical treatment is needed to improve the appearance and functions of the affected limb. Surgery is performed to release the contracture of volar skin soft tissues of the second to fifth fingers. Proximal or distal flaps are used for the release of the first fingerweb, and Kirschner silk is used to maintain the normal width of the first fingerweb. As for the moderate fingerweb contracture, four-flap method can be used; if necessary, the adductor and the first interosseous muscle can be disjuncted. The treatment of tendons and ligaments includes centralized treatment of extensor tendons and crossed displacement of hand internal muscles (grafting the ulnar hand internal muscles and fixing them to the radial side of the adjacent finger). The Z-plasty method is used to lengthen the tendon of the shorted thumb breviflexor. The main bone processing method is performance of wedge-shaped osteotomy below the head of metacarpal bone, the metacarpal bones are fixed with steel plates and screws after operation, and dynamic splints are used for 4 months.

6.7.7.5 Treatment of Windblow Hand Deformity Induced by Abnormal Muscles

Lanz U. et al. (1994) [77] reported three cases of congenital ulnar deviation hand deformity, and abnormal muscles were found to end at the ulnar side of the metacarpophalangeal joint in all of them. Removal of these abnormal muscles during operation, subsequent phase II surgery including crossed displacement of hand internal muscles, and postoperative treatment with dynamic splints have all produced good treatment effects.

To sum up, the etiologic factors and clinical manifestations of each patient with congenital windblow hand deformity vary; therefore, the Wood's viewpoint that "each patient needs an individualized therapeutic regimen" is correct and can serve as the reference for hand surgeons [83].

6.7.7.6 Dynamical, Pathological, and Anatomical Analyses

Dynamical, pathological, and anatomical analyses are the bases for the selection of surgical methods of windblow hand deformity.

Windblow hand deformity is a type of syndrome with the manifestations of multifunctional anatomical defects. According to the author's many years of experiences in clinical treatment and the review of relevant literature, people still have a lot to study on windblow hand deformity. Therefore, each patient should be carefully examined, and dynamical, pathological, and anatomical analyses and all-round evaluation of hand functions should be conducted. These are also the bases for decision-making on the correcting surgery of windblow hand deformity.

6.7.8 Typical Case

6.7.8.1 Case One: Correction of Mild Windblow Hand Deformity

A male pediatric patient visited the hospital for the first time at the age of 4 and a half, showing the manifestations of windblow hand deformity in both hands, thumb flexion and adduction deformities, and ulnar deviation of metacarpophalangeal joint. After the release and lengthening of thumb breviflexor tendon, thumb web expansion, rotation and grafting of pedicle skin flap at the dorsal side of the index finger, transplantation and repair of free skin grafts at the donor sites, and application of postoperative scaffolds, the volar skin and soft tissue contracture of each finger is released, and the palmar fascia is released. After a 7-month follow-up visit, the windblow hand deformity was basically corrected (Fig. 6.67).

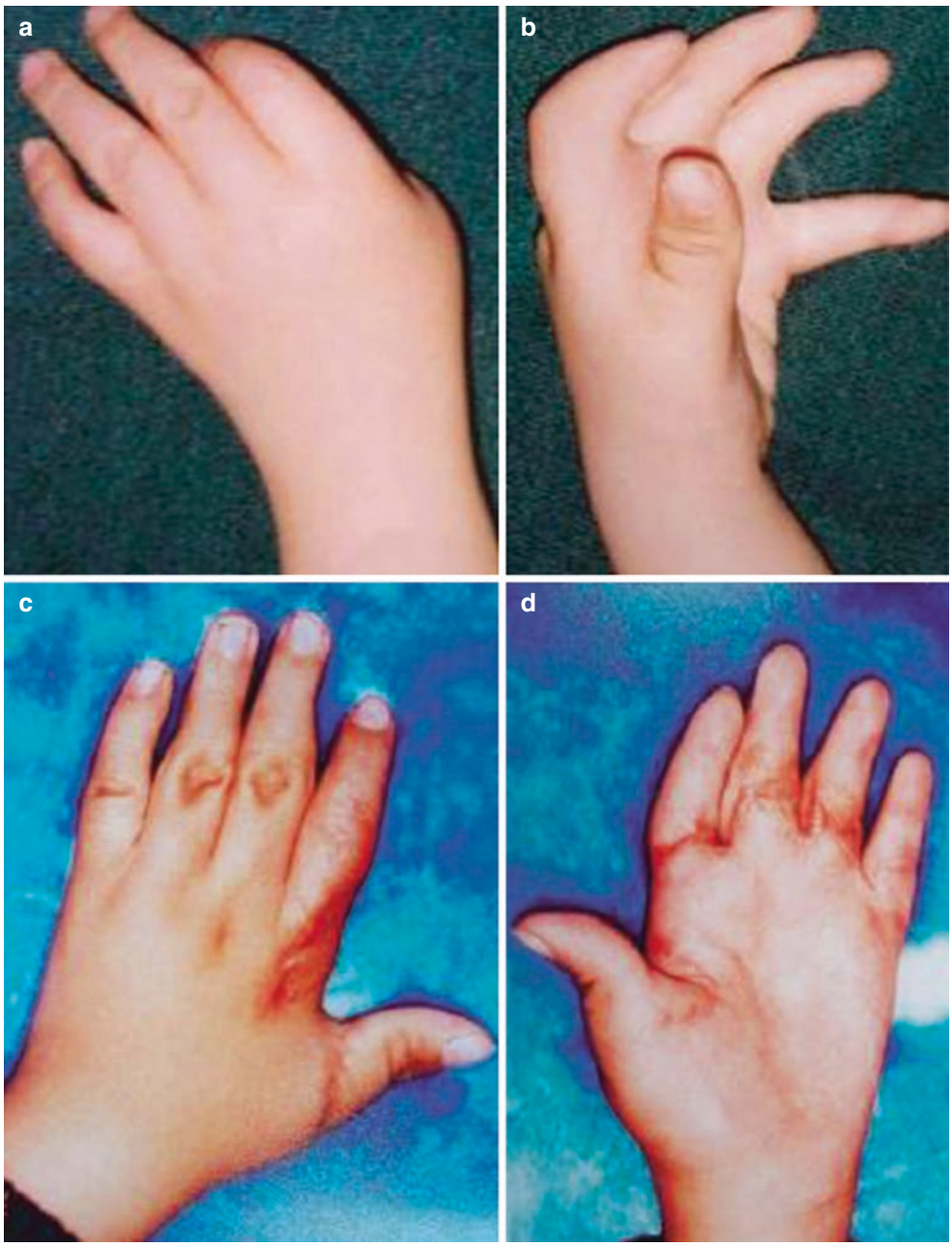


Fig. 6.67 Case one: correction of mild windblow hand deformity. (**a**, **b**) Before operation on windblow hand deformity of the left hand. (**c**, **d**) The hand dorsal view and dorsal view after a 7-month follow-up visit after operation

The thumb flexion and adduction deformity is not only expressed as shortening of the skin but also expressed as shortening of the thumb vascular nerve bundles. The contracture of the thumb breviflexor is another important cause. During the surgery, it is often necessary to lengthen the thumb breviflexor tendon, but the range of lengthening of this surgery is limited, so the insertion of this muscle is always peeled off to antedisplace the insertions of the muscle. After the antedisplacement of the insertions of the thumb breviflexor, the passive extension of the thumb can create a high tension. One of the methods the author would adopt is to antedisplace the volar plate of the metacarpophalangeal joint to correct thumb flexion deformity. After the operation, the Kirschner silk can be used for temporary fixation for 3 weeks when the metacarpophalangeal joint is in the extension position; after the Kirschner silk is pulled out, the splints are used for fixation for 4 weeks until the deformity is improved. As for serious thumb flexion deformity, the Z-plasty of thumb long flexor can also be considered, and it is suitable to lengthen the thumb long flexor tendon at the forearm. If the tension of the thumb long flexor is still too high, it is necessary to fold and shorten the thumb extensor. If the thumb still cannot be abducted completely, the inherent extensor tendon of the index finger can be grafted to the thumb long extensor tendon. In order to completely correct the thumb adduction surgery, the partial insertion of the thumb adductor at the third metacarpal bone can be disjuncted and released during the operation; if correction is still impossible, the partial insertion of the first dorsal interosseous muscle at the first metacarpal bone can be disjuncted and lengthened simultaneously, and the thumb is passively extended and fixed with Kirschner silk in the abduction position for 3–6 weeks. Before operation, function examination and accurate evaluation of the affected hand are very important. In case of thumb adduction and flexion deformities induced by thumb extensor and thumb abductor dysplasia, then the dynamical tendon grafting method can be adopted to repair the dysplasia of dynamical muscle. As for the reconstruction of the palm opposition functions of the thumb, the method of finger superficial flexor tendon grafting of the index finger can be used for opposition dynamical reconstruction; another method is Royte-Thompson method, including rotational osteotomy and the radial grafting of finger superficial flexor tendon of the ring finger [80]. The author adopted the less invasive contracture correction and dynamical reconstruction, both of which have produced good effects.

6.7.8.2 Case Two: Correction of Moderate Windblow Hand Deformity

A 6-year-old female pediatric patient visited the hospital with the manifestation of windblow hand deformity in both hands, thumb adduction and flexion deformities, thumb web stenosis, palmar contracture, flexion deformity of the remaining four fingers, the ulnar deviation of metacarpophalangeal joint of over 30°, and obvious drop in the muscular strength of thumb extensor and abductor; her mother has similar symptoms. The pediatric patient underwent the surgical correction of windblow hand deformity of the left hand in October 2003, during which the radial wrist long extensor grafting was used for the functional reconstruction of the thumb long extensor and the ulnar wrist flexor grafting was used for the functional reconstruction of the thumb abductor, the reshaping of the metacarpophalangeal joint of the index, middle, and ring fingers, and the finger extensor tendon hoods, including the resection of the triangular ligaments, the release of lumbrical muscles, the correction of military salute hand, the radial incision and folding of the tendon hoods of the index, middle, and ring fingers, and the ulnar longitudinal incision and release; finally, the finger ulnar deviation deformity was effectively corrected. The finger ulnar deviation of the windblow hand deformity is mainly induced by hand internal muscle-lumbrical muscle congenital dysplasia. The lumbrical muscle originates from the profound digital flexor and ends at the finger extensor tendon hoods. Once the triangular ligaments are resected, the tendon of the lumbrical muscles is released and then the contracture can be corrected. After the release of the tendon hoods of the index, middle, and ring fingers and the lumbrical muscle tendons and correction of the ulnar deviation of the metacarpophalangeal joint of the three fingers, surgery on the extensor tendon of metacarpophalangeal joint of the little finger can be avoided, and the ulnar deviation of the metacarpophalangeal joint of the little finger can be passively corrected. In the meantime, the thumb web expanding and palmar aponeurosis releasing surgeries can be performed. A follow-up visit was paid in Mar. 2004, and in the meantime, the surgical correction of windblow hand in the right hand was performed (Fig. 6.68).

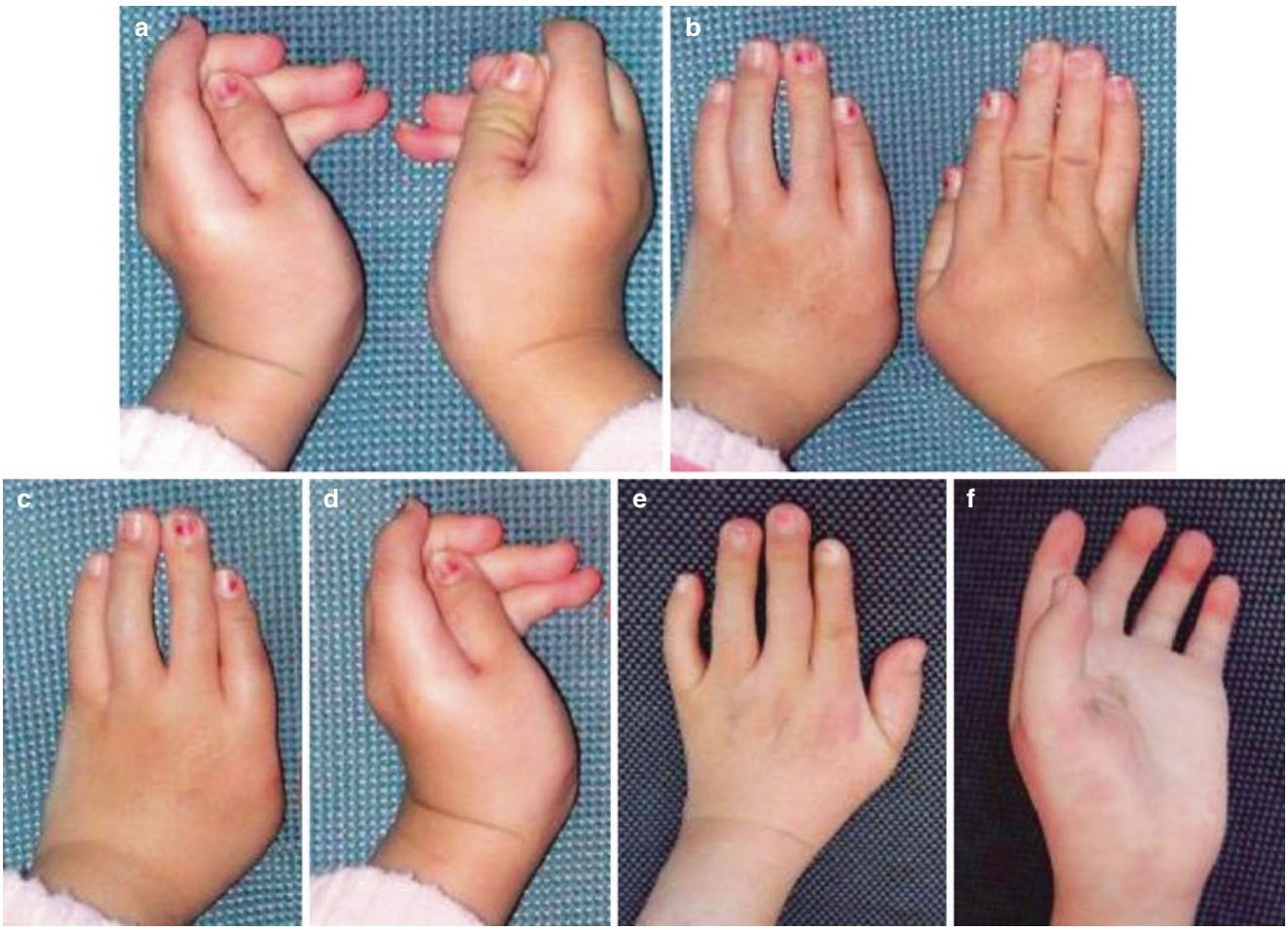


Fig. 6.68 Case two: correction of moderate windblow hand deformity. (**a**, **b**) Before the surgery of windblow hand deformity in both hands. (**c**, **d**) Before the surgery of windblow hand deformity in the left hand. (**e**, **f**) After the surgery of windblow hand deformity in the left hand. (**g–i**) Before the surgery of windblow hand deformity in the right hand. (**j**, **k**) Windblow hand deformity in the right hand, immediately after operation and before operation. (**l**) During the surgery, the removal of the triangular ligaments of the index finger is displayed. (**m**) Removal of the triangular ligaments of the middle finger extensor tendon, radial folding and shortening of the extensor tendon hoods, ulnar longitudinal incision and release, and fixation with Kirschner silk. (**n**) Schematic diagram of correcting surgery of ulnar deviation of metacarpophalangeal joint of the fingers. (**o**) Partial tendon grafting of the hand, dynamical reconstruction of thumb extension and thumb abduction, and fixation with Kirschner silk

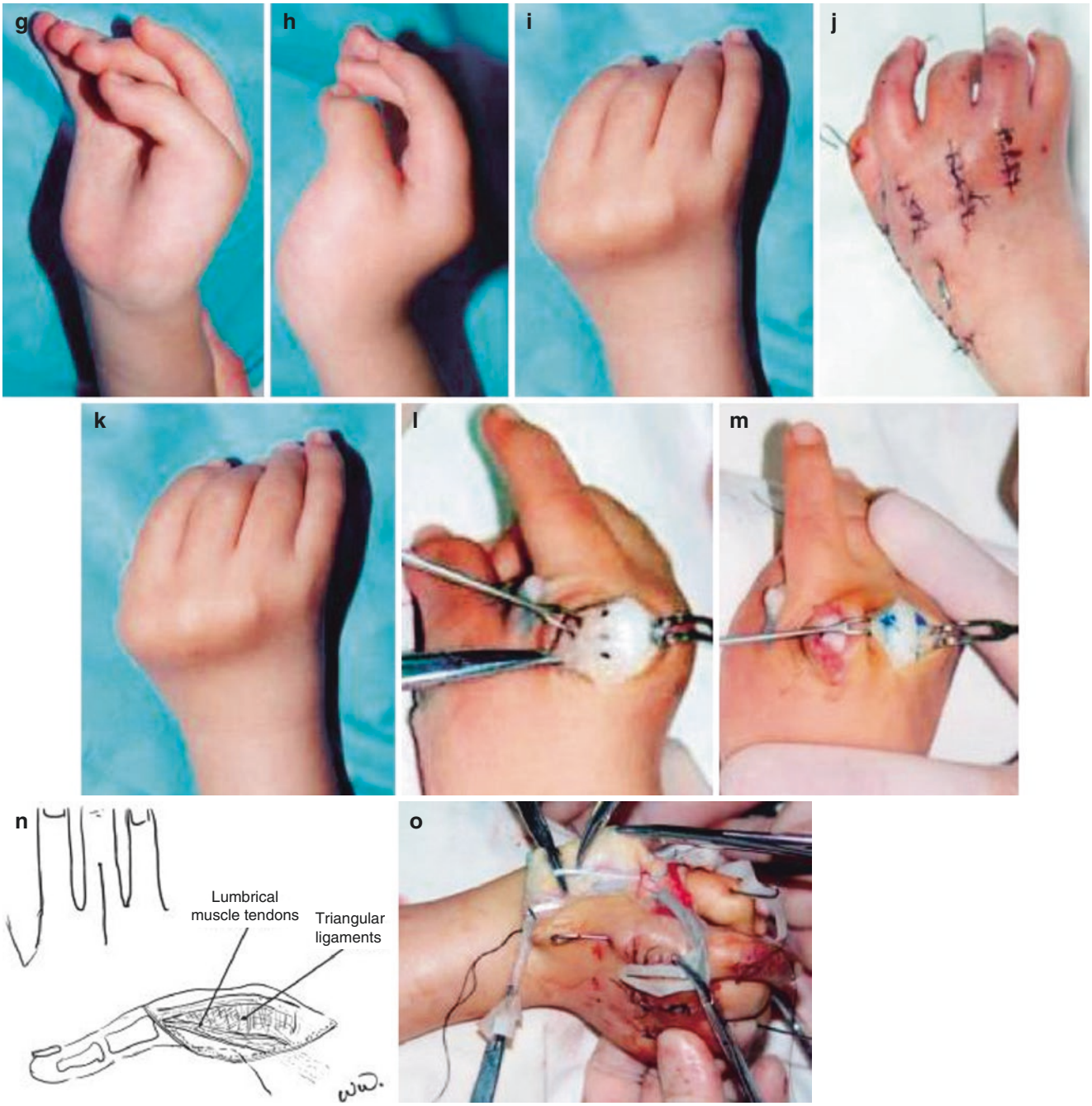


Fig. 6.68 (continued)

6.8 Arthrochalasis

Zhenyu Bian and Liulong Zhu

6.8.1 Overview

Joint laxity (arthrochalasis), also called joint hypermobility syndrome and hypermobile joint syndrome, is one of the reasons for pain in four limbs [84]. Kirk et al. [85] first reported the joint hypermobility syndrome [86] in 1967 and subsequently used different names to call this syndrome, such as hypermobile joint syndrome [87] and benign joint hypermobility syndrome (BJHS) [88, 89]. This disease refers to the appearance of musculoskeletal symptoms on the basis of joint hypermobility, but it is not accompanied by generalized rheumatic diseases. This disease is a genetic connective tissue disease [90] and is initially considered as the orthopedical clinical manifestations of Ehlers-Danlos syndrome (EDS). The most important manifestations of this disease are the wide relaxation of generalized joints, concomitantly with the thin skin, an obviously higher extensibility than normal people, and varicose. The manifestations during the infancy are abnormal limb activities, late learning of walk, frequent fall, and the medical history of multiple sprains and joint dislocation during growth. The pediatric patients are often accompanied by deformities such as equinovarus, flat foot, scoliosis, and chicken breast [85].

As extensive and in-depth studies have been carried out on this disease during the recent 40 years, Beighton et al. [91] described this disease as familial articular hypermobility syndrome in the international classification of hereditary connective tissue disorder (INHDCT), and in the meantime the hereditary connective tissue disorders that involved joint hypermobility were ruled out, such as Ehlers-Danlos syndrome, Marfan syndrome, and osteogenesis imperfecta. Because the arthrochalasis does not have fatal complication [92] and only manifests the significantly higher joint mobility, some scholars call it benign joint hypermobility syndrome [93]. At present, this concept has been increasingly adopted by more and more surgeons.

6.8.2 Epidemiology

The systemic joint laxity is very common in the population without chief complaint, and the incidence of joint hypermobility without systemic diseases among the population is 4–13% [94, 95]. With the increase in age, the joint hypermobility gradually subsides, and it is associated with gender and race [95]. On the whole, females' joints are laxer than males' joints, and among the healthy women, only 5% suffer from joint hypermobility symptoms, and the proportion in men is only 0.6% [96]. Populations of African descent, Asian descent, and Middle Eastern descent have relatively lax joints [97–99].

The epidemiological survey indicates that, due to the influence of such factors as race, age, and gender, the proportion of this disease among the populations varies greatly. In the fourth century BC, Hippocrates noticed that the joints of the Scythians from the Black Sea and Caspian coastal regions had a very high range of motion, and these people could not draw the bows or throw the javelin due to joint laxity [100]. The epidemiological survey on the arthrochalasis among the Caucasians indicates that the incidence of the arthrochalasis of those with a soft physique is 5–7% [88, 101]. The results of the investigation on the population in India, Iraq, and Africa carried out by Beighton et al. indicated that, among the Asian and African population, the incidence of this disease was significantly higher than that among the Caucasian populations [102]. An epidemiological survey carried out by Seckin et al. [95] on arthrochalasis among the American senior high school students indicated that the male patients with arthrochalasis accounted for 7.2% of all the tested males and the female patients with arthrochalasis accounted for 16.2% of all the tested females. The epidemiological survey on soft physique carried out by Jansson et al. [103] on 1845 students in Sweden indicated that the proportion of those with a soft physique in the 15-year-old male group was 15.5% and that in the 15-year-old female group was 53.3%. The result was the same as the result of this study, and all of the above investigations supported the gender-related difference of arthrochalasis in epidemiological survey, that is, the proportion of the arthrochalasis of females with a soft physique was significantly higher than that of males.

6.8.3 Etiologic Factor

Joint laxity is an autosomal dominant inheritance that affects the coding of the connective tissue protocollagen [104]. Some studies reveal that type III collagen and type I collagen ratio of the patients with arthrochalasis is abnormal [105]. The tensile strength of type I collagen is high, and it is the commonest collagen in vivo and has a high content in tendons, joint capsules, skin, demineralized bones, and neuroreceptors; present in cartilage, type II collagen can bear compressive stress; the extensibility of type III collagen is stronger, and it is arranged in disorder and present in intestinal tracts, skin, and vessels [106], which can explain the laxity in heredity or drop in tissue hardness [107]. Recent studies have indicated that this disease is associated with the mutation of the genes that encode type V collagen [108]. Under the normal circumstances, type V collagen can interact with type I collagen during collagen fiber formation, the diameter of collagen fibers can be adjusted, the changes

during this process can potentially cause the collagen fibers to become thin, and the arrangement is more in disorder. The fibroblast biopsy analysis enables researchers to further know the differences in fiber structures of the hereditary connective tissue disorders. Malfai et al. [108] predicted that the processing of N terminal propeptide of α chain (α1 or α2) of type I collagen was interrupted, resulting in Ehlers-Danlos syndrome like symptoms, including skin laxity and joint semiluxation and luxation. The nerve system of this patient was also involved, and some scholars reported that the injection or the surface application of local anesthetics turned out ineffective on this patient and its mechanism is unknown [109, 110]. Studies revealed that this patient's ability to repeat the proximal interphalangeal joint angle was not as accurate as that of healthy people [111]. Other studies indicated that the patient's knee joint topesthesia decreases, especially the ability of locating the range of extension terminal decreases [112]. The laxity and brittleness of connective tissues, the decreased sensitivity of proprioception, and the changes in the reflectivity of the nerve muscles are the possible reasons why the patients with arthrochalasis are apt to injuries [113, 114].

6.8.4 Clinical Manifestation

The symptoms and physical signs of arthrochalasis are various. The patient's original chief complaint is arthralgia, which may involve one or multiple joints. The symptoms can be systemic or symmetric, and the pain has a wide range and a long duration which ranges from 15 days to 45 years [115]. In addition, the patient will describe many joint-related symptoms, such as rigidity, snap, semiluxation, luxation, stability, and sensation of weak joint; there are also many symptoms that affect other tissues, such as paresthesia, fatigue, asthenia, sensory discomfort, and flu-like symptoms [116]. Sometimes, it is very difficult to match the patient's chief complaint with his/her appearance and behaviors [117] because the patient's appearance and behaviors are often good. Under very rare circumstances, the patient will be misunderstood [105]. Under the very extreme circumstances, the patient can be considered to suffer from decompression and is labeled as a patient with psychological problems. During the first visit, the surgeons can use five simple questions to recognize the joint laxity [118]: ① Can you or could you lay your hands flat on the ground under the condition that your knees are not bent? ② Are you or were you completely able to use a thumb to touch the forearm? ③ When you were a child, were you able to wrist your body into a strange shape to amuse your friends, or can you do the splits? ④ During your childhood or adolescence, were your shoulder joints or knee joints often dislocated? ⑤ Do you think you have two-segment joints?

Arthrochalasis can occur at any age, and many patients are grafted to department of orthopedics, department of rheumatism, or department of rehabilitation. The typical manifestation is that self-limiting pain at multiple joints during the childhood can occur, the pain can last for a relatively long period of time, and the pain can become continuous when the patient becomes an adult. The pain can involve any of the systemic joints, but the commonest one is the knee and ankle, and it is predicted that they are associated with burden bearing. Physical activities or repeated use of involved joints will aggravate the pain; therefore, the pain will occur at later time of each day, and early morning stiffness is not common. Rare symptoms include joint rigidity, muscular soreness, muscular spasm, and limb pain in non-joint sites. The patients often believe that they have two-segment joints, and the patients can wrist their body into strange shapes (namely, autonomously controlled semiluxation) or do the splits easily. However, these chief complaints are not necessary for the diagnosis of arthrochalasis (including differential diagnosis). Such patients may have the medical history of shoulder or patellar dislocation or may have the family history of two-segment joints or repeated luxation, and other manifestations include easy tendency of bruising, ligament or tendon rupture, congenital hip joint dysplasia, and functional disorder of temporomandibular joint [97, 117, 119]. The extraarticular manifestations of this disease include skin brittleness and laxity, dysautonomia, blepharoptosis, varicose and cyanosis, genitourinary tract prolapse, Raynaud's phenomenon, delayed developmental movement coordination, changes in neuromuscular reflexes, neuropathy, tarsal tunnel and carpal tunnel syndrome, fibromyalgia, low bone density, anxious and panic status, and decompression.

Arthrochalasis does not necessarily result in problems. Sometimes it can be considered to be a gift [120]. However, for those unfortunate patients, joint hypermobility and tissue laxity may be the reasons for multiple debility symptoms. Symptoms often occur during the childhood and can last until the adult stage [121]. According to a study report, three-fourths of the patients with arthrochalasis will show symptoms at the age of 15 [85]. According to Lewkonia [122] and Murray et al. [123], joint laxity was one of the commonest reasons for musculoskeletal symptoms among the children and adolescents aged between 13 and 19, especially the girls.

The physical examination results can vary due to differences in the involved joints. The appearance of pain after joint activities is very common, and mild dropsy can occur and is not common. The patients with this disease will not clinically be advised of significant tenderness with inflammation, reddening, edema, and fever [94, 97], but may show typical connective tissue disease physical signs, including scoliosis, flat foot, genu valgum, lumbar lordosis, patellar semiluxation or luxation, Marfan syndrome, varicose, rectal or uterine prolapse, thin skin, etc. The connection between

Table 6.5 Neuromusculoskeletal symptoms and sequelae of joint laxity

Acute (traumatic)	Chronic (non-traumatic)
Sprain	Soft tissue rheumatism
Repeated ankle sprain	Tendinitis
Meniscal tear	External humeral epicondylitis
Acute or habitual luxation or semiluxation of the following joints	Rotator cuff syndrome
Shoulder joint	Synovitis
Knee joint	Adolescent occasional synovitis
Metacarpophalangeal joint	Bursitis
Temporomandibular joint	Chondromalacia
Traumatic arthritis	Lumbago
Cyanosis	Scoliosis
Fracture	Fibromyalgia
	Functional disorder of temporomandibular joint
	Nerve entrapment syndrome
	Carpal tunnel syndrome
	Tarsal tunnel syndrome
	Acroparesthesia
	Thoracic outlet syndrome
	Raynaud syndrome
	Flat foot and its sequelae
	Non-specific joint pain or joint (foot, ankle, knee, hip, neck, shoulder, elbow, wrist, and finger) effusion
	Osteoarthritis Motor function retardation
	Congenital dislocation of the hip

arthrochalasis and mitral valve prolapse is controversial; the early studies indicate that arthrochalasis and mitral valve prolapse are correlated [124], but subsequent studies question the relevancy due to the presence of stricter diagnosis standard for the echocardiogram for mitral prolapse [89]. During the first visit, the surgeons should graft joint laxity patients with the manifestation of mitral prolapse to the department of cardiology to further evaluate or rule out more serious heart abnormalities and connective tissue disorders (Table 6.5).

6.8.5 Diagnostic Criterion

The diagnosis criteria of arthrochalasis develop gradually with the constant deepening of the understanding of it, and they are being constantly discussed. Although the concept of "hypermobility syndrome" was proposed in 1967, the relation of joint injuries and diseases with joint laxity was reported earlier. In 1958, Carter and Sweetnam found that females' joints were laxer than males', so that females were more apt to knee joint dislocation [125]; in 1960, they noticed the same factors that made women more apt to shoulder joint dislocation [126]. Carter and Wilkinson examined 285 normal British school-age children and found that those showing joint hypermobility accounted for 7% [101]. Through the above work, they formulated the diagnostic criterion that was initially used to determine the degree of joint laxity. Although this criterion was easily subject to the interference of subjective factors so as to affect the determination results, during the early stage, the criteria are still applied to the majority of investigations on arthrochalasis. In 1969, Beighton et al. modified this diagnostic criterion, and the modified diagnostic criterion is called Beighton's scale [127] (Table 6.6). It is considered to be the most suitable criterion and has been widely applied. Beighton's scale not only contains the tests on the range of activities of big and small joints but also contains the comprehensive tests on the range of activities of the body trunk and hip joints. As for the determination items, Beighton et al. selected the items with the criterion easy to be accurately mastered; the determinations of ankle joint dorsal extension and strephexopodia degree were canceled as they were hard to accurately master. In the tests on fingers, Beighton et al. believed that the measurement only on the middle finger or all the five fingers was inaccurate, so they only measured the passive dorsal extension of

Table 6.6 Beighton's diagnostic criteria on arthrochalasis

• Primary criteria
Beighton's scale ≥4 points
Pain in four or more joints for more than 3 months
• Secondary criteria
Beighton's scale ranges from one to three points
Pain in one to three joints for over 3 months, or lumbago for over 3 months, or with cervical spondylopathy, rupture of isthmic portion of the pedicle of vertebral arch, and olisthe of lumbar vertebrae
With the history of joint luxation or semiluxation in more than one site or suffering joint luxation for more than once in one site
At least three soft tissue lesions, such as external humeral epicondylitis, tendovaginitis, and bursitis
Marfan syndrome: tall, slim body figure, arm span > height (arm span/height > 1.03), upper part < lower part (upper body/lower body <0.89), and arachnodactyly syndrome
Skin streaks, abnormal increase in skin elasticity, thin skin, or abnormal scarring
Eye signs, such as ptosis, myopia, and antimongoloid slant
Varicose, hernia, uterine or rectal prolapse
Mitral valve prolapse
• The diagnosis can be confirmed in case of the presence of the following circumstances in the diagnostic criteria
Two primary criteria
One primary criterion + two secondary criteria
Four secondary criteria
Two secondary criteria and family history (confirmed patients among the first-level direct relatives)

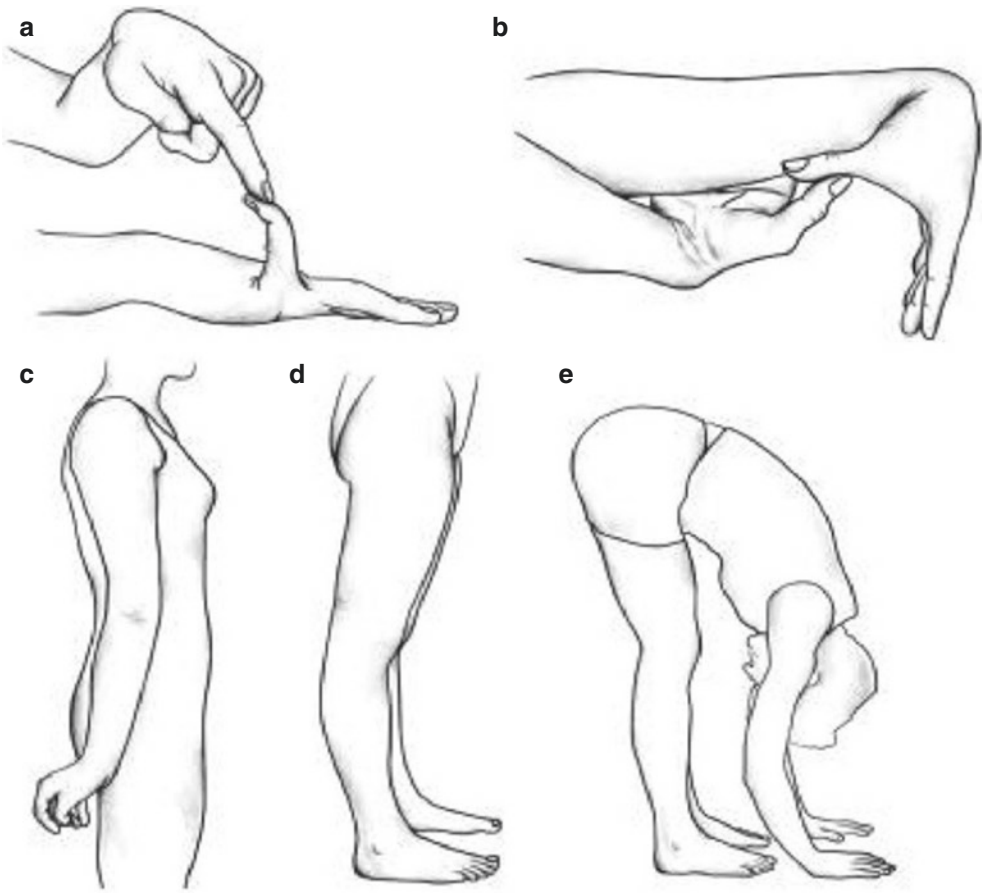


Fig. 6.69 Beighton examination on arthrochalasis. (**a**) Passive dorsal extension of the little finger of over 90°. (**b**) During passive motion, the thumb can palpate the flexion side of the forearm. (**c**) The hyperextension of the elbow joint of over 10°. (**d**) The hyperextension of the knee joint of over 10°. (**e**) The knee joint is maintained in the extension position, and when bending forward, the palm can more easily touch the ground

the little finger. The Beighton's diagnostic criterion is easily subject to the interference of the subjective factors, so many surgeons diagnose the arthrochalasis through the measurement of the range of motion of the joints.

Beighton's scale is necessary for the confirmed diagnosis of arthrochalasis. The first step is to calculate the Beighton's scale, namely, examine the degree of laxity of systemic joints. The surgeons can get this scale through letting the patient perform five simple actions, and these actions can be performed within 45–60 s (Fig. 6.69). One point is scored for each positive action [128], and four or more points of the Beighton's scale indicate the presence of systemic joint laxity. The judgment can then be made based on the patient's medical history (family history), symptoms, and physical signs and according to the diagnostic criteria for the purpose of confirming the diagnosis of arthrochalasis. Adopting these criteria can also help the surgeons to distinguish the arthrochalasis with other connective tissue disorders [94, 117, 119].

The diagnosis of arthrochalasis is preclusive; as for the patients with joint swelling or pain, what is important is to rule out the possibility of infection or autoimmunity. Clinical examination includes detection of whole blood cell count, blood sedimentation rate, rheumatoid factors, antinuclear antibody, serum complement (e.g., C3, C4, and CH50), and serum immune globulin (e.g., IgG, IgM, and IgA), and these examination results are not within the normal reference value range, which indicates other diagnoses.

The joint laxity of most patients with arthrochalasis is symmetric. Sometimes, the patients with arthrochalasis have articular hydrops, which is induced by meniscus or articular cartilage being simulated, and the joint puncture fluid is noninfective.

Arthrochalasis needs to be differentiated with other diseases that share the same characteristics, such as Marfan syndrome, EDS, and osteogenesis imperfecta. The systemic joint laxity is the common feature of all hereditary connective tissue disorders, and these features overlap, but there are also some distinguishable features that facilitate the differentiation of these diseases.

6.8.6 Treatment

The first step to treat arthrochalasis is pointing out to the patient that this disease is a nonprogressive and noninflammatory connective tissue disorder [117, 119, 129]. Effective therapies include change of lifestyles, adjustment of mode of exercise, protection of joints, and proper gymnastic exercise.

6.8.6.1 Treatment During the Acute Stage

Nonsteroidal anti-inflammatory drug (NSAID) or acetaminophen is commonly used to relieve pain. It is generally believed that the joint pain is not induced by inflammation; therefore, NSAID is only used for pain treatment, and it is not recommended to treat the symptoms other than pain [97, 98, 117]. NSAID cannot be used by patients with peptic ulcer, serious liver and renal lesions, serious heart functional insufficiency, and serious blood system abnormalities or the lactating women. As for moderate or severe pain, rest and the discontinuation of intense exercise can improve symptoms. Physiotherapy and joint protection can also be beneficial [98, 117].

6.8.6.2 Treatment During the Intermediate Stage

Usually, attention should be paid to change of the lifestyles, and the activities that can induce symptoms should be avoided. Joint hypermobility is associated with the appearance of the symptoms of arthrochalasis, and intense and repeated activities are often considered to be the factors that can aggravate this disease. Excessive training, excessive performance, or sports competition and activities that emphasize the flexibility rather than stability of the joints will increase the risk of joint pain and injuries [130]. If the patients cannot accept the suggestions to avoid these activities, the surgeons should adopt other methods. Before competitive sports, administration of NSAID can often relieve the symptoms. In addition, acceptance of the training plans that provide muscular and joint stability can be beneficial. The extension activities that apply the stress to the tense muscles instead of the adjacent joints can improve the symptoms through improving the balance and control force [117]:

1. Treatment plan. The Russet's report [117] points out the importance and necessity of education, therapeutic exercise, and accommodation and adjustment of work and lifestyles. For most patients, establishment of a list of the problems that should be preferentially solved as well as the short-term, medium-term, and long-term treatment objective agreed to by the patient is the key to successful treatment. The initial treatment often includes recuperative medical care of pain or injuries [116] during the acute stage by means of resting, walking, joint maintenance, use of certain physical treatment devices (including ultrasound and transcutaneous neurostimulation, bandages and splints, heat and ice), gentle application of relevant hypomobile regions, massage, muscle energy techniques (MET), and acupuncture and moxibustion. MET refers to the active contraction and relaxation of a certain designated muscle according to the instructions of the surgeon (therapeutist). The contraction intensity can vary according to the patient's conditions, and it can be adjusted according to the different counter forces given by the surgeon (therapeutist). MET can lower the excessively high muscular tension, lengthen the shortened fascia in muscles, increase the range of motion of the limited joints, and help the integration of the sensation and movement.
2. Physical therapy. As the rehabilitation and cure are often slow [131], special attention should be paid when the physical therapy is adopted because the pain is usually hidden and can often become aggravated. The doctors must be aware of the increase in the brittleness of the connective tissues, which should be considered in deciding the treatment dose. As an effective arthrochalasis-treating method, massage can relax joints, relieve pain, and improve blood supply, lymphatic return, and proprioception [124].
3. Proper exercise. Including the combination of open kinetic chain (the distal limbs can move freely) and closed kinetic chain (the distal limb is subject to resistance) exercise. During the closed kinetic chain exercise, the functions of limbs are often simulated, and the open kinetic chain exercise is more effective in providing oriented strength exercise [117]. The exercise that improves joint proprioception (e.g., application of swing plates) may improve the symptoms [112, 132]. Sometimes, the combination of supporting splints with suitable shoes can protect the joints, and supporting joint bandages can improve the proprioception of joints [132]. The special exercise that improves muscular strength, balance, and coordination helps improve joint stability. The improvement of proprioception can reduce the sprain of the ligaments around the joints and avoid further injuries [112, 132], including ① exercise on the stability of the trunk and limb joints as well as enhanced exercise of proprioception, ② systemic physical strength exercise to offset or reverse poor somatic tendency, and ③ adoption of motor technology to recover the natural overexercise of the joint or spinal column segments lost due to the disturbance and the fear for damages [133].
4. Others. The treatments should also include behavioral correction to some extents, e.g., walking techniques, coping strategies, handling of the problems in human engineering, work and lifestyles, as well as dietary suggestions on irritable bowel syndrome, nutritional supplement, and weight management. Arthrochalasis is commoner in females; therefore, attention should be paid to incontinence, pregnancy, care of infants, etc. As is described above, due to fear and decreased motions, the patients are often at the disturbance status and can easily become the high-risk group with other diseases [104]; therefore, this can be avoided through regular exercise to improve the level of health. Because decreased physical activity is a known risk factor for many systemic diseases, seeking and establishing the methods to encourage the patients to

engage in physical activities is an important part of arthrochalasis treatment [133].

As for the patients with chronic pain, analgesics are often ineffective [134]. The pain-treating procedures implemented by specially trained psychologists on the basis of cognitive behavior techniques can ease pain and anxiety and reduce the influence of pain on daily life [135]. As for patients with problems in both hands and feet, they can be grafted to the specialist in hand and foot diseases so that the doctor can perform mechanical evaluation of feet or grafted to the occupational therapist who can assist them with the writing and other work-related hand problems.

6.8.7 Prognosis

As arthrochalasis is nonprogressive and the arthrochalasis degree decreases with the increase in age, the prognosis of this disease is often good. However, the patients should be aware of some of the potential complications of this disease, including acute ligament and soft tissue injuries, overuse injury, unstable joints, bone fracture, scoliosis, and uterine and rectal prolapse, and their incidence is increased. In addition, these patients are apt to suffer from osteoarthritis due to long-term joint hypermobility [98]. Studies reveal that arthrochalasis has a connection with panic disorder [136]. In spite of the presence of these sequelae, the patients should try to do more activities as far as possible. Changing the exercise plan can avoid chronic joint pain. Good exercise strategies include slow and regular exercise, conformity with correct biomechanical principles, and improvement of proprioception [130]. The potential complications of arthrochalasis indicate the importance of early diagnosis and patient education.

6.8.8 Relevant Disease

Arthrochalasis is considered to be benign, but there are some diseases accompanied by joint hypermobility, such as serious complications involving the cardiovascular system, nerve system, and skeletal system [137–141]. There are also some joint laxity-related genetic diseases, and most of them are familial.

6.8.8.1 Marfan Syndrome

Marfan syndrome, first described by Marfan in 1896, is a group of clinical syndromes induced by congenital interstitial tissue defects. It has a potential fatality and can involve the skeletal system, visual system, and cardiovascular system. The common symptoms include tallness, slim body figure, multiple joint hypermobilities, thin and long fingers, myopia, and dislocation of lens [138].

6.8.8.2 Osteogenesis Imperfecta

Osteogenesis imperfecta, also called brittle bone disease, is a type of systemic connective tissue disorder. Its lesions are not limited to skeletons because other connective tissues such as the skin, fascia, tendon, ligament, artery, and cornea are also usually involved. The common genetic mode of this disease is autosomal dominant inheritance, the rare mode is autosomal recessive inheritance, and the genetic mode varies greatly with individual. Its incidence is low, it is not obviously race related, and it is common in females. This disease features multiple fractures, blue sclera, progressive deafness, dental changes, joint laxity, and abnormal skin. The patients often visit a doctor after bone fracture. Bone brittleness is the outstanding manifestation of this disease, and mild trauma, even muscular contraction, can also induce bone fracture. Clinically, it can be divided into two categories: congenital dysostosis and delayed dysostosis. Patients with congenital dysostosis can suffer fracture in over 100 sites at birth; the patients with delayed dysostosis do not suffer fracture until childhood. The later the symptoms occur, the milder the clinical manifestations are [140].

6.8.8.3 Ehlers-Danlos Syndrome

Ehlers-Danlos syndrome is a type of connective tissue disease that includes joint hypermobility, purple papyrus-shaped scar, abnormal skin elasticity, and skin brittleness and easily causes injuries. Its genetic mode, like that of arthrochalasis, is also autosomal dominant inheritance, leading to connective tissue collagen defects and functional abnormalities. Arthrochalasis is the most similar to the type III (the joint hypermobility type), including joint pain, outstanding joint hypermobility, mild extraarticular symptoms, mild skin changes, and scars. Some researchers believe that arthrochalasis may be a benign classification of Ehlers-Danlos syndrome because they have similar genetic modes and similar clinical symptoms [137, 141].

6.8.8.4 Others

Other joint laxity-related diseases include Achard syndrome, homocystinuria, and hyperlysinemia.

As is verified by relevant studies, the anterior and posterior displacement of shinbones has a close relationship with the laxity of knee joint ligaments and the injuries in anterior cruciate ligaments [142–144], so when the cruciate ligaments are reconstructed in patients, attention should be paid to screening the arthrochalasis. As is validated by studies, under the conditions of the same surgeons and the same surgical mode, the effect after the reconstruction on the injuries of posterior cruciate ligaments in the arthrochalasis group is poorer than that of the normal people group [144]. In 1979, Bird et al. [145] studied 35 students with sports talent and 36 students without sports talent who were at the same age group without gender-related difference and then concluded

that long-term physical exercise would not lead to arthrochalasis. In some sports with a requirement for flexibility (including gymnastics, dancing, football, basketball, etc.), female athletes are more apt to athletic injuries than male athletes, and injuries in anterior cruciate ligaments are especially common [146]. In spite of no related results of epidemiological survey on arthrochalasis, all the researchers believe that the large range of motion of women's joint is one of the main pathogenic factors [147]. When selecting athletes, we should screen the patients with arthrochalasis. Although some athletes have some advantages in performance of some technical actions, they are more apt to motor injuries such as joint dislocation and ligament rupture than average people [85, 101, 102, 125, 126], so supplementary exercise should be taken during their training. Take the knee joint as an example. In order to strengthen joint stability, athletes with arthrochalasis should strengthen exercise of hamstring muscle to consolidate protective effects on cruciate ligaments [148].

Arthrochalasis is closely associated with joint diseases and injuries such as shoulder joint dislocation [102, 126], knee joint dislocation [102, 126], ligament injuries [101, 102], and osteoarthritis [92, 101, 102]; at the same time, the patients are accompanied by symptoms such as scoliosis, flat foot, and varicose [102], and it is very necessary to carry out a profound study on them.

6.9 Onychodysplasia

Jianmin Yao, Yizhen Chen, and Jinghong Xu

The fingernails, located at the tip of the finger, possess the function of fixing and protecting fingertips, preventing the rotation of finger pulp soft tissues, consolidating finger pulp tactile sensation, and strengthening and supporting finger functions (e.g., clutching, pinching, and scratching). The fingernails also have cosmetological functions. In case of absence or deformities, the appearance and functions of the fingers will be affected. Onychodysplasia is rare clinically but has a lot of varieties; sometimes it can result in patient's psychological disorder.

Fingernails are skin's derivatives, and the matrix cells at the nail root grow and advance closely along the nail bed and they constantly accept the proliferative and thickened epithelial cells on the nail beds during the process. From the perspective of section, the nail bed becomes thickened from the root to the anterior margin, with a growth of about 1 mm per week (the normal growth speed of fingernails of adults is 0.5–1.2 mm per week). It can be seen that the maintenance of the growth and shape of fingernails is co-completed by the nail root and nail bed. When predicted according to histological study, the various factors that affect, change, and damage the nail root matrix or the normal growth and development of epithelial cells on the nail bed during the embryonic period will lead to nail deformities.

6.9.1 Etiologic Factor

Onychodysplasia is mostly induced by environmental factors. It can also be caused by drugs, food, virus infection, and air pollution. In rare cases, it is the result of genetic factors.

6.9.2 Histological Basis

Lewis (1954) [149] divided a fingernail into three parts according to its color and luster: ① the nail dorsum formed by the fold epidermis of the proximal fingernail; ② the middle part of the fingernail from the nail root margin to the lunular distal epithelia, namely, the semilunar arch; and ③ the ventral part of the fingernail formed by the nail bed epithelia.

Instead, Zaias (1968) [150], after using radioactive imaging techniques, believed that the fingernail was only derived from onychostroma; Norton (1971) [151] and Shufuku Takefumi (1972) [152] adopted the nuclear medicine research methods and reached the same conclusion with Zaias; Suzuki Tateotto (1980) [153] found from the research on fetal nail formation that fingernails were only derived from onychostroma and were not associated with nail bed or proximal fold epithelia. The above views are the unitary theory on the source of fingernails.

It was previously believed that the nail bed could not be regenerated per se, but more studies and clinical observation have validated that the nail bed has the abilities of growing toward the distal side, but the speed is slower than that of fingernail. Norton [151] performed the nuclide amino acid labeling on the onychostroma and found that the cells absorbing this marker moved from the onychostroma to the nail bed and moved from the proximal side of the nail bed to the distal side. Zaias [154] labeled the silver nitrate on the nail bed and the root of the fingernail. Five weeks later, it could be seen that the silver nitrate at these two sites move toward the distal side. Masanori Watanabe [155] removed most of the nail beds of the apes, and the wounds were covered with skin flaps. Two to four months later, they were histologically observed, and it was found that granular layer cells and cuticular layers which were originally absent grew out of the residual nail beds, and they grew to distal position until the tip of phalange was reached, and gradually the fingernails emerged. The author found among the fingernails of patients with burns, due to superficial position, the nail root is apt to be injured, so that the forward growth of fingernails is affected, but the nail bed is not apt to be injured and can grow

constantly. At this time, its growth speed is faster than that of the nail root, and this is a manifestation called onychauxis. As is evident, under normal conditions, the growth speed of the nail root is faster than that of the nail bed.

6.9.3 Clinical Manifestation

Ungual dysplasia has many categories with varying clinical manifestations. When classified according to the number of fingernails, it includes fingernail absence, polyonychia, ectopic fingernails, etc.; when classified according to the size of fingernails, it includes megalonychia and unguiculus; when classified according to the involvement of nail root, it includes fingernail absence, polyonychia, ectopic fingernails, split fingernails, and black nevus in fingernails; when classified according to the involvement of nail bed, it can include various deformities of fingernail contours, such as carination, scaphoid, and shell shape. The clinically common symptoms include no fingernail in fingers, unguiculus, ectopic fingernails, polyonychia, neoplastic fingernails, etc. The deformity of fingernail excavation means that the fingernails take the shell shape with the presence of black lines in the fingernail (Fig. 6.70). This disease is often accompanied by finger bed defects and other deformities, such as microcheiria, absence of phalange, wide hands, syndactylia, or hyperdactylia. The ungual dysplasia mentioned generally refers to the abnormality of osteoarticular absence, occasionally combined with too short middle phalange of the little finger, but the hand functions were basically normal. However, the length of the distal dactylopodite will affect or decide the length or appearance of the fingernails.

6.9.3.1 No Fingernail or Absence of Fingernail

The patients may have immature (residual) fingernails at birth. Subsequently the residual fingernails will come off and then do not grow out. This is the simple no fingernail, which falls into the category of autosomal dominant inheritance, recessive inheritance, or occasional genetic abnormality. It can be manifested as partial or whole absence of fingernails, but the latter is very rare. The number of the involved finger can be one or more, and sometimes one hand or one foot is

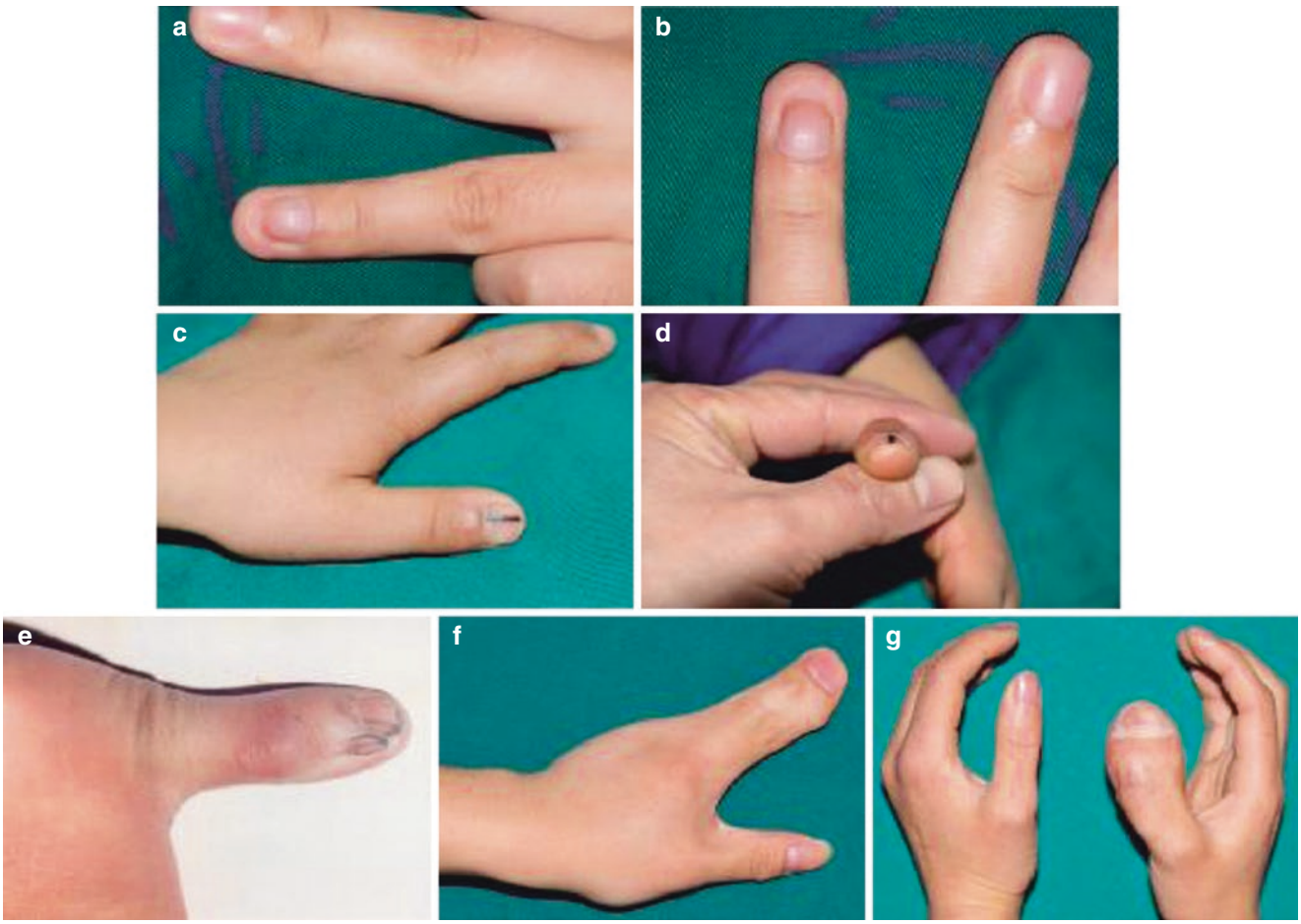


Fig. 6.70 Common types of ungual dysplasia. (**a**, **b**) Deformity of short and small fingernails (shell shape). (**c**, **d**) *Black lines* of fingers (melanocytic nevus). (**e**) Deformity of multiple fingernails. (**f**) Syndactylous wide fingernails. (**g**) Megalonychia (abnormal bones)

involved. Sometimes, the symptom of no fingernail can be accompanied by dental dysplasia, such as recessive genetic no fingernail accompanied by abnormalities in dominant genetic upper teeth or dysplasia, large schizodontia, and absence of bruxism; sometimes it can be accompanied by abnormal development of the ear and agenesis of the fibula; it can also appear in some syndromes, such as the relatively rare glossopalatine rigidity syndrome, familial dominant genetic no-finger syndrome accompanied by deformed shape at the flexor aspect, chromatosis and paratrichosis syndrome, mandibular eye-face-skull deformity syndrome, familial fifth finger absence accompanied by intelligence development retardation, rough facial features, oral and lip swelling, and hypotrichosis syndrome.

6.9.3.2 Periodic Loss of Fingernail

Periodic loss of fingernail can be manifested as the periodic loss of one or more fingernails. In spite of replacement by new fingernails, the new ones that grow out are often defective. This disease is autosomal dominant inheritance and is only limited to genetic disease of fingernails.

6.9.3.3 Fingernail Atrophy

The fingernail atrophy is manifested as thin and small fingernails. Some people call the residual fingernails in the no-fingernail syndrome "fingernail atrophy." This disease is induced by congenital ectodermal dysplasia and is an autosomal dominant inheritance.

6.9.3.4 Congenital Pachyonychia

The clinical manifestations of congenital pachyonychia are fingernail thickening, discoloration and no luster, presence of longitudinal groove and ridge, as well as nail margin bulge due to hyperkeratosis subungualis. Congenital pachyonychia syndrome is a rare epiblast defect, first reported by Jadassohn and Lewandowski in 1906, so it is also called Jadassohn-Lewandowski syndrome. This disease falls into the category of autosomal dominant inheritance.

6.9.3.5 Leuconychia

1. Simple leuconychia. It can be clinically classified into spot-shaped leuconychia, strip-shaped leuconychia, partial leuconychia, and whole leuconychia. The fingernails are cream white or porcelain white, the tips of some fingernails have a pink area as long as 2–4 mm; the strip-shaped leuconychia has longitudinal or transverse belts. The mode of inheritance of strip-shaped leuconychia and whole leuconychia is autosomal dominant inheritance.
2. Leuconychia-koilonychia. The fingernails are cream white or porcelain white accompanied by key-shaped deformity. It falls into the range of autosomal dominant inheritance.
3. Leuconychia-brittle fingernail-hypoparathyroidism-odontopathy-cataract syndrome. The fingernails are cream white or porcelain white, accompanied by brittle fingernails, hypoparathyroidism, odontopathy, and cataract. It falls into the range of autosomal recessive inheritance.
4. Leuconychia-koilonychia-deafness-knuckle pads-keratoderma palmaris et plantaris syndrome. The fingernails are cream white or porcelain white accompanied by koilonychia, deafness, knuckle pads, and keratoderma palmaris et plantaris. It falls into the range of autosomal dominant inheritance [156].
5. Leuconychia-steatocystoma multiplex-renal calcification syndrome. The fingernails are cream white or porcelain white accompanied by steatocystoma multiplex and renal calcification. It falls into the range of autosomal dominant inheritance [157].
6. Leuconychia-duodenal ulcer-bladder stone syndrome. The fingernails are cream white or porcelain white accompanied by duodenal ulcer and bladder stone.
7. Nail-patella syndrome. Also called nail-patella dysplasia, it is a rare kind of autosomal dominant disease. In 1920, Chatelain first described this disease, and in 1946, Foug reported this disease again. This disease is the functional absence induced by gene mutation and mainly involves the mesodermal and ectodermal structures during embryonic development, 98–100% have fingernail problems, 70% have iliac angles, 30–60% have kidney disease, and 15% evolve into renal failure. The main clinical features are ungual dysplasia, patellar dysplasia or absence, iliac angles, and radial capitulum dislocation tetrad, with the fingernails, iliac angles, and knee and elbow deformities as the typical characteristics [158, 159] (Fig. 6.71).

6.9.4 Treatment

6.9.4.1 Surgery Timing

Fingernail deformities or absence affects the functions and appearance of the patient's finger. As the patient grows, it will have an impact on the patient's mental status. Therefore, generally the age of 2–7 (preschool) is the best surgical timing. However, since the limbs of patient for tissue transplantation suffer hypoplasia, the repair materials are not many or insufficient, and the cost of the donor sites is too high; the advantages and disadvantages of surgery should be weighed. Perhaps the surgery should be performed when the patient becomes an adult.

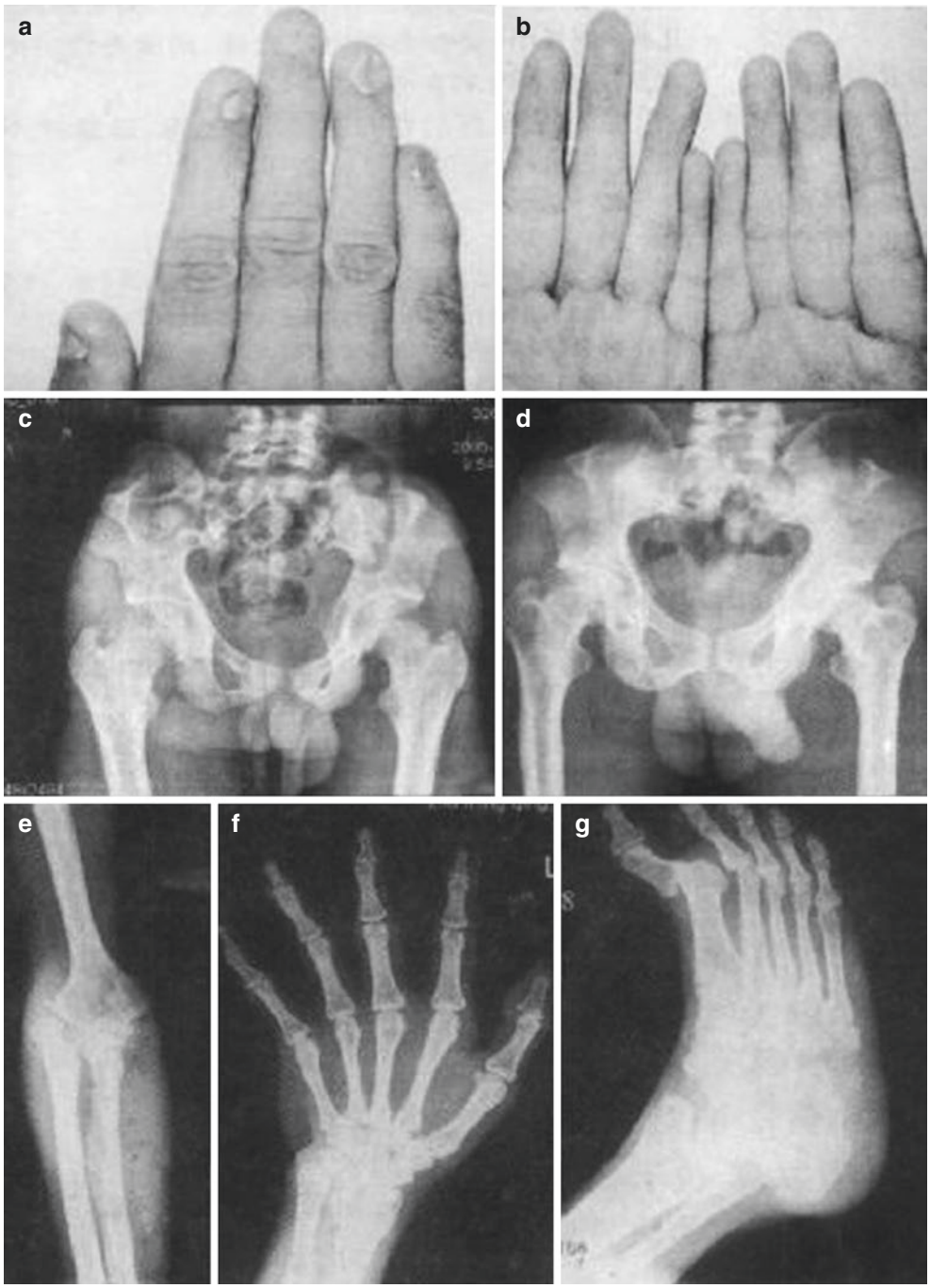


Fig. 6.71 Nail-patella syndrome. (**a**, **b**) Small and flat fingers, excavation in the center, visible longitudinal fissures, and disappearance of fingerprints of the final segments of interphalangeal joint of the third and fourth fingers. (**c**, **d**) Osseous protuberance at the posterior aspect at the rear of the bilateral iliac bones, namely, iliac angles, visible epiphysis at the iliac angles, and rhomboid sacroiliac articular surface. (**e**) The patellar bones have a small development and migrate toward the exterior and upper regions, and the femoral medial condyle becomes enlarged. (**f**) The lower end of the ulnar bone is excessively long, and the wrist joint suffers ulnar deformity. (**g**) Deformity of strephenopodia, dislocation of the first to third metatarsophalangeal joints

6.9.4.2 Surgical Method

1. No fingernail (little fingernail). As for the fingernail transplantation on the deficient fingernails, free transplantation is mainly adopted with the following four methods: ① transplantation of the toenail, nail bed, and nail matrix in the center; ② toenail, nail bed, and partial nail matrix transplantation; ③ posterior nail wall, toenail, nail bed, nail matrix, and phalange surface layer transplantation; and ④ toenail, nail bed, nail matrix, and phalange transplantation with the vessel. These surgeries need to be performed under the microscope.

   According to the report of Soeda and Shugo (1988) [160], due to the nail bed and onychostroma defects, the transplantation of fingernails and the comprehensive tissues of their accessory structures is adopted to treat

congenital fingernail defects. The toenail, nail bed, nail matrix, nail fold, nail epithelium, and nail subcutaneous tissues are completely resected, one fish mouth-shaped incision is made at the affected end, stealth separation is performed on them into the bag shape, the graft is implanted, the incision is sutured, and embedding and fixation are performed. About 1 month later, the skin on the surface of implanted fingernails becomes necrotic and detached, and the viable implanted finger is exposed; in several months, this fingernail may become detached again to grow the newly emerging fingernail.

When free transplantation is performed on the entire nail matrix, the fingernail generally will not become deformed after operation. In removal of the toenail, some part of the phalange should be removed together, including the free transplantation of the toe dorsal side of posterior nail wall of the phalange with a length of more than 6 mm. The bone cortex of the finger dorsum of the recipient bed in the recipient site is resected to expose the bone medulla, and then it is closely fixed with the lower layer of bone surface of the donor site. There are many fixation-related methods: the surgeons can open the skin at the transplantation site in a fan shape and after the toenail is inserted, the mattress suture is adopted for fixation; the fingernail, phalange, and finger pulp are passed with a nylon line and fixed with fasteners at the two sides; a skin flap is made at the digital dorsal transplantation site to lift it up, and after the fingernail is transplanted, the flap is used to restore and suture it. The performing of microvascular anastomosis under the surgical microscope can greatly improve the success rate of fingernail transplantation, which may benefit from the replantation of amputated finger. During the operation, microsurgery technology can be used to anastomose the vessels for the transplantation of the phalange, toenail, toe matrix, and skin, and the ring-shaped flaps are used to prolong the fingers, so that the functions can be recovered properly to make the appearance close to normal. This surgery can be used for the amputation and explantation of the fingertip, and it can be used for treatment of fingertip defects and fingernail defects including the thumb.

After free nail bed transplantation, pressure dressing is needed for 10–14 days; anti-infectious, anticoagulation, and anti-spasm therapies are given according to the microvascular anastomosis procedures; and drugs are given to dilate vessels and observe blood circulation [160–162].

2. Large fingernail, megalonychia, and deformed fingernails. The resection can be performed on the nail finger or nail bed stromal layer tissues, and the recombination and correction are performed on the nail beds.
3. Duplication of fingernails (fingertip bifurcation). Among the congenital fingernail abnormalities, fingertip bifurcation is the commonest, and it falls into the category of one type of supernumerary finger. The common treatment method is Bilhaut-Cloquet surgery, in which the furcal last segment of the finger is divided into two portions according to a proper size, and then the cross section of the two phalangettes is sutured. In surgery, the nail matrix and the nail wall on both sides should be at the same height, and after operation, the two pieces of fingernail become mutually healed until they form one piece of fingernail. The disadvantage is the easy formation of one longitudinal sulcus at the fingernail anastomosis line, and the fingernails do not have a normal curvature and take flat shape. In order to prevent this phenomenon, the fingernails can be made to have normal transverse bending during operation, and when the phalangeal cross section is processed, attention should be paid to make sure that it and the fingernail should have corresponding bending planes. In addition to the suturing with steel silk, one fine Kirschner silk can be inserted from the lateral aspect of the phalangettes; in order to prevent the formation of longitudinal sulcus at the fingernail, the fingernail should be pulled out, and suturing should be performed in proximity with the nail bed.
4. Melanocytic nevus of the fingernail. Melanocytic nevus should be removed at the onychostroma at the nail root.
5. Others. For the congenital wide nail, deviating nail, and hypertrophic nail, the nail bed can be reconstructed or repaired according to shape; nonsurgical methods can also be adopted, such as placement of nails and ornamentation of nails.

6.10 Kirner Deformity

Bin Wang and Feng Ni

Kirner deformity was first reported by Kirner in 1972. Its characteristics are progressive bending of digital distal phalange toward the volar side and radial side, and the commonly involved finger is the little finger; in case the finger distal segment bends to the lateral side, it is called reverse Kirner deformity. This deformation occurs on two planes, different from the unitary radial-ulnar plane deformation of the finger deviation deformity.

6.10.1 Epidemiology

The incidence of Kirner deformity is 0.15–0.25%, and the rate for females is twice that for males. The mode of inheritance of this deformity is autosomal dominant inheritance and presents incomplete penetrance, and the homozygotic state decides the expression of deformities in multiple fingers

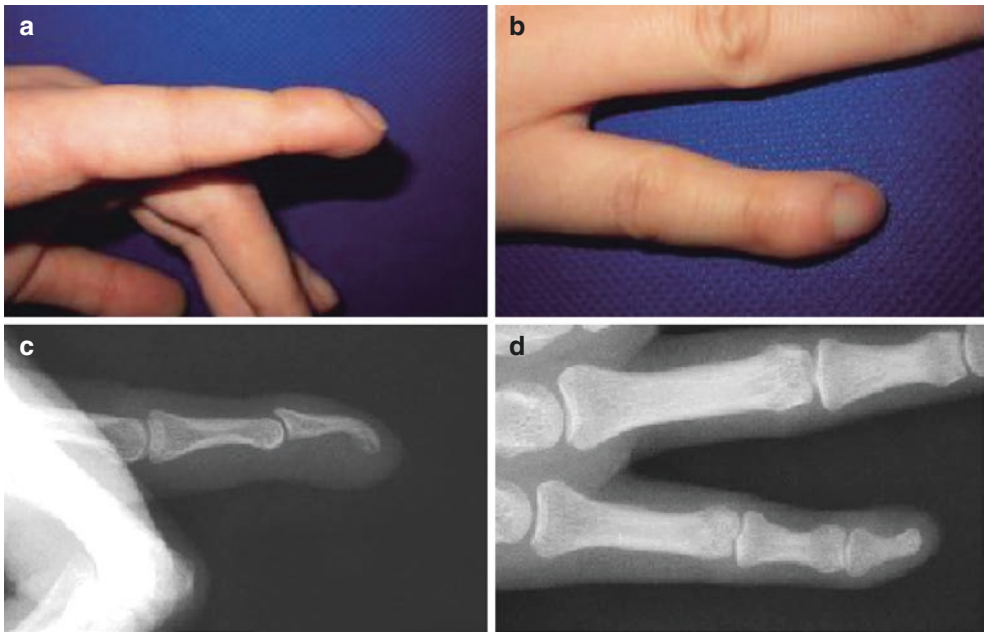


Fig. 6.72 Kirner deformity of the little finger of the right hand. (**a**, **b**) Finger appearance. (**c**, **d**) X-ray findings

and two hands. At least half of the cases are sporadic, and one of the differences between the familial inheritance and sporadic cases is the age of onset. Based on this, Song and Koh proposed a classification method, premature or congenital Kirner deformity has manifestations at birth, and the patients have related family history and the disease will not evolve with the patient's growth; delayed Kirner deformity occurs in a sporadic manner during the childhood or adolescence. The first manifestation is the swelling of the dorsum of the distal phalange or little finger phalange of the single hand or bilateral hands and slight discomfort but no pain; subsequently, the distal fingertip bends toward the volar side and radial side, the characteristic manifestation is the club-shaped or rostrum fingernails, and the distal interphalangeal joint activities are not affected [163, 164] (Fig. 6.72). Once formed, the deformity is relatively stable.

Kirner deformity is common in patients with Cornelia de Lange syndrome [165, 166], Silver syndrome, Turner syndrome, and Down syndrome. The differential diagnosis of unilateral deformity includes the delayed manifestation of the distal phalangeal tumor, especially when the epiphysis fracture is accompanied with volar angulation; the finger burns or the cold injury involves epiphysis growth plates. Diagnosis can usually be confirmed through all-round medical history inquiry in combination with physical examination and X-ray examinations.

6.10.2 Etiology and Pathology

Kirner deformity is attributable to the dysplasia of epiphyseal plate of the distal phalange, and the traction of asymmetric epiphysis growth or flexor tendon on diseased epiphyseal plates results in deformities of the distal side of the phalange. Another theory holds that the abnormal insertions at the distal end of finger profound flexor tendon lead to excessive traction force during the bone growth process [164, 167] or the normal tendons act on the osteoporotic distal phalange and hence result in deformities. In addition, there is also the hypothesis that vascular factors result in deformities.

The characteristics of Kirner are deformation and widening of the distal phalangeal epiphyseal plate accompanied by bending of the backbones. Imaging examinations can show abnormal epiphysis, and the volar margin increases and the margin of the metaphysis is rough and hardened. The epiphyseal plate closure is delayed and starts from half side of the dorsal aspect. The phalangeal histological examination during this stage indicates dissolution of the connection between the metaphysis and the epiphyseal plate. Subsequently, backbone bending is formed. During the stage of maturity, the epiphyseal plates are completely fused, and the density of the deformed phalanges returns to normal.

6.10.3 Treatment

The influence of Kirner deformity on hand functions is slight, and musicians and keyboardists can recognize the specific discomforts correlated with the little finger. The first thing that matters during the surgery is to reconstruct the appearance of fingers. The potential risk and expected results of the surgery should be made known to the patients.

According to reports, the use of splints to fix the little finger in the extension position during the early swelling of

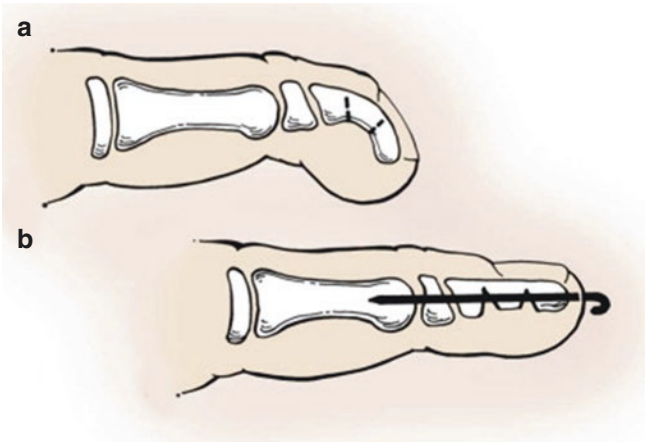


Fig. 6.73 Corrective osteotomy: wedge-shaped osteotomy on multiple planes at the volar side and Kirschner silk used for fixation of the distal phalange. (**a**) Before operation. (**b**) After operation (Reprint with permission from NILS CARSTAM, ODDVAR EIKEN, Kirner's Deformity of the Little Finger. Journal of Bone & Joint Surgery, 1970)

deformity can effectively relieve discomforts. Other researchers report that early application of delayed and continuous splints for fixation before the completion of growth helps to correct deformities. Benatar reported the release of abnormal components at the flexor tendon insertions to correct deformities [167]. Another method is to resect the dorsal wedge-shaped tissues (including dorsal growth plates) to prevent deformity progression. As for mature deformity, corrective osteotomy can be applied to improve the phalangeal alignment and the growth of corresponding fingernails (Fig. 6.73). During operation, the dorsal enclosed wedge-shaped osteotomy or the open wedge-shaped osteotomy with two to three volar incisions can be utilized to expose the phalange. Traction and lengthening can also be used to correct deformities [168].

We recommend application of splints for fixation in the early stage of deformity, but strict monitoring is needed for effective prevention and correction of deformity. As is recommended by Benatar [167] during early surgery, distal separation at the insertions of flexor tendon can have certain effects on the relief of the deformation force of developing skeletons. However, the corrective surgery must be delayed until the patient becomes able to participate in decision-making, because the surgery mainly aims to improve appearance instead of functions. The author corrects the phalangeal bending through volar osteotomy, during which the nail plates are removed, the radial-dorsal incision is made, the nail folds are retained, and the distal phalange is exposed. The osteotomy is performed from the dorsal side to the flexor tendon according to the design, and the surgery can be performed for two to three times according to the size of the phalange. Open the volar periosteum and retain the dorsal periosteum because it can play the hinge role for the corrective effects and at the same time avoid injuring onychostroma. Apply sharp blades for osteotomy. Fix and correct phalangeal bending through the axial Kirschner silk, and retain the Kirschner silk in the original position until healing is achieved at the site of osteotomy. During healing, use the splints to fix and protect the fingers.

6.11 Congenital Proximal Radioulnar Joint Fusion

Xu Gong and Laijin Lu

Congenital proximal radioulnar joint fusion is a rare kind of upper limb deformity (Fig. 6.74). Most patients with it have the manifestation of pronation and fixation of the forearm (>60° in most cases), and their daily motions such as eating, face washing, and writing are affected.

6.11.1 Clinical Manifestations and Classification

As the flexion functions of the elbow joint and functions of the wrist joint are normal, most pediatric patients can perform the rotation action of the forearm through the compensation of radiocarpal joint and midcarpal joint. If at the school age the pediatric patients cannot perform personal hygiene actions such as washing the face, they will then be found to suffer forearm rotation disorder. Therefore, congenital proximal radioulnar joint fusion is mostly diagnosed at school age. In other cases, the pediatric patients are found to suffer forearm proximal radioulnar joint fusion due to traumas. In case of examination of forearm rotation functions when the shoulder is in the joint adduction position and elbow joint is in the elbow bending position, most patients suffer pronation and fixation deformity.

The entire forearm X-ray film can reveal the presence of synostosis at the radial and ulnar proximal ends, but sometimes the forearm rotation disorder is derived from the ulnar and radial interosseous cartilaginous or fibrous connection. Usually the humeroulnar joint and distal radioulnar joint are normal.

Based on the shape and position of radial capitulum, Cleary and Omer classified proximal radioulnar joint fusion into four types: type I, normal radial capitulum and radial head joint and presence of fibrous fusion between the radioulnar bones; type II, normal appearance of radial capitulum and paraposition between it and humeral capitellum but presence of bony fusion between the radial bones; type III, presence of bony fusion between radial bones and radial capitulum dysplasia accompanied by posterior dislocation; and type IV, presence of bone fusion between the radial bones and forward dislocation of radial capitulum.

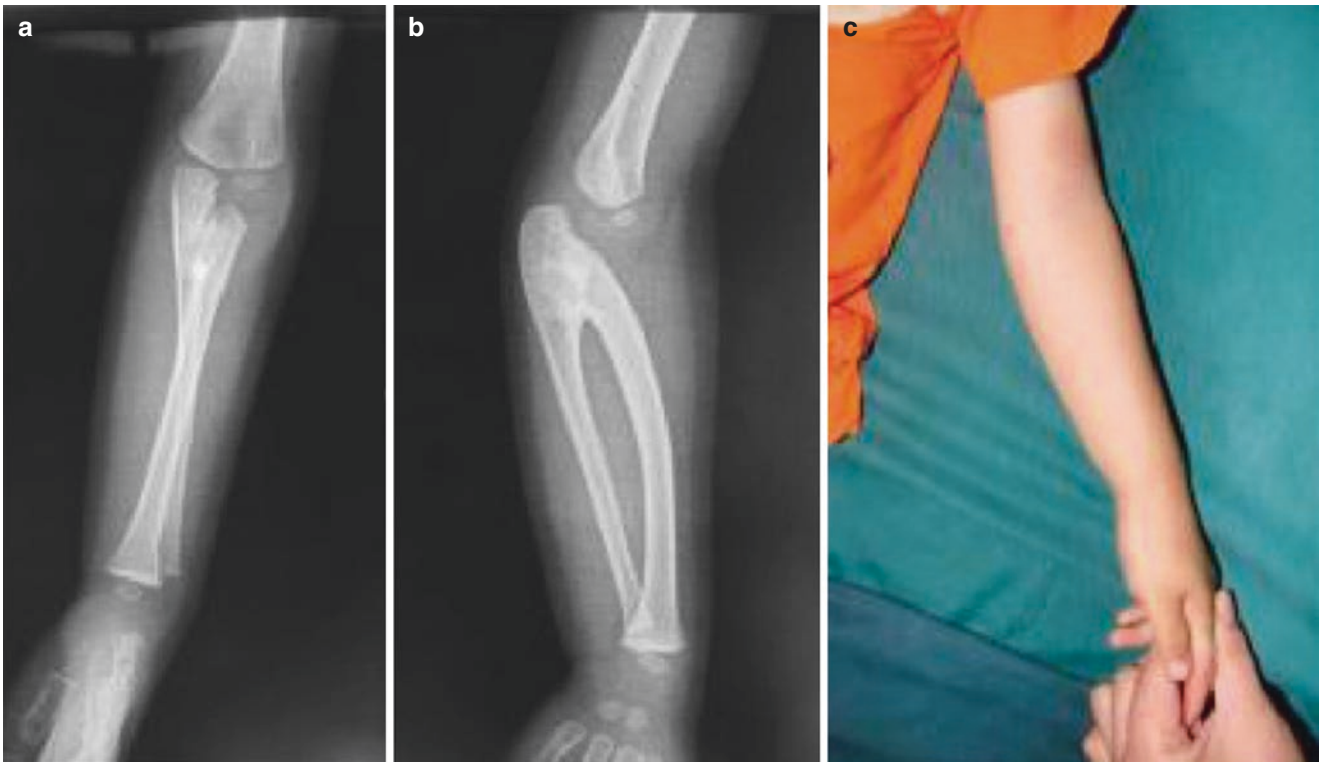


Fig. 6.74 Congenital proximal radioulnar joint fusion (Cleary III type). (**a**, **b**) X-ray findings. (**c**) Deformed appearance

6.11.2 Treatment

The pediatric patients with unilateral forearm pronation deformity of over 60° or obvious bilateral forearm pronation deformity that seriously affect daily life should receive surgical treatment. For patients with only unilateral forearm involved and insignificant pronation deformity, treatment may be unnecessary.

At present, there are mainly two surgical modes in the surgical treatment of congenital proximal radioulnar joint fusion: The first one is separation of ulnar-radial fusion site to recover the rotation functions of the forearm. The second one is the performance of osteotomy at the site of ulnar-radial fusion for derotational correction, namely, performing of osteotomy through the radial and ulnar bones and then correction of the forearm in the position suitable for the child's daily activities. For the dominant upper limb, place the forearm in the position of pronation (10°–20°); for the nondominant upper limb, place the forearm in the neutral position. During the first surgical mode, since it is necessary to place the fascia valves with blood circulation in the separated radioulnar interosseous positions to prevent the occurrence of re-fusion, it is difficult to recover the paraposition and alignment of humeroradial joint, and the surgical effects are poor in most reported cases, scholars prefer the second surgical mode.

The surgical method of performing derotational osteotomy at the ulnar-radial fusion site (Willian Green surgery): after exsanguination at the upper limb, place the tourniquets at the middle segment of the forearm. With the one horizontal finger at the proximal side of the dorsal ulnar olecranon process apophysis of the forearm, make a longitudinal incision about 7–10 cm along the radial side of the ulnar dorsal ridge, incise the skin and cutaneous tissues, expose the ulnar-radial bone, and perform detachment to expose the proximal fusion of the ulnar-radial bones. Implant a 1.5 mm Kirschner silk along the ulnar shaft nerve cavity at the ulnar olecranon process to prevent the displacement of fracture end after osteotomy. The osteotomy line is located at the distal segment at the ulnar-radial fusion and the site of ulnar-radial fusion is transversely disjuncted along the osteotomy line. After the completion of osteotomy, rotate the forearm to the position of pronation (10°–20°), and use one Kirschner silk to obliquely fix the fracture site. Suture the periosteum, perform the preventive incision on the forearm deep fascia to prevent the occurrence of osteofascial compartment syndrome (Fig. 6.75). After operation, use long arm plaster support to fix the forearm in the position of pronation of 10°–20°. Examine the incision, take out the suture lines, and continue to use the long arm plaster support to fix the upper limb in the elbow bending (90°) and pronation (10°–20°) positions for 4–6 weeks [169, 170].

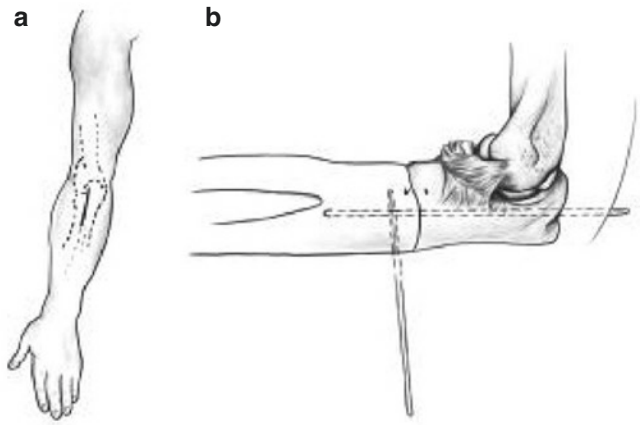


Fig. 6.75 Willian Green surgical mode. Redraw from Green WT, Mital MA. Congenital radio-ulnar synostosis: surgical treatment. J Bone Joint Surg Am. 1979 Jul;61(5):738–43 (Reprint with permission from WOLTERS KLUWER HEALTH, INC. LICENSE TERMS AND CONDITIONS)

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Twin Deformity

7

Wei Wang, Bin Wang, Hongxing Zhang, Yongkang Jiang, Yi Cao, Feng Ni, and Chuanchang Dai

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7.1 Duplication of the Thumb

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7.1.1 Overview

Manifested as twin thumb or radial and ulnar hyperdactylia of the thumb, duplication of the thumb falls into the category of deformity of hyperdactylia and is a relatively common kind of congenital hand deformity. It is classified as twin deformity or polydactylism. The twin thumb suffers dysplasia to varying extents in most cases.

Polydactylism is common among the upper limb congenital deformities, and its incidence is 5/10,000 to 19/10,000 of the neonates [1, 2]. Polydactylism can exist along and may be one of the symptoms of multiple syndromes, such as Beckwith-Wiedemann syndrome, blood syndrome, Ellis-van Creveld syndrome, Holt-Oram syndrome, and Klippel-Trenaunay-Weber syndrome [1].

Duplication of the thumb is also called radial hyperdactylia and preaxial hyperdactylia. Although to be exact it cannot be classified as polydactylism, it is common in literature. The author agrees to the viewpoints of many scholars who think it should be called duplication of the thumb. The duplication of the thumb has the following features:

1. Duplication of the thumb is mainly thumb deformity. For example, type 1 duplication of the thumb is only manifested as wide thumbnail. The structure, shape, and functions of thumb with such deformity are the same as those of normal thumbs.
2. Most patients with the duplication of the thumb have relatively well-developed thenar muscle group and are often accompanied by broad thumb web.
3. Most of the patients with duplication of the thumb have the first metacarpal bone, which is thumb-type metacarpal bone, and it means that the epiphysis is located at the proximal end of the metacarpal bone.
4. In most cases, duplication of the thumb refers to the thumb-type two-segment phalanx but with different degrees of dysplasia, and the finger-type phalangeal duplication of the thumb is mostly common in type VI and type VII.
5. Among the five fingers in duplication of the thumb, one finger is shorten than other fingers and is located in the palm-opposition position.

Therefore, it is necessary to separate duplication of the thumb from polydactylism and describe it in a separate section. Obviously, duplication of the thumb is a more exact term for radial polydactylism with thumb shape, structure, and functions. The incidences of duplication of the thumb reported vary greatly. It can be 0.08‰, 0.18‰, or 0.33‰; the incidence of polydactylia as reported is 1.4‰ [1, 3–5].

Although duplication of the thumb falls into the category of twin deformity or polydactylism, it is different from the latter in terms of shape, structure, diagnosis, and treatment method (Fig. 7.1).

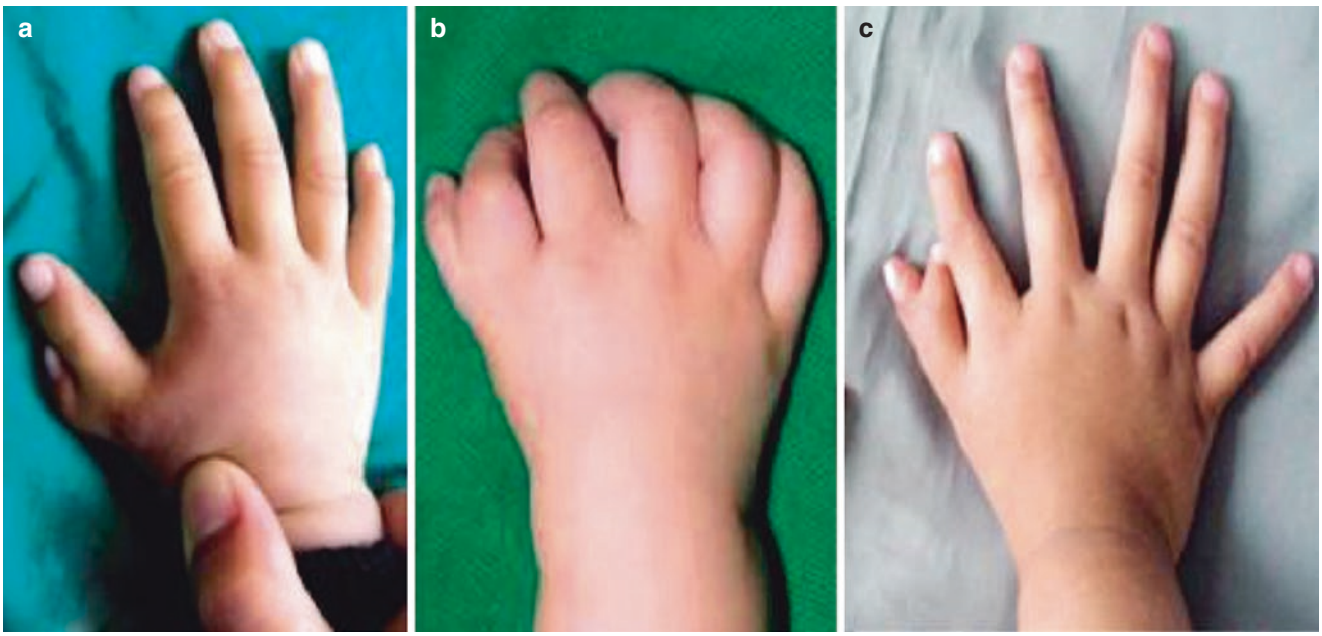


Fig. 7.1 Duplication of the thumb and polydactylism. (**a**) Six-fingered polydactylism—duplication of the thumb. (**b**) Six-fingered polydactylism—type VI thumb dysplasia. (**c**) Seven-fingered polydactylism—polydactylism

7.1.2 Clinical Manifestation

Duplication of the thumb refers to the superfluous growth of one or more thumbs or fingers on the radial or ulnar side of the thumb. It can occur to one hand or two hands. Sometimes, more than one member in the same family may suffer duplication of the thumb of varying degrees.

The clinical manifestations of duplication of the thumb are various but regular with the thumb in the palm-opposition position as well as superfluous growth of the thumb; the structure, shape, and functions of other fingers are basically normal.

Duplication of the thumb can occur to the radial side or ulnar side of the thumb or is manifested as superfluous growth at two sides of the thumb. The two or three thumbs in duplication of the thumb are not equally large, with dysplasia and deformity to varying extents. It can be biphalangeal duplication of the thumb or triphalangeal or triangular phalangeal duplication of the thumb.

The author calls the duplication of the thumb with two thumbs that are not equally large as the "major-minor duplication of the thumb," and the size, shape, structures, and functions of the larger thumb are close to normal; the size, shape, structures, and functions of the smaller thumb are greatly different from those of the normal thumb. In terms of the treatment methods, the thumb with complete functions should be retained and is called retained thumb (main thumb); the thumb with incomplete functions should be removed and is called superfluous thumb [4, 5].

Sometimes, the two thumbs suffering duplication of the thumb are twin thumbs with similar size and shape and are called mirror image thumb deformity. The two thumbs suffering duplication of the thumb are accompanied by lateriflection, volar flexion, or angulation deformity in the interphalangeal joint and metacarpophalangeal joint; their shape is like a lobster tong, so it is called lobster-tong-shaped duplication of the thumb. When the number of the thumbs in duplication of the thumb is three, it is called three-thumbed deformity.

7.1.3 Classification

There are many classification methods for duplication of the thumb. Wassel (1969) [6] classified duplication of the thumb into seven types (Fig. 7.2), but this classification can hardly summarize the complexity and changes of the several hundred cases of duplication of the thumb clinically observed by the author. In early years, the author classified the duplication of the thumb into 12 types. This method was similar to that of Wassel classification. Later, Wood (1978) [7] proposed the subtypes of duplication of the thumb on the basis of the abovementioned method (Fig. 7.3), and the classification method was made up for the deficiency of Wassel

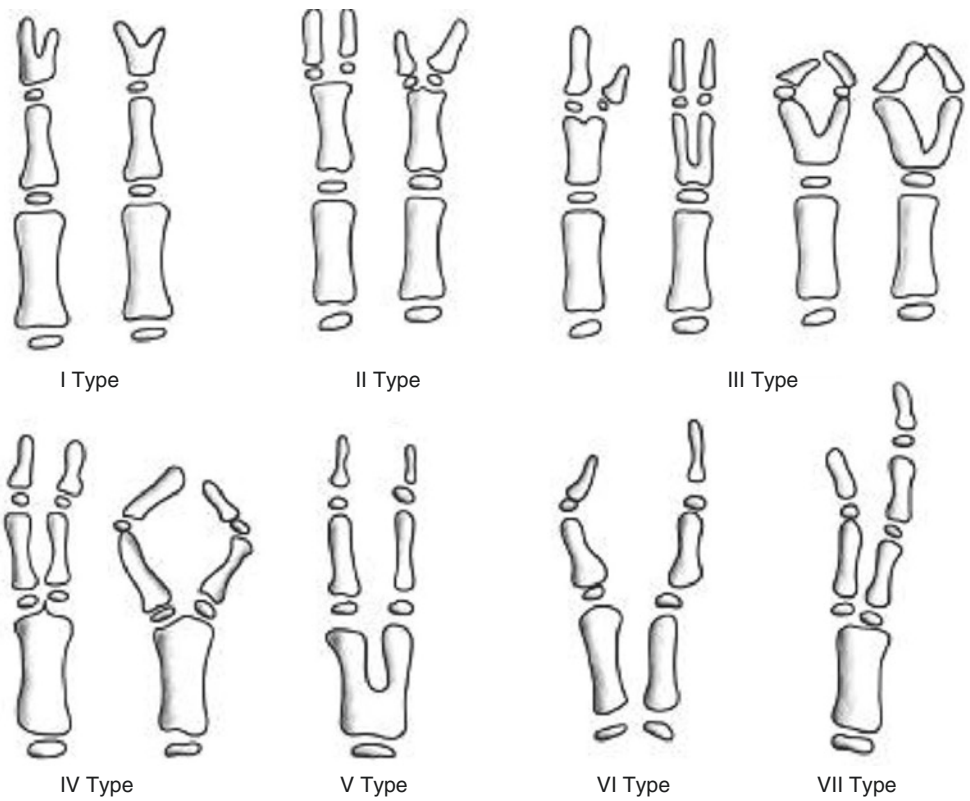


Fig. 7.2 Wassel (1969) classification for duplication of the thumb (Redrawn from Wassel HD: The results of surgery for polydactyly of the thumb: a review, Clin Orthop Res 64:175, 1969)

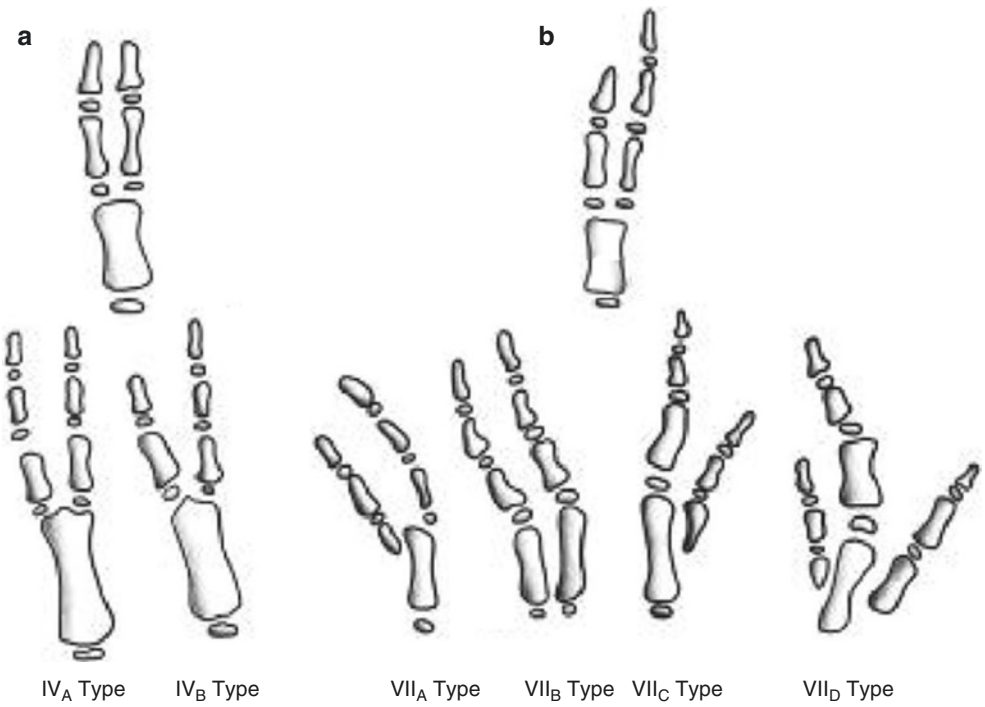


Fig. 7.3 The subtypes of duplication of the thumb supplemented by Wood (1978). (**a**) IV subtype. (**b**) VII subtype (Redrawn from Wood VE: Polydactyly and the triphalangeal thumb. 1978, J Hand Surg 3:436–444)

classification so that the Wassel-Wood classification method for duplication of the thumb was formed.

Wang Shuhuan classified it into five types: ① distal phalanx type (including complete cleft type and incomplete cleft type), ② proximal phalanx type (including complete cleft type and incomplete cleft type), ③ metacarpal type (including complete cleft type and incomplete cleft type), ④ triphalangeal type, and ⑤ floating thumb type. The deformity is classified on the basis of anatomic sites. The author believes that the Wassel (1969) classification and Wood (1978) supplementation cannot cover all types of duplication of the thumb, and they just basically cover the categories of the duplication of the thumb.

7.1.3.1 Type I Duplication of the Thumb (Phalangette Bifurcation Type)

Type I duplication of the thumb refers to the duplication of the thumb with phalangette bifurcation, the twinning of the distal end into two, and the fusion of the proximal end into one, namely, the separation of the distal end of the thumb phalangette into two and the presence of the fingertips of the phalangettes of two thumbs, or only the thumb phalangettes become widened and take flat shape, the traces of separation into two can be seen in finger pulp or fingernail, and single thumb is still found at the root of thumb phalangette and the proximal thumb. The X-ray film indicates that the distal end of the phalangettes is divided into two, the proximal ends are abreast, and the phalangettes take Y shape. The clinical manifestations are wide and flat thumb phalangettes, widened and flat fingernails, and the presence of one groove separated into two in the center and equally large thumb at the two sides of the groove (perhaps one big and one small), equally or unequally big nail arcs of the two thumbs, accompanied by the nail fold deformity of the thumb (Figs. 7.4 and 7.5).

7.1.3.2 Type II Duplication of the Thumb (Phalangette-Pairing Type)

Type II duplication of the thumb refers to the duplication of the thumb with twin and paired phalangettes, namely, separation of thumb phalangette into two and the formation of two interphalangeal joints at the thumb proximal phalanx. Type II duplication of the thumb has two fingernails, the two fingernails are obviously separated, and the two thumbs may be equally or unequally large. X-ray film indicates that the thumb phalangettes are completely separated and constitute two interphalangeal joints with one proximal phalanx. There is one protruding ridge in the center of the interphalangeal joint surface of proximal phalanx so that the distal phalanxes of the two clefts are separated. The duplicated thumb has two interphalangeal joints, there are accessory ligaments at the radial side and ulnar side of the interphalangeal joint, and there is no accessory ligament on the adjacent surface of the two separated phalanges. There may be a joint at the basal parts of two adjacent distal phalanxes. The thumb takes Y shape, and the two interphalangeal joints are located at the distal part or the lateral aspect of the proximal phalanx, one

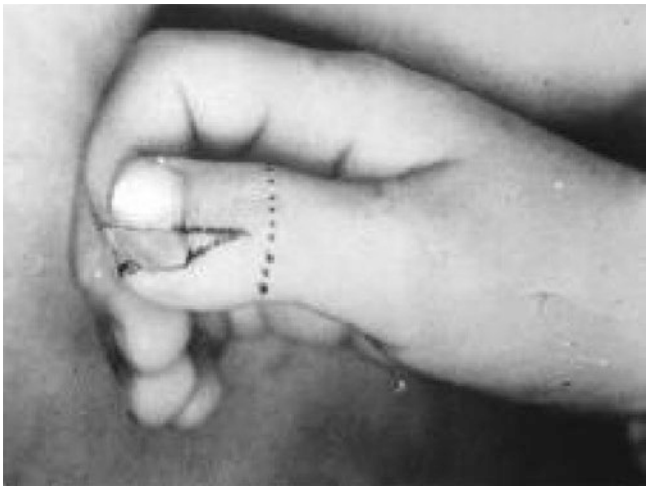


Fig. 7.4 Right hand type I duplication of the thumb; thumb phalangette bifurcation, incomplete cleft

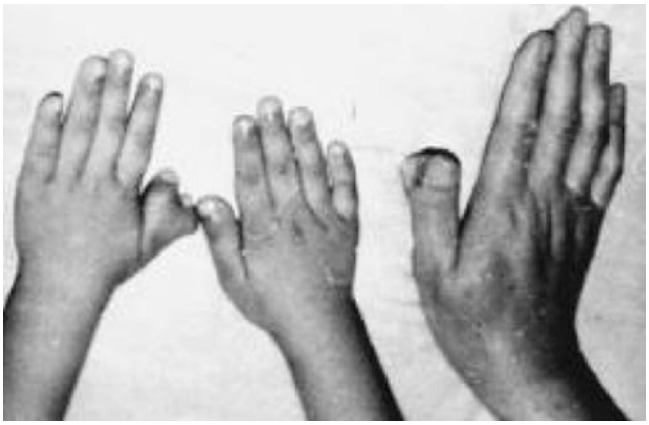


Fig. 7.5 Both the son and the mother suffer duplication of the thumb and fingernail deformity. The mother suffers type I duplication of the thumb in the right hand (*right*), and the son suffers type III duplication of the thumb in the left hand (*left*)

may be located on the central axis line of the thumb, and the other may be located at the radial side of the thumb proximal phalanx, often accompanied by the deformities in thumb extensor tendons and thumb flexor tendons. The twin two phalangettes and the distal ends of the proximal phalanx constitute the two interphalangeal joints, and there might be formation of interphalangeal joints at the lateral side of the proximal phalanx (Figs. 7.6, 7.7, 7.8, and 7.9).

In 1998, Dror Y. reported the rare phalangette duplication of the thumb accompanied by Shwachman-Diamond syndrome (SDS) with the manifestation of incomplete pancreatic external secretion functions, pancytopenia to varying extents, and metaphysis achondroplasia.

7.1.3.3 Type III Duplication of the Thumb (Proximal Bifurcation Type)

Type III duplication of the thumb is the twin and duplicated thumb of phalangette pairing and proximal phalanx bifurcation, namely, the separation of phalangette into two; the distal end of the proximal phalanx is separated, the proximal ends are connected and take a Y shape, and it works together with metacarpal bone to form a metacarpophalangeal joint. The two thumbs in type III duplication of the thumb may be equally or unequally large, and the degree of phalangeal development also varies. The growth direction of the two thumbs can be along the direction of central axis, and the possible manifestation is the abduction of the interphalangeal joint and mutual opposition of angles. The shape of the duplicated thumb is like lobster-tong shaped, and deformities in thumb extensor tendons and thumb flexor tendons can often be found (Figs. 7.10 and 7.11).

7.1.3.4 Type IV Duplication of the Thumb (Proximal Pairing Type)

Type IV duplication of the thumb refers to the duplicated thumb with paired and twin digital proximal phalanxes and phalangettes, namely, the separation of phalangettes and proximal phalanxes into two and the formation of two

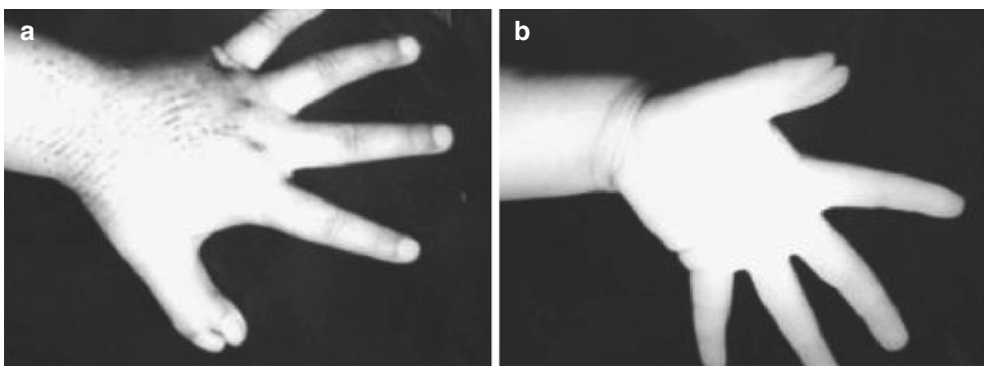


Fig. 7.6 Left hand type II duplication of the thumb: the phalangettes are twin and paired, and the thumb phalangette is separated into two

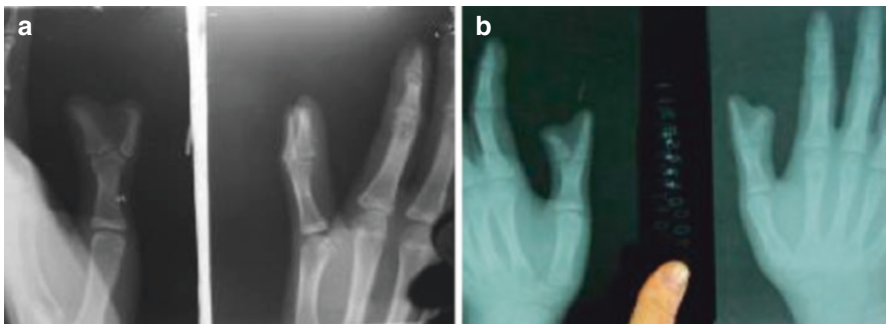


Fig. 7.7 The X-ray findings of type II duplication of the thumb of the right hand: the phalangettes are twin and paired, the distal end is separated, there is a ridge (snake-head shaped) in the center of the interphalangeal joint surface of the distal end of proximal phalanx, and the two interphalangeal joints make the thumb take Y shape

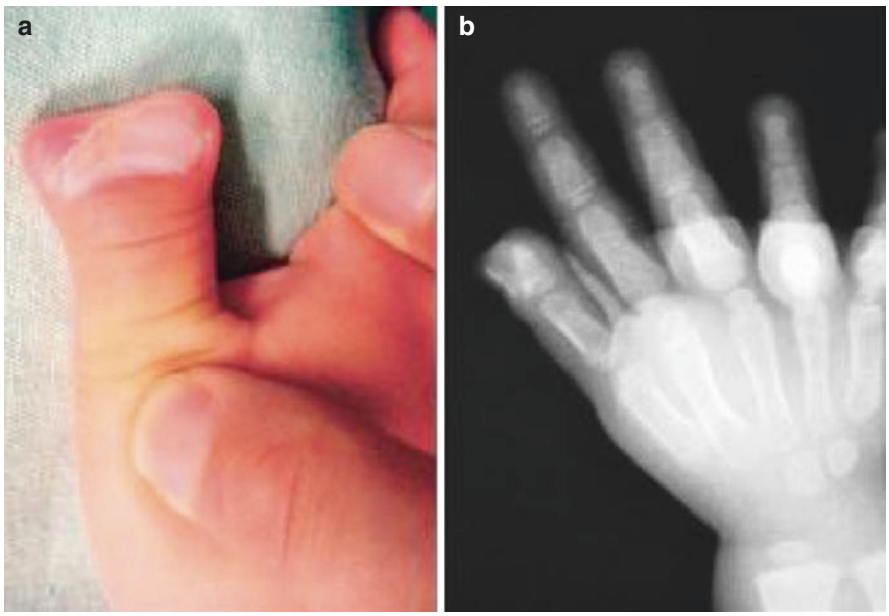


Fig. 7.8 Right hand type II duplication of the thumb: the phalangettes are twin and paired, and the thumb phalangette is separated into two. (**a**) Hand appearance. (**b**) X -ray film indicates the phalangettes are separated, the distal end is separated, there is a ridge in the center of the interphalangeal joint surface of the distal end of proximal phalanx, and the two interphalangeal joints make the thumb take Y shape

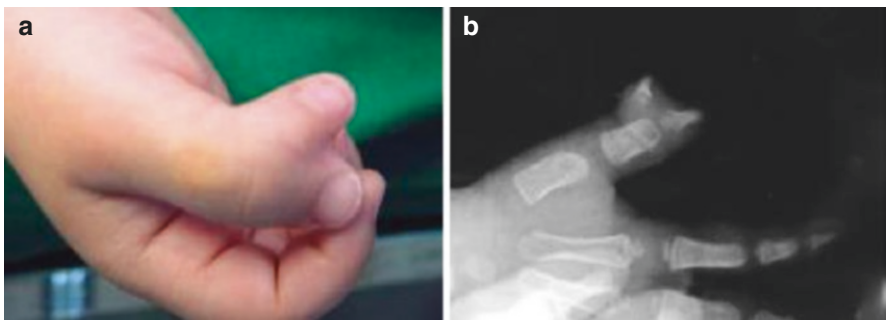


Fig. 7.9 Right hand type II duplication of the thumb: the phalangettes are twin and paired, one of the interphalangeal joints are located in the central axis line of the thumb, and the other is located at the radial side of the thumb proximal phalanx. (**a**) Hand appearance. (**b**) X-ray findings

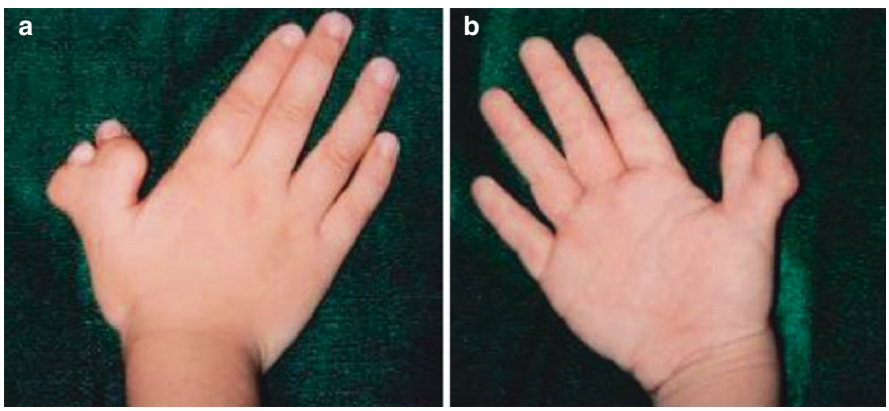


Fig. 7.10 Right hand type III duplication of the thumb: the phalangettes are in pairs, the proximal phalanxes are bifurcated and twin, the thumb is separated into two, and the interphalangeal joint suffers angulation deformity and takes lobster-tong shape

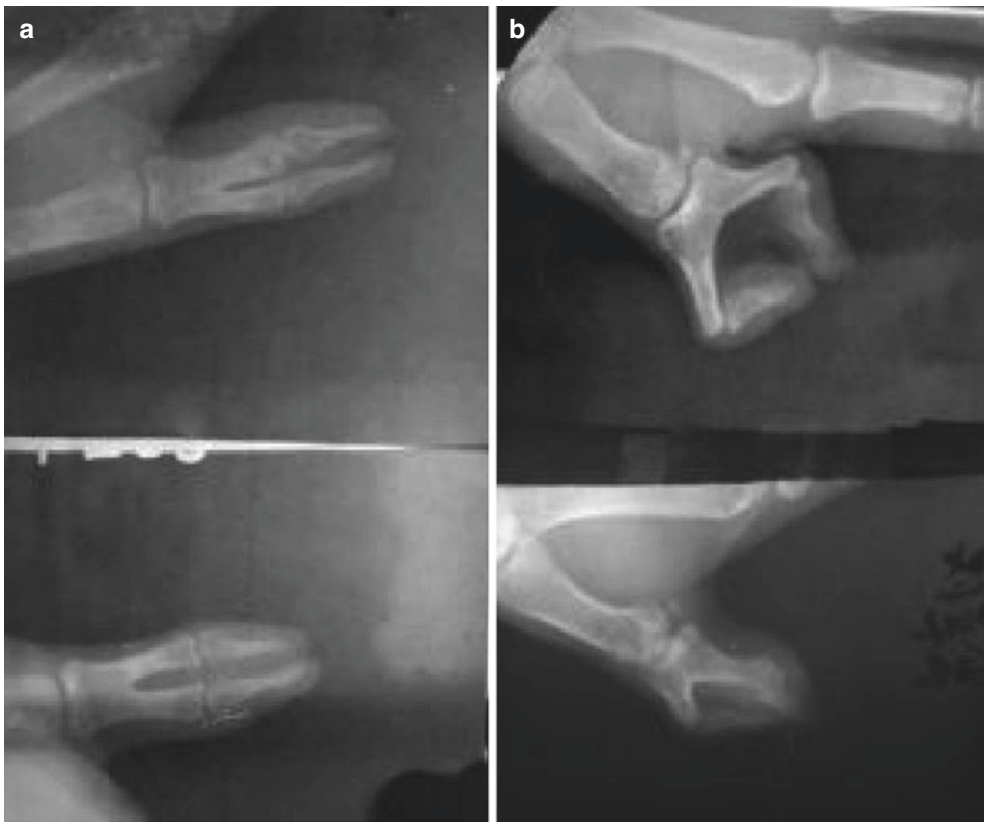


Fig. 7.11 X-ray findings of type III duplication of the thumb: the distal and proximal phalanxes are twin, the thumb is separated into two, the proximal phalanx is separated into two, the distal end of proximal phalanx is bifurcated, and a Y shape is formed with the metacarpal central axis. (**a**) Parallel growth of interphalangeal joints of the two thumbs. (**b**) The interphalangeal joints of the two thumbs suffer angulation deformity and take lobster-tong shape

metacarpophalangeal joints with one metacarpal bone. The bifurcated thumbs may be equally large and take the shape of image mirror, but in most cases they are not equally large. The accompanying symptoms are the dysplasia of the extensor and flexor tendons of the thumb; the interphalangeal joints of the two thumbs become flexed to the opposing surfaces so that the thumb is lobster-tong shaped. The thumbs present the Z-plasty opposition. The radial side and the ulnar side of the metacarpophalangeal joint have complete accessory ligaments, and accessory ligaments are absent between the two adjacent proximal phalanxes and metacarpal bones (Figs. 7.12 and 7.13).

Some authors divide the type IV duplication of the thumb into four subtypes according to the morphological and anatomical characteristics (Table 7.1).

Clinically, type I to type IV are the commonest duplication of the thumb. Among the 391 cases of duplication of the thumb reported by Lu Laijin [8], 46 patients had the phalangette bifurcation type (type I), 49 patients had the phalangette pairing type (type II), 38 patients had the proximal bifurcation type (type III), and 202 patients had the proximal pairing (type IV). The incidence of type I to IV duplication of the thumb occupies 85.7% (335/391) of the total cases of duplication of the thumb, and type IV is the commonest.

7.1.3.5 Type V Duplication of the Thumb (Phalange and Metacarpal Cleft Type)

Type V duplication of the thumb refers to the division of phalangette and proximal phalanx of thumb into two as well as the pairing and twining of the deformity; the first metacarpal bone has incomplete division, the distal end of the metacarpal bone is separated into two, and the proximal ends are combined. Type V duplicated thumb has two metacarpophalangeal joints and two interphalangeal joints. The proximal metacarpal bone constitutes a first metacarpophalangeal joint, the two thumbs are dysplastic with varying sizes, or the two thumbs grow in parallel or take lobster-tong shape (Figs. 7.14 and 7.15). In the type V duplication of the thumb, sometimes, although the first metacarpal bone is not separated into two, the metacarpophalangeal joint formed by the neoplastic thumb proximal phalanx and the first metacarpal bone is located at the lateral side of the first metacarpal bone (Fig. 7.16); sometimes, the midline of the first metacarpal bone is vertical.

The feature of this type of duplication of the thumb is that the duplicated thumb has two metacarpophalangeal joints, which can grow either on the same plane or on different planes. If they grow on different planes, the thumb phalangette and proximal phalanxes are separated into

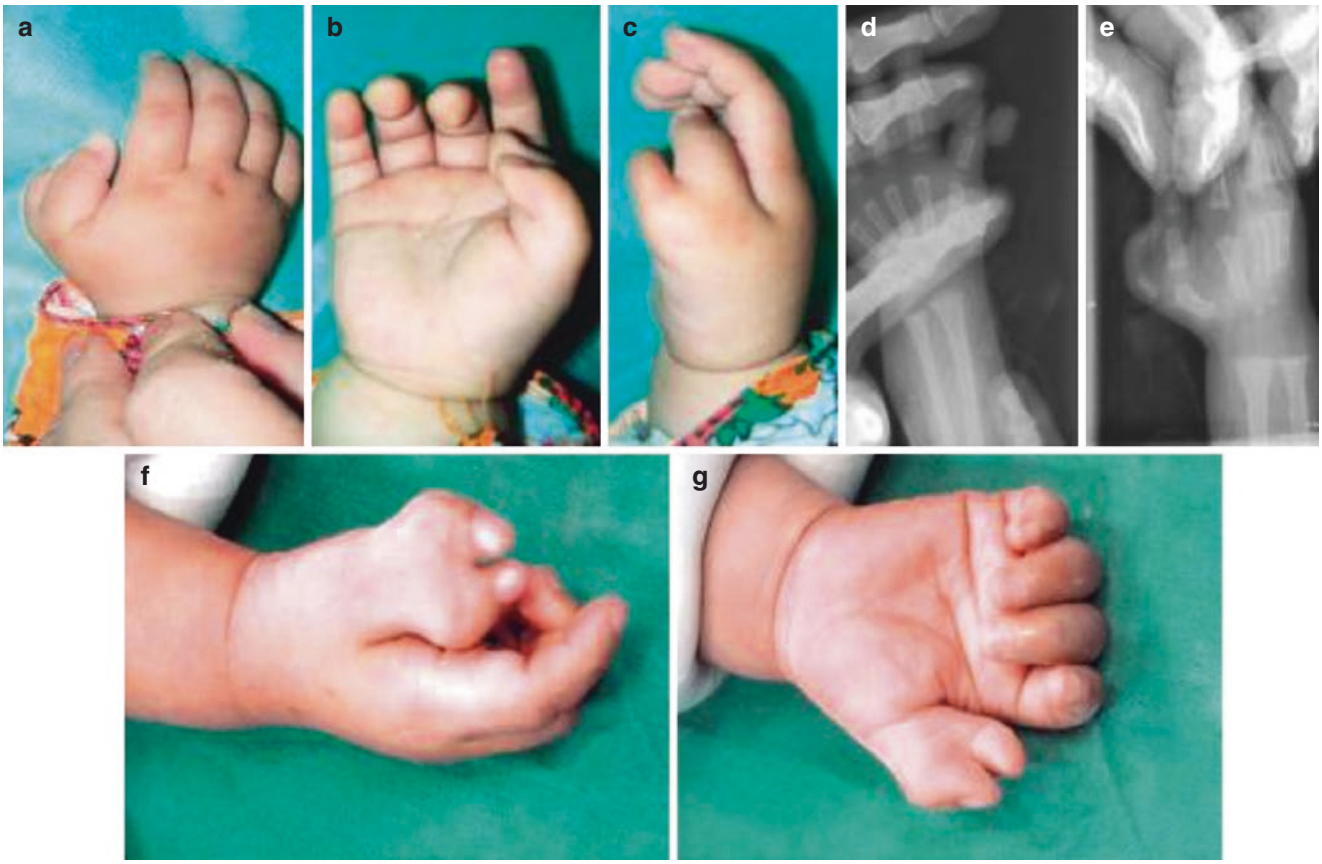


Fig. 7.12 Right hand type IV duplication of the thumb; the proximal and distal phalanxes are paired and twin. (**a**–**e**) Parallel growth of interphalangeal joints of the duplicated thumb. (**f**, **g**) The interphalangeal joint of the duplicated thumb suffers deformity of lateral curvature and takes lobster-tong shape (type VI_D duplication of the thumb)

two. Though the first metacarpal bone does not present typical Y-shaped cleft, there are metacarpophalangeal joints formed by the thumb proximal phalanx and the first metacarpal bone: one is at the distal end of the radial side of the first metacarpal bone; the other is in the middle of the radial side of the shaft of the first metacarpal bone. When the metacarpophalangeal joint of this type of thumb deformity appears at the distal end of the radial side of the first metacarpal bone, it is called the floating thumb, and the name is easily confused with the floating thumb in the type IV thumb dysplasia. For the convenience of recognition, the author suggests not to use the name of floating thumb.

7.1.3.6 Type VI Duplication of the Thumb (Phalange and Metacarpal Pairing Type)

Type VI duplication of the thumb refers to duplication of the thumb where the thumb phalange and metacarpal bone exist in pairs and have clefts, that is, the two thumbs exist independently with four phalanxes and two metacarpal bones, and the skin, subcutaneous tissues, tendons, ligaments, and joints of the two thumbs suffer dysplasia. The shape and histological structure of the two thumbs are various, and the size may be the same or one is larger and one is smaller; they may exist in parallel or are in lateriflection and opposition position (Figs. 7.17 and 7.18).

Although type VI duplication of the thumb and type VI thumb dysplasia fall into the category of six-fingered deformity in appearance, there are differences between the two: the former has the thumb in the palm-opposition position with the presence of the thenar muscles, but the latter has no thumb in the palm-opposition position and no thumb web, which is actually congenital thumb absence. In addition, type VI thumb dysplasia is the hyperphalangeal thumb absence, which is different from type VI duplication of the thumb (Fig. 7.19).

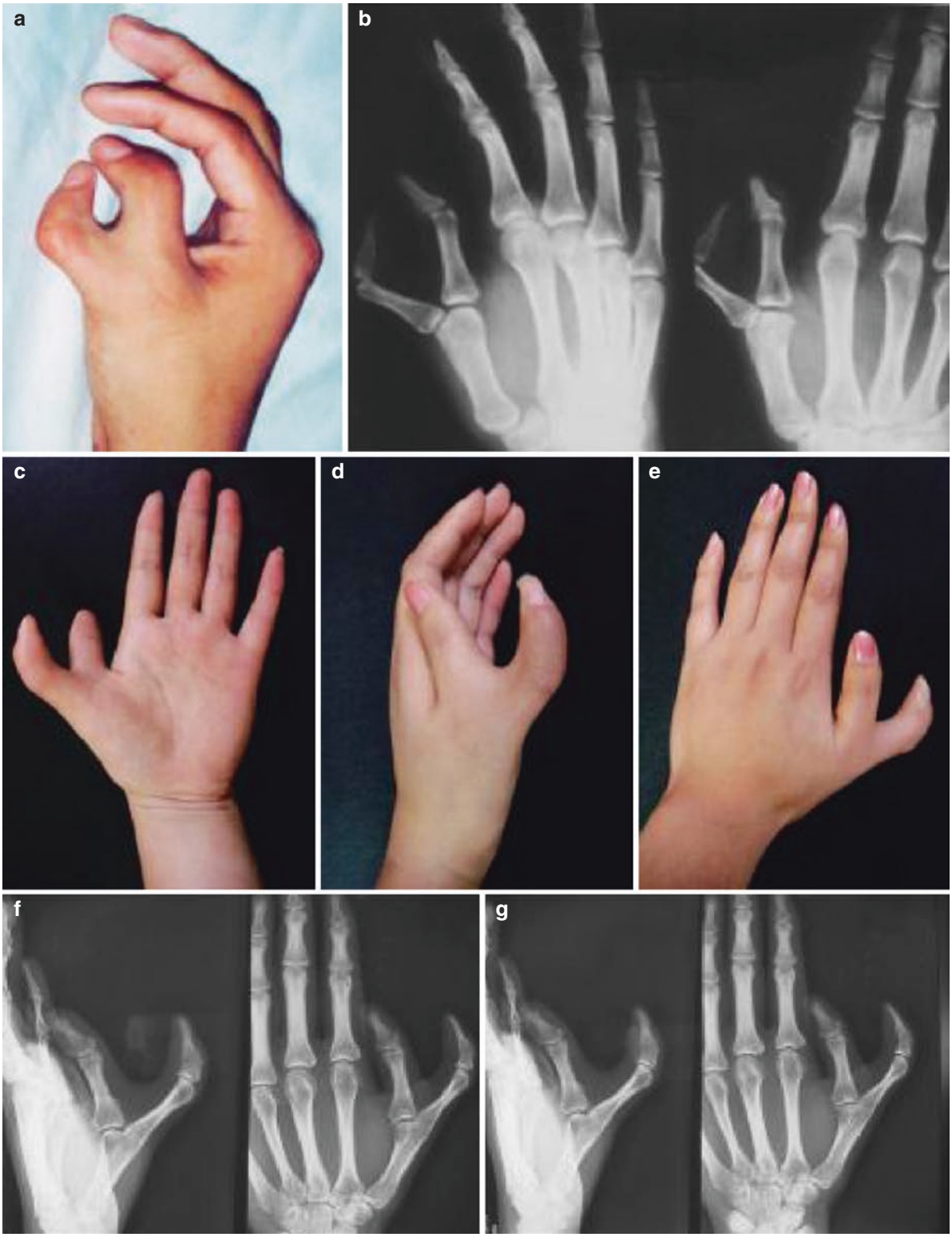


Fig. 7.13 Right hand type IV duplication of the thumb; the proximal and distal phalanxes are paired and twin. (**a**–**e**) The interphalangeal joint of the duplicated thumb suffers deformity of lateral curvature and takes lobster-tong shape, and the proximal and distal phalanxes form two interphalangeal joints with the first metacarpal bone, respectively. (**f**, **g**) X-ray film indicates the fusion between the proximal phalanx of the radial duplicated thumb and the first metacarpal bone

7.1.3.7 Type VII Duplication of the Thumb (Thumb Irregular Superfluous Growth Type)

Type VII duplication of the thumb refers to the duplication of the thumb not covered in type I to IV. The phalanges and metacarpal bones of this type of duplicated thumb feature irregular cleft and irregular superfluous growth, with the following manifestations:

Table 7.1 Subtypes of type IV duplication of the thumb

Classification	Description	Incidence (%)
Type IV_A	Hypoplastic	12
Type IV_B	Ulnar deviated	64
Type IV_C	Divergent	15
Type IV_D	Convergent	9

1. The duplicated thumb is triphalangeal or triphalangeal-like fingers appear at the site of thumb.
2. The duplicated thumb has two or more metacarpal bones that exist independently.
3. There are 2–3 thumbs in the palm-opposition position, and the number of hypoplastic carpometacarpal joint may exceed 1.
4. There are 6–7 fingers in the entire hand; although the finger suffers dysplasia, it is in the palm-opposition position, with the presence of broad first fingerweb (thumb web) and thenar muscles.
5. There are extensor and flexor of the thumb, the position is normal, and the patient can engage in functional activities (Figs. 7.20, 7.21, and 7.22).

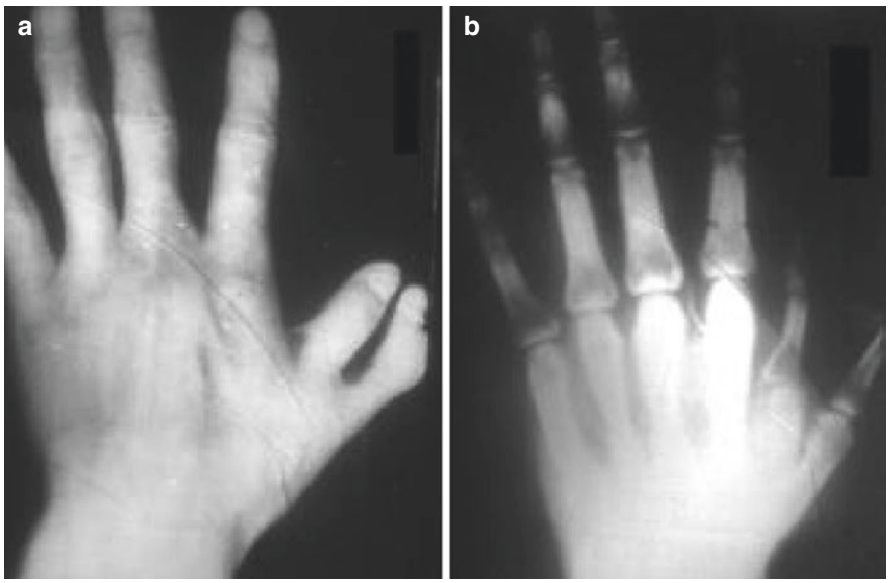


Fig. 7.14 Left hand type V duplication of the thumb: in adult male, the thumb phalangettes and the proximal phalanx are separated into two, the distal end of the first metacarpal bone is separated into two, and the proximal ends are combined. (**a**) Hand appearance. (**b**) X-ray findings

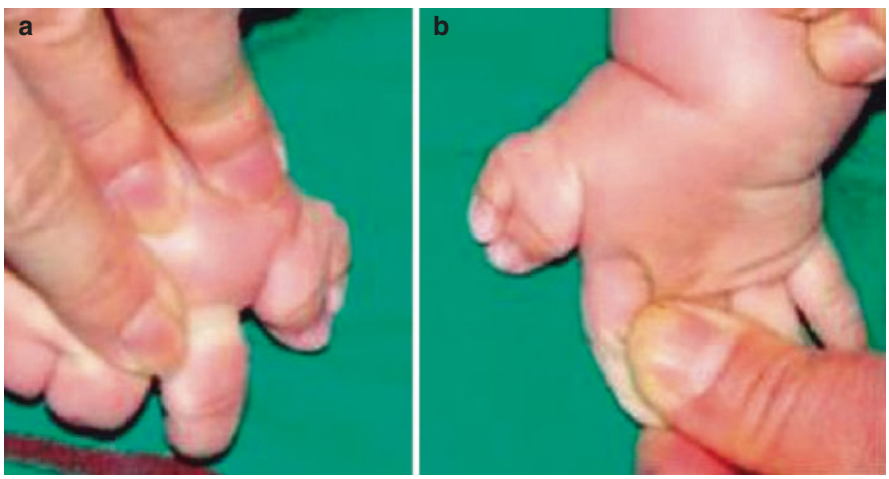


Fig. 7.15 Type V duplication of the thumb of bilateral hands: a 3-month-old male infant; both thumb phalangette and proximal phalanx are separated into two, the distal end of the first metacarpal bone is separated into two, and the proximal ends are combined

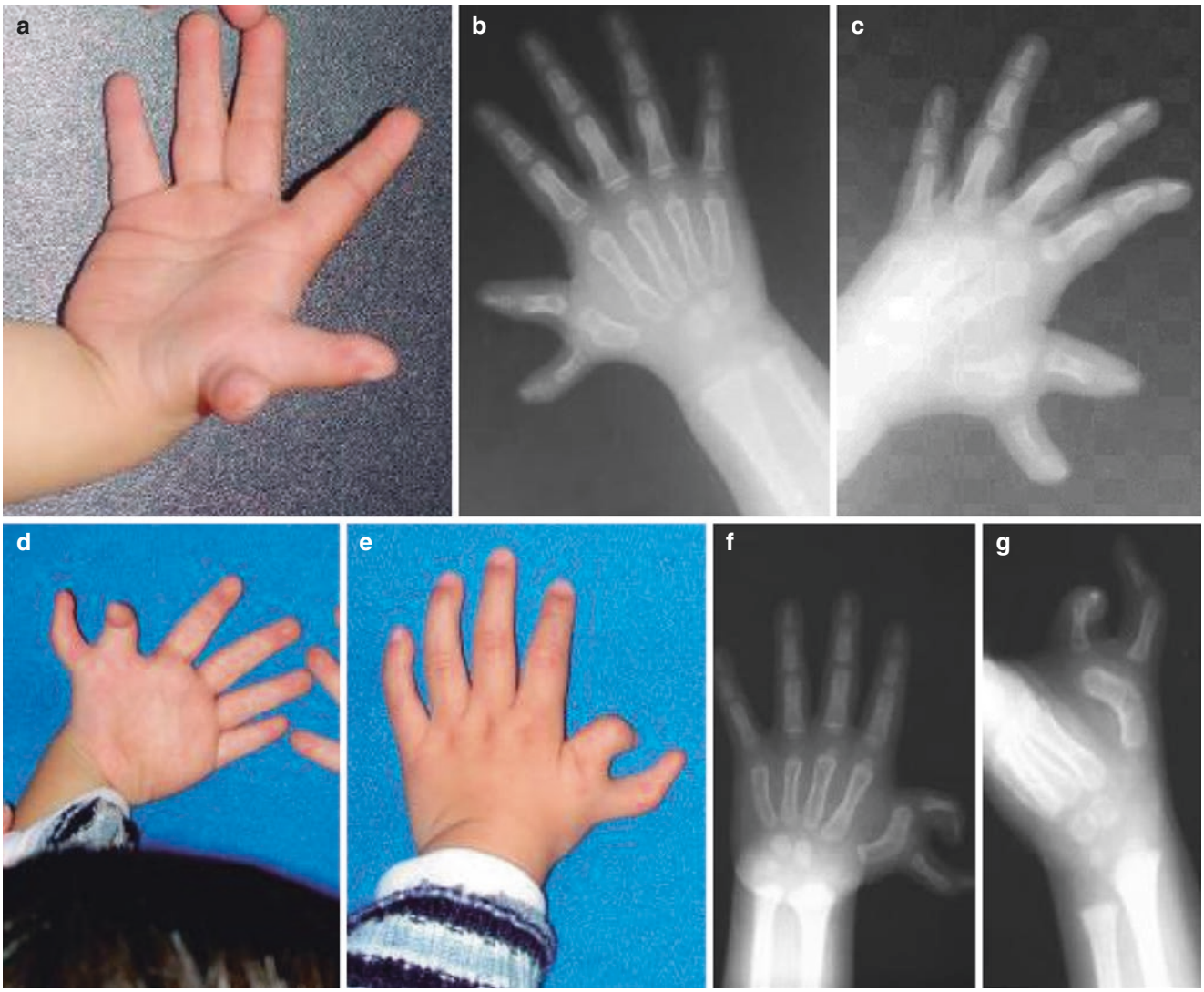


Fig. 7.16 Type V duplication of the thumb: the thumb phalangette and proximal phalanx are separated into two; although the first metacarpal bone is not separated, the metacarpophalangeal joint formed by the neoplastic thumb proximal phalanx and the first metacarpal bone is located at the lateral side of the first metacarpal bone

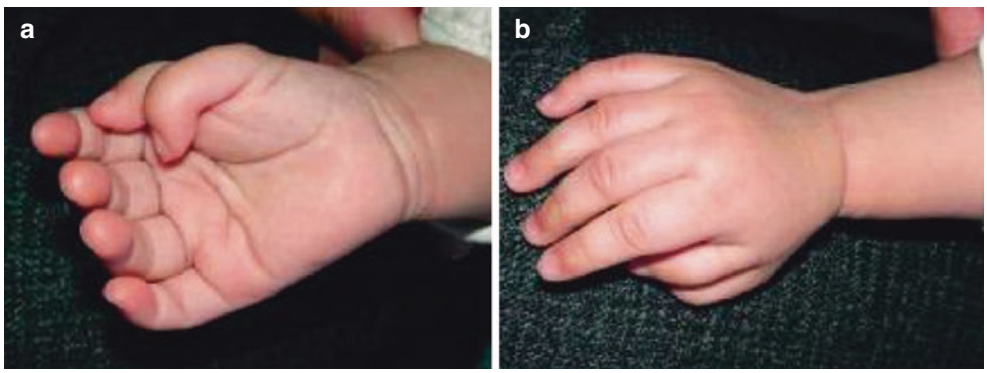


Fig. 7.17 Right hand type VI duplication of the thumb: both the phalanges and metacarpal bones have cleft, the two thumbs suffer dysplasia, but the two thumbs are in the palm-opposition position, the thumb web develops well, and there are thenar muscles

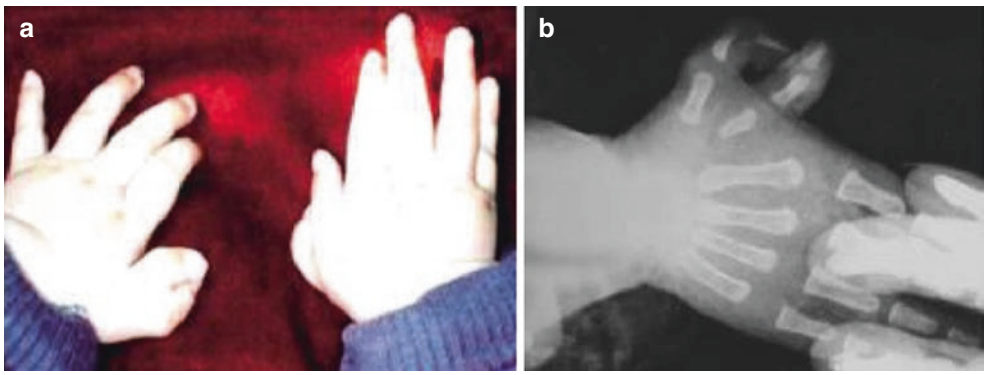


Fig. 7.18 Duplication of the thumb in both hands: the left hand suffers type V duplication of the thumb, and the right hand suffers type VI duplication of the thumb. (**a**) Hand appearance. (**b**) X-ray film indicates that the proximal phalanx, distal phalanx, and metacarpal bone of the two thumbs all exist in pairs accompanied by dysplasia

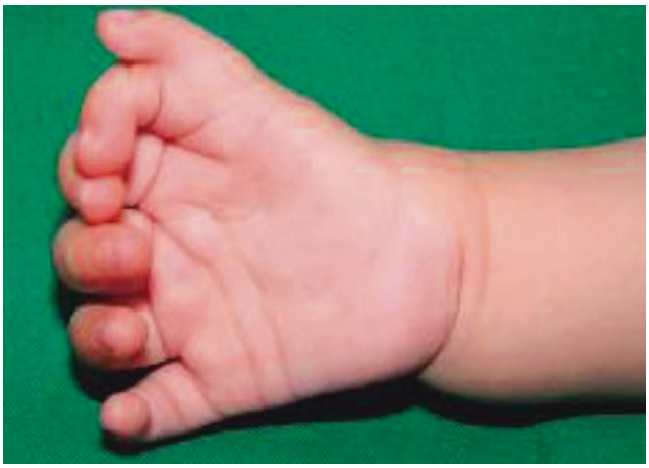


Fig. 7.19 Type VI thumb dysplasia: although it is six-fingered deformity, it has no thumb in the palm-opposition position and no thumb web, which is different from type VI duplication of the thumb

7.1.4 Pathological Manifestations and Functional Assessment

The pathological manifestations and functional assessment of duplication of the thumb are the basis for selection of therapeutic regimen and also the basis for assessment of surgical therapeutic effects. Due to the deformity changes in tissue components of neoplastic thumb, the pathological manifestations of duplication of the thumb are various.

7.1.4.1 Morphological Abnormality

Morphological defects, deformities, or variation are important factors in hand function defects. Duplication of the thumb is a serious thumb dysplasia, which can affect thumb functions and hand functions. Serious hand morphological defects can affect expression of hand functions and can also affect the patient's psychological developmental and social interactions. Although most patients with duplication of the thumb can perform the daily actions of the hand, due to thumb deformities, the patients find it can be a problem in social interactions. For example, a 46-year-old famous male engineer suffered duplication of the thumb in the right hand, and his two thumbs were lobster-tong like. Since the affected hand functions, he did not offer to receive treatment until the late 1970s when he found surgery was necessary due to the need to interact with the Westerner peers.

Hand appearance abnormality is a kind of obvious damage. As current hand functional assessment is limited to assessment of sensation and motor functions and there is no morphological assessment, many domestic scholars, when describing and reporting duplication of deformity, often consider that duplication of the thumb (especially type I to IV duplication of the thumb) does not involve obvious functional disorder; the flexion, extension, adduction, abduction, rotation, and palm-opposition functions of the duplicated thumb are basically normal; the finger-to-finger pinch, finger lateral pinch, and grip strength are also present; or compensation functions are nearly normal and finger sensations are good. But assessment on hand shape is neglected. In fact, morphological defect is a kind of hand function disorder.

The author proposes to assess morphological defect during assessment of the functional defects of duplication of the thumb. The preoperative and postoperative appearance defects of the duplicated thumb are divided into three categories (Table 7.2): category one (mild deformity, mild abnormality in thumb shape and size, and joint angulation < 15°), category two (moderate deformity, obvious abnormality in thumb shape and size, obviously visible deformity, and joint angulation >16°), category three (severe deformity, serious abnormality in thumb shape and size, ugly shape, or joint angulation >30°).

The assessment of the influences of these three categories of appearance defects on hand functions: category one

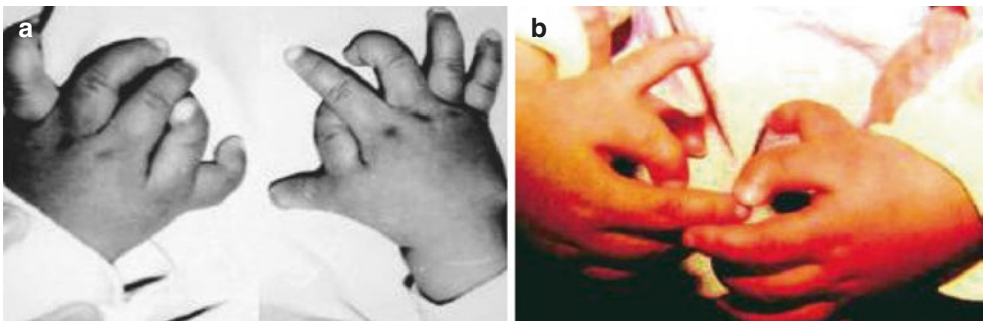


Fig. 7.20 Type VII duplication of the thumb: the phalanges and metacarpal bones of the duplicated thumb suffer irregular superfluous growth, the radial thumb is the abduction type, and the ulnar thumb is the adduction type; the duplicated thumb has broad thumb web, with good palm-opposition functions and presence of thenar muscles; the ulnar interphalangeal joints of the duplicated thumb suffers angulation deformity, there are digital flexor and extensor tendons, and the patient can engage in functional activities

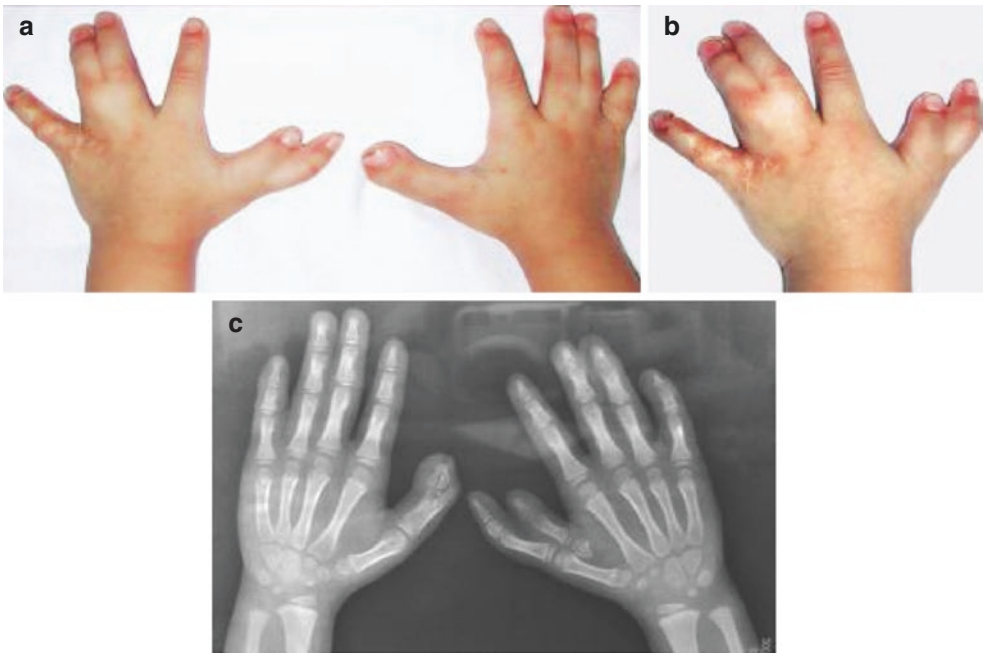


Fig. 7.21 The duplication of the thumb in both hands is accompanied by syndactylia: the left hand suffers type VII duplication of the thumb, the thumb suffers triphalangeal deformity, but the palm-opposition is good, with the presence of thumb web and thenar muscles and the radial interphalangeal joint of the duplicated thumb suffers angulation deformity, there are extensor and flexor tendons, but the position is abnormal. (**a**, **b**) Hand appearance. (**c**) X-ray findings

appearance defects result in 10% of hand functional defects, category two appearance defects result in 20% of hand functional defects, and category three appearance defects result in 30% of hand functional defects.

Take a pediatric patient, for example. The extension, flexion, adduction, and abduction functions of its thumb are normal, but its thumb proximal and distal phalanxes suffer serious deformity, namely, category three appearance defects. Because thumb functions occupy 40% of hand functions (refer to 2–45) and category three appearance defects result in 30% of thumb functional loss, the entire hand functions of the pediatric patient is rated to be 30% × 40% = 12%.

7.1.4.2 Structural Abnormality

1. Phalangeal and interphalangeal joint structural abnormalities:
   (a) Phalangeal abnormalities: in case of duplication of the thumb, the phalanges suffer abnormalities in quantity, shape, and structure. It may be biphalangeal thumb or triphalangeal thumb. In the triphalangeal thumb, the phalanges may have a normal shape, and the middle phalanx may also take triangular shape; in the normotrophic phalanges, there are small, short, flat, or thickened phalanges. The phalanges with residual thumb and neoplastic thumb can

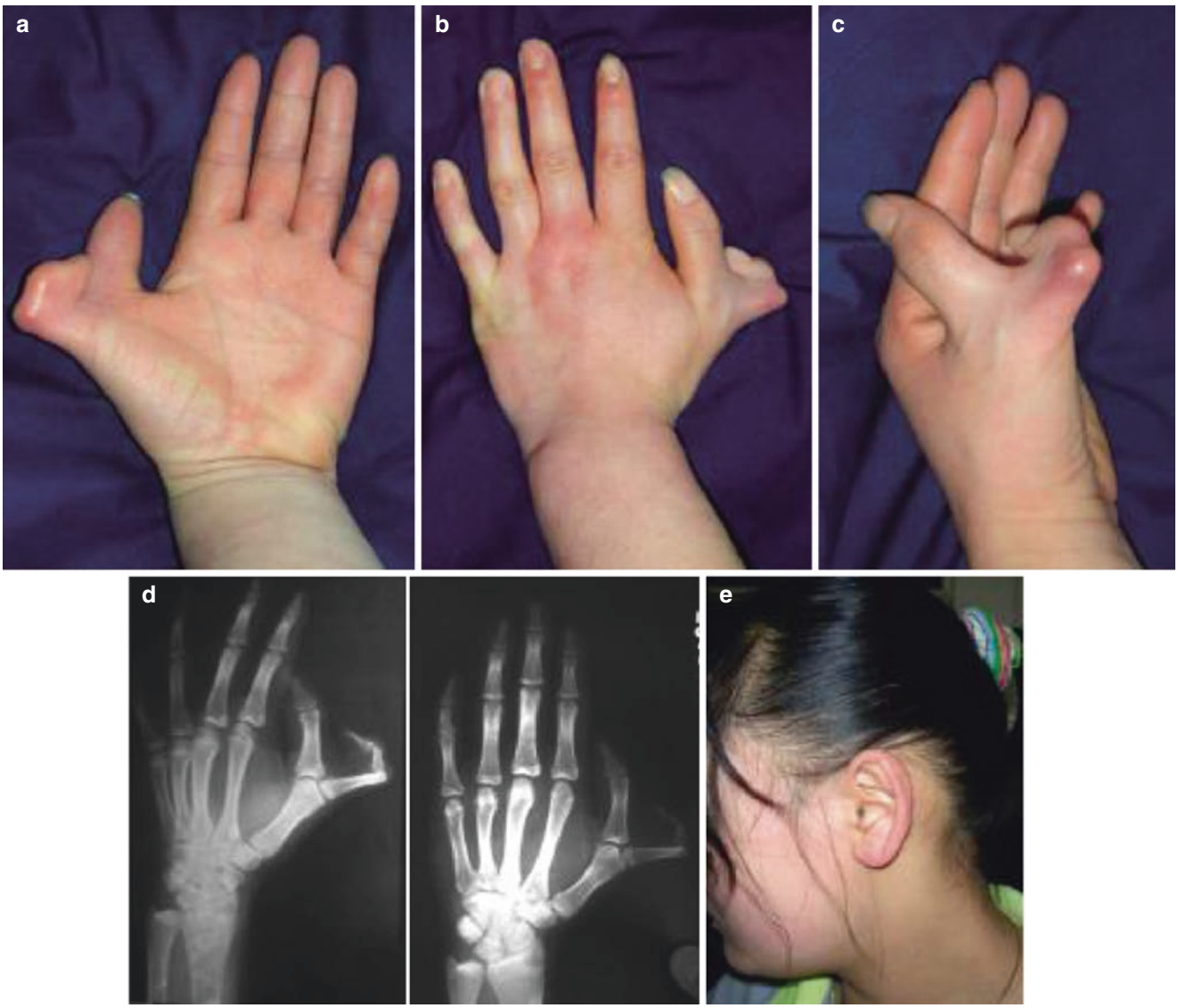


Fig. 7.22 The left hand type VII duplicated thumb is accompanied by bilateral external auricular deformities: the patient is an adult female, the radial thumb is triphalangeal, the ulnar thumb has two segments of phalange, and the head of metacarpal bone becomes irregularly enlarged; the duplicated thumb has broad thumb web, the palm-opposition is good, and there are thenar muscles; the ulnar and radial interphalangeal joint of the duplicated thumb suffers angulation deformity, there are digital extensor and flexor tendons, and the position is abnormal. (**a**–**c**) Hand appearance. (**d**) X-ray findings. (**e**) Auricle appearance

exist independently and can also have fusion to varying degrees.

(b) Structural abnormality in interphalangeal joint: the articular surfaces of interphalangeal joint may be normal hinge type and can also suffer deviation, displacement, angulation, and dislocation to varying degrees. The soundness and tension of accessory ligaments on bilateral sides of the joints are inconsistent, often manifested as relaxation of the radial accessory ligaments of the radial thumb or the ulnar accessory ligaments of the ulnar thumb. As for the duplication of the thumb, the interphalangeal joint of the radial thumb can become flexed to the ulnar side to varying extents, the interphalangeal joint of the ulnar thumb can present lateriflection toward the radial side to varying extents, the interphalangeal joint plane does not take horizontal position but inclines toward one side, and the shape of the two thumbs is lobster-tong like.

2. Structural abnormalities in the phalange, metacarpal bone, and metacarpophalangeal joint. The phalanges of the duplicated thumb may have abnormal changes in quantity, shape, and structures:

(a) The metacarpal bone and phalange: the number of metacarpal bones of the duplicated thumb is one in at least half of the cases, and it can be seen in type I to

Table 7.2 Assessment of appearance defects of the duplication of the thumb

Category	Deformity degree	Assessment criteria	Type I (thumb, finger nail)		Type II, III, and IV		Type V, VI		Type VII	
			Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative
Category one	Mild deformity	Shape and size are close to normal and joint angulation <15°								
Category two	Moderate deformity	Shape and size are obviously abnormal and joint angulation >16°								
Category three	Severe deformity	Shape and size are severely abnormal or joint angulation >30°								

IV duplication of the thumb; it can be paired and twined and common in type V to VII duplication of the thumb. The structure of metacarpal bone can be thumb type or phalange type, e.g., the ossification center of the metacarpal bone is located at the distal end of the metacarpal bone, or it is shortened metacarpal bone. If there are two metacarpal bones, fusion to varying extents may be present, such as type V duplication of the thumb.

(b) The metacarpophalangeal joint: there can be normal metacarpophalangeal joint, or two metacarpophalangeal joints can be formed at the distal end of one metacarpophalangeal joint. Under the latter situation, the articular surfaces of the metacarpophalangeal joints are located on the radial surface or ulnar surface at the distal end of the metacarpal bone, and, respectively, incline to the radial side or ulnar side by 15°–60°. There is a ridge in the center of the metacarpophalangeal joint at the distal end of the metacarpal bone, making the two thumbs of the duplicated thumb incline to the radial side and the ulnar side, respectively. The metacarpophalangeal joint accessory ligaments of type I to III duplication of the thumb develop well, and type IV or higher level of duplication of the thumb often suffers dysplasia or relaxation in the radial accessory ligaments (including the sarciniform part and scallop part of accessory ligaments). Type IV duplication of the thumb often suffers absence of ulnar accessory ligaments in the radial thumb metacarpophalangeal joint and absence of radial accessory ligaments of ulnar thumb metacarpophalangeal joint, which should be reconstructed in corrective surgery.

3. Structural abnormalities in muscles and tendons:

(a) Hand internal muscle dysplasia: the duplication of the thumb, especially the finger-type metacarpal duplication of the thumb is often accompanied by thenar muscular dysplasia and has great effects on the thumb palm-opposition functions. It features not only different degrees of dysplasia of hand internal muscles but also abnormalities in thumb short abductor insertion, with the manifestation of relaxation to varying extents. Muscular insertions should be reconstructed during correction.

(b) Hand external dysplasia: the distal end of thumb long extensor tendon and thumb long flexor tendon of the duplicated thumb is separated into two, the insertion positions of the thumb long extensor tendon and the thumb long flexor tendon are abnormal, and instead of ending in the center of the phalangettes, it ends at the adjacent side of the radial and ulnar duplicated thumb phalangettes. Due to the displacement of the thumb long extensor tendons and thumb long extensor tendons, it inclines to the opposite side of the duplicated thumb, and the interphalangeal joint or the metacarpophalangeal joint of the two thumbs can also be made to become laterally flexed to the opposite side. The abnormal tendinous junction between the thumb long extensor and thumb long flexor tendons is the anatomic basis of lateriflection, volar flexion, or digital distortion deformities of the duplicated thumb, and in the meantime, limitations in thumb extension and flexion functions or dysplasia in extension and flexion strength result. The abnormalities in these tendon structures will gradually result in the structural abnormalities in duplicated thumb interphalangeal joint or metacarpophalangeal joint, which is also the important reason why the author recommends it be performed during the infantile period. In reconstructive surgery, it is suitable to disjunct, remove, or displace the abnormal symphysic structures or choose dynamic reconstruction.

In order to assess the hand functions before and after the duplicated thumb surgery, the author proposes that relevant records be made before and after the operation so that the surgical therapeutic efficacy can be assessed (Tables 7.3 and 7.4).

7.1.5 Treatment

Treatment of the duplicated thumb is aesthetical reconstruction of thumb shape and functions.

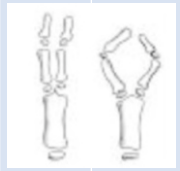
7.1.5.1 Selection of Treatment Timing

The author proposes that, if the pediatric patient suffers no serious deformities, such as cardiovascular deformities, is under good physical condition, and there is no surgical contraindications, the corrective surgery of duplicated thumb should be performed before the development period of the palm-opposing function, that is, the first surgery for correction should be completed within 6 months after birth, but the osteoarticular epiphysis of the thumb should be well protected; the treatment of secondary deformities should be performed before school age. If the functions of the affected hand should be accurately determined, the determination often needs to be made when the patient is 2 years old.

7.1.5.2 Treatment of Type I and II Duplication of the Thumb

1. Anaplasty to combine duplicated thumbs. It is the combination of the thumbs and used for the mirror image thumb deformity in type I and II duplication of the thumb, and some type III and IV duplicated thumbs are combined

Table 7.3 Records of function determination of duplicated thumb

Function												
Type interphalangeal joint				Metacarpophalangeal joint				Carpometacarpal joint				
Position of central axis		Flexion/extension		Position of central axis		Flexion/extension		Adduction/abduction		Opposition		
Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative	
I												
II												
III												
IV												
V												
VI												

(continued)

Table 7.3 (continued)

Functions												
Type interphalangeal joint				Metacarpophalangeal joint				Carpometacarpal joint				
Position of central axis		Flexion/extension		Central axis	Position	Flexion/extension		Adduction/abduction		Opposition		
Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative	
VII												

Table 7.4 Records of muscular strength determination of duplicated thumb

Muscular strength						
Type finger-to-finger pinch			Finger lateral pinch		Grip	
Preoperative		Postoperative	Preoperative	Postoperative	Preoperative	Postoperative
I						
II						
III						
IV						
V						
VI						
VII						

into two. It is generally called Bilbaut surgery, first proposed by Bilhaut M. in 1889 in one French academic report. The combining surgery of duplicated thumb involves resection of redundant tissues in the middle of the two thumbs, e.g., redundant skin, fingernails, phalanges, articular surface, and tendons are removed; the two residual structures are combined into one; and the reconstructed thumbs reach the completely normal status in shape and functions, so it is a very complicated aesthetical reconstructing surgery for thumbs.

Take type I and II duplicated thumb combined anaplasty, for example. The surgical procedures are as follows:

(a) Design of skin and fingernail incisions: at the most bulging site of the fingernail, namely, the central region, make a longitudinal incision from the nail margin to the nail root and make a triangular incision on the thumb dorsal skin at the nail root to correct the tip of the thumb with the deformity of being wide. Make a Z-plasty or sawtooth-shaped skin incision at the finger pulp to reach the phalange, and

resect the finger pulp of the wide thumb. In resection of the skin, retain excessive subcutaneous tissues to make the reconstructed finger pulp full in shape, and protect the finger nerve vessels and fingerprints (Fig. 7.23).

(b) Phalangeal osteotomy: dissect the distal phalanx, amputate the ulnar half of the radial thumb distal phalanx and the radial half of the ulnar thumb distal phalanx on the middle line of longitudinal axis of the phalanx, and use two Kirschner silk for transverse fixation, or use fine steel-silk or nylon-silk to ligate the two distal phalanxes and combine them into one. Rotate the radial thumb phalange to the volar side by 10°–15° and rotate the ulnar phalange to the dorsal side by 10°–15° so that the bone surfaces of the two sides of thumb phalange can achieve close apposition, to achieve a good nail arch of the reconstructed thumb.

(c) Skin suture: perform suturing on the thumb that is cut into half so that the formed new thumbs have full finger pulp.

(d) Repair and reconstruction of tendons and articular ligaments: as for type II duplication of the thumb, interphalangeal joint accessory ligament reshaping should be performed during the combined anaplasty of the duplicated thumb to achieve the stability of interphalangeal joints. It is also necessary to combine the thumb flexor and extensor tendons into one (Fig. 7.24).

This surgical mode can also be adopted for the treatment of some type III and IV duplication of the thumb (Fig. 7.25), but the interdigital and metacarpophalangeal joint may be damaged after operation, which will easily result in poor postoperative joint extension and flexion.

2. Partial resection of neoplastic thumb and anaplasty of residual thumb. As for type II duplication of the thumb, the two-thumb combined anaplasty can be adopted, the partial resection of neoplastic thumb and anaplasty of residual thumb can be also adopted, and the indication is that the shape and functions of one thumb of the duplicated thumbs are close to normal. The surgical method is resect neoplastic thumb, perform interphalangeal joint, and perform radial accessory ligament resection and aesthetical skin repair (Fig. 7.26.)

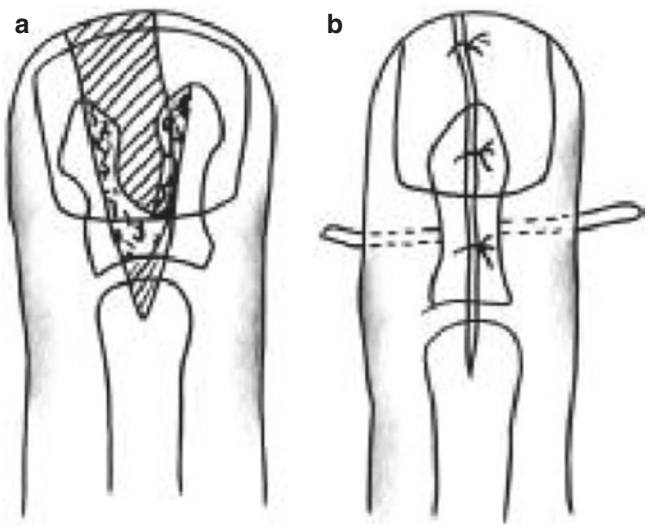


Fig. 7.23 Design of type I duplicated thumb anaplasty

7.1.5.3 Principle of Comprehensive Anaplasty of Type III to VII Duplication of the Thumb

Resection of neoplastic thumb and anaplasty of residual thumb are the basic skills of the plastic surgery of duplicated thumb and applicable to the comprehensive reshaping of type III to VII duplication of the thumb. This technology covers multiple basic techniques in the reshaping, reconstruction, and aesthetical reconstruction of congenital hand deformities [8–10].

1. Surgical principle. The aesthetical microscopic reconstructing surgical technique is utilized for the reshaping, beautification, and reconstruction of the duplicated thumb so that the shape and functions of the reconstructed finger are close to normal thumbs. Sometimes, after the corrective surgery of duplicated thumb, the degree of fine resemblance can be reached compared with normal thumbs. Comprehensive anaplasty of type III to VII duplication of the thumb should be understood to involve not only resection of neoplastic thumb but also, and more importantly, reconstruction of the functions and shape of the residual thumb.
2. Postoperative evaluation. The content of preoperative evaluation includes determination of residual thumb, profound understanding of the functional and morphological defects of the residual thumb, determination of the transplantable tissues of neoplastic thumb, and selection of surgical modes.

   Before the operation, the size, circumference, length, radian, and osteoarticular deformity status of all parts of the deformed thumb should be studied in detail, the central axis line of each joint should be measured and recorded, and the motor functions of flexion, extension, adduction, abduction, and rotation of all parts of the thumb should be measured; if possible, the muscular strength and sensation should be measured, the project of repair and reconstruction of the residual thumb should be designed, and the transplantable tissues of the neoplastic thumb should be evaluated.
3. Basic surgical skills. The duplicated thumb plastic surgery falls into the category of microscopic surgery, which should be performed during the infancy in most cases. As for the pediatric patients aged within 6 months, the minimally invasive technique should be adopted for aesthetical reconstruction:

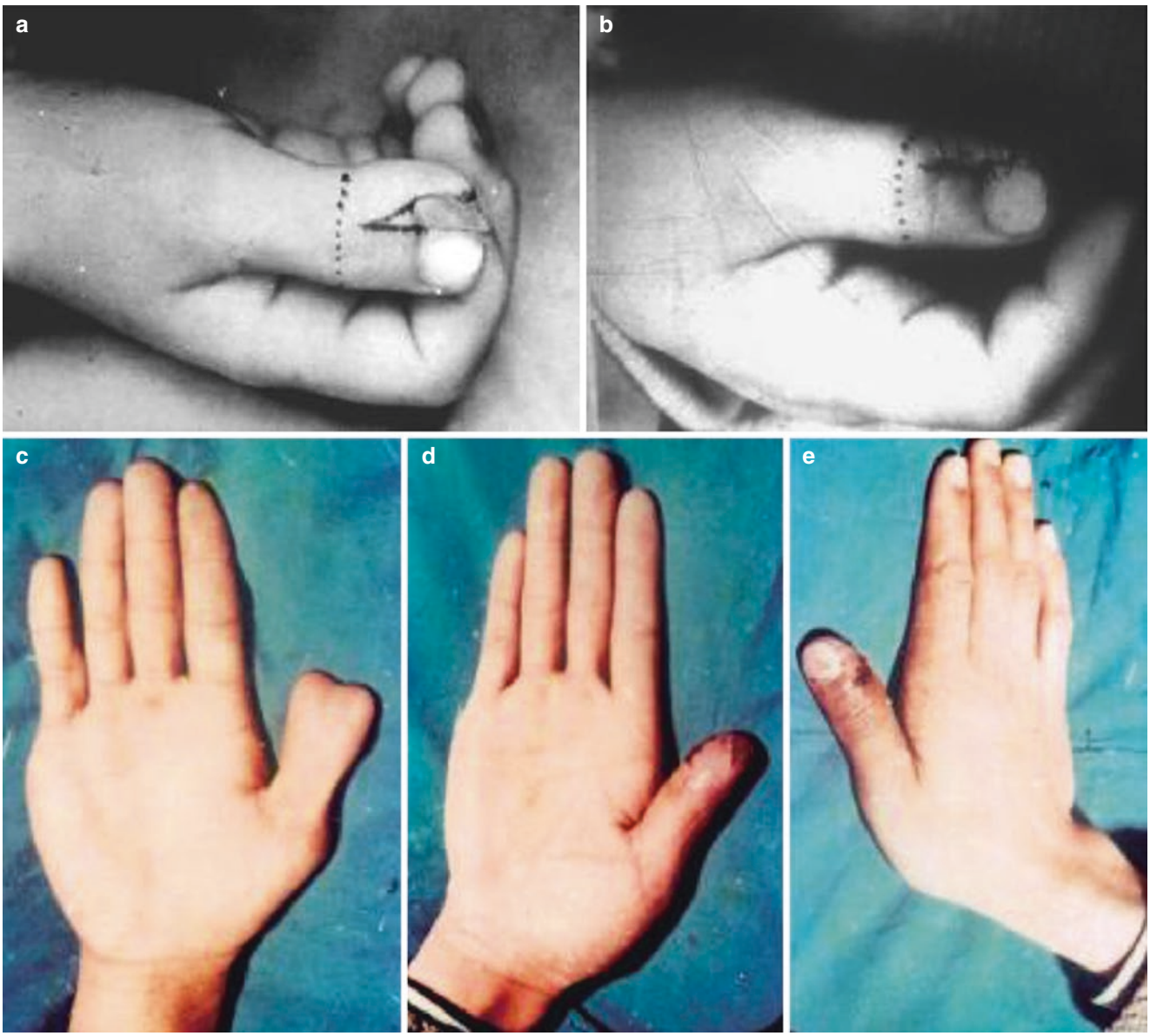


Fig. 7.24 Treatment of type I duplication of the thumb. (**a**, **c**) Before operation. (**b**, **d**, **e**) After operation

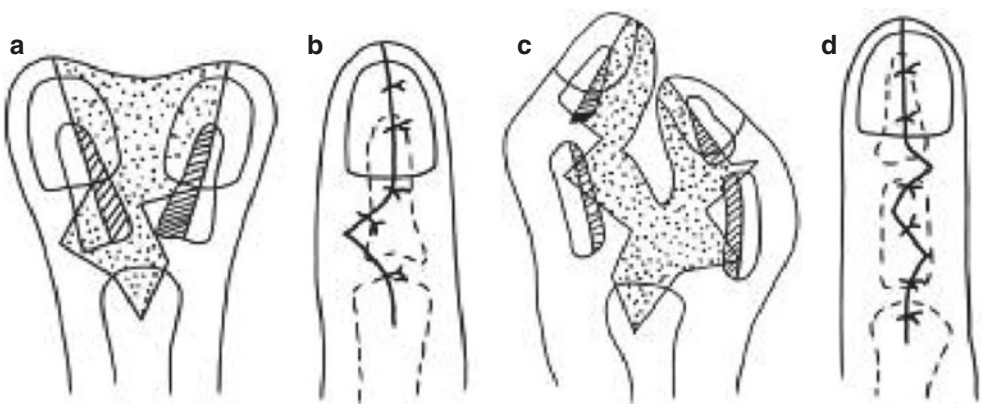


Fig. 7.25 Schematic diagram of anaplasty of type II and IV duplication of the thumb (the *dash line* region is the resected skin and subcutaneous tissues, and the oblique line region is the marker for the phalangeal osteotomy). (**a**) Surgical design of anaplasty of type II duplication of the thumb. (**b**) Surgical effects of anaplasty of type II duplication of the thumb. (**c**) Surgical design of anaplasty of type IV duplication of the thumb. (**d**) Surgical effects of anaplasty of type IV duplication of the thumb

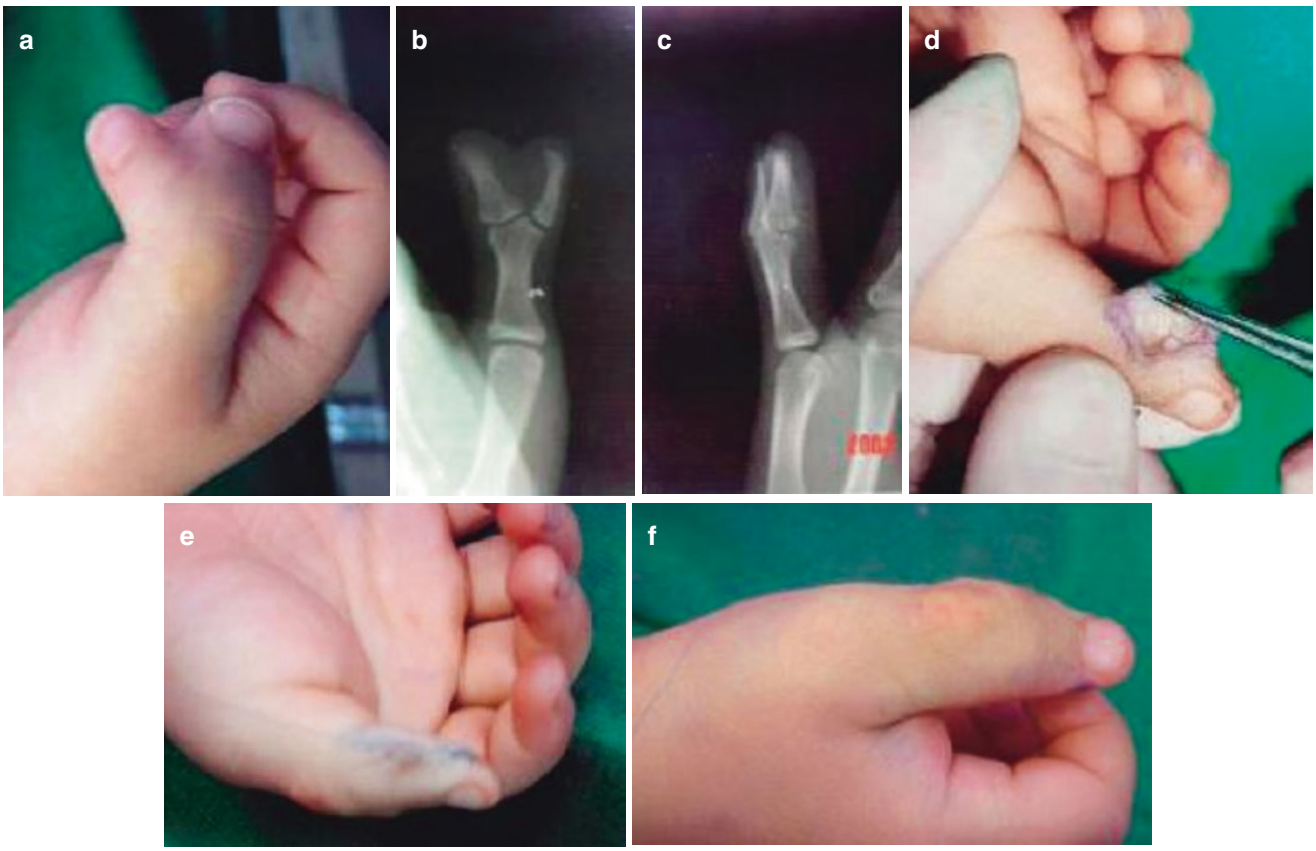


Fig. 7.26 Plastic treatment of type II duplicated thumb anaplasty. (**a**) Shape of the thumb before operation. (**b**, **c**) X-ray films before operation. (**d**) Collateral ligament reconstruction of interphalangeal joint of the duplication of the thumb during the operation. (**e**, **f**) Thumb deformity corrected after operation

(a) Design of skin incisions: make a Z-shape or curve-shaped skin incision, and do not make a straight incision to avoid cicatrical contracture of incision after operation.
(b) Plastic surgery of phalange and interphalangeal joint: amputate the bulging part of the interphalangeal joint of the neoplastic distal and the proximal phalanx and retain the lateral periost and accessory ligaments in resection of phalanx, for the purpose of repairing the accessory ligaments of interphalangeal joint of the residual thumb. Resect the epiphysis of the residual thumb to prevent local phalangeal proliferation. In order to guarantee the stability of the repaired interphalangeal joint, Kirschner silk can be used to fix the interphalangeal joint in the extension position for 3 weeks.
(c) Skin suture: the tension-free, curve-shaped (or Z-plasty), accurate, and fine suturing techniques are adopted.

4. Microsurgical aesthetical reconstructive technology:
(a) Transplantation of neoplastic thumb: it is not suitable to completely resect the neoplastic thumb. Its flaps with blood supply can be used for transplantation for the use of morphological reshaping on the extra large thumb, such as fabrication of wide, flat, and full finger pulp and vivid fingernails and folds, correction of skin defects after angulation deformity of thumb interphalangeal joint, and fabrication of an almost normal thumb web.
(b) Osteotomy and bone grafting: it can be used to correct the angulation deformity or central axis deviation of interphalangeal joint or metacarpophalangeal joint of the residual thumb.
(c) Repair and reconstruction of joint stability: in resection of the neoplastic thumb at the interphalangeal joint or metacarpophalangeal joint, the joint accessory ligaments or partial periost of the thumb should be retained for the repair and reconstruction of the joint accessory ligaments of the thumb.
(d) Functional reconstruction of the residual thumb: the thumb extensor or thumb flexor tendons of the neoplastic thumb should be retained according to the specific needs for the reconstruction of the extension or flexion functions of the residual thumb or the correction of the joint lateral deviation deformity.
(e) Postoperative treatment: protect the blood supply of the grafted flaps. For those that need tendon transplantation or bone and joint correction, fixation and braking for over 3 weeks are needed.

During plastic surgery of the duplicated thumb, prepare the neoplastic thumb that is to be resected and use

the original bones, joints, skin, tendons, and ligaments for tissue transplantation as the donor sites of the thumb morphological and functional reconstruction of the residual thumb. Before the operation, the surgeon should carry out detailed visual, physical, and X-ray examinations on the shape, functions, and structures of the residual thumb and neoplastic thumb; establish plans on the aesthetical and functional reconstruction of the residual thumb; and, in the meantime, design the transplantable skin flaps, ligaments, tendons, and bones. Therefore, correction of the duplicated thumb is a technology that applies microsurgical techniques for the morphological and functional reconstruction of the thumb (Figs. 7.27 and 7.28).

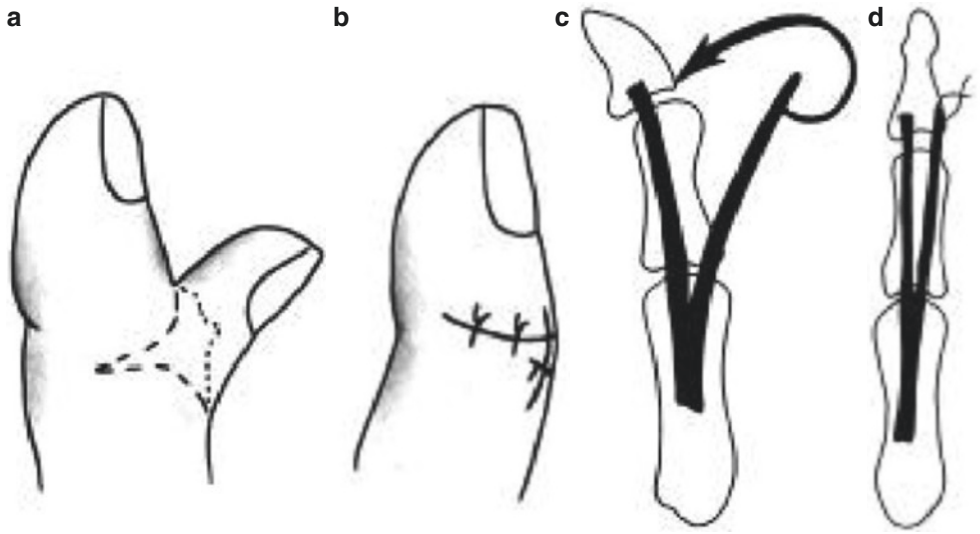


Fig. 7.27 Surgical design and surgical simulation process of type II to IV duplicated thumbs and neoplastic thumbs. (**a**) Design of skin incisions: design V-shaped incision at the dorsal side of the radial interphalangeal joint of the residual thumb, design a V-shaped incision at the ulnar side of the volar surface of the neoplastic thumb, and fabricate the V-shaped skin flaps of the neoplastic thumb for repairing and beautifying the central axis deviation of the interphalangeal joint of the residual thumb or fabricate one vessel (or nerve) containing island flaps for the deformity correction and morphological and aesthetical reconstruction of the residual thumb. (**b**) Surgical effects. (**c**, **d**) Grafting the thumb long extensor tendon or thumb long flexor tendon of the neoplastic thumb to correct the ulnar deviation deformity of the thumb interphalangeal joint; in case of the presence of the central axis deviation of the interphalangeal deformity, it is necessary to perform osteotomy for correction

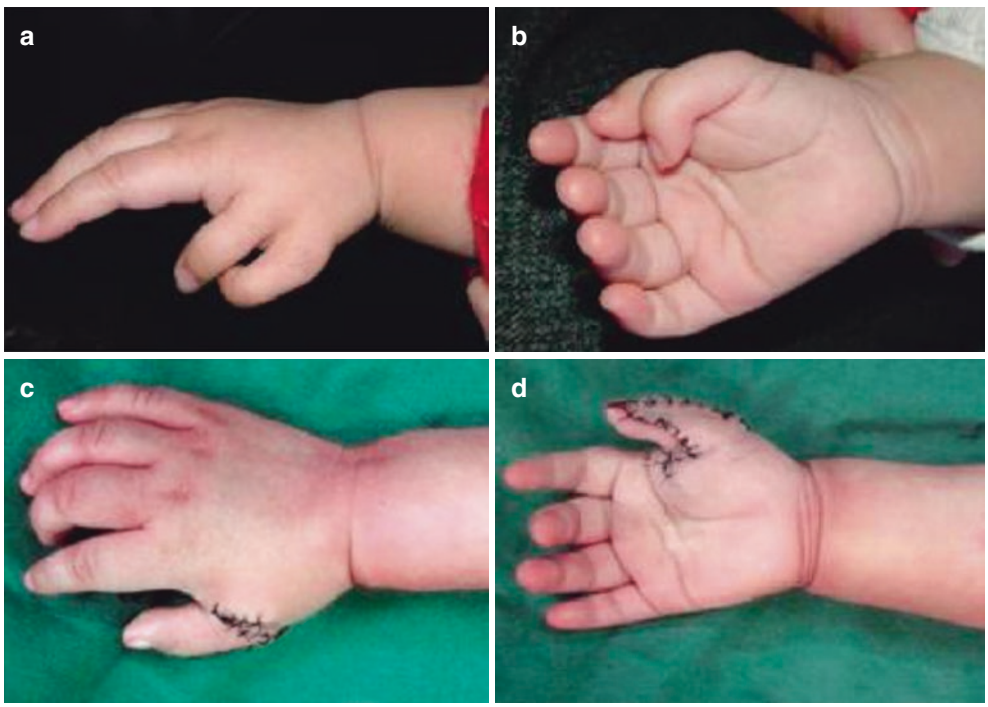


Fig. 7.28 Plastic surgery of type V duplicated thumb. (**a**, **b**) Before operation, type V duplication of the thumb is suffered; the four fingers develop well; the preoperative examination indicates that the position, shape, and functions of the ulnar thumb are normal; and the thumb is diagnosed as residual thumb. As the residual thumb is fine and small with functional insufficiency of extension and flexion, neoplastic thumb plans to be used as the donor site for tissue transplantation to enlarge the residual thumb and perform functional reconstruction. (**c**, **d**) After operation

5. Surgical design and implementation error. During the treatment of duplication of the thumb, surgical design and implementation errors are common. The commonest error is simple resection of one of the duplicated thumbs as the treatment of the duplication of the thumb, or although the residual thumb was previously repaired, no complete plastic and aesthetical reconstruction of the skin, bones, joints, ligaments, and tendons is performed; therefore, poor surgery will have pessimistic effects on the phase II repair of type III to VII duplication of the thumb (Fig. 7.29). In addition, the surgeons should pay sufficient attention; if they do not master corresponding plastic surgery knowledge, the pediatric patient with duplication of the thumb should be grafted to the specialist for treatment.

7.1.5.4 Common Surgical Methods of Plastic Surgery of Type III to VII Duplication of the Thumb

1. Resection of neoplastic thumb. It is applicable to the major-minor type duplication of the thumb. The phalanges of neoplastic thumb and the corresponding interphalangeal joints are surgically resected. If the deformity is type IV to VII duplication of the thumb and its metacarpal bone also suffers deformity, neoplastic metacarpal bone should be resected when the neoplastic thumb is being resected. Under normal circumstances, as for type IV and VII duplicated thumb, after the basal part of the proximal phalanx of the neoplastic thumb is resected, the protruding part of the metacarpal bone should be resected,

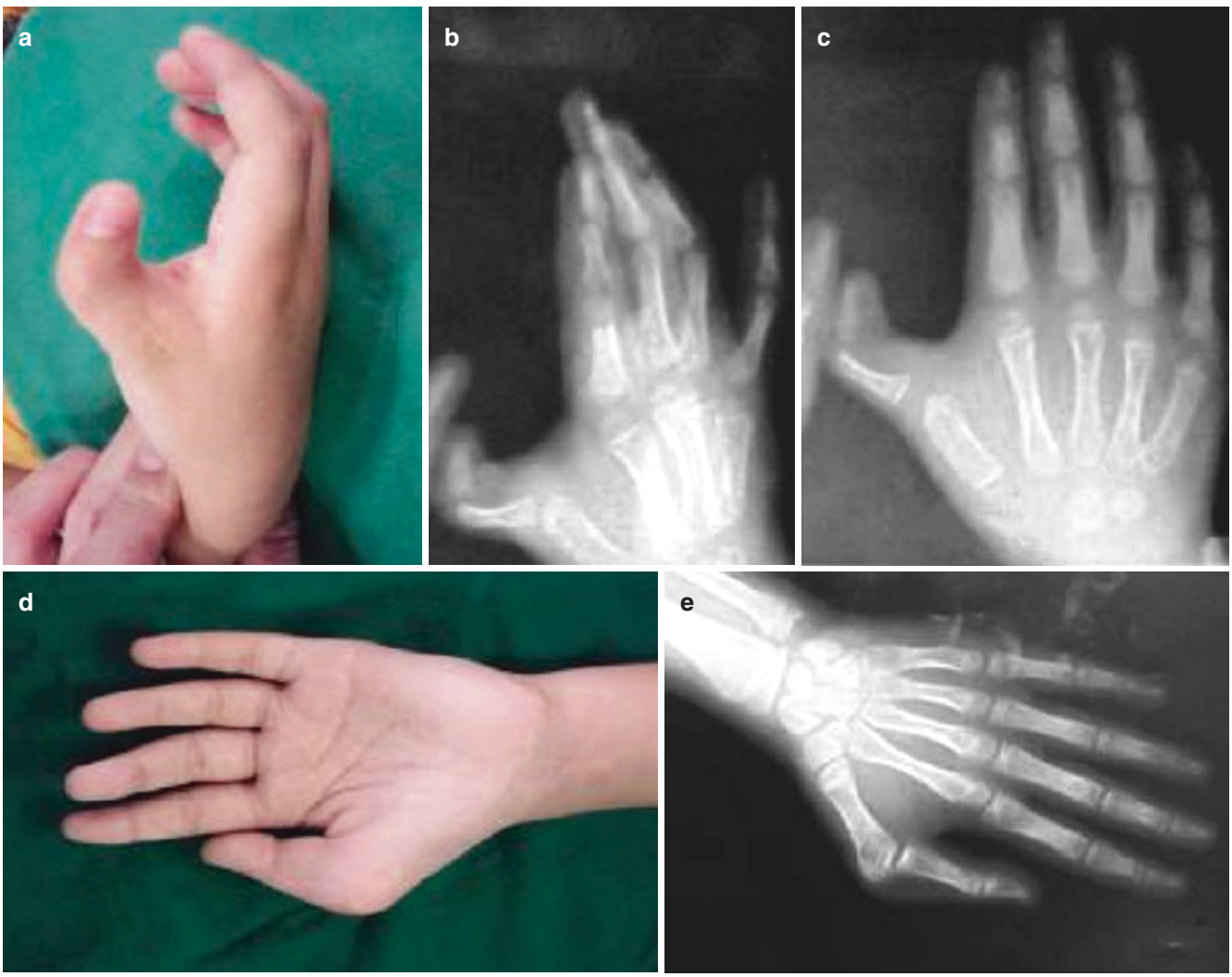


Fig. 7.29 Surgical errors of lobster-tong-shaped thumb in the type IV duplication of the thumb. (**a**–**c**) Perform simple surgical resection of one thumb, keep the residual thumb deformity after the operation; the thumb metacarpophalangeal joint and interphalangeal joint suffer angulation deformity, and phase II repair and reconstruction are needed. (**d**, **e**) Type IV duplication of the thumb, the patient once underwent resection of one thumb; after the operation, the patient experienced middle axis lateral deviation of the metacarpophalangeal joint of the residual thumb, metacarpal bone head protrusion, and metacarpophalangeal joint deformity

and then the metacarpophalangeal joint accessory ligaments should be repaired or reconstructed to prevent the metacarpal bone head protrusion and metacarpophalangeal deformities after the operation.

2. Transplantation of distal thumb with vascular vessels. It is applicable to the major-minor type duplication of the thumb. The major-minor type duplication of the thumb can be manifested as good development of the free part at the distal end of the thumb, but poor metacarpal bone formed with the metacarpal bone; the development at the proximal end of the other thumb is relatively good, and a broad thumb web can be formed. At this time, the thumb at the distal end that develop well can be utilized to transplant the distal thumb with vascular nerves to the basilar part of the amputated thumb with good metacarpophalangeal joint at the proximal end (Fig. 7.30).
3. Aesthetical and functional comprehensive anaplasty. The comprehensive anaplasty includes resection of neoplastic thumb; utilization of vascular nerve island flaps, tendons, bones, and joints and periarticular accessory ligaments of the thumb for dynamical functional reconstruction; and morphological aesthetical modeling of the skin, bones, joints, ligaments, and tendons of the residual thumb. The comprehensive anaplasty is applicable to type III to VII duplication of the thumb:

   The core technology of comprehensive plastic surgery of the duplicated thumb is applied for the treatment of type IV and VII duplication of the thumb. The thumb shapes of type IV duplicated thumb include lobster-tong shape, parallel shape, and quasi-lobster-tong shape, and there are differences in terms of the complexity of the surgery. Take the treatment and design of the lobster-tong deformities as an example. The technology of clinical application is as follows:

   (a) Preparation of vascular nerve island skin flaps of neoplastic thumb: design Z-plasty skin incision at the finger dorsum and finger pulp of the neoplastic thumb and design oval-shaped, rhomboid, or polygonal vascular nerve island flaps at one side of the finger pulp. One to two skin flaps should be designed according to the needs to enlarge the finger pulp, fingernails, and finger folds of the residual thumb and repair the skin defects after correction of interphalangeal joint and metacarpophalangeal joint lateriflection and flexion deformities (Fig. 7.31).

   (b) Correction of finger flexion and lateriflection deformities: after the resection of the neoplastic thumb, the flexion and lateriflection deformity of metacarpophalangeal joint or interphalangeal joint of the residual thumb should be corrected. The joint flexion and lateriflection deformities are induced by following factors: ① abnormalities in the quantity and shape of the bones and articular surface and capsule deformities, ② dysplasia of accessory ligaments at two sides of the

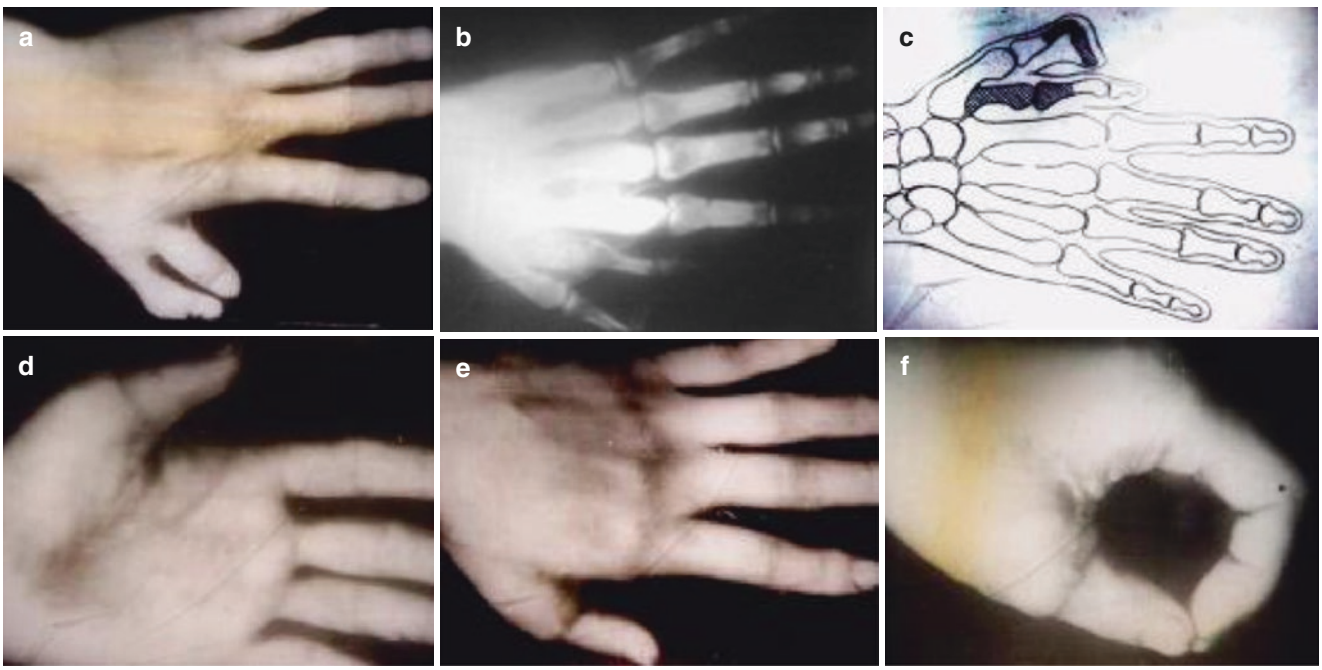


Fig. 7.30 Transplantation of distal thumb with vascular nerves of the left hand type VI duplication of the thumb. (**a**–**c**) Before operation, type VI duplication of the thumb, thumb web stenosis; although the medial thumb is short and small, the shape of the distal thumb is normal, the lateral thumb is in the good palm-opposition position, and the thumb interphalangeal joint suffers deformity of flexion. (**d**) Surgical design: resect the distal end of the flexed thumb of the lateral interphalangeal joint, take the distal end of the proximal phalanx of the medial thumb to perform grafting with vascular nerves, repair and reconstruct the lateral thumb, and repair the thumb web. (**e**, **f**) After operation

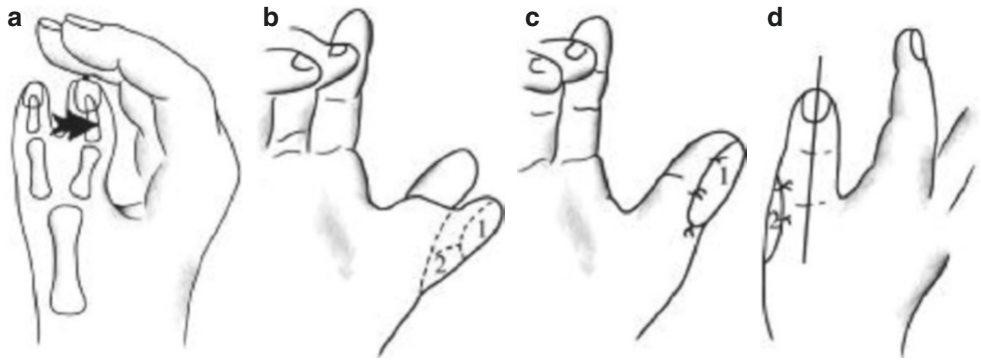


Fig. 7.31 Design of vascular nerve island flap grafts of the neoplastic thumb. (**a**) Resect the neoplastic thumb and perform transplantation of relevant tissues as reconstruction of the functions and shapes of the residual thumb. (**b**) Design vascular nerve island flaps 1 and 2 on the neoplastic thumb, and plan to graft them on the residual thumb. (**c**) Grafting the vascular nerve island flap 1 of the neoplastic thumb to the finger tip of the residual thumb to enlarge the residual thumb for the plastic surgery of the finger pulp of the residual thumb. (**d**) Use the vascular nerve island flap 2 of the neoplastic thumb to repair the joint lateriflection deformity of the residual thumb

joints and imbalanced sustaining strength at two sides, ③ abnormalities in insertion position of flexor and extensor tendons and tendon labyrinth, and ④ structural and distribution abnormalities of skin and subcutaneous fascia. In the twist-type duplication of the thumb, the above factors often coexist, which constitute the diversity of the duplication of the thumb.

Reshaping of the phalanx and metacarpal bone: after the osteoarticular resection of neoplastic thumb, the twist deformity of the residual thumb should be corrected. What is important is to perform osteotomy of phalange and metacarpal bone, but in osteotomy of phalange or metacarpal bone, joint epiphysis and cartilage injuries should be prevented as much as possible. Often, wedge-shaped osteotomy or bone grafting of proximal phalanx and metacarpal bone should be performed so that the deviating interphalangeal joint or metacarpophalangeal joint planes are in the horizontal position, the articular surface is smooth, the cartilage apposition of the articular surface is good, the middle axis of the joint and the middle axis of the thumb are consistent and in the palm-opposition position, and they are opposite to other fingers. After osteotomy or bone grafting, Kirschner silk are used for fixation. Steel silk can also be used for ligation or micro-steel plates can be used for fixation. During the osteotomy, the joint accessory ligaments should be protected to serve as the tissue source of the joint stability and deformity repair.

Reshaping of interphalangeal joint and metacarpophalangeal joint: type III to type VII duplication of the thumb can result in the poor or unstable shape of interphalangeal joint or metacarpophalangeal joint after resection of neoplastic thumb. Type III duplicated thumb has thick proximal phalanx, which forms unstable interphalangeal joint with thin distal phalanx of the residual thumb; type V duplicated thumb has thick metacarpal bone, which forms unstable metacarpophalangeal joint with thin proximal phalanx of the residual thumb. For these two types of deformity, it is necessary to amputate the distal interphalangeal joint of the bulging proximal phalanx (type III), or amputate the metacarpophalangeal joint at the distal end of bulging metacarpal bone (type V), so that the reconstructed interphalangeal joint or metacarpophalangeal joint can take streamline shape. The middle axis of the joint conforms to the thumb growth direction without local abnormal bulge. However, in amputation, the joint accessory ligaments should be protected so that the accessory ligaments can be repaired after the resection of neoplastic thumb to enhance joint stability.

In the reshaping of interphalangeal joint and metacarpophalangeal joint, it is necessary to judge before operation the factors such as imbalanced tension of bilateral accessory ligaments, and volar plate tension or relaxation, as well as perform repair and correction during operation.

(c) Correction and repair of tendon deformities. Type III to VII duplications of the thumb often suffer abnormalities in thumb long flexor tendon and thumb long extensor tendon insertions. The insertion is not located in the middle of the phalangettes but at the adjacent lateral sides of the two thumbs, which is the reason why the interphalangeal joint suffers angulation deformity. At this time, the two tendons not only serve to extend and flex the interphalangeal joint and metacarpophalangeal joint of the thumb but also lateriflex them. To correct this deformity, two surgical modes can be adopted: one is to cut the thumb long flexor tendon and the thumb long extensor tendon of the neoplastic thumb, rotate and graft it to the opposite side of the phalangette of the residual thumb, and reconstruct the

insertions to balance the dynamical direction of flexor and extensor tendons of the thumb and the other is to remove the thumb long extensor and the thumb long flexor tendons of the neoplastic thumb and the residual thumb from the insertions, combine them into one, and replace the insertions in the middle of the phalangette. As the pediatric patients with duplication of the thumb are very young and the dysplasia makes tendons very small, the surgeons are required to possess proficient microsurgical techniques to perform minimally invasive and accurate anatomy and separation and perform accurate repair and reconstruction so that this surgery can be finished.

Displacement of hand internal muscle tendons: in the resection of neoplastic thumb, detach the thumb short abductor with the periost at the fundus of the proximal phalanx and retain sufficient length, but the metacarpophalangeal joint accessory ligaments are not contained. Fix it together with the basal part of the proximal phalanx of the residual thumb, reconstruct the insertions, and use Kirschner silk to perform temporary fixation of the metacarpophalangeal joint.

7.1.5.5 Typical Cases

1. Case I. A male patient with right hand type III duplication of the thumb has duplicated thumb taking lobster-tong shape and nearly normal thumb web. The ulnar thumb is chosen as the residual thumb, this thumb is small, the interphalangeal joint suffers angulation deformity toward the radial side, and the muscular strength of the thumb is weak. Surgical design: incise the skin at the radial margin of the residual thumb, separate the thumb volar and dorsal skin, disjunct the thumb long extensor tendon, perform phalangeal osteotomy to correct the middle axial angulation deformity of the thumb interphalangeal joint, and shorten the radial hypertrophy of the first metacarpal bone head. Partially resect the neoplastic thumb, take the flap grafts of the neoplastic thumb, and enlarge the residual thumb; take the extensor tendon grafts of the neoplastic thumb, and strengthen the thumb long extensor; use the metacarpophalangeal joint accessory ligaments and periosteal flaps of the neoplastic thumb to repair and reconstruct the radial accessory ligaments of the thumb metacarpophalangeal joint. After operation, use the Kirschner silk for fixation (Fig. 7.32).
2. Case II. Plastic treatment of lobster-tong-shaped thumb in the type IV duplication of the thumb.

   In the plastic treatment of duplication of the thumb, the repair of the lobster-tong-shaped thumb is difficult, and there are many deformities of the residual thumb to be corrected, including thin thumb, interphalangeal joint deformity, phalangeal deformity, joint angulation deformity, thumb extensor and thumb flexor tendon dysplasia, etc. (Figs. 7.33 and 7.34).
3. Case III. Plastic treatment of parallel thumbs in the type IV duplication of the thumb.

   The pediatric patient suffers type IV major-minor type duplication of the thumb, the two thumbs are parallel, and their sizes are different. The radial thumb is relatively small with few functional applications; the ulnar thumb is relatively large and it is frequently used. The thumb is in the palm-opposition position with nearly normal thumb web. Therefore, the surgeons decide to resect the neoplastic thumb of the radial thumb, and take the skin flaps and tendons for transplantation; the ulnar thumb is the residual thumb, and the tissue transplantation of the neoplastic thumb is performed for functional and morphological repair and reconstruction.

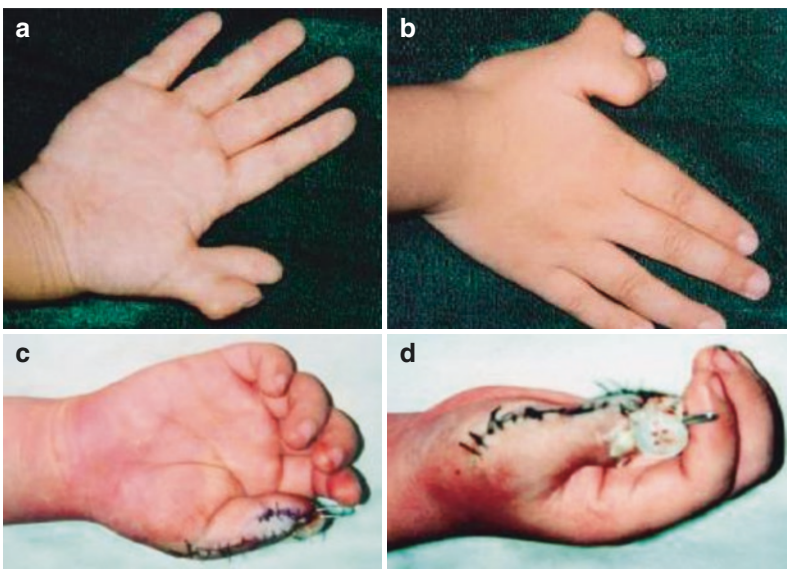


Fig. 7.32 Case I. (**a**, **b**) Before operation. (**c**, **d**) After operation

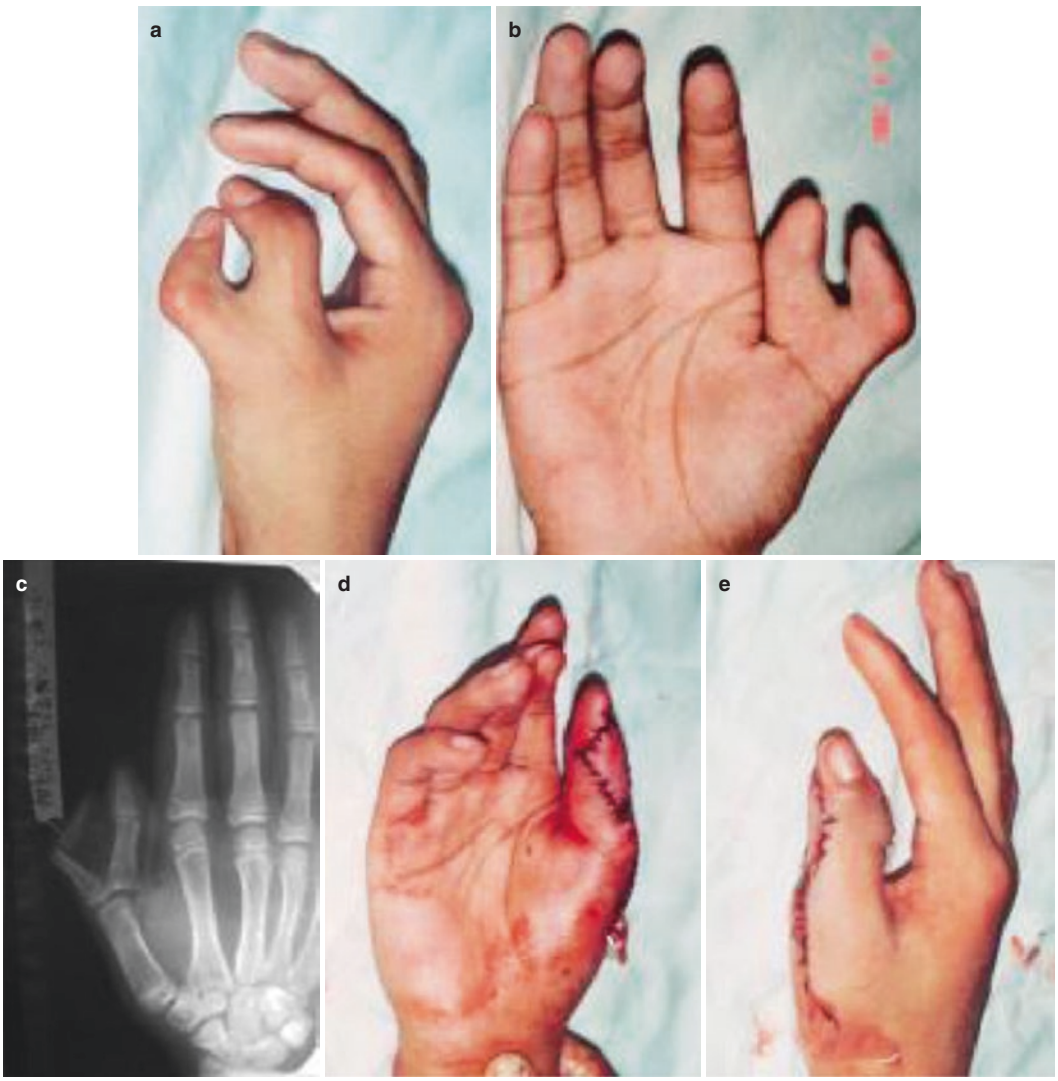


Fig. 7.33 Case II. (**a**, **b**) Hand appearance before operation. (**c**) X-ray film before operation. (**d**, **e**) Hand appearance after operation

The tissues of the radial neoplastic thumb are adopted to repair the ulnar residual thumb. When the residual thumb is large-sized, the vascular nerve island skin flaps of the neoplastic thumb are only used to repair the skin defects of the metacarpophalangeal joint of the residual thumb (Fig. 7.35).

4. Case IV. The repair and reconstruction of metacarpophalangeal joint accessory ligaments of the quasi-lobster-tong-shaped thumb of type IV duplication of the thumb.

   The manifestation of quasi-lobster-tong-shaped thumb in the category of type IV duplication of the thumb is the mild deviation or flexion deformity in interphalangeal joint or metacarpophalangeal joint. In the deformities of type IV duplication of the thumb, the quasi-lobster-tong-shaped deformity is milder than the lobster-tong-shaped deformity, but it is more severe than the parallel deformity. This treatment method of this type of deformity is similar to the lobster-tong-shaped deformity, and the corrective surgery is relatively simple. The correction process is shown in Fig. 7.36.

5. Case V. Plastic treatment of floating duplicated thumb deformity

   Treatment method of floating duplicated thumb deformity is relatively easy. For patients with type V to VII floating duplication of the thumb, good surgical effects can be produced as long as the neoplastic thumb is resected and the skin and bone joints of the residual thumb are repaired and reconstructed (Fig. 7.37).

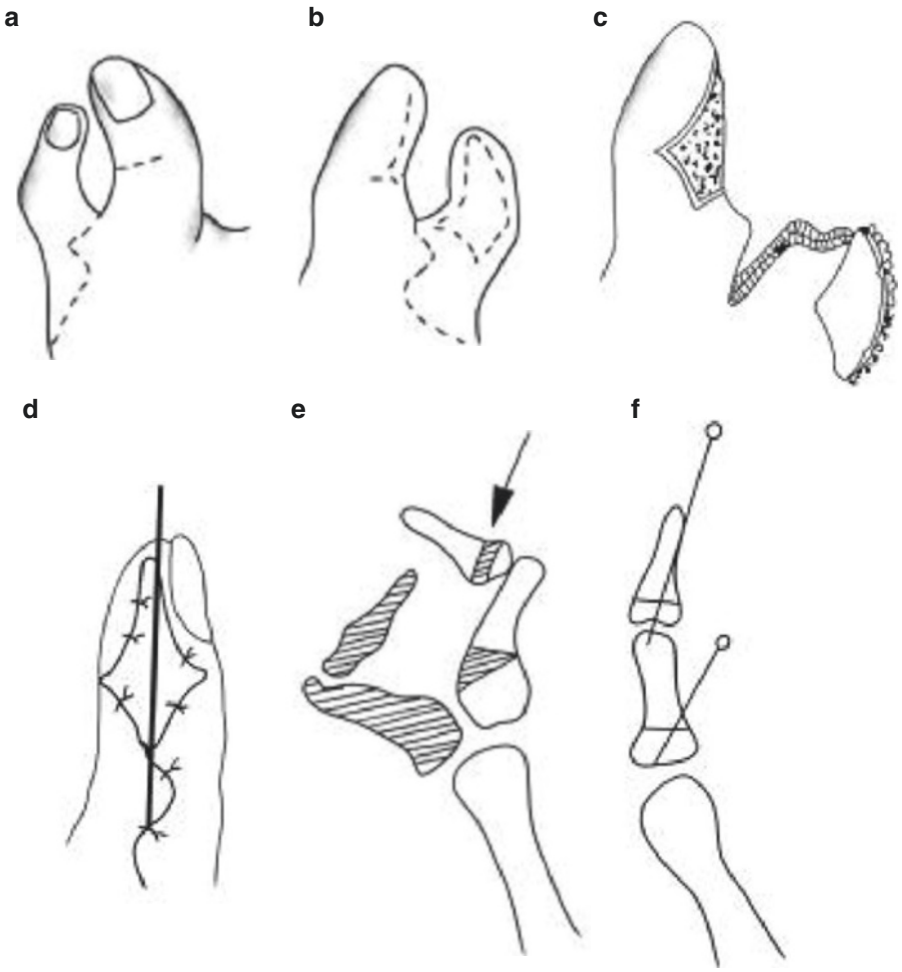


Fig. 7.34 Schematic diagram of design of comprehensive plastic surgery of type IV duplication of the thumb. (**a**, **b**) Skin incision design. (**c**, **d**) Vascular nerve island flap preparation of neoplastic thumb and the transplantation effects. (**e**, **f**) Reshaping of bones and joints

7.1.6 Phase II Repair of Residual Deformities

The simple resection of duplication of the thumb often results in residual deformities and secondary dysplasia, and the residual deformities of the thumb will become aggravated progressively with the increase in age, but the evolution can be slow. Therefore, the plastic and reconstructive therapy of the residual deformities after the simple resection of duplication of the thumb is necessary, and the treatment should be given as early as possible.

7.1.6.1 Clinical Manifestations of Residual Deformities

1. Thumb dysplasia. The thumb suffers dysplasia to varying extents, manifested as residual thumb smaller than healthy thumb.
2. Thumb adduction deformity. The manifestations of residual thumb are as follows: adduction deformity, excessively superficial and small first fingerweb, limited thumb abduction activities, and disorder of palm-opposition or finger-opposition functions.
3. Joint deviation. It includes the deviation of thumb interphalangeal joint toward the ulnar side or the radial side, the deviation of metacarpophalangeal joint toward the ulnar side or radial side, or the deviation of the two joints toward the opposite direction. The phalange and metacarpal bone are not in the central extension position, making the transmission force of extensor and flexor tendons decreased and further leading to disorder of extension and flexion function. The ulnar deviation or radial deviation of the interphalangeal joint coexists with the radial deviation or ulnar deviation of the metacarpophalangeal joint; in clinical examination, the bony protrusion or projection deformity is palpable at the site of deviation.
4. Weakness of thumb extension. Due to the internal and external muscle dysplasia of the thumb, the thumb extension, flexion, abduction, and adduction muscular strength decrease; the grip strength, finger-to-finger pinch, and three-finger pinch strength decrease; and the thumb extensor function is often lost or deteriorates.

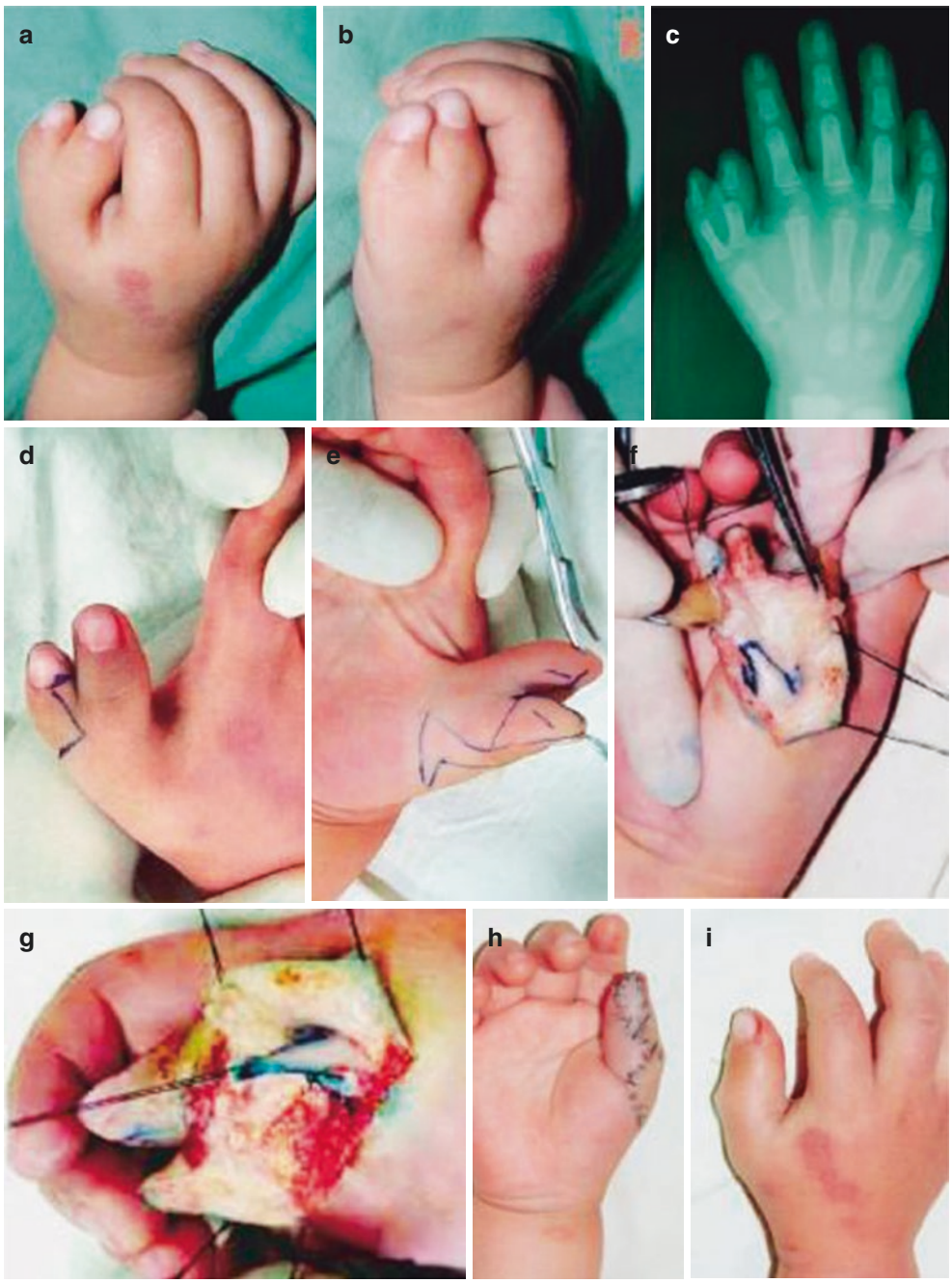
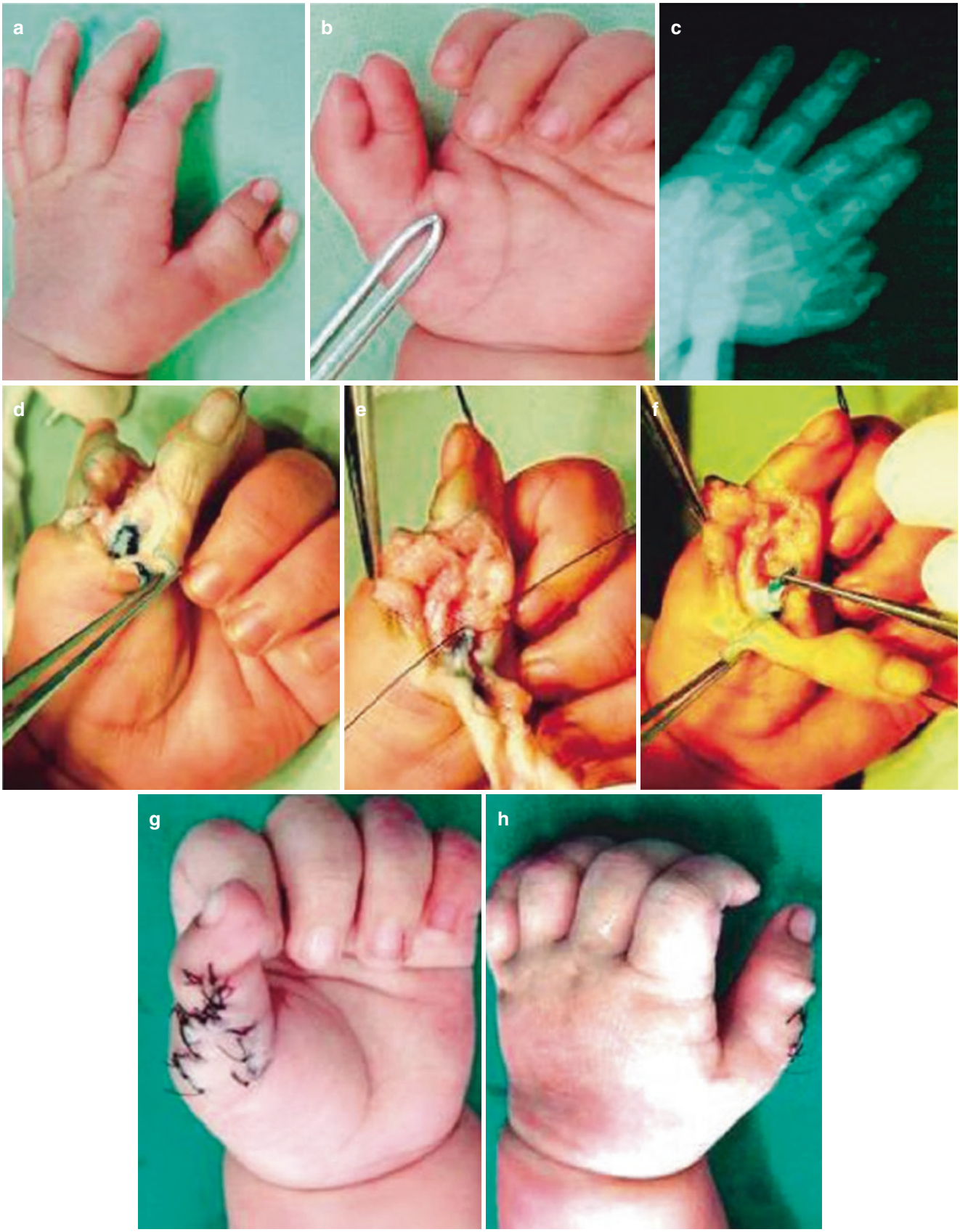


Fig. 7.35 Case III. (**a**, **b**). Preoperative hand appearance. (**c**) Preoperative X-ray film. (**d**) Design Z-shape skin incision. (**e**) Design graft flaps of neoplastic thumb. (**f**) Grafting of thumb extensor tendon of neoplastic thumb. (**g**) Reshaping of metacarpophalangeal joint accessory ligaments: design linguiform tendon flaps (pulled with black silk) on the radial thumb long extensor tendons of the metacarpophalangeal joint of the neoplastic thumb, design and take the radial metacarpophalangeal joint accessory ligaments at the radial side of the metacarpophalangeal joint of the neoplastic thumb, use the silk to elevate the linguiform tendon flaps and the radial metacarpophalangeal joint accessory ligaments, and repair and reconstruct the metacarpophalangeal stable structures of the residual thumb. (**h**, **i**) Hand appearance after operation

Fig. 7.36 Case IV. (**a**, **b**) Hand appearance before operation. (**c**) X-ray film before operation. (**d**) Neoplastic thumb pulled by silk, some periost and joint accessory ligaments retained that are marked by *methylene blue*, used to repair the metacarpophalangeal joint radial accessory ligaments of the residual thumb. (**e**) Thumb extensor tendon, some periost, accessory ligament tissue flap and skin flap remaining after the resection of neoplastic thumb that are marked by *methylene blue*; metacarpophalangeal joint radial accessory ligaments, thumb extensor tendon, and skin of residual thumb for repair. (**f**) Metacarpophalangeal joint accessory ligaments marked in *methylene blue*, for the repair of the metacarpophalangeal joint radial accessory ligaments for the residual thumb. (**g**, **h**) After operation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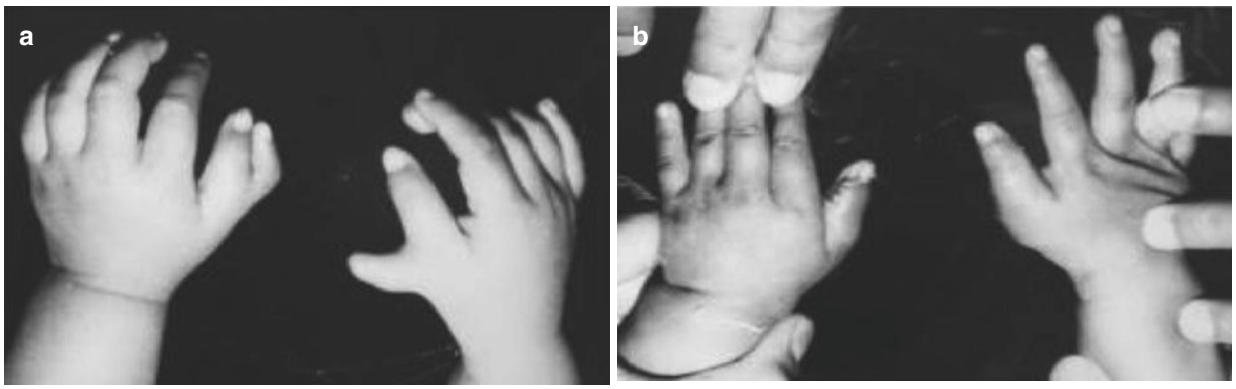

Fig. 7.37 Case V. (**a**) Before operation. (**b**) After operation

5. Others. Thumb phalangeal and metacarpal dysplasia, unstable interphalangeal joint and metacarpophalangeal joint, etc.

7.1.6.2 Treatment of Residual Deformities

The residual deformities from the simple resection of duplicated thumb are often the continuous development of the abnormal anatomic structure of the duplicated thumb, and the simple surgical resection usually cannot solve the problem of normalized changes of abnormal anatomic structures. The surgeons often find the following abnormal anatomic structures during the diagnosis and treatment of the residual deformities of the duplicated thumb: ① phalangeal and metacarpal bones suffer dysplasia; ② the residual epiphysis and duplicated thumb have dual articular surfaces; ③ the insertions of the thumb short abductors become displaced downward; ④ the thumb long extensor is fine, and its insertion becomes displaced toward the lateral side; and ⑤ joint capsule becomes lax. These abnormal structures can exist independently and can also coexist with other deformities, which is the anatomic basis for generation of residual deformities.

As for the abnormal anatomic structures, the main measure should be comprehensive plastic and reconstructive therapy, including ① reshaping of skin soft tissues, ② refixation of abnormal muscular insertions, ③ reconstruction of joint capsule accessory ligaments, ④ repair and maintenance and osteotomy of articular surfaces, ⑤ necessary tendon grafting and replacement, and ⑥ utilization of toes or partial toe-to-hand free grafting for repair and reconstruction.

1. Plastic and reconstructive therapy of adduction deformity Z- or multi-Z reshaping method is adopted for the correction (the details are shown in relevant content of this chapter).
2. Correction of joint deviation. First, the causes of joint deviation and pathological anatomical defects should be identified: induced by osteodysplasty, induced by articular ligament relaxation and dysplasia, induced by dysplasia of hand internal muscle and hand external muscle, or the combination of the above three. Generally, the relaxation of joint capsule accessory ligaments, abnormal displacement of muscular insertion, residual epiphysis, and dual articular surface abnormalities are common. The treatment should be based on anatomical abnormalities, and the correction methods include the following two:
   (a) Ligament-periosteum flap combination: the joint accessory ligaments and proximal phalange or metacarpal periosteum are utilized to constitute the accessory ligament-periosteum combined flap. The pedicle is designed at the proximal end, shaped like a reverse U. Elevate the combined flap at the distal end of the joint; after the manual reposition of joint, tighten up and re-suture the combined flap (refer to Fig. 7.36). As for the relatively mild joint deviation deformity, this method produces good repair effects.
   (b) Osteotomy and correction: perform wedge-shaped resection on the deviating phalange or metacarpal bone, or cut the residual articular surface; after manual reposition, make the phalangeal and metacarpal bone be in the central extension position and fix them with Kirschner silk.
3. Repair of extensor functions. In case of duplication of the thumb, the thumb long extensor is relatively thin, decreasing the thumb extending muscle strength, or the insertions are located at the lateral side of the phalange. In surgery, dissect the residual thumb long extensor tendon at the proximal end of the incision of the original duplicated

thumb; after one segment is dissected and dissociated toward the proximal side, pass through the subcutaneous regions and reach the residual thumb, perform suturing with the thumb long extensor tendon of the residual thumb, combine the two tendons and consolidate the thumb extending strength, and after operation, use Kirschner silk or plaster support to fix the thumb in the extension position for 6 weeks. In case the thumb extensor tendon is excessively thin or absent and cannot be adopted, the index finger inherent extensor tendon or the radial wrist long extensor tendon can be adopted for grafting and replacement. In case of dysfunction of the thumb long extensor, the anatomy indicates that this muscular development is thin; it may also be found that this muscular insertion can become abnormally displaced toward the posterior side and the lateral side and ends at the proximal side or ulnar side of the distal end of the proximal phalanx. In surgery, the abnormal insertions should be separated and should be re-sutured to the dorsal side of the phalangette fundus to recover the functions of thumb long extensor.

4. Repair of thumb dysplasia. There is no ideal method for it except the comprehensive plastic treatment during the phase I repair of the duplication of the thumb. The author once utilized the second toe complex tissue for free grafting and thumb reconstruction to improve the appearance and functions of the thumb.
5. Surgical precautions. It includes ① during reshaping of skin contracture due to thumb adduction, the downward shift of the insertions of thumb short abductor can often be observed, so the insertions should be reconstructed so that the thumb abduction functions can be improved; ② when the ligament-periosteum flap is adopted to correct joint deviation, see whether there is abnormal change in thenar muscular insertions; ③ after the reduced phalangeal and metacarpal bone are in the central extension position, they should be refixed to the normal position; ④ after the two thumb long extensors are combined and sutured, examinations should be made to check whether this muscle becomes displaced toward the posterior side and the lateral side; and ⑤ as for the patients with serious joint deviation, the cutting should not be too deep in order to avoid exposing the nerve cavity and increasing the cicatrization inside the joint capsule, and after the joint chipping, use the combined flaps for repair.

The phase II repair of the duplicated thumb is similar to the comprehensive reshaping of the duplicated thumb, the tissue transplantation should be selected according to the degree of defects of skin and subcutaneous tissues, and the bone, joint, tendon, and ligament deformities as well as the joint deviation should be correspondingly corrected.

7.1.6.3 Typical Cases

1. Case I. Type IV duplication of the thumb. During the first phase, the neoplastic thumb is surgically removed; after surgery, there are lateriflection deformity of the thumb interphalangeal joint and thumb web stenosis, so phase II repair is needed. During the phase II surgery, proximal phalanx osteotomy and correction, expansion of thumb web, repair and reconstruction (grafting and insertion reconstruction) of the thumb extensor tendons, and the repair and reconstruction of the metacarpophalangeal joint and interphalangeal joint accessory ligaments are performed (Fig. 7.38).
2. Case II. After the correction of type IV duplication of the thumb, the residual deformities include fineness of residual thumb, lateriflection of metacarpophalangeal joint, and hypertrophy at the distal end of the metacarpal bone and thumb web stenosis, and phase II repair is needed. During the phase II surgery, the neoplastic part of the first metacarpal bone head of the left thumb is resected, the radial metacarpophalangeal joint accessory ligaments are shortened, the ulnar accessory ligaments of the metacarpophalangeal joint are prolonged, the tendon of the reconstructed thumb is grafted and repaired, the thumb extensor tendon of the neoplastic thumb is grafted, the radial accessory ligaments of the metacarpophalangeal joint and the thumb web are expanded, etc. (Fig. 7.39).
3. Case III. The residual deformities after the correction of lobster-tong-shaped thumb of type IV duplication of the thumb are manifested as lateriflection of thumb proximal phalanx toward the ulnar side and ulnar-ward angulation deformity of metacarpophalangeal joint, lateriflection of the thumb distal phalanx toward the radial side and radial-ward angulation deformity of the interphalangeal joint, and joint deformities, but no obvious deformity in phalange and metacarpal bone.

Surgical design: ① prolong the radial accessory ligaments of the thumb interphalangeal joint, and shorten the ulnar accessory ligaments; ②graft the insertion of thumb long extensor tendons, and correct the lateral deviation deformity of the interphalangeal joint; ③ shorten the radial accessory ligaments of the metacarpophalangeal joint, and correct the ulnar deviation deformity of thumb metacarpophalangeal joint; ④ correct the abnormal displacement of thumb extensor tendon

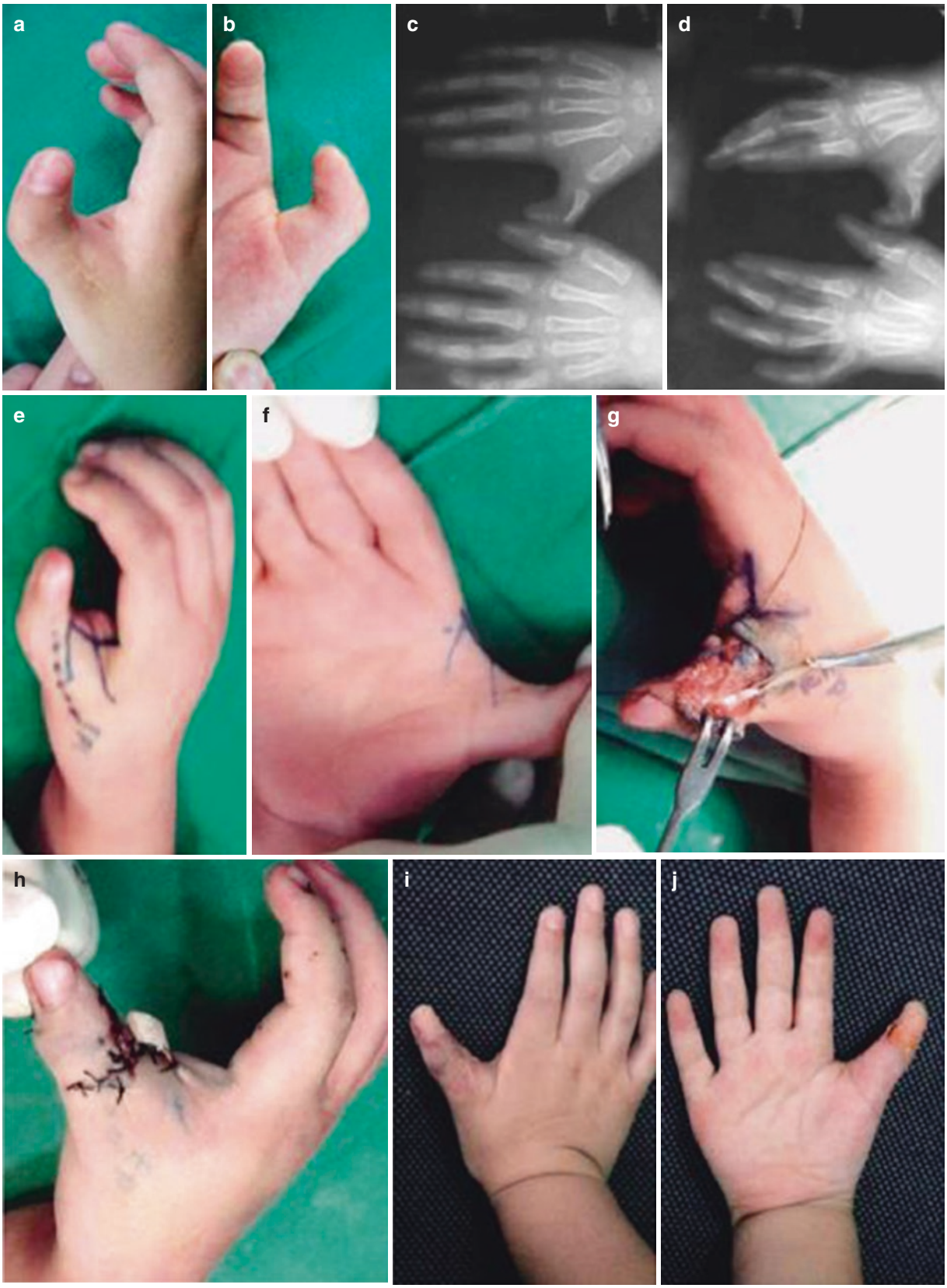

Fig. 7.38 Case one. (**a**, **b**) Hand appearance before operation. (**c**, **d**) X-ray findings before operation. (**e**, **f**) Design of skin incisions for thumb web expansion during the surgery. (**g**) Phalangeal osteotomy and correction and tendon repair and reconstruction. (**h–j**) Hand appearance after operation

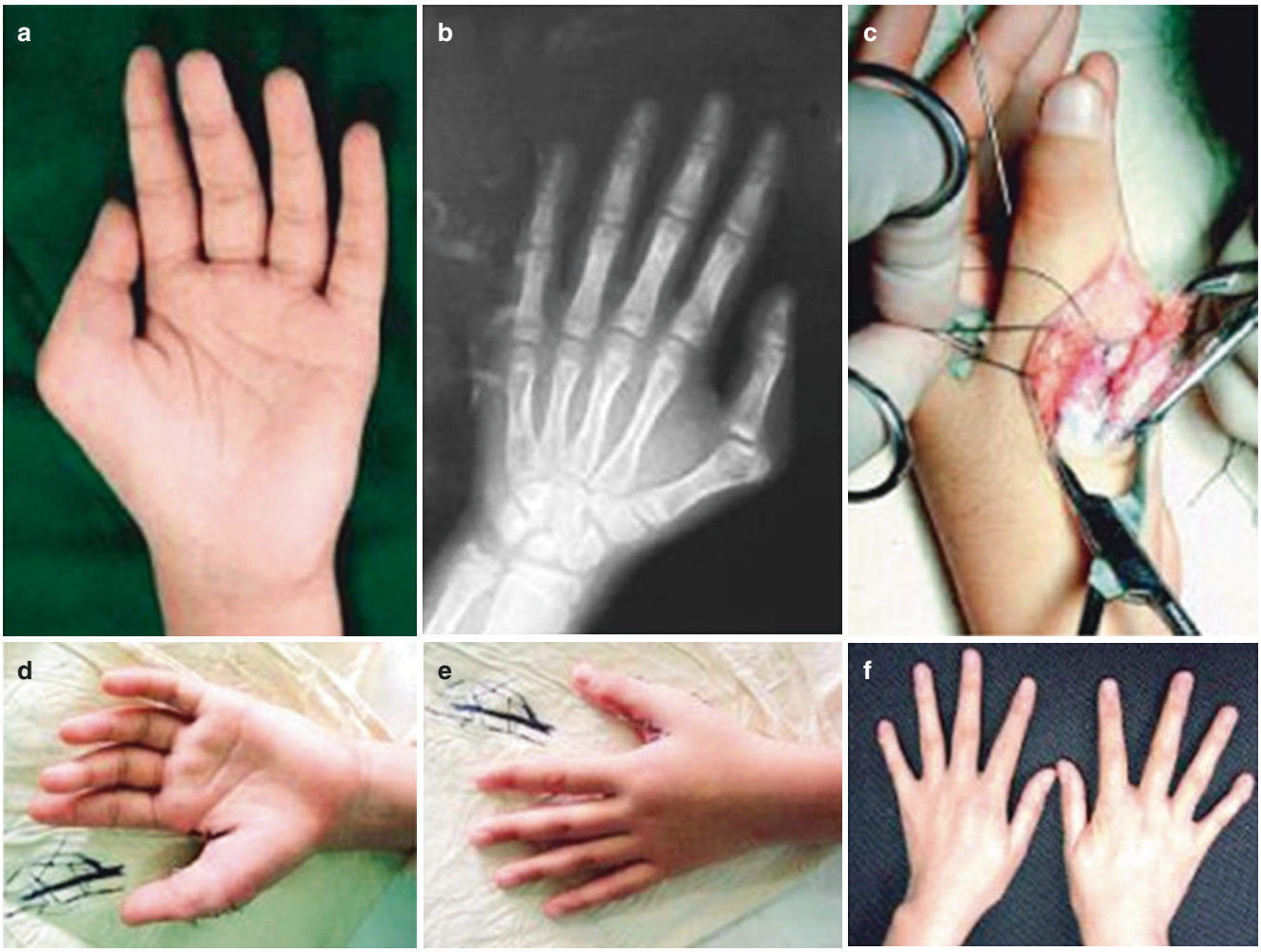

Fig. 7.39 Case two. (**a**, **b**) Residual deformities after correction of type IV duplication of the thumb. (**c**, **d**) Residual deformities of phase II surgery correction. (**e**) Immediate after operation. (**f**) At 2 months after operation

insertions; ⑤ graft the thumb extensor tendons of the neoplastic thumb, correct deformity of inclination toward the side of interphalangeal joint, and strengthen thumb extensor; and ⑥ make Z-plasty skin incisions at the radial side of the residual thumb, and make a skin incision at the ulnar side of the interphalangeal joint to expose the region from the metacarpophalangeal joint to the interphalangeal joint. The phalangeal and metacarpal osteotomy and correction are not adopted, and serious lateral deviation deformities of interphalangeal joint and metacarpophalangeal joint are also effectively corrected (Fig. 7.40).

7.1.7 Postoperative Effect Evaluation

Corrective surgery of duplication of the thumb is a typical plastic surgery, including resection of neoplastic thumb and aesthetical reshaping of residual thumb. After aesthetical reshaping, especially after osteotomic reshaping of deformed phalange and correction of joint lateral deviation, usually there is a decrease in the range of joint activities and even joint rigidity; therefore, the treatment of duplication of the thumb should focus not only on morphological correction but also on functional repair and reconstruction. Therefore, postoperative morphological and functional assessment for the duplication of the thumb is a yardstick of surgical effectiveness. The assessment of postoperative effects of corrective surgery of duplication of the thumb as designed by the author is shown in Table 7.5.

7.1.8 Duplication of the Thumb and Syndrome

Clinically, duplication of the thumb is one of many syndromes. The author once treated one rare patient: a girl that is 6.5 years old, with congenital bilateral facial nerve

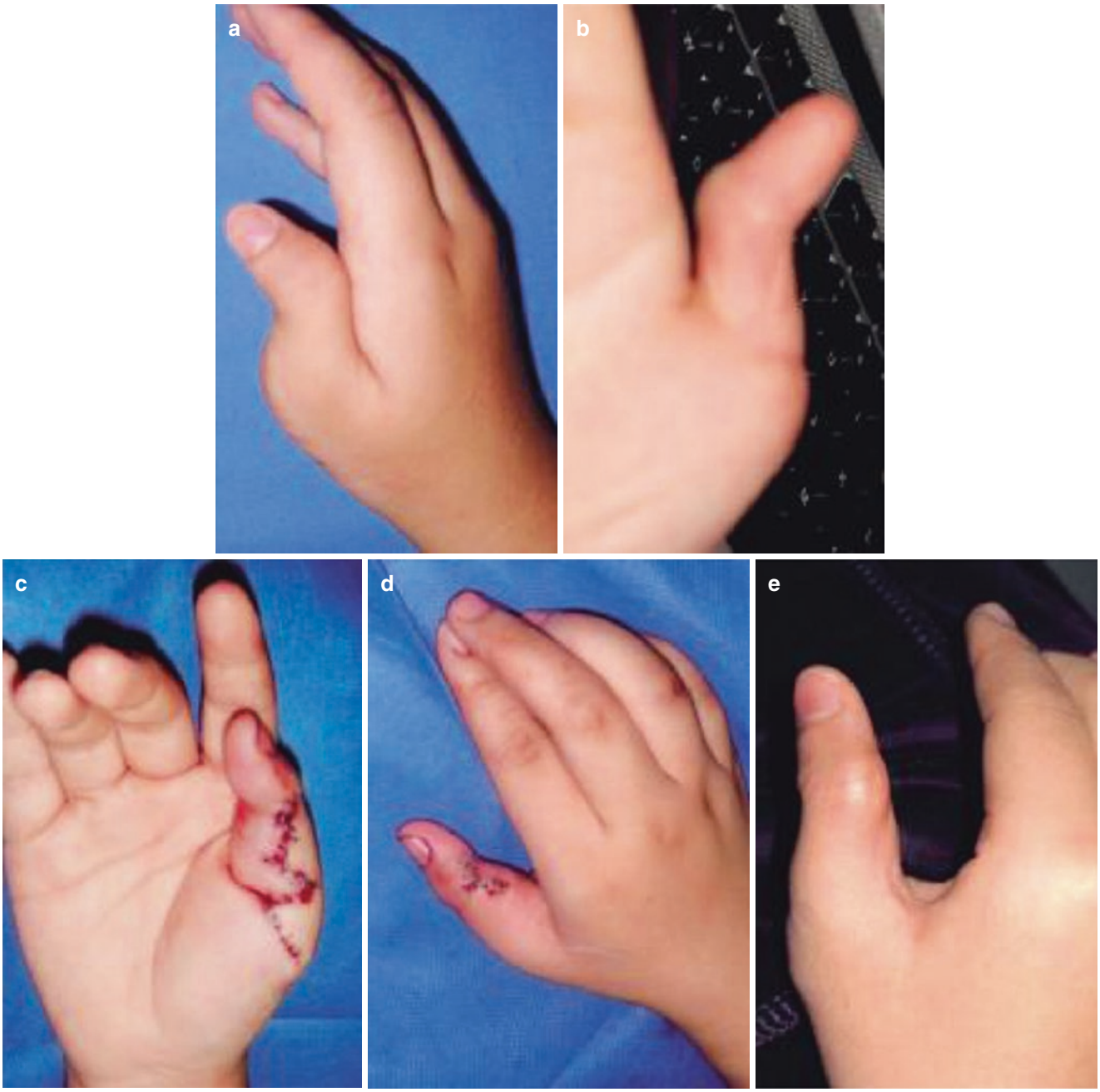

Fig. 7.40 Case three. (**a**, **b**) Residual deformities after correction of lobster-tong-shaped thumb of type IV duplication of the thumb. (**c**–**e**) Post phase-II surgical correction

paralysis, convergent strabismus of both eyes (treatment has been given), eyeballs set in the middle, dysraphism of eyelids, paralysis of bilateral frontal muscles, inability to bend and descend the brow, inability to whistle, cauliflower-shaped deformity of the bilateral external ear auricles, and deformity of duplicated thumb of the right hand (Fig. 7.41). Then, she visited our hospital for resection of radial thumb, and after operation, the residual thumb had adduction and flexion deformities, and the right foot had equinovarus.

7.2 Triphalangeal Thumb

Bin Wang, Yongkang Cai, and Hongxing Zhang

Triphalangeal thumb can either be an independent deformity or be associated with systemic deformities (e.g., Holt-Oram syndrome, Fanconi syndrome). It can occur simultaneously with duplicated thumb (Wassel type VII) and take various forms [11, 12]. The triphalangeal thumb is often accompanied by other congenital abnormalities, such as polydactylism, thumb hypoplasia, or central ray absence [13, 14].

7.2.1 Clinical Types

The triphalangeal thumb can be divided into two different types: the firs type, the thumb has normal shape and extra phalanges with varying sizes, and the extra phalanges can take the shape of triangle, trapezoid, or rectangle [15, 16]; the second type, phalanges develop fully on the finger plane; this type is considered to be five-fingered deformity and can also be the index finger polydactylism in thumb absence [17].

Like most classifications, the manifestations of the above two types of triphalangeal thumb have their own and unique characteristics. In addition, the types of triphalangeal thumb can vary according to the differences in genetic generations.

Both types of triphalangeal thumb fall into the category of autosomal dominant inheritance with obvious variability and high penetrance [15, 16, 18, 19]. Given that different spectrums of variation will have different phenotypes, the parents need to receive proper genetic consultations. The triphalangeal thumb can also be sporadic.

Table 7.5 Assessment of postoperative effects of corrective surgery of duplication of the thumb

Effects	Manifestation
Excellent+	There is no residual morphological and functional defect in the deformed thumb, and its shape is similar to that of normal thumb
Excellent	There is no residual morphological and functional defect in the deformed thumb, but it suffers dysplasia unlike normal thumb
Good	Mild deformities in thumb appearance or finger nails, joint angulation <15°, and the range of joint activities <15°
Fair	Obvious morphological or functional defects in thumb or finger nails, joint angulation >15°, and the range of joint activities >15°
Poor	Serious thumb deformities, serious functional disorder, obvious joint angulating deformity, and joint rigidity

7.2.2 Treatment

7.2.2.1 Selection of Methods and Timing

The treatment of triphalangeal thumb varies with individual [20], and the method depends on the type of triphalangeal thumb and whether any other congenital abnormality is suffered, such as duplication of the thumb. Multiple factors need to be considered in the treatment of triphalangeal thumb, including the length abnormalities and angulation of the fingers, extra interphalangeal joints, fingerweb between the thumb and index finger, and thumb palm-opposing functions.

The selection of treatment timing also depends on whether any other congenital abnormality is suffered. The palm-opposition and pinching functions of thumb develop well when one becomes 12–18 months old, and the thumb

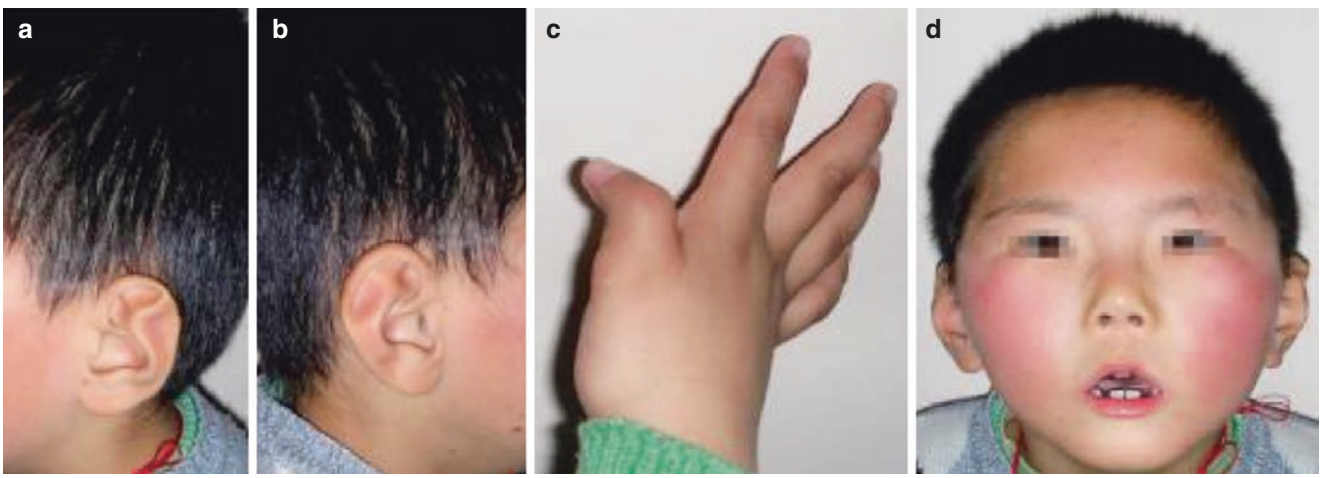

Fig. 7.41 Syndromic duplication of the thumb, bilateral syndromic paralysis, and convergent strabismus of both eyes. (**a**, **b**) Auricle appearance. (**c**) Hand appearance. (**d**) Facial appearance

should be reconstructed before this age so that it can serve such functions; slight length abnormality or angulation treatment can be delayed to a later period of time, and it is generally suggested that it be completed before the patient's school age.

7.2.2.2 Treatment of Length Abnormality and Angulation

Extra phalanges take different shapes and have different development processes, and their length and shape depend on the length and arrangement of the thumb. There are some differences in the growth speed of the extra phalanges, and their growth potential should be inspected and evaluated during preliminary treatment. In the meantime, the motions of each interphalangeal joint need to be evaluated on a regular basis, which is important to reconstruction. The extra phalanges may not develop eventually and not grow basically, so treatment is unnecessary; on the contrary, the continuous growth or asymmetric growth will lead to the excessive length of the thumb or the progressive angulation, and angulation often occurs at ulnar direction of the top.

As small wedge-shaped middle phalanx will lead to progressive deviation, it is necessary to perform ligament reconstruction [20, 21], and it is recommended that the surgery be performed at the age of around 1. Choose the dorsal longitudinal incisions or the lateral incisions that pass through the extensor structures, separate the extra phalanges, and record the size. Elevate the periosteal sleeve including the accessory ligaments, remove the extra phalanges, and reattach the accessory ligaments. Appose the distal and proximal phalanxes, and use Kirschner silk for longitudinal fixation and braking until the ligament heals. Use absorbable sutures to close the incisions and fix them with long-arm thumb spica casts; 6 weeks after operation, remove the casts and Kirschner silk, and fabricate and use short-arm thumb spica splints; they are removable during the treatment and mild activities, and the splint can be removed at 6 weeks after the operation. During the first 3 months, the pinching and holding should be reduced to prevent the radial deviation at the interphalangeal joint. The seemingly compatible surgical method will lead to multiple problems, including joint instability, joint rigidity, and insufficient range of motion. In fact, the author tries to avoid simple resection and chooses to retain one piece of middle phalanx, and its effects on the thumb function can be neglected. If this phalange affects the arrangement and functions of thumb, then joint reduction should be performed.

7.2.2.3 Treatment of Supernumerary Interphalangeal Joints

Supernumerary interphalangeal joints have different ranges of motion, and such joints have a slight influence on hand functions. Massive wedge-shaped supernumerary phalange will result in excessively long fingers with or without deviation. Generally, phalangectomy is not performed as it will concomitantly cause joint instability. The usual practice is to fuse the abnormal phalange with the distal or proximal phalanx, and at the same time, shorten the bones to remove the supernumerary joint, rearrange the thumb, and correct the length; retain the joint with the largest range of motion; and fuse the joint with the smallest range of motion.

Choose the dorsal longitudinal incisions that pass through the extensor structures, and separate the supernumerary phalanges and adjacent joint. Perform joint incision at the joint with the smallest range of motion, and avoid performing it on the joint with the largest range of motion. Resect this phalange from the two ends of the joint to shorten and rearrange the thumb. Asymmetric ostectomy can correct angulation deformity. After ostectomy, it is necessary to appose the bone and have the Kirschner silk retrograde from the fingertip to the osteotomy site for internal fixation. Adjust the extensor structure, use the absorbable suture to close the skin incisions, and use the long-arm thumb for spica cast fixation. Five weeks after operation, remove the casts and Kirschner silk, and fabricate and use short-arm thumb spica splints, which are removable in functional exercise.

7.2.2.4 Treatment of Fingerweb Between Thumb and Index Finger

Stenotic fingerweb will impede thumb functions. Mild syndactylia of thumb and index finger can be corrected through deepening the fingerweb. Four-flap Z-plasty can be used to deepen the fingerweb and can produce more smooth appearance than the two-flap method [22, 23]. As for the mild fingerweb between the thumb and index finger, it is necessary to perform local rotation to advance the skin flap to serve the purpose of sufficient deepening. The treatment strategy of fingerweb is similar to the treatment method of the fingerweb stenosis between the thumb and index finger in case of thumb dysplasia.

The five-fingered hand deformity has no fingerweb, pollicization can provide the finger in the palm-opposition position and the fingerweb between the thumb and the index finger [22, 24], and the surgical method is similar to that for type V thumb dysplasia or thumb absence. Some scholars describe the phase II surgical reconstruction of fingers [25], the phase I surgery includes joint reduction,

and the phase II surgery includes rotary ostectomy in combination with the reconstruction of palm-opposition function. As for the dysplastic triphalangeal thumb, thumb resection is needed and then pollicization of index finger is performed.

7.2.2.5 Reconstruction of Thumb Palm-Opposition Functions

The fiver-fingered deformity affects hand functions due to the absence of finger in the palm-opposition position, and the reconstruction method of palm-opposition functions is shown in Chap. 5 "Congenital Thumb Dysplasia." In case of fingerweb stenosis between the thumb and index finger, it is necessary to perform fingerweb expanding surgery simultaneously. Joint reduction can be performed together with tendon grafting or regarded as phase II surgery.

7.2.2.6 Treatment of Concomitant Deformities

The treatment of triphalangeal thumb concomitantly with duplication of the thumb should follow the principle of thumb function reconstruction [7]. Usually, the dominating group should be retained, the nondominating should be reserved, and any important elements (e.g., accessory ligaments) should be retained. If the triphalangeal bone is retained, the treatment of supernumerary phalange just follows the abovementioned principle. The treatment of the triphalangeal part in the concomitant deformities can be given simultaneously with or later than treatment of duplication of the thumb.

As for hand deformities with both triphalangeal thumb and central ray absence, treatment should be given by following both the longitudinal ray principle and triphalangeal thumb reconstruction principle. The severity of longitudinal ray formation and the status of the fingerweb between the thumb and index finger are the key in treatment. As for complete syndactylia between the thumb and index finger, syndactylous fingers should be separated during the early stage. The skin at the longitudinal ray can be grafted to the connection of the thumb and index finger to recover the fingerweb (also called Snow-Littler skin flap) [26, 27].

7.3 Ulnar Hyperdactylism

Ulnar hyperdactylism, also called postaxial hyperdactylism, is rarer than radial hyperdactylism but is not as various as radial hyperdactylism [28, 29]. Among the 403 cases of hyperdactylism reported by Flat (1994) [12], 142 were ulnar hyperdactylism accounting for 35%. Ulnar hyperdactylism is relatively common in the African race, and some reports indicate that its incidence is 1/300 to 1/143 of infants. It can occur to one side or two sides, and bilateral deformity is common; in case of occurrence at one side, deformity at the left side is common, accompanied by or not accompanied by ulnar polydactyly (Fig. 7.42). It is generally autosomal dominant inheritance (Fig. 7.43), but the diseased shape is often various and can be manifested as multiple clinical types.

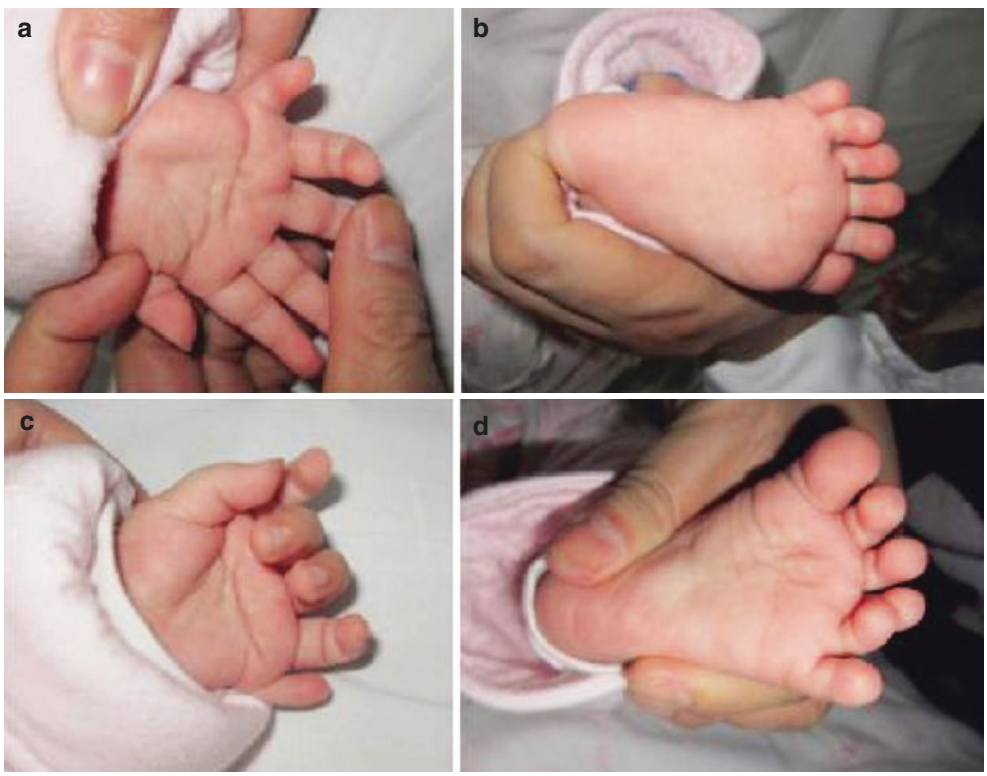

Fig. 7.42 Multiple ulnar polydactylia of four limbs

7.3.1 Clinical Manifestation and Classification

Temtamy and McKusick (1969) [30] divided the ulnar hyperdactylism into two types according to the development of hyperdactylism and the gene phenotype: type A, type of good development, and type B, dysplasia type or neoplastic finger type. Type A hyperdactylism often has a complete phalangeal and tendon system and even the completely or partially fused metacarpal bones. The ulnar hyperdactylism with entire ray of complete and independent metacarpal bones is relatively rare, and in most cases, the proximal phalanxes of dominating and accessory fingers form joints jointly with a thick fifth metacarpal bone head. Type B is sometimes only a tiny hyperdactylism with very incomplete development which is connected with a skin pedicle. Type B is commoner than type A, and reports indicated that the incidence of type A is only 0.014%.

Rayan and Frey (2001) [31] further classified the clinical types of ulnar hyperdactylism into five types:

1. Type I. It is the soft tissue tubercle (wart-shaped finger) without finger nail (Fig. 7.44).
2. Type II. It is the pedicled nonfunctional finger (lollipop-shaped finger); type IIA has narrow pedicle (the pedicle width is less than 3 mm) and type IIB has wide pedicle (the pedicle width is larger than 3 mm) (Fig. 7.45).
3. Type III. It is the functional finger with good appearance which forms a joint with fifth metacarpal bone (type IIIA) or become fused with it (type IIIB) (Fig. 7.46).
4. Type IV. It is the functional finger with good appearance, with independent sixth metacarpal bone (Fig. 7.47).
5. Type V. It is the compound hyperdactylism, such as ulnar hyperdactylism and syndactylia or three-little-fingered deformity (Fig. 7.48).

In the classification of Rayan-Frey [31], type I is the soft tissue hyperdactylism without tissues such as bones and tendons in the hyperdactylism; type II and type III are simple hyperdactylism, and bones and tendons are contained in hyperdactylism; type IV and V are compound hyperdactylism, which not only contains tissues such as bones and tendons but also contains twin metacarpal bones, etc.

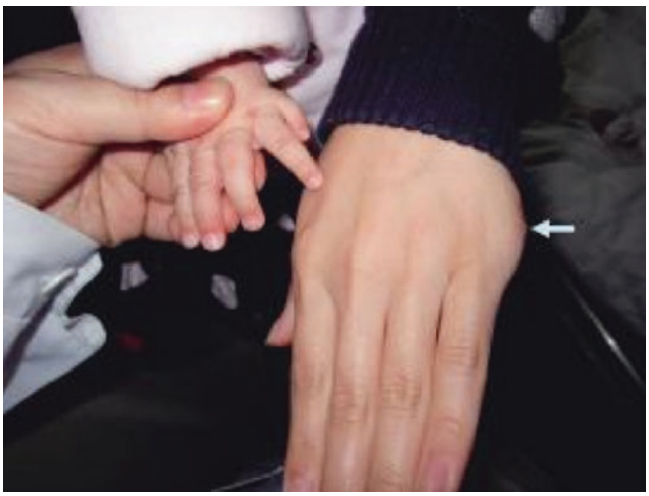

Fig. 7.43 Both the mother and the daughter suffer ulnar hyperdactylism (what is indicated by the arrow is the cicatrix residual from the resection of the mother's ulnar supernumerary finger)

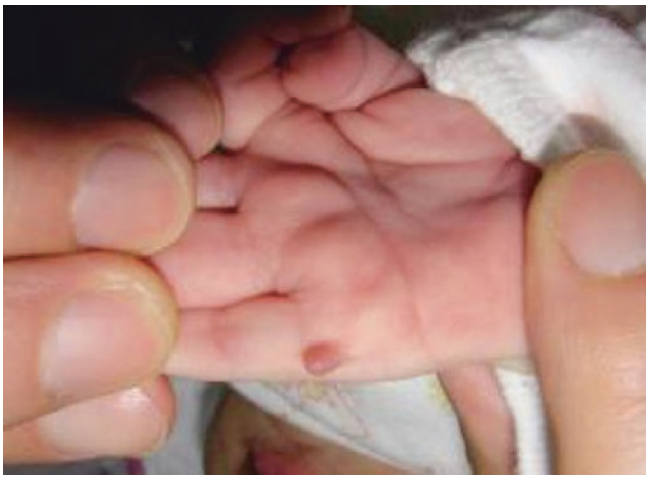

Fig. 7.44 Type I ulnar hyperdactylism

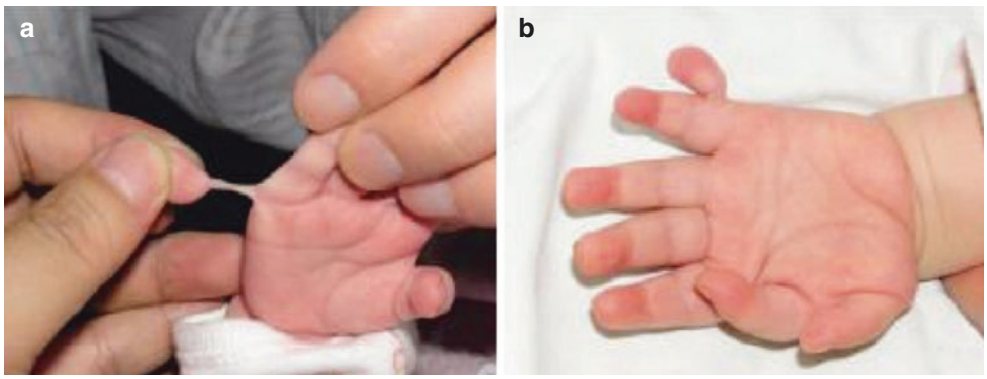

Fig. 7.45 Type II ulnar hyperdactylism. (**a**) Type IIA. (**b**) Type IIB

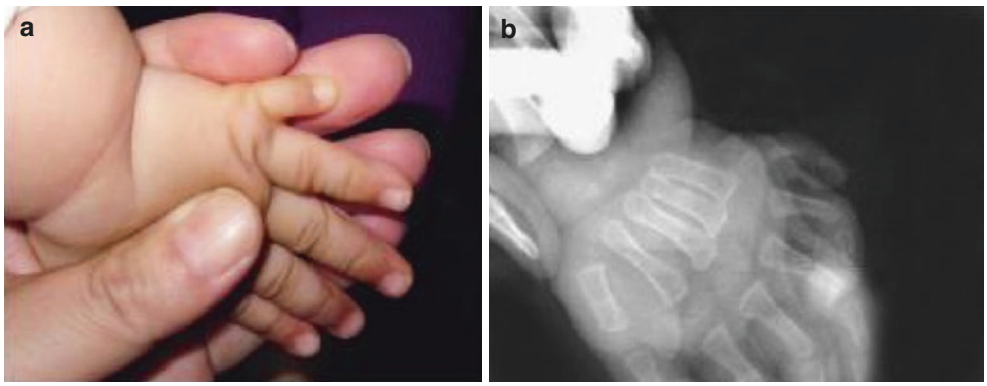

Fig. 7.46 Type IIIA ulnar hyperdactylism

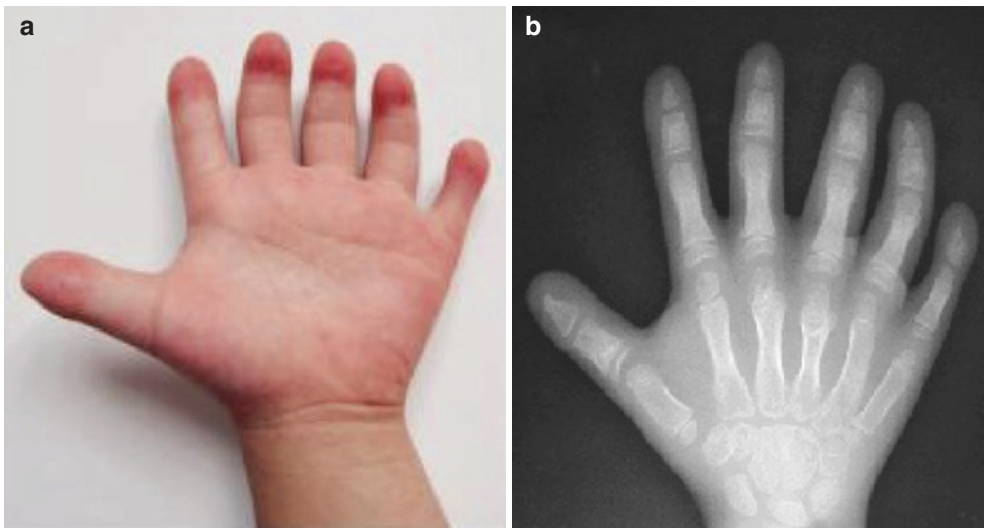

Fig. 7.47 Type IV ulnar hyperdactylism

Fig. 7.48 Type V ulnar hyperdactylism

Ulnar hyperdactylism may be accompanied by other deformities, among which syndactylia is relatively common, and it can also be accompanied by 13 and 18 Patau syndrome, Ellis-Van Creveld syndrome, triphalangeal thumb (Fig. 7.49), tibial defects, spinal column deformity, chapped lips, polycystic disease, exstrophy of the bladder, anal atresia, heart disease, feeblemindedness and dwarf, and careful identification is clinically needed.

7.3.2 Treatment

7.3.2.1 Neoplastic Finger Without Metacarpal Bone or Metacarpophalangeal Joint

As for the neoplastic finger without metacarpal bone or metacarpophalangeal joint, the method of adopting suture to ligate the pedicle can be adopted to allow it to become black, necrotic, and desquamation, but this method will result in sequelae, such as occurrence of head-shaped incisura or nerve tumor at the site of hyperdactylism, and a second surgery is needed. If the pedicle of the neoplastic finger is relatively wide, it is not suitable to adopt the ligation method because it may lead to incapability of autotomy or excessive bleeding.

7.3.2.2 Ulnar Simple Hyperdactylism

For the ulnar simple hyperdactylism, it is suitable to resect the relatively small duplicated thumb within 3–6 months. Compared with the resection of radial supernumerary finger, generally very complicated reconstructing surgery is not necessary for the retained finger. If the duplicated thumbs have a similar size, the doctor can wait until the patient is over 1 year old to observe which finger is the nonfunctional finger and decide which one needs to be resected. When the ulnar supernumerary finger is resected, as for the insertion of the hypothenar muscle (the little finger abductor) attached to the supernumerary finger, it should be detached during the surgery and then refixed at the ulnar side of the basal part of the proximal phalanx of the retained finger to facilitate the retention of the abduction function of the little finger; the tendon is disjuncted from the bifurcation of the supernumerary finger and then abandoned. In resection of the supernumerary finger, it can be used for the disengagement at the metacarpophalangeal joint; before the disengagement, the ulnar accessory ligaments should be detached and retained to repair the metacarpophalangeal joint capsule of the little finger (Fig. 7.50). As for the patients with hyperdactylism with simple syndactylia, the usual practice is to make a Z-plasty incision at the ulnar margin of the hand to expose and resect the supernumerary secondary finger and then perform plastic surgery on local skin. If any soft issue is reconstructed, after the operation, it is necessary to use gypsum for external fixation of the diseased hand for 3 weeks.

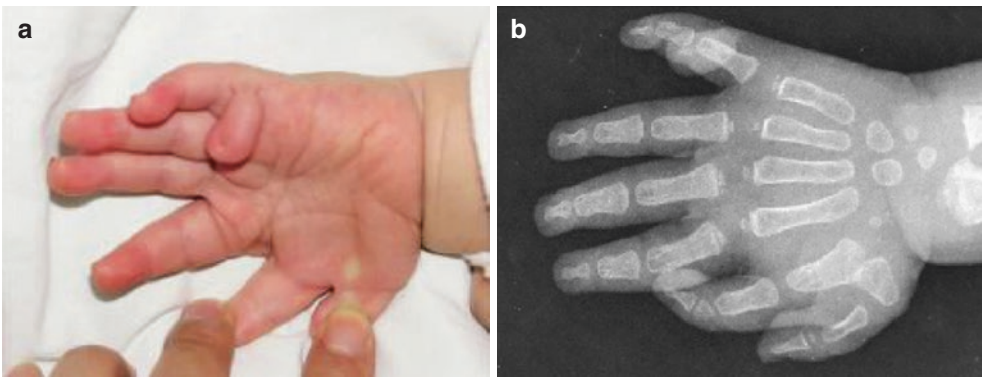

Fig. 7.49 Ulnar hyperdactylism with triphalangeal duplication of the thumb

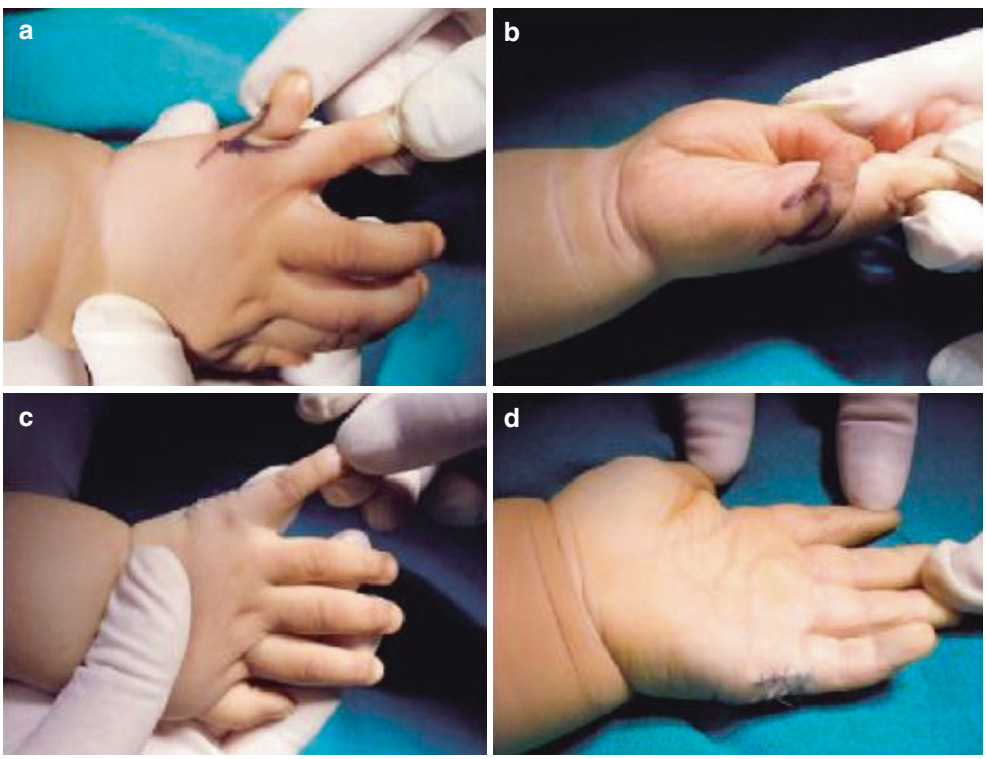

Fig. 7.50 Surgical correction of ulnar simple hyperdactylism. (**a**, **b**) Surgical design. (**c**, **d**) Hand appearance immediate after operation

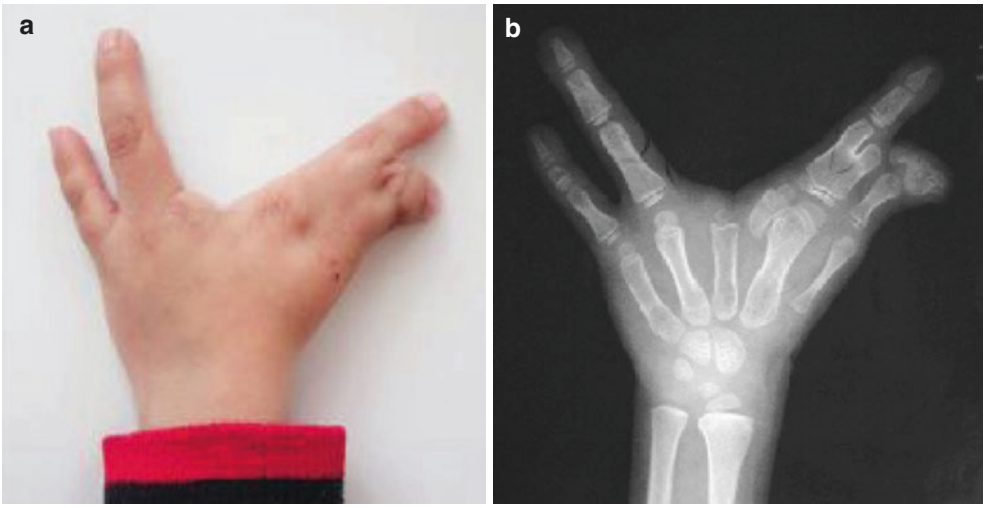

Fig. 7.51 Central hyperdactylism accompanied by syndactylia and cleft hand

7.3.2.3 Ulnar Compound Hyperdactylism

For the patients with compound hyperdactylism, the surgical time can be postponed, but the surgery should be completed before the patient becomes 2 years old so that the parents can feel relieved as early as possible and the pediatric patient can be free from the psychological disorder. If the ulnar supernumerary finger has complete or partially fused metacarpal bone, in addition to the resection of the supernumerary finger, it is also necessary to perform total or partial section of the twin metacarpal bone, and how much is resected depends on the requirements on morphological and functional reconstruction of the affected hand. During the resection of supernumerary finger, it is sometimes necessary to correct the joint and bone deformities, and the repair and skin plastic repair of joints and ligaments should be considered during the surgery.

7.4 Central Hyperdactylism

Bin Wang and Yi Cao

Central polydactyly refers to the deformity of the duplication that mainly involves the index finger, middle finger, and ring finger; the involvement of ring finger is the commonest, followed by the middle finger, and then the index finger. Among the 403 cases of hyperdactylism reported by Flat (1994) [12], the central hyperdactylism accounts for 19%, and its incidence is lower than that of radial and ulnar hyperdactylism. Ogino et al. [32, 33] found that central hyperdactylism, syndactylia, and cleft hand had similar morphological manifestations during the early stage of embryonic development, and they reported that three patients in the same family had five sites of central hyperdactylism, syndactylia, and cleft hand and considered that the three deformities could be included into the same category. We also observe that the patients with central hyperdactylism accompanied by syndactylia and cleft hand often present autosomal dominant inheritance (Fig. 7.51).

7.4.1 Clinical Manifestation

Clinically, the central hyperdactylism can be expressed as single symptom and can also be the concomitant symptom of certain syndrome, such as Grebe chondrodysplasia syndrome. As the central hyperdactylism can be covered by the accompanying syndactylia, careful examination is clinically needed, and imaging measures are relied on to assist in the diagnosis. Muragaki (1996) [34] reported that the patients with polysyndactyly of the ring finger often have a family history accompanied by the variation in HOXD13 gene on the chromosome 2.

Central polydactyly takes various forms and occurs at bilateral sides. It may be accompanied by central polydactyly (Fig. 7.52) and cleft foot deformity (Fig. 7.53).

Steling and Turek divided central polydactyly into three types: type I, only a mass of supernumerary soft tissues, without bone, articular cartilage, or tendon; type II, duplication or partial duplication of the fingers, with normal finger structures, including furcal metacarpal bone or phalange, connected with joints, and this type can be also subclassified into type II a (non-syndactylous) and type II b (syndactylous) (Fig. 7.54); and type III, very rare, the supernumerary finger with complete structures of metacarpal bone, phalange, and various soft tissues (Fig 7.55).

7.4.2 Treatment

The surgeon should decide the surgical mode according to the finger conditions, the syndactylia degree, and the functions of the central syndactylous finger, with the functional and mechanical stability as the guideline [35, 36].

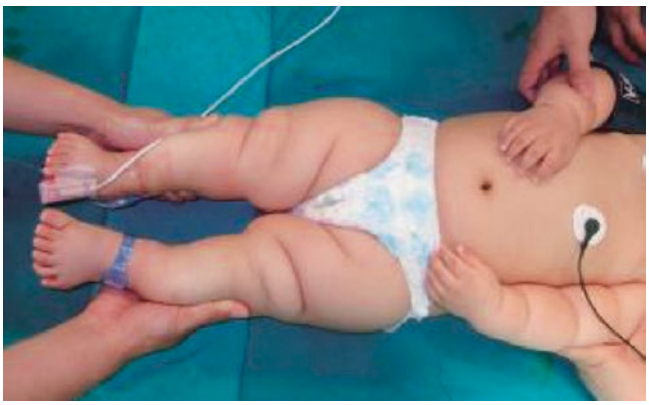

Fig. 7.52 Central polydactyly with central hyperdactylia

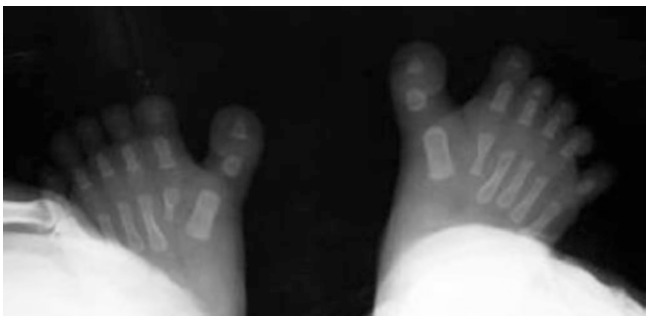

Fig. 7.53 Central polydactyly with cleft foot deformity

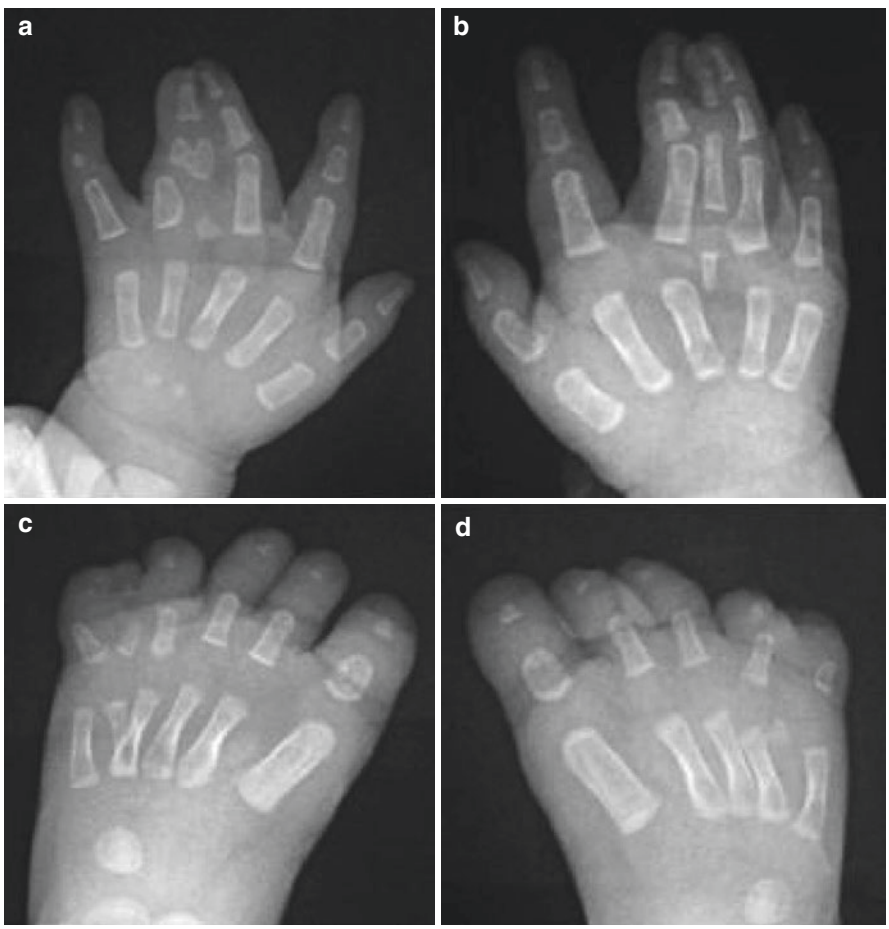

Fig. 7.54 Type II central polydactyly (hyperdactylia). (**a**, **b**) Hand X-ray findings. (**c**, **d**) Foot X-ray findings

1. Type I central hyperdactylism. It only has supernumerary soft tissues without bone or joint; therefore, resection alone is enough.
2. Type II central hyperdactylism. The supernumerary finger is often overlapped with the adjacent finger, and the finger-separating dermoplasty can be performed after the supernumerary finger is resected.

   At the proximal basal part of the syndactylous finger, design one isosceles triangular flap at the volar side and dorsal side, respectively, and on the skin with the finger-web, make two Z-plasty incisions at the distal end of triangular flaps along the volar side and dorsal side, whose directions are opposite, similar to the surgical incisions in finger-separating surgery for syndactylia. Incise the skin and subcutaneous tissues according to the surgical design until the fundus is reached; at the same time, resect the supernumerary finger and trim the supernumerary subcutaneous tissues. Let the triangular flaps intersect at the

basal part of the finger and suture them to form the new fingerweb, suture the flaps at the lateral side of all fingers, and obtain the intermediate split thickness skin graft to cover the residual wounds.

3. Type III central hyperdactylism. As for the fingers with complete structures of metacarpal bone, phalange, and various soft tissue, it is necessary to resect the entire digital ray including the metacarpal bone and then draw the adjacent two fingers close to each other. During the surgery, not only should attention be paid to the reconstruction of fingerweb, but attention should also be paid to the reconstruction of transverse ligaments of head of metacarpal bone after the supernumerary metacarpal bone is resected (Fig. 7.56).

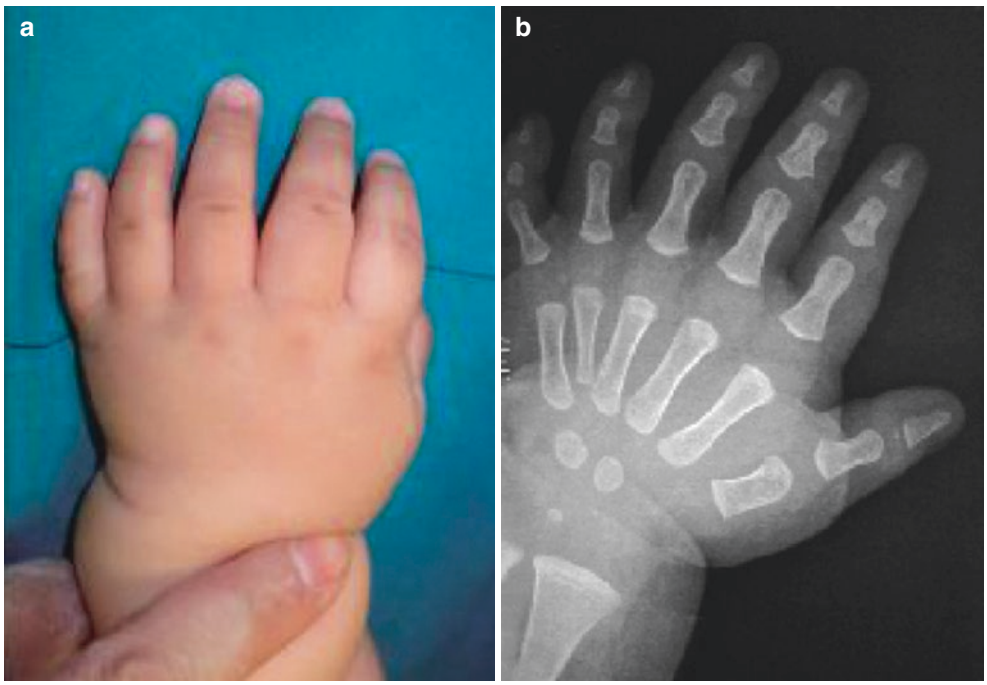

Fig. 7.55 Type III central hyperdactylism

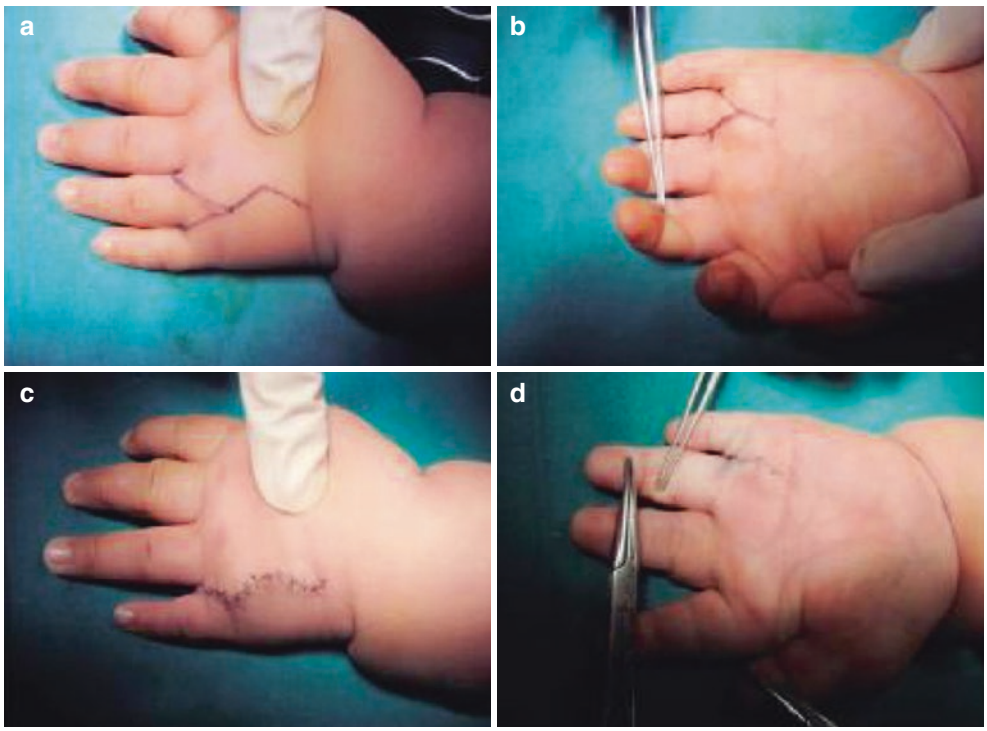

Fig. 7.56 Correction of central polydactyly. (**a**, **b**) Surgical design. (**c**, **d**) Hand appearance immediate after operation

4. Precautions
   (a) In resection of supernumerary finger, avoid damaging the adjacent digital nerve vascular bundles to prevent the blood supply of the retained finger from being affected.
   (b) The syndactylous should be completely separated until the fundus of the fingerweb is reached.
   (c) It is necessary to trim the wide articular surface; in case of finger deviation, osteotomy is necessary for correction.
   (d) It is necessary to centralize or rebalance the extensors and flexors and reconstruct or retain the accessory ligaments.
   (e) It is necessary to reconstruct the transverse ligament of head of metacarpal bone, and attention should be paid to fingerweb gap.

As the central hyperdactylism is often accompanied by syndactylia as well as the variation in vessels and nerves, during resection of supernumerary finger, avoid damage to the blood supply and innervation of the retained finger to prevent necrosis of the retained finger. As for the resection of the central supernumerary finger, some adopt the method of resecting it in two separate surgeries: in the first surgery, the supernumerary finger is resected when the pediatric patient is 6 months old; as for the second surgery, perform osteotomy for correction as well as repair and reconstruction of accessory ligaments when the pediatric patient is older. If the surgeon has good microsurgical skills and has sufficient understanding on the finger blood supply before the operation, the purpose of resecting supernumerary finger and correcting bone, joint, ligament, and tendon deformities can be served just through one surgery.

Usually, supernumerary finger-separating surgery is difficult, and separation of syndactylous fingers and resection of the hidden syndactylous finger may lead to incomplete resection of supernumerary phalange, resulting in instability of finger joints and movement disorder, so, for the structurally complex polysyndactyly, detailed surgical regimens should be made before the operation so that the functions of the affected hand will not be damaged.

7.5 Mirror Hand

Wei Wang and Bin Wang

Mirror hand or mirror hand polydactyly refers to the twin deformities in most structures of the hand, including the neoplastic, syndactylous, and digital flexion deformities of more than three three-segmented fingers and metacarpal bones, accompanied by dual ulnar deformities and ulnar dysplasia, and it is a congenital deformity with duplication of the fingers in hands, like a mirror image, so the name is given. This deformity, also called single-arm dual ulnar deformity or dual ulnar deformity, is a rare kind of deformity in the twin deformities of the upper limb.

7.5.1 Classification

Mirror hand is a rare kind of congenital hand deformity, characterized by symmetric duplication with the limb midline as the axis. The typical mirror hand is manifested as symmetric arrangement of three dactylopodites that represent the middle finger, ring finger, and little finger at the two sides of the central finger. In spite of the existence of multiple fingers, the thumb is absent; there is no radial bone in the forearm with two ulnae, and the ulnae support the ulnar carpal components arranged like a mirror. However, this deformity takes many forms, making its classification (Table 7.6) and treatment more complicated. Very rare polycheiria also falls into the category of mirror hand deformity [37, 38].

Barton et al. summarized the abnormal manifestations: usually the preaxial ulna is short and small, and the hand deviates toward the radial side; the soft tissue structures are abnormal and complicated, and unknown anatomical variation is common [38].

7.5.2 Etiologic Factor

The onset of the mirror hand is attributed to the duplication of the signal centers that regulate the radial-ulnar development. The zone of polarizing activity (ZPA) at the rear of the limb bud polarizes the limb into the radial-ulnar axis and dominates the preaxial and postaxial limb development. Migration of ZPA or its signaling molecules and hedgehog factor lead to the mirror image duplication of the ulnar side of the limb [39]. Currently, no gene abnormality directly correlated with the mirror hand has been found. The examination of mirror hand starts from the number and functions of the fingers; the range of motion of the wrist, forearm, and elbow are recorded; and wrist extension limitation secondary to the defect of wrist extensor tendon can be observed. As the two ulnae block the normal movement of the joint, the range of motion of the

Table 7.6 Classification of mirror hand

Type	Name	Clinical feature
1	Ulnar dimelia	Hyperdactylism, two ulnae Type A: each ulna develops well Type B: the preaxial ulna develops well
2	Middle type	Hyperdactylism, two ulnae, radial hyperdactylia
3	Middle type	Hyperdactylism, single ulna, single radial bone Type A: the radial bone develops well Type B: the radial bones suffers dysplasia
4	Syndromic type	Bilateral mirror foot, nasal defects Type A: Sandrow syndrome—two ulnae Type B: Martin syndrome—single ulnar-radial bone
5	Polycheiria	Including the mirror image of the whole hand, normal forearm

forearm and the elbow is also limited. The changes in range of motion of bending the elbow are correlated with the deformity degree of the elbow anatomic structure.

7.5.3 Clinical Manifestation

The feature of mirror hand deformity is the appearance of two ulnae in one forearm, manifested as radial hyperdactylism, with 7–8 three-segmented fingers or hypoplastic fingers, as if the fingers of two hands grew on one hand. Such deformed hands are often accompanied by deformities in the lower end of the humerus; radial absence; and two ulnae, one of which suffers dysplasia, or the coexistence of two ulnae, accompanied by one dysplastic radial bone. Mirror hand deformity is induced by dysplasia of embryonic upper limb bud.

The author treated eight pediatric patients with mirror hand deformity during the several decades' medical practice. A mirror hand may have 7–8 fingers, in which four three-segmented fingers at the ulnar side develop well, and the 3–4 three-segmented fingers at the radial side suffer dysplasia to varying extents. Its manifestation can also be that all the fingers suffer dysplasia or all fingers suffer syndactylia and flexion to varying extents. Due to no development of the thenar muscle, the patient only has flat palms, the wrist flexion strength is relatively weak, the hand is in the radial micro-flexion position, the wrist extending strength is also weak, the forearm pronation and supination are limited, the elbow joint often suffers flexion and extension function insufficiency due to the deformity of two ulnae, the radial forearm wrist extensor is absent, the biceps muscle of arm suffers dysplasia, and the ipsilateral shoulder joint, when in the abduction position, is apt to suffer shoulder joint incomplete dislocation; the patient can concomitantly suffer from the mirror foot like deformity in the lower limb. These symptoms are reported in author's cases and literature (Fig. 7.57) [40–42].

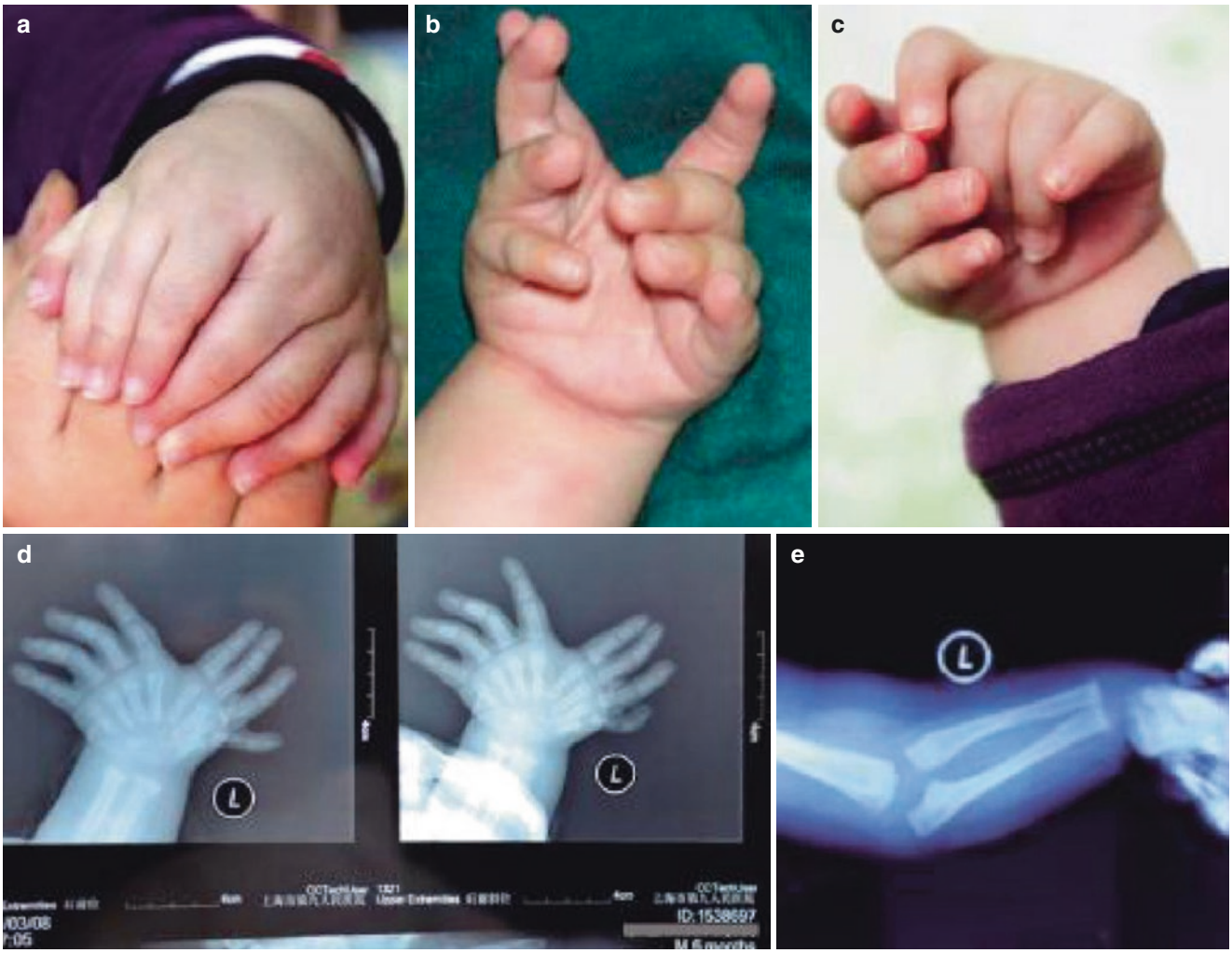

Fig. 7.57 A male infantile patient, with mirror hand deformity in the left hand and deformity of two ulnae in the forearm, visited the doctor at the age of 3 months and received surgical correction at the age of 6 months. (**a**–**c**) Preoperative hand appearance. (**d**) X-ray indicates the presence of four-fingered triphalangeal hyperdactylism at the radial side of the left hand, concomitantly with the neoplasm of four metacarpal bones. (**e**) X-ray indicates the deformity of two ulnae

In 1966, the author once admitted one 14-year-old girl with eight fingers in the right hand, the syndactylia between the seventh and eighth fingers, six fingers in the left hand, concomitantly with syndactylia and finger flexion deformity and the deformity of polydactyly in 2 ft. From the childhood, the pediatric patient always put two hands into her pockets. After admission, the patient received surgical treatment (Fig. 7.58). Such deformities are called by Laurin-Sandrow syndrome (LSS) in the literature, manifested as deformity of two ulnae and deformity of two fibulae, mirror hand and mirror foot, and radial and tibial absence or dysplasia, concomitantly with syndactylia and brachydactylia [43].

Mirror hand deformity is relatively rare upper limb deformity, and according to the description of Upton J. (1990), a total of 60 cases were reported in the historical literature [44].

The author includes the hyperdactylism with seven, eight, or more fingers below the forearm into this type of deformity; the number of metacarpal bones may be seven or eight8, the forearm is often short and small, the ulnae are in the twin status, and the thumb is absent. Most of the mirror hands described in the literature are unilateral, and six of the eight cases of mirror hands of the author are bilateral. Among them, one was a 12-year-old male with both hands being mirror hands and both feet being mirror feet, but he did not receive any treatment. The shape of mirror hand deformities is various. Bhaskaranand (2003) from India reported one rare case of mirror hand deformity. The pediatric patient was a 3.5-year-old boy with hyperdactylism in the left hand and mirror hand; his fourth finger at the radial side had two segments and the remaining fingers had three segments. X-ray film indicated that his forearm has two dysplastic radial bones which are almost fused as well as one ulnar bone, the shoulder joint was normal, the elbow joint became flexed by 40° with fixation deformity, and the right upper limb was normal [45].

Mirror hand deformity can also be manifested as six-fingered hand deformity, and the shape is generally the same as supernumerary finger, so it can be misdiagnosed as hyperdactylism. The author once treated one case of a 9-month-old boy, one of his hands suffers six-fingered incomplete syndactylia, while the other one suffers seven-fingered incomplete syndactylia; the two hands were obviously

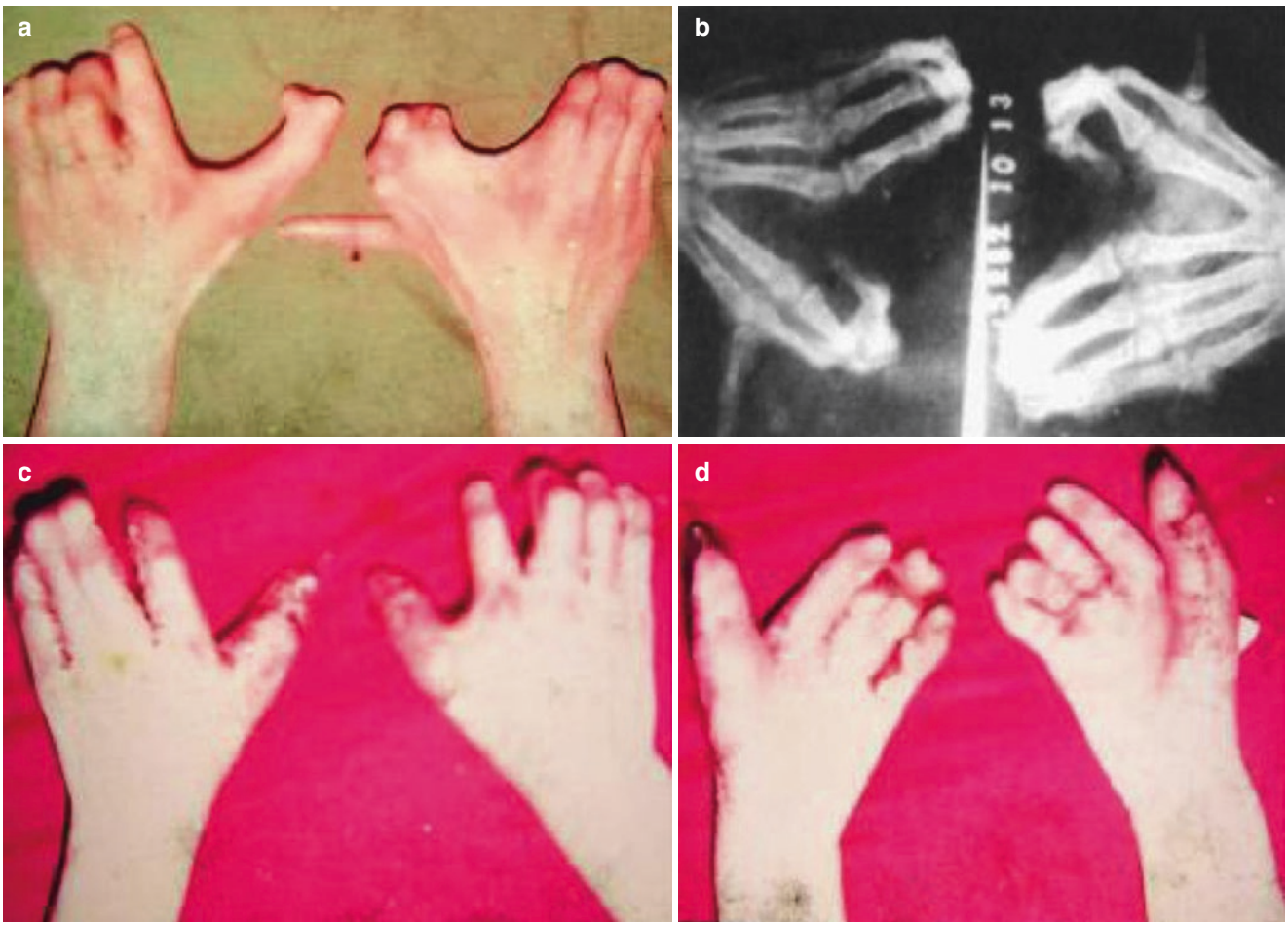

Fig. 7.58 Before and after surgery for bilateral mirror hands. (**a**) Hand appearance before operation. (**b**) X-ray findings before operation. (**c**, **d**) After the first surgery

separated in the middle; the two sides were symmetric; the two feet presented duplication and neoplasm; and he had two mirror hands and two mirror feet (Fig. 7.59).

7.5.4 Treatment

The treatment of mirror hand often needs individualized design according to the anatomic structures of the deformed hand.

As for the selection of surgical phase, the author suggests that phase I surgery be performed when the palm-opposing function of the thumb begins to develop; in other words, the phase I surgery should be performed when the infant is 6 months old. The surgical contents include resection of supernumerary finger, morphological and functional reconstruction of the thumb, reconstruction of the first fingerweb, or simultaneous reconstruction of the wrist extending function at the carpal part. In addition to these, palmar and hand dorsal skin plastic surgery can be performed simultaneously,

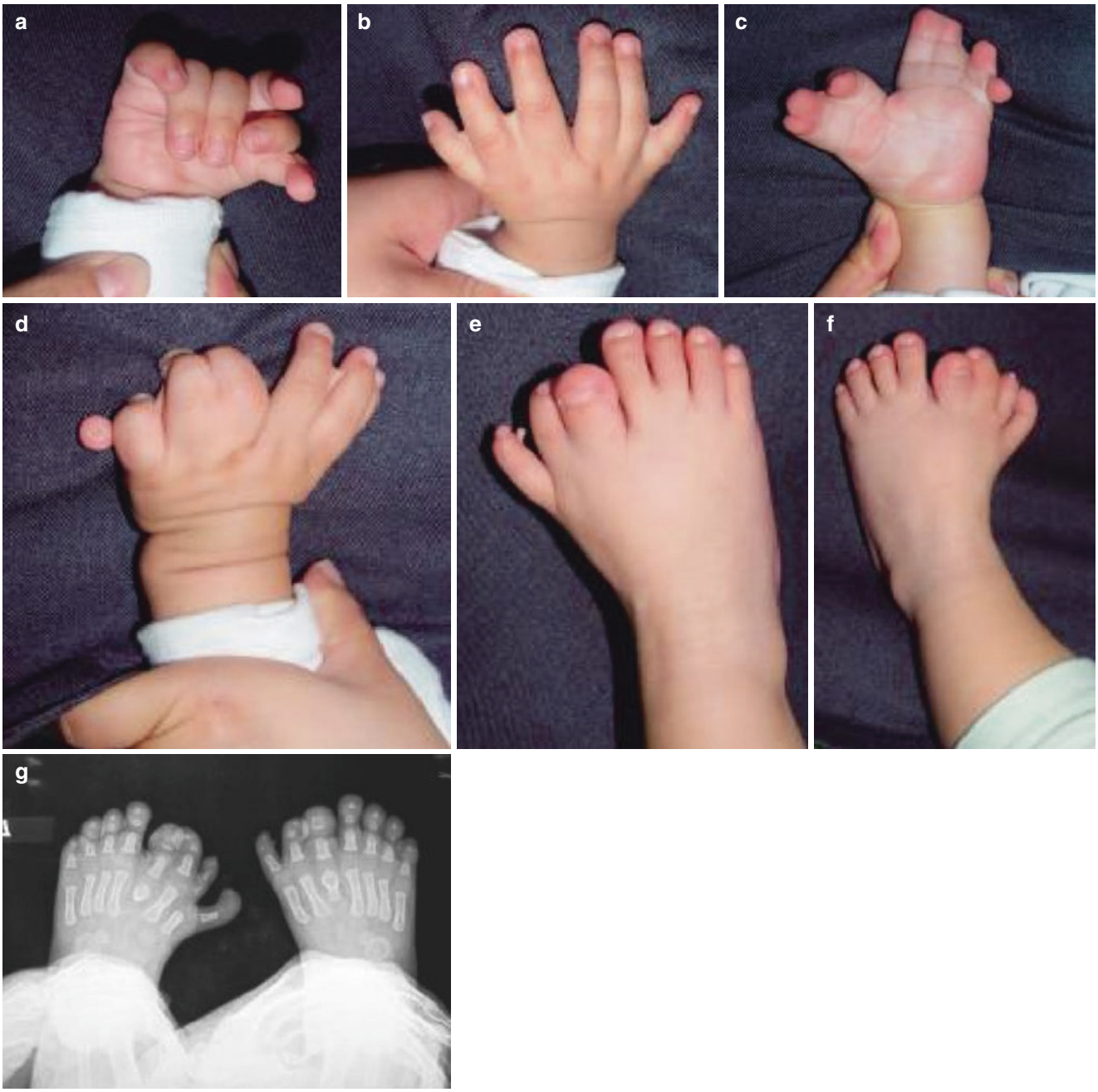

Fig. 7.59 Bilateral mirror hands and bilateral mirror feet. (**a**–**d**) Hand appearance. (**e**, **f**) Foot appearance. (**g**) Foot X-ray findings

the tendons and hand internal muscles of fingers can be grafted to repair and reconstruct the dynamic functions of the reconstructed thumb, and the purpose is to make sure that the shape and functions are close to normal after the surgery on mirror hand. The repair of relevant wrist extension, forearm pronation and supination, and elbow joint functional disorder is to be finished at the late stage. The corrective surgery on the supernumerary finger with syndactylous finger can be performed in different stages: thumb reconstruction, supernumerary finger resection, and separation of some syndactylia deformities; the corrective surgery of bent fingers can first be performed, and then 2–3 months later, correction of the syndactylia of other fingers can be performed. During the phase II surgery, dynamical reconstruction of the radial wrist extensor and the pronation functional reconstruction of the forearm can be performed. During the phase III surgery, functional reconstruction of the elbow joints can be performed.

The treatment method is mainly resection of supernumerary finger, and one finger of them is utilized for thumb reconstruction. Selective resection of supernumerary finger and thumb reconstruction is the key to surgery. Thumb web must be reconstructed during the surgery, and the tendons can be grafted to increase the range of motion of the retained thumb. The thumb reconstruction includes the utilization of supernumerary fingers for pollicization. Generally, the fingers with the optimal range of motion at the radial side and the best appearance are selected for pollicization, the relatively rigid fingers are removed, and the skin of the removed fingers can be used to expand thumb web. According to the author's years of mirror hand treatment experience as well as the corresponding literature, this is an effective treatment method (Fig. 7.60) [46, 47].

Application of Buck-Gramcko technology for the reconstruction of the first carpometacarpal joint still remains the key step. The author has applied Buck-Gramcko technology to make multiple modifications and highlighted the reconstruction of the metacarpophalangeal joint of the finger to be pollicized into carpometacarpal joint, namely, reconstruction of the head of metacarpal bone of the finger to be pollicized into the trapezium. Simply put, this is to perform osteotomy below the head of metacarpal bone on the fingers to be pollicized, rotate the head of metacarpal bone to the dorsal side, suture the broken end of the metacarpal bone and the dorsal fundus of the proximal phalanx, correct the flaccid volar plate, shorten the joint accessory ligaments, and form the stable first carpometacarpal joint. During the surgery, adjust the reconstructed thumb in the palm-opposition position (e.g., abducting the reconstructed thumb by 80°–90°), make the finger pulp coronary position

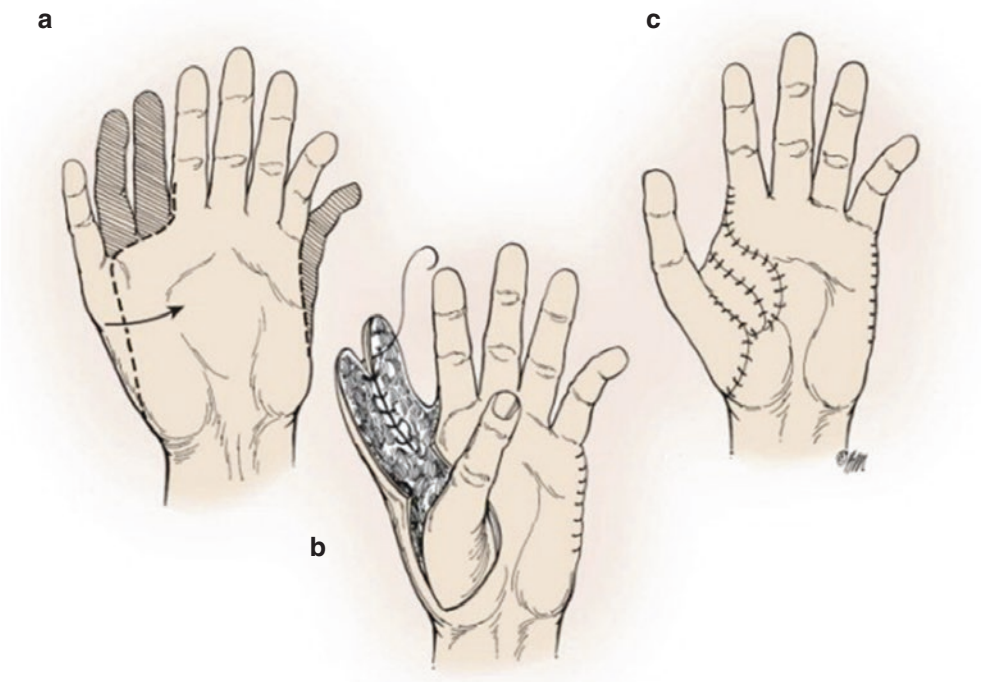

Fig. 7.60 Treatment of mirror hand. (**a**) Select the functionally and morphologically optimal fingers for pollicization. (**b**) Form the supernumerary into filleted flaps for thumb web reconstruction. (**c**) The postaxial supernumerary finger can be removed to improve the seriations and appearance of the whole hands (originate from Barton NJ, Buck-Gramcko D, Evans DM, et al.: Mirror hand treated by true pollicization. J Hand Surg [Br] 1986; 11:320–336) (Reprint with permission from ELSEVIER LICENSE TERMS AND CONDITIONS)

of the reconstructed thumb pronate by 45°, and take palm-opposing position, so that it is opposite to the other four fingers. The fixation method of carpometacarpal joint and the aesthetical reconstruction and dynamical reconstruction of the reconstructed thumb include the shortening of extensor tendon of the pollicized finger, the replacement of thumb adductor with volar interosseous muscles, the replacement of thumb short abductor with dorsal interosseous muscle, and the replacement of thumb long abductor with inherent extensor tendon; the details are shown in Chap. 5 "Congenital Thumb Dysplasia."

7.5.5 Typical Case

A boy, with left mirror hand and the deformity of two ulnae, received the surgery in three stages, and the first surgical correction was performed at the age of 6 months.

7.5.5.1 Phase I Surgery

1. Surgical design. Among the four fingers at the radial side, the third finger is relatively thick and used as the finger for thumb reconstruction, and the supernumerary fingers and metacarpal bones are resected. The thumb reconstruction method and the surgical method of thumb dysplasia are similar.
2. Finger grafting for thumb reconstruction. Select the third finger at the radial side as the finger for thumb reconstruction, and resect the first, second, and fourth fingers and metacarpal bones at the radial side. Design one linguiform flap at the volar side of the proximal interphalangeal joint of the third finger at the radial side, and design two flaps at the back of hand. As the reconstructed thumb is thin, the flaps with vascular nerves at the finger pulp of the adjacent fingers can be selected to enlarge the reconstructed thumb. The surgical design is seen in Chap. 5 "Congenital Thumb Dysplasia."
3. Finger shortening for thumb reconstruction. Partially resect the third metacarpal bone at the radial side, rotate the dorsal side of the metacarpophalangeal joint by 90° for fixation, and fabricate the first carpometacarpal joint of the reconstructed thumb; or perform partial resection of the proximal end of the proximal phalanx of the third finger at the radial side and the distal end of the third metacarpal bone, fabricate it into the biphalangeal thumb (Fig. 7.61), and resect other supernumerary fingers.
4. Reconstruction of the palm-opposition position of the reconstructed thumb. After shortening the phalanx and metacarpal bone of the third finger at the radial side, rotate it to the palm-opposing position for fixation as the reconstructed thumb.
5. Dynamical reconstruction of the reconstructed thumb. Retain the tendon of the first finger at the radial side or the flexor brevis of the little finger as the thumb short abductor of the reconstructed thumb. Shorten the digital extensor tendon of the third finger at the radial side, use it as the thumb long extensor of the reconstructed thumb, and make the thumb flexor become retracted after the surgery. Retain the lumbrical muscle or interosseous muscles of the fourth finger at the radial side as the adductor of the reconstructed thumb. Grafting the digital extensor of the first and second fingers at the radial side as the thumb long abductor of the reconstructed thumb.
6. Hemostatic suture. Trim the supernumerary skin, carefully perform hemostasis, and suture the skin accurately.
7. External brace fixation of reconstructed thumb. Use the brace to control the thumb web in the expanding position, and fix the thumb in the palm-opposition position for 1 month (Figs. 7.61 and 7.62).

7.5.5.2 Phase II Surgery

Correct the wrist joint, reconstruct the wrist extensor at the radial side, and reconstruct the pronation and supination functions of the forearm.

7.5.5.3 Phase III Surgery

Reconstruct the elbow joint functions (Fig. 7.63).

Tsuyuguchi (1982) [47] reported that phase II and phase III surgeries can be implemented together.

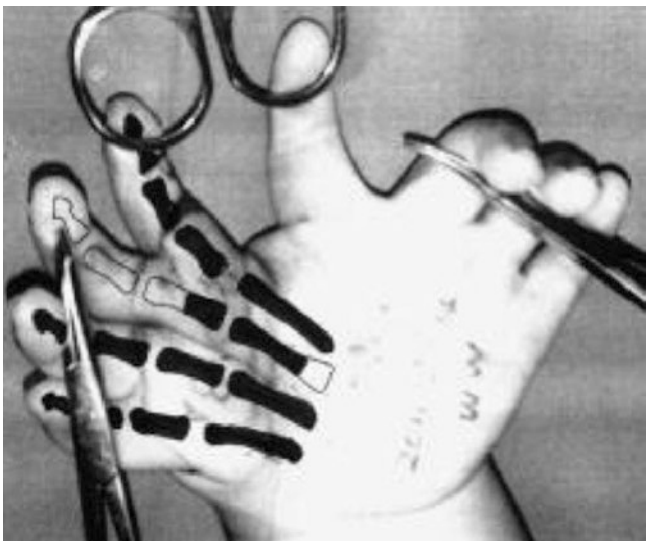

Fig. 7.61 Perform partial section of the proximal end of the proximal phalanx and the distal end of the third metacarpal bone of the third finger at the radial side, and fabricate it into the biphalangeal thumb (the black substance is the resected part of phalangeal metacarpal bone, and the black hollow is the retained part of the phalangeal metacarpal bone)

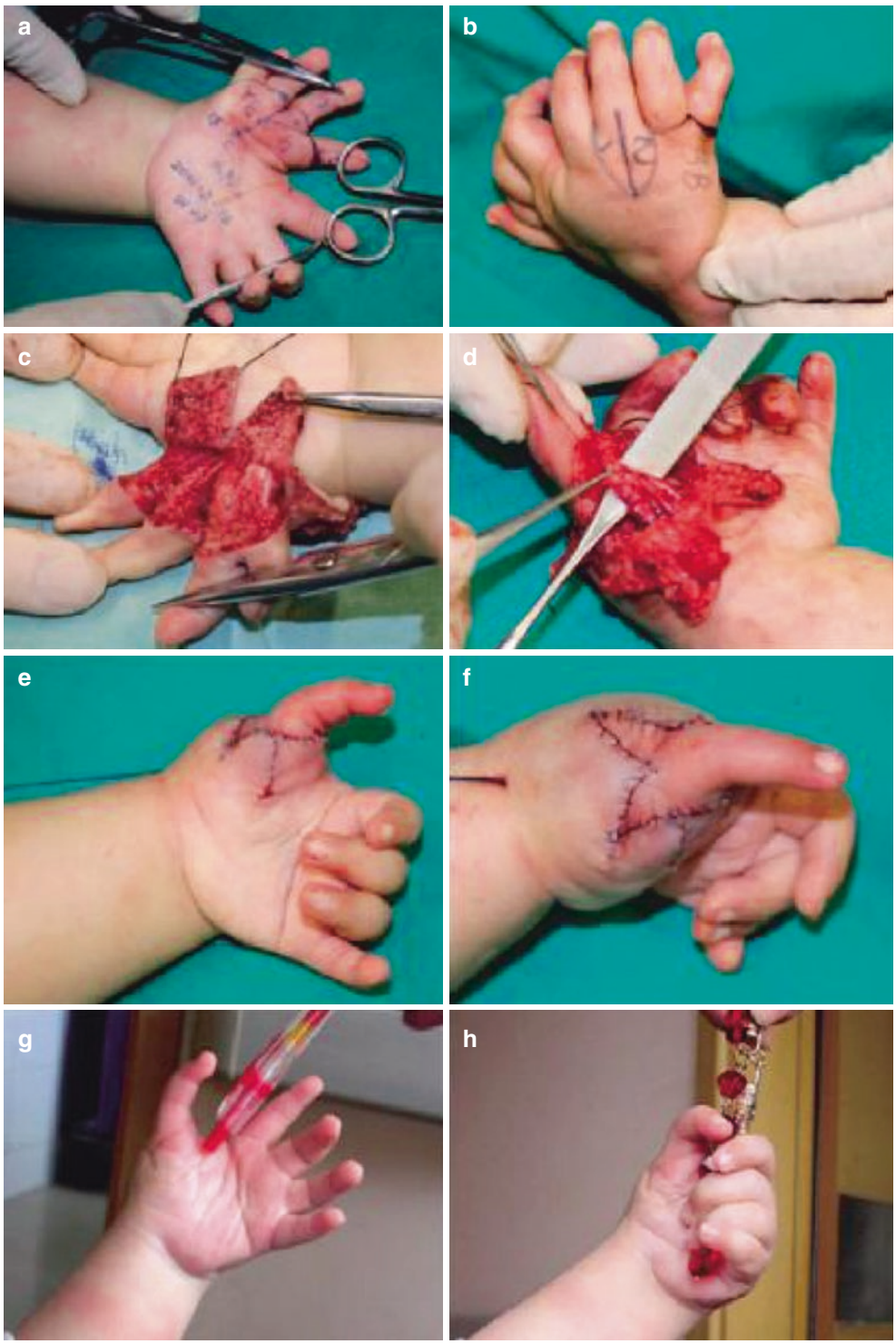

Fig. 7.62 Correction of mirror hand deformity. (**a**, **b**) Surgical design. (**c**) Lifting of volar flaps during operation. (**d**) Protection of nerve vascular bundles. (**e**, **f**) Hand appearance immediate after operation. (**g**, **h**) Hand appearance and functions at 6 months after operation

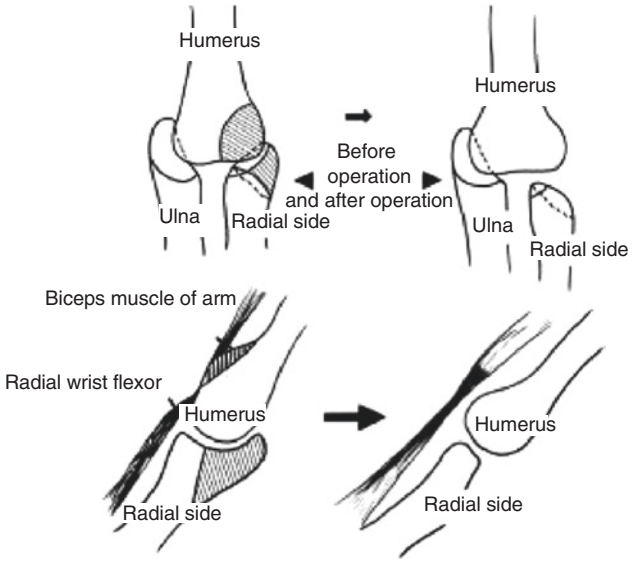

Fig. 7.63 Schematic diagram of elbow joint functional reconstruction in phase III surgery on mirror hand (the oblique line represents the resected part)

7.6 Congenital Superfluous Hand Deformity

Congenital superfluous hand is a very rare congenital limb deformity, with the manifestations of neoplasm of upper-limb-like or handlike structure on the trunk, and the neoplastic part may be the entire limb or part of the limb part. The neoplastic limb is often located at the abdomen or the back, and the neoplasm on the face is very rare.

7.6.1 Clinical Manifestation

Congenital superfluous hand is the partial or whole hand-like structures growing on the trunk, and the patient often has normal hands. The neoplastic hand has partial or whole characteristics of the hands, but they are not functional and are often connected with the deep tissues. This disease is very rare. Dai Chuanchang once treated one case: the superfluous finger was located at the face; the finger sagged toward the buccal part and was connected with skin pedicle and auricles, and in the meantime, the patient concomitantly suffers from syndactylia (Fig. 7.64).

7.6.2 Etiological Factors and Pathology

The limb bud or the ectodermic layer is subject to special damages during the early stage and part of it experiences division. This might be the reason for deformity of superfluous thumb. The superfluous hand has all or some structural characteristics of the hand (e.g., bone scaffolds), but it has no dynamical system of the hand (e.g., tendons). It falls into the category of deformity of limb duplication according to the Swanson's classification.

7.6.3 Treatment

The treatment of congenital superfluous hand should aim to improve appearance and correct psychological disorder to resect and repair the superfluous hand. If the hand is connected with deep tissues, ensure the deep tissues are not injured during the surgical resection and just resect the superfluous hand; if it forms a joint with spinal column or is accompanied by cleft spine, this should be clarified before surgery, and the surgery should be completed under the guidance of relevant disciplines.

When the superfluous hand is located at the abdomen, the flaps of the superfluous limb should be used to repair the abdominal defects after it is resected.

7.6.4 Typical Case

The author once treated one case of superfluous hand at the neck during the 1980s. The patient was a 14-year-old boy with one superfluous dysplastic limb at the neck, which was shaped like a seal hand and attached on the cervical vertebra VI-VIII and thoracic vertebra I (C6–8 and T1) and T1 regions. The patient was accompanied by spina bifida. The sensations of the superfluous hand are present, but the superfluous hand does not have the motor functions. No deformity was seen in other systemic organs, the patient was free from history of familial inheritance, and the parents said that the intelligence of the pediatric patient developed very well. The superfluous hand was resected under the condition of general anesthesia. Due to the presence of spina bifida, the material part of the cervical and thoracic vertebrae was not involved, and the appearance of the neck and spinal column was improved after surgical resection (Fig. 7.65).

Hua Mingxue once reported one 11-month-old girl, who had a soft tissue lump with a diameter of about 5 cm on the back (T7 to 9 segments), and there were five lacunae with

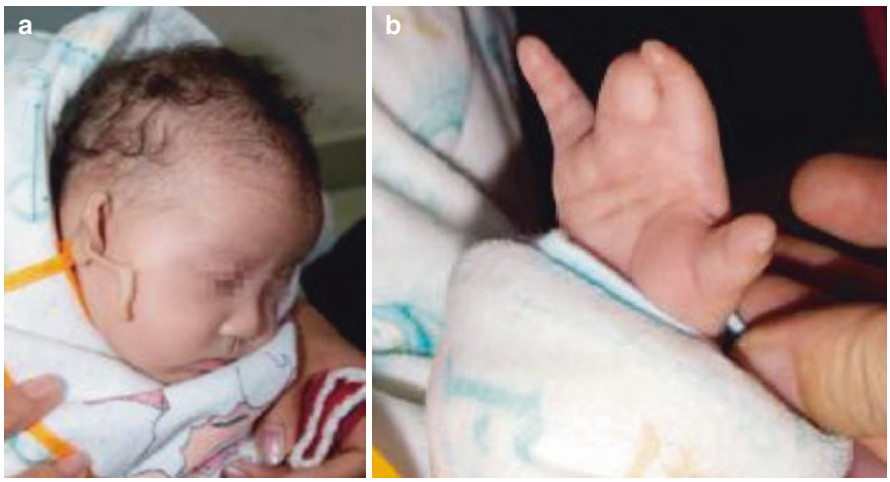

Fig. 7.64 Facial superfluous finger with deformity of syndactylia of index, middle, and ring fingers

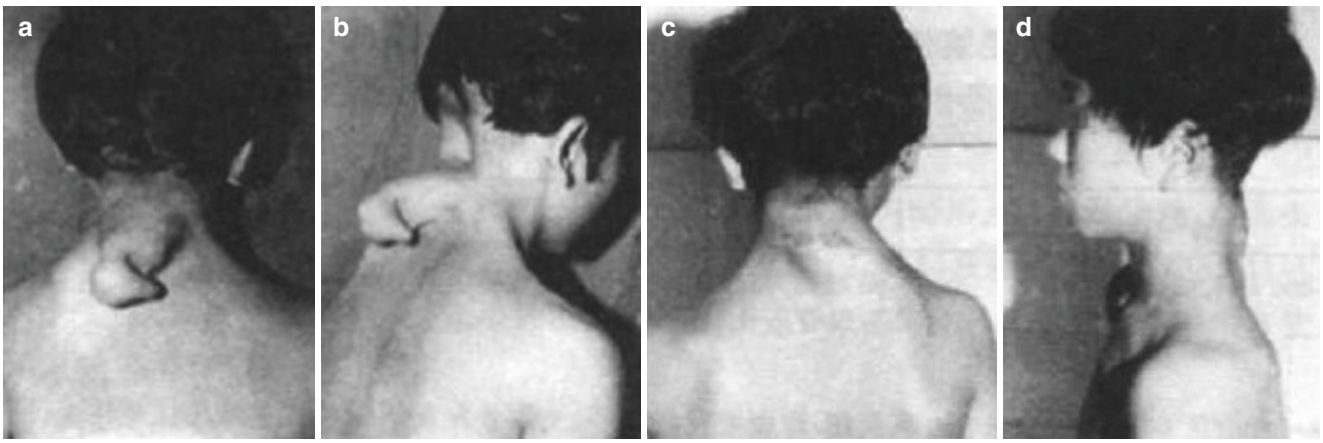

Fig. 7.65 Congenital superfluous hand deformity at the neck. (**a**, **b**) Before operation. (**c**, **d**) After operation

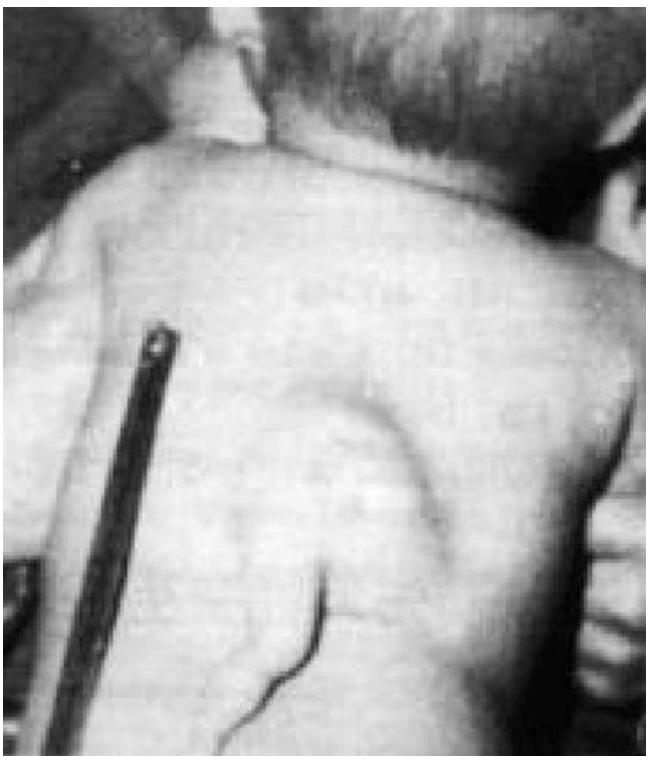

Fig. 7.66 Congenital superfluous hand deformity on the back

varying sizes and depths on the surface. One finger-shaped object was found at its upstream, the fingertip was downward with a length of 8 cm, the appearance presented three segments, the finger transverse striations were clear, there was no active movement and function, and the fingernails and finger pulp at the terminals were the same as those of normal fingers. The X-ray film indicated T7 to T9 had bifid vertebrae, and T6 to T11 had bifid spine accompanied by clubbed spinous processes (Fig. 7.66).

The superfluous limb can also grow on the abdomen, and the patient is like one of conjoined twin babies with incomplete growth.

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Low Development

8

Jinghong Xu, Bo Chen, Xu Gong, Liang Ma, Dongping Li, Shengjun Sun, Jianmin Yao, Wei Wang, Yijia Yu, Qingping Xie, and Laijin Lu

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8.1 Anomaly of Brachydactylia

Brachydactylia is a type of autosomal dominant inheritance deformity with the main manifestation of short and deformed phalanges induced by phalangeal and metacarpal dysplasia (Fig. 8.1). Because it was induced by the low

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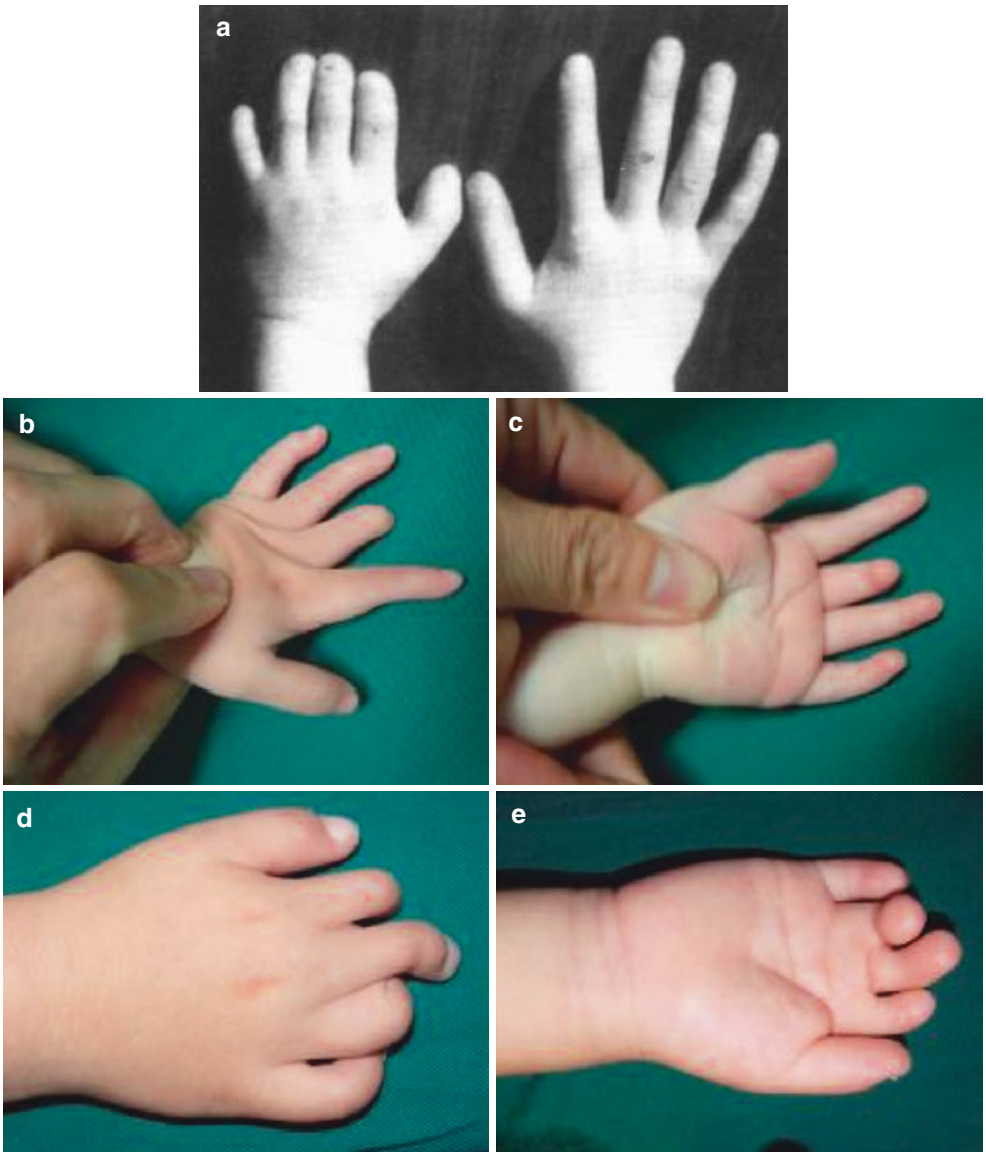

Fig. 8.1 Anomaly of brachydactylia

development of hands and fingers, it is also called hand and finger dysplasia. It is commonly believed that brachydactylia is mainly associated with genetic factors and environmental factors, and the drug-induced brachydactylia cannot be overlooked. The hand and finger dysplasia can occur independently or appear in many syndromes, such as Apert syndrome and Poland syndrome [1]. This deformity can be manifested as the partial absence of single finger or as the absence of one or more fingers. It can also be accompanied by metacarpal absence, resulting in cleft hand; in severe cases, the whole hand can be absent. In addition to macromelia, nearly all congenital upper limb deformities can be accompanied by hand and finger dysplasia to varying extents. The phalangeal growth plate injuries induced by trauma, infection, etc. often induce brachydactylia. Thumb dysplasia also falls into the category of hand and finger dysplasia. Based on the specificity of the treatment, it is described as a special subject.

The single-fingered spade hand or the hand dysplasia in Apert syndrome and Poland syndrome all show brachydactylia or syndactylia, and this deformity is sometimes divided into the range of syndactylia.

8.1.1 Etiologic Factor

Brachydactylia is mainly induced by genetic factors and environmental factors, or induced by compression, such as intrauterine amputation induced by amniotic band or cord entanglement and compression, which can result in the ectromelia of the whole hand or limb. Drug-induced deformity is more important. For instance, the thalidomide incident that occurred during the early 1960s resulted in a large number of fetal limb deformities, especially deformities of brachydactylia and adactylia. There were reports that brachydactylia is caused by congenital syphilis and endo-

crine functional disorder, because phalangeal endochondral ossification during the embryonic middle period is subject to interference, the endochondral growth suffer disorder or discontinuation, so that the phalange is shortened and the fingers are short, and even deformity of phocomelia is caused.

The familial inheritance is often manifested as autosomal dominant inheritance. The first case of Mendelian autosomal dominant genetic disease, i.e., type A1 brachydactylia, was first reported by Farabee in 1903 and was quoted by most genetics and biology textbooks all over the world. During the recent 100 years, scientists from all over the world effortfully studied its pathogenesis, but no substantial breakthrough has been made.

With the completion of human genome during the recent years, one after another breakthrough has been made in genetic diagnosis. The scholars in China have accurately located the disease-causing gene (the site is located at region 35–36 of No. 2 chromosome) and first found that the IHH gene and the three mutation sites on this gene are the direct reason that induces type A1 brachydactylia. Foreign scholars also successfully elucidated the mutator gene of brachydactyly in 2001, which provides a good foundation for the prevention and treatment of such a disease.

8.1.2 Classification

8.1.2.1 Blauth and Gekeler Classification [2]

In 1971, Blauth and Gekeler classified brachydactylia according to the severity.

1. Short finger type (type I). Some or all of the middle phalanxes are shortened, and the central digital ray is often involved; the thumb still retains its normal size and shape, but the root of the thumb is often located at the metacarpal level; the wrist joint is dysplastic and loses the functions of rotation and flexion. The whole hand often becomes homogeneously shortened, and all fingers are involved; compared with the normal hands, the affected hand is significantly shortened and grows with age (refer to Fig. 8.1).
2. Cleft hand type (type II). One digital ray or multiple digital rays in the center suffers serious dysplasia, which is common at the ulnar side; in severe cases, one or more fingers are just one limb-bud-like neoplasm, including the remnants of bones and cartilages. The adjacent fingers can suffer the deformity of bending, the hypoplasia of phalanges and joints, and the dislocation or semi-dislocation of interphalangeal joints.
3. Single-fingered type (type III). The second to fifth fingers become shortened to one residual finger with a nail; the thumb can retain its shape and functions, but it is relatively small and accompanied by limited activity.
4. Ectromelus type (type IV). All of the fingers are absent only with one-hump eminence.

8.1.2.2 Bell's Classification

In 1951, Bell [3] analyzed 124 families with dominantly inherited brachydactylia. According to the anatomic shape, the genetic non-comprehensive brachydactylia was classified into five types:

1. Type A: Reclassified into four subtypes:
   (a) Type A1: the middle phalanxes and the thumb proximal phalanx of the second to fifth fingers are short and small.
   (b) Type A2: the middle phalanx and thumb phalanxes of the second finger are short and small, the epiphysis is absent, and the S-shaped phalange bends toward the radial side.
   (c) Type A3: the middle phalanx of the fifth finger is short and small with the deformity of radial deviation.
   (d) Type A4: the middle phalanxes and the thumb proximal phalanx of the second to fifth fingers are short and small, bifurcation is seen in the thumb proximal phalanx, and the fifth finger has no function.
2. Type B: The middle phalanxes of the second to fifth fingers are short and small, and the phalangette is short and small or absent.
3. Type C: The middle phalanxes of the second, third, and fifth fingers are short and small, and the proximal phalanxes of the second and third fingers are excessively long.
4. Type D: The proximal segment of the thumb is short and thick.
5. Type E: The middle phalanxes of the third to fifth fingers are short.

8.1.3 Clinical Manifestation

This disease is characterized by short and small hands and fingers, and it can be simple short and small phalanxes or the brachydactylia induced by short metacarpal bones. According to the site of shortening, the brachydactylia can be classified into brachytelephalangia, brachymesophalangia, brachybasophalangia, and brachymetacarpia. In addition, the deformities include hypophalangism and hyperphalangism, and

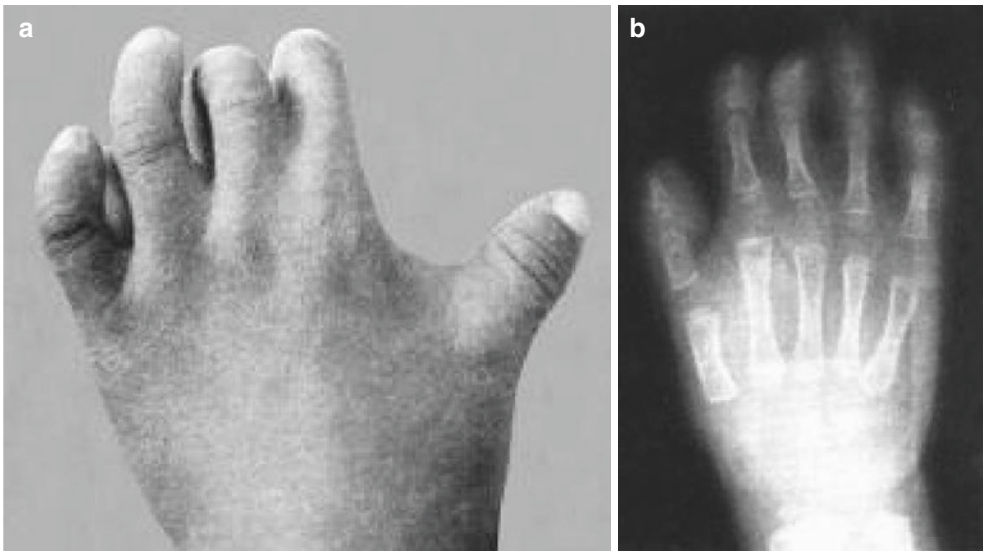

Fig. 8.2 Brachydactylia and syndactylia deformity: two tri-dactylopodited short fingers are abreast

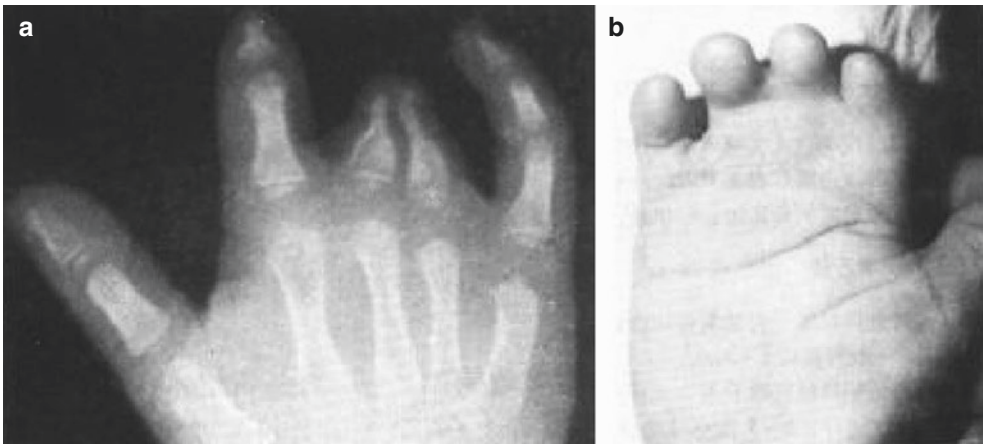

Fig. 8.3 Brachydactylia and syndactylia deformity: two mono-dactylopodited short fingers are abreast

the latter deformity had superfluous one phalange at the proximal segment. Brachydactylia can coexist with syndactylia and take multiple forms; two tri-dactylopodited short fingers can be abreast (Fig. 8.2), or two mono-dactylopodited short fingers can be abreast (Fig. 8.3); the thumb can be normal, and the four short fingers (second to fifth fingers) are abreast (Fig. 8.4); the manifestations can be found in the whole hand, and the thumb and all the short fingers (second to fifth fingers) are abreast (Fig. 8.5). If the deformity is only limited to one or more fingers and the corresponding metacarpal bone, it is called brachydactylia; if the deformity involves all the fingers, it is called microcheiria (Fig. 8.6). The shape or functions of mild short fingers are close to normal with only one phalange or metacarpal bone or with dysplasia of a certain tendon or muscle. In the case of severe hand dysplasia, the finger is like a bean-shaped wart and attached to the distal end of the palm, and it has no bone,

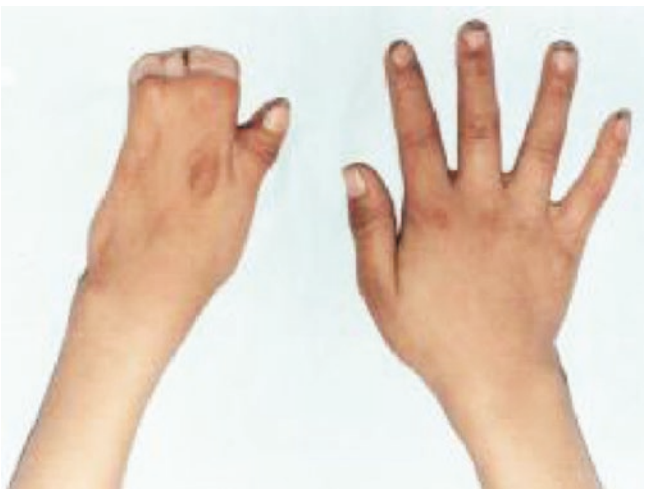

Fig. 8.4 Brachydactylia and syndactylia deformity: the thumb can be normal, and the four short fingers (second to fifth fingers) are abreast

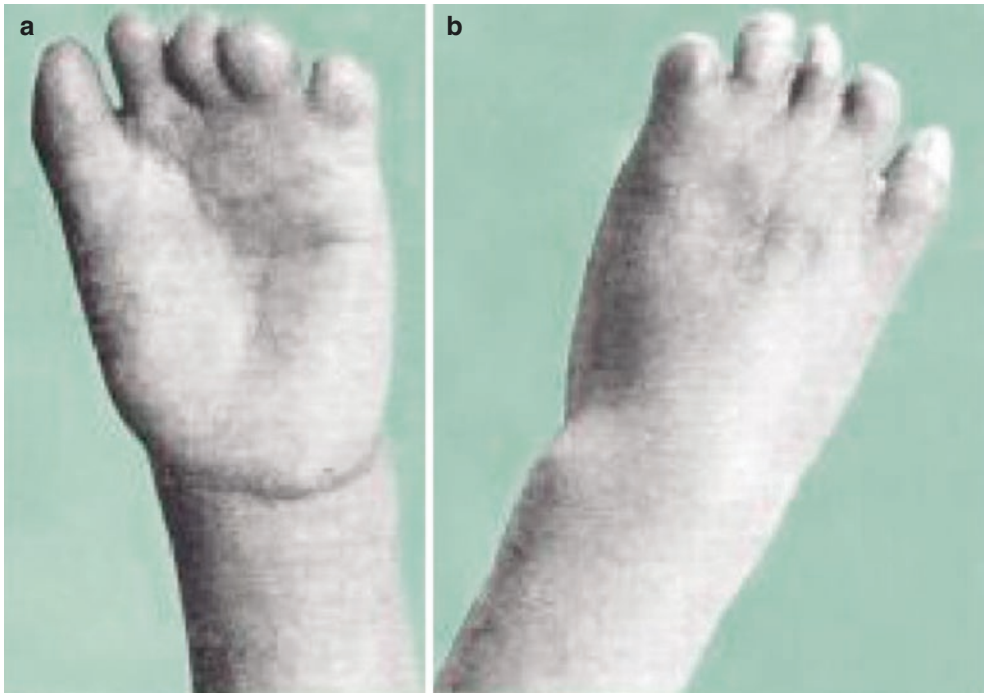

Fig. 8.5 Microcheiria and syndactylia: the short fingers of the whole hand are abreast

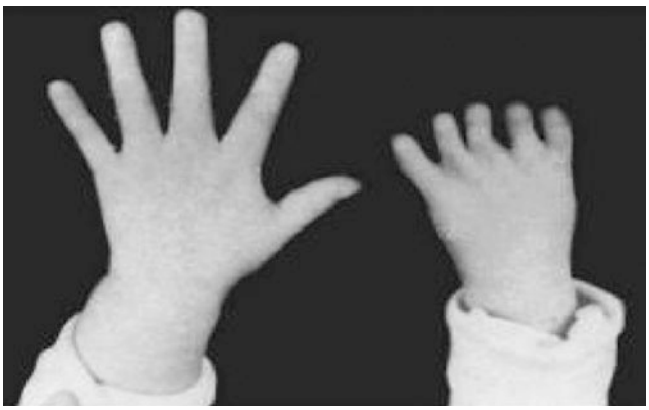

Fig. 8.6 Microcheiria deformity

joint, tendon, or component except skin and subcutaneous tissues; or the brachydactylia exists, and the finger tips suffer syndactylia to varying extents (Fig. 8.7). This disease often involves chest wall (e.g., Poland syndrome), and during examination, whether the chest wall is abnormal should be observed [4].

8.1.4 Treatment

The treatment method of this disease depends on the degree of finger shortening, the status of residual finger and the abnormalities in other parts of the body [3].

8.1.4.1 Nonsurgical Therapy

The treatment method of this disease depends on the degree of finger shortening, the status of residual finger and the abnormalities in other parts of the body [4].

8.1.4.2 Surgical Therapy

Except some single brachydactylia, almost all the other types of brachydactylia suffer functional disorder; therefore, surgeries are needed to recover and reconstruct the finger functions. As for the patients with degree II to IV brachydactylia, the main purpose of surgeries is to improve the grasping and finger to finger functions of the hands.

Preschool is the best surgical timing for degree I to II deformities, and the joint cannot be fixed until the age of epiphyseal arrest. The surgical methods are as follows:

1. Metacarpal bone extension. Perform longitudinal incision at the dorsal side of the shortened metacarpal bone, pull the finger extensor tendon to one side, expose the shaft of metacarpal bone, perform two V-shaped osteotomy surgeries at the 1/3 junction of the proximal-middle part of the metacarpal bone, expose the deep transverse metacarpal ligament to the distal side and perform the incisions, and detach the interosseous muscles at two sides of the metacarpal bone. Pull the metacarpal bone, pick bones at the iliac crest, insert them into the space after molding, and fix them with Kirschner silk. Refix the interosseous muscles at the metacarpal bone or bone graft in the proper positions, suture the skin, and perform external fixation

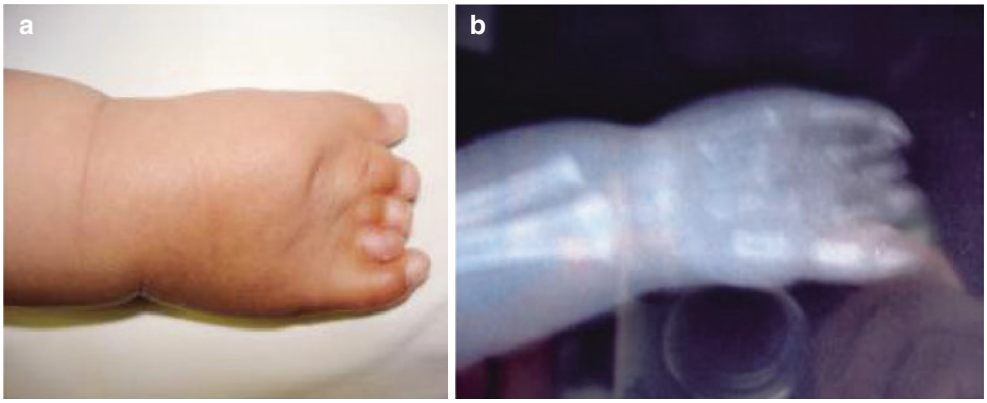

Fig. 8.7 Severe hand dysplasia

with plastic splints. Three weeks after operation, start using the fingers, and 6 weeks after operation, remove the Kirschner silk.

2. Traction and lengthening. Arslan and Miyawaki [5, 6] once reported the application of bone tractors to treat the brachydactylia, which is currently the common method. The surgeries need to be performed in different phases: phase I, keep pulling the interosseous space until a proper length is reached or the limit of nerves, vessels, and skin is reached; phase II, pick bones from the sites of iliac crest, ulnar bone, and fibula and insert them into the interosseous space, and fix them with Kirschner silk.
3. Grafting of phalanges of foot. First pick the second; if necessary, the doctors can choose other toes. As for the degree III brachydactylia patients with an intact thumb but absent second to fifth fingers, this method is relatively ideal; the reconstruction of one to two fingers can recover the finger motor functions, and the appearance is satisfying. However, the difficulty of the surgery is high [4].
4. Arthroplasty. For those with stiffness of finger joints or metacarpophalangeal joints that affect the functions, arthroplasty can be performed.
5. Dactylolysis. For the patients concomitantly suffering syndactylia, dactylolysis can be performed according to the specific circumstances; especially for the patients with microcheiria accompanied by syndactylia, dactylolysis should be performed as early as possible to improve the hand appearance and better exercise finger functions.
6. Bone grafting and skin tube lengthening. Pick the iliac bone blocks to lengthen the fingers, and then adopt the skin tubes for repairing. This surgical method is simple, but the reconstructed finger is fat and unpleasant, its functions are not satisfying, so this kind of surgery is seldom adopted at present.

This disease is a genetic disease, so prevention is very important; whether the new prevention and treatment methods can be found through the gene engineering technology needs further exploration.

8.2 Brachysyndactyly

Brachysyndactyly is a complicated deformity with coexistence of finger shortening and syndactylia. Brachysyndactyly can be found in many syndromes, such as Apert syndrome and Poland syndrome [7, 8].

Poland syndrome is a rare type of congenital deformity that has been reported long ago [9], including unilateral sternal rib dysplasia and unilateral major pectoral muscle, minor pectoral muscle, and ipsilateral upper limb dysplasia. It often occurs to the right side of girls, who concomitantly suffer from breast dysplasia. The manifestation of hand dysplasia is short and small hand, syndactylia, and brachydactylia. The patients often visit the doctor due to congenital syndactylia. The etiologic factors are often considered to be correlated with deformities in subclavian arterial series.

8.2.1 Selection of Treatment Timing

The treatment timing of syndactylia should depend on the patient's general conditions, hand functional lesion, syndactylous site, safety of anesthesia, and requirements of parents. If conditions permit, the surgery should be performed before the patient becomes 5–6 months old; in case of the bone fusion type syndactylia, the surgery can be postponed but should be finished before the patient becomes 2 years old; if it is necessary to postpone the surgery for some other reasons, the treatment should be given before the patient's school age.

The surgery on skin syndactylia deformity only involves separation of fingers, so the surgical method is simple. But it is not easy to make the affected fingers nearly functionally normal. If microsurgical techniques are adopted, even though syndactylous finger separating surgery is performed on newborns, the surgery is not technologically difficult. The key is to choose the accurate skin to make Z-plasty incisions, achieve tension-free suture, and use the flaps to repair the fingerweb. As for the skin defects, skin graft transplantation

is needed. If possible, experienced surgeons can arrange syndactylous finger separating surgery for the pediatric patients that were newly born. During correction of complicated syndactylia, as for the patients with bone fusion deformity, the surgical time can be postponed. It is proper to perform the plastic surgery of crossed brachydactylia when the patient is 6 months to 2 years old. The surgery aims to repair and reconstruct the shape and functions of main fingers. If necessary, the length of fingers with less functions needs to be sacrificed, and the length of important fingers will be increased. The finger separating surgery of the syndactylia of four fingers needs to be performed in different times.

8.2.2 Surgical Method

The surgical methods include: ① finger separating surgery of syndactylia, which is the surgery to which top priority should be given; ② bone lengthening, including the lengthening of phalanges and metacarpal bones, which is the main surgery for brachydactylia correction; ③ the spade hand that is often accompanied by the presence of thenar muscles and pollicization of the parallel radial fingers which is a simple and effective method; ④ and, as for the finger absence due to serious dysplasia, toe transplantation which can be performed to reconstruct the thumb and the middle finger or reconstruct the thumb, middle finger, and ring finger.

Zhang Pingyuan et al. adopted finger separating and upward shift of fingerweb to treat the complicated short and small syndactylia and achieve satisfying effects. As the flap dissociation range is wide and fingerweb reconstruction position is high, some space is reserved for further growth. The follow-up visit after a long period of time indicated that the fingerweb shifted upward after surgery and the purpose of finger separation and finger lengthening as well as treatment of brachysyndactyly was served.

8.2.3 Typical Case

An 8-year-old pediatric patient with Poland syndrome-type spade hand deformity. The surgery is performed under the condition of generalized anesthesia to separate the spade hand, pollicize the first metacarpal bone, rotate the osteotomized bone to the palm opposition position, use the local flaps to repair thumb web, and use free skin grafts to repair the deficiencies (Fig. 8.8) [10–14].

8.3 Hand Flexor and Extensor Dysplasia

Bo Chen, Liang Ma, Dongping Li, Shengjun Sun, Jianmin Yao, and Wei Wang

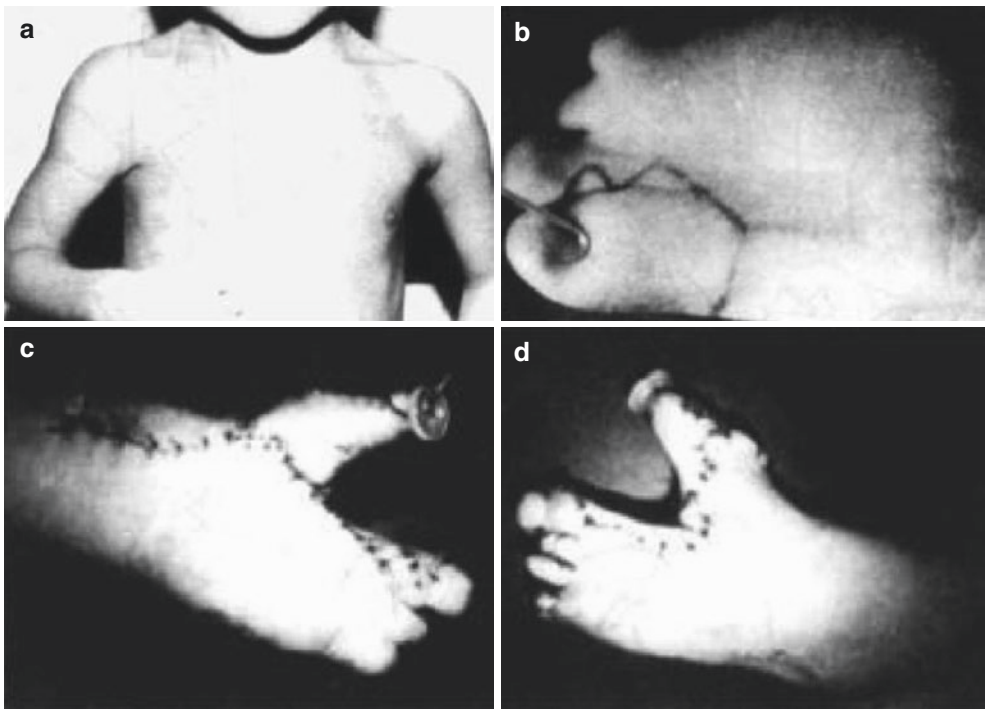

Fig. 8.8 Finger separating surgery of Poland syndrome-type spade hand deformity. (**a**) Before operation. (**b**) Design of skin incision. (**c**, **d**) After operation

8.3.1 Abnormality of Superficial Digital Flexor

8.3.1.1 Classification

There are three types of abnormality of superficial digital flexor

1. Long muscle type. The belly of superficial digital flexor extends from the forearm to the distal end of carpal canal.
2. Short muscle type. The superficial digital flexor does not originate from the forearm but from the wrist transverse ligament and palmar aponeurosis; or one abnormal muscle belly originates from itself.
3. Biventer type. Abnormal muscle belly is located at the palm and substitutes some tendons, and the tendons at the two ends are connected [15].

8.3.1.2 Clinical Manifestation

Abnormality of superficial digital flexor often shows no symptom. The reported cases are often misdiagnosed as soft tissue tumor due to the occurrence of local lumps or found during the carpal tunnel syndrome surgery [16].

8.3.1.3 Treatment

The asymptomatic patients may not be treated; in case of protruding local lumps or compression of surrounding tissues, especially occurrence of the symptom of nerve compression, it can be resected. As for those with intact profound digital flexor functions, the tendons may not be repaired after the superficial digital flexor is resected.

For those concomitantly suffering absence of thenar muscle and thumb long flexor, retain and then dissociate the proximal vascular nerve of this abnormal muscle belly, disjunct it from the tendon of the two segments, and displace it to replace the thumb short abductor; that is, suture the proximal end of this muscle belly at the wrist transverse ligament, and suture the distal end to the radial lateral tendon bundle of the thumb through the subcutaneous channels so that the thumb can be located in the abduction palm opposition position. In the meantime, resect the superficial flexor tendon of the ring finger, displace it, and fix it at the end of the thumb to replace the thumb long flexor tendon.

8.3.2 Abnormality of Thumb Long Flexor

The muscle that is the most frequently reported in the flexor tendon is the thumb long flexor. Anatomically, the abnormal tendon connection between the thumb long flexor and the index finger profound flexor muscle is the commonest. When the thumb interphalangeal joint is flexed, the proximal and distal sides of the index finger are also flexed.

8.3.2.1 Classification

1. Simple absence of thumb long flexor. The simple absence of thumb long flexor was first reported in 1895. During the process, the formation and growth of thumb long flexor are separated from profound digital flexor. There have been many reports about the simple absence of thumb long flexor; residual fibrous tissues can be found in the absent tendons, and the traction of them can cause flexion of interphalangeal joint [17].
2. Complicated abnormality of thumb long flexor. It is the thumb long flexor abnormality accompanied by other abnormalities: the abnormal thumb long flexor is mutually connected with the radial margin of the thumb long extensor, impeding the flexion of interphalangeal joint and abducting the thumb; the thenar muscles suffer dysplasia, and the thumb long flexor tendon is located at the carpal canal volar side or passes through the wrist transverse ligament; the ulnar collateral ligaments are relaxed with functional insufficiency; the thumb long flexor and the profound digital flexor share a muscle belly [18].

8.3.2.2 Clinical Manifestation

There are mainly two kinds of clinical manifestations of abnormal thumb long flexor: one is the simple type, e.g., interphalangeal joint flexor disorder is suffered during the infancy or childhood, which can be unilateral or bilateral, and some patients have a family history [19]; another manifestation is the thumb long extensor tendon is attached to the insertion of the thumb long extensor tendon, the thumb is abducted when the interphalangeal joint is flexed and it is often accompanied by thumb dysplasia, and the dermal ridges of thumb interphalangeal joint disappear.

8.3.2.3 Treatment

1. Simple absence of thumb long flexor. There should be two prerequisites for the surgical treatment: ① the passive activity of finger joint is good; ② the pediatric patient is cooperative during rehabilitation.
2. Thumb long flexor absence accompanied by tendon sheath absence. The thumb long flexor is often accompanied by tendon sheath absence; the reconstruction of tendon also needs the reconstruction of tendon sheath, especially A2 trochlea, and extensor retinaculum can be adopted to reconstruct the A2 trochlea. If the thumb long flexor has no muscle belly, the ring finger superficial flexor tendon can be displaced to reconstruct the thumb long flexor tendon.

8.3.3 Abnormality of Thumb Long Extensor

The abnormality of finger extensor is relatively rare. In 1934, Zadek first reported the absence of thumb long extensor in both hands, and genetic predisposition was found in one family [20].

8.3.3.1 Clinical Manifestation

The manifestation of the typical simple dysplasia or absence of the thumb long extensor is that the thumb is in the flexion position and the interphalangeal joint suffers disorder in extension; but in some cases, as the thumb short extensor is present and the insertion is at the distal end of thumb, the interphalangeal joint may be able to be extended, and attention should be paid to this during the examination.

8.3.3.2 Treatment

The treatment methods of simple absence of thumb long extensor:

1. Displacement of index finger inherent extensor tendon. Disjunct the index finger inherent extensor tendon from the dorsal side of the interphalangeal joint, extract it from the incision of dorsal part of the wrist, pull it to the dorsal side of the thumb interphalangeal joint through the subcutaneous channels, and fix it at the dorsal side of fundus of the thumb phalangette.
2. Displacement of radial wrist extensor tendon. As for the cases of abnormalities in index finger inherent extensor tendon, the doctor can disjunct the radial wrist long extensor at its insertion, lengthen it through tendon transplantation, pull it to the dorsal side of the thumb interphalangeal joint through the subcutaneous channels, and fix it at the dorsal side of fundus of the thumb phalangette.

8.3.4 Abnormality of Finger Extensor

8.3.4.1 Clinical Manifestation

The sequence of muscles according to incidence of finger extensor abnormalities from high to low is common extensor of fingers, index finger inherent extensor, thumb long extensor, and little finger inherent extensor. The clinical manifestation is that the involved finger metacarpophalangeal joint suffers disorder in extension [21].

8.3.4.2 Treatment

Pull the ring finger superficial flexor tendon to the dorsum of wrist through the interosseous membrane to repair the absence of thumb long extensor. Disjunct the radial wrist short extensor from its insertion, and resect the volar long tendon for transplantation, and lengthen it to repair the absence of index finger and middle finger common extensor muscles.

8.4 Madelung Deformity

Madelung deformity is also called congenital distal ulnar-radial joint semi-dislocation and wrist joint progressive semi-dislocation. In 1839, Dupuytren first reported this deformity; in 1878, Madelung first gave a detailed description of the clinical manifestations of this deformity, so it is called Madelung deformity. The mechanism of this deformity is as follows: the ulnar and volar partial epiphysis aplasia at the distal end of the radial bone and the continuous normal development of ulnae cause the distal ends of ulnar and radial bones not to be on the same plane, which leads to semi-dislocation [22–24].

The incidence of this disease in females is higher than that in males, and it is commonly suffered by children aged between 6 and 13. The onset often involves two sides, but the degree of the deformity at the two sides are not always the same. The nosogenesis of Madelung deformity is not very clear. It is generally believed to be the hereditary disease with chondrodysplasia, and it is inherited through dominant genes with incomplete penetrance.

8.4.1 Clinical Manifestation

Madelung deformity is induced by disorder in the growth and development of distal epiphysis of some radial bones during the embryonic period, so this disease is often not obvious and some patients do not visit the hospital until the wrist is painful. The clinical manifestations of Madelung deformity mainly include wrist joint deformity and pain, wrist joint instability, and wrist joint movement disorder [25–27].

8.4.1.1 Deformity

Deformity is the main symptom of Madelung deformity, the volar and ulnar osteochondrodysplasia at the distal end of the radial bones is the main reason for deformity, ulnae protruding toward the dorsal side can be observed at the ulnar side of the wrist, the distal ulnar and radial joints can present the manifestation of dislocation, and the hand inclines toward the ulnar side (Fig. 8.9).

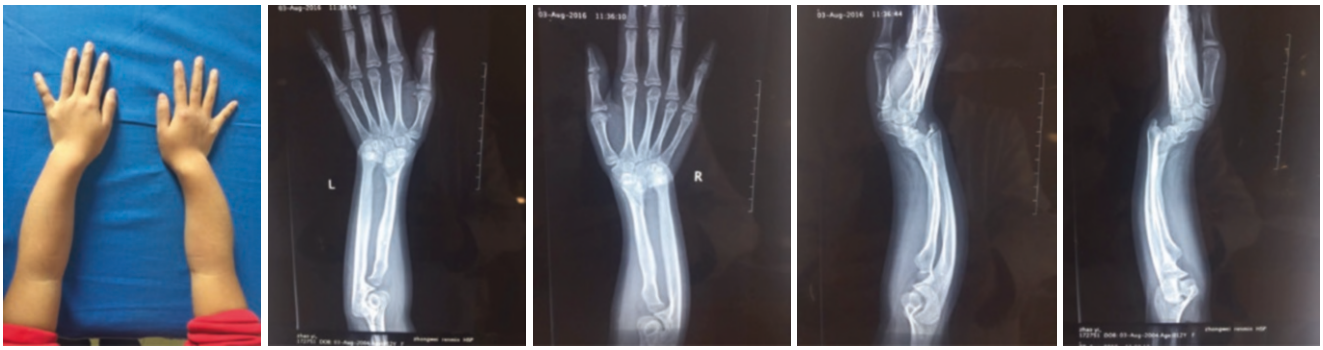

Fig. 8.9 Madelung deformity (Offer by Ningxia Hui Autonomous Regional Corps Hospital, Chinese People's Armed Police Force)

8.4.1.2 Pain

As the wrist joint is in the nonfunctional position for a long period of time, abnormal development of articular cartilage will make the joint apt to wear and gradually develop traumatic arthritis, and pain will be felt in case of movement of wrist joint. With the gradual aggravation of traumatic arthritis, the pain will also become aggravated.

8.4.1.3 Wrist Joint Instability

The ulnar inclination angle of articular surface at the distal end of radial bones of patients with serious deformity is greater than 45°, the hand seems to be suspending at the distal end of the radial bones, the ulnar and radial joint has dislocation, and the wrist joint becomes unstable. Due to the wrist joint instability, the hand suffers fatigue, which becomes gradually aggravated.

8.4.1.4 Movement Disorder

The general manifestation is that the wrist joint dorsal extension and ulnar deviation are limited, the pronation and supination of forearm are limited with obvious limitation in supination, and the movement of range in wrist flexion is increased.

8.4.2 X-ray Findings

The diagnosis of Madelung deformity is mainly based on the clinical symptoms, physical signs, and X-ray examinations.

The early X-ray film indicates that the bone shaft of radial bones presents arch-shaped bending, the metaphysis takes amputation shape, and the epiphyseal nucleus is sharp and fine as if a part were cut. With the increase in age, the arch-shape bending is more obvious, the development of the distal end of the radial bone is obviously asymmetric, the development of ulnar part and volar part is poor, the metaphysis takes triangular shapes, and chondrodysplasia appears. The epiphyseal arrest of the ulnar and volar parts of the radial bone is earlier than the normal radial part, and it can be advanced by several years. The typical X-ray film can indicate that the distal articular surface of the radial bone and the osteoepiphyseal line become inclined toward the ulnar side, the angle of ulnar inclination can be greater than 45°, and the medial part has bone union; the ulnar distal end is rotated and hardened, and the osteoepiphyseal line and the articular surface become inclined toward the radial side; the proximal end of the proximal end of carpal bone changes from the curved arch-shaped circle to the apex of cusp, and the carpal bone angle can be lower than 90°. X-ray lateral film indicates that the radial bone presents the arch-shaped bending, the articular surface at the distal end of radial bones becomes inclined toward the volar side, the angle of volar inclination can be greater than 25°, the lunar bone is hidden inside, and the distal end of the ulnae becomes eminent toward the dorsal side. The degree of carpal changes is positively correlated with the severity of conditions, but the corresponding X-ray film has the above basic characteristics [8, 26, 28] (Fig. 8.10).

8.4.3 Treatment

The treatment of Madelung deformity mainly focuses on surgical treatment, and the surgery on pediatric patients is recommended to be performed after they become 12 years old. The surgical mode should be selected according to the deformity degree, lesions, and doctor's experience [8, 23].

8.4.3.1 Indications for Surgery

Wrist joint pain, obvious deformity, and limited activities are the indications of surgical treatment.

8.4.3.2 Anesthesia

Generally, brachial plexus block is selected for anesthesia. If it is unsuitable to select brachial plexus block for anesthesia because the patient is a child or due to other reasons, general anesthesia can be adopted.

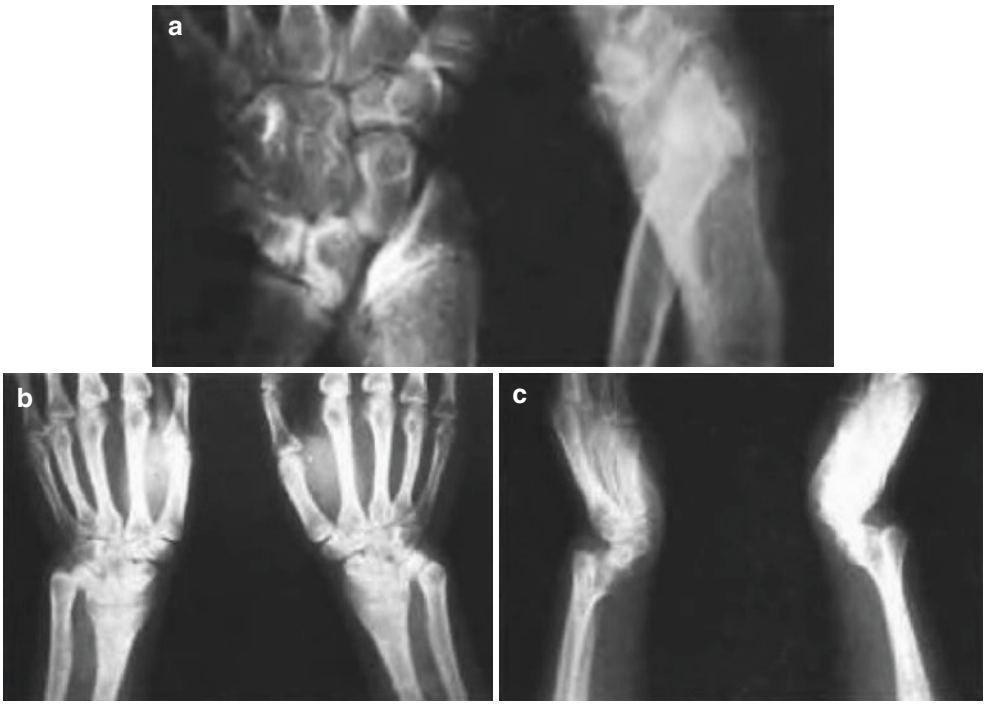

Fig. 8.10 X-ray findings of Madelung deformity. (**a**) Unilateral deformity. (**b**, **c**) Bilateral deformity

8.4.3.3 Surgical Method

1. Wrist joint lysis. Make a longitudinal incision at the site about 1.5 cm away from the proximal end of volar wrist cross striation of the forearm and the median of the upper part of wrist joint; expose the volar long muscle, radial wrist flexor, and median nerve after the skin and subcutaneous tissues are incised; pull them toward the radial side; expose the finger flexor tendon at the incision; pull it toward the ulnar side; and protect the median nerve. Perform dull separation toward the deep part, incise the distal quadrate pronator muscle, expose the lesion area of the distal end of radial bones, and at this time the abnormally hypertrophic radiolunate ligaments that connect the proximal end of lunar bone and the distal end of radial bone as well as other abnormal soft tissues can be observed. Then disjunct all of them, and then perform longitudinal resection of a small amount of cortical bone by about 5 mm by starting on the radial bone surface of the radiocarpal joint along the radial bone surface of the distal ulnoradial joint.

   When the abnormally thickened ligaments are released, attention should be paid to avoid injuring the median nerve and finger flexor tendons. When the quadrate pronator muscle is incised, attention should be paid to avoid the volar interosseous artery. Release the tourniquet, perform thorough hemostasis, and prevent postoperative hematoma formation and tendon adhesion.

   This surgery is applicable to the early lesions; the release can make the development of radial bone improved so that the deformity can be relieved.
2. Ulnar distal resection and posterior interosseous neurectomy. Make an L-shaped incision at the site 1/3 of the distal end of the forearm, incise the skin and subcutaneous tissues, perform separation along the region between the ulnar wrist extensor and the ulnar wrist flexor, and expose the ulnar distal end. At this time, the head of ulna protrudes toward the dorsal, and it is located on the deep surface of ulnar deep fascia at the proximal end of the hand dorsum. Incise the ulnar periost, perform dull separation, amputate the ulnae at the site about 4 cm away from styloid process of ulna, and resect the distal end (Fig. 8.11).

   After the ulnar distal end is resected, in order to main the stability of the proximal end of the ulna, cut the ulnar wrist flexor tendon into two halves at its insertion, take a proper length of it at the proximal side, and suture the broken end of the proximal end that encircles the ulna (Fig. 8.12).

   In this incision, find the posterior interosseous nerve distal part at the interosseous membrane of ulnar-radial bone, and resect about 2 mm of it to relieve the wrist pain. Wash the wounds, perform thorough hemostasis, and suture the wounds after the rubber strip is placed for drainage. At 24 h after the operation, pull out the drainage strip; at 2 weeks after the operation, take out the stitches and perform functional exercise.

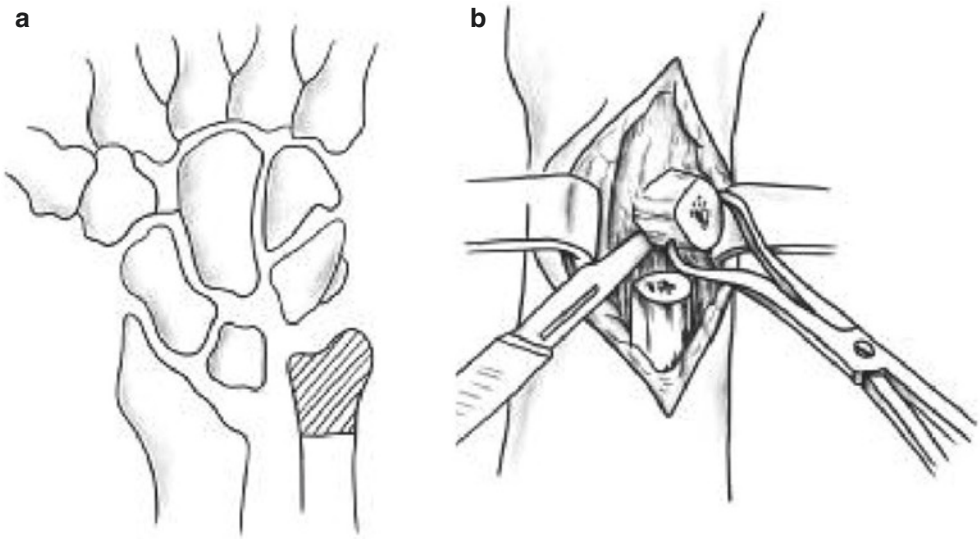

Fig. 8.11 Ulnar distal resection. (**a**) Surgical design. (**b**) Ulnar distal resection

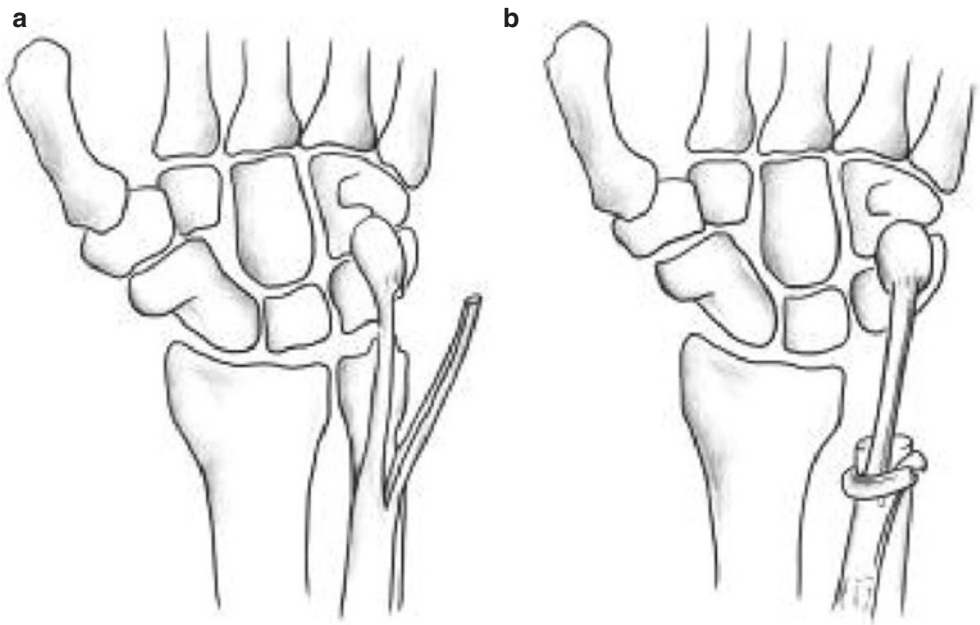

Fig. 8.12 Utilization of wrist flexor tendon strip for fixation after ulnar distal resection. (**a**) Sever the ulnar wrist flexor tendon strip. (**b**) Pass the tendon strip by the residual end of ulna for fixation

Surgical precautions: ① the amount of ulnar distal resection should not be too little; otherwise, the rotation functions cannot be improved due to insufficient resection range ; ② if the wrist pain is not serious, the posterior interosseous nerves may not be resected; ③ during the operation, do not injury the dorsal branch of ulnar nerve; ④ in case the articular surface of the radiocarpal joint has been damaged or traumatic arthritis is suffered, it is not suitable to perform this surgery, but the fusion of radiocarpal joint can be performed; ⑤ this surgery cannot improve the stability of wrist joint.

3. Ulnar distal resection and radial distal wedge-shaped osteotomy. Make longitudinal incisions at the ulnar and radial sides of the distal end of the dorsal side of forearm. Expose the ulnar distal end inside the ulnar incisions, and resect about 4 cm of it. Incise the skin and subcutaneous tissues inside the radial incisions, perform dull separation along the region between the brachioradial muscle and radial wrist long extensor, expose the radial bone, incise the periost at the site about 5 cm away from the styloid process of radius and perform dull separation, and perform wedge-shaped osteotomy below the periost. The size of the radial bone block to be cut should depend on the degree of deformity, the radial distal joint after osteotomy should be maintained in the position of volar inclination of 15°, and ulnar deviation of 30° approximately and steel plate screws are used for fixation. Wash the wounds, perform thorough hemostasis, and suture the wounds after the rubber strip is placed for drainage. At 24 h after the operation, pull out the drainage strip; at

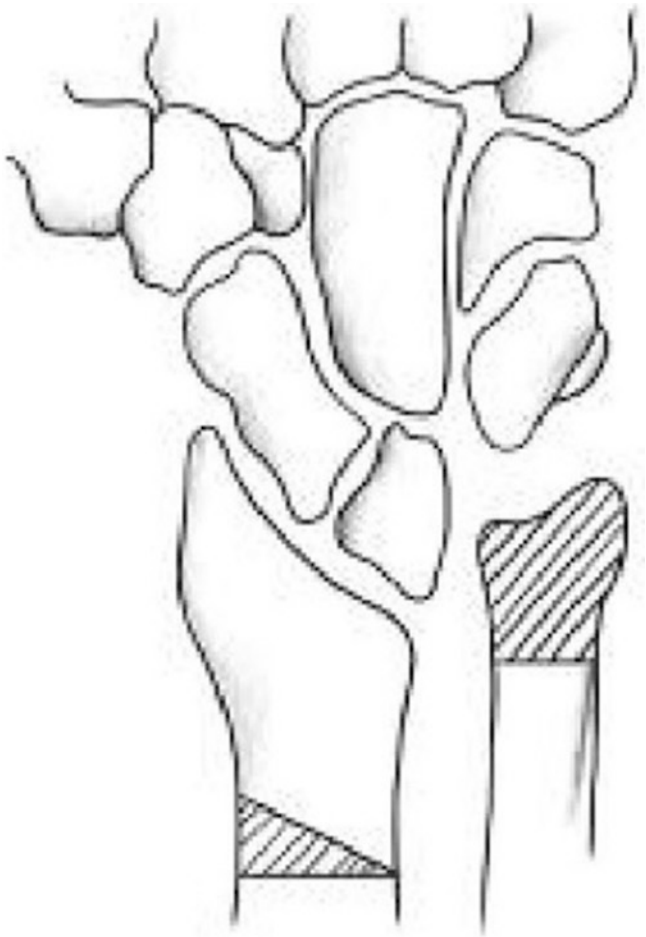

Fig. 8.13 Ulnar distal resection and radial distal edge-shaped osteotomy

2 weeks after the operation, take out the stitches; at 6 weeks after the operation, remove the external fixation, and perform functional exercise (Fig. 8.13).

Surgical precautions: ① before the surgery, properly design the angle for wedge-shaped osteotomy; ② during osteotomy, completely retain a small amount of bone cortex and periost at the ulnar side of radial bone, to avoid difficulty in surgery or radial displacement after complete amputation; ③ in case the apposition of osteotomy surface is not tight, shred the cut bone blocks and return them to the original place.

4. Radial distal wedge-shaped osteotomy and ulnar shortening. Make longitudinal incisions at the two sides of the radial and ulnar distal ends, and expose the ulnar distal segment between the ulnar wrist flexor and ulnar wrist extensor. Amputate a segment of ulna at the site 4 cm away from the styloid process of ulna, set the osteotomy volume according to the deformity degree, make the head of ulna parallel with the ulnar margin of the radial distal articular surface, and draw the proximal and distal ends of the ulna together and then fix them with steel plates [29] (Fig. 8.14). Perform wedge-shape amputation on the radial bone according to the above method, and fix it with steel plates. Wash the wounds, perform thorough hemostasis, place one rubber strip separately at the two sides, suture the wounds, and fix it with plaster after bandaging. At 24 h after the operation, pull out the drainage strip; at 2 weeks after the operation, take out the stitches; 6 weeks after operation, remove the external fixation, and perform functional exercise.

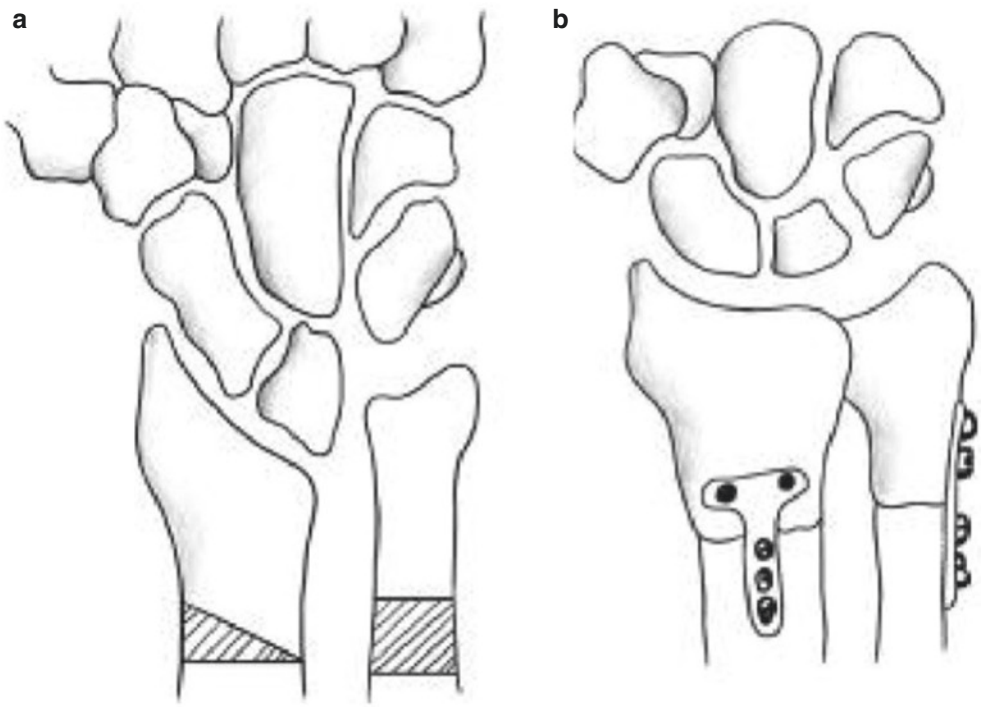

Fig. 8.14 Radial wedge osteotomy and ulnar shortening

Surgical precautions: ① after the ulna is shortened, recover the head of ulna and reconstruct the distal ulnoradial joint; ② when there is a difficulty in ulnar shortening, release the soft tissues around the head of ulna; ③ this surgery is applicable to teenagers or children; for those with insignificant protrusion of head of ulna toward the distal end, it can obviously correct deformity, relieve pain, maintain the wrist joint stability, and improve the wrist joint function; ④ for those with unstable distal ulnoradial joint, the effect of surgical reconstruction of ulnoradial joint is not exact, and the patients are apt to traumatic arthritis.

5. Distal ulnoradial joint fusion and ulnar false joint formation make a reverse L-shaped incision at the 1/3 ulnar side of the distal side of forearm, expose the ulnae along the region between the ulnar wrist extensor and wrist flexor tendon, and resect 1.5–2 cm of the ulna (along with the periost) at the site 2 cm away from the ulnar proximal end to form bone defects. Recover the proximal end of the ulnar capitulum to the radial incisure, chisel a tuberosity bone surface separately at the radial and ulnar capitulum to achieve the apposition of two bone surfaces, and fix them with screws (Fig. 8.15). Wash the wounds, perform thorough hemostasis, place one rubber strip, and suture and bandage the wounds. At 24 h after the operation, pull out the drainage strip; at 2 weeks after the operation, take out the stitches and perform early functional exercise.

   Surgical precautions: ① the ulnar defects caused during the operation should range from 1.5 to 2 cm and should not be less than that; otherwise, the ulna will reform the bone union; ② if necessary, the doctors can use the tissue flaps near the quadrate pronator muscle for stuffing to guarantee the formation of ulnar false joint to replace the distal ulnoradial joint for the purpose of reconstructing the forearm rotation functions; ③ if the distal ulnoradial joint has formed the bone union, or there is little ulnar displacement with stability, refixation may not be necessary; ④ if the angle of ulnar deviation of articular surface of the distal side of the radial bone is too large, radial wedge-shaped osteotomy can be performed simultaneously to correct the deformity and improve the angle of the radial deviation of the wrist joint.

   This method can obviously correct deformity, maintain the stability of wrist joint, eliminate pain, and improve rotation functions.

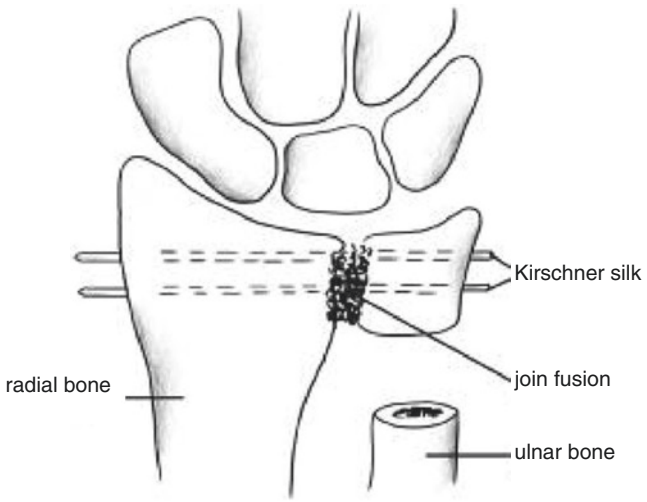

Fig. 8.15 Distal ulnoradial joint fusion and ulnar false joint formation

6. Vickers ligament release + epiphyseal plate lysis. In 1992, Vickers and Nielson [20] found abnormal ligament (Vickers ligament) was present between the volar ulnar side of epiphysis of the distal end of radial bone and carpal bone. This ligament originated from the volar ulnar side of the epiphysis of the distal end of radial bone and ended at the lunar bone and triangular fibrocartilage complex. The histological examination verifies that Vickers ligament consists of fibrous tissues and fibrous cartilage tissues. Murphy et al. found that the quadrate pronator muscle of patients with Madelung deformity had two insertions, but other scholars believed that the occurrence of these abnormal structures might be associated with the compensatory response that occurs because the soft tissues need to support carpal bone. Currently, there is no literature that can answer the question whether Madelung deformity is the initial factor or the secondary soft tissue reaction.

   Vickers ligament lysis is applicable to the Madelung deformity during the early childhood, and the purpose of this surgery lies in the recovery of growth of radial bone. Make longitudinal incision VI (Henry incision) on the surface of the radial wrist flexor tendon, pull the radial wrist flexor tendon toward the ulnar side, incise its tendon sheath, expose the quadrate pronator muscle, incise its insertion along the radial margin, and retain some soft tissues for suturing. Detach the quadrate pronator muscle below the periost toward the ulnar side, expose the thick Vickers ligament at the ulnar side of the distal end of the radial bones, and set the thickness as 5–7 mm. Disjunct the Vickers ligament, and release the constraint of soft tissues. According to the preoperative X-ray plain film and intraoperative fluoroscopy, erase the bone bridge of the volar ulnar side of distal epiphyseal plate of the radial bone, and during the operation, the injuries to the adjacent epiphyseal cartilages should be avoided. In the meantime, erase the surrounding fatty tissues or quadrate pronator muscle to prevent the occurrence of bone bridge. Repair the quadrate pronator muscle, enclose the incisions, and use the short-arm plaster cast to fix the joint for 2 weeks. At 6 months after operation, observe the growth of radial bones through the X-ray plain film.

7. Radial distal arch-shaped osteotomy + Vickers ligament lysis. For the young patients with Madelung deformity, obvious deformity, and limited potential of radial epiphyseal plate growth, the radial distal arch-shaped osteotomy + Vickers ligament lysis are adopted [21].

Under the condition of general anesthesia, place tourniquets at the forearm. Make a longitudinal incision along the distribution of radial wrist flexor tendon from the volar wrist transverse striation to the proximal side with a length of about 8 cm. Separate the gap between the radial artery and radial wrist flexor tendon to expose the quadrate pronator muscle on the deep surface. Incise the insertion of quadrate pronator muscle at the radial bone and retain partial soft tissues to repair the quadrate pronator muscle. Detach the quadrate pronator muscle toward the ulnar side, and expose radial metaphysis, thickened volar radiolunate ligaments, and triangular fibrocartilage complex. Incise the joint capsule at the radial side of lunar bone, and disjunct the Vickers ligament at the volar ulnar side of distal end of the radial bone. After the ring-shaped detachment of radial metaphysis periost, perform crescent-shaped osteotomy at the proximal side of the metaphysis and distal radioulnar joint, and the crescent arch protrudes toward the proximal side. Use the arch-shaped bone chisel to complete osteotomy through the radial volar osteotomy line, metaphysis, and dorsal cortex so that the proximal end of the bone blocks at the distal end of the radial bone takes arch shape. The bone block at the distal end of the radial bone is connected to the ulnar bone through the carpal bone and triangular fibrocartilage complex and is connected to the radial bone blocks at the proximal side through dorsal periosteum. Perform traction along the axial direction, and restore the position relation between the hand and the ulnae so that the radial bone blocks at the radial side produce radial deviation and dorsal extension. During the operation, use the thumb to squeeze the radial bone blocks at the distal side to make it displaced toward the dorsal side. After correction, use two Kirschner silk to fix them via the styloid process of radius (Fig. 8.16). Utilize the dorsoventral and lateral X-ray fluoroscopy to evaluate the position of the hand with the forearm axial line, the paraposition of osteotomy surface, and the Kirschner silk. Use a bone rongeur to remove the protruding volar bone cortices, and use them as the bone grafting materials, and staff them in the position below the dorsal periost. Perform preventive incision of the deep fascia of the forearm before the closure of the incisions, suture the quadrate pronator muscle, and close the incisions.

For the patients with serious deformity or with growth potential in ulnae, epiphysis fixation or ulnar shortening can be performed simultaneously. Make an incision between the ulnar wrist extensor tendon and wrist flexor tendon, protect the dorsal branches of ulnar nerve inside the subcutaneous tissues, and expose the epiphyseal plate at the distal end of ulnae. For those without obvious plus variant of ulnae, expose the epiphyseal plate to erase it, and stuff the gap with cortical or cancellous bone; for those with obvious plus variant of ulnae, resect the corresponding ulnae and fix them with the bone plates. The osteotomy plane should be close to the metaphysis of the ulnae, but the distal radioulnar joint should not be involved. Use two screws for fixation separately at the upper and lower regions of the osteotomy surfaces. After the operation, use the plaster cast for fixation by passing by the elbow position; after 6 weeks, first examine the union of fracture, then remove the plaster, remove the needles, and perform functional exercise.

8.5 Congenital Hand Dysplasia

Laijin Lu and Xu Gong

8.5.1 Apert Syndrome

In 1906, Eugene Apert reported nine deformed patients with similar characteristics: the forehead was wide and high, the occiput was flat, the eye span was widened, the outer canthus was lower than the inner canthus, and the submaxilla was protruding and the maxillary bone was short; the hand completely took cucullar shape; the tip became thin; the bilateral sides were symmetric; the index, middle, and ring fingers presented the bony syndactylia with only one nail; the little and ring fingers presented the simple syndactylia; the thumb and index finger also presented the syndactylia or was manifested as short and small thumb; and it bended toward the radial side. Apert named it acrocephalosyndactyly. Apert syndrome is clinically rare with an incidence of 1/200,000 [30].

8.5.1.1 Manifestations and Classification of Hand Deformities

As for Apert syndrome, hand deformity is obvious, the second to fourth phalangettes become fused to form complicated syndactylia, the nails of three fingers form synonychia, the fundus of the fourth and fifth metacarpal bones form bony fusion after the patient becomes 5 years old, but the fourth to the fifth fingers are manifested as simple syndactylia to varying extents, and the proximal interphalangeal joint of the second to the fifth fingers and the interphalangeal joint of the thumb have adhesion. Although X-ray indicates the presence of separation between the phalanges, it is actually not a joint; therefore, the fingers of the pediatric patients gradually develop the disorder of flexion from infancy to childhood. In addition to metacarpophalangeal joint and the proximal interphalangeal joint of the little finger, all other

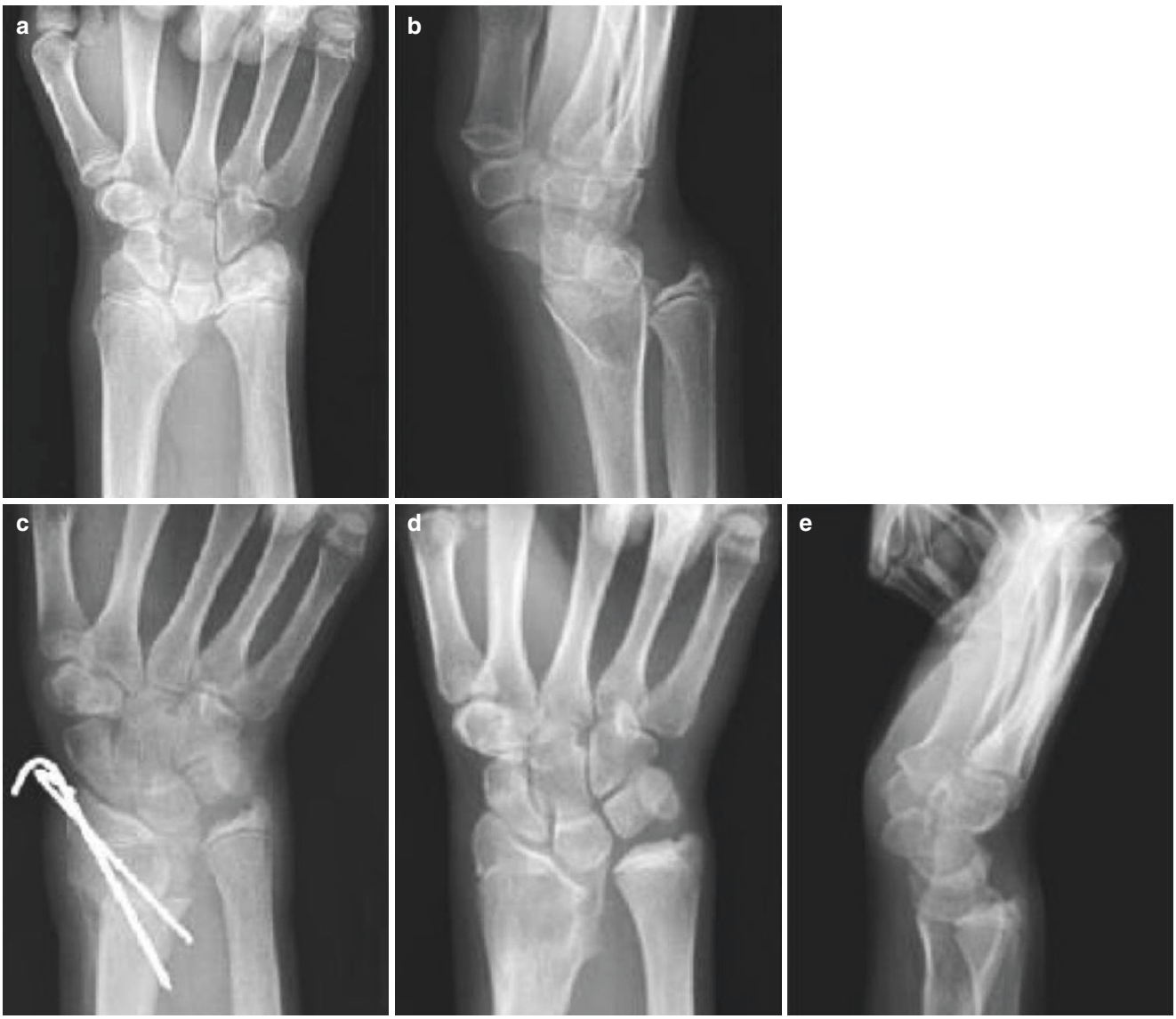

Fig. 8.16 Arch-shaped osteotomy at the distal end of radial bone. (**a**, **b**) X-ray findings before operation. (**c**) Perform arch osteotomy at the radial bone, and utilize the Kirschner silk to fix them via the radial styloid process. (**d**, **e**) Radial bone union 6 weeks after operation

joints suffer stiffness. The occurrence of hemi-closure to the epiphyseal plate of the proximal phalanx is the reason for lateral curvature deformity of index finger or little finger.

In the pediatric patients with Apert syndrome, the thumb and index finger often form syndactylia, and the metacarpophalangeal joint is always manifested as serious radial lateral curvature. Fereshetian and Upton verify that the lateral curvature deformity of the thumb is induced by the thumb short abductor having abnormal insertions at the tip of the thumb; therefore, as for the lateral curvature deformity of the thumb, the main measure is to release the thumb short abductor instead of adopting the proximal osteotomy for correction.

In 1991, Upton classified the hand deformities of Apert syndrome into three types [31]:

1. Type I. The hand takes spade shape, there is thumb web between the thumb and the index finger, but the thumb web gap is superficial; the index, middle, and ring fingers present complicated syndactylia, and the ring and little fingers present simple syndactylia (Fig. 8.17).
2. Type II. The hand takes cucullar shape; the thumb and index finger present partial or complete simple syndactylia; the index, middle, and ring fingers present complicated syndactylia; and the ring and little fingers present complete simple syndactylia (Fig. 8.18).
3. Type III. The hand takes rose bud shape, the thumb and the index finger present complicated syndactylia, the distal end has bony fusion, and one wide nail is covered at the dorsal side of the distal end of the thumb, index finger, middle finger, and ring finger; there is no bony fusion

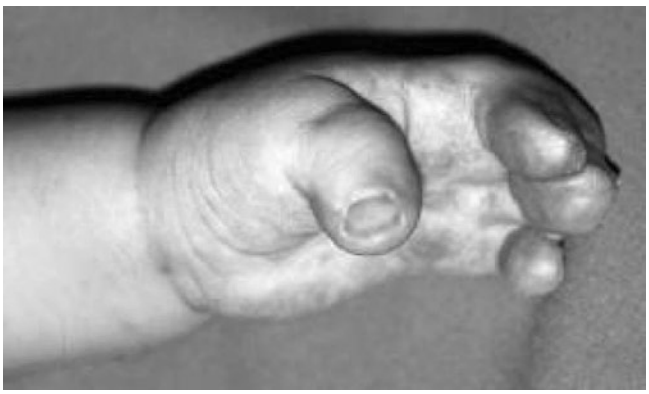

Fig. 8.17 Type I: spade hand

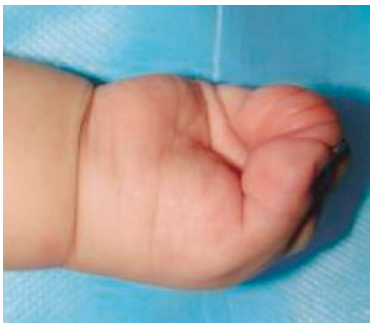

Fig. 8.18 Type II: cucullar hand

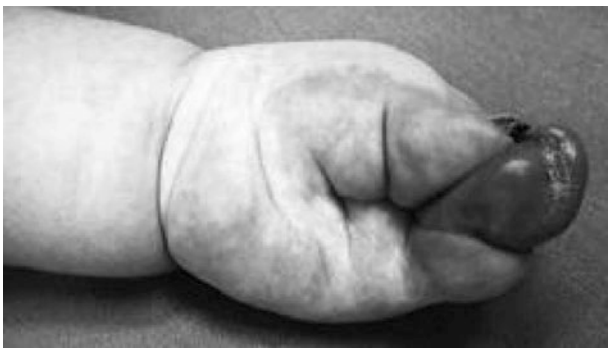

Fig. 8.19 Type III: rose bud-shaped hand

between the little finger and the ring finger, but they present complete simple syndactylia. Some patients have bony fusion at the distal end of five fingers (Fig. 8.19).

8.5.1.2 Treatment of Hand Deformity

Although the current medical technology cannot change the appearance of deformed hand of Apert syndrome as well as the stiff joint, the active correction of hand deformities can obviously improve the functions of the affected hands. Especially for type III hand deformity, in case of no correction, the pediatric patient can only perform the actions of gripping and grasping with two hands [32].

The correction of the hand deformities of Apert syndrome mainly includes expansion of the first and fourth fingerweb. Although the motor functions of the thumb are limited, the clamping between the thumb and the index finger after the thumb web is expanded is of vital importance to the functions of the affected hand. As the motor functions of the little finger of patient with Apert syndrome are optimal, expansion of the fourth fingerweb can effectively enhance the griping and grasping functions of the affected hand. The principle of syndactylous finger separating surgery of Apert syndrome is consistent with that of the ordinary syndactylous finger separating surgery; in most cases, the skin at the hand dorsum is adopted at the tissue donor site, the local flaps are designed to form thumb web or fingerweb, and full thick skin grafts are adopted for grafting to cover the lateral margins of adjacent fingers. The pediatric patients with Apert syndrome have colliquative sweating in hands; after the operation the sweat in hands immerses the skin grafting area, which often leads to necrosis of skin grafts; therefore, attention should be paid, and the treatment should be given in non-hot seasons. As for the synonychia of the second to fourth finger tips, the last segment of the finger pulp can be adopted to perform Z-plasty to form perionychium.

Correction of the lateral curvature deformity of the thumb is of no significance to the improvement of the functions of the affected hand, but it can effectively improve appearance. Currently, the method often adopted is releasing the thumb short abductor insertions at the tip of the thumb, then shortening them, and, finally, suturing them at the fundus of the proximal segment of the thumb.

The separating surgery of the stiff third and fourth fingers is of no significance to the improvement of hand functions, but it is of some significance to the improvement of the appearance. Dobyns et al. suggested removing index finger ray and increasing the thumb web distance to improve hand functions.

Correction of the hand deformities of Apert syndrome needs to be completed in several surgeries, and the recurrent syndactylia or contracture rate after the second lysis can be up to 13–18%; therefore, how to develop therapeutic strategies to reduce the surgery time(s) is of vital importance.

As for the type I and type II of hand deformities of Apert syndrome, five fingers can be retained in surgical treatment; as for type III, as the skeletal deformity is serious, it is necessary to resect one finger ray to reduce the surgery times. After the classification of deformity is confirmed, whether four fingers or five fingers are retained can be determined so that the surgery times can be limited to three.

As for type I and type II, the first surgery should be performed before the pediatric patient is 12 months old to complete the thumb web expansion of both hands, the formation of the third fingerweb, and the correction of the thumb lateral curvature deformity; the second surgery should be performed

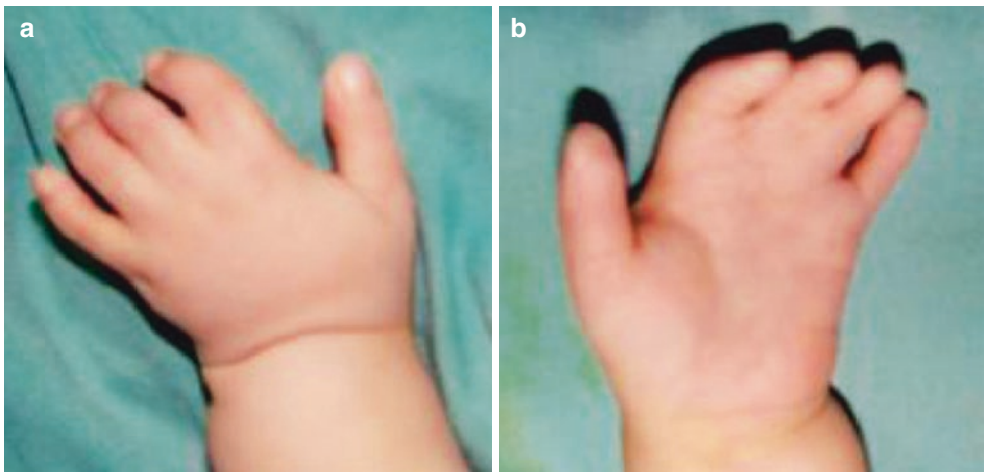

Fig. 8.20 Type III: monoarticular finger

6 months later to separate the second and the fourth fingerwebs of one hand; after another 6 months, the third surgery should be performed to complete the second and fourth fingerweb formation of another hand.

As for type III, the first surgery should be performed before the pediatric patient is 12 months old to complete the thumb web expansion of both hands, the formation of the fourth fingerweb, and the correction of the thumb lateral curvature deformity, and in the meantime, the fourth finger ray or the third finger ray should be resected; the second surgery should be performed 6 months later to finish the formation of the second fingerweb of one hand; after another 6 months, the third surgery should be performed to complete the second fingerweb formation of another hand.

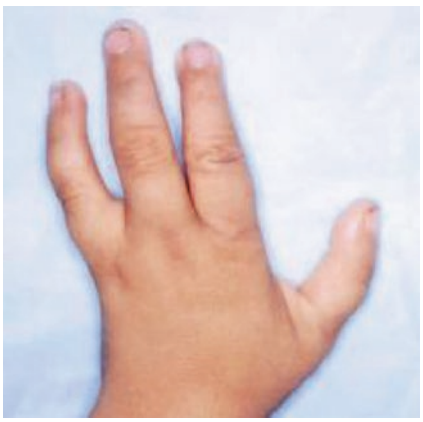

Fig. 8.21 Type IV: finger absence

8.5.2 Poland's Syndrome

Poland syndrome was first reported by Alfred Poland 150 years ago. Its incidence is 1/30,000 of live neonates. Its manifestations are dysplasia of thoracic costal head of unilateral major pectoral muscle and dysplasia of ipsilateral hand accompanied by simple syndactylia and brachydactylia [33]. Most patients have no family history, the incidence in males is higher than that of females, and the incidence at the right side is higher than that at the left side.

8.5.2.1 Manifestations and Classification of Hand Deformities

Poland syndrome can be diagnosed based on the dysplasia or absence of major pectoral muscle as well as hand deformity. Al-Qattan [34] divided the hand deformities of Poland syndrome into seven types:

1. Type I. The hand development is normal.
2. Type II. The function of the hand is normal, but is relatively small compared with the healthy side.
3. Type III. The five rays of the hand exist but suffer hand dysplasia to varying extents, the manifestations are short and small fingers and syndactylia, and the syndactylia is simple. The shortening deformity of forearm and upper arm can occur, and the shortening degree is positively correlated with dysplasia. Another feature of this type is dysplasia or absence of the middle phalanx of the fingers, and the defect degree is positively correlated with the degree of finger shortening. In severe cases, the middle phalanx of the fingers is completely absent and monoarticular finger is observed (Fig. 8.20).
4. Type IV. One-ray or multi-ray absence of the fingers (Fig. 8.21) which can be classified into five subtypes according to the degree of absence: type A, absence of radial ray finger, with floating thumb or thumb absence; type B, absence of index finger; type C, absence of index finger and middle finger; type D, absence of central ray finger, possibly with formation of cleft hand; type E, absence of ulnar ray finger.
5. Type V. All fingers are nonfunctional and take tubercle-like shape (Fig. 8.22).

Fig. 8.22 Type V: nonfunctional fingers

6. Type VI. It is the acheiria; the transverse defect plane can be beyond the wrist joint, with residue metacarpal vestige; it can also be near the wrist joint.
7. Type VII. It is the phocomelia-like deficiency.

8.5.2.2 Treatment of Hand Deformity

Type I and II hand deformities of Poland syndrome need not be treated. For type III, syndactylous finger separating surgery and thumb web deepening treatment are needed; the postoperative effects are relatively good, but to what degree the appearance is improved depends on the degree of hand development. For type IV, thumb reconstruction is needed for type A (toe-to-hand free grafting or pollicization of index finger); for type D, the hand cleft needs to be deepened to improve the gripping and grasping functions of the fingers. For type V, toe-to-hand free grafting is needed to reconstruct the finger. For type VI and type VII, artificial extremities are needed.

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Overgrowth

9

Wei Wang, Wen Tian, Ping Yao, Suting Fang,
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9.1 Congenital Macromelia

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and Jianmin Yao

The overgrowth of single or multiple fingers or in palms and limbs is manifested as abnormal growth and hypertrophy of fingers and limbs. It is called concomitantly macromelia or megalomelia, and it is one of the clinically rare upper limb deformities.

9.1.1 Etiologic Factors and Classification

Macromelia is a rare kind of congenital overgrowth deformity, whose etiologic factors have been unclear. Inglis considers that it may be caused by innervation abnormality, blood supply abnormality, and humoral system abnormality. According to some opinions, it occurs because the external factors (e.g., X-ray, hormone, and nitrogen mustard) or the lack of microelements interferes with the development of intraembryonic germ layer, and some consider that it is one type of degradation of neurofibromatosis. Macrodactylia can occur to one single finger or multiple fingers, mostly in index fingers, it is limited to one hand, and the several fingers simultaneously involved are all adjacent fingers; it can also

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occur to the toes. The incidence in males is a little higher than that in females. The severity of the deformity varies without a pattern to follow, so it is difficult to classify it. Patients with macromelia have no family history, and no abnormality can be found by the examination on the patient's chromosome [1].

Barsky and Brotherston classified congenital macromelia into static type and progressive type: the static type does not evolve with the development of the children, and the progressive type becomes enlarged disproportionately with the normal development.

9.1.2 Clinical Manifestation

The macromelia is clinically divided into two types: ① true macromelia, or called primary macromelia, and ② secondary macromelia, or called acquired macromelia. This disease can lead to symptoms such as ugly appearance, dysfunction, and nerve compression.

9.1.2.1 True Macromelia

True macromelia refers to the ordinary overgrowth and abnormal growth and hypertrophy of various finger components including the skin, subcutaneous adipose tissues, nerves, vessels, and bone tissues. It can be the overgrowth of a single finger or multiple fingers or the overgrowth of the entire limb, limb segments, or a part of the body (Fig. 9.1).

True macromelia can also be divided into normal macromelia and progressive macromelia; the former refers to the appearance of finger thickening and lengthening immediately after birth; the latter means that the finger is unnecessarily hypertrophic immediately after birth, but it becomes rapidly thickened and enlarged in early childhood. Clinically, true progressive macromelia is often seen, and macrodactylia of the index finger or the macrodactylia of more than two fingers is relatively common.

9.1.2.2 Secondary Macromelia

Secondary macromelia refers to the abnormal development and overgrowth of the limb induced by generalized or regional diseases, such as limb hypertrophy induced by hyperpituitarism and overgrowth of fingers and limbs induced by space-occupying lesions such as upper limb angioma, lymphangioma, fibroneuroma, AV fistula, and adipose tissue hyperplasia.

9.1.3 Treatment

Currently there is no effective nonsurgical method that can control the growth of macromelia, and the surgical indications include thickening and angulation of limbs (fingers), carpal tunnel syndrome, and causalgia. The purpose of surgical treatment is to reduce the difference of macromelia in length and circumference, correct the lateral curvature through fat-reducing surgery carried out in different stages, as well as highlight the retention of the sensation at the tip and the movement of the metacarpophalangeal joint. Therefore, the treatment must be individualized, and many factors should be considered, such as type of macromelia, progression, and age. As for the treatment time, it is suggested that it be given at 6 months after birth, because at this time the range and mode of deformity involvement can be objectively evaluated so that the optimal surgical regimen can be developed. The common surgical methods include the following types [2].

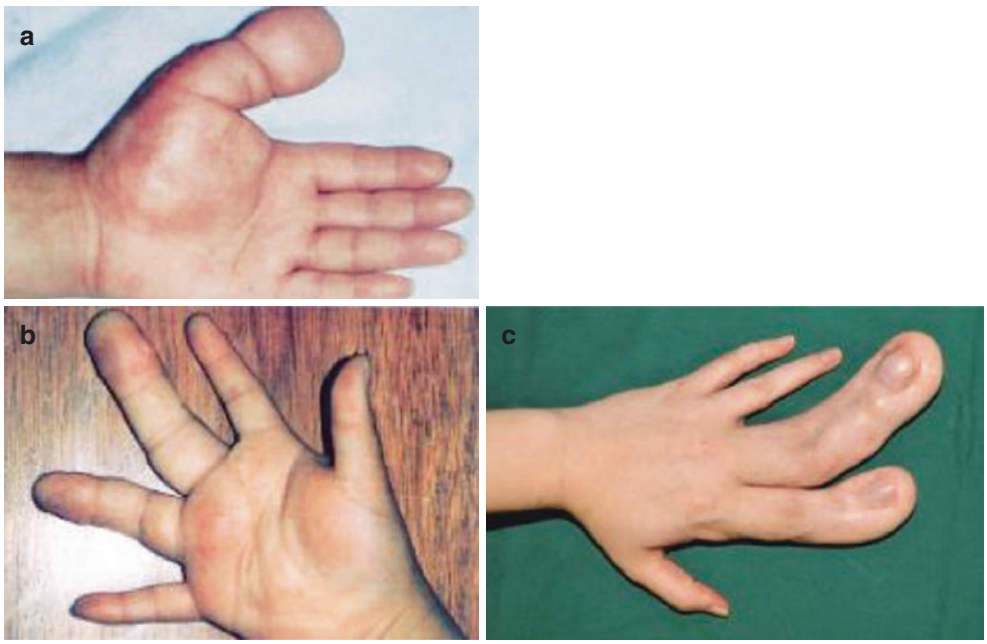

Fig. 9.1 Congenital macromelia. (**a**) Macrodactyly of thumb. (**b**) Macrodactyly of middle and ring fingers. (**c**) Macrodactyly of index and middle fingers

9.1.3.1 Soft Tissue Resection

Soft tissue resection is only applicable to patients with static macromelia, namely, those with obvious hypertrophy in the skin and subcutaneous tissues but little involvement in bones. Soft tissue resection can be used as the effective supplement for ostectomy. As for serious macromelia, soft tissue resection alone cannot effectively reduce the volume of hands and foot but only temporarily shorten them, and many patients need retreatment. Due to the possibility of tissue thickening, ugly appearance, and partial defect of functions after soft tissue resection, clinically this method is seldom adopted [3].

9.1.3.2 Plastic Shortening Surgery of Tip of the Fingers

1. Classic Barsky's method: As shown in Fig. 9.2.
2. Tsuge method: As shown in Fig. 9.3.
3. Uemura method: The classic Barsky's method and Tsuge [4] method are applicable to local finger enlargement, but both of the methods need to be implemented in two stages (during the first stage, shorten the giant finger or giant toe and fabricate the fingernail flaps with pedicles, and during the second stage, resect the superfluous skin tissues from

Fig. 9.2 Barsky's method. (**a**) Make an L-shaped bilateral median incision and remove the superfluous soft tissues at the dorsal side, the distal end of middle phalanx, and the proximal end of the distal phalanx (*shadow area*). (**b**) Prepare bone ends, such as pencil-cap-like shortening and reposition. (**c**) Restore the distal phalanx to the middle phalanx, and use Kirschner wire for fixation (Reprint with permission from ARTHUR J. BARSKY, Macrodactyly, Journal of Bone & Joint Surgery, 1967, 47:7)

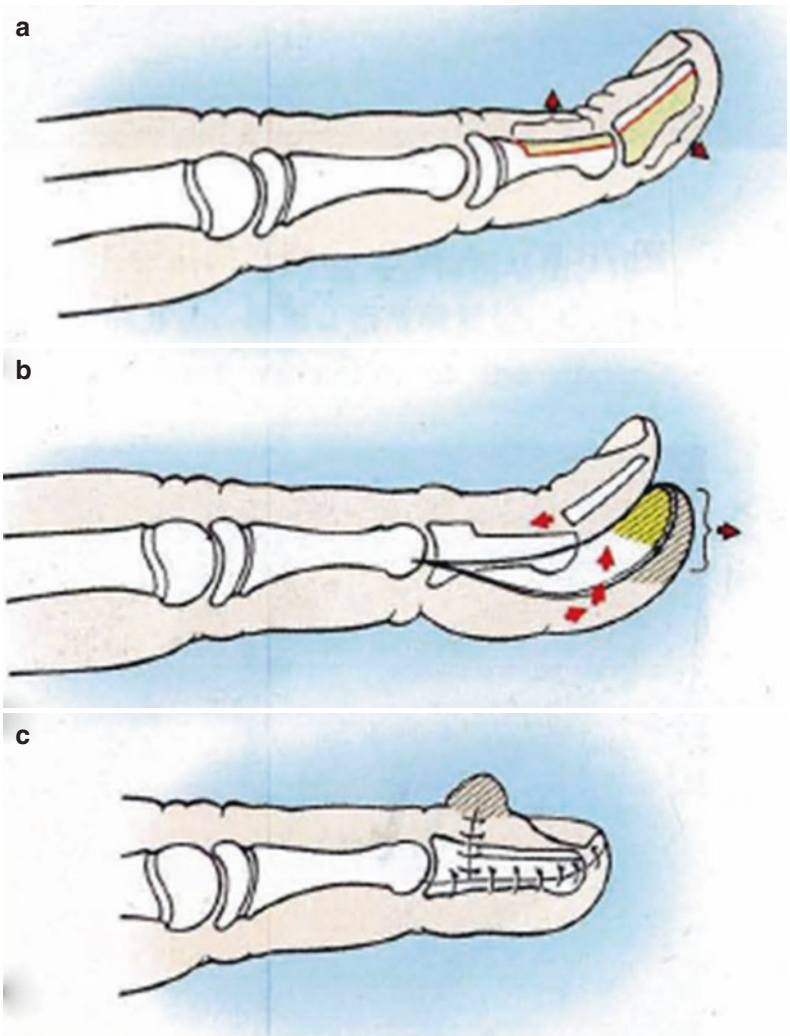

Fig. 9.3 Tsuge method. (**a**) Resect the corresponding part of the volar semi-region of the distal phalanx and the dorsal semi-region of middle phalanx (*shadow areas*). (**b**) Place the distal phalanx on the middle phalanx, retain the dorsal skin connection, and remove the superfluous tissues. (**c**) Close the wounds, and retain the supernumerary soft tissues at the dorsal side (Redrawn from Tsuge K: Treatment of macrodactyly, J Hand Surg 10A:968, 1985)

the suturing [5]); therefore, Uemura designed a method of shortening the fingernail flaps with vessel pedicles and reconstructing the fingernails that can be finished during the phase I surgery. During the phase I surgery, simultaneously resect some of the distal and unilateral fingernails and the skin on the surface of some middle finger joints; the bone shortening method is the same as Tsuge method. The advantage of this surgery is that the fingernail size can be adjusted at the doctor's will. However, this method is inapplicable to children at the stage of progression. As for serious macrodactylia, the constriction width can be increased.

4. Classic Millesi method: It can be performed on the basis of Uemura method. Resect the middle 1/3 part of the distal phalanx as well as the fingernail on its surface and the middle 1/3 part of the nail bed through the longitudinal incision at the dorsal side of the proximal and distal phalanxes, and then resect the middle 1/3 part of the proximal phalanx through parallel oblique osteotomy. Traverse the two residual longitudinal parts of the distal phalanx with Kirschner wire for fixation, shorten the distal end and proximal end of the proximal phalanx for paraposition, and use Kirschner wire for oblique fixation (Fig. 9.4).
5. Bertelli method: As the above methods cannot be used to change the transverse widening of the fingers, Bertelli designed a method of longitudinal-transverse semi-finger resection with transplantation of accessory ligaments of the interphalangeal joint of the proximal end [4, 6]. The surgical method is Z-plasty longitudinal resection of the ulnar part on the volar surface and dorsal surface, including the skin, fat tissues, and neurovascular bundle; perform longitudinal resection of about 1/3 of the articular surface and bone tissues, including the proximal, middle, and distal finger joint; when transverse shortening of giant fingers is performed, the fingertips and some fingernails will be retained. The fingertip resection includes transverse resection of the distal 1/3 fingernail level, and some volar skin is retained to cover the fingertip defects. The fingertip of the distal phalanx is not removed. The accessory ligaments harvested from the resected fingers are transplanted to the defect site of the proximal interphalangeal joint so as to guarantee joint stability. However, Bertelli [4] believes that this surgical mode can induce the bending of the distal interphalangeal joint toward the lateral side due to the cicatricial contracture.
6. Others: There are also some fingernail retaining surgical methods. For example, Sabapathy method is the method of resecting enlarged fingers or toes to make the fingernails become completely free fingernail flaps. This method is simple and can be widely applied, but the blood supply cannot be completely guaranteed, and if so, atrophy and deformation of fingernails will result; the transplantation of fingernails with vessels as reported by Koshima [7] needs the anastomosis with vessels and the technical requirements are high, but good blood supply can be guaranteed.

The above several surgical methods have advantages and disadvantages, so individualized treatment must be adopted in clinical work and the optimal surgical methods must be selected; in the meantime, the patient's will must be fully respected. The surgeon must know whether the patient actively requires retention of fingernails and is willing to receive phase II surgery.

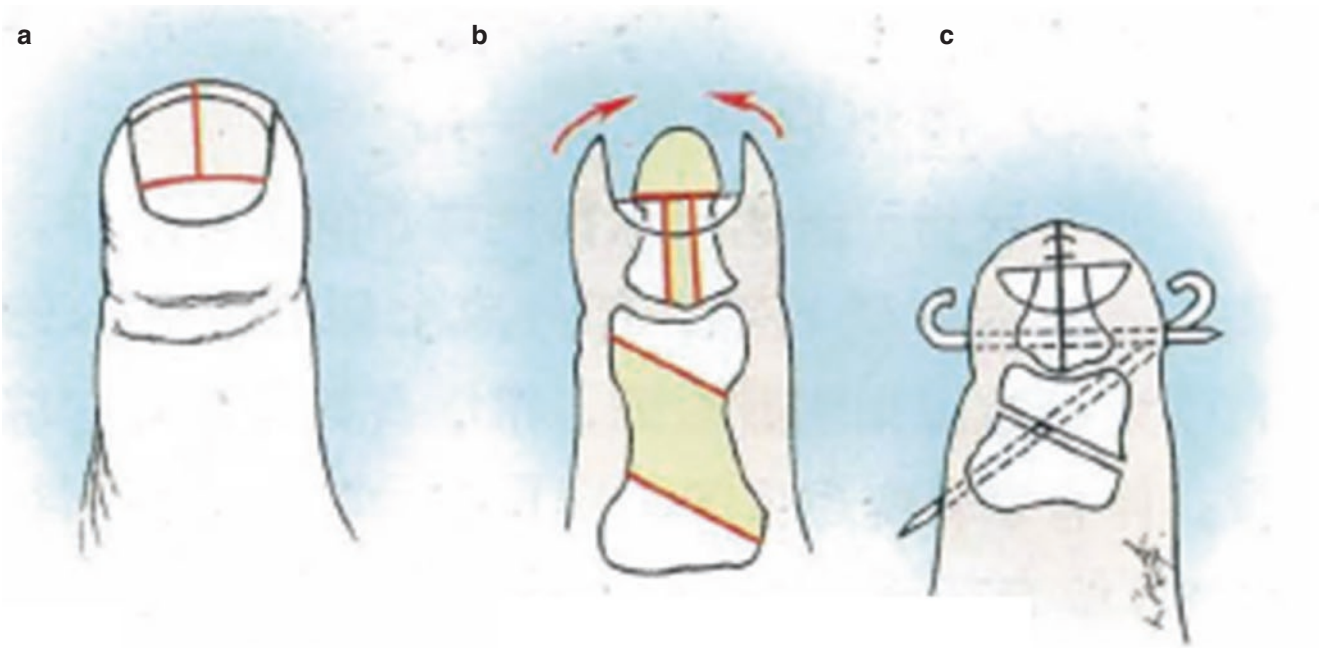

Fig. 9.4 Millesi method. (**a**) Resect the distal parts of the phalange and fingernails, and retain the nail bed. (**b**) Perform shortening and osteotomy through the dorsal side. (**c**) Shorten the residual bones, and fix them with Kirschner wire (Redrawn from Millesi H: Macrodactyly: a case study. In Little JW, Cramer LM, Smith JW: Symposium on reconstructive hand surgery, St Louis, 1974, Mosby)

9.1.3.3 Epiphyseal Arrest

The epiphyseal arrest is applicable to progressive macrodactylia to block the longitudinal growth of bones (including metacarpal bone and metatarsal bone) [8, 9]. During the operation, expose the proximal, middle, and distal phalangeal epiphyseal plates; use the high speed drill, curette, or fulguration technique to fuse the epiphyseal plates; and use splints for fixation for 3 weeks after the incisions are closed. The unilateral epiphyseal arrest can be used for the correction of lateral curvature; wedge-shaped osteotomy can also be utilized to treat the lateral curvature, and currently this method is often clinically used. The osteotomy can be simultaneously performed together with the joint fusion [10].

9.1.3.4 Nerve Resection and Decompression

Tsuge believes that finger nerve hypertrophy is the reason for macrodactylia, so resection of finger nerve is effective, and the surgery performed during childhood has little influence on the nerve functions. He emphasizes that partial or total resection of involved hypertrophic nerves is not very effective. During resection of hypertrophic nerves, the excessive fatty tissues around them should also be removed, so reduction of the volume of the giant finger is an effective measure. In order to retain the sensory function, only the finger nerve branch is resected, the nerve trunk is retained, and all branches are dissociated from the nerve trunk and resected together with the fatty tissues. If the finger nerves become obviously thickened, half of them should be resected. If there is excessive nerve bending of fingers, first perform segment resection according to the Kelikian method and then perform end-to-end anastomosis. Resect the volar half of the distal phalanx and the dorsal half of the middle phalanx, and then superpose the residual bone flaps until paraposition is achieved. However, this method is still controversial. Kalen et al. [11] reported that finger neurectomy was unnecessary. Minguella et al. [12] reported that partial finger neurectomy did not produce any effect on the treatment of three patients with macrodactylia. Even so, we still believe that resection of hypertrophic nerve branches and soft tissues is one of the methods to treat macrodactylia.

When the macrodactylia is accompanied by loss of control over the growth of the median nerve, which result in the compression of median nerve at the carpal canal, release of carpal canal can decompress the nerves [13]. Yoshida [14, 15] recently reported the utilization of endoscopic technique for the release and decompression of the carpal canal to treat the carpal tunnel syndrome of patients with macrodactylia, which produced good effects.

9.1.3.5 Osteotomy

The osteotomy of phalanges can be used to shorten bones, and it is one of the commonest clinical methods. Tsuge [16] also recommends utilization of wedge-shaped osteotomy to correct angulation deformity. Tan et al. [3] designed the shortening surgery of removing the joint from the middle phalanx, during which the middle phalangeal joint was removed, and the interphalangeal arthroplasty was utilized to connect the proximal and distal phalanxes. As the two are anatomically similar, the difference in the size of joint surface is not significant, and the sufficient lacuna can form functional joints. The flexor tendons and extensor tendons are shortened to achieve good joint activities. The nerve vascular bundles must be retained, but the supernumerary soft tissues can be removed. This method can retain the basic functions of fingers such as lifting, pinching, and grasping, but the appearance is good. However, this method is inapplicable to thumbs and hallux.

9.1.3.6 Dactylolysis

Dactylolysis is applicable to the giant fingers that are too large, lose their functions, and affect the functions of other fingers, but dactylolysis means sacrifice of the entire finger (toe). This radical method will lead to serious appearance defects and functional loss [8, 17]. In the meantime, we believe that, due to the various shapes of the affected fingers (toes), even the radical operation cannot ensure beautiful appearance, and dactylolysis cannot guarantee local tissues no longer grow, so a second surgery may be needed. Because the resected fibrofatty tissues affect the local blood supply after the intraoperative utilization of skin flaps to wrap the residual ends, the postoperative wound healing is often delayed (according to the reports, 4 of 7 experienced delay in healing); therefore, dactylolysis is often the method that will be eventually adopted, and careful consideration and full doctor-patient communication must be conducted.

9.1.3.7 Radiatiform Resection

The radiatiform resection of phalanges covers the hypertrophic metatarsal bones as well as the bone tissues and the surrounding soft tissues such as proximal, middle, and distal interphalangeal joints; the surgery can effectively shorten the enlarged metatarsal bone and correct the increased width and height of toes, and it is applicable to the patients with macrodactylia in toes rather than the big toe involving metatarsal bones. The phalangeal distal resection or epiphyseal backbone formation can only shorten toes but is of no help to the correction of excessive width and height of the toes, so radiatiform resection can produce better beautifying effects than toe amputation at the level of the metatarsophalangeal joint. When the patient's metatarsal extension angle is over 100°, radiatiform resection can be performed. As for the macrodactylia of the big toe, surgical resection is not suitable because the big toe and the first metatarsal bone play an important role in weight loading and maintenance of normal gait. When the macrodactylia of the big toe only involves one side [13], it is recommended that the phalangeal and metatarsal backbone shortening be performed repeatedly. Metatarsal epiphysiodesis is another option, and it is recommended that the epiphysiodesis be performed when the metatarsal bone develops into the size of a normal adult [8, 18, 19].

9.2 Congenital Unilateral Limb Myogenic Hypertrophy Syndrome

Congenital unilateral limb myogenic hypertrophy syndrome is a clinically rare type of congenital disease with the main manifestation of hypertrophy in one side of the upper limb or lower limb and the obvious symptom of hand hypertrophy. The main morphological changes of unilateral upper limb myogenic hypertrophy syndrome are the number of skeletal muscles increases and volume increases, the deformity of hand muscles is more serious, the hand internal muscles and external muscles are both involved, and the appearance and functions of the hand are seriously affected. Similar deformities are also found in the lower limb, but the incidence is lower than that in the upper limb. Up to now, the author treated the patients with such deformities that were diagnosed and treated in the center; 13 patients suffered the disease in the upper limbs and 2 patients suffered the disease in the lower limbs. This deformity was first reported by a Japanese scholar named Mizuoka in 1962 [15]. After the author reviewed the literature, there have been 31 reports up to now [15, 20–28]. The bony structures of deformed upper limbs are enlarged to varying extents, and the deformities of muscular tissues are relatively mild. As the deformities in the bony structures of the forearm and upper arm are mild, the functional disorder in shoulders and elbow joints is mild. The main reason for serious hand functional disorder is accumulation of a large quantity of abnormally hypertrophic muscles, as well as abnormality in the volume, quantity, and distribution, which leads to abnormal hand osteoarticular movement and dynamic mechanism and produces a series of hand deformities. When the hands are used, incoordinate abnormal contraction occurs between the variant muscles, leading to mutual antagonism, counteraction of effects, and changes in stress directions so that the hand functions cannot be performed normally.

This congenital disease has other different names, such as vasovagal muscle syndrome and accessory muscle syndrome; the author also calls it windblown hand deformity or atypical windblown hand deformity. Currently, it is a kind of independent congenital limb deformity with its own morphological characteristics. Its exact pathogenesis is still unclear, and the patient has no family history of genetic diseases.

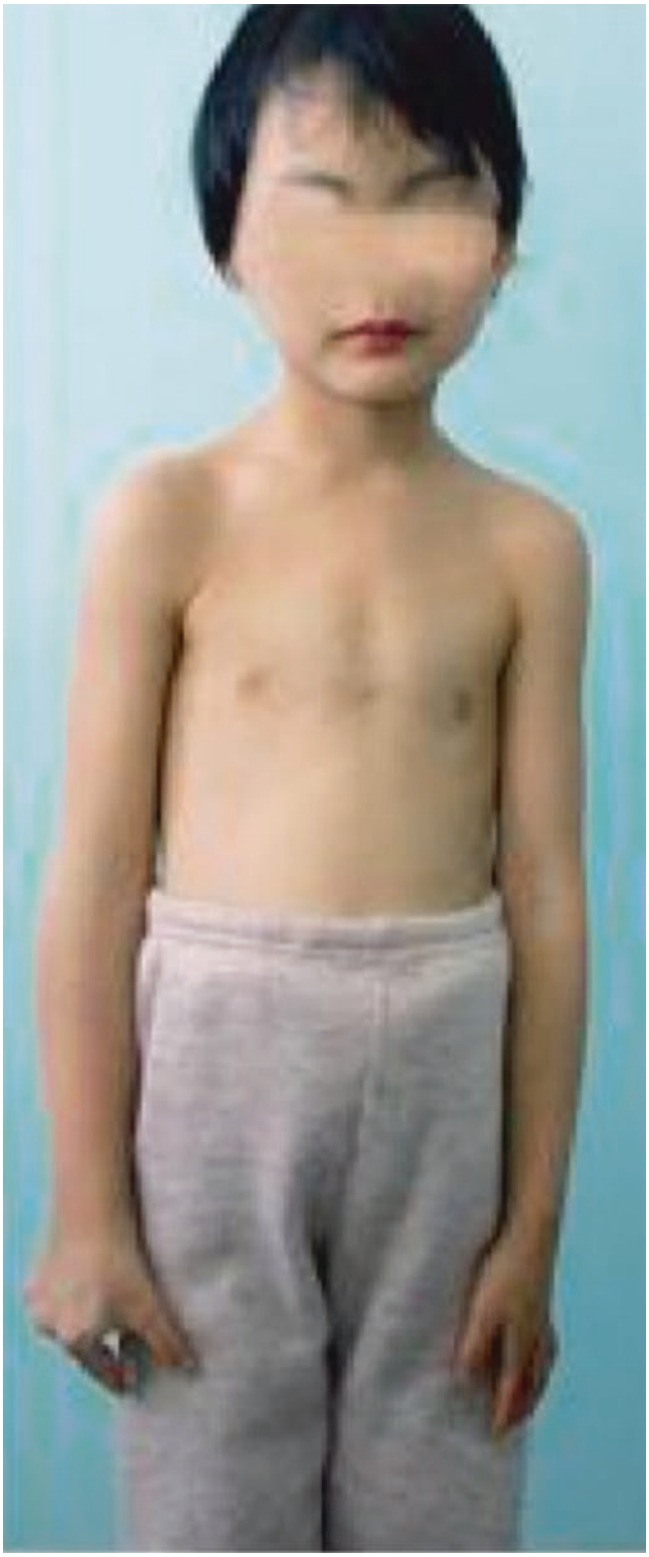

Fig. 9.5 Congenital unilateral upper limb myogenic hypertrophy syndrome (right side)

9.2.1 Congenital Unilateral Upper Limb Myogenic Hypertrophy Syndrome

9.2.1.1 Clinical Manifestation

The main manifestations of congenital unilateral upper limb myogenic hypertrophy syndrome (Fig. 9.5) include: ① the upper limb hypertrophy exists at birth; ② the upper limb hypertrophy is unilateral (flexion-extension side), the hand hypertrophy is more serious than that in the upper arm and forearm, and the increase is nonprogressive; ③ the hand functions are subject to serious influences.

9.2.1.2 Morphological Characteristic

1. When the fingers are extended by force, the fingers are abducted excessively (Fig. 9.6).
2. The thumb is excessively abducted to the radial and volar side, and the thumb web is extremely wide (Fig. 9.7).
3. The palm becomes widened (Fig. 9.8).
4. The metacarpophalangeal joint has ulnar deviation and flexion (similar to windblown hand), with severe symptoms in index and middle fingers, and the metacarpophalangeal joint also has radial deviation under the dynamic status (Fig. 9.9).

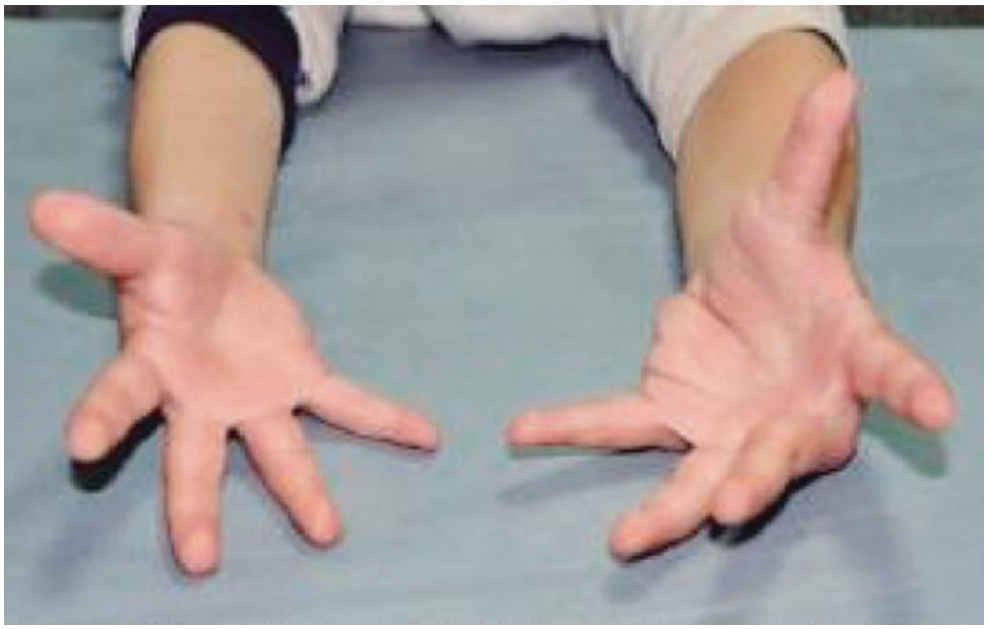

Fig. 9.6 When the affected (left) hand is extended, the abduction amplitude of the fingers is obviously enlarged when compared with the healthy side

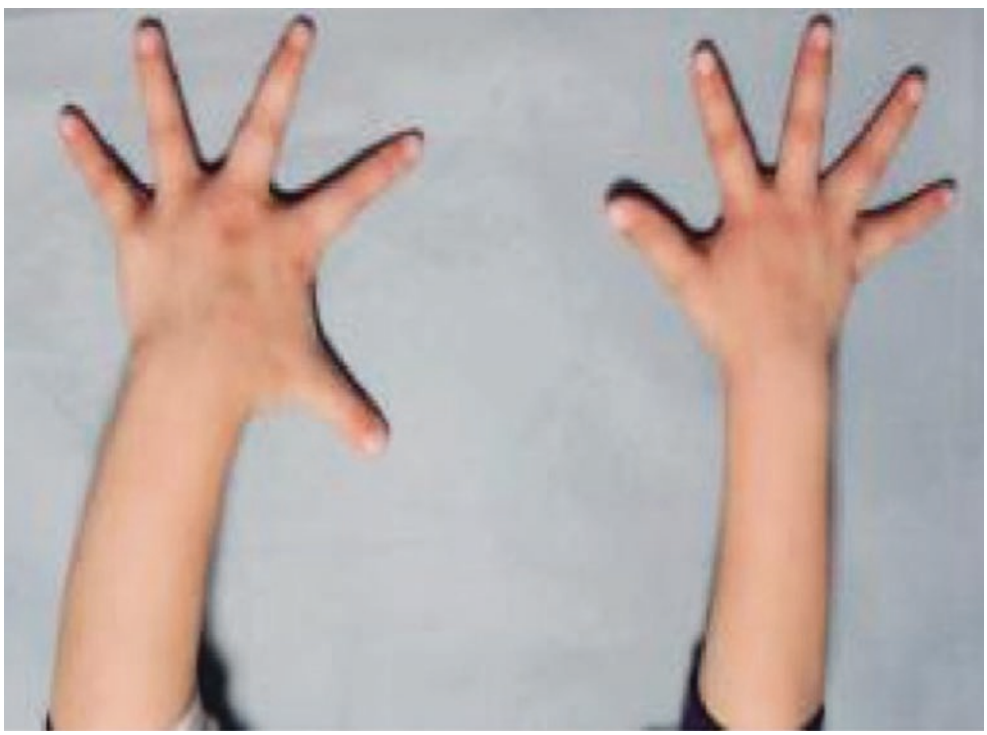

Fig. 9.7 The thumb of the affected (left) hand becomes abducted to the radial side, and the thumb web is wide

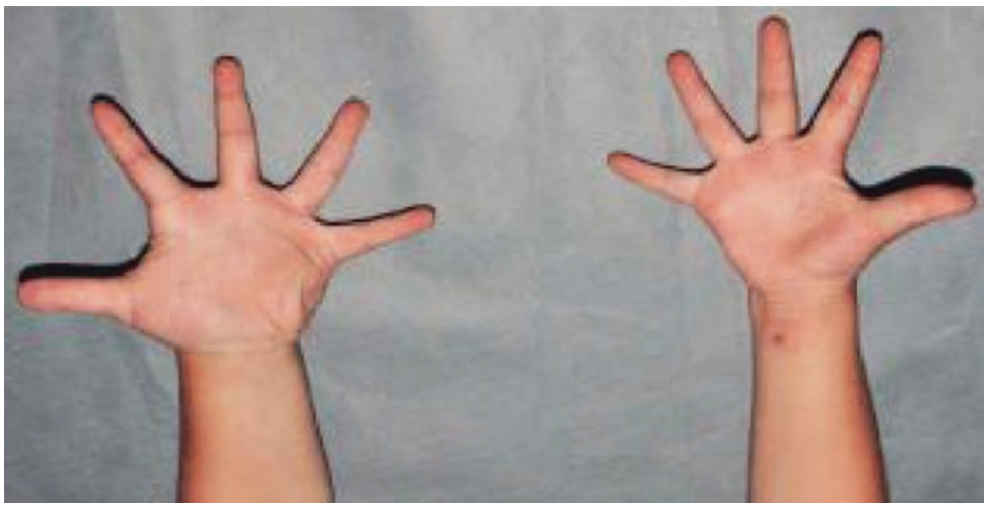

Fig. 9.8 The palm of the affected (left) hand is wide

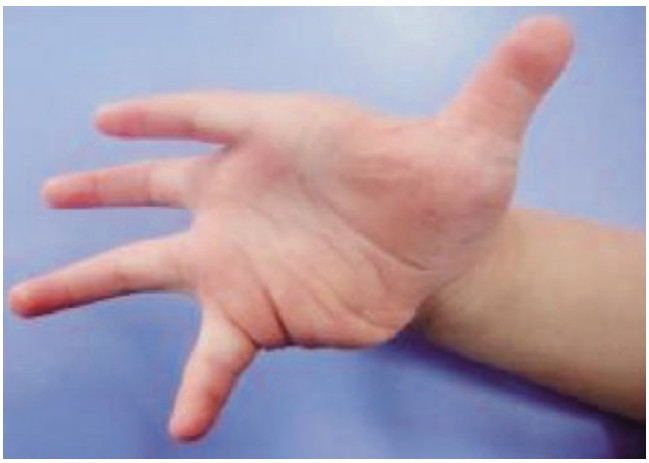

Fig. 9.9 The metacarpophalangeal joint of index, middle, ring, and little fingers have ulnar deviation and flexion

9.2.1.3 Basis of Diagnosis

1. With obvious morphological characteristics.
2. In addition to changes in appearance, the hand functions are obviously affected (Fig. 9.10).
3. Rule out upper limb hypertrophy induced by diseases such as primary macromelia (Fig. 9.11), Oller disease (Fig. 9.12), angioma (Fig. 9.13), neurofibromatosis, Proteus syndrome (Fig. 9.14), and windblown hand deformity (Fig. 9.15).
4. The surgical exploration can indicate the presence of increased number, enthesis, abnormal distribution, and internal muscles and external muscles with enlarged volume (Fig. 9.16).

9.2.1.4 Other Morphological Manifestations

1. Increase in skin folds, and no regularity in distribution (Fig. 9.17).
2. Abnormal palm prints (Fig. 9.18).
3. Relaxation of metacarpophalangeal joint accessory ligaments (Fig. 9.19).
4. The central tendon bundle is unstable, and the finger extension with excessive physical exertion can lead to flexion and radial deviation of the metacarpophalangeal joint.
5. The metacarpophalangeal joint of the index finger has obvious ulnar deviation, resulting in crossing deformity of the index and middle fingers (Fig. 9.20).

9.2.1.5 Imaging Examination

1. X-ray findings
   (a) The humeral bone, radial bone, ulnar bone, metacarpal bone, and phalanges is slightly thickened, and changes in length are not great (Fig. 9.21).

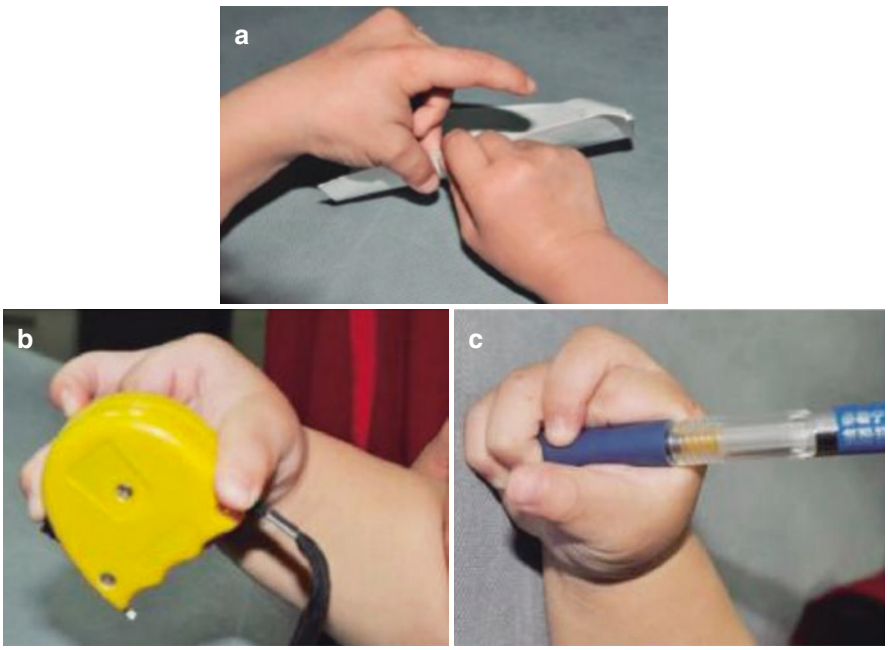

Fig. 9.10 Hand functional disorder. (**a**) Object pinching functional disorder. (**b**) Object grasping functional disorder. (**c**) Pencil gripping functional disorder

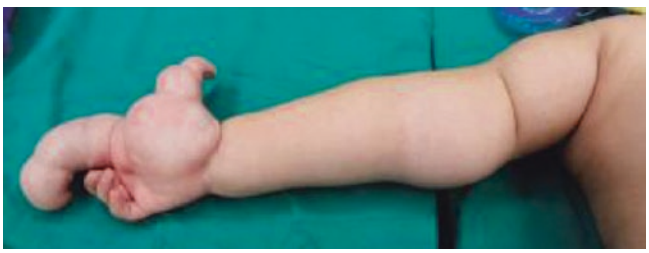

Fig. 9.11 Primary macromelia

(b) The gaps among the heads of metacarpal bone are thickened, especially the gap between the head of the second and the third heads of metacarpal bone. In addition, shadow of muscle soft tissues appears (Fig. 9.22).
(c) Subluxation of metacarpophalangeal joint (Fig. 9.23).
(d) Abnormal shape of the head of metacarpal bone (Fig. 9.24), induced by volar epiphysis dysplasia.

2. MRI manifestations: There is wide hypertrophy of internal and external muscles; the muscle volume increases; the upper arm, forearm, and hand are involved; and the muscle imaging signals are similar to those of the healthy side (Fig. 9.25).

9.2.2 Congenital Unilateral Lower Limb Myogenic Hypertrophy Syndrome

The incidence of unilateral lower limb myogenic hypertrophy syndrome is far lower than that of the upper limb. Similar to the upper limb, the morphological hypertrophy of the foot is more obvious than the thigh and shank, but the

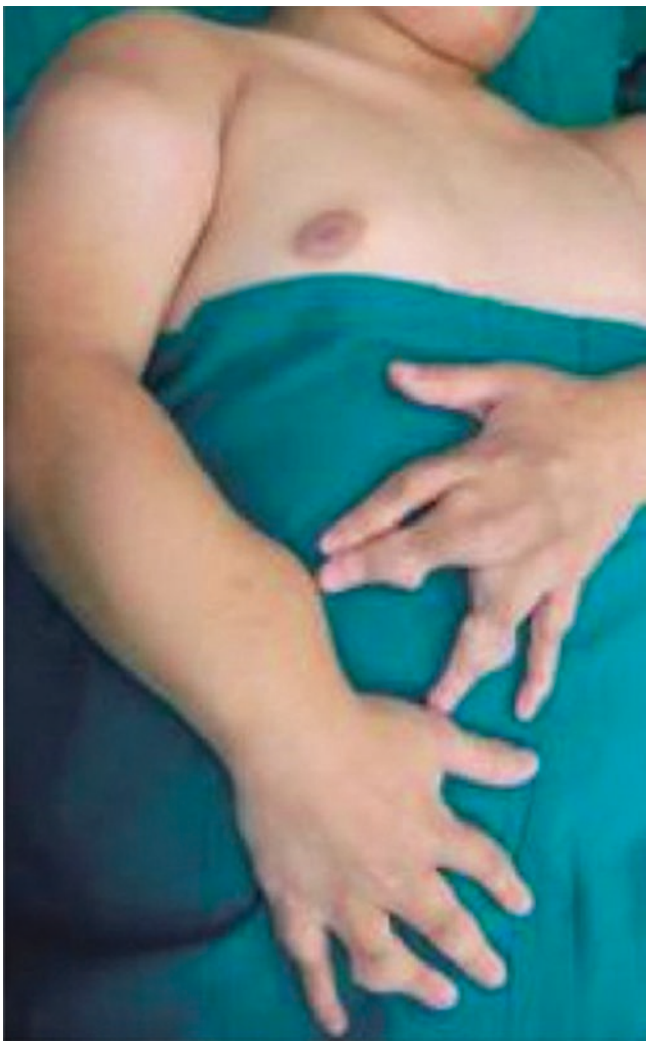

Fig. 9.12 Oller disease

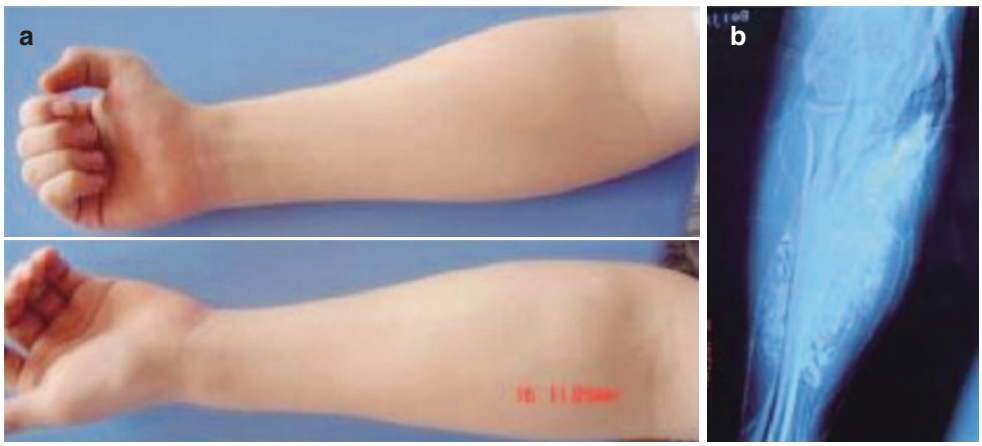

Fig. 9.13 Hemangioma. (**a**) Left upper limb angioma. (**b**) Findings of angiography

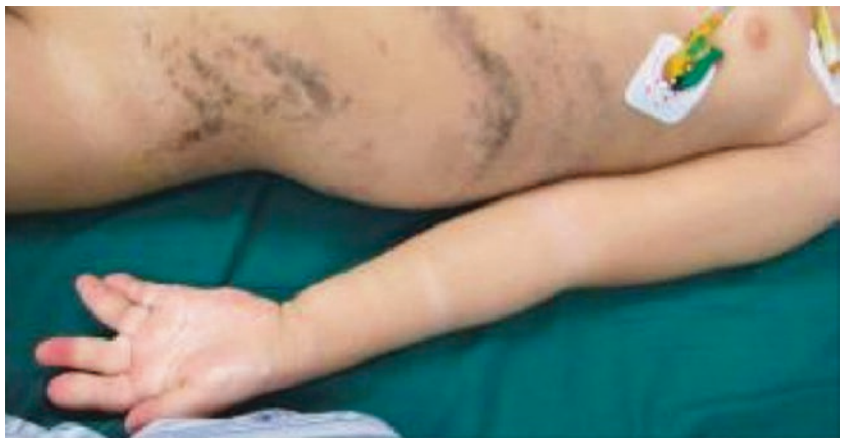

Fig. 9.14 Proteus syndrome

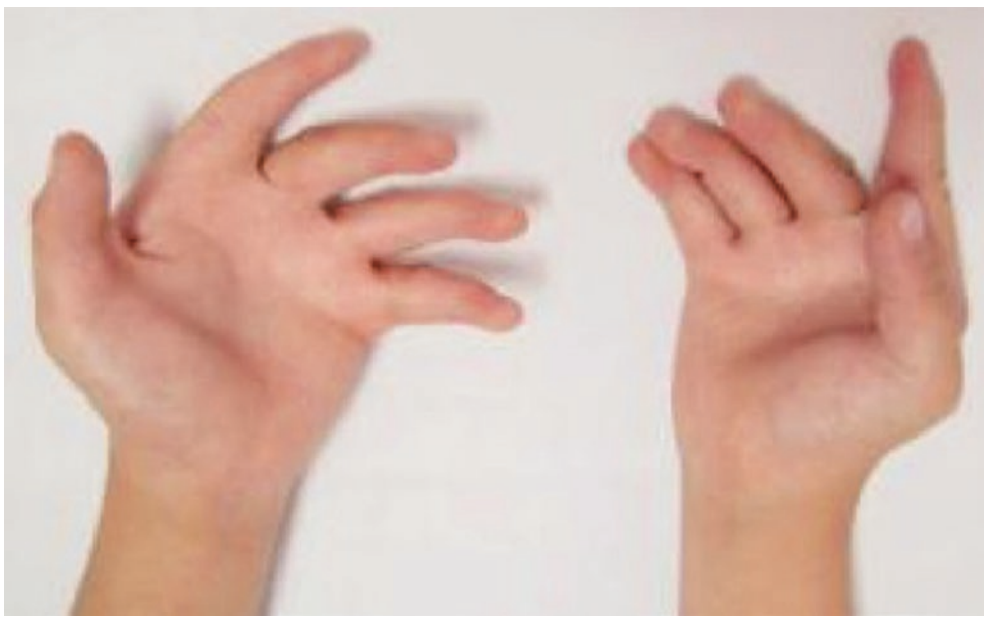

Fig. 9.15 Windblown hand deformity of both hands

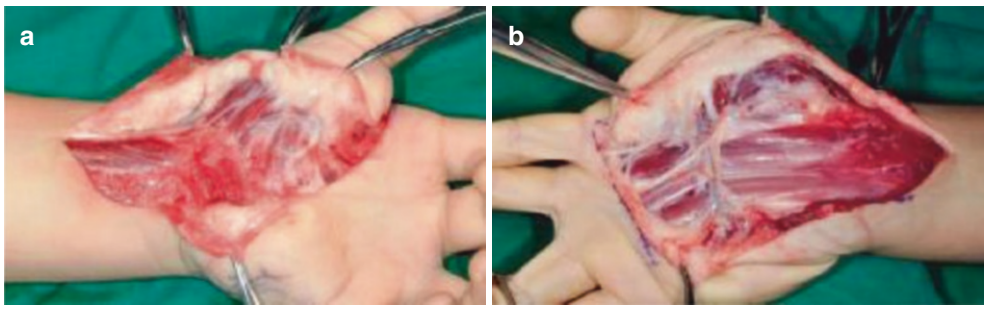

Fig. 9.16 Surgical exploration findings. (**a**) A large quantity of abnormally hypertrophic internal muscles and external muscles could be seen in the palm, carpometacarpal part, and forearm. (**b**) The abnormal muscles that stem from the forearm and end at the fingers can be seen in the hands

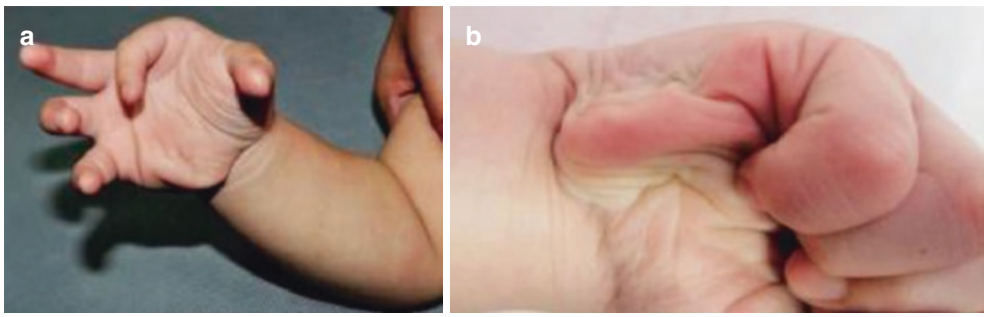

Fig. 9.17 Increase in skin folds. (**a**) Increase in hand skin folds. (**b**) In contraction of hypothenar muscles, a large number of irregular and messy wrinkles are visible in the skin

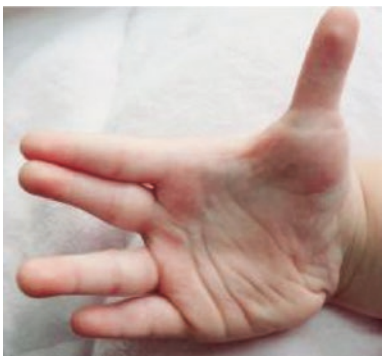

Fig. 9.18 Abnormal arrangement of palm transverse striation

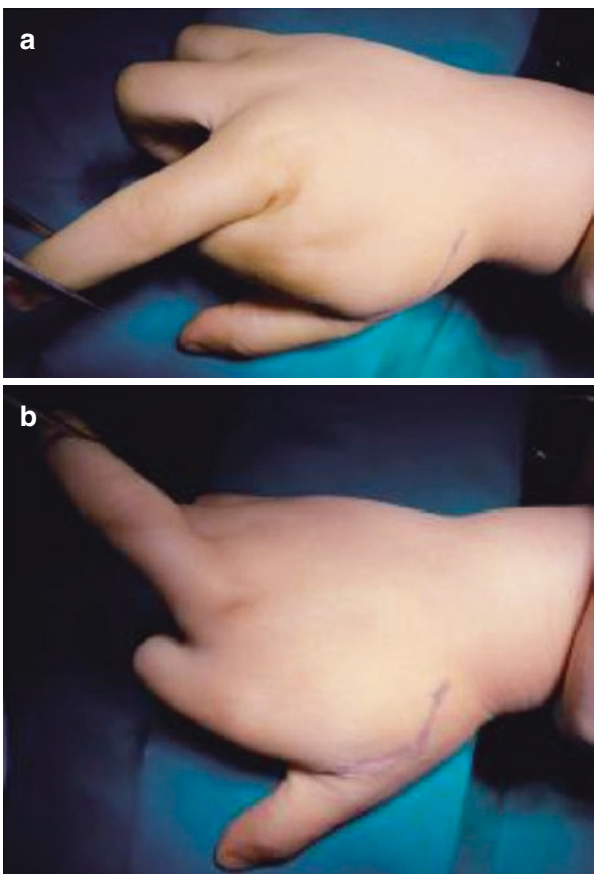

Fig. 9.19 Relaxation of metacarpophalangeal joint accessory ligaments. (**a**) Relaxation of ulnar accessory ligaments of metacarpophalangeal joint. (**b**) Relaxation of radial accessory ligaments of metacarpophalangeal joint

influence of the deformity on functions is not as serious as that on the upper limb. As the sole of the foot has the filling of a large quantity of muscular tissues with abnormal growth, the instep disappears or becomes superficial. Slight instability is felt in quick walking or running, or obvious sensation of weakness is felt after walking for a long period of time (Fig. 9.26).

9.2.3 Treatment

As it is very rare, the law of aberration of congenital unilateral limb myogenic syndrome is not very clear, and the knowledge on the exact etiologic factors and biomechanical mechanism is limited; therefore, currently the corresponding treatment regulations have not been formed, and the uniform treatment principles have not been established. Currently, a certain surgical intervention is given according to the treatment intentions of the patient's family and the knowledge on

deformities; only the surgical resection to varying extents can be performed on the variant muscles, and there is almost no possibility in anatomical resection. The purpose of treatment should be to recover certain hand functions and improve the appearance. Patients need to receive multiple different surgical treatments so that they can obtain limited morphological and functional modification. The author once paid follow-up visits with a mean duration of 2.5 years on 12 patients, and the results indicated that: the thumb radial abduction decreased by 25° on average after surgery; a total of 12 fingers were involved in the corrective surgery of lateral deviation of the metacarpophalangeal joint and four fingers experienced recurrence; a total of seven sites were involved in the metacarpal rotary ostectomy and all of the fractures were healed; and a total of 11 patients with flexion deformity in metacarpophalangeal joint were corrected, with an average decrease of 12°. The family's satisfaction degree on postoperative appearance was 60%, and the dissatisfaction degree was 40%; the satisfaction degree on postoperative functions was 50%, and the dissatisfaction degree was 50%.

The surgical treatment methods adopted by the author are as follows.

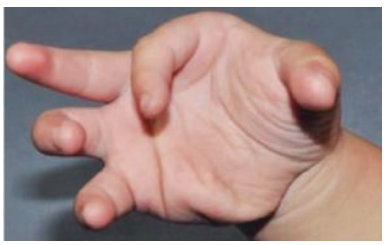

Fig. 9.20 Crossing deformity of index and middle fingers

9.2.3.1 Partial Resection of Variant Muscles

The partial resection of variant muscles and superfluous skin is performed to improve the appearance of thenar and hypothenar muscles. Shorten the abnormally wide thumb fingerweb, and relieve the pinching and grasping functional disorder induced by the excessively wide thumb fingerweb (Fig. 9.27).

9.2.3.2 Resection of Hypertrophic Muscles of Palm and Forearm

The hypertrophic muscles with a large volume, a large quantity, and abnormal distribution become accumulated at the palm, wrist, and forearm, leading to abnormalities in hand osteoarticular movement and dynamic mechanism; incoordinate abnormal contraction occurs between the variant muscles, leading to muscular mutual antagonism, counteraction of effects, and changes in stress directions so that the hand shape experiences aberration and the hand functions cannot be completely normal. A large quantity of muscle tissues become accumulated at the wrist volar side, affecting the hand pinching and griping functions. Limited partial resection of aberrant muscles can effectively reduce the influence of aberrant muscles on the functions (Fig. 9.28).

9.2.3.3 Release the Skin Soft Tissues of the Palm, and Perform Transplantation with Free Skin Grafts

During the early stage, as the author has insufficient knowledge on the aberration principle of this syndrome, the common practice is to correct the metacarpophalangeal joint ulnar deviation and flexion through the resection of skin soft tissues, and in the meantime resect some skin tissues of the thumb fingerweb to

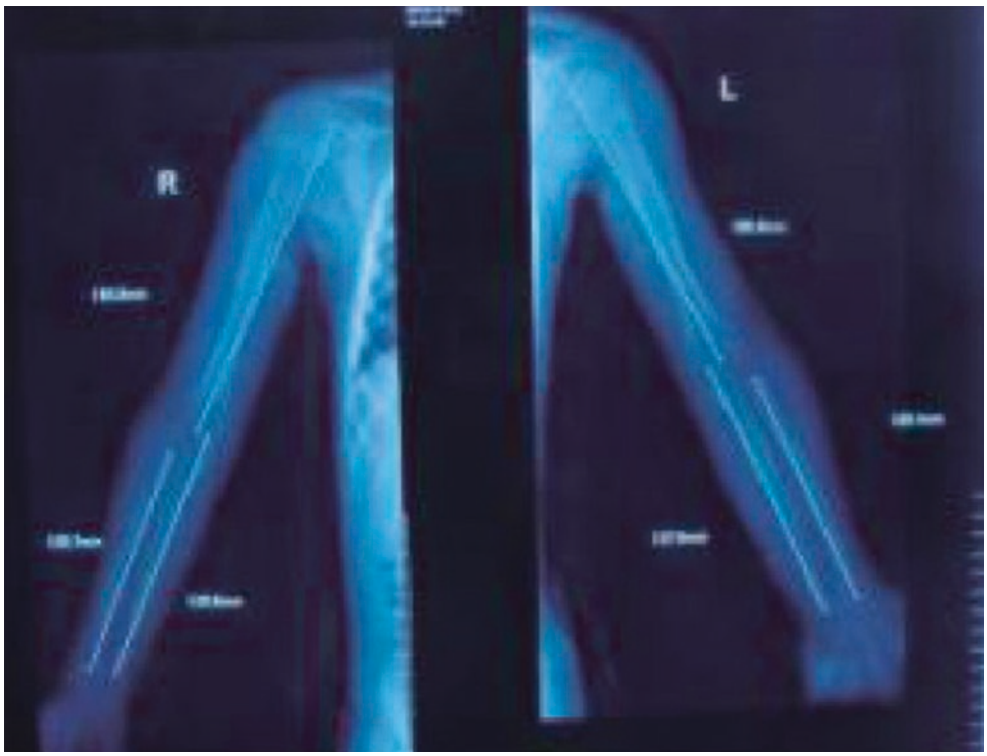

Fig. 9.21 Morphological comparison of upper limit joint of healthy and diseased (left) sides

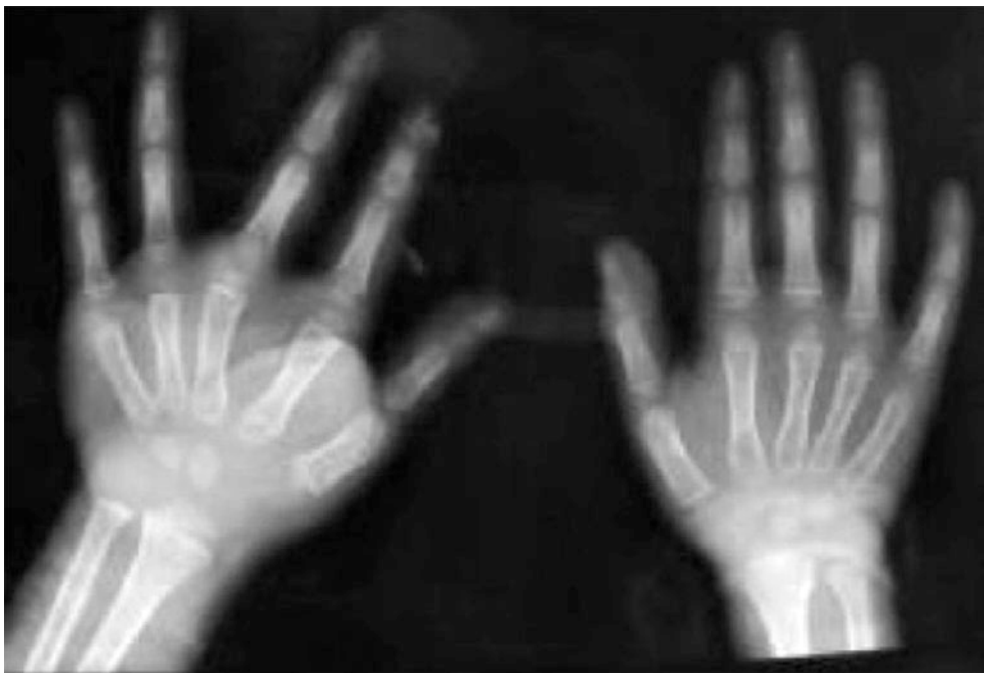

Fig. 9.22 The gaps of the first to fifth heads of metacarpal bone of the left hand become obviously widened, and muscle soft tissue shadows appear

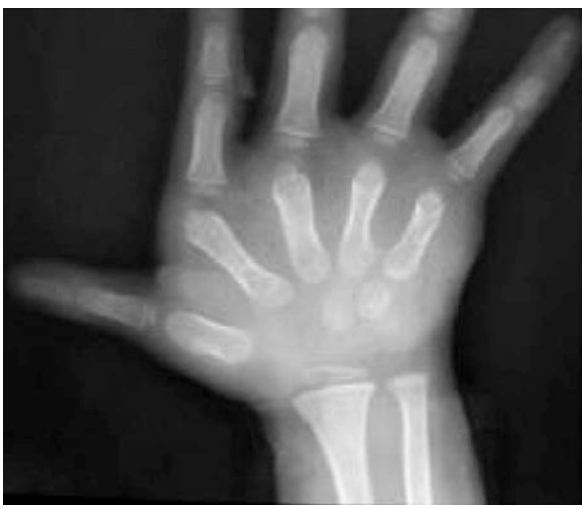

Fig. 9.23 Subluxation of the second and third metacarpophalangeal joints which is particularly obvious in the second metacarpophalangeal joint

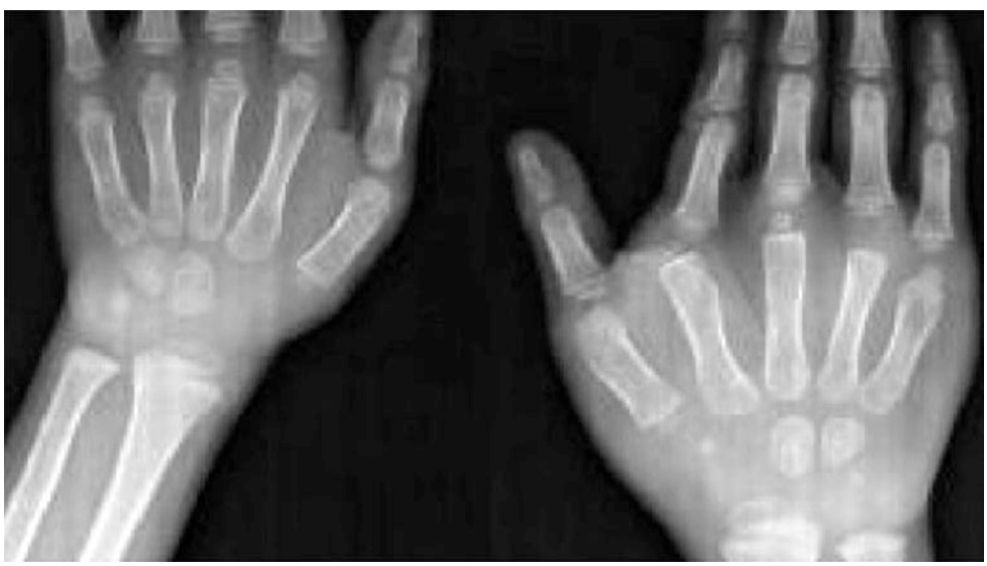

Fig. 9.24 Abnormal shape of the second to fifth heads of metacarpal bone of the right hand

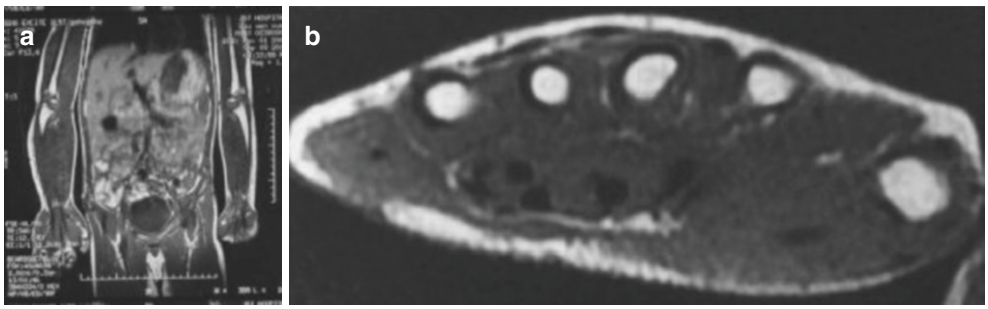

Fig. 9.25 MRI manifestations. (**a**) Comparison between healthy side and affected (left) side in terms of MRI imaging. (**b**) Transverse section MRI of the affected hand indicates that the abnormally hypertrophic muscles are distributed at the volar interoseous regions, hand palm, and thenar and hypothenar muscles

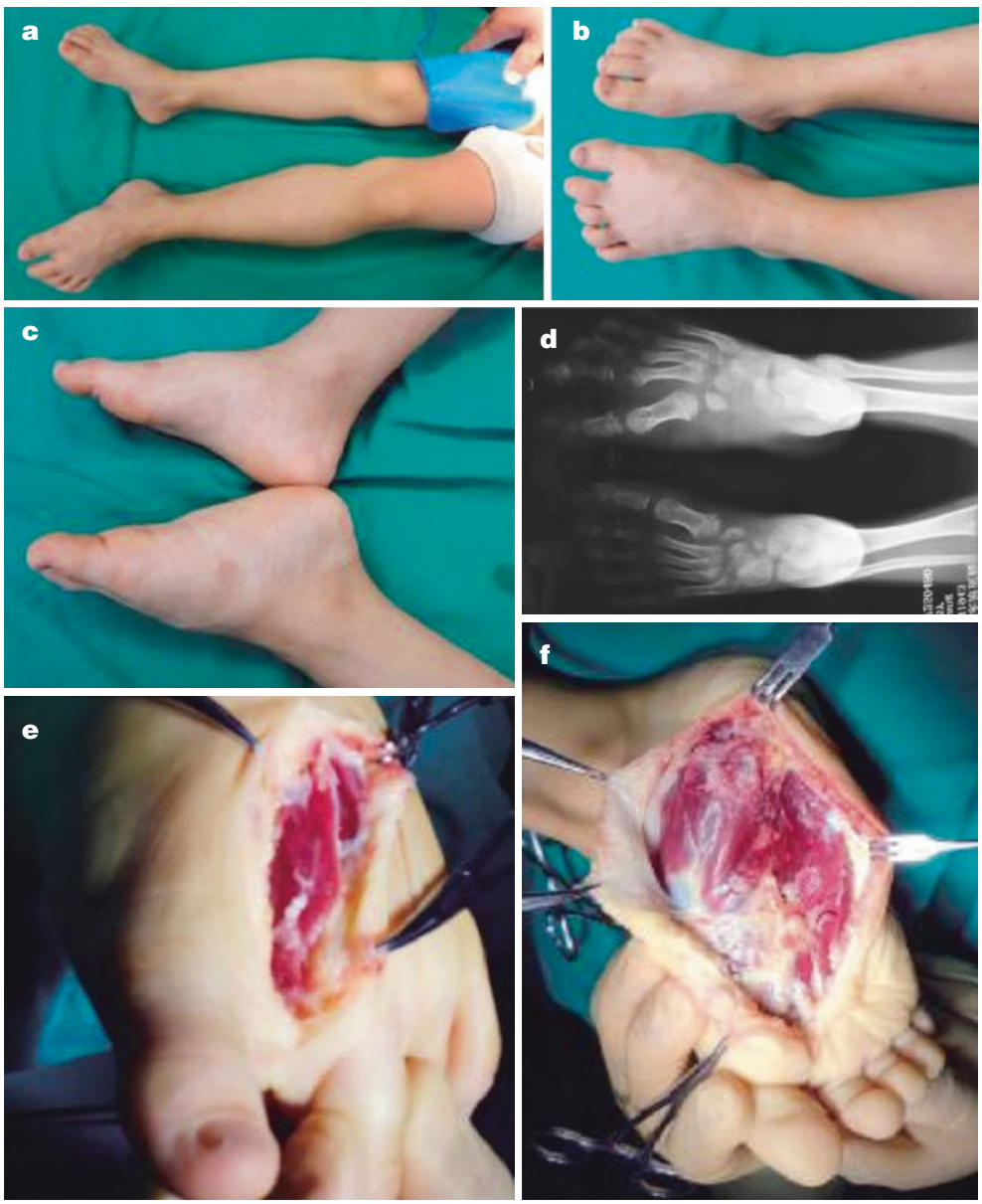

Fig. 9.26 Congenital unilateral lower limb myogenic hypertrophy syndrome. (**a**) Left lower limb myogenic hypertrophy. (**b**) Obvious foot hypertrophy and widening of gaps of heads of metatarsal bone. (**c**) Instep disappears. (**d**) The osteoarticular variation is similar to that of the upper limb. (**e**) Abnormal hypertrophy of internal muscles in the dorsum of the foot. (**f**) Increased quantity and volume of muscles at the sole of the foot

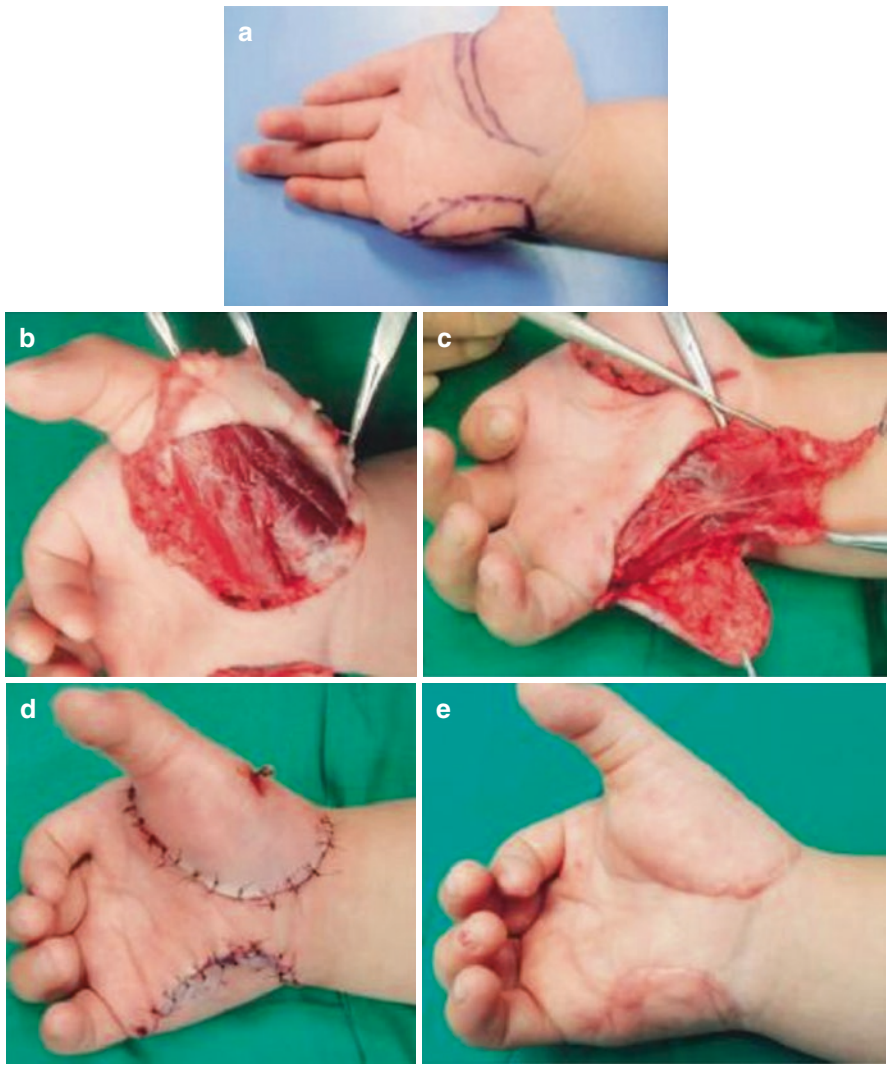

Fig. 9.27 Partial resection of variant muscles. (**a**) Incision design. (**b**) Expose the variant muscles of thenar muscles. (**c**) Expose the variant muscles of hypothenar muscles. (**d**) After removal of variant muscles and superfluous skin. (**e**) Hand appearance at 6 months after operation

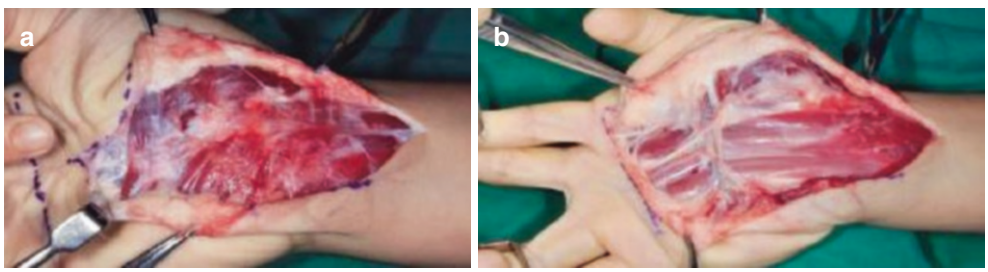

Fig. 9.28 Resection of hypertrophic muscles of palm and forearm. (**a**) Resect the variant muscles of forearm and palm. (**b**) After resection of superficial layer muscle, further expose the variant muscles at the deep parts

shorten the thumb web, but during the second stage the unstable dorsal central slip of the metacarpophalangeal joint needs to be further corrected. Although the palm skin soft tissues are released, the unresected aberrant muscles with free skin grafts are transplanted, this surgery can, to a certain extent, improve the metacarpophalangeal joint ulnar deviation and flexion and the excessive abduction of the thumb (Fig. 9.29).

9.2.3.4 Resect the Variant Muscles at the Dorsal Side of the Metacarpophalangeal Joint, and Tighten the Central Slips and Extensor Tendon Hood

After the release of palm skin soft tissues, resect the dorsal side of the metacarpophalangeal joint and tighten the central slips and extensor tendon hood so that the dynamic equilibrium of the metacarpophalangeal joint can be further recovered and the central slips of finger extensor tendons can be stabilized (Fig. 9.30).

9.2.3.5 Perform Rotary Ostectomy on the Second Metacarpal Bone or Second and Third Metacarpal Bones

The reasons for the occurrence of crossing deformity in index and middle fingers include not only mechanical equilibrium changed by the variant muscles but also joint dislocation or semi-dislocation induced by the epiphyseal dysplasia of the second metacarpal bone or the second and third metacarpal bones. After the soft tissue factors are partially released, it is still necessary to perform rotary ostectomy to correct the line of force of the second metacarpal bone or the second or the third metacarpal bones so that the semi-dislocated metacarpophalangeal joint can be restored; in the meantime, the shortening of metacarpal bone head gap in conjunction with soft tissue surgery can effectively improve and control finger crossing and lateral deviation deformities (Fig. 9.31).

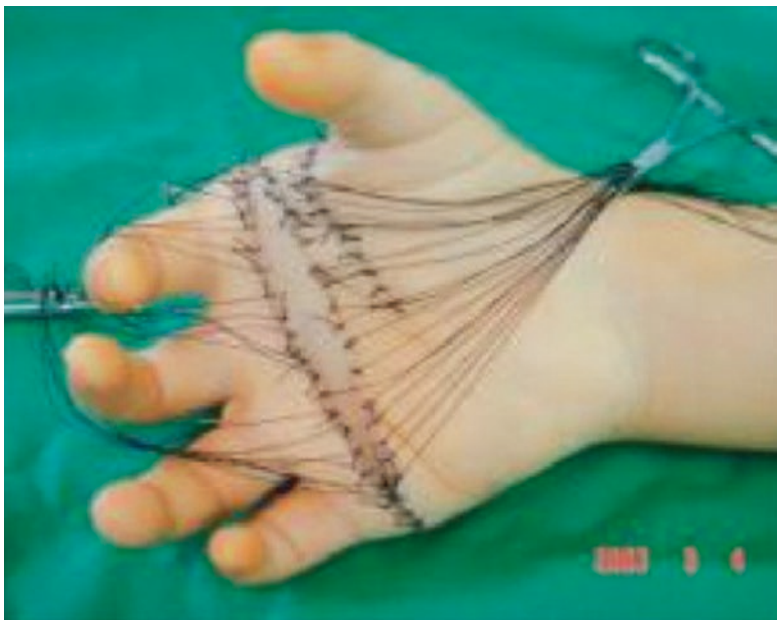

Fig. 9.29 Release the skin soft tissues of the palm, perform transplantation with the free skin grafts, and constrict the thumb fingerweb after the superfluous skin is resected

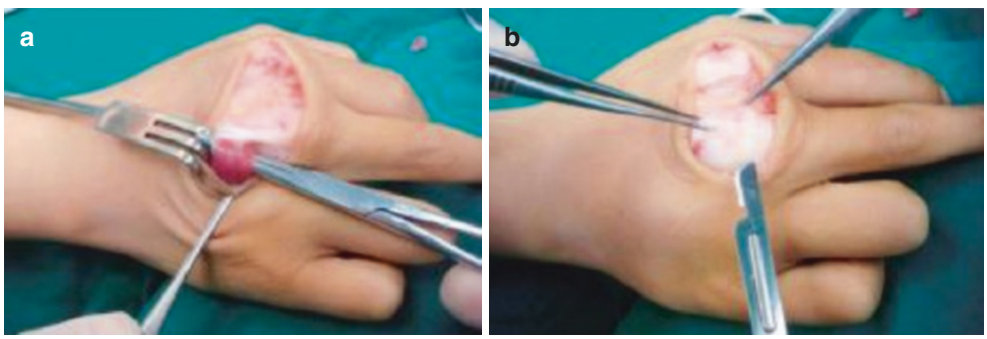

Fig. 9.30 Resect the variant muscles at the dorsal side of the metacarpophalangeal joint, and tighten the central slips and extensor tendon hood. (**a**) Expose the variant muscles at the dorsal side of metacarpophalangeal joint. (**b**) Displace the central slip for fixation after the variant muscles are resected

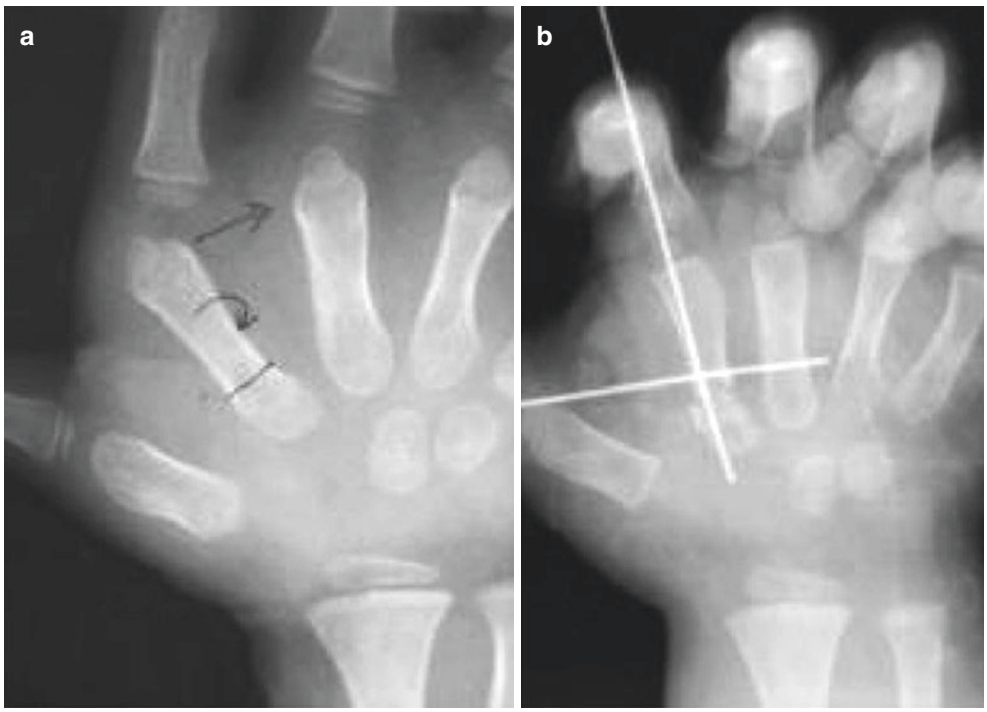

Fig. 9.31 Perform rotary ostectomy on the second metacarpal bone or second and third metacarpal bones. (**a**) Before ostectomy. (**b**) Use Kirschner wire for fixation after ostectomy

9.2.4 Summary

Currently, foreign literature has different names for this kind of congenital deformities such as macromelia, accessory muscle syndrome, vasovagal muscle syndrome, and congenital unilateral limb muscle hypertrophy. According to the morphological characteristics and etiologic factor analysis, this deformity is obviously different from the primary or secondary macromelia (e.g., Proteus syndrome, osteochondromatosis, angioma, and neurofibromatosis), has its own uniqueness, and should become an independent category of congenital limb deformity. In combination with the clinical features of this deformity such as possible pathological anatomical mechanism, nature, range, main morphological characteristics, and etiologic factors, the author suggests the adoption of "congenital unilateral limb myogenic hypertrophy syndrome" for the naming, and it is more reasonable for diagnosis, treatment, or scientific research and teaching [29].

Further in-depth researches need to be done on the morphological and etiologic factors of congenital unilateral limb myogenic hypertrophy syndrome, a clinically rare clinical limb deformity, especially the aberration law of variant muscles and the biomechanical mechanism that induces deformities and functional disorder. The analysis on the results of the author's treatment indicates that the current treatment method is not mature and no scientific and reasonable law has been formed; therefore, it can improve the appearance and functions of the limb only to some extent, and constant studies are needed during future diagnosis and treatment.

The materials in this section are provided by Guang Lei and Liu Bo.

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Congenital Constriction Band Syndrome

10

Bin Wang and Shengbo Zhou

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Constriction band syndrome, also called amniotic band sequence syndrome or amniotic band syndrome, and as many as 34 names are reported by literature (Table 10.1).

This syndrome is characterized by complete or incomplete ring constriction of four limbs and fingers, and the clinical manifestations include acrosyndactyly, brachydactylia, or ectrodactylia and the local swelling and finger (toe) lymphedema beyond the constriction band (Fig. 10.1).

Table 10.1 Different names of constriction ring syndrome

Constrictive bands	Congenital constriction ring syndrome
Constriction bands	Congenital ring constriction syndrome
Constriction rings	Congenital bands
Constriction grooves	Congenital amputations
Amniogenic bands	Congenital ring constriction
Amniotic bands	Congenital annular constrictions
Amniotic disease	Congenital annular defects
Amniotic constrictions	Annular defects
Amniotic band syndrome	Annular constricting bands
Amniotic band disruption complex	Amniochorionic mesoblastic
Ring constriction	Fibrous strings
Ring constriction syndrome	Circular constricting scars
Constriction ring syndrome	Focal deficiency
Congenital ring syndrome	Streeter dysplasia
Congenital constricting band	Acrosyndactyly
Congenital constriction band	Intrauterine amputations
Congenital constriction band syndrome	Fetal amputations

10.1 Etiologic Factor

There are two theories: internal cause and external cause. The theory of internal cause was first proposed by Streeter in 1930 who holds that the etiologic factor was that the vessels in the embryo were discontinued and blastodermal development was subject to interference [1]; Van Allen utilized RMA and TCA to observe the artery at the limb ring constriction of the newborns which was bifurcation-shaped or fine without branches, which also supports this theory [2]. The representative in favor of the theory of external cause holds that, due to intrauterine amniorrhexis, the fetal limb or partial limb is wined and strangled by the released amniotic bands; other hypotheses hold that the limb may be stuck at the cleavage of amniotic membrane wall, but this theory cannot explain the cases with other congenital diseases (e.g., syndactylia, cheilopalatognathus, and anal atresia). In 1975, Kino utilized embryonic rats and duplicated the animals with ring constriction with distal syndactylia [3, 4] (Fig. 10.2).

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DOI 10.1007/978-981-10-5101-2_10

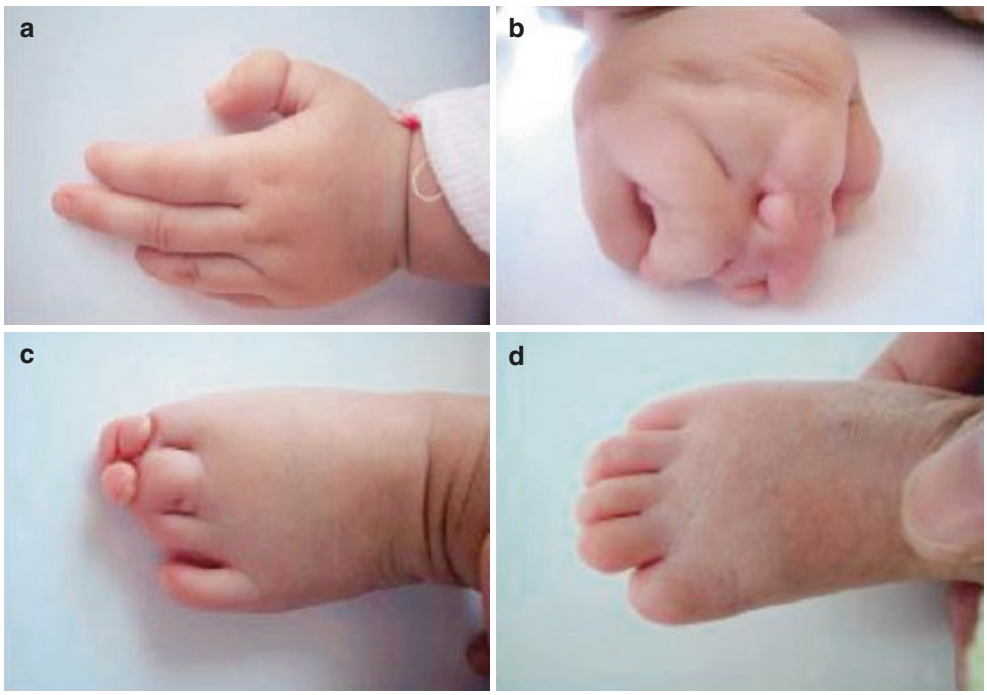

Fig. 10.1 Congenital constriction ring syndrome. (**a**) Thumb ring constriction with lymphedema at the distal end of finger body. (**b**) Fusion of the distal ends of the index, middle, and ring indexes and the presence of acrosyndactyly on the skin at the proximal end. (**c**) Fusion at the distal end of the first to fourth toes and the presence of acrosyndactyly on the skin at the proximal end. (**d**) Toe amputation induced by ring constriction

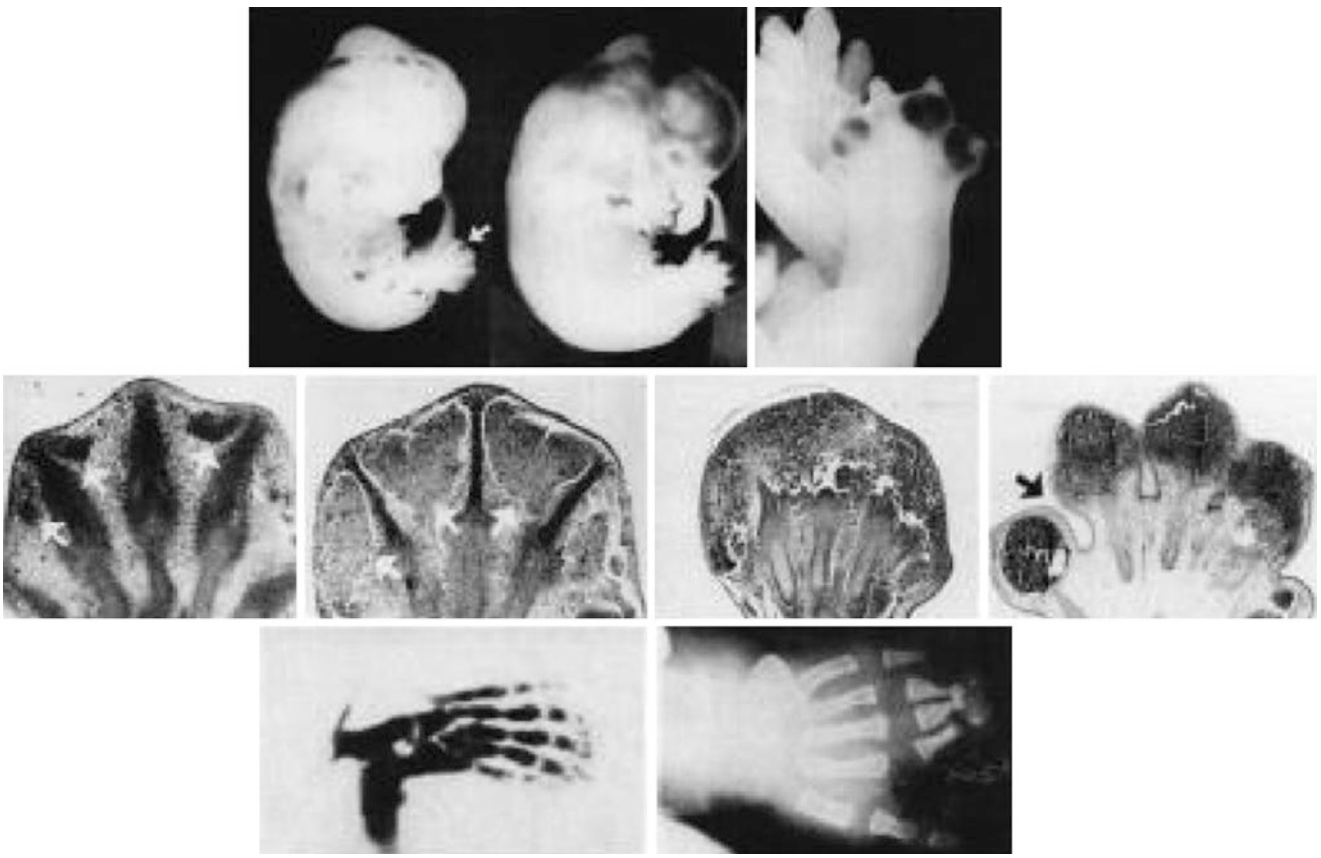

Fig. 10.2 Establishment of animal models with ring constriction. (Reprint with permission from Kino Y. Clinical and experimental studies of the congenital constriction band syndrome, with an emphasis on its etiology. J Bone Joint Surg 1975;57A(5):636–43)

10.2 Clinical Manifestation

The constriction can be complete ring constriction or incomplete ring constriction. It can occur to any site of the body, but the commonest site is the limb. These constrictions can cause congenital amputation or insufficient mutilation with distal edema, and in severe cases, limb development can be affected; the constriction can also lead to fusion of adjacent or nonadjacent fingers and toes, therefore forming combined syndactylia and acrosyndactyly, and the distal ends of the two fingers are fused, and residual sinus tract can be observed at the proximal end (Figs. 10.3 and 10.4). However, the limb near the constriction ring is almost normal. The amputation can occur to any level of the limb and even to the head, neck, and abdomen as reported. Smaller constriction ring can result in some rare facial clefts. The residual limb can be manifested as follows: the skin is tightly covered on the skeletons which become thin at the tips. As for the residual limb beyond the constriction ring, the skin may be hard and presents nonpitting edema. The nerve injuries can be sequelae induced by constriction ring and can manifest symptoms immediately after birth. Through surgical exploration, the author finds that these symptoms are correlated with the absence of distal nerves. The ring constriction can be accompanied by cheilopalatognathus, cleft foot, and even radial longitudinal ray hypoplasia (Figs. 10.5, 10.6, and 10.7).

MRI is of significance to the judgment of the involvement degree of deep tissues and can display the distribution of important vessels. Our studies also reveal that, in most cases of ring constriction, the deep vessels are rarely involved, which provides solid imaging support for the one-time resection (Fig. 10.8).

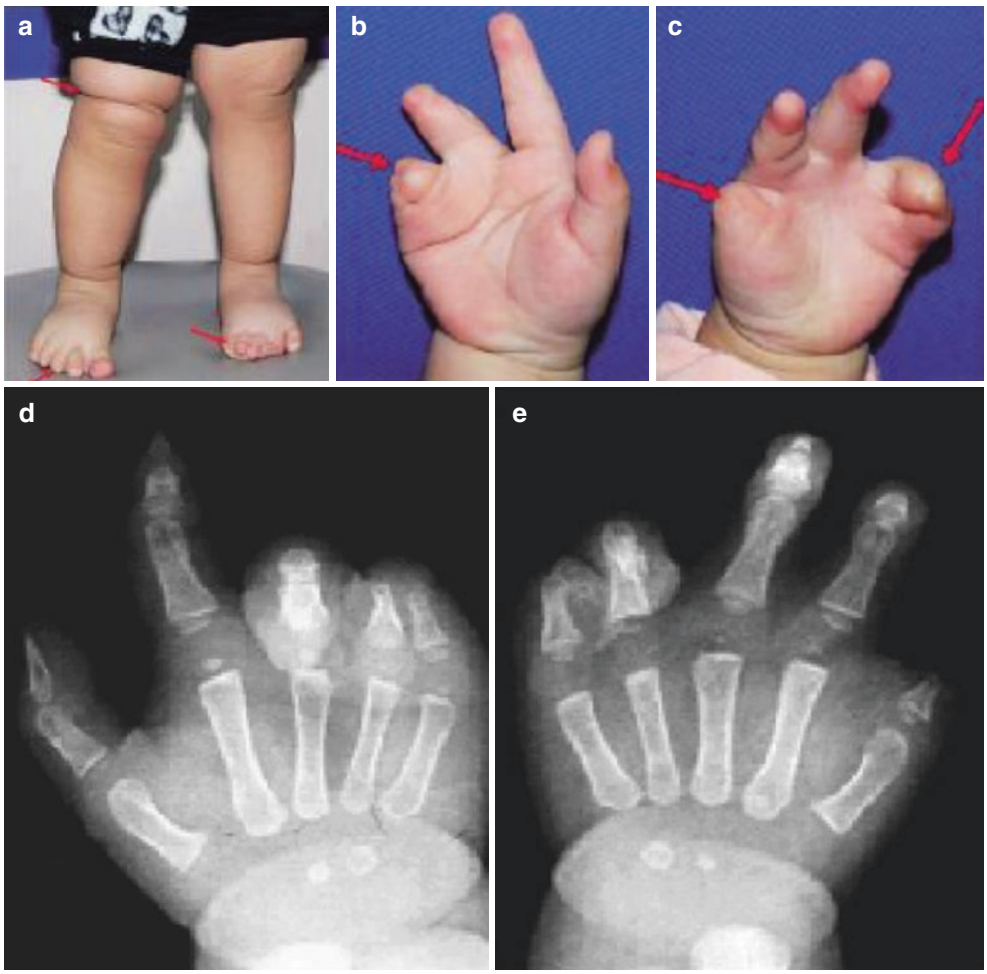

Fig. 10.3 Right thigh ring constriction with brachydactylia, amputation, and acrosyndactyly

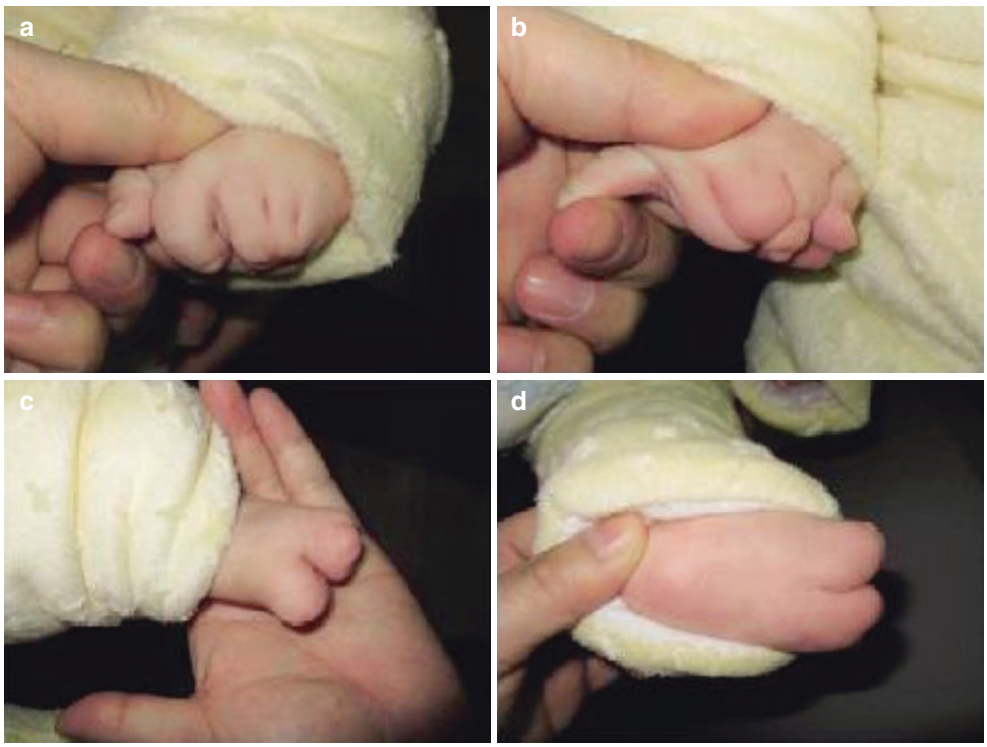

Fig. 10.4 Ring constriction with cleft foot, brachydactylia, amputation, and acrosyndactyly

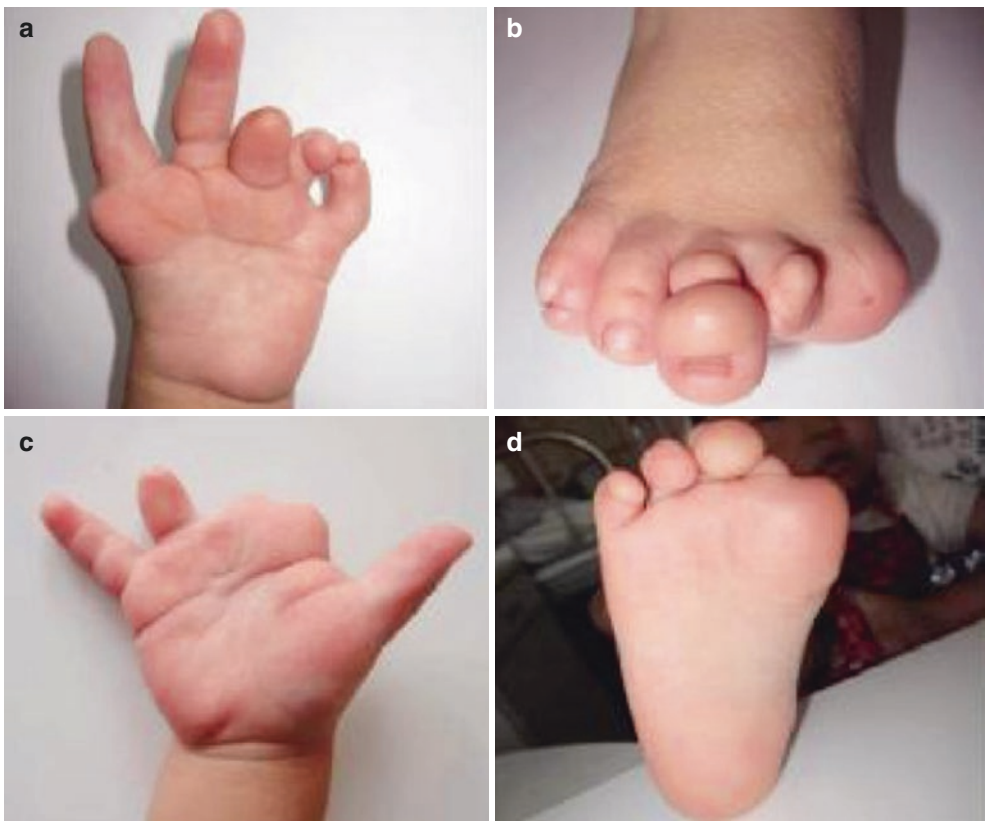

Fig. 10.5 Ring constriction with toe lymphedema, finger radial longitudinal ray hypoplasia, and brachydactylia, syndactylia, and amputation

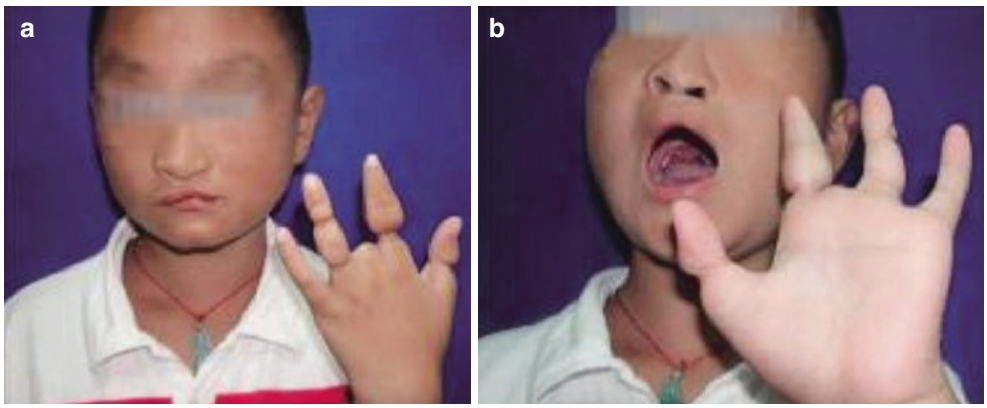

Fig. 10.6 Ring constriction with recessive cheilopalatognathus and amputation

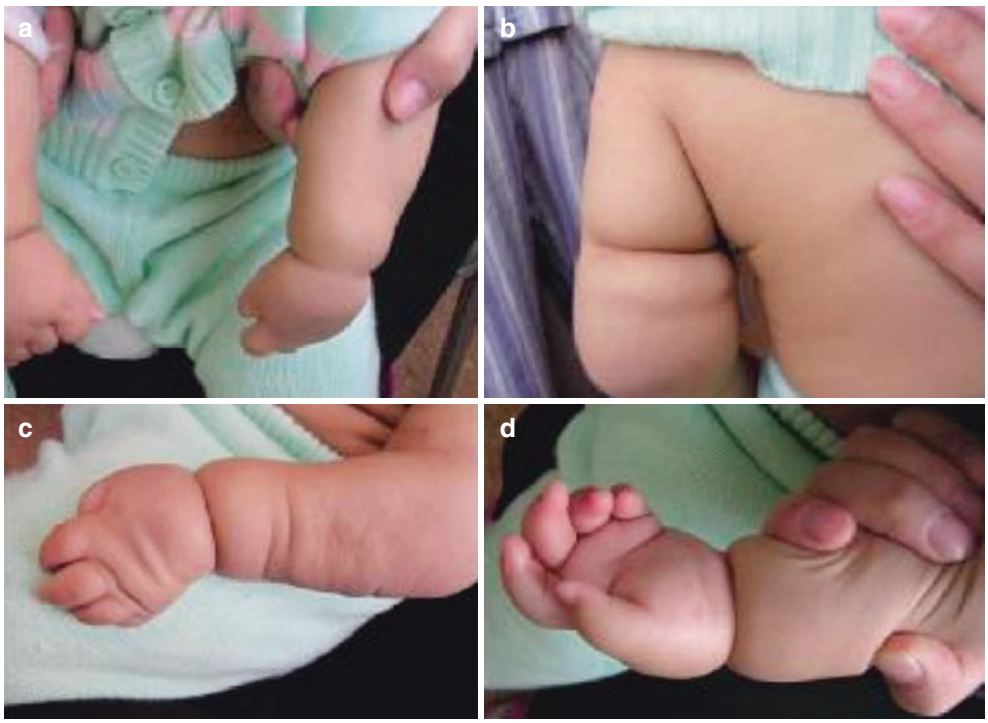

Fig. 10.7 Ring constriction with the forearm and wrist with radial nerve injuries and hand lymphedema

10.3 Classification

Constriction ring syndrome is defined as a whole and can be further classified according to the deformity manifestations of the limb [5, 6] (Tables 10.2 and 10.3).

10.4 Treatment

10.4.1 Amniotic Band Release Before Birth

Under very rare circumstances, the constriction ring will lead to distal ischemia. At this time, surgical release of the compression of constriction band is necessary, but postoperative survival of the limb is often a problem, and surgical amputation is needed in most cases. The surgery of intrauterine release of lower limb constriction ring has been successfully performed, and the affected limb can be retained. When antenatal ultrasound examination indicates the presence of severe constriction, this treatment can be considered; however, the risk on pregnant women and fetuses must also be taken into consideration. Currently, the release of amniotic band under the fetoscope is only limited to the cases with progressive edema and cycle discontinuation which can lead to limb defects. The surgeons must inform the patients of the possibility of spontaneous abortion, and the incidence is 6–10% [7].

10.4.2 Nerve Transplantation

Ring constriction can lead to peripheral nerve injuries. There is still some question concerning the value of electrophysiological estimation. Some reports claim that good efficacy can be obtained after the release of nerve compression, but the

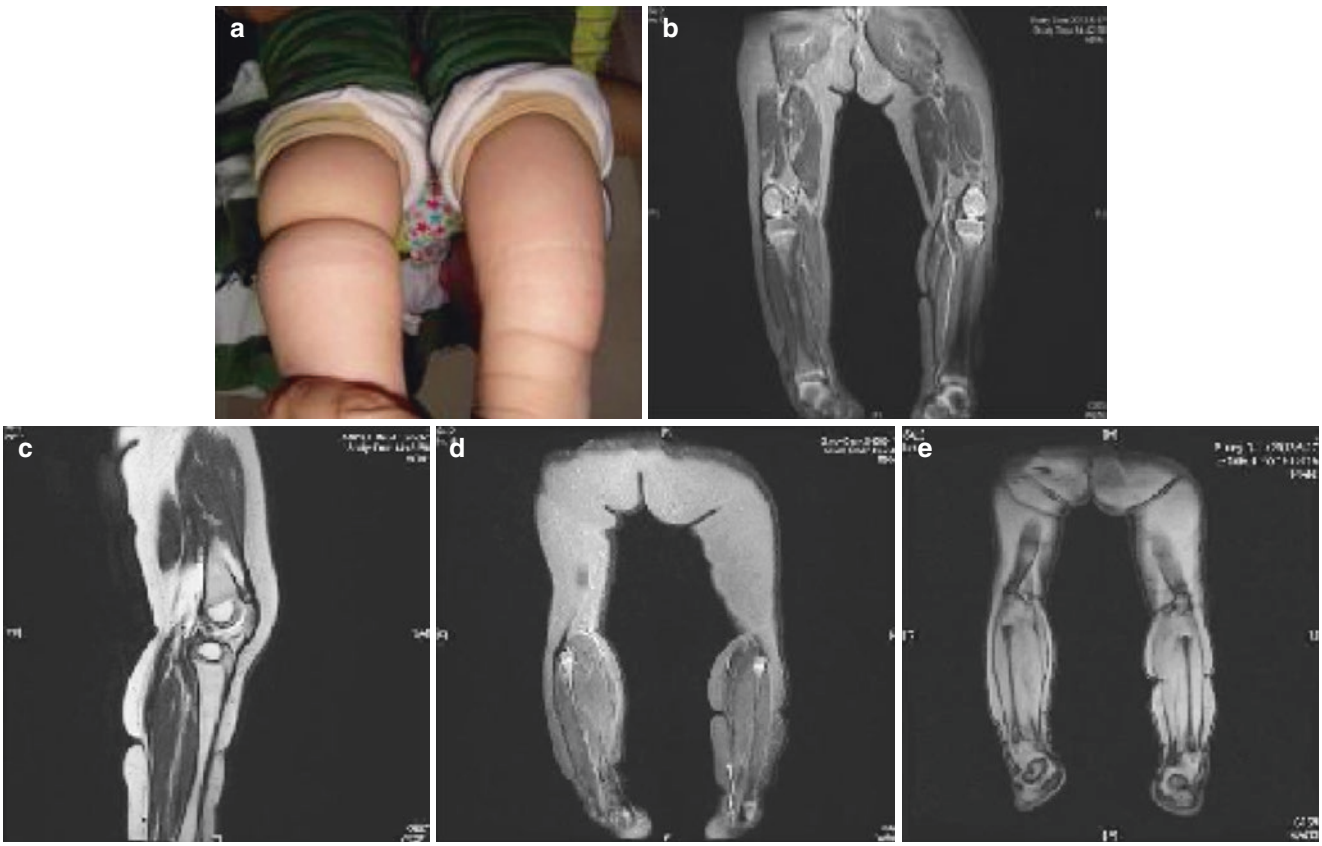

Fig. 10.8 Left shank constriction ring is deep and present, and MRI displays that the deep vessels are not involved

Table 10.2 Patterson classification of constriction ring syndrome

Type	Manifestation
Class 1	Simple ring constriction
Class 2	Ring constriction with distal deformities, complicated by (or not complicated by) lymphedema
Class 3	Ring constriction with distal fusion, acrosyndactyly
Type I	Fingertip fusion
Type II	Fingertip fusion, relatively far fingerweb
Type III	Fingertip fusion, no fingerweb, combined syndactylia with proximal sinus tract
Class 4	Intrauterine amputation

Originate from Patterson T: Congenital ring-constrictions, Br J Plast Surg 1961:14:1–31

Table 10.3 Isacsohn classification of constriction ring syndrome

Type	Manifestation
Class 1	The band is only a groove in the skin
Class 2	The band extends to the subcutaneous regions and muscles
Class 3	The depth reaches the skeletons
Class 4	There is a pseudoarthrosis
Class 5	Uterine amputation occurred

Originate from Isacsohn M. Congenital annular constrictions due to congenital constriction bands. Acta Obstet Gynecol Scand 1976;55:179–182

cases with the presence of nerve continuity but no improvement in nerves still dominate, and a majority of pediatric patients still need nerve transplantation.

10.4.3 Treatment of Constriction Ring

The purpose is functional and aesthetical improvement, mainly by means of resection of the skin and subcutaneous tissues around the constriction ring, and performance of ring bundle Z-plasty or W-plasty. The techniques can be used to redesign the scars so that the constriction ring can be released and eliminated (Figs. 10.9 and 10.10). Upton emphasizes the importance of application of subcutaneous fascial flap reposition to correct contracture (Fig. 10.11). Mutaf [8] recently reported a type of rectangle flap technique: the dermic fat flap transposition at the fissure can be used to increase the tissue thickness so that the skin scars will not be lengthened (Fig. 10.12). It was previously considered that one-time resection of the entire girdle was unnecessary, but it is currently considered that one-time resection of the entire girdle is safe. In case two girdles are present and they are mutually adjacent, it is recommended that separate surgeries be adopted, and one girdle be resected each time. The author girdles proposes that

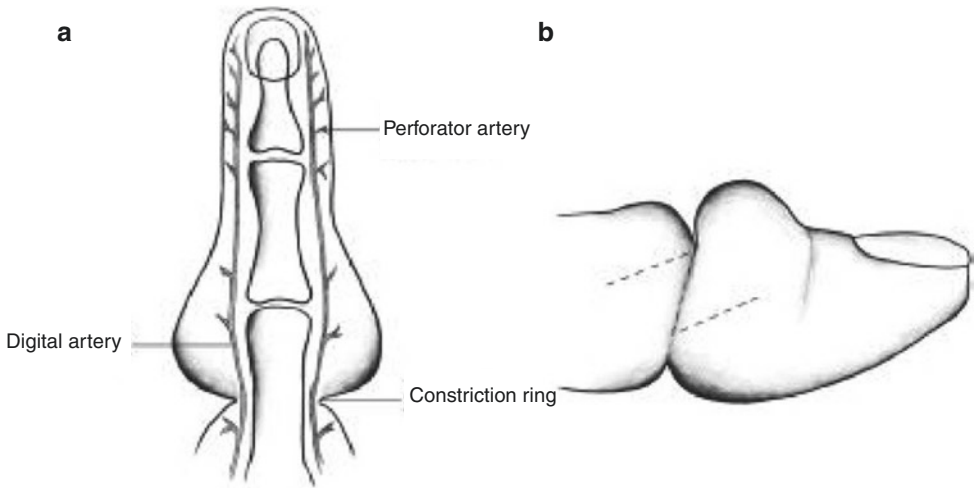

Fig. 10.9 Anatomical characteristics and surgical design of finger ring constriction

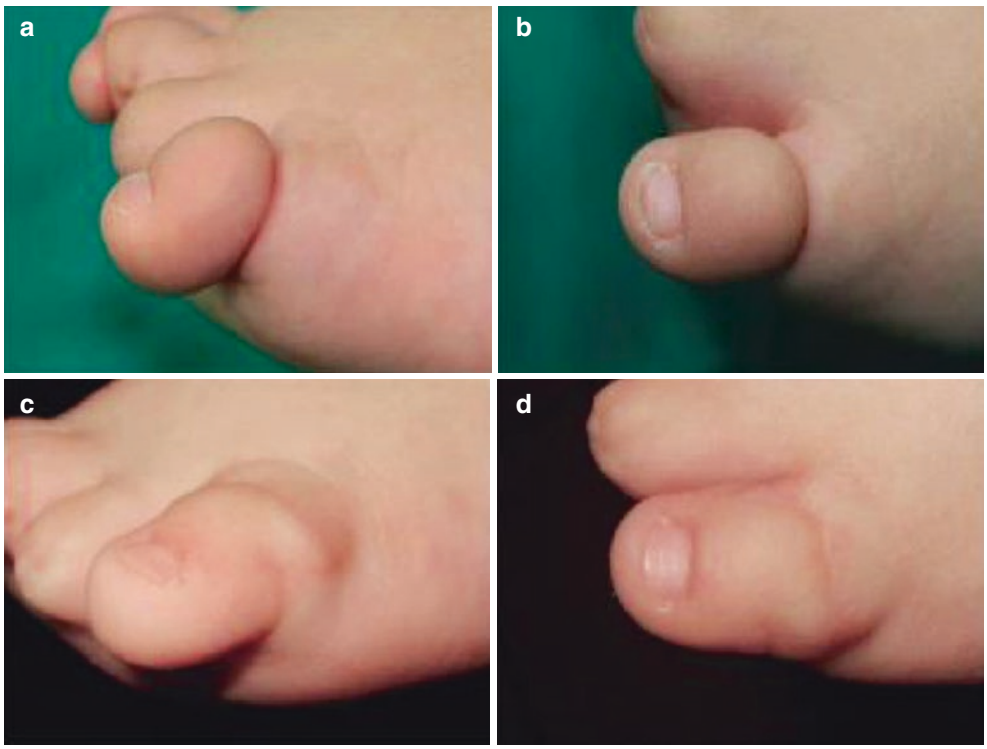

Fig. 10.10 Surgical effects of one-time resection of ring constriction. (**a**, **b**) Before operation. (**c**, **d**) At 6 months after operation

one-time resection characterized by complete resection of constriction ring, fascial flap reposition, and formation of skin triangular flaps can not only relieve the lymphedema but also achieve minimization of the scars (Fig. 10.13).

10.4.4 Separation of Acrosyndactyly

The treatment principle of ring constriction complicated by syndactylia is the same as that of other types of syndactylia. In finger separation, attention should be paid to reconstruction of fingerweb, skin grafting of finger skin defects, and formation of finger nails and finger pulp. Usually, the distance from the sinus tract is far, and the combination with the flaps at the junction is difficult, so full thick skin grafts are used for transplantation after the resection. In addition, attention should be paid to the release of the accompanying constriction ring. The time of finger separating surgery is very important because the connected fingers are usually not equally long, and angulation deformity will be induced with the growth. The release of the adhesion at the tip of the syndactylous fingers can relieve such tethered effect, and the separation of proximal fingers and the reconstruction of fingerweb can be delayed until school age.

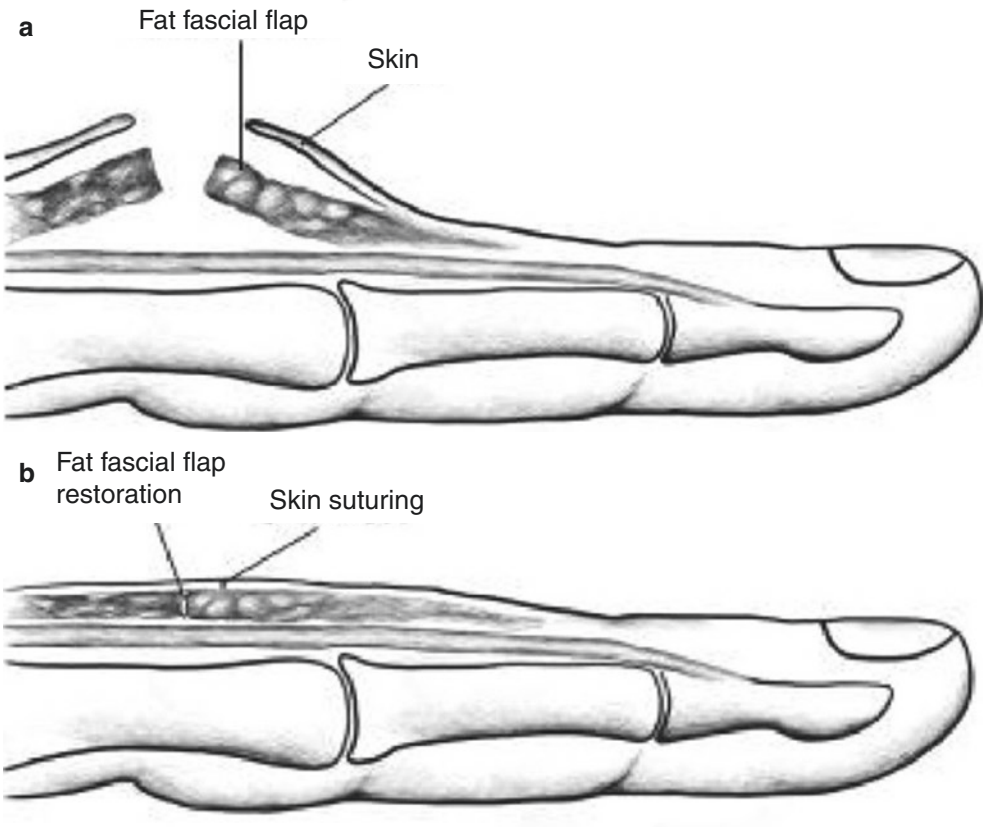

Fig. 10.11 Advancement and reposition of fascial flaps

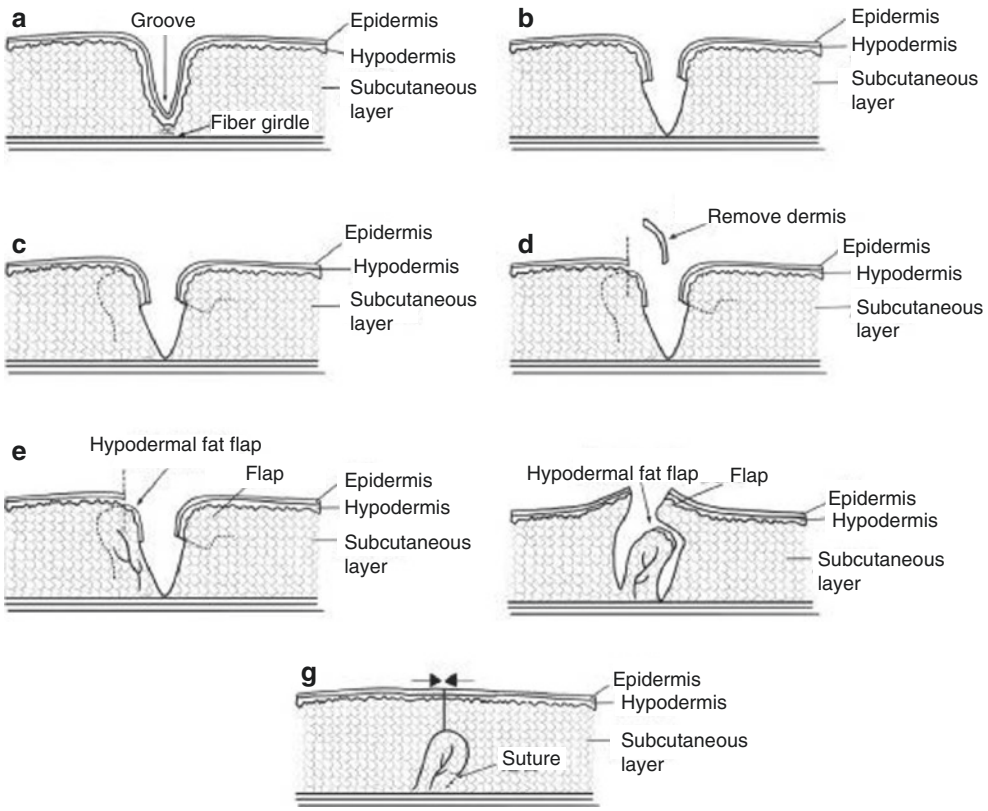

Fig. 10.12 Mutaf rectangle flap technique. (Redrawn from Mutaf M, Sunay M. A new technique for correction of congenital constriction rings. Ann Plast Surg 2006;57(6):646–52)

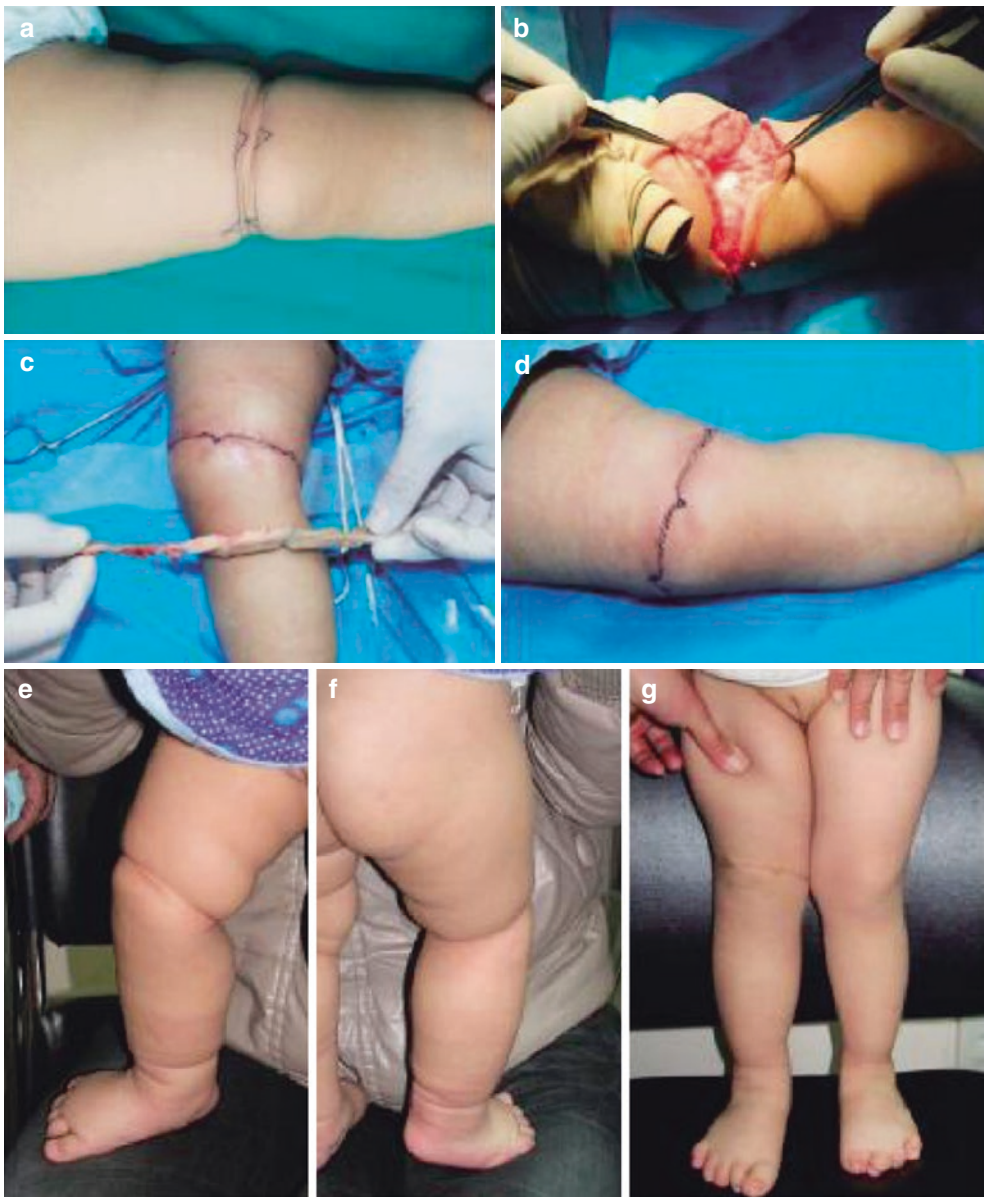

Fig. 10.13 The one-time resection characterized by complete resection of constriction ring, fascial flap reposition, and formation of skin triangular flaps

10.4.5 Skin Nodule

Skin nodules are common in constriction ring syndrome; these lumps are often located at the digital dorsum, and the position is fixed with edema. These skin nodules can be treated using many methods, but Z-plasty is often not effective. The common method is complete resection; if necessary, local full thick skin grafts can be used for transplantation.

10.4.6 Finger and Toe Transplantation

Finger and toe absence are common in constriction ring syndrome, the number of absent fingers and toes varies, and the appearance is similar to transection absence. As the proximal structures of the level of the absence are normal, finger and toe transplantation can be performed.

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Congenital Deformities of the Hands and Upper Limbs and Associated Syndromes

11

Jinghong Xu, Hui Shen, Wei Wang, Bo Chen, and Sheng Ding

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The congenital deformities of the hands and upper limbs can appear in both various syndromes and generalized skeletal deformity syndromes, and they are descried, respectively, as follows.

11.1 Congenital Deformities of the Hands and Upper Limbs and Various Syndromes

11.1.1 Marfan Syndrome

Marfan syndrome, also called arachnodactyly syndrome, is a congenital disease in generalized connective tissues, and it is an autosomal dominant inheritance. This disease can be divided into four types [1]: weak type, non-weak type, contracture type, and joint hypermobility type, and it mainly involves the muscles, skeletons, cardiovascular regions, and eyes. Abnormalities can be found immediately after the pediatric patient is born with the manifestation of arachnodactylia; the patient is relatively tall; the limb, especially the distal limb, is tall and slender; and the lower part of the body is longer than the upper part; the face is long, and the palatine arch is high; the patient may have chicken chest or funnel chest and lateral curvature or kyphosis; the ligaments and joint capsules are relaxed with dislocation of hip joint and knee joint and flatfoot. The vascular deformities include

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distention of aorta, mitral prolapse, and dissection of aorta. The eye symptoms can be accompanied by serious myopia, heterotropy, glaucoma, etc. No especially effective treatment is available for this disease, and the main measures are symptomatic treatment and corrective surgery [2–4].

11.1.2 Ehlers-Danlos Syndrome

Ehlers-Danlos syndrome, also called cutis hyperelastica symptom, is one of the commonest genetic connective tissue diseases. This disease is induced by collagen metabolism disorder with the main presentations of cutis hyperelastica, fragile skin and vessels, and joint hyperactivity. It has nine clinical types:

Type I (severe type), which is autosomal dominant inheritance with the manifestation of cutis hyperelastica, joint hyperactivity, and subcutaneous calcified nodules

Type II (mild type), whose symptoms are the same as type I but milder, and the joint hyperactivity is only limited to the hands and feet

Type III (benign joint hyperactive type), which is autosomal dominant inheritance, with the manifestations of joint hyperactivity and normal skin cicatrization

Type IV (ecchymosis type), where the skin is thin, the subcutaneous bruise is obvious, the joint activity is normal, and the artery is apt to rupture

Type V which is sex-linked inheritance and whose symptoms are the same as those of type II

Type VI (eye type), where in addition to skin and joint symptoms, there can be rupture of cornea and sclera as well as detachment of retina

Type VII (congenital multiple arthrochalasis type), with the manifestation of joint laxity accompanied by dislocation or semi-dislocation in hip, knee, and other joints

Type VIII (periodontitis type), where the skin is apt to contusion with progressive periodontitis and premature dedentition

Type IX (occipital angle type), where the skin is fragile and relaxed, joints feature hyperactivity, and dislocation of radial capitulum, occipital angle, hernia, and bladder diverticulum can be accompanied [2, 3, 5]

There is no especially effective treatment for this disease. Surgical correction can be performed in case of deformities and dislocation of four limbs, but the postoperative scars are wide, thin, and bright. Due to the laxity of articular ligament, there may be still semi-dislocation after the operation, but the influence on the function is not great.

11.1.3 Larsen Syndrome

Larsen syndrome, also called cleft palate, poker face, or multiple congenital joint dislocations, is an autosomal recessive or dominant inheritance. The aplasia of generalized connective tissues induces the excessive laxity of generalized joints and the subsequent dislocation, facial changes, and deformities in the hands, feet, spinal columns, and heart, with the manifestations of protruding forearm, flat face, widened eye span, low and flat nose bridge, angulus oculi, and cleft palate and dislocation of the hip, knee, and elbow joints, which is symmetric; the fingers take cylindrical shape, the metacarpal bone is short, and the fingernails are wide and short; the feet present equinovarus or equinovalgus (Fig. 11.1). Joint dislocation of four limbs and foot deformities can be surgically restored and corrected, and the effects are good [6].

11.1.4 Down Syndrome

Down syndrome, also called 21-trisomy syndrome, is the congenital development abnormality induced by the presence of one more chromosome 21 with dysgnosia as the main symptom with the manifestations of short head; with

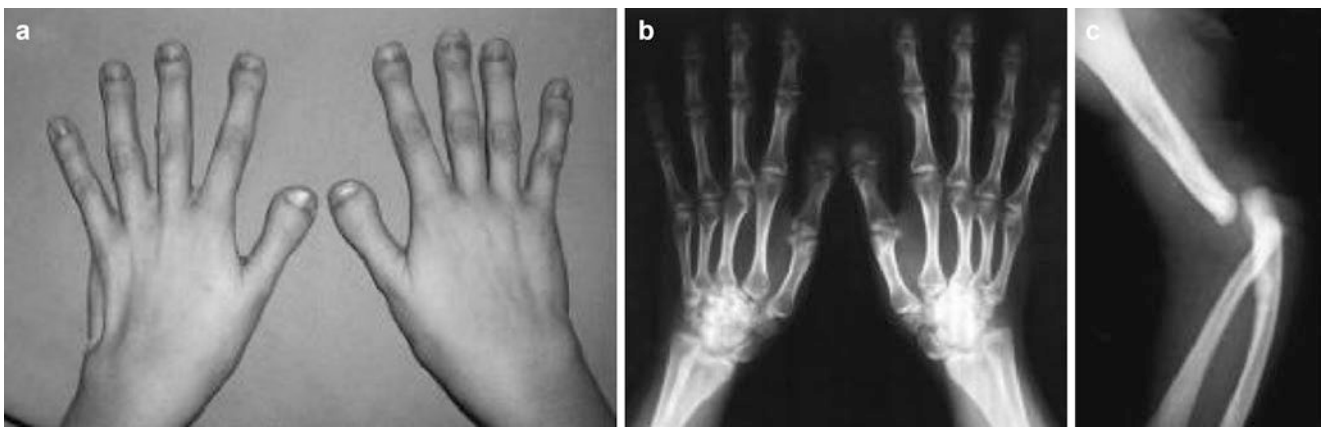

Fig. 11.1 Larsen syndrome. (**a**, **b**) The fingers take cylindrical shape, the metacarpal bone is short, and the fingernails are wide and short. (**c**) Dislocation of elbow joint (Reprint with permission from Bicknell LS. A molecular and clinical study of Larsen syndrome caused by mutations in FLNB. Journal of Medical Genetics. 2007; 44 (2):89–98)

the third fontanel and dysplasia in maxillary bone and nasal bone, the palpebral fissure becomes oblique toward the external upper region, the eye span is widened, the tongue is thick, and the ears are small; the muscular tension of four limbs is low, the joints are soft, the articular ligaments are relaxed, and hyperactivity is possible; the hands are short and wide, there is always one palm print, it is the simian line, and the tip of the little finger bends inward; the feet are wide and thick, and the affected toe is far away from the remaining four toes; the patient often suffer from lateral curvature, the wing of ilium becomes abducted, and acetabulum often tends to be flat; the patient often suffer from dysgnosia. The diagnosis can be confirmed through chromosome examination. No special treatment is needed for this disease, and the main measure should be long-term education training [2, 3].

11.1.5 Edwards Syndrome

Edwards syndrome, also called 18-trisomy syndrome, is induced by one more chromosome 18. Karyotype analysis indicates the presence of chromosome 47, and one supernumerary chromosome can be found in the position of chromosome 18, so trisomy is formed. The clinical manifestations are multiple deformities. After the patient is born, the fingers become flexed and take clenching shape, the second and third fingers are overlapped, and the little finger becomes bent and sloped inward; the feet present equinovarus or platypodia; the skulls are narrow and long, the occipital bones are protruding, the ear position is low, the low jaw is short and small, the neck is short, the chest bones are short, the pelvis is stenotic and the spinal column is deformed; the patient often concomitantly suffers from congenital cardiovascular deformities and low intelligence. The diagnosis can be confirmed through cell chromosome examination. The prognosis of this disease is poor, and 90% of the pediatric patients die within the age of 1 [2, 3].

11.1.6 Patau Syndrome

Patau syndrome, also called 13-trisomy syndrome, is induced by the presence of one more chromosome 13, so the patient has 47 chromosomes. The clinical manifestations are small skulls, microphthalmia or anophthalmia, cheilopalatognathus, short neck with neck web, capillary angioma, deafness, low ear position, syndactylia and hyperdactylia, flexion contracture of interphalangeal joint, foot deformities, and dislocation of ankle bone, with severe deformities in nerve system and cardiovascular deformities and low intelligence. The diagnosis can be confirmed through chromosome karyotype examination. The prognosis of this disease is poor and the patients often die during the infancy [2, 3].

11.1.7 Trisomy-8 Syndrome

Trisomy-8 syndrome, also called 8-trisomy-syndrome, is induced by the presence of one more chromosome 8, so the patient has 47 chromosomes. The clinical manifestations are large skulls; dysplasia of external ears; low ear position; short neck; tall and slender trunk; pitting of chest bones; deformity of equinovarus; flexion of the big toe; tall, slender and bent fingers; limited movement of four limbs and joints; and dislocation of hip joint; the patient can concomitantly suffer from cryptorchidism, congenital heart disease, and low intelligence (Fig. 11.2). The diagnosis can be confirmed

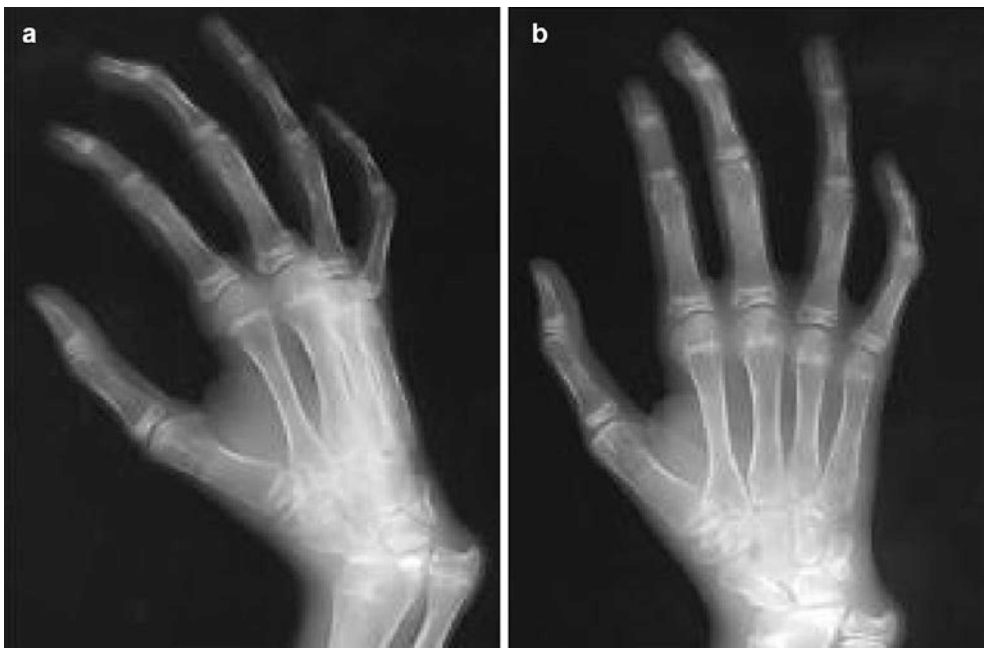

Fig. 11.2 Hand deformities induced by Trisomy-8 syndrome (Reprint with permission from Alvi F. Upper limb abnormalities in mosaic trisomy 8 syndrome. Archives of Orthopaedic and Trauma Surgery. 2004; 124 (10):718–9)

through chromosome examination. The prognosis of this disease is poor and only a few of them can survive until they become adults [2, 3, 7].

11.1.8 Turner Syndrome

Turner syndrome, also called primary ovarian function insufficiency syndrome, is induced by congenital chromosome combination abnormalities. It only occurs to women with the manifestations of short body, neck web, cubitus valgus, sexual infantilism, ovarian atrophy, small ovary, breast nondevelopment, and primary amenorrhea; the patients often suffer from cardiovascular system deformities and low intelligence. X-ray displays deformity of cubitus valgus, abnormal arrangement of carpal bone, shortened fourth metacarpal bone, and delayed bone age. The diagnosis can be confirmed through chromosome examination. Sexual infantilism can be treated by hormone replacement therapy. If necessary, neck web and cubitus valgus can be corrected through surgery [2, 3].

11.1.9 Klinefelter Syndrome

Klinefelter syndrome, also called small testopathy, is a disease suffered by patients who has one more X chromosome, and the sex chromosome is XXY. During the infancy, the symptoms are not obvious. After puberty, the manifestations are small testes, incapability of childbearing, presentation of feminized personality and posture, and mild intellectual disturbance; the skeletal deformity has ulnar-radial fusion and shortened fourth metacarpal bone, and the tip of the fifth finger is bending inward. The diagnosis can be confirmed through chromosome examination. This disease can be treated by testosterone replacement therapy. When the skeletal deformity influences the functions, surgical correction can be performed [2, 3].

11.1.10 Klippel-Trenaunay-Weber Syndrome

Klippel-Trenaunay-Weber syndrome is also called vasodilatory limb hypertrophy. The etiological factors are not clear, and the manifestations are that one side of the limb is thick and long and the skin temperature rises with hyperhidrosis or hypohidrosis, accompanied by skin angioma and hearable vascular murmur; the patients can be accompanied by generalized vascular deformity, syndactylia, hyperdactylia, arachnodactyly syndrome, spina bifida, and microcephaly, and some patients may suffer from feeble mindedness (Fig. 11.3). X-ray can indicate the thickening of cortex of the bone. During the early stage, pressure bandages can be used for symptomatic treatment. The prognosis of this disease is relatively good [2, 3, 8].

11.2 Congenital Deformities of the Hands and Upper Limbs and Generalized Skeletal Deformity Syndrome

11.2.1 Apert Syndrome

Apert, a French psychiatrist, first reported Apert syndrome in 1906. This is a rare disease characterized by complete syndactylia in both hands and both feet; its incidence is about 1/65,000, and there is no obvious difference between males and females. Late childbirth is correlated with the onset. Data indicate that the incidence of Asians is high and the incidence of Spaniards is low [9].

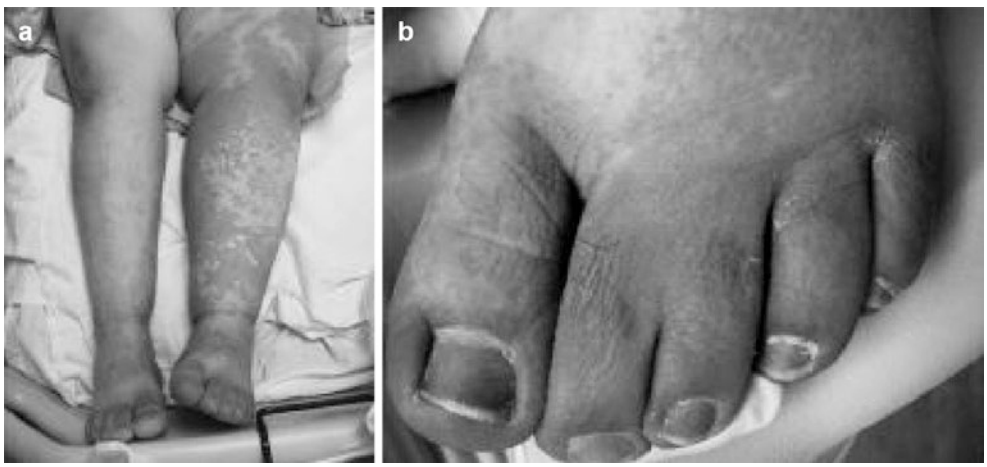

Fig. 11.3 Klippel-Trenaunay-Weber syndrome. (**a**) Angioma at one side of the limb. (**b**) Syndactylia (Reprint with permission from Klippel-Trenaunay syndrome and cerebral haemangiopericytoma: a potential association. Acta Neurochirurgica. 2008; 150 (4):399–402)

Apert syndrome, also called acrocephalosyndactyly, is a clinical syndrome mainly manifested as retroposition of middle face, parrot mouth and nose, craniosynostosis, and mirror image syndactylia. Due to unknown reasons, the severity of cranial deformity is inversely proportional to the severity of hand deformity [9–11]. The hand deformities include the complicated syndactylia of the index, middle, and ring fingers, the simple syndactylia of the ring and little fingers, the stenosis of the first fingerweb to varying extents, and the lateral deviation of the thumb, which seriously affect the hand gripping functions. In severe cases, complete fusion at the distal end of the phalange can occur, the distal end of the fingers takes bud shape, the palm becomes sunken inward, and infection often occurs to the nail grooves.

The pediatric patients with Apert symptom have two mutation sites on the gene that encodes the FGF receptor 2 (FGFR2), namely, the Ser252Trp and Pro253Arg mutations, and both of the two sites are located at the III A exon region of FGFR2 locus. According to clinical observation, it is found that the manifestation of Ser252Trp mutation is mild cranial deformity and severe hand deformity, and the manifestation of Pro253Arg mutation is mild hand deformity and severe cranial deformity [12].

11.2.1.1 Clinical Manifestation

1. Acrocephalia: The early closure of coronal suture impedes the development of the cranium perpendicular to the coronal suture, so that the anteroposterior diameter of skull cannot be increased, but the development is along the transverse diameter; therefore, the manifestation is the high and wide forehead.
2. Facial deformity: The eye span is increased, the nose bridge is low and flat, the maxillary bone becomes protruding due to dysplasia, and the patient can also suffer from cleft palate.
3. Syndactylia: There is cutaneous syndactylia or bony fusion to varying extents; the symptoms in the middle, ring, and little fingers are common, and they occur symmetrically in both hands. Due to the syndactylia of phalangettes, the interphalangeal joint makes no movement. The phalange is short and the fingernails are wide and large and they cover the entire distal phalanx.
4. Others: The thumb is short with only one segment of phalange. The metacarpal bone is short, and the fundus of the fourth and fifth metacarpal bones becomes fused sometimes. In most cases, the patients suffer from the backwardness and internal organ deformities. There is an increase in intracranial pressure, and the accompanying symptoms can be headache, tic, exophthalmus and visual loss.

11.2.1.2 Classification

The Apert syndrome hand deformities can be divided into three types according to the degree of soft tissue and skeletal deformities. Different clinical surgical regimens can be decided according to the classification [1].

1. Type I [obstetrician's hand]: The plane of syndactylous fingers is flat; the thumb is independent; the index, middle, and ring fingers form the syndactylous complex; the syndactylia of fifth finger is relatively separated; and the tip can move. The thumb suffers radial lateral curvature deformity (auto-stop hand).
2. Type II [cup hand]: The thumb is relatively independent with the presence of radial lateral curvature, the transverse arch of hand becomes collapsed into the cup shape, the interdigit length reaches the gap of the metacarpophalangeal joint, and the syndactylia of the fifth finger is relatively separate.
3. Type III [rosebud hand or hoof hand]: All syndactylous fingers are closely connected into a mass, and endogenous fingers often occur, leading to infection. The thumb phalange is small and is surrounded by the index, middle, and ring fingers, but the thumb does not necessarily suffer radial lateral curvature deformity.

The hand deformity of Apert syndrome is special; compared with other hand deformities, it has a special therapeutic regimen. Its treatment should conform to the following principles: ① due to finger joint adhesion and no relative activity of the index, middle, and ring fingers, straight incisions can be made in separating the syndactylous fingers; ② the thumb and the fifth finger should be first separated; ③ finger separation can be performed at both sides simultaneously before the pediatric patient becomes 1 year old; ④ before the finger separation surgery, it is necessary to first deal with the infection problems induced by endogenous fingers; ⑤ triangular flaps can be used for nail groove formation; ⑥ the wound regions of fingers can be covered with skin grafts; ⑦ the thumb should be lengthened to the level of phalange; ⑧ the union of the fourth and fifth metacarpal bones should be separated to improve the object gripping functions.

11.2.1.3 Surgical Method

1. Reconstruct the first fingerweb so that the thumb has sufficient abduction space: The method such as continuous skin fascia release, adherent muscle lengthening, and carpometacarpal joint capsule incision can be adopted to increase the fingerweb area, and the allowed thumb abduction can be up to 45°. The stenosis of the first fingerweb of juveniles can be repaired through local skin flaps, such as four-flap Z-reshaping. As for the patients

with relatively severe stenosis of the first fingerweb with skin tissues, the hand dorsum rotation and advancement flaps or hand dorsum dilating flaps can be utilized for repair [13, 14].

2. Correction of thumb bending: Open wedge-shaped osteotomy is the first option to lengthen the thumb. Release of thumb fingerweb and thumb osteotomy should be performed simultaneously during the same surgery. After the fingerweb release and thumb wedge-shaped osteotomy for lengthening surgery, the patients often concomitantly suffer from skin defect at one side of the thumb, and Z-reshaping can be performed for the repair [3].
3. Syndactylia separating: The release is usually completed in different stages; the release of fingerweb and the position of index finger decide the surgical regimen, and the nerve vascular bundles in syndactylous fingers generally do not affect the formulation of surgical regimen. The severe syndactylia deformity is mainly treated through separating the phalanges and nail beds with fusion at the tips, the treatment is achieved through surgeries in different stages, and the complex syndactylia is transformed into the simple syndactylia, so that thorough correction can be achieved. If no finger with sufficient functions can be formed after the seriously deformed fingers are separated, then consideration can be given to removal of the fingers.

   As for type I syndactylia, separation of the first fingerweb is seldom needed; if necessary, flaps can be designed to improve the thumb web functions. As for type III syndactylia with serious deformities, the phalanges and fingernails are very small, and there is no skin tissue that can be used; at this time, it is very necessary to fully utilize the skin around the fingerweb through design. The lengthening of the thumb at the age of 3–5 can effectively increase the thumb web functions. As for the surgical design of each fingerweb, there is no fixed optimal regimen, and the surgeons can select the design regimen according to the basic principles. As the skin at the palm and hand dorsum is insufficient, it is necessary during the design to leave sufficient skin to the anteroposterior junction of the fingerweb. The usual practice is to design flaps at the palm and hand dorsum and insert them into the opposite side [1].
4. Phase II surgery: After dactylolysis, with the growth and development of hands, it is necessary to reshape the scars in the skin grafting region and the asymmetric growth that occurs to other tissues. The thumb radial lateral curvature should be corrected before the patient is of 4–5 years old. Phase II osteotomy is also needed to separate the fourth and fifth metacarpal synostosis (Table 11.1).

Table 11.1 Treatment of hand deformities in Apert syndrome

Age	Surgery
1–6 months	Treat the nail groove infection, release the thumb web, and transform type III surgery into type I surgery
6–18 months	Separate the syndactylous fingers and release the joints
4–6 years	Correct the thumb lateral curvature, perform osteotomy and correction of metacarpal synostosis, reshape the skin scars, and reshape the nail beds
≥7 years	Correct the phalangeal deformities, reshape the skin scars, release the fingerweb, and reshape the joint

11.2.2 Poland Syndrome

11.2.2.1 Clinical Manifestation

This deformity was first reported in 1841, when Alfred Poland described the anatomic deformities of thorax and four limbs of a corpse, including the absence of major pectoral muscle, sternal head, and some serratus anterior muscles and the finger hypoplasia with web-shaped adhesion. Hundred years later, this file was browsed and studied in the same hospital, and this deformity was named Poland syndrome. The two manifestations of Poland syndrome are partial absence of major pectoral muscle and dysplastic web-shaped fingers (Fig. 11.4). The patients often suffer from the skeletal structural abnormalities in the ipsilateral thorax, including rib absence and scoliosis, and the patient can also suffer from thumb triphalangeal deformity [1–3, 15].

The etiological factors of Poland syndrome are unclear, and there is no genetic predisposition. The hand dysplasia degree can range from web-shaped finger deformities of the middle and ring indexes to complete loss of the fingers so as to affect the forearm and upper arm. Usually, the fingers in the middle (the index, middle, and ring fingers) are more apt to deformities; when serious dysplasia involves the first and fifth fingers, only the globular bulges of varying sizes are observed in the middle.

11.2.2.2 Treatment Principle

As the deformities in the pectoral muscles and thorax generally do not cause respiratory problems, the correction of thorax mainly serves the purpose of reshaping. For females, the absent breast can be selected for reconstruction or mammaplasty; for males, the broadest muscles of back can be selected for transposition to reconstruct pectoral muscles.

As the deformity degree varies, there are great differences in the treatment of hand deformities in Poland syndrome. For the patients with good finger development and

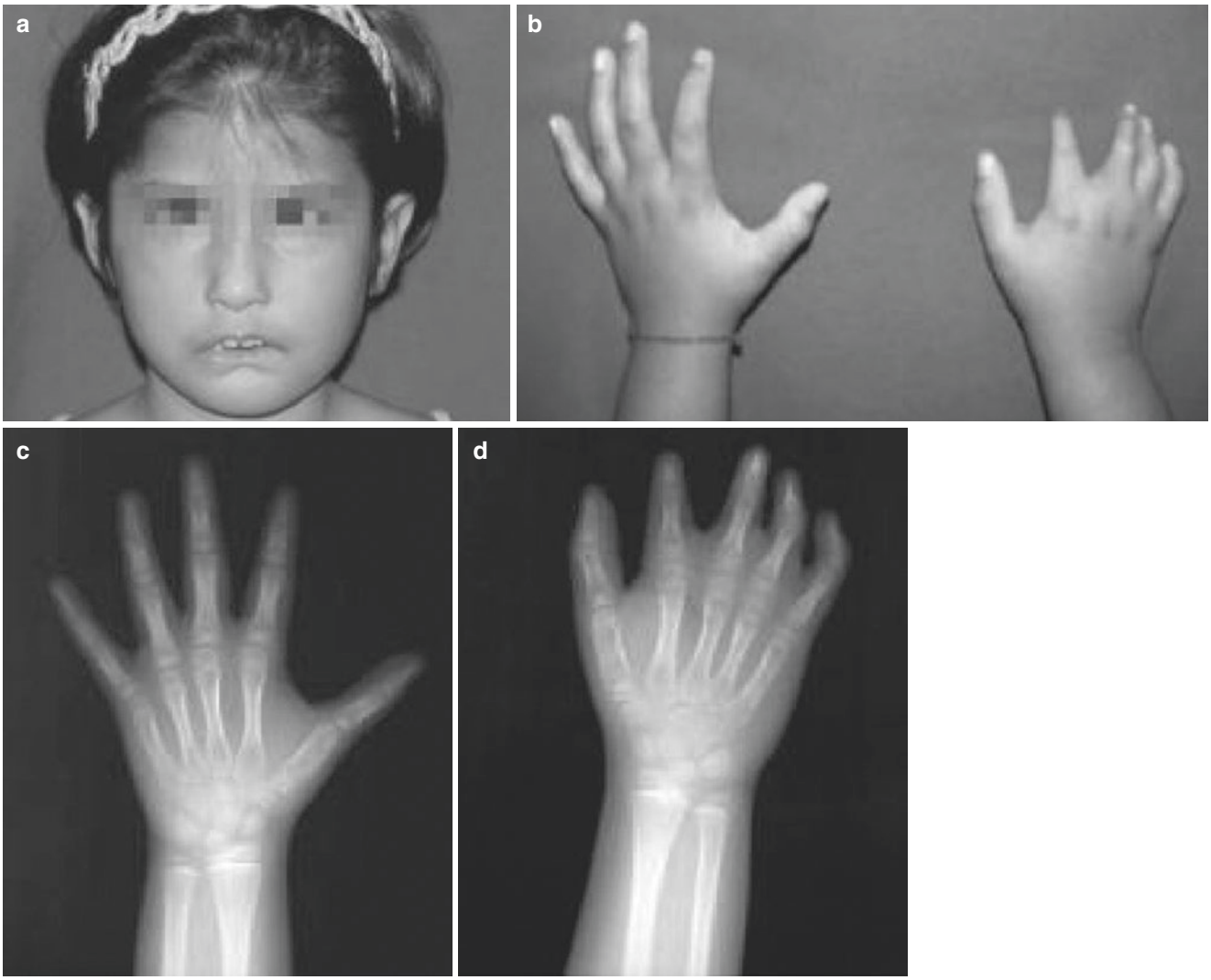

Fig. 11.4 Poland syndrome. (**a**) Facial deformity. (**b**–**d**) Hand deformities (Originate from Domingos AC. Poland-Moebius syndrome: a case with oral anomalies. Oral Diseases. 2004;10(6): 404–7). (Reprint with permission from JOHN WILEY AND SONS LICENSE TERMS AND CONDITIONS)

normal thumb, the fingers must be separated, and in the meantime the thumb web should be retained maximally; for the patients with serious dysplasia in the middle finger, transposition of marginal fingers can be performed for transplantation; for the fingerless or thumbless deformities, transplantation of toes can be performed to reconstruct the fingers.

There is no very special requirement on the syndactylous finger separation of Poland syndrome, and for the patients with relatively short fingers, it is necessary to design and fabricate deeper fingerwebs to lengthen the fingers; for the complicated deformities that need reconstruction, it is necessary to design individualized sequential treatment, and there is no fixed regimen for this type of surgery, the design by each expert varies, and many complicate techniques need to be employed [1–3].

11.2.3 Holt-Oram Syndrome

The patients with Holt-Oram syndrome have an autosomal dominant inheritance predisposition, and 85% of the patients have TBX5 gene mutation. Its incidence in females is high with a high severity, and the main manifestations are upper limb skeletal system deformity and cardiovascular system deformities.

As for the cardiovascular deformities, atrial septal defects are common, and the commonest type is type II ostium defect, followed by interventricular septal defect; other types include patent ductus arteriosus, transposition of great vessels, tubular artery abnormalities, aortic stenosis, pulmonary arterial stenosis, and tricuspid atresia. Arrhythmia includes degree I atrial ventricular block, incomplete right bundle branch block, atrial or ventricular premature contraction, etc.

The skeletal deformities are manifested as hyperdactylism and syndactylia of upper limbs. The radial bones are often involved with the manifestations of short radial bone and fusion of radio-ulna. The thumb mutation has such characteristics: the thumb is at the same level with the other four fingers, pollicization is observed, and the palm-opposition function disappears; one thumb is absent and at the opposite side is multi-segmented thumb. Carpal bones and metacarpal bones may suffer dysplasia. The patients may concomitantly suffer from deformities in other sites except the upper limbs, such as asymmetry of bilateral clavicles and scapulae, chicken breasts, chonechondrosteron, scoliosis, humpback, scapula, cheilopalatognathus, and bow-shaped palate. There are also deformities in digestive system and urinary system.

In terms of treatment, the surgical time should be decided according to the severity of heart defects, and if necessary, interventional therapy can be selected. As for the ulnar-radial fusion, the surgery is unnecessary for the patients with pronation of less than 30°, because it can be compensated; for the patients with pronation of more than 60°, due to the presence of obvious functional lesions, it is necessary to perform the separation of ulnar-radial fusion; in case of pronation of 30°–60°, it is necessary to decide whether surgery should be performed or when to perform the surgery according to the degree of functional lesion and the requirements on appearance. The surgically separated ulnar and radial bones can experience readhesion, and the fat or fascia obstruction for end breaking can be adopted after the intraoperative separation to reduce readhesion [1–3, 15, 16].

Hand deformities can be classified according to the status of thumb development, and corresponding surgical modes can be chosen.

11.2.4 Whistling Face Syndrome

Whistling face syndrome is also called cranio-carpo-tarsal syndrome; its deformities include microstomia, windmill armlike hand flexion deformity, and golf club foot deformity; the patients also suffer from limitation in height development, and the patients have autosomal dominant inheritance predisposition (chromosome 11p15.5). Due to the enhanced muscular tension, the patients often have the manifestations of forehead fullness and visor-like face, with the whistling-like microstomia, deep pitting of both eyes, wide nose bridge, epicanthal fold, loxophthalmus, small palpebral fissures, small nose, low development of nasal wing, medium length of the body, H-shaped skin dimples on the face, high dome of palate, lingula, limited lingula movement, and nasal sound; the hand and foot deformities include hand ulnar deviation, external cutaneous thumb, finger flexion, thick skin covering the curved surface of adjacent digits, equinovarus, toe contracture, and vertical talus. The patients often concomitantly suffer from inguinal hernia and incomplete orchiocatabasis, and the intelligence is within the normal range in most cases. The fetuses are generally at the breech presentation, and labor is difficult. Vomiting and dysphagia induce insufficient fetal growth, which can induce early death.

The treatment should focus on the improvement of quality of life, including expanding the oral fissure to improve food intake and respiration functions, correcting crossfoot to produce some walking functions, and correcting the spinal column deformities until the patient can sit. As for the correction of hand deformities, the release of flexor insertion and the incision and release of the anterior side of the wrist joint can be performed during the early stage [2, 3, 17].

11.2.5 Thrombocytopenia-Absent Radius Syndrome

The patients with thrombocytopenia-absent radius syndrome often have autosomal recessive inheritance predisposition, and the current pathogenicity still remains unclear. It is apt to occur to females. The pediatric patients may have normal routine blood test at birth, but there is a decrease in platelet count, and the drop is sharp within the first year after birth. The pediatric patients have the manifestations of body shortness, developmental retardation, and complete absence of radial bone, possibly with ulnar defects; dysplasia in humeral bone, scapula, or clavicle; syndactylia; hip joint dislocation; and deformities in femoral bone, shank bone, and feet (Fig. 11.5). Compared with patients with other radial bone absence syndrome, the pediatric patients with this syndrome often have the thumb with relatively complete functions, and the thumb bends and extends toward the lateral side; the patients often have hand internal muscles with a certain abduction function; the ulnar bone is short and deformed, and 10–20% of the patients have the ulnar absence to varying degrees. Fifty percent of the patients suffer various kinds of lower limb deformities; in addition, there are congenital deformities in cardiovascular system. First, the thrombocytopenia and bleeding should be treated; when the pediatric patients grow older, corrective surgery can be performed according to relevant principles [2, 3, 19].

11.2.6 Carpenter Syndrome

Carpenter syndrome, also called acrocephalopolysyndactyly, falls into the category of autosomal recessive inheritance. The pediatric patients have the manifestations of acrocephalosyndactyly, widened eye span, obesity, and anorchia. Due to the long-term increase in intracranial pressure, headache, diminution of vision, tic, and dysgnosia can occur. Preaxial hyperdactylia is observed, syndactylia is mild, and soft tissues are connected in most cases [2, 3, 20] (Fig. 11.6).

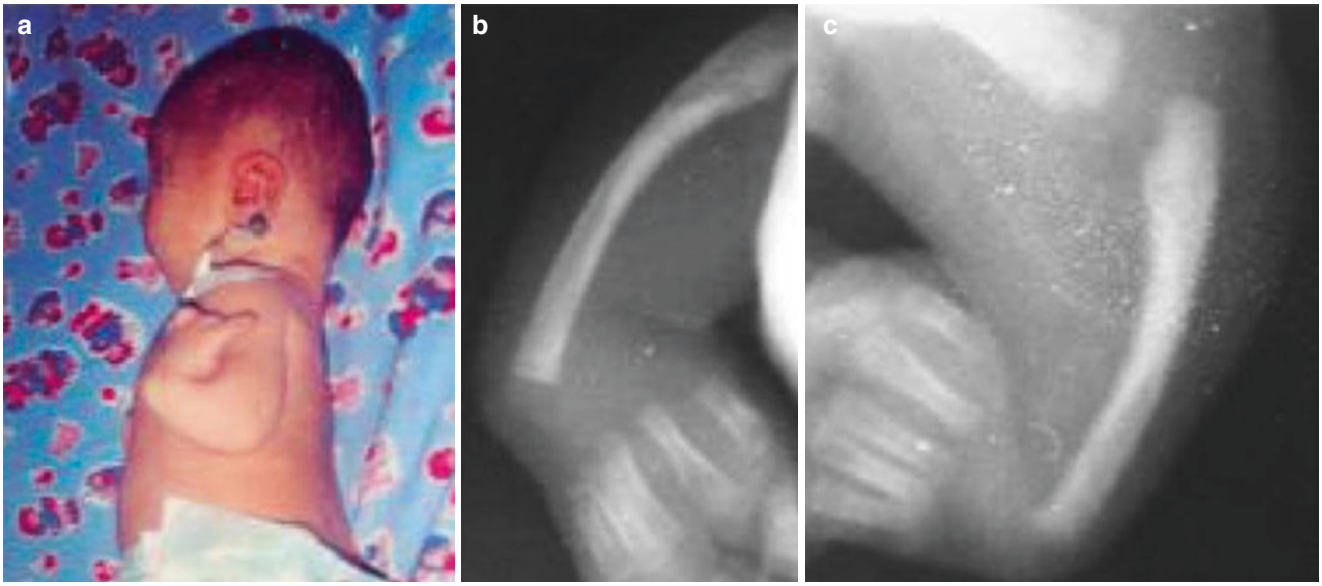

Fig. 11.5 Thrombocytopenia-absent radius syndrome. (**a**) Deformed appearance. (**b**, **c**) X-ray findings (Originate from Pavlenishvili IV. Thrombocytopenia-absent radius - TAR-syndrome. Georgian Medical News. 2011; 193: 86–8). (Reprint with permission from OXFORD UNIVERSITY PRESS LICENSE TERMS AND CONDITIONS)

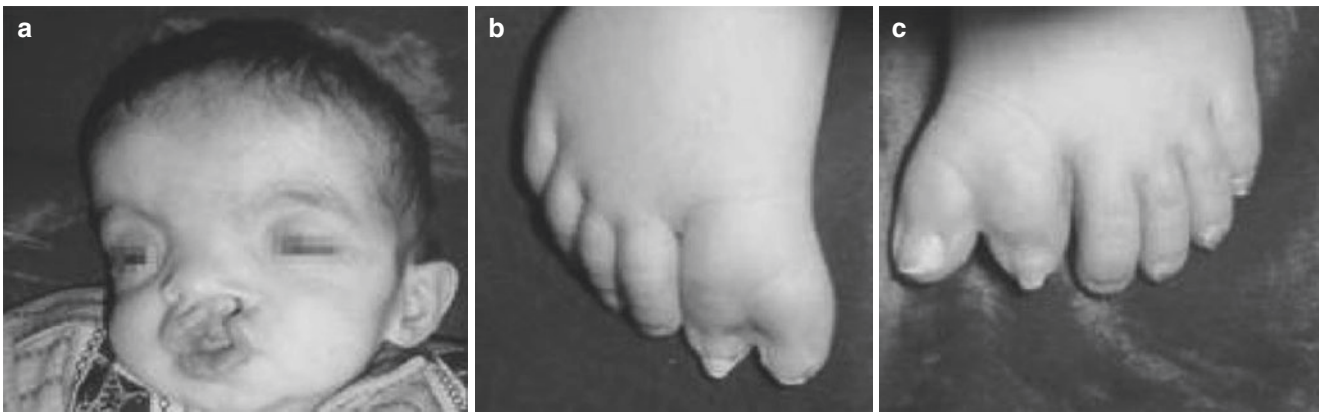

Fig. 11.6 Carpenter syndrome. (**a**) Head and face deformities. (**b**, **c**) Limb deformity (Originate from Wani, Abrar A. Carpenter's syndrome: A rare craniofacial dysmorphic syndrome. INDIAN JOURNAL OF PEDIATRICS. 2009;76(9): 972–972). (Reprint with permission from JOHN WILEY AND SONS LICENSE TERMS AND CONDITIONS)

The treatment should focus on the craniotomy to lower the intracranial pressure. Syndactylia and hyperdactylia can be treated according to the treatment principle of hand deformities.

11.2.7 Treacher Collins Syndrome

Treacher Collins syndrome, also called lower mandible facial bone dysplasia, falls into the category of autosomal dominant inheritance. The pediatric patients have the manifestations of downward inclination of palpebral fissures, dysplasia of cheek bone and jaw bones, small external auditory canal, and cecum sinus tract between the mouth angle and external ear. The patients often concomitantly suffer from upper limb deformities, such as syndactylia, thumb dysplasia or absence, upper ulnoradial joint fusion, and radial bone absence.

The treatments include the pinnaplasty to improve hearing; facial reshaping can be performed to improve the facial deformities, and the upper limb and spinal column deformities can be treated according to relevant treatment principles [3].

11.2.8 Pierre Robin Syndrome

Pierre Robin syndrome, also called lower mandible dysplasia, is genetically related, and it falls into the category of dominant inheritance concomitantly with different penetrances.

The pediatric patients have the manifestations of small low mandible, which shrinks back. In severe cases, the patients are accompanied by airway obstruction induced by glossoptosis. The patients often suffer cleft palate, cardiovascular deformities, and abnormalities in eyes and brain. The limb deformities include syndactylia, brachydactylia, lateriflection of fingers, and contracture of multiple joints.

The corresponding surgical correction can be adopted to treat the deformities in four limbs according to the patient's conditions [3].

11.2.9 Oculo-Mandibulo-Facial Syndrome

Oculo-mandibulo-facial syndrome is also called Hallermann-Streiff syndrome. Its etiological factors are clear. It may be induced by the embryonic developmental disorder at 5 weeks of development. The pediatric patients have the manifestations of skull dysplasia, bulging forehead, little birdlike appearance, and retrognathia; the eyes have the characteristic of cataract, microphthalmus, visual disorder, and nystagmus; the patients suffer dental deformities; the patients can also have dwarf manifestations to varying extents; the limb deformities include claw hand, syndactylia, radial and ulnar bony connection, spina bifida, and hip join dislocation.

There is no especially effective treatment for this disease. As for the patients with affected functions in four limbs, corrective surgery can be performed according to relevant principles [3].

11.2.10 Oculo-Dento-Digital Syndrome

Oculo-dento-digital syndrome falls into the category of autosomal dominant inheritance and is induced by ectosermic developmental disorder. The pediatric patients have the manifestations of capitulum, small and thin nasal wing, anteversion of nostrils, microphthalmus, small palpebral fissure, narrowed eye span, visual disturbance, and adamantine dysplasia; common hand deformities are syndactylia, hyperdactylism, middle finger absence, or flexion deformity [3].

For patients with affected functions in four limbs, corrective surgery can be performed according to relevant principles.

11.2.11 Oro-Facial-Digital Syndrome

Oro-facial-digital syndrome is a genetic disease and only occurs to women. The pediatric patients have the manifestations of olecranon, dysplasia of nasal alar cartilage, cleft lip, lobulated tongue, and thickening and proliferation of frenulum linguae; hand deformities include syndactylia, brachydactylia, and deviating fingers.

Treatment includes facial reshaping and hand corrective surgery [2, 3, 21].

11.2.12 Oto-Palato-Digital Syndrome

Oto-palato-digital syndrome is hereditary with unclear etiological factors and only occurs to males. With multiple bone dysplasias as the basis, it often has the manifestations of conductive hearing loss, cleft palate, body shortness, and dwarf; hand and foot deformities include short, small, and flat thumb (big toe), enlarged tips of the three fingers in the middle, and the little finger bending toward the lateral side. X-ray film can indicate the changes in epiphysitis at the fundus of the second and third metacarpal bones, and abnormalities are seen in the size and shape of metacarpal bones and phalanges.

Surgery is not necessary if the hand and foot deformities do not influence their functions [3].

11.2.13 Weill-Marchesani Syndrome

Weill-Marchesani syndrome falls into the category of autosomal recessive inheritance. The eye symptoms often occur before the patients become 10 years old, manifested as myopia and crystalline lens deformities; the four limbs and fingers (toes) are short, and the X-ray can show symmetric shortening and widening of metacarpal bones and phalanges and delayed ossification of carpal bone, metatarsal bone, and phalanges; the patients are often short (Fig. 11.7).

The correction of deformities in four limbs can be in accordance with the functions of the limbs and by referring to the corresponding principles [3, 22].

11.2.14 Rubinstein-Taybi Syndrome

Rubinstein-Taybi syndrome, also called broad thumb-hallux syndrome, may be genetically related. The pediatric patients have the manifestations of wide and short thumb (big toe), which take spoon shape or short club shape, and thick and large phalangettes. X-ray can show the thickening of the first metacarpal bone, the first metatarsal bone, and the thumb (big toe), rib fusion, and short neck of femur (Fig. 11.8). The facial deformities include low ear position and ear deformities, downward inclination of palpebral fissure, many eyelashes, eyeball downbeat, possible cataract, small nose, and sharp chin. Most of the pediatric patients are short with low intelligence [3, 23].

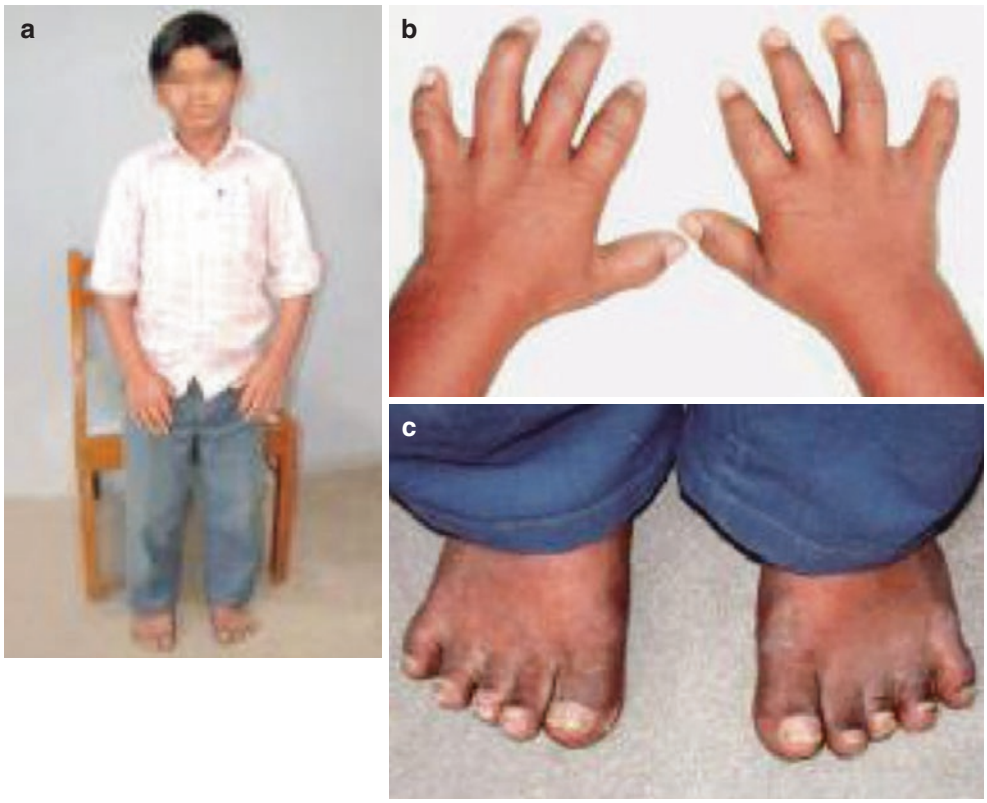

Fig. 11.7 Weill-Marchesani syndrome. (**a**) Microsoma. (**b**) Brachydactylia of fingers. (**c**) Brachydactylia of toes (Originate from Chu BS. Weill-Marchesani syndrome and secondary glaucoma associated with ectopia lentis. Journal of the Australian Optometrical Association. 2006; 89 (2): 95–9)

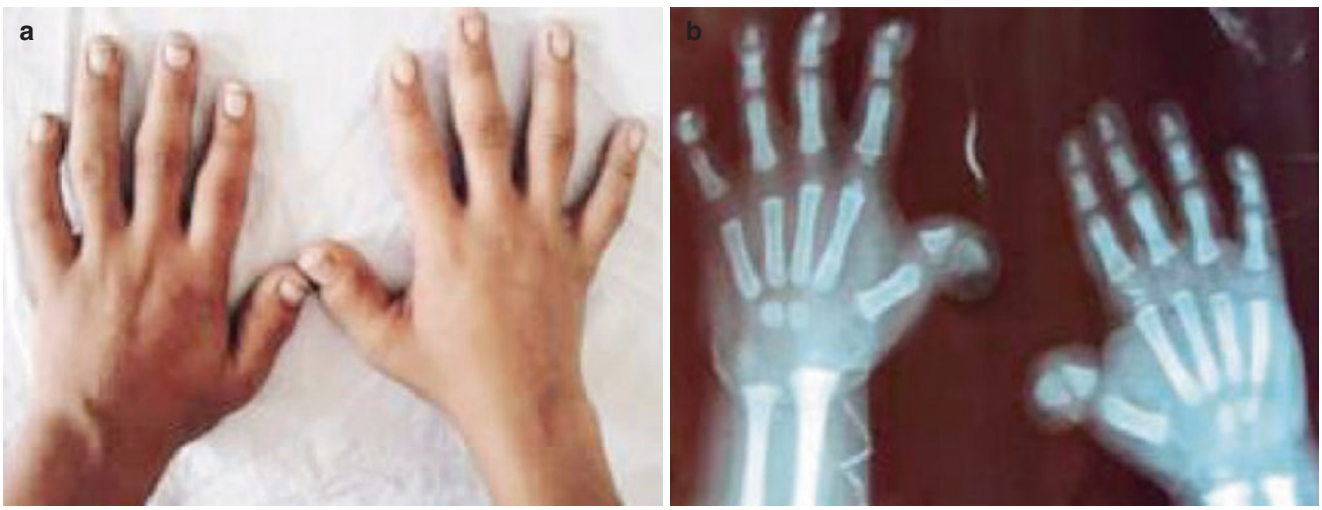

Fig. 11.8 Hand deformities of Rubinstein-Taybi syndrome (Reprint with permission from Kumar, Suresh. Rubinstein-Taybi syndrome: Clinical profile of 11 patients and review of literature. Indian Journal of Human Genetics. 2012;18(2):161–166)

11.2.15 Smith-Lemli-Opitz Syndrome

Smith-Lemli-Opitz syndrome, also called capitulum, micrognathia, and dactylion syndrome, falls into the category of autosomal recessive inheritance. The pediatric patients have the manifestations of dwarf, low intelligence, capitulum, wide nose bridge, anteversion of nostrils, ptosis, small and back-shrinking of lower mandible, high pillars of fauces, and scoliosis. Hand and foot deformities include too short and small thumb and middle finger, deviation of little finger, syndactylia of the second and third toes, and adduction and entropion of metatarsal bone (Fig. 11.9).

11.2.16 Laurence-Moon-Bardet-Biedl Syndrome

Laurence-Moon-Bardet-Biedl syndrome, also called sexual infantilism, retinitis pigmentosa, or hyperdactylism, falls into the category of autosomal recessive inheritance. It can be the secondary sex gland hypofunction caused by functional disorder of hypothalamus, with the manifestation of sex hypofunction and no appearance of secondary sex characters during the puberty. The pediatric patients often suffer from attenuation in visual acuity and even loss of sight, shortness of body, hyperdactylia, or syndactylia, with intelligence development disorder (Fig. 11.10).

The treatment should be mainly the sex hormone replacement therapy, and the deformities of four limbs can be surgically corrected according to relevant principles [3, 25].

11.2.17 Cornelia de Lange Syndrome

Cornelia de Lange syndrome, also called Amsterdam type dwarfism, may fall into the category of autosomal dominant inheritance. The pediatric patients may have the manifestations of dwarf, low intelligence, capitulum, hairiness, short upper limbs, and abnormal texture of hand skin. X-ray film can show delayed development of generalized epiphysis,

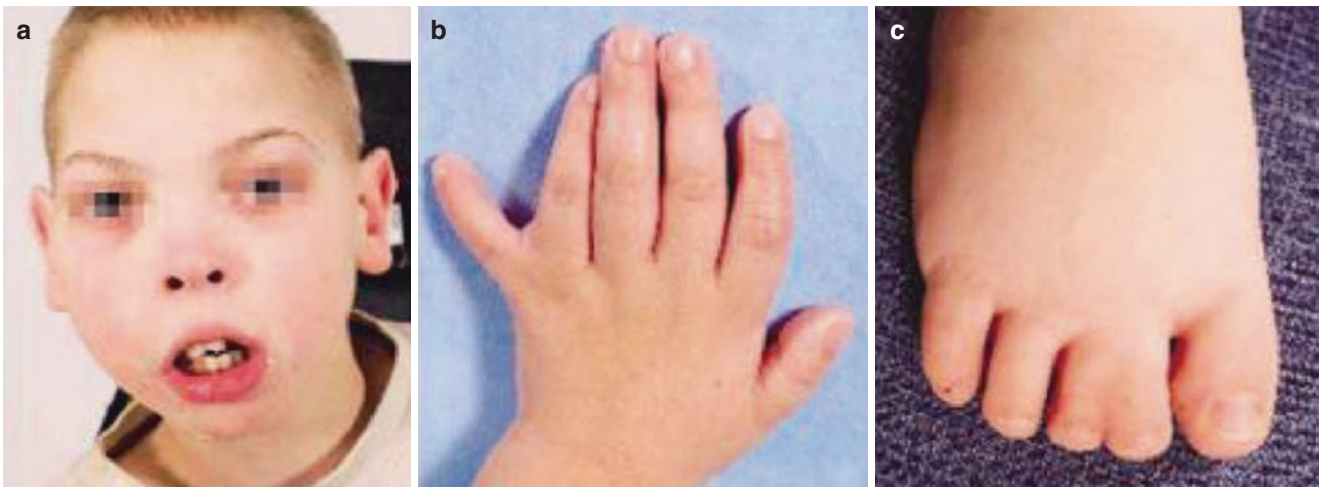

Fig. 11.9 Smith-Lemli-Opitz syndrome. (**a**) Face. (**b**) Hand deformities. (**c**) Foot deformity. The limb deformities can be surgically corrected according to relevant principles [3, 24] (Reprint with permission from Porter FD. Smith-Lemli-Opitz syndrome: pathogenesis, diagnosis and management. European Journal of Human Genetics. 2008;16 (5):535–41)

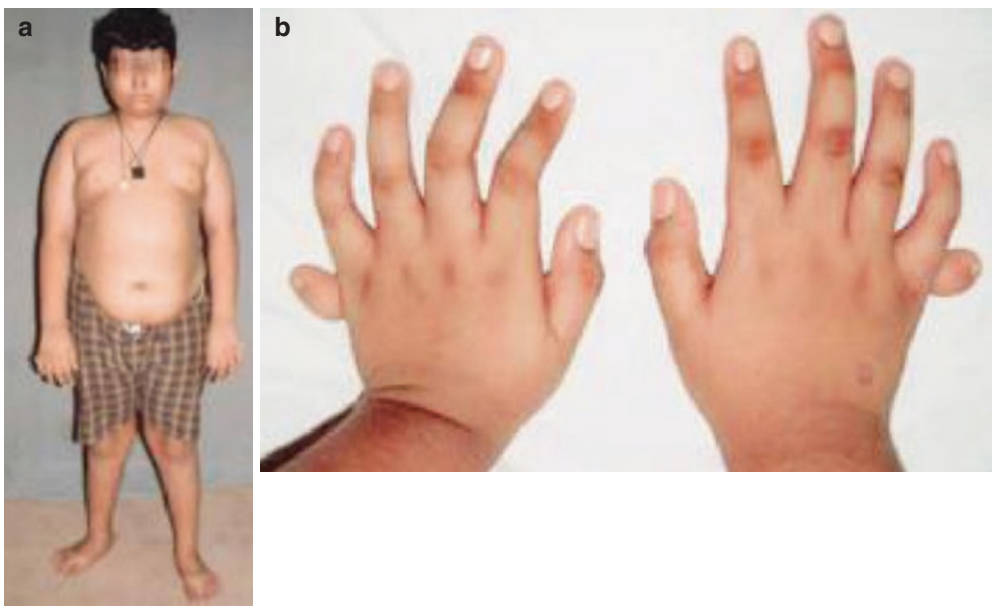

Fig. 11.10 Laurence-Moon-Bardet-Biedl syndrome. (**a**) Body form. (**b**) Hyperdactylism (Originate from Sahu JK. Laurence-Moon-Bardet-Biedl syndrome. Journal of the Nepal Medical Association. 2008; 47 (172): 235–7)

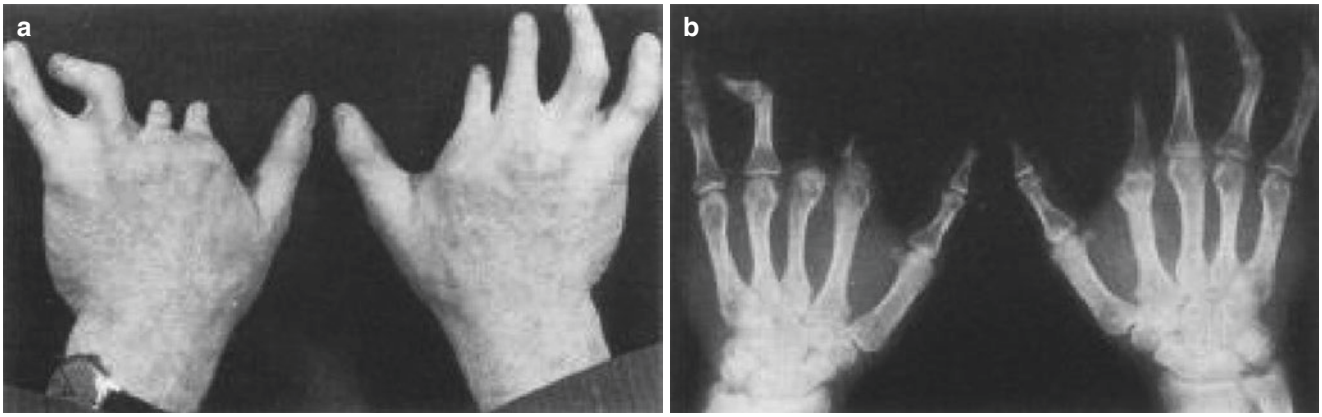

Fig. 11.11 Hand deformities of Aglossia-adactylia syndrome (Reprint with permission from Nevin NC. Aglossia-adactylia syndrome. Journal of Medical Genetics. 1975; 12 (1): 89–93)

dislocation of radial capitulum, flexion contracture or rigidity of elbow joint, deviation of little finger, partial absence of finger (ulnar side), fusion of carpal bone, dislocation of the proximal segment of the thumb, and dislocation of hip joint.

Surgical correction can be performed if the deformities of four limbs and joints can affect their functions [3].

11.2.18 Aase-Smith Syndrome

Aase-Smith syndrome, also called congenital anemia-triphalangeal thumb syndrome, may fall into the category of sex-linkage recessive heredity. It only occurs to males with the manifestations of anemic complex, presence of triphalangeal thumb, mild radial hypoplasia, narrow shoulders, interventricular septum, delayed fontanel closure, hepatosplenomegaly, and poor bone marrow hyperplasia.

Surgical correction can be performed according to relevant principles if the deformities of four limbs can affect their functions [3].

11.2.19 Aglossia-Adactylia Syndrome

Aglossia-adactylia syndrome has unclear etiological factors. The pediatric patients have the manifestations of facial deformities; small, sharp, and narrow faces (like a bird-shaped face); and ateloglossia or aglossia, concomitantly with partial or complete absence of fingers and toes or fingers with absent fingernails (Fig. 11.11).

Corrective surgery can be performed according to the need in functions [3, 26].

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Hand Hemangiomas and Vascular Malformations

12

Hui Chen, Xiaoxi Lin, Lei Chang, Gang Ma, Yunbo Jin, and Xi Yang

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Hemangioma and vessel deformities (including vascular malformations and lymphatic vessel deformity) are very common congenital or acquired lesions and are visible in any site of the body surface. About 60% of them are distributed in the head and neck, about 20% of them are distributed in the four limbs, and the proportion of hand lesions is not explicitly reported.

As the onset sites are wide, the patients with such diseases cannot be widely found in different departments, such as department of plastic surgery, department of oral and maxillofacial surgery, department of vascular surgery, dermatological department, department of interventional radiology, department of orthopedics, and department of hand surgery. In the past, the interdisciplinary exchange and fusion were not close, limitations to varying degrees could be found in the strategies of diagnosis and treatment, so most patients did not have access to accurate diagnosis and proper treatment; during recent years, with the constant deepening of the research on

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these diseases, a transdisciplinary consensus has been reached in many problems; therefore, it has become an international tendency that the medical teams from different disciplines work together to complete the diagnosis and treatment of such diseases. Currently, although the domestic professional academic conferences on hemangioma and vessel deformities tend to be frequent, a great number of professional academic organizations have been successively established, jointly devoted to the promotion of new viewpoints and new technologies, and they have promoted the cross-disciplinary research and fusion, but a quite number of surgeons in multiple disciplines still have a vague or old understanding on such lesions, obviously wrong diagnosis and treatment occur commonly, resulting in delay and aggravation in patient's conditions and the difficult-to-repair secondary deformities [1, 2].

Hands have very delicate histological structures and important functions; therefore, the diagnosis and treatment of hand hemangioma and vessel deformities have their uniqueness, and particular attention should be paid to the balance between functions and appearance. Improper treatment often leads to the loss of both hand functions and appearance, severely decreasing the quality of patient's survival, and this is what both the doctors and the patients are unwilling to encounter. Therefore, this chapter will give an introduction to the basic problems and latest updates in the diagnosis and treatment of hand hemangioma and vessel deformities for the purpose of common exchange and improvement.

12.1 Classification of Hemangioma and Vessel Deformities

Hui Chen and Xiaoxi Lin

Before the 1980s, people's understanding of hemangioma and vessel deformities is vague. According to modern opinion, the greatest confusion at that time is no differentiation between the vascular endothelial cell-derived true neoplasm and vascular and lymphatic malformations induced by vascular dysplasia. "Hemangioma" is used to collectively call various diseases and cannot accurately reflect the nature of lesions, resulting in obvious confusion in treatment and big disorder in academic exchange. Therefore, when the early literature (even some recent domestic literature) is reviewed, there must be a very clear understanding of the modern opinion so that the types of the cases reported in the literature can be discriminated among and the reliability of the method or opinion can be judged [3–5].

The confusion in the understanding of the nature of tumor and deformities also results in the absence of scientific and uniform criteria to classify vascular lesions, and this situation remained until 1982 when a fundamental change occurred. That year, Professor Mulliken from the Department of Plastic Surgery of Children's Hospital Boston of Harvard University, USA, first proposed the classification method on the basis of the biological characteristics of vascular endothelial cells; he classified vascular lesions into hemangioma and vascular deformity and expounded the most fundamental differences between the two: the vascular tumor had abnormal proliferation in vascular endothelial cells, while vascular malformations had no such a phenomenon. This opinion is widely accepted and becomes the basis of modern classification criteria, and this book has become one of the most classic ones in the history of plastic surgery [6, 7].

In 1992, the International Society for the Study of Vascular Anomalies (ISSVA) was established in Budapest, the capital city of Hungary. At the conference in 1996, ISSVA established a relatively sound classification system (Table 12.1) which was widely adopted and has become a common language for different disciplinary researchers all over the world.

In this classification system, the vascular lesions are divided into vascular tumor and vascular malformation, and the latter is further categorized into low-flow, high-flow, and complex

Table 12.1 ISSVA classification of vascular lesions

Vascular tumor	Vascular malformation
Infantile hemangioma	Low-flow vascular malformation
Congenital hemangiomas (RICH and NICH)	Capillary malformations: including port wine stain, telangiectasia, and angiokeratoma
Tufted angioma (with or without Kasabach-Merritt syndrome)	Venous malformations: blue rubber bleb nevus syndrome, familial cutaneous and mucosal venous malformation, glomuvenous malformation, and Maffucci syndrome
	Lymphatic malformation
Kaposiform hemangioendothelioma (with or without Kasabach-Merritt syndrome)	High-flow vascular malformations
	Arterial malformation
Spindle cell hemangioendothelioma	Arteriovenous fistulae
Rare hemangioendothelioma: epithelioid hemangioendothelioma, composite hemangioendothelioma, retiform hemangioendothelioma, polymorphous hemangioendothelioma, Dabska tumor, lymphangioendotheliomatosis	Arteriovenous malformations
	Complex combined vascular malformations
Dermatologic acquired vascular tumors: pyogenic granuloma, targetoid hemangioma, glomeruloid hemangioma, and microvenular hemangioma	CVM, CLM, LVM, CLVM, AVM-LM, and CM-AVM

Originate from Odile Enjolras, Michel Wassef and Rene Chapot. Introduction: ISSVA Classification. Color Atlas of Vascular Tumors and Vascular Malformations. Cambridge University Press. 2007, 6

Note: ① *RICH* rapidly involuting congenital hemangiomas. ② *NICH* non-involuting congenital hemangiomas. ③ *C* capillary, *A* artery, *V* vein, *L* lymph, *M* malformation

Table 12.2 A comparison between the modern naming and traditional naming of angioma and vessel deformities

Modern naming	Traditional naming
Infantile hemangioma	Strawberry hemangioma
Port wine stain	Capillary hemangioma
Venous malformation	Cavernous hemangioma
Arteriovenous malformations	Racemose hemangioma
Lymphatic malformation	Lymphangioma

combined vascular malformations according to the differences in hemodynamics. In vascular lesions, five kinds of lesions are the commonest clinically; the naming methods go through the evolution from tradition to modern (Table 12.2), and these are lesions which will be mainly expounded in this chapter.

12.1.1 Infantile Hemangioma

Infantile hemangioma, previously called strawberry hemangioma, is the commonest infantile benign tumor. The patients with it have the history of characteristic natural diseases of rapid proliferation observed at birth or soon after birth and spontaneous subsidence at the age of about 1. Its typical manifestation is the bright red bulging enclosed mass. But some deep skin on the surface of angioma is nearly completely normal.

12.1.2 Port Wine Stain

Port wine stain, previously called capillary hemangioma, and also called naeus telangiectaticus and nevus flammeus, is a congenital capillary malformation or venular malformation with the manifestations of pink to purple patches with defined borders, and the lesions at the head and face will become proliferative and nodular after the patient becomes an adult.

12.1.3 Venous Malformation

Venous malformation, also called cavernous hemangioma, is formed due to the dilation by abnormally communicated thin-walled veins with the manifestations of purple-blue soft masses with a sensation of compression, and the volume can change with body position.

12.1.4 Arteriovenous Malformation

Arteriovenous malformation, previously called racemose hemangioma, is the tortuous and dilated vascular mass formed by the direct communication of arteries and veins. The surface skin temperature is high; the pulsation or tremor is obvious; serious complications can occur such as tissue necrosis, a large amount of bleeding, or congestive heart failure; and it is the most-difficult-to-treat type with a highest hazard and the highest risk.

12.1.5 Lymphatic Malformation

Lymphatic malformation, previously called lymphangioma, is induced by abnormally dilated lymph canals, and the percutaneous puncture can indicate the presence of light, yellow, and bright lymph fluid. It can be divided into macrocyst type and microcyst according to the size of cyst, and they are closely correlated with therapeutic effects.

12.1.6 Congenital Hemangiomas

Congenital hemangioma is classified into rapid involuting congenital hemangioma (RICH) and non-involuting congenital hemangioma (NICH) with the manifestations of obvious lesions just at birth, which presented the blue-purple hemisphere-shaped bulge and defined borders, red dilated vessels are visible on the surface, white areolae with lightened pigment are visible in the surrounding margins, and it subsides also completely at the age of around 1 or it does not subside for life long (Fig. 12.1). Its appearance and pathological and imaging manifestations are obviously different from those of infantile hemangioma. The main treatment is surgical resection (Fig. 12.2) [8, 9].

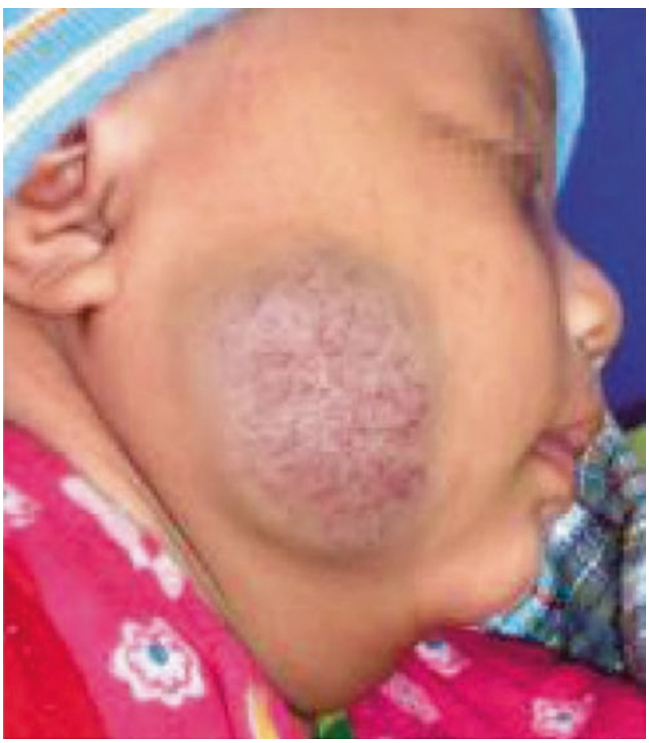


Fig. 12.1 Typical appearance of congenital hemangioma

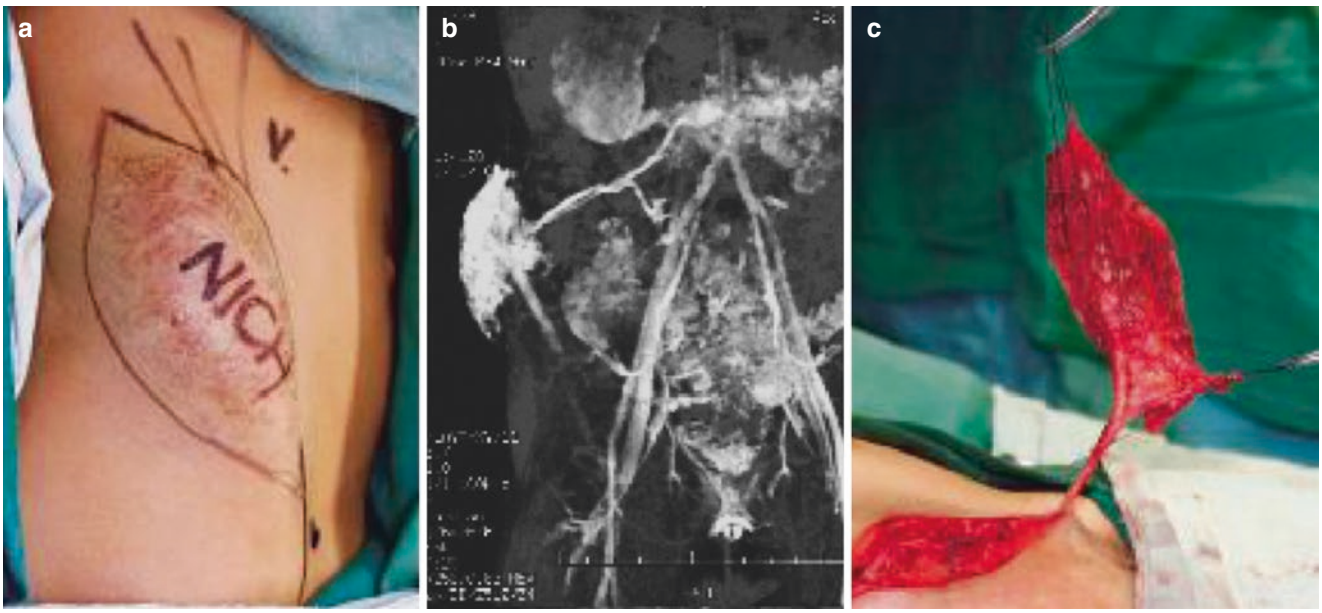


Fig. 12.2 Surgical resection of non-involuting congenital hemangiomas. (**a**) Non-involuting congenital hemangioma of the right iliac region. (**b**) MRA indicates the two fat blood-supplying arteries within the lesions. (**c**) Blood supply arteries are dissected during the operation, consistent with the MRA findings

12.1.7 Kaposiform Hemangioendothelioma

Kaposiform hemangioendothelioma (KHE) and tufted angioma (TA) are closely correlated with KM phenomenon. They present purplish red plaques or lumps, and the appearance has various manifestations (Figs. 12.3 and 12.4). The big solid tumor will capture and filter the platelets, resulting in thrombopenia; in severe cases, life can be threatened. A minority of patients can have natural subsidence, but the subsidence is slow and incomplete. In case of clinical detection of suspicious patients, routine blood test should be conducted. The treatment methods include local compression, hormonal therapy, antitumor drug (vinblastine and cyclophosphamide) treatment, and surgical treatment [10].

12.1.8 Pyogenic Granuloma

Pyogenic granuloma, also called lobulated capillary hemangioma, has no inflammatory changes or granuloma-like characteristics in pathology. It is apt to occur after mild local injuries or during the pregnancy, with the manifestations of fresh red soft bulge or nodules, generally the size does not exceed 2 cm, and the surface has ulcer and there is an easy tendency of bleeding after touch (Fig. 12.5). The treatment methods include laser therapy, electrochemistry treatment, and surgical resection, but there is a possibility of postoperative recurrence.

12.1.9 Angiokeratoma

Angiokeratoma is the cutinized purple plaques on the surface with defined borders, it is limited to the dermic superficial layer, and it does not invade the deep tissues (Fig. 12.6). According to the sites and ranges of the lesions, it can be classified into local type, acra type, scrotum type, papule type, and diffuse type. The treatment mainly includes laser and surgical resection.

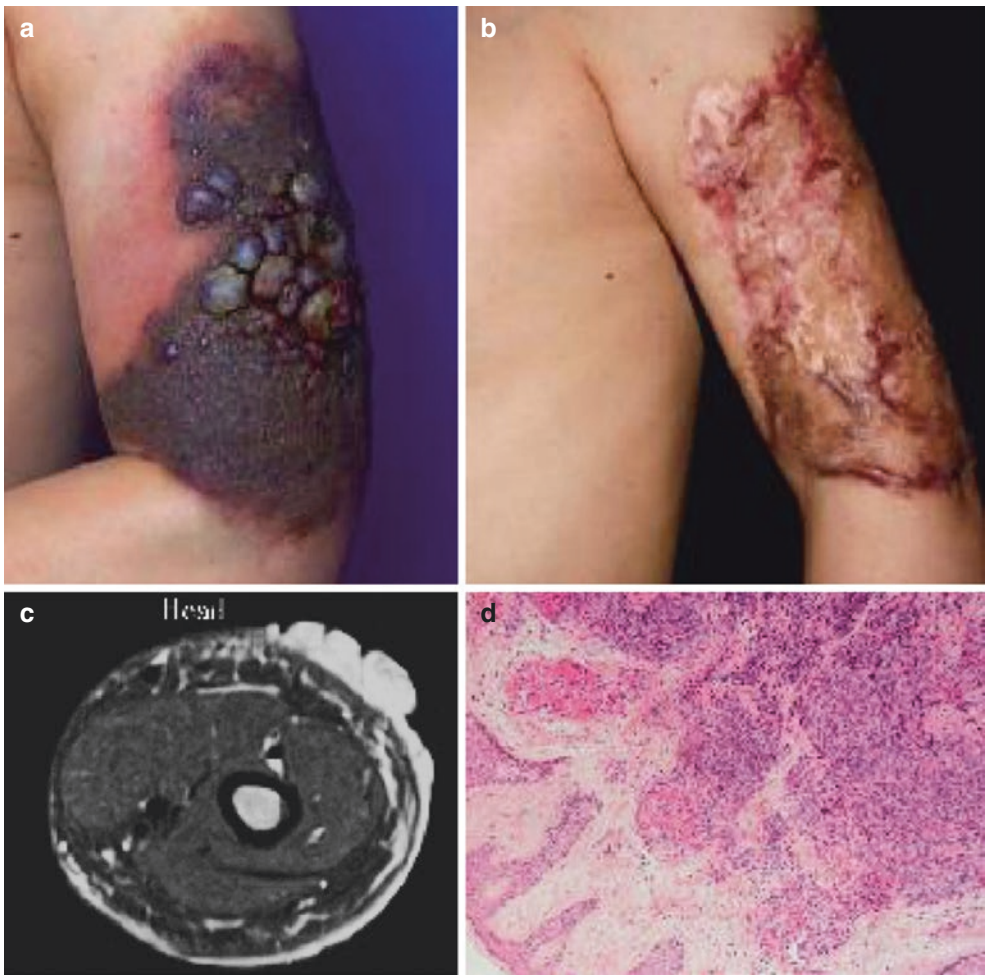


Fig. 12.3 Kaposiform hemangioendothelioma at the left upper arm with KM phenomenon. (**a**) The prunosus nodules at the left upper arm present cluster-shaped distribution with defined borders. (**b**) The patient underwent resection and dermatoplasty (the intraoperative bleeding was 1000 mL and the patient once experienced a large amount of repeated bleeding after operation). (**c**) MRI indicates that the lesions involve the superficial fascia layer. (**d**) The pathological examinations indicate the presence of vascular endothelial cell mass in corium and subcutaneous tissues

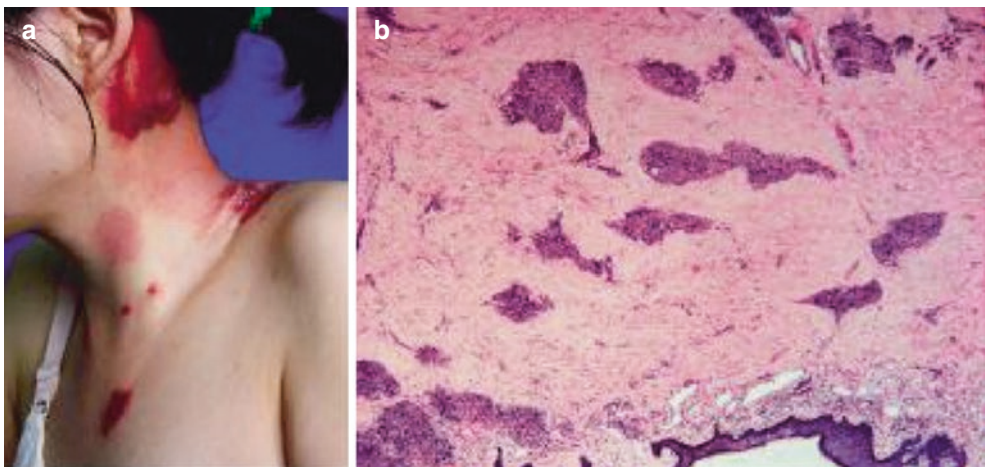
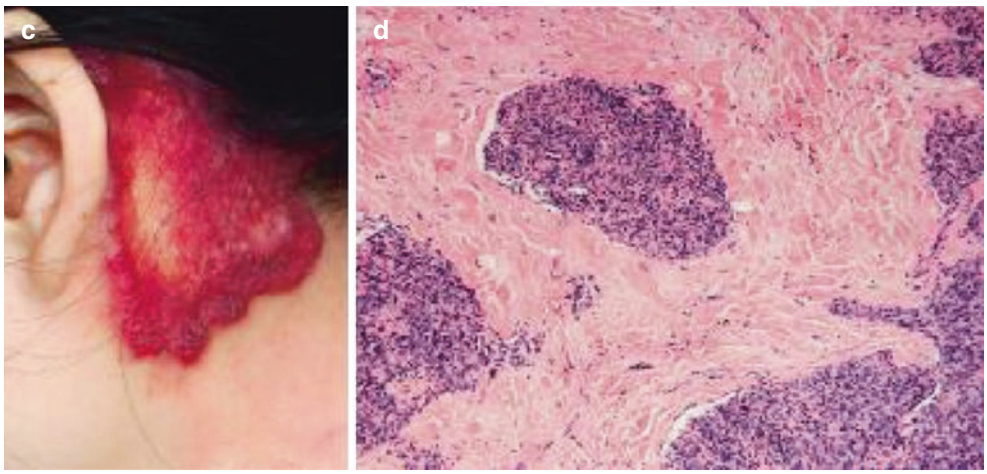


Fig. 12.4 Plexiform angioma at the posterior border of the left ear and the neck. (**a**, **c**) Red plaques are present at the posterior border of the left ear, the surrounding margins are bulging, the central region is plat, the borders are defined, and obvious tenderness exists. (**b**, **d**) The pathological examinations are characteristic, with the manifestations of endothelial cell mass with cluster-shaped distribution and defined borders on the background of collagenous fibers

Fig. 12.4 (continued)



12.1.10 Glomuvenous Malformation

Glomuvenous malformation (GM), previously called vascular malformation, is named due to the appearance of globuli cells on the deformed venous walls, and it can have the nature of both vascular malformations and tumor. The size ranges from several millimeters to several centimeters, and obvious tenderness or paroxysmal pain is its obvious characteristics. Solitary foci are mainly distributed in nail bed and can also be distributed in the limb ends, head, and neck; multiple foci are the autosomal dominant inherited diseases (Figs. 12.7 and 12.8).

The above classification is still in the middle of improvement because new lesions will be reported or new clinical phenomena and experimental studies will be found. Some lesions with close appearance should be differentiated cautiously to avoid wrong diagnosis and improper treatment (Fig. 12.9). There are still many unknown fields in the study of hemangioma and vessel deformities and further profound explorations are needed.

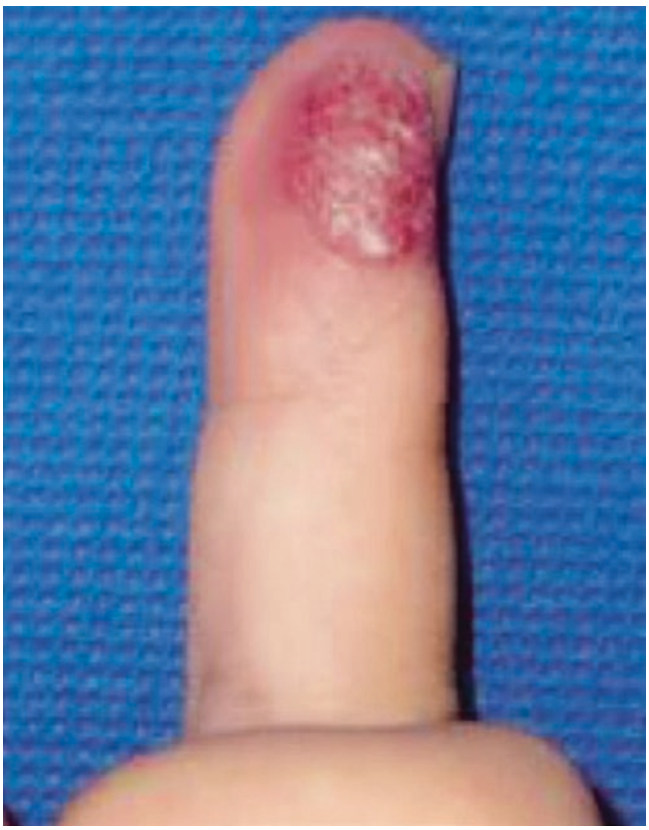


Fig. 12.5 Purulent granuloma is observed at the tip of the right index finger, presents purple lump, and is apt to bleed after touch

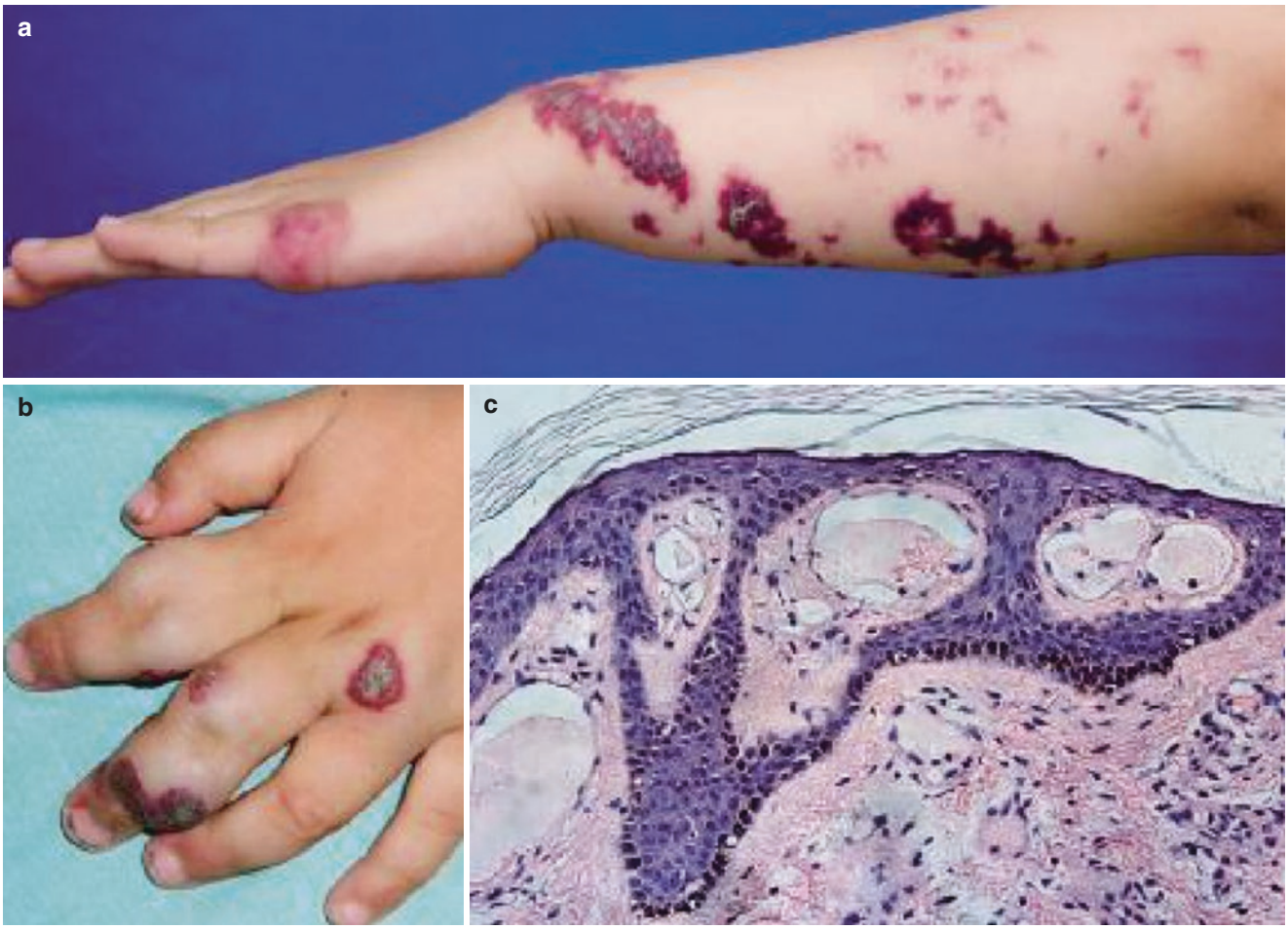


Fig. 12.6 Angiokeratoma. (**a**, **b**) The scatteredly distributed lesions at the forearm and hands are purple with defined borders and locally bulging plaques, obvious keratinization is observed on the surface, and they take white multilayer shape and become aggravated with increase in age. (**c**) The pathological examination indicates that the lesions are composed of a number of dilated capillaries and distributed at the dermal superficial layers and bulging toward the epidermis, and covering keratosic substances are visible

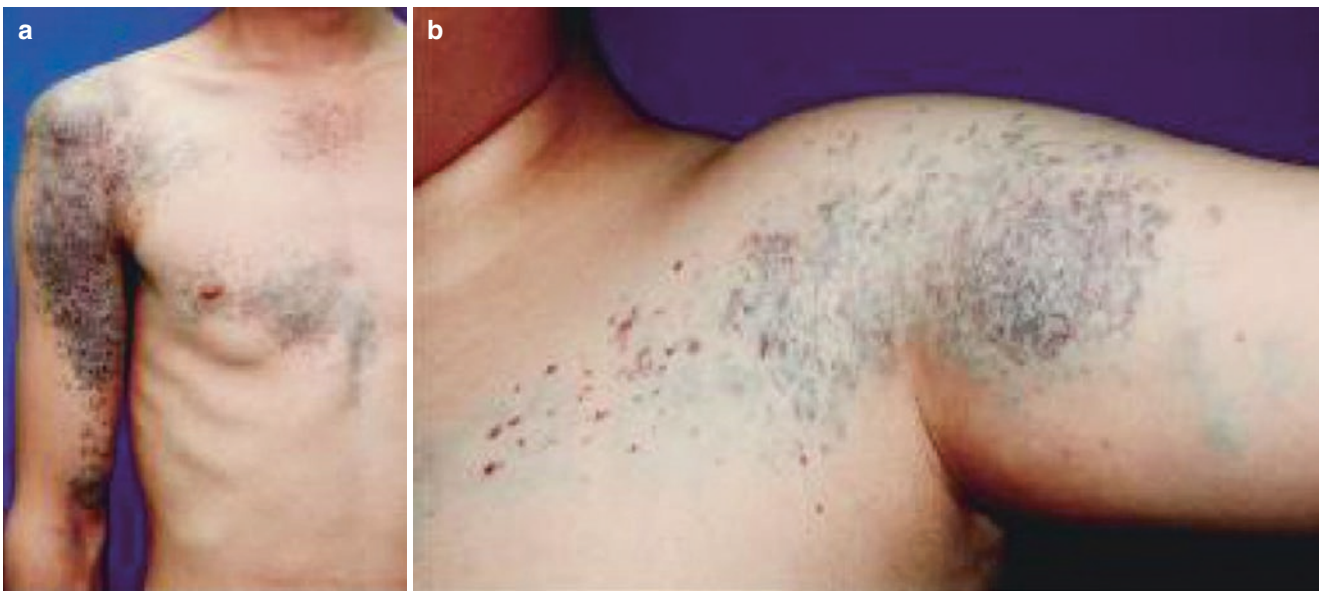


Fig. 12.7 Glomuvenous malformation venous malformation. (**a**–**c**) As for the foci at different body sites, purple cluster-shaped bulge is visible on the surface of deep lesions, and the body position test turns out negative. (**d**) Pathological sections can indicate multiple layers of globuli cells in the vessel walls

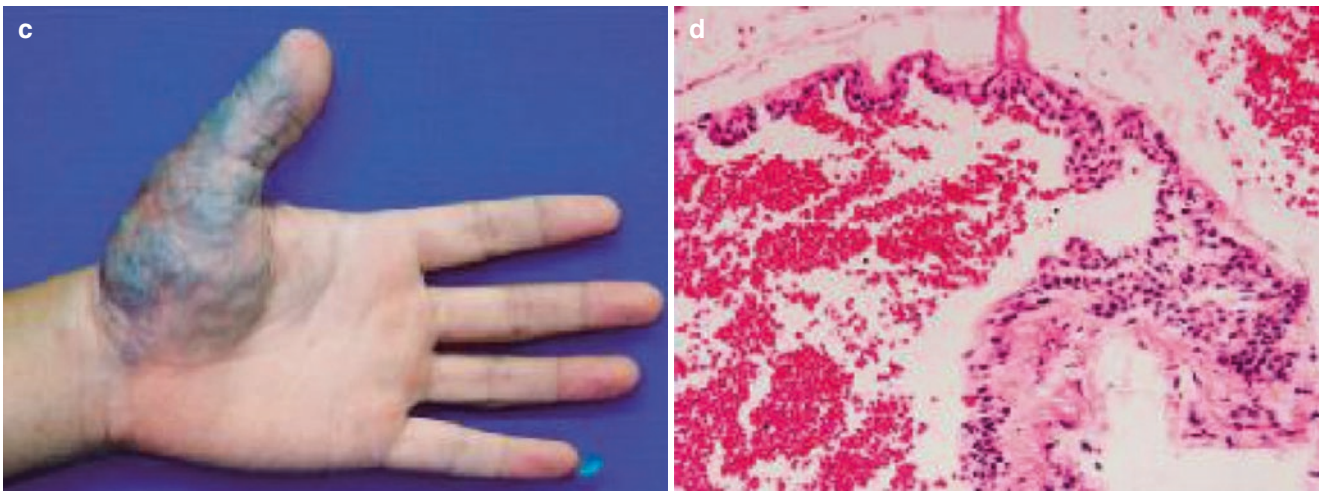


Fig. 12.7 (continued)

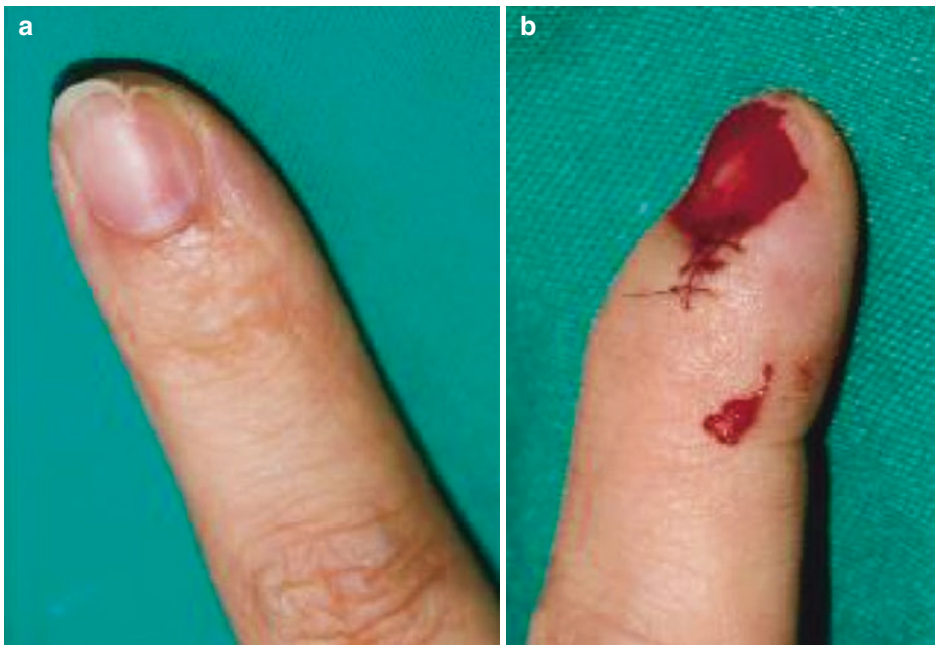


Fig. 12.8 Surgical resection of subungual glomuvenous malformation. (**a**) The subungual focus presents the color of lavender-blue, and sharp pain is felt after compression. (**b**) The complete focus is surgically resected and mung bean sized with a hard and tough texture

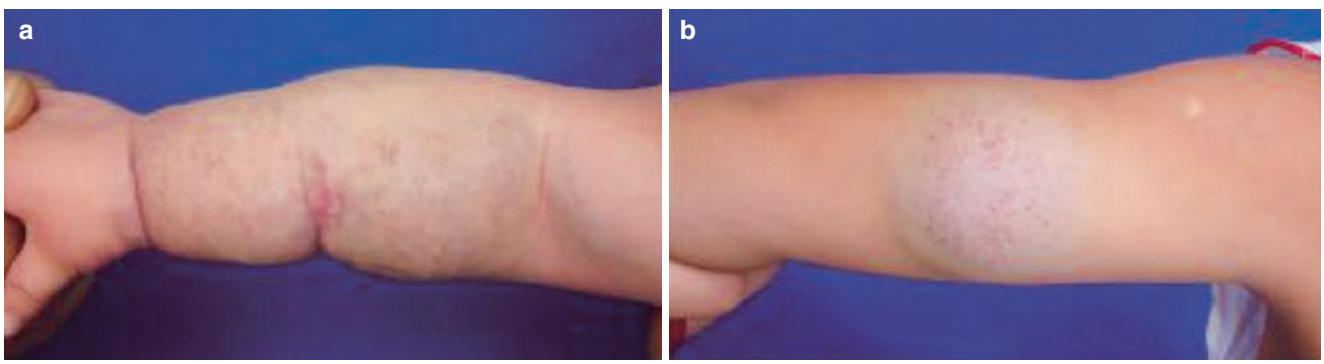


Fig. 12.9 Four kinds of hemangioma located at the limb. (**a**) The infantile hemangioma is located at the right upper limb, most of the foci are located subcutaneously, they are purple-blue as a whole, and they locally take on atypical bright red color. (**b**) Non-involuting congenital hemangioma is located at the left upper limb and presents blue-purple round mass, white halos are found in the surrounding margins, and bulging purple-red lesions with a cluster-shaped distribution are scatted in the center. (**c**) Kaposiform hemangioendothelioma is observed in the right upper limb with purple-red bulging lesions, less-defined borders, and a hard texture. (**d**) Plexiform angioma is observed in the right thigh and presents brick red plaques with still defined borders, a hard texture, and tenderness

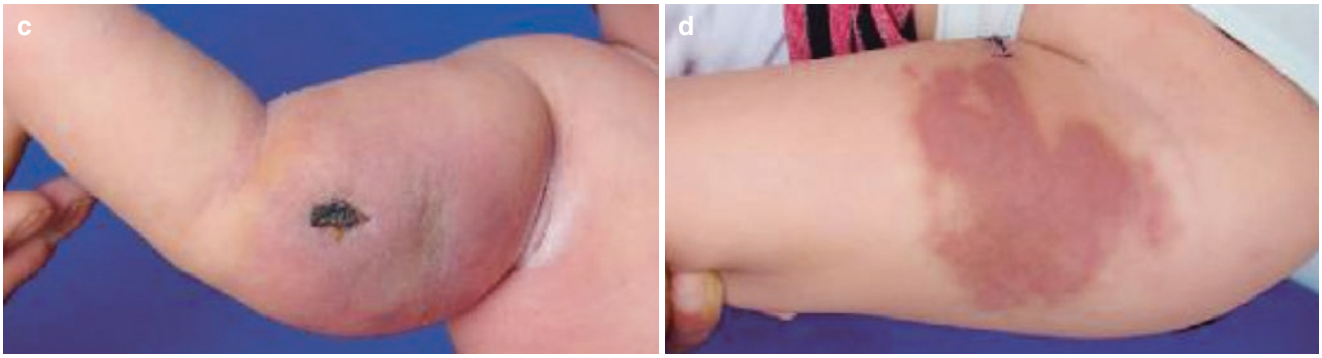


Fig. 12.9 (continued)

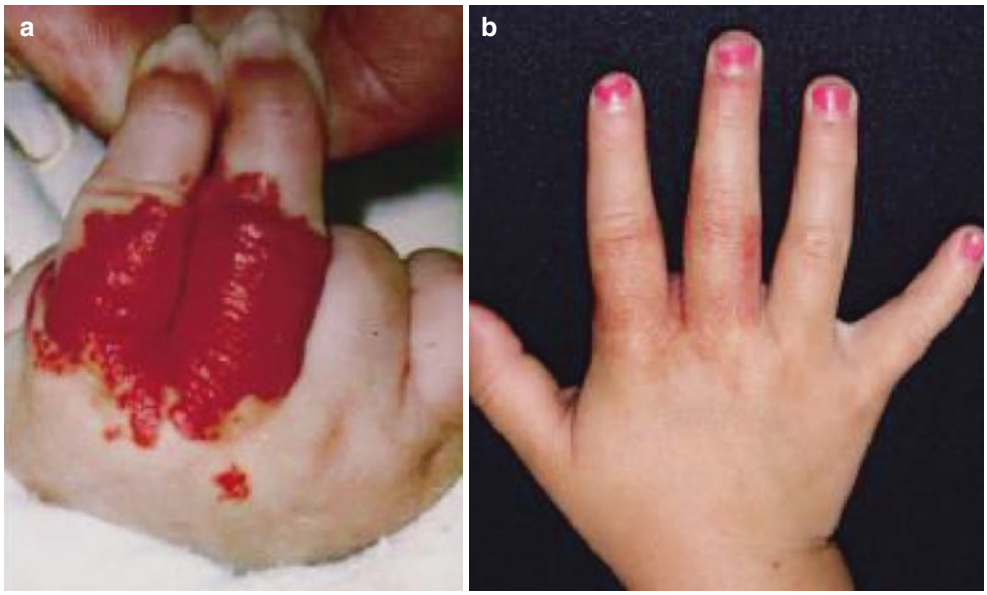


Fig. 12.10 Natural subsidence of hand hemangioma. (**a**) At the age of 3 months, the hemangioma is at the proliferative stage, the foci are fresh red, and the borders are defined. (**b**) At the age of 2, the foci almost subside, only slight skin pigmentation and telangiectasis remain, and hand functions are normal

12.2 Infantile Hemangioma

Hui ChenLei Chang, and Xiaoxi Lin

Infantile hemangioma is the commonest infantile true benign tumor with an incidence of 3‰ to 1%. The foci can be distributed all over the body, but it often occurs to the face and neck. The ratio of male to female incidence is 1:5 to1:3.

12.2.1 Clinical Manifestation

Infantile hemangioma has very a distinctive and unique natural disease course and can subside on its own, which is of vital significance to diagnosis and treatment. The focus often occurs at 2 weeks after birth and becomes enlarged rapidly. After proliferative phase for about 1 year, it starts to subside slowly for 3–5 years. The typical appearance is the presence of fresh red sharply marginated lumps which are protruding out of the skin; they are shaped like strawberries and do not become discolored when compressed. However, the early foci can be manifested as needle tip-like red spots, red patches, or plaques, and then they become enlarged, bulging or fused to form the above typical appearance. When the central part of the foci starts to turn white gradually and becomes fused and dilated, the lumps start to become soft, indicating that the lumps enter the subsidence stage. After subsidence, pigmentation, cicatrization, telangiectasis, and fiber and fatty deposition will be observed (Figs. 12.10 and 12.11). A minority of foci that have a large volume or grow rapidly can have spontaneous ulcer, especially in the places that are apt to form skinfolds such as joints. The changing of dressings can slow the healing, but obvious scar will remain (Figs. 12.12 and 12.13).

Infantile hemangioma can be seen in any site of hands or limb, and there is an obvious difference in size and appearance, ranging from the solitary soybean-sized bulge to the plaque penetrating the entire limb. It often involves skin or superficial fascia, rarely inducing functional disorder.

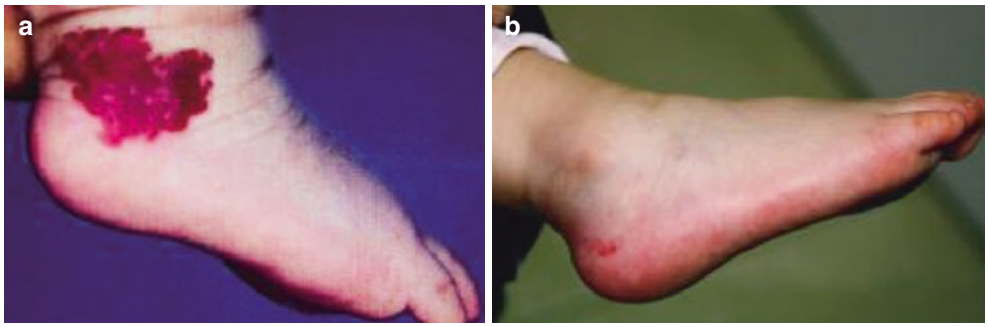


Fig. 12.11 Natural subsidence of foot hemangioma. (**a**) At the age of 4 months, the hemangioma is at the proliferative stage. (**b**) At the age of 3 years, the foci subside, only slight telangiectasis remains, and the skin color and texture return to normal

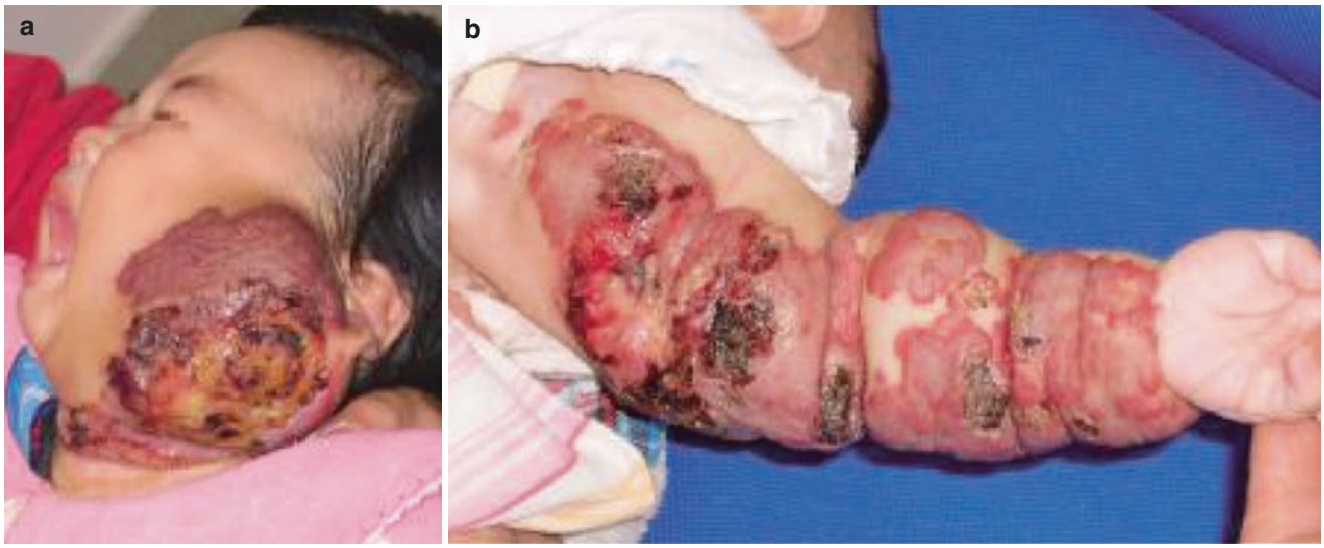


Fig. 12.12 Spontaneous ulcer of hemangioma; its occurrence may be correlated with the excessively rapid growth of tumor and local tissue hypoxia. (**a**) Giant hemangioma ulcer at the left face and neck. (**b**) Hemangioma ulcer in the left upper limb and obvious skinfolds

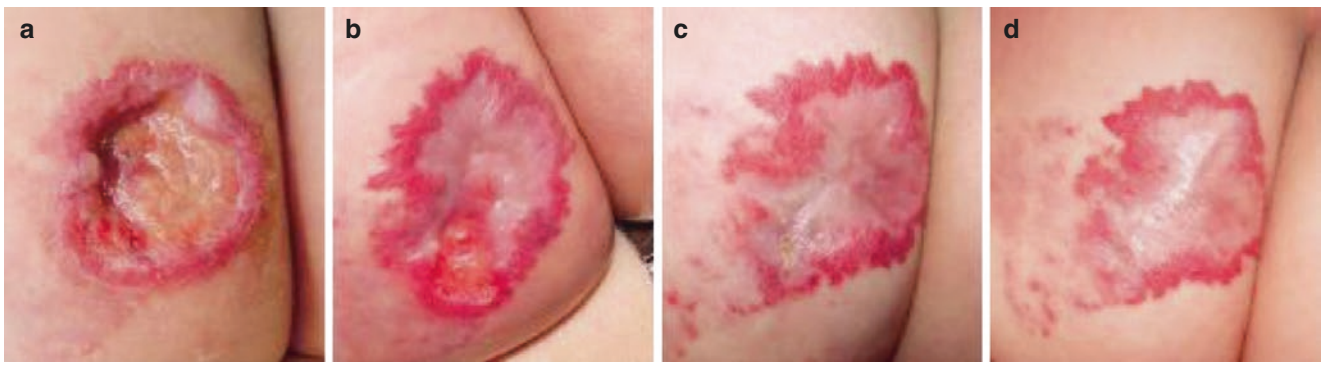


Fig. 12.13 Healing process of hemangioma ulcer in the left hip; lasting 4 months

Not all hemangiomas have typical strawberry-like appearance; for example, the deep hemangioma is only manifested as subcutaneous mass while the skin surface is completely normal; however, the strawberry-shaped red foci may not be hemangioma (Fig. 12.14). During the transformation from the proliferative stage to the subsidence stage, the pathological manifestations of hemangioma will change significantly; therefore, the history of natural diseases is also an important diagnosis basis.

12.2.2 Pathogenesis

The pathogenesis of infantile hemangioma has not been unclear currently, and there is no good explanation on the phenomena of proliferation and subsidence. There are many theories on the mechanism of proliferation, such as angioblast theory, placental origin theory, vasculogenesis imbalance theory, cytokine regulation pathway mutation theory,

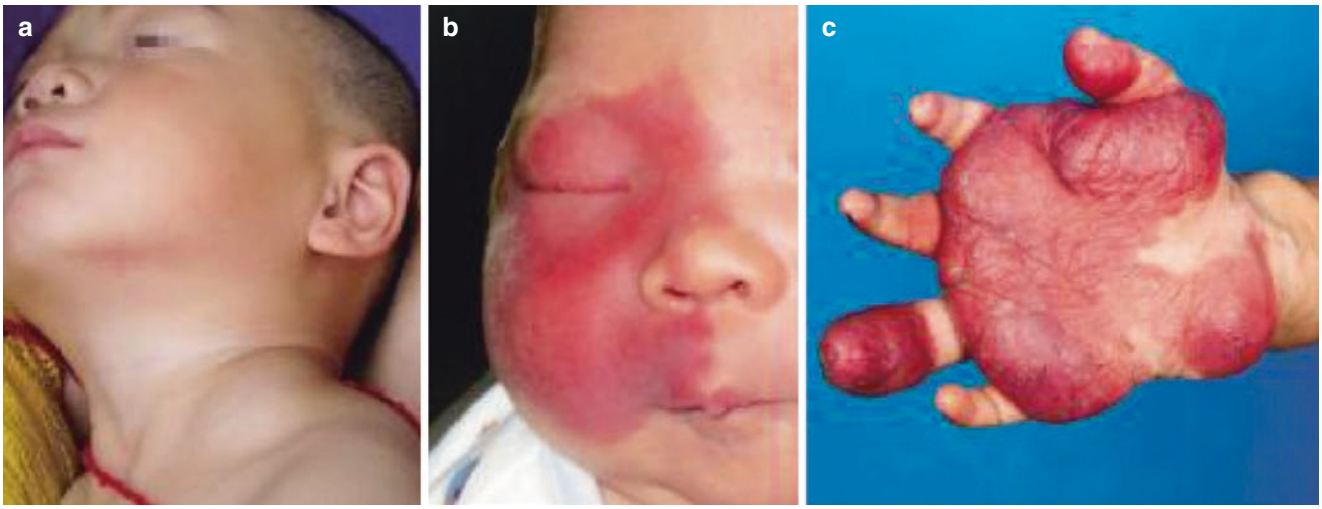


Fig. 12.14 Deep hemangioma and strawberry non-hemangioma lesions. (**a**) Deep hemangioma at the root of the left neck; it is the subcutaneous mass with a hard texture, its surface skin is normal, and CT and angiography can distinguish between them. (**b**) Port wine stains in the right facial part; it is fresh red, which is present immediately at birth; it proportionally grows with the body development, and there is no history of proliferation. (**c**) One arteriovenous malformation in the capillaries of the right hand, which is present at birth and rapidly grows, tortuous vessels can be locally seen, the skin temperature is high, and pulsation is palpable, with the characteristics of arteriovenous malformation

development area defect theory, theory of non-endothelial cells in hemangioma formation, and theory of immunity and inflammation; the subsidence is considered to be correlated with the factors such as changes in cell components and changes in expression of cytokine and apoptosis. With the constant accumulation of epidemiology of hemangioma characteristics, basic biology, and cytological information, significant progress has been made in the study of the mechanism of proliferation and subsidence, but there is no ideal animal model, and there is some impedance in the further implementation of studies. The current research focus is still on the aspects of angioblast, placenta implantation, angiogenesis imbalance, and development defects, and the mechanism of hemangioma subsidence is less studied, so the future studies should focus on this, which provides new ideas for early prevention of hemangioma and interventional treatment and finds new insights for the study of vascular development biology [11, 12].

12.2.3 Treatment

As hand hemangioma rarely induces functional disorder and can subside on its own; surgical resection should be avoided during the proliferative stage. The postoperative effect is often not as good as that of subsidence, and there will be obvious scar, resulting in the secondary sexual functional disorder.

A nonsurgical method is the first choice, the therapies such as drug and laser can promote the tumor subsidence, and obvious residual secondary deformities can be avoided. As for the local foci with a small area in the limb, if obvious fibrofatty will remain after subsidence, surgical resection can be performed to improve the appearance.

The currently often used drugs that can promote hemangioma subsidence include propranolol, corticosteroids, imiquimod, and antitumor drug (bleomycin). The application of laser or radioactive nuclide can be used as the adjunctive therapy, but its use should be cautious.

12.2.3.1 Drug Treatment

1. Propranolol: Currently as the first-line drug, it can be taken orally or externally. It was first reported by French Bordeaux Children's Hospital in 2008. When it uses propranolol to treat one pediatric patient with hypertrophic cardiomyopathy, they found that the accompanying nasal hemangioma would subside obviously. This is an extremely occasional but vitally significant discovery.

   As a type of nonselective beta blocker, it has been several decades since propranolol was first clinically used to treat hypertension, hypertrophic cardiomyopathy, and arrhythmia. Its safety has been recognized; the history of use of it to treat hemangioma is very short, and its mechanism of action is not very clear.

   (a) Indications of oral administration: ① the tumor grows rapidly, and within 1–2 weeks the area or the volume is enlarged and becomes over two times as large as that of the original tumor; ② multiple hemangioma; ③ the hemangioma is complicated by ulcer or is located at the site prone to ulcer, such as neck, axillary region

and perineal position; ④ parotid region and breast gland (female baby) hemangioma; ⑤ the hemangioma is complicated by functional disorder or located at the high-risk site, such as aural regions, lips, periocular regions, and nose; ⑥ hemangioma in airway or below the glottis (not complicated by abnormal respiration; laryngoscope or MRI indicates that the airway obstruction is less than 50%).

(b) Contraindications: ① the hemangioma that may endanger life, such as complication with respiratory distress and abnormal heart; ② internal organs, digestive tract, and intracranial hemangioma; ③ family history of bronchial asthma or asthma; ④ heart block (degree II to III atrial ventricular block); ⑤ severe or acute heart failure; ⑥ sinus bradycardia; ⑦ liver and renal functional disorder; ⑧ the guardian of the pediatric patient does not sign the Informed Consent Form (over-the-counter medications).

(c) Usage: in terms of medication methods, Siegfried and Lawley proposed an internationally recognizable gradient therapeutic regimen; for the infants that first used propranolol, the blood pressure, heart rate, and blood glucose were monitored within 48 h after admission; afterward the patients received further relevant and necessary examinations (e.g., ECG, heart color ultrasound, and consultation at the department of infantile cardiology) to rule out heart lesions, airway highly sensitive diseases, and other lung diseases; and the initial dose was 0.17 mg/kg·8 h and then gradually increased to the maximal dose of 0.67 mg/kg·8 h; for the patients incapable of being admitted for monitoring, the initial dose was also set as 0.17 mg/kg·8 h; 1 h after the initiation of administration, the vital signs and blood glucose level should be monitored, and in case of no obvious abnormality, the dose could be increased by one time every 3 days until the dose reaches 0.67 mg/kg·8 h. The majority of pediatric patients can tolerate this type of therapeutic regimen, and relatively good effects can be produced.

(d) Adverse reactions: the reported adverse reactions that occur during treatment of hemangioma using propranolol include decreased blood pressure, temporary respiratory distress, recurrence of foci, slowed heart rate, hyperkaliemia, hypoglycemia, nausea and vomiting, lethargy, increased aminopherase, irritability, icy limbs, pavor nocturnus, and rashes, and most of the symptoms can become relieved after symptomatic and supporting treatment or become relieved on their own. Currently, there is no report on pediatric death induced by administration of propranolol or serious adverse reactions.

The author's department carried out a prospective study on the application of the method of oral administration of propranolol at the outpatient service to treat 395 cases of infantile hemangioma from September 2008 to September 2012. Before the medication, all pediatric patients underwent routine blood, liver and renal function, blood glucose tests, myocardial enzymogram, ECG, and heart color ultrasound examination. The initial dose for treatment was 0.5 mg/kg. After the first administration, the doctors observed whether the patient showed the symptoms of cold acral coldness and wetness, dispiritedness, dyspnea, and obvious fidget. If the pediatric patients could tolerate them, the administration was continued 12 h later, and the dose still remained at 0.5 mg/kg. If the pediatric patients still showed no obvious abnormality, the dose was increased to 1.5 mg/kg the next day, the patient finished the oral administration in two times, and the patient was closely observed. If the pediatric patients still had no abnormal reactions, the dose was increased to 2 mg/kg on the third day, the patient finished the oral administration in two times, and this dose was maintained in the subsequent treatment. The method of gradually decreasing the dose was adopted to withdraw the drug, the dose on the last but 3 days was 2 mg/kg, the patient finished the oral administration in two times, and the interval was 12 h; the dose on the last but 2 days was 1.5 mg/kg, the patient finished the oral administration in two times, and the interval was 12 h; the dose on the last day was 1 mg/kg, the patient finished the oral administration in two times, and the interval was 12 h. The adverse reactions included decreased sleep (23 cases), increased sleep (5 cases), diarrhea (9 cases), decreased appetite (5cases), vomiting (5 cases), intermittent constipation (1 case), deepened respiration (3 cases), rash (1 case), and acral coldness (1 case). All of them improved per se within 1 month after administration. Another pediatric patient withdrew the drug 1 month after the initiation of the administration due to an elevation in aminopherase. Serious adverse reactions induced by propranolol have not been reported (Fig. 12.15) [13, 14].

Currently, the external preparation of β-blocker has been widely applied, such as timolol eye drops and propranolol gel, it is especially applicable to superficial hemangioma with a small area, and the application is safer and more feasible (Fig. 12.16). Combined medications can also produce satisfying effects (Fig. 12.17).

2. Corticosteroid hormone: It tends to be less-frequently used after the appearance of propranolol. The patient can orally take prednisone or receive local injection of triamcinolone acetonide. The oral administration is applicable to the focus with a large area or with a fast proliferation speed, and the local injection is mostly applicable to the local mass-like foci. The conventional regimen to orally take prednisone is 4 mg/kg each time, once a day, taken at

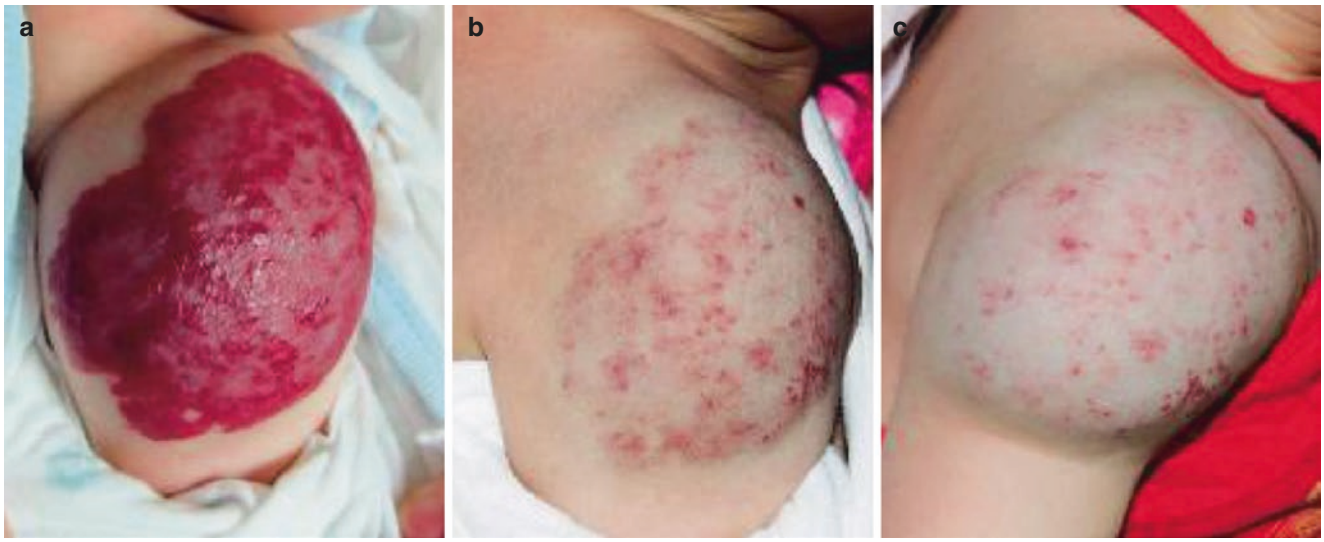


Fig. 12.15 Utilization of propranolol to treat infantile hemangioma. (**a**) An 18-month-old pediatric patient with giant mixed infantile hemangioma in the right shoulder. (**b**) Oral administration of propranolol for 45 weeks and during drug withdrawal. (**c**) At 2 months after drug withdrawal, slight recurrence in the deep foci

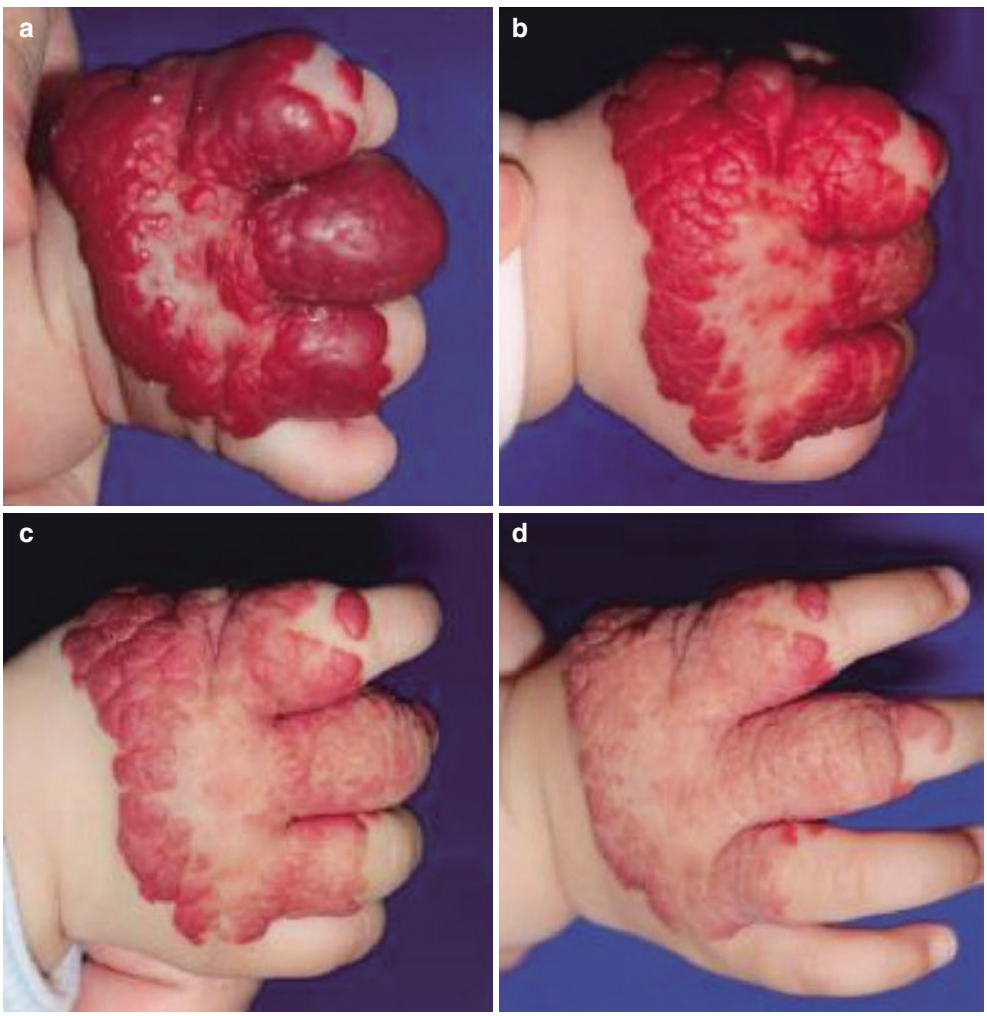


Fig. 12.16 Utilization of 0.5% timolol eye drops to treat infantile hemangioma. (**a**) The pediatric patient was 2 months old; the tumor in the right hand became proliferative rapidly and had red bulges. (**b**) After 4-month treatment, the tumor starts to become atrophic and subside. (**c**) After 8-month treatment, the tumor became obviously thinned. (**d**) After 10-month treatment, the majority of the foci had subsided; at 2 months after drug withdrawal, the tumor further subsided, and no recurrence was observed

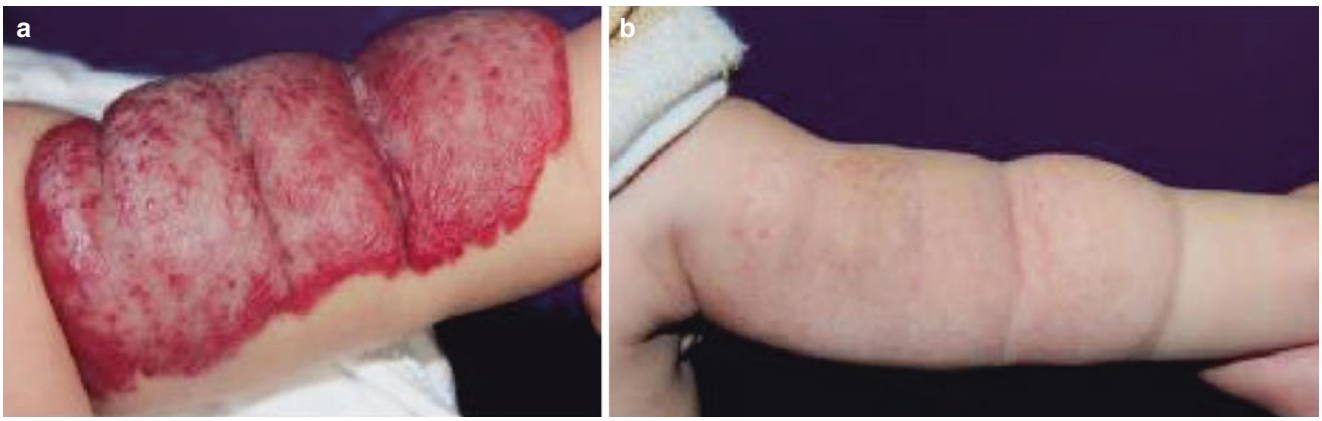


Fig. 12.17 The combined medications of oral propranolol and external timolol eye drops to treat the hemangioma at the left upper limb. (**a**) Before treatment. (**b**) The foci subsided after administration for 20 weeks

a draft in the morning, for a total of 8 weeks; afterward, the dose is reduced to half every week, and the administration continued for 1–2 weeks is a one treatment course. Usually the administration does not exceed two treatment courses, and the interval of two treatment courses is 2–3 weeks. However, not all proliferative hemangioma is sensitive to hormones, and the effectiveness rate is about 85%. As for the hemangioma without efficacy during the first treatment course which is insensitive to hormones, large-dose hormonal therapy should not be continued. As for the hemangioma that has entered the involuting phase, hormonal therapy should be not used; therefore, the process of angioplasty has ended already. Judging from the results of large-sample treatment, obvious or severe complications rarely occur to the patients who take the medicines according to the regulations; even in case of complications such as poor appetite, diarrhea, and obesity, they can be gradually recovered after drug withdrawal [15].

In injection treatment, injection of drug into normal tissues should be avoided, so the injection treatment is applicable to the foci with a certain thickness. Multiple treatments at small doses help to improve the safety. The adverse reactions of injection treatment may be more than those induced by oral medication.

3. Imiquimod: As a type of immunomodulator, it can directly induce α-interferon and tumor necrosis factors and indirectly induce the aggregation of γ-interferon at the foci and even induce cell apoptosis. It is applicable to the treatment of diseases such as condyloma acuminatum, squamous epithelial carcinoma, basal cell carcinoma, etc. It was reported to be used for treatment of infantile hemangioma as early as 2002. As for the treatment of infantile hemangioma, the prospective own control study has verified its efficacy [16]. It is mainly applicable to the flat foci during the early stage and also to those with a large area, and the focal effectiveness with obvious thickness is poor. Its common administration method is once a day, application to the foci before sleep, and cleaned with clear water in the morning. The main adverse reactions are local irritative responses, such as skin reddening and pruritus around the foci and scabs in foci to varying degrees, and even ulceration in severe cases, which should be avoided as far as possible; otherwise, superficial scars will remain.
4. Antitumor drug: The clinical application of antitumor drug for local injection is also very wide. The most-frequently reported drugs are pingyangmycin and bleomycin, which have obvious therapeutic effects on the proliferative hemangioma and can inhibit the proliferation of hemangioma at a relatively low concentration. It is the method commonly used in China to treat hemangioma. The common concentration of pingyangmycin is 8 mg diluted by 8–16 mL of normal saline, and the common concentration of bleomycin is 15 mg diluted by 10–20 mL of normal saline.

When the drug concentration is excessively high or the dosage is too much, local soft tissue necrosis can be induced. A large number of cases followed up for a long term indicate that local tissue development may be subject to the inhibition of drugs, deformities of defects or pitting are formed, so attention should be paid when the injection is at the oral lips, eyelids, or facial region.

12.2.3.2 Laser Therapy

The main principle is dependence on the selective photothermy of laser. The selective photothermy of laser means utilization of the characteristics of hemoglobin in the capillaries having high peaks of absorption near the wavelength of 580 nm and the surrounding tissues having little heat absorption as well as the principle of loss of heat during the pulse to produce high selectivity thermocoagulation effects

on the hemoglobin and eventually lead to vascular occlusion.

The first choice of treatment is laser of pulse dye (585 nm), and generally cicatrization and pigment alteration seldom occur. As the actual penetration power of light is relatively weak within the range of this wavelength, which is usually less than 1.5 mm, it cannot act on the full layer of foci of the majority of hemangiomas; so it is only applicable to some foci which are superficial, have a small area, grow slowly, or stop growing; and the prerequisite is no formation of any scar and no occurrence of permanent pigment alteration. Therefore, currently only proper cases and operation by experienced surgeons can satisfy this requirement.

Nonselective photothermal lasers such as Nd:YAG (1064 nm) and CO_2 (10,600 nm) are not suitable to be used for treatment of angioma as ulceration easily occurs and the risk of scar is high. As for the patients with residual obvious telangiectasis after subsidence, the laser of pulse dressings or Nd:YAG laser therapy is very suitable.

12.2.3.3 Applicator Therapy of Radionuclides

The vascular endothelial cells of proliferative hemangioma are at the immature proliferative status. It is highly sensitive to the radiation therapy, the angiogenesis ends after treatment, the capillaries have lesions and occlusion, the manifestations similar to subsidence appear, and the effects are reliable and objective. However, this method is only applicable to the superficial hemangioma with a small area which is located at the face, and particular attention should be paid to the control of radiation dose; otherwise, secondary changes such as hypopigmentation of the skin, cicatrization, and telangiectasis are apt to occur, which even lead to chronic radioepidermitis, the appearance is far worse than the appearance after spontaneous subsidence, and repair is difficult. Therefore, this treatment method is not suggested unless it is given by experienced nuclear medicine surgeons who have a good mastery of the indications.

12.2.3.4 Surgical Therapy

Surgical therapy is mainly applicable to the following two circumstances: ① the hemangioma is located at the special site, such as upper and lower eyelids, external nose, or oral lips, which may result in obvious functional disorders, such as amblyopia, strabismus, and disturbance in respiration or difficulty in food intake, and total or partial resection can be considered to improve the functions; ② the residual skin laxity, fiber fatty deposition, and tissue and organ displacements after the hemangioma subsidence can be repaired through surgeries.

12.2.3.5 Follow-Up Visits

As for the hemangiomas whose proliferation is not obvious or have entered the stable phase or involuting phase, too active treatment should not be given because what naturally remains are the basically normal skin structures, it is even hard to perceive after subsidence, and even the residual relaxed surface skin can be corrected through late phase reshaping. By contrast, if non-specific treatment measures that can cause large damages are adopted, there is no efficacy on the shrunk foci; further adverse results such as cicatrix or pigment alteration can be caused. Therefore, as for the involuting foci which are difficult to operate on or with poor postoperative appearance and relatively slow expected growth and even the proliferative hemangioma at a close-to-static status, follow-up visit is a relatively ideal choice.

12.3 Port Wine Stain

Hui ChenXiaoxi Lin, and Gang Ma

Port wine stain, commonly called nevus flammeus, falls into the category of congenital capillary deformities or venular malformations, and its incidence is as high as 0.3–2.1%. As surgical treatment is unnecessary for hand foci, such patients are very rare in the department of hand surgery.

12.3.1 Clinical Manifestation

The port wine stain has the manifestations of red or pink skin plaques at birth, which become discolored when compressed, and the borders are defined. The foci can be distributed all over the body, 75% of them occur to the face and neck, the size is various, and the largest one can involve nearly half of the body surface areas. With the increase in age, the color of the foci gradually becomes deep and takes on prunosus color or dull red. When the patient is at the age of 20–30, most of the foci at the face and neck can have obvious proliferation, and multiple nodules of varying sizes are formed on the basis of this; in patients with extreme symptoms, the nodules can be egg sized, or multiple nodules experience cluster-shaped proliferation and prolapse, forming the grape-bunch-like appearance and seriously affecting the patient's look (Fig. 12.18). However, only the foci at the hands can experience thickening and will not form nodules (Fig. 12.19) [17, 18].

12.3.2 Pathogenesis

The pathogenesis of port wine stain has not been cleared up to now. Currently, the research in this regard is only limited to histopathological observation. It is preliminarily found that the innervations of the diseases vessels are exceedingly reduced and even absent, and the blood flow gradually

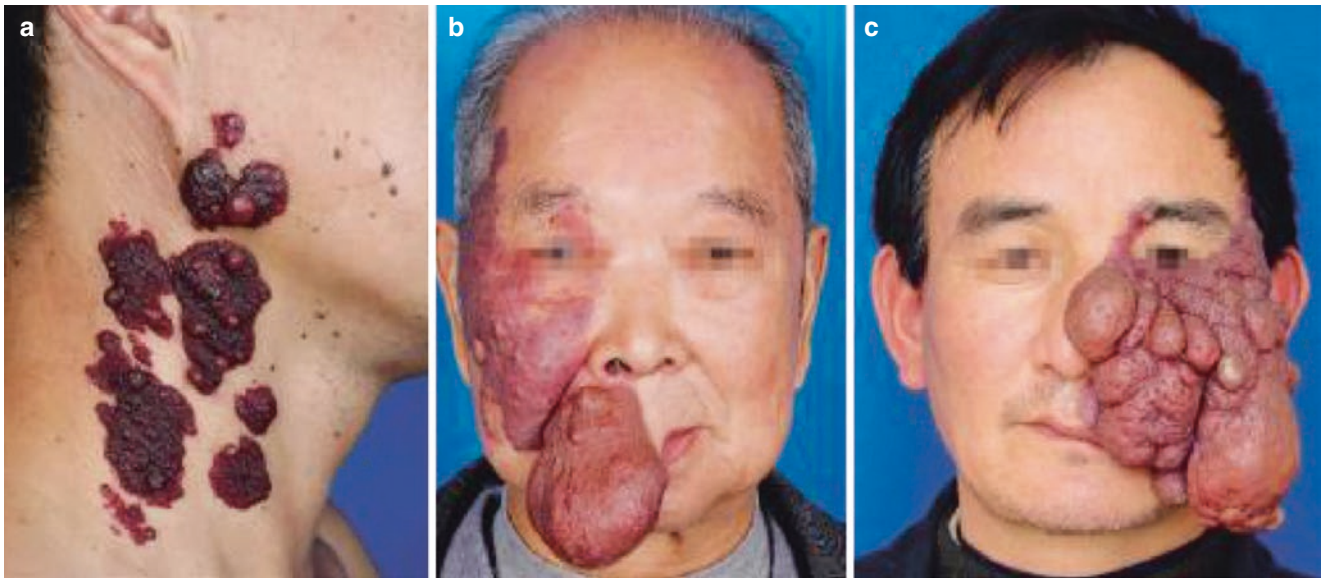


Fig. 12.18 Nodular-like appearance formed by port wine stain in face and neck

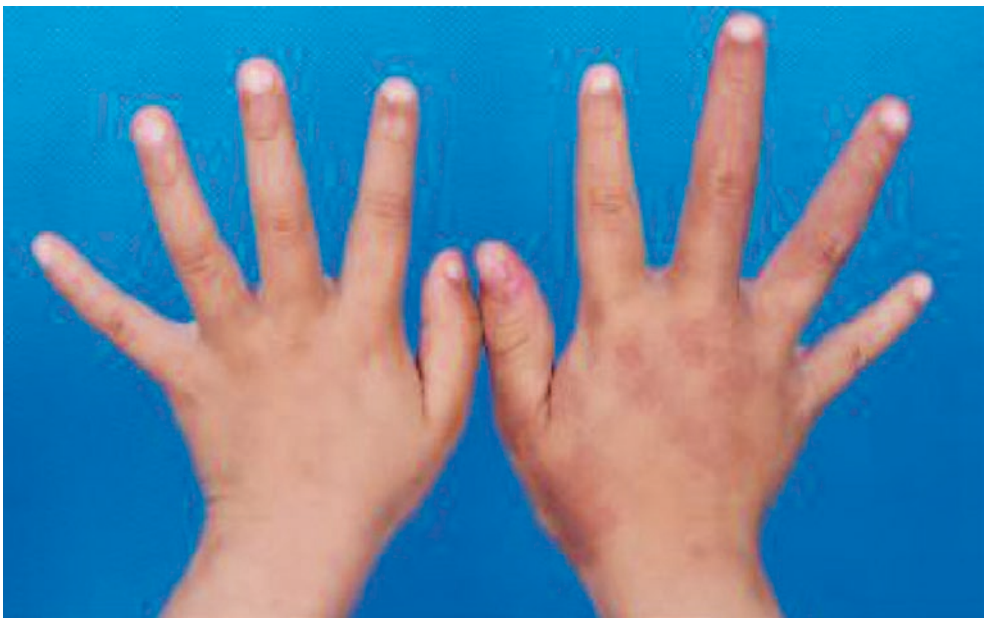


Fig. 12.19 Finger macroplasia induced by port wine stain in the right hand

impacts the vessels without nervous tension regulation to cause abnormal dilation of diseased vessels.

12.3.3 Pathological Characteristic

Abnormally dilated capillary angiomas or venulas appear inside the corium. Not only abnormally dilated vascular components are present in the thickened foci, but also a large number of epithelia, nerves and mesenchymal components that present falsification-like changes are distributed [18, 19].

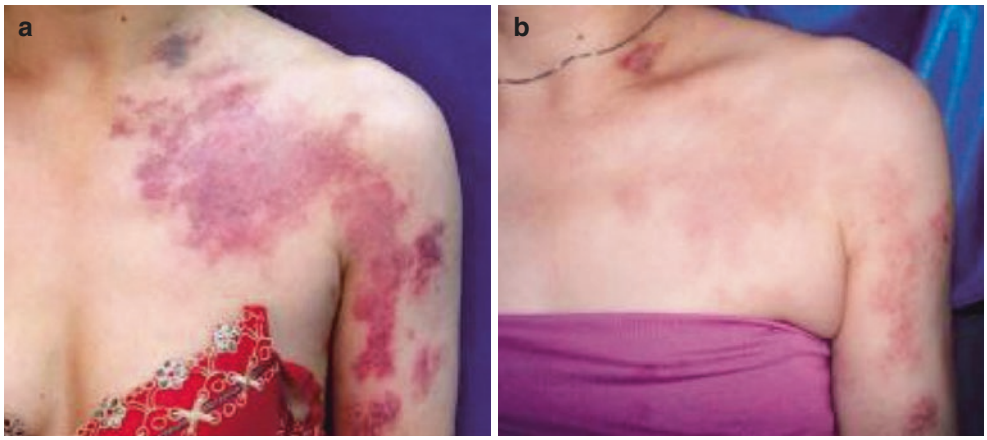


Fig. 12.20 Laser therapy of port wine stain. (**a**) Port wine stain of the left upper limb. (**b**) The majority subside after one pulse dye laser treatment

12.3.4 Treatment

There is no impeccable method for treatment of port wine stain. The previous treatments included freezing, artificial tattooing, drug injection, sclerotherapy, electric coagulation, dermabrasion, Chinese medicine application, nonselective photothermy laser, etc., but the above methods cannot specifically destroy the vessels or damage the normal skin structures, leading to cicatrization and poor appearance. Therefore, the laser therapy based on the selective photothermy principle is currently the mainstream [20, 21].

12.3.4.1 Laser Therapy

With the constant development of laser technology, the theory of selective photothermy revolutionarily appears, pulse dye laser emerges correspondingly, and it becomes the gold standard for the international treatment of port wine stain. However, although the wavelength and pulse width of laser are constantly modified and auxiliary cooling devices, multiple overlapping treatments, and the combination of various wavelength lasers are provided, only about 20% complete clearance rate is reached as for the overall efficacy. In addition, due to the relatively small laser light spots, it is difficult to homogeneously act on the skin, and the treated skin may take on an obvious variegation-shaped look. As the foci cannot be completely cleared, at the place of the deep residual vessels, new vessels and dilation can occur in the superficial regions through revascularization so that posttreatment recurrence of can be induced (Fig. 12.20).

12.3.4.2 Intense Pulsed Light

Different from laser system, intense pulsed light (IPL) can produce 515–1200 nm incoherent light. The light filter can be used to filter the unwanted wavelength light to treat deeper vessels. It can reduce the purpura after treatment; as for the patients with trunk foci with a large area or the patients tolerant against the pulse dye laser treatment, it can be used as the preferred treatment method, but there has been no systematic evaluation on its efficacy.

12.3.4.3 Photochemical Therapy

In the 1970s, photochemical therapy began to be used for the treatment of malignant tumor, and its mechanism was the destruction to the photosensitivity of tumor vessels. The principle is as follows: after the intravenous injection of photosensitizer, it can be accumulated selectively in the target tissues within a certain phase; at this time the laser radiation of a certain intensity is given; after the photosensitizer molecules are provoked, they react with the substrates to generate a series of photochemical reactions and produce some intermediate active compounds, including singlet molecular oxygen, eventually leading to the irreversible biological destruction of important intracellular structures. Port wine stain consists of capillaries with the deformity of intradermic dilation, the vascular capillaries absorb the photosensitizers the most rapidly, and its concentration within a certain phase is far higher than that of the epidermal tissues; under the optical excitation, it can highly selectively destroy the endothelial cells of dilated capillary network so that they will become deformed, necrotic, and thrombopoietic, leading to luminal atresia and subsidence of erythema; however, the covering epidermis is not injured. The currently used light sources mainly include copper steam laser (578 nm) and semiconductor laser (637 or 532 nm). Due to the large size of light spots with a diameter of 7–10 cm, it has obvious advantages in the treatment of port wine stain with a large area, the color subsidence is homogeneous and natural, the treatment frequency is relatively low, a certain therapeutic effect can be obtained in the foci with deep colors and mild thickening, and the incidence of cicatrix is less than 1%. In the author's study on 118 patients, 27.1% of the patients can almost achieve nearly complete subsidence, and the patients with complete ineffectiveness only occupy 1.7%. The photochemical therapy has a high dependence on the surgeon's experience. With the constant development of photosensitiz-

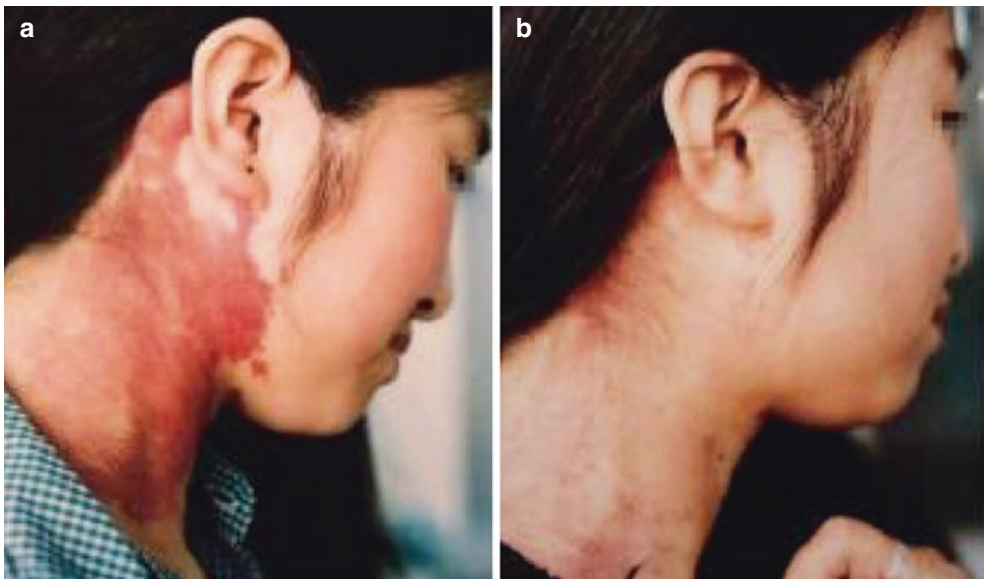


Fig. 12.21 Photochemical therapy of port wine stain. (**a**) Port wine stain in right neck and shoulder. (**b**) Subsidence of most of them after two photochemical therapies

ers and matched light sources, it will become the new development direction of the study and treatment of port wine stain (Fig. 12.21).

12.3.4.4 Surgical Therapy

The surgical treatment is mainly used in the following patients: ① the patients with secondary deformities of residual permanent scars, pigment alteration, and tissue atrophy after receiving improper treatment and ② untreated patients with foci which have obvious thickening and nodules who cannot receive nonsurgical treatment; ③ when the port wine stain involves the eyelids, nasal alae, or oral lips, proliferation and thickening in the above structures can be induced, affecting the vision, speaking, or food intake, and the surgical repair can obviously improve the above deformities. The methods of adopting skin grafts and free flaps often result in poor effects; therefore, when conditions permit, the first choice is the skin dilating techniques for repair, the successful dilation and repair can achieve the ideal matching between skin color and texture, and obvious cicatricial proliferation will not appear.

12.4 Venous Malformation

Hui Chen and Xiaoxi Lin

Venous malformation, previously called cavernous hemangioma, is low-flow cavernous vascular malformation. It has a low incidence, and there is no obvious difference between males and females. In most cases, it is sporadic, very few of them manifest familial aggregation, and it falls into the category of autosomal dominant inheritance.

12.4.1 Clinical Manifestation

Venous malformation is observed at birth and proportionally grows with the body; as for some patients, the foci are not obvious at birth and can appear only after the patient becomes an adult. The foci can be observed all over the body and is common in the head and neck and presents local or diffuse growth. It can involve skin and subcutaneous tissues and can penetrate muscles, joint capsules, and bones. Typical superficial foci take on blue-purple color and are soft lumps with an obvious compression sensation. The skin temperature is not high without tremor or pulsation. The size of the foci can change with body position; the filling is the maximal when the foci are in the lowest position of the body. In the foci with a large volume and a long disease course, smooth nodules with varying sizes, hard texture, and an easy tendency of movement are palpable, and they are the phleboliths formed after thrombus organization in the foci [22, 23].

The venous deformities of head and neck can lead to obvious deformities in appearance and induce the deformation and displacement of eyelids and oral lips, and giant foci can lead to enlargement or hypertrophy in face and bones; if the foci are located near the tongue and the pharyngeal or paratracheal regions, the patient will have difficulty in food intake and suffer airway obstruction.

In the patients with slight venous deformities in hands or limbs, only the local skin and subcutaneous soft tissues are involved; in severe cases, skin, cutaneous tissues, all hand internal muscles, or muscles of four limbs can be widely involved, but the involvement in skeletons is rare. The manifestations of focus appearance are various, and the typical patients can have the blue-purple soft masses; they can also be local nodular bulges with varying sizes, and the texture is hard; the patients can also have deep masses, the surface skin

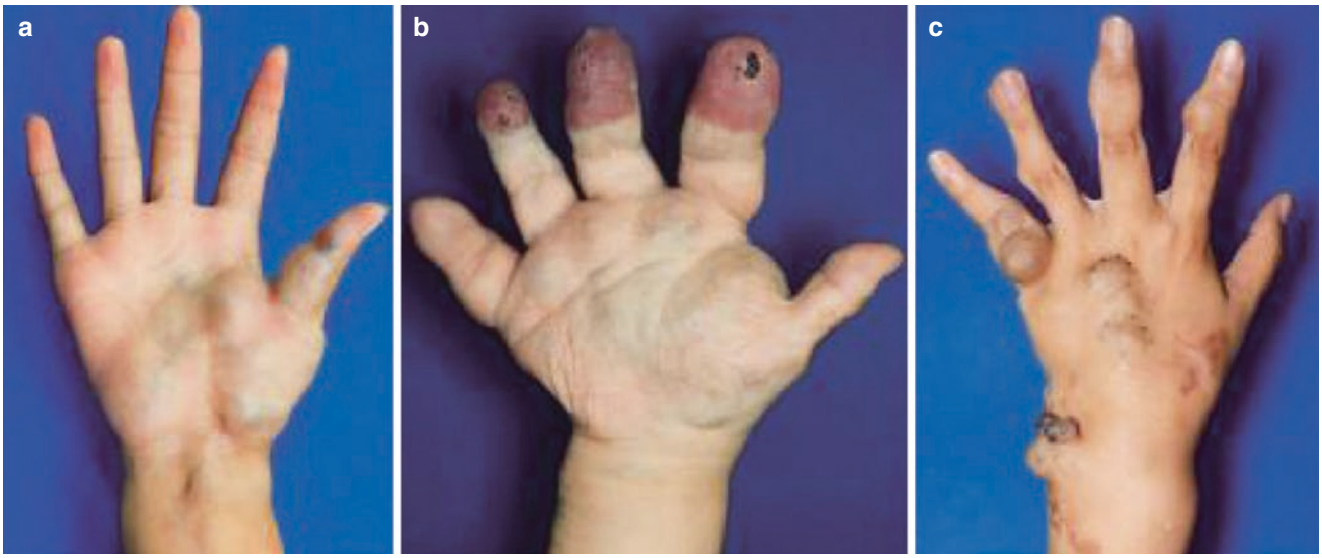


Fig. 12.22 Different appearances of hand venous malformations. (**a**) The typical hand venous malformations present the blue-purple masses with a soft texture. (**b**) The fingertip is red, with capillary deformities. (**c**) Nodular venous malformation, obvious bulge, and hard texture

is completely normal, and the patient can be accompanied by capillary deformities, etc. (Fig. 12.22).

The hand or limb venous malformations can have influences to varying degrees on the functions, so attention should be paid. The focal bleeding in muscles, periost, or joint cavity can induce pain to varying extents; frequent pain can make the joint take protective posture for long time, which is apt to induce joint stiffness or rigidity and results in functional disorder. The severity of functional disorder has no direct correlation with the widespread of the foci. As for the single muscles around the joint, such as local foci at the distal end of the four quadriceps muscle of thigh, the proximal end of gastrocnemius muscle, and the proximal end of the brachioradialis muscle, in case of long-term bleeding, severe functional disorder will occur early. If the foci only involve the flexor group or the extensor group, they can result in dysplasia of muscle group and decreased muscular strength, and this can lead to skeletal dysplasia and joint dislocation; if all limb muscles are involved, the functional disorder will be insignificant due to the balance in development or progressions in patient's conditions. It should be noted that the iatrogenic functional disorder induced by improper treatment is very common.

12.4.2 Pathogenesis

Venous malformation falls into the category of congenital vascular dysplasia, the molecular mechanisms of the onset of sporadic venous malformation still remain mysterious, but some advancements have been achieved in the recent studies on familial venous malformation. From 1994 to 1995, in two families with venous malformations in skin mucosa characterized by autosomal dominant inheritance, genetic analysis

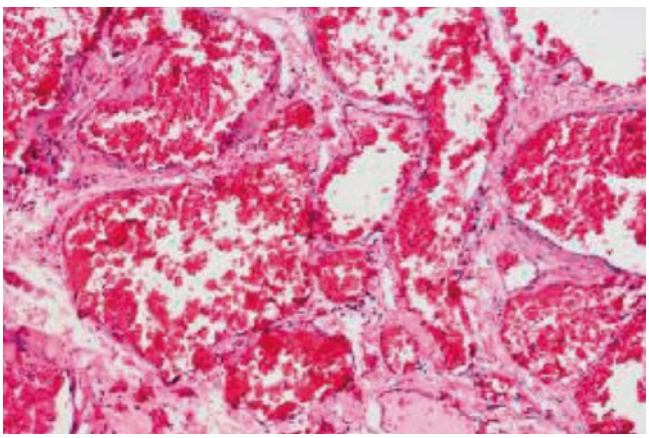


Fig. 12.23 The pathological sections of the venous malformations can show the presence of a large number of red blood cells in the blood sinus

indicated that the gene mutation was located at chromosome 9p21. Tie2 signal pathway plays an important role in the formation of activity bed; the above two venous malformation families have the following manifestation: the arginine in the Tie2 receptor kinase region that encodes the endothelial cells is replaced by tryptophan, inducing the hyperphosphorylation and the increased activity of the receptors and further leading to abnormal vascular development.

In 1999, there was a foreign report on four venous malformation families with autosomal dominant inheritance, one of them had the same mutation with the above two families, and the gene mutation site of other families was irrelevant to chromosome9p21, indicating that hereditary venous malformation has a genetic heterogeneity. Other studies also contain the detection of the ligand of Tie2 receptor, angiogenin 1, and angiogenin 2, and these proteins are considered to play an important role in the formation of vascu-

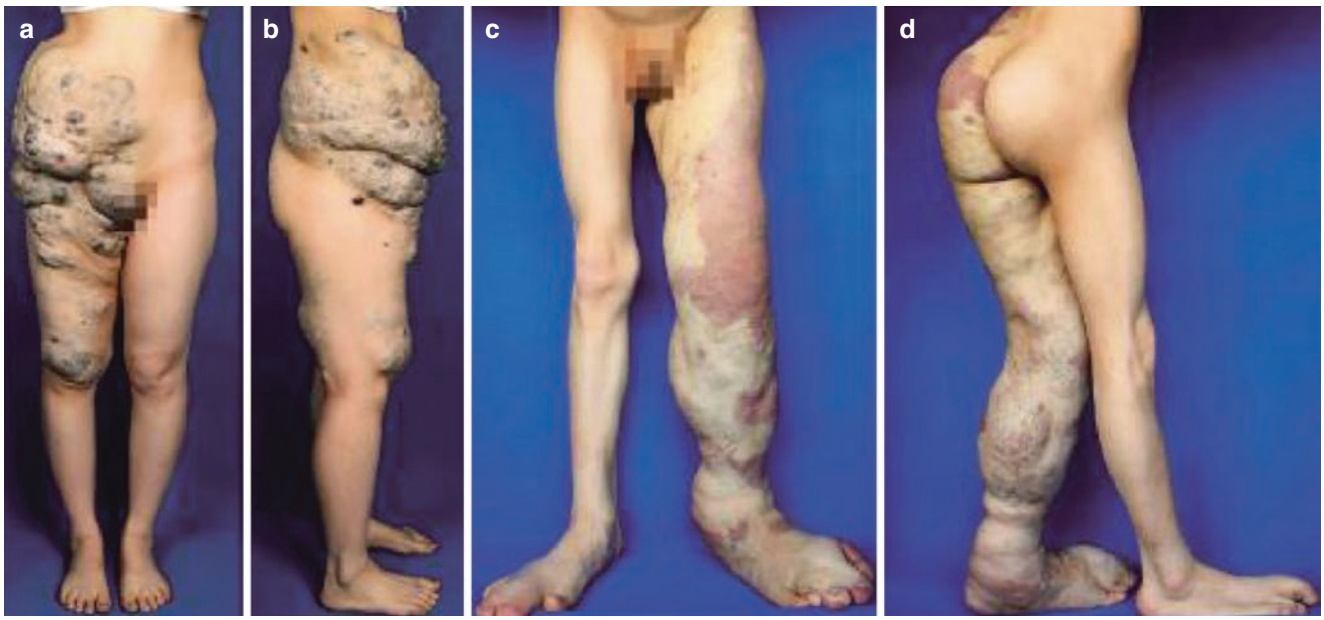


Fig. 12.24 Difference between common limb venous malformation and KTS. (**a**, **b**) Giant venous malformation in the right lumbar region and lower limb, no appearance of limb macroplasia, and equal length of the two lower limbs. (**c**, **d**) KTS in the left lower limb, with the typical triad with venous malformation, port wine stain, and limb macroplasia

lar non-endothelial components (including smooth muscles).

12.4.3 Pathological Characteristic

The inner linings of the foci consist of a large number of irregular lumens, no characteristics of proliferation are observed in the endothelial cells, the layers of smooth muscles of the lumens are thin or absent, and a large number of red blood cells are present in the blood sinus (Fig. 12.23).

12.4.4 Syndromes Correlated with Limb Venous Malformation

Limb venous deformity is one of the manifestations of multiple vascular lesion syndromes, such as Klippel-Trenaunay syndrome (KTS), Maffucci syndrome (MS), and blue rubber-bleb nevus syndrome (BRBNS).

12.4.4.1 Klippel-Trenaunay Syndrome

This syndrome is a congenital sporadic vascular deformity, but there is also a report on the familial onset. The typical triad is manifested as limb port wine stain, venous malformation or varicose, and macroplasia of bones and soft tissues. In the skin foci, the lymphatic vessel components are relatively common, they are often mixed with capillary deformities, map-like dull red plaques are formed around the knee joint, the surface has follicular nodules with varying sizes, and they become thickened with ulcer and bleeding.

As for the pediatric patients with erythema in a large area, they are suspected to suffer from KTS. During the growth and development, the affected limb will become thickened and longer gradually. Some pediatric patients feel pain and are unwilling to walk or move due to the occurrence of pain, leading to joint stiffness or Achilles tendon contracture. MRI is a necessary examination to determine the range and depth of the deep foci. Currently, there has been no effective treatment method for KTS, continuous pressurization with elastic garment with can delay the progression in the patient's conditions, and local sclerotherapy can facilitate the mitigation of pain. As for the patients with obvious joint functional disorder, they can be given surgical treatment accompanied by active functional exercise (Figs. 12.24 and 12.25).

12.4.4.2 Maffucci Syndrome

The foci often appear during the infancy with the main manifestations of subcutaneous venous malformation, chondrodysplasia of four limbs, and multiple enchondroma. The diagnosis is mainly dependent on the clinical manifestations, the venous malformations are visible all over the body, but they are most commonly seen at the distal ends of four limbs with the manifestations of blue-purple soft masses with occasional presence of tenderness. However, pathologically, venous malformations and hemangioendothelioma of spindle cells are jointly present. Enchondroma often appears asymmetrically at both sides, X-ray indicates that the bone cortex is bulging and becomes thin, it shows orbicular-ovate

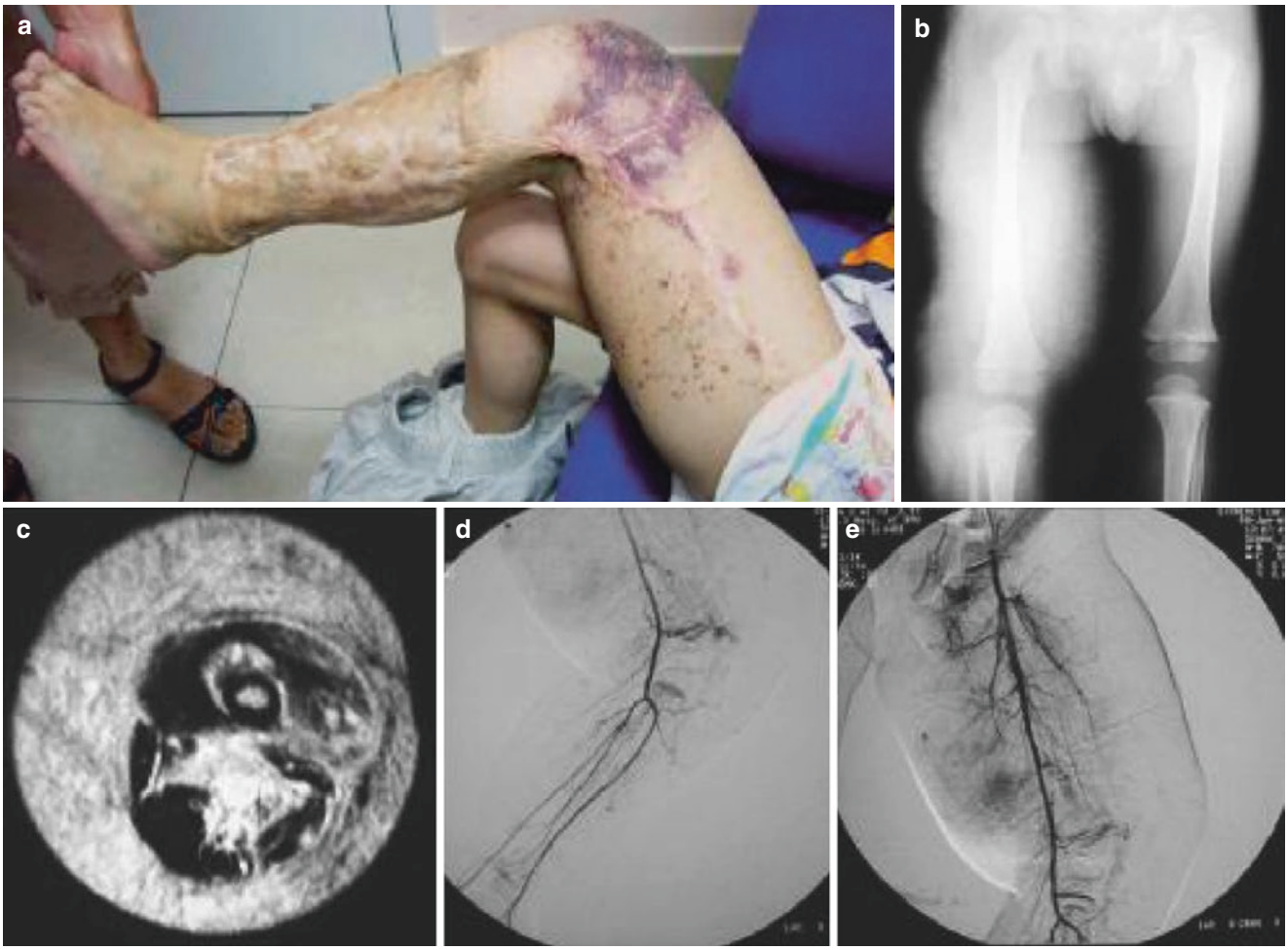


Fig. 12.25 KTS in the left lower limb. (**a**) The patient received multiple surgical resections at other hospitals, and the functions and appearance were not obviously improved. (**b**) X-ray film indicates that the skeletons in the left lower limb become obviously thickened and long. (**c**) MRI indicates the involvement of full layers of superficial fascia and some muscles. (**d**, **e**) No obvious arteriovenous fistulae were observed in the digital subtraction angiography after arterial catheter, and the foci of venous malformation cannot be completely displayed

defects, and grit-shaped small calcification spots are visible in the venous malformations.

The most serious complications of MS include multiple fracture during the childhood, nerve system abnormality induced by cranial enchondroma, and enchondroma canceration (about 15% of the canceration is enchondroma). If the venous malformation is asymptomatic, generally nonsurgical treatment is adopted. If the cartilage lesion is applicable to surgical treatment, biopsy must be performed to rule out the canceration.

12.4.4.3 Blue Rubber-Bleb Nevus Syndrome

BRBNS has the main manifestations of multiple venous malformations in the skin and digestive tract, most of them are sporadic, and some may fall into the category of autosomal dominant inheritance. Foci often occur at birth or during the early stage of childhood. Skin venous malformations are commonest in the trunk and upper limbs, they can exist in single form or the number can be several hundred, the size ranges from several millimeters to several centimeters, and the color can be prunosus, blue, and even black; the venous malformations of the digestive tract can be easily detected by endoscope or MRI, the size varies, most of them are multiple and located in the lower layer of the mucosa, the region from the small intestines to the end of the colon can be involved, and it has no connection with the distribution and range of venous malformations on the body surface. Venous malformations can be seen in other organs, such as the liver, lungs, urethra, brain, and muscles (Fig. 12.26).

The main treatment is to control the chronic bleeding of the digestive tract. In case of serious bleeding, endoscopic electric coagulation or surgical resection can be adopted. If the skin foci affect the appearance, sclerotherapy or surgical treatment can be adopted.

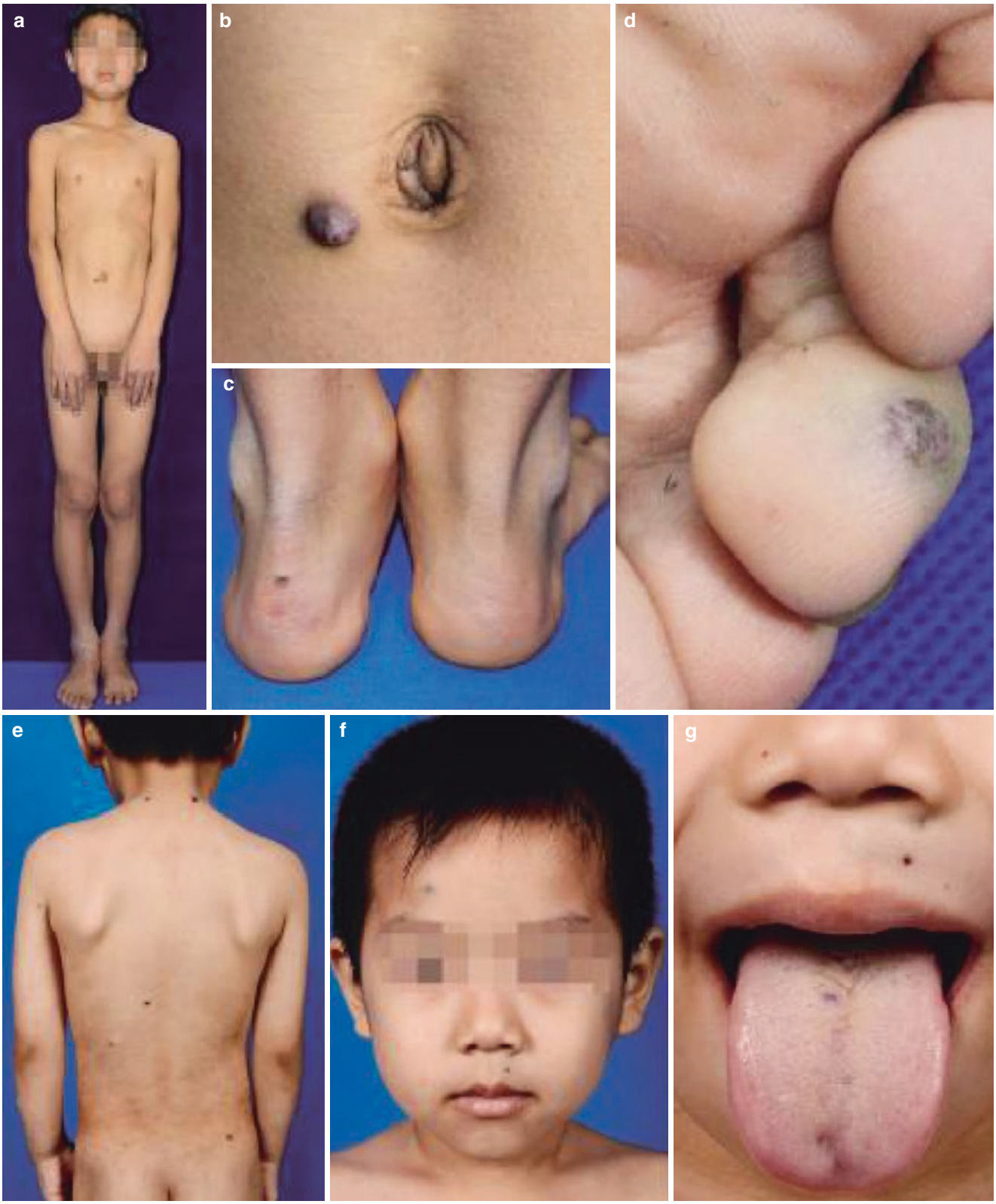


Fig. 12.26 Two pediatric patients with blue rubber-bleb nevus syndrome; blue-purple nodules are visible all over the body

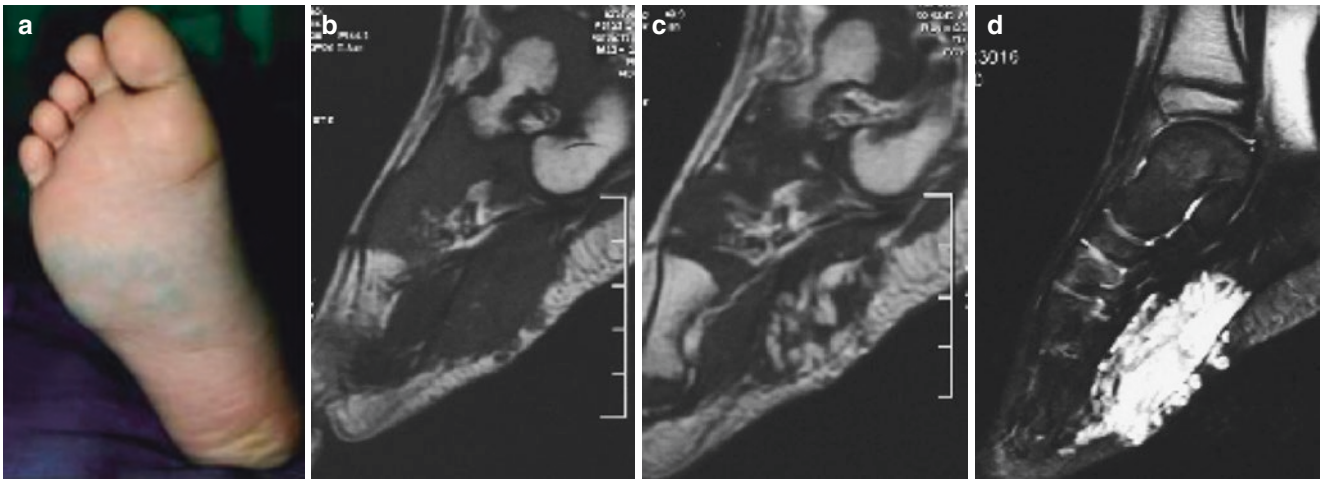


Fig. 12.27 MRI characteristics of venous malformation at the sole of the foot. (**a**) Venous malformation at the sole of the foot. (**b**) T1-weighted images; the foci present equal signals, and the borders are defined. (**c**) T1-weighted images become enhanced, and heterogeneous intensification is visible in foci. (**d**) T2-weighted images; the foci present homogeneous high signals

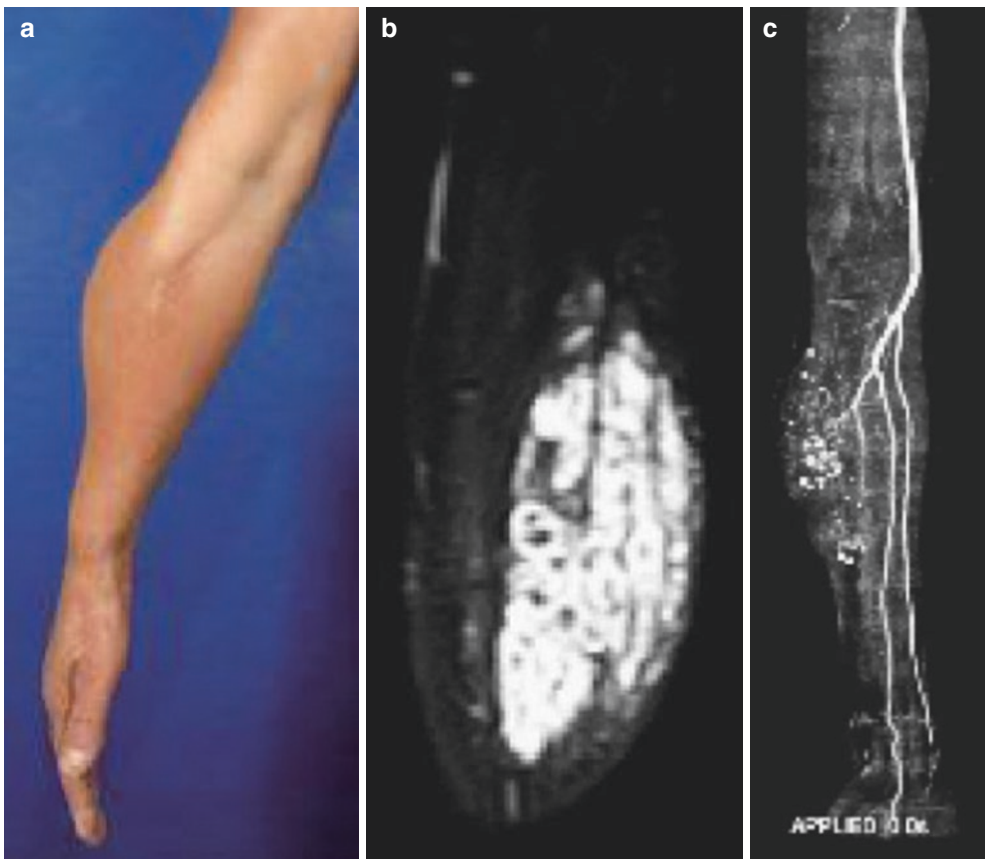


Fig. 12.28 Imaging characteristics of intra-focal phlebolith of venous malformations. (**a**) Intramuscular venous malformations of the right upper limb. (**b**) MRI indicates that, in the high-signal foci, the phlebolith presents dense circular low signal shadows. (**c**) CT can show the presence of multiple nodule shadows of a high density inside the foci

12.4.5 Diagnosis

The diagnosis of most patients can be confirmed only by relying on medical history and clinical manifestations. MRI is the most important imaging examination that determines the range and depth of foci and their relation with the adjacent histological structures, venous malformations present moderate or low signals in the T1-weighted images and present high signals in the T2-weighted images, and intensification to varying degrees can be seen after enhancement

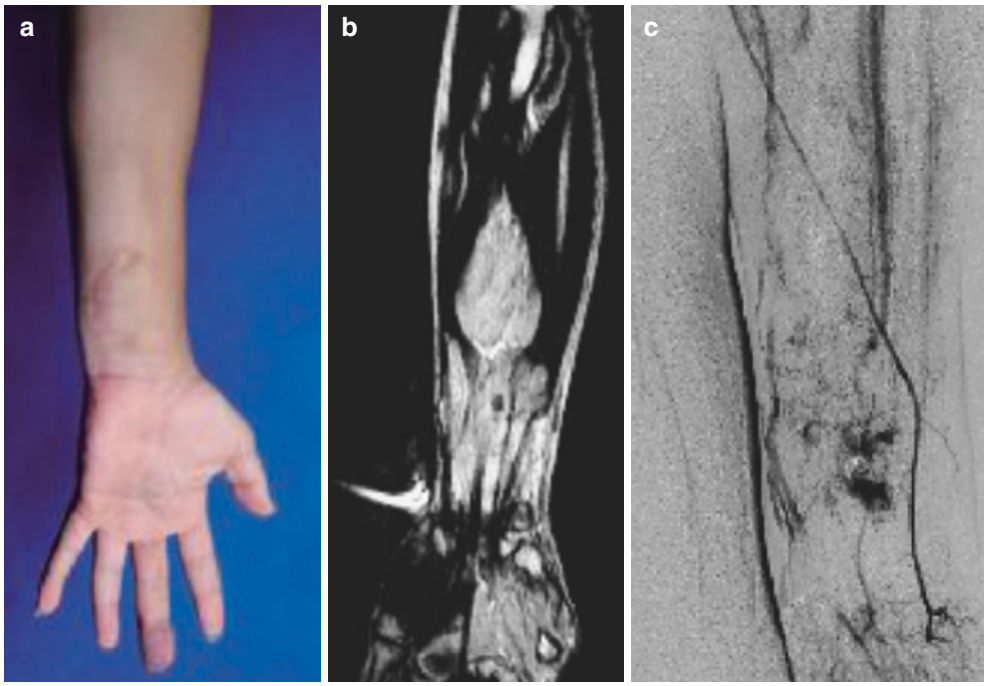


Fig. 12.29 The per-arterial digital subtraction angiography cannot fully display the foci of venous malformations. (**a**) Venous malformations in the left upper limb. (**b**) MRI indicates that the foci are located at the forearm flexor group, the borders are defined, and the display of foci is complete. (**c**) Digital subtraction angiography can only show the concentration of patchy less-defined contrast medium and cannot display the whole picture of the foci

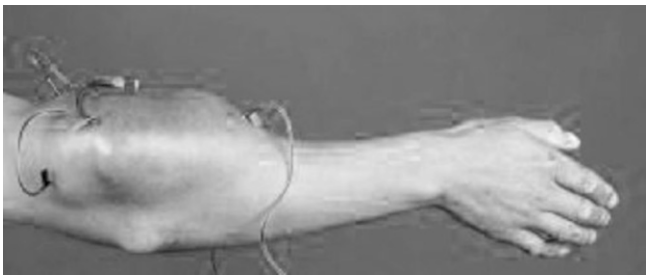


Fig. 12.30 Multi-spot puncture of venous malformations can indicate the effusion of dull red venous blood, which can serve as the important diagnosis evidence

(Fig. 12.27). Phlebolith and bone invasion can be determined through X-ray plain film and CT (Fig. 12.28). The percutaneous tumor cavity puncturing angiography can properly indicate the foci and their hemodynamic characteristics. It is very difficult for per-arterial digital subtraction angiography to display the venous malformations, and it is often not used as the conventional examination (Fig. 12.29). Percutaneous focal puncture can indicate the return of dull red blood, which facilitates the diagnosis (Fig. 12.30).

12.4.6 Treatment

Except very few local foci, other hand or limb venous malformations cannot be eradicated completely. Therefore, while appearance repair should be taken into account, preservation or improvement of functions should be the first consideration in treatment. In addition, functional lesions

Table 12.3 Common hardeners and their characteristics

Hardener	Characteristic
Absolute ethanol	It can destroy the vascular endothelial cells and vascular walls, inducing a large amount of thrombosis, and the efficacy is exact. The pain is acute in injection. The incidence of serious adverse reactions is high, and the allergic reactions rarely appear
Pingyangmycin	It is the purified bleomycin A5. It can inhibit the proliferation of endothelial cells. The action is mild, and the pain is slight in injection. It has additive effects, and the use at a high dose during a short term is avoided. Allergic reactions occasionally occur
Bleomycin	It can inhibit the proliferation of endothelial cells. The action is mild, and the pain is slight in injection. It has additive effects. The allergic reactions rarely appear
Sodium morrhuate	It is a biological extract and the components are unstable. It can injure the intravascular endothelial cells and promote thrombosis. The pain is acute in injection. In case of overflow from the vessels, it is apt to induce wide tissue necrosis. Allergic reactions occasionally occur
Polidocanol	It can destroy the lipid bimolecular layers of the cell membrane, injure the cells, and promote the thrombosis. The pain is mild in injection. The efficacy when it is prepared into foams is better, but the adverse reactions correlated with gas embolism may occur
Sodium tetradecyl sulfate	It decomposes the inter-endothelial matrix so that the cells become detached and thrombosis is promoted
Ethanolamine oleate	It can injure vascular endothelial cells. Its viscosity is large and injection is difficult

and exacerbation induced by improper selection of treatment methods as well as destruction of appearance should be avoided. Currently, the mainstream treatment of venous malformation all over the globe is intravascular treatment (sclerotherapy), and a strict mastery of indications is needed for surgical resection [23–26].

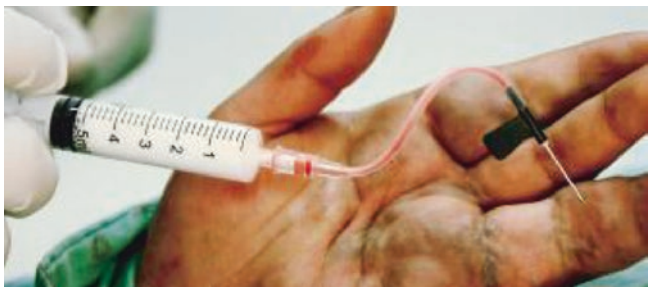


Fig. 12.31 As for the intravascular treatment of hand venous malformations, the used drugs are the polidocanol prepared into foams

12.4.6.1 Intravascular Treatment

Intravascular treatment refers to the treatment that utilizes such drugs as absolute alcohol, pingyangmycin, bleomycin, sodium morrhuate, or foam sclerosing agents (e.g., polidocanol and sodium tetradecyl sulfate) (Table 12.3) to destroy the vascular endothelial cells and cause the blockage of focal vessels and the atrophy of volume, so as to improve the appearance and functions and ensure low probability of recurrence. However, the wide and diffuse foci may need multiple therapies, and the efficacy is relatively poor (Figs. 12.31, 12.32, 12.33, 12.34, and 12.35).

The main adverse reactions of intravascular treatment included pain, swelling, cutaneous pigmentation, subcutane-

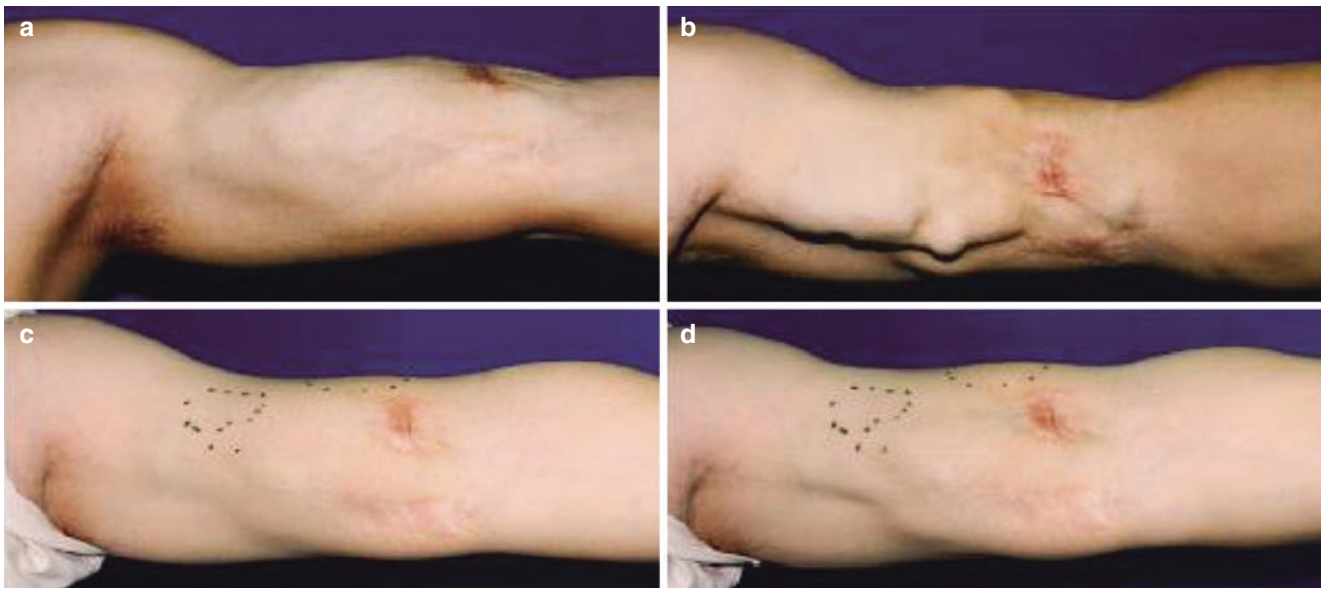


Fig. 12.32 Sclerotherapy of venous malformations at the left upper arm using absolute alcohol. (**a**) Subcutaneous and intra-bicipital venous malformations of the left upper arm; they present blue-purple bulging masses, and residual scars are observed after incomplete resection at other hospital. (**b**) When the bicep muscle of arm is contracted, the foci are squeezed and protruding nodules are formed. (**c**) After sclerotherapy with absolute alcohol for three times, the volume of foci shrinks obviously, and the functions of the upper limb are not affected. (**d**) After treatment, in case of the contraction of bicep muscle of arm, the protruding nodules become obviously relieved

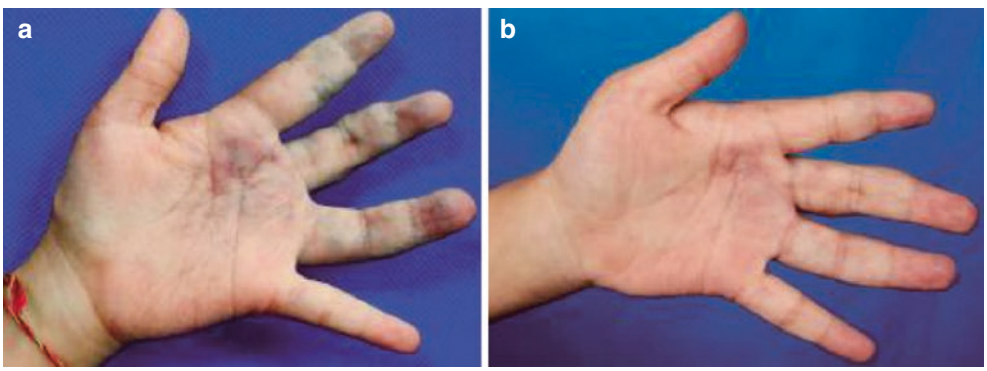


Fig. 12.33 Sclerotherapy of venous malformations at the left hand arm using absolute alcohol. (**a**) The left hand has wide foci; thenar muscle, center of the palm, and index and middle fingers are involved. (**b**) After sclerotherapy treatment for three times, the foci subside obviously and the hand functions are normal

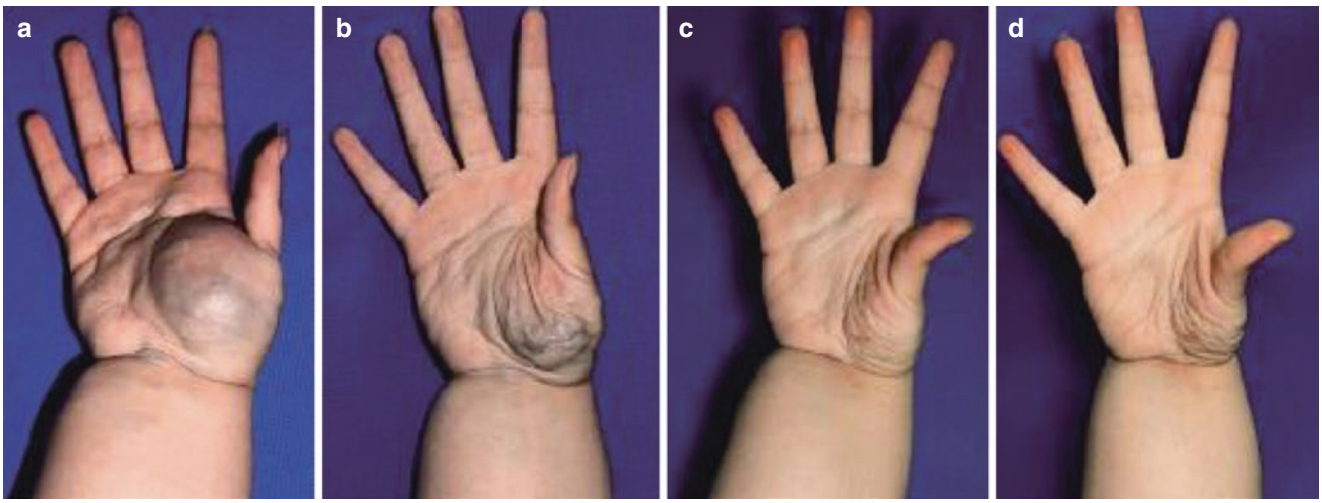


Fig. 12.34 Sclerotherapy of venous malformations in the thenar muscle of the right hand using absolute alcohol. (**a**) The right hand droops, and the foci become obviously engorged and bulging. (**b**) The right hand is superducted, the blood refluxes, and the foci collapse. (**c**) After sclerotherapy with absolute alcohol for four times, the right hand droops, the foci at the thenar muscles shrink obviously without more filling, and the foci in the center of the palm still have filling without treatment. (**d**) After treatment, the right hand is superducted; due to the disappearance of most foci, the appearance has no significant difference from that in case of drooping

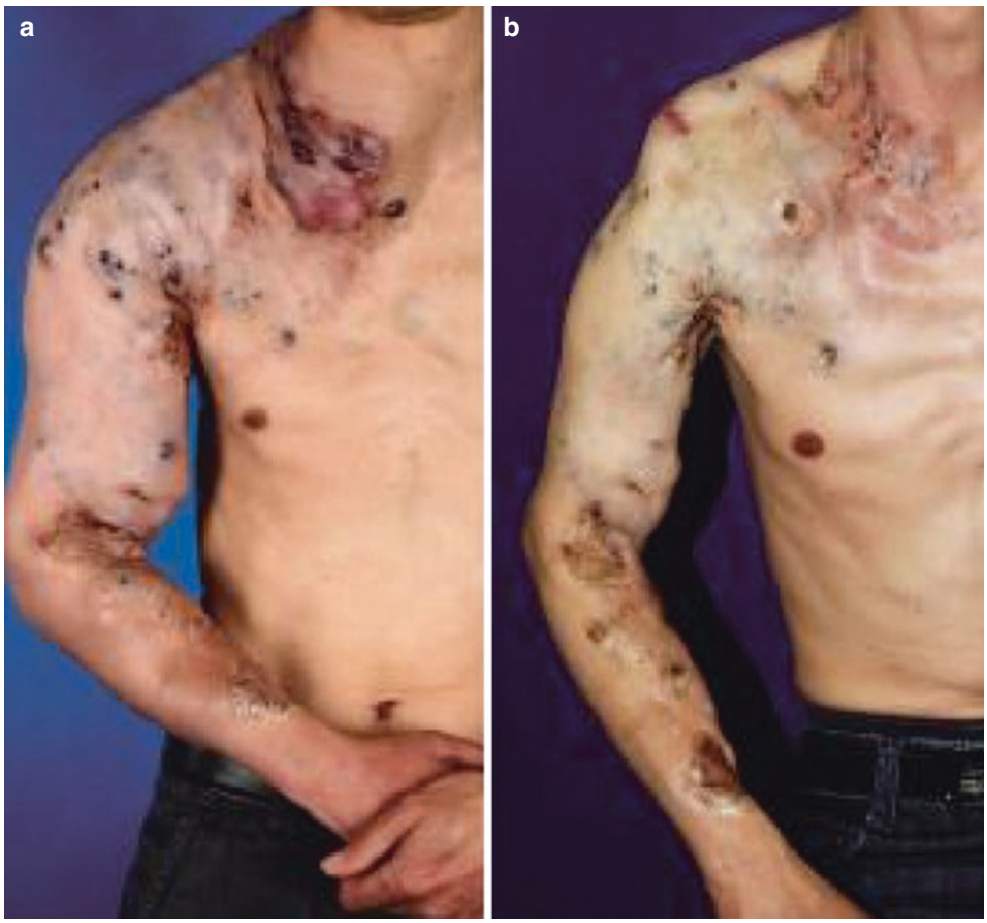


Fig. 12.35 Giant venous malformation in the right upper limb. (**a**) Congenital venous malformations in the right upper limb; multiple local resections at other hospital; the diagnosis was intra-focal bleeding of venous malformation, obvious swelling in appearance and ulceration, and bleeding of local skin. (**b**) After sclerotherapy for ten times, the foci had obvious atrophy, and no ulceration or bleeding was observed in skin

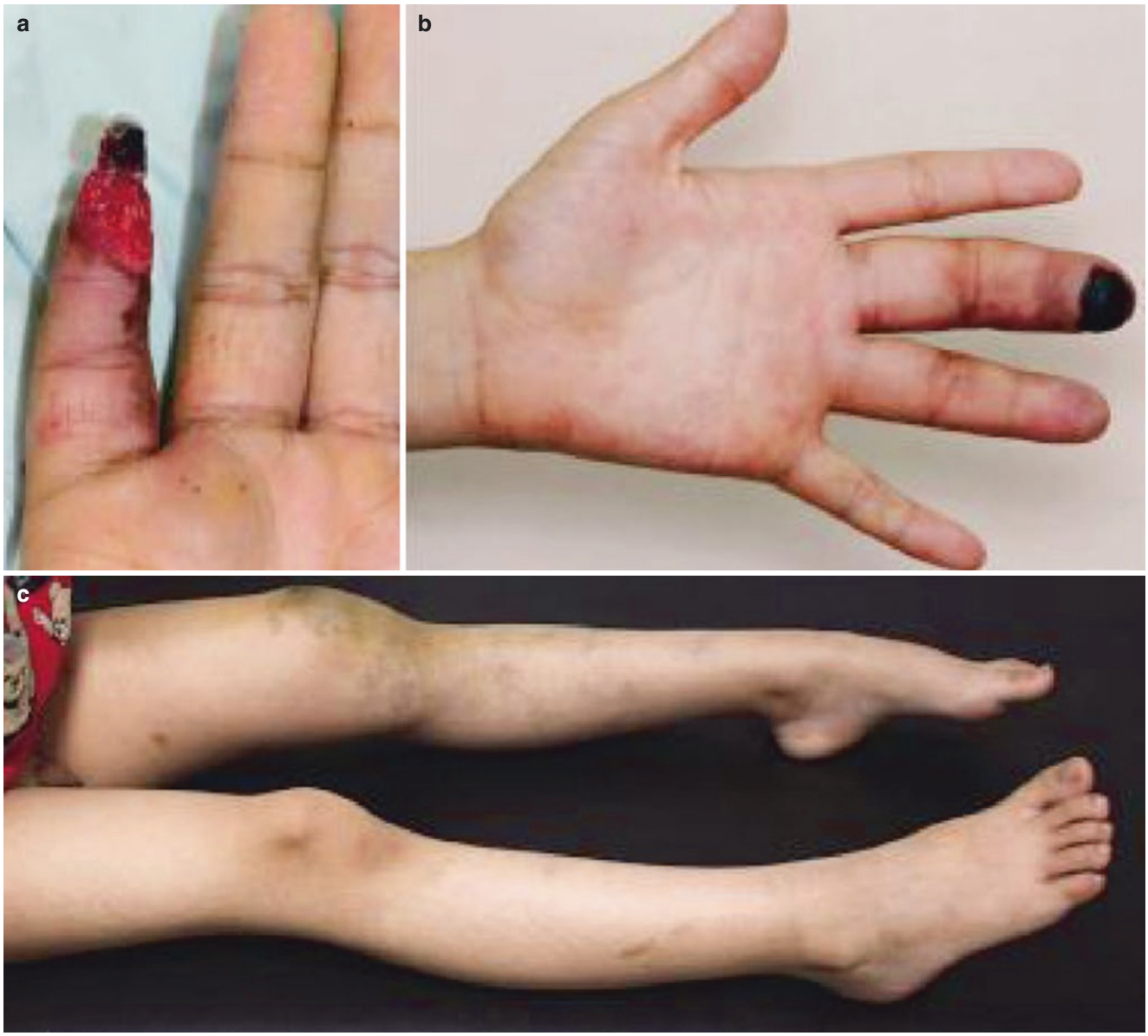


Fig. 12.36 Adverse reactions of sclerotherapy of limb venous malformations. (**a**, **b**) Local tissue necrosis of the fingertips. (**c**) Common peroneal nerve injuries of the left lower limb, with the presence of drop foot deformity (recovered after 6 months)

ous induration formation, local tissue necrosis, nerve injuries, anaphylactic shock, pulmonary arterial spasm, pulmonary embolism, and cerebral embolism. The serious complication that is the most apt to occur after the treatment of hand or limb venous malformations is nerve vascular injury, further leading to tissue necrosis or functional disorder. Therefore, the patients should be very familiar with this anatomical structure and should perform a comprehensive evaluation of focal distribution, and the treatment should be given with caution (Fig. 12.36).

12.4.6.2 Other Treatments

Other treatments include surgery, electrochemistry, laser, and compression of affected limb. Surgery is not the ideal therapeutic method of hand or limb venous malformations; due to functional consideration and less-defined focal borders, complete resection is usually difficult, and the residual foci will subsequently experience dilation and lead to recurrence. In addition, the intraoperative bleeding volume is large; after operation, a large amount of difficult-to-control continuous capillary hemorrhage may occur, continuous compression of the tissues will lead to tissue necrosis, and the patient will end up with amputa-

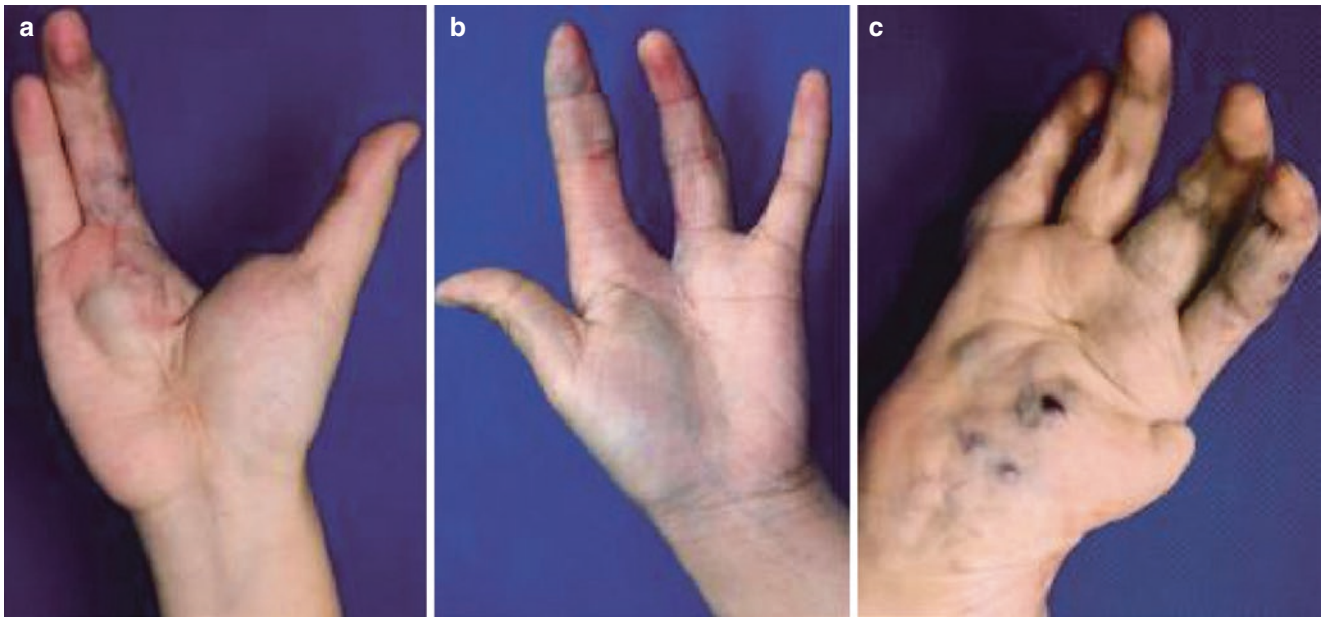


Fig. 12.37 The finger venous malformations are amputated, resulting in permanent functional absence

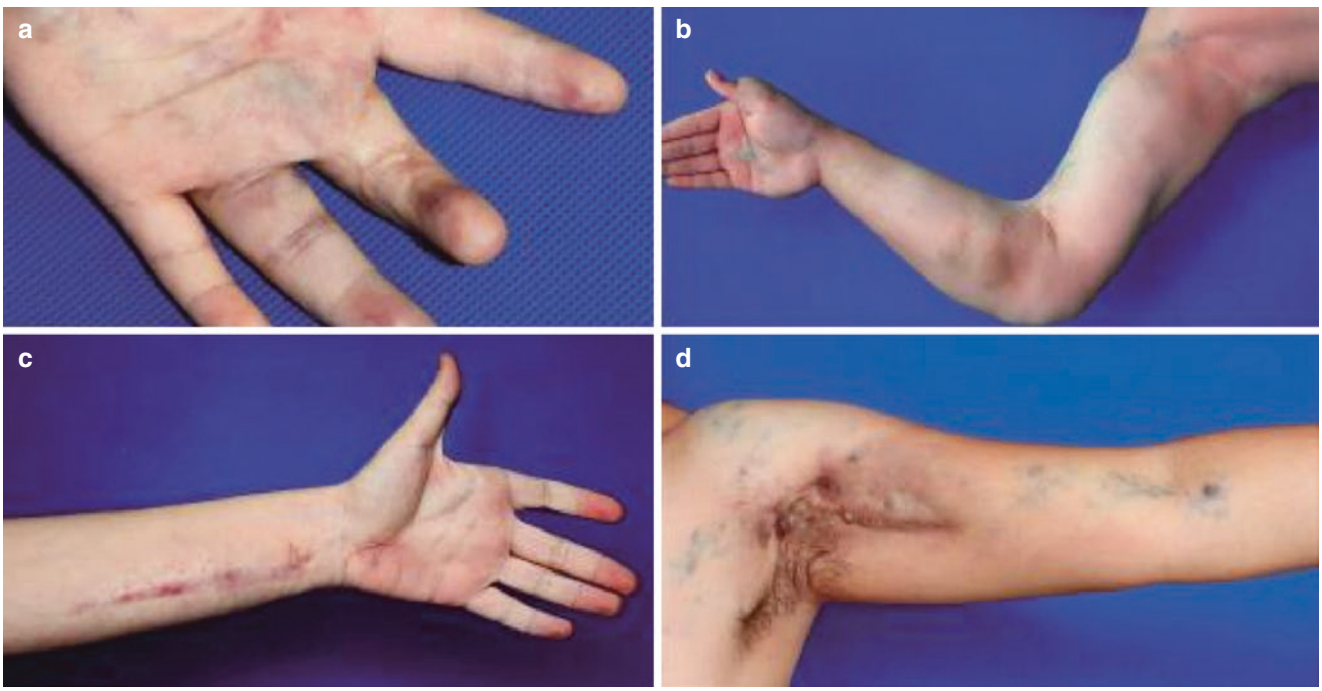


Fig. 12.38 Surgical treatment of upper limb venous malformation is apt to result in secondary joint functional disorder and obvious cicatricial hyperplasia, and the foci cannot be resected completely

tion (Fig. 12.37). The postoperative scars are often obvious, and the scars near the joints can easily lead to secondary functional disorder (Fig. 12.38). If the local bulging foci at hands and feet have a significant influence on appearance or walking or shoe wearing, surgical resection can be performed on the basis of comprehensive evaluation (Fig. 12.39).

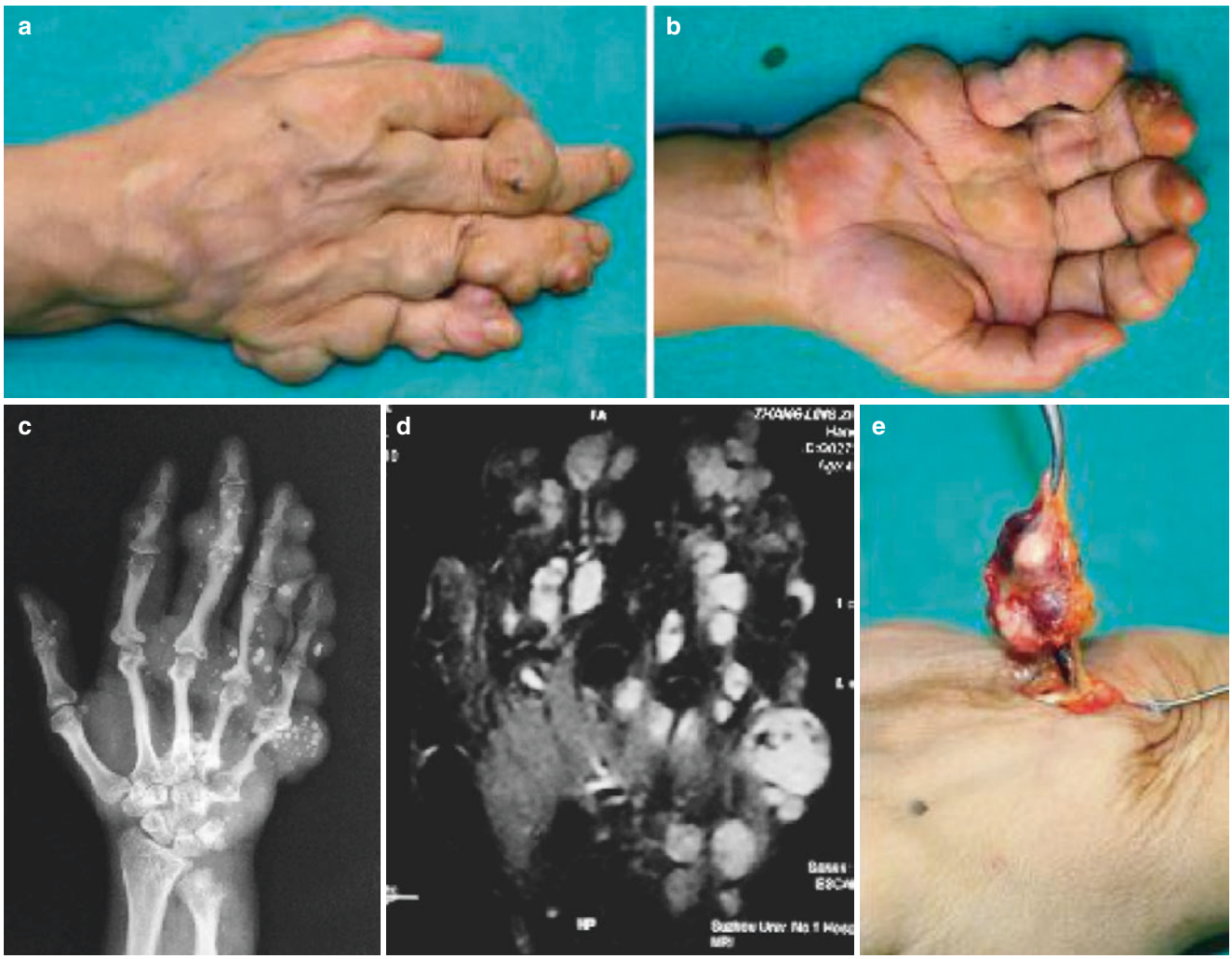


Fig. 12.39 Partial surgical resection of multiple venous malformations in the right hand. (**a**, **b**) The foci take multiple nodular shape of varying sizes, the borders are defined, and the texture is hard. (**c**) X-ray films can show the dense phlebolith shadows inside the foci. (**d**) MRI shows the presence of multiple circular-low-signal shadows in the foci, and they are phleboliths. (**e**) After the foci are resected, blue-purple blood sinuses and multiple white phleboliths can be observed

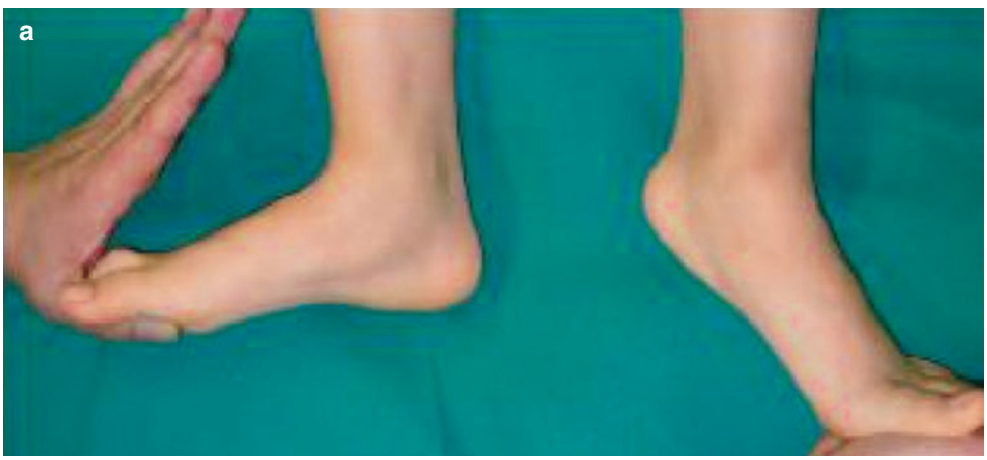
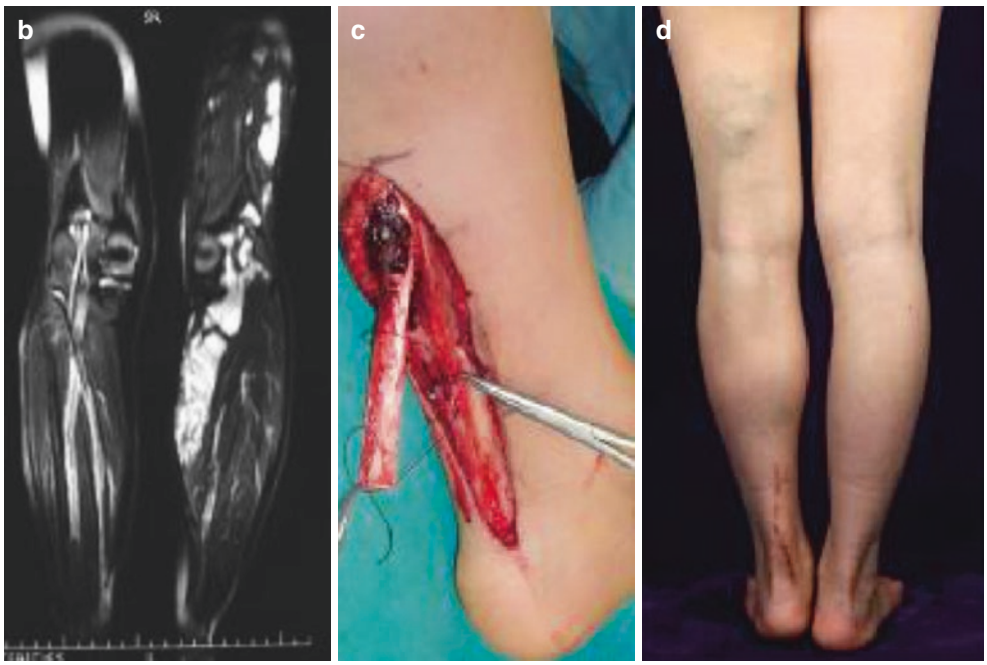


Fig. 12.40 Surgical treatment of contracture of Achilles tendon induced by venous malformations in the left lower limb. (**a**) Preoperative contracture of left Achilles tendon. (**b**) MRI can indicate the foci are located at the posterior muscle groups of thigh and shank. (**c**) Blue-purple foci are observed during the operation. (**d**) At 3 months after operation, the contracture shows obvious improvement

Fig. 12.40 (continued)



Electrochemotherapeutics refer to the method of insertion of positive electrodes into the foci and then utilization of electrochemical reactions to induce the degeneration and necrosis of focal tissues to eliminate the foci. As its treatment efficacy is not high, normal tissues are often damaged and pinprick scars often remain; this treatment method is gradually eliminated.

Laser therapy can be adopted for the superficial venous malformations of skin or mucosa, and the long pulse width Nd:YAG (1064 nm) can produce good effects.

As for the patients with wide focal distribution, if they keep wearing elastic garment during the early stage, the progression of the patient's conditions can be delayed. If the patient suffers obvious functional disorder during infancy, scientific and continuous functional exercises are needed, so that the limb disorder after the patient becomes an adult can be mitigated; the surgical treatment at necessary time can improve functions (Fig. 12.40).

12.5 Arteriovenous Malformation

Yunbo Jin and Xiaoxi Lin

Arteriovenous malformation, previously called hemangioma racemosum, is a type of high-flow congenital vascular malformation, formed by tortuous and dilated arteries and veins which are interlocked and communicated, and there is no normal capillary bed between the abnormal arteries and veins. Arteriovenous malformation has a low incidence, and there is no obvious difference between males and females [27].

12.5.1 Pathogenesis

The pathogenesis of arteriovenous malformation has not been clear. It may be caused by incomplete communication and degeneration of arteries and veins in the original vascular plexus during the embryonic development. This embryological theory explains why arteriovenous malformations often occur to neck and neck, because the early embryo mainly consists of cranial structures. In addition, the theoretically consistent clinical phenomenon is that the facial arteriovenous malformations often occur to the buccal division and ears and the two sites occupy more surface areas than other facial regions in the early embryonic structures.

Most arteriovenous malformations are sporadic, but a minority of patients manifest familial aggregation. The recent molecular genetic studies have indicated that, in the patients with capillary deformities complicated with arteriovenous malformations, mutation occurs to the RASA1 gene that expresses p120-rasGAP33 on chromosome 5q. Although the reasons for the progression of patient's conditions of arteriovenous malformations remain unknown, they can be predicted; the arteriovenous communication originally at the resting state will be reopened due to the hemodynamic changes and local ischemia induced by trauma, which may be the reasons for aggravation in patient's conditions.

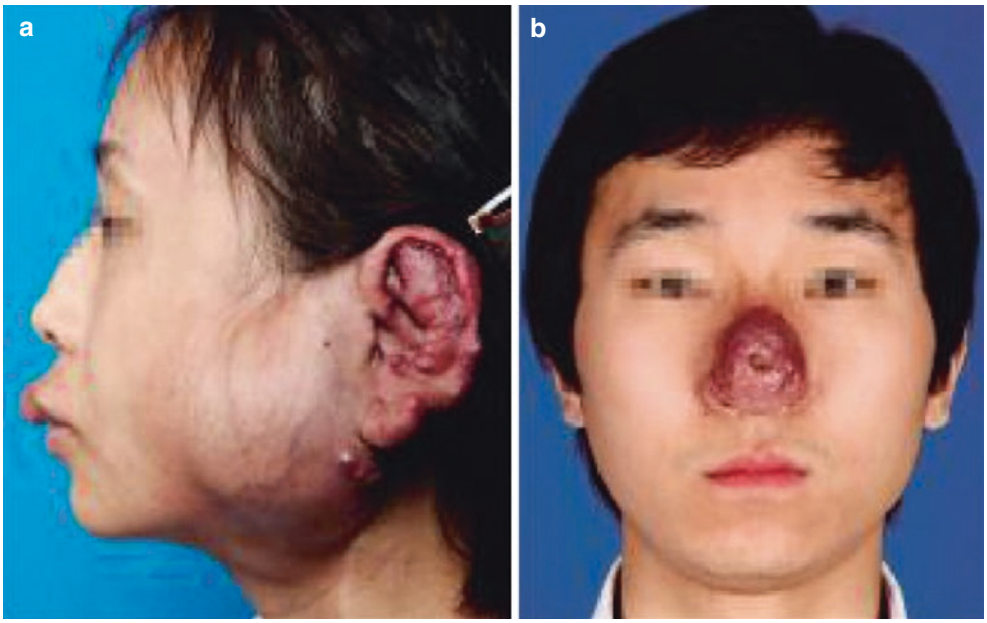


Fig. 12.41 Arteriovenous malformations. (**a**) Arteriovenous malformations of left ear. (**b**) Arteriovenous malformations of nose

12.5.2 Clinical Manifestation

Forty to sixty percent of the patients have already had the foci at birth, about 30% have visible skin injuries during the childhood, and the predilection sites are the head and neck (Fig. 12.41).

According to the severity of progression of patient's conditions, the arteriovenous malformations can be divided into four phases:

1. Resting stage: It is asymptomatic usually from birth to puberty. The foci during this phase are inconspicuous, or are only manifested as skin erythema, but skin temperature can be elevated. Some foci can stay at the testing phase for long.
2. Dilating stage: It usually starts from puberty, the foci become enlarged progressively, the color is deepened, skin and deep tissues are involved, pulsation or tremor is palpable, and blood flow murmur can be heard. The changes in the hormone secretion during the puberty, improper surgical resection and embolotherapy, trauma, and pregnancy are the main factors that drive the resting stage to progress into the dilating stage.
3. Destruction stage: The foci show symptoms such as pain, spontaneous skin necrosis, chronic ulcer, and difficult-to-control bleeding, and they are the results of the long-term progression and destruction of foci.
4. Decompensation stage: Due to long-term hemodynamic abnormalities, the patients suffer congestive heart failure and in severe cases it can be fatal.

The clinical manifestations of hand arteriovenous malformations are various, such as swelling, pulsation, high skin temperature, pain, bleeding, ulcer, and necrosis. The hemodynamic abnormalities of hand arteriovenous malformations with obvious fistulous orifice and a high flow rate may lead to heart overload and congestive heart failure. The conditions of some patients may still be stable when they become adults. Trauma, surgery, and hormones can induce hemodynamic changes and further promote the progression in patient's conditions.

12.5.3 Diagnosis

The main diagnosis evidence of arteriovenous malformations is clinical manifestations and imaging examinations. Ultrasound and color Doppler often serve as the initial examinations so that the hemodynamic characteristics of the foci can be generally known. MRI can display the range of foci, and the flowing void effect helps to determine the presence of high flow vessels. Computed tomographic arteriography can display the vascular components of arteriovenous malformation foci and their relation with bone tissues. Digital subtraction angiography (DSA) is an imaging examination necessary for the diagnosis of

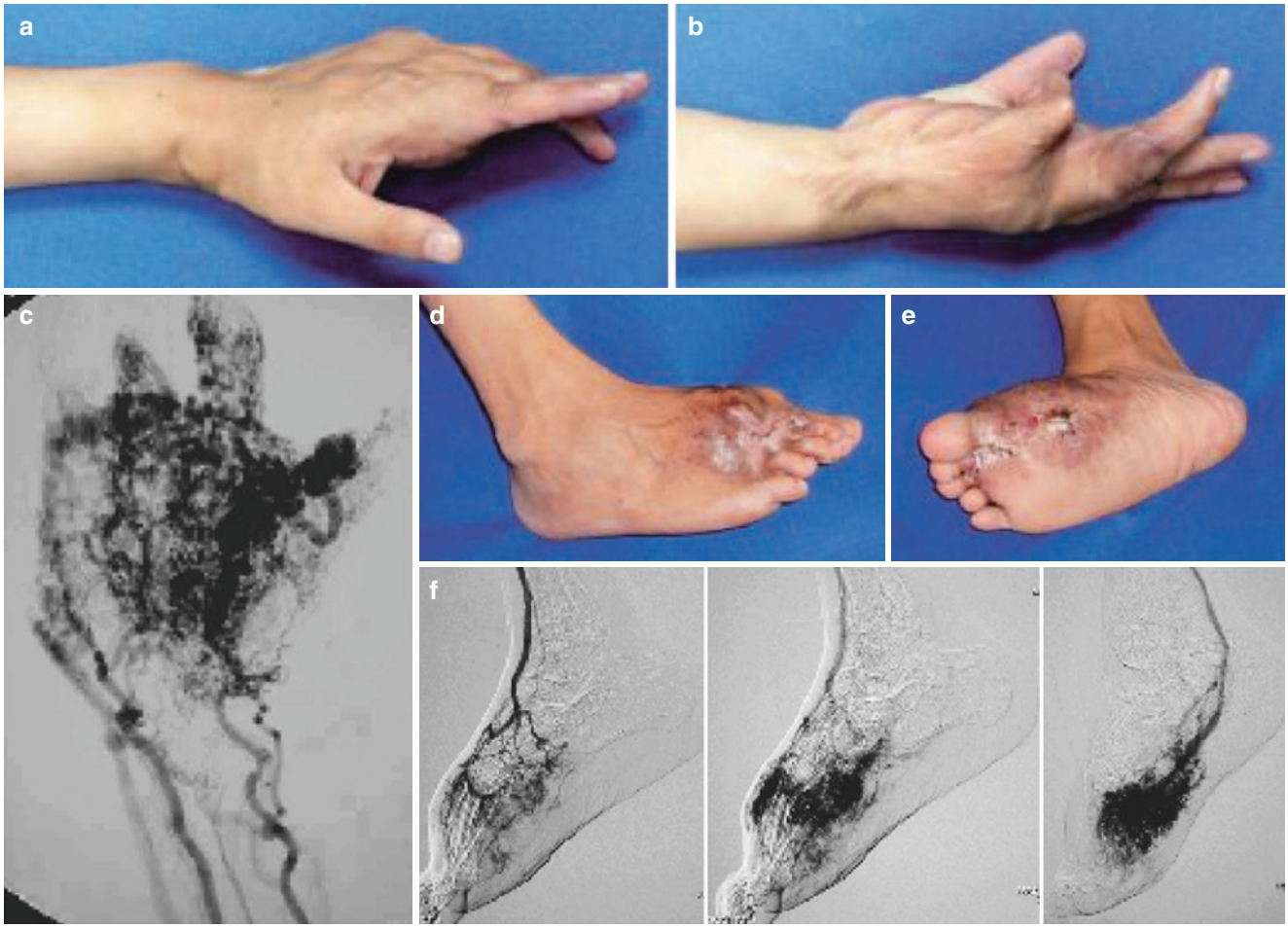


Fig. 12.42 Arteriovenous malformations of hand and foot and their DSA contrastographic pictures

arteriovenous malformations, which is also the evidence for the selection of proper treatment methods (Fig. 12.42).

12.5.4 Treatment

1. Selection of treatment methods: The main treatment measures of arteriovenous malformations are interventional embolotherapy and surgical resection, and a reasonable choice needs to be made according to the size, site, and staging of the foci [28–33].
   (a) For the patients at the resting stage and some patients at the dilating stage, it is necessary to perform superselective embolotherapy to be as close as possible to the foci so that the purpose of complete embolism of foci can be served.
   (b) For the patients at the destruction stage and the patients at the dilating stage that cannot receive superselective embolization, embolotherapy can first be performed and then the foci can be resected completely to repair the wounds.
   (c) For the patients at the resting stage who cannot receive superselective embolization, the patient can suspend the follow-up visits for observation.
   (d) For the foci which are surgically unresectable and to which superselective embolization is inapplicable, the patients can only receive palliative embolotherapy.
2. Typical case.
   (a) Case one: A 19-year-old female patient suffered congenital left sole erythema, which gradually becomes enlarged into red lumps; 1 year ago, the patient's lump suffered ulceration and bleeding; at a local hospital, she received partial resection and skin-grafting twice, which still became aggravated, and she could not walk. After admission, the patient underwent surgery in combination with interventional treatment (Fig. 12.43).

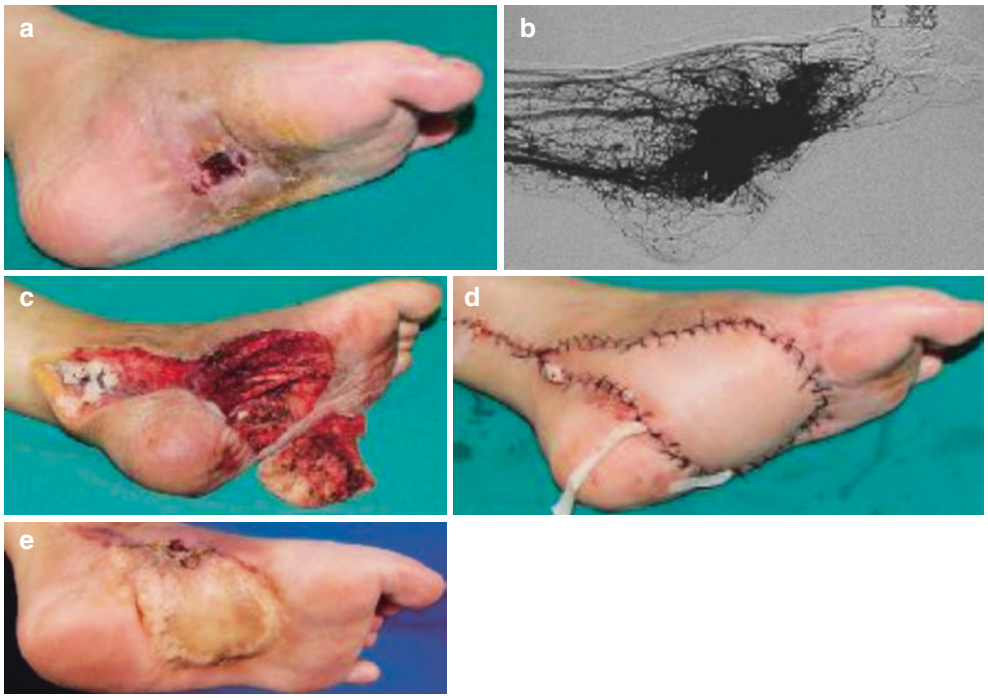


Fig. 12.43 Surgery in combination with interventional therapy for arteriovenous malformations of left sole. (**a**) Preoperative ulceration and incrustation of foci at the left sole. (**b**) DSA indicates that a large amount of abnormal vascular imaging is observed in the left sole, and the blood supply is from the posterior tibial artery. (**c**) During the operation, the foci should be completely resected as much as possible. (**d**) Use the right femoral anterior lateral free skin flaps to repair the wounds. (**e**) After operation, the wounds of the medial incisional margins of the sole did not heal after a period of time; DSA indicates residual foci but another resection may result in the possibility of foot necrosis; after the patient underwent the absolute alcohol interventional treatment once on the marginable foci of the wounds, the wounds became gradually healed

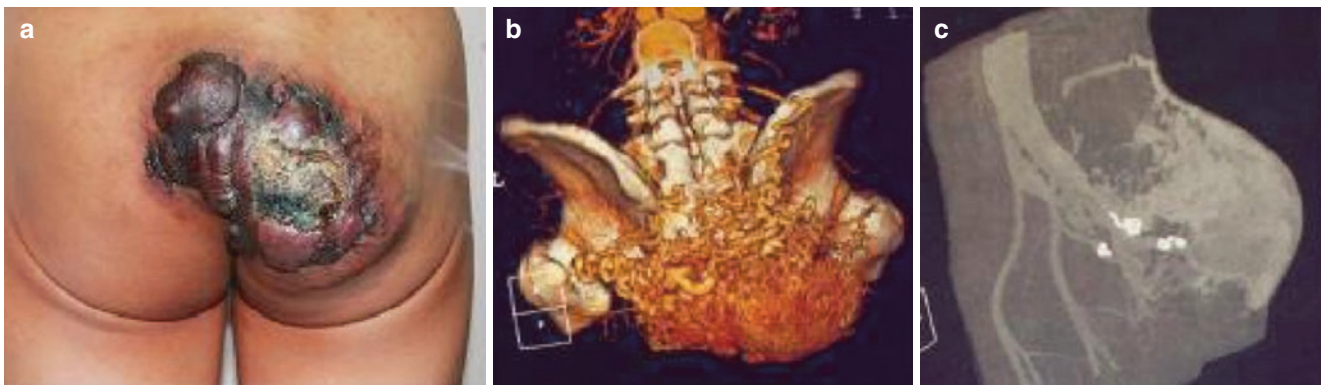


Fig. 12.44 Surgical treatment of hip arteriovenous malformations. (**a**) Preoperative ulceration and incrustation of hip foci. (**b**) CTA indicates the relation between the deformed vascular masses and the pelvis. (**c**) CTA indicates the presence of emboli. (**d**) CTA indicates the relation between foci and blood-supplying arteries. (**e**) DSA indicates the presence of a large quantity of arteriovenous fistulous orifice, whose blood is supplied by superior and inferior gluteal arteries. (**f**) Postoperative DSA indicates the disappearance of deformed vascular masses. (**g**, **h**) After complete resection of foci, the patient underwent the repair with free flaps at broadest muscle of back; 3 years after operation, the flap survival was good, but local swelling and sagging are observed, and the patient refused to receive further reshaping of the appearance

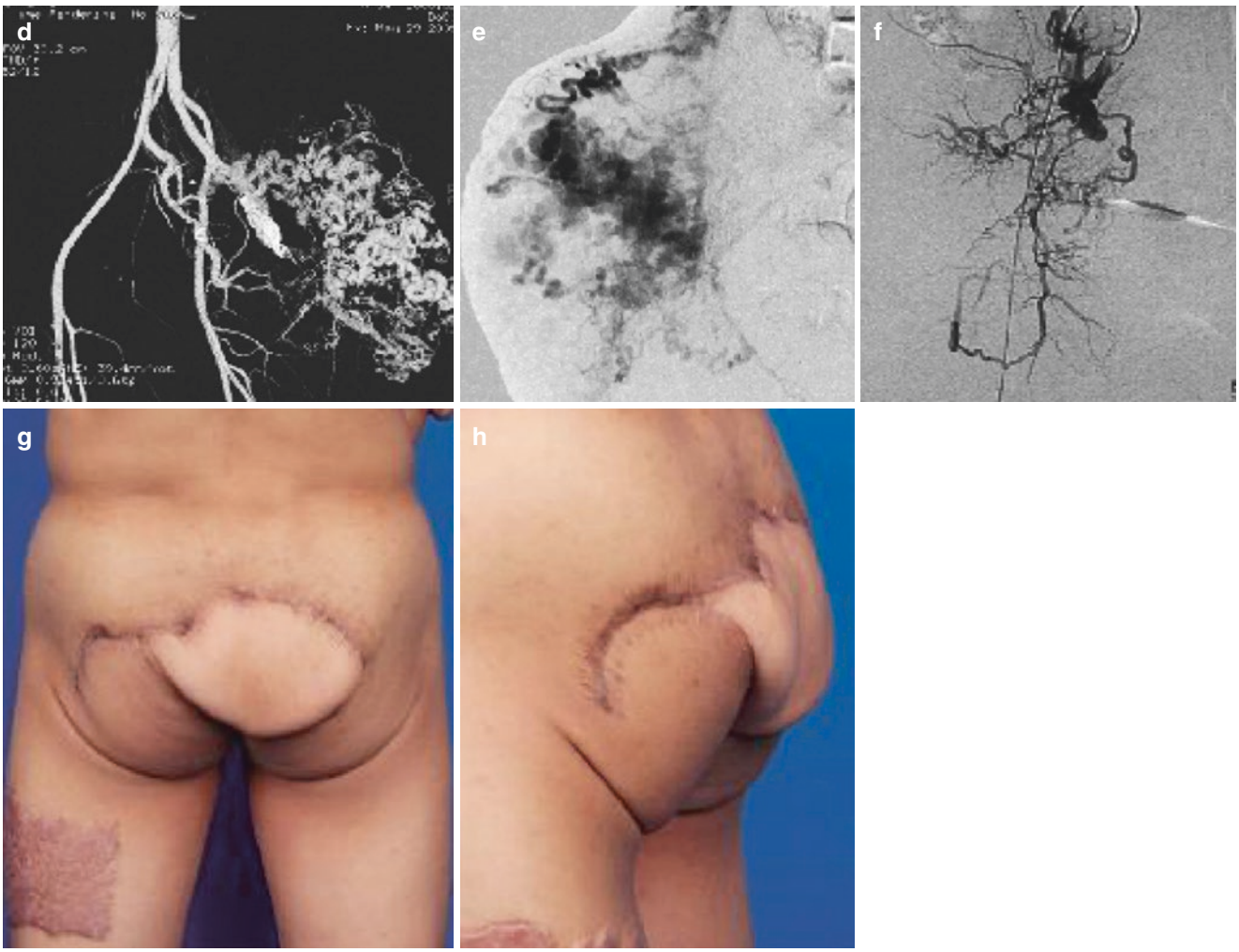


Fig. 12.44 (continued)

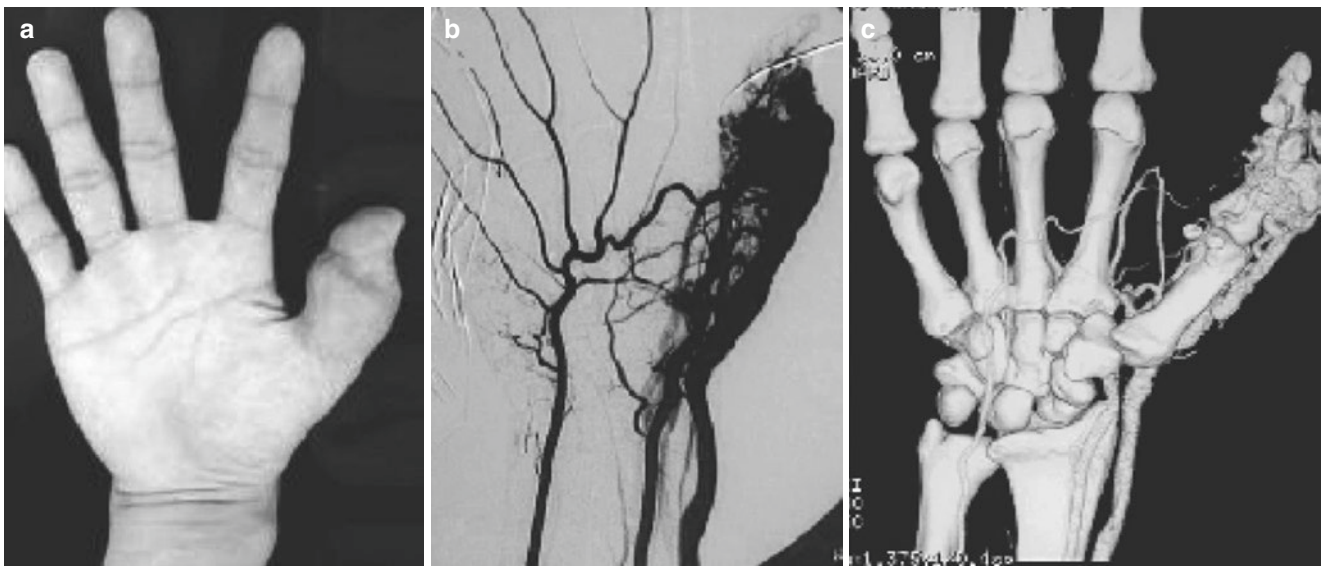
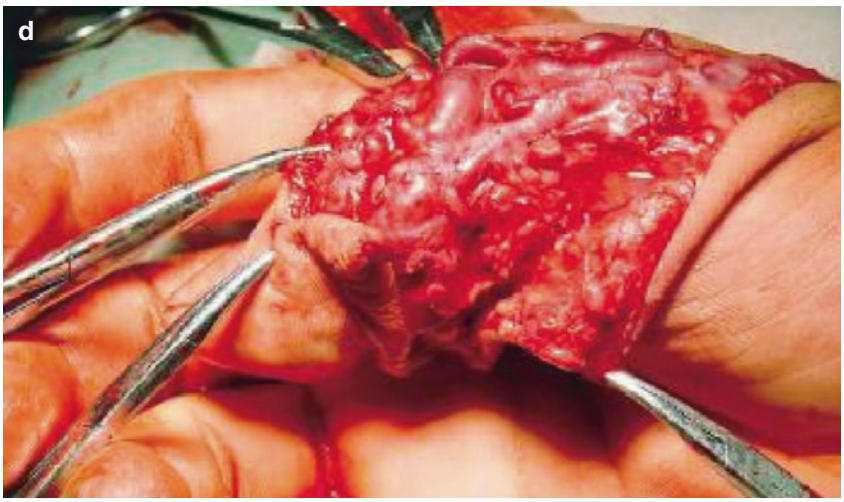
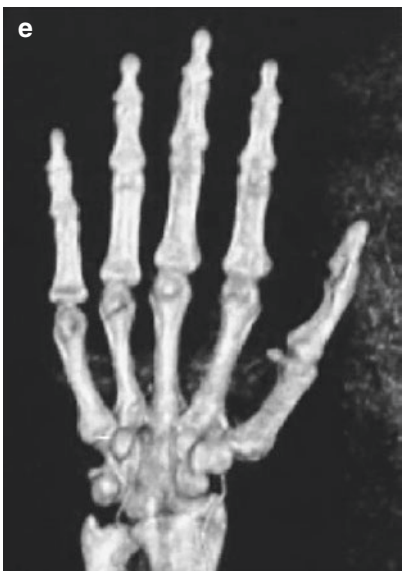
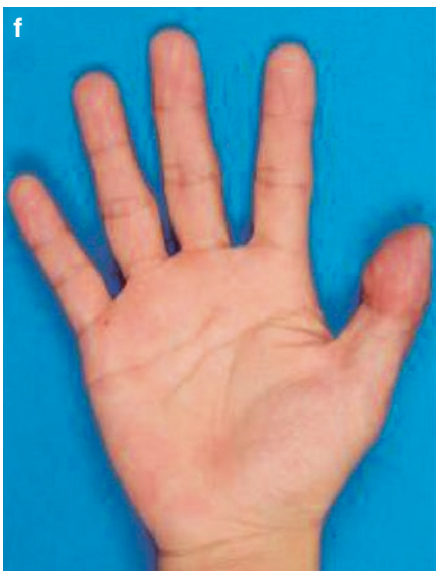


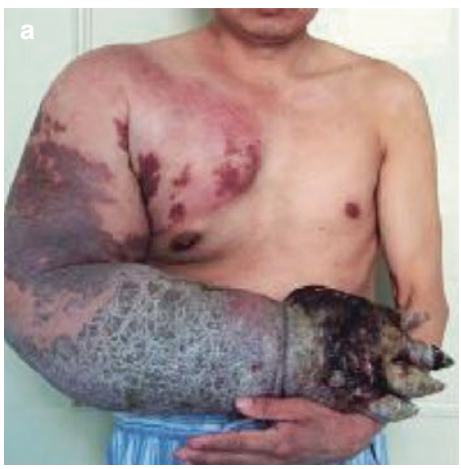
Fig. 12.45 Surgical treatment of arteriovenous malformations of the right thumb. (**a**) Preoperative pulsatile masses in the right thumb. (**b**) DSA indicates the presence of a large amount of abnormal vascular imaging. (**c**) CTA indicates the presence of deformed vascular masses. (**d**) During operation, a large quantity of tortuous and deformed vessels were observed. (**e**) Postoperative CTAs indicate no presence of obvious residual foci. (**f**) The operative 10-year follow-up visit indicated that the appearance and functions of thumb were normal

Fig. 12.45 (continued)









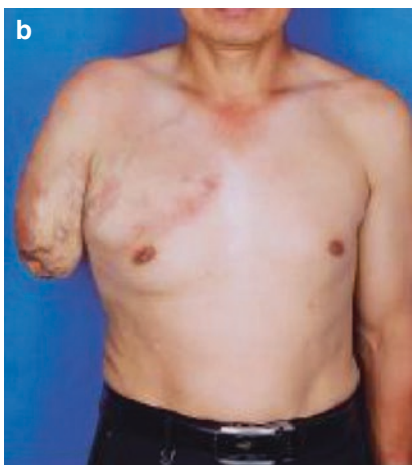


Fig. 12.46 Amputation of giant arteriovenous malformations in the right upper limb. (**a**) The preoperative appearance of foci of the right upper limb. (**b**) At 7 years and 8 months after the amputation, the patient recovered well and no recurrence occurred. (**c**) CTA indicates the presence of diffuse foci at the forearm. (**d**) CTA indicates the presence of a large quantity of tortuous and thickened blood-supplying arteries. (**e**) MRI indicates the presence of a large quantity of flowing void shadows. (**f**) The subclavian artery becomes obviously thickened. (**g**) DSA indicates the foci above the elbow joint were not obvious, and only the dilated blood-supplying arteries were observed. (**h**) DSA indicates the presence of wide diffuse foci in the forearm and hands. (**i**) Angiography indicates the presence of a large quantity of abnormally proliferative vessels in the forearm and hand

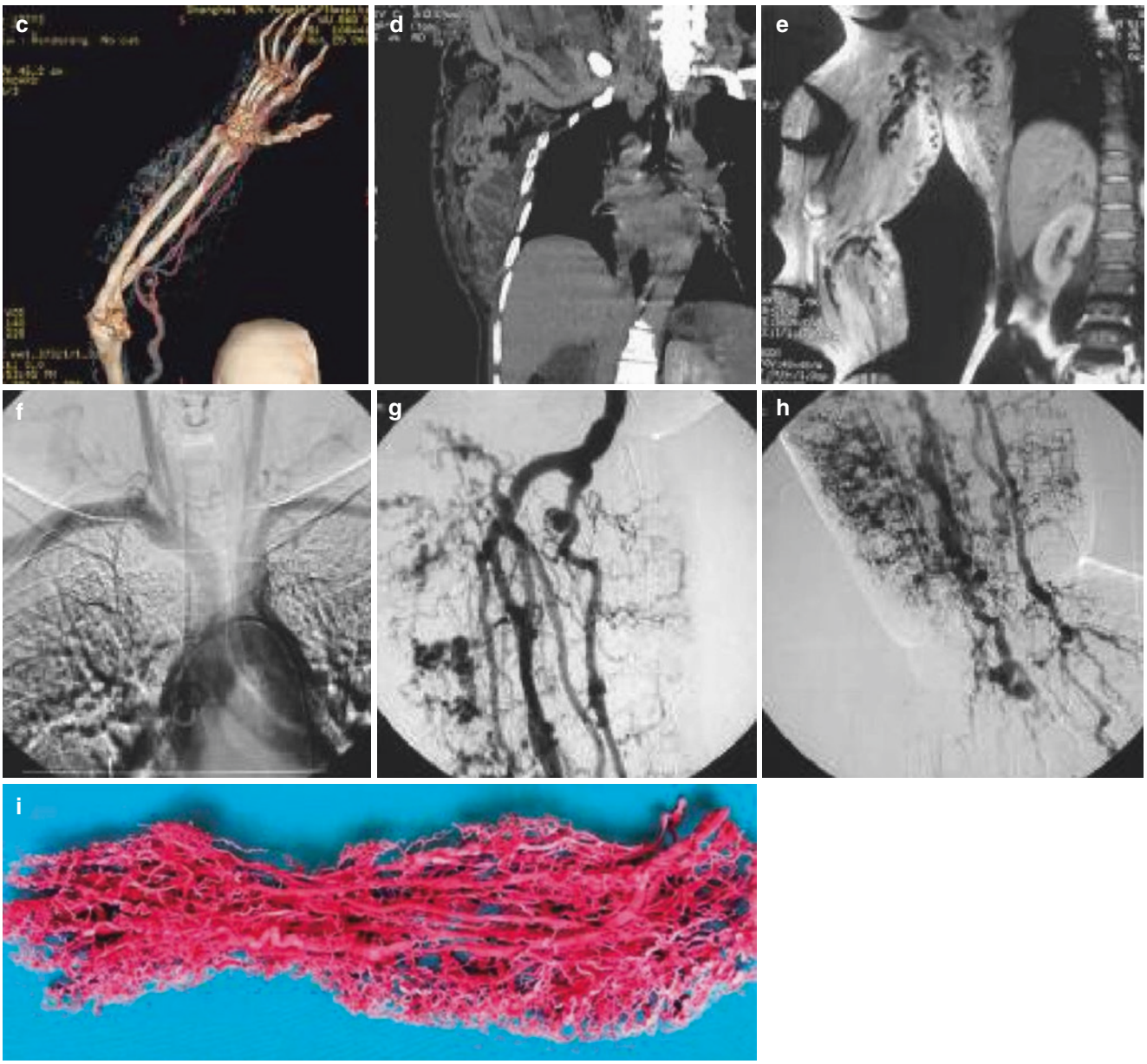


Fig. 12.46 (continued)

(b) Case II: A 38-year-old male patient had congenital hip lumps, which became aggravated after receiving the spring coil embolism at a local hospital, the foci had frequent ulceration and bleeding, he could not take plain sitting posture, and the life was threatened; after admission, the patient underwent surgical treatment (Fig. 12.44).
(c) Case III: Surgical treatment of right thumb arteriovenous malformations (Fig. 12.45).
(d) Case IV: A 51-year-old male patient suffered giant arteriovenous malformations with infection and necrosis of the right upper limb; he once underwent the ligation of blood supplying arteries at other hospital; after operation, the patient's conditions became progressively aggravated; after admission, the patient underwent amputation (Fig. 12.46).

It should be noted that, when interventional or surgical treatment is given, the only embolism of the main blood-supplying artery or the ligation at the proximal end of the blood-supplying artery should be avoided; otherwise, it may lead to a large amount of patency of the collateral circulation, which may further aggravate the original conditions. Due to the constant progress of embolism materials and technology,

the indications of intravascular treatment are being expanded: not only effectively reducing the volume of foci and controlling the symptoms of pain and bleeding but also improving the appearance to some extents.

The treatment of hand arteriovenous malformations is difficult because in treatment, the hand functions should be protected as much as possible, and the incidence of postoperative complications is high. The treatment modes of hand arteriovenous malformations include conservative treatment, focus resection, amputation, and interventional embolism, and the therapeutic regimen should be comprehensively evaluated by multidisciplinary treatment teams in good mastery of surgeries and interventional techniques; the decision of treatment should be made only after the evaluation indicates that the benefits outnumbers the risks.

For the patients without obvious symptoms and with stable conditions, conservative treatment should be selected, such as using elastic gloves for local compression.

There are many risk involved in focal resection, such as the difficult-to-control intraoperative bleeding, difficult-to-completely-resect foci, and high postoperative recurrence. The focus residue or ligation of blood-supplying arteries will aggravate the progression of patient's conditions, and this will make the subsequent treatment more difficult. As for the hand arteriovenous malformations with serious complications, such as tissue necrosis and deformities, functional loss, or bleeding, if the foci are limited to the fingers, amputation may be a good choice.

Intravascular embolotherapy can serve as not only an independent treatment method but also a supplement to surgical operation. Embolic agents include N-butylcyanoacrylate, absolute alcohol, N-butylcyanoacrylate, etc.; among them, absolute alcohol is the most powerful embolic agent, has clear effects on hand arteriovenous malformations, and can impeccably retain functions and improve appearance. However, the operation on this technique has very high requirements, and the operation should be performed on experienced specialists; otherwise, serious complications are apt to occur. Systemic complications include acute alcoholism, erythrocatalysis, and cardiopulmonary collapse, and local complications include skin blister, necrosis, and injuries of adjacent injuries.

12.6 Lymphatic Malformation

Hui Chen Xi Yang and Xiaoxi Lin

Lymphatic malformation, previously called lymphangioma, is a group of diseases characterized by congenital lymphatic vascular development deformities and lymph fluid circulation disorder, and the etiologic factors still remain unknown. According to the morphological features of lymphatic malformations, lymphatic malformation is often divided into microcapsular lymphatic malformation and macrocapsular lymphatic malformation, and the mixed foci shared by both of the two types are not uncommon. Its classification is closely correlated with the selection of the treatment methods.

12.6.1 Pathological Characteristic

The lymphatic malformations mainly consist of dilated lymphatic vessels, it is considerably distributed in the dermic superficial layers or mucosa, and it can invade the subcutaneous regions, submucosal regions, and muscles. The dilated lymphatic vessels are lined with nonproliferative endothelial cells and form the thin-walled capsular space with varying sizes; the cavity is full of lymph fluid and rich in lymph cells, blood red cells, and neutrophils. The capsular space of microcapsular lymphatic malformation is small and dense and takes a honeycomb shape; macrocapsular lymphatic malformation consists of single or several vascular lumens with a large volume.

12.6.2 Clinical Manifestation

Lymphatic malformations mostly occur during the infancy; and nearly 50% of the foci are located in the head and neck, and generally the growth is slow. The foci can present local or diffuse distribution, they can involve the cutaneous and subcutaneous regions, and they can invade muscles and internal organs. The microcapsular lymphatic malformations are relatively common in sites such as eyelids, neck, proximal ends of four limbs, tongue, and mouth floor with the manifestations of bulging soft masses, not obvious sensation of compression, and negative body position test. The microcapsular lymphatic malformation located at the skin is often manifested as multiple skin vesicles or wart-shaped nodules, can be accompanied by skin hyperkeratosis, and can involve a large range of skin; when it is located at the mucosa, prunosus or dull red miliary micro-lymph follicles can be formed. Macrocapsular lymphatic malformation, previously called capsular hydroncus, often occurs to the neck region, hydroncus, inguinal groove, and chest wall, presents cystic lumps, and has fluctuation sensation and can extend to the mouth floor and retroclavicular regions along the nerves, vessels, and tissue space and reach the mediastinal septum. In a minority of patients with a rapid growth, the trachea and esophagus can be compressed. Most of the hand lymphatic malformations are the microcapsular type.

The commonest complications of lymphatic malformations are infection and bleeding; in case of bleeding, the vol-

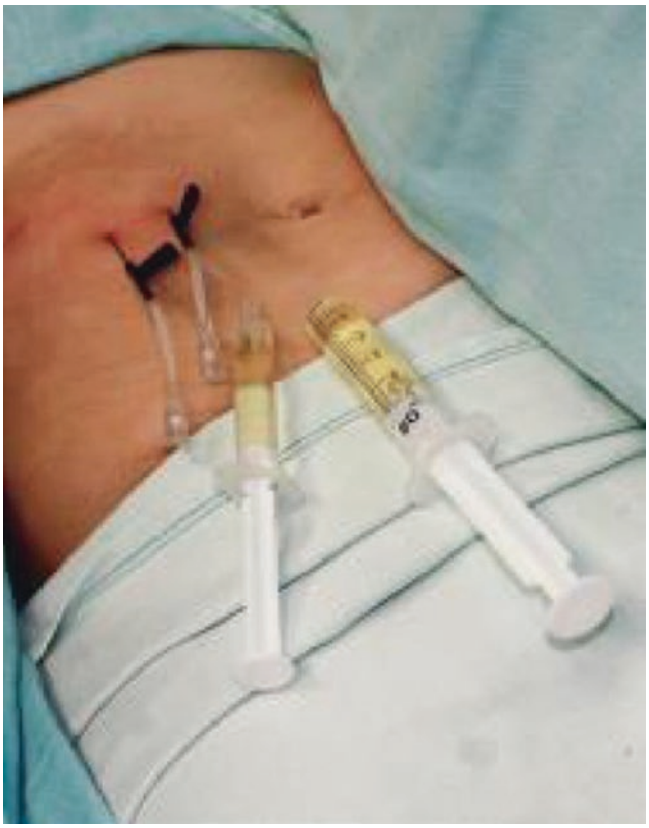


Fig. 12.47 Abdominal wall macrocapsular lymphatic malformation; after puncture, light yellow lymph fluid can be sucked out

ume of foci suddenly becomes enlarged, and the skin experiences ecchymosis. As for the lymphatic malformations located at special sites, in case of repeated occurrence of infection and swelling, functional disorders can be induced such as impairment in visual acuity and dysphagia. The wide lymphatic malformations that occur to the limb can be accompanied by lymphedema with changes in skin and appearance disorder. Wide lymphatic malformations can be accompanied by hyperproteinemia and hypolymphemia. In case of coexistence of lymphatic malformations and vascular malformations, the disease can be called capillary lymphatic malformation or venous lymphatic malformation according to the pathological characteristics of vascular deformities.

12.6.3 Diagnosis

The diagnosis of lymphatic malformations is mainly dependent on physical examination, diagnostic puncture, and imaging examination. After diagnostic puncture, light yellow bright liquid (Fig. 12.47) can be sucked out, and in case of intracapsular bleeding or complication with venous malformations, blood can be sucked out. The transillumination test of macrocapsular lymphatic malformation is positive. Enhanced MRI is the most important diagnosis and differential diagnosis method, obvious high signals are presented in the T2-weighted images, but there is no inten-

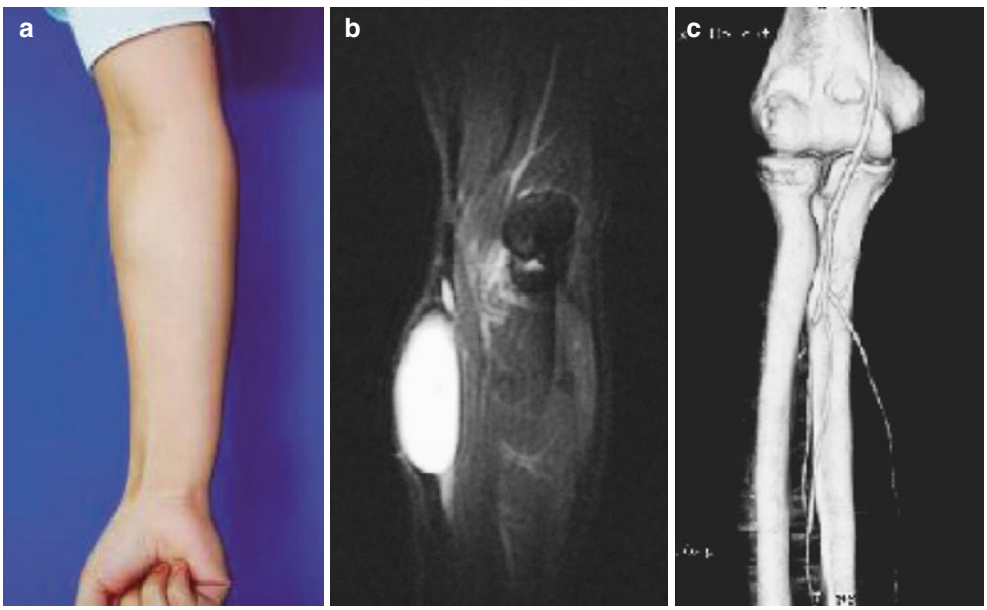


Fig. 12.48 Lymphatic malformation in the right forearm. (**a**) Bulging mass of the right forearm with a hard texture. (**b**) MRI indicates that foci in the T2-weighted images present homogeneous and well-defined high signals. (**c**) Computed tomographic arteriography cannot display the foci

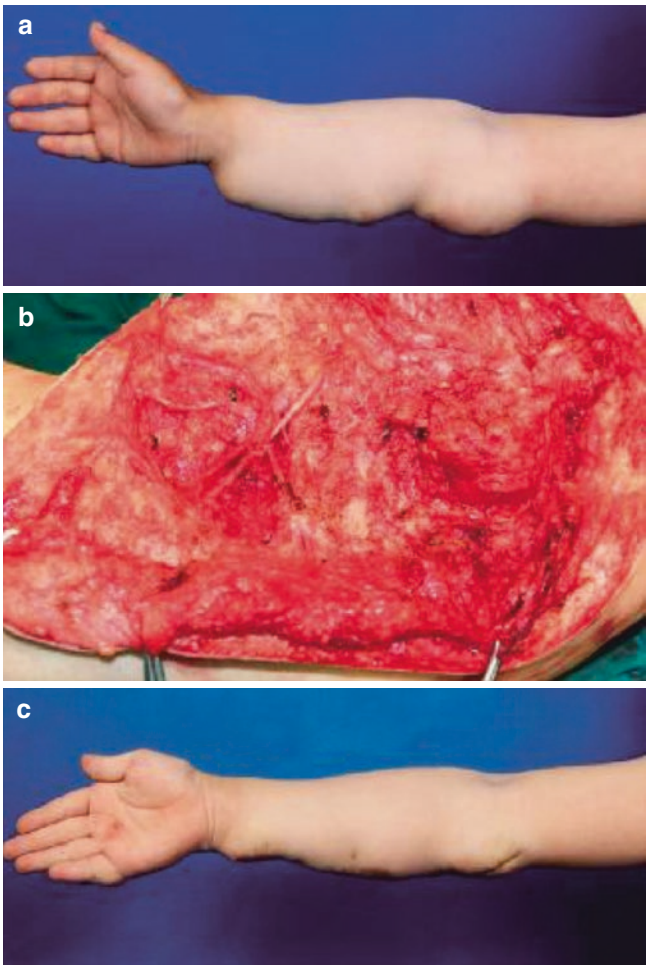


Fig. 12.49 Surgical treatment of microcapsular lymphatic malformation in the right forearm. (**a**) Bulging foci at the ulnar side of right forearm. (**b**) The intraoperative findings are a large quantity of fibrous connective tissues, mixed micro-lymphatic vessels, and the presence of a large amount of lymph fluid exudation in the wounds. (**c**) The postoperative appearance is obviously improved

sification (Fig. 12.48). In case of intra-focal bleeding, obvious fluid-fluid level can be observed. According to the above characteristics, it can be differentiated with infantile hemangioma and venous malformations.

12.6.4 Treatment

The current treatments are mainly injection of sclerosing agents and surgical resection. As for macrocapsular lymphatic malformations, sclerotherapy can often produce satisfactory effects. The currently common drugs are pingyangmycin and hemolytic streptococcus preparations, proper injection methods can be selected according to the sites of foci, and tissue necrosis and injuries to important nerves and glands should be avoided. For microcapsular lymphatic malformation, sclerotherapy is generally not apt to make the foci shrink obviously, and the combined therapy can be surgical resection (Fig. 12.49). It is difficult to resect completely the hand microcapsular lymphatic malformation, and such malformation is apt to recur. If the foci are located on the upper eyelid and frontal part and affect eye opening, it is a good surgical indication, but most foci cannot be resected completely. The lymph follicles located at the mucosa can be removed by using Nd:YAG laser or pulse dye laser [34, 35].

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Congenital Tumor of the Hand and Upper Limb

13

Jinghong Xu, Jialiang Chen, Qifang He, Liulong Zhu, and Jianmin Yao

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The bone tumor is classified into two types: primary and secondary. The incidence of primary bone tumor is 2/100,000 to 3/100,000, accounting for about 2% of all the tumors, and it can also be classified into tumor-like lesions, benign type and malignant type. The primary bone tumor is uncommon during the childhood; the commonest type is osteochondroma, followed by chondroma, osteoma, and ossifying fibroma. Generally, osteochondroma grows and becomes large within the initial 10 years, and the development process will end due to the closure of growth plates during the puberty. Generally, it takes a pedicled shape or wide basal shape, the difference in size is great, it is located at the endochondral bone of the long bone, it is commoner in the lower limb bones than in the upper limb bones, and there is a tendency of symmetric occurrence. It is the commonest at the lowest segment of the femur and the upper end of the tibia, followed by the upper end of the humeral bone, the lower end of the radial bone, the lower end of the tibia, and the upper end of the calf bone, it rarely occurs at the short bones and the irregular bones, the occurrence is occasionally seen in the spinal column, spinous process is often involved, and sometimes it can be complicated by other limb deformities. During the first onset, the pediatric patient generally shows no obvious symptoms, so it is often found during the childhood and puberty. Most of the patients visit a doctor due to deformities, hard painless tumor is palpable locally or in multiple sites, and when the tumor squeezes the surrounding nerves and vessels, pain and joint movement disorder can be induced.

Generally speaking, the diagnosis and treatment of benign tumor are simple, and early diagnosis and radical treatment are needed for malignant tumor. This section will focus on the discussion of the etiologic factors, pathology, clinical manifestations, diagnosis, and treatment of congenital tumors.

13.1 Osteochondroma

Jinghong Xu, Jialiang Chen, Qifang He, Liulong Zhu and Jianmin Yao

Osteochondroma is a common benign tumor. During the early stage of the nineteenth century, osteochondroma was first described. According to the WHO statistics, osteochondroma accounted for 19.89% of the primary tumor and accounted for 43.80% of the benign tumor. Osteochondroma is a common tumor during the childhood and puberty, the incidence before the age of 20 accounts for 70–80%, and it is common in boys.

Osteochondroma can be divided into single type and multiple type, the hereditary characters of single osteochondroma are not obvious, and it is often occasional; multiple osteochondroma is an autosomal dominant hereditary disease, and therefore, it is commonly called hereditary multiple exostoses (HME).

The incidence of HME is about 1/50,000 [1], the ratio between male and female is about 1.5:1, and about 62% of the patients have a clear family history [2]. The mutation of EXT1 and EXT2 genes located at the 8q24 and 11p11-p12 is closely correlated with the occurrence of HME [3], they contain 11 and 16 exon coding segments, the onset of 44–66% of HME families is correlated with EXT1 [4], and 27% of the onset is correlated with EXT2 [5]. EXT serial genes fall into the category of tumor-suppressing gene, and how the mutation or absence of the two induces the tumor differentiation of cartilage cap is currently in the middle of research [6].

13.1.1 Solitary Osteochondroma

13.1.1.1 Site of Onset

Solitary osteochondroma generally occurs to the site of chondrified bone; metaphyses at the distal femur, proximal humerus, proximal tibia, and proximal fibula are often involved; and it is gradually away from epiphyseal plate with the growth and development [7].

13.1.1.2 Etiologic Factor

The pathogenesis of solitary osteochondroma remains unclear, and lesions and injuries around the growth plates may also be the reasons for its onset. The studies of animal experiments indicate that injuries in growth plates can generate typical osteoid osteoma. Other scholars indicate that solitary osteochondroma is formed by the separation of epiphyseal cartilage plates during the development and the formation of ectosteal traverse growth [8]. Some scholars also point out that solitary osteochondroma is a periosteal abnormal development. The displaced cartilage nests continue to grow, inducing developmental epiphyseal growth defect.

13.1.1.3 Pathology

The lesions present hill-shaped protrusion or are pedicled lumps, the diameter ranges from 1 to 10 cm, and the surface is uneven. The appearance of exostosis can be hill shaped, semicircular, cauliflower shaped, or pedicled long tubular shaped.

Solitary osteochondroma is the bony bulge with cartilage cap, it occurs to osseous external surface, it has marrow cavity, and it is connected with the marrow cavity of the basal bone. The lesions can be divided into three layers; the outermost layer is the fibrous perichondrium, connected with the periost of the basal bones; the second layer is the cartilage cap, whose thickness is often over 2 cm (it becomes thin with the increase in age), the superficial cartilage cells in the cartilage cap present cluster-shaped distribution, the cartilage cells in the adjacent bone migration regions are arranged in the cord shape, which is similar to the epiphyseal plates, and ossification is observed inside the cartilages; bones are located in the innermost. The cartilage structure disappears, the fibrous zone becomes widened, the lesions are mucus-like, the density of cartilage cells is increased, the division activities are increased, and the obvious cartilage cell heteromorphism and necrosis indicate the possibility of canceration. In case of accompanying fractures, fibrocyte reactions can be locally displayed.

13.1.1.4 Clinical Manifestation

Solitary osteochondroma is common in children or teenagers, and the patients that suffer it before the age of 20 account for 70–80%. This disease is generally asymptomatic in clinical manifestations. The patient visits the hospital because the parents of the pediatric patient find the bony mass that becomes gradually enlarged around the joint. Slow growth of the tumor can last for as long as 30 years, but sometimes, the growth speed is unstable. In the symptomatic cases, the symptoms are often correlated with the size and site of the lesions, and some tumors can cause pain, which are induced by repeated injuries of protruding sites and compression of local muscles and vascular bundles.

The physical examination indicates that one hard mass without tenderness is palpable, and as for the slightly large osteochondroma, one subcutaneous bulge is palpable. Scapular osteochondroma is often located in front of scapulae, and pain and snap can be caused during joint movement; due to the overlap between the scapula and lungs, the intrapulmonary nodules are misdiagnosed by chest X-ray radiography. A minority of patients will be found with the phenomenon of spontaneous absorption of osteochondroma during the childhood or adolescence to reduce the shrinkage or disappearance of tumor, the reason might be that the pedicle of osteochondroma will experience bone fracture after injuries, and due to active bone absorption, the tumor can be absorbed; the reason might also be that the osteochondroma is gradually fused into the enlarged metaphyseal bone during growth and development.

The complications of solitary osteochondroma are common, including bone fracture, skeletal deformity, vascular nerve injuries, synovial cyst formation, and canceration.

13.1.1.5 Imaging Examination

1. X-ray examination: The X-ray characteristics of osteochondroma located on the long backbones is the presence of bony bulge on the bone surface, which is connected with metaphysis, and the lump consists of cortex of bone and cancellous bone. As the shape of tumor basilar part varies, it can be divided into pedicled type (the stem is narrow, the top is wide) and pedicleless type (the fundus is wide and flat). Osteochondroma often occurs to the adhesion of the tendon ligaments at the metaphysis, the growth tendency is consistent with the direction of force generated by tendon ligaments, and the growth is along the direction from the metaphysis to the backbone. The top of the tumor is covered by the cartilage cap, whose thickness varies, the patients with the thin cap only present linear-shaped transparent regions, and it cannot be observed easily; the patients with the thick cap present cauliflower-shaped dense shadows. In case the cartilage cap is thin, the borders are defined with regular spot-shaped calcification, and benign growth is observed; in case the cartilage cap is thick and large, the borders are less defined with irregular calcification, and attention should be paid to the possibility of canceration (Fig. 13.1).

   The X-ray findings of osteochondroma in hand and foot short bones and long tubular bones are similar; it should be noted that small osteochondromas may be present in finger and toe tips, which are also called subungual osseous wart.
2. CT or MRI examination: As for typical manifestations of solitary osteochondroma, the needs of diagnosis and treatment can be satisfied only by the X-ray examinations; as for a minority of osteochondroma cases with the

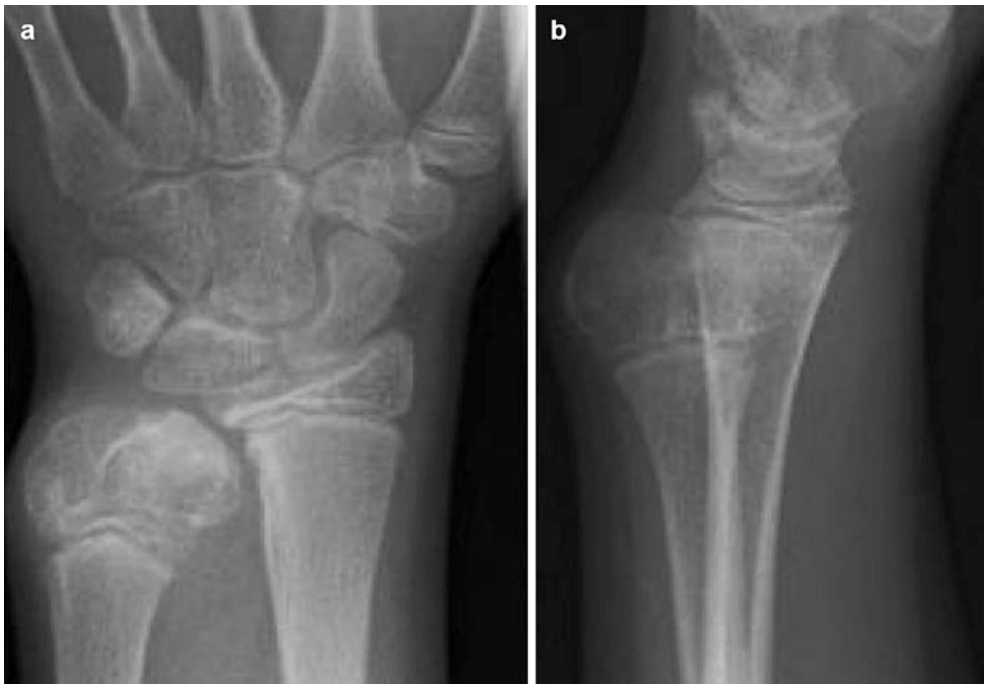

Fig. 13.1 Osteochondroma at the ulnar metaphysis. (**a**) Anteroposterior position. (**b**) Lateral position

atypical manifestations or complicated sites in anatomical structures (e.g., scapulae and pelvic cavity), CT or MRI can be adopted to ascertain the position of lesions and their relation with the surrounding tissues. As for the osteochondroma of long tubular bones, CT examination can provide the relation between tumor and affected bone, type of diseased matrix, calcification, and thickness of the cartilage cap and facilitate the differentiation of osteochondroma and periosteal chondrosarcoma. It should be noted that normal variations such as internal process of humerus (it is the congenital dysplasia) and intra-tibial hip bone osteochondrosis (also called teratogenous osteomalacia and tibial introversion) should be not misdiagnosed with osteochondroma.

13.1.1.6 Diagnosis

In addition to the clinical manifestations, the typical X-ray findings are mainly based to determine the diagnosis of this disease, and for a minority of atypical patients, CT or MRI can be adopted to facilitate diagnosis.

13.1.1.7 Treatment

For the patients who are asymptomatic or have a small-sized tumor and develop slowly and whose finger functions are not affected, treatment is usually unnecessary, and changes in the fingers should continue to be observed closely.

The surgical treatment indications of childhood osteochondroma are: ① the growth speed of tumor is higher than what is expected and presents progressive increase; ② the tumor compresses the adjacent nerve vascular bundles or the covering muscles to produce symptoms; ③ the tumor located near the joints exerts influences on joint movement.

The osteochondroma of fingers and metacarpal bones in hands often leads to hand deformities, the relatively large lump is apt to disturb the activities and functions of fingers, so treatment is needed. The treatment principles of hand solitary osteochondroma are the following:

1. For the patients with a relatively large tumor which leads to finger deformities and affects finger functions, or which compresses the important tissues such as peripheral nerves and vessels and further induces corresponding symptoms, surgical resection should be performed. Under the conditions that the finger vessels and nerves are protected, during the operation the surgeons should perform the extra-fibrous-capsular separation of osteochondroma, the tumor should be fully exposed, and the normal bone margins around the tumor basal part are selected for resection of the entire lump. The results of pathological examination should cover complete fibrous capsule, cartilage cap lid, and bone fundus of the osteochondroma; otherwise, it is apt to induce tumor recurrence.
2. The resection of osteochondroma during the childhood is apt to induce injuries of adjacent growth plates, so for the clinically asymptomatic solitary osteochondroma with a small (less than 2 cm) tumor, surgical resection is unnecessary.
3. If the tumor is excessively large, or the basal part range is wide, and it is difficult to resect, the entire segment of bones invaded by the tumor can be resected, and the autogenous bones can be adopted to transplant and repair the formed bone defects.

4. For the patients suffering recurrence after tumor resection, the possibility of canceration should be ruled out according to the clinical manifestations and X-ray findings and in combination with biopsy if necessary. If the tumor is still benign, another local resection should be performed.

13.1.2 Hereditary Multiple Osteochondroma

13.1.2.1 Site of Onset

Hereditary multiple osteochondroma is also called metaphyseal aclasis, osteochondromatosis, or Ehrenfried disease. This disease can involve all systemic skeletons, but the involvement in the skull and vertebrae is rare; the typical sites of onset are the distal and proximal ends of the femoral bone, tibia and fibula, and the proximal end of humeral bones, and the onset in the radial and ulnar distal ends is slightly rare [9]. Experience shows that, if there is no exostosis around the knee joint, the diagnosis of hereditary multiple osteochondroma cannot be established. Compared with solitary osteochondroma, hereditary multiple osteochondroma tends to occur at the sites of the scapula, iliac bone, and ribs.

13.1.2.2 Etiologic Factor

Multiple osteochondroma has a genetic predisposition with the manifestations of autosomal dominant inheritance; if the parents suffer from this disease, about half of their children may suffer this disease.

13.1.2.3 Pathology

The hereditary multiple osteochondroma, sharing the same pathological appearance with solitary osteochondroma, is the bony bulge with the cartilage cap, it occurs to osseous external surface, it has marrow cavity, and it is connected with the marrow cavity of the basal bone. The lesions are divided into three layers: perichondrium, cartilage, and bones.

13.1.2.4 Clinical Manifestation

Male patients with this disease outnumber female patients, the incidence between males and females is about 3:1, and it is common in children and young people at the age of about 20. Its incidence is not high, only 5–10% of that of the solitary osteochondroma.

Hereditary multiple osteochondroma often presents asymmetric distribution and the number of lesion varies [10]. The patients may have tumor at 3–4 sites, but the tumor can occur at 10–15 sites at more sites, and the number of sites may be even over 100. The shape of the tumors is various, they are mainly distributed near the metaphysis of the long bones, and they can also occur to the spinal column, ribs, pelvic cavity, and scapulae. The clinical manifestations can be abnormal bulge at multiple sites of the limb or palpable bony mass.

The patients with multiple osteochondroma are generally short, but their height is within the normal height range of this age group, the regions beside the joints may appear bulging due to the existence of tumor, and the long bones of four limbs may suffer skeletal shortening and curvature deformity.

The pediatric patients can suffer disorder of some functions of the adjacent joints, which affects the rotation of the forearm, the flexion and extension of the elbow joint, the adduction and abduction of the hip joint, and the introversion of ankle joints.

Most of the pediatric patients with hereditary multiple osteochondroma have the characteristic X-ray findings, namely, the femoral neck is thick and short and attached with multiple bony neoplasms. The patients with hereditary multiple osteochondroma may suffer secondary osteochondroma after becoming 30 years old, and secondary osteochondroma is very rare during the childhood.

The complications of hereditary osteochondroma are common, including bone fracture, vascular nerve injuries, and synovial cyst in addition to skeletal deformities.

13.1.2.5 Imaging Examination

The X-ray signs of multiple osteochondroma are basically the same as solitary osteochondroma, and their lesions are very wide. The characteristics of hereditary multiple osteochondroma are as follows: it can induce defects in bone formation and skeletal deformities, the wrist joint gradually experiences ulnar deviation, the ulnar bones are relatively shortened, and the radial-ulnar separation deformity can be observed (Fig. 13.2). The typical manifestation of CT or MRI is the connection of the marrow cavity of basal part into the lesions.

13.1.2.6 Treatment

The treatment of hand hereditary multiple osteochondroma is the same as solitary osteochondroma. No treatment is needed for those showing no symptoms; in case of the presence of pain, limb functional disorder, and skeleton development deformities or complications, surgical treatment should be given. The correction of skeletal deformities should proceed after the bones are mature to avoid the occurrence of secondary deformities or the recurrence of deformities. The chance of canceration of hereditary multiple osteochondroma is high; if the tumor becomes enlarged rapidly within a short period of time, X-ray findings have the sign of canceration; for the patients with canceration validated by biopsy, the treatment given should be based on the diagnosis of osteochondroma, and amputation should be performed.

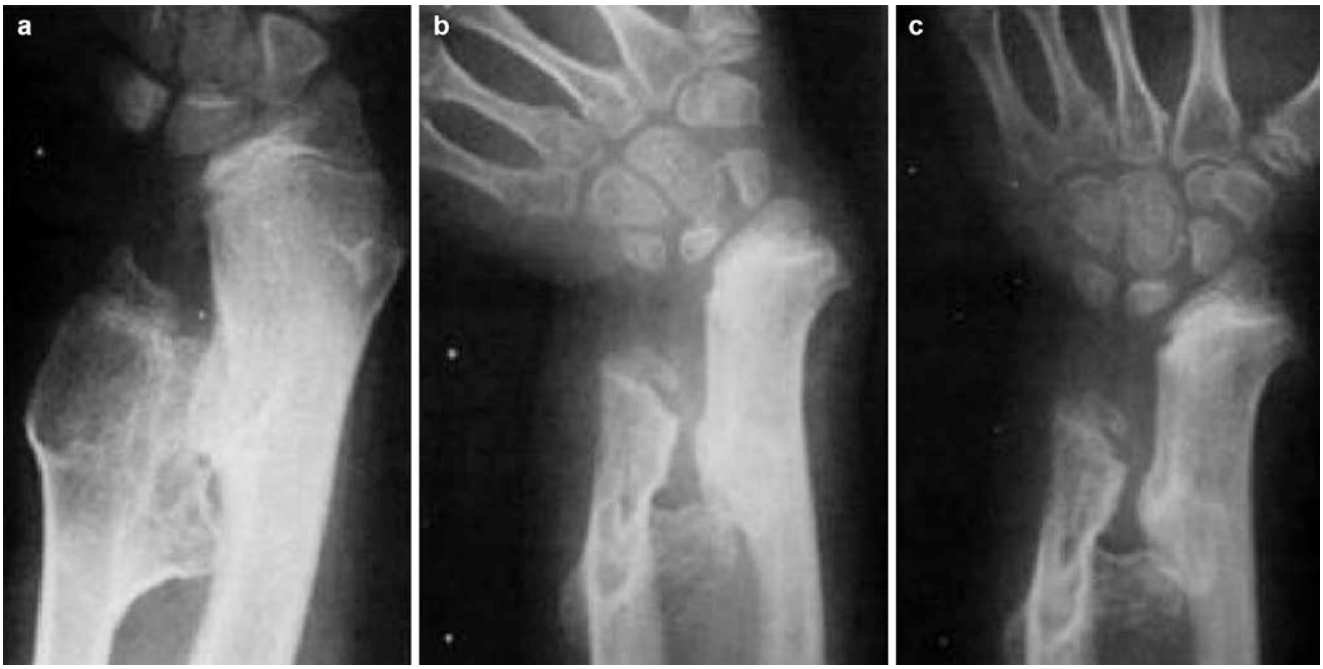

Fig. 13.2 Hereditary multiple osteochondroma complicated by skeletal deformities

13.2 Enchondroma

Chondroma is a common benign tumor, accounting for about 11% of the benign tumor. It can be divided into enchondroma, periosteal chondroma, and enchondromatosis. Chondroma is a hyaline cartilage tumor with multiple similar histological characteristics, but the onset sites and clinical features are different. Enchondroma and periosteal chondroma are sporadic cases, and enchondromatosis often occurs to the cases with congenital tumor syndrome.

Enchondroma is the intraosseous benign cartilage tumor; the tumor which invades the single skeleton is called single enchondroma, and the tumor which invades the multiple skeletons is called multiple enchondroma. As for the single enchondroma, the lesions are located inside the marrow cavity; as for the multiple enchondroma, the lesions are derived from the periosteum and subsequently penetrate into the marrow cavity.

13.2.1 Single Enchondroma

13.2.1.1 Site of Onset

The distribution of enchondroma is very characteristic. It often occurs to short tubular bones, and 2/3 of them are located in hands. The single enchondroma occurring to hands occupies 40–56% of all the enchondromas; the hand proximal phalange is the most frequently attacked site, accounting for 40–50% of hand enchondroma; the sequence is the metacarpus, middle phalange, and phalangette according to the frequency of occurrence, and it occurs most infrequently to the pollical phalange and rarely to the metacarpal bone. The enchondroma rarely attacks the foot, and enchondroma occurring to foot accounts for 6% of all enchondromas.

13.2.1.2 Etiologic Factor

The exact etiologic factors of chondroma still remains unknown, like those of most tumors. Enchondroma is the benign hyaloenchondroma that occurs to medullary substance bones, most of them are solitary, and more than one bone or multiple sites of the same bone can be occasionally involved.

13.2.1.3 Pathology

1. Macroscopical findings: Tumor grows inside the bones, making the cortex of bone present bulge-shaped changes and therefore become thin. As the internal periost is invaded by the tumor, its margins take lobulated shape. The volume of short tubular shape osteochondroma is small, and the volume of long tubular shape osteochondroma is large.
2. Microscopic findings: The enchondroma consists of hyaline cartilage lobules, and the cartilage cells gather in the well-formed lacuna. The typical cartilage cells are small sized, the cytoplasm is unclear, the nucleus is small sized and round, and the color is deep. The enchondroma often has calcification areas, the inner cells in the region can

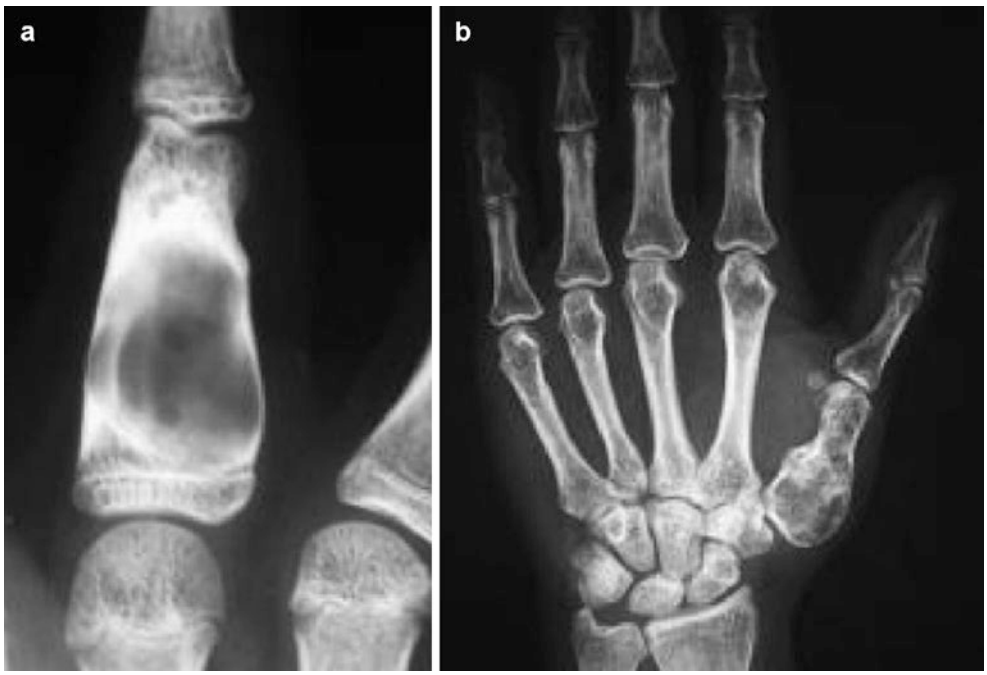

Fig. 13.3 Phalangeal single enchondroma. (**a**) One local and lobulated radiolucent shadow is noted in the ring finger of the right hand, and the endosteum is eroded (anteroposterior position). (**b**) The metacarpal bone of the left hand is bulging and the cortex of bone becomes thin (anteroposterior position)

manifest degenerative changes or necrosis, and there are enlarged and irregular substantial nuclei. The changes in the maturity of cells in enchondroma are great; especially in children and teenagers, a large number of cells are often observed, and atypical cell nuclei and dicaryocyte nuclei increase in number. The above histological features can be used to distinguish between benign enchondroma and chondrosarcoma, but they cannot serve as the evidence for the diagnosis of hand and foot cartilage tumor. As for the single enchondroma, the occurrence of invasion and destruction of the cortex of bone as well as the bulging of tumor into the soft tissues are the signs of canceration.

13.2.1.4 Clinical Manifestation

The age distribution of chondroma patients is wide: patients aged between 5 and 80 may all suffer chondroma, but most of the patients are aged between 10 and 40, and there is no significant difference between males and females in incidence. The typical manifestation of enchondroma of short bones in hands and feet is palpable bulges, sometimes pain. In case of local intense pain, the possibility of canceration should be suspected. As the lesions make the ossicle bulge and the cortex become thin, the patient may pay the first visit to the doctor due to pathological fracture. The tumors of long tumors are usually asymptomatic, and many patients are occasionally found during X-ray photography or bone scan due to other reasons.

As for the phalangeal and metacarpal enchondroma, local lumps can occur due to its superficial position, the surface is smooth, the texture is hard, and sometimes mild tenderness is suffered.

13.2.1.5 X-Ray Examination

The typical X-ray finding of phalangeal single enchondroma is one local, marginally smooth, and lobulated oval-shaped transparent shadow, the tumor is located in the center of the bone, the cortex of bone becomes thin and bulges toward the exteriors, and there is a thin layer of proliferative and sclerotic margins around the invaded inner periost (Fig. 13.3). The X-ray characteristics of metacarpal single enchondroma are similar to those of phalangeal single enchondroma, but as the tumor shadow is large and deviates to the epiphysis, the bulge of the cortex of bone is significant. The long bone diaphyseal single enchondroma presents central or eccentric growth in the marrow cavity with varying sizes, the main symptom is osteolytic change accompanied by dense calcification shadows, and the margins of the cortex of bone present lobulated erosion. The single enchondroma of flat bones or irregular bones has no typical X-ray findings [11].

13.2.1.6 Treatment

The treatment principles of enchondroma are the following: ① surgery may not be performed temporarily for the asymptomatic patients with a small range of lesions, but they should be closely observed on a regular basis; ② if the tumor tends to be enlarged, or the tumor range is wide, the deformities are obvious, the substance of bone becomes thin, and surgery should be performed immediately; ③ for the patients with pathological fracture, the treatment is dependent on specific conditions, fracture should be treated first, surgery can be

performed after the fracture is healed, or the surgery should be immediately performed; ④ for the patients with serious deformities, complete loss of finger functions, and still the tendency of recurrence and canceration after multiple surgeries, amputation can be performed; ⑤ for the patients with benign tumor after postoperative recurrence, another surgery can be performed.

The surgical methods of hand enchondroma include simple curettage, curettage and bone grafting, ostectomy, and amputation.

1. Simple curettage: It is applicable to those with small-sized lesions. The surgery can be performed on the outpatient axillary region under the condition of nerve block anesthesia. After the operation, there can be new bone formation and reconstruction of lesion areas, and generally the patient is free from complications such as bone fracture, infection, and recurrence. The endoscopic simple tumor curettage without grafting is an effective method to treat hand enchondroma.
2. Curettage and bone grafting: It is applicable to most patients. Through phalangeal windowing, the surgery can completely scrape off the diseased tissues; cancellous bone is implanted after the bone cavity is washed. The main insert is autogenous bone, and iliac bone, fibula, distal radius, and olecranon process of the ulna can be taken according to the demands; bone cement can be used for filling, but they should be taken out in case of tissue reactions.

   The key of this surgery is: ① in case of presence of segregation, it should be opened, and the diseased tissues should be completely scraped; ② after curettage, washing or burning is performed and tumor cavity is cleaned to prevent the residue of tumor tissues; ③ the insert should be closely padded without residual lumens.
3. Ostectomy: For the patients with a large lesion range and serious osteoclasia, phalangectomy can be performed on the diseased segments.
4. Amputation: It is applicable to the patients with giant lesions, serious deformity, loss of finger functions, multiple recurrences, or the tendency of canceration.

The therapeutic effects of hand enchondroma are good. The evaluation criteria of postoperative functions include: ① the appearance of the affected finger is normal or close to normal; ② the range of active movements of the affected finger occupies over 80% of the healthy finger; ③ the pinching function is up to over 80% of the normal functions; ④ after bone union, X-ray verifies no shortening, deformity, osteoarthritis, or tumor recurrence. The criteria of efficacy evaluation include: excellent, satisfying four of the above conditions; good, satisfying three of the above conditions; intermediate, satisfying two of the above conditions; and poor, satisfying one of the above conditions.

13.2.2 Multiple Enchondroma

This disease, also called dyschondroplasia or Ollier disease, is a relatively rare kind of dysplasia, its onset has not been proven to be genetically related, and there is no obvious difference between males and females. Generally, Ollier disease refers to the enchondroma with mainly unilateral onset; dyschondroplasia, however, is a developmental defect and does not fall into the category of tumor. This etiology of this disease remains unknown, and the reason may be that the metaphyseal vessels phagocytize the calcified cartilages and further lead to the aggregation of noncalcified cartilages.

The multiple enchondroma complicated by scattered amalgamation and phleboliths in skin and other soft tissues is called Maffucci syndrome, and the patients with this syndrome can concomitantly suffer from internal organ angioma, skin superficial vein dilation, multiple pigmented spots, and leukoderma.

13.2.2.1 Pathology

Multiple enchondroma is generated due to the falsification and proliferation of intra-metaphyseal cartilage cells, and the pathological feature is that globular or bead-like cartilages are contained in multiple intraosteal regions. The distribution and range of lesions are various, some are located in the unilateral or bilateral hand, one limb is involved, most of them occur to the bones of lower limb, and the involvement in the upper limb skeletons is mainly located at one side.

Macroscopical findings indicate that the long tubular bones are short and bent, and the metaphysis is widened. The foci are longitudinally cleaved, multiple circular or orbicular-ovate gray-white regions are observed in the cartilage masses, and bone diaphragm is seen. Histological examination can indicate small cartilage cells and large vacuolar cartilage cells are alternating, and the arrangement is in disorder; they are correlated with single enchondroma in addition to the poor intercellular matric calcification.

13.2.2.2 Clinical Manifestation

The clinical symptoms appear early, and it often occurs to the metacarpus, phalange, upper and lower regions of knee joints, and the distal ends of the ulna and radius. When the lesions involve the hands, the results are enlarged fingers, deformed hands, and damaged functions. The involvement of femur and tibia will produce knee introversion and extroversion. The number and range of lower limb lesions are often asymmetric, often resulting in the unequal length of lower limbs. For a 3-year-old infantile patient, the difference between the two lower limbs is 2–4 m, and the mean difference after bone maturity can be up to 5–15 cm. The pediatric patients with involved lower limb can suffer claudication, and the pediatric patient with involved forearm can suffer bending of upper limb, limitation in pronation, and hand ulnar deviation.

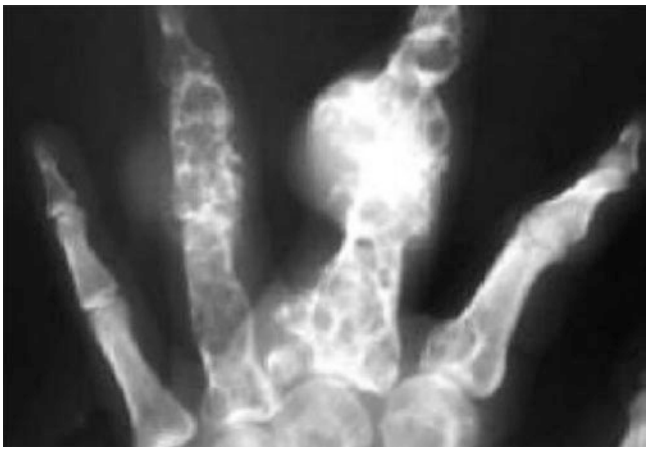

Fig. 13.4 X-ray findings of multiple enchondroma in the left hand

13.2.2.3 X-Ray Examination

X-ray films can show that the short tubular bones of hands and feet are often dilated and take globular shape, the cortex of bone becomes thin and expanded to the surrounding regions, and calcification is often observed (Fig. 13.4); the long tubular bones are manifested as metaphyseal longitudinal lucent streaks and extension into the diaphysis, and the tumor has spot-shaped calcification. The cortex of bone at the tumor site becomes thin, and as a result pathological fracture can occur.

One oval-shaped homogeneous lucent region is visible in the proximal phalange of the index finger, and the range is limited; multiple cystic lucent regions are visible in the middle and ring fingers, the margins are irregular, the cortex of bone becomes thin due to erosion, the surrounding soft tissues are involved, and irregular calcification is observed during the phase.

13.2.2.4 Treatment

As the lesions are multiple, it is difficult to give treatment to each enchondroma. Treatment may not be necessary for asymptomatic patients, but the patient should be closely observed. For symptomatic diseased sites, curettage can first be performed and then skin grafting can be carried out. For obvious limb deformities, osteotomy can be performed for correction. For the treatment method, refer to single enchondroma.

13.2.3 Maffucci Syndrome

Maffucci syndrome is a rare type of congenital nonhereditary mesodermal dysplastic disease characterized by multiple enchondroma and soft tissue hemangioma, and it was first reported in 1881 [12, 13].

13.2.3.1 Clinical Manifestation

The male and female incidences of Maffucci syndrome are the same, most of the patients are children under 10, but some patients may be found to suffer it at birth.

The evidence of diagnosis of Maffucci syndrome includes the following two aspects:

1. Abnormal skeletons: The patients with mild conditions may have no obvious clinical manifestations, and in severe cases, deformities to varying extents can be caused to the limb. About 20% of the patients can suffer malignant changes in cartilages [14]. The hand metacarpal bones and phalanges are the predilection site, those with invaded fingers account for about 5% [12], and long tubular bones can also be involved. The distribution of multiple enchondroma can be limited to unilateral or bilateral upper limbs and unilateral or bilateral lower limbs, and the patients with onset in unilateral limb account for about 50% [15–17]. The lesions can result in unequal length of bilateral limbs or scoliosis.
2. Soft tissue lesions: Most of soft tissue lesions and skeletal lesions occur to the adjacent sites, but they do not necessarily occur to the same site. Most of the soft tissue lesions are cavernous hemangioma or capillary hemangioma, and a minority of the soft tissue lesions are lymphangioma. The soft tissue lesions may have no clinical symptoms, and sometimes, mild local discomfort or elevated skin temperature can be felt. Hemangioma can be located in cutaneous and subcutaneous regions and can also be located in mucosa and internal organs.

13.2.3.2 X-Ray Examination

The typical X-ray findings of Maffucci syndrome are the metaphyseal centric or eccentric lucent regions, anisometric calcification foci are observed, and phleboliths can be seen in the soft tissues (Fig. 13.5). As for the osteodysplasty induced by Maffucci syndrome, it is found in the X-ray film that the bilateral limbs are unequally long and asymmetric, the ulnae are shortened, and the lower ulnoradial joint has semi-location.

Multiple eccentric lucent regions are observed in the substantia ossea, and circular-shaped local calcification foci are visible in the soft tissues, namely, the typical phleboliths.

For the patients aged over 40 or with wide lesions, X-ray shows the erosion and destruction of the cortex of bone, or the disappearance of calcification shadows in the chondroma, or the occurrence of pathological fracture, or the enlargement of the soft tissue masses, and the possibility of canceration should be considered.

13.2.3.3 Treatment

The treatment principle should be the same as that of multiple enchondroma.

13.3 Epiphyseal Abnormalities

13.3.1 Dysplasia Epiphysealis Punctata

Dysplasia epiphysealis punctata, also called congenital calcified cartilage dysplasia or stippled epiphysis, is character-

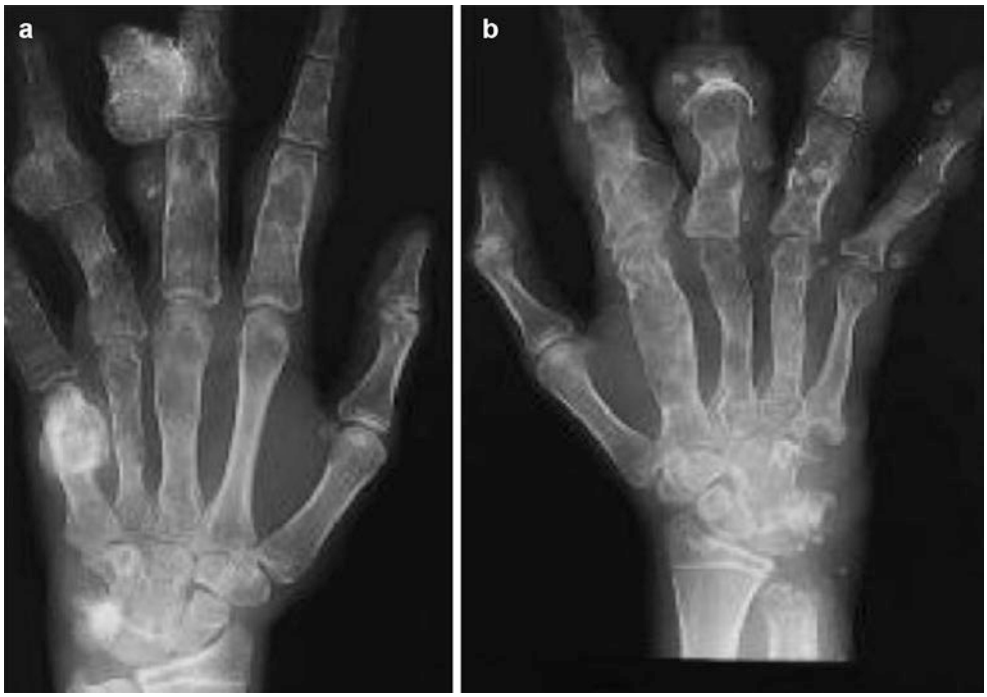

Fig. 13.5 X-ray findings of Maffucci syndrome

ized by poor skeletal growth and punctate calcification of epiphyseal cartilage [18]. The patient often suffers dysplasia of four limbs and joint deformities, and the patient can be accompanied by skin impairment or vascular malformations.

13.3.1.1 Etiologic Factor

The etiologic factors of this disease remain unknown and may be genetically related. Autosomal recessive inheritance can lead to type I of this disease, autosomal dominant inheritance can lead to type II of this disease, and the deformities of the former are more severe than the latter [19]. Sex chromosomal dominant inheritance is manifested as death of male infant and onset of female infant.

13.3.1.2 Pathology

The generalized cartilage primordium has a large number of scatted calcification foci, most of which gather at the site of epiphyseal cartilage and close to the metaphyseal part, mucus-like changes and cystic degenerative changes are distributed between the calcification foci, subsequently the metaphysis is expanded, and the long bones are shortened.

It still remains unclear whether the above pathological changes are induced by vascular dystrophy or induced by cartilage developmental defect. The epiphyseal cartilage primordium displays rich vessels and punctate mucus-like degenerations, inducing cartilage fracture. Each cartilage fragment, with the vessels as the center, constitutes a center of proliferation, calcification, and final ossification, the fissures between the fragments are connected, the epiphyseal vascular circle and the metaphyseal cancellous bone are connected, and the cartilage proliferation and ossification of the epiphyseal plates are not hindered [20].

13.3.1.3 Clinical Manifestation

1. Poor skeletal growth: The onset usually appears before the age of 3, with the manifestations of dysplasia of four limbs, such as micromelia, hyperdactylia, hip, knee, and elbow joint contracture, and hip joint dislocation, and some patients may suffer from cardiovascular deformities.
2. Special appearance: The head is small or large, the forehead is bulging, the eye span is widened, the bridge of the nose collapses, the palate is high or cleft, the neck is short, and the patient has mental retardation.
3. Ocular abnormalities: Cataract, optic atrophy or dysplasia, strabismus, and nystagmus.
4. Skin abnormalities: Scaly keratosis, erythroderma, and shedding of hair.

13.3.1.4 X-Ray Findings

Multiple scattered small circular or oval-shaped, sharply marginated calcification spots are visible in the epiphysis, their diameters are mostly several millimeters, they can be fused into patches, and these calcification spots often appear earlier than the epiphyseal ossification center. These changes are common in long bones, tarsal bones, and carpal bones of the lower limb, and they can also be found in other long tubu-

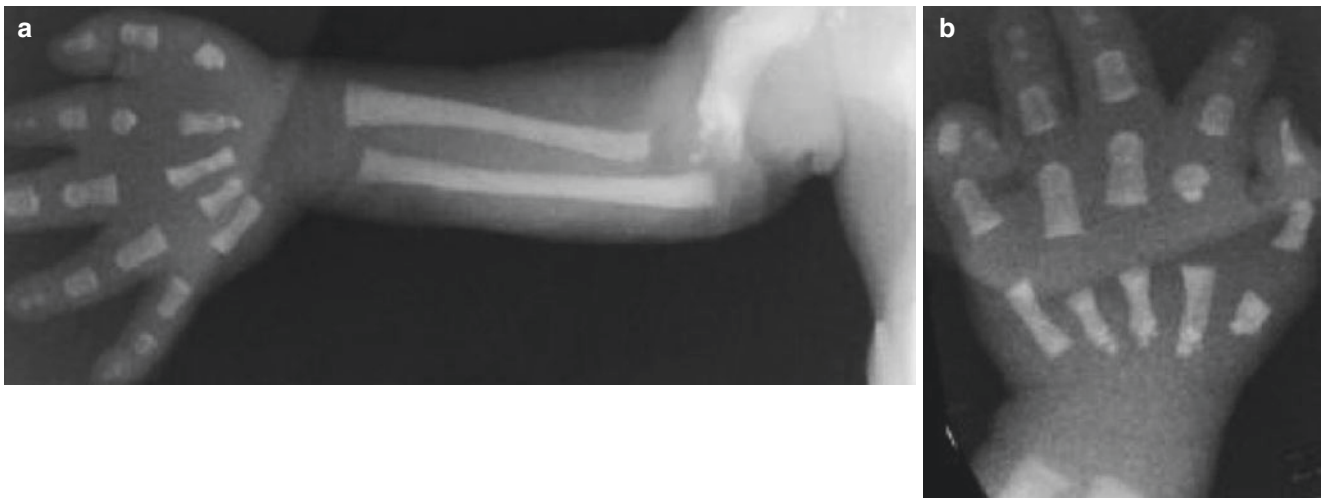

Fig. 13.6 X-ray findings of dysplasia epiphysealis punctata. (**a**) The humeral bone is shortened and bent, and multiple scattered calcification spots are observed in the osteoepiphyseal portion. (**b**) The first, second, and fourth metacarpal bones are shortened and widened, and the second phalange is shortened and widened with punctate calcification

lar bones (Fig. 13.6). The calcification can extend the epiphyseal range and involve the adjacent muscles, ligaments, and synovium of joint, and the cartilages of carpal bones and tarsal bones have no such changes. These calcification spots will not increase with age and generally will disappear in 3–4 years, and subsequently skeletal deformities may appear.

13.3.1.5 Diagnosis

1. Medical history: If the patient has a family history, inquiry on his/her family history will help discover this disease.
2. Symptoms or physical signs: Eye abnormalities such as micromelia, hyperdactylia, multi-joint deformity, special appearance, cataract, optic atrophy and strabismus, keratinization of skin scales, and shedding of hair will indicate this disease.
3. X-ray radiography: The typical X-ray finding during the infancy is the epiphyseal multiple punctate calcification.

13.3.1.6 Differential Diagnosis

1. Multiple epiphyseal dysplasia: In addition to epiphyseal punctate calcification, this disease features pain mainly in the hip and knee joints, no special appearance and cataract.
2. Pituitary dwarfism: This disease is induced by decreased growth hormone secretion, with the manifestations of well-balanced dwarfism with proportional shrinkage of the body without joint contracture, cataract, or skin lesions.
3. Cretinism: The epiphyseal changes of this disease are similar to the dysplasia epiphysealis punctata, but the face and skull are normal; the patient is free from cataract, optic atrophy, or dysplasia, the joint function is not limited, and the symptoms will improve after the administration of thyroxine. The epiphyseal punctate calcification of this disease will appear after the age of 2, the calcification spots are large, and the distribution is not wide. In spite of the late appearance of epiphysis, the shape is normal.
4. Congenital joint contracture: After birth, the patients with this disease can suffer activity limitation, contracture, and deformities of multiple joints, and no calcification spot is observed in the X-ray film at the epiphyseal sites.

13.3.1.7 Treatment

If the general conditions of pediatric patients are good, the patients with unequal length of lower limbs can undergo epiphyseal arrest surgery; the patients with serious deformities can receive osteotomy for correction; for the patients with severe joint contracture but with intact joint surface, soft tissue release surgery, lengthening of tendon, or joint capsule decollement can be performed.

13.3.1.8 Prognosis

Due to weak anti-infectious ability, pediatric patients often die before they become 1 year old or die on the vine; for a minority of patients that pass the childhood, the manifestations can be microsomia as well as shortening and deformities in long bones.

13.3.2 Multiple Epiphyseal Dysplasia

Multiple epiphyseal dysplasia, also called Fairbank disease or dysostosis epiphysealis multiplex, is a rare kind of heredi-

tary epiphyseal dysplastic disease and falls into the category of autosomal dominant inheritance. Characterized by abnormal ossification of multiple epiphyses, it leads to limb growth retardation with the manifestations of thick and short fingers, microsomia, and joint deformities. It is commoner in males than in females, and the onset is mainly observed in infants and teenagers. It often occurs to hip, shoulder, and ankle joints, followed by knee, wrist and elbow joints.

13.3.2.1 Etiologic Factor

Chromosomal hereditary defects lead to epiphyseal dysplasia, late appearance of ossification center, and irregular calcification in the epiphyseal plate, and subsequently the long bone growth and joint development are influenced. The appearance of multiple irregular ossification centers make the epiphysis enlarged and sometimes extended to the diaphysis. During the late stage, the ossification centers are irregularly fused, resulting in non-smoothness of the joint surface and leading to osteoarthritis.

13.3.2.2 Pathology

The pathological changes are irregular epiphysis and epiphyseal plates, lack of osteoid tissues, irregular arrangement of cartilage cells, disorder in bone trabecula, and abnormal ossification in the majority of epiphyses.

13.3.2.3 Clinical Manifestation

At birth, generally no obvious abnormality is observed; after the age of 2, symptoms can be gradually observed. The early chief complaint is pain in the hip, knee, and shoulder joints, leading to limited functions, and the manifestations are difficulty in walking, unstable gait, or swaying gait. The four limbs of the patient become short, the fingers become thick, the patient is undersized, but the facial and cranial development is normal, and the intelligence development is not affected.

13.3.2.4 Imaging Examination

The ossification centers of most epiphyses appear late and are divided into many parts, like many mulberries surrounding one principal ossification center. These ossification centers are finally fused into one, and the epiphyseal line is closed at normal time. The epiphyseal margins are smooth, but become flat after compression. After entry into the adulthood, the patient's joint space becomes narrow, with early occurrence of joint degeneration. Multiple ossification centers make the epiphysis become enlarged, and the tibia, ulna, carpal bone, and metacarpal bone can undergo corresponding epiphyseal changes (Fig. 13.7). With increase in age, epiphyseal changes will gradually disappear, but the deformity of flatness is still present, and in severe cases, secondary degenerative osteoarthritis can occur. The skull and teeth are normal. It can be divided into two types: ① Ribbing type (mild type), with multiple epiphyseal dysplasia, flat epiphysis, and mild invasion of hand substantia ossea, and ② Fairbank type (severe type), with small epiphysis, delayed ossification of irregular carpal bones, and obvious changes in metacarpal bones and phalanges.

13.3.2.5 Diagnosis

1. Medical history: If the patient has a family history, inquiry on his/her family history will help discover this disease.
2. Symptoms and physical signs: The fingers are thick and short, the four limbs are short, the patient is undersized, but the face and skull are normal. The gait is unstable, the knees have introversion and extroversion, the lower limbs are not equally long, and the epiphysis is thick and large; these symptoms and signs will indicate this disease.
3. X-ray examination: It can serve as the main diagnosis basis. The characteristic manifestations of X-ray are irregular ossification and increased density of ossification center; they are spot shaped or flat shaped or mulberry shaped, divided into many small parts, and small and scattered ossification centers are present and surround the large ossification centers.

13.3.2.6 Differential Diagnosis

During the early stage, the disease should be differentiated with Kashin-Beck disease, which is an endemic disease, and there are many patients of the same type around him/her. During the late stage, the disease should be differentiated from the primary proliferative arthritis, which is common in old women who have thick finger joints.

13.3.2.7 Treatment

This disease is self-limiting, but the deformed joint is apt to degenerative joint disease. During the childhood, external fixation is unnecessary, and it is not suitable for the patient to receive surgery. When the conditions are unstable, the weight loading should be reduced, and the patient should be engaged in the occupation with little walking and standing. The treatment principle of adult osteoarthritis should be the same as that of general osteoarthritis. The patients with severe degenerative joint disease can select joint replacement or joint debridement. The patients with limited finger movement can undergo metacarpophalangeal or proximal interphalangeal joint capsule incision and release to improve the finger grasp functions. After the multiple epiphyseal dysplasia is healed, the epiphyseal density can return to normal, but the irregular shape can be partially retained.

13.3.2.8 Prognosis

This disease generally has no influence on the general health, but the involved joint will experience proliferative changes during the late stage.

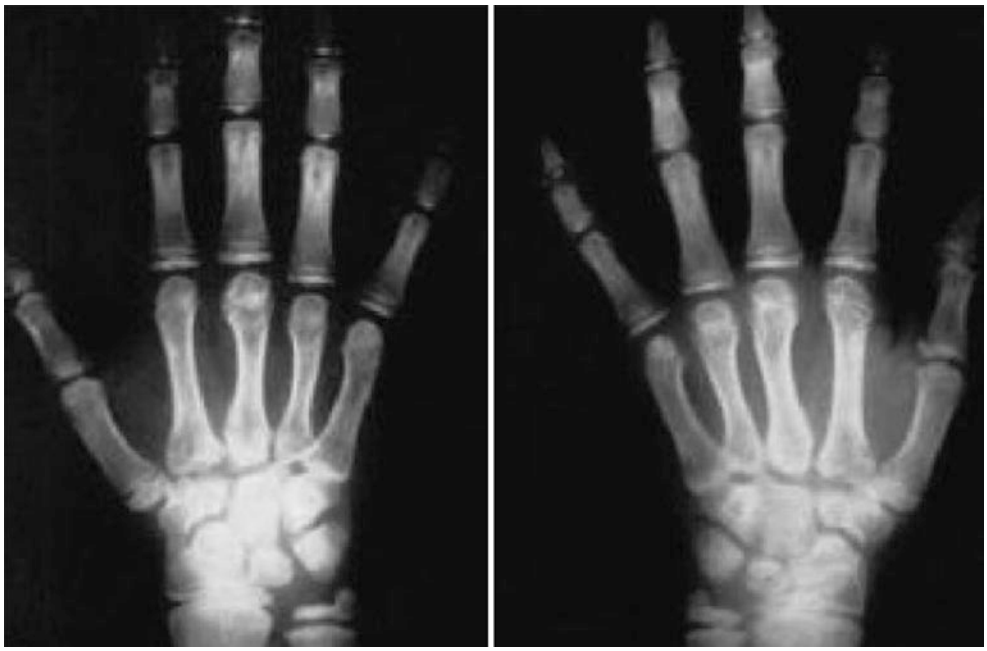

Fig. 13.7 X-ray findings of multiple epiphyseal dysplasia of metacarpal bones and carpal bones

13.3.3 Trevor Disease

Trevor disease, also called dysplasia epiphysealis hemimelica (DEH), is a type of epiphyseal growth deformity, and its feature is that one or multiple epiphyses of the half body present asymmetric excessive growth [21]. This disease is not common in China, male patients with this disease outnumber female patients, and the incidence ratio between males and females is about 3:1.

13.3.3.1 Etiologic Factor

The patients of this disease are sporadic, but it may be correlated with gene mutation and dominant heredity. Some studies reveal that this disease is autosomal hereditary in a family.

13.3.3.2 Pathology

The lump is connected with epiphysis, and there is no obvious border between the two. Similar to the osteochondroma, its surface is covered by a layer of light blue, well-lubricated, and lustrous cartilages. In the middle is the normal cartilage calcification region, and the region near the epiphysis takes the shape of irregular ossification [22].

13.3.3.3 Clinical Manifestation

The symptoms are mainly shown during the childhood from birth to the age of about 8, and the patients often have the chief complaint of the appearance of unilateral painless lump in knee, ankle, or wrist joints. The lump has the osteoid hardness without tenderness, it is connected with epiphysis, the activity is poor, there is no adhesion with the skin, and the skin also has no manifestation of inflammation. Located at one side of the epiphysis, the lump induces the overgrowth at this side of epiphysis, generating deformities of knee introversion and extroversion [23]. The activities of the involved joints are limited, and mild atrophy is observed in the adjacent muscles.

13.3.3.4 X-Ray Examination

During the early stage, one side of the epiphysis becomes enlarged, the appearance is irregular, and a large number of circular or irregular ossification centers are visible in the adjacent soft tissues. The articular surface opposite to the enlarged epiphysis is not smooth, and there is a phenomenon of condensing. Subsequently, these ossification centers will form bony connections with the epiphysis. Finally, most of the lumps will be ossified, and the appearance is similar to that of osteochondroma (Fig. 13.8).

13.3.3.5 Diagnosis and Differential Diagnosis

According to the clinical manifestations and X-ray characteristics, the diagnosis of this diagnosis is not difficult, but it should be differentiated with osteochondroma and chondrosarcoma. The osteochondroma grows in metaphysis instead of epiphysis. Most of the chondrosarcomas grow rapidly, the subcutaneous venous engorgement is present, pain is obvious, no bone texture is observed in calcified mass, and the destruction in bone cortex and the formation of subperiosteal new bones are common.

13.3.3.6 Treatment

The infantile bone molding capabilities are strong, and the early resection of diseased tissues can make the joint return to normal. After entry into adult stage, the patients with a

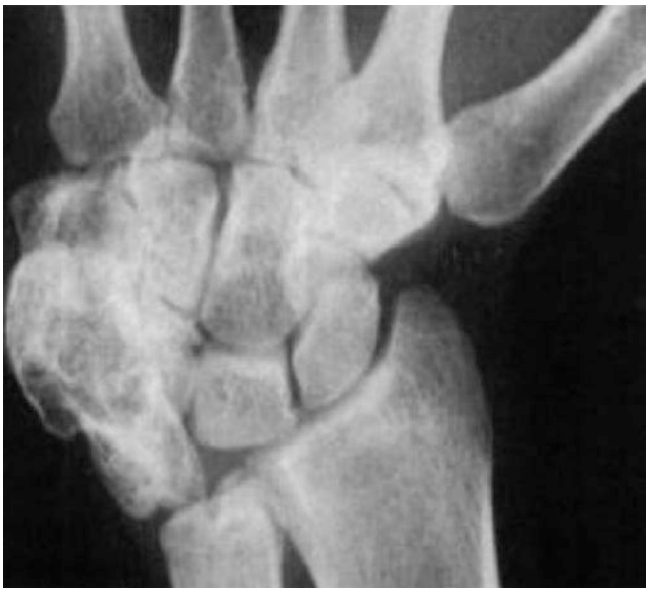

Fig. 13.8 X-ray findings of the left wrist with Trevor disease

relatively small lesion and without joint functional disorder may not necessarily receive any treatment; if the lump influences the patient's functions greatly, it can be completely resected; if the joint deformity is obvious, osteotomy can be performed for correction; if the patients concomitantly have non-smooth articular surface, joint fusion can be performed after lump resection.

13.4 Fibrous and Osteofibrous Dysplasia

13.4.1 Fibrous Dysplasia

Fibrous dysplasia, also called fibrous abnormal proliferation, is a type of benign tumor-like differentiation of fibrous bone tissues, and it is the intramedullary benign fibro-osseous lesions. This disease can be divided into two types, namely, monostotic fibrous dysplasia and polyostotic fibrous dysplasia [24]. The monostotic type often occurs to the ribs, femoral bone, and tibia, the onset age ranges from 10 to 70, and it is common in patients aged between 10 and 30. The polyostotic type is rarer than the monostotic type often with one-sided distribution, its onset age is younger than that of the monostotic, and 2/3 of the patients develop the clinical symptoms before the age of 10. The polyostotic type is often accompanied by unilateral distribution triad characterized by the yellow skin in the trunk or brown chromatosis (cafe-au-lait patch), endocrine disorder, and polyostotic injuries, which is called McCune-Albright syndrome. The reason for the occurrence of McCune-Albright syndrome is that the somatic mutation of C-fos tumor gene in the involved tissues leads to the activation of the signal transduction pathway of cyclic adenosine.

13.4.1.1 Etiologic Factor

As is verified, the activation and mutation of G protein are present in monostotic and polyostotic fibrous dysplasia, and it may be correlated with the onset. The GNAS1 gene that encodes the α-subunit of stimulatory G protein is proven to experience activation and mutation in the monostotic and polyostotic fibrous dysplasia.

Polyostotic fibrous dysplasia is closely correlated with McCune-Albright syndrome, and there is some connection with Mazabraud syndrome (intramuscular myxoma).

13.4.1.2 Pathology

1. Macroscopical findings: The basic manifestations are the presence of gray-white-like tough tissues, with internally scattered fine trabecula, which induces a gravel sensation in the cross section. The cortex of bone swells and becomes thin. Cysts with varying sizes may be formed inside the lesions [25].
2. Microscopic findings: A large quantity of irregular mixed woven bones are produced in the intercellular substances, the fibrous matrix presents turbine-shaped or lamellar arrangement, and they are rich in vessels inside. There is no osteoblast segregation in the irregular osteoid components, and there are immature bone islands without histological regularity formed inside the membranes instead of the lamellar structure in the benign foci of fibrous structures.

13.4.1.3 Clinical Manifestation

The fibrous dysplasia often has no subjective symptoms, and it is intentionally found during X-ray examinations in most cases. Pathological fractures often occur to the foci that bear the load of bones, and delayed union and disunion are often refractory complications.

13.4.1.4 Imaging Examination

The typical X-ray findings of fibrous dysplasia are as follows: the long tubular bones present ground glass-like changes, the stromal layers present lamellar changes, and the structures of the tissues around the lesions are normal. The foci of long tubular bones are located in the intramedullary regions with the benign tumor manifestations of sharp margins, map-like bone destruction, and some sclerotic margins and are characterized by transparence, sclerotic margins, and distention of the cortex of bone. The repeated fracture and union of the proximal end of the femoral bone form the typical "shepherd crook sign." Generally the patient has no periosteal reaction or soft tissue infiltration, unless fracture occurs (Fig. 13.9). ECT is of significance in determination of single or multiple lesions and CT and MRI can further ascertain the range of lesions.

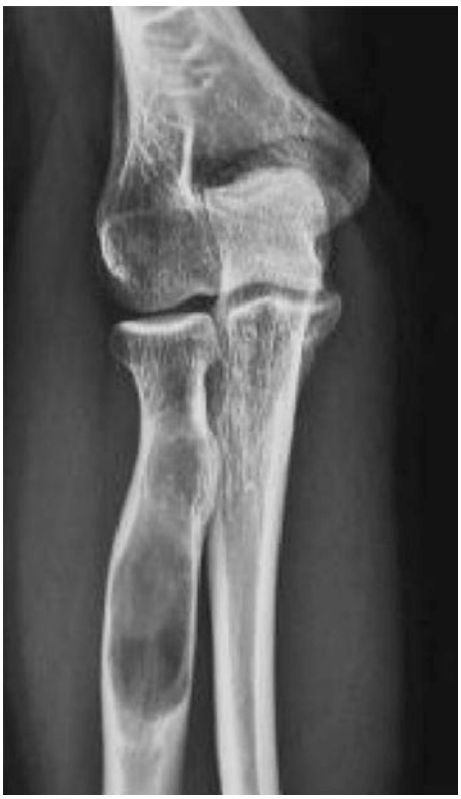

Fig. 13.9 X-ray findings of radial osteofibrous dysplasia

13.4.1.5 Treatment

Palliative surgery is the only option for the treatment of fibrous dysplasia, and focal curettage, bone grafting, osteotomy, and internal fixation can produce some effects.

If the patients are asymptomatic or have no risk of fracture, surgery may not be necessary, and the patients with pathological fracture are treated as general fracture due to the capability of self-union. During the childhood, most of the implanted bones can be absorbed due to the easy recurrence and frequent expansion, and the surgery should be performed with caution and should be limited to osteotomy and internal fixation to correct deformities; the probability of success and failure during the puberty is equal; most of the surgeries can be a success after the patients become adults. When the lesions are widespread, the resection of diseased tissues can be very difficult. After the curettage of long bone lesions, the transplantation of cancellous bones and cortical bones has no advantage compared with single osteotomy because the implanted bones will be absorbed in the foci. The implanted bones and adjacent bones can be invaded by the dysplastic tissues, which is induced by the complete expansion of the incomplete total resection.

13.4.1.6 Prognosis

The prognosis of fibrous dysplasia depends on the range and degree of initial skeletal lesions and the degree of ectosteal lesions. Generally, mono-osseous lesions will not be converted into poly-osseous lesions. The size and quantity of most osseous lesions will not increase compared with the early onset stage. Mono-osseous or poly-osseous lesions will be static during the puberty, but continue to exist. Some deformities can further evolve, especially for those with wide lesions during the early stage, the patients can suffer fracture and serious deformities, and the prognosis is poor. In the beginning, the patients with local lesions evolve slowly, the prognosis is good, and this is irrelevant to the age at onset. During the pregnancy and the estrin treatment, the fibrous dysplasia lesions may be activated.

13.4.2 Osteofibrous Dysplasia

Osteofibrous dysplasia (OFD), also called Kempson-Campanacci lesion and cortical fibrous dysplasia, is a benign self-limiting fibro-osseous lesion and specifically involves the cortex before the middle segment of infants' and children's tibia [26].

13.4.2.1 Etiologic Factor

It is very difficult to distinguish between OFD-like adamantinoma and osteofibrous dysplasia, and it indicated that there is some inevitable connection between OFD and adamantinoma. It has been verified that OFD has mutation in various chromosomes, especially the trisomy of no. 7 and no. 8, and fos and jun proto-oncogene products, and the feature is no mutation in signal transduction G protein α-subunit that induces the increase of cAMP.

13.4.2.2 Pathology

1. Macroscopical findings: OFD is a solid lesion, and the foci take on the color of gray-white, gray-yellow, or red, and it is soft with a gravel sensation. The periost is intact, but the cortex of bone becomes thin or disappears. There are defined sclerotic marginal borders between the lesions and medulla [27].
2. Microscopic findings: Irregular mixed woven bones are visible, the margins consist of lamellar bones and sharply marginated osteoclasts, and osteoclasts may also exist. Fibrous components consist of spindle cells and the collagenous fibers they generate, and the degree of matrix can be mucus-like to moderate fibrosis. The lesions present girdle-shaped distribution, the central parts are fine bone trabecula or components with fibrous tissues as the majority, the surrounding regions mainly consist of lamellar bones, and the posterior regions are mutually fused with the surrounding normal osseous tissues.

13.4.2.3 Clinical Manifestation

OFD is common in males under 20, particularly common in males under 10, and most patients with it are attacked by it before they are 5 years old. The typical pathogenic site is the tibia. The proximal end and the middle segment are the commonest, and sometimes the ipsilateral fibula can also be involved, and other pathogenic sites include ulnar and radial bones. The typical pathogenic site of long bone osteofibrous dysplasia is the diaphysis, and the metaphysis is rarely invaded.

The commonest symptom is swelling or painless deformation and abnormal curvature involving the osseous segments. Sometimes pathological fracture (often the complete bone fracture) can occur, and sometimes pain and a small amount of displacement can be observed.

13.4.2.4 Imaging Examination

The X-ray findings are the eccentric osteolytic lesions in the cortex of bone, the focal borders are defined, and more or less swelling is observed in the surrounding cortex of bone. The swelling cortex of bone is often surrounded by one clear bone sclerosis line, and the osteolytic lesion becomes narrow. The osteolytic foci are single or multiple, and the involvement in the full circumference is very rare. The supratibial foci are multiple, the lesions that invade the entire backbone are rare, and sometimes the polynesic or giant confluent lesions which are arranged longitudinally along the cortex are visible (Fig. 13.10).

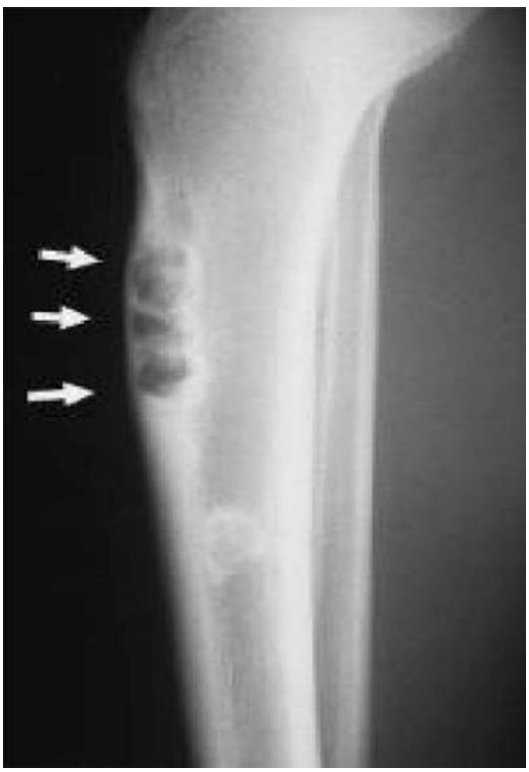

Fig. 13.10 X-ray findings of tibial osteofibrous dysplasia

13.4.2.5 Treatment

The selection of treatment methods largely depends on the age of the infantile patient. The younger the surgical age is, the higher the recurrence rate is; the recurrence rate of patients under 5 is almost 100%; there is almost no recurrence in patients aged more than 15. In addition, the patients with osteofibrous dysplasia in long bones have the tendency of self-union before they are 5 years old, so surgical treatment should not be performed before this age. For the patients aged between 5 and 10, whether the surgery should be performed depends on the specific circumstances; for the patients with expanded lesion range or decreased bone strength, surgery can be considered, the surgical treatment mainly aims at expansion and curettage, and the curetted substances need to be pathologically examined to rule out the possibility of adamantinoma; for the patients with formed false joints, tensile internal fixation can be adopted, and the bone grafts with a high strength can be adopted. As for the patients with serious abnormal curvature in long bones, osteotomy can be performed for correction after the patients become 10 and 12 years old.

13.4.2.6 Prognosis

The natural course of this disease is long, the progression is slow, and the prognosis is good. Some scholars believe that this disease is induced by dysostosis, and it is not the real tumor; after the arrest of growth and development, the lesions will become static subsequently. OFD generally grows gradually before the age of 10; at the age of about 15, it gradually subsides and becomes rehabilitated. In a minority of patients, OFD-like adamantinoma will evolve into typical adamantinoma.

13.5 Fibrous Tissue Tumors

13.5.1 Fibroma

Fibroma is the benign tumor consisting of mature fibrous connective tissues, the tissue source is the mesenchymal and parenchymal tissue, and it is common in body surface and uncommon in hands. The patient with one symptom suffers from fibroma, and the patients with multiple symptoms suffer from fibromatosis. The etiological factors remain unclear.

13.5.1.1 Pathology

1. Macroscopical findings: The fibroma is the circular or elliptic hard and simple benign nodule or lump with complete envelope, and it can be easily separated from normal tissues. There is no reaction zone between the tumor and its surrounding tissues; after the tumor is incised, white spiral patterns are invisible, and the medial surface of the tumor resembles the shape of cicatricial tissues.

2. Microscopic findings: Fibroma consists of irregular and collaterally arranged mature fibrocytes and fibers, the internal newly emerging vessels are rare, and the adipose tissues are occasionally visible. Some tumors have mucous degenerative foci inside, and the infiltration of phagocytes is observed around. The desmocyte has little cytoplasm, the color is reddish, the nuclei are large and date shaped, and the nucleoli and nuclear membrane are unclear. The size of fibrocyte nuclei is small and the cells are spindle shaped. The fibers are mainly the collagenous fibers and distributed in the extracellular regions, and the arrangement is parallel, intricate, or vortex shaped. There are few lattice fibers and elastic fibers.

13.5.1.2 Clinical Manifestation

Fibroma often occurs to the sites of skin, subcutaneous regions, and nail bed and also to sites such as myolemma, tendon sheath, joint capsule, ligament, and periost, the growth is slow, and generally there is no subjective symptom. In case of occurrence to tendon sheaths or muscle tendons, snapping finger can occur; in case of occurrence to the subungual region, the nail deformation and phalangette compression atrophy can occur. The palm part in the teenagers can have hard and ill-defined lumps, inducing flexion contracture of the finger, which is called calcifying aponeurotic fibroma. The surface of the dorsal or lateral fibroma of the infantile fingers or toe is smooth and hard without activity or pain, and this tumor is called infantile digital fibroma.

13.5.1.3 Staging

Most fibromas have been in the phase I of the sneaking status when detected, the tumor can develop to phase II, but the tumors are within the envelope. X-ray plain films indicate the presence of single lump, which shares the same density with muscles. Radioisotope scanning indicates no increase in absorption. The arteriography indicates that the surrounding regions of the tumor are the normal vascular shadows; unless the tumor is close to the great vessels, generally there is no induced increased in new vessel. The tumor that becomes protruding into the fatty tissues can be displayed in CT.

13.5.1.4 Treatment

The main treatment is surgical resection with skin grafting. Rare recurrence is observed in the extracapsular margins, and there is an easy tendency after intracapsular resection. Generally, the size of the recurrent tumor does not exceed that of the original tumor, but in cicatricial tissues, it is difficult to distinguish between the margins and cicatricial tissues of the recurrent tumors, making another resection very difficult, so a layer of healthy tissues around the tumor should be resected during the first surgery for the purpose of reducing recurrence. The juvenile adolescence has the tendency of shrinking with the growth of fingers, and the local use of hormone ointment can diminish the infantile digital fibroma, so the juvenile or pediatric patients can be closely observed when the lump is small sized or at the static status.

13.5.2 Calcifying Aponeurotic Fibroma

Calcifying aponeurotic fibroma is a rare type of benign soft tissue tumor. In 1953, Keasbey first described the calcifying aponeurotic fibroma as juvenile aponeurotic fibroma; because he got to know that this disease could occur to the patients with a wide range of onset age, he called it aponeurotic fibroma [28–30]. Calcifying aponeurotic fibroma often occurs to the patients aged between 10 and 20. Although the onset age ranges from birth to 64 as reported by literature, the median age of the patients with confirmed diagnosis is 12. Since the first report, over 150 cases have been reported.

13.5.2.1 Pathogenic Site

The typical pathogenic sites of such a tumor are the finger, palm, and sole, the commonest site is the palm (67%) and sole, and there is rare report on its occurrence to the back, knee, forearm, elbow, and skull and face. This disease is common in children and adolescents and uncommon in adults; it often occurs to males, and the ratio of incidence of males and females is about 2:1.

13.5.2.2 Pathology

The histopathological examination finds that the calcifying aponeurotic fibroma consists of hypertrophic fibrocytes, and the nuclei are round or orbicular-ovate. The tumor cells have the tendency of growing toward the surrounding fatty tissues and muscles. The tumor cells form the fence-like structures by surrounding the scattered calcification spots, and cartilaginoid tissues are occasionally found. Although the cells are dense, the mitosis activity is insignificant.

13.5.2.3 Clinical Manifestation

Calcifying aponeurotic fibroma is a local benign tumor, whose histological structures are similar to those on the aponeurosis and skeletons. It has the fibrous-cartilaginous appearance, and the cartilaginoid cells participate in the calcification of the foci [31]. The characteristic manifestations of calcifying aponeurotic fibroma are painless and fixed lump with a slow growth, the diameter is less than 3 cm, and there is no adhesion with the skin. There are reports on some tumors which are fine and long shaped with a longitudinal diameter of about 5 cm.

During the clinical examination, lumps are painless and palpable with less-defined margins, a hard texture, and slow growth. After development for a period of time, it can become defined small nodules with a harder texture [32]. It

tends to be at the standstill status at the end of the body growth, and important local disorders such as limited joint activities are never induced.

13.5.2.4 Imaging Examination

X-ray examinations show that the tumors often have punctate calcification to varying degrees, and nonspecific soft tissue lump shadows can be observed (Fig. 13.11). Morii et al. find that, on the T1-weighted images of MRI, the calcifying aponeurotic fibroma is displayed as equal signals or low strength; on the T2-weighted images, the manifestation is the strength of multiphase high signals and relatively low-strength equal-signal areas in them. Calcifying aponeurotic fibroma and giant cell tumor of tendon sheath often grow near the fascia and tendons; however, after MRI contrast enhancement, it can be found that the giant cell tumors of tendon sheath are lobulated, local, and homogeneously enhanced lumps; the calcifying aponeurotic fibromas are the ill-defined heterogeneous lumps with punctate calcification [33].

13.5.2.5 Treatment and Prognosis

As the foci are infiltrative, the surgery is often performed inside the foci, and they cannot be completely resected, so they are apt to recur after operation. As the growth tends to be at a standstill at the end of body growth, surgical resection can be proper and limited. The surgery should be performed when the tumor tends to be mature and enters the static stage. At this time, the recurrence rate obviously decreases.

The local recurrence rate of calcifying aponeurotic fibroma is over 50%; this is because the tumor has the feature of growing by infiltrating into the surrounding tissues, but there are reported cases without postoperative recurrence for over 6 years. According to the description of some scholars, the calcifying aponeurotic fibroma has dual-directional characteristics: during the early stage, the tumor presents infiltrative and destructive growth, but there is no calcification; during the late stage, the tumor cells are dense and present nodular growth with local margins and scattered calcification spots. With the increase in age, the cell activity decreases and the collagen matrix increases, which may cause the tumor to become mature. If the tumor is resected during the mature stage of tumor cells, the local recurrence rate is relatively low even if the tumor resection is not complete. The possibility of canceration of calcifying aponeurotic fibroma is low; in case of local recurrence, local resection can be performed after clear histopathological examinations.

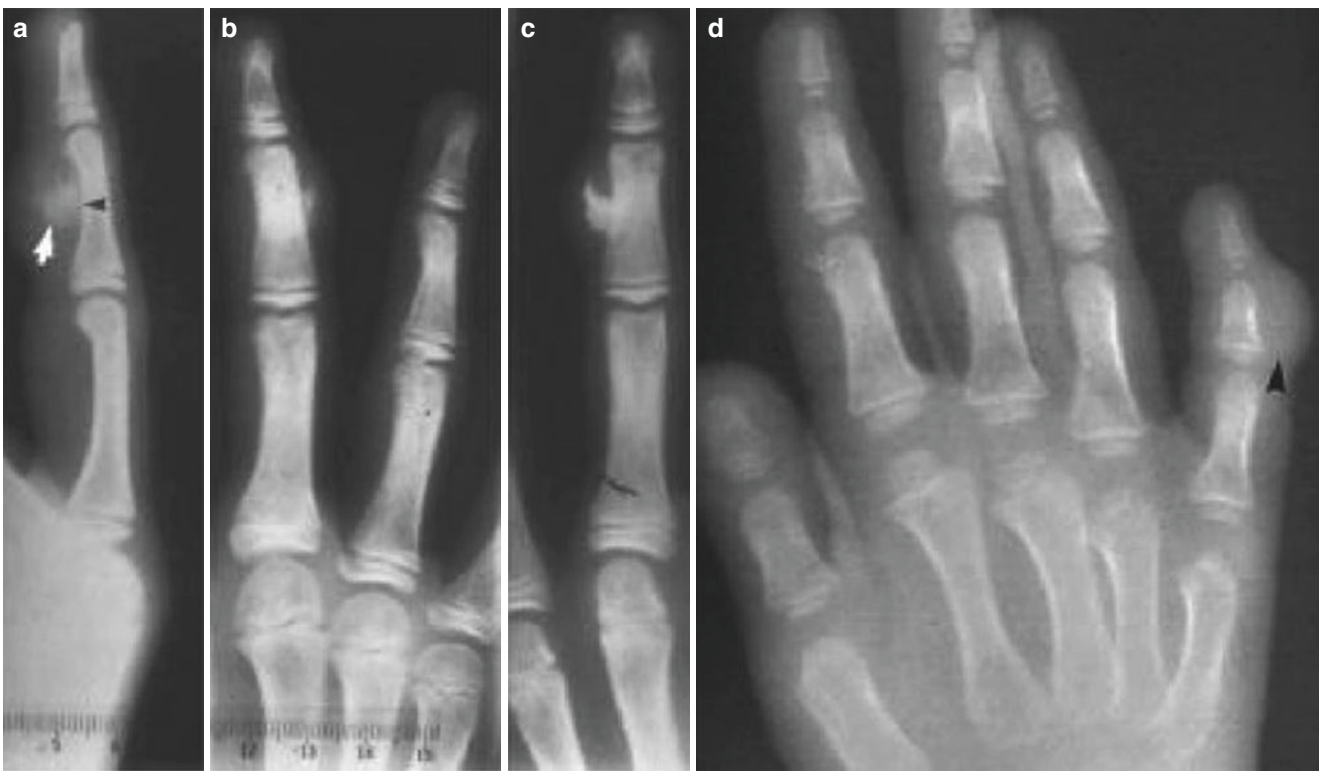

Fig. 13.11 X-ray findings of calcifying aponeurotic fibroma. (**a**–**c**) Calcifying soft tissue shadows are visible (*white arrows*), and mild phalangeal erosion is observed (*black arrows*). (**d**) Soft tissue swelling shadows are observed in the little fingers without bone erosion (*black arrows*)

13.5.3 Infantile Fibrous Hamartoma

Hamartoma is the abnormal proliferation of tissue that occurs during embryonic development or subsequent physical development, and this kind of congenital abnormal proliferation falls into the category of histological structural defects. Hamartoma can grow independently, and its histological structures are of no functional significance; this is because it is derived from embryonic tissues and the regular or functional activities of the human body will not be influenced. The hamartoma with similar growth to tumors has the same proliferative changes; during the adulthood, it tends to arrest growth and becomes completely mature. Hamartoma is a tumorous deformity, it is the abnormal mixture of normal organ components, and the so-called abnormality is the change in the relative content, arrangement, or mixture degree.

Fibrous hamartoma of infancy (FHI) is a kind of benign lesions which fall into the category of dysontogenesis or hamartoma. In 1956, Reye first reported this disease and considered that it was a kind of infantile intradermal fibromatosis tumor [34]. It mainly occurs to the infants with characteristic organ-like microscopic manifestations, including three components: fibroblasts, juvenile mesenchymal cell, and mature fat cells [35]. The ultrastructural display shows the content of a large number of myofibroblasts, so scholars suggest that it be named "infantile subcutaneous myofibroblastoma."

13.5.3.1 Pathology

1. Macroscopical findings: The lumps are round or irregularly shaped with obscure boundaries and located in dermis or subcutaneous regions; it is mixed with the surrounding subcutaneous adipose tissues. Their diameters range from 3 to 5 cm, the largest diameter of the individual lump can be over 10 cm, and there is also the report of those with a diameter of up to 12 cm. Solid, gray-white, and bright, it is intermingled with some island-shaped yellow fatty tissues, sometimes there are many fatty tissue components, and gray-white cords traverse them irregularly.
2. Microscopic findings: The lump structures are special and the obscure and irregular organ-like structures are formed: ① crisscrossed fiber bundles, which consist of spindle fibrocytes and collagenous fibers; ② the small nests consisting of primitive mesenchymal cells, which have a small volume; take circular, orbicular-ovate, or astroid shape; contain mucus-like matrix; and are loosely arranged into vortex-shaped or globular islands with little cytoplasm and deeply stained nuclei; and ③ immature anisometric mature fibrocytes, which can not only be located around the lesions but also occupy the main part of tumor.

The above three components are mutually mixed, the proportion varies, there is a defined border mutually, but there is a phenomenon of mutual transition between the fibrous tissues and primitive mesenchymal-like islands. In addition, diffuse fibrosis to varying degrees can occur, which means that the juvenile mesenchymal cell islands and adipocyte can be replaced by diffuse collagenous fibers and scattered fibroblasts.

13.5.3.2 Clinical Manifestation

This disease is common in males and the incidence of males is about twice that of females. The onset age ranges from birth to 4 with a mean of 10 months, the period within the age of 2 is the crest time, and 15–20% of the patients are found immediately at birth. The lesions mainly occur to the armpit, followed by the upper arm, thigh, inguen, suprapubic regions, shoulder and back, and forearm; according to the report, they can occur to the foot, scalp, perianal regions, and scrotum. The lump is dermic or subcutaneous, the volume is generally small, the texture varies with the proportion of adipose tissues or fibrous tissues, it is movable, but some lesions are fixed due to the adhesion with the fascia or the muscles below them [36, 37].

13.5.3.3 Differential Diagnosis

As the structures and components of the tumor are special, diagnosis is generally not difficult, but when the myofibroblasts are the main component, attention should be paid to differentiate with the following tumors:

1. Infantile fibroma: It can be located in the subcutaneous regions and is rich in collagen, but it often occurs to the muscular fascia, and there are no organ-like structures of fibrous hamartoma.
2. Diffuse myofibroma: It can be the typical single or multiple nodules and can be differentiated according to the nodular presentation due to the division up by hemangiopericytoma-like vascular regions.
3. Calcifying aponeurotic fibroma: It also has fiber bundles and rich collagen elements, little or no calcification is observed during the early stage of growth, and it can be easily confused with FHI. However, calcifying aponeurotic fibroma often occurs to children or young people, and the tumor is often located at the palm or wrist, which is an important diagnostic characteristic.
4. Embryonal rhabdomyosarcoma: The cells with characteristic organ-like structures and immature cells should be differentiated from the embryonal rhabdomyosarcoma to avoid mistreatment caused by the misdiagnosis of this kind of benign fibrous proliferative disease as malignant. In some infants, the fibrous hamartoma can occur to the scrotum, but the spindle cell components are easily misdiagnosed as the juvenile parts of embryonal rhabdomyo-

sarcoma, but this type of lesions often occur to the older children, and the juvenile parts can be clearly differentiated due to the presence of obvious cell specificity and nuclear mitotic figure and the presentation of invasive growth.

13.5.3.4 Treatment and Prognosis

Generally local resection is selected to treat infantile fibrous hamartoma; due to the incomplete resection of tumor, about 16% of the patients have recurrence, and the delay of surgery will not increase the risk of surgical complications. The infantile fibrous hamartoma is manifested as the benign clinical process, and the prognosis is good.

13.6 Neurogenic Tumor

Jinghong Xu and Jialiang Chen

13.6.1 Fibroneuroma

The generally so-called fibroneuroma, also called solitary neurofibroma, as the name suggests, refers to the locally single fibroneuroma without the manifestation of neurofibromatosis. Because the diagnosis of type I neurofibromatosis cannot be ruled out in patients with the occurrence of local fibroneuroma during the childhood and the patients without family history, it is very difficult to accurately obtain the incidence; however, solitary neurofibroma significantly outnumbers the fibroneuroma. As early as 1935, after carrying out a series of relevant studies, Geschikter found that about 90% of the neurofibromatosis was single, and the remaining falls into the category of type I neurofibromatosis; therefore, the only dependence on the solitary neurofibromatosis foci cannot obviously support the diagnosis of neurofibromatosis.

As the chromocytes and Schwann cells are both derived from the nervous crest, the fibroneuroma and neurofibromatosis have the manifestations of pigment anomaly. Histologically, these chromocytes and the cutaneous chromocytes have the same characteristics in staining and ultrastructures.

The incidence of solitary neurofibroma for males and females are the same, and it often occurs to the patients aged between 20 and 30. Most of the fibroneuromas are distributed in the superficial sites of dermic or subcutaneous regions, the chances of distribution in all body sites are equal, such as the sites of the head and face and four limbs and trunk, and the frequent manifestation is a painless nodule or lump with a slow growth; when the lump becomes enlarged to a certain degree, its looseness and softness often leads to the displacement of dropping of local and adjacent organs, resulting in obvious deformities and also some symptoms and functional disorder. When the fibroneuroma occurs to the scalp, the manifestations are very loose and soft masses in the local regions; when it occurs to the face, it often expands from unilateral frontotemporal region to the lower region; in case of involvement of the upper eyelid, the visual lines can be blocked due to the excessive hypertrophy and dropping of the upper eyelids; in case of involvement of the middle part of the face, the nose and oral lips can be displaced downward thereby. The fibroneuroma in the trunk is often located in sites such as the back, lumbus, and hips, the number is big, and the patients feel like bearing a load.

The solitary neurofibroma takes on the gray-white color on the gross specimens, and the sections are smooth and bright, except that gel-like substances are present outside of the dense brittle and tender tumor tissues; many vascular sinus cavities with varying sizes and sparse cellular tissues can be found inside some tumors, the blood supply is abundant, no function of contraction is observed in sinus cavity walls, the control is difficult in case of bleeding, and there is no manifestation of secondary degenerative lesions in the neurilemoma. The fibroneuroma that mainly occurs to the nerve trunk present fusiform intumescence, and normal nerves get in and out of the lumps. When epineurium is present outside the fibroneuroma, obvious capsular sacs can be formed, but the fibroneuroma is often derived from large nervous ramifications and apt to grow into the soft tissues, the range of involvement is limited, and no envelope is formed.

The histological manifestations of fibroneuroma vary with the different quantities of contained cells, mucoprotein and collagen. The most distinctive manifestation of fibroneuroma is that the nuclei are wave shaped, the deeply stained fine long cells are interwoven into bundles, these cells are closely arranged with the collagen, a small quantity of mucus-like substances are visible, and hypertrophic cells, lymphocytes, and a very small number of yellow tumor cells are occasionally seen in the matrix of the tumor foci. In addition, some fibroneuromas have no mucus-like substances, which are Schwann cells and homogeneous collagen cells, and the intra-tumor cells are arranged into cord or vortex shape. A large number of mucus-like substances can be seen in the rarest fibroneuroma, which is apt to be confused with the myxoma, and these fibroneuromas often occur to the limbs.

The blood supply of fibroneuroma is very rich, and characteristic differentiated substances such as Wagner-Meissner corpuscles can be found. S-100 protein can be separated from the fibroneuroma, but the content is not as high as neurilemoma.

13.6.2 Neurofibromatosis

Neurofibromatosis (NF), also called Von Recklinghausen disease, was named after the person, Von Recklinghausen, who first described this disease in 1882. It is currently believed that it is a kind of autosomal dominant hereditary disease with multiple pathological lesions in bones, soft tissues, nerve system, and skin tissues. Generally, it is found soon after the birth, the progression of disease course is slow, and the development during the puberty or pregnancy can be very rapid. According to the clinical and genetic differences, neurofibromatosis can be divided into two types: type I neurofibromatosis (NF1), previously called peripheral neurofibromatosis, and type II neurofibromatosis (NF2), also called bilateral acoustic neurofibromatosis. The clinically common type is type I neurofibromatosis, and the rare type is type II neurofibromatosis.

13.6.2.1 Etiologic Factors and Pathogenesis

Type I neurofibromatosis (NF1) is a common heredity disease, and its incidence among the population is 1/3000 to 1/2500 [38]. Type I neurofibromatosis falls into the category of autosomal dominant hereditary disease with a high penetrance, and half of the patients have a family history. The onset of type I neurofibromatosis is associated with the deletion, insertion, and mutation of type I neurofibromatosis. It has been clear that this gene is an anti-oncogene located in the long arm (17q11.2) of no. 17 chromosome with a length of 300 kb, and it encodes the neurofibromin that acts on the microtubule system. Currently, the functions of neurofibromin have not been fully known, but it has been known that it has a significant homology with the activating protein of GTP enzyme of RAS [39–41]. Under the normal circumstances, neurofibromin regulates cell proliferation through the mutual reaction with the RAS protein, and the mutated neurofibromin loses this type of regulating function, which leads to improper cell growth and tumor formation and further induces various manifestations of neurofibromatosis.

Type II neurofibromatosis (NF2) is a kind of autosomal dominant hereditary disease with a high penetrance (95%), and its incidence is about 1/50,000. The gene of type II neurofibromatosis is located in no. 22 chromosome and encodes the Merlin protein, and it has a homology with one type of protein moesin-ezrin-radixin that promotes the bind of cell membrane with intracellular matrix. This disease has many gene mutation types, but there is no clear mutational hot spot. The studies carried out during the recent years have indicated that the protein product Merlin has tumor-inhibiting functions.

13.6.2.2 Pathology

The histological structural characteristic of neurofibromatosis is no presence of connective tissue envelope; it consists of wave-shaped fibrillae, which are sparse and arranged into bundles and take vortex shape or helical shape; there are many spindle- or oval-shaped nuclei between them, the size is homogeneous, the color is light, no elastic fiber is observed, mucus-like degeneration can be observed in some, and the nuclei are embedded into the homogeneous light blue matrix [42–44].

13.6.2.3 Clinical Manifestation

Type I neurofibromatosis has many characteristic clinical symptoms and physical signs, with the main manifestations of nervous system tumor, skin cafe-au-lait macule, and abnormal skeletal development.

Type II neurofibromatosis often occurs during the puberty or slightly later, the disease course is long, the duration lasts for as long as several years from the onset to the treatment, and the clinical manifestations include tinnitus, loss of hearing, nystagmus, dizziness, and vertigo. Most acoustic neuromas occur to the vestibular canals of the acoustic nerves, the bilateral occurrence is basically the local manifestations of type II neurofibromatosis, and the tumor is circular with a slow growth, complete envelope, and few adhesions with peripheral tissues. There is also the presence of cafe-au-lait macule, which is rarer than the type I neurofibromatosis. The patients can also be easily accompanied by other cerebral neurilemoma.

13.6.2.4 Diagnosis and Auxiliary Diagnosis

For the diagnosis criteria of neurofibromatosis, please refer to Table 13.1.

CT or MRI can clearly display the site and source of the tumor, and MRI can especially provide accurate diagnosis basis. In vivo histopathological examination can help rapidly confirm the diagnosis.

13.6.2.5 Treatment

Due to a large number of foci, the scattered distribution, and the frequent involvement of deep tissues, surgical resection cannot be relied on to clear all the foci of neurofibromatosis, and the surgical treatment is mainly targeted at the symptoms of tumors with a large volume and that can induce pain or the tumors with a tendency to induce functional disorder. However, these foci often have a large volume without clear borders and envelope but with abundant blood supplies; in the meantime, whether the normal tissues are maintained or removed should be considered, so reasonable estimation and necessary preparations should be made before the surgical resection. Before the surgery, ultrasound examination can be made to learn the distribution of great vessels and blood sinuses and roughly estimate the possible bleeding; for a minority of especially large foci, even angiography can be performed to learn the conditions of the communicating vessels; if conditions permit, attempts can be made before opera-

Table 13.1 Diagnosis criteria of neurofibromatosis

Type I neurofibromatosis (meeting over two of the following conditions)	Type II neurofibromatosis
More than six cafe-au-lait macules (> 5 mm before puberty and >15 mm after puberty)	Bilateral vestibular neurilemoma
Two or more neurofibromas of any type or one plexiform neurofibroma	There is some connection with NF2, with unilateral eight cranial nerve tumors or one of the following items:
Freckle-like pigmented spots in armpit or inguinal region	Cutaneous or subcutaneous acoustic neuroma
Optic neurogliocytoma	Plexiform neurofibroma
Characteristic bony lesions (sphenoid osteodysplasty, formation of tibial pseudarthrosis, and thin cortex of long bone)	Neurogliocytoma
Patients with neurofibromatosis in one generation of blood relation (parents, siblings, and children) have received regular standard diagnosis	Juvenile posterior subcapsular cataract
Two or more iris hamartomas (Lisch nodules)	

tion to adopt other measures such as transcatheter embolization or electrochemical treatment to reduce intraoperative bleeding; in the meantime, sufficient blood should be prepared for contingent use. Because foci can still reoccur occasionally after the resection of most of them and the bleeding cannot be properly controlled after direction resection of foci, resection can be performed within the peripheral normal tissues. The repair after this focal resection should be considered according to the size and depth of wounds, and then skin-grafts, island flaps, and free flaps can be selected to perform the phase I repair. If the focal area is excessively large but the donor site area is insufficient, resection and harvesting of normal skin on the tumor surface can be considered for back-grafting. For some special patients, if the estimated repair effect is very poor, partial resection can be performed to serve the purpose of reducing weight and improving appearance. For the patients with canceration verified by tissue biopsy, radical operation should be performed immediately.

As for therapies in other aspects such as skin cafe-au-lait macule, laser selective photothermal therapy can be selected. The spinal deformity complicated by neurofibromatosis is always serious, and its treatment is difficult. Usually the entire spinal column needs to be fixed. Tibial pseudarthrosis is a relatively stubborn disease; although dozens of surgical methods have been available, only half of the patients eventually achieve bone union. During recent years, free fibular grafting of long segment of anastomotic vessels has been adopted to treat congenital pseudarthrosis, the rate of success in bone union increases greatly, and this method is considered to be the first-choice method.

13.6.2.6 Prognosis

The patients with neurofibromatosis rarely suffer canceration of sarcoma. For the patients with rapid increase in tumor volume, attention should be paid to the possibility of canceration. If most of the metastases are hematogenous, the prognosis is poor. As postoperative recurrence is common, the first surgery should be very thorough.

13.6.3 Neuroblastoma

The neuroblasts are derived from primordial nervous crest cells, similar to the developmental sympathetic nerve system and the primordium of the adrenal medulla, and they are composed of neuroblasts in different developmental stages [45].

In most cases, this disease is sporadic, and some cases have the tendency of familial occurrence. Neuroblastoma is a malignant tumor commonly suffered in childhood [46]. Its incidence accounts for 10–12% of that of all kinds of malignant tumors suffered by children, second only to those of leukemia and brain tumor. One patient in every 1000 newborn infants can be found to suffer from it, and about 1/4 of the neuroblastoma suffered is congenital. Half of the cases are attacked by it before they are 5 years old, and the age of 1.5 is the age when such disease attacks most. Few patients are juveniles and adults. The ratio of incidence of male to female is 1.22:1 to 1.26:1.

13.6.3.1 Pathogenic Site

The tumor occurs to the sympathetic nervous system, and it is mainly located beside the midline from the cranial base to the pelvis. The previous reports indicate that those occurring to adrenal gland, abdominal sympathetic chain, pelvic sympathetic chain, and cervical and thoracic sympathetic chain occupy 1/3 each [47], but recent reports indicate that those at the retroperitoneal regions are common. Among the 212 patients reported by DeLorimier et al., 134 patients had the tumor at the retroperitoneal region, 33 patients had the tumor at the mediastinum, 5 patients had the tumor at the neck, and 6 patients had the tumor at the sacral region. Nearly half of the retroperitoneal tumors are derived from the adrenal gland. In addition, the neuroblastoma is also seen in the skin and soft tissues, small intestines, uterus, scrotum, inner ears, nasal cavity, maxilla, mandible, retina at the fundus, and posterior cranial fossa.

13.6.3.2 Pathology

1. Macroscopical morphology: It is a lobulated lump with a diameter of 6–8 cm (sometimes up to over 15 cm); it has thin capsules or has no capsule and becomes infiltrative to the adjacent tissues. The lump is soft and crisp, the resec-

Table 13.2 Shimada histological classification

Prognosis		
Mesenchymal manifestations	Histological classification of good prognosis	Histological classification of poor prognosis
Abundant mesenchyme	Differentiated type	Nodular type
	Mixed type	
Deficient mesenchyme	MKI < 200/500	MKI < 100/5000
Age < 18 months	MKI > 100/5000	MKI < 100/5000
Age of 18–60 months	Differentiated type	Undifferentiated type
Age > 5 years	N/A	All

tions are fish meat-like, wide bleeding, necrosis and cystic degeneration are common, and calcification is seen in a minority of cases.

2. Organization form: The main component is neuroblastoma, and gangliocyte differentiation to varying degrees is displayed; it can be divided into the undifferentiated type and the poorly differentiated type according to the degree of differentiation.

The undifferentiated type consists of primordial neuroblasts without visible differentiation of gangliocyte; the tumor cells are small circular or orbicular-ovate with relatively consistent cell sizes, little cytoplasm, circular nuclei, deep staining, and a large amount of visible nuclear mitotic figure; the cells are diffusely arranged in patches with little mesenchyme, and it can only be found that the fine fibrous septum separates the tumor tissues into lobulated shape or nest shape. In the poorly differentiated type, except that most cells are similar to the undifferentiated type, some tumor cells have a slightly large volume, the shape is round and orbicular-ovate and with a short spindle, the karyotin is light, the chromatin is scattered, small nucleoli are visible, some tumor cells present mulberry-shaped arrangement, and these are the early manifestations of chrysanthemum-shaped mass; with the differentiation in cells, the tumor cells present fine cell processes, the tumor cells present radiatiform arrangement to form pseudorosette, the central part consists of tenellous neurofibrilla, and the chrysanthemum-shaped masses are distributed in a scattered way or relatively densely in some regions of tumor; in addition, feltwork can be found inside the matrix and it consists of cell processes. Some tumor cells display partial or complete ganglion cell differentiation; the manifestations are as follows: the cell volume is large, the shape is oval or polygonal, the cytoplasm is abundant and acidophilic, the nuclei are large and orbicular-ovate, the nuclear membranes are obvious, the chromatin takes the foam shape or is scattered on the chromatin, the nucleoli are obvious, and binuclear cells are visible.

13.6.3.3 Biological Indicator

Many biological indicators in the neuroblastoma are correlated with the prognosis. In most of the prognosis factors, age has always been an important indicator, and the children under 18 months tend to have a good prognosis. Important biological variables include Shimada histological classification, DNA ploidy, and N-MYC oncogene amplification. In the Shimada histological classification, the neuroblastoma is divided into two types: good prognosis type and poor prognosis type (Table 13.2) according to the mesenchymal quantity of tumor specimens, the cell differentiation degree, and mitotic karyorrhexis index (MKI). In older children, DNA index is a more important prognosis factor, and DNA polyploid is correlated with the good prognosis of infants. N-MYC oncogene amplification (>10 copies) is correlated with the poor prognosis, and less than three copies are considered to mean no amplification; on the contrary, N-MYC oncogene expression (protein) is not a prognosis indicator. There are also some other prognosis variables, such as Trk-A (one gene that encodes neurotrophic factor receptor), serum lactate dehydrogenase, and serum ferritin, and some chromosome abnormalities of prognosis value.

13.6.3.4 Clinical Manifestation

The clinical symptoms are dependent on the patient's age, tumor site, and related clinical symptom. In 80–90% of the patients, the content of catecholamine and its metabolites, vanillylmandelic acid (VMA) and homovanillic acid (HVA), in urine increases. Neuroblastoma complicated by type I neuroblastoma is very rare, but some people observe that neuroblastoma can be transformed into multiple neuroblastoma or ganglioneuroma through subcutaneous dissemination.

1. Generalized symptoms: As the neuroblastoma is characterized by early metastasis and diffusion, the patients often have the chief complaint of metastatic tumor symptoms. What first appears are the symptoms of metastasis from distant sites such as the bone marrow, liver, skull, and brain via the blood channels, so the first-visit symptoms in clinical practice include arthralgia, low fever, emaciation, and anemia, and they are often misdiagnosed as arthritis, anemia, and leukemia. Although the generalized symptoms of neuroblastoma have no specificity, the lesions can become aggravated

within a short period of time due to the short disease course. The skeletal lesions often lead to severe pain, and some pediatric patients refuse to walk due to the manifestations of pain induced by osseous metastasis of tumor. Under the occasional circumstances, during the progression stage, the pediatric patients have the manifestation of bleeding tendency, which is correlated with the thrombopenia induced by a large amount of bone marrow metastasis as well as the clotting factor dyspoiesis induced by hepatic metastasis. During the 4S phase of infancy, the patients with neuroblastoma can experience liver enlargement and subcutaneous multiple nodules. Brain metastasis is manifested as headache and epilepsy in older children.

The chest plain film finds that only 4% of the patients suffer pulmonary metastasis, which results from the direct invasion of mediastinal lymph nodes into the lungs or the diffuse hematogenous spread, and the radiological manifestations can be easily confused with the pulmonary edema or interstitial pneumonia. Under occasional circumstances, pulmonary lymph node metastasis can be found during biopsy.

2. Lump characteristics: They vary with tumor origin. As for the tumor derived from the adrenal gland, the lump is located at the retroperitoneal hypochondriac deep surface; during the early stage, the shape is circular; it is smooth but fixed and solid; it grows rapidly due to the infiltration and quickly bulges out of the envelope, inducing the non-smoothness of the surface; and it grows toward the midline and crosses the midline. If it is derived from the cervical ganglions and nerve plexuses, it is the cervical lump and is often misdiagnosed with lymphoid tuberculosis or Hodgkin disease due to the early lymph node metastasis, and during the early stage, sympathetic nerve compression syndrome (Horner sign) appears. If the lump is derived from pelvic retrorectal presacral sympathetic chain, the manifestation is the symptom of compression rectum and urinary bladder and the appearance of urinary retention, anal sphincter dilation, and fecal incontinence; the rectal digital examination indicates the presence of retrorectal lumps, and the giant lumps can compress the bilateral ureters to further induce renal dropsy and compress the iliac veins to induce regurgitation disorder so as to cause lower limb edema. If the lump is derived from the sympathetic chain, the thoracic segment or the first and second lumbar vertebrae often present dumbbell-shaped growth, some extend toward the spinal canals through the intervertebral space, and the manifestation is weakness in the lower limb, eventually leading to the paralysis.
3. Diarrhea: Refractory diarrhea, also called watery diarrhea, is a special clinical manifestation of some patients with neuroblastoma. The tumor cells secrete vasoactive intestinal peptide (VIP) and inhibit the intestinal mucosa's absorption of water. The pediatric patients often have the manifestations of chronic watery diarrhea and water loss sign, which is often misdiagnosed as autumn diarrhea and summer diarrhea and hence delays the treatment.
4. Characteristics of metastatic tumor: In case of osseous metastasis, it is often misdiagnosed as arthritis due to the fixed osteoarticular pain. However, this tumor often becomes metastatic to the eye socket. What is most noticeable is the early manifestation of hyperemia and congestion of the soft tissues around the eye socket. The infiltration of the retrobulbar optic nerves induces exophthalmos, eyelid edema, and congestion in skin of eye corner, which is called cat's eye. Blindness is induced due to lump compression. Although sometimes tumor resection and chemoradiotherapy can make the cat's eye disappear, the visual impairment is often permanent.

The hepatic and subcutaneous metastatic tumors are manifested as giant liver and smooth, hard, and solid surface; in the meantime, hard subcutaneous active nodules are palpable in multiple sites, and the surface is mild purple-blue; as the patients often suffer from serious anemia, thrombopenia, and 24-h urine VMA/HVA significant increase, they are often diagnosed with nephroblastoma. In this case, 90% of the tumors are derived from adrenal gland with the intrahepatic infiltration and subcutaneous nodules of special giant tumor cells.

13.6.3.5 Clinical Staging

The criteria for the clinical staging of neuroblastoma formulated by International Neuroblastoma Staging System (INSS) in 1997 are as follows:

1. Stage 1 is limited to the tumor in the primary region, it can be removed completely, with or without microscopic residue, and microscopic metastasis can be observed in the ipsilateral or contralateral lymph nodes.
2. The unilateral tumor at stage 2A cannot be removed completely, but there is no microscopic metastasis in the ipsilateral lymph nodes.
3. The unilateral tumor at stage 28 can be removed completely or incompletely, but there is no microscopic metastasis in the ipsilateral lymph nodes.
4. At stage 3, the tumor invades to cross the midline, or without local lymph node metastasis, there is ipsilateral lymph node metastasis as for the unilateral tumor, or there is ipsilateral metastasis at the bilateral lymph nodes as for the midline tumor.
5. At stage 4, the tumor spreads to distal lymph nodes, bones and bone marrow, liver, and other organs.
6. At stage 4S, as is verified, the primary tumor occurs at stage 1 or stage 2, and the distal metastasis is limited in the liver, skin, or bone marrow, and it is only limited to the baby under the age of 1.

13.6.3.6 Diagnosis and Differential Diagnosis

Neuroblastoma is a high malignant tumor with early metastasis, and sometimes the metastasis symptoms occur (the osseous metastasis has the manifestations of pain of limbs, arthralgia, and low fever), but the main tumor does not appear, and it is apt to be misdiagnosed as arthritis and rheumatic fever. As the tumor cells secrete vasoactive intestinal peptide and lead to refractory diarrhea, diagnosis is often delayed due to its misdiagnosis as diseases such as autumn diarrhea and summary fever; therefore, the diagnosis should be made according to the age of diarrhea and some before-cancer syndromes in combination with the corresponding clinical laboratory examinations and imaging examinations.

Neuroblastoma often occurs to children aged under 5, the site of lump is closely associated with the sympathetic chain and adrenal medulla, it mainly consists of small round cells, and pseudorosette can be formed. The catecholamine and its metabolites in urine increase, the cytogenetics displays the deletion and rearrangement of no. 1 chromosome short arm, which is the main basis for the diagnosis of neuroblastoma, and it should be differentiated with diseases such as ectosteal Ewing sarcoma, embryonal rhabdomyosarcoma, and lymphocytic lymphoma.

13.6.3.7 Treatment

Neuroblastoma is highly malignant solid tumor during the childhood, and currently its overall survival rate is not more than 70% internationally. Its treatment principle is based on the patient's age, clinical staging, seroenzyme status, and pathological classification of the tumor cells as well as the molecular biological and genetic characteristics. It is divided into high-risk group and low-risk group, and different treatment methods and measures can be selected (Table 13.3).

1. Surgical treatment principle.
   (a) Low-risk group tumor: the first visit is usually paid at the advanced stage clinically; if the tumor is local tumor according to B-scan ultrasound, CT, and MRI, it is proposed that chemotherapy and radiotherapy should be selected according to the pathological classification after early surgical treatment. As for the surgery, it is suggested that large transverse incisions be made at the abdomen to fully expose the tumor. In tumor resection, the kidneys should be protected as much as possible to avoid separation and stimulation of the renal hilum vessels and to minimize the renal injuries from the renal ischemia induced by vasospasm.
   (b) High-risk group tumor, namely, the advanced neuroblastoma: the recommended therapy is the triple and quadruple chemotherapies; after four to eight treatment courses, the delayed or phase II surgical treatment can be performed after the tumor shrinks and the metastasis foci disappear and the tumor envelopes are more clear, thickened, and apt to be separated for the purpose of improving the resection rate of tumor.
2. Radiotherapy: Studies show that neuroblastoma has a high sensitivity to X-ray, which is applicable to the phase 3+ patients with residual tumors in primary foci or the patients with difficult-to-radically-remove dumbbell-shaped tumors inside the vertebral canals. For the patients with postoperative recurrence, radiation therapy can be given after the surgery.
3. Chemotherapy: After 10-year epidemiological studies, Japanese scholars found in 1985 many advanced patients, so Japan unified the chemotherapy regime for advanced neuroblastoma, namely, cyclophosphamide, vincristine, Adriamycin, and cisplatin.

Table 13.3 Risk factors of neuroblastoma

Factor	High-risk group	Low-risk group
Age	>1, especially >2	<1
Clinical staging	Stage INSS4, stage 3 (partially)	Stage INSS1, stage 2, stage 3, stage 4S
N-MYC amplification	>10 copies	<10 copies, especially <3copies
1P36 deletion	Present	N/A
Cell classification	UH	FH
DNA ploidy	Amphiploid tumor	Multiploid tumor
Trk-A	No expression	High expression
(In diagnosis) ferritin	>143–150 ng/mL	<143–150 ng/mL
CD44	No expression	High expression

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14 Applications of Microsurgical Reconstructive Surgery Technique in the Repair of Hand Congenital Deformities

Tsu-Min Tsai, Y. Tien Huey, and Dong Han

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14.1 Psychological Preparations of Pediatric Patients and Parents

The pediatric patients and parents must have enough psychological preparations before considering adopting microsurgical techniques to repair hand congenital deformities. For the parents, their child has lost one hand or one pair of perfect hands; if they consider undergoing toe transplantation or using free flaps to repair the deformed hand, the pediatric patient will lose the complete toes or other histological structures, and new injuries will arise; in the meantime, the surgeons cannot guarantee that the microsurgery is bound to succeed; therefore, the parents always have many concerns and are under great pressure, which will unconsciously influence the surgeons; under this circumstance, many surgeons will passively select microsurgery. Therefore, for the surgeons, pediatric patients (if possible), and their parents, before the microsurgery, an in-depth discussion will be conducted on various conditions before the surgery, during the surgery, and after the surgery; the child's family will be asked to make the final choice and the surgeons will respect this choice, which is very important.

Parents always feel dejected, self-blamed, and even angry when seeing a pair of imperfect hands after the child's birth because children are the hope of parents. If they lose the perfect hands, the parents feel that they will not have a bright future. This psychological manifestation is similar to the sensation of losing family members and friends, but what is more cruel is that the pain of losing family members is transient and will subside with the lapse of time, but the child's parents will suffer the pain and torture for as long as they see the hands every day. Most patients will ask why this happens and whether they have done anything wrong when taking the pediatric patients to see a doctor at the outpatient service; at the same time, they will be worried whether their next child, if they continue to bear, will also suffer from the disease.

Some parents can quickly make psychological adjustments, accept this fact, and actively seek for treatment, but

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some parents will be in a bad mood for a long time. A hand surgeon should be good at observing the reactions of the family, pay attention to the parent's emotions, and kindly show their concern. If necessary, psychologists should be invited to give interventional treatment.

Hands are the windows for the pediatric patients to explore and sense the world. With the increase in age, they will be gradually aware that their hands are different from those of others. As is often described by the pediatric patients' parents, they will look at that deformed hand for a long time at the age of 1–2; when they become 3–4 years old, they will ask why their hands are different from those of others, which is very painful for the parents. They not only need to face this frustrating fact but also worry that the deformed hands will traumatize the children. In fact, the parents should straightforwardly answer this question to let their children know that this is a discussable question that does not have to be deliberately avoided so that the children can feel relaxed and get out of the predicament gradually more easily. When they go to kindergarten or go to school at the age of 5–6, the children will have a more profound understanding on hand deformities; even if they are not teased by their classmates, they still feel sad and often hide their hands, so they are unhappy during this period of time. Some pediatric patients will gradually overcome this psychological barrier, some will become coward and weak, and some will adopt other methods to make up for the deformities; during this period, the parents' correct guidance is of vital significance. After entry into middle school, the child's environment becomes more complicated; additionally, teenagers tend to rebel; some introverted pediatric patients will be faced with the mental preparation; at this time, the parents should give them more help and guidance and encourage them to make more friends instead of hiding themselves at home. Teenagers pay special attention to their physical changes and are worried about their future due to the deformities and about whether the deformities will influence the interaction with the opposite sex. During this period, they become alert; for the hand surgeons, the communication between them and the pediatric patients becomes difficult during this period of time. Their entry into the adulthood does not mean the end of the problem because they will be faced with important life choices, such as making friends, getting married, and seeking a job. At this time, they will consider surgical treatment to make a change. As a hand surgeon, they should be clearly aware that circumstances change with the passage of time and new problems will keep emerging, and they should be asked to carefully consider the necessity of the surgery.

The postoperative effects on some deformities are good, such as syndactylia and duplication of the thumb, but the surgical effects on some deformities may not be obvious, such as microsurgery, the parents and the pediatric patients have to sacrifice a lot for the surgery, there will be obvious postoperative scars, and the appearance is not beautiful. As for whether the surgery should be performed and when to perform it, the hand surgeons should actively communicate with the parents and the pediatric patients (if they are old enough), let them know the loss and gain after the operation, decide which is more important, and allow them to make the decision on their own. Especially when multiple surgeries need to be done, find a way to let the pediatric patient agree and accept; otherwise, the surgeries will become increasingly difficult, thus affecting the postoperative effects. What is gratifying is that, one investigation indicates, the pediatric patients that receive toe-to-hand free grafting for finger reconstruction are very satisfied with the postoperative functions, aesthetic extent, appearance of donor site, and mental health, and the satisfaction of the pediatric patients even exceeds that of the parents.

When the surgery should be performed is one of parents' top concerns, and pediatric patients at different age groups have different responses to surgery. The performing of surgery before the age of 1 does not have a great psychological effect on the pediatric patients. The age of not less than 1, especially between 15 months and 13 years, is a period when the patient can face lots of difficulties, so it is required that the parent should give more guidance and properly divert the child's attention. The pediatric patients aged between 3 and 6 have been capable of knowing the significance of the surgery to them; if pediatric patients have a close relation with the parents and the postoperative analgesic effect is good, they can better handle the surgery, so microsurgical treatment can be considered during this period. Many parents also wish to have the surgery completed before the school age of their child, which is beneficial to the children's mental health. If they are older, the pediatric patients are unwilling to receive the surgical treatment for this or that reason. The complicated micro-replantation should be avoided during the puberty because during this period the patients undergo rapid changes in psychology and physiology, they are not psychologically mature enough, and the surgery often leads to their inner panic, the fear of death, and even the strangeness and aversion to their body after the operation. Since adults have a sensation of fear toward the surgery, the children have the same reasons to feel frightened, so the parents should play an active role during the perioperative stage in minimizing various kinds of predicted damages and lowering the child's fear of the hospital and surgery to the minimum.

During the previous treatment, the surgeons and parents always emphasized the surgical improvement of the hand functions and appearance, but they neglect the influence exerted by the surgery on the child's psychological growth. During this period, the parents play the most important role. In case the parents have a good state of mind, their child can develop a very healthy attitude; if the parents do not have a good state of mind, the surgery may have a negative influence on the children's psychology, even if the operative effect is good. Therefore, when the surgeons are treating the pediatric

patients, they should not only observe the hands but also pay attention to the entire state of the infantile patient, the parents' state of mind, and their family conditions; if necessary, a psychologist should be invited for the intervention; only in this way can a good therapeutic effect be achieved [1].

14.2 Transplantation of Free Tissues to Treat Hand Deformities

Currently, for the patients of different age groups, transplantation of free tissues to treat hand deformities is feasible and reliable. In spite of a certain risk, the failure rate is relatively low in terms of the upper limbs. Traditional therapeutic techniques, such as pedicled flaps and dilators, often include multistage therapies; as a mature technique, microsurgery can be flexibly selected according to the needs, such as free grafting of skeletons and joints with epiphysis; after the vessels are anastomosed, the original growth capabilities can be maintained, so that the problem can be solved reliably once and for all. Techniques such as skin grafting can always limit the growth and development of the corresponding sites of infantile patient; however, free skin flap almost has nearly no such side effects as it can lay a good soft tissue foundation for various reconstructive surgeries in the late stages along with the growth and development of the pediatric patients. In addition, the regeneration capability especially that of nerves of children is very powerful; no matter if it is a direct nerve repair or transplantation, good effects can be obtained; even if nerve repairing surgery cannot be performed, good sensation recovery can be eventually obtained, which is incomparable by adult surgeries. Therefore, the transplantation of free tissues is beneficial to children in multiple aspects. Microsurgery for children is slightly different from that for adults. Long-term anesthesia is more apt to cause problems, and vascular anastomosis is more difficult; therefore, an experienced microsurgeon should be selected as the operation surgeon. However, successful microsurgery does not always solve all the problems, and sometimes it is necessary to perform phase II flap thinning, scar plastic surgery, and re-repair of the covered tissues; therefore, a sufficient and detailed communication (including problems arising from surgical failure) should be made between the pediatric patients (especially the pediatric patients aged above 5) and the parents before the operation for the purpose of winning the child's trust so that the pediatric patients will not resist or evade the subsequent treatment. If conditions permit, the surgeons should complete all the needed surgeries once and for all to reduce physical and mental damages to the infantile patient. During the first several months after the birth of the infantile patient, microsurgery is not suggested, and the surgery should be completed before the school age, which is the conclusion drawn by most hand surgeons after long-term clinical observation and overall measurement. As for the estimation of surgical effects, consideration should be based on multiple functions. Actually, the appearance and functions of the hands are of vital importance to the mental health and growth of the children. Therefore, the professional hand surgeon should try their best to improve the appearance to obtain a pair of beautiful hands.

14.3 Application of Free Tissue Flaps

The commonest free tissue (skin) flaps for the treatment of infantile hand congenital deformities include back broadest muscle (skin) flap, serratus anterior muscle (skin) flap, gracilis muscle (skin) flap, groin skin flap, arm lateral skin flap, fibula flap, and toes. The muscle flaps can be used to cover the wounds with tissue defects and serve the functions of padding dead space, controlling infection, and improving appearance. If only the muscle flaps are used, it is necessary to perform split-thickness skin graft on the muscle surface for transplantation to close the wounds. If the muscle flaps are used for free grafting to reconstruct the functions, higher requirements are proposed for the surgical team, including the selection of donor sites, the design of enthesis, the regulation of tension, the high-quality anastomosis of nerve vessels, the good balance of blood circulation, and the postoperative rehabilitation, which are of vital importance; in addition, the growth capabilities of the muscle flaps are poor, which should be noted. The selection of muscle flaps need to conform to the following principles: ① if the functions of the limb and the trunk at the donor site are affected, the consideration should be careful; ② if the vessels adopted by the flaps are the main vessels of the limb, the selection should be cautious, especially the upper limbs with congenital deformities, and radial and ulnar arteries of the forearm may be the only blood supply source; once adopted, limb necrosis will result.

In the infantile hand congenital deformities, the free tissue flaps are mainly used for the plastic surgery and repair of the contracture of the first finger web and the deformities of the dorsal side of the palm. The local transposition using the index digital dorsal flaps is often adopted to repair the contracture of the first finger web (thumb dysplasia with finger web contracture) to serve the purposes of widening the finger web space and improving the hand functions. However, for some seriously deformed hands, such as mitten-like deformities, a large number of skin soft tissues are needed to widen the first finger web; as the local flap grafting always cannot satisfy the requirements, the retrograde island-shaped flaps in the forearm can be adopted to widen the finger web, but the blood supply at the end of the pedicled flaps is usually not reliable, and the shape and area of the harvested flaps are often limited. Free flaps can overcome these disadvantages, the needed soft skin tissues can be fully resected and harvested at the donor site, the recipient site will not have addi-

tional scars, the blood supply of the flaps is reliable, this can solve the problems once and for all, and flaps such as groin flaps and arm lateral free flaps can be used to widen the first finger web. The traditional flaps that repair the palm dorsum mainly include the pedicled groin flaps, radial arterial and ulnar arterial retrograde island-shaped skin flaps, dorsal interosseous arterial retrograde island-shaped skin flaps, and ulnar arterial supra-carpal retrograde island-shaped skin flaps. Generally, forearm skin flaps should not be used because this will leave an obvious scar, and the blood supply of the forearm and hands will be affected under some circumstances. The forearm lateral lower skin flaps and the groin free skin flaps are very good choices; in case the wounds are large sized or accompanied with tissue defects, the free flaps at the broadest muscle of the back can be selected for the repair. From the perspective of injuries at the donor sites, the groin free skin flaps are the most ideal, the injuries at the donor sites are slight, and the positions are very hidden. The resection and harvesting of forearm lateral lower free skin flaps are often accompanied with the injuries with cutaneous nerves, so it is not the first choice.

14.4 Common Free Tissue Flaps

14.4.1 Back Broadest Muscle (Skin) Flaps

Back broadest muscle (skin) flaps can be used to pad the wounds covered with tissue defects and also used to perform pedicled (vascularized) muscle grafting for the reconstruction of limb functions. The resection and harvesting of back broadest muscle (skin) flaps are easy; the pedicles are constant; a large area can be resected; the muscles are thick, solid, and powerful; and this wins warm praise from the surgeons. The disadvantage is that the donor sites have large scars; in case of only the need of muscle flap resection and harvesting, the transplantation of autologous skin flaps is often needed to cover the muscle flaps. Based on the vascular anatomical characteristics of this skin flap, it can be designed into the two pedicles for transplantation and repair. The pediatric patient is often lying in lateral recumbent position during resection, but some surgeons can complete the resection and harvesting when the pediatric patient is lying in the dorsal position. The lateral decubitus position may induce brachial plexus paralysis, so the duration of surgery at this position should be shortened as far as possible, and every hour the limb at the surgical side is placed at the lateral side of the body for 5 min to prevent paralysis. Whether the back broadest muscle skin flaps or the simple muscle flaps should be resected and harvested is determined according to the conditions of the donor site and actual needs. In case of resection and harvesting of muscular skin flap, the skin can be partially designed at the position close to the lateral middle axial line; if 50% of the skin flaps exceed the muscular borders, usually no blood supply problem will be caused, and this design will make the scars at the donor sites of the skin flaps covered by the upper limbs. First, the anterior margins of the broadest muscle of the back should be searched and located, and the thoracodorsal nerve vascular bundles are often 2–3 cm away from the medial side of the anterior margins; when the retractor is used to separate the broadest muscle of the back and the serratus anterior muscle, the nerve vascular bundles and their position in the broadest muscle of the back (the position is often at the nipple line for men and children) can be observed, and at this time, the surgeon can ascend the exploration upward along the nerve vascular bundle. Generally speaking, the ascending of the vascular pedicles may not necessarily reach the level of axillary artery because this will not significantly increase the length of vascular pedicles and will compromise the resection and harvesting of scapular skin flaps. If the thoracodorsal nerves with muscles are not needed, the surgeon should retain the nerve branches as many as possible under the microscope and only harvest the vascular pedicles; in this way, the motor function of the remaining broadest muscles of the back can be maximally retained. After the resection and harvesting of muscle (skin) flaps, the surgeon should try his/her best for direct suturing. In case the tension is excessive, local rotation flap can be designed for the coverage to address the problem because skin grafting will make the appearance of this region ugly and bring trouble to the nursing. Postoperative conventional drainage for 3–5 days can prevent the formation of hematomas until the patients can move [3–6] (Figs. 14.1 and 14.2).

14.4.2 Forearm Lateral Lower Skin Flaps

The vascular source of forearm lateral lower skin flaps is the posterior descending branch of the deep brachial artery which runs at the position 1/3 beyond the upper arm lateral intermuscular septum, and the anatomical projection is from the triangular muscular nodules to lateral epicondyle of the humerus. The starting point of this skin flap contains much fat and the distal end contains little fat, so the skin flaps become thinner and thinner from the initial point to the lateral condyle of humerus. When necessary, the skeletal flaps near the lateral condyle of humerus, some muscle tissues, and a small number of the tendons of triceps muscle of the arm can be resected and harvested according to the repair needs. The resection and harvesting of the skin flaps are often from the posterior side to the anterior side, and it is performed along the sarcolemma of the triceps muscle of the arm. When the lateral intermuscular septum is reached, the triceps muscle of the arm is pulled toward the posterior side; the skin flap vessels can be seen; then from the front to back, the skin flaps are resected and harvested at the same layer until the position of intermuscular septum is reached. The distal ends of the skin

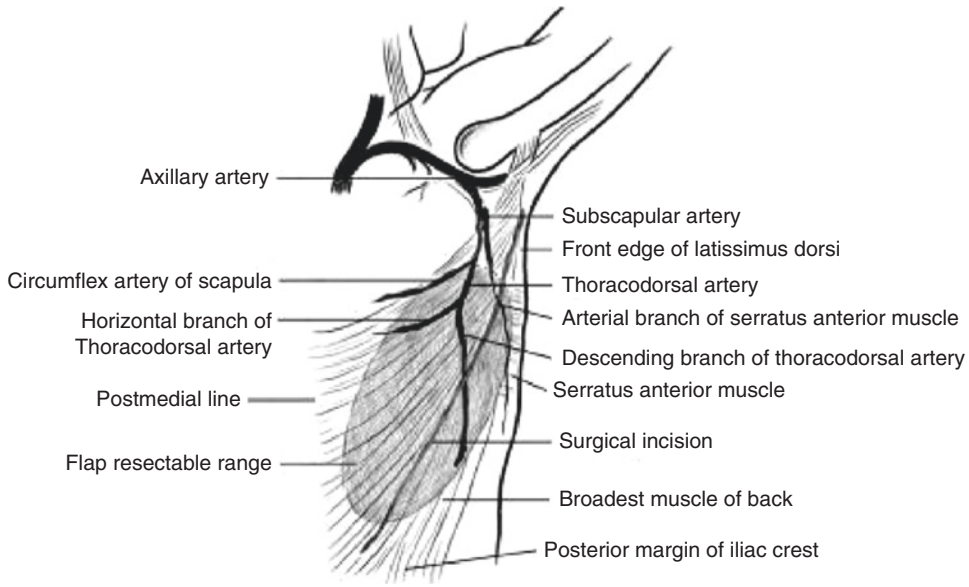

Fig. 14.1 The thoracodorsal artery often enters the muscle tissues at deep surface of the broadest muscle of the back after being sent out from the circumflex scapular artery. Muscle (skin) flaps are often designed at the anterior lower part of the broadest muscle of the back; when no skin flap is contained, the surgical incisions are parallel to the anterior margins of the muscles and at the region 2–3 cm of the posterior margins

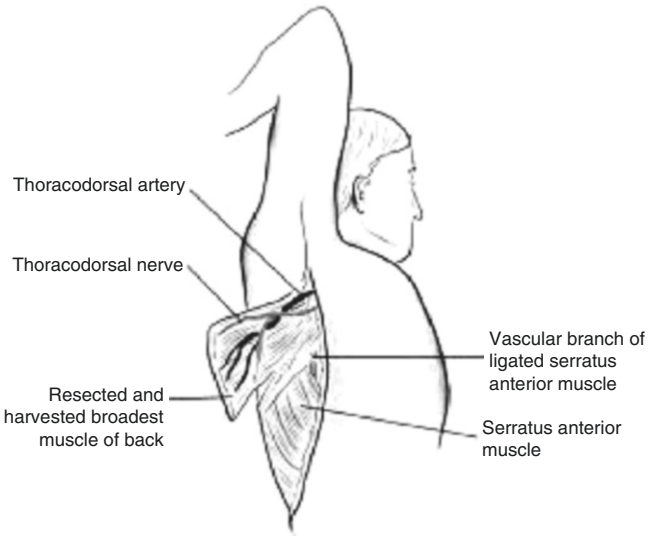

Fig. 14.2 Distal margin, anterior margin, and posterior margin of the muscle flaps have been separated, and the ligation to the vascular branches of serratus anterior muscles can separate the thoracodorsal artery toward the proximal end

flaps can be resected and harvested at the superficial layers of the membrane around the extensor tendons. When three directions of the skin flaps have been resected and harvested, the intermuscular septum that contains the flap vessels is resected at the superficial layers of the periost from far to near, the exploration is ascended toward the proximal end, and the maximal resection length of the vascular pedicles can be up to 6 cm. During the process of pedicle separation, the skin nerves of the forearm are first seen, then radial nerves are observed after the separation toward the proximal end, and the radial nerves should be protected when the tissues are pulled. If possible, suturing should be performed directly in the donor sites of the skin flaps; if the tension is excessive, the free skin grafts can be used for transplantation to cover the wounds. The advantage of this skin flap is the clear anatomical hierarchy, the blood supply is reliable, the resectable and harvestable area can vary according to the specific needs (the minimum is 10 cm^2 and the maximum is within the 12 cm at the proximal end of the forearm), and the length of the vascular pedicle is sufficient; as the distal part of the skin flap is thin, the appearance can easily be accepted after the repair. As this flaps supplies the blood in the axial manner, it can be designed into long and narrow thin skin flap; after combination, they can be used to cover the wounds, and the donor site can be sutured directly. The color and texture of this skin flap are close to those of hands; therefore, repair of the sites such as the back and thumb web is proper. In addition, some scholars propose that the lateral cutaneous nerve of the forearm can be contained inside the skin flaps and can be anastomosed with the sensory nerves in the donor site to improve the sensation of the skin flaps; or it can be treated as vascularized nerves for transplantation. However, some scholars propose the opposite opinion that the sensation of the forearm skin nerves is very important, and sensory functions at the lateral side of the forearm should not be sacrificed. In fact, the posterior skin nerves of the arm contained in this skin flap can be used to anastomose the donor site nerves to obtain the sensation of the skin flaps.

The disadvantage of this skin flap is the difficult direct closure of the donor site, and in case of skin grafting the scars are obvious; if the skin nerves are contained in the skin flaps, anesthesia can be observed in most of the radial side of the forearm. As for the specific resection and harvesting methods of the skin flaps, there are many related books and articles, which will not be described here. It should be noted that the smaller skin flaps means the increasingly finer design and resection and harvesting and the higher requirements on the operators. If it is necessary

to resect and harvest the micro free flaps at the lower part of the lateral side of the arm, the most reliable position is the lateral condyle of humerus, and the vascular anatomy at this site is constant [3, 7] (Figs. 14.3, 14.4, 14.5, 14.6, 14.7, 14.8, 14.9, and 14.10).

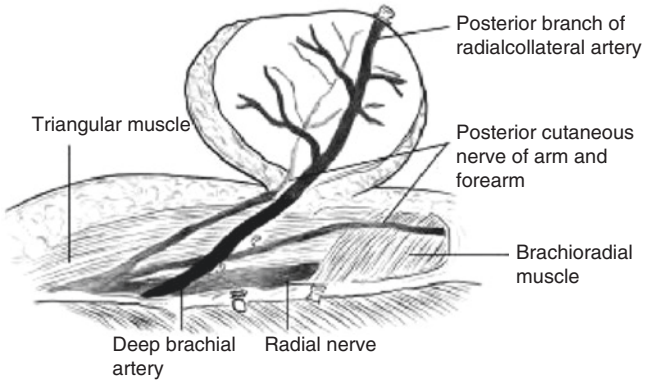

Fig. 14.3 Design of skin flaps that contain radial collateral arterial posterior branch and forearm posterior cutaneous nerve

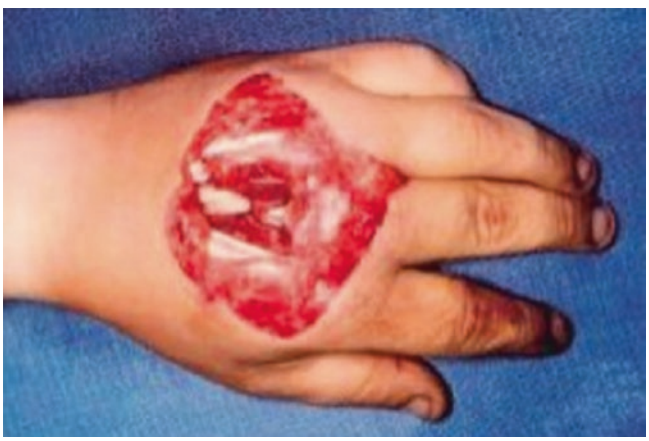

Fig. 14.4 Hand dorsum wounds complicated by the defect of the third metacarpal bone

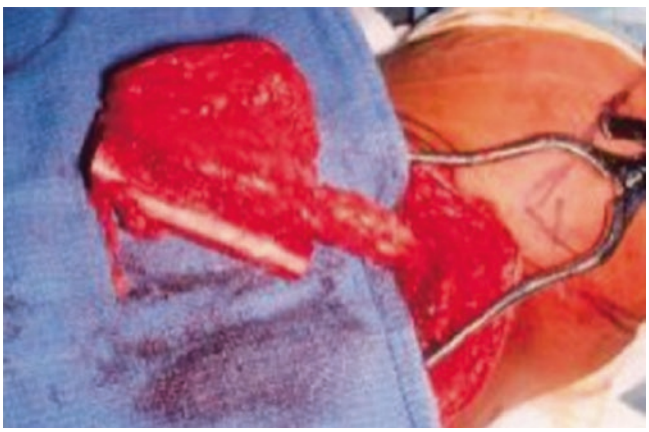

Fig. 14.5 Dissociation of bone skin flaps at the lower part of the forearm lateral side that contains some skeletal flaps

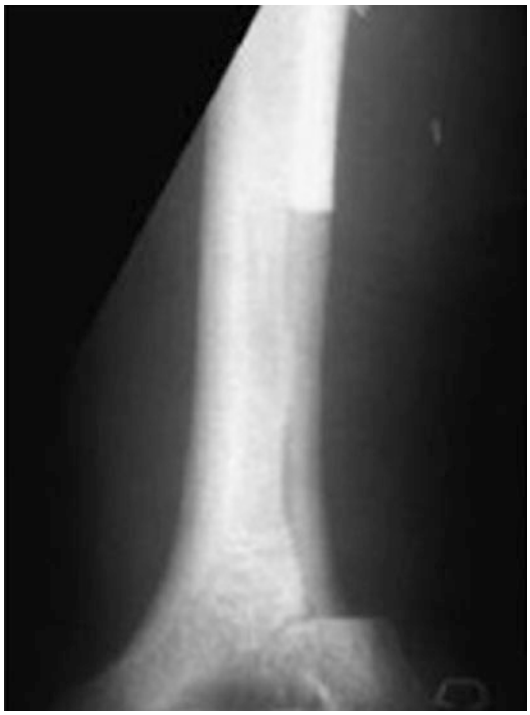

Fig. 14.6 Skeletal flaps removed from the lower part of the humeral bone as indicated in X-ray

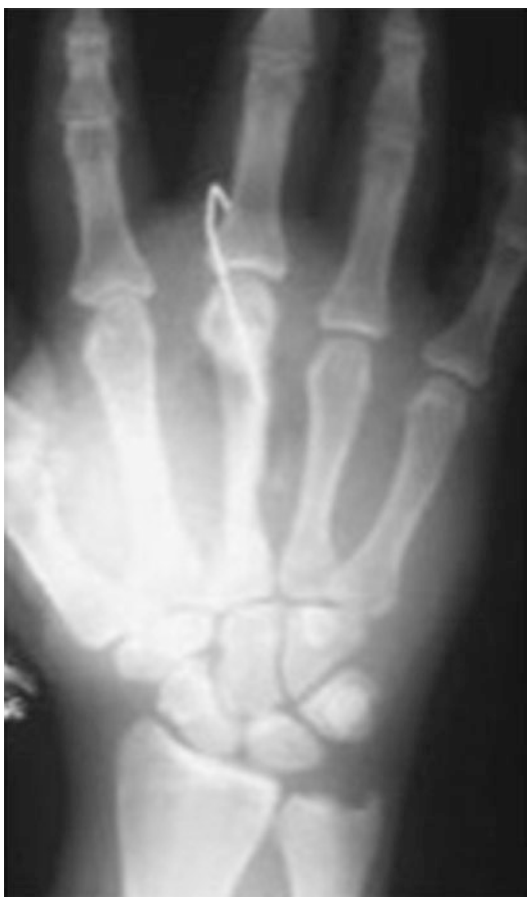

Fig. 14.7 Skeletal flaps are used for bridging and repair of metacarpal defects, and skin flaps are used for covering the soft tissue defects

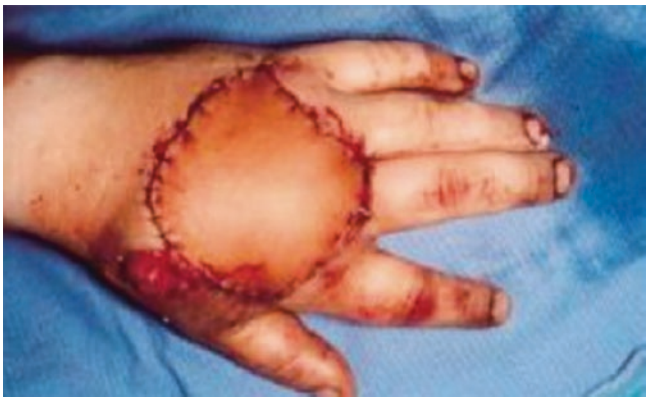

Fig. 14.8 Immediately after the bone skin flap repair

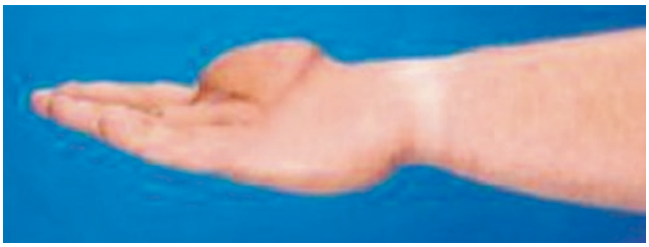

Fig. 14.9 Good healing after the bone skin flap repair

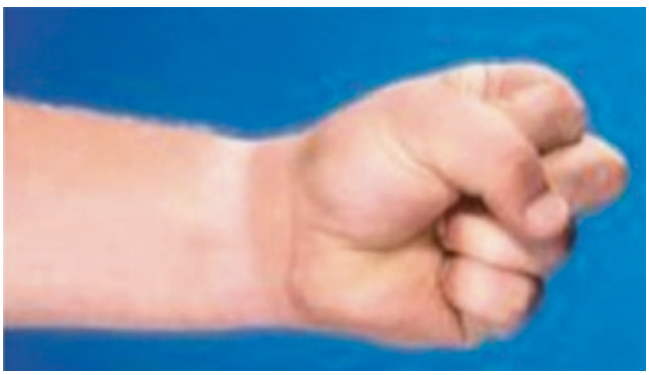

Fig. 14.10 Good functions after the bone skin flap repair

14.4.3 Groin Flap

Groin flap is usually the narrow and long skin flap with the superficial circumflex iliac vessel as the design, but this skin flap is not perfect due to the following disadvantages: the vessels are thin, the anastomosis is difficult, and there are some variation in the anatomy (the incidence is up to 96%); its arterial pressure is not as high as that of other famous vessels, the circulating blood volume is not sufficient, the length of vascular pedicles is limited, and there are no proper skin nerves inside the skin flaps for the anastomosis. For children, this kind of flap has a prominent advantage: the position is hidden, and the influence on the donor site is little. In the world today where super microsurgery and perforator flap enjoy a great development, the microsurgical technique and patience should not impede our steps, and we should have more reasons to emphasize this skin flap. For children, this skin flap is not swelling and can be harvested in the inguinal flexion crease regions, but the pudendal region should be avoided in case the appearance might be ugly due to the growth of pubic hair after the pediatric patient develops and becomes mature.

In the specific design of skin flaps, small free skin flap is a good choice, and the region can be from the femoral artery to the median margin of the sartorius muscle and also can be lengthened to the regions beyond anterior superior iliac spine according to the specific circumstances. As the superficial circumflex iliac vessel is thin with anatomical variation, some scholars propose that exploratory discrimination be performed on the pedicle vessels before the resection and harvesting and then the skin flaps be further resected and harvested. However, some scholars oppose this practice and believe it is unnecessary because there is always a group of vessels in this region, excessive exploration will destroy the blood supply of the skin flaps, and the skin flaps can be resected and harvested from the lateral side to the medial side. Usually, after the superficial circumflex iliac artery is sent from the femoral artery, at the distance of one to two fingers (the child's fingers) below the inguinal ligament and parallel to the ligament, move to the anterior superior iliac spine. Lateral cutaneous nerve of the thigh is often below the inguinal ligament or in the position 1 cm below it and through the median side of the anterior superior iliac spine, and it is crossed with superficial circumflex iliac vessel; in resection and harvesting of skin flaps, attention should be paid not to injure this nerve; otherwise, this will induce refractory neuralgia. In resection and harvesting of skin flaps, with the superficial circumflex iliac vessel as the central axis line, it should be enlarged by 20% according to the actual needs for resection and harvesting, and trimming is then performed according to the needs. The skin flaps are elevated from the lateral side to the medial side; in case the lateral cutaneous nerve of the thigh is met, the separation should be careful to avoid injuries. When passing the sartorius muscle, the superficial circumflex iliac artery comes out of the superficial layer of the deep fascia at the median margin of muscles, and some branches enter the muscle tissues; then ligation is performed and the sarcolemma should be carried. In the meantime, pay attention to the thorough hemostasis of the skin flaps; otherwise, the postoperative bleeding may compress the vascular pedicles, resulting in surgical failure. Under the microscope, exploration can be ascended to the vascular pedicle until the femoral arterial branch is reached. During this process, the vascular muscle branches are carefully ligated, and then the vascular pedicles are harvested according to the needs. Then

the accompanying veins at the medial side of the pedicles are harvested for anastomosis; although the superficial veins inside the skin flaps have a large bore, they do not belong to the reflux branches of the venous network inside the veins, so they are not selected. Although the vascular pedicle of this skin flap is small, the matching with the bore of the deep branch at the dorsal side at the small radial arterial nasopharyngeal fossa is good. The harvested skin flaps can be thinned through properly defatting, especially the peripheral parts of the skin flaps, to avoid injuring the vessels, and attention should be paid not to remove the adipose tissues between the deep fascia and the skin [3] (Figs. 14.11 and 14.12).

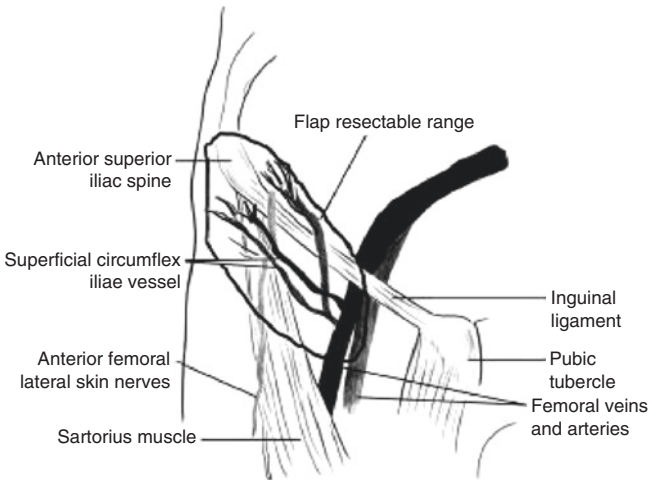

Fig. 14.11 Design skin flaps with the sartorius muscle of superficial circumflex iliac vessel as the centers; the passing of the anterior femoral lateral skin nerves can be observed in the skin flaps, they should be retained, and they should not be injured

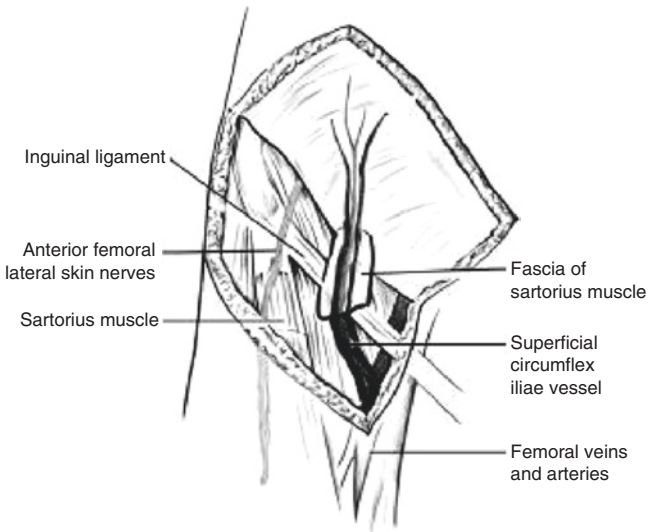

Fig. 14.12 Separate the skin flaps, including the sartorius muscle sarcolemma, and retain the skin nerves in their original position

14.5 Vascularized Bone Graft

When the bone transplantation of anastomosed vessels is used to repair the large segmental bone defect, the healing can be achieved in the way similar to fracture union, and the healing is more rapid compared with traditional bone transplantation; the postoperative rehabilitation and exercise are early, and the functional recovery is satisfying. In addition, the vascularized bone grafting can retain the epiphysis so that the capabilities of bony longitudinal growth and development are retained. For children, the advantages and disadvantages should be weighed before vascularized bone grafting, to see whether the traditional bone grafting can produce similar therapeutic effects and whether the bone lengthening technique can replace the vascularized bone grafting; if yes, vascularized bone grafting should not be considered. In the utilization of traditional bone grafting to treat hand congenital deformities, the commonest type is phalangeal transplantation to treat brachydactylia and syndactylia deformity and implantation of the entire phalanges with the complete periost and epiphyseal plates into the end of short fingers. Currently, the scholars believe that this surgery should be performed before the patient is 18 months old. If the pediatric patient is older than 18 months, the survival rate and the growth abilities of the patients after nonvascularized bone grafting will drop. Therefore, when the patient is too old for nonvascularized bone grafting, vascularized bone grafting can be considered. For the forearm congenital false joint and the large segmental bone defect after tumor resection and trauma, if the traditional bone transplantation technique is adopted, problems such as slow growth replacement, too long external fixation, and obvious scars will arise, and at this time vascularized tubular bone grafting is an option [8].

The fibula is very suitable to be used to repair the diaphysis due to its structures and appearance. It is currently the commonest donor site for vascularized bone grafting and the widely applied well-developed surgical technique. The skeletal flaps can be designed by taking the middle 1/3 part of the fibula with a length which can be up to 26–30 cm together with the skin. The blood supply of the fibular shaft mainly consists of the periost, fibular artery, and intramedullary artery in the middle of the diaphysis, and the fibular artery sent out from the tibial artery is clinically used to design the fibular flaps. The skin supply of the fibular surface is mainly from the perforating vessels at the fibular side of the gastrocnemius muscle; the fibular arterial perforating branches often fall on the connecting lines between the perforating branches and the fibular head. If they are designed into fibular flaps, careful observation should be made along the direction of arrangement and distribution of the vascular perforating branches when the deep fascia is

resected. Before the operation, it is best to use three-dimensional ultrasound to locate the vascular position and distribution. Due to the great differences between different individuals, the design of skin flaps can be adjusted under the direct vision according to the specific conditions of the perforating branches. If the skeletal flaps are resected, it should be noted that the skin perforating branches of the fibular extensors and the soleus muscle intermuscular septum should not be destroyed; the usual practice is to resect the dissociated fibular flaps and fibular arteries from the anterior and then resect the skin flaps from the posterior and protect the perforating branches of fibular artery, and the remaining method is the same as the resection and harvesting of fibular flaps. In resection and harvesting of fibular flaps, the lateral approach should be adopted and its operation is easier than the posterior approach. The long fibular muscle is a wide tendinous tissue; one fatty tissue can be observed at the rear of the long fibular muscle and is the marker to locate the space between the long fibular muscle and soleus muscle intermuscular septum, which can be easily differentiated. After this intermuscular septum is dully separated, it can be found that the skin perforating branches come out of the superficial layers of the anterior region of soleus muscle or fibular long and short muscle and soleus muscle intermuscular septum, and these vessels can be traced until the facies posterior fibularis is reached. During the separation of the fibula, the anterior fibular superficial fibular nerves and deep fibular nerves should be protected. The distal segment of the fibula at a length from 7 to 8 cm should be retained; for children, tibiofibular interosseous space at the fused distal end of iliac block should be taken to prevent the instability of the ankle joint and the developmental ankle joint extroversion. In the disjunction of the fibula, the fibular vascular bundles at the posterior side of the fibular bone should be protected. After the two ends of the fibular flaps are disjuncted and the skeletal flaps are rotated, the distribution of posterior fibular vascular pedicles can be clearly seen. The interosseous membrane should be carefully disjuncted, and attention should be paid to distinguish between the fibular arteries and veins and the anterior tibial arteries and veins. When the vascular pedicles are resected and harvested in a position close to the distal end, pay attention not to injure the common fibular nerves. Usually, the vascular pedicles are not long, but it suffices for the repairing surgery of the upper limb in most cases. In the simple harvesting of fibular flaps, the fibula can first be cut and then the fibula is rotated, which can more easily expose the fibular arteries and veins. In case of harvesting of skeletal flaps, the donor site should be covered by skin grafts through transplantation, and direct suturing may result in excessively high tension and osteofascial compartment syndrome [3, 9] (Figs. 14.13, 14.14, 14.15, 14.16, 14.17, 14.18, and 14.19).

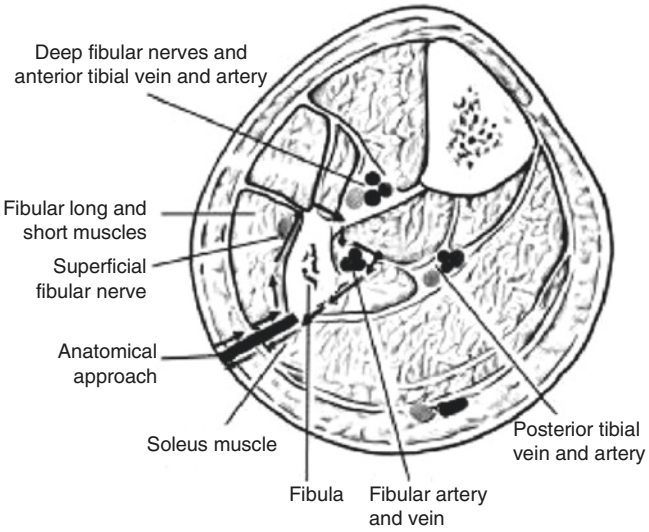

Fig. 14.13 Approach and range of fibular valve resection and harvesting (transverse section)

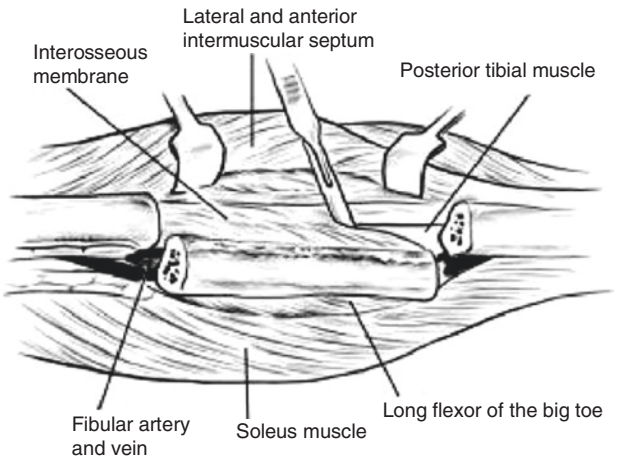

Fig. 14.14 Harvest the fibular flaps from the lateral approach, and protect the fibular nerves and anterior tibial arteries and veins. When the distal and proximal ends of fibular bones are disjuncted, the distal ends of the fibular arteries are ligated, and at this time, the intermuscular septum at the medial side of the fibular bones can be disjuncted

14.6 Free Grafting of Functional Muscles

The main functions of muscles are contraction and generation of force, and the effect and the length are closely related. When free grafting of functional muscles is considered, the morphological types of the donor site muscles are the key; in case of contraction of ribbon muscles, relatively long sliding distance can be produced, and the bipennate muscle contraction can produce large force. The free grafting of functional muscles has its special indications; this technique is only considered after the methods of traditional muscular transposition are inapplicable; in the meantime, there should be proper motor nerves available for phase I anastomosis. For

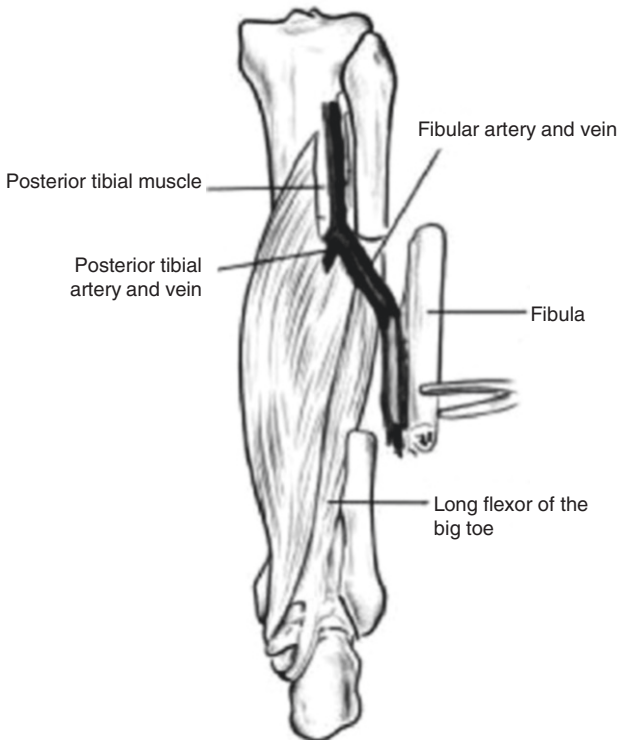

Fig. 14.15 The muscular tissues around the fibular bones are completely dissociated, and the vascular pedicles can carry some muscular tissues to avoid injuring the fibular arteries and veins

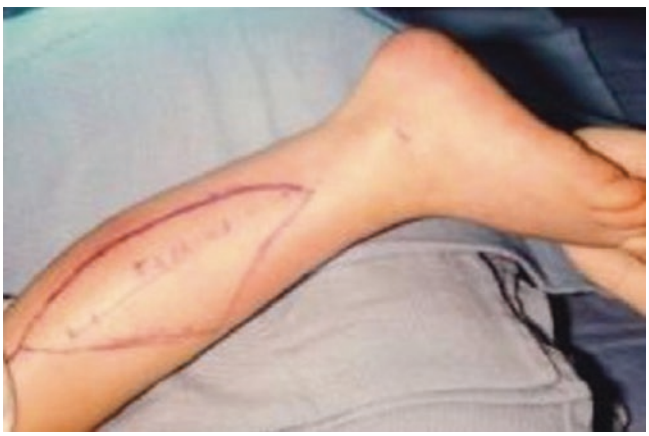

Fig. 14.16 Design of fibular skeletal flaps with skin flaps

children, free grafting of functional muscles is mainly used for the treatment of birth palsy under special conditions and the Volkmann ischemic contracture induced by humeral supracondylar fracture. If the pediatric patients suffer birth palsy and elbow functional disorder, their shoulder functions are usually poor. In that case, performing translocation of the broadest muscle of the back or major pectoral muscle to reconstruct the elbow bending functions is bound to make

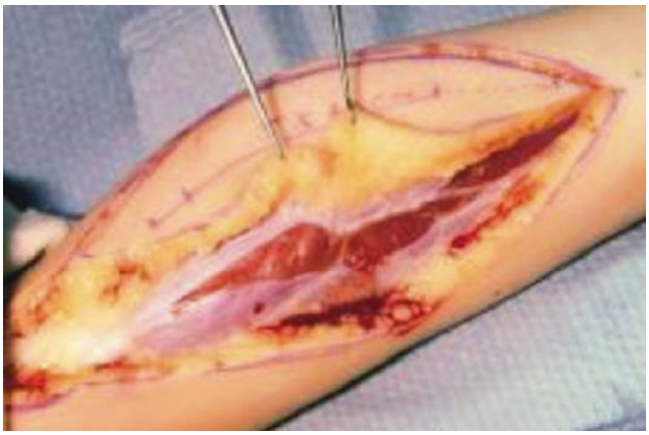

Fig. 14.17 The skin flaps are incised from the anterior to the posterior, and the intermuscular septum is separated for entry

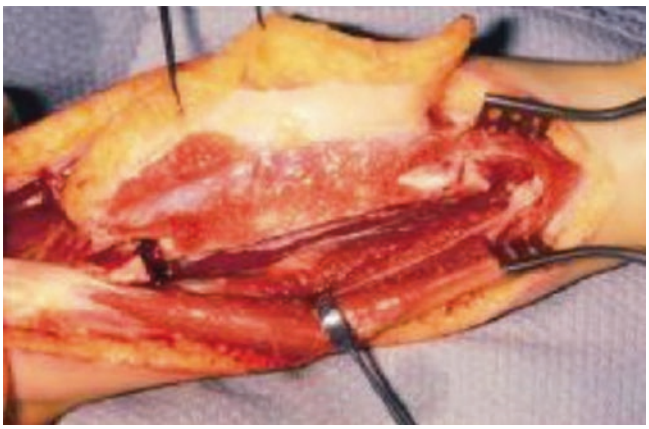

Fig. 14.18 The two ends of the fibular bones have been amputated

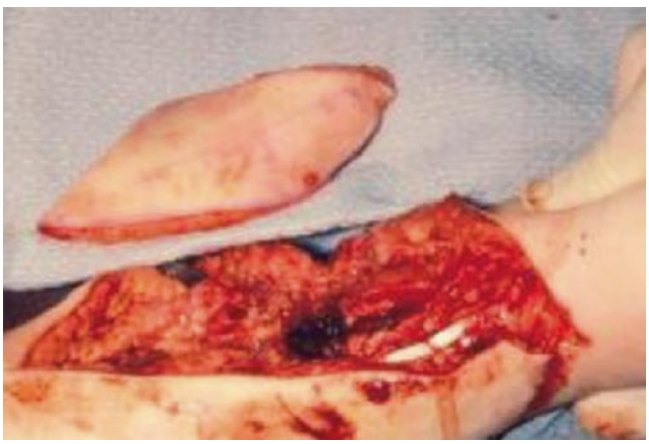

Fig. 14.19 After the resection and harvesting of skeletal flaps

the function of the shoulder joint poorer. In this case, free grafting of functional muscles is a solution. The motor nerves at the donor sites can be provided through the transposition of intercostal nerves or other nerves. Attention should be

paid to the following aspects in free grafting of the functional muscles: ① The postoperative appearance at the donor sites can be accepted; ② the muscles have sufficient length and width so that enough strength and sliding distance can be produced; ③ phase I motor nerves and vessels for anastomosis should be provided at the donor sites. The free grafting of functional muscles is still in the middle of exploration. Although some hospitals have made great achievements in free grafting of functional muscles, it is too early to draw a conclusion. Usually, the muscles from three donor sites, i.e., gracilis muscle, broadest muscle of the back, and gastrocnemius muscle, can be considered for the grafting. The free grafting of functional muscles is often adopted to recover the functions of finger flexion, elbow joint flexion, and shoulder abduction.

14.6.1 The Gracilis Muscle

Located at the medial side of the thigh, the gracilis muscle is the most suitable donor site for the free grafting of functional muscles; in the meantime, it can carry one skin island at the proximal side of the muscle, and the vascular perforating branches of skin island and the vascular pedicles of muscles are about at the same level and relatively constant. For most of the patients, the fat at the medial side of the thigh is relatively thick and seems to be swelling if carrying skin flaps; therefore, they prefer to add skin flaps to cover the muscles during the simple free grafting of functional muscles. The enthesis of the gracilis muscles are aponeurotic and tendinous, the length is sufficient, the span is large, usually there is a functional contraction distance ranging from 12 to 15 cm, the length of the vascular nerve pedicles is sufficient, and it is suitable to be anastomosed with the vessels at the donor sites. In addition, the main function of the gracilis muscle is to maintain the body balance in walking. As there are many muscles with the same functions, those with few influences on the donor site functions and with insignificant scars after the resection should be selected; therefore, the gracilis muscle is undoubtedly the most ideal choice. However, it should be noted that the blood supply of the skin flaps carried on the gracilis muscle is not reliable, especially the lower 1/3; this might be correlated with the few perforating branches whose vascular net enters the skin. The gracilis muscle starts from the pubic body, and some ischiadic ramus in the form of aponeurosis is distributed at the posterior side of the great adductor muscle; the upper 2/3 of it is the muscle belly, the lower 1/3 of it is the long muscle tendon, and the posterior medial side at the knee joint ends at the medial surface of the upper end of the tibia. The gracilis muscle is dominated by obturator nerve (L2/L3), this nerve (it is a motor nerve) can be divided into two bundles, each bundle can independently dominate part of muscle bundles, and this is of great help to the construction of the independent flexion functions of the thumb and other fingers. The accompanying obturator vessels mainly supply the proximal muscles (the nerves enter the muscle tissues at the proximal end of the vessels); another main blood supply source is the branch of deep femoral artery, which runs between the long and short femoral arteries and mainly supplies the distal blood of muscles. In free grafting of functional muscle, some conditions must be satisfied: (1) The bone structures must be stable and reliable, and the elbow, wrist, and hand joints should have the range of motion close to normal. (2) The estimated site of tendon sliding, namely, the distal 1/2 of the gracilis muscle, should have good soft tissue coverage and healthy sliding connection. (3) The hands should have good sensory functions.

(4) What is the most important? There should be intact motor nerves and arteries and veins that provide anastomosis, the distance between the nerves and muscles should be as short as possible, and in this way the time of neuranagenesis can be shortened.

The patient is taking the supine position with the lower limb abduced, the tendon of the long adductor can be touched in the anterior inferior region of the pubic bone, and the connection line between this spot and the long adductor muscle nodules of the femoral internal condyle is the posterior margin of the long adductor and also the anterior margin of the gracilis muscle. Please remember the long adductor is an important locating marker to seek the gracilis muscle and the great saphenous vein often runs in the prezone of the gracilis muscle. This connection line can serve as the surgical approach for the resection and harvesting of the gracilis muscle or the anterior margin of the muscular skin flap (the skin flap should be designed to be at 2/3 of the thigh, and the skin perforating vessels should be protected), and during the operation, it should not be confused with the long muscle and sartorius muscle. In resection and harvesting of the muscular skin flap at the proximal end, the performing of forward separation can find the obturator nerve vascular bundles, the traction of long adductors can indicate that the branches of the obturator nerves and deep femoral artery can obliquely run from the lateral side to the medial side and enter the gracilis muscle, and the projection position is about 1/4 of the thigh. At most 6–8 cm of the vascular pedicles can usually be resected and harvested, and the outer diameter of the artery is 1.5–2 mm. There are many vascular plexuses at the starting point of the vessels, the separation should be careful, and in the meantime, the accompanying veins should be protected. The motor nerves of the gracilis muscle can be carefully separated, and reverse ascending exploration can be performed until the position near the foramen obturatum is reached, and then they are disjuncted. After the pedicles are prepared, downward separation can be performed at the

lateral side of the gracilis muscle. Before resection and harvesting of the muscles, the suture line is needed to mark the length of the muscles. When the hip joint is in the abduction position and the knee joint is in the extension position, the marker line is sutured every 5 cm for the reference when the tension is adjusted in the donor sites. Before disjunction of the muscles, the enthesis of the donor sites and the nerve vessels for anastomosis should be prepared. In resection and harvesting of initial point, separation should be performed carefully at the superficial layers of the periost at the lower branch of pubic bone, the pubic body, and the ramus inferior ossis ischii to harvest the aponeurosis and paratenon; in the disjunction of tendons at the terminal site, properly protect the para-tendinous membrane and the sarcolemma at the muscular migration, and these sliding structures are of very vital importance to the prevention of postoperative adhesion.

During the surgery, reduction of muscular ischemia time as much as possible is of great importance. Before resection and harvesting of the muscles, the tourniquet is often released to let the blood perfuse for over half an hour so as to reduce the muscular ischemia time. Clinical practice indicates that the ischemia time within 3 h is within the allowable range. The anastomosis of nerves and vessels is started after the muscular enthesis is repaired at the donor sites, and usually 10-0 or 11-0 microscopic suture line is used. The repair of nerves and vessels must be perfect, meaning the thrombus cannot occur after the angiostomy. In case of embolism, immediate exploration should be performed; otherwise, disastrous consequences will arise. The nervous anastomosis should be accurate to bundle membrane suture, and there should be no fibrous tissues inside the nerves. Take the transplantation and repair of the forearm as an example. The gracilis muscle appears to be slightly long, nonabsorbable surgical sutures are generally used to suture the initial point to the humeral supracondylar ridge, the tendons at the donor sites are woven and sutured to the muscular parts of the gracilis muscle tendons for adjustment, and the space of the marking points of the muscle belly is re-elongated to 5 cm. Vascular anastomosis is easy no matter if it is end-to-end anastomosis or end-to-side anastomosis. When the vascular length is insufficient, this problem can be solved through pretransplantation of veins; in case of mismatch of the bores, the methods invented by Dr. Tsai et al. can be adopted to address the problems. After the completion of the surgery, the tendons must be covered by intact skin soft tissue or skin flaps, and in the meantime, the transplanted muscles do not bear extra tension. After the operation, external fixation is performed in the flexion position (the relaxation position of transplanted muscles), ligation is performed with external dressings, windowing is opened subsequently, the patient is asked to lie in bed for rest for 5–7 days with the affected limb slightly elevated, and attention should be paid to the intake and output volume of the liquid; ensure that the environment is warm and the perfusion is sufficient, and, if necessary, anticoagulant therapy plan should be given. After 3-week braking, physiotherapy and rehabilitator training are started gradually, and the rehabilitation stage usually lasts for 6–12 months. Usually, at 2–4 months after the operation, the muscles regain the innervation with palpable muscle contraction. At this time, the patients should be encouraged to do active muscle contractions for training to achieve good tendon sliding and overcome the adhesion so that the maximal muscular strength can be achieved. Multiple literature reports indicate that, at 2–3 years after the operation, the muscular strength of most patients can return to about 20% of the normal level and, under the best situation, it can return to 50% of the normal level, and the effects on adults are better than those on children [3, 5, 10] (Figs. 14.20, 14.21, and 14.22).

14.6.2 Broadest Muscle of the Back

For a wide range of soft tissue defects and functional disorder, free grafting of functional muscles can be considered. This muscle has sufficient strength, resection and harvesting are easy, and it can carry long nerve vascular pedicles; when

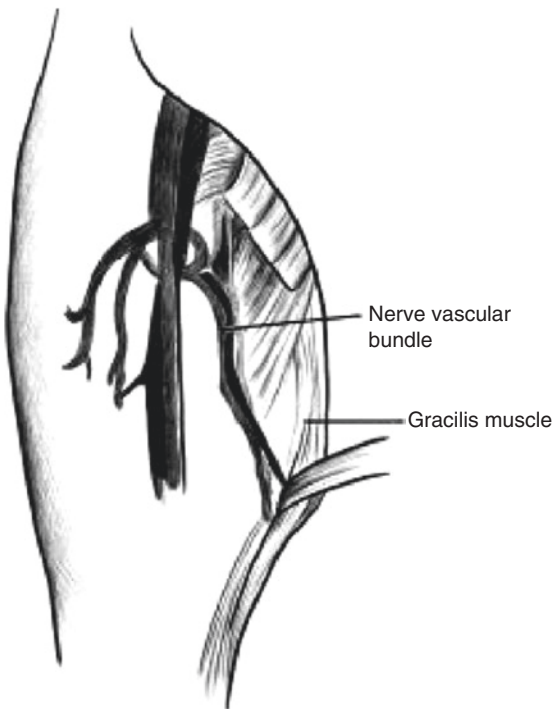

Fig. 14.20 The main nerve vascular pedicles of the gracilis muscle enter the muscular tissues at the upper 1/4 position of the thigh and consist of the medial branch of the femoral circumflex artery and the anterior branch of the obturator nerves

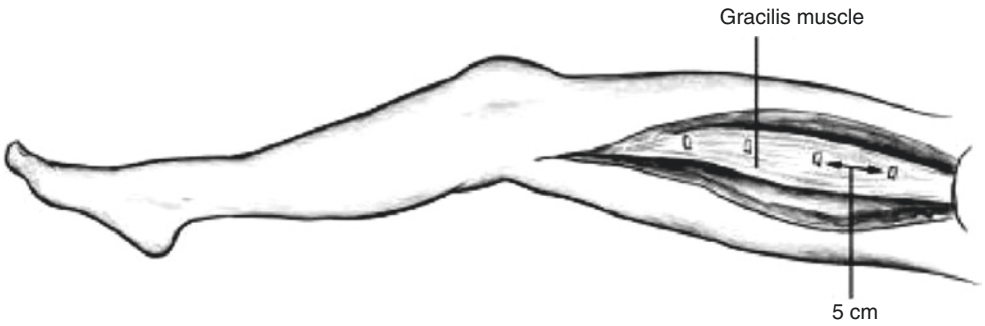

Fig. 14.21 The length (5 cm) before the gracilis muscle transplantation is confirmed using a marker line

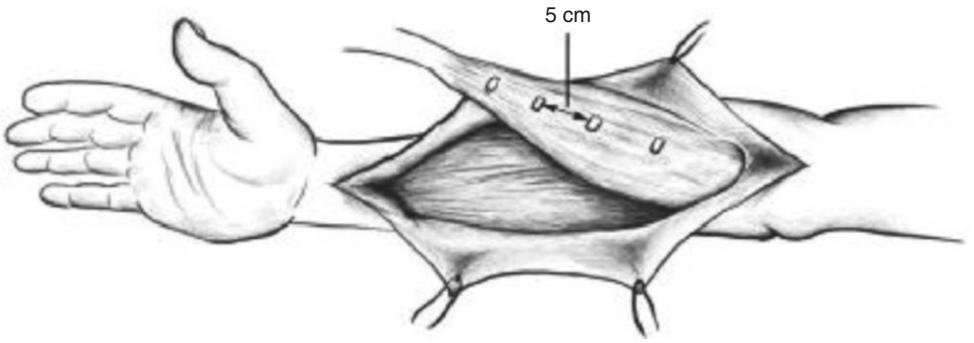

Fig. 14.22 After the gracilis muscle transplantation, the proximal end is sutured and fixed; then the muscle tissues are pulled to recover the marking point of the original length (5 cm), and then it is fixed

carrying skin flaps, the blood supply is stable and reliable, and the scars at the donor site are not obvious; after the resection, the influence on the upper limb functions is not obvious. However, this muscle flap has obvious disadvantages: it is the fat muscle type; the length of muscular fibers is not equal; the muscular flap becomes swollen; there is no obvious aponeurosis at the initial point; the transplantation, construction, and suturing are difficult; and there is no long muscle tendon for sliding. After a comprehensive balance, this muscular flap is not the first choice.

14.6.3 Gastrocnemius Muscle

Some surgeons proposed using the gastrocnemius muscle to perform free grafting of functional muscles, and good effects have been produced. This muscle has many advantages: the aponeurosis at the initial point is obvious, the muscle tissues are powerful, each half has single nerve vascular pedicles, and they can be harvested together with the skin flaps on them. The disadvantage is as follows: If the shank at the donor site is not treated properly, the appearance is very ugly.

Overall, as for the selection of free tissues for repair of upper limbs during childhood, repair performed during this period can produce good effects; and there is no obvious difference between children and adults in terms of the rate of success. However, to successfully complete the children's microsurgical operation, the following aspects should be considered: whether the appearance of the donor site and recipient site is satisfying with the child's development, whether hospital departments meet the conditions of its performance (e.g., professional nursing team and pediatric anesthetists), the postoperative analgesia, and the future phase II surgery. The main complication of repair of free tissue flaps is still complete or partial necrosis of the tissue flaps, the children's microsurgical operation has higher requirements on the entire medical team, and the surgeons should try their best to avoid the poor blood circulation of tissue flaps and develop a set of surgical techniques applicable to the infants so that the purpose of treatment can be served.

14.7 Toe-to-Hand Free Grafting

Toe-to-hand free grafting occupies an important part in the history of microsurgery. Up to now, there are many specific methods that are derived from it, but in the field of children's congenital deformities, especially adactylia and brachydactylia, this method is not widely applied because surgeons, parents, and pediatric patients have different understandings of this surgery. The indications are the absence of thumb and finger and the functional loss induced by longitudinal and transverse dysplasia, and the rate of success is usually higher than 95%. As for transverse dysplasia, the histological structures at the proximal end, such as skeletons, nerves, vessels, muscles and tendons, are basically normal, and the toe-to-hand free grafting can reconstruct the

needed functions; as for longitudinal dysplasia, the development of the histological structures at the proximal end is usually poor or absent, and it is necessary to graft the needed tissues from the adjacent finger rays for the construction and repair. The thumb reconstruction is the main indication of toe-to-hand free grafting. In addition, it can reconstruct other fingers than the thumb and the common fingers are the middle finger and the ring finger. The ultimate goal of treatment is to establish the relatively complete grasping functions, including the lateral pinching, the three-finger pinching, and the capabilities to grip large objects. Some pediatric patients or parents have higher requirements on the hand appearance; sometimes, the surgeons will consider the transplantation of toes or some toes to reconstruct the fingers, but generally multi-toe transplantation of one leg is not recommended currently because this will result in poor appearance in the donor site. The toe appearance after the transplantation of the second toe is the most acceptable; therefore, the transplantation of the second toe is relatively common. There are varying opinions on the age suitable for the transplantation, and analysis should be made according to the specific circumstances. In case of dysplasia in both hands, the surgical timing can be relatively late; in case of dysplasia in one hand, the surgical timing can be early. Otherwise, with the development of the infantile patient, the functions of the defected hand will be replaced by those of the normal hand; even if another toe transplantation is performed for the treatment, the surgical effects will be greatly reduced. The previous study materials indicate that the age between 2 and 5 may be a relatively good age period for the surgery. During this period of time, histological structures especially the vessels have further development, the rate of success of microsurgical operation is higher, the children in this age group can actively take rehabilitation exercises, and they can also have sufficient upper limb strength to do the exercise, so good postoperative effects can often be produced.

When there are only two fingers in the reconstructed hand, the hand can only hold an object like a pair of forceps and the appearance is not very pleasant; is it still necessary to replant the fingers? Even if the patients are still willing to undergo reconstruction, the carpometacarpal joint of thumb should have a good range of motion; if there is no good carpometacarpal joint, the hand functions cannot obviously improve even if one more finger is replanted. If the carpometacarpal joint of the residual thumb is good, it can do the circumduction but the thumb is dysplastic, the phalanges of toes can be adopted for free grafting (before the age of 18 months), or the bone lengthening can be used to consolidate and lengthen the thumb ray, and the ulnar ray finger is used to perform toe-to-hand free grafting for reconstruction to reconstruct the object-pinching functions such as finger opposition. On the contrary, if the thumb ray lacks the motor abilities (rigid thumb), grafting of thumb ray to the ulnar side can be considered, and the toe is then replanted to the thumb position, and the joint of the transplanted toe is used to enhance the object-pinching functions of the hand.

When the toe-to-hand free grafting for hand function reconstruction is considered, the objectives should be made clear and the design should be accurate to finally serve the purpose of enhancing hand functions, and such a surgery is worth considering. Two teams should be assigned to perform the surgery to respectively anatomize various histological structures of the hands and feet. To first ascertain the usable vessels, nerves and tendons are of vital importance. What structures should be used and how to harvest them are dependent on the specific conditions of the deformed hands in the operation. This is different from some microsurgical operations simultaneously performed by two teams. Usually, a transverse incision is designed at the dorsal carpometacarpal side. In this way, the structures such as nerves, tendons, and vessels can be clear at a glance after the incision, and the postoperative scars are not obvious. As for the extensor tendons, that with the longest sliding distance is often selected; when it is necessary to add another new tendon to make the replanted fingers have independent motor functions, it is proper to select the inherent extensor tendons of the index finger or the little finger. The palmer incision is designed into the Z-plasty incisions which go in the same direction as dermal ridges; the nerves, tendons, and vessels are sequentially distinguished; and if necessary, differentiation can be made at the level of the carpal canal. In addition to the anastomosis with the palmar artery for the toe transplantation, the anastomosis with the deep branch of radial artery at the nasopharyngeal fossa can be considered. The toe harvesting technique has been very mature. In the transplantation and repair of the deformed hands, the resected and harvested histological structures, range, and modes should be dependent on the conditions of the hand.

Take transplantation of the second toe as an example. A V-shaped incision is often made; the vessels are sought from far to near; the first finger web should be first explored; the first and second dorsal digital arteries should be determined; the common initial point, dorsal metatarsal arteries, and perforating branches at the sole are found at the proximal end; and which vessel should be used is determined according to the vascular bore and distribution. If possible, the dorsal metatarsal arteries should preferably be used, and exploration should be ascended to the proximal end. The arterial accompanying veins should be selected, and the reflux is reliable. If it is necessary to retain one extensor tendon, the one that can effectively extend the interphalangeal joint should be selected. The branch of the anterior tibial nerves should be close to the dorsal metatarsal arteries and should be retained inside the transplanted toe. It is necessary to resect and harvest the proximal end of the toe nerves at the toe pulp after

the intra-nerve separation so that sufficient length can be obtained. After the traction near the metatarsophalangeal joint tendon sheath, the flexors are resected and harvested according to the needs. The osseous disjunction planes may be mutilated at the levels of metatarsal bone and metatarsophalangeal joint or the level of proximal phalanx according to the needs of donor sites, and they are usually mutilated at the metatarsophalangeal joint. There are many methods to fix the bone tissues after transplantation: The single longitudinal Kirschner silk fixation has few injuries to the epiphyseal plates, the operation is easy, and the fixation effects have been sufficient because the infants have a very quick union of bone tissues; dual steel silk can be used for crossing and fixation and the epiphysis should be avoided. After the skeletons are fixed, the extensor tendons should first be repaired, and the extensor tendons are usually repaired near the level of wrist joint. If conditions permit, two extensor tendons are repaired; for the second one, the inherent extensor tendon of the index finger or the little finger can be used for repairing to increase the independent motor abilities of the fingers. When the tendons are sutured, the extensor tendons should have a certain tension to give full play to the roles of tendons. When the repair of the flexor tendon is difficult, the flexor system always suffers dysplasia, the effectively sliding distance is insufficient, and the hand functions after replantation is affected. The sutured flexor tendon is fixed reliably using the Tsai's six-coil method, the tension of the flexor tendon should not be too large, the adjustment can be made in the future, and usually repair of one profound flexor tendon is enough. As many nerves as possible should be anastomosed under the microscope because good nerve distribution can harvest good hand functions. The vascular anastomosis methods and techniques have been described a lot, and no more will be discussed here.

As for the transplanted histological structure, its growth and development problems should be considered; in case of no epiphyseal plate in skeletons, some of the transplanted bone tissues will not grow longitudinally, which should be clearly considered for children. In addition, the warm ischemia time of the transplanted toes should be reduced as far as possible, the epiphyseal plates are sensitive to ischemia, and long ischemia time will affect the development of the transplanted toes. Absorbable sutures are usually adopted for skin suturing, and the edge distance should be as small as possible with little suturing. If the closed wounds have tension, thin split-thickness skin grafts can be used to cover the wounds. The postoperative 24-h observation and nursing are of great importance, and sedatives and continuous regional anesthesia can be given for anesthesia. Four weeks after the operation, the steel needles can be pulled out and external stabilization splints can be provided. During the daytime, external fixation devices are taken off, and active and passive functional exercise is done under the guidance of parents, and during the night, the devices are worn. The final range of motion of the transplanted toes is unexpected, usually the range of passive movement is larger than the range of active movement, and DIP usually has a very small range of active movement; PIP lacks the extending abilities and is eventually manifested as fixed flexion deformity. The lysis of adhesions of tendon almost has no effect in improving the functions of transplanted toes.

In most cases, the transplanted toe serves hand grasping functions through serving as a fixed support. Whether the transplanted toe has certain growth capabilities is dependent on multiple factors including surgical techniques, ischemia time, stable blood supply, mode of bone connection, nerve degeneration, and infection. However, the transplanted toes can be finally fused into the functional units of the hands and produce good effects [11–14] (Figs. 14.23 and 14.24).

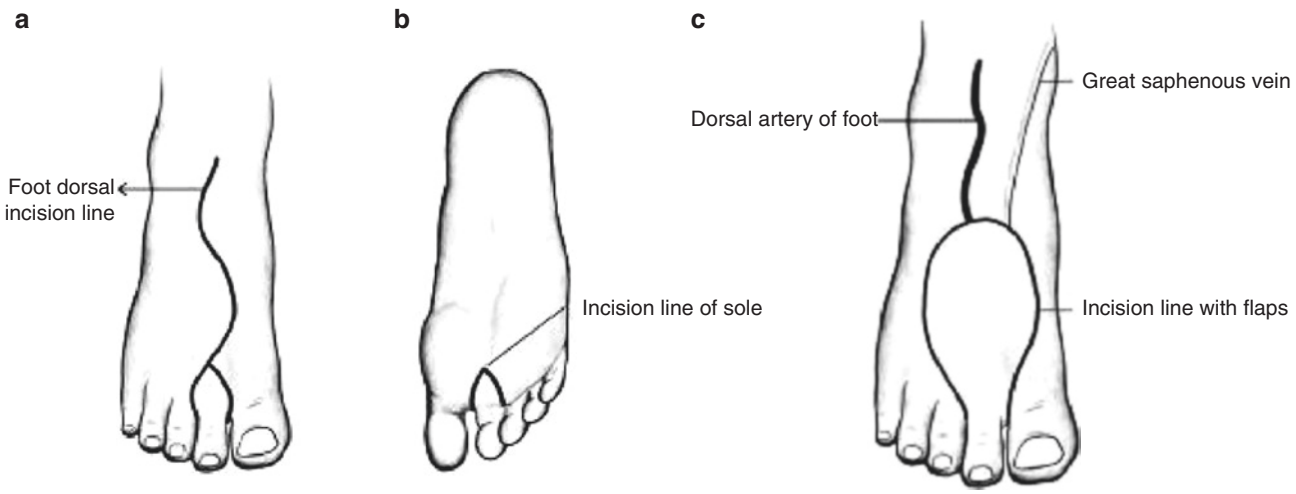

Fig. 14.23 Surgical design of toe-to-hand free grafting of the second toe. (**a**) Incision line and resection range of the foot dorsum. (**b**) Incision line and resection range of the sole. (**c**) Incision line and resection range when the second toe contains some foot dorsal skin flaps

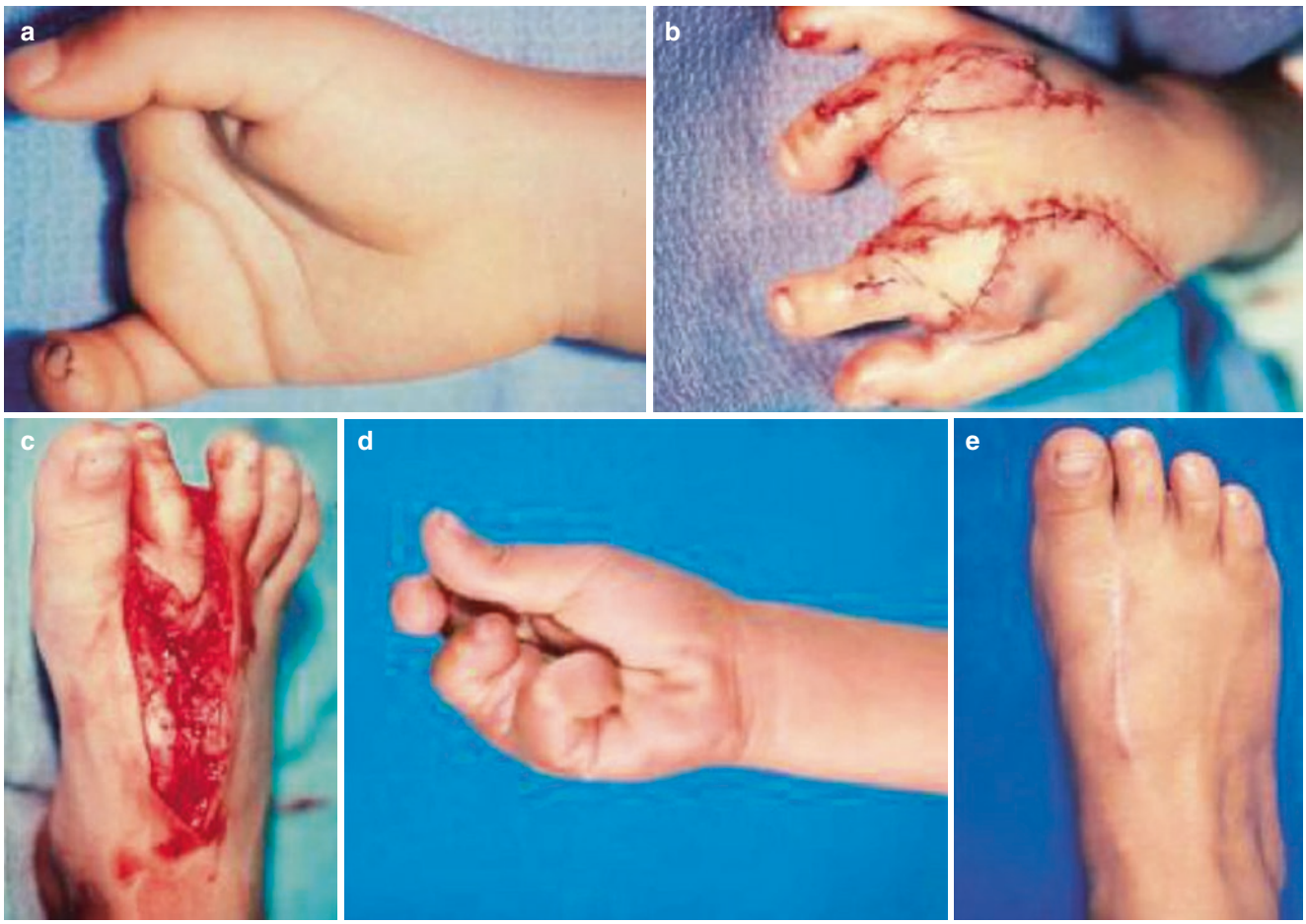

Fig. 14.24 Congenital absence of index, middle, and ring fingers as well as little finger dysplasia and bilateral toe-to-hand free grafting of the second toe for the reconstruction of index finger and ring finger. (**a**) Hand appearance before operation. (**b**) Immediately after operation. (**c**) Resection range of the second toe. (**d**) Normal hand functions after operation. (**e**) Insignificant scars at the foot donor site and good appearance

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Genetic Research of Hand Congenital Deformities and Advancement in Plastic and Reconstructive Treatment

15

Jinghong Xu, Yang Wang, and Jianmin Yao

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15.1 New Progress in Studies of Pathogenesis and Etiologic Factors of Congenital Hand Deformities

Jinghong Xu and Yang Wang

15.1.1 Embryonic Development of Upper Limbs

Limb bud is composed of mesodermal mesenchyma or mesodermal center and one layer of dermal layer on the external surface, and it is the initial phase of upper limb development. The development of limb bud is the result of gradual changes of various signaling molecular mechanisms in three-dimensional space, and the three axes are proximo-distal, anteroposterior, and dorsoventral, which correspond to the shoulder-finger direction, thumb-little finger direction, and forearm-palm direction in limb morphology. In order to guarantee the correct development and growth of limb bud, three categories of cell groups are of vital importance: they are apical ectodermal ridge (AER) at the lateral side of the limb bud, the progress zone (PZ) at the medial side of the limb bud, and the zone of polarizing activity (ZPA) at the posterior side of the limb bud. The signaling molecules generated by the cells in these regions decide the growth directions of the adjacent cells so that the normal growth and development of upper limb is maintained (Fig. 15.1).

During the development process from the proximal end to the distal end, the formation of limb is subject to the dual influences of the embryonic lateral plate and the somatomic mesoderm [1]. Soon after the formation of limb bud at two sides of the embryo, the cells from the lateral margins of the adjacent somite invade the limb bud, forming the muscles, nerves, and vessels of the limb. The limb bud grows gradually, and various skeletal factors are arranged on the *Y*-axis. Different signal centers control the growth and formation of limb from three directions (Fig. 15.2). The growth of proximal and distal axes of the limb is subject to the control of the

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covering ectoderm, and it forms a special epithelial structure at the tip of the end—AER. In the experiment, the removal of AER at different time points can result in mutilation deformities to varying extents.

The formation of the anteroposterior part of the limb is subject to the regulation of the polarizing activity zone (ZPA), and ZPA consists of mesenchymal cells at the margin of the posterior part of the limb bud. The main functions of ZPA decide the number and structure of fingers. As is verified by the studies, the sonic hedgehog (SHH) signals regulate the target genes through controlling the balance between Gli3 inhibitors and activators, and it is the key pathway in ZPA regulation.

The ectoderm except the AER regulates the formation of limb dorsoventral part, including the upper limb muscles, tendons, and finger nails. Studies indicate that the dorsal part of the limb bud is controlled by genes WNT7a and LMX1, and the ventral part is controlled by gene En1.

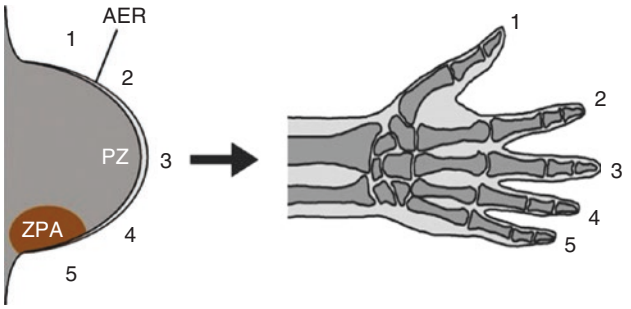

Fig. 15.1 The AER, PZ, and ZPA cell groups in the limb bud; *1–5*, respectively, represent the developmental regions of five fingers (Reprint with permission from Pascal H.G. Duijf, Hans van Bokhoven, Han G. Brunner, Pathogenesis of split-hand/split-foot malformation, 2003)

15.1.2 Progress of Studies on the Molecular Mechanism of Congenital Hand Deformities

AER, ZPA, and PZ constitute the main structure of limb bud, and they are of vital importance to the formation and differentiation of the limb bud. Currently, the known key factors that regulate the growth of limb bud include SHH signaling molecules, WNT signaling molecules, fibroblast growth factor (FGF), bone morphogenetic protein (BMP), and homeobox (HOX)-related protein. The recent studies mainly focus on the following aspects.

15.1.2.1 WNT Signaling Pathway

WNT signaling pathway is a highly conserved signal pathway widely present in the multicellular eukaryotes and plays an important role in the development of embryonic limb. WNT signaling pathways include typical signal pathway and atypical

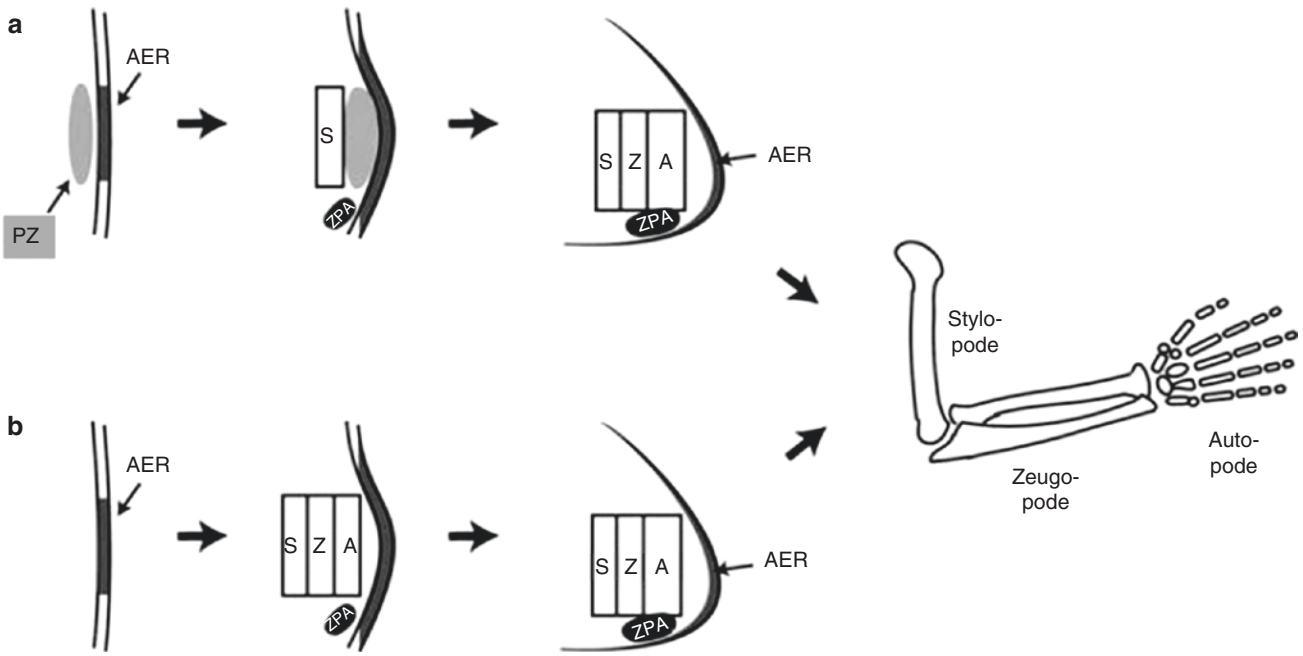

Fig. 15.2 Two possible modes of limb bud development (Reprint with permission from G. C. Schwabe, S. Mundlos, Genetics of Congenital Hand Anomalies, Handchirurgie · Mikrochirurgie · Plastische Chirurgie, 2004). (**a**) The mesodermal mesenchymal cells first leave the progress zone (PZ) to become differentiated and sequentially form the proximal end (S), the middle end (Z), and the distal end (A) of the limb. (**b**) During the early stage, the mesodermal mesenchymal cells first become differentiated to form the proximal end (S), the middle end (Z), and the distal end (A) of the limb and then multiply and grow into complete upper limb

signal pathway. In the typical WNT signaling pathway, the ligand protein binds with the Frizzled receptor family on the cell surface to activate the key component (dishevelled, DSH) of cell membrane-related WNT receptor compound, and DSH is separated from Frizzled receptor to inhibit the downstream proteins, including Axin, GSK-3, and APC protein. Axin/GSK-3/APC compound can promote the degradation of intracellular signaling molecule β-catenin. After the β-catenin degradation compound is inhibited, the β-catenin inside the cytoplasm stably exists; some β-catenins enter the cell nuclei and act with the Tcf/Lef transcription factor family to promote the expression of specific genes (Fig. 15.3).

R-spondins are secretory ligands that bind with the WNT/β-catenin surface cell receptors and start this signal path. Its functional defects will result in serious dysplasia of embryonic limb. The four proteins of R-spondins (Rspo1~4) are, respectively, encoded by Rspo1~4 genes. Currently, some studies show that the homozygous mutation of Rspo 2 will lead to defects in anterior limb phalanges and finger nails as well as abnormalities in finger shape [2], and the simultaneous mutation of Rspo3 and Rspo2 will lead to more serious mutilation deformities [3].

Mammals have four kinds of Tcf/Lef genes, namely, Tcf1(Tcf7), Tcf3(Tcf7L1), Tcf4(Tcf712), and Lef1, which

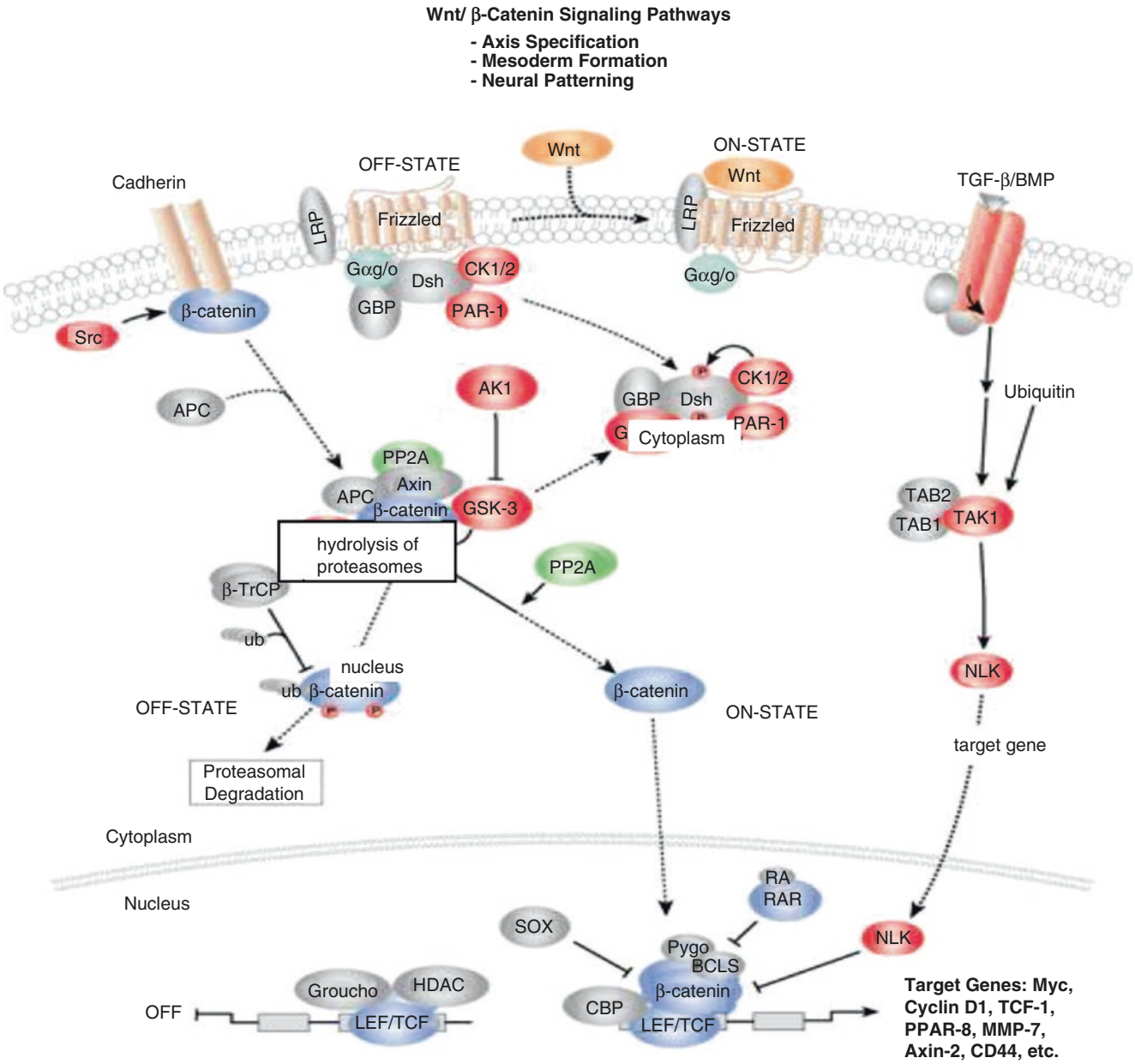

Fig. 15.3 Typical WNT signaling pathway (Reprint with permission from Stanley Neufeld, Jessica M. Rosin, Anshula Ambasta, Kristen Hui, Venessa Shaneman, Ray Crowder, Lori Vickerman, John Cobb, A conditional allele of Rspo3 reveals redundant function of R-spondins during mouse limb development. Genesis, 2012)

have almost the same DNA binding domain and can bind on the same sequence (A/TA/TCAAAG) to further regulate the transcription of target genes. After the β-catenin enters the cell nuclei, it will bind with Tcf/Lef to turn its transcription inhibition effects into transcription activation effects. Current studies reveal that Tcf3 mainly serves as a kind of transcription inhibitor, this effect does not need the binding with the β-catenin, and the Tcf3 defects will lead to the overexpression of WNT signals to further induce hyperdactylism; however, after Tcf3 binds with β-catenin, the transcription inhibition effect will disappear, and it is of vital importance to the limb growth and the formation of posterior fingers after the formation of gastrula [4].

Kremen1 (Krm1) and Kremen2 (Krm2) are the transmembrane coreceptor of Dickkopf1 (Dkk1, the inhibition factor of WNT/β-catenin signal pathway). As the regulating factor of WNT, Kremen protein of mammals plays the role of signal switch, and the formation of ternary complex that contains Krm, Dkk, and Lrp5/6 (coreceptor of WNT protein) inhibits the WNT/β-related signal pathway. Recent studies have revealed that Krm bi-mutation can present enhanced WNT signaling pathway, with AER expansion and finger deformities at the posterior side of upper limb, and an increase in bone volume and bone formation parameters; the Krm1(−/−) Krm2(−/−) Dkk1(+/−) tri-mutation can present the hyperdactylism at the posterior side of the limb [5, 6].

In addition, in the currently found WNT protein, some can produce endogenous β-catenin, while some do not produce β-catenin stimulating signals. Multiple WNT proteins including WNT5a and WNT11 transduce the signals through other modes, and this signal is called atypical WNT signal. Different from typical signals, atypical WNT signaling pathways regulate the cell polarity and intracellular Ca^{2+} functions through multiple branches. Recent studies have revealed that, during limb skeletal development, WNT5a can bind with transmembrane protein receptor ROR2 to activate the planar cell polarity (PCP) pathway so as to promote skeletal development; the mutation of Vang 12, a gene that encodes PCP pathway, will induce serious phalangeal absence [7], and the mutation of WNT5a and ROR2 is the direct cause of brachydactyly type B1 (BDB1) and Robinow syndrome [8].

15.1.2.2 Hedgehog Signal Pathway

SHH belongs to the intracellular signal protein hedgehog (Hh) family; it regulates the cell growth through dose-dependent effects and plays an important role in finger growth, formation, and morphogenesis of the upper limb. Many signal mechanisms of Hh are highly conserved between different species. After the synthesis of hedgehog protein, the N-terminal will rupture under the action of autocatalysis; in the meantime, the C-terminal has a covalent bonding with the cholesterol and is secreted outside of the cells in an active form. Hedgehog protein binds with its receptor Patched (Ptc, forming a kind of compound with seven-pass receptor Smo, thereby blocking the downstream signal pathway) to release the receptor binding protein Smo and further activate the transcription factors Ci/Gli, serine/threonine protein kinase Fused (Fu), Fu inhibitor (SuFu), motor-like protein Costal-2 (Cos2), protein kinase A (PKA), and other downstream signals so that a series of biological effects can be produced (Fig. 15.4). Ci/Gli and Fu produce positive regulation effects, and Cos2 and PKa produce negative regulation effects. Gli family protein members are large multifunction transcription factors and belong to C2H2-type zinc finger structural protein.

Studies show that, during the development of limb bud, the spatial expression range of SHH is only limited within the range of ZPA range, and this expression mode is subject to the remote control of cis-regulatory element ZRS on the upstream; this regulating mechanism has the following manifestation: in the ZPA internal region, ZRS promotes SHH expression through binding with GABPα/ETS1; in the external region of ZPA, ZRS inhibits the expression of SHH through binding with ETV4/ETV5 [9]. In many patients with skeletal deformities in the upper limb, the point mutation of ZRS sequence can always be found, including preaxial polydactylism 2 (PPD2), triphalangeal thumb-polysyndactyly syndrome (TPTPS), and type IV syndactylia (SD4). The reasons are as follows: this type of point mutation makes the SHH abnormally expressed at the anterior part of the limb bud, further inducing the disorder in the regulation of SHH on limb bud development. Other studies reveal that [10] there is a segment of highly conserved sequence in the upstream of the SHH encoding sequences,MFCS1, containing the cis-acting element that regulates SHH expression; the function is to limit the expression of SHH only at the ZPA. In case MFCS1 experiences mutation, the affinity between the mutant and the hnRNP U will significantly increase, further influencing the SHH expression and inducing hyperdactylism. In addition, it has been believed that SHH signal is only expressed in ZPA; however, other studies reveal that [11] Hh signal pathway is also present in the ectosermic AER and regulates its growth; the knockout of Smo in AER will seriously affect the formation of upper limb fingers, leading to the ossification of supernumerary cartilages.

Gli zinc finger protein is the intracellular transcription factor in the Hh signal pathway, and SHH, after activating the Gli factor, can promote the expression of target genes. In the mammals, Gli2 and Gli3 play a key role during the formation of anterior-posterior axis of the limb bud. Recent studies reveal that [12] Gli3 can directly limit the expression of factors (e.g., cdk6) that regulate the transition of cells from phase G(1) to phase S to further prevent anterior hand plate digit progenitors from entering into phase S; in the meantime, it can promote the differentiation of proliferative

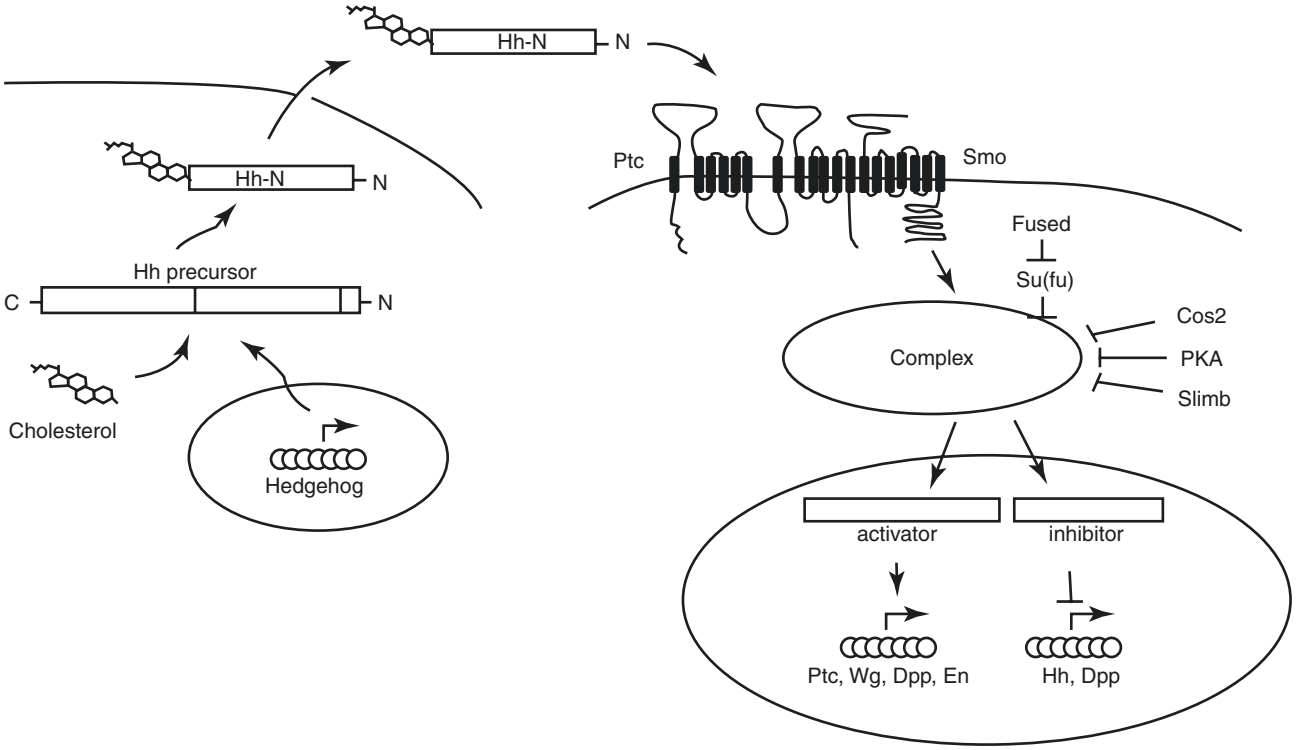

Fig. 15.4 Hedgehog signal pathway (Reprint with permission from Motoko Aoki, Hiroshi Kiyonari, Harukazu Nakamura, Hitoshi Okamoto, R-spondin2 expression in the apical ectodermal ridge is essential for outgrowth and patterning in mouse limb development, 2007)

precursor cells to cartilages through temporally and spatially inhibiting Gremlin1 (the antagonizing factor of BMP) to further guarantee the structures and number of normal fingers. In case of mutation in Gli3, preaxial and postaxial hyperdactylism as well as congenital dysplasia such as Pallister-Hall syndrome [13] or Greign head-hyperdactylism-syndactylia deformity will be induced. Some scholars find that Zic3 (a neuroglioma-related oncogene homologen superfamily member) can regulate the expression level of Gli3 and SHH and further regulate the number of limb fingers. The inhibition of Zinc 3 in Gli 3 mutational mice will significantly improve the deformity degree [14]. In addition, during the recent years, a new negative regulatory factor Tulp3 (tubby-like protein 3) has been found, and this protein is preliminarily found in the nervous system development. As is verified, Tulp3 can react with the Gli3 in the downstream of SHH pathway to inhibit the intensity of Hh signal [15]. The animals without Tulp3 have neural canal defects and hyperdactylism, and the reason is that the inhibition effects disappear and further induce the abnormal enhancement in the Hh signals [16].

Cilia are a highly conserved microtubular structural cell organ and it projects the cell surface onto the centrosome. The main function of cilia is mediation of cell movement and sensor functions, such as light, gas, and protein ligand. Recent studies show that [17], during the growth of limb bud, cilia can mediate the transformation of SHH signals through regulating the processing of Gli transcription factors in it. Researchers find that [18] the Meckel-Gruber syndrome (accompanied by hyperdactylism) is induced by the following reasons: the Mks1 (del64-323) gene experiences mutation; the cilia cannot be located on the centriole, so that the planar cell polarity is disturbed; and the Hh signals decrease. In addition, the products encoded by Ift172 gene are part of the complex that mediates the intraflagellar transport (IFT) in the cilia. Some studies reveal that [19, 20] the mutation of Ift172 gene will lead to cilia dyspoiesis and decrease in SHH signals and induce serious hyperdactylism. In addition, orofacial-digital type I (OFD I) has always been considered to be induced by the cilia formation disorder arising from Odf1 gene defects; however, recent studies have indicated that [21] the serious hyperdactylism accompanied by preaxial-postaxial finger absence and long-bone shortening induced by Ofd1 gene defects has no clear relation with the cilia formation disorder but is mainly correlated with the gradual decline of SHH signals and the decrease in Gli3 activity. The defects of limb extension are induced by IHH signal defects and ossification disorder in the intrachondrial bone formation.

15.1.2.3 Tp63 Gene

Tp63 gene is one of the necessary genes that maintain AER growth. It has been verified that Tp63 mutation is the molecular basis that induces ectrodactyly-ectodermal dysplasia-

cleft lip/palate syndrome (EEC syndrome) or limb-mammary syndrome (LMS). The replacement of the highly conserved amino acid in the DNA binding domain of Tp63 induced by this mutation affects the specificity and affinity of DNA binding [22], to further result in the decrease in the interstitial cell proliferation and the increase in apoptosis [23]. In addition, other studies indicate that P-cadherin gene (CDH3) is the transcription target gene of Tp63 and plays a key role in the formation of hair and hair follicles as well as the AER growth of human limb buds [24].

15.1.2.4 Homeobox Gene

Homeobox is the conserved motif shared by the homeotic gene. The genes with homeobox are called homeobox genes, and they are collectively known as homeobox family gene. Nearly all of these genes are correlated with development regulation. Human homeobox genes contain four groups of 39 genes (HOXA-HOXD), and they determine the limb shape and structural characteristics of formed bones during the development of limb bud. HOXD13 is of vital importance to the formation of limb tips, but the mechanism controlling their growth remains unclear. Studies conducted in recent years reveal that, when the limb anteroposterior formation is involved [25], HOXD13 binds on the Hand2 site; when the limb proximal-distal axes are involved, it binds on the Meisl and Meis2 sites; when skeletal formation is involved, it binds on the Sfrp1, Barx1, and Fbn1 sites, and additionally it binds on the Emx2 site of Dach1, Bmp2, and Bmp4 genes. The wrong expression of HOXD13 changes the expression of most of the above genes, further leading to synpolydactyly (SPD). HOXA gene mutation is considered to be the main cause of hand-foot-genital syndrome. Recently, some scholars have found that [26] this mutation is mainly the absence of the sequence with a length of 5.6 Mb in the chromosome 7p15 to p15.3 regions (including HOXA gene and adjacent development genes), further leading to insufficient dosage of HOXA13 and affecting the normal embryonic growth and development.

15.1.2.5 T-Box Gene

T-box (TBX) gene family belongs to the developmental regulation-related transcription factor family and has over 20 members. Their common characteristics are as follows: they have the transcription factors that encode the highly conserved N-terminal DNA binding domain (T region), have important development regulation functions, and play an important role in the morphogenesis and histogenesis of vertebral and non-vertebral animal embryos. Recent studies indicate that [27] TBX 5 gene mutation will lead to a significant increase in the transcriptional level of mesenchymal cell apoptosis-related genes (bad, bax, and bcl2) and cell cycle-related genes (cdk2, pcna, p27, and p57) and result in the enhancement in embryonic cell apoptosis to cause such congenital deformities as Holt-Oram syndrome. In addition, TBX gene is also the action targets of some drugs such as white atractylodes rhizome and retinoic acid, and these drugs can lead to ulnar finger absence, humeral and radial bone shortening, ulna-breast syndrome, and congenital micromelia through lowering the expression [28, 29].

15.1.2.6 Twist1 Gene

Twist1 gene is a transcription factor that contains basic helix-loop-helix (bHLH); it can bind on the Nde1 E-box element in the form of a dime and activates or inhibits its target gene. Some diseases induced by heterozygous mutations of human Twist1 gene include Saethre-Chotzen syndrome [genuine acrosphenosyndactylia]. Studies have revealed that [30] the Twist1 key residue phosphorylation changes their abilities to form active dimers; in addition, the DNA combining power of Twist1 dimer can be influenced through a type of cis-acting element-dependent behaviors. These changes are small but complicated. They not only directly influence the transcription process of Twist1 but also influence the transcription results of any bHLH factor (can form dimeric factors with TwiSt1). In addition, Twist1 also involves the epithelial-mesenchymal transition (EMT) and tumor cell metastasis process, and relevant mechanism should be further expounded [31].

15.1.2.7 Sister Chromosome Adhesion-Related Proteins

Esco2 is a type of acetyl transferase and it is necessary for the sister chromosome adhesion. This adhesion is mediated by a type of multi-subunit complex called cohesin (it is located at the chromosome through cell cycle protein NIPBL). Under the action of acetylization of Esco2, the binding between cohesion and chromosome is stable during phase S. Cornelia de Lange syndrome (CdLS) and Robertson syndrome (RBS, reflex iridoplegia) are both induced by the mutation in the proteins needed by sister chromosome stickiness, CdLS is induced by the heterozygous mutation of NIPBL or cohesin subunits SMC1A and SMC3, and RBS is induced by the homozygous mutation of Esco2. However, the manifestations of the two syndromes are different, and the reasons are that the different downstream genes of cohesion are affected, and Rad21 of downstream gene of cohesion is manifested as obvious transcription factor enrichment, and the downstream genes regulated by Esco2 (Esco2 regulates the downstream gene through cohesion, but it is not Rad21) mainly involve cell cycle or apoptosis, and the high-level cell apoptosis produces the shape with Esco2 deletion, the characteristics similar to RBS, mitosis defects, facial deformities, and amputation deformities [32]. In addition, other studies reveal that Esco2 inhibits the transcription activity of Notch (one cell differentiation inhibiting signals)

protein through the method nondependent on transacetylase to further promote cell proliferation; in case of excessive expression of Esco2, the differentiation of P19 embryonal carcinoma cells and C17.2 nerve precursor cell neurons will be promoted [33].

15.1.2.8 Others

In addition, some congenital hand deformities and relevant syndromes can be induced by abnormalities of specific molecular mechanisms; these syndromes include:

1. Coffin-Siris syndrome: Coffin-Siris syndrome (CSS), also called the fifth digit syndrome, is induced by insufficient ARID1B expression [34].
2. Adams-Oliver syndrome: Adams-Oliver syndrome (AOS), also called scalp and cranial absence and defects and congenital scalp defects with limb distal amputation, is the shortening and mutation in the DOCK6 gene that encodes one type of atypical guanidine exchange factor (GEF), which leads to the impossible formation of actin cytoskeletons in cells [35].
3. Mesomelia-synostoses syndrome: Mesomelia-synostoses syndrome, also called limb middle part shortening, distal synostosis, and repeated congenital deformities, is induced by deletion with a length of 582–738 kb in the chromosome 8q13. The two genes, SULF1 and SLC05A1, in the absent sequence play an important role in human skeletal development and cardiac and cerebral development [36].
4. Spondylocarpotarsal synostosis syndrome: Spondylocarpotarsal synostosis syndrome, namely, short stature and fusion of vertebrae, carpal bones, and tarsal bones, is a syndrome induced by the lack of actin B in the cell substance due to the nonsense mutation of FLNB gene that encodes the filamin B. Filamin B is a type of pluripotent cytoplasmic protein and plays a key role in skeletal formation [37].

Wong et al. [38] find that BMP is a secretory signal protein which serves to regulate AER functions and inter-digital programmed cell death of limb during development. This effect is co-regulated by Smad1/Smad5. The bi-mutation of Smad1/Smad5 is manifested as decreased tissue and cell apoptosis of limb during the development, which further induces syndactylia.

Pontual et al. [39] find that the absence of MIR17HG gene of polycistron miRNA that encodes miR-17-92 will induce microcephaly, short stature, and finger deformities in addition to deafness, which indicates that miR-17-92 has regulatory effects on the growth and skeletal development.

Klopocki et al. [40] detect one deletion with a length of about 900 kb in the genome of patients with autosomal-dominant inherited brachydactyly type E (BDE), and it contains PTHLH gene [which encodes parathyroid hormone-related protein (PTHRP)]. PTHRP can promote cartilage cell proliferation and hypertrophic differentiation, and the devitalization of mouse PTHRP will lead to brachydactylia and microsoma. In further experiment, they get to know that the PTHLH in BDE contains two missense mutations (L44P and L60P), one nonstop mutation [X178WextX(*)54], and one nonsense mutation [K120X]. Missense mutation L60P is the main cause that leads to the functional loss of PTHLH.

Mucopolysaccharide hyaluronic acid is an important component of extracellular matrix, and it can directly affect the cell behaviors through interactions with cell surface receptor. Matsumoto et al. [41] conditionally inactivate the gene of hyaluronic acid synthetase (Has 2) and find that the skeleton and joint capsule suffer serious deformities in the limb of mutant, which result in deformities such as serious limb shortening and hyperdactylism.

P450 oxidoreductase (POR) of cytochrome is the specific electron donor of cytochrome P450 of all microsomes and can catalyze the metabolism of many exogenous and endogenous compounds, including retinoic acid, cholesterol, and polyunsaturated acids. The point mutation of POR has been recently found in the Antley-Bixler-like syndrome, and it leads to skeletal deformity. Schmidt et al. [42] knocked out the POR from the interstitial cells of mouse limb bud, and the results indicated that the anterior limb and posterior limb of the mouse embryo had shortening and skeletal thinning and joint fusion, and the occurrence to the anterior limb was earlier. In further studies, they found that the excessive content of retinoic acid and the lack of cholesterol in the POR-deficient embryos could induce the changes in limb growth, cell apoptosis, and skeletal separation.

15.1.3 Progress of Studies on the Etiologic Factors of Congenital Hand Deformities

The etiologic factors of congenital upper limb deformities are very complicated, and they are mainly classified into the genetic factor and environmental factor. The genetic factor includes chromosome abnormalities and gene mutation, and the progress on the latest studies of relevant mechanisms has been described before. The environmental factor is the teratogen, and it is the most apt to induce congenital deformities during the embryonic period. The common factors include ① biological factors; ② physical factors, such as ionizing radiation; ③ chemical factors, such as thalidomide and retinoic acid; and ④ other factors, such as anoxia, parent diabetes mellitus, chronic ethylism, and lack of nutrient, which can induce fetal deformities. The recent studies mainly focus on the following aspects.

15.1.3.1 Thalidomide

Thalidomide, commonly called Fanyingting, was once used as a type of sedative to relieve morning sickness, but it caused several thousand infantile cases of deformities such as amelia and micromelia in Europe especially in German from 1957 to 1961. This event also promoted the etiological and pathological studies on congenital deformities, but the teratogenetic mechanism still remains unknown at present, and anti-revascularization and oxidative stress are considered to be the two main mechanisms.

Recent studies have found that the main target of thalidomide is the cereblon protein. Cereblon protein can catalyze one type of E3 ubiquitin ligase complex that contains DDB1, Cu14A, and Roc1, and this complex is of vital importance to the expression of fibroblast growth factor 8 (FGF8), a key regulator of limb development. The binding between thalidomide and wild-type cereblon inhibits the functions of E3 ubiquitin ligase, then downregulates the expression of FGF8 [43] and leads to the changes of actin cytoskeletons and upregulation of insulin signals, downregulation of the vascular system development, and inflammatory reaction route, and interferes with the regulation process of embryonic shape, inducing multiple deformities [44] such as micromelia (amelia), limb bending, hyperdactylia, syndactylia, and brachydactylia. Knobloch et al. find that thalidomide inhibits the activity of redox-sensitive transcription factor NF-kB in the limb bud, changes the balance between FGF and Bmps, and makes it tend to be the pre-apoptotic Bmps [45]; through Bmps, it protects the active PTEN (phosphatase and tensin homolog deleted on chromosome ten, a product of anti-oncogene) from being degraded by protease [46], inhibits WNT/β-related protein and Akt-dependent cell survival signals, and makes the caspase-dependent apoptosis activated by the internal Fas apoptosis receptor routine, thus greatly reducing the number of cartilage precursor cells, resulting in poor bone formation and causing mutilation deformity.

15.1.3.2 Retinoic Acid

Retinoic acid (RA) is the metabolic intermediate product of in vivo vitamin A, mainly influences bone growth, and can promote epithelial cell proliferation, differentiation, and keratin dissolution. RA is a substance needed for the normal development of embryonic limb and will more or less induce limb deformities. Some studies reveal that [47] the RDH10 (the main enzyme that synthesizes vitamin A during the embryonic period)-deficient embryo will suffer upper limb developmental disorder and poor inter-digit tissue degeneration. The mechanism of too much RA leading to deformities still remains unknown. It is currently believed that RA can directly or indirectly modify the expression of one HOX gene with known changes to produce its effects [48], and it can also influence limb growth through promoting the expression of heat shock proteins (HSPs) [49]. It is recently found that azole antifungal agents will change the RA metabolism and lead to limb deformities [50]; CYP26 enzyme can degrade RA and prevent teratogenesis [51].

15.1.3.3 Cadmium

Cadmium (Cd) is a type of heavy metal, and the exposure to cadmium during the embryonic period will increase the incidence of adactylia in the anterior limb. Studies show that the exposure of pregnant women to cadmium will significantly upregulate the expression of glucose-regulated protein 78 (GRP78, a type of endoplasmic reticulum chaperonin), make the eIF2α in the placental PERK signal pathway phosphorylated, and increase the CHOP level, further increasing the cellular endoplasmic reticulum stress and inhibiting the growth of placental cells and promoting their apoptosis [52]. The results indicate that the endoplasmic reticulum stress mediated by reactive oxygen species may participate in the Cd-induced influences on the placental and embryonic growth. As a drug, antioxidants can be used to protect the Cd-induced embryonic deformities and growth [53]. Other studies find that the CdCI(2)-induced apoptosis is p53 (a type of anti-oncogene) dependent; it is p53-nondependent in AER, and this effect is irrelevant to the dose of p53 gene expression [54].

15.1.3.4 Valproic Acid

Valproic acid (VPA) is a widely used anticonvulsant. The use of VPA during the first trimester will increase the risk of congenital fatal deformities, including heart deformities, facial deformities, and skeletal limb deformities, but the mechanism still remains unclear, which may be correlated with oxidative stress. Recent studies have revealed that the use of VPA to treat the in vivo cultured embryos can significantly elevate the intracellular reactive oxygen species (ROS) [55]; the addition of peroxidase can weaken the VPA-induced ROS formation and apoptosis. There are also studies indicating that the reason for VPA-induced deformities may be that it inhibits the activity of histone deacetylase to further change the expression of relevant genes [56]; another reason is the inhibition of the activity of in vivo folic acid because the folic acid insufficiency will lead to the growth defect of AER [57].

15.1.3.5 Chlorcyclizine

Chlorcyclizine will lead to deformities of embryo such as cleft palate, high-vaulted arch, microrhinia, micrognathia, high head, adactylia, brachydactylia, or micromelia, and histamine H1 can antagonize this effect [58].

15.1.3.6 Cholesterol

Cholesterol is the precursor to synthesize steroid hormones and bile acid. It is very important to the synthesis of Hh signal protein, so cholesterol is also important to embryonic

development. After birth, cholesterol can be obtained through synthesis in the human body or through external intake; but before birth, it can only be obtained through synthesis in the body; therefore, all the factors that induce abnormalities in the fetal cholesterol synthesis will lead to developmental disorder, such as limb defect syndrome [59].

15.1.3.7 Anoxia

Anoxia is often induced by vascular injuries, can influence the normal cell surface ion exchange, and leads to osmosis imbalance, increase in free radicals, and increase in cell apoptosis. If the state of anoxia lasts for long, the hemoglobin F (HbF) will decrease, eventually leading to tissue necrosis and exerting serious influence on embryonic growth and development. The drugs that lead to embryonic anoxia include misoprostol, adrenalin, potassium channel-blocking agent, Dilantin, cocaine, and hydroxycarbamide, and the reasons why they induce anoxia vary. Misoprostol will induce poor embryonic angiopoiesis and lead to fetal blood circulation disorder [60]; cocaine, misoprostol, and serious shock will lead to uterine arterial temporary spasm; potassium channel-blocking agent will induce serious slow heart rate and arrhythmia, affect the heart ejection function, and subsequently lead to the hypoxia. This harm will be enhanced when Dilantin is involved [61]. In addition, in the patients with hereditary thrombocytosis, an increase in the thrombopoietin (TPO) will seriously affect embryonic vascularization and lead to congenital limb transverse developmental disorder [62]. Hydroxyurea (HU) will cause cells to produce ROS, and ROS will react with polyunsaturated fatty bimolecular film to produce 4-hydroxy-2-nonenal (4-HNE). The latter can lower the activity of glyceraldehyde-3-phosphate dehydrogenase (GAPDH), glutamic-oxaloacetic transaminase 2 and aldolase 1, and A isodynamic enzyme (three kinds of enzymes that involve energy metabolism) and lead to the decrease in cell energy production. In addition, HU can also induce the intranuclear transposition of embryonic GAPDH; this intranuclear transposition will initiate the cell apoptosis in the P53-dependent route and eventually lead to multiple fetal deformities [63].

15.1.3.8 Ionizing Radiation

Ionizing radiation is considered to be an intense teratogenic factor of embryo. Currently, there are studies indicating that, when the embryos are exposed to ionizing radiation, the overexpression of pre-apoptosis gene Bax will manifest chondriosome-mediated cell apoptosis. In the meantime, overexpression of MKK3 and MKK7 (members of stress MAP enzyme family) can also be observed. The latter gets involved in the ray-induced apoptosis through the activation of p38 and JNK pathways. In addition, the oafs induced by ionizing radiation also have obvious telomere shortening, and amniotic fluid detection indicates that the inflammatory reaction cytokine level is significantly improved. It can be found that apoptosis, inflammation, stress, and DNA injuries can be observed during the early stage after ionizing radiation; they can induce increase in the amniotic fluid cytokine level and shortening of the telomere, eventually leading to upper limb deformities [64].

15.2 Progress of Plastic and Reconstructive Treatment of Congenital Hand Deformities

Jinghong Xu, Yang Wang, and Jianmin Yao

There are various kinds of congenital hand deformities, the mutation is big and complicated by deformities in other sites, the conditions are very complicated, and the treatment method has always not been very satisfying. The main treatment principles are described according to the following classification description.

15.2.1 Poor Formation

The main manifestations of congenital poor formation are the local or complete absence of the limb, and certain methods need to be adopted to reconstruct the defective sites. As the soft tissue engineering is still in the middle of study, currently it is very difficult to fabricate an artificial tissue that can be used for transplantation. Therefore, in light of the currently limited medical technology, the usual practice is to adopt autologous or allogenic transplantation to reconstruct the defective sites, but the autologous bone grafting is often subject to the adverse reactions of the harvesting site and the limitation in the source of usable bones. The reconstructed fingers through the toes or the proximal fibular transplantation have the growth potentials, but relevant technologies are not perfect, and there is a great risk in failure. Allogenic transplantation has the problem of rejection, and after the operation, the patients may need to receive lifelong anti-immunologic rejection therapies.

As for the poor longitudinal formation, such as radial defects, osteotomy and tendon grafting and repairing surgery are usually adopted to improve the functions of the affected limb. In addition, the skeletal lengthening and tissue transplantation have been used to replace the absent bones, joints, or muscles. The results of one recent long-term follow-up study show that osteomere forearm surgery can effectively improve the long-term functions of the affected limb while avoiding autogenous bone transplantation [65]. In the poor transverse formation, during the recent period of time, some scholars attempt to use autogenous iliac growth plate for transplantation to treat the congenital wrist transverse defects

and have produced good effects [66]. In the treatment of congenital central cleft hand deformities, Oberlin et al. [67] adopted the anti-pollicization surgical method to repair the functions of index finger, grafting skin flaps from the hand dorsum to repair the defects so as to avoid the injury to the palm nerve vascular network. It has proven to be a safer surgical method.

15.2.2 Excessive Formation

For excessive formation, supernumerary tissues used to be removed to produce satisfactory therapeutic effects; sometimes, some extra surgeries are needed, such as rearrangement of tendons and corrective osteotomy to realign the residual fingers. The joint fusion can be used to reduce the supernumerary bones and joints in polydactylism. For the duplication of the thumb, the Bilhaut-Cloquet method and the modified operation method are used to combine two small fingers to reconstruct a stable thumb with a normal size [68].

15.2.3 Poor Differentiation

As for poor differentiation, such as palmature (longitudinal), syndactylia (transverse), and carpal bone fusion (mixed), separating surgery is always needed, but patients suffering from it are always accompanied by incomplete or deformed bones, joints, and muscles so that they need extra repairing surgeries.

Bone fusion can be defined as formation of fixed unmovable joints due to imperfect joint development. As we cannot reconstruct a completely normal joint that can slide, the joint junction site can only be separated, namely, increasing its mobility through fabricating a false joint. As the soft tissues around the false joint are not complete, the artificial false joints have the disadvantages of refusion, instability, and incomplete movement, and sometimes osteotomy is needed to obtain more functional shape. In the poor differentiation of soft tissues, such as congenital finger flexion deformity, the corresponding soft tissues (e.g., abnormal skin, mucosa, ligaments, or tendons) can be removed in case of no obvious functions.

In the syndactylia patients with Apert syndrome, Piza-Katzer et al. [69] placed one piece of silicone rubber sheet between two fingers immediately after syndactylous finger separation; 2–3 weeks later when the new vessels and tissues grew out, the silicone rubber sheet was removed, and full thick skin graft was used to cover the gap. This method combines the original multiple surgeries used to treat syndactylia patients with Apert syndrome and considerably reduces the times of surgery.

15.2.4 Overdifferentiation

The treatment principle for overdifferentiation is resection of superfluous tissues and consolidation of residual structures. Take congenital joint dislocation, for example. Some methods are always needed to remove the supernumerary tissues at the joints, and in the meantime the joint should be fixed until it becomes stable. When the joint suffers dysplasia or semi-dislocation, as this joint tends to develop early osteoarthritis or complete dislocation, attempts should be made to use the intra-articular or extra-articular osteotomy to reconstruct a more stable joint; when restoration is impossible, only arthroplasty or joint fusion can only be carried out to improve its functions.

15.2.5 Poor Growth

The limb manifestation of poor growth is that part of it has a small shape but the functions are close to normal; theoretically, only a little expansion is needed; however, it is very difficult to expand the affected limb to the normal level through current therapeutic therapies. When the needed length increase ranges from 1 to 2 cm, osteotomy can be performed, and transplantation of supporting bones for the internal fixation can be performed under the condition of traction; when the needed length increase is over 2 cm, external fixation and traction can only be performed to gradually elongate it, but the effects of these methods are not very satisfying.

For the toe transplantation used to treat congenital brachydactylia, longitudinal or arch-shaped incisions at the hand dorsum are always adopted; after the operation, spasm will often occur to the palm soft tissues and functional recovery will be affected. Recently, Sammut et al. [70] attempted to adopt palmar surface V-Y incision suturing and toe transplant to repair the brachydactylia, which increases the length of hand palmar surface soft tissues, lowers tissue tension, and thus reduces the incidence of postoperative flexion and spasm.

15.2.6 Overgrowth

The overgrowth such as fibroneuroma, hemangioma, lymphangioma, or arteriovenous malformations is sometimes accompanied by deformities in skeletons, joints, muscles, and ligaments. The treatment objective is to resect or compress the sites of deformities, including the normal and abnormal tissues, to reduce the length and volume and maintain the normal similar functions and appearance. However, it is very difficult to decrease the affected limb to the normal shape, and tissue necrosis will occur after the operation.

15.2.7 Structural Abnormality

The main treatment objective of systematic structural abnormalities is to extend the survival time and prevent the continued development of deformities. In the systematic structural abnormalities induced by hormones or enzymes, replacement therapy can reach treatment objective. In most cases, the etiologic factors of such diseases remain unclear. If the deformities are so obvious that they affect the daily walking or life, symptomatic treatment can be given to improve life, through supporting, fixing, or surgical methods. As for the local structural abnormalities, simple orthopedic procedure can be used to achieve the long-term treatment objective. For example, in the treatment of congenital middle phalangeal serious bending, Ali et al. [71] adopted the closed wedge-shaped orthopedic procedure and fully corrected the bending bone deformities; in the meantime, they repaired most of the functional and appearance abnormalities in fingers and produced good effects.

Although various current treatment methods have covered all the congenital upper limb deformities, the overall therapeutic efficacy is still not very satisfying. With the profound exploration on the genetic and molecular mechanism of upper limb growth and development, our understanding of the birth defects will be constantly improved, and the treatment methods will be constantly improved. We believe, in the near future, we can prevent and cure the diseases more effectively.

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