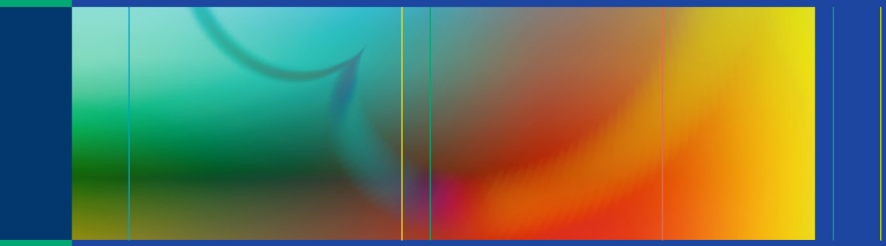
Prasad P. Godbole
Martin A. Koyle
Duncan T. Wilcox *Editors*



Guide to Pediatric Urology and Surgery in Clinical Practice

Springer

Guide to Pediatric Urology and Surgery in Clinical Practice

Prasad P. Godbole • Martin A. Koyle
Duncan T. Wilcox
(Editors)

Guide to Pediatric Urology and Surgery in Clinical Practice

Springer

Editors
Prasad P. Godbole
Department of Pediatric Surgery
and Urology
Sheffield Children's Hospital
Sheffield, UK

Duncan T. Wilcox
Department of Pediatric Urology
Denver Children's Hospital
University of Colorado at Denver
Denver, CO
USA

Martin A. Koyle
Department of Urology
and Pediatrics
University of Washington
and
Department of Urology
and Pediatrics
Seattle Children's Hospital
Seattle, WA
USA

ISBN 978-1-84996-365-7 e-ISBN 978-1-84996-366-4
DOI 10.1007/978-1-84996-366-4
Springer Dordrecht Heidelberg London New York

British Library Cataloguing in Publication Data
A catalogue record for this book is available from the British Library

Library of Congress Control Number: 2010933607

Cover design: eStudio Calamar, Figueres/Berlin

Printed on acid-free paper

Springer is part of Springer Science+Business Media (www.springer.com)

Preface

Primary care physicians (general practitioners or family physicians & in North America pediatricians and emergentologists) are often the first port of call for children with non-emergency, but potentially serious surgical or urological problems. During the training period as primary care physicians, while most clinicians will gain adequate exposure to general pediatric medicine, very few will obtain a sound knowledge or clinical experience of general pediatric surgical or urological problems that they may commonly encounter in their primary care environment. This ready reference book addresses these issues.

All chapters in this book give a synopsis of a particular condition, its management in primary care, indications for referral and timing of referral. Complications of a particular procedure and its management if presenting to primary care is also discussed. The format is standardized throughout the book with illustrations wherever appropriate. This will enable primary care physicians in providing a continued high quality of service to children with surgical problems.

The contents include all those pediatric and urological conditions that are commonly encountered in primary care and are divided region wise. The list of conditions included is based on referral letters received in a tertiary care setting over many years.

The editors thank the contributors spanning all continents in their efforts to enable this book to be published in a timely fashion. We are also grateful to Denise Roland and Melissa Morton at Springer, for their support and encouragement throughout the administrative process. And finally

once again, this book would not have been possible without the sacrifices of our families to whom we are deeply indebted.

Prasad P. Godbole
Martin A. Koyle
Duncan T. Wilcox

Contents

Contributors

Carolina Acuña, MD
Department of Pediatric Surgery
Hospital San Juan de Dios, and
Universidad del Desarrollo
Santiago, Chile

Angela M. Arlen, MD
Department of Urology, University of Iowa
Iowa City, IA, USA

Paul F. Austin, MD
Department of Surgery and Pediatric Urology Research
Washington University School of Medicine
St. Louis, WA, USA

J. Christopher Austin, MD
Division of Urology, Oregon Health and Science University
Portland, OR, USA

Özgü Aydogdu, MD
Department of Urology, Ankara University
Ankara, Turkey

Andrew Barker, FRACS MBBS
Department of Surgery, Princess Margaret Hospital
Perth, WA, Australia

Neil D. Bateman, BMedSci BM BS FRCS (ORL-HNS)
Sheffield Children's Hospital, Sheffield, UK

Spencer W. Beasley, MBChB (Otago)
MS (Melbourne) FRACS
Department of Paediatric Surgery
Christchurch Hospital, Christchurch
New Zealand

Adriana M. Behr, MSN RN CPNP
Urology Department, The University of Colorado/The
Children's Hospital, Aurora, CO, USA

David I. Campbell, MD MB BS MRCPCH
Department of Gastroenterology, Sheffield Children's
Hospital, Sheffield, UK

Christopher S. Cooper, MD
Department of Urology, University of Iowa, Iowa City
IA, USA

Joseph Ignatius Curry, MBBS FRCS (Eng)
FRCS (Paed Surg)
General Paediatric Surgery Department
Great Ormond Street Children's Hospital
London, UK

Tom P. V. M. de Jong, MD PhD
Pediatric Renal Center, University Children's Hospitals
UMC Utrecht and AMC Amsterdam,
The Netherlands

Prasad P. Godbole, FRCSFRCS (Paed) FEAPU
Department of Paediatric Surgery and Urology
Sheffield Children's Hospital, Sheffield, UK

Kenneth W. Gow, MSc MD
General and Thoracic Surgery Department,
Seattle Children's Hospital, Seattle, WA, USA

Mohan S. Gundeti, MD MCH FRCS (Urol) FEBU FEAPU
Urology Department, University of Chicago Pritzker School of Medicine, Chicago, IL, USA

Richard S. Hurwitz, MD
Department of Urology, Kaiser Permanente Medical Center
Los Angeles, CA, USA

Stephanie A. Jones, DO
Department of Pediatric Surgery, University of Colorado
Denver, CO, USA

Martin A. Koyle, MD FAAP FACS FRCS (England)
Department of Urology and Pediatrics
University of Washington, and
Department of Urology and Pediatrics, Seattle Children's Hospital, Seattle, WA, USA

Michael C. Large, MD
Department of Surgery, University of Chicago
Chicago, IL, USA

Pedro-Jose Lopez, MD
Pediatric Surgery and Urology, University of Chile, and
Department of Pediatric Urology
Hospital Exequiel Gonzalez Cortes & Clinica Alemana
Santiago, Chile

Sean S. Marven, MBChB, FRCS (Paeds)
Department of Paediatric Surgery, Sheffield Children's Hospital, Sheffield, UK

Travis J. McKenzie, MD
Department of Research, Mayo Clinic, Rochester
MN, USA

Andrew Neilson, MB ChB MRCS
Surgical Paediatrics Department, Royal Hospital for Sick Children, Glasgow, UK

Stuart O'Toole, MD FRCS (Paeds)
Surgical Paediatrics Department, Royal Hospital for Sick Children, Glasgow, UK

Robert T. Peters, BSc (Hons) BM (Hons) MRCS (Eng)
Paediatric Surgical Unit, Sheffield Children's Hospital Sheffield, UK

Garrett D. Pohlman, MD BS
Division of Urology, University of Colorado Health Sciences Center, Aurora, CO, USA

D. Dean Potter, MD
Department of Pediatrics, Mayo Clinic, Rochester MN, USA

Christian Radmayr, MD PhD
Urology Department, Medical University Innsbruck Innsbruck, Austria

Naeem Samnakay, FRACS MMedSci MBBS
Department of Surgery, Princess Margaret Hospital Perth, Western Australia, Australia

John A. Sandoval, MD
Pediatric Surgery Department, St. Jude Children's Research Hospital, and
College of Medicine of the University of Tennessee Health Science Center, Memphis, TN, USA

Roly Squire, MBBS FRCS (Eng) DCH
Department of Paediatric Surgery, Leeds Teaching Hospitals NHS Trust, Leeds, UK

Marianne A. W. Vijverberg, MD
Pediatric Renal Center, University Children's Hospitals
UMC Utrecht and AMC Amsterdam,
The Netherlands

Ashish Wakhlu, MS MChir (Paediatric Surgery)
Paediatric Surgery Department, CSM Medical University
Lucknow, India

Alan R. Watson, MB ChB FRCP (Ed) FRCPH
Children's Renal Urology Unit, Nottingham University
Hospitals, Nottingham, UK

Duncan T. Wilcox, MD MBBS
Department of Pediatric Urology, Denver Children's
Hospital, and University of Colorado,
Denver, CO, USA

Mark R. Zaontz, MD
Department of Urology, Temple University School
of Medicine, Philadelphia, PA, USA

Part I
Pediatric Urology

Chapter 1
Urinary Tract Infection: USA

Angela M. Arlen and Christopher S. Cooper

Key Points

- UTI must be suspected in febrile infants.
- Catheterized or suprapubic urine specimens are superior, but mid-stream clean catch samples can be obtained in toilet-trained children.
- Routine follow-up culture is not necessary if organism is sensitive to originally selected antibiotic.
- Neurogenic bladders that are frequently instrumented will have colonization; urine culture should be obtained only if child is symptomatic.
- Evaluation of febrile UTI includes renal US and VCUG.

1.1 Introduction

Diagnosis, treatment and management of urinary tract infections in the pediatric population remain controversial. Pediatric UTIs are common and constitute a significant health burden; it is estimated that 7% of girls and 2% of boys under age 6 will be diagnosed with a urinary tract infection. In addition to appropriate treatment, the goal of managing UTIs in children should be to identify and modify factors that may affect renal parenchyma and function.

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_1,

1.2 Risk Factors

1. Age: prevalence of bacteruria is higher in first year of life than other times during childhood.
2. Race: Caucasians are more likely than Hispanic or African-American children to develop UTI.
3. Uncircumcised foreskin in males 6 months of age or younger increases the incidence of urinary tract infection ten times compared to circumcised males.
4. Incomplete bladder emptying or infrequent voiding: stasis allows growth of bacteria that would normally be flushed out with voiding.
5. Difficulty relaxing pelvic floor during voiding.
6. Constipation/encopresis: approximately 1/3 of children with recurrent UTIs have associated bowel problems.
7. Neurogenic bladder (managed with catheterization):
   - Bacteruria and pyuria occur in most children.
   - *E. coli* strains in this population commonly reflect colonization.

1.3 Presentation

1. Infants: in febrile infants, a urine specimen must be obtained even if signs point elsewhere; UTI accounts for 4.1–7.5% pediatric fevers
2. Young children (<2 years): fever, irritability, poor appetite, vomiting and diarrhea
3. Older children: dysuria, suprapubic pain, urgency, frequency, incontinence and abdominal/flank pain

1.4 Diagnosis

1. Examination of urine:
   - Infants and young children: a catheterized specimen should be obtained.
   - Toilet-trained children: a mid-stream, clean-catch urine specimen can be obtained but contamination can be difficult to rule out.

- A bagged specimen is only useful if negative; if positive, a catheterized specimen should be obtained. Bagged urine specimens are unreliable and unacceptable for diagnosis of UTI in high-risk population and infants younger than 2 months.
- Positive leukocyte esterase or nitrite with five or more white blood cells and presence of bacteria on microscopy is highly predictive of infection.

2. Urine cultures with greater than 10^5 colony-forming units are considered positive; however, if the urine is obtained in a sterile fashion, fewer colony-forming units can constitute an infection.

1.5 Common Pathogens

1. *Escherichia coli* (54–67%)
2. *Klebsiella* (6–17%)
3. *Proteus* (5–12%)
4. *Enterococcus* (3–9%)
5. *Pseudomonas* (2–6%)

1.6 Treatment

1. Infants younger than 90 days:
   - Consider IV antibiotics (ampicillin and gentamicin vs. third generation cephalosporin), especially with infants 30 days and younger.
   - Need for admission depends on clinical status.
2. Young children (<2 years):
   - Outpatient therapy with parenteral third generation cephalosporin (once daily).
   - If less ill and capable of taking oral fluids, treat with 7–10 days of antimicrobials with broad genitourinary pathogen coverage.
   - No follow-up culture is required if organisms are sensitive to selected antibiotic.

3. Older children: if uncomplicated UTI, oral broad-spectrum antimicrobial agents are well-tolerated and clear the infection after a 3–5 day course if there is no known genitourinary pathology.

1.7 Imaging

All children with culture-documented UTI and fever greater than 38.5°C should have follow-up imaging to determine any anatomic anomalies.

1. Retroperitoneal ultrasound: should include evaluation of kidneys and bladder and can be obtained at any time after the infection
   - Children younger than 5 years of age with febrile UTI.
   - Males of any age with first UTI.
   - Females younger than 2 years of age with first UTI.
   - Children with recurrent UTIs.
2. Voiding cystourethrogram (VCUG): should be obtained after confirmation that infection has cleared
   - Children younger than 5 years of age after febrile UTI.
   - Males of any age after first UTI.
   - Females younger than 2 years of age with first UTI.
   - Children with recurrent urinary tract infections.
3. DMSA renal scan: differentiate true pyelonephritis and severity of infections
   - Useful in high risk patients for prognostic purposes.
   - Renal scarring is a risk factor for future scarring.

1.8 Indications for Referral

1. Febrile UTI in infant
2. UTI + anatomic anomaly on US or VCUG
3. Recurrent urinary tract infections
4. Voiding dysfunction unresponsive to timed voiding and bowel regimen

Suggested Reading

1. Bauer R, Kogan BA. New developments in the diagnosis and management of pediatric UTIs. *Urol Clin North Am*. 2008;35:47-58.
2. American Academy of Pediatrics, Committee on Quality Improvement, Subcommittee on Urinary Tract Infection. Practice parameter: the diagnosis, treatment, and evaluation of the initial urinary tract infection in febrile infants and young children. *Pediatrics*. 1999;103:843-852.
3. Dairiki Shortliffe LM. Infection and inflammation of the pediatric genitourinary tract. In: Wein AJ, Kavoussi L, Novick AC, et al., eds. *Campbell-Walsh Urology*, vol. 4. 9th ed. Philadelphia: Saunders; 2007: 3232-3268.
4. Hinds AC, Holmes NM. Urinary tract infections in children. In: Baskin LS, Kogan BA, eds. *Handbook of Pediatric Urology*. 2nd ed. Philadelphia: Lippincott Williams & Wilkins; 2005:58-68.

Chapter 2
Urinary Tract Infection: United Kingdom

Andrew Neilson and Stuart O'Toole

Key Points

- Urinary stasis due to anatomical or functional obstruction predisposes to UTI.
- Dysfunctional voiding is common, but underlying anatomical abnormalities of the urinary tract should be considered and excluded.
- Anatomical abnormalities can predispose to renal scarring, which can lead to hypertension, renal impairment or renal failure.
- UTI should be considered in all febrile infants and children where another cause is not clearly established.
- A renal tract ultrasound scan is the first line investigation after diagnosis of UTI but it can miss vesico-ureteric reflux and renal scarring.
- Try to prove the diagnosis of UTI before undertaking more invasive investigations, and target such investigations to those most at risk.

2.1 Introduction

Urinary tract infection (UTI) is common in childhood. Three percent of boys and eleven percent of girls have had at least one UTI before their 16th birthday. In the first

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_2,

3 years of life the sex related incidence is approximately equal, but thereafter UTI becomes more common in girls. Correct diagnosis, treatment and subsequent targeted investigation of UTI in children is important because of the associations between UTI, underlying urological abnormalities, subsequent progressive renal damage and associated hypertension.

2.2 Pathogenesis

- The mode of infection in most cases is bacterial ascent into the urinary tract from below.
- The risk of infection is elevated if there is urinary stasis because organisms are cleared less efficiently from the urinary tract. Urinary stasis can be due to anatomical factors (anatomical obstruction, vesico-ureteric reflux), or functional interference with bladder emptying (neuropathic bladder, dysfunctional voiding, constipation).
- Dysfunctional voiding in girls, often in association with constipation, is a common clinical picture.
- The causative organism is *Escherichia coli* in the majority of cases.
- Other causative organisms include *Proteus vulgaris*, *Klebsiella*, *Enterobacter*, and *Pseudomonas*.
- Certain host factors modulate the risk of UTI. Breast feeding reduces the risk. Prematurity increases the risk, as does the presence of a foreskin in males.
  - In normal boys, circumcising 111 boys would prevent one UTI.[1]
  - In boys with severe vesico-ureteric reflux, the number needed to treat (NNT) to prevent one UTI is 4.[1]
  - In boys with posterior urethral valves, the NNT to prevent one UTI is less than one, therefore circumcision is recommended in these boys.[2]
  - Circumcision should be considered in boys with "at risk" urinary tracts, and those having recurrent UTIs.

2.3 Establishing the Diagnosis

Clinical features:

- The clinical features of UTI vary with age[3] (Table 2.1).
- A high index of suspicion is important, especially with pre-verbal children.
- UTI should be considered in any unwell infant and any child with failure to thrive or prolonged jaundice.

Upper tract or lower tract infection:

- Differentiating upper from lower tract infection can be difficult.
- The absence of fever does not preclude renal scarring, and children who have afebrile UTIs should also be treated and investigated.[4]
- Features suggestive of upper or lower tract infection are shown in Table 2.2.

Table 2.1. Clinical features of UTI in infants and children.

Age group	Most common	Less common	Least common
<3 months	Fever Vomiting Lethargy Irritability	Poor feeding Failure to thrive	Abdominal pain Prolonged jaundice Hematuria Offensive urine
>3 months			
Preverbal	Fever	Abdominal pain Loin tenderness Vomiting Poor feeding	Lethargy Irritability Hematuria Offensive urine Failure to thrive
Verbal	Frequency Dysuria Nausea	Dysfunctional voiding Changes to continence Abdominal pain Loin tenderness Fever	Malaise Vomiting Hematuria Offensive urine Cloudy urine Suspected sexual abuse Hypertension

Modified from NICE guideline: UTI in children, 2007[3]

Table 2.2. Clinical features used to distinguish upper tract from lower tract infection.

Upper tract features	Lower tract features
Temperature >38°C	No systemic features
Loin pain	Frequency
Upper abdominal pain	Urgency
Malaise	Nocturia
Vomiting	Secondary enuresis
	Dysuria
	Hesitancy
	Supra-pubic pain

Differential diagnosis:

- One fifth of children with appendicitis have urinary symptoms.[5]
  - Most children with appendicitis have some abnormality on dipstick urinalysis.[6]
  - The risk of perforation is elevated and the length of stay is prolonged in children who have received antibiotics for suspected UTI before a correct diagnosis of appendicitis is made.[6]
  - The clinical signs of appendicitis become less obvious after antibiotics have been given.[7]
- Foreskin problems in boys and vulvo-vaginitis in girls can mimic lower urinary tract infection.

The urine sample:

- A urine specimen should be collected and analyzed in all children with fever and in afebrile children with urinary symptoms.[8]
- Older children can provide a mid-stream urine sample into a sterile bowl if clearly instructed. A clean catch sample can be obtained from younger children with patience and perseverance on their parents' part. Catheter samples should be considered, but they may give false positive results. Bag samples have unacceptably high false positive rates. Samples from cotton wool or gauze placed in the nappy should be avoided.[9]

- In unwell, hospitalized infants where empirical treatment is going to be commenced before culture the result is available, the sample should be obtained by supra-pubic aspiration (which should be ultrasound guided).
- If immediate culture cannot be performed, samples can be stored for up to 24 h at 4°C, or in a tube containing boric acid.

Dipstick urinalysis:

- Testing for protein and blood is not useful in the diagnosis of UTI. These abnormalities are frequently present with other pathologies (e.g., appendicitis).
- Testing for nitrites and leukocyte esterase is useful. Not all dipsticks test for the presence of these so beware if someone else runs the test and tells you the urine is "clear." Did they test for nitrites and leukocyte esterase?
- When testing for nitrites, it is important to test a fresh urine sample.
- Nitrites are the product of bacterial conversion of nitrates. Many gram-positive cocci are not capable of this conversion, so give a false negative.
- Leukocyte esterase is an indirect marker of pyuria, but this too can be caused by conditions outside the urinary tract (e.g., appendicitis).
- If urine is positive for both nitrites and leukocyte esterase UTI is likely.
- If urine is negative for both nitrites and leukocyte esterase, UTI can be excluded and other causes of symptoms should be considered. The exception is children under 3, in whom a sample should still be sent for culture.
- Table 2.3 summarizes recommended actions to be taken based on clinical features and dipstick findings in children aged 3 years and older.

Urine microscopy and culture:

- Microscopy performed on fresh, unspun urine can reveal the presence of motile bacteria and the presence of pyuria. Significant pyuria is defined as >10 white cells/mm^3.

Table 2.3. Recommended actions based on dipstick findings in children aged 3 years or older

Dipstick urinalysis findings		Treat as a UTI with antibiotics?	Send urine for culture?	Comment
Leukocyte esterase positive	Nitrite positive	Yes	Yes[a]	[a]If high or intermediate risk of serious illness, or previous UTI
Leukocyte esterase negative	Nitrite positive	Yes[b]	Yes	[b]If fresh sample was tested
Leukocyte esterase positive	Nitrite negative	Only if clinical evidence of UTI	Yes	May indicate infection outside urinary tract which may need different management
Leukocyte esterase negative	Nitrite negative	No	No	Explore other causes of illness

Modified from NICE guideline: UTI in children, 2007[3]
a and b refer to the corresponding sentence in the comments column

- Pure growth of $>10^5$ bacterial colony forming units per ml is the traditional criteria used for diagnosis of UTI, though this was based on adult series.
- On SPA samples, any growth of gram negative bacteria, or >500 gram positive colony forming units per ml is significant.
- The necessity of performing culture in all cases has recently been questioned. However, in children, the benefit of confirming the diagnosis, and identifying the causative organism and its sensitivities should outweigh cost concerns.

2.4 Acute Management

- The choice of antibiotic, route of administration and duration of course are all controversial, and few studies have found significant differences when comparing regimes. Many units have their own local protocols; other units follow national guidelines.

Which antibiotic:

- Local patterns of sensitivity and resistance should guide your practice.
- Trimethoprim remains effective against many common organisms.
- Many organisms are resistant to amoxicillin alone, but are sensitive to co-amoxiclav.
- Aminoglycosides or cephalosporins are appropriate alternatives.

Route of administration:

- IV is preferred in early infancy because infants are at the greatest risk of renal scarring.
- In older infants and children, oral antibiotics are appropriate so long as they are tolerated.
- Initial IV therapy followed by subsequent oral therapy once the child is tolerating enteral feeds may be required.

Duration of course:

- Short courses are appropriate where lower tract infection is suspected.
- Longer courses should be prescribed where upper tract infection is suspected.
- Any systemically unwell child should have a longer course.

Failure of initial management:

- Parents or carers should seek medical advice if their child fails to respond to treatment within 24–48 h. They may be on the wrong antibiotic, or the wrong diagnosis may have been made initially.
- If there is failure of treatment whilst on the correct antibiotic, an intervention to drain the infected urine may be required. In bladder outlet obstruction and vesico-ureteric reflux this can be achieved by urethral or suprapubic catheter insertion. In VUJ or PUJ obstruction, a nephrostomy may be required.

2.5 Once the Diagnosis Is Established

The urological history:

1. Voiding history: volume, frequency, stream, urgency, incomplete emptying
2. Fluid intake: volume, type
3. Bowel habit: constipation, dietary information
4. Drug history: prophylaxis, breakthrough UTI on prophylaxis, laxatives
5. Antenatal history: any abnormal antenatal scans
6. Family history: renal disease

The urological examination:

1. Abdomen: renal masses, palpable bladder, fecal loading
2. Anus: inspection for position and fissure if constipation or fecal loading

3. Spine: any stigmata of spina bifida occluta, palpable sacral abnormality
4. Lower limbs: neurology
5. Blood pressure

Radiological investigations:

- Investigation of UTI places a burden on the child, their family and health care services. However, investigation of UTI in children allows:
  1. Identification of anatomical anomalies that require treatment
  2. Identification and documentation of scarring & damage to the kidneys
  3. Identification of dysfunctional voiding that predisposes to further UTI
- Renal tract ultrasound should routinely include pre-void and post-void images if the child is able to comply.
- Ultrasound can demonstrate gross renal scarring, obstruction, high grade vesico-ureteric reflux and dysfunctional voiding. It fails to detect lower grades of vesico-ureteric reflux, and lesser degrees of renal scarring.
- DMSA is the gold standard investigation for renal scarring. It also measures differential function. Scarring is more likely to be present in children who have had upper tract infections, recurrent infections, and those with a family history of vesico-ureteric reflux. Remember, it is difficult to differentiate upper from lower tract infection in younger children.
- Clinicians can target DMSA scans to those most at risk.
- In children under 2 years of age who have a single febrile UTI, ultrasound and cystogram alone are poor predictors of long-term renal damage. A DMSA scan is therefore recommended in this group. It should be considered in older children.[10, 11]
- Children over a year of age who have a normal DMSA and normal ultrasound do not require cystogram investigation for reflux because any reflux in these children is likely to be low grade, is likely to resolve spontaneously, and is unlikely to cause subsequent renal scarring.

- A catheter cystogram is the gold standard investigation for vesico-ureteric reflux, though it is invasive and may be poorly tolerated.
- An indirect cystogram using MAG3 can also detect vesico-ureteric reflux in children able to void upon request, although low grade reflux may be missed on this investigation. The MAG3 also measures differential function.

2.6 Long Term Management

Underlying abnormalities:

- A small number of children with history of UTI have an underlying urinary tract abnormality, some of which will require follow-up or intervention.

Modifiable risk factors:

- It is important to identify and manage poor fluid intake, constipation and dysfunctional voiding.
- This is the most important conservative measure in prevention of recurrent UTIs in children.

Prophylactic antibiotics:

- Which children should receive prophylactic antibiotics is controversial.[12]
- The objective of their use is to prevent renal scarring associated with UTIs.
- Prophylaxis should be considered in all children who have had a UTI, especially those who experienced upper tract symptoms.
- Prophylaxis is often continued until investigations are complete.
- Children with an underlying renal tract abnormality may continue on prophylaxis for several years.
- The timing of a trial off antibiotics often involves an element of parental preference, and may be postponed until the child is toilet trained.

Other measures:

- Cranberry juice has been shown to be as effective as antibiotic prophylaxis in children with vesico-ureteric reflux, though large studies are lacking.[13]
- Probiotics may have a role in prophylaxis of UTIs in children, but robust evidence is lacking.

References

1. Singh-Grewal D, Macdessi J, Craig J. Circumcision for the prevention of urinary tract infection in boys: A systematic review of clinical trials and observation studies. *Arch Dis Child*. 2005;90:853-858.
2. Mukherjee S, Joshi A, Carroll D, Chandran H, Parashar K, McCarthy L. What is the effect of circumcision on risk of urinary tract infection in boys with posterior urethral valves? *J Pediatr Surg*. 2009;44:417-421.
3. National Institute for Health and Clinical Excellence. Urinary tract infection in children: diagnosis, treatment and long-term management. Royal College of Obstetricians and Gynaecologists; National Collaborating Centre for Women's and Children's Health. 2007.
4. Coulthard MG, Lambert HJ, Keir MJ. Do systemic symptoms predict the risk of kidney scarring after urinary tract infection? *Arch Dis Child*. 2009;94:278-281.
5. Jones PF. Active observation in management of acute abdominal pain in childhood. *BMJ*. 1976;2:551-553.
6. Neilson A, Newland A. Prevalence of abnormalities on dipstick urinalysis in children with appendicitis and the adverse effects of misdiagnosis as urinary tract infection. Presentation, BAPS & EUPSA combined congress, Graz. 2009.
7. England RJ, Crabbe DC. Delayed diagnosis of appendicitis in children treated with antibiotics. *Pediatr Surg Int*. 2006;22:541-545.
8. National Institute for Health and Clinical Excellence. Feverish illness in children: Assessment and initial management in children younger than 5 years. National Collaborating Centre for Women's and Children's Health. 2007.
9. Long E, Vince J. Evidence behind the WHO guidelines: hospital care for children: what are appropriate methods of urine collection in UTI? *J Trop Pediatr*. 2007;53:221-224.
10. Montini G, Zucchetta P, Tomasi L, et al. Value of imaging studies after a first febrile urinary tract infection in young children: data from Italian renal infection study. *Pediatrics*. 2009;123:e239-e246.

11. Christian MT, McColl JH, MacKenzie JR, Beattie TJ. Risk assessment of renal cortical scarring with urinary tract infection by clinical features and ultrasonography. *Arch Dis Child*. 2000;82:376-380.
12. Mattoo TK. Are prophylactic antibiotics indicated after a urinary tract infection? *Curr Opin Pediatr*. 2009;21:203-206.
13. Nishizaki N, Someya T, Hirano D, et al. Can cranberry juice be a substitute for cefaclor prophylaxis in children with vesicoureteral reflux? *Pediatr Int*. 2009;51:433-434.

Chapter 3
Urinary Tract Infection: Europe

Özgü Aydogdu and Christian Radmayr

Key Points

- Urinary tract infection (UTI) is one of the most common bacterial infections seen in children.
- Urine culture should be obtained for diagnosis, if there is clinical suspicion or positive urinalysis.
- After a maximum of two UTI episodes in a girl and one episode in a boy, investigations should be undertaken.
- Dimercapto-succinic acid (DMSA) scan is the gold standard in the prediction of renal scarring.
- Main goal of the treatment should be the elimination of symptoms in the acute episode and prevention of renal deterioration in the long term.
- Oral treatment with fluids and oral antibiotics should be preferred in case of uncomplicated UTI.
- Prophylactic antibiotics may be used to reduce the risk of recurrent UTI.
- Prevention of potential UTI is the most critical step in the management of UTI in a child.

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_3,

3.1 Introduction

Urinary tract infections (UTIs) remain a significant cause of serious bacterial infections in children. It represents the most common bacterial infection in children less than 2 years of age. The incidence of UTIs varies depending on age and sex. In the first year of life, mostly the first 3 months, UTIs are more common in boys (3.7%) than in girls (2%). Later the incidence changes and approximately 3% of prepubertal girls and 1% of prepubertal boys are diagnosed with UTIs.[1-3]

Especially younger than 2 years, UTIs have been associated with significant morbidity and long term medical problems, such as hypertension, impaired renal function and chronic kidney disease. Thus, prompt diagnosis and treatment are critical in preventing the possible pathologic sequelae of UTIs. Pediatric UTIs should be considered as complicated until proved otherwise. Potential genitourinary abnormalities should also be considered subsequent to a diagnosis of UTI, and early diagnosis and accurate management can provide patients with an improved long-term prognosis.[3]

3.2 Aetiology

1. The common pathogenic sources are gram-negative, mainly enteric, organisms. *Escherichia coli* is responsible for more than 80% of episodes of UTIs.[2, 4, 5]
2. *Klebsiella* and *Proteus* with *Enterococcus* and *Staphylococcus* represent 5–7% of cases.[2, 4, 5]
3. Groups A and B streptococci are relatively common in the newborn.[2, 4, 5]

3.3 Pathogenesis and Risk Factors

1. Normally the urinary tract is a sterile space and retrograde ascent is the most common mechanism of infection particularly after the third or later months of life.[4, 5]

2. Nosocomial infection and involvement as a part of a systemic infection are less common.[4,5]
3. Evidence on risk factors for UTI in children is limited. The most common risk factors for UTI in infants and children are age, sex, periurethral or colonization factors, genitourinary abnormalities and native immunity.[5-13]
   - In girls, bacteria can gain access to the urinary tract more easily than in boys because of the perineal location of the urethral orifice and the shorter urethra.
   - Vaginal voiding can lead to increased moisture in the perineum allowing for bacterial overgrowth.
   - Phimosis predisposes to UTI. In boys, the prepuce can serve as a reservoir for potentially uropathogenic bacteria. *E. coli* expressing P fimbriae which adhere to the inner layer of the preputial skin and to uroepithelial cells colonize the preputial sac.
   - Obstruction is one of the most common causes of UTI. A wide variety of congenital urinary tract abnormalities (posterior urethral valves, ureteropelvic junction obstruction, and ureterovesical junction obstruction) can cause UTIs through obstruction.
   - Non-obstructive urinary stasis (e.g., vesico-ureteral reflux (VUR), prune belly syndrome) is another potential cause of UTIs.
   - Dysfunctional voiding may result in infrequent bladder emptying and residual urine in the bladder, providing the bacteria a suitable place for multiplying enough to cause infection.
   - Neurogenic bladder caused by spina bifida, spinal lesions or injury, may lead to post void residual urine and therefore is a potential risk factor for UTI.
   - Children who have been catheterized and patients on clean intermittent catheterization are potential candidates of UTIs.
   - Some children may have a susceptible urothelium that allows an increase in bacterial colonization. These patients may possibly have a decrease in cellular immunity.
   - Other significant causes of UTIs are chronic constipation/encopresis and labial adhesion.

3.4 Classification

1. Urinary tract infections can be classified according to either number of episodes (first episode or recurrent) or severity (simple or severe).[2, 14]
2. Recurrent UTIs may be due to unresolved infection, bacterial persistence or reinfection.[5, 15]
3. Recurrent UTI (any of the following)[14, 16]
   - Two or more episodes of UTI with acute pyelonephritis (APN) or upper UTI.
   - One episode of UTI with APN or upper UTI plus one or more episode of UTI with cystitis or lower UTI.
   - Three or more episodes of UTI with cystitis or lower UTI.
4. Severe and simple forms UTI should be differentiated clinically (Table 3.1) to decide the degree of urgency with which diagnostic tool and treatment are to be undertaken.

3.5 Signs and Symptoms

Signs and symptoms suggestive of UTI include;

1. Non-specific symptoms, such as irritability, lethargy, malaise, vomiting, diarrhea, failure to thrive, poor feeding, jaundice (especially in infants).[16, 17]
2. In the first weeks of life, 13.6% of patients with fever have a UTI.[17]

Table 3.1. Clinical classification of UTIs in children.

Severe UTI	Simple UTI
Fever ≥ 39°C	Mild pyrexia
Persistent vomiting	Good fluid intake
Moderate or serious dehydration	Not or slightly dehydrated
Poor treatment compliance	Good expected level of compliance

Taken from EAU guidelines, update March 2009

3. Specific symptoms include dysuria, frequency, hesitancy, urgency, small-volume voids, hematuria, cloudy urine and suprapubic, lumbar or abdominal pain with or without fever.[2, 14, 16]
4. Unusual odor of the urine is generally not helpful in predicting UTI.[15, 16]
5. Other conditions, such as acute urethritis and vulvovaginitis may mimic UTI symptoms.[14, 16]

The presenting symptoms of UTI may differ according to the anatomic site of the infection.[14-17]

1. Fever, nausea, vomiting and lumbar pain may be observed in upper UTI.
2. A lower UTI generally presents with symptoms such as dysuria, frequency, urgency and sometimes urinary incontinence.
3. The only way to correctly differentiate upper and lower urinary tract infection is performing a DMSA scan at the time of diagnosis. However this is not needed in most of the cases.[18, 19]

3.6 Diagnosis

A complete history and physical examination of the patient are just as critical in the evaluation of the patient with suspected UTI as the laboratory tests.

1. *Physical examination*
   - Physical examination should be the first step and may be alone enough to discover underlying pathology.
   - Clinician should be cautious for phimosis, signs of pyelonephritis or epididymo-orchitis, labial adhesion and congenital abnormalities (e.g., stigmata of spina bifida).
   - Fever may or may not exist with aforementioned signs.
2. *Laboratory tests*
   - Diagnosis of a suspected UTI is based on the examination of the urine.[14, 16, 19]
   - Most investigators define a UTI as the presence of organisms in the urine combined with signs and symptoms of UTI in the patient.[2, 18, 19]

A. *Method of urine collection*

- The technique of obtaining urine for urinalysis or culture affects the rate of contamination and thus influences the interpretation of the result.
- The diagnostic threshold depends on the method of urine collection.[20-22]
- In older, toilet-trained children who can void on command, the use of clean catch, especially the midstream urine, after carefully retracting the foreskin and cleaning the glans penis in boys and spreading the labia and cleaning the periurethral area in girls was found to be an acceptable technique for obtaining urine.[22]
- Some studies have compared the results of cultures of urine obtained by both clean catch and catheterization. As a result it was reported that clean voided specimens had similar rates of contamination to those obtained by catheterization.[20, 22]
- For urine collection from infants and young children, suprapubic aspiration or transurethral catheterization should be preferred.[19, 20]
- Prospective studies proved that collection from bags or pads had a high incidence of false positive predictive value.[21, 22]
- Urine collection from a bag is helpful when the culture result is negative and has a positive predictive value of 15%.[21, 22]

B. *Urinalysis: microscopic examination/dipstick analysis*

- There is a need for a more rapid determination of the probability of the presence of a UTI to guide the clinician in the decision to treat empirically while waiting for the culture result.[19, 23, 24]
- The most frequent markers are nitrite and leukocyte esterase usually combined in a dipstick test.
- Combination of the presence of leukocyte esterase and nitrite is highly predictive of a positive urine culture.[24]

- Nitrite is the degradation product of the nitrates of bacterial metabolism, particularly of gram-negative bacteria.[22, 24]
- Clinician should realize that many gram-positive cocci does not produce nitrites and may yield a false-negative result.
- The nitrite test has a sensitivity of 45–60%, and a specificity of 85–98%.[20, 24]
- Leukocyte esterase is produced by the activity of leukocytes.
- Leukocyte esterase test has a sensitivity of 48–86% and a specificity of 17–93%.[20, 24]
- In patients who have a negative dipstick and microscopic analysis, especially if there is an alternative source of fever, further urine culture is generally not necessary.
- If the tests are positive, it is better to confirm the results in combination with the clinical symptoms and urine culture.
- Bacteriuria without pyuria may be found when urine is collected before the onset of the inflammatory reaction, in bacterial contamination and colonization.[19, 24]
- Pyuria without bacteriuria may be due to incomplete treatment, urolithiasis or foreign bodies in the urinary tract and infections caused by *Mycobacterium tuberculosis* or *Chlamydia trachomatis*.[19, 24]
- Either bacteriuria or pyuria may not be considered reliable parameters to diagnose or exclude UTI.
- In a febrile child with both bacteriuria and pyuria (the findings of ≥10 WBC/mm^3 and ≥50,000 cfu/mL in an urine specimen), the possibility of UTI increases significantly.[17, 24]

C. *Urine culture*
- Culture of the urine remains the gold standard for the diagnosis of the UTIs.[20-23]

Table 3.2. Interpretation of the urine culture results in the diagnosis of UTIs in children.

Suprapubic aspiration	Transurethral catheterization	Midstream void
Any number of cfu/mL	≥1,000 –50,000 cfu/mL	$\geq 10^4$ cfu/mL (with symptoms) $\geq 10^5$ cfu/mL (without symptoms)

Taken from EAU guidelines, update March 2009

- Urine cultures in infants and children smaller than 6 months of age, without another focus of infection, show an incidence of UTI in approximately 8% with an axillary temperature of greater than 38.3°C.[20-23, 25]
- As aforementioned concentration of bacteriuria in urine and thus the criteria of UTI in children mainly depend on the method of urine collection (Table 3.2). Diuresis, method of the storage and transport of the urine are also critical.

3.7 Imaging Studies

Ultrasound scan (USG), voiding cystourethrography (VCUG), and DMSA are the three main imaging options for the investigation of UTI in children.[18, 26-29] According to some recent studies imaging work-ups for children with a first uncomplicated UTI may not improve the patient management.[27,28]

However other studies advocate that, although only minority of children with a UTI has an underlying urological disorder, morbidity can be disruptive for diseased children. Therefore after a maximum of two UTI episodes in girls and one episode in boys, imaging studies should be undertaken.[18, 27]

- Use of all three renal imaging techniques is neither realistic nor practical.
- A gold standard imaging technique should be painless, non-invasive, safe, reliable enough to detect any structural

anomaly and cost-effective. There is not any present radiological technique, fulfilling all these requirements.
- Urinary USG; for the detection of congenital abnormalities, obstruction, and renal scarring.
- VCUG; for the identification of VUR.
- DMSA; for the determination of renal scarring.

3.8 Ultrasound Scan (USG)

1. Safe, rapid, cost-effective and high accuracy in identifying the anatomy and size of the renal parenchyma and collecting system.[27,29]
2. Subjective and provides no information on renal function.
3. USG alone is not enough to rule out VUR. A child with high grade reflux in VCUG may have a totally normal urinary USG.[27,28]
4. Bladder USG can be used to determine the post void residual urine volume.

3.9 Voiding Cystourethrography (VCUG)

1. Urine sterility is mandatory to perform VCUG.
2. It is considered to be essential in the evaluation of UTIs in children less than 1 year of age.[25,28]
3. It is mandatory in the assessment of febrile UTIs in children.
4. Up to 23% of children with febrile UTI and normal USG may reveal VUR in VCUG.[28]
5. Disadvantages include the risk of infection, invasive and radiation exposure.[27,28]
6. Fluoroscopic VCUG may be used instead of conventional VCUG in order to minimize the radiation exposure.
7. Timing of VCUG in the investigation of UTI remains controversial. In some studies it was highlighted that VCUG should be performed 4–6 weeks after the acute infection.[28]
8. According to some recent studies VCUG can be performed as long as the child is free of infection.[27,28]

3.10 Dimercapto-Succinic Acid Scan (DMSA)

1. DMSA scan rightly remains the gold standard in the prediction of renal scarring. In a study its specificity and sensitivity for renal scarring were found as 100% and 80% respectively.[27]
2. This technique is helpful in determining functional renal mass and cortical scarring.
3. A star shaped defect in the renal parenchyma may indicate an acute episode of pyelonephritis.
4. A focal defect in the renal cortex often indicates a chronic lesion.
5. DMSA scan may help to differentiate between the good and the bad prognosis of UTIs in children.[18, 27]

3.11 Treatment

Management of a child with possible UTI is critical due to potential irreversible morbidities which can be avoided with proper treatment. Figure 3.1 summarizes the management of UTIs in children.

The main goal of the treatment in the acute period is to eliminate the symptoms and get rid of the bacteria. In the long term the treatment should aim firstly to preserve renal function by preventing possible renal scarring, prevent recurrent UTIs and correct underlying urological disorders.[14, 16, 19]

1. Antimicrobial treatment should be initiated on an empirical basis and should be adjusted according to the urine culture results.
2. The child should be reevaluated with a repeat urine culture and urinary USG if clinical improvement does not occur within two days.
3. If UTI is considered to be simple, oral rehydration and oral antibiotics on an outpatient basis may be sufficient. Oral empirical treatment with trimethoprim (TMP), cotrimoxazole (TMP plus sulphamethoxazole), cephalosporin or amoxycillin/clavulanate is recommended. The

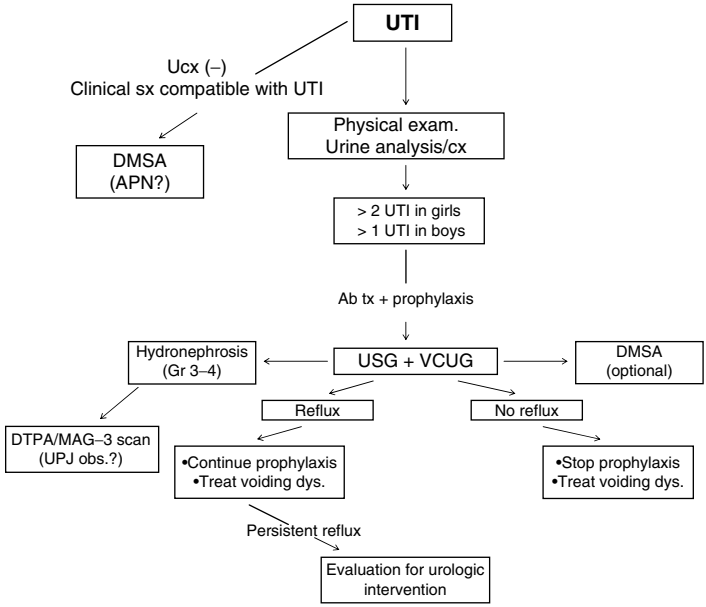


FIGURE 3.1. Management of UTIs in children.

preferred oral medication may differ due to the local resistance pattern.

4. Although there can be some variations due to different regions, the resistance rates in UTIs caused by *E. coli* are highest for ampicillin (39–45%) and co-trimoxazole (14–31%) and lowest for nitrofurantoin (1.8–16%) and fluoroquinolones (0.7–10%).
5. The duration of treatment in case of simple UTI should be 5–7 days.
6. If the child is not able to tolerate oral fluid intake or the response to oral antibiotics is poor, intravenous hydration and antibiotics should be administered in the hospital.
7. Preferred intravenous antibiotic in case of severe UTI should be a third generation cephalosporin. If a gram positive bacterium is suspected, it is better to use aminoglycosides (serum levels should be monitored) in combination with ampicillin or amoxycillin/clavulanate.

8. In case of sensitivity or allergy to cephalosporins, aztreonam or gentamicin may be preferred.
9. Parenteral therapy should be continued for 24–36 h. After this period if the child becomes afebrile and able to take fluids, oral antibiotic treatment as aforementioned may be administered. In case of severe UTIs, oral antibiotic treatment should be continued 10–14 days.

3.12 Prophylaxis and Prevention

1. Prophylactic antibiotics may reduce the risk of recurrent UTIs.
2. In case of increased risk of pyelonephritis (VUR, recurrent UTI) and until the diagnostic studies are completed antibiotic prophylaxis should be given.[19, 23]
3. Most preferred and effective agents are TMP, cephalexin, nitrofurantoin and cefaclor.
4. Cranberry juice and related products may be administered for prevention, but the evidence base is too limited to support the use of cranberry juice alone.
5. Breastfeeding has been shown to offer significant protection against UTIs by the protective factors such as lactoferrin and oligosaccharides in human milk.[19, 23]
6. Underlying conditions which increase the risk of a potential UTI such as voiding dysfunction and constipation should be treated properly.[11, 12]
7. Significant abnormalities in the urinary tract (e.g., VUR, obstruction), should be corrected with appropriate urological intervention.
8. Boys with underlying urinary system disorders such as posterior urethral valve and VUR may benefit from neonatal circumcision. However this remains controversial.[6, 7, 10]

References

1. Qigley R. Diagnosis of urinary tract infections in children. *Curr Opin Pediatr*. 2009;21:194-198.
2. Riccabona M. Urinary tract infections in children. *Curr Opin Urol*. 2003;13:59-62.
3. Foxman B. Epidemiology of urinary infections: incidence, morbidity, and economic costs. *Am J Med*. 2002;113(Suppl 1A):5S-135S.
4. Jodal U. The natural history of bacteriuria in childhood. *Infect Dis Clin North Am*. 1987;1(4):713-729.
5. Ma JF, Shortliffe LM. Urinary tract infection in children: Etiology and epidemiology. *Urol Clin North Am*. 2004;31(3):517-526.
6. Craig JC, Knight JF, Sureshkuman P, Mantz E, Roy LP. Effect of circumcision on incidence of urinary tract infection in preschool boys. *J Pediatr*. 1996;128(1):23-27.
7. To T, Agha M, Dick PT, Feldman W. Cohort study on circumcision of newborn boys and subsequent risk of urinary-tract infection. *Lancet*. 1998;352(9143):1813-1816.
8. Shapiro ED. Infections of the urinary tract. *Pediatr Infect Dis J*. 1992;11(2):165-168.
9. Richards MJ, Edwards JR, Culver DH, Gaynes RP. Nosocomial infections in pediatric intensive care units in the United States. National Nosocomial Infections Surveillance System. *Pediatrics*. 1999;103(4):e39.
10. Fussell EN, Kaack MB, Cherry R, Roberts JA. Adherence of bacteria to human foreskins. *J Urol*. 1988;140(5):997-1001.
11. Wan J, Kaplinsky R, Greenfield S. Toilet habits of children evaluated for urinary tract infection. *J Urol*. 1995;154(2):797-799.
12. Schulman SL. Voiding dysfunction in children. *Urol Clin North Am*. 2004;31(3):481-490.
13. Abrahamsson K, Hansson S, Jodal U, Lincoln K. Staphylococcus saprophyticus urinary tract infections in children. *Eur J Pediatr*. 1993;152(1):69-71.
14. Mori R, Lakhanpaul M, Verrier-Jones K. Diagnosis and management of urinary tract infection in children: summary of NICE guidance. *BMJ*. 2007;335:395-397.
15. Struthers S, Scanlon J, Parker K, Goddard J, Hallett R. Parental reporting of smelly urine and urinary tract infection. *Arch Dis Child*. 2003;88:250-252.
16. Alper BS, Curry SH. Urinary tract infection in children. *Am Fam Physician*. 2005;72(12):2483-2488.
17. Lin DS, Huang SH, Lin CC, et al. Urinary tract infection in febrile infants younger than eight weeks of Age. *Pediatrics*. 2000;105(2):E20.

18. Luk WH, Woo YH, Au-Yeung AWS, Chan JCS. Imaging in pediatric urinary tract infection: A 9-year local experience. *AJR Am J Roentgenol*. 2009;192:1253-1260.
19. Zorc JJ, Kiddoo DA, Shaw KN. Diagnosis and management of pediatric urinary tract infections. *Clin Microbiol Rev*. 2005;18:417-422.
20. Long E, Vince J. Evidence behind the WHO guidelines: hospital care for children: what are appropriate methods of urine collection in UTI? *J Trop Pediatr*. 2007;53:221-224.
21. Cavagnaro F. Urinary tract infection in childhood. *Rev Chilena Infectol*. 2005;22(2):161-168.
22. Whiting P, Westwood M, Watt I, et al. Rapid tests and urine sampling techniques for the diagnosis of urinary tract infection in children under five years: a systematic review. *BMC Pediatr*. 2005;5:4.
23. Watson AR. Pediatric urinary tract infection. *EAU Update Ser*. 2004;2:94-100.
24. Stamm WE. Measurement of pyuria and its relation to bacteriuria. *Am J Med*. 1983;75(1B):53-58.
25. Hoberman A, Chao HP, Keller DM, Hickey R, Davis HW, Ellis D. Prevalence of urinary tract infection in febrile infants. *J Pediatr*. 1993;123(1):17-23.
26. De Sadeleer C, De Boe V, Keuppens F, Desprechins B, Verboven M, Piepsz A. How good is technetium-99m mercaptoacetyltriglycine indirect cystography? *Eur J Nucl Med*. 1994;21(3):223-227.
27. Kass EJ, Kernen KM, Carey JM. Paediatric urinary tract infection and the necessity of complete urological imaging. *BJU Int*. 2000;86(1):94-96.
28. Mabant S, To T, Friedman J. Timing of voiding cystourethrogram in the investigation of urinary tract infections in children. *J Pediatr*. 2001;139:568-571.
29. Ransley PG, Risdon RA. Renal papillary morphology in infants and young children. *Urol Res*. 1975;3(3):111-113.

Chapter 4
Urinary Tract Infection: Australasia

Naeem Samnakay and Andrew Barker

Key Points

- Always check the antenatal history in a child with a UTI.
- A renal USS must be obtained in all children after an initial UTI. It will not exclude VUR.
- An MCU is an important test that should be done selectively looking for VUR, bladder anomalies or posterior urethral valves. It is performed after the first UTI in those with an abnormal USS, after UTI requiring IV antibiotics or if there is a strong family history of VUR; or after a second UTI in a child who had a previous normal USS. A male child post-UTI with a history or USS suspicious for posterior urethral valves should have an MCU.
- VUR can be treated surgically and surgical management has been shown to significantly reduce the risk of febrile UTIs in children with VUR compared to medical management alone.

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_4,

4.1 Epidemiology

Urinary tract infections (UTIs) are a common childhood condition. It is estimated that 2% of boys and 7% of girls will be diagnosed with a urinary UTI by age 6.[1, 2]

Under 1 year of age, boys are more likely to present with a UTI than girls; after this age, girls get UTIs more commonly than boys.[1]

4.2 Presentation

The very young infant and the preverbal child with a UTI usually present generally unwell with fever, vomiting and lethargy. Up to 13% of infants with fever of unknown origin will have a UTI, so one must always suspect UTI as a possible diagnosis in the unwell child.[3]

The older child may present with localized signs and symptoms more specific to a UTI, such as loin pain, suprapubic pain or dysuria.

4.3 Diagnosis and Workup

Apart from the general history of the presenting complaint, specific aspects of history in a child with a UTI are important:

- Antenatal history
  In particular, any antenatal hydroureteronephrosis or bladder abnormalities at any stage of gestation. As these findings are usually filed in the mother's obstetric notes, they may not be available without an effort to trace them. Management will change if there is a known antenatally diagnosed urinary tract anomaly. On the other hand, we feel that a normal antenatal scan does *not* obviate the need for a renal tract ultrasound in a child with a UTI.
- History of previous UTIs
- History of predisposing conditions such as renal stones, congenital structural anomalies and spina bifida

- Urinary stream and voiding pattern – is there dysfunctional voiding?
- History of constipation
- Family history of urinary tract anomalies and vesicoureteric reflux (VUR)

Examination should also include looking for loin tenderness, a palpable bladder or kidneys and the appearance of the external genitalia. The back should be inspected for evidence of occult spinal dysraphism.

The diagnosis of UTI is made with a urine sample. The acutely unwell baby or infant with fever and poor feeding in Australia is often referred to the emergency department of a secondary or tertiary centre, where a full septic screen is performed. As much as possible, a urine sample should be obtained as a clean catch. A bag specimen has a high false positive rate due to contamination from the skin. A bag specimen is useful to exclude a UTI if the collected urine is culture negative; however, one should be suspicious of a UTI if bacteria, protein and white cells are present in the bag specimen and obtain a clean catch if possible. If a clean catch cannot be obtained, a suprapubic aspirate or catheter specimen should be obtained.

Urine should be sent off for urgent microscopy and culture in the unwell young infant; in the older child, it may be sent off for routine microscopy, culture and sensitivities. Bacteria seen on microscopy are 93% sensitive and 95% specific for a UTI. Positive nitrites on a dipstick are 99% specific in predicting a UTI, but if nitrites are negative, they are only 60% sensitive in ruling out a UTI.[4]

4.4 Management

Empirical treatment with antibiotics is usually commenced after specimen collection, and adjusted accordingly once results are through. The majority of UTIs (>80%) are due to *Escherichia coli,* with the remaining being due to *Proteus, Klebsiella,* and *Enterococcus. Pseudomonas* and *Staphylococcus* UTIs are uncommon and considered atypical.[5]

Generally children with temperatures over 38°C, loin pain and tenderness with positive urine cultures are considered to have an upper tract infection and are treated with intravenous antibiotics; older children with temperatures under 38°C, dysuria and frequency with positive urine cultures and no loin pain or tenderness, are considered to have cystitis and treated with oral antibiotics.

4.5 Investigations after First UTI in a Child

Studies suggest that 21–50% of children with a UTI will have an underlying abnormality.[3] It is estimated that about 10% of children presenting with a UTI will have an abnormal ultrasound finding which may or may not affect management.[6] This includes abnormalities such as pelviureteric junction obstruction, vesicoureteric junction obstruction and posterior urethral valves in boys. For this reason, we recommend that all children presenting with an initial UTI should have a renal tract ultrasound (USS). A renal tract USS after initial UTI should be performed even if there is a history of normal antenatal ultrasound scans.

Up to 30% of children presenting with a UTI will have underlying vesicoureteric reflux (VUR).[6] 90% of this VUR is lower grades, I to III. USS is not useful for assessing the presence or grade of VUR. A micturating cystourethrogram or MCU is the most useful test to assess for and grade VUR. An MCU is performed by catheterizing the bladder urethrally and filling it with contrast, followed by imaging during voiding. Sedation may be required in some cases.

VUR and UTIs are associated with renal damage, scarring and hypertension. It is estimated that 5% of children presenting with UTI will have associated renal scarring.[7] Delay in the diagnosis and treatment of UTIs, and recurrent UTIs correlate more with renal scarring. However, it is now well known that renal scarring in children with VUR may be present even before a clinical UTI, suggesting pre-existing scarring or dysplasia of the kidney. This is especially

common in young male infants with VUR. On the other hand, children with VUR and UTIs may not acquire renal scarring, whilst some children with no proven VUR may acquire renal scarring after UTIs. With these variations, it is difficult to obtain data about the true risks of UTIs and the true role of associated VUR in the development of renal scarring.

The American Academy of Pediatrics recommends USS and MCU for all children presenting with their first UTI between ages of 2 months and 2 years.[8] When significant VUR is found it is treated surgically with either endoscopic ureteric injection or open ureteric reimplantation.

The Royal College of Physicians in the UK in 1991 recommended screening children under 1 year of age with a renal USS, MCU as well as a Dimercaptosuccinic acid scan (DMSA) after a first UTI.[9] The more recent UK NICE guidelines from 2007 have changed this traditional view. In essence, the guidelines recommend imaging after a first UTI in children should be more directed and selective, based on factors such as the age of the child, whether the UTI was typical or atypical and the response to treatment.[10] One must remember that guidelines such as NICE are guides and are not meant to be prescriptive, so individual cases must be dealt with on their merits.

There are no set guidelines in Australia, and Australian practice in terms of imaging children after their first UTI varies from centre to centre and practitioner to practitioner.[11]

Our practice currently is to always obtain a renal USS after an initial UTI. An MCU should also be obtained in the following situations:

- Known anomaly on antenatal ultrasound scan – especially if dilated ureters are visible or a bladder or urethral abnormality is suspected
- Abnormality noted on the post-UTI renal ultrasound scan
- In any boy with any degree of bilateral hydronephrosis, or history of poor stream or voiding difficulties – to exclude the possibility of posterior urethral valves (Fig. 4.1)

- Strong family history of VUR and reflux nephropathy
- Recurrent UTIs
- Atypical UTI organism
- Severe UTI requiring IV antibiotics
- Geographical factors – Children in rural and remote areas of Australia who have to travel long distances to specialist health care, often need to be transferred to regional centers for their post-UTI investigations. In such children, it may be prudent to perform an MCU whilst they are at the regional centre even if they have a normal USS after a first UTI.

A DMSA scan is performed in children who have documented VUR on the MCU or the appearance of renal scarring suggested on ultrasound scan. The DMSA scan will give

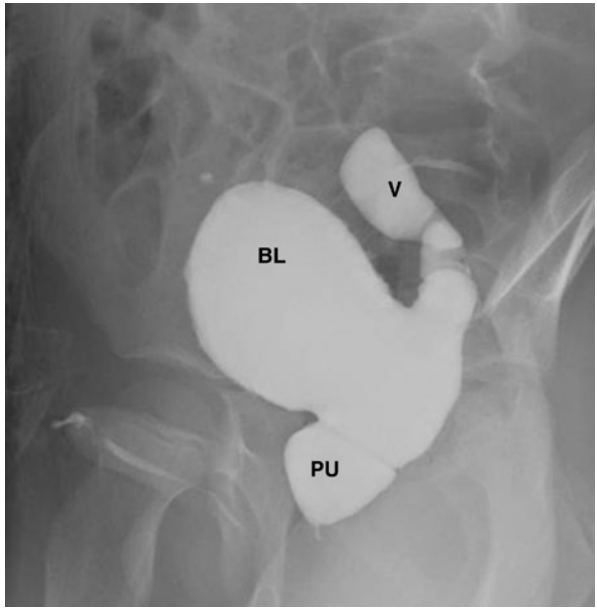


FIGURE 4.1. MCU of 6 year old boy whose first UTI in infancy was treated but not followed up with imaging. It shows a severely dilated posterior urethra (PU), trabeculated bladder (B) and VUR on the left (V). The boy had posterior urethral valves.

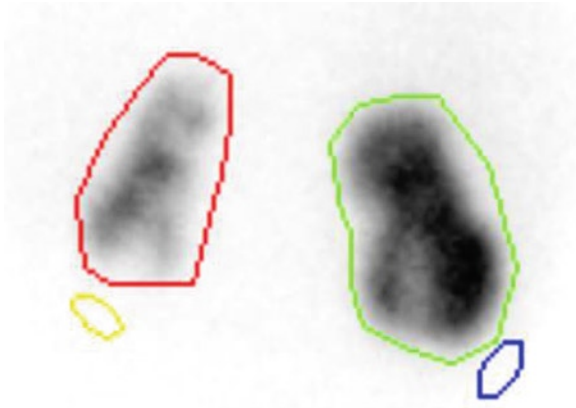


FIGURE 4.2. DMSA scan of 10 year old girl with persistent grade II VUR bilaterally and recurrent UTIs managed medically showing bilateral renal scarring. The black areas are functioning renal tissue and the pale punched out areas are scars. Left kidney (*red ring*) functions poorly compared to the right kidney (*green ring*).

a clear idea of differential renal function and distribution of renal scarring. It can also been used acutely to diagnose acute pyelonephritis (Fig. 4.2).

4.6 Prevention of UTIs

Important practices to minimize the risk of UTIs include:

- *Prophylactic antibiotics*

  Up to now, there was no strong evidence to support the role of prophylactic antibiotics after a single UTI in preventing recurrences. A recent Australian randomized control trial is the first to definitively show a reduction in the risk of UTIs in the 12 months after the index UTI in the group of children placed on daily trimethoprim/sulphamethoxazole.[5]

However, the protective effect seemed to drop off with time, and the long term benefit of antibiotic prophylaxis in children with UTIs remains unproven.

- *Managing dysfunctional elimination*

  Dysfunctional voiding, with raised post-void residual volumes and constipation, are associated with an increased risk of urinary tract infections and hence must be carefully investigated and managed, particularly in children who present with recurrent UTIs.

- *Cranberry juice*

  Contains anthocyanidin/proanthocyanidin moieties that are potent antiadhesion compounds and thought to prevent I and P-fimbriated uropathogens such as *E. coli* from adhering to the urothelium. There is randomized evidence that cranberry helps reduce the risk of recurrent UTIs in adult women, and its value in children is extrapolated but not proven.[12]

- *Probiotics*

  Thought to promote the balance of bowel organisms in favor of non-pathogenic commensals.

- *Circumcision in male infants*

  Studies suggest that circumcised boys have a tenfold lower risk of UTI compared with uncircumcised boys.[13, 14] However, there is no evidence to recommend circumcision for all boys after a first UTI. Circumcision is certainly considered in boys who get recurrent UTIs and have underlying urinary tract anomalies such as posterior urethral valves or VUR.

- *Drinking plenty of fluids*

- *Good toileting habits*

  These include regular 3 hourly voids, and wiping the perianal area away from urogenital area.

4.7 Managing VUR and UTIs

Most low grade VUR resolves spontaneously. For higher grade VUR, parents are given the choice of prophylactic antibiotics and waiting for possible spontaneous resolution, or surgical intervention to correct the VUR.

Data from the Cochrane Review show a 50% reduction in the risk of febrile UTIs in children after surgical treatment for VUR.[15] Surgical treatment options for VUR include:

- Endoscopic injection of ureteric orifice with substances like Deflux gel. This has an 80% success rate in correcting the VUR in correcting VUR after the first injection. It is done as day-case surgery under general anesthetic and can be repeated if VUR persists.
- Open surgical ureteric reimplantation. This has a 98% + success rate in correcting VUR. It involves a small suprapubic incision and 1–3 days in hospital recovering.

References

1. Marild S, Jodal U. Incidence rate of first-time symptomatic urinary tract infection in children under 6 years of age. *Acta Paediatr*. 1998;87(5):549-552.
2. Hellstrom A et al. Association between urinary symptoms at 7 years old and previous urinary tract infection. *Arch Dis Child*. 1991;66(2):232-234.
3. Linda S. Urinary Tract Infections in Infants and Children. In: Walsh P, ed. *Campbell's Urology*, vol. 3. Baltimore: Saunders; 2002.
4. Bauer R, Kogan BA. New developments in the diagnosis and management of pediatric UTIs. *Urol Clin North Am*. 2008;35(1):47-58. vi.
5. Craig JC et al. Antibiotic prophylaxis and recurrent urinary tract infection in children. *N Engl J Med*. 2009;361(18):1748-1759.
6. Giorgi LJ Jr, Bratslavsky G, Kogan BA. Febrile urinary tract infections in infants: renal ultrasound remains necessary. *J Urol*. 2005;173(2):568-570.
7. Coulthard MG, Lambert HJ, Keir MJ. Occurrence of renal scars in children after their first referral for urinary tract infection. *BMJ*. 1997;315(7113):918-919.

8. American Academy of Pediatrics. Committee on Quality Improvement. Subcommittee on Urinary Tract Infection. Practice parameter: the diagnosis, treatment, and evaluation of the initial urinary tract infection in febrile infants and young children. *Pediatrics*. 1999;103(4 Pt 1):843-852.
9. Royal College of Physicians. Guidelines for the management of acute urinary tract infection in childhood. Report of a working group of the research unit. *J R Coll Physicians Lond*. 1991;25(1): 36-42.
10. National Collaborating Centre for Women's and Children's Health. *Urinary Tract Infection in Children: Diagnosis, Treatment and Long-Term Management*. London, UK: National Institute for Health and Clinical Excellence; 2007.
11. Williams G et al. Ordering of renal tract imaging by paediatricians after urinary tract infection. *J Paediatr Child Health*. 2007;43(4): 271-279.
12. Jepson RG, Craig JC. Cranberries for preventing urinary tract infections. *Cochrane Database Syst Rev*. 2008(1):CD001321.
13. Wiswell TE et al. Declining frequency of circumcision: implications for changes in the absolute incidence and male to female sex ratio of urinary tract infections in early infancy. *Pediatrics*. 1987;79(3): 338-342.
14. Schoen EJ, Colby CJ, Ray GT. Newborn circumcision decreases incidence and costs of urinary tract infections during the first year of life. *Pediatrics*. 2000;105(4 Pt 1):789-793.
15. Hodson EM et al. Interventions for primary vesicoureteric reflux. Cochrane Database Syst Rev. 2007(3):CD001532.

Chapter 5
Abnormalities of the Scrotum

Paul F. Austin

Key Points

- The diagnosis of most acute abnormalities of the scrotum can be made by the history and physical exam.
- Testicular torsion is a urologic emergency whereas torsion of the testicular appendages may be managed expectantly.
- Most conditions causing an acute scrotum can be managed conservatively in primary care.
- A painless, non-acute testicular mass is considered a tumor until proven otherwise. The majority of pre-pubertal testicular tumors are benign.

5.1 Introduction

Abnormalities of the scrotum are frequently encountered in children and adolescents. Certain scrotal conditions require emergent care while others are non-emergent. Sorting through the many different conditions that result in an "abnormal appearing" scrotum is critical in determining the acuity of care and in choosing the appropriate treatment. This chapter will outline the different conditions that result in

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_5,

scrotal abnormalities and assist the primary care provider in diagnosis and management.

5.2 Common Abnormalities of the Scrotum

1. Testicular torsion: Testicular torsion is a urologic emergency and typically occurs in pubescent boys but may also present in the pre-pubescent period. The diagnosis is primarily a clinical determination that is made from the medical history and the physical exam. Radiologic imaging with scrotal ultrasonography is beneficial when the diagnosis is in doubt. There is a critical six hour time period after which irreversible testicular damage ensues with continued torsion. Key diagnostic symptoms and signs include: nausea, vomiting, acute onset, diffusely tender and palpably hard testis. Other helpful findings include a high-riding testis (Fig. 5.1), a horizontal lie of the testis and no cremasteric reflex. A colicky type of pain is typically seen early in the time course of testicular torsion. With delayed presentation, no tenderness in a palpably hard testis is an ominous sign.
2. Torsion of the appendix testis or epididymis: Torsion of the appendage of the testis or epididymis is common in the prepubescent child and is a great "mimicker" of testicular torsion. The onset of pain usually is acute, but may develop slowly over time. The intensity of pain ranges from mild to severe. The pain is generally located in a specific location such as at the superior pole of the testicle corresponding to the most common location of the appendix testes. However with prolonged duration, diffuse tenderness develops from the resultant inflammatory response. The pain is worse with movement and better with rest. If left untreated, diffuse scrotal erythema and edema ensues similar to testicular torsion (Fig. 5.2). Systemic symptoms such as nausea and vomiting are absent with a torsed appendage. The presence of a cremasteric reflex is a helpful physical finding. Generally, the testis is soft in consistency on palpation.

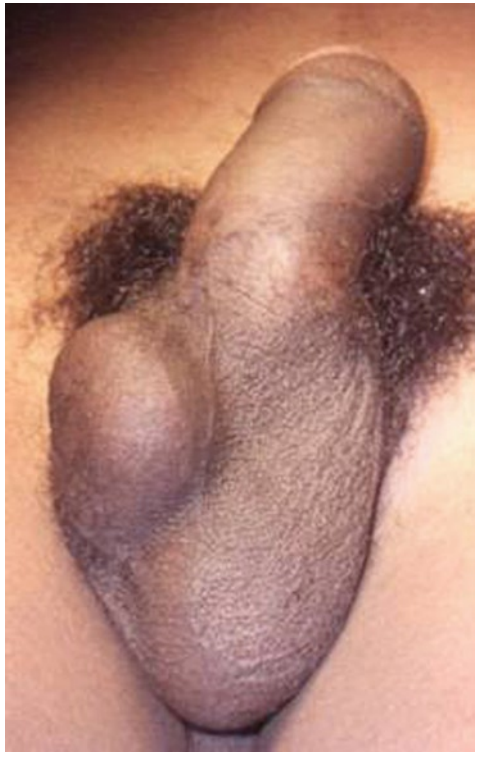


FIGURE 5.1. High riding right testis.

A hard palpable, paratesticular nodule in the groove between the superior aspect of the testis and the adjacent epididymis may be present. Additionally this infarcted appendix testis may demonstrate the "blue dot sign" underneath the scrotal skin.

3. Epididymo-orchitis: Bacterial epididymo-orchitis is rare in children. Bacterial epididymo-orchitis occurs secondary to ascension of a urinary tract infection (UTI) up the vas deferens into the epididymis. Irritative voiding symptoms would be present in setting of a UTI e.g., urinary urgency, frequency and dysuria. The child may have other symptoms such as a fever and the testicle and epididymis are

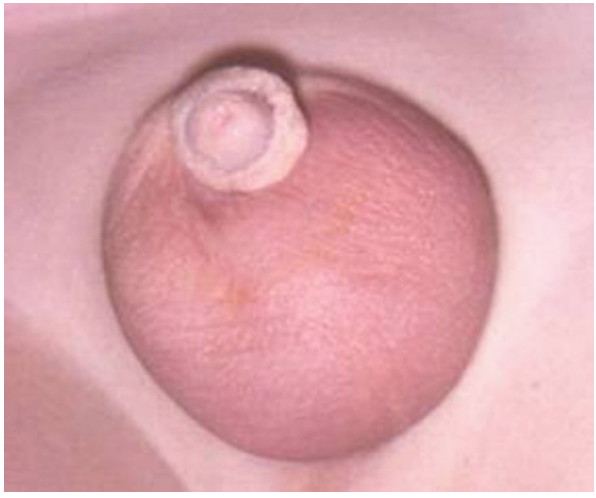


FIGURE 5.2. Diffuse left scrotal edema with torsed appendix testis.

exquisitely tender to touch with variable erythema. When bacterial epididymo-orchitis is present, it may indicate an underlying anatomical abnormality of the genitourinary system. The anatomical abnormality generally results in urinary stasis leading to the UTI. Lower urinary tract dysfunction is another cause for developing a UTI that could lead to epididymo-orchitis. More frequently, there is no evidence of a UTI upon lab testing and this term is often applied as a "wastebasket" diagnoses when there is uncertainty of the etiology of the acute scrotum. In a sexually active adolescent, the diagnosis is made after the history, physical exam and laboratory findings are supportive of a sexually transmitted disease.

4. Hernia/Hydrocele: Hernias and hydroceles in children are usually congenital rather than acquired as seen in adults. Pediatric hernias and hydroceles represent a patent processus vaginalis that allows either peritoneal fluid, omentum or bowel to descend into the inguinal canal or scrotum.

The history of a fluctuating mass that is reducible on exam is characteristic of the diagnosis. Hydroceles will transilluminate on exam. Hernias and hydroceles are generally not painful; however an incarcerated segment of bowel that becomes trapped or strangulated will be painful and represents a surgical emergency.

5. Spermatic varicocele: Spermatic varicoceles are common in the adolescent population and are frequently discovered during annual physical exams for school or camp. Spermatic varicoceles invariably occur on the left side because of the insertion of the gonadal vein into the left renal vein rather than the vena cava. The majority are asymptomatic however spermatic varicoceles may elicit pain. The pain is usually dull and achy in nature rather than sharp or knife-like. The adolescent will typically complain of a heaviness feeling of the scrotum. Testicular atrophy may be associated with a spermatic varicocele.
6. Spermatocele: Spermatoceles are well circumscribed, round cysts that are located in the epididymis. Spermatoceles are generally benign conditions and asymptomatic. They are generally located superiorly at the head of the epididymis or inferiorly at the tail of the epididymis. Spermatoceles are frequently discovered during annual physicals for school or camp.
7. Trauma: Most injuries to the testicle(s) are secondary to impact to the scrotum from rough play or sports. The testes are sore and tender to touch. Trauma to the testes may result in a contusion to the testes or rarely, there can be a disruption of the covering of the testes or tunica albuginea.
8. Insect bite: Insect bites to the scrotum may cause generalized erythema, edema and tenderness to the scrotal skin. There may be a localized, raised papule where the insect bite occurred on the skin.
9. Testicular tumors: A painless, hard testicular mass is presumed cancerous until proven otherwise. Testicular tumors are generally benign in the pre-pubescent child.

5.3 Treatment of Abnormalities of the Scrotum

1. Testicular torsion: Any acute and disabling testicular pain should be triaged quickly in the emergency room or office. A quick and expeditious diagnosis is paramount because of the ensuing ischemia. There is a 6–8 h window before complete infarction and necrosis of the testes occurs (Fig. 5.3). The diagnosis is typically made from the clinical history and the physical exam. The classic history is a post-pubertal child with sudden onset of pain accompanied by nausea and vomiting. The testicle is high-riding and diffusely tender and the child exhibits colicky pain. There is diffuse and increased firmness of the testicle. The cremasteric reflex is absent. A delayed presentation is frequent because the adolescent may not initially disclose their complaints. With a late presentation, the testicle becomes painless and hard. Scrotal sonography is useful in equivocal cases where the clinical history and physical exam are not definitive. An initial intervention by manual

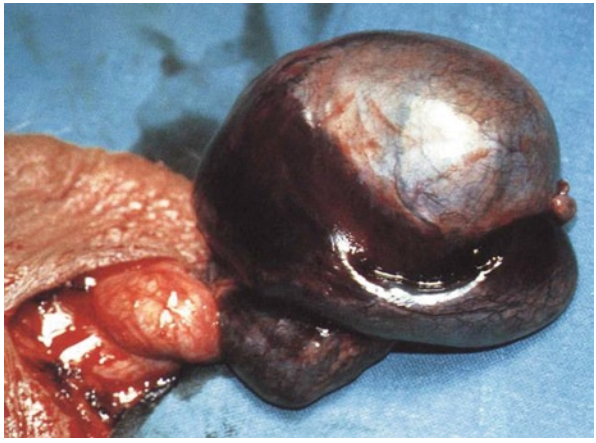


FIGURE 5.3. Necrotic testicle secondary to testicular torsion.

detorsion under sedation can be attempted depending on the comfort level of the physician. Manual detorsion is typically performed in a medial to lateral rotation however testicular torsion does not always occur in a uniform direction. Referral to a pediatric urologist should be promptly made when the diagnosis of torsion is suspected to plan for emergent exploration and treatment. Treatment is scrotal exploration with detorsion of the testes and either orchiopexy vs. orchiectomy with contralateral orchiopexy.

2. Torsion of the appendix testis or epididymis: Treatment is directed toward symptomatic relief and the resultant inflammation. Patients are advised to take non-steroidal inflammatory drugs (NSAIDs), use scrotal supportive undergarments and to limit physical activity or sports for one week.
3. Epididymo-orchitis: Treatment is directed toward the etiologic cause. If the epididymo-orchitis is idiopathic, then treatment is similar to treatment for torsion of the appendix testis e.g., NSAIDs, scrotal support and limited activity for 7 days. If the etiology is infectious then treatment includes empiric antibiotics until definitive cultures with sensitivity results are available that confirm the empiric treatment or direct new and different antibiotics. If there is suspicion that the patient has a sexually transmitted disease, the adolescent is treated with Ceftriaxone 125 mg or 250 mg intramuscular in a single dose plus Doxycycline 100 mg orally twice a day for 10 days. For acute epididymitis most likely caused by enteric organisms or with negative gonococcal culture or nucleic acid amplification test, treatment is Ofloxacin 300 mg orally twice a day for 10 days or Levofloxacin 500 mg orally once daily for 10 days.
4. Hernia/hydrocele: Treatment for hernias and hydroceles is surgical. If the hernia is reducible, then surgical treatment may be scheduled electively as an outpatient procedure. If the hernia is non-reducible with strangulation of a segment of bowel, then emergent surgical intervention is warranted.

5. Spermatic varicocele: The indications for treatment for varicoceles in an adolescent are different than in an adult. Fertility issues are the primary indication for treatment of varicoceles in adults and are not applicable to adolescents. The indications for treatment for adolescents would include (1) scrotal or testicular pain or (2) testicular atrophy of the affected side. Another consideration for treatment would include an extremely large spermatic varicocele such that the child is self-conscious and has self-esteem issues. The treatment for spermatic varicoceles is either through ligation/division of the spermatic veins or embolization of the spermatic veins.
6. Spermatocele: Spermatoceles are typically innocuous and pose no concern. Observation is the usual course. Rarely, spermatoceles may become quite large and elicit pain or become quite unsightly. If so, then surgical excision is reasonable.
7. Trauma: The treatment for scrotal trauma is dependent upon the underlying resultant injury. If there is testicular or scrotal wall contusion, then treatment is supportive care with NSAIDs, scrotal support and limited physical activity. If there is a testicular rupture of the testes, then emergent scrotal exploration with testicular debridement and closure is warranted.
8. Insect bite: Treatment is no different than for insect bites at other skin sites. For scrotal edema, treatment is supportive care with NSAIDs, scrotal support and limited physical activity.
9. Testicular tumors: Treatment is dependent on staging. Staging workup includes serum testing with a complete blood count, complete metabolic panel including a hepatic panel, alpha fetoprotein level and beta human chorionic gonadotropin level. Radiologic imaging includes computerized tomography (CT) of the chest, abdomen and pelvis or a chest x-ray with a CT of the abdomen and pelvis. Ultrasonography may be helpful in the surgical planning of pre-pubertal testes tumors which are more likely to be benign tumors.

5.4 Indications for Referral

1. Testicular torsion: Once the diagnosis is suspected, referral to a pediatric urologist is mandatory.
2. Torsion of the appendix testis or epididymis: Referral is dependent on the comfort level of the primary care provider with the diagnosis.
3. Epididymo-orchitis: If an anatomical congenital anomaly of the urinary tract is found, then a referral to a pediatric urologist is warranted.
4. Hernia/hydrocele: Referral to a pediatric urologist or surgeon is warranted after the diagnosis is made.
5. Spermatic varicocele: Referral to a pediatric urologist or interventional radiologist is warranted if there is an indication for treatment as listed above.
6. Spermatocele: Referral is not necessary if the patient is asymptomatic.
7. Trauma: Referral is dependent on the degree of injury. Contusions can be managed by the primary care provider.
8. Insect bite: Referral is generally not necessary.
9. Testicular tumors: Referral to a pediatric urologist and an oncologist is warranted once the diagnosis is suspected.

Suggested Readings

1. Gatti JM, Murphy JP. Acute testicular disorders. *Pediatr Rev*. 2008;29:235.
2. Trojian TH, Lishnak TS, Heiman D. Epididymitis and orchitis: An overview. *Am Fam Physician*. 2009;79:583.
3. Ross JH. Prepubertal testicular tumors. *Urology*. 2009;74:94.
4. http://www.cdc.gov/std/treatment/2006/updated-regimens.htm, 2009
5. Homayoon K, Suhre CD, Steinhardt GF. Epididymal cysts in children: Natural history. *J Urol*. 2004;171:1274.
6. Sessions AE, Rabinowitz R, Hulbert WC, et al. Testicular torsion: Direction, degree, duration and disinformation. *J Urol*. 2003; 169:663.
7. Diamond DA. Adolescent varicocele: Emerging understanding. *BJU Int*. 2003;92(Suppl 1):48.

Chapter 6
Disorders of Male External Genitalia: Problems of the Penis and Foreskin

Prasad P. Godbole

Key Points

- A non retractile foreskin at birth is normal. Spontaneous retractility begins around 2 years of age.
- Majority of boys will have a retractile foreskin by 10 years of age and 95% by 16–17 years of age.
- The only absolute medical indication for circumcision is penile malignancy and balanitis xerotica obliterans.
- Most inflammatory conditions of the foreskin can be managed conservatively in primary care.

6.1 Introduction

The management of foreskin conditions varies amongst medical practitioners from observation to circumcision. A number of conditions may affect the foreskin and may lead to a specialist referral. This chapter deals with common foreskin problems, their etiology and management in primary/emergency care. Indications for referral will be highlighted. Circumcision will be dealt with in another chapter.

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_6,

6.2 Common Foreskin Conditions

1. Non retractile foreskin: Almost all boys have a non retractile foreskin at birth. The inner foreskin is attached to the glans. Foreskin adhesions break down and form smegma pearls presenting as yellowish white cysts under the foreskin which are then extruded. Foreskin retractility begins after 2 years of age. This process is spontaneous and does not require manipulation. A non retractile foreskin on gentle attempted retraction pouts like a flower-physiological phimosis (Fig. 6.1). The majority of boys will have a retractile foreskin by 10 years of age and 95% by 16–17 years of age.[1]
2. Balanoposthitis: inflammation of the glans and the foreskin.
3. Balanitis: inflammation of the glans that often spreads along the shaft and may occur in the circumcised population.
4. Posthitis: inflammation restricted to the foreskin itself.
5. Balanitis Xerotica Obliterans(BXO): a lesion akin to lichen sclerosus et atrophicus. This causes true phimosis – pathological phimosis. This causes a shutter type foreskin with no pouting of the inner foreskin on gentle retraction (Fig. 6.2). It is rare before 5 years of age.
6. Paraphimosis: results when the narrow tip of the foreskin is retracted behind the glans at the coronal sulcus causing edema of the glans and foreskin and inability to manipulate the foreskin back over the glans (Fig. 6.3).
7. Hooded foreskin: is an abnormal dorsal hemiforeskin which is deficient ventrally and is usually associated with hypospadias (Fig. 6.4).

6.3 Treatment of Conditions of the Foreskin

1. Non retractile foreskin: This does not require any treatment if the foreskin is healthy. Topical steroids are known to hasten retractility of the foreskin and may be considered.[2, 3] On no account should the parents be asked to forcibly retract

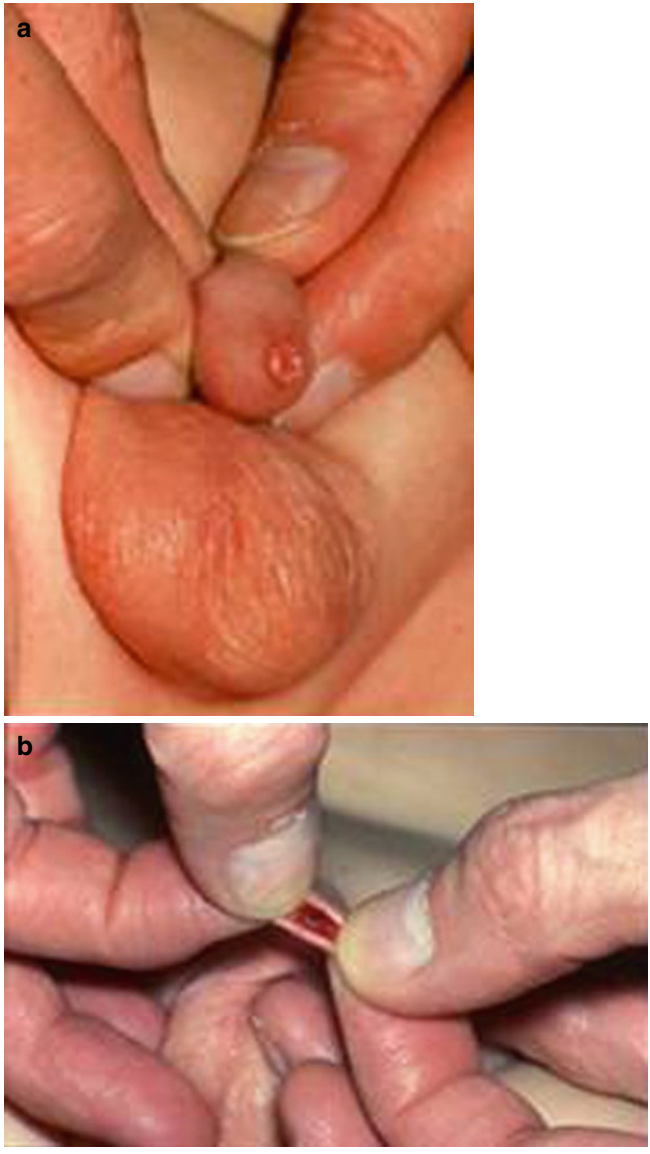


FIGURE 6.1. (**a, b**) Healthy non retractile foreskin-physiological phimosis: note the inner foreskin "pouts like a flower".

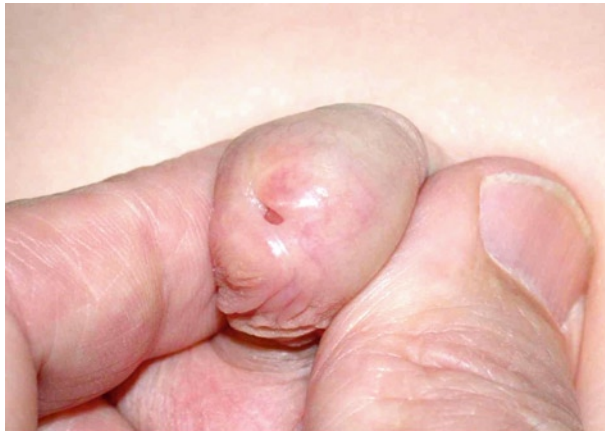


FIGURE 6.2. Balanitis Xerotica Obliterans: shutter type foreskin with obvious sclerotic margin that does not pout on gentle retraction.

the foreskin. In older pubertal children with a healthy non retractile foreskin, a preputioplasty may be considered.[4]

2. Localised collection of smegma pearls: No intervention is necessary.
3. Inflammatory conditions: balanoposthitis, balanitis, posthitis: Simple bathing, topical steroids, and antibiotics. If recurrent disabling attacks of balanoposthitis occur despite conservative management, a circumcision may be considered.
4. Balanitis Xerotica Obliterans: Circumcision.
5. Paraphimosis: Reduction with or without an anesthetic.[5]
6. Hooded foreskin: without hypospadias: no treatment, modified circumcision or foreskin reconstruction. If with hypospadias: no treatment, modified circumcision or foreskin reconstruction with hypospadias repair.

6.4 Indications for Referral

1. Recurrent severe balanoposthitis where conservative management has not been successful.

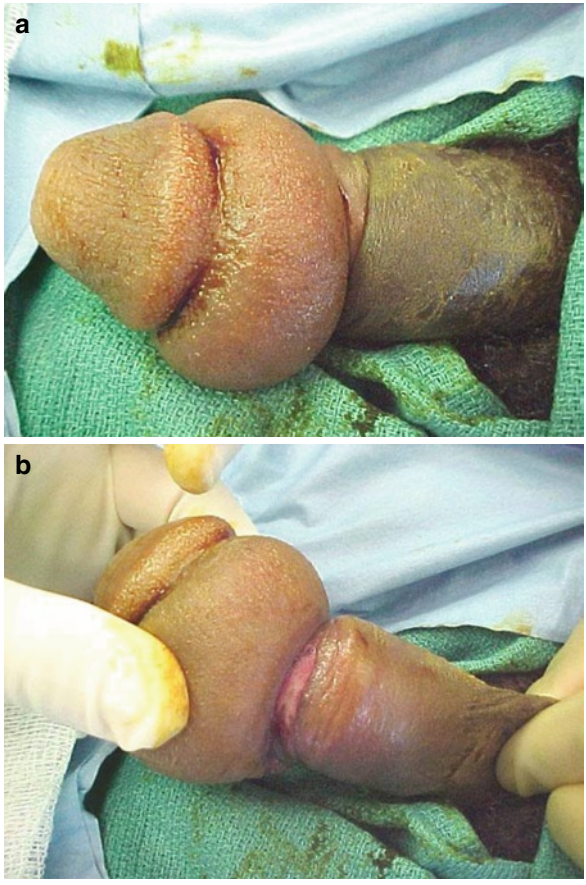


FIGURE 6.3. (**a, b**) Paraphimosis. Note the constricting band at the coronal sulcus.

2. Balanitis xerotica obliterans.
3. Paraphimosis where reduction is not possible with simple manual reduction.
4. Hooded foreskin with or without hypospadias: If parents wish surgery for cosmetic/functional reasons.

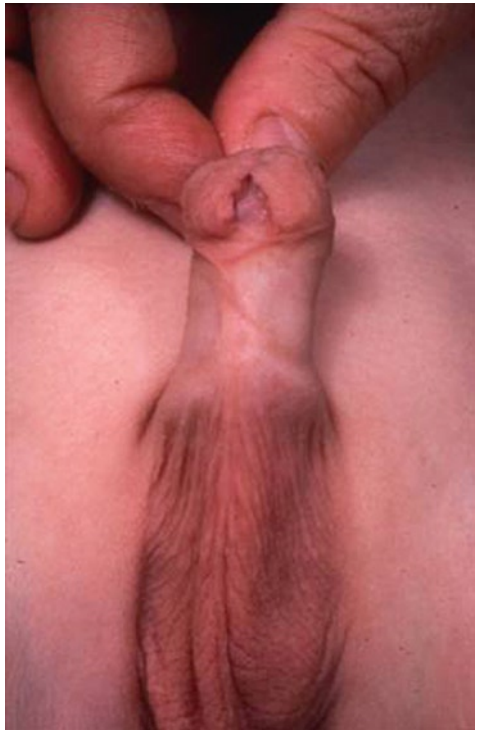


FIGURE 6.4. Hooded foreskin with distal hypospadias.

References

1. Gairdner D. The fate of the foreskin. *BMJ*. 1949;2:1433-1437.
2. Escala JM, Rickwood AM. Balanitis. *Br J Urol*. 1989;63:196-197.
3. Ashfield JE, Nickel KR, Siemens DR, et al. Treatment of phimosis with topical steroids in 194 children. *J Urol*. 2003;169(3):1106-1108.
4. Cuckow PM, Rix G, Mouriquand PD. Preputialplasty: A good alternative to circumcision. *J Pediatr Surger*. 1994;29:561-563.
5. Barone JG, Fleisher MH. Treatment of paraphimosis using the puncture technique. *Pediatr Emerg Care*. 1993;9:298-299.

Chapter 7
Disorders of Male External Genitalia: Hypospadias, Epispadias, Concealed Penis, and Urethral Disorders

Mark R. Zaontz

Key Points

- Diagnosis should be made at birth. Hypospadias is easily recognized by an incomplete hooded foreskin appearance and urethral opening anywhere beneath the tip of the penis. Hypospadias may or may not be associated with penile curvature.
- Circumcision is contraindicated when the diagnosis of hypospadias is made.
- The most common associated anomalies include undescended testes, hydroceles and/or hernias. Screening renal ultrasound is not routinely recommended unless there are other midline defects and/or if the hypospadias is proximal.
- In cases with undescended testes it is important to obtain a karyotype at birth, as there is a high incidence of intersex in this scenario.
- Beware of a complete foreskin hiding a hypospadias condition. Hence, there must be full foreskin retraction prior to performing circumcision to properly identify the meatal location.

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_7,

- Not all cases of hypospadias require surgical correction particularly those contained well within the glans in the absence of penile curvature.
- Boys with associated penile curvature, misdirected downward urinary stream and or abnormal positioning of the scrotum with respect to the penis require surgical correction.
- Timing for surgery is usually performed around 6 months of age as a single stage outpatient procedure in the majority of cases although, more severe cases may need to be completed in two stages.
- The most common complications of surgery include urethrocutaneous fistula, urethral/meatal stenosis and recurrent penile curvature, all of which are correctable surgically.

7.1 Hypospadias

7.1.1 Introduction

Hypospadias (Fig. 7.1) is one of the more common conditions treated by pediatric urologists. Its incidence is around 8–16/1,000 live births.[1] The penis is fully developed within the first 12–16 weeks of embryonic life so there is no associated increased risk of having hypospadias in premature neonates. This entity is usually referred after recognizing the defect at birth. The examiner will note in 95% of the cases that boys with hypospadias will have an incomplete hooded foreskin that is mostly dorsally located and that the opening of the penis is somewhere beneath the normal orthotopic location at the tip of the glans. The opening can be anywhere from 1 mm below the glanular tip to the level of the perineum in the most severe cases. The majority of hypospadias defects are distal in about 70% of the cases.

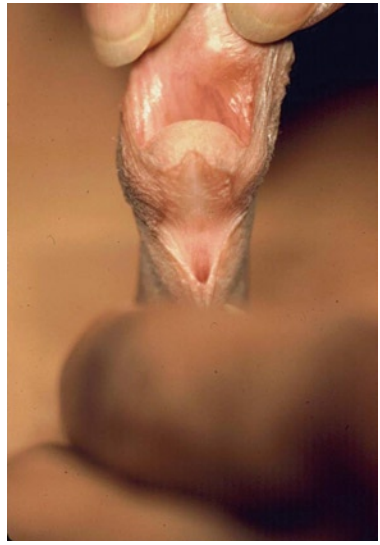


FIGURE 7.1. Example of classic distal hypospadias.

Practitioners need to beware of the boys born with a normal prepuce and have hidden beneath the foreskin hypospadias (Fig. 7.2a and b). This is noted in about 5% of hypospadias boys. Thus, it is imperative that all boys undergoing circumcision have their foreskins fully retracted before cutting, to determine where the urethral meatus is located, and if abnormal avoid circumcision.[4]

Penile curvature (Fig. 7.3), which commonly accompanies hypospadias, may or may not be appreciated at birth. Without seeing an erection, this is a difficult assessment especially in the boys with mild hypospadias. Curvature and/or ventral tethering of the penis is often more obvious visually in the boy with more proximal meatal locations.

On occasion the examiner may note an incomplete foreskin that has an orthotopic meatus. This boy has the hypospadias complex without hypospadias and, may or may not have penile curvature. Circumcision should be avoided in these boys.

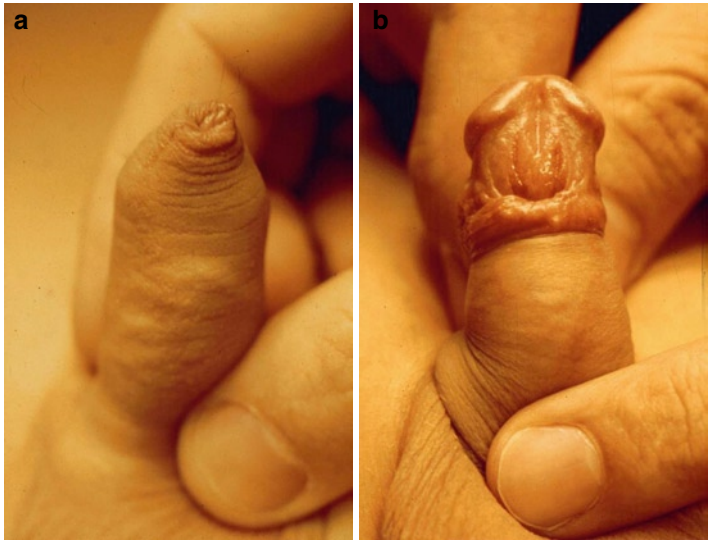


FIGURE 7.2. (**a**) Complete prepuce. (**b**) Same boy showing distal megameatus hypospadias after retracting foreskin.

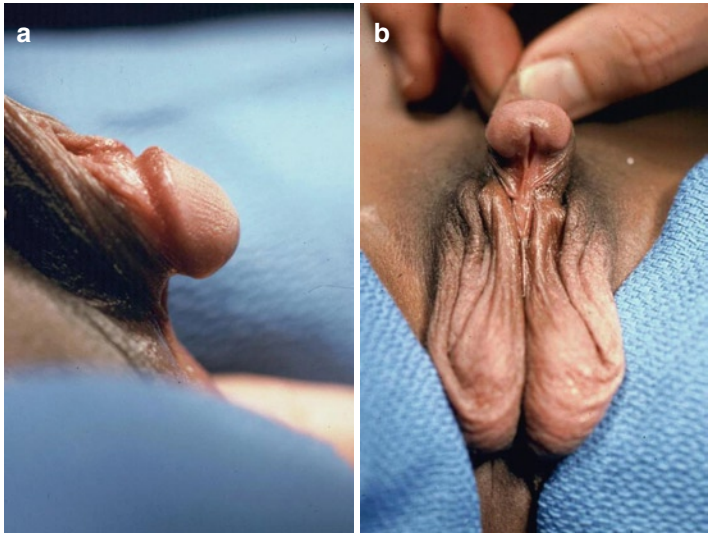


FIGURE 7.3. (**a, b**) Proximal hypospadias with ventral chordee.

Associated anomalies found in about 10–15% of boys with hypospadias include undescended testes, hernia and hydrocele. In boys that have associated undescended testes or nonpalpable testes, it is imperative to obtain a serum karyotype, as intersex conditions are common. Ultrasound screening of the kidneys are not routinely recommended for the majority of boys with hypospadias. However should he have a penoscrotal or more proximal meatus or associated midline anomalies, then it is reasonable to perform the ultrasound exam to rule out potential renal anomalies.

Circumcision should be avoided in all of the aforementioned scenarios so that the pediatric urologist can operate on a virginal, nonscarred penis and also has the ability to use the foreskin as needed for reconstructive purposes.

Fertility in boys with hypospadias, to our knowledge is not different from that of boys without hypospadias.

7.1.2 Management Issues

Assuming that the boy with hypospadias was born at term and had no significant respiratory or hematological issues, surgical timing for repair of hypospadias is recommended for around age 6 months as an outpatient procedure. In cases of severe prematurity, a history of respiratory problems, especially a history of apnea, surgery is generally held off until the infant is one year of age.

In the majority of cases, hypospadias repair is done as a single stage procedure. However, in cases of proximal hypospadias with severe chordee with or without penoscrotal transposition, a two-stage approach may be used. Depending on the appearance of the penis preoperatively and in particular the size of the glans, preoperative depotestosterone may be given intramuscularly in the thigh or given as a cream directly to the penis to enhance growth and help facilitate surgery. This is generally taken care of by the pediatric urologist's office but certainly the primary care physician can arrange to give the shots in their office if desired.

Not all infants with hypospadias require surgery, especially if the meatus is well within the glans and the urinary stream is full and well directed. Also if there is no evidence of penile curvature in these same boys, surgery can be avoided.

The goals of surgery are to bring the meatus to the tip of the glans which will enable normal controlled voiding, to straighten the penis if curvature is present, to create a cosmetically "normal" appearance of the penis similar to that of a normal circumcised male and to allow for normal sexual function.

7.1.3 Indications and Timing of Referral

All boys with hypospadias or questionable hypospadias and/or penile curvature should be referred to pediatric urologist at the time of diagnosis shortly after birth or while in the hospital. This will allow the specialist to counsel the family as to future plans and expectations for their child. Typically the patient will be seen between 3 and 4 months post natally for a second visit to assess him for penile growth and then to determine the need for hormonal supplement preoperatively. Typically if they are to receive depotestosterone, it will be administered 5 weeks before the procedure and repeated 2 weeks preoperatively such that the surgery will be performed as close to the 6-month mark as possible.

From a psychological standpoint it is recommended that genital reconstructive surgery be performed between 6 and 18 months of life with the ideal being under 1 year. There is no question that successful hypospadias surgery can be done beyond that recommended age, but the patients generally fare better emotionally and psychologically if done within the recommended time frame.

7.1.4 Complications of Surgery

Unfortunately, surgery has the risk of complications. Among the most common complications from hypospadias surgery is a urethrocutaneous fistula (Fig. 7.4), which presents as a leak

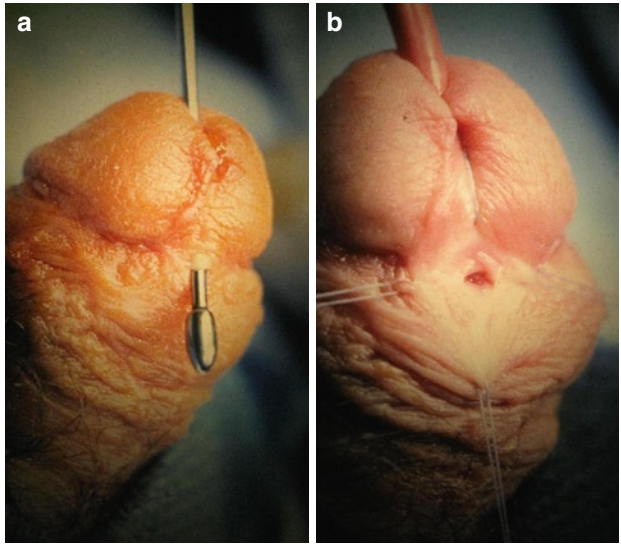


FIGURE 7.4. (**a, b**) Urethrocutaneous fistula noted after hypospadias repair.

somewhere below the newly reconstructed new urethral opening. This may be difficult to notice especially when the infant is still in diapers and is the fistula is very small. Typically the parents will notice a small drip or fine spray from an abnormal location and bring it to the doctor's attention. Depending on when the surgery had taken place will usually determine how long one needs to wait to perform outpatient excision and closure of this fistula. As long as the baby is comfortable, voids without straining and has no evidence of infection, a wait and see attitude is appropriate. Some fistulas close spontaneously but, the ones that do not can be readily corrected after the surrounding tissue softens up to allow success closure usually around 6 months after the initial surgery.

Meatal and/or urethral stenosis is another common complication heralded by the parents noting a fine thin or dribbling urinary stream or noting the baby straining to void. This requires prompt referral to prevent other significant problems

such as infection. Depending on the scenario this may require simple office dilatation or minor meatotomy. However a more proximal stricture will require definitive surgery that may in fact include reopening the repair to where the urethral is normal and then reconstruct the urethra at a later date.

Recurrent or persistent penile curvature will be brought to the pediatric urologists' attention by the parents after seeing an erection or referred by the primary care giver. Repeat correction is usually delayed at least 6 months from initial surgery.

Urethral diverticulum (Fig. 7.5) will present as a distinct bulge below the glans again usually seen by the parents while watching their child void. The mild diverticulum can be observed as long as the voiding pattern is good and there is no infection. However the large diverticulum require surgical reconstruction. These are far more common in proximal hypospadias repairs.[6]

Glans/urethral dehiscence (Fig. 7.6) is another complication that fortunately is uncommon but requires referral when seen and may or may not require reconstruction at a later date depending on the degree and associated symptoms.

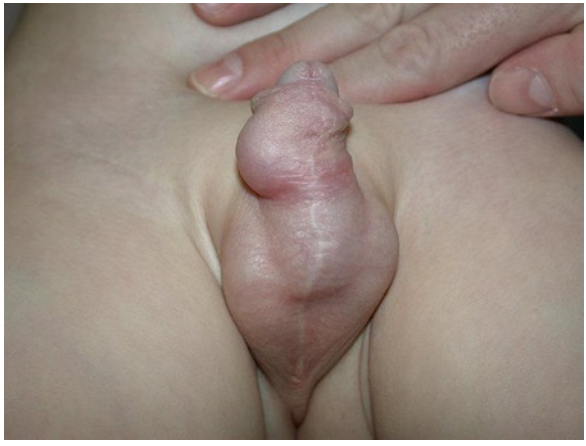


FIGURE 7.5. Urethral diverticulum after proximal hypospadias repair. Note ventral bulging.

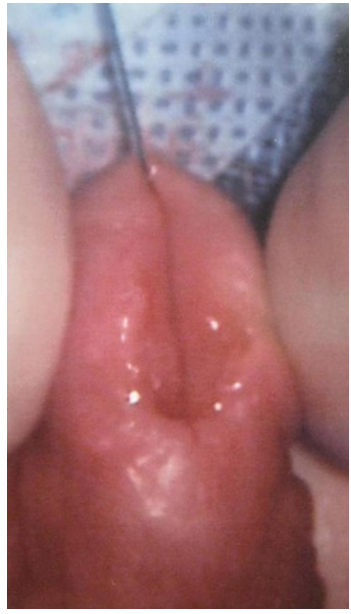


FIGURE 7.6. Glans dehiscence after hypospadias repair.

7.2 Epispadias

Key Points

- Epispadias is a rare condition that presents with a dorsal urethral opening below the normal meatal opening. Cosmetically the penis appears split when looking down on it and the urethra appears as an open defect like a trench.
- This is a rare condition, which presents as a spectrum ranging from very mild glanular epispadias to the most severe form, which is associated with bladder exstrophy.
- Children born with the complete exstrophy-epispadias complex will need prophylactic antibiotics to prevent urinary tract infection in the face of existing vesicoureteral reflux.

- In most boys with epispadias the penis is generally wider, foreshortened and is associated with dorsal chordee where the penis bends back toward the abdomen.
- The meatal location in epispadias determines whether or not urinary incontinence is an issue secondary to an incompetent bladder neck.
- Associate anomalies of other organ systems are very uncommon such that routine screening studies are not performed for isolated epispadias that does not involve the bladder neck. Epispadias-exstrophy complex requires radiographic screening to rule out vesicoureteral reflux and renal anomalies.
- Retrograde ejaculation is common in men with a penopubic epispadias and can lead to fertility problems.
- Penile size and dorsal penile curvature can adversely affect sexual intercourse.
- Correction of epispadias can be done anywhere from the first few days of life in the presence of exstrophy to 6 months post natally for the less severe cases.
- Complications are similar to hypospadias except for the issues associated with urinary continence, overall smaller penile size and difficulty correcting penile curvature.

7.2.1 Introduction

Epispadias (Fig. 7.7) is a rare condition that affects boys in about 1:100,000 live births.[2] It represents a condition where the foreskin is ventrally situated and the penis is widened, foreshortened and bends upward toward the abdomen. Because the penis is attached proximally to the pubic bones

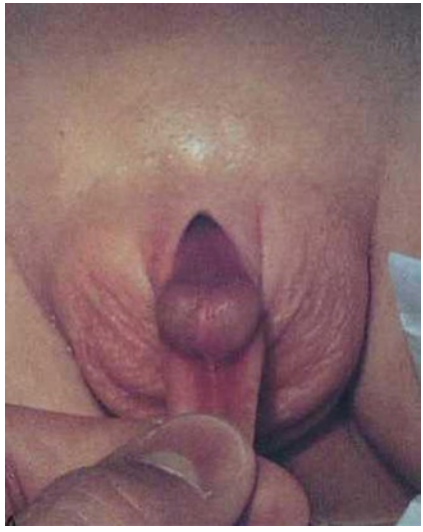


FIGURE 7.7. Epispadias.

that are separated in both the penopubic and the more severe cases of epispadias, the penis is pulled inward toward the abdomen. The urethral opening can be anywhere from just below the tip of the penis dorsally in the glans to the penopubic location and in the most severe cases presents as the exstrophy-epispadias complex (Fig. 7.8) where the entire bladder and the penis is "split open " as if cut across in a complete transverse plane.

This condition is readily diagnosed at birth, although frequently incorrectly diagnosed in the nursery setting. In that scenario, the foreskin is complete and because the preputial opening turns upward, consults are inappropriately made to rule out epispadias. This is in fact a common and normal finding for normal foreskin. It is important to recognize that in epispadias the foreskin is hooded and only on the ventral surface of the penis. Hence the epispadias defect is very apparent.

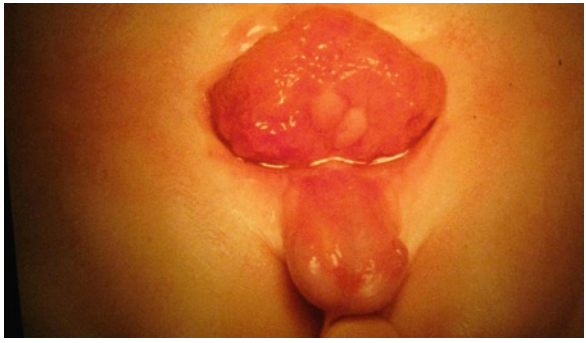


FIGURE 7.8. Exstrophy-epispadias complex. Note the filleted appearance of the penis.

7.2.2 Management Issues

Referral to a pediatric urologist should be made at the time of diagnosis, which is usually while the baby is in the nursery. While isolated epispadias is not a urologic emergency and can wait several months prior to surgical reconstruction, complete bladder exstrophy-epispadias is a more urgent matter. This condition is usually surgically managed within the first few days of life. In addition because of the presence of vesicoureteral reflux in these severe cases, prophylactic antibiotics are recommended from day one of life.

Fortunately, babies born with isolated epispadias and even the exstrophy-epispadias complex are quite healthy at birth. Isolated epispadias does not usually require any screening study, as the incidence of associated anomalies is very low. However, those boys with the exstrophy-epispadias complex require radiographic evaluation especially because of the known coexisting incidence of vesicoureteral reflux and the risk for renal anomalies such as an absent kidney.

Penopubic epispadias or exstrophy-epispadias will have problems associated with urinary incontinence as they get into toilet training even after primary surgical reconstruction because this population has an incompetent bladder neck.

Early surgical reconstruction in this group has many advocates because this will help the bladder capacity expand which may ultimately benefit urinary continence and increase the chances for normal bladder control.

Fertility is another issue that affects the penopubic epispadias patients. As a result of the bladder neck not being able to close completely, retrograde ejaculation is very common. Additionally, a congenitally short broad penis with dorsal penile curvature may increase difficulty in having sexual intercourse.

7.2.3 Surgery, Common complications, and Postoperative Issues

Today epispadias is repaired as a single stage procedure that attempts to repair the penis in an anatomically correct fashion. However, as with hypospadias certain complications are common such as a urethrocutaneous fistula that usually presents with a separate urethral opening either ventrally or dorsally depending on the reconstructive technique employed. This may or may not close spontaneously and if not, will require surgical correction. Persistent dorsal penile curvature (Fig. 7.9) has been a very common complication post epispadias repair. However, using the newer Mitchell disassembly technique, it is less common than before.[8]

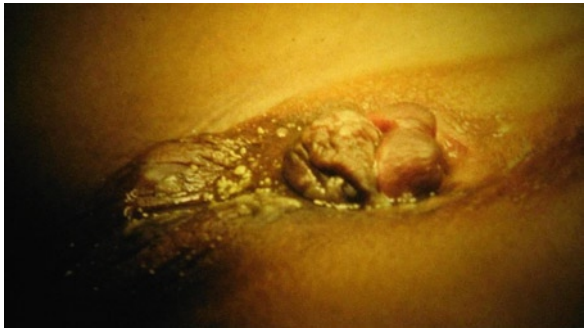


FIGURE 7.9. Dorsal chordee as seen in epispadias.

Persistent urinary incontinence due to an incompetent bladder neck is a difficult problem in boys with a history of proximal penopubic epispadias or the exstrophy-epispadias complex. Secondary procedures are often necessary to try to improve continence. These include the use of minimally invasive endoscopic bulking agents to the use of bladder neck open reconstruction, slings and/or artificial urinary sphincters each of which carry their own potential risks and complications.

7.3 Concealed Penis

Key Points

- The penis at birth may be partially concealed due to a variety of factors including: (1) Excessive deposits of fat suprapubically, (2) Penoscrotal web, (3) Bands of tissue deep to the penis pulling under the abdominal wall, (4) Poor penile suspension, (5) Result of a circumcision, or (6) Congenital micropenis.
- Endocrine consult must be obtained in cases of true micropenis.
- In the other presentations of concealed penis, the phallus is generally normal in size and length but appears small due to its hidden appearance.
- Newborn circumcision is contraindicated in cases of significant concealment and the infant will undergo a formal correctable surgical reconstruction between 6 and 12 months of life as an outpatient procedure.
- In cases where the penis is clearly concealed or there are questions regarding possible concealment, circumcision should be deferred until consulted by the pediatric urologist.

7.3.1 Introduction

The hidden or concealed penis (Fig. 7.10) represents a concern to the parents of their newborn son. If this is not handled properly, an ill-advised circumcision may compound this problem and, if not attended to properly, may lead to both mechanical and psychosocial problems in the older boy. Fortunately, most boys with a concealed penis can be recognized at birth and appropriate urological consultation obtained before circumcision is approved.

There are a variety of causes for the concealed penis some of which are combinations of etiologies.[3, 5] These may include the presence of a large suprapubic fat pad or mons pubis (Fig. 7.11), the presence of a penoscrotal web, bands of tissue deep to the penis that pull the penis inward, poor skin suspension where the suprapubic fascia fails to anchor the penile skin to the deeper fascia preventing the normal penile contour, complication of circumcision and the presence of a micropenis.

Micropenis (Fig. 7.12) represents a congenitally small penis that is readily apparent by measuring the penis on

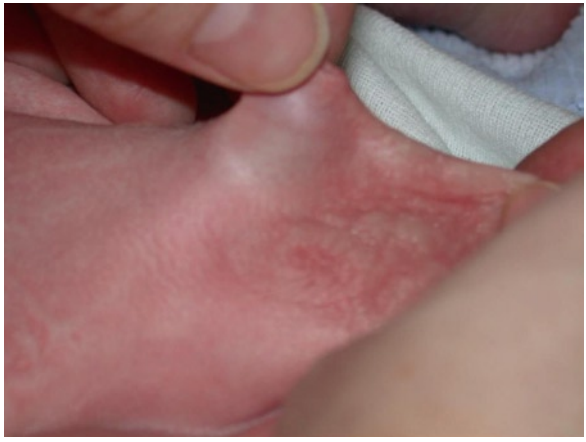


Figure 7.10. Concealed penis with penoscrotal webbing.

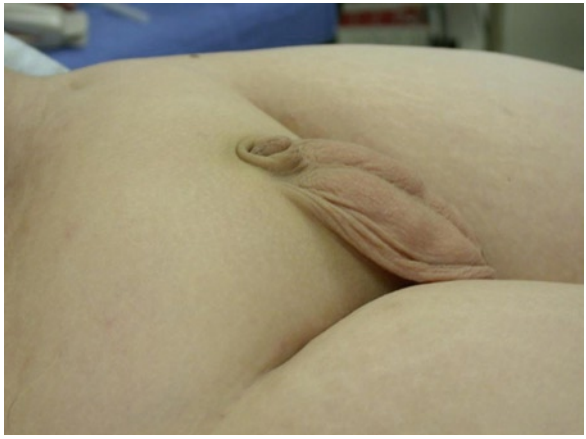


FIGURE 7.11. Obese male with concealed penis.

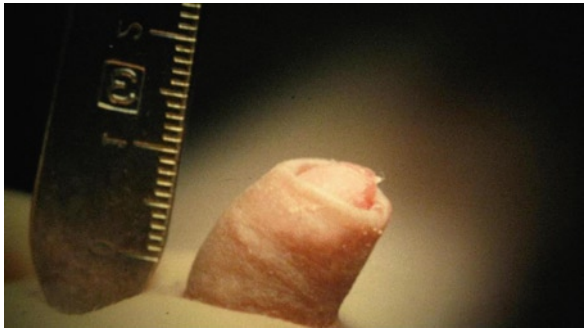


FIGURE 7.12. Micropenis in boy with Fanconi's syndrome.

stretch from the tip of the penis to the pubic bone. If the penis measures in the newborn less than 2.8 cm (2 standard deviations below the norm), then the diagnosis is made and endocrinology need to be consulted.[7]

The concealed penis that is due to the etiologies other than congenital micropenis is generally of normal size and length.

7.3.2 Referral and Treatment

In the newborn period while the baby is in the hospital, the pediatrician needs to make the determination as to whether there is a penile issue, which would preclude circumcision until evaluated by a specialist. It is far better to consult a specialist especially in situations where there is concern of potential penile concealment but not completely obvious.

Many times a consult is called for a concealed penis that is webbed. While in many cases this is correctly diagnosed, one can perform a simple test to see if circumcision should be deferred. By placing the index and middle finger along each side of the penis (without actually touching the penis) and then pushing down toward the pubis, the penis will become exposed. If there is no ventral tethering of the penile shaft and one sees a good shaft with the penoscrotal web being obviated, then circumcision can be performed. The parents however must be taught postoperative care because many of these boys will have penile retraction inward post operatively. In those cases, one needs to "pop" the penis out during each diaper change as already described to prevent adhesions and other post circumcision issues.

If the pediatrician or family practitioner performs the newborn circumcisions in the hospital then it is imperative to use the proper technique. In all of these children, the foreskin needs to be totally freed up from its adhesions to the glans and the meatal location recognized first. This can be done with a small hemostat and no dorsal slit need be done. Next the penis needs to be fully exposed after reducing the foreskin over the glans, again using the two-finger technique described. Next a marking pen needs to be used to outline the coronal margin of the glans. Finally whether one uses a Mogen Clamp or Gompco clamp, the foreskin is pull up to the marking pen line and then the clamp tightened and skin removed. If the doctor charged with the circumcision is uncomfortable, then the referral should be made to see the pediatric urologist in the hospital or in the office within the first 30 days of life for the procedure.

The boy who has significant concealment preventing newborn circumcision will generally be scheduled for surgical reconstruction, which includes the circumcision and correction of his concealment between 6 and 12 months of age.

There are a number of techniques in the literature for correction of the concealed penis.[5] One of the more popular approaches in the absence of significant webbing is to use internal suture fixation at various points at the base of the penis and affix them to the corresponding dermis of the abdominal wall-shaft skin juncture. In the presence of a penoscotal web where the ventral penile shaft is deficient, a scrotoplasty is performed and the lateral skin edges are transferred ventrally to create a cosmetically normal ventral shaft. Sometimes there is an obese prepubescent or adolescent male who might benefit liposuction to remove the large suprapubic fat pad. Often in those situations, the liposuction by itself allows improved exposure of the previously hidden penis. However, these children need to be placed on a diet regimen and exercise program to prevent recurrent problems.

7.3.3 Complications

Penile adhesions may be treated by the use of topical steroid creams.

Penile skin bridges may require surgical division, many of which can be done in the surgeon's office.

Recurrent penile concealment may improve spontaneously on its own with tincture of time but, in cases where a cicatrix scar forms and prevents penile exposure, first try topical steroid treatment. If that were to fail to loosen up the adhesions and expose the penis, then repeat surgery is necessary.

7.3.4 Benign Urethral Lesions in Boys

Urethral polyps are congenital benign growths of fibrous stalks of tissue and may present with gross hematuria, symptoms of a urinary tract infection or problems voiding.

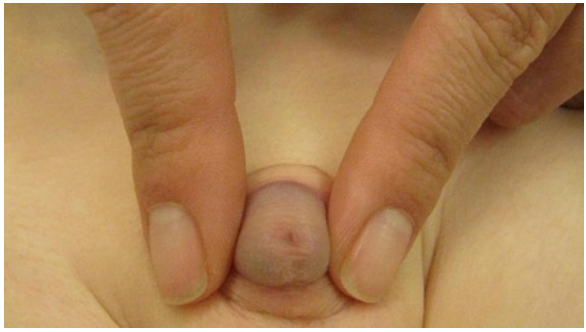


FIGURE 7.13. Meatal stenosis in a circumcised boy.

Diagnosis is made by voiding cystourethrography (VCUG) and cystoscopy.

Meatal stenosis (Fig. 7.13) occurs in only males and represents a tiny urethral opening in the orthotopic position. This is most commonly diagnosed during puberty when the parents note a thin and/or misdirected urinary stream. Dysuria is also a common symptom and bloody spotting may occur. This is seen predominantly in circumcised males.

Congenital urethral fistula is rare but the findings are similar to fistula seen after hypospadias repairs. Observation and physical exam make the diagnosis.

Anterior urethral diverticuli are pouch like enlargements of the urethra. They may present either as a saccular form where the diverticulum arises from the floor of the urethra or the megalourethra, which involves the entire anterior urethra. Both entities are discovered by observation during voiding whereby a weak urinary stream is observed in conjunction with visible expansion and bulging of the urethra. Boys with Prune Belly Syndrome are at risk for having megalourethras.

Cowper's Duct cysts result when the bulbourethral glands, which are located beneath the prostate gland, expand into cystic like structures due to abnormal narrowing of the glands' exit passageway. They can cause discomfort during voiding, bloody spotting and a weakened urinary stream with associated post void dribblig. The diagnostic test of choice is a VCUG.

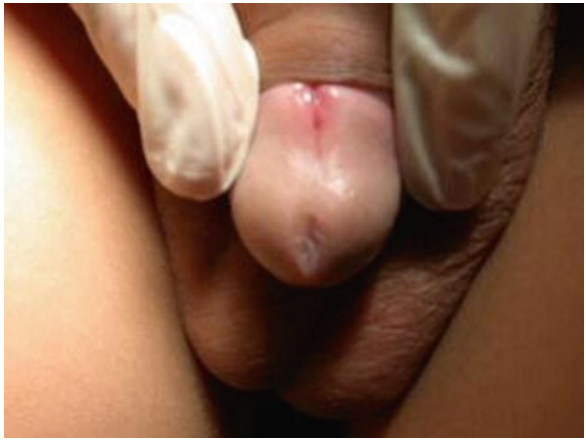


FIGURE 7.14. Urethral duplication. Note the dorsal located "extra" meatus.

Urethral duplication (Fig. 7.14) is rare and is represented by a spectrum from simply having a dorsal blind ending sinus tract below the normally located meatus to where there is not only a complete duplicated urethra but a duplication of the bladder as well. They may present as a serendipitous physical finding or by noting two distinct separate urinary streams. They may also present with infection and/or obstructing symptoms in the other urethra.

7.3.5 Treatment

When urinary symptoms as noted above occur, referral to pediatric urology is indicated to complete the workup. Once a diagnosis is made surgical correction is individual tailored to the condition.

Urethral polyps are excised and the base fulgurated by using minimally invasive cystoscopy.

Meatal stenosis can be corrected either as a meatotomy in the office setting with local topical anesthetic in a cooperative child or, as a short outpatient procedure by performing a

urethromeatoplasty to enlarge the urethral opening. It is important to provide instructions for the patient to apply ointment at least twice a day and to spread the raw edges apart until healing occurs in about 2 weeks.

Congenital urethral fistula is repaired using the same techniques for fistula resulting after hypospadias surgery as outpatient procedures.

Congenital urethral diverticulum is corrected much the same way as that done for diverticulum occurring after hypospadias repair. In addition a complete urological evaluation is necessary since there is a high association of other urologic problems.

Cowper's duct cysts are treated by endoscopic resection to relieve symptoms. If this is impossible, then open surgery is done.

Urethral duplication does not require treatment for a blind ending sinus or incomplete duplication that is not causing any adverse symptoms. A simple technique when there are two streams and the urethral openings are close to one another would be to connect the openings together to create one single urinary stream. Some urethral duplications are associated with penile curvature, which then needs correction and the extra urethra excised.

7.3.6 Follow-Up After Treatment

Urethral polyps once successfully resected should result in resolution of preoperative symptoms.

After surgery for meatal stenosis, the child may experience dysuria for a few days that may be easily treated with a urinary analgesic in the older children. In the young boy who cannot swallow pills, placement in a warm tub of water will allow him to void with less discomfort. Also by spreading the meatus and placing ointment over the raw edges a few times per day will also improve any discomfort. The key to preventing a recurrence is the frequent and active spreading of the raw edges after surgery for up to 2 weeks until there is no more crusting over the wound and the meatus is well healed.

Urethral fistula follow-up is similar to hypospadias. Patients are generally seen at 2 weeks, 3 months, 1 year, and puberty.

Urethral diverticulum secondary to the megalourethral fusiform variant are not going to have a functional penis except for the passage of urine due to the congenital absence of corporal tissue. The milder varieties should have acceptable penile function after successful repair.

Cowper's duct cysts after resection are essentially symptom free.

Connecting the urethral meati should relieve the symptoms in urethral duplications when surgery is needed.

References

1. Zaontz MR. Nuances of hypospadias. In: Kramer SA, Paulson DF, eds. *Problems in Urology*, vol. 4. Philadelphia: JB Lippincott Co; 1990:705-717.
2. Zaontz MR, Packer MP. Abnormalities of the external genitalia. *Pediatr Clin North Am*. 1997;44(5):1276-1297.
3. Maizels M, Zaontz MR, Donovan J, et al. Surgical Correction of the buried penis: Description of a classification system and a technique to correct the disorder. *J Urol*. 1990;136:268-271.
4. Hatch DA, Maizels M, Zaontz MR, Firlit CF. Hypospadias hidden by a complete prepuce. *Surg Gyn Obst*. 1989;169(3):233.
5. Zaontz MR. The concealed penis. *Dialog Pediatr Urol*. 2006;28:1-9.
6. Zaontz MR, Kaplan WE, Maizels M. Surgical repair of anterior urethral diverticula after hypospadias repair in children. *Urology*. 1989;33:39-42.
7. Zaontz MR. Congenital microphallus. In: Seidmon EJ, Hanno PM, eds. *Current Urologic Therapy*. 3rd ed. Philadelphia: WB Suanders Co; 1994:436-437.
8. Zaontz MR, Steckler RE, Shortliffe, LMD et al. Multicenter experience with the Mitchell technique for epispadias. *J Urol*. 1998;160(1): 172-176.

Chapter 8
Disorders of Male External Genitalia: Undescended Testis

Michael C. Large and Mohan S. Gundeti

Key Points

- Cryptorchidism, or undescended testis, occurs in 3% of full term males.
- Spontaneous descent rarely occurs after age 6 months.
- Surgical therapy minimizes the risk of infertility in men with an undescended testicle.
- Orchidopexy improves the patient's ability to perform testicular self-examination, and when done at an early age, may marginally decrease the relative risk of testicular cancer in an undescended testicle.

8.1 Introduction

Three percent of term male infants have cryptorchidism, or undescended testis, while approximately 17% of premature males are affected.[1] The rate of spontaneous descent is debatable, with incidence ranges of 10–70% reported.[1,2] The testis may be abdominal, inguinal, scrotal or ectopic. The rationales for repair are many:

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_8,

1. Men with an undescended testis appear to have higher infertility rates and lower sperm quality and counts when compared to males with bilateral descended testes.[3]
2. Spermatic indices in undescended testes reach critically low levels by 9 months age, and spontaneous descent is very rare after age 6 months.[2,4] Thus, repair is frequently performed during this time.
3. Infertility rates worsen if both testes are affected.[5]
4. Improved physical examination of the testis allows earlier detection of malignancy.[6]
   a. The contralateral descended testis appears to have no increased risk of malignancy.[7]
   b. The relative risk of testicular cancer in a cryptorchid testis repaired by age 12 is 2–3, but 2–6 when repaired after age 12. As such, orchiectomy should be considered in patients between 12 and 50 years of age.[7]
5. A vast majority of undescended testes are associated with a patent processus vaginalis and thus increased likelihood of hernia.[8]
6. Torsion may occur relatively more frequently in the undescended testis.[9]

8.2 Common Conditions

Three types of nonpalpable testicle exist: undescended, ectopic and retractile. Radiographic studies for the undescended testicle are not warranted as their negative predictive values are shy of the requisite 100%.[10]

1. Undescended testes have a normal gubernacular attachment but have failed to reach the scrotum during descent. The examiner gently sweeps from the internal inguinal ring caudally, sometimes with the aid of lubricant, and frequently will feel the oval testis roll under the finger and then retract inguinally when released (Fig. 8.1).
2. Ectopic testes have an abnormal gubernacular attachment and may be found in the thigh, prepubic or abdominal regions. If maneuvered into the scrotum, the testis will immediately retract to its ectopic position once released.

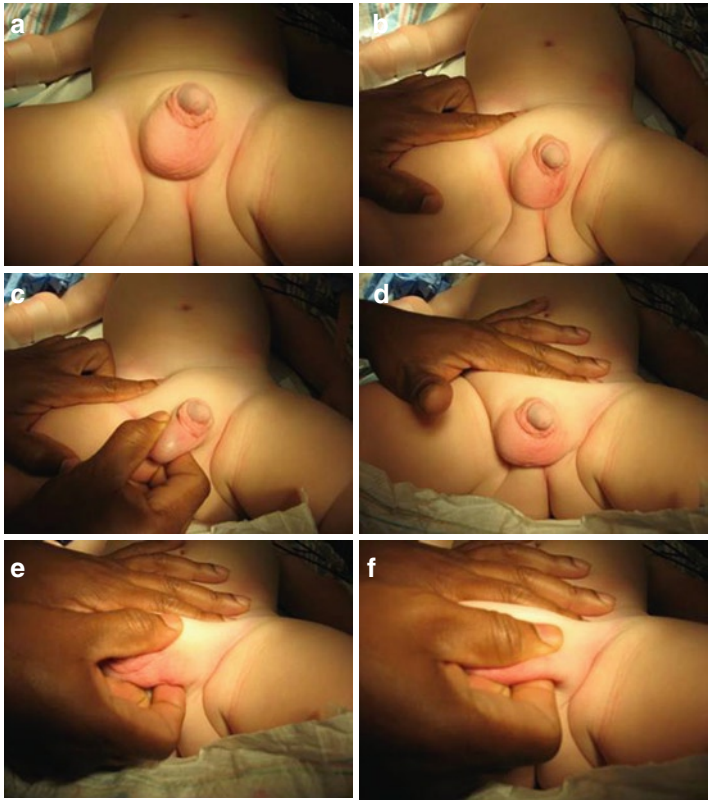


FIGURE 8.1. Physical examination of the testicles. (**a**) Appearance of empty left hemi-scrotum. (**b**) Application of pressure at external inguinal ring, directed toward scrotum. (**c**) Palpation of right testicle. (**d**) Application of pressure at the left external ring. (**e**). Attempt at palpating the left testis between index finger and thumb. (**f**). Milking movement of finger and thumb distally in an attempt to delineate testis (here, no left testis was palpable).

3. Retractile testes have an exaggerated cremasteric response. If manipulated into the scrotum, they will remain there until the cremasteric reflex is stimulated.
4. Risk factors include family history, prematurity, low birth weight or gestational size, multiple gestation, Eagle-Barrett syndrome, or in vivo exposure to estrogen.

8.3 Treatment of Undescended Testis

1. Hormonal therapies have poor success rates as a high local concentration is required to induce descent.[11]
2. Surgical repair may be scrotal, inguinal, abdominal, laparoscopic or robotic-assisted. The approach is dictated by the location of the testis and surgeon's preference (e.g., an inguinal testicle may be repaired through an inguinal or high-scrotal incision).
3. Occasionally, nonpalpable testes that are visualized intraabdominally are not able to be placed intrascrotally, in which the testicular artery and vein are ligated and the child is reexplored in 6 months in hope that collateral supply will allow increased mobilization.
4. Blind-ending vessels indicate absence of the testis (a testis may be present in spite of a blind-ending vas deferens).
5. The treatment of small testicular nubbins is debatable: some surgeons remove them citing rare reports of CIS developing, while others will leave them in situ.[7]

8.4 Indications for Referral

1. Examination by a urologist should be performed by age 3–6 months, with repair indicated at age 6 months if spontaneous descent has failed to occur.
2. Presence of ambiguous genitalia or severe hypospadias alongside undescended testes mandates a karyotype and multi-disciplinary workup with urologic involvement. Elevation of LH and FSH with absence of Müllerian inhibiting substance strongly suggests absence of testes.

References

1. Berkowitz GS, Lapinski RH, Dolgin SE, et al. Prevalence and natural history of cryptorchidism. *Pediatrics*. 1993;92:44-9.
2. Wenzler DL, Bloom DA, Park JM. What is the rate of spontaneous testicular descent in infants with cryptorchidism? *J Urol*. 2004;171:849-51.

3. Kogan SJ. Fertility in cryptorchidism. An overview in 1987. *Eur J Pediatr*. 1987;146(Suppl 2):S21-S24.
4. Wilkerson ML, Bartone FF, Fox L, et al. Fertility potential: a comparison of intra-abdominal and intracanalicular testes by age groups in children. *Horm Res*. 2001;55:18-20.
5. Friedman RM, Lopez FJ, Tucker JA, King LR, Negro-Vilar A. Fertility after cryptorchidism: a comparative analysis of early orchiopexy with and without concomitant hormonal therapy in the young male rat. *J Urol*. 1994;151:227-33.
6. Swerdlow AJ, Higgins CD, Pike MC. Risk of testicular cancer in cohort of boys with cryptorchidism. *BMJ*. 1997;314:1507-11 [Published erratum appears in BMJ 1997;315:1129].
7. Wood HM, Elder JS. Cryptorchidism and testicular cancer: Separating fact from fiction. *J Urol*. 2009;181:452-61.
8. Elder JS. Cryptorchidism: Isolated and associated with other genitourinary defects. *Pediatr Clin North Am*. 1987;34:1033-53.
9. Schultz KE, Walker J. Testicular torsion in undescended testes. *Ann Emerg Med*. 1984;13:567-9.
10. Dessireddi NV, Liu DB, Maizels M, et al. Magnetic resonance arteriography/venography is not accurate to structure management of the impalpable testis. *J Urol*. 2008;180(4 Suppl):1808-9.
11. Rajfer J, Walsh PC. Hormonal regulation of testicular descent: experimental and clinical observations. *J Urol*. 1977;118:985-90.

Chapter 9
Disorders of Male External Genitalia: *Circumcision*

Garrett Pohlman and Duncan Wilcox

Key Points

- The majority of boys will have a fully and easily retractable foreskin by physical maturity.
- Benefits of circumcision include lower risk of UTI's in male infants, a protective affect against invasive penile cancer, and reduced incidence of STD's.
- Circumcision should not be performed in newborns with hypospadias, epispadias, ambiguous genitalia, hidden penis, chordee without hypospadias, webbed penis, micropenis, or dorsal hood deformity.
- The complication rate for newborn circumcision ranges from 0.2% to 3%.

9.1 Natural History of the Prepuce

The prepuce is apparent prenatally at 8 weeks of gestation as a ridge of thickened epithelium. The prepuce grows forward over the glans to completion by 16 weeks of gestation. Early in gestation there is no separation between the epithelium lining the prepuce and glans. Preputial adhesions

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_9,

are a characteristic of normal development with initiation of spontaneous separation by desquamation late in gestation. The rate of preputial separation after birth is variable with up to 70% of boys having some degree of preputial adherence at 5 years of age. Consequently a "non-retractile foreskin" or more appropriately described "physiologic phimosis" at birth is normal. The majority of boys will have a fully and easily retractable foreskin by physical maturity.

9.2 Benefits of Circumcision

1. UTI's: Circumcised males infants are at a lower risk of UTI's than uncircumcised infants. The effect of circumcision on UTI's has been studied primarily in infants because they have a higher prevalance of UTI's than older males. The incidence of UTI in circumcised male infants has been reported at 0.10% versus 1% in uncircumcised male infants. Despite the decreased risk of UTI's in circumcised infants it has been reported that as many as 195 circumcisions would be needed to prevent one hospital admission for UTI in the first year of life.
2. Cancer: Neonatal circumcision has a protective effect against invasive penile cancer.
3. STD's: Circumcision has been demonstrated to reduce the incidence of HSV-2 infection and the prevalence of HPV infections. Circumcision is also associated with a reduced risk of HIV infection.

9.3 Absolute Indications for Circumcision

1. Paraphimosis refractory to manual reduction
2. Recurrent balanoposthitis with failed conservative management
3. Balanitis xerotica obliterans

9.4 Relative Indications for Circumcision

1. Persistent phimosis
2. To reduce the rate of STD's and HIV particularly in sub Saharan Africa
3. Cultural and Religious preference

9.5 Surgical Options

Circumcision is most often performed on neonates using the Gomco clamp or one of the plastic disposable devices such as the Hollister Plastibell. Circumcision should be performed on an infant no earlier than 12–24 h old. Various options are available for analgesia ranging from allowing the infant to suck on a sucrose solution, topical anesthetics (e.g., EMLA cream), to various local blocks (e.g., dorsal penile nerve block or ring block) using 1% local anesthetic without epinephrine. It is important to free the glans completely from the inner mucosal layer of the foreskin. It is essential to observe that an equal amount of foreskin is present circumferentially when pulling the foreskin into the clamp. Care should also be taken to pull a proper amount of foreskin into the clamp to avoid taking too much or too little foreskin with the circumcision. Routine post-circumcision care includes application of an ointment (e.g., triple antibiotic) to the wound for several days and resumption of normal bathing.

After several months of age circumcision should be performed as a formal procedure with general anesthesia in addition to a local anesthetic. Several different surgical techniques are employed and are a matter of individual preference. Circumcision clamps are not recommended in the older patient as the vessels become larger and are not easily sealed by compression. As in the neonate, care is taken to separate the prepuce from the underlying glans to prevent formation of persistent skin bridges between the glans and penile skin. The outer and inner layers of preputial skin are excised separately watching closely to excise sufficient but not too much

preputial skin. During the procedure meticulous hemostasis is maintained (e.g., bipolar diathermy). Several methods of skin closure are described including the use of rapidly absorbable sutures to avoid suture tracts, subcuticular sutures, as well as the use of tissue glues.

9.6 Contraindications to Circumcision

Patients with hypospadias, epispadias, or ambiguous genitalia should not undergo neonatal circumcision as their foreskin may be required for reconstructive purposes. Circumcision should also be avoided in those with chordee without hypospadias, hidden penis, webbed penis, micropenis, dorsal hood deformity, and megaprepuce. If an anomaly is detected after the dorsal slit is made it may be better to stop the procedure rather than proceeding with the circumcision, however if the anomaly is minor circumcision should proceed as the foreskin is very rarely needed if further reconstruction is necessary. One should also inquire about a family history of bleeding disorders prior to proceeding with circumcision to avoid excessive postoperative hemorrhage.

9.7 Complications of Circumcision

1. Bleeding: usually from the frenulum or from a vessel along the penile shaft which is generally controlled with compression.
2. Wound infection.
3. Penile adhesions.
4. Meatal stenosis: most common late complication.
5. Too much penile skin removed: if this occurs, apply triple antibiotic ointment to the open wound. Usually most of the skin will grow back bridging the defect and rarely is immediate skin grafting necessary.
6. Too much penile skin left: revision circumcision is often requested by parents for an incomplete circumcision.
7. Urethral injury.
8. Partial removal of glans: has been reported with the Mogen clamp and should be immediately sutured back in place.

9. Urethrocutaneous fistula: low incidence, thought to be secondary to poorly placed suture at frenulum in an attempt to control bleeding.
10. Penile necrosis: rare, can result from thermal injury secondary to cautery.

9.8 Conclusion

Circumcision is the most common operation performed worldwide. In a few boys circumcision is clinically necessary but in the vast majority of boys circumcision is performed for cultural or religious reasons. However circumcision should not be performed without the families understanding that there is a significant complication rate and reoperation rate.

References

1. Elder JS. Abnormalities of the genitalia in boys and their surgical management. In: Wein AJ, Kavoussi LR, et al., eds. *Campbell's Urology*. 9th ed. Philadelphia: W.B. Saunders Company; 2006:3747-3749.
2. Hutton K. The prepuce. In: Thomas D, pohlman G, et al., eds. *Essentials of Paediatric Urology*. London: Informa Healthcare. 2008. 232-245.
3. Schoen EJ et al. The highly protective effect of newborn circumcision against invasive penile cancer. *Pediatrics*. 2000;105:E36.
4. Shoemaker C. Circumcision: Risks and benefits. UpToDate; January 2009.
5. To T et al. Cohort study on circumcision of newborn boys and subsequent risk of urinary-tract infection. *Lancet*. 1998;352:1813-1816.
6. Tobian AA et al. Male circumcision for the prevention of HSV-2 and HPV infections and syphilis. *N Engl J Med*. 2009;360:1298-1309.
7. Warner TE et al. Male circumcision and risk of HIV infection among heterosexual African American men attending Baltimore sexually transmitted disease clinics. *J Infect Dis*. 2009;199:59-65.
8. Weismiller D. Procedures for neonatal circumcision. UpToDate; January 2009.
9. Wiswell TE, Enzenauer RW, Holton ME, et al. Declining frequency of circumcision: implications for changes in the absolute incidence and male to female sex ratio of urinary tract infections in early infancy. *Pediatrics*. 1987;79:338-342.
10. Wiswell TE, Geschke DW. Risks from circumcision during the first month of life compared with those for uncircumcised boys. *Pediatrics*. 1989;83:1011-1015.

Chapter 10
Disorders of the Female External Genitalia

Richard S. Hurwitz

Key Points

- Labial adhesions usually resolve spontaneously. Extensive adhesions may be misdiagnosed as vaginal agenesis or as a disorder of sex development. When treatment is required, betamethasone cream 0.05% works faster and with fewer recurrences and side effects than estrogen creams.
- Interlabial masses have a characteristic appearance.
  - Paraurethral cyst – whitish or yellowish mass that displaces the urethral meatus
  - Imperforate hymen/hydrocolpos – bulging vaginal mass covered by thin grayish membrane. Abdominal mass may be present
  - Prolapsed ectopic ureterocele – smooth round mass bulging through the urethral meatus, usually with congested purplish mucosa
  - Urethral prolapse – swollen donut-shaped lesion with congested red to purple mucosa
  - Urethral polyp – single pedunculated structure from urethral meatus
  - Vaginal rhabdomyosarcoma – fleshy polypoid mass with grape-like clusters

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_10,

› Pre-pubertal girls with persistent vaginal discharge and all young girls with vaginal bleeding should be referred immediately for evaluation under anesthesia with cystoscopy and vaginoscopy.
  – Persistent vaginal discharge: Rule out vaginal foreign bodies and sexual abuse
  – Vaginal bleeding: Rule out vaginal malignancy, foreign bodies, benign papillomas, and sexual abuse

10.1 Introduction

In this chapter on the disorders of the female external genitalia, identification and treatment of labial adhesions and recognition of the classic interlabial masses is emphasized. The significance of vaginal discharge and vaginal bleeding in the prepubertal female is discussed.

10.2 Labial Adhesions

Labial adhesions occur when the inner edges of the labia minora, labia majora, or both in continuity somehow become excoriated causing opposing raw surfaces to adhere in the midline. The epithelial breakdown may be due to irritated inner labial skin from chronic wetness, ammoniacal inflammation, or recurrent vaginitis.

Labial adhesions may be asymptomatic and discovered during a diaper change or a routine physical examination (Figs. 10.1a and b). Patients may also present with symptoms of frequency, UTI, vaginitis, or post void dribbling due to trapping of urine.

Closure of the introitus by labial adhesions may be alarming when first recognized. The degree of introital closure is variable, but in some cases can be nearly complete with only a tiny subclitoral patency remaining. This appearance can

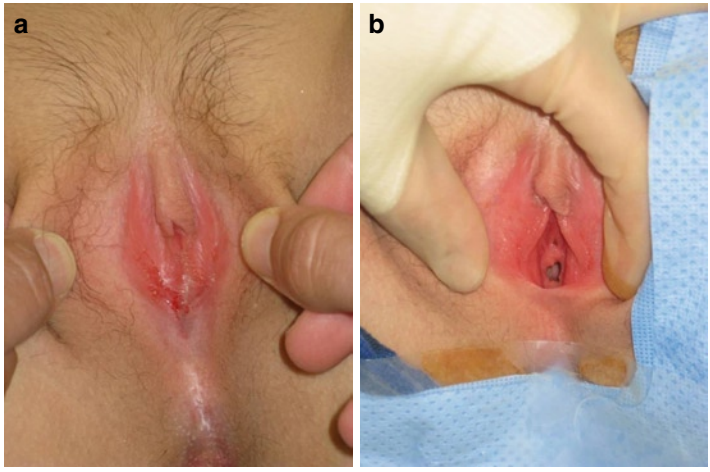


FIGURE 10.1. (**a, b**) Labial adhesions: Presumed urogenital sinus with posterior labial fusion in a patient with non-classical CAH. This unsuspected extensive adhesion was opened by the pressure of the cystoscope during endoscopic evaluation.

lead to misdiagnoses such as vagina atresia, vaginal agenesis or urogenital sinus anomaly raising the possibility of CAH or other forms of DSD. There is no associated clitoral enlargement. Sometimes a fine, vertical grayish membrane can be seen representing the thin midline fusion of the labia.

Labial adhesions tend to resolve spontaneously over time. Treatment is indicated for symptomatic and/or more extensive adhesions. Treatment options include topical estrogen cream, betamethasone cream, manual separation, or surgery. In a recent report of 151 patients with labial adhesions, 0.05% betamethasone cream resulted in separating the adhesions quicker (average 1.3 months) than did topical estrogen (premarin) therapy (average 2.2 months). Rates of recurrence were lower for patients treated with betamethasone. Side effect from estrogen treatment included breast budding and vaginal bleeding while side effects from betamethasone were limited to local irritation.[1] A success rate of 68% has been reported.[2] As more experience with betamethasone has accumulated, it seems to be favored for primary therapy. Surgical

lysis is occasionally required in cases resistant to medical therapy.

10.3 Interlabial Masses

An interlabial mass is occasionally encountered when inspecting the female genitalia. Some of these may cause discharge and/or bleeding. Differential diagnosis includes paraurethral cysts, imperforate hymen with hydrocolpos, prolapsed ectopic ureterocele, urethral prolapse, urethral polyp, and vaginal malignancy. Understanding the characteristic appearance and typical anatomic relationship to the urethral meatus and vaginal opening of each of these interlabial masses will greatly help in making the correct diagnosis.

10.4 Paraurethral (Skene's Duct) Cyst

Paraurethral cysts or Skene's duct cysts typically present as an asymptomatic interlabial mass in newborns. The typical appearance is that of a whitish or yellowish mass that displaces the urethral meatus to an eccentric position. The normal vaginal opening should still be visible. The cyst may cause deviation of the urinary stream or deform the anterior vaginal wall. It is thought that these cysts form because of an obstruction of the paraurethral gland ducts (Fig. 10.2).

Paraurethral cysts in newborns usually resolve by spontaneously rupturing during the first few weeks of life. Incision or needle aspiration may be needed to resolve persistent cysts.

10.5 Imperforate Hymen with Hydrocolpos

Hydrocolpos due to an imperforate hymen presents as a bulging vaginal mass covered by a thin pearly grey (hymeneal) membrane. The urethral meatus should be seen in its normal position just above the mass (Fig. 10.3).

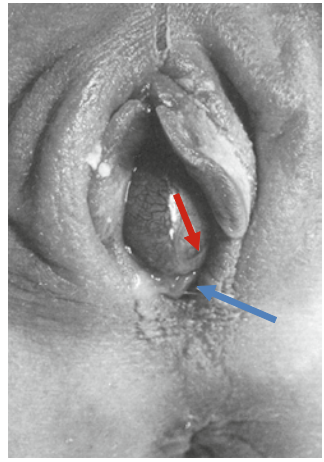


FIGURE 10.2. Paraurethral (Skene's duct) cyst: Right sided cyst is pushing and flattening urethral meatus to left (*red arrow*). Vaginal opening visible (*blue arrow*). (courtesy of Stephens 1983).

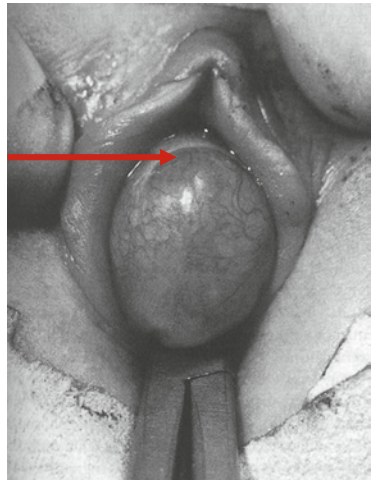


FIGURE 10.3. Imperforate hymen with hydrocolpos in double vagina: Urethral meatus splayed above bulging mass (*red arrow*). Hemostat in second patent hemi-vagina. (courtesy of Cartwright et al. 2002).

The imperforate hymen traps the naturally occurring mucous secretions created by maternal estrogen stimulation during in utero development. In some cases, the build-up of mucous secretions can result in massive vaginal dilation creating a palpable abdominal mass. Compression of the bladder and ureters may lead to difficulty voiding and hydronephrosis. The uterus

may also become distended (hydrometrocolpos), and urinary retention, constipation, and edema or cyanosis of the lower extremities have been known to occur in extreme cases.[3]

Simple incision of the imperforate hymen drains the distended vagina and relieves the compressive effects on adjacent organs. Renal and bladder ultrasound should be performed to evaluate the urinary tract status after vaginal drainage has been accomplished.

10.6 Prolapsed Ectopic Ureterocele

A prolapsed ureterocele presents as a smooth, round interlabial mass. If the prolapse is recent, the mucosa may still be pink. Spontaneous reduction of the prolapse may occur in the early pre-congestion stage. The mass will be large and purplish if the prolapse is seen after significant congestion and strangulation have occurred. Since the ureterocele has prolapsed through the urethra, a distinct urethral meatus may be difficult to visualize. The normal vaginal opening may be obscured, but should be detectable posteriorly (Fig. 10.4a).

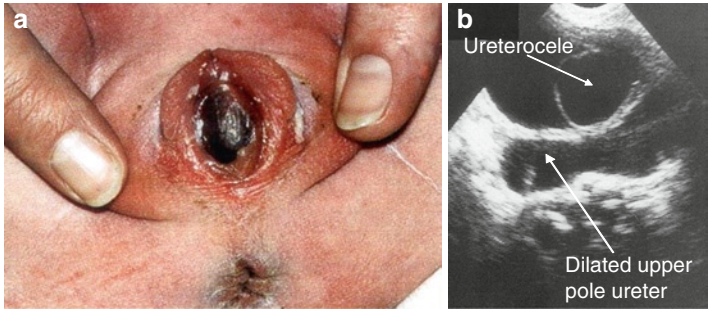


Figure 10.4. (**a**) Prolapsed strangulated ectopic ureterocele. (**b**) Ultrasound showing ectopic ureterocele in bladder and dilated upper pole ureter. (courtesy of Thomas et al. 2002).

Ectopic ureteroceles are associated with and cause obstruction of the upper pole system of a duplex kidney.

The diagnosis of an ectopic ureterocele can be confirmed by a renal and bladder ultrasound that will show a dilated upper pole collecting system and an intravesical cystic lesion in the area of the bladder neck (Fig. 10.4b). Urgent urological consultation is indicated when a prolapsed ureterocele is identified.

10.7 Urethral Prolapse

Urethral prolapse has a very characteristic donut-shaped appearance with a congested, red to dark purple discoloration of the mucosa. The urethral meatus is in the middle of the swollen, prolapsed mucosa. The mass created by the prolapsed tissue can be quite large and occupy most of the introitus in younger girls, obscuring the normally positioned vaginal opening (Fig. 10.5).

The typical presentation is with spotting of blood in the underwear. Sometimes this is accompanied by dysuria, perineal discomfort, and rarely urinary retention. The combination of the dark, swollen appearance and bleeding has sometimes led to the erroneous diagnosis of sexual abuse.

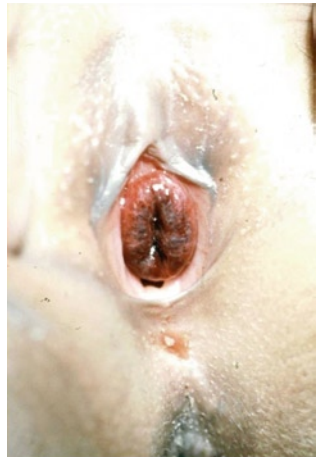


FIGURE 10.5. Urethral prolapse: Note the congested donut appearance around the centrally located urethral meatus.

Urethral prolapsed is more common in black girls 4–5 years of age and is often associated with coughing or constipation.

Treatment is usually conservative with sitz baths, elimination of constipation, and a short course of topical estrogen cream. Rarely, surgical excision is required.

10.8 Urethral Polyp

A urethral polyp is a smooth, mucosa – covered mass that protrudes from the urethral meatus. They are usually single pedunculated structures, either wide based or on a thin stalk averaging 1–3 cm in length. Although rare, large fleshy urethral polyps up to 6 cm in size have been reported in newborns.[4] The vaginal opening should be normal (Fig. 10.6).

Urethral polyps usually arise from the posterior wall of the urethra. They are not site-specific and can arise from the proximal, mid, or distal urethra. They commonly present with "vaginal" bleeding and some have associated symptoms of vulvitis, frequency, dysuria, or UTI.[5]

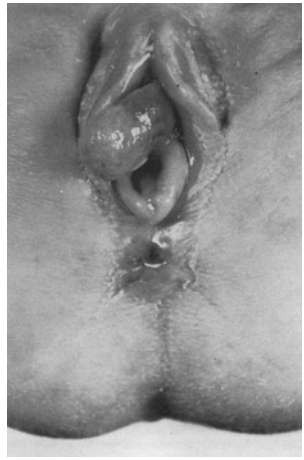


Figure 10.6. Urethral polyp. (courtesy of Stephens 1983).

Urethral polyps are benign lesions. Most are fibroepithelial polyps. Inverted papilloma and hamartomatous variations have been reported. The etiology thought to be related to prolapsing urothelium that undergoes an inflammatory response and then evolves into a polyp.[5]

Treatment is by simple excision and fulguration or suturing of the base.

10.9 Vaginal Rhabdomyosarcoma or Endodermal Sinus Tumor

Rhabdomyosacroma or sarcoma botryoides is the most common primary malignant tumor of the vagina in children and usually occurs in the first 5 years with a peak incidence before age 2. It may present as vaginal bleeding, passage of tissue fragments, or as an interlabial polypoid mass protruding from the vagina. The urethral meatus is usually normal. The polypoid masses typically appear as grape-like clusters. The rare endodermal sinus tumor, which carries a worse prognosis, may have a similar presentation (Figs. 10.7a–b).

Urgent urological referral is indicated. Biopsy will confirm the diagnosis. Treatment usually involves a combination of surgical excision, chemotherapy, and radiation therapy.

10.10 Vaginal Discharge and Vaginal Bleeding

Vaginal discharge and vaginal bleeding in prepubertal girls are indicators of a potentially serious social or medical problem. Vaginal discharge is most commonly due to a benign infectious process. It is rarely, if ever associated with malignancy. Development of new vaginal discharge in a child should suggest the possibility of sexual abuse. Initial evaluation should include an external genital exam and vaginal cultures that include analysis for chlamydia and gonorrhea. Persistent discharge after antibiotic treatment requires further investigation. In a referral population of 24 girls less than

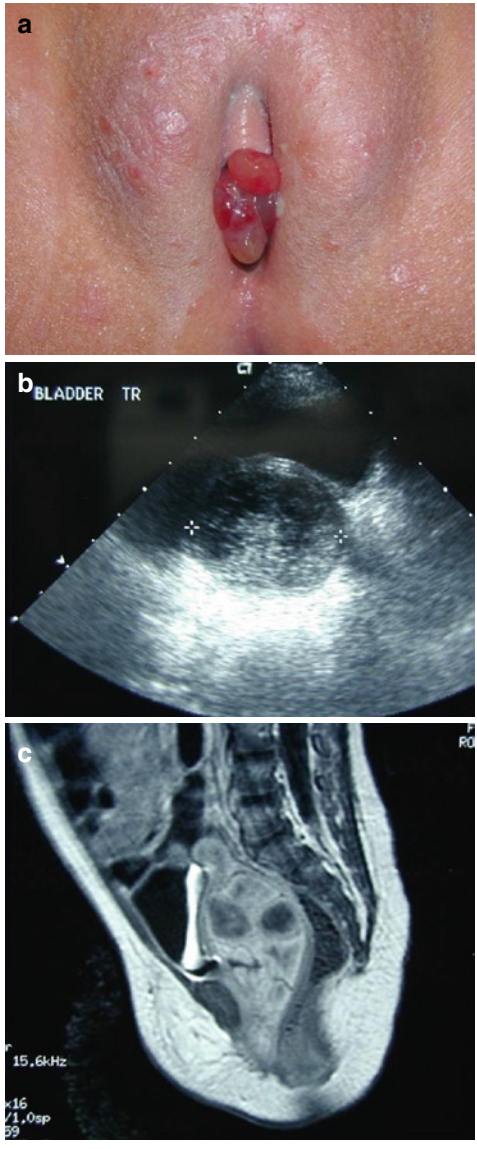


FIGURE 10.7. (**a**) Vaginal rhabdomyosarcoma: Fleshy grape-like clusters of tissue. (**b, c**): US and MRI of vaginal rhabdomyosarcoma.

6 years of age with vaginal discharge or bleeding, 11 who had persistent vaginal discharge after antibiotic treatment underwent evaluation under anesthesia, vaginoscopy, and cystoscopy. Five (45%) were found to have vaginal foreign bodies and two (18%) had evidence of sexual abuse.[6]

Vagina bleeding in young girls is relatively rare, but is often associated with severe underlying pathology. Vaginal bleeding can occur in up to 10% of normal female neonates due to maternal estrogen withdrawal, but vaginal bleeding after 10 days of age is not physiological and requires investigation. Differential diagnosis includes infection, vulvar lesions, trauma, foreign bodies, benign and malignant tumors, urethral prolapse, endogenous or exogenous hormonal exposure, and precocious puberty. In the above referral population of 24 girls, 13 were referred for vaginal bleeding and underwent evaluation under anesthesia with vaginoscopy, cystoscopy, and biopsy as needed. Six (45%) were found to have vaginal malignancies, two (15%) had foreign bodies, two (15%) had benign Müllerian papillomas, and two (15%) had evidence of sexual abuse.[6]

The usefulness of radiographic imaging in the evaluation of vaginal discharge or bleeding is not well-defined. Prepubertal girls with severe or persistent vaginal discharge and all young girls with vaginal bleeding should be referred immediately for pelvic evaluation under anesthesia, vaginoscopy, and cystoscopy.

References

1. Mayoglou L, Dulabon L, Martin-Alguacil N, et al. Success of treatment modalities for labial fusion: a retrospective evaluation of topical and surgical treatments. *J Pediatr Adolesc Gynecol*. 2009; 22(4):247-250.
2. Myers JB, Sorensen CM, Wisner BP, et al. Betamethasone cream for the treatment of pre-pubertal labial adhesions. *J Pediatr Adolesc Gynecol*. 2006;19(6):407-411.
3. Nussbaum AR, Lebowitz RL. Interlabial masses in little girls: Review and imaging recommendations. *AJR Am J Roentgenol*. 1983;141: 65-71.

4. Bertolotti A, Gonzalez SGT, Etcheverry R, et al. Giant congenital urethral polyp. *J Pediatr Surg*. 2008;43:1211-1212.
5. Ben-Meir D, Yin M, Chow CW, et al. Urethral polyps in prepubertal girls. *J Urol*. 2005;174:1443-1444.
6. Striegel AM, Myers MD, Sorensen PD, et al. Vaginal discharge and bleeding in girls younger than 6 years. *J Urol*. 2006;176(6 Pt 1): 2632-2635.

Chapter 11
Disorders of Elimination: Voiding Dysfunction

Tom P.V.M. de Jong and Marianne A.W. Vijverberg

Key Points

- Between 7–10% of school age children have lower urinary tract symptoms (LUTS).
- Common manifestations of LUTS are overactive bladder (OAB), underactive bladder (UAB) voiding postponement and dysfunctional voiding (DV).
- Evaluation of LUTS should include a full history and examination and assessment of micturition and defecation.
- Treatment is multifaceted , depends on the underlying cause and is tailored to individual children.

11.1 Introduction

Between 7% and 10% of school age children are seen by a specialist for recurrent urinary tract infections and/or urinary incontinence based on non-neurogenic lower urinary tract dysfunction.

According to the International Continence Society definition, any method that gives information on the function of the lower urinary tract may need to be used in dealing with

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_11,

these children, such as voiding history, physical examination, micturition and defecation diary, static and dynamic ultrasound of the lower urinary tract, uroflowmetry with ultrasonographic measurement of residual urine, and finally invasive studies such as voiding cystourethrography and urodynamic studies (UDS). The second report of the International Consultation on Incontinence also includes measurements of the function of the lower gastrointestinal tract in this definition.[1] A voiding and defecation history and voiding and defecation diary are, therefore, the most important sources of information on the function of the lower urinary and gastrointestinal tracts,[2–5] and can direct the course of subsequent therapy and investigations.

Any initial diagnostic investigations on children that wet during day-time or suffer recurrent urinary tract infection are done to discriminate between those children with functional voiding problems, those with neuropathic bladders and those with anatomic anomalies who may need surgery. Neuropathic conditions should be ruled out at the first clinical visit by physical examination and any suspect finding must lead to further investigations.[6,7] In boys with overactive bladder (OAB) and incontinence, urethral obstruction must be evaluated as the possible cause. Incontinence resulting from functional lower urinary tract symptoms (LUTS) is very common in girls and can be accompanied by urinary tract infection (UTI). In girls with dysfunctional voiding or underactive bladder (UAB), exclusion of other anomalies needs to be done because for many of these girls LUTS is a chronic condition that needs life-long attention to their voiding behavior.

11.2 Functional LUTS

Between 7% and 10% of children at school age have functional LUTS.[8,9] LUTS can manifest as urgency, frequency, incontinence, or recurrent UTI. Urgency typically comes with hold-up maneuvers during detrusor contractions. The

child uses all surrounding muscles to give extra urethral closure or mechanically compresses the urethra by hand or by squatting on a heel. A 3-day voiding diary and a 2-week defecation diary are recommended before the first visit to a pediatric urologist. A complete voiding and defecation history, a physical examination with special attention to lumbo-sacral neurological function, and at least two free uroflowmetries with ultrasonographic measurement of the post void residual volume should be done. Ultrasonography of the urinary tract is routinely advised[2,10–12]; in the upper tract, this technique can indicate double systems or can show dilatation or scarring. Bladder ultrasonography gives information on wall thickness; a thick-walled bladder is suspicious for anatomic or functional obstruction, while an open bladder neck in girls is commonly present in dysfunctional voiding.[10–12]

The transverse diameter of the rectum can be determined on bladder ultrasonography and a dimension of >3 cm in the absence of urge to defecate is a strong sign of constipation.[13,14] Advanced static and dynamic ultrasonography of the perineum can give additional information on the mobility of the bladder neck, the ability to contract the puborectalis muscle and sphincter at will, the guarding reflex (S3 neurological pathway) and the length of the urethra.[15–17] Hypermobility of the bladder neck can be seen in the 15% of children with generalized hyperlaxity of joints and might be associated with congenital stress incontinence. Inability to control the pelvic floor might indicate physical therapy before urotherapy. Absence of S3 reflexes might point to spinal dysraphism and rarely a congenital very short urethra of less than 15 mm can predict failure of conservative therapy.

The possible diagnoses in children with LUTS are OAB, dysfunctional voiding, UAB and incontinence with UTI caused by voiding postponement. An overview of the diagnostic steps in children with LUTS is provided in Fig. 11.1. Most of the children with LUTS need to be treated by urotherapy, when needed supported by pharmacologic therapy.

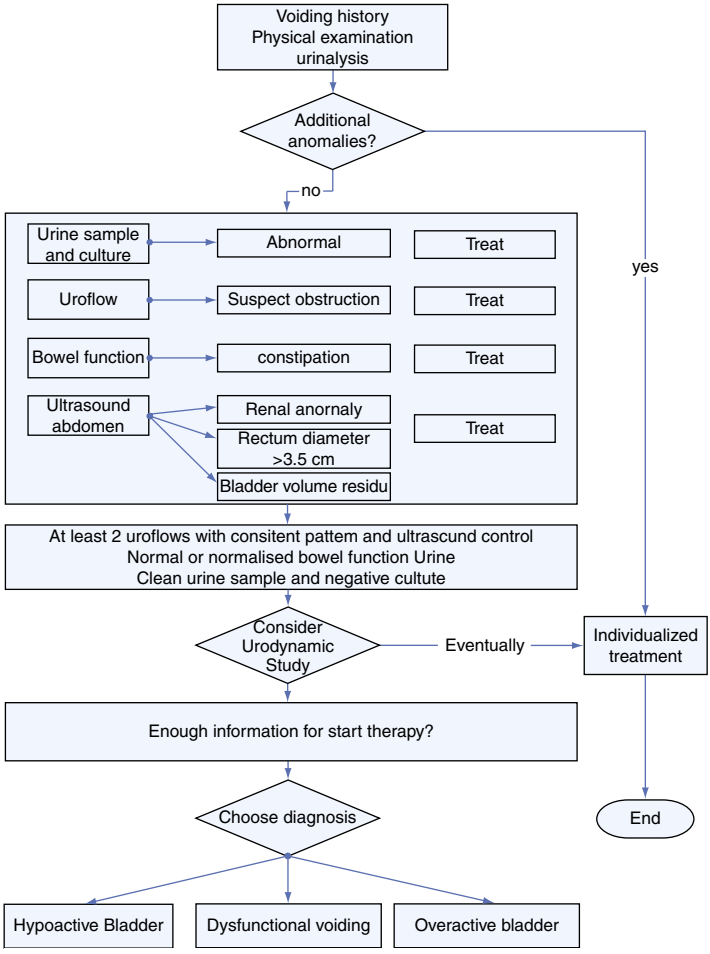


FIGURE 11.1. Diagnostics and diagnoses.

Urotherapy is defined by the ICCS as non-surgical, non-pharmacologic treatment for LUT malfunction, synonymous with the term LUT rehabilitation that is used for adults.[2,18]

11.2.1 Overactive Bladder

In school-aged boys with urge complaints and incontinence resulting from OAB, the major differentiation is between secondary overactivity caused by urethral obstruction and primary absence of central control of bladder behavior, which probably both present with a small voided volume compared to expected bladder capacity for age.[19] Obstruction in boys can occur anywhere from the bladder neck to the tip of the urethra, and will often be undetected by voiding cystourethrography.[19–21] A positive reaction – in terms of relief of symptoms – to antimuscarinic therapy, even when it only lasts a few weeks, is a strong predictor of urethral obstruction being the primary cause of the OAB in boys.[22] Based on this study we routinely recommend a 3-month period of antimuscarinic pharmacotherapy to point to obstruction before deciding whether a boy needs a UDS – and potentially endoscopic surgery – or needs alternative treatments for bladder overactivity.

Girls with LUTS predominantly need pharmacologic and behavioral therapy. Pharmacologic therapy can be chemoprophylaxis for recurrent UTI's, laxatives for the, often present, constipation and antimuscarinics in case of detrusor overactivity. In a third line referral setting, approximately one-third of girls with LUTS appears to have a urethral meatus anomaly that prohibits relaxed voiding in the ideal toileting position. They wet the toilet rim and buttocks because of an anteriorly deflected urinary stream and thus may need to have a meatus correction before urotherapy.[23,24] Sometimes, UDS are needed to identify meatal anomalies and a strongly elevated maximal detrusor pressure during voiding can typically be found in these cases.

11.2.2 Dysfunctional Voiding

Dysfunctional voiding occurs often in girls and rarely in boys. Inappropriate relaxation of the pelvic floor during voiding results in staccato and interrupted stream on uroflowmetry

with residual urine seen on subsequent ultrasonography. Loss of feeling of the filling state of both the bladder and the rectum characterizes dysfunctional voiding and incontinence caused by voiding postponement is frequently observed.[2] A urethral meatus anomaly, as described earlier, can be present; approximately half of girls with an anteriorly deflected urinary stream will be free of complaints after treatment of the meatus deformity while the other half will require cognitive and biofeedback training.

11.2.3 Underactive Bladder

UAB presents as a decompensated form of dysfunctional voiding that occurs predominantly in girls, but sometimes in otherwise healthy boys.[25] In boys with a history of severe urethral obstruction, loss of feeling of the filling state of the bladder and loss of sensation of the full bladder combined with low bladder compliance can be dangerous for the upper tracts, especially when night-time polyuria is present. The same problem can be present in girls after severe obstruction by a large ureterocele.[26–30]

11.2.4 Uroflowmetry

In general, voided volumes of less than 100 mL, or less than 50% of the expected functional volume for age, cannot be interpreted reliably. In all cases, it is important to note the voided volume, the recorded urinary flow and the flow-time. Urinary flow can be described in terms of rate and pattern and can be continuous, interrupted (fractionated), or staccato (fluctuating with peaks and troughs). The calculation of average flow rate (voided volume divided by flow time) is only meaningful if flow is continuous. The parameters used to characterize continuous flow can be applicable, if care is exercised, in children with fractionated or staccato flow patterns. Patterns and rates should be consistent over repeated studies

to allow evaluation, and therefore, several recordings are needed to obtain consistency.

Low rate continuous flow with extended flow-time can point to anatomical infravesical obstruction.[31] Staccato and interrupted flow is seen in patients with dysfunctional voiding and underactive bladder.[2] Normal continuous flow rate within expected time does not exclude urethral obstruction because no information is present on the pressure that the detrusor generates to produce this flow. Specific literature on this subject, other than expert opinion, is sparse.[3, 32–34]

11.2.5 Treatment

Medical treatment can be tried for an episode of 3 months by the specialist. Any treatment should start with explanation of bladder and sphincter function combined with explanation of the condition that is causing the LUTS.

In case of urge complaints and OAB, in the absence of constipation, antimuscarinics can be given. At this moment, oxybutynin is the only anticholinergic that is accepted for pediatric use although many also prescribe detrusitol. Oxybutynin has a fast effect, within 30 min, that lasts for 6–8 h. This gives the opportunity to tailor medication to the urge complaints. Many children suffer predominantly from urge complaints in the late afternoon and early evening. In those cases, 1 dosage of oxybutynin around 3 pm can be sufficient to give relief of complaints. Thus, also the most important complication of oxybutynin treatment, constipation, can be prevented as much as possible. For daytime only complaints, dosage of oxybutynin is around 8 am and around 3 pm. A dosage in the morning and the evening, in most patients, gives no relief of complaints in the afternoon and early evening.

Oxybutynin causes no problems in the vast majority of patients but parents must be warned that, sometimes, central side effects like loss of concentration or psychological changes can occur that force to stop medication. Also, in high temperatures, temperature control can derange, even causing a heath stroke.

This is caused by a less effective transpiration due to the anticholinergic effect. We advise the parents, when possible, to interrupt medication with outside temperatures exceeding 30°C.

Much discussion exists around the use of chemoprophylaxis in patients with dilated upper urinary tracts and primary vesico-ureteric reflux. In our opinion, girls with recurrent UTI's based on DV or UAB can really be depending on prophylaxis although good scientific evidence is not available. Preferably, we prescribe trimethoprim or nitrofurantoin MC 2 mg/kg/day in one gift. In girls with recurrent break-through infections we commonly use trimethoprim and nitrofurantoin alternating day by day. Girls with a low number of UTI's can use self test strips at home (leucocytes and nitrite) and do self medication with nitrofurantoin MC 6 mg/kg/day in 3 or 4 gifts for 3 days. Nitrofurantoin MC should be prescribed in capsules or powder. Nitrofurantoin solution is not suitable for prophylaxis because all children will develop stomach complaints after several weeks or months.

For laxative therapy nowadays polyethylene glycols (PEGS) are preferred. Since constipation in children with LUTS does not always follow strictly the Rome III criteria for constipation but suffer mostly from a dilated rectum that they cannot feel, oral laxative therapy is not always successful. In those cases we use rectal water enemas comparable to those used in spina bifida patients.

After the 3 months run-in period, in boys with OAB and in girls with meatal anomalies, first step must be to think about the need to correct an urethral obstruction or do a meatus correction to get a normal urinary stream. Once this has been done the next step will be out-patient urotherapy.[24,35]

11.2.5.1 Standard Outpatient Urotherapy

Instructions are given about the bladder and sphincter function, proper voiding pattern, and the use of voiding charts and proper toileting position. Prophylactic antibiotics are continued and instructions are given for a regular defecation pattern with or without laxative therapy the child needs to sit on the toilet

with a book for 5 min after breakfast and after dinner. This needs to be controlled with a calendar for at least 6–8 months. Weekly follow-up by the urotherapist is done by telephone and visits to the outpatient clinic are planned at 4 and 8 weeks after the first visit. At these visits, uroflowmetry with ultrasound residual urine assessment is done. Training results are evaluated and instructions are repeated. In children with OAB, special attention is given to central control of bladder overactivity. The training has been divided into two steps. The instruction mainly consists of the competition to stay dry with the help of maximum mental concentration. At this stage, it is not important how many times the child will void: it can go as often as necessary. If the child has been able to stay dry, than the next step is started. By using a balloon and drawings, the children are taught to experience the difference between holding up using the 'emergency brake' (contraction of the pelvic floor muscles), or holding up using the "normal brake" (contraction of the urinary sphincter). Originally the brain directs the urinary sphincter in a subconscious fashion. These children have to learn this skill now in a conscious way by cognitive techniques.

Once a day they have to try and hold up their urine as long as possible. Once a week they measure the amount of urine and put the results in a graph. They have to achieve a rising line. (Break a record) Besides they have a voiding list. With a fluid intake of 1½ L, they have to achieve a final goal of 7 micturitions per day.

In patients with DV, voiding postponement and UAB the emphasis is on timed voiding and relaxation of the pelvic floor musculature.

Many alternatives exist for biofeedback training. Biofeedback can be given with Uroflowmetry, wetting alarm, pelvic floor EMG etc.[36–41]

11.2.5.2 The Failed Training

Failure in training, especially recurrent UTI's in girls with DV or underactive bladder, often is caused by failure of treatment of constipation. When a wide rectum persists after

adequate laxative and toileting therapy we have adopted rectal wash-out enema therapy for this group. The child is taught to take rectal enema with 20 mL tap-water/kg. First to weeks, daily enemas are taken, followed by 3 months of once every 2 days. When the child passes stools on the day that no enema is given, it can start doing it once every 3 days. Most children can stop the therapy after 12 months; some children remain depending on the enema therapy but, in general, keep performing it once a week or even less. Advantage of the enema therapy is that most children regain the feeling to discriminate between a distended or empty rectum.[13,14,42]

For therapy resistant patients some centers offer an intensive, hospitalized training program for 2 weeks that consists of daily training by biofeedback, wetting alarm, psychological support etc.[43]

When all these therapies fail, one should be aware of the possibility of structural anomalies of the bladder neck and urethra that need surgery to get cured.[44]

11.2.6 Giggle Incontinence, Incontinentia Risoria

It is well-known that directors of theatres dislike comic spectacles because of the fact that many seats are wet after such an event. Giggle incontinence can be a socially very debilitating condition for children. No good therapy exists. One can look specifically to the normal functioning of the pelvic floor and offer pelvic floor physical therapy to the children that appear to have insufficient strength. We are offering a training program that aims at awareness by a self-administered shock during moments of laughter.[45,46] Success is approximately 60%. Ephedrine-like substances with an α-mimetic effect can sometimes reduce wet incidents. Prescription of 10 mg ephedrine will reduce accidents for 4 h. Methylphenidate, used in the treatment of ADHD, also acts on the bladder neck and can be tried.[47] Even botulinum toxin injection into the detrusor muscle has been tried.[48]

References

1. Homma Y, Batista J, Bauer S, Griffiths D, Hilton P, Kramer G, Lose G, Rosier P. In: Abrams P, Cardozo L, Khoury S, Wein A, eds. *Urodynamics. Incontinence: 2nd International Consultation on Incontinence*. 2002. Plymouth UK.
2. Neveus T, Von Gontard A, Hoebeke P, et al. The standardization of terminology of lower urinary tract function in children and adolescents: report from the Standardisation Committee of the International Children's Continence Society. *J Urol*. 2006;176(314):314.
3. Tekgul S, Nijman RJ, Hoebeke P, Canning D, Bower W, von Gontard A. Diagnosis and management of urinary incontinence in childhood. In: Abrams P, Cardozo L, Khoury S, Wein A, eds. in Incontinence, Health Publications Plymouth, 2009.
4. Rasquin A, Di LC, Forbes D, et al. Childhood functional gastrointestinal disorders: Child/adolescent. *Gastroenterology*. 2006;130:1527.
5. Hyman PE, Milla PJ, Benninga MA, Davidson GP, Fleisher DF, Taminiau J. Childhood functional gastrointestinal disorders: neonate/toddler. *Gastroenterology*. 2006;130:1519.
6. Mosiello G, Capitanucci ML, Gatti C, et al. How to investigate neurovesical dysfunction in children with anorectal malformations. *J Urol*. 2003;170:1610.
7. De Gennaro M, Rivosecchi M, Lucchetti MC, Silveri M, Fariello G, Schingo P. The incidence of occult spinal dysraphism and the onset of neurovesical dysfunction in children with anorectal anomalies. *Eur J Pediatr Surg*. 1994;4(Suppl 1):12.
8. Heron J, Joinson C, Croudace T, von Gontard A. Trajectories of daytime wetting and soiling in a United kingdom 4 to 9-year-old population birth cohort study. *J Urol*. 2008;179:1970.
9. Soderstrom U, Hoelcke M, Alenius L, Soderling AC, Hjern A. Urinary and faecal incontinence: A population-based study. *Acta Paediatr*. 2004;93:386.
10. Dogan HS, Akpinar B, Gurocak S, Akata D, Bakkaloglu M, Tekgul S. Non-invasive evaluation of voiding function in asymptomatic primary school children. *Pediatr Nephrol*. 2008;23:1115.
11. Yeung CK, Sreedhar B, Leung YF, Sit KY. Correlation between ultrasonographic bladder measurements and urodynamic findings in children with recurrent urinary tract infection. *BJU Int*. 2007;99:651.
12. Yeung CK, Sreedhar B, Leung VT, Metreweli C. Ultrasound bladder measurements in patients with primary nocturnal enuresis: a urodynamic and treatment outcome correlation. *J Urol*. 2004;171:2589.
13. Klijn AJ, Asselman M, Vijverberg MA, Dik P, de Jong TP. The diameter of the rectum on ultrasonography as a diagnostic tool for constipation in children with dysfunctional voiding. *J Urol*. 2004;172:1986.

14. Joensson IM, Siggaard C, Rittig S, Hagstroem S, Djurhuus JC. Transabdominal ultrasound of rectum as a diagnostic tool in childhood constipation. *J Urol*. 2008;179:1997.
15. de Jong TP, Klijn AJ, Vijverberg MA, de Kort LM, van Empelen R, Schoenmakers MA. Effect of biofeedback training on paradoxical pelvic floor movement in children with dysfunctional voiding. *Urology*. 2007;70:790.
16. de Jong TP, Klijn AJ, Vijverberg MA, de Kort LM. Ultrasound imaging of sacral reflexes. *Urology*. 2006;68:652.
17. de Kort LM, Verhulst JA, Engelbert RH, Uiterwaal CS, de Jong TP. Lower urinary tract dysfunction in children with generalized hypermobility of joints. *J Urol*. 2003;170:1971.
18. Abrams P, Cardozo L, Fall M, et al. The standardisation of terminology of lower urinary tract function: report from the Standardisation Sub-committee of the International Continence Society 5. *Neurourol Urodyn*. 2002;21:167.
19. Schober JM, Dulabon LM, Woodhouse CR. Outcome of valve ablation in late-presenting posterior urethral valves. *BJU Int*. 2004;94:616.
20. de Jong TP, Radmayr C, Dik P, Chrzan R, Klijn AJ, de Kort L. Posterior urethral valves: search for a diagnostic reference standard. *Urology*. 2008;72:1022.
21. de Kort LM, Uiterwaal CS, Beek EJ, Jan Nievelstein RA, Klijn AJ, de Jong TP. Reliability of voiding cystourethrography to detect urethral obstruction in boys. *Urology*. 2004;63:967.
22. de Kort LM, Klijn AJ, Dik P, Uiterwaal CS, de Jong TP. Oxybutynin for diagnosis of infravesical obstruction in boys with urinary incontinence. *Urology*. 2003;62:127.
23. Hoebeke P, Van LE, Raes A, van Gool JD, Vande WJ. Anomalies of the external urethral meatus in girls with non-neurogenic bladder sphincter dysfunction. *BJU Int*. 1999;83:294.
24. Klijn AJ, Uiterwaal CS, Vijverberg MA, Winkler PL, Dik P, de Jong TP. Home uroflowmetry biofeedback in behavioral training for dysfunctional voiding in school-age children: A randomized controlled study. *J Urol*. 2006;175:2263.
25. Hinman F, Baumann FW. Vesical and ureteral damage from voiding dysfunction in boys without neurologic or obstructive disease. *J Urol*. 1973;109:727.
26. De Gennaro M, Capitanucci ML, Mosiello G, Caione P, Silveri M. The changing urodynamic pattern from infancy to adolescence in boys with posterior urethral valves. *BJU Int*. 2000;85:1104.
27. De Gennaro M, Capitanucci ML, Capozza N, Caione P, Mosiello G, Silveri M. Detrusor hypocontractility in children with posterior urethral valves arises before puberty. *Br J Urol*. 1998;81(Suppl 3):81.
28. Sherman ND, Stock JA, Hanna MK. Bladder dysfunction after bilateral ectopic ureterocele repair. *J Urol*. 2003;170:1975.

29. Abrahamsson K, Hansson E, Sillen U, Hermansson G, Hjalmas K. Bladder dysfunction: an integral part of the ectopic ureterocele complex. *J Urol*. 1998;160:1468.
30. Beganovic A, Klijn AJ, Dik P, de Jong TP. Ectopic ureterocele: long-term results of open surgical therapy in 54 patients. *J Urol*. 2007;178:251.
31. Vanderbrink BA, Gitlin J, Palmer LS. Uroflowmetry parameters before and after meatoplasty for primary symptomatic meatal stenosis in children. *J Urol*. 2008;179:2403.
32. Farhane S, Saidi R, Fredj N, Touffahi M, Lefi M, Saad H. Uroflowmetry in children: Prospective study of normal parameters. *Prog Urol*. 2006;16:598.
33. Szabo L, Fegyverneki S. Maximum and average urine flow rates in normal children – the Miskolc nomograms5. *Br J Urol*. 1995; 76:16.
34. Wese FX, Gaum LD, Liu TP, Wong AK, Hardy BE, Churchill BM. Body surface related flow rate nomograms in a normal pediatric population 1. *Acta Urol Belg*. 1989;57:467.
35. Shei Dei YS, Wang CC. Outpatient biofeedback relaxation of the pelvic floor in treating pediatric dysfunctional voiding: a short-course program is effective Home uroflowmetry biofeedback in behavioral training for dysfunctional voiding in school-age children: a randomized controlled study. *Urol Int*. 2005;74:118.
36. Van Laecke E, Wille S, Vande WJ, et al. The daytime alarm: a useful device for the treatment of children with daytime incontinence. *J Urol*. 2006;176:325.
37. Hoebeke P. Twenty years of urotherapy in children: what have we learned? *Eur Urol*. 2006;49:426.
38. Hoebeke P, Vande WJ, Theunis M, De PH, Oosterlinck W, Renson C. Outpatient pelvic-floor therapy in girls with daytime incontinence and dysfunctional voiding. *Urology*. 1996;48:923.
39. de Jong TP, Klijn AJ, Vijverberg MA, de Kort LM, van ER, Schoenmakers MA. Effect of biofeedback training on paradoxical pelvic floor movement in children with dysfunctional voiding. *Urology*. 2007;70:790.
40. van Gool JD, Vijverberg MA, Messer AP, Elzinga-Plomp A, de Jong TP. Functional daytime incontinence: non-pharmacological treatment 10. *Scand J Urol Nephrol Suppl*. 1992;141:93.
41. van Gool JD, Kuitjen RH, Donckerwolcke RA, Messer AP, Vijverberg M. Bladder-sphincter dysfunction, urinary infection and vesico-ureteral reflux with special reference to cognitive bladder training. *Contrib Nephrol*. 1984;39:190.
42. Chrzan R, Klijn AJ, Vijverberg MA, Sikkel F, de Jong TP. Colonic washout enemas for persistent constipation in children with recurrent urinary tract infections based on dysfunctional voiding. *Urology*. 2008;71:607.

43. Vijverberg MA, Elzinga-Plomp A, Messer AP, van Gool JD, de Jong TP. Bladder rehabilitation, the effect of a cognitive training programme on urge incontinence. *Eur Urol*. 1997;31:68.
44. de Kort LM, Vijverberg MA, de Jong TP. Colposuspension in girls: clinical and urodynamic aspects. *J Pediatr Urol*. 2005;1:69.
45. Elzinga-Plomp A, Boemers TM, Messer AP, Vijverberg MA, de Jong TP, van Gool JD. Treatment of enuresis risoria in children by self-administered electric and imaginary shock. *Br J Urol*. 1995;76:775.
46. Richardson I, Palmer LS. Successful treatment for giggle incontinence with biofeedback. *J Urol*. 2009;182:2062.
47. Berry AK, Zderic S, Carr M. Methylphenidate for giggle incontinence. *J Urol*. 2009;182:2028.
48. Wefer B, Seif C, van der Horst C, Junemann KP, Braun PM. Botulinum toxin A injection for treatment-refractory giggle incontinence. *Urologe A*. 2007;46:773.

Chapter 12
Disorders of Elimination: Nocturnal Enuresis

Adriana Marie Behr

Key Points

- The primary goal in treating enuresis is to teach the child to self-awaken to the sensation of a full bladder and independently go to the bathroom.
- Enuresis alarms are associated with the highest cure rates and lowest relapse rates of any single treatment option.
- Enuresis alarms are simple for children to use and cost-effective for families.
- Combination therapy (enuresis alarms and pharmacologic management) may increase cure rate.
- Early management of enuresis is important for social development and self-esteem of the child.

12.1 Introduction

12.1.1 Definition

Enuresis or nocturnal enuresis is the involuntary, intermittent release of urine during sleep. There are two types of nocturnal enuresis. Primary enuresis occurs in children aged 5 years or older who are neurologically and developmentally normal, but have not yet achieved 6 months or more of nighttime dryness.

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_12,

Secondary enuresis is the relapse of nighttime wetting in children who have achieved at least 6 months of nighttime dryness. Enuresis is further classified based on the presence or absence of additional symptoms. Monosymptomatic or physiologic enuresis is nighttime wetting in children without any other lower urinary tract symptoms, except nocturia, and without a history of dysfunctional voiding. Children with enuresis and any other symptoms of the lower urinary tract or complicating organic factors are classified as having nonmonosymptomatic enuresis.[1]

12.1.2 Prevalence

Nocturnal enuresis is a common condition. Of school-aged children aged 5 to 16 years, 6–10% experience nocturnal enuresis.[2] Most primary enuresis is thought to be caused by a maturational delay. Therefore, the prevalence should decrease as children mature. Each year 15% of enuretic children cease wetting without intervention.[3] Boys tend to be affected more often than girls. About 60% of intermittently enuretic children and 90% of nightly bedwetters are male.[4]

12.1.3 Causes

The etiology of nocturnal enuresis is unclear and is likely multi-factorial, involving genetic, sleep, urological, neurological, and psychological components. As previously stated, it can be classified as monosymptomatic (physiologic) or nonmonosymptomatic (organic) in nature.

12.1.4 Monosymptomatic Enuresis

12.1.4.1 Genetics

For many families, bedwetting is a familiar disturbance. Research has demonstrated a higher incidence of bedwetting

in children whose parents were affected when compared to families with no parental history. Geneticists have identified possible markers for primary nocturnal enuresis on chromosomes 12, 13, and 22.[5] We know that enuresis is an autosomal dominant trait, so the likelihood of enuresis if both parents were affected is 77% and about 44% if one parent was affected. Children still have a 15% chance of acquiring enuresis even if neither parent was affected.[6]

12.1.4.2 Sleep

Children with nocturnal enuresis have been shown to have normal sleep patterns; however, parents of bedwetting children report much difficulty awaking the child from sleep, and when awakened, these children are often disoriented. Bedwetting is not caused by deep sleep however and is not considered a sleeping disorder. Nonetheless, it seems as though children with nocturnal enuresis have more difficulty arousing from sleep and are not able to wake up when their bladders are full. Instead, their muscles relax and the bladder empties while they sleep. For instance, a study performed by[7] Wolfish demonstrated that children with enuresis awoke to increasing tone intensity 8.5% of the time compared to the control group, who awoke about 40% of the time. For that reason, it is believed that enuretic children have a slow-to-mature sleep arousal mechanism within the central nervous system.

Nocturnal enuresis can occur in all sleep stages, but has shown to be more likely during non-rapid eye movement. Cystometric studies performed on children during sleep have shown that they have more difficulty arousing from sleep during non-rapid eye movement, which predominately occurs during the first two-thirds of the night. If spontaneous bladder contractions occur in that phase, children are less likely to wake and are therefore more likely to have an enuretic episode. Children spend the last one-third of sleep in rapid eye movement and have fewer enuretic episodes during that time.[7]

12.1.4.3 Sleep-Disordered Breathing

Obstructive sleep apnea has been studied for its association with nocturnal enuresis. It is well known that regular sleep disruptions due to upper airway obstruction in sleep apnea cause a number of daytime concerns, like sleepiness, decreased school performance, headache, and growth disturbance. Incongruously, these children are very difficult to wake from sleep. It is believed that the brain puts such a high value on sleep integrity that the arousal threshold is actually increased in patients with sleep apnea because of the constant stimuli from the airways. Therefore, these children sleep more heavily and do not wake easily to less obvious stimuli, like bladder fullness. Therefore, in addition to the higher incidence of polyuria in this population, decreased sleep arousal is another cause for the relationship between enuresis and sleep apnea.[8]

12.1.4.4 Small Functional Bladder Capacity

Though most children with nocturnal enuresis have normal bladder function and a normal structural bladder capacity, they may exhibit small *functional* bladder capacity that leads to nighttime wetting. Functional capacity is the point at which the brain receives the signal that the bladder needs to empty. This is in contrast to estimated bladder capacity (EBC) which is projected by the formula, [30 + (age in years × 30)] (in milliliters) and structural bladder capacity, which identifies the actual volume of the bladder.

The International Children's Continence Society (ICCS) has replaced the term functional capacity with voided volume to reflect bladder function versus bladder anatomy. To obtain functional capacity or voided volume, the child should urinate in a measuring container or toilet "hat" at home. Parents should collect about 10 measurements over the course of 3 days to get an average voided volume for the child. Evidence of small functional capacity is primary enuresis, wetting the bed every night, several wetting episodes per

night, and frequency of urination during the daytime. Reduced functional capacity compared to EBC, in addition to decreased sleep arousal, is a common finding in bedwetting children and can make treatment more complicated.

12.1.4.5 Psychological/Behavioral

Psychological stressors do not cause nocturnal enuresis but may play a role in secondary enuresis, or the relapse of physiologic enuresis. These children often display daytime symptoms as well, including daytime incontinence, urgency, encopresis, decreased school performance, and social withdrawal. Associations have been made between nocturnal enuresis and psychological or behavioral problems and have been attributed to low self-esteem of bedwetters and an overrepresentation of children suffering from ADHD in the enuretic group. Once these physiological and psychological causes have been addressed, the enuresis can be treated with the same methods as children with primary enuresis. Another possible association is that children suffer from psychosocial problems because of poor sleep quality secondary to detrusor overactivity at night, which is another reason to consider early and efficient therapy for bedwetters.

12.1.5 Nonmonosymptomatic (Organic) Enuresis

12.1.5.1 Urological Conditions: Urgency of Urination and Dysfunctional Elimination

The detrusor muscle contracts to signal the initial urge to urinate. Some children may have difficulty suppressing this urge by inhibiting the detrusor contraction until they can get to the bathroom. These children may show daytime urgency but maintain continence by using the bathroom more often. Children who have severe bladder contractions often have associated daytime (urge) incontinence and may have difficulty suppressing this urge at night while sleeping. These children usually have a normal bladder capacity, but a small

functional bladder capacity, and waking in time to void at night can be difficult.[4]

Bladder irritation can also lead to urgency and incontinence. One of the most common causes of bladder irritation is a urinary tract infection (UTI). Urinary tract infections can decrease bladder function and inflame the normal bladder tissue making it difficult to expand and contract normally. Antibiotics can heal infections, but the secondary enuresis may not automatically improve. Once the irritation and wetting starts, it seems to become a cycle. If children present with enuresis after a period of being dry, UTI should be ruled out or treated before the enuresis is addressed.

Children with nocturnal enuresis may be affected by elimination disorders that can cause irritation of the lower urinary tract. Elimination disorders include delayed voiding or incomplete bladder emptying and irregular or infrequent bowel movements. These children prolong urination during the daytime, void only partially, and do no empty their bladders before bedtime. These habits increase the possibility for nocturnal enuresis (Table 12.1).

Irregular and infrequent stooling can also affect the ability of the bladder to fill and empty properly. A full rectum can put pressure on the bladder, preventing it from functioning well. During bladder filling, expansion of the bladder may be

Table 12.1. Causative conditions for secondary enuresis.

Condition	Mechanism
Constipation	Reduced bladder capacity
Cystitis	Reduced bladder capacity
	Detrusor overactivity
Diabetes (mellitus/insipidus)	Nocturnal polyuria
Sleep-disordered breathing	Impaired arousal
Neurogenic bladder	Reduced bladder capacity
	Detrusor overactivity
Psychological stress	–

restricted by a full rectum. The limited expansion causes the bladder to contract even though there may be room for additional urine. Then, instead of contracting smoothly and uniformly, the bladder will spasm, causing wetting to occur. Additionally, when the rectum chronically intrudes upon the bladder, it can decrease the sensation of bladder fullness and lead to day and nighttime enuresis.[9]

12.1.5.2 Polyuria

Excessive urine production may cause the bladder to fill one or more times during the night, requiring the child to wake to urinate. If the child does not arouse from sleep with the sensation to void, he or she will likely wet the bed. Polyuria can arise from several disease states, including diabetes mellitus, diabetes insipidus, and isothenuria associated with sickle cell anemia. It can exist in the absence of disease in children who drink excessive amounts of fluid in the evening hours or get up to drink overnight. Habit polydipsia is the most common cause of polyuria. Children or parents should be asked to report the amount of fluids at dinnertime, before bedtime, and if fluids are consumed in the middle of the night. It is important to ask about the intake of caffeine and any medications in the evening, as these factors can cause transient polyuria.

12.1.5.3 ADH Secretion

Normally, antidiuretic hormone (ADH) production increases at night causing the kidneys to concentrate urine and the bladder to fill more slowly. Some children do not secrete adequate amounts of ADH and their kidneys continue to produce dilute urine in quantities consistent with daytime amounts, overfilling the bladder and causing nocturia or enuresis. These children may benefit from the addition of desmopressin (DDAVP), the synthetic version of this hormone. Children who have normal ADH levels can still respond to DDAVP administration, which further reduces their urine production and may allow them to be dry at night.

12.1.5.4 Food Sensitivity

Diet may also play a role in enuresis. Although scientific evidence showing a relationship between diet and nighttime wetting is lacking, anecdotal data demonstrate that avoiding certain foods and beverages can decrease wetting. For instance, dairy products like milk or ice cream contain tryptophan, the amino acid that produces serotonin, which slows down nerve traffic in the brain and may lead to decreased sleep arousal. Other foods may stimulate urine production, like carbonated and caffeinated beverages, citrus fruits and drinks, melons, licorice and Vitamin C. Bladder irritants, like spicy foods, can cause the bladder to contract more often, signaling the need to void. Therefore, the presence of these foods in a child's diet may cause more bladder contractions that can lead to nocturia or enuresis.

12.2 Investigations

12.2.1 History

A thorough history is essential in the evaluation of enuresis. Most school-aged children can give a good school-day history of voiding. For those families who are not reliably accurate, a more objective measure is the voiding diary. The voiding diary is a tool kept by the parents to assess the time and volume of voids and their association with certain events, such as incontinence episodes, urgency, bowel movements, and play or distraction activities. The history should also include screening for symptoms like cystitis and constipation, since both are associated with reduced bladder capacity. Parents are often unaware of their child's bowel habits once they reach school age and children should be directly interviewed about the amount and consistency of stool, the frequency of bowel movements, and any pain or soiling.

A history of soaking undergarments and sheets suggests nocturnal polyuria. A daily diet and fluid intake can be

helpful in this instance. Many children do not get adequate amounts of fluid during the school day and some purposefully dehydrate themselves to prevent daytime incontinence episodes. These children will compensate once they arrive home and through the evening, which leads to a higher nighttime production of urine.

12.2.2 Physical Examination

The physical examination should focus on the bladder and bowel and include assessment of the genital and neurological systems. The abdomen should be palpated for stool and bladder distention. The spine should be examined for any cutaneous defects, including sacral dimples off the midline or above the gluteal cleft, hair tufts or pigmentation. Genitals should be evaluated for abnormalities (ie., urethral position, meatal stenosis, adhesions, and signs of abuse or trauma). The urine stream should be observed if it sounds abnormal by history. A digital rectal exam could be performed if the history is consistent with encopresis or constipation, although this is very rarely needed, as the history is usually sufficient. The anus should be evaluated for sphincteric reflex. Assessment of strength, tone, reflexes and sensation should be performed for possibility of neurogenic bladder.

12.2.3 Laboratory Tests

A urinalysis should be conducted in every child with enuresis. This study will help to evaluate for glycosuria in clinical diabetes mellitus. Specific gravity should be assessed to rule out diabetes insipidus or low fluid intake. If symptoms of UTI are present or if the urine is malodorous, a urine culture should also be performed, especially if the urinalysis contains nitrites, leukocyte esterase, or the patient has a history of UTI.

12.2.4 Imaging Studies

Radiographic studies are not routine for children with nocturnal enuresis alone, but may be helpful in children with daytime symptoms or in specialty assessment. Ultrasound of the bladder is performed to evaluate bladder wall thickness and post-void residual volumes. Voiding cystourethrogram (VCUG) may be performed in children with recurrent UTIs, signs of urinary tract obstruction or neurogenic bladder.

12.2.5 Evaluation of Functional Capacity

Measurement of the bladder capacity is often underestimated, but can be very useful in the evaluation of enuresis. Ask the family to document about ten voided volumes and average this volume to obtain an approximation of the child's functional capacity. Compare this to the estimated bladder capacity to determine if the child has a physiologic cause for enuresis. The parents of these children are often very happy to learn of a simple explanation for the wetting.

12.3 Conventional Treatment

Just as there is no simple cause for bedwetting, there is no easy solution. Conventional approaches to nocturnal enuresis include behavioral modifications, alarm therapy, and pharmacologic therapy. These treatment interventions should be chosen based on the type and severity of symptoms, the benefits and disadvantages of the treatment, and ultimately, what the family's goal for treatment is. Nonetheless, one of the most important considerations is the child's self-esteem. Though enuresis can improve over time with maturational development, treatment should not be deferred based on that belief alone. These children deserve evaluation and

intervention by the age of seven and treatment should be guided by the degree of concern and motivation of the child.

12.3.1 Behavioral Therapy

The basis of behavioral therapy is the attainment of good bladder and bowel habits. Essentially, this means the child should be encouraged to void frequently enough to avoid urgency and incontinence and have a bowel movement daily without difficulty. This method requires consistency and effort, a supportive parent to encourage a motivated child, and patience. A behavioral approach will require at least 2–3 months of effort.[13]

Children with associated diurnal enuresis, urgency, or constipation will need to have these symptoms addressed before bedwetting therapy can begin. Timed voiding and proper toileting posture are important for regular and complete emptying of the bladder and preventing incontinence and urgency. Optimal posture for relaxing the pelvic floor muscles and complete emptying of the bladder and bowel includes sitting on the toilet with feet flat on the floor or stool, legs apart, and a slight forward lean. Children should be encouraged to have privacy, take their time in the bathroom, relax to void, and practice good perineal hygiene. Biofeedback therapy is another behavioral approach that has been shown to improve outcomes. It consists of pelvic floor muscle re-training for isolation and relaxation of the pelvic floor and sphincter. Irregular or infrequent bowel movements may require the addition of increased fluids and fiber intake or laxatives to soften or pass stool through the bowel more easily.

Once concurrent symptoms have resolved, nighttime behavioral management can begin. Parents should be informed of the causes of nocturnal enuresis and understand that bedwetting is not intentional. They should be advised of the importance of positive reinforcement and avoidance of punishment for wetting episodes. In addition, they should be

encouraged to discuss the wetting with siblings of the affected child, to prevent teasing and shameful feelings.

Establish with the child the goal of getting up at night to urinate in order to stay dry. Explain to the child that he is in charge of his dryness and that you and his parents are there to support him. Include the child in the preparation of the bed before bedtime and clean-up the next day, including waterproofing the bed, helping to wash soiled sheets, and showering the next morning. Encourage avoidance of fluids two hours before bedtime, especially those containing caffeine, carbonation, and dairy. The child should also void twice before bedtime. These behavioral activities will be more easily remembered and progress can be tracked with the use of voiding diaries or charts. These charts can be reviewed during regular follow up every 1–3 months (Table 12.2).

Table 12.2. Recommendations for behavioral therapy.[a]

Encourage voiding at least once every 2–3 h, to avoid urgency and incontinence
Boys and girls should void with pants to ankles. Girls should sit on toilet with feet flat on floor or stool, knees open, forward lean with elbows on knees
Facilitate easy access to bathroom at school by writing a note to the teachers or school RN
Adequate water intake during daytime hours (1 oz/kg/day), preferably morning and early afternoon
Minimize intake of fluids before bedtime unless evening sports
Avoid bladder irritants before bedtime
Encourage child to empty bowels at least once a day
Discuss high fiber foods and water intake to soften stool
Use teamwork for nighttime preparation of the bed
Encourage double voiding before bedtime
Use progress charts to track activities being used and program success

[a]These recommendations are based on clinical experience

12.3.2 Alarm Therapy

Alarm therapy is fundamentally classic conditioning. Treatment is designed to teach the enuretic child to awaken to the sensation of a full bladder. A device is used that provides a strong sensory signal immediately upon the occurrence of incontinence. If the child learns to wake as quickly as possible to the alarm, he will begin to recognize the feeling of bladder fullness and eventually awaken before the alarm goes off.

Recent designs are small, portable, and are worn on the clothing. They include a small box that attaches to the shoulder or waist and clip-like sensor worn on the underpants. These differ from the older bell-and-pad type alarms, which require the child to wet through his clothing and the sheet to reach the alarm. They have also been known to be activated by perspiration. Although these alarms are still available, they are no longer the best options for treatment. Most alarms emit an 80-db sound, many include a vibration feature, and some only use vibration to wake the child when the sensor gets wet. Vibration-only alarms, while seemingly attractive to parents, may make the learning process more difficult since the burden of responsibility falls solely on the sleeping child.

Enuresis alarms have the highest cure rate of any treatment option for bedwetting. In a recent Cochrane Review, when compared to no treatment, about 70% of children become dry with the use of alarm therapy and almost half who persisted with alarm use stayed dry after treatment was discontinued, compared to almost none of those who had no treatment at all. Unfortunately, there was not enough data to distinguish between types of alarms or how alarms compared to other behavioral treatment. When overlearning was added to therapy after initial success, relapse rates decreased.[10] Overlearning is the process of giving extra fluids before bedtime, after a child has successfully become dry using the alarm.

Parents should be reminded that a new behavior is progressively learned and the parent's response is very

important, especially during the first few weeks of use. It may be necessary for parents to wake the child, remind the child what the alarm is for, physically swing the child's feet to the side of the bed, and encourage them to employ the proper behavior – using the bathroom to empty the bladder. The alarm should not be turned off until the child is awake and standing on the floor. The child will gradually learn to awaken to the alarm without reminders or parental help. Finally, the child will use this new sensation to wake himself and use the bathroom without the use of the alarm. This process takes 2–3 months on average but may take up to 6 months of practice and patience.

Parents should also consider modifying sleeping arrangements to make the best of the learning process. If the child sleeps in a bunk or far away from the parent, promote moving the child to a position that allows for easier access to the bathroom or near enough for a parent to intervene if there is no response from the child. A baby monitor is another alternative. Make sure the route to the bathroom is clear of obstacles, including toys. A night light or flashlight may also be helpful for the child to find his way.

Alarms are simple to use and offer a permanent cure if used properly and for the length of time required. This therapy alone may not work for every enuretic child, but it will for the majority of children when used consistently and employed with additional treatments available. In addition, overlearning can be used to increase success with alarm therapy. This fluid challenge has been shown to reduce relapse rates among children using alarm therapy.[10]

12.3.3 Pharmacologic Therapy

Medications for the treatment of bedwetting include desmopressin, anticholinergics, and tricyclic agents. The most frequently prescribed medication for enuresis is desmopressin acetate or DDAVP. The synthetic form of the natural anti-diuretic hormone (ADH) vasopressin works by

concentrating urine, thereby decreasing the quantity of urine produced after dosing. For enuretic children, this allows them to sleep dry or wet less at night and can be very beneficial for social events, like sleepovers or overnight camps. A review by Moffatt[11] concluded that in 18 controlled studies on desmopressin, only 24.5% of children were completely dry on the medication and 94.3% relapsed after discontinuation of the drug.

Desmopressin is available in nasal spray and tablet form; however, the spray now has a black-box warning from the Food and Drug Administration (FDA) and is no longer recommended due to the reported episodes of water intoxication. The nasal spray has a longer half-life, but is also associated with inconsistent absorption rates because of the changes to the nasal mucosa. Though the tablet has a shorter duration of pharmacologic action, it still works within the average sleep duration of an elementary school-aged child. As with any drug, it is important to discuss potential side effects and contraindications. Reinforce with the parents the importance of stopping fluids 2 hours before and 8 hours after administration to prevent side effects of headache, nausea, abdominal discomfort, and vomiting, which are symptoms suggestive of water intoxication. In addition, the medication should not be given during illness or on evenings when there is excessive water intake.

Dosage of DDAVP needs to be individualized. The dose can be slowly increased from one tablet (0.2 mg) if no effect after several nights. One to three tablets (0.2 mg) each night before bedtime is the recommended dosage. Some studies have shown a reduced pharmacodynamic effect after 6 months of continuous use. Parents should be informed that DDAVP does not cure enuresis, but may stave off bedwetting for a period of time.

Desmopressin can also be prescribed as combination therapy with an enuresis alarm. The dose will need to be decreased to allow enuresis to occur in order for the alarm to activate. The wetting should then occur in the early

morning hours closer to waking and will be less in quantity. Most children who wet the bed do so within the first 90 minutes to 2 hours after going to sleep, which coincides with non-REM sleep. This makes them more difficult to awaken. By adding desmopressin to the enuresis alarm therapy and shifting the wetting episode to early morning, the child may become more successful with the alarm treatment. After a period of time, the medication can be reduced and then discontinued.[12]

Anticholinergic agents, such as oxybutynin, are also used for the treatment of enuresis, especially when there are concurrent symptoms of small bladder capacity, frequency, or urgency. Oxybutynin works by relaxing the smooth muscle of the bladder, allowing more urine to be stored and delaying the urge to void. Because of this effect, children should be followed closely for constipation or increased post-void residual, both of which can make enuresis worse. Tolterodine is another anticholinergic medication but has not been approved by the FDA for use in children.

For children with daytime symptoms, twice-a-day dosing may be helpful and can be increased to up to three times daily. For children without daytime symptoms, but a small bladder capacity, administration at bedtime is recommended. Immediate release tablets or syrup are the most useful options for these symptoms. As with desmopressin, oxybutynin can be used in conjunction with an alarm. As the child learns to wake to the alarm, the medication can be weaned.

Imipramine and other tricyclic antidepressants have been used to reduce bedwetting. The mechanism of action is an anticholinergic effect to increase bladder capacity combined with a noradrenergic effect that decreases the excitability of the bladder. In spite of this, due to unfavorable adverse effects such as mood changes and sleep disturbances and the risk of death with overdose, the International Children's Continence Society does not recommend the use of imipramine unless all other therapies have failed.[1]

12.4 Alternative Treatment

Other forms of treatment are widely available for nocturnal enuresis, including hypnosis, acupuncture, chiropractic medicine, energy therapy, and homeopathic or herbal therapy; however not all of these forms of therapy have published evidence to support their use. For families who desire another approach, alternative therapies may offer additional options for treatment. In terms of overall support, these options may provide a first-line treatment that is comparatively better than pharmacologic therapy, especially considering long-term cure rates after discontinuation of treatment.

12.5 Conclusion

Bedwetting is a common childhood condition that should be discussed as part of every child's examination. Once nocturnal enuresis has been identified as a problem and the child demonstrates a desire for dryness, initiating management is imperative. Prolonging treatment can be detrimental to the child's self-esteem and social development. A thorough history and exam are necessary in order to decide and address the underlying cause of the enuresis. Once the cause is identified, the treatment approaches can be introduced. Families are entitled to a full and balanced presentation of the treatment options available. This should include cost of those options, realistic expectations of time and effort, any negative effects, and probability of cure. In order for a family to make an informed decision and ensure the best possible adherence to therapy, all of the options must be offered.

Treatment options are available that will enable a child to sleep dry at night or wake to use the bathroom. Enuresis alarms and behavioral changes have much to offer enuretic children and appear to be more effective as first-line treatment when compared to pharmacological options. The benefit

of supportive management like medication, absorbent undergarments, or waterproof bedding should still be considered however, because treatment alone has been shown to improve a child's self-esteem, regardless of the type and success of the therapy. Needless to say, greater success results in greater improvement of self-esteem. No matter which option is chosen however, the provider, child, and the family need to outline a plan, including a system for measuring progress and regular follow-up visits.

12.5.1 Areas of Uncertainty

More research may help us better understand relationships between causative factors and enuresis. Children with enuresis do not arouse from sleep well. There is very limited evidence that these children may have abnormalities in the areas of the brain that mediate sleep arousal and micturation.

Constipation is another factor that plays a role in enuresis, but the mechanism is not well understood. Children have a smaller pelvic volume. Retained stool may cause decreased capacity of the bladder leading to enuresis. It has also been thought that a dilated rectum might stimulate colonic motility, which causes a contraction of the detrusor at lower than normal bladder volumes.

Lastly, behavioral therapy and alarm use are known to be helpful in enuresis. Better quality studies comparing alarms with other treatments are necessary, especially with follow-up to determine relapse rates.

12.5.2 Guidelines

The International Children's Continence Society (ICCS) has published recommendations for the evaluation and treatment of enuresis. The information presented in this chapter is consistent with the recommendations of the Society (Fig. 12.1).

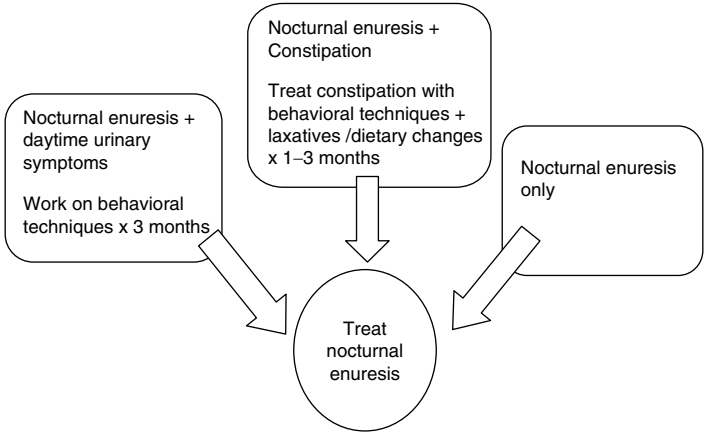


FIGURE 12.1. Treatment algorithm.

References

1. Neveus T, von Gontard A. The standardization of terminology of lower urinary tract function in children and adolescents: Report from the standardisation commitee of the international children's continence society. *J Urol*. 2006;176:314-324.
2. Hjalmas K, Arnold T, et al. Nocturnal enuresis: an international evidence-based management strategy. *J Urol*. 2004;171:2545-2561.
3. Feehan M, McGee R, Stanton W, Silva PA. A six year follow up of childhood enuresis: prevalence in adolescence and consequences for mental health. *J Paediatr Child Health*. 1990;26:75-79.
4. Schmitt BD. Nocturnal enuresis. *Pediatr Rev*. 1997;18:183-191.
5. Culbert TP, Banez GA. Wetting the bed: Integrative approaches to nocturnal enuresis. *Explore*. 2008;4:215-220.
6. Mercer R. *Seven Steps to Nighttime Dryness: A Practical Guide for Parents of Children with Bedwetting*. Ashton: Brookeville Media LLC; 2004.
7. Wolfish NM. Sleep/arousal and enuresis subtypes. *J Urol*. 2001;166:2444-2447.
8. Neveus T. Enuretic sleep: deep, disturbed or just wet? *Pediatr Nephrol*. 2008;23:1201-1202.
9. Maizels M et al. *Getting to Dry: How to Help Your Child Overcome Bedwetting*. Boston: The Harvard Common Press; 1999.
10. Glazener CMA et al. Alarm interventions for nocturnal enuresis in children. *Cochrane Database Syst Rev*. 2009:1-108.

11. Moffatt MEK, Harlos S, Kirshen AJ, et al. Desmopressin acetate and nocturnal enuresis: how much do we know? *Pediatrics*. 1993;92:420.
12. Mercer R. Dry at night: Treating nocturnal enuresis. *Adv Nurse Pract*. 2003;11:26-32.
13. Robson WL. Evaluation and management of enuresis. *N Engl J Med*. 2009;360(14):1429-1436.

Chapter 13
Disorders of Elimination: Constipation

J. Christopher Austin

Key Points

- The majority of children who present with urinary tract infections and problems with daytime and nighttime incontinence will have associated bowel symptoms such as constipation and encopresis.
- In children presenting with the complaint of constipation up to 2/3 will also have associated symptoms of bladder dysfunction.
- About 95% of patients with constipation have functional constipation and will respond to therapy with behavior modification (timed evacuation after meals) and laxative therapy.
- The treatment of constipation will improve or cure some children with daytime incontinence and recurrent urinary tract infections.

13.1 Introduction

Constipation is commonly seen in pediatric patients. It is a frequent complaint seen by pediatric gastroenterologists as well as primary care providers. Constipation is very relevant to pediatric urology patients as it is seen frequently in

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_13,

patients presenting with urinary tract infections and/or voiding dysfunction. *Dysfunctional elimination* is the combination of bowel and bladder dysfunction that was initially described in patients with recurrent urinary tract infections and vesicoureteral reflux. *It is important to seek out symptoms of bowel dysfunction in patients seen with urinary complaints and vice versa*. The treatment of constipation is an integral part of the treatment regimen in children with voiding dysfunction. Encopresis, or fecal incontinence associated with constipation and chronic fecal impaction may also be present in patients with voiding dysfunction and recurrent urinary tract infections. In children with constipation an organic cause should be sought out during the evaluation including neurologic, endocrine, or anatomic causes. In 95% of patients no etiology is found and they are classified as having functional constipation.

13.2 Definition of Constipation

There are multiple definitions of constipation. For research studies the Rome definition is often used, however, for clinical practice this is too restrictive. For simplicity constipation can be defined as symptoms lasting for more than 2 weeks of infrequent (less than 3 per week), delay or difficulty in having bowel movements, large hard or painful bowel movements, or fecal soiling (encopresis). *Given the relatively benign nature of the treatment of constipation with stool softeners or laxatives treating patients with bowel or urinary complaints should be considered when the diagnosis is suspected.*

13.3 Evaluation

The first step in evaluating a child with constipation is a thorough *elimination history.* This should include whether the child passed meconium within 48 h of birth, the frequency, size and consistency of bowel movements. The presence of

associated symptoms such as abdominal pain, vomiting, blood in the stool, withholding bowel movements, fever, and abdominal distention should be noted. The grading of bowel movements using an objective scale such as the Bristol stool scale may be useful confirming if the child is passing hard stools. Large diameter stools or stools which clog the toilet are a signs of constipation as well. The history should probe for possible organic causes of constipation such as hypothyroidism, botulism, medication side effects, or neurological causes as well as failure to thrive. As mentioned above there is a high incidence of concomitant bladder and bowel symptoms and thus the frequency of urination, presence of urgency, posturing, daytime or night-time incontinence, and urinary tract infections should be sought.

A *physical exam* should be performed with emphasis on the abdominal (palpable stool, tenderness, abdominal distention, rectal exam and anal position), neurological (reflexes, strength, anal wink and perineal sensation), and back exam (sacral dimple, lipoma, or hair tuft). Any suspicious neurological findings should lead to evaluation for a spinal cord abnormality such a tethered spinal cord. Plain abdominal films have been used to assess the fecal load and evaluate the spine for an occult dysraphism. There use is considered optional although many clinicians find them useful and objective in assessing for retained fecal load. Patients with no suspicious findings in the history and exam likely have functional constipation and should be treated.

13.4 Treatment of Constipation

Classically, the therapy for constipation has consisted of behavior modification (sitting to have a bowel movement for 5–10 min shortly after meals) and laxative therapy. The first step is bowel cleanout by disimpaction, which has traditionally been achieved with enemas but many practitioners now use oral laxatives and stool softeners with similar results. Once disimpaction is complete then the child is maintained on laxatives to keep the stools soft and allow the chronically

over distended rectum to regain its normal tone and function. There is evidence for the efficacy of laxative and stool softener therapy in constipation. No treatment is clearly superior. The most common treatments and their usual dosages are listed below. The use of other stool softeners such as docusate sodium has not been well studied in children and the efficacy is not known.

The therapy should be continued for the duration of at least 4–6 months before discontinuation with the goal of 1–3 bowel movements per day that are of a milkshake consistency during treatment. If the child has difficulty with fecal incontinence during therapy it needs to be ensured that they are adhering to the scheduled bowel evacuations and not trying to hold their stools. Of the following agents PEG 3350 is a safe and well tolerated choice to start with.

- *PEG 3350* (Miralax™, Glycolax): Effective dose for constipation has been reported in the 0.5–0.84 gm/kg/day given daily. Total dose may be divided in half and given twice daily which can help improve the stool consistency. The powder may be mixed with any good tasting fluids. The solution should be mixed with 17 gm of powder (filled to the mark inside the cap) into 240 mL (8 oz) and the appropriate amount of this dilution taken (i.e., a dose of 8.5 g would require drinking 120 mL (4 oz) of the solution). The mechanism of action of PEG 3350 is an osmotic agent which retains water in the stool to soften it and increase the frequency of bowel movement. The dose can be adjusted up or down every 3–5 day by an ounce until the desired stool consistency is reached.
- *Milk of magnesia* (magnesium hydroxide): Effective dose for treating constipation is 1–2 mL/kg/day given in a single or divided into a twice daily dose. The maximum recommended amount is 60 mL/day. Compliance with daily doses is less than with PEG 3350. It can be mixed or flavored with applesauce, chocolate syrup, or other flavorings to improve the taste. Its laxative effect is due to a hyperosmotic effect drawing fluid into the stool and the stimulation of increased peristalsis in the small intestine.

- *Mineral oil*: Dosage for children 6–12 years old is 5–15 mL/day. After age 12 years 15–45 mL/day. Not recommended for younger children due to the risk of aspiration. The oil helps soften stools by coating them and the intestine to reduce the colonic absorption of water and lubricate hard stools to facilitate passage.
- *Lactulose*: Supplied in a solution of 10 g/15 mL. The mechanism of action due to the synthetic disaccharide being degraded in the colon by bacteria producing lactic acid, acetic acid, and formic acid, which increase the osmotic pressure drawing water into the stool softening it.
- *Senna*: A laxative that stimulates peristalsis and is available as both oral syrup (218 mg/5 mL) and a tablet (167 mg). For children 1–5 years of age the recommended dose is 2.5–5 mL of syrup (109–218 mg) given at bedtime. For children age 5–15 years old a dose of 5–10 mL of the syrup can be used initially. Alternatively 1–2 tablets can be given at bedtime. Senna should only be used short term.

13.5 Indications for Referral

Patients with constipation that is suspected to be due to an organic cause based on the history or physical exam should be referred for further testing which may include rectal manometry, barium enema, or spinal MRI. Patients with persistent symptoms after adequate treatment for 3–6 months should be referred to pediatric gastroenterologist.

Suggested Readings

1. Allen HA, Austin JC, Boyt MA, et al. Evaluation of constipation by abdominal radiographs correlated with treatment outcome in children with dysfunctional elimination. *Urology*. 2007;69:966-969.
2. Baker SS, Liptak GS, Colletti RB, et al. Constipation in infants and children. *J Pediatr Gastro Nutr*. 1999;29:612-626.
3. Chase JW, Homsey Y, Siggaard C, et al. Functional constipation in children. *J Urol*. 2004;171:2641-2643.

4. Erickson BA, Austin JC, Cooper CS, Boyt MA. Polyethylene glycol 3350 for constipation in children with dysfunctional elimination. *J Urol*. 2003;170:1518-1520.
5. Hadjizadeh N, Motamed F, Abdollahzade S, Fafiei S. Associate of voiding dysfunction with functional constipation. *Indian Pediatr*. 2009;46:1093-1095.
6. Heaton KW, Ghosh S, Braddon FEM. How bad are the symptoms and bowel dysfunction of patients with the irritable bowel syndrome? A prospective, controlled study with emphasis on stool form. *Gut*. 1991;32:73-79.
7. Koff SA, Wagner TT, Jayanthi VR. The relationship among dysfunction elimination syndromes, primary vesicoureteral reflux, and urinary tract infections in children. *J Urol*. 1998;160:1019-1022.
8. Loening-Baucke V. Urinary incontinence and urinary tract infection and their resolution with treatment of chronic constipation of childhood. *Pediatrics*. 1997;100:228-232.
9. Loening-Baucke V, Pashenkar DP. A randomized, prospective comparison study of polyethylene glycol 3350 without electrolytes and milk of magnesia for children with constipation and fecal incontinence. *Pediatrics*. 2006;118:528-535.
10. Pashankar DP, Bishop WP, Loening-Baucke V. Long-term efficacy of polyethylene glycol 3350 for the treatment of chronic constipation in children with and without encopresis. *Clin Pediatr*. 2003;42:815-819.
11. Voskuil WP, Heijmans J, Heijmans HS, et al. Use of Rome II criteria in childhood defecation disorders: applicability in clinical and research practice. *J Pediatr*. 2004;145:213-217.

Chapter 14
Hematuria and Proteinuria

Alan R. Watson

Key Points

- Macroscopic hematuria is a rare symptom in childhood and should be referred for evaluation to the pediatric department.
- Commonest cause for macroscopic and microscopic hematuria (on dipstick) is urinary tract infection and the major causes in adults of stones and cancer are rare in childhood.
- Dipsticks are very sensitive for blood and urinalysis should be repeated before referral. If persistent dipstick hematuria test family members as benign familial hematuria is not uncommon.
- Heavy proteinuria on dipstick (+++ or more) needs prompt referral but lesser degrees need confirmation with repeat testing. Orthostatic (postural) proteinuria is common in older children and can be deduced from testing early morning and late afternoon urines.
- A combination of hematuria with proteinuria are more likely to be significant than isolated microscopic hematuria and low level proteinuria alone.[1]

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_14,

14.1 Hematuria

The dipsticks are very sensitive for blood and can be positive at <5 RBCs per high power field. Evidence of red blood cells in the urine should be confirmed by microscopy which should be performed on fresh urine. Contrast microscopy has been advocated with anatomically normal red blood cells suggesting bleeding of lower urinary tract origin as distinct from dysmorphic red cells suggesting a higher (glomerular) source. However, the test is very rarely applied in pediatric practice. Microscopy of urine can help in diagnosis with the presence of red cell *casts* indicating glomerular bleeding.

Causes of "red urine" need to be excluded:

- Urate crystals, especially in young infants giving "pink" nappies
- Food coloring, e.g., beetroot or blueberries
- Hemoglobinuria and intravascular hemolysis
- Myoglobinuria, e.g., in rhabdomyolysis
- Drugs, e.g., Rifampicin
- External source, e.g., menstrual blood loss
- Fictitious – consider if no cause found.

14.1.1 Important Points in the History

- Symptoms suggestive of UTI, e.g., fever, frequency, and dysuria. UTI quoted as commonest cause for macroscopic and microscopic hematuria.
- Urine red (more likely to be a local cause) or tea/coke colored (more likely to be glomerular with oxidized blood)
- Is the hematuria at the beginning or end of the stream (bladder or urethral cause)
- Is there colicky abdominal pain (suggesting stones)
- History of trauma
- Is there history of coagulopathy such as easy bruising
- Is there a family history of hematuria or of renal disease and deafness (Alport's syndrome) or sickle cell disease

- Examination will include palpation of the abdomen for abdominal masses, skin rashes and blood pressure

14.1.2 Causes of Hematuria

- Urinary tract infections:
  - Bacterial
  - Viral (e.g., adenovirus in outbreaks)
  - Schistosomiasis (history of foreign travel)
  - Tuberculosis
- Glomerular
  - Post-infectious glomerulonephritis
  - IgA nephropathy, Henoch-Schonlein purpura, SLE
  - Hereditary – thin basement membrane, Alport's syndrome
- Urinary tract stones: hypercalciuria
- Trauma
- Other renal tract pathology
  - Renal tract tumor
  - Polycystic kidney disease
- Vascular
  - Renal vein thrombosis
  - Arteritis
- Hematological: coagulopathy/sickle cell disease
- Drugs – cyclophosphamide
- Exercise induced

14.1.3 Investigations

When evaluating childhood hematuria it is important to identify serious and progressive conditions. If macro- or microscopic hematuria is found in the setting of an acute illness such as UTI then it needs to be confirmed by urine culture and the urine tested after the acute illness. Glomerulonephritis will require a full "nephritis work-up." However, if the child presents with asymptomatic "benign" hematuria without growth failure, hypertension, edema, proteinuria, urinary casts or renal impairment then investigations can be arranged in the outpatient clinic.

- Urine
  - Microscopy to confirm RBCs and look for casts
  - Culture for bacteria and possibly viruses
  - Protein : creatinine ratio to define level of proteinuria if present (normal <20 mg protein/mmol creatinine) on an early morning urine
  - Calcium : creatinine ratio on second morning urine to exclude hypercalciuria (normal <0.7 mmol calcium/mmol creatinine)
- Bloods
  - U&E/creatinine/albumin/calcium/phosphate/alk phos
  - FBC/clotting
  - Complement: C3/C4
  - ASOT and antihyaluronidase B titers
  - ANA/anti-dsDNA and ANCA titers if nephritis suspected
- Radiology
  - Ultrasound of urinary tract of all patients with hematuria
  - Further radiology will depend upon clinical situation and initial ultrasound
- Test urine of parents and other children in clinic (hereditary causes)
- Cystoscopy – *not routine* and rarely indicated in children. Heavy isolated hematuria, recurrent fresh bleeding and abnormal findings on ultrasound may provide indication

14.1.4 Management

- If obvious cause such as UTI treat with antibiotics but will require at least renal tract ultrasound.
- If impaired renal function, proteinuria or family history refer to pediatric nephrology unit.
- If suspicious lesion on ultrasound of urinary tract or intermittent macroscopic hematuria (with IgA nephropathy excluded in an older child) then discuss with radiology/urology.
- If no cause found and normal renal function, BP and no proteinuria monitor in clinic at 6-monthly intervals.
- If persistent microscopic hematuria after one year or change in any of the above parameters then refer to pediatric nephrology unit for consideration of renal biopsy.[2]

Pediatric nephrologists debate the role of renal biopsy in benign persistent microscopic hematuria. One approach is shown in Fig. 14.1. One cause of persistent microscopic hematuria is thin basement membrane nephropathy with abnormal thinning of the glomerular basement membrane. This is probably the commonest cause of benign familial hematuria. When thinning is combined with thickening of the glomerular basement membrane (basket weave appearance) and a

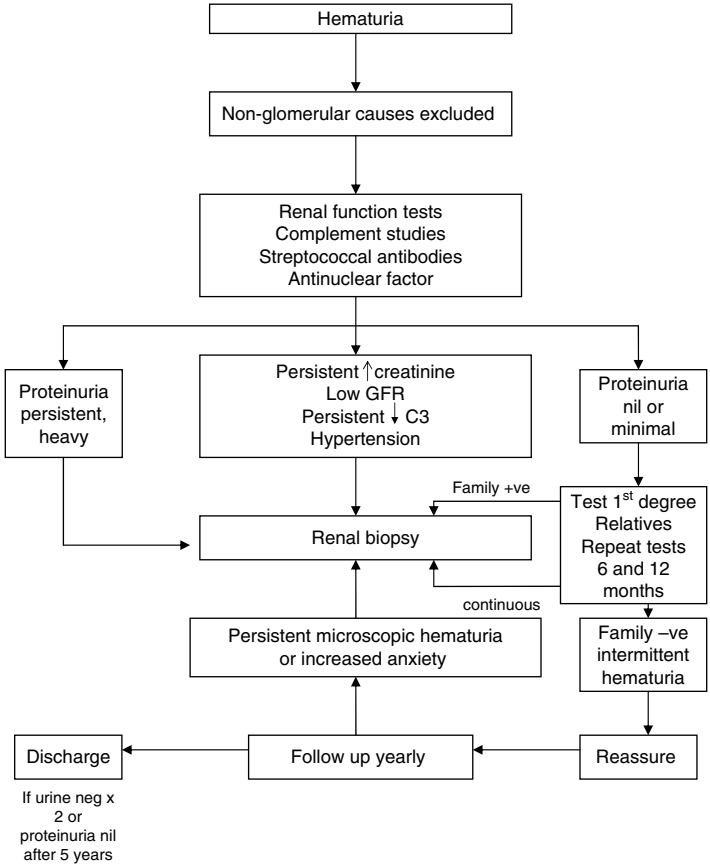


FIGURE 14.1. A scheme for the management of children with hematuria.

family history of deafness or progressive nephritis then Alport's syndrome is a likely diagnosis. This can lead to progressive nephritis needing dialysis and transplantation, especially in boys as 80–90% of cases are X-linked dominant.

14.2 Proteinuria

Protein may be found in the urine of healthy children and usually does not exceed 0.15 g/24 h. It is usually detected on dipstick testing which predominantly detects albumin in the urine (Table 14.1).[3]

14.2.1 Quantification of Proteinuria

We have largely abandoned 24 h urine collection in children but instead collect an early morning urine (EMU) for measurement of the urinary protein : creatinine ratio. (normal <20 mg protein/mmol creatinine)

Microalbuminuria will not be detected by dipsticks but is often assessed in patients with diabetes and is defined as UA:UCr of >2.5 mg/mmol.

14.2.2 Causes of Proteinuria

Proteinuria may be due to benign or pathological causes.

Table 14.1. Urinalysis by dipstick testing.

Test result	Equivalent protein estimate (g/L)
+	0.2
++	1.0
+++	3.0
++++	≥20

14.2.2.1 Non-Pathological Proteinuria

- Transient
- Fever
- Exercise
- Urinary tract infection

14.2.2.2 Orthostatic Proteinuria (Postural Proteinuria)

This is a common cause of referral in older children. There is usually no history of significance and examination is normal. The UP/UCr ratio in the early morning urine should be normal with an elevated level in the afternoon specimen. If confirmed and no other findings on history and examination then the proteinuria should be regarded as a benign finding and requires no investigation or treatment.

14.2.2.3 Pathological Proteinuria

- Nephrotic syndrome is recognized as heavy proteinuria (>200 mg/mmol), hypoalbuminemia (<25 g/L) and edema
- Glomerulonephritis is recognized by associated hematuria, clinical features and investigation
- Chronic kidney disease is associated with glomerulosclerosis or a reduced nephron mass from any cause resulting in hyperfiltration

Current consensus is to treat proteinuria associated with suspected low nephron mass with ACE inhibitors which may slow the rate of CKD progression. Hence documentation of persistent proteinuria and referral to pediatric nephrology is important.

- Tubular disease may give rise to proteinuria and require measurement of urine NAG and urine retinal binding protein to creatinine ratios.

14.2.3 Investigations

- Early morning urine protein : creatinine ratio and afternoon specimen if orthostatic proteinuria suspected
- Urine microscopy and culture
- If proteinuria combined with hematuria then investigations directed at causes of hematuria and nephritis
- Renal tract ultrasound
- Patients with proteinuria (>20 mg/mmol on EMU) for a period of 6–12 months should be referred to pediatric nephrology centre for consideration for renal biopsy (Fig. 14.2)

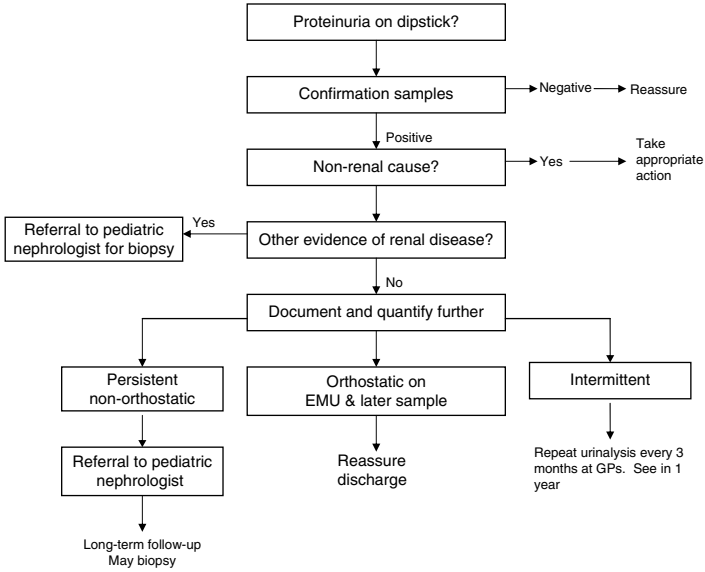


FIGURE 14.2. Scheme for assessment of a child with proteinuria.

References

1. Milford DV, Robson AM. The child with abnormal urinalysis, haematuria and/or proteinuria. In: Webb N, Postlethwaite R, eds. *Clinical Paediatric Nephrology*. 3rd ed. 2003:1-29.
2. Hussain F, Watson AR, Hayes J, Evans J. Standards for renal biopsies: comparison of inpatient and day care procedures. *Pediatr Nephrol*. 2003;18:53-56.
3. Christian M, Watson AR. The investigation of proteinuria. *Curr Paeds*. 2004;14:547-555.

Chapter 15
Abdominal Pain – Urological Aspects

Pedro-Jose Lopez and Carolina Acuña

Key Points

- Abdominal pain is a common symptom in pediatric practice.
- Different pathologies may cause abdominal pain including a number of urological conditions.
- It is relevant to have these possible conditions in mind when studying pediatric patients.
- This chapter will review some of these pathologies and their initial management.

15.1 Introduction

Abdominal pain is one of the most common symptoms in pediatric practice. Etiologies behind it are multiple; a number of urological conditions present themselves with abdominal pain which may be classified as follows:

a. Obstructive
b. Nonobstructive pathologies: infectious diseases, malignancies and gynecological conditions

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_15,

A. *Obstructive urological pathologies*:

1. The most typical of them all is undoubtedly urolithiasis Fig. 15.1. Nevertheless, the classical acute abdominal or flank pain that characterizes this pathology in adults occurs in only 50% of children. Pre-school children are rather asymptomatic and are usually diagnosed after a UTI. Other clinical manifestations are hematuria, dysuria and/or urinary frequency. Evaluation of these patients include a complete clinical history with emphasis on dietary habits, metabolic evaluation and, ideally, stone analysis. Diagnosis in children is generally achieved with ultrasound, which may detect stones at the renal pelvis, ureteropelvic junction, proximal and distal ureter; nonetheless, it does not visualize stones at the mid ureter. Signs due to obstruction may suggest the presence of urolithiasis. In older patients plain abdominal x-ray may be diagnostic. Non enhanced abdominal computed tomography is the gold standard in the diagnosis of this pathology, although a relevant issue in children is the possible risk of accumulated radiation exposure. Therapeutic alternatives are multiple; expectant approach, extracorporeal shockwave lithotripsy and surgery. In turn, surgery offers

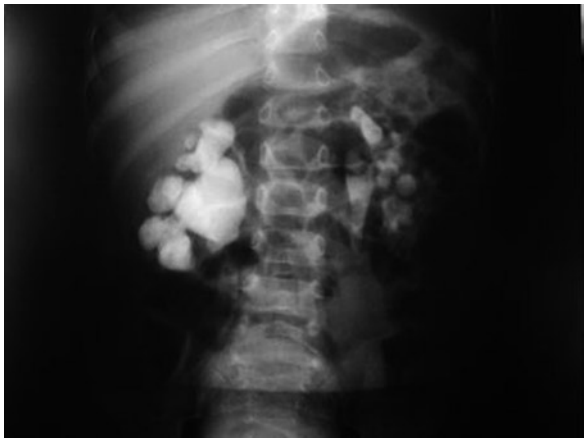


FIGURE 15.1. Urolithiasis: multiple radio opaque stones in both kidneys.

various approaches, especially minimally invasive surgery with endourologic procedures such as ureteroscopy, percutaneous nephrolithotomy and laparoscopic procedures.

2. Pelvicureteric junction (PUJ) obstruction Figs. 15.2 and 15.3 is the most common congenital urinary obstruction. Before the spread of prenatal diagnosis, the most common presenting symptom in patients with PUJ obstruction was abdominal pain, followed by UTI and hematuria. Initial study should include ultrasonography which may show the dilation caused by the PUJ obstruction, renal parenchyma thickness and suggest if renal dysplasia exists. Dynamic renal scintigraphy is very useful in the assessment of possibly obstructed kidneys. Some of these obstructions are non progressive and asymptomatic making relevant the need to determine which kidneys will require a surgical management. Also, in these patients, you should not discard concomitant problems like vesicoureteral reflux or urinary stone disease, especially in adolescents. The surgical repair consists of a pieloplasty either open or laparoscopically approached, which has a high rate of success and a low rate of complications.

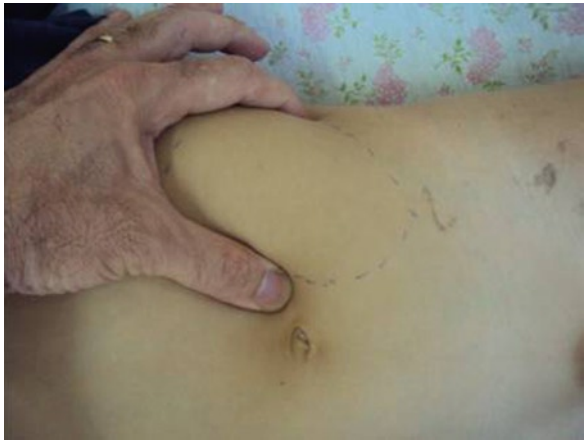


FIGURE 15.2. Palpable abdominal mass in a 4 year old patient, who presented with abdominal pain and constipation.

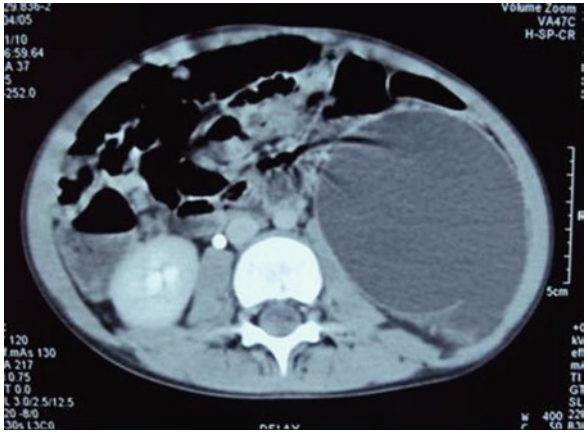


FIGURE 15.3. Abdominal CT from same boy showing mass with no renal tissue observed: diagnosis of Right PUJ obstruction.

3. The wide ureter or megaureter is caused by either obstruction or reflux occurring at the ureterovesical junction. They can be classified in primary megaureters, ureteral defects, and secondary, defects whose "cause" is extraureteral. This explains why other forms of megaureters appear such as non obstructive – non refluxing or refluxing obstructed megaureters. Megaureters, especially primary ones, should be suspected in prenatal ultrasound. If not, they usually are diagnosed while studying a patient with UTI, hematuria, abdominal mass or cyclic abdominal pain. Work up must include ultrasound, voiding cystourethrogram, dynamic nuclear renography and, in specific cases, cystoscopy. When anatomy is unclear, an enhanced computed tomography should be requested in order to determine the specific cause of the wide ureter and plan the appropriate surgical approach.
4. Urinary tract obstructions at other levels, such as bladder outlet and urethra, have different clinical manifestations. They may sometimes include abdominal pain but clearly not as a prime symptom.

B. *Nonobstructive urological pathologies*:

1. Urinary tract infections are the most prevalent bacterial disease during the first three months of life and account for almost 10% of febrile episodes in infants. Clinical presentation is variable, especially considering age, gender, pathogen and associated anatomical malformations. Pyelonephritis is generally characterized by fever, abdominal pain, dysuria, frequency, and hematuria. Symptoms of peritoneal irritation may be present. Considering that symptoms are usually non specific, UTI has to be ruled out in a febrile infant. Diagnosis is made with a properly obtained urine specimen; eventually a DMSA is useful to demonstrate acute pyelonephritis and secondary scarring.
2. Renal tumors also may present with abdominal pain. The most common primary malignant renal tumor of childhood is Wilm's tumor Figs. 15.4 and 15.5 and the most common

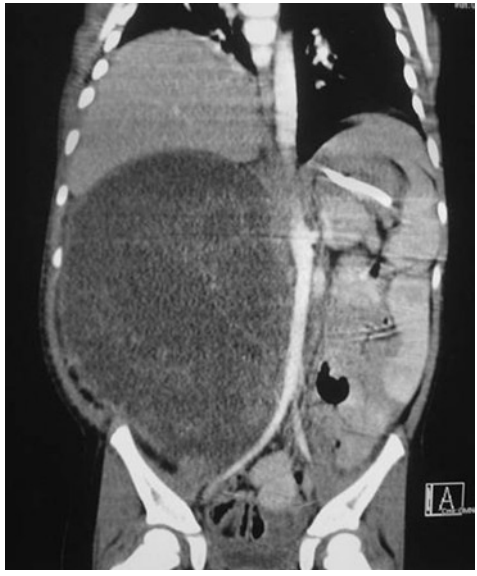


FIGURE 15.4. MRI: Large heterogeneous mass at the right flank showing a kidney tumor.

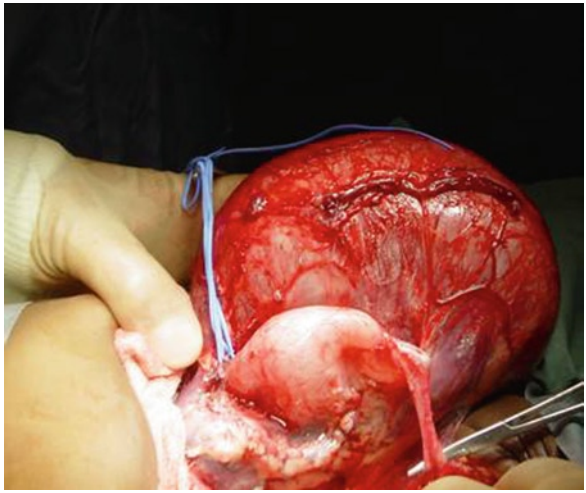


FIGURE 15.5. Same Wilm's Tumor from Fig. 14.4 at surgery.

benign solid tumor is mesoblastic nephroma. Generally, renal tumor presentation is through palpable abdominal mass. Additional findings include abdominal pain, hematuria, and sometimes pyelonephritis among others. Diagnostic evaluation should include routine laboratory tests, tumor markers, imageneologic studies for accurate staging and in search for metastasis. These patients should be referred to the specialist as soon as possible.

3. Symptoms that may lead you to suspect a vesico ureteric reflux (VUR) in a pediatric patient are generally related to urinary tract infections. In newborns and pre-school patients symptoms are vague such as irritability, failure to thrive, poor feeding, vomiting and fever. Classic presentation occurs in older children. In this group, patients with VUR and UTI present abdominal pain as a prime symptom.
4. Considering the intraabdominal location of the female reproductive tract, different gynecological conditions should be considered as possible etiologies of abdominal pain. A characteristic example are ovarian masses, especially ovarian torsion. Abdominal pain, nausea, vomiting and palpable abdominal

mass is the classical presentation of ovarian torsion, whose diagnosis still remains as one of exclusion. None of the actual imaging studies is definitive in the diagnosis, although ultrasound is useful. The best diagnostic modality in ovarian torsion is laparoscopy; it is also the treatment of choice.

5. Finally, multiple genital anomalies may include abdominal pain as a presenting symptom. Congenital vaginal obstruction as a result of an incomplete canalization of the vagina is a diagnosis to consider. An imperforate hymen can result in hydrocolpos, distension of the vagina, and sometimes with distension of the vagina and uterus, known as hydrometrocolpos Fig. 15.6. These patients are usually diagnosed at newborn age with a palpable lower abdominal mass, urinary tract obstruction, and abdominal pain. If no abdominal mass is present at birth, these patients may remain asymptomatic until adolescence when they present amenorrhea, cyclic abdominal pain and abdominal mass (hematometrocolpos). All these patients should be referred promptly to the specialist.

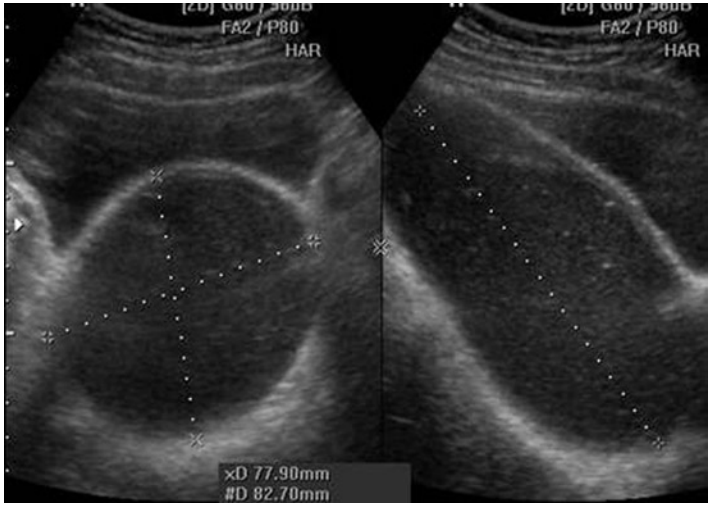


Figure 15.6. Ultrasound: Hypoechoic fluid content distending the uterus and vagina.

15.2 Indications for Referral

a. Abdominal pain with urinary tract dilatation at any level.
b. Urinary tract infection diagnosed during evaluation of a patient with abdominal pain.
c. Patients with urolithiasis.
d. Renal mass, ovarian mass and/or pelvic mass which may present with abdominal pain.
e. Pelvicureteric junction obstruction ideally at an early age when prenatal diagnosis has been made. In older children it should be considered in patients with pain and/or with abdominal mass.

References

1. Gillenwater Y, Grayhack Y, Howards S, Duckett Y. *Adult and Pediatric Urology*, vol. 3. 3rd ed. St. Louis: Mosby; 1996.
2. Tekgul S, Riedmiller H, Gerharz E, Hoebeke P, Kocvara R, Nijman R, Radmayr Chr, Stein R. *Guidelines on Paediatric Urology*. European Society for Paediatric Urology © European Association of Urology 2008.
3. The American Urological Association Pediatric Vesicoureteral Reflux Clinical Guidelines Panel. Report on The Management of Primary Vesicoureteral Reflux in Children. Copyright © 1997 American Urological Association, Inc.
4. Boccon-Gibod L. Pathological evaluation of renal tumors in children: International society of pediatric oncology approach. *Pediatr Dev Pathol*. 1998;1:243-248.
5. Oltmann S, Fischer A, Barber R, et al. Cannot exclude torsion, a 15 year review. *J Pediatr Surg*. 2009;44:1212-1217.

Part II
Pediatric Surgery

Chapter 16
External Angular Dermoid and Pilomatrixoma

Ashish Wakhlu

Key Points

- External angular dermoids may have intracranial extension.
- Complete resection is curative.
- Pilomatrixoma usually occurs in a hair bearing area on the front of the trunk.
- Recurrence after resection of pilomatrixoma is rare.

16.1 Introduction

This is the commonest site of a dermoid swelling in children. External angular dermoids are congenital swellings occurring at the outer corner of the eye typically appearing as a mass beneath the outer end of the eyebrow. They form at the sites of the lines of embryonic fusion where ectodermal elements either get trapped beneath the skin or fail to disconnect from the developing neural tube. The swelling, despite being attached to the underlying periosteum is mobile and non tender. Untreated, the swelling grows in size, eventually impinging on opening of the eye.

The dermoid cyst occupies a shallow depression in the underlying bone and occasionally may be dumbbell shaped,

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_16,

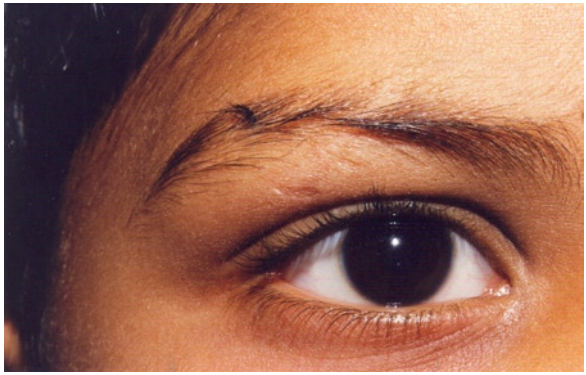


FIGURE 16.1. External angular dermoid with punctum.

protruding intra cranially through a defect in the bone. Such cysts are less mobile than normal.

Clinically the dermoid presents as a mobile non tender swelling under the lateral end of the eyebrow, occasionally there is a punctum, signaled by deformity of a few eyebrow hairs (Fig. 16.1). X-ray of the skull and CT scan is necessary if intracranial protrusion is suspected.

16.2 Treatment of Angular Dermoid

The treatment of external angular dermoid is excision, with care to remove the whole lining of the cyst to prevent recurrence. This can usually be accomplished through a skin crease incision. For intracranial extension a craniotomy may be accomplished by lowering a frontal flap.

Pathologically angular dermoids contain sebaceous material with hair follicles and glandular epithelium. Recurrence is uncommon unless part of the lining has been left behind.

16.3 Indications for Referral

Presence of a swelling under the lateral end of the eyebrow with or without an opening.

16.4 Pilomatrixoma (Calcifying Epithelioma of Malhebre)

16.4.1 Introduction

This is a rare lesion, occurring in middle aged men, in a hair bearing area (usually the front of the trunk). It presents as a painless slowly enlarging subcutaneous nodule, mobile and non tender. Average size of the lesion is 0.5–1 cm in diameter. With larger lesions thinning of the overlying skin may occur. The tumor is hard and may feel gritty.

Treatment of the Pilomatrixoma is excision, usually accomplished under local anesthesia. The tumor is easily removed in its entirety.

Pathological examination shows large pale cells resembling epidermal keratinocytes and empty "shadow" cells devoid of nuclei. There is a fibrocollagenous stroma with multinucleated giant cells and small areas of calcium deposition.

Recurrence of a pilomatrixoma is uncommon.

Indications for Referral – presence of a hard mobile non tender subcutaneous nodule.

Suggested Reading

1. Smith CD. Cysts and sinuses of the neck. In: Grosfeld JL, O'Neill JA, Fonkalsrud EW, Coran AG, eds. *Pediatric Surgery*. 6th ed. St. Louis, MO: Mosby Elsevier; 2006:861-874.

Chapter 17
Neck Swellings/Lumps: Midline Neck Swellings

Neil Bateman

Key Points

- Thryoglossal duct cysts are the commonest congential abnormality in the neck. They can usually be easily diagnosed clinically.
- The diagnosis of a thyroglossal duct cyst can be confirmed, and the presence of normal thyroid can established, be ultrasound.
- A midline block excision of tissue from the level of the thyroid up to the tongue base, including the middle third of the hyoid bone is the treatment of choice in thyroglossal duct cysts.
- Dermoid cysts can be distinguished from thyroglossal duct cysts by their failure to move on tongue protrusion.
- Dermoid cysts can be easily treated by simple surgical excision. If there is any doubt over the diagnosis then the lesion should be treated as a thyroglossal cyst.

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_17,

17.1 Introduction

Neck lumps occurring in children can be usefully divided into those occurring in the midline of the neck, discussed here, and those situated laterally. While it is very uncommon for midline neck lumps represent sinister pathology it is very important that these lesions are accurately diagnosed and treated by those with a specialist interest as the treatment of those children receiving initial inappropriate treatment can be very challenging.

17.2 Differential Diagnosis of Midline Neck Lump

17.2.1 Thryoglossal Duct Cyst

This is a developmental abnormality and represents the commonest congenital abnormality in the head and neck. The thyroglossal duct is formed by the descent of the thyroid gland in the fetus from the foramen cecum in the tongue base to its familiar position anteriorly in the neck. Under normal circumstances the duct involutes between the 8th and 10th week of gestation. Cysts forming in a persistent duct are clinically known as thyroglossal duct cysts. The thyroglossal duct is closely related to the hyoid bone. This gives rise to the cardinal clinical sign of these lesions, namely elevation on tongue protrusion.

Thyorglossal duct cysts can present at any age although it is very rare for them to be present at birth. They are almost all in the midline (Fig. 17.1) although they can occur more laterally in the neck. When this happens, it is more commonly on the left. Recurrent episodes of infection may occur and, if sufficiently severe and frequent, this may lead to cyst rupture and subsequent sinus formation.

It is theoretically possible, although very rare, that a lesion with the appearance of a thryoglossal duct cyst is, in fact, an incompletely descended thyroid gland. Under these circumstances it is possible that the lesion represents the only viable thyroid tissue. There are some case reports of thyroid carcinoma occurring in these lesions.

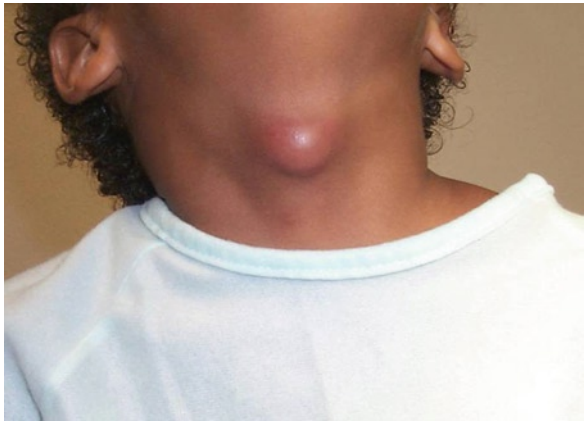


FIGURE 17.1. Thyroglossal cyst in the midline.

17.2.2 Midline Dermoid Cyst

These are developmental abnormalities caused by abnormalities in midline fusion in the fetus. They appear clinically as midline lesions occurring at any level in the neck. They do not move on tongue protrusion.

17.2.3 Lymph Nodes

While lymphadenopathy in the neck is most commonly situated laterally, submental nodes especially may present in the midline.

17.2.4 Thyroid Nodule

Pyramidal lobe nodules or those occurring in the thyroid isthmus may present as a midline lump, although thyroid nodules occur most commonly laterally in the neck.

17.2.5 "Plunging" Ranula

A ranula is an extravasation pseudocyst caused by obstruction of a minor salivary gland in the floor of the mouth.

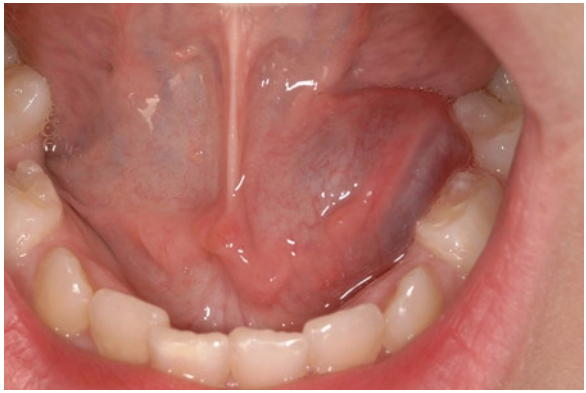


FIGURE 17.2. Ranula in the floor of the mouth.

Typically they present as a easily recognizable lesion in the mouth (Fig. 17.2). A "plunging" ranula is one which has become so large as to present in the neck. The diagnosis is usually apparent on intra-oral examination.

17.2.6 Investigations

In most children a presumptive clinical diagnosis can be confirmed with ultrasound. This can differentiate between cystic and solid lesions. While ultrasound is a highly user dependant modality it is often possible to positively identify lymph nodes using it as well as other common lumps such as lipomata. In the case of thyroglossal duct cysts ultrasound is also used to identify the presence of a normal thyroid gland. Occasionally other imaging modalities such as MRI can be helpful although these are more commonly used in larger, more complex neck masses.

Fine needle aspiration, while in adults the investigation of choice for many neck lumps, is very rarely helpful in investigating neck lumps in children.

17.3 Treatment

17.3.1 Thryoglossal Duct Cyst

The treatment is surgical, with excision of the cyst and any associated tract, including the mid-third of the hyoid bone. The classical operation, known as Sistrunk's procedure, where the cyst and tract are identified and followed to the tongue base is, in many people hands, being superceded by a midline block dissection of neck tissues up to the foramen cecum. The rationale behind this is that the thyroglossal duct is a complex branching structure and this technique is less likely to leave portions of duct remaining which carry the risk of recurrence.

17.3.2 Midline Dermoid Cyst

Simple excision is curative.

17.3.3 Lymph Nodes

These can be managed as any other lymph node in the neck (see Chap. 18).

17.3.4 Plunging Ranula

These me be excised although often marsupialization of the cyst intra-orally with excision of the sublingual glands is sufficient. Excision carries a risk of damage to structures in the floor of mouth including the hypoglossal nerves.

Chapter 18
Neck Swellings/Lumps: Lateral Neck Lumps

Neil Bateman

Key Points

- Lymphadenopathy in children is the commonest cause of laterally placed neck lumps.
- The majority of children with a lymph node in the neck have self limiting viral induced reactive lymphadenopathy.
- Referral and/or biopsy is indicated where there is persistent adenopathy greater than 2 cm in diameter, systemic symptoms, nodes in multiple sites or where the lump has a sinister clinical appearance.
- Cancer is the second commonest cause of death in children. Where there is any doubt about the cause of a neck lump then biopsy should be urgently considered.
- Hemangiomata are an entirely distinct clinical entity from vascular malformations and require different treatment strategies.

18.1 Introduction

The majority of laterally placed neck lumps in children originate in lymph nosed. Cervical lymphadenopathy is very common in children and in the vast majority of children will be

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_18,

self limiting and benign. Clinicians must therefore be aware of the potential causes of lymph node enlargement in children and the indicators of serious pathology.

18.2 Differential Diagnosis of Lateral Neck Lumps

18.2.1 Lymph Nodes

The causes of lymphadenopathy in the neck are

18.2.1.1 Infective

Viral: Upper respiratory viral infections may cause lymphadenopathy which is usually self limiting. Other common childhood viral infections such as measles, mumps, and rubella can also cause lymphadenopathy.

Infectious mononucleosis, caused by Epstein Barr virus infection, can cause massive lymphadenopathy, often accompanied by tonsillitis, fever, lethargy, and malaise. The diagnosis can often be confirmed with a serological test (such as the Monospot or Paul Bunnell) and there are often atypical lymphocytes on the blood film.

Human Immunodeficiency Virus may present with persistent generalized lymphadenopathy. The transmission in children is usually vertical from mother to child. They may be a history of recurrent or opportunistic infections. This is a diagnosis which could be considered in any patient with an unknown cause of lymphadeopathy.

Bacterial: Acute lymphadenitis may occur with infection with pyogenic organisms, most commonly streptococci and staphylococci. Occasionally gram negative organisms may be responsible. If untreated this may progress to an acute inflammatory mass and/or suppuration to form an abscess (Fig. 18.1). Often these are relatively superficial and require an external incision and drainage. Infection within the deep neck spaces can present more of a management challenge

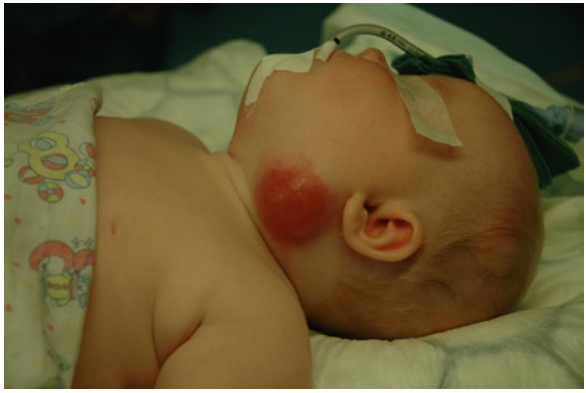


FIGURE 18.1. A superficial neck abscess in a baby.

and carry a risk of airway obstruction or progression to medistinal infection.

Mycobacterial infection: Tuberculosis presenting in the neck is rare in UK practice. More common is non-tuberculous mycobacterial infection, sometimes referred to as atypical mycobacterial infection or incorrectly as atypical TB. These organism cause infection via an oral route and tend to infect those lymph nodes draining the oral cavity (i.e., submandibular and intraparotid nodes most commonly). They most commonly present as a painless mass in an otherwise entirely healthy child. If left untreated the skin over the lump develops a violaceous discoloration and given more time eventually breaks down to form a discharging sinus (Fig. 18.2). If left untreated this situation is likely to continue for some months or years before the wound heals, often with an unpleasant puckered scar. Treatment of this condition is either medical (and a variety of antibiotic have been employed including clarithromycin, azithromycin, ciprofloxacin as well as more traditional antituberculous drugs) or surgery. The resistance of mycobacteria to antibiotic treatment means that, if medical treatment is planned, antibiotic treatment often requires several weeks or months of treatment. There is little doubt that surgery is technically easier and has a greater chance of a good outcome in early disease.

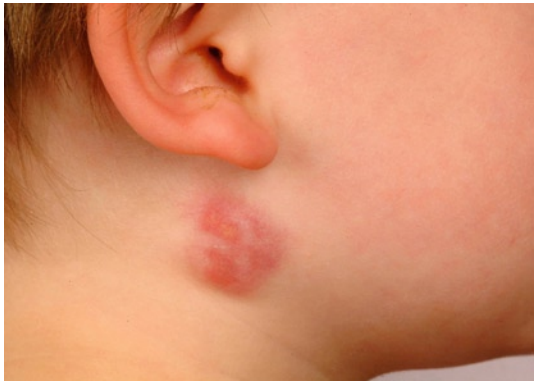


FIGURE 18.2. Non-tuberculous mycobacterial infection in the parotid gland showing the characteristic skin discoloration.

Other bacterial infection which can present with lymphadenopathy include brucellosis, cat scratch disease, actinomycosis, and syphilis.

18.2.1.2 Inflammatory

Kawasakis disease is an inflammatory, non infective disease which presents with cervical lymphadenopathy, pyrexia, desquamation of the hands and feet, erythema of the oral cavity and conjunctivitis. It is an important diagnosis to consider as there is a potential mortality of 1–2% due to cardiac complications. The treatment is by gamma globulin administration.

18.2.1.3 Neoplastic

Fortunately head and neck malignancy is rare in children but cancer is the second commonest cause of childhood death in the UK and a significant proportion of pediatric cancers involve the head at some stage. Early diagnosis and appropriate management in a specialist centre is of vital importance to a good outcome.

Hodgkin's disease occurs most commonly in teenagers and young adults and characteristically affects lymph nodes rather than extra-nodal sites. The typical history is progressive enlargement of cervical lymph nodes. There may be systemic symptoms such as anorexia, weight loss and night sweats. Hepatosplenomegaly indicates more advanced spread of the disease. Diagnosis is by lymph node biopsy and treatment by chemotherapy and/or radiotherapy.

Non-Hodgkin's Lymphoma is a heterogeneous group of malignancies distinguished pathologically from Hodgkin's disease by the absence of Reed Sternberg cells. Extra-nodal sites (e.g., tonsil, adenoids) may be affected in the head and neck rather than lymph nodes. A younger age group (2–12 years) is more typically affected. Diagnosis is by a biopsy and treatment modalities similar to that of Hodgkin's disease.

Other malignancies presenting as neck lumps include rhabdomyosarcoma, thyroid malignancy and salivary gland tumors. Congenital teratomata may present as large masses in the head and neck.

18.2.2 Vascular Malformations and Vascular Tumors

Hemangiomata should be clearly distinguished from vascular malformations. Hemangiomata are benign neoplasms of endothelium which grow rapidly in the first few weeks of life, often having been undetectable at birth. They commonly affect the parotid area and can appear alarming to parents and doctors alike. Although they are known to respond to systemic steroids or propranolol treatment as well as being amenable to surgical excision they will, if left alone, involute spontaneously so treatment is only really required for lesions causing a functional problem (such as swallowing or airway problems, or in facial lesions where there is closure of one eye).

Vascular malformations are present at birth and are a developmental abnormality. Lymphatic malformations (cystic hygroma, lymphangioma) may be considered with this

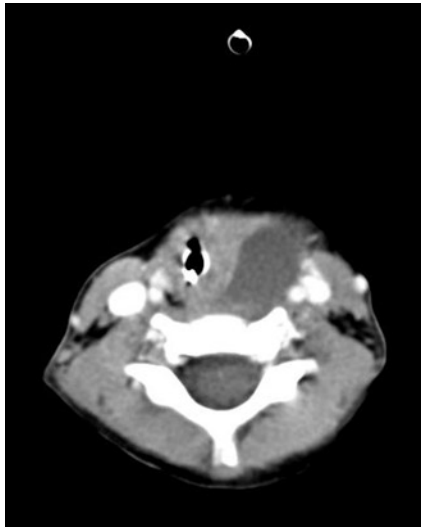


Figure 18.3. CT scan showing mulitioculated lymphatic malformation in the parapharyngeal space extending inferiorly on the left side of the neck. There was airway obstruction and the child presented with snoring.

group of lesions (Fig. 18.3). They do not proliferate and do not resolve spontaneously. Treatment is either conservative, surgical (where the functional or cosmetic problem outweighs the implications of surgery), or, in the case of lymphatic malformations sclerotherapy may be considered (Fig. 18.4).

18.2.2.1 Investigations

Investigations for lymphadenopathy are aimed at determining the likelihood of there being any sinister underlying pathology in order to identify those children who should undergo excision biopsy. Lymph nodes in the neck are a very common problem and many children would undergo unnecessary surgery if they were all biopsied.

Ultrasound is a very useful modality to identify numbers and size of nodes. In experienced hands (and this is a very

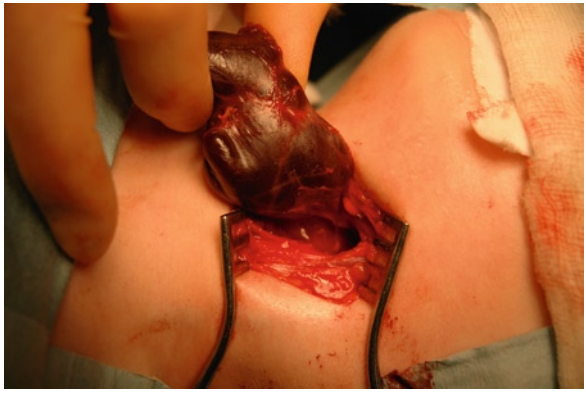


FIGURE 18.4. The lesion in Fig. 18.3 at surgical excision.

operator dependant modality) abnormalities in node morphology can indicate sinister pathology.

CXR may indicate mediastinal lymphadenopathy in children with lymphoma.

FBC may be abnormal in patients with hematological malignancy.

Serology may be helpful in establishing a diagnosis and thereby avoiding unnecessary biopsies.

Fine needle aspiration is uncommonly performed in children. It is less acceptable for patients and parents than in adults and FNAC has limited usefulness in the diagnosis of lymphoma.

Excision biopsy is, in general, indicated in those children:

- Who have multiple nodes in many areas of the neck, especially those with supraclavicular nodes (which have an increased likelihood of sinister pathology)
- Who have a fixed node or one with obvious skin involvement
- Who have persistent nodes of greater than 2 cm diameter
- Who have abnormalities on CXR or FBC
- Who have unexplained systemic symptoms (e.g., anorexia, weight loss, night sweats, etc.)

Chapter 19
Neck Swellings/Lumps: Torticollis

Stephanie Jones

Key Points

- Torticollis, or wryneck, is a congenital or acquired condition of infants and children that causes tilting of the neck to one side secondary to contraction of the neck muscles.
- Congenital torticollis can resolve spontaneously, but rarely resolves spontaneously if infant >1 year old.
- Children with torticollis may have associated bony or intracranial abnormalities.
- Lack of treatment may lead to deformational plagiocephaly, cosmetic deformity, or functional impairment.

19.1 Introduction

Torticollis is a condition seen in newborns and children that results in an effective shortening of the sternocleidomastoid muscle (SCM) causing the head and neck to rotate toward the affected side, the chin to rotate opposite. This condition can be congenital or acquired. The incidence can be as high as 1:250 infants in the newborn period for congenital muscular torticollis, most studies show an incidence of 1–2% in the general population. Acquired torticollis has an incidence of

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_19,

that varies according to the etiology. This chapter will define the two types of torticollis and discuss the evaluation and treatment of both.

19.2 Etiology and Types of Torticollis

1. Congenital muscular torticollis (CMT): The etiology is controversial with proposed causative factors being birth trauma, an ischemic event, venous occlusion, intrauterine malposition, genetic factors, constitutional growth arrest, infective myositis, neurogenesis, and intrauterine compartment syndrome.
   a. CMT is divided into three groups, the most common being Group 1 – the sternocleidomastoid tumor (SMT). This consists of torticollis and a palpable SCM tumor, called fibromatosis colli, which is palpable in the body of the SCM and present at birth. Group 2, or muscular torticollis (MT) is torticollis with a tightness of the SCM, but no palpable tumor. Group 3, or POST (postural torticollis) has no mass or tightness of the SCM. The usual age of presentation is 2–4 months of age, with SMT presenting earliest and POST presenting later.
   b. Evaluation begins with physical exam showing the head tilted toward the shortened SCM and the chin rotated opposite. If a palpable tumor is not present in SCM, ultrasound or MRI may be used to identify a fibrotic lesion in the SCM and to differentiate from other pathologies in the neck. Any bony abnormalities seen on radiographs is a contraindication to manual therapy.
2. Acquired or non-muscular torticollis (NMT): Torticollis described from as many as 80 different etiologies resulting in the characteristic head and neck tilt. In a retrospective study of 288 patients with confirmed torticollis, the incidence of NMT was 18.4% of all cases of torticollis. NMT was subdivided into the following categories:
   a. Klippel-Feil and congenital scoliosis – 5.6% of all torticollis, 30.2% of all non-muscular causes

b. Ocular disorders – 4.2%, 22.6% respectively
c. Central nervous system abnormalities (posterior fossa and cervical spine tumors, syringomyelia or Arnold-Chiari malformation) – 2.1%, 11.3%
d. Obstetrical palsies – 3.1%, 17%
e. Clavical fractures – 0.7%, 3.8%
f. C1-C2 rotary subluxations – 0.7%, 3.1%
g. Inflammatory conditions – 0.7%, 3.1%
h. Idiopathic or unexplained – 1.4%, 7.6%

3. Work-up of non-muscular causes of torticollis can be performed based on the algorithm described by Ballock et al. (Fig. 19.1). After a history and physical exam excludes CMT, based on a palpable mass of the SCM and early age of presentation, inflammatory causes or birth trauma are ruled out. Cervical and bony radiographs define any bony abnormalities. An eye exam defines abnormalities causing ocular torticollis. A neurologic examination, possibly including a MRI, identifies CNS or spinal cord abnormalities. Finally, pain indicates the need for evaluation of an osteoma or osteoblastoma. If CMT or the above causes of torticollis are ruled out, observation and physical therapy are indicated.

19.3 Treatment of Torticollis

1. Positioning and handling of infant should encourage infant to rotate head toward affected side during feeding or crib placement.
2. Manual stretching – best outcome is achieved when conducted by a trained physiotherapist. First line of therapy for initial treatment or for mild cases. May incur a snapping sound that is thought to be tearing of the SCM body or fibrotic band. No studies have shown adverse outcomes when such a sound is heard during therapy.
3. Orthotics – tubular orthosis for torticollis (TOT) collar – used in conjunction with physiotherapy or after surgery. This soft collar is best used for children around 4–6 months of age.

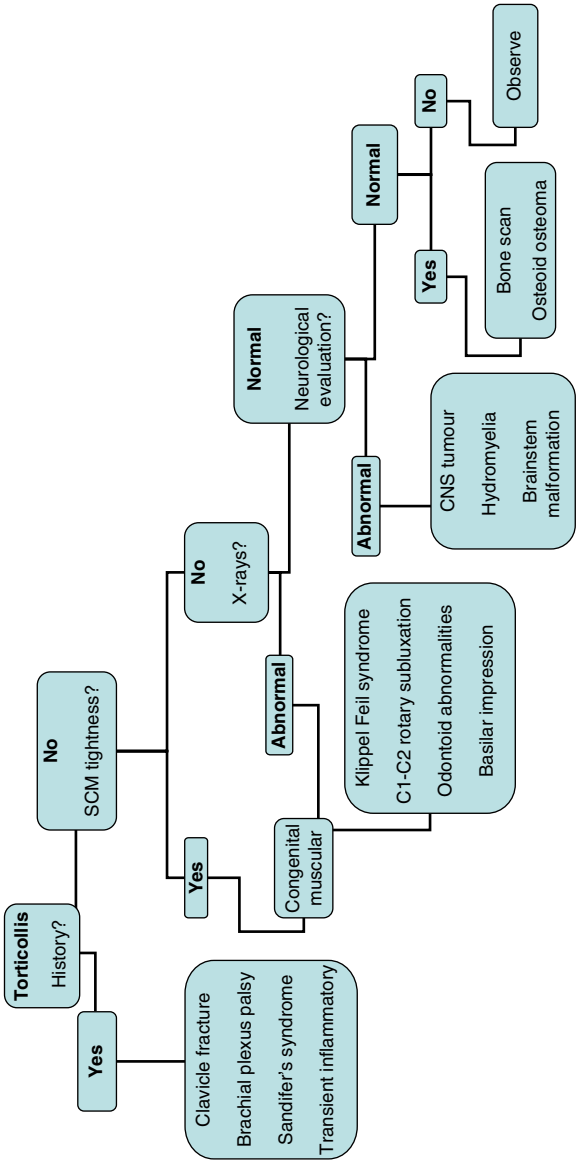


FIGURE 19.1. Algorithm for the management of torticollis (Adapted from Ballock et al. 1996).

4. Surgical therapy – definitive therapy of refractory torticollis. Surgeon preference dictates surgery performed, as no studies have proven benefit of one type of operation over another. Surgical options include simple myotomy, uni- or bipolar release of the SCM, z-plasty or myoplasty, subperiosteal lengthening of the SCM at its origin and insertion, or resection of the SCM itself.
5. Non-conventional therapy – botox injection – not well tested or commonly used.

19.4 Indications for Referral

1. Most CMT will resolve 2–6 months of age. If it persists, referral to a physiotherapist is indicated.
2. If NMT is suspected, the underlying etiology should be investigated.
3. If deficit of passive rotation >10°, refer to physiotherapist for manual stretching.
4. If patient has significant head tilt and deficits of passive rotation and side flexion >10°–15°, the presence of a tight band or mass in the SCM, has not responded to 6 months of physiotherapy manual stretching, refer to a surgeon for surgical treatment.

Suggested Readings

1. Do TT. Congenital muscular torticollis: Current concepts and review of treatment. *Curr Opin Pediatr*. 2006;18:26-29.
2. Tang SP, Cheng J. Outcome of surgical treatment of congenital muscular torticollis. *Clin Orthop Relat Res*. 1999;362:190-200.
3. Ballock RT, Song KM. The prevalence of nonmuscular causes of torticollis in children. *J Pediatr Orthop*. 1996;16(4):500-504.
4. Cheng JC, Tang SP, Chen TM, Wong MN, Wong EM. The clinical presentation and outcome of treatment of congenital muscular torticollis in infants – a study in 1086 cases. *J Pediatr Surg*. 2000;35:1091-1096.
5. Canale ST, Griffin DW, Hubbard CN. Congenital muscular torticollis. A long-term follow-up. *J Bone Joint Surg*. 1982;64:810-816.

6. Ohman A, Nilsson S, Lagerkvist AL, Beckung E. Are infants with torticollis at risk of a delay in early motor milestones compared with a control group of healthy infants? *Dev Med Child Neurol*. 2009;51:545-550.
7. Chen JC, Wong MW, Tang SP, Chen TM, Shum SL, Wong EM. Clinical determinants of the outcome of manual stretching in the treatment of congenital muscular torticollis in infants. *J Bone Joint Surg*. 2001;83A(5):679-687.
8. Rogers GF, Oh AK, Mulliken JB. The role of congenital muscular torticollis in the development of deformational plagiocephaly. *Plast Reconstr Surg*. 2009;123:643-652.
9. Lin JN, Chou ML. Ultrasonographic study of the sternocleidomastoid muscle in the management of congenital muscular torticollis. *J Pediatr Surg*. 1997;32(11):1648-1651.
10. Rosman NP, Douglass LM, Sharif UM, Paolini J. The neurology of benign paroxysmal torticollis of infancy: Report of 10 new cases and review of the literature. *J Child Neurol*. 2009;24(2):155-160.
11. Mezue WC, Taha ZM, Bashir EM. Fever and acquired torticollis in hospitalized children. *J Laryngol Otol*. 2002;116:280-284.
12. Koumanidis S, Per H, Gismos H, et al. Torticollis secondary to posterior fossa and cervical spinal cord tumors: report of five cases and literature review. *Neurosurg Rev*. 2006;29:333-338.

Chapter 20
Umbilical Disorders

Spencer W. Beasley

Key Points

- Umbilical hernias are common in small children, occurring in about 10% of infants after separation of the umbilical cord, but the vast majority of them involute spontaneously.
- Umbilical hernias virtually never ulcerate, rupture, strangulate or give pain.
- Umbilical hernias only need to be repaired if they are still present beyond the age of 3–4 years: this is done as a day case under a short general anesthetic.
- A moist lump at the umbilicus is usually an umbilical granuloma or ectopic bowel mucosa, and can be treated with topical application of silver nitrate.
- Discharge of air, or ileal content in the presence of a small opening suggests a vitellointestinal tract; and discharge of urine from an opening at the umbilicus indicates a urachus.

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_20,

20.1 Introduction

The umbilicus can be the site of a variety of abnormalities, most of which are minor. The most common is the umbilical hernia, and about 90% of these involute spontaneously, usually in the first few years of life. In contrast to inguinal hernias, strangulation of an umbilical hernia is extremely uncommon.

20.2 Common Umbilical Conditions

1. Umbilical hernia: Umbilical hernia presents as a skin-covered swelling at the umbilicus, first seen after separation of the cord (Fig. 20.1). It increases in size with increasing abdominal pressure, (e.g., with crying, straining) but reduces when the infant is relaxed.
2. Umbilical granuloma: this appears as a fleshy pink protuberance which is moist, causing yellow staining of the overlying clothing. It becomes apparent after separation of the umbilical cord, and has no sinus opening (Fig. 20.2a).
3. Ectopic bowel mucosa at the umbilicus: this has an appearance similar to an umbilical granuloma but tends to be more "cherry red" and may bleed more readily.

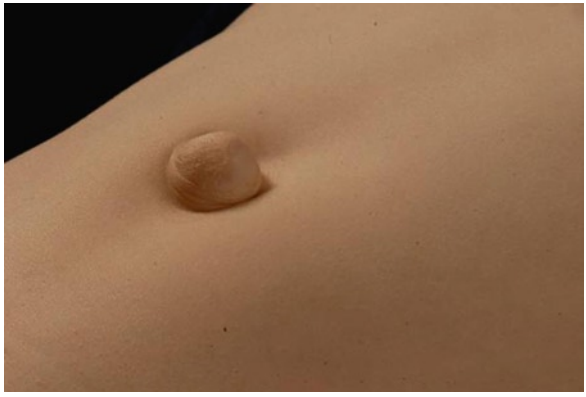


FIGURE 20.1. Typical appearance of an umbilical hernia.

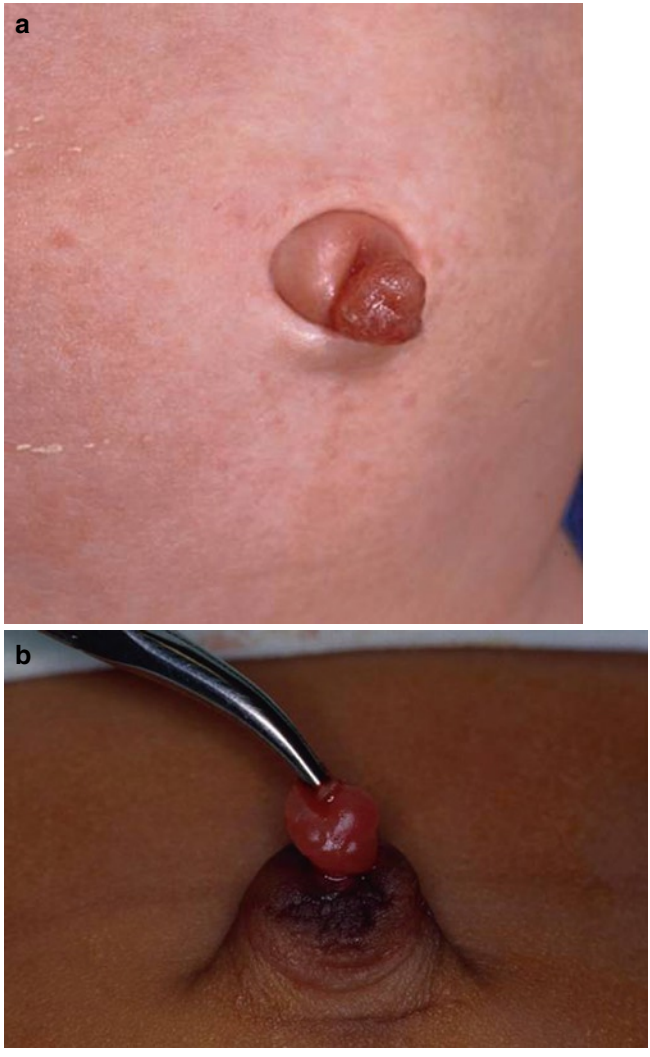


FIGURE 20.2. (**a**) An umbilical granuloma presents as a moist and fleshy protuberance at the umbilicus that has no sinus opening. (**b**) An umbilical granuloma often has a "stalk." Here it is being held up by forceps.

4. Patent urachus: there is an opening at the umbilicus through which urine passes (because there is a connection with the bladder). A urachal remnant may also present as a blind ending sinus or as a urachal abscess inferior to the umbilicus.
5. Patent vitellointestinal tract: there is an open communication between the ileum and the umbilicus. Gas and fluid ileal contents can pass through it, appearing from a small opening on the umbilicus (Fig. 20.3). There are a number of variants of this condition including a sinus, Meckel's diverticulum and Meckel's band.
6. Exomphalos (or omphalocele): the umbilicus is widely open, and bowel and liver protrude through the broad umbilical cicatrix, but are contained by a sac consisting of peritoneum and an amniotic layer. Often the infant has associated abnormalities such as cardiac and renal defects, or Beckwith-Wiedemann Syndrome.
7. Gastrochisis: there is a relatively small defect immediately to the right of the umbilicus through which bowel protrudes antenatally, without any evidence of a sac (thus distinguishing it from exomphalos). The bowel is often

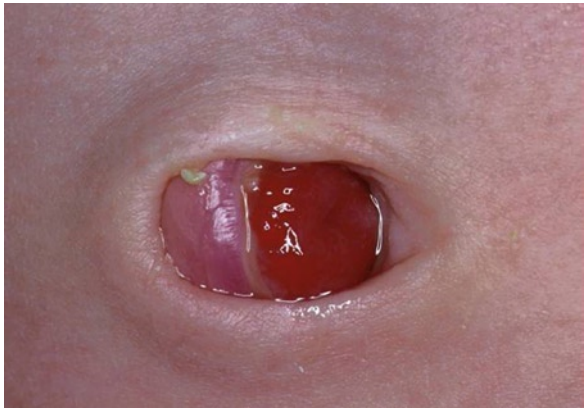


FIGURE 20.3. A patent vitello-intestinal tract has an opening through which fluid ileal contents and air can leak.

edematous and thickened, and there may be an associated bowel atresia.

8. Omphalitis: this is a neonatal infection emanating from the umbilicus, causing streaks of erythema radiating from the umbilicus.

20.3 Treatment of Conditions of the Umbilicus

1. Umbilical hernia: Those that do not resovle spontaneously are repaired after 3 years of age as a day case under a short general anesthetic. The scar is within the umbilicus, and recurrence is unlikely.
2. An umbilical granuloma and ectopic bowel mucosa are treated either with topical application of silver nitrate to the base of the stalk, or by ligation of the (often narrow) base with a suture tied tightly. Care should be taken to ensure that there is no sinus opening, as this may indicate an underlying vitellointestinal tract or urachus.
3. Where a patent urachus is suspected, imaging with a sinugram or ultrasonography may reveal the extent of the abnormality. Treatment involves surgical excision.
4. Vitellointestinal tract: this also can be confirmed by a sinugram. Once confirmed, treatment is by surgical excision of the tract.
5. Exomphalos (or omphalocele): this is often first diagnosed on antenatal ultrasonography. Its characteristic appearance makes it easily diagnosed at birth. Associated abnormalities (e.g., renal, cardiac, and chromosomal) should be excluded. Emergency management involves prevention of excessive heat loss (wrap the torso including the defect in plastic film and place the infant in a warm humidicrib), insertion of an intravenous line, and nasogastric decompression. These children are then treated in a neonatal unit. Surgery returns the bowel contents to the peritoneal cavity and the defect is closed. For extremely large defects, preliminary non-surgical treatment by external compression may be required.

6. Gastrochisis: this is a surgical emergency. Immediate management at birth involves protection of the bowel by wrapping it in plastic film, prevention of heat loss, and insertion of a nasogastric tube. The bowel should be returned to the abdominal cavity as soon after birth as possible, and the defect closed.

20.4 Indications for Referral

1. An umbilical hernia that has not resolved by the age of 3–4 years.
2. A presumed umbilical granuloma/ectopic bowel mucosa unresponsive to ligation or topical application with Silver Nitrate.
3. Suspicion of a patent vitellointestinal tract or urachus (both will have a sinus opening at the umbilicus).
4. All exomphalos and gastrochisis infants: antenatal diagnosis allows transfer to a pediatric surgical centre prior to birth; failing that, the newborn infant is transported to the tertiary centre as soon after birth as possible.

20.5 Epigastric Hernia

20.5.1 Introduction

An epigastric hernia becomes apparent as a small midline lump, usually midway between the xiphisternum and umbilicus (Fig. 20.4). The swelling is often less than a centimeter in diameter. The bulge is due to extra-peritoneal fat protruding through a small defect in the linea alba.

The parents often notice a small non-tender swelling in the midline or adjacent to it. It may be more prominent or cause discomfort after meals.

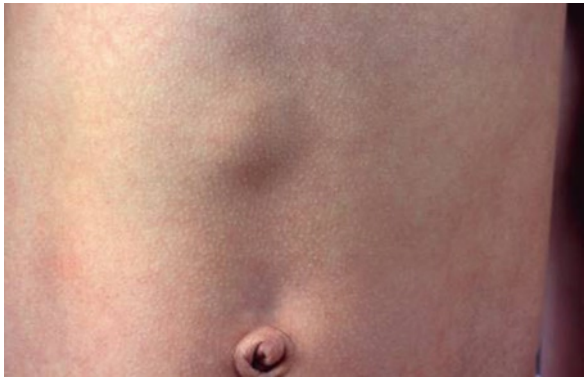


FIGURE 20.4. An epigastric hernia appears as a midline lump usually midway between the xiphisternum and umbilicus.

It is a relatively harmless condition, but if it causes discomfort, it is best repaired under a short general anesthetic as a day case.

20.6 Divarication of the Rectus Abdominus Muscle

This is best considered a variant of normal, rather than an abnormality. Often, in infants and small children, there is a longitudinal bulge between the xiphisternum and the umbilicus that is most obvious when the abdominal muscles (rectus abdominus) are contracted. This can be demonstrated by getting the child to lift his/her head up while lying supine (Fig. 20.5).

It is of no significance, other than it often causes concern to parents or a medical practitioner unfamiliar with the condition.

No treatment is required. It has no long term sequelae. It never causes discomfort.

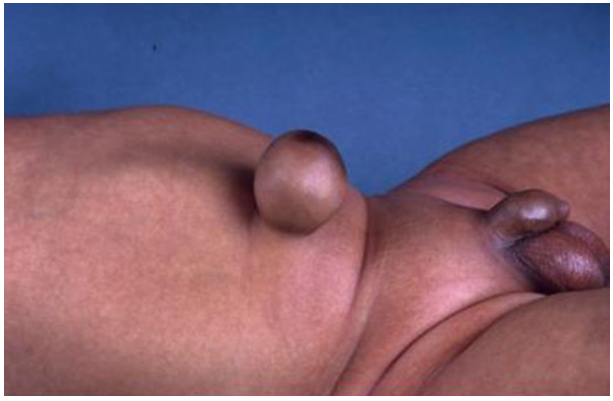


FIGURE 20.5. Divarication of the rectus abdominus in association with an umbilical hernia.

References

1. The umbilicus. In: Hutson JM, O'Brien M, Woodward AA, Beasley SW, eds. *Jones' Clinical Paediatric Surgery: Diagnosis and Management*. 6th ed. Blackwell Publishing; 2008:117-120.
2. Campbell J, Beasley SW, McMullin ND, Hutson JM. Umbilical swellings and discharges in children. *Med J Aust*. 1986;145:450-453.

Chapter 21
Surgical Aspects of Abdominal Pain

John A. Sandoval

Key Points

- Appendicitis, intussusception, and malrotation with volvulus may be among the most elusive diagnoses in children.
- Appendicitis may occur together with another illness (gastroenteritis).
- Midgut volvulus, intussusception, and ovarian torsion are the three surgical conditions that have acute onset of pain as the initial symptom.
- There is a need to arrive at a diagnosis of acute abdomen promptly, as a delay may have devastating consequences to the child.

21.1 Introduction

Abdominal pain remains a common complaint in children and accounts for a frequent number of surgical consultations. Rapid, accurate diagnosis of abdominal pain in children reduces the morbidity of common causes of pediatric abdominal pain. Clinical evaluation may help identify which children with abdominal pain should undergo immediate surgical consultation and which children with equivocal presentations

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_21,

should undergo further diagnostic evaluation The important clues provided by a thorough history and physical examination along with an understanding of the multiple sources of abdominal pain and the formulation of a limited differential diagnosis is crucial for the pediatric patient presenting with either acute, subacute, or chronic abdominal pain (Fig. 21.1). A number of conditions may cause abdominal pain and may lead to a specialist referral. This chapter deals with common sources of abdominal pain among pediatric patients, their etiology and management in primary/emergency care. Indications for referral will be highlighted.

21.2 Common Sources of Abdominal Pain

21.2.1 Children

1. Gastroenteritis: Acute gastroenteritis (AGE) is inflammation of the mucous membranes of the gastrointestinal tract, and is the most common cause of abdominal pain in children. The most common causes are viruses (rotavirus, Norwalk virus, adenovirus, and enterovirus), but bacterial (*Escherichia coli*, *Yersinia*, *Campylobacter*, *Salmonella*, and *Shigella*), protozoal and helminthic GE may be responsible for AGE in developing countries
2. Constipation: A common and distressing pediatric problem. Cause is usually functional (stool retention) but may also have organic etiologies such as Hirschsprung's disease, appendicitis, pseudo-obstruction, spinal cord abnormality, hypothyroidism, diabetes insipidus, cystic fibrosis, gluten enteropathy, or congenital anorectal malformation.
3. Appendicitis: The most common surgical disease of the abdomen in children. Approximately one in 15 people develop appendicitis and the disease may vary considerably in its clinical presentation, contributing to delay in diagnosis and increased morbidity.
4. Intussusception: The most common cause of intestinal obstruction in children between 3 months and 6 years.

Right upper quadrant
Biliarydisease: Biliary colic, cholecystitis, choledocholithiasis, cholangitis
Hepatic: Hepatitis, neoplasm, abscess, congestive hepatopathy
Colonic: Colitis, right-sided diverticulitis
Pulmonary: Pneumonia, subphrenic abscess, embolism, pneumothorax
Abdominal wall: Herpes zoster, muscle strain
Renal: Nephrolithiasis, pyelonephritis, perinephri cabscess

Right lower quadrant
Small intestine: Meckel's diverticulum, IBD, mesenteric adenitis, intussusception
Colonic: Appendicitis, colitis diverticulitis, IBD, IBS
Gynecologic/Testicular: Ectopic pregnancy, ovarian tumor/torsion, endometriosis, hematocolpos, PID, testicular torsion/tumor
Renal: Nephrolithiasis, pyelonephritis, ureteropelvic junction obstruction
Abdominal wall: Herpes zoster, muscle strain, incarcerated/strangulated hernia

Hematologic/Metabolic/ Drugs
Sickle cell anemia, Henoch-Schonlein purpura, hemolytic uremic syndrome, diabetic ketoacidosis, hypoglycemia, porphyria, acute adrenal insufficiency, uremia, hypercalcemia, erythromycin, salicylates, lead poisoning, venoms, opiate withdrawl

Miscellaneous
Infantile colic, functional pain, pharyngitis, hereditary angioedema, malingering, lactose intolerance, Famial Mediterranean fever

Left upper quadrant
Gastric: gastritis, esophagitis, peptic ulcer
Spleen: infarction or rupture,
Pancreas: pancreatitis or mass
Pulmonary: Pneumonia, subphrenic abscess, embolism, pneumothorax
Renal: Nephrolithiasis, pyelonephritis, perinephric abscess

Left lower quadrant
Colonic: Colitis, diverticulitis, IBD, IBS
Gynecologic/Testicular: Ectopic pregnancy, ovarian tumor/torsion, endometriosis, hematocolpos, PID, testicular torsion/tumor
Renal: Nephrolithiasis, pyelonephritis, ureteropelvic junction obstruction
Abdominal wall: Herpes zoster, muscle strain, incarcerated/strangulated hernia

FIGURE 21.1. Selected differential diagnosis of pediatric abdominal pain by anatomic region.

Intussusception occurs when a more proximal portion of bowel invaginates into more distal bowel. These patients often present with a wide range of non-specific symptoms, with less than one quarter presenting with the classic triad of vomiting, abdominal pain, and bloody stools.

5. Bowel obstruction: Intestinal obstruction in the newborn infant and older child may be due to a variety of conditions, including atresia and stenosis, annular pancreas, malrotation, duplication cyst, meconium ileus, meconium plug syndrome and neonatal small left colon syndrome, Hirschsprung's disease, neoplasia, trauma, and other rarer causes.
6. Incarcerated hernia: results when bowel becomes swollen, edematous, engorged, and trapped outside of the abdominal cavity. Incarceration is the most common cause of bowel obstruction in infants and children and the second most common cause of intestinal obstruction in North America (second only to intra-abdominal adhesions from previous surgeries).
7. Meckel's diverticulum: The most common congenital malformation of gastrointestinal tract. It can cause complications in the form of ulceration, hemorrhage, intussusception, intestinal obstruction, perforation and, rarely, vesicodiverticular fistulae and tumors.
8. Ovarian torsion: A problem in the pediatric age group that must be included in the differential diagnosis of any girl with abdominal pain or a pelvic or abdominal mass.
9. Ureteropelvic junction (UPJ) obstruction: Frequent in pediatric age and is a common cause of upper urinary tract obstruction that can be clinically silent or lead to symptoms such as pain, chronic urinary tract infections, and urinary stone disease.

21.2.2 Infants

1. Colic: a syndrome of persistent crying in infants, with multiple causes.
2. Hypertrophic pyloric stenosis: Is a gastrointestinal tract disorder common in infancy. The disorder causes projectile vomiting, weight loss, and fluid and electrolyte abnormalities.

3. Malrotation with midgut volvulus: A life-threatening surgical emergency in which the intestine becomes twisted as a result of malrotation of the intestine during fetal development. Because the consequences of malrotation associated with midgut volvulus may be catastrophic, prompt diagnosis and treatment are required to prevent mortality and short-gut syndrome.
4. Necrotizing enterocolitis (NEC): The most common disease process of the gastrointestinal (GI) tract of premature neonates that results in segmental or total bowel necrosis.

21.3 Treatment of Conditions

1. Nonsurgical conditions: Gastroenteritis, constipation, colic, and other extra-abdominal conditions require treatment directed at the underlying cause.
2. Surgical conditions: In general, prompt surgical intervention is required for pathologies associated with:
   a. Vascular compromise (malrotation with volvulus, irreducible incarcerated hernias, nonreduced intussusception, adhesive bowel obstruction, solid organ torsion, and trauma)
   b. Perforated viscus (acute appendicitis, NEC, Meckel's diverticulum, trauma, and foreign bodies)
   c. Acute hemorrhage (Trauma, Meckel's diverticulum, and ectopic pregnancy)

21.4 Indications for Surgical Referral in Children with Abdominal Pain

1. Increasing abdominal pain with progressive clinical deterioration
2. Bilious vomiting
3. Involuntary abdominal guarding or rigidity
4. Rebound abdominal tenderness
5. Marked abdominal distension with tympany
6. Acute fluid or blood loss into the abdomen

7. Abdominal trauma
8. Abdominal pain without clear etiology

References

1. Acute abdomen. *Semin Pediatr Surg* 1997;6:57-111.
2. Ashcraft KW. Consultation with the specialist: acute abdominal pain. *Pediatr Rev*. 2000;21:363-367.
3. Leung AK, Sigalet DL. Acute abdominal pain in children. *Am Fam Physician*. 2003;67:2321-2326.
4. Mason JD. The evaluation of acute abdominal pain in children. *Emerg Med Clin North Am*. 1996;14:629-643.
5. Buchert GS. Abdominal pain in children: An emergency practitioner's guide. *Emerg Med Clin North Am*. 1989;7(3):497-517.

Chapter 22
Approach to Abdominal Masses

Kenneth W. Gow and Martin A. Koyle

Key Points

- Most abdominal masses originate from the retroperitoneum with the kidney accounting for more than half of the lesions.
- Most lesions in infants are benign and malignant in older children.
- Diagnostic imaging commonly begins with an ultrasound followed by a CT scan to characterize the lesion and determine the anatomical relationships.

22.1 Introduction

Pediatricians commonly see abdominal masses in their patient population. While most abdominal masses will not require surgery, it is estimated that 40% will. The key task for the clinicians is to determine which need further work-up. Most disorders originate from the retroperitoneum with the kidney accounting for more than half of the lesions. Only 10% will arise from intraperitoneal organs. While most lesions in infants are benign in nature (Fig. 22.1), there is a higher risk of malignancy in older children (Fig. 22.2). The method of determining which abdominal masses need surgical attention involves a thorough history and physical, followed by selected laboratory tests, and focused diagnostic imaging (Fig. 22.3).

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_22,

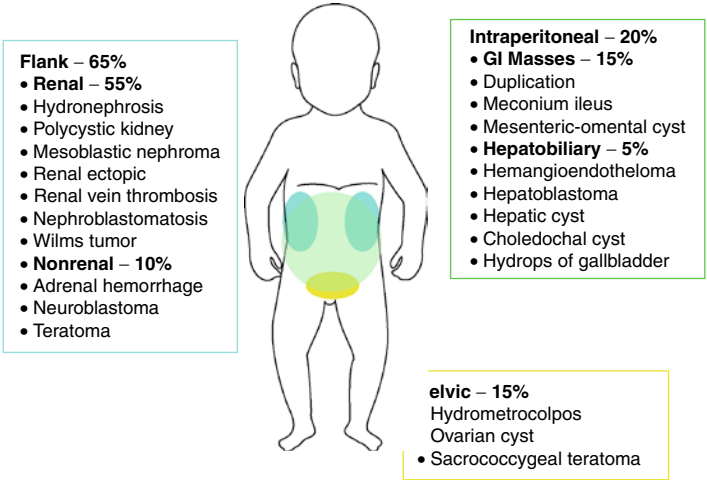


FIGURE 22.1. The common causes of abdominal masses in infants.

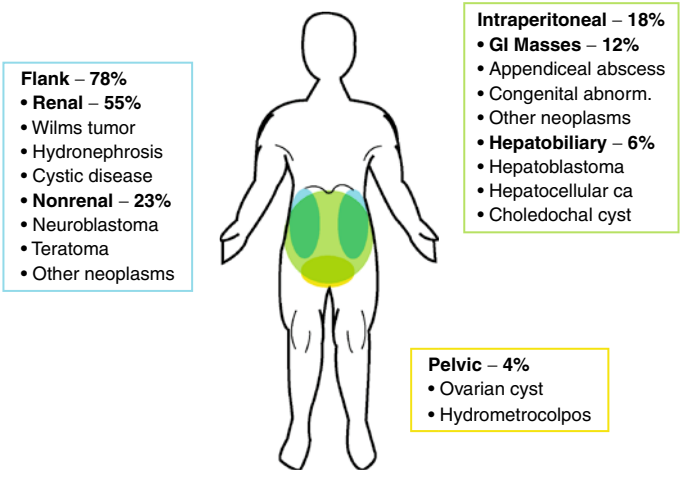
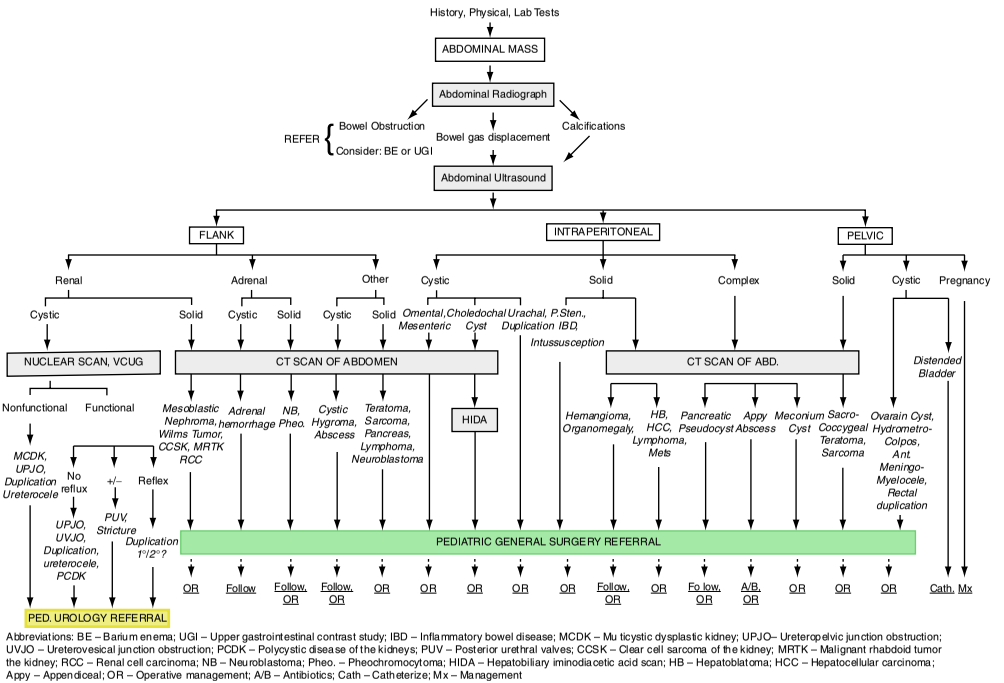


FIGURE 22.2. The common causes of abdominal masses in children and adolescents.



Abbreviations: BE – Barium enema; UGI – Upper gastrointestinal contrast study; IBD – Inflammatory bowel disease; MCDK – Mu tcystic dysplastic kidney; UPJO– Ureteropelvic junction obstruction; UVJO – Ureterovesical junction obstruction; PCDK – Polycystic disease of the kidneys; PUV – Posterior urethral valves; CCSK – Clear cell sarcoma of the kidney; MRTK – Malignant rhabdoid tumor the kidney; RCC – Renal cell carcinoma; NB – Neuroblastoma; Pheo. – Pheochromocytoma; HIDA – Hepatobiliary iminodiacetic acid scan; HB – Hepatoblatoma; HCC – Hepatocellular carcinoma; Appy – Appendiceal; OR – Operative management; A/B – Antibiotics; Cath – Catheterize; Mx – Management

FIGURE 22.3. A flowchart for work-up of abdominal mass in pediatric patients.

22.2 History

Specific questions regarding the mass may help isolate the origin: duration of its presence; associated pain; changes in eating and elimination patterns; and history of trauma. Other aspects of the history are also salient: polyhydramnios may indicate intestinal obstruction; prematurity is associated with hepatoblastoma; a difficult birth is associated with adrenal hemorrhage; syndromes in the family may be associated with tumors (Beckwith-Wiedemann Syndrome (BWS)); Denys-Drash, Wilms/aniridia/GU anomalies/mental retardation (WAGR)); and a review of symptoms may elicit B-symptoms (fevers, night sweats, weight loss) associated with lymphoma.

22.3 Physical Examination

Some tumors secrete hormones that lead to tachycardia and hypertension. Also, particularly large tumors may limit diaphragmatic excursion which can lead to respiratory embarrassment. Such tumors may also obstruct blood flow thus leading to abdominal compartment syndrome. A head-to-toe exam may reveal the disease of origin; overgrowth syndromes such as BWS ("blueberry muffin" lesions) occur when a neuroblastoma metastasizes to the skin; aniridia is a part of WAGR; periorbital lesions such as raccoon eyes and proptosis may also represent NB deposits; upper thoracic chain impingement may lead to Horner's syndrome; tumors with high vascular flow may lead to congestive heart failure; and WT is associated with ambiguous genitalia, hypospadias, and cryptorchidism. The mass itself should be palpated to characterize the location, configuration, size, consistency, mobility, and associated tenderness.

22.4 Laboratory Tests

There are some specific blood tests for pediatric malignancies: vanillylmandelic acid (VMA) and homovanillic acid (HVA) for neuroblastoma; alphafetoprotein (AFP) for

hepatoblastoma, hepatocellular carcinoma, and germ cell tumors; and beta-HCG in pregnancy and for germ cell tumors. Other non-specific blood tests include a complete blood count (CBC) for leukemia and lymphoma; liver function tests (LFTs) for liver masses; BUN/Cr for renal function with renal masses; and amylase/lipase for pancreatic masses.

22.5 Diagnostic Imaging

Diagnostic imaging pinpoints the site of origin and defines tumor characteristics that help identification (invasiveness, heterogeneity, calcifications, cystic spaces, rupture, and metastases). X-rays are good to perform if the mass is thought to be either air-filled or stool-filled loops of bowel. Otherwise, in most situations, an ultrasound should be the first test ordered. It is inexpensive, easy to obtain, and it does not expose the child to radiation or contrast agents. Ultrasound provides very good identification of the likely region of origin (flank, intraperitoneal, or pelvic) and characterizes the mass as either cystic or solid. The constraints of ultrasound include the need for skilled ultrasonographers, limited anatomic detail, and image occlusion by bowel gas, bone, and barium. A computed tomogram (CT) scan provides more detailed characterization of the lesion; helps with staging; reveals anatomic relationships; and provides a prediction of the histological origin (Fig. 22.4). However, a CT should not be the first test of choice because it is more expensive, requires contrast agents, may involve sedation for younger patients, and it exposes the patient to radiation. Despite current interest in using magnetic resonance imaging (MRI) in lieu of CT, its application has been limited due to cost, accessibility, the need for contrast agents, and the need for sedation. Therefore it is usually selected for special situations, specifically biliary imaging (MRCP) and angiograms (MRA). Other tests that may be required to help guide therapy but are seldom required for the initial diagnosis include: Tc-99m sulfur colloid liver-spleen scans for hemangioendothelioma; bone scans to document bony metastases; and Iodine-131-meta-iodobenzylguanidine (MIBG) for detecting the spread of NB (Table 22.1).

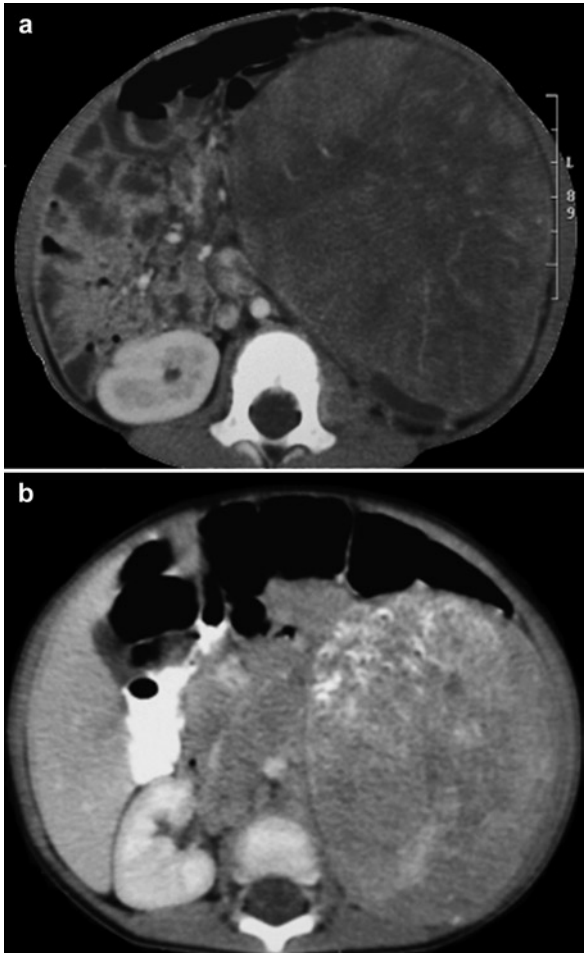


FIGURE 22.4. (**a**) Left-sided neuroblastoma. Note the aorta and inferior vena cava are surrounded by tumor, and are raised off of the retroperitoneum. There are also calcifications within the tumor. (**b**) Left-sided Wilms' tumor. Note the aorta and inferior vena cava are being displaced by the tumor but remain in the retroperitoneum. There are no calcifications within the tumor.

Table 22.1. Common abdominal masses.

Disease	Age range	Location	Type of mass	Best test(s)	Comments
Renal – cystic lesions (Fig. 22.5)					
Multicystic kidney	Infants	Flank	Congenital	US	Usually genetic; mx depends on type
Ureteropelvic jx Obstruction	Infants	Flank	Congenital	US	Pyeloplasty
Ureterovesical jx Obstruction	Infants	Flank	Congenital	US	Reimplantation
Neurogenic bladder	Infants	Flank	Congenital	US	Catheterize; urodynamics
Posterior urethral valves	Infants	Flank	Congenital	US; VCUG	Catheterize; vesicostomy; valvotomy
Renal – solid lesions					
Mesoblastic Nephroma	1st year	Flank	Benign neoplasm	CT	Complete excision
Nephrogenic Rests	Children	Flank	Premalignant	MRI	Multiple = Nephroblastomatosis
Nephroblastoma (Wilms')	3–5 years	Flank	Malignant	CT and US	Stage; May be bilateral

(*continued*)

TABLE 22.1 (continued)

Disease	Age range	Location	Type of mass	Best test(s)	Comments
Clear cell sarcoma	<2 years	Flank	Malignant	CT and US	Bony metastases
Malignant rhabdoid tumor	<2 years	Flank	Malignant	CT and US	Brain lesions; aggressive disease
Renal cell carcinoma	Older	Flank	Malignant	CT and US	Local – nephrectomy; mets fatal
Adrenal gland					
Adrenal hemorrhage	Infants	Flank	Trauma	US	Allow resolution
Neuroblastoma	Young	Flank	Malignant	CT	Stage; consider surgery or chemotherapy
Pheochromocytoma	Older	Flank	Neoplasm	CT	Medical mx; resection after stable
Liver (Fig. 22.6)					
Hepatoblastoma	Young	RUQ	Malignant	CT	AFP; surgery and chemotherapy
Hepatocellular carcinoma	Older	RUQ	Malignant	CT	Often multiple lesions
Hemangioma	Young	RUQ	Benign neoplasm	CT, MRI	Platelets; Kassabach-Merritt possible

Pelvic (Fig. 22.7)					
Ovarian teratoma	Older	Pelvic	Neoplasm	CT	Most benign; solid lesions concerning
Sacrococcygeal teratoma	Infant	Pelvic	Neoplasm	CT	May be benign or malignant
Hydrometrocolpos	Infant	Pelvic	Congenital	US	Error in development; Surgical drainage
Anterior meningomyelocele	Infant	Pelvic	Congenital	CT	Neurosurgical consult
Intraperitoneal					
Appendiceal abscess	Any age	Lower abd.	Inflammatory	CT	Drainage; antibiotics
Choledochal cyst	Any	RUQ	Congenital	US, CT, HIDA	Complete excision; intestinal conduit
Cystic hygroma	Any	Mid abd.	Congenital	CT	Lymphatic
Duplication cyst	Young	Any	Congenital	CT	May occur along any portion of intestine
Inflammatory bowel disease	Children	Any	Inflammatory	CT	Crohn's more likely to cause mass

(*continued*)

Table 22.1 (continued)

Disease	Age range	Location	Type of mass	Best test(s)	Comments
Intussusception	Young	RUQ	Inflammatory	BA enema	Enema is dx and therapeutic
Lymphoma	Older	Any	Malignant	CT	May originate from bowel
Meconium pseudocyst	Infant	Mid abd.	Inflammatory	CT	Due to in utero intestinal perforation
Omental/mesenteric cyst	Any	Mid abd.	Congenital	CT	Failure of lymphatic drainage; excise
Organomegaly	Any	Upper abd.	Multifactorial	CT	Management depends on origin
Pancreatic pseudocyst	Any age	Upper abd.	Inflammatory	CT	Drainage
Pyloric stenosis	Infants	Upper abd.	Congenital	US	Palpable “olive”; pyloromyotomy
Sarcoma	Any	Any	Malignant	CT, MRI	Prognosis depends on histology
Teratoma	Any	Mid abd	Neoplasm	CT	May be benign or malignant; AFP, b-hCG
Urachal cyst	Infants	Lower abd.	Congenital	US	May have urine drainage; excise

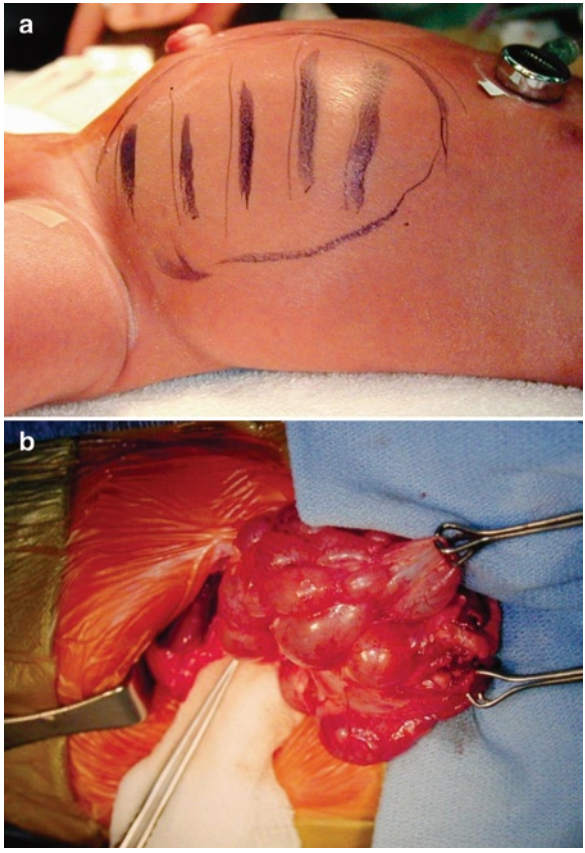


FIGURE 22.5. (**a**) An infant noted with a large left sided mass causing respiratory embarrassment. (**b**) Resection demonstrated a large multicystic dysplastic kidney.

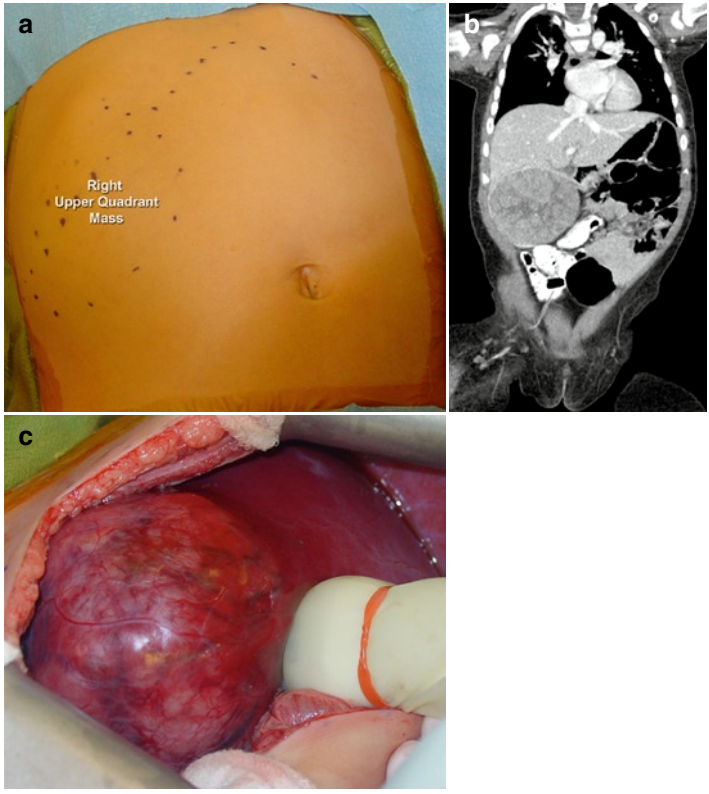


FIGURE 22.6. Six-month-old with (**a**) large right upper quadrant mass, (**b**) arising from the liver on CT scan, and (**c**) resected with intraoperative ultrasound guidance.

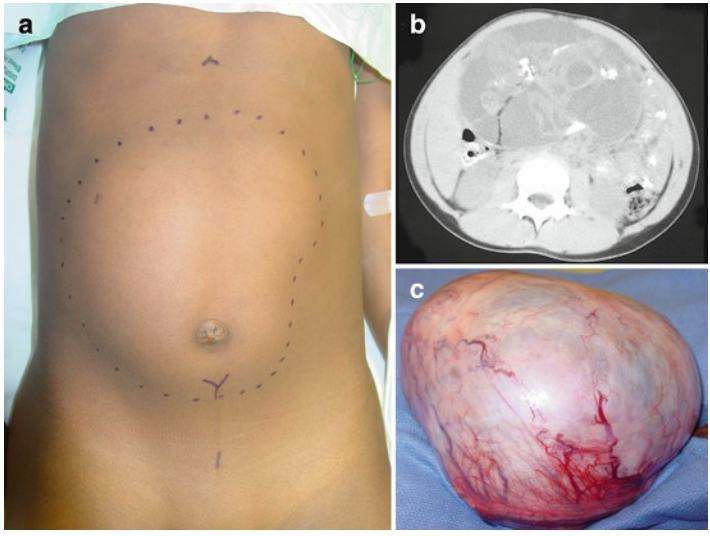


FIGURE 22.7. Sixteen-year-old female with (**a**) a large abdominal mass, (**b**) with a heterogenous appearance on CT scan, (**c**) resected via a laparotomy with a mixed solid and cystic appearance which was determined to be an ovarian teratoma on pathology.

Suggested Readings

1. Golden CB, Feusner JH. Malignant abdominal masses in children: quick guide to evaluation and diagnosis. *Pediatr Clin North Am*. 2002;49(6):1369-1392.
2. Seidel FG. Imaging of a pediatric abdominal mass. *Semin Surg Oncol*. 1986;2(3):125-138.
3. Kaste SC, McCarville MB. Imaging pediatric abdominal tumors. *Semin Roentgenol*. 2008;43(1):50-59.
4. Caty MG, Shamberger RC. Abdominal tumors in infancy and childhood. *Pediatr Clin North Am*. 1993;40(6):1253-1271.
5. Pearl RH, Irish MS, Caty MG, Glick PL. The approach to common abdominal diagnoses in infants and children. Part II. *Pediatr Clin North Am*. 1998;45(6):1287-1326.

Chapter 23
Gastro-Esophageal Reflux Disease

David I. Campbell

Key Points

- Gastro-esophageal reflux disease (GORD) may present in a variety of ways and the presentation may vary according to age.
- Simple GORD may be treated empirically depending on age of the child.
- Troublesome GORD will require investigations such as endoscopy and pH run or a contrast study.
- Severe GORD is oftern associated with cow's milk protein intolerance in children.

23.1 Introduction

Consider infants and young pre-school children as different from older children. The vomiting child is very obvious, but the consequences of reflux esophagitis can be marked with a lesser grade of reflux (GORD without vomiting) (Tables 23.1 and 23.2).

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_23,

Table 23.1. Consider differential diagnoses in suspected GORD with the following components of GORD. Presentation and differential diagnosis of GORD in young children.

Profuse vomiting	3 months pyloric stenosis (dry FTT, inexorable history). UTI Raised intracranial pressure Metabolic diseases
Failure to Thrive	Due to calorie loss (i.e., vomiting, see above) Due to insufficient calorie intake (i.e., feed aversion (see below)). General medical clerking to identify other body systems involved. Always consider Cystic Fibrosis, coeliac in the weaned child and neglect.
Feed aversion	GORD may be co-existent with milk allergy Any cause of feed induced pain (peptic ulcer, small bowel stricture or ulcer, esophagitis)
Feeding problems (Gagging or choking)	Neurological problems such as CP/bulbar or pseudobulbar palsies
Hematemesis	Esophageal varices Peptic ulcer Swallowed blood (pharyngitis or cracked nipples in breast fed babies)
Poor sleep	Raised intracranial pressure
Sandifer Syndrome	Dystonia syndromes, epilepsy (rare situations)
Drooling	Causes of hypotonia
Colic and abdominal pain	UTI, constipation, milk allergy (often co-exists with GORD)
Chest infections	Cystic fibrosis Primary immunodeficiency

Table 23.2. Presentation and differential diagnosis of GORD in the older child/adolescent.

Dyspepsia	Peptic ulcer disease, non-ulcer dyspepsia (gastritis), gallstones (cholecystitis), chronic pancreatitis
Waterbrash	Causes of upper GI dysmotility (coeliac, cholecystitis
Vomiting/regurgitation	Migraine, esophageal stricture General medical clerking to identify other body system disorders
Dysphagia	Esophageal webs, peptic stricture (due to GORD usually), esophageal tumors
Nausea, halitosis	Any cause of vomiting
Laryngitis and dysphonia	Vocal cord palsy (consider Raised intracranial pressure)
Respiratory problems (dry cough)	Asthma Endobronchial infection

23.2 Investigations

Infants with symptoms suggestive of simple gastroesophageal reflux can be empirically treated with feed thickeners (alginate preparations such as Gaviscon®) or formula feeds changed with thickeners pre-added i.e., (Enfamil AR® Mead Johnson Nutrition). It is reasonable to empirically try an antacid (Ranitidine 2 mg/kg tds) and consider Domperidone 0.4 mg/kg tds (although efficacy of latter is poor).

Children with eczema or strong family history of atopy, or with symptoms of fore gut disturbance (GORD) plus midgut disturbance (bloating, colic, iron deficiency) or hind gut disease (diarrhoea) should be considered to have possible cow's milk protein intolerance. How these children should be treated will be dealt with elsewhere, but will include a dairy (or a dairy and soya free diet) together

with the use of an extensively hydrolyzed milk (Pepti® and Peptijuniour® both Danone Baby Nutrition or Nutramigen I or II® Mead Johnson Nutrition). More resistant cases may require an amino acid based formula (Neocate® SHS Nutrition.[1]

Children with troublesome symptoms or failure to thrive should have diagnosis confirmed, but also it should be defined what the cause of reflux is (hiatus hernia, dysmotility due to enteropathy, gastritis, etc.). This should be done by endoscopy and ph study (catheter based as a combined impedance study or wireless Bravo Ph®).

23.3 Treatment

Empirical treatments can be continued if a positive response is seen.

We do not recommend more potent acid suppression (using a proton pump inhibitor) beyond 3 months duration without a confirmed diagnosis (see above). Treatment with any form of PPI beyond 3 months increases the risk of community acquired *Clostridium difficile* colitis, which can present acutely as a fulminating disease.[2]

PPI such as Omeprazole at 0.7 mg/kg od (max dose 20 mg/day) in <2 year olds, or 10 mg od up to 20 kg body weight or 20 mg od for body weight above 20 kg. Higher doses should be used for shorter periods.[3]

23.4 Indications For Pediatric Gastroenterology Referral

Severe disease with FTT or anemia. Requirements for PPI. Confirm diagnosis. More complex food allergy than simple dairy intolerance.

References

1. Vandenplas Y, Brueton M, Dupont C, et al. Guidelines for the diagnosis and management of cow's milk protein allergy in infants. *Arch Dis Child*. 2007;92(10):902-908.
2. Thachil J, Time for a hospital antacid policy on *Clostridium difficile*. BMJ. 2008; 336(7636):109-a.
3. Children BNFf. Proton Pump Inhibitors. In: BNFC, editor. BNFC. London: BMJ Group; 2009:63–64.

Chapter 24
Rectal Bleeding

David I. Campbell

Key Points

- Bleeding from proximal to the duodeno jejunal flexure will give rise to hematemesis and malaena but may give rise to passage of bright red stool in cases of rapid transit.
- Necrotizing enterocolitis is a common cause in premature neonates.
- Dairy allergy, infections such as *Salmonella*, *E. coli*, *Shigella*, intususception are common causes in infants and toddlers.
- Inflammatory bowel disease, rectal polyps, anal fissures are common in older children.
- Treatment depends on underlying etiology.

24.1 General Principles

The following issues need to be considered in the history as they affect diagnosis and hence management:

1. Is this bloody diarrhoea?
2. Is this acute or chronic (acute on chronic)?
3. Are there any features to suggest upper GI bleeding (hematemesis and melaena)?

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_24,

Sites of bleeding proximal to first loops of jejunum likely to have signs of upper GI bleeding, including melaena and hematemesis. Rapid transit would lead to bright red bleeding. This phenomenon is quite common in pediatrics. Causes of upper GI bleeding vary from trivial (swallowed blood in breast fed infant, or nose bleeds to catastrophic hemorrhage from esophageal varices). If in doubt consult with a pediatric gastroenterologist.

Consider the following differential diagnosis in

Older children: inflammatory bowel disease, infection, polyps, fissures.

Fissures: Usually secondary to constipation or idiopathic. If associated with induration, multiple or with skin tags, consider Crohn's disease. Also a feature of child sexual abuse.

24.2 Neonates and Newborn

In newborns and young infants rapid transit can deliver unaltered blood to the terminal large bowel, obscuring the fact that the source of bleeding may be the upper GI tract.

Hemorrhagic disease of the newborn (HDN) is a vitamin K deficiency state, more common in breastfed infants and newborn. Bleeding can be from any site, but is often the GI tract. This condition is rare in babies given supplemental vitamin K at birth. HDN is a cause of hemorrhagic cerebrovascular accidents in young infants.

Necrotizing enterocolitis (NEC) is rare in term babies, but should be considered in any form of rectal bleeding in neonates. NEC is an idiopathic condition; most prevalent in the extremely premature when enteral feeding begins. Translocation of bacteria from gut lumen, vascular thrombosis leads to gut ischemia and infarction with pneumatosis coli. When advanced the abdomen is discolored, the baby shocked and peritonitic. Unless babies are fasted and treated with IV antibiotics, it progresses to intestinal perforation and peritonitis with a high mortality rate. Suspected cases should be referred to a pediatric surgeon.

Newborns with rectal bleeding may have rare diseases such as arterio-venous malformations affecting the GI tract,

these babies usually have cutaneous hemangiomas as well. Most cases of rectal bleeding in new born should be investigated by a pediatric medical team.

24.3 Infants and Young Toddlers

Defining whether the rectal bleeding is part of a bloody diarrheal illness is important.
Causes of bloody diarrhoea:

1. Infection (bacterial: *Salmonella*, *Shigella*, *Campylobacter* and more notably *E. coli*). Usually early on there is a history of fever, pus or mucus are passed in the stool. Stools should be sent for microscopy and culture for three reasons, even if the treatment is conservative. Firstly, for public health reasons, secondly to identify specifically *E. coli 0157* which is a causative agent for hemolytic uremic syndrome (HUS). Children with D+ HUS (i.e., HUS with diarrhoea), usually carry *E. coli 0157* and often need renal support or expert assessment by pediatric nephrologists. The final reason is that treatable causes such as *Entamoeba histolytica*, are diagnosed on microscopy (and can be treated with oral metronidazole).
2. Dairy allergy (allergic enterocolitis). Usually these are not well young, but young infants that appear well that have mucousy, blood stained, loose frequent stools. The child usually is thriving and may be breast feeding. It is now known that microgram quantities of lactalbumin, lactoglobulin and casein proteins from cow's milk are found in human breast milk within a few hours of ingestion.[1] Any form of cow's milk infant formula will produce the same reaction in the susceptible infant. A 2 week dairy free diet using an extensively hydrolyzed formula, or an amino acid based formula, is the gold standard test for dairy allergy.[2]
3. Intussusception. This is a life threatening condition more common from 6 months of age, but can occur at any age (but the etiology is different). Children have usually started weaning. The history is of episodes of crying (screaming) associated with pallor (rather than flushing which is normal).

The child is unwell and the stools are classically described as redcurrant jelly. An abdominal mass on the right hand side is a classical finding, but smaller intussusceptions may not be palpable and an abdominal ultrasound should be performed to identify the intussusception employed to detect the apex of the intussusception. Children must be urgently referred to see a pediatric surgeon.

4. Perianal fissure. If the history is of a child straining to pass a large hard stool and intermittently there may be signs of bright red blood separate to the stool, then there should be a perianal fissure. Treat such children with laxatives.

24.4 Older Children

1. Inflammatory bowel disease. Either UC, Crohn's or indeterminate colitis will cause bloody diarrhoea. In all forms of IBD causing bloody diarrhoea the bleeding will be more prominent if the distal colon is affected. Associated symptoms of colitis/proctitis will be present: urgency, mucus, tenesmus, nocturnal diarrhoea, gastrocolic reflex (passage of stool after eating). Symptoms are chronic (or acute on chronic). Abdominal pain is a prominent feature.
2. Polyps. Painless rectal bleeding is the cardinal features on history. Usually a solitary juvenile polyp is the cause of painless rectal bleeding. Treatment is by therapeutic endoscopy and will require pediatric gastroenterology or surgical assessment. Rarely do forms of familial adenomatous polyposis cause bleeding.
3. Infection: as above
4. Perianal fissures. Treat with laxatives and GTN paste.

24.5 Conclusion

Age of child, presence of signs of upper GI bleeding and whether the bleeding is acute or chronic alter differential diagnosis and hence management.

References

1. Sorva R, Mäkinen-Kiljunen S, Juntunen-Backman K. Beta-lactoglobulin secretion in human milk varies widely after cow's milk ingestion in mothers of infants with cow's milk allergy. *J Allergy Clin Immunol*. 1994;93(4):787-792.
2. Vandenplas Y, Brueton M, Dupont C, et al. Guidelines for the diagnosis and management of cow's milk protein allergy in infants. *Arch Dis Child*. 2007;92(10):902-908.

Chapter 25
Intestinal Obstruction

Travis J. McKenzie and D. Dean Potter

Key Points

- Pediatric intestinal obstructions are divided into neonatal versus childhood.
- Neonatal intestinal obstructions are divided into proximal versus distal.
- Proximal obstructions require an upper contrast study, lower require lower contrast study.
- Bilious emesis or bilious aspirates in a neonate mandates an evaluation for malrotation.
- Pediatric intestinal obstructions frequently require surgical therapy.

25.1 Introduction

Pediatric intestinal obstructions are divided by age group: neonatal versus childhood. Neonatal obstructions are further divided into proximal versus distal obstructions to help guide further evaluation (Table 25.1). In a newborn, any bilious emesis, bilious aspirates, or a gastric aspirate of 20 mL or more is highly suggestive of intestinal obstruction. Importantly, childhood intestinal obstructions are often a surgical disease. As such, any patient suspected of having intestinal obstruction

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_25,

Table 25.1. Etiology of intestinal obstruction.

Neonatal (Proximal)
Rotational anomalies/malrotation
Duodenal atresia
Duodenal stenosis
Hypertrophic pyloric stenosis
Annular pancreas
Antral atresia
Pyloric atresia
Esophageal atresia
Neonatal (Distal)
Hirschsprung's disease
Meconium ileus
Jejunoileal atresia and stenosis
Imperforate anus
Colonic atresia or stenosis
Small left colon syndrome
Omphalomesenteric remnant
Milk curd syndrome
Childhood
Postoperative adhesions
Adynamic ileus
Intussusception
Incarcerated Hernia
Inflammatory Stricture
Pseudo-obstruction
Alimentary tract duplication

should be referred to a medical center with the radiologic and surgical capabilities necessary to care for these patients. This chapter presents some common causes of pediatric intestinal obstruction.

25.2 Neonatal Intestinal Obstruction (Proximal)

1. Malrotation/Volvulus: Presents with feeding intolerance or bilious emesis to cardiovascular collapse secondary to acute volvulus and intestinal ischemia/infarction. Commonly a well baby that presents to emergency department with bilious emesis. Plain abdominal radiograph is often non-diagnostic but findings of proximal obstruction (double bubble) with distal intestinal gas mandates evaluation for malrotation (Fig. 25.1). Diagnosis by upper contrast radiography or emergency laparotomy. Treatment is operative.
2. Duodenal Atresia: Presents within hours of birth with repeated bilious or non-bilious emesis without abdominal distention. Double bubble on abdominal radiograph is classic finding. If delayed repair planned, upper contrast study to evaluate for malrotation with midgut volvulus is advised. Treatment is elective surgical repair after resuscitation.
3. Duodenal Stenosis: Double bubble with distal intestinal gas. If distal gas is present, must rapidly evaluate for rotational anomaly. Older children may present with duodenal web and perforated diaphragm. Upper contrast study to evaluate obstruction is useful. Surgical intervention is commonly required.
4. Hypertrophic Pyloric Stenosis: Progressive non-bilious emesis that becomes projectile often followed by hunger. Significant dehydration with hypochloremic, hypokalemic, metabolic alkalosis. Resuscitate with chloride rich fluids. Abdominal exam may reveal mass in the right upper quadrant (olive). Diagnosis by ultrasound. Treatment is pyloromyotomy after fluid resuscitation.

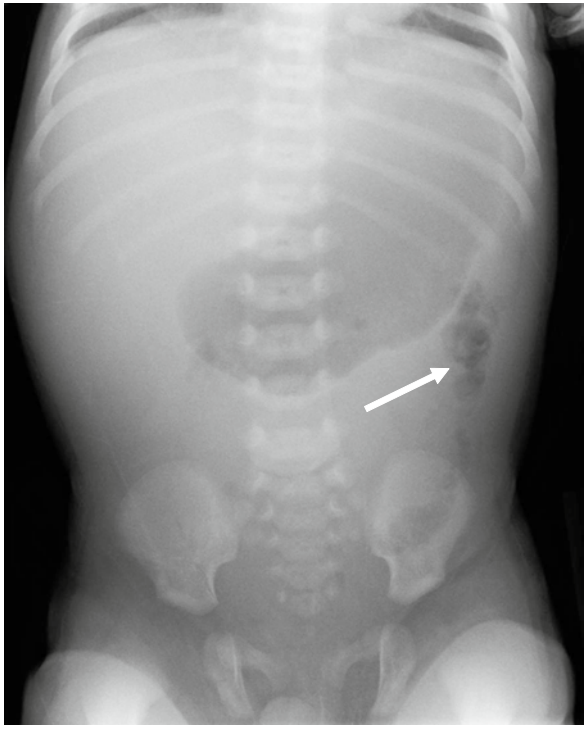


FIGURE 25.1. Abdominal radiograph demonstrating proximal intestinal obstruction with distended stomach and distal intestinal gas (*arrow*). This mandates evaluation for malrotation.

25.3 Neonatal Intestinal Obstruction (Distal)

1. Hirschsprung's Disease: Diagnosis frequently made in neonatal period. Failure to pass meconium in first 24 h of life or constipation requiring suppositories or rectal stimulation is suggestive. Baby may have bilious emesis and abdominal distention. Rectal exam reveals a tight anus and explosive diarrhea. Diagnostic studies include plain radiographs, contrast enema, and rectal biopsy. Bilious emesis mandates upper contrast study for malrotation. Treatment

is surgical and depends on the extent of aganglionosis. Baby may present with sepsis secondary to Hirschsprung's associated enterocolitis. Treat with hydration, intravenous antibiotics including metronidazole and colon irrigations.

2. Meconium Ileus: Associated with Cystic Fibrosis. Neonates are often born with abdominal distention. Uncomplicated form presents with bilious emesis, abdominal distention, and failure to pass stool. Complicated form includes intestinal ischemia/infarction. Plain radiograph shows obstruction with "soap-bubble" or "ground-glass" appearance in the right lower quadrant secondary to inspissated meconium. Contrast enema shows microcolon of disuse. Uncomplicated meconium ileus managed with solubilizing contrast enemas. Need to demonstrate reflux of contrast into dilated loops of bowel. Complicated forms require laparotomy.
3. Jejunoileal Atresia and Stenosis: Patients present early with bilious emesis, abdominal distention, jaundice, or failure to pass meconium. Obstruction may be complete or partial. Plain abdominal radiographs demonstrate many loops of dilated bowel. Contrast enema shows microcolon of disuse. Treatment is surgical repair after resuscitation.
4. Imperforate Anus: Usually present neonatally with abnormal perineal examination. Associated anomalies including vertebral malformations, cardiac anomalies, tracheoesophageal fistula, and genitourinary malformations. Patients should be referred to an appropriate center with pediatric surgical capabilities as early surgical repair may be indicated.

25.4 Childhood Intestinal Obstruction

1. Postoperative Adhesive obstruction: Most common etiology of bowel obstruction. Present with bilious emesis and abdominal distention in the setting of previous surgery. May occur anytime following abdominal operation. Plain

abdominal radiographs with dilated bowel loops with air-fluid levels. Treat with intravenous hydration, gastrointestinal decompression with replacement of lost GI fluid, and delayed surgery if needed. Surgery early if signs of bowel ischemia/infarction.

2. Adynamic Ileus: Present in a similar fashion to those with mechanical obstruction (bilious emesis, abdominal distention) in the setting of other physiologic stress (pneumonia, postoperative, inflammatory conditions). Differentiating ileus from mechanical obstruction can be difficult. Plain radiographs may not differentiate between the two diagnoses. Contrast-enhanced gastrointestinal series may be necessary. Treatment is conservative/non-operative similar to adhesive obstruction.
3. Intussusception: Invagination of one part of the intestine into another. Most common in 3–12 month olds, but can be found in all ages. Severe colicky abdominal pain that lasts only a few minutes. The child will commonly bring up their legs (in infants). Possible abdominal mass and rectal bleeding (current jelly stools). Ultrasonography is the initial diagnostic modality of choice. Treatment is radiologic reduction. Operative intervention for signs of bowel ischemia/infarction or failed radiologic reduction.
4. Incarcerated Inguinal Hernia: Commonly present during the first year of life, more common in males and premature babies. Patient may have a history of hernia or intermittent inguinal bulge. Incarceration results in pain and irritability with progressive obstructive symptoms including bilious emesis and abdominal distention. Late presentation may include peritonitis. Attempt at manual reduction may be made if the patient is stable and without evidence of intestinal compromise. Repair is operative.
5. Inflammatory stricture: Relatively common complication of necrotizing enterocolitis. Presents with feeding intolerance, abdominal distention or persistent diarrhea. Work-up includes an upper and lower contrast study. Treatment is operative.

References

1. Gingalewski C. Other causes of intestinal obstruction. In: Grosfeld J, ed. *Pediatric Surgery*. 6th ed. Philadelphia: Mosby; 2006:1358-1368.
2. Akgur FM, Tanyel FC, Buyukpamukcu N, et al. Adhesive small bowel obstruction in children: The place and predictors of success for conservative treatment. *J Pediatr Surg*. 1991;26:37-41.
3. Andrassy RJ, Mahour GH. Malrotation of the midgut in infants and children. *Arch Surg*. 1981;116:158.
4. Calder FR, Tan S, Kitteringham L, et al. Patterns of management of intussusception outside tertiary centres. *J Pediatr Surg*. 2001;36:312.

Chapter 26
Unique Considerations in the Neonate and Infant: Bile-Stained Vomiting in the Neonate

Robert T. Peters and Sean S. Marven

Key Points

- Bile in vomit or gastric aspirate is green.
- Any child with bilious vomiting/bile-stained aspirates should be referred to a pediatric surgeon urgently.
- Malrotation with midgut volvulus is a life-threatening emergency

26.1 Introduction

It is normal for babies in the first few months of life to bring up small amounts of milk during or following feeds (possetting). As long as they are otherwise well and continue to grow then nothing further need be done.

This chapter concerns bile-stained, i.e., green vomiting in the first month of life. Any neonate with bilious vomiting or bile-stained aspirates should be referred urgently to a pediatric surgeon to rule out an obstructive cause of the vomiting. Over one third of neonates admitted to a pediatric surgical unit with bilious vomiting may have a surgical cause.[1] Bile enters the gut in the second part of the duodenum and obstruction at any point below this level can result in bilious

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_26,

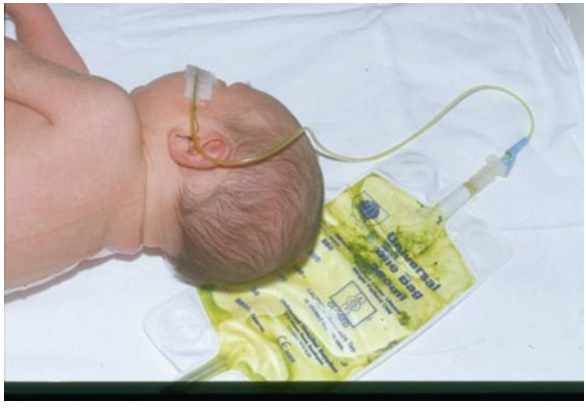


FIGURE 26.1. Neonate with bilious aspirate.

vomiting. It is possible that yellow vomit/aspirate may be seen early in intestinal obstruction but it would normally become green. Obstruction proximal to the second part of the duodenum, e.g., esophageal atresia or pyloric atresia (rare) will therefore not cause bilious vomiting.

Amongst parents and healthcare professionals there is little agreement about the color of bile in the newborn. When shown four shades of yellow and four shades of green as part of a questionnaire study, 56% of parents, 25% of GPs and 10% of midwives did not choose any of the shades of green as a possible match for bile.[2] It is therefore paramount when referred or seeing a neonate with reported vomiting that a clear history is taken from the parents or referring clinician about the actual color of the vomit/aspirate (Fig. 26.1).

26.2 Assessment of the Neonate With Bilious Vomiting

Assessment of the neonate includes a detailed maternal, pregnancy and labor history with a search for clues that may

give the cause for the vomiting. Points from the history include:

History	
Maternal • Medical conditions • Medications • Family history • Consanguinity Pregnancy • Antenatal scans – Timing – Bright or dilated Bowel	Labor • Mode of delivery • Risk factors for sepsis • Condition at birth • Meconium-stained liquor • Green liquor (fetal vomiting) Early neonatal period • Vitamin K given? • Any feeds given/tolerated? • Delayed/failure to pass meconium • Onset of vomiting/color

The examination of the neonate includes an assessment of its general condition with a search for co-existing abnormalities and it may provide an estimate of the level of obstruction if present and a search for co-existing abnormalities:

Examination	
Respiratory condition	External appearance of
Hydration/perfusion	perineum/anus
Jaundice	Sacrum/spine normal?
Dysmorphic features	Hernia?
Degree of abdominal distension	

Initial investigations will be guided by your assessment of the child so far and the clinical setting you are in. The

following should be considered but are by no means a prescriptive list nor exhaustive:

Investigations	
Bedside	Radiological
• Urine dipstix	• Supine plain abdominal x-ray
• Capillary blood gas	• Contrast studies (usually in tertiary pediatric centre
• Bilirubin	– Upper or lower GI contrast
Laboratory	– (Ultrasound)
• Full blood count (↓ platelets NEC/sepsis)	
• Lactate (↑ NEC/ischemia)	
• Biochemistry (note renal function is maternal in first 24 h)	

The purpose of the assessment is to determine the cause of the vomiting and guide initial management. The key features in neonatal bowel obstruction are:

1. Bile-stained vomiting
2. Abdominal distension
3. Failure to pass or delayed passage of meconium

The level of obstruction determines the degree of abdominal distension. A neonate with a high obstruction may have no or a small degree of abdominal distension. They may also pass meconium initially and are likely to have onset of vomiting early. Conversely, a neonate with more distal obstruction will usually not pass meconium and may tolerate feeds initially. Over the first 24 h of life the abdomen will become progressively distended and they will then begin to vomit. At birth there is no air in the gastrointestinal tract but in a normal child it can take as little as 6 h for swallowed air to reach the rectum.

Ninety-five percent of term neonates pass meconium within the first 24 h and almost all by 48 h. Premature babies may have delayed passage of meconium. A neonate born in meconium-stained liquor has passed meconium but beware of green stained liquor. This may represent fetal vomiting of green bile into the amniotic fluid and therefore obstruction.

Neonates who may pass meconium but who may still have bowel obstruction include:

1. Babies with Hirchsprung's may pass meconium, especially after rectal examination
2. Some sticky meconium pellets may be passed in meconium ileus
3. Onset of symptoms in malrotation with volvulus may be delayed for some time after birth[3]
4. A missed anorectal malformation with a fistula

26.3 Initial Management

At this stage the diagnosis may or may not be known but there are certain management steps that are common to all neonates with bilious vomiting:

Initial management	
Pass a nasogastric tube • 8 or 10 Fr in a term baby • Aspirate and leave on free drainage (Septic screen ± lumbar puncture)	Commence iv fluids Commence iv antibiotics • as per local guidance

26.4 Causes of Neonatal Bilious Vomiting

The following is a (non-exhaustive) list of causes of bilious vomiting in the neonate. If a malrotation with volvulus is missed then the consequences, i.e., dead midgut, are disastrous and it is for this reason that any child with bilious vomiting should be referred urgently to a pediatric surgeon.

Causes	
Medical	High obstruction
• Necrotizing enterocolitis	• Malrotation ± volvulus
• Sepsis	• Duodenal atresia/stenosis
– Urinary tract infection	• Proximal jejunal atresia
– Meningitis	
– Group B streptococcus	Low obstruction
• Raised intracranial pressure	• Hirschsprung's disease
• Hypothyroidism	• Ileal/colonic atresia
• Prematurity	• Anorectal malformation
• Idiopathic bilious vomiting	• Meconium ileus
	• Meconium plug syndrome
	• Small left colon syndrome
	• Hernia

26.5 Malrotation With/Without Midgut Volvulus

Malrotation is failure of the gut to complete anti-clockwise rotation resulting in the entire small bowel lying attached by a narrow stalk rather than a broad based mesentery to the posterior abdominal wall. It is therefore prone to twist around on this stalk which compromises the blood supply (superior mesenteric vessels) of most of the small bowel - a midgut volvulus. Up to 80% of cases of malrotation present in the first month of life and most of these in the first week.[4] Therefore a previously apparently normal neonate (or child of any age) can present with a midgut volvulus. This is one of the reasons for urgency of referral

in bilious vomiting. If a midgut volvulus is missed then the entire midgut can become unviable. At best this leads to short gut and lifelong parenteral nutrition, at worst, death. Plain abdominal x-ray may show dilated stomach and proximal duodenum but may also be entirely normal in malrotation. The diagnostic modality of choice is an upper GI contrast study. This demonstrates the duodenal-jejunal flexure lying to the right of the spine and inferior to the duodenal bulb. Treatment involves urgent laparotomy with resection of any dead gut and Ladd's procedure (Figs. 26.2 and 26.3).

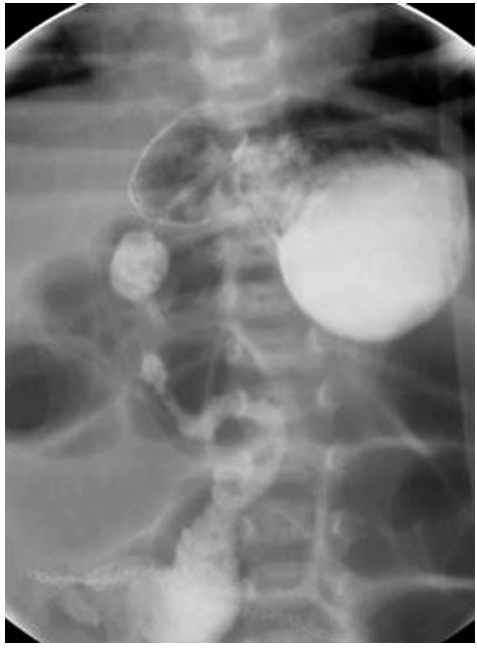


FIGURE 26.2. Upper gastrointestinal contrast study demonstrating malrotation. The duodenal-jejunal flexure lies to the right of the spine.

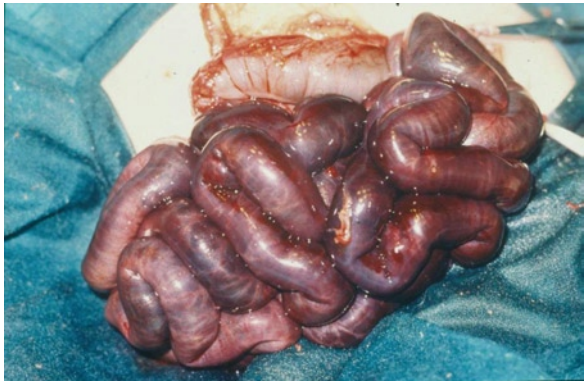


FIGURE 26.3. Dead gut found at operation for malrotation with midgut volvulus.

26.6 Necrotizing Enterocolitis

This is almost exclusively a disease of preterm infants (although it can occur at term) and is therefore uncommon outside of the neonatal unit. Necrotizing enterocolitis (NEC) consists of bowel wall inflammation and ulceration which may proceed to perforation. Neonates present generally unwell with apneas and bradycardias, green vomiting and temperature instability. The abdomen may be distended and shiny or erythematous and there may be rectal bleeding. Abdominal radiograph features include distended bowel loops, intramural gas (*pneumatosis intestinalis*), portal vein gas and pneumoperitoneum. Management is mostly undertaken by the neonatology team and consists of stopping enteral feeds, a nasogastric tube on free drainage and antibiotics. Complicated NEC may require surgical intervention including perforation and stricture formation. Mortality for NEC with perforation remains between 30% and 50%.[5]

26.7 Duodenal Atresia

Traditionally it is reported that a third of infants born with duodenal atresia have Down syndrome.[6] With a decrease in the number of Down children being born, this figure may be as low as 1 in 10. Conversely, 1 in 20 Down infants are born with duodenal atresia.[7] Approximately half of cases are diagnosed antenatally with the appearance of a fluid-filled double-bubble on ultrasound. Over half have associated congenital abnormalities including Down's syndrome, cardiac anomalies, VACTERL, malrotation and other bowel atresias.[8] Vomiting may or may not be green depending on the site and configuration of the abnormality. The typical appearance on plain abdominal film is a double-bubble which represents a dilated, air-filled stomach and duodenum. If there is stenosis causing the obstruction, there may be distal air also present on the plain film. However, in this case it is important to consider that malrotation may be the diagnosis. Surgical treatment involves creating an anastomosis between the proximal and distal duodenum without disturbing the atresic or stenotic segment as the common bile duct commonly enters the bowel here (duodenoduodenostomy) (Fig. 26.4).

26.8 Small Bowel Atresia

Atresias may occur more distally in the jejunum, ileum or rarely in the colon. There may be a single atresia, a large atresic segment like an apple-peel or multiple atresias. The level of the most proximal atresia determines the degree of abdominal distension, the timing of onset of vomiting, and the number of distended bowel loops that will be evident on plain abdominal film. Associated abnormalities include gastroschisis, cystic fibrosis and cardiac anomalies as well as atresias in other parts of the GI tract. Surgical management involves resection of the atresic segment and either primary anastomosis or temporary stoma formation. Extensive

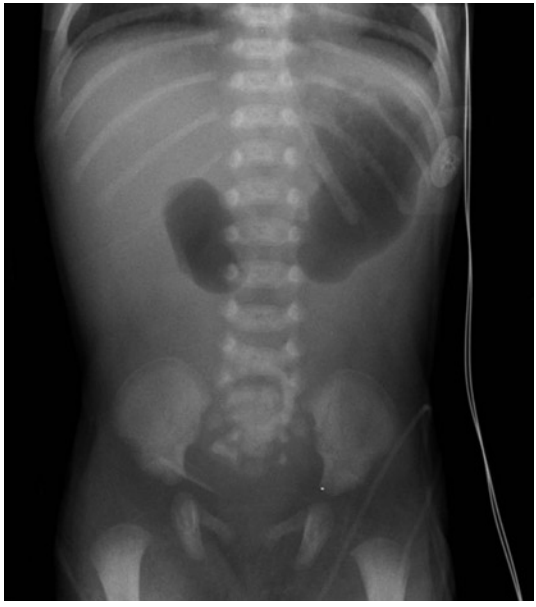


FIGURE 26.4. Plain abdominal film demonstrating typical double-bubble appearance in duodenal atresia with no distal gas.

atresia(s) may leave a child with short gut syndrome (Fig. 26.5).

26.9 Meconium Ileus

Meconium ileus is the presenting feature in up to 1 in 5 children with cystic fibrosis.[9] However, 1 in 5 of neonates presenting with meconium ileus will not have cystic fibrosis.[10] This condition occurs when abnormally thick meconium impacts in and obstructs the distal ileum. Abdominal distension is a key feature and plain abdominal film may show a soap bubble appearance of the meconium. Diagnosis and treatment is with (high osmolar) water-soluble contrast medium. Should this fail then surgical intervention is required.

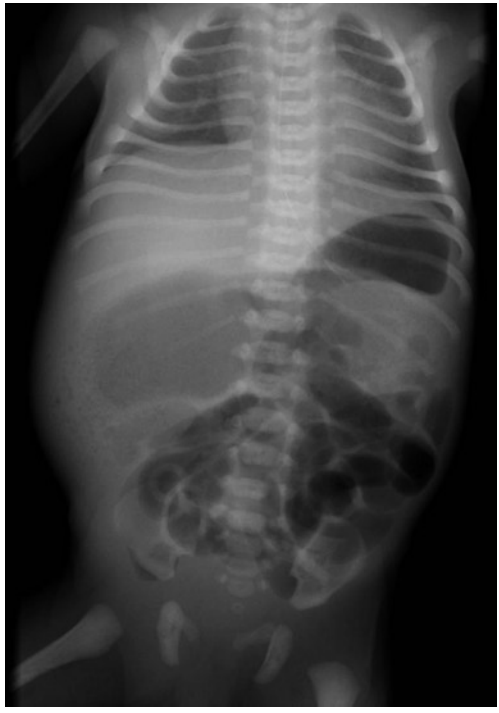


FIGURE 26.5. Plain abdominal film of distal atresia (colonic atresia).

Interestingly, the long term pulmonary outcome of cystic fibrosis children does not differ between those who present with meconium ileus and those who do not.[11]

26.10 Hirschsprung's Disease

Agangliosis of a continuous segment of the large bowel extending for a variable distance proximally from the rectum leads to a functional bowel obstruction. The aganglionic bowel maintains a tonic contraction with dilated normal bowel proximal to this. Delayed passage of meconium or constipation in the first 4 weeks of life should raise the

suspicion of Hirschsprung's disease.[12] Contrast enema may be suggestive but diagnosis is confirmed by rectal suction biopsy demonstrating an absence of ganglion cells and presence of acetyl cholinesterase (AChE)-positive hypertrophic nerve fibers. Initial management involves rectal washouts (proximal to the contracted bowel) to decompress the bowel or stoma formation until definitive procedure(s).

26.11 Anorectal Malformations

Babies with early discharge after birth may go home with an unrecognized anorectal malformation and it is vital that a proper inspection of the perineum and anus occurs before discharge. The passage of meconium does not exclude abnormal anatomy as there may be a fistulous tract with the rectum communicating with the urinary tract in boys, the vestibule or vagina (as a cloaca) in girls or the perineum in either. The finding of an anorectal malformation should trigger a search for associated abnormalities, particularly belonging to the VACTERL association (V-vertebral, A-anorectal, C-cardiac, TE-tracheo-esophageal, R-renal, L-limb). After a thorough pre-operative work-up, initial surgical management involves either a procedure to restore correct anatomy or a defunctioning colostomy.

26.12 Conclusion

Bile-stained vomiting in the neonate may herald intestinal obstruction. Delay in referral or in diagnosis may result in perforation, ischemia or death. Happily, all the anomalies may be corrected with little or no long term effects.

References

1. Godbole P, Stringer MD. Bilious vomiting in the newborn: How often is it pathologic? *J Pediatr Surg*. 2002;37(6):909-911.

2. Walker GM, Neilson A, Young D, Raine PAM. Colour of bile vomiting in intestinal obstruction in the newborn: Questionnaire study. *BMJ*. 2006;332(7554):1363.
3. Jones' Clinical Paediatric Surgery: Diagnosis and Management. 6th ed. Hutson JM, O'Brien M, Woodward AA, Beasley SW, eds. Wiley Blackwell, 2008.
4. Williams H. Green for danger! Intestinal malrotation and volvulus. *Arch Dis Child Educ Pract Ed*. 2007;92(3):ep87-ep91.
5. Henry MCW, Moss RL. Neonatal necrotizing enterocolitis. *Semin Pediatr Surg*. 2008;17(2):98-109.
6. Escobar MA, Ladd AP, Grosfeld JL, et al. Duodenal atresia and stenosis: Long-Term follow-up over 30 years. *J Pediatr Surg*. 2004;39(6):867-871. discussion 867–71.
7. Torfs CP, Christianson RE. Anomalies in down syndrome individuals in a large population-based registry. *Am J Med Genet*. 1998;77(5):431-438.
8. Choudhry MS, Rahman N, Boyd P, Lakhoo K. Duodenal atresia: Associated anomalies, prenatal diagnosis and outcome. *Pediatr Surg Int*. 2009;25(8):727-730.
9. Mushtaq I, Wright VM, Drake DP, Mearns MB, Wood CB. Meconium ileus secondary to cystic fibrosis. The east london experience. *Pediatr Surg Int*. 1998;13(5-6):365-369.
10. Fakhoury K, Durie PR, Levison H, Canny GJ. Meconium ileus in the absence of cystic fibrosis. *Arch Dis Child*. 1992;67(10 Spec No): 1204-1206.
11. Kappler M, Feilcke M, Schröter C, Kraxner A, Griese M. Long-Term pulmonary outcome after meconium ileus in cystic fibrosis. *Pediatr Pulmonol*. 2009;44(12):1201-1206.
12. Ghosh A, Griffiths DM. Rectal biopsy in the investigation of constipation. *Arch Dis Child*. 1998;79(3):266-268.

Chapter 27
Unique Considerations in the Neonate and Infant: Pyloric Stenosis

Joseph Ignatius Curry and Sinead Hassett

Key Points

- Common cause of non bilious vomiting in infancy
- Occurs due to hypertrophy of the pyloric muscle resulting in partial gastric outlet obstruction.
- The typical biochemical picture is a hypochloremic, hypokalemic metabolic alkalosis.
- Treatment is surgical with extra mucosal splitting of the pyloric muscle.

27.1 Introduction

Pyloric stenosis is a common pediatric surgical condition with an incidence of 2–4 per 1,000 live births.[1] It affects males four times more than females. The etiology of this condition is unclear. It is postulated to occur due to failure of relaxation of the pyloric muscle secondary to abnormal innervation. The prolonged contraction causes hypertrophy of the muscle fibers and development of a pyloric mass. There is a definite genetic component to the condition. 1 in 20 of male offspring and 1 in 50 of female offspring from a male parent with pyloric stenosis will be affected.[2]

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_27,

27.2 Presentation

The diagnosis of pyloric stenosis can often be elicited from the history. It typically presents as non bilious projectile vomiting after feeding in infants between 2–8 weeks of age. In the initial stages of the condition the babies are well and hungry to feed. As the condition progresses they become dehydrated and demonstrate weight loss. Common conditions mimicking pyloric stenosis are gastro esophageal reflux (GOR) and over feeding. In gastro esophageal reflux while the vomiting may be projectile, the dehydration and weight loss noted in the later stage of pyloric stenosis does not occur. A careful feeding history is helpful to identify if the vomiting is from over feeding or feeding too concentrate a formula. The other important point to elicit is a family history of pyloric stenosis.

Examination of a child with suspected pyloric stenosis should begin with an assessment of their hydration status. Increased capillary refill time, sunken fontanelle, loss of skin turgor and reduced alertness are all signs of dehydration which can occur secondary to persistent vomiting of feeds. The classical abdominal signs in a child with pyloric stenosis are the presence of reverse gastric peristalsis on inspection of the abdomen and palpation of an olive shaped mass in the right hypochondrium. Palpation of the pyloric mass requires time and patience. The baby needs to completely relax their abdominal musculature. This can be facilitated by giving the baby a feed and flexing the hips during the examination. Palpation from the left hand side is also helpful as this helps to avoid the caudate lobe of the liver, which can sometimes be mistaken for the pyloric mass.

27.3 Investigations

If pyloric stenosis is suspected from the history, blood should be taken for blood gas and biochemical analysis. Pyloric stenosis results in a hypochloremic hypokalemic metabolic

alkalosis. This occurs due to vomiting of gastric fluid rich in hydrogen and chloride ions. Sodium ions are reabsorbed in the renal tubule, to maintain intravascular volume, in exchange for potassium ions. High bicarbonate levels are present due to reabsorption along with sodium ions. Some potassium may be retained in exchange for hydrogen ions resulting in paradoxical aciduria. In the initial stages of the condition the biochemical abnormalities may not be present as the body compensates by producing a more alkaline urine.

Unconjugated hyerbilirubinema is present in 2% of patients due to a deficiency of hepatic gluconryl transferase which resolves post operatively.[3]

While history and clinical examination are often sufficient to confirm the diagnosis of pyloric stenosis, a pyloric ultrasound has become part of the work up for a vomiting baby in many pediatric surgical units. The signs of pyloric stenosis on an ultrasound are a thickened pyloric channel of >3 mm, an elongated pyloric channel length of >15 mm and failure of relaxation of the pyloric canal (Fig. 27.1). Pyloric ultrasound performed by an experienced observer has a sensitivity and specificity approaching 100%.[4] UGI contrast has now been superceded by ultrasound as the radiological investigation of choice where the diagnosis of pyloric stenosis is suspected.

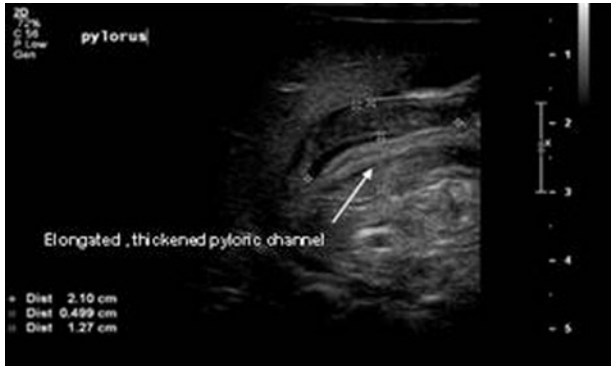


Figure 27.1. Ultrasound appearance of pyloric stenosis.

27.4 Management

The mainstay of treatment in pyloric stenosis is correction of electrolyte abnormalities prior to definitive surgical treatment. Intravenous fluids (0.45% normal saline and 5% dextrose) should run at one and a half times maintenance with supplemental potassium added to correct the hypokalemia. An NG tube should be passed, aspirated regularly and losses replaced with intravenous 0.9% normal saline. The baby should be fasted until surgery. Venous blood gases and electrolytes should be measured approximately every 8 h until they have normalized. Failure to correct electrolyte abnormalities prior to surgery can lead to myocardial dysfunction following anesthetic induction and respiratory depression post operatively.[5]

Surgical techniques in the management of pyloric stenosis have evolved since the modern day operation was first described by Ramstedt in 1912. While the main aim of the surgery, i.e., extra mucosal splitting of the pyloric muscle, has not changed, the approach has modified. The classic approach is a right upper quadrant incision to gain access to the pylorus. In the 1980s a circumumbilical incision was described to deliver the pylorus.[6] This technique grew in popularity due to the excellent cosmetic result post operatively. The first laparoscopic pyloromyotomy (Fig. 27.2) was reported in 1991 and has now been adopted by many centers as the technique of choice.[7] A large multi centre randomized control trial has recently demonstrated a shorter hospital stay and quicker time to full enteral feeds using the laparoscopic approach over the circumumbilical approach.[8]

Feeding can begin approximately 6 h post surgery with a gradual build up of feeds as tolerated. Vomiting is common in the first 24 h post surgery and parents should be reassured. Children are discharged once full feeds are tolerated which is usually on the second post operative day.

Overall morbidity rates from pyloromyotomy are approximately 10%.[9] Complications include intra operative mucosal perforation, wound infection, wound dehiscence and inadequate

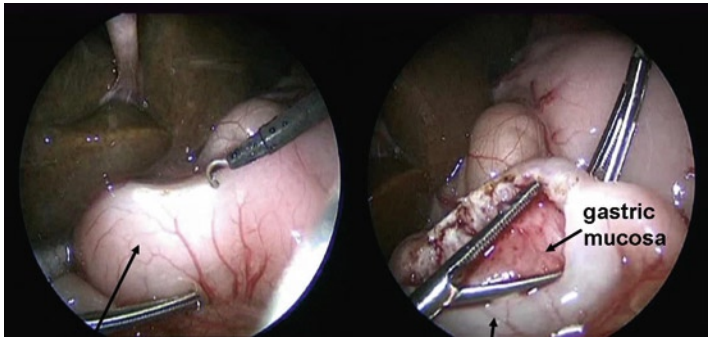


FIGURE 27.2. Laparoscopic view of pyloric mass and pyloromyotomy.

splitting of the pyloric muscle resulting in prolonged vomiting and recurrence of pyloric stenosis. Mortality from pyloric stenosis is rare and results from an unrecognized mucosal perforation and subsequent leak of gastric contents.

Medical therapy using IV and oral atropine to treat pyloric stenosis has not gained widespread acceptance due to the longer length of hospital stay required and the 15% failure rate associated with the treatment.

Long term follow up of babies with pyloric stenosis demonstrate no significant side effects or symptoms from this condition.

References

1. To T, Wajja A, Wales PW, et al. Population demographic indicators associated with incidence of pyloric stenosis. *Arch Pediatr Adolesc Med*. 2005;159:520-525.
2. Carter CO, Evans KA. Inheritance of congenital pyloric stenosis. *J Med Genet*. 1969;6:233-254.
3. Woolley MM, Felsher BF, Asch J, et al. Jaundice, hypertrophic pyloric stenosis, and hepatic glucuronyl transferase. *J Pediatr Surg*. 1974;9: 359-363.
4. Hernanz-Schulman M, Sells LL, Ambrosino MM, et al. Hypertrophic pyloric stenosis in the infant without a palpable olive: accuracy of sonographic diagnosis. *Radiology*. 1994;193:771-776.

5. Steven IM, Allen TH, Sweeney DB. Congenital hypertrophic pyloric stenosis: the anaesthetist's view. *Anaesth Intensive Care*. 1973;1: 544-546.
6. Tan KC, Bianchi A. Circumumbilical incision for pyloromyotomy. *Br J Surg*. 1986;73:399.
7. Alain JL, Grousseau D, Terrier G. Extramucosal pyloromyotomy by laparoscopy. *Surg Endosc*. 1991;5:174-175.
8. Hall NJ, Pacilli M, Eaton S, et al. Recovery after open versus laparoscopic pyloromyotomy for pyloric stenosis: a double-blind multicentre randomised controlled trial. *Lancet*. 2009;373:390-398.
9. Hulka F, Harrison MW, Campbell TJ, et al. Complications of pyloromyotomy for infantile hypertrophic pyloric stenosis. *Am J Surg*. 1997;173:450-452.

Chapter 28
Unique Considerations in the Neonate and Infant: Intussusception

Joseph Ignatius Curry and Sinead Hassett

Key Points

- Peak incidence between 5 and 9 months of age.
- Triad of abdominal colic, vomiting and bleeding PR in 30% of children.
- Delayed diagnosis common due to subtle signs and symptoms.

28.1 Introduction

Intussusception occurs where there is invagination of bowel into its neighboring distal segment (Fig. 28.1a and b) As bowel invagination progresses, its mesentery becomes incorporated and venous outflow is obstructed. Engorgement and swelling of the bowel follows which obstructs the arterial supply with ischemia leading to necrosis. The etiology of this invagination is thought to be due to hyperplastic intestinal Peyer's patches which act as a lead point. In many cases this hyperplasia occurs secondary to viral infection, typically following an upper respiratory tract infection o r gastroenteritis.

An identifiable lead point is found in only 10% of cases of intussusception. A Meckels diverticulum is the commonest reported lead point.[1] Other examples are the appendix, small bowel lymphoma, intestinal polyps and following submucosal

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_28,

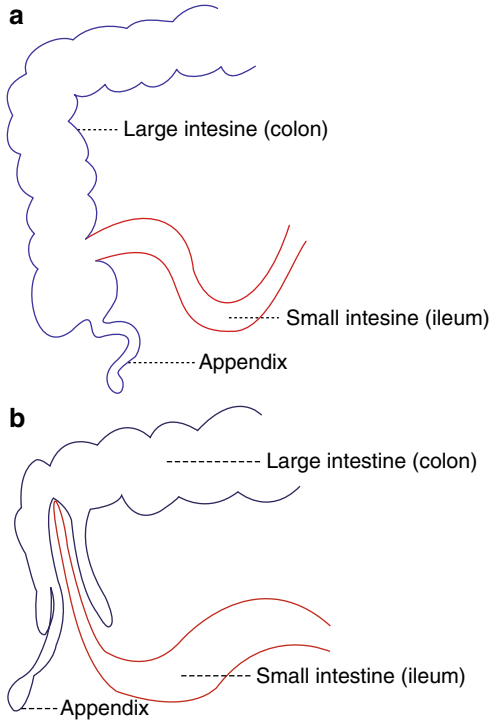


FIGURE 28.1. (**a**) normal ileo-caecal region, (**b**) appearance during intussusception.

intestinal hemorrhage in children with Henoch–Schonlein purpura. 80% of intussusceptions involve a portion of distal ileum invaginating into the colon (ileocolic). Ileoileal, caecocolic, colicocolic and jejenojejunal intussusceptions are less common. The incidence if intussusception is approximately 2 per 1,000 children and is the commonest cause of bowel obstruction in children under 5.[2]

28.2 Presentation

While intussusception can occur at any age the incidence is greatest between 5 and 9 months. The winter months are the commonest time of presentation due to the higher rate of

viral respiratory tract infections. The typical history is of a child experiencing episodes of severe abdominal colic with drawing up of their legs and non billous vomiting. In between these episodes of colic they are quieter than normal and often falls asleep. As the intussusception progresses the child passes a mixture of blood and mucus per rectum, the so called "red currant jelly" stool. If the condition remains untreated there is progression to small bowel obstruction indicated by billous vomiting and worsening lethargy secondary to dehydration.

The classical triad of abdominal colic, vomiting and passage of a red currant jelly stool is present in only 30% of children. The most common symptom is colicky abdominal pain while atypical presentations include diarrhoea, lethargy, irritability or simply a reluctance to feed.[3] There may be a history of a pyrexial illness a few days prior to the development of symptoms.

Examination of a child with a suspected intussusception should focus on their hydration status. If symptoms have been present for longer than 24 h it is likely that signs of dehydration such as increased capillary refill and tachycardia will be present.

In 30% of children a sausage shaped mass will be palpable in the right upper quadrant. Abdominal tenderness or guarding indicates the presence of bowel ischemia, necrosis or perforation. A rectal examination is useful to determine the presence of blood if there is no history of passage of a bloody stool. Rarely an intussusception may be noted protruding from the rectum.

28.3 Investigations

If a diagnosis of intussusception is suspected IV access is obtained, full blood count and biochemical analysis performed and a cross match taken. Fluids are run at maintenance rate. If the child is dehydrated 20 mL/kg boluses of 0.9% normal saline should be administered as appropriate. They should not be moved to the radiology department for imaging until their clinical status improves. Mortality in

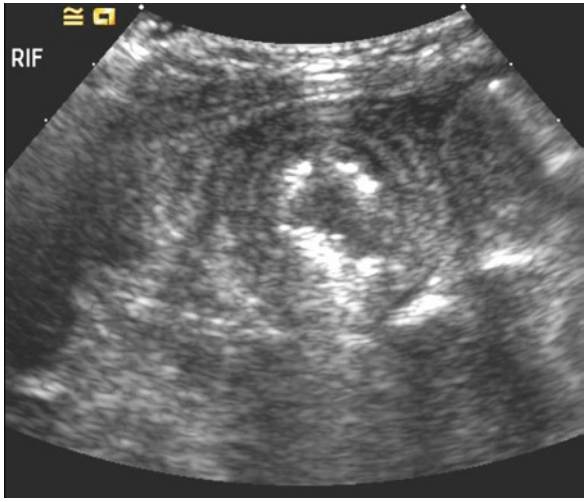


Figure 28.2. Ultrasound picture of intussusception.

intussusception can be due to hypovolemic shock secondary to inadequate fluid resuscitation. If there is significant abdominal distension an NG tube is passed. A dose of broad spectrum antibiotics should be given.

The gold standard in the diagnosis of intussusception is an abdominal ultrasound. In experienced hands the sensitivity and specificity approaches 100%. The appearances of an intussusception on ultrasound are a target or donut sign (Fig. 28.2). This pattern represents the walls of the intussusception. There is little role for an abdominal x-ray in the diagnosis of intussusception. The sensitivity of an x-ray is 45% with a false negative rate of 20%.[4] While an x-ray may show the presence of free air indicating a perforation, this will often be apparent on examination.

28.4 Management

Once the diagnosis of intussusception is made the treatment is reduction by air enema. Contraindications are the

presence of peritonitis, indicating bowel necrosis or perforation. A catheter is inserted into the child's rectum and air is introduced under measured pressure control. Fluoroscopy is performed to track the progress of the reduction. The rule of threes is employed, namely three effective attempts at air reduction lasting 3 min each. If the intussusception does not reduce with this protocol further attempts at reduction are unlikely to be successful. The success rates with air enema reduction are approximately 70–80% and are often related to the duration of symptoms. Reasons for failure of intussusception reduction are the presence of ischemic or necrotic bowel, an identifiable lead point or technical failure related to inability to generate significant intra luminal pressure. The perforation rate with air enema reduction is 0.8%.[5] Ultrasound guided air and water reduction of intussusception has been reported although no direct comparison has been made with the fluoroscopic technique.

28.5 Surgical Management

Open reduction of an intussusception is through a right illac fossa incision. Gentle pressure is applied to the distal portion of bowel to squeeze out the invaginated proximal portion. If significant ischemia or frank necrosis is present, reduction should not be attempted and resection and primary anastamosis is indicated. Occasionally a negative laparotomy is performed as the intussusception has spontaneously reduced in the interval between the failed air enema reduction and surgery. In view of this laparoscopy prior to open surgery has been advocated (Fig. 28.3). The results of laparoscopic reduction of intussusceptions are less convincing with success 70% in comparison to 100% with the open technique.[6]

There is a 10% recurrence rate with intussusception, usually within a year of the first episode, and is more likely to be associated with a pathological lead point.

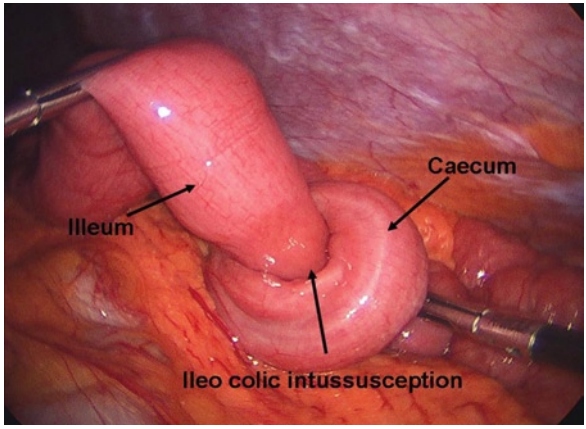


Figure 28.3. Laparoscopic view of ileocolic intussusception.

References

1. Kaiser AD, Applegate KE, Ladd AP. Current success in the treatment of intussusception in children. *Surgery*. 2007;142:469-475. discussion 475–477.
2. Huppertz HI, Soriano-Gabarro M, Grimprel E, et al. Intussusception among young children in Europe. *Pediatr Infect Dis J*. 2006;25: S22-S29.
3. O'Ryan M, Lucero Y, Pena A, et al. Two year review of intestinal intussusception in six large public hospitals of Santiago, Chile. *Pediatr Infect Dis J*. 2003;22:717-721.
4. Best evidence topic reports. Bet 4. Role of plain abdominal radiograph in the diagnosis of intussusception. Emerg Med J. 2008;25: 106-107.
5. Daneman A, Navarro O. Intussusception. Part 2: An update on the evolution of management. *Pediatr Radiol*. 2004;34:97-108. quiz 187.
6. Bonnard A, Demarche M, Dimitriu C, et al. Indications for laparoscopy in the management of intussusception: A multicenter retrospective study conducted by the French Study Group for Pediatric Laparoscopy (GECI). *J Pediatr Surg*. 2008;43:1249-1253.

Chapter 29
Hemangioma and Lymphangioma

Roly Squire

Key Points

- The vast majority of hemangiomas involute spontaneously and do not need treatment.
- Early treatment for hemangiomas can be required for: obstruction of the airway, oropharynx, vision, hearing; ulceration causing infection; pain; persistent bleeding. Initial treatment is with oral propranalol or prednisalone.
- Lymphatic malformations are generally only of cosmetic importance, but their natural history is unpredictable, and they seldom resolve spontaneously.
- Venous malformations generally only need treatment for symptoms, mostly pain with activity but occasionally phlebitis.
- Vascular malformations are best treated in collaboration with an interventional vascular radiologist

29.1 Introduction

Vascular anomalies are common, and the majority present in childhood. The terminology has been confusing, but there is now a consensus classification, which distinguishes vascular

P.P. Godbole et al. (eds.), *Guide to Pediatric Urology and Surgery in Clinical Practice*, DOI: 10.1007/978-1-84996-366-4_29,

tumors, such as hemangiomas, from vascular malformations, which can be capillary, lymphatic, venous or arterial. Most vascular anomalies are of cosmetic importance only, but symptoms can be both distressing and dangerous, and choice of treatment depends upon accurate diagnosis and careful assessment of the extent of the abnormality. This is often not apparent on simple clinical examination.

29.2 Types of Vascular Anomalies

1. Hemangioma: this is a common congenital abnormality, occurring in up to 5% of infants. It may only show as a small red mark at birth, but proliferates over the first two weeks of life to produce a firm, red, raised benign tumor (Fig. 29.1), often referred to as a strawberry naevus.

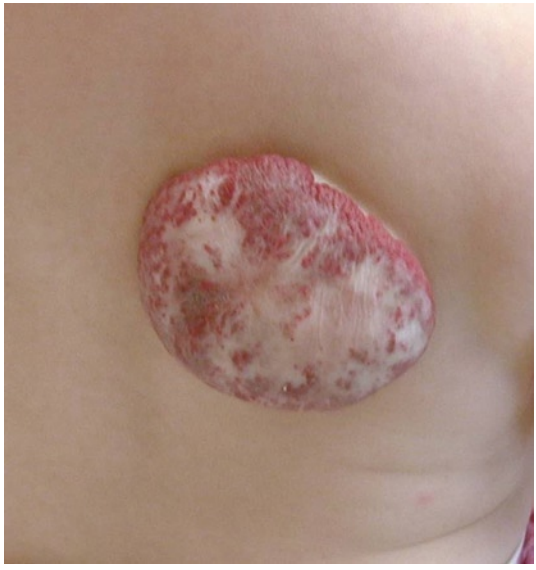


FIGURE 29.1. A large hemangioma on the back, showing early signs of involution, but also some scarring from healed ulceration.

Proliferation continues variably during the first year of life, but generally a hemangioma remains asymptomatic, simply a cosmetic abnormality, with a favorable natural history: 50% will involute by age 5, and in more than 90% involution will mean that there is no need for intervention, though in up to 40% a faint blemish will always be visible.

Early treatment can be required for hemangiomas that occur at sites that interfere with normal bodily functions, such as peri-orbital or oropharyngeal lesions. Complex lesions around the mouth can extend into the pharynx or airway. Treatment can also be required for complications such as ulceration and infection, which lead to pain and scarring. Bleeding can be a nuisance, but is not dangerous, and can be controlled by simple pressure. Some large hemangiomas can have prominent arteriolar feeding vessels, but are still likely to spontaneously involute. The high flow component is only important in the very rare situation where there are signs of a hyperdynamic circulation causing heart failure or venticulomegaly.

Rarely hemangiomas are fully developed at the time of birth (congenital hemangioma), and only some of these will show the same tendency to involute as the classic infantile hemangioma.

2. Lymphangioma: a benign soft tissue tumor which can occur practically anywhere in the body. Typical sites are the neck, axilla or groin, but lymphatic malformations of the trunk or limbs are also regularly encountered. Lymphangioma can be present at birth, and 70% have presented by 1 year of age. However an occult lymphangioma can suddenly present at any age, even in an adult, as new soft tissue swelling appearing over a few hours in association with increased lymphatic flow. This would usually be as a result of an intercurrent infection, or an acute inflammatory process in the same lymphatic drainage area as the anomaly. Lymphangiomas can be macrocystic, often soft and flaccid making the diagnosis quite straightforward, or microcystic, in which case they can be firm and ill defined. Microcystic or mixed lesions can be difficult to distinguish clinically

from other soft tissue tumors, including malignant tumors as well as other vascular anomalies. The history of rapid onset and/or fluctuation in size in association with viral infections is often diagnostic. The natural history of lymphatic malformations is unpredictable; although spontaneous resolution is rare some become less prominent with time, yet others enlarge unexpectedly.

3. Lymphangioma circumscriptum: the cutaneous form of lymphangioma, presenting as small blisters in the skin overlying a subcutaneous lymphatic swelling (Fig. 29.2), nearly always on the trunk. The blisters are often painful and can sometimes leak clear fluid. They can occur as part of the initial presentation of a lymphatic anomaly, but are more commonly a complication of attempted resection.
4. Venous (low flow) malformations: these may present as a visible varicosity or a soft tissue mass. Reduction of the swelling with pressure can be diagnostic. Unlike hemangiomas they do not tend to proliferate in the first few months, but also do not involute with time. Some venous malformations may not have a significant cosmetic impact, but may have an occult component that causes symptoms

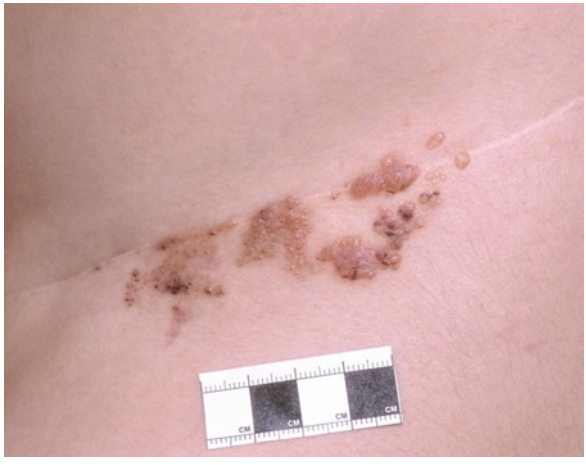


FIGURE 29.2. Lymphangioma circumscriptum.

as the child becomes older. Common symptoms are pain or swelling with activity, and rarely phlebitis. Malformations extending into joints can be a particular problem. Venous malformations can also be complicated by phleboliths, calcified thrombi sitting within the abnormal veins, presumably caused by turbulent flow.

5. Arterial (high flow) malformations: often called arteriovenous malformations (AVMs) these are rare in children, and are more likely to be congenital than post-traumatic. Heart failure and tissue overgrowth are the main complications in childhood, whereas adult AVMs tend to cause pain.
6. Kaposiform hemangioendothelioma: this rare form of vascular tumor is distinct from congenital or infant hemangioma because of its characteristic brawniness and infiltration (Fig. 29.3). Recognition is important because it can be complicated by platelet sequestration, resulting in profound and life-threatening thrombocytopenia, known as Kasabach Merritt Syndrome (KMS).

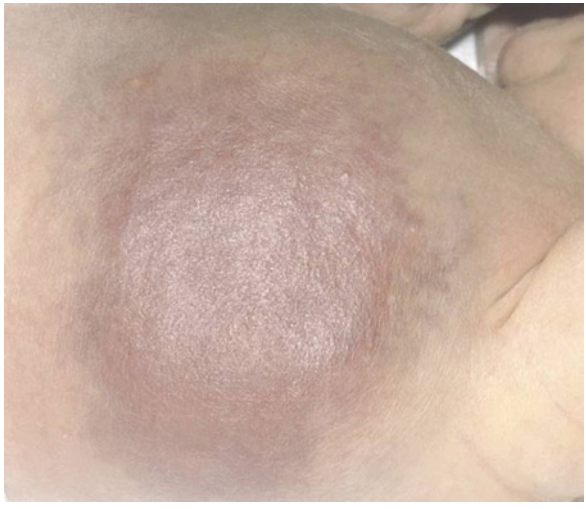


FIGURE 29.3. Kaposiform hemangioendothelioma (complicated by profound thrombocytopenia).

29.3 Investigation of Vascular Anomalies

Cutaneous hemangiomas seldom require any investigation, but for other soft tissue swellings Doppler ultrasound is generally the best initial imaging modality, to establish a diagnosis and to assess the extent of the lesion. MR scan provides additional information and is particularly important in delineating the margins of an infiltrative lesion, identifying the proximity or involvement of adjacent structures, and assessing resectability. However children under 6 years of age usually require general anesthesia or sedation for satisfactory MR imaging, so it is recommended that an MR scan should only be requested when it will impact upon management decisions. Generally contrast MR imaging, using Gadolinium, is recommended for investigating vascular anomalies.

Childhood malignant soft tissue tumors can be very vascular, and so if the diagnosis remains unclear from the history, examination and imaging, biopsy should be considered. If a hemangioma is biopsied it can be useful to look for expression of GLUT-1, an endothelial marker for infant hemangiomas that are likely to involute spontaneously.

Interventional radiologists will sometimes require angiography to identify the presence of vessels suitable for sclerotherapy or embolization, but since these procedures also generally require general anesthesia in children it is often best to carry out angiography as a preliminary component of a treatment episode, under a single anesthetic. Aspiration under anesthesia can be a useful ultimate diagnostic procedure, with a view to injecting the sclerosant appropriate for the diagnosis through the same needle.

29.4 Treatment of Vascular Anomalies

1. Hemangioma: Because of the favorable natural history of hemangiomas treatment is generally not required. Parents can be reassured that the lesion has a high likelihood of spontaneous resolution, generally leaving only a minor

visible abnormality, which will not have any significant cosmetic impact. Despite this, early treatment can be required for specific sites, such as periorbital lesions, hemangiomas that become ulcerated and painful, or sites which are prone to persistent trauma resulting in bleeding. The treatment strategy starts with simple options, which are easy to administer and have a low potential for side-effects, with more complicated treatments being reserved for symptomatic and unresponsive lesions. The most straightforward options are oral propranalol (2 mg/kg daily) or oral prednisalone (2–4 mg/kg daily), which can be prescribed in primary care. A more immediate response might be required for lesions such as a hemangioma obstructing vision, where intralesional steroid injection is usually rapidly effective. Infected hemangiomas are very painful, and so as well as treating the underlying abnormality it is important to give adequate analgesia, targeted antimicrobials, and non-adherent dressings. Some hemangiomas are easily amenable to excision, and this can be carried out early if the lesion is causing symptoms. Further treatment escalation is seldom required, and is discussed below for treatment of kaposiform hemangioendothelioma.

It is wise to wait until the child develops self-awareness before considering treatment for cosmesis. It is unusual for a child to develop anxiety or embarrassment about an unsightly vascular anomaly until age 4 or 5, by which time most hemangiomas will be showing signs of involution. Late options include surgery or laser therapy; laser can be effective to treat persistent discoloration, but has a minimal effect on residual swelling.

2. Lymphangioma: Most lymphangiomas are asymptomatic and of minor cosmetic importance, and so treatment is not required. If treatment is requested, usually because the swelling is unsightly, surgery is the traditional approach, but over the last two decades there has been a steady move towards injection sclerotherapy. A number of sclerosants have been reported to be effective, including OK432, bleomycin, tetracycline, doxycycline or even ethanol. It seems likely on

current evidence that the choice of sclerosant is less important than patient selection- sclerotherapy works most effectively on macrocystic lesions, often unconvincingly on mixed lesions, and barely at all on microcystic lesions. Sclerotherapy may need multiple treatment episodes, usually requiring general anesthesia. Sclerotherapy causes pain and swelling in the early postoperative period, and so should be avoided in lesions where this may result in compromise of the airway. However sclerotherapy does reduce the risk of complications associated with surgical excision/debulking, namely unsightly scarring, damage to adjacent structures, recurrent lymphangioma and lymphangioma circumscriptum. Macrocystic lesions are the most amenable to both surgery and sclerotherapy; the real difficulty is how to treat unsightly microcystic lesions. Surgical debulking may be the only option, especially if the airway is compromised, but results are seldom satisfactory, and so new developments are awaited. Lymphangiomas may also develop a bacterial infection, requiring urgent treatment using antibiotics with a spectrum that covers the most common soft tissue infection organisms, such as staphylococcus and streptococcus.

3. Lymphangioma circumscriptum: Individual blisters are usually transient, lasting for a few weeks before spontaneous resolution, but laser therapy can be effective treatment for lesions causing distress.
4. Venous malformations: Treatment of phleboliths is usually straightforward- they can be removed from within the lesion with a small incision, using an intraoral approach for lesions in the cheek. Some venous malformations can be excised, though many, particularly intramuscular lesions, are unresectable. They are amenable to sclerotherapy which is best carried out using radiological guidance, and so referral to an interventional vascular radiologist is advised. Recurrence is common.
5. Arteriovenous (high flow) malformations: high flow lesions causing overgrowth, heart failure or pain require treatment. Embolization is usually recommended to occlude the feeding vessels, but unfortunately recurrence is almost inevitable, so to avoid the need for repeated treatments excision should be considered in the early post-embolization period.

6. Kaposiform hemangioendothelioma: These lesions generally need urgent treatment not just because of the cosmetic and functional impact, but also because of the profound thrombocytopenia (KMS). Platelet counts below 25×10^3/mL are associated with spontaneous hemorrhage, intracranial bleeding being of particular concern in small infants. Platelet transfusions should be avoided, and so initially immediate treatment with propranalol and steroids should be commenced. If no response is seen within 14 days treatment should be escalated. Because of the potential neurological sequelae of interferon-α, cytotoxic chemotherapy using intravenous vincristine is now generally accepted as being the first choice for escalation therapy. Meanwhile MRI +/– angiography may demonstrate options for interventional radiology, and as a last resort kaposiform hemangioepithelioma usually responds to low-dose radiotherapy.

29.5 Indications for Referral

1. Any vascular anomaly that is interfering with breathing, feeding, vision, hearing.
2. Hemangioma complicated by ulceration, infection or persistent bleeding.
3. Lymphangioma causing concern regarding cosmesis.
4. Venous malformation causing symptoms.
5. Uncertain diagnosis, particularly if possibility of malignant tumor.

Suggested Readings

1. Adams DM, Lucky AW. Cervicofacial vascular anomalies. I. Haemangiomas and other benign vascular tumours. *Sem Pediatr Surg*. 2006;15:124-132.
2. Elluru RG, Azizkhan RG. Cervicofacial vascular anomalies II. Vascular malformations. *Sem Pediatr Surg*. 2006;15:133-139.
3. Okazaki T, Iwatani S, Yanai T, et al. Treatment of lymphangioma in children: our experience of 128 cases. *J Pediatr Surg*. 2007;42: 386-389.

4. Mulliken JB, Glowacki J. Haemangiomas and vascular malformations in infants and children: A classification based on endothelial characteristics. *Plast Reconstr Surg*. 1982;69:412-422.
5. Manunza F, Syed S, Laguda B, et al. Propranalol for complicated infantile haemangiomas: a case series of 30 infants. *Br J Dermatol*. 2010;162:466-467.

Index