ABC of urology
Common paediatric problems

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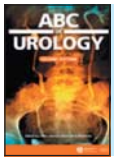
Many paediatric problems can present to medical professionals. This article covers some of the common ones including phimosis, undescended testis, retractile testis, vesicoureteric junction reflex, hypospadias, neonatal hydronephrosis, obstruction of the pelviureteric junction, and some types of tumour.



This article is adapted from the second edition of the ABC of urology (Blackwell Publishing), available from all good medical bookshops, including www.hammicksbma.com

Phimosis

Phimosis is the most common reason for circumcision, although recurrent balanitis is also an indication. Circumcision may also be performed for religious or social reasons.

At birth, adhesions are present between the glans penis and foreskin, but separation begins to occur immediately and continues thereafter. The prepuce normally becomes retractile after the age of two years, but many adolescent boys retain some adhesions. Preputial adhesions are a common reason for referral to a urologist, but adhesions are normal and should be treated only if "physiological phimosis" persists into adolescence and causes problems with masturbation or sexual intercourse. A non-retractile foreskin is free of symptoms and self limiting, and circumcision is not needed. Parents often say that the prepuce "balloons" when the child urinates, but this is a sign of a non-retractile foreskin rather than phimosis. Careful examination will show that the urethral meatus is visible through the narrowed preputial opening, and, with time, this opening widens to allow the foreskin to retract normally. True or "pathological phimosis" is rare, but it may cause appreciable problems in childhood or adolescence. Treatment is usually circumcision, whereas alternative treatments are preputioplasty or application of steroid creams.

Indications for circumcision in patients with phimosis

Recurrent infection under foreskin
Appreciable restriction of urine flow

Undescended testis

The incidence of undescended testis ranges from 3.4% to 5.8% in full term boys but decreases to 0.8% in boys aged about one year. Why testes fail to descend into the scrotum is unclear, but recent evidence suggests that descent occurs in two distinct phases and that androgens may have an important role, possibly acting via the genitofemoral nerve. An undescended testis can be classified by its location in the upper scrotum, superficial inguinal pouch, inguinal canal, or abdomen. In 80% of cases, the undescended testis will be palpable in the inguinal canal. Patients with undescended testes have two major concerns: increased incidence of testicular cancer and heightened risk of subfertility.

For treatment purposes, the main distinction that needs to be made is whether the testis is palpable. If the testis is palpable in the inguinal canal, an orchidopexy should be carried out. The correct timing of orchidopexy has been debated. Spontaneous descent of undescended testis is rare after the age of one year.

Every attempt should be made to locate an impalpable testis. Ultrasound, computed tomography, and magnetic resonance imaging have been used, but laparoscopy is the current investigation of choice. If blind ending spermatic vessels are noted, further evaluation is not needed; the patient and parents should be counselled and hormonal replacement and a testicular prosthesis may be needed. If the testis is

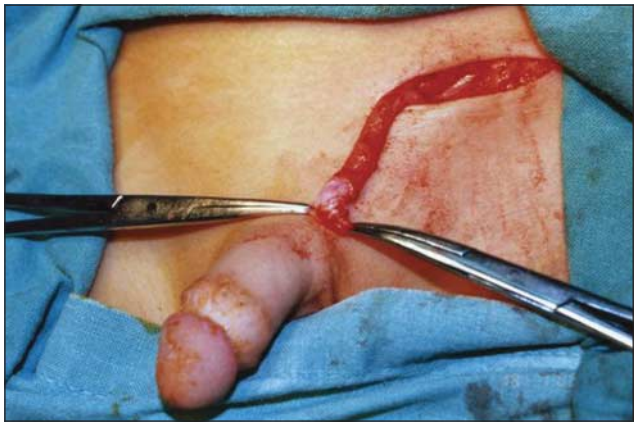
Sites of undescended testis

- Inguinal canal (80%)
- Intra-abdominal (19%)
- Other (1%):
  —Suprapubic
  —Femoral
  —Perineal
  —Contralateral scrotum

Main findings on laparascopy to locate an impalpable testis

- Blind ending spermatic vessels above internal inguinal ring but no testis
- Intra-abdominal testis
- Cord structures that enter internal ring

Fertility of an undescended testis becomes compromised after the age of two years



Inguinal exploration of undecended testis

intra-abdominal in a prepubertal child, orchidopexy should be performed as soon as possible. If an intra-abdominal testis is detected after puberty, orchidectomy should be performed, as the testis is incapable of spermatogenesis and the risk of malignancy is up to 10 times higher than in a normal testis. If the cord structures enter the internal ring, inguinal exploration is warranted. In boys with bilateral undescended testis in whom neither testis is palpable, chromosomal and endocrine evaluation is needed.

Retractile testis

Retractile testis is common in general practice and is often confused with undescended testis. The key to distinguishing a retractile testis from an undescended testis is to show that the testis can be delivered into the scrotum. A retractile testis will stay in the scrotum after the cremaster muscle has been overstretched, whereas a low undescended testis will immediately pop back to its undescended position after being released. If any doubt exists, the child should be seen in follow-up for a repeat examination. If doubt exists as to whether the testis is retractile or undescended, referral for a urological opinion should be arranged.

Vesicoureteric junction reflux

Reflux stops spontaneously in a large proportion of patients, although the degree of resolution is inversely proportional to the severity of the reflux. For children with reflux of grades 1-2, antibiotic prophylaxis is the recommended initial treatment. In all children with reflux of grades 3-5 and those with persistent reflux despite a trial of observation on prophylactic antibiotics, surgical correction is recommended. Dysfunctional voiding as a result of bladder instability should be treated with anticholinergic agents.

In the neonatal period, reflux is likely to be the result of anatomical abnormalities; the incidence of reflux is equal in the sexes. In later childhood, the condition predominantly occurs in girls with voiding disturbances. Much evidence shows that reflux should not be considered in isolation and that dysfunctional voiding has a large role in the development of symptoms.

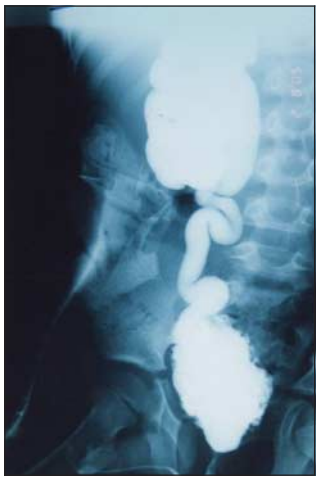
A vicious cycle of symptoms may also exist, because reflux may lead to infection, which itself may lead to bladder instability, dysfunctional voiding, and further reflux. These three elements thus should be considered equally in the treatment of reflux. Reflux alone does not to lead to renal damage—infection must also be present. Many urologists believe that renal damage occurs early in the natural course of the disease, and in many cases it is not progressive.

Recent advances in management of reflux

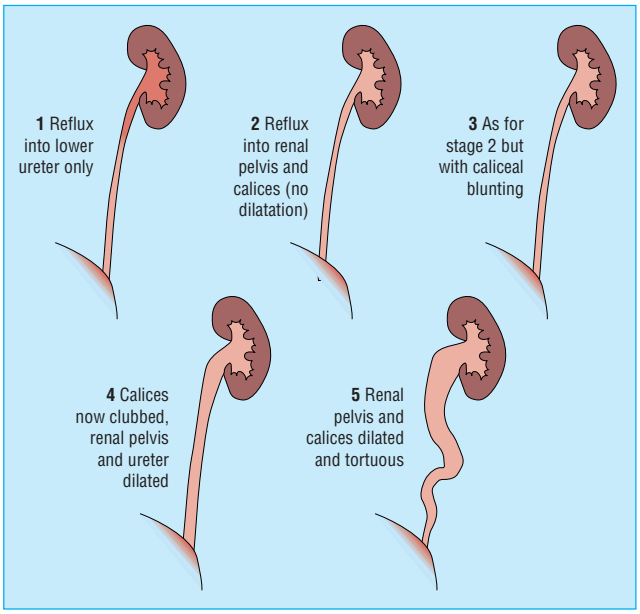
- Past treatment for reflux centred on ureteric reimplantation
- Recently, endoscopic injection of tetrafluoroethylene polymer (Teflon) into the submucosa of the ureter has been used with some success
- Concern about the risks of migration of particles of tetrafluoroethylene polymer has prevented universal acceptance of the technique
- Other agents, such as bovine crosslinked collagen, autologous chondrocytes, dextranomer plus hyaluronic acid copolymer and polydimethylsiloxane have been suggested for injection
- Success rates vary between 65% and 90%

Hypospadias

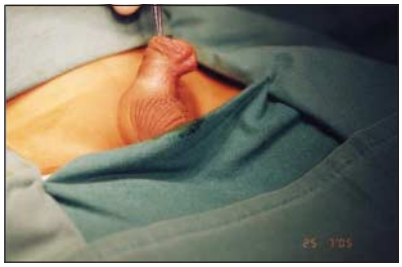
Hypospadias is a congenital condition that affects three in 1000 male infants and results in underdevelopment of the urethra.



Right sided vesicoureteric junction reflux with gross dydronephrosis



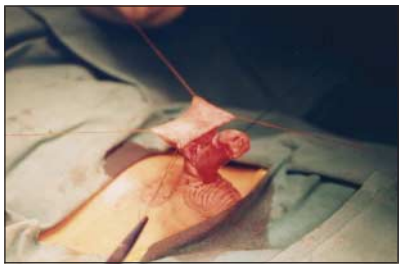
Grading of vesicoureteric reflux



Mid-penile hypospadias

Mid-penile hypospadias–urethral opening calibrated

The penis may be deviated by chordee, and the urethral opening may be situated anywhere from the perineum to the glans on the ventral surface (in contrast to epispadias in which the opening is on the dorsal surface). The child should be referred for urological assessment and surgical treatment. The ideal age for surgery is 6-12 months.



Hypospadias surgery—transverse preputial island flap

Neonatal hydronephrosis

The use of routine ultrasound examination in pregnancy has identified a number of fetuses with hydronephrosis. Postnatal evaluation and management depend on the severity and laterality of hydronephrosis

Fetal urinary tract anomalies are common; they occur in 0.2-0.9% of all pregnancies. Hydronephrosis accounts for more than 50% of these anomalies. Antenatal hydronephrosis may be caused by ureteropelvic junction obstruction, ureterovesical junction obstruction, multicystic kidney, primary obstructive megaureter, vesicoureteral reflux, or posterior urethral valves.

In cases of mild unilateral hydronephrosis (< 15 mm in diameter) with normal appearing renal parenchyma, further prenatal follow-up is seldom useful, and surgery is unnecessary. A postnatal check is important to confirm the hydronephrosis has resolved.

In cases of moderate unilateral hydronephrosis (15-19 mm), ultrasound and a micturating cystogram should be performed at two months and subsequently at intervals of six months. Surgery is unlikely to be needed in these cases.

In cases of severe unilateral hydronephrosis (> 20 mm), ultrasound, a micturating cystogram, and an isotopic renal scan should be performed at one month. Severe unilateral hydronephrosis is eventually most likely to need surgery.

In neonates with severe bilateral hydronephrosis, ultrasonography and a micturating cystogram should be performed within a week. Early surgery is often indicated.

Presentation of pelviureteric obstruction

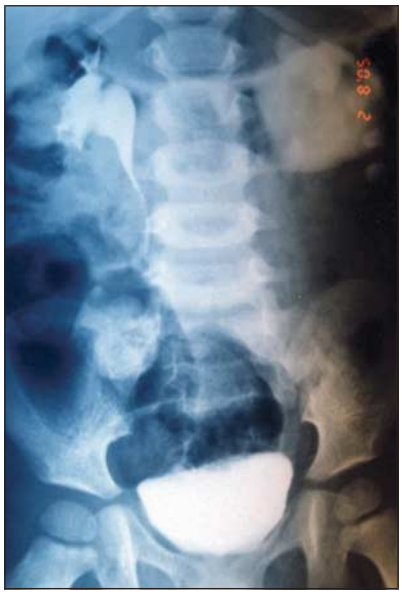
Obstruction of the pelviureteric junction may occur at any time (before birth, in childhood, or in adulthood)
Infants typically present with an abdominal mass
Older children may have abdominal pain
The condition often presents with haematuria after fairly minor abdominal trauma

Obstruction of pelviureteric junction

The essential defect seems to be an aperistaltic segment of ureter, from which the normal musculature is congenitally absent. The role of "aberrant" vessels in causing obstruction recently has been questioned. These vessels are usually normal variants, often pass behind the ureter, and are not generally thought to cause obstruction.

Obstruction of the pelviureteric junction is usually diagnosed by intravenous urography, which shows delay in appearance of contrast on the affected side and dilated renal pelvis and calices. The ureter, when seen, is usually not dilated. Differential renal function and confirmation of obstruction should be obtained with isotope renography.

Surgery is indicated for obstructive symptoms, stone formation, recurrent urinary infection, or progressive renal impairment. Pyeloplasty is the treatment of choice, but if the affected kidney possesses < 10% of total renal function, nephrectomy should be performed. Minimally invasive alternative techniques include antegrade endopyelotomy and laparoscopic pyeloplasty. Laparoscopic pyeloplasty is becoming the treatment of choice, and open procedures usually are reserved for patients in whom laparoscopic surgery is contraindicated.



Congenital obstruction of left pelviureteric junction

Common paediatric tumours

Wilms' tumour

15% of children with Wilms' tumour have congenital abnormalities, including musculoskeletal and other genitourinary anomalies (4.4%)
Bilateral disease is seen in 5-7% of children with Wilms' tumour

Wilms' tumour

Wilms' tumour (nephroblastoma) is the most common primary malignant renal tumour of childhood. It typically affects young children (median age 3.5 years), with more than 80% of the patients being identified before the age of five years. The most common presentation of Wilms' tumour is an abdominal mass,

although haematuria is the presenting feature in up to 15% of cases. Wilms' tumour is usually diagnosed with ultrasonography, computed tomography, or magnetic resonance imaging.

Wilms' tumour is treated by radical nephrectomy; chemotherapy is usually given after surgery, with the exact protocol depending on the stage of the disease. Radiotherapy is needed only if residual tumour has been left behind at surgery and for patients with lymphatic and pulmonary metastases. Neoadjuvant chemotherapy is beneficial for patients with bilateral involvement.

Renal cell carcinoma

This tumour is rare in children and is not usually diagnosed until confirmed by histological examination of a presumed Wilms' tumour. Some tumours are chemosensitive, and radiotherapy may be needed for microscopic residual disease, but radical nephrectomy remains the mainstay of treatment.

Rhabdomyosarcoma

This sarcoma commonly presents with lower urinary tract symptoms, particularly haematuria or urinary retention. Tumours of the vagina may cause a foul vaginal discharge, and pelvic tumours may cause a large mass.

Rhabdomyosarcoma is treated effectively with chemotherapy. The role of radical surgery is diminishing and currently is reserved for children who fail to respond to chemotherapy or who develop a pelvic relapse.

Further reading

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The ABC of urology is edited by Hugh N Whitfield, consultant urological surgeon, Royal Berkshire Hospital, Reading, and Chris Dawson, consultant urologist, Edith Cavell Hospital, Peterborough. The book will be published in autumn 2006.

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Retrospective blues: Robert Johnson—an open letter to Eric Clapton

Let us hope the medical profession can continue to learn from all the other worlds that surround it, and which it ultimately serves. Blues aficionados among you may have enjoyed Eric Clapton's *Sessions for Robert J*—a personal tribute to Robert Johnson, "the most important blues musician who ever lived."

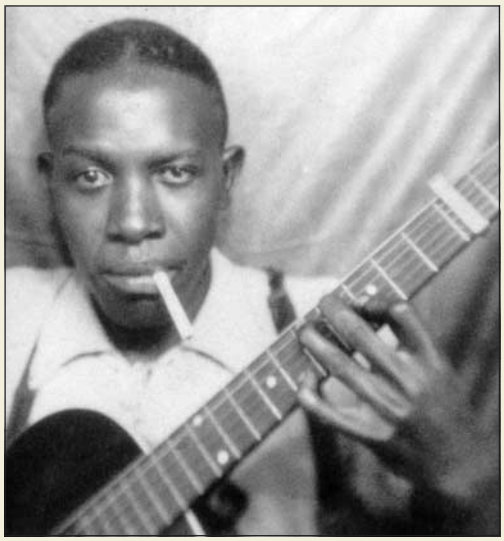
Session IV of the material featured marvellous acoustic playing by Clapton, emulating his favourite influence, and also his enlightening commentary regarding the difficulty of reproducing the material. The scene was carefully set up with the long familiar (to those of you in the know) photograph of Johnson in the background with his unnaturally long fingers clutching his guitar neck.

It dawned on me then that perhaps Johnson's guitar playing gift related to unfair advantage from this arachnodactyly. Eric pay heed.

So what else do I know? Closer inspection of his face revealed a slight left ptosis, and he was known to have "one bad eye"—a small cataract afflicted him from time to time, but later disappeared. Dislocated?

The film played on, recounting some events surrounding Johnson's premature death, at the early age of 27 years. What was the cause of his death way back then in the 1930s? His death certificate was displayed, revealing the cause of death as "No doctor." Legend has it that he was poisoned by strychnine-laced whisky or succumbed to syphilis, but neither rang true. One account from the evening of his demise described him "on his hands and knees howling and barking like a dog." Dissecting?

So it seems possible that this tall, slim, loose jointed guitar wizard with a history of "resolving cataract" and unexplained sudden death had Marfan's syndrome.



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So what does looking back teach us? Now coming up to 20 years in general practice, I can reflect that in my personal experience I have seen Marfan's syndrome on four occasions, most recently, sadly, in retrospect, after the sudden premature death of one of our young patients.

And then it dawned on me; that in my 20 years of practice, to my knowledge I have seen only one death due to aortic dissection—on a cold dark winter night many years ago, "on her hands and knees howling like a dog."

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