Testicular problems in children

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Abstract

Testicular problems in children can be either congenital or acquired. These problems are often difficult to diagnose and carry significant sequelae if untreated. Early surgical consultation is often needed for correction of the problem. This article reviews the pathophysiology of the most common paediatric testicular abnormalities with emphasis on the diagnostic modalities employed and discusses the current treatment choices.

Keywords acquired anomalies; congenital; cryptorchidism; epididymal cyst; epididymo-orchitis; testis; torsion; tumours; varicocele

Introduction

The diagnosis and treatment of many paediatric testicular abnormalities can be challenging and frustrating. Moreover, testicular problems, if not evaluated and treated in a timely manner, can have significant sequelae. This article focuses on the diagnosis, evaluation, indications for referral and treatment for some of the more common paediatric testicular problems that are encountered in paediatric practice.

Congenital causes of testicular problems

Undescended testis

An undescended testis (cryptorchidism) occurs when normal testicular descent is arrested along its normal path (Figure 1). Undescended testis occurs in about 1% of term male infants at 1 year of age and in 20% of cases undescended testes are impalpable.

Aetiology: the testis develops from the bipotential gonad at 6 weeks' gestation under the influence of the SRY gene. Sertoli cells and Leydig cells secrete Mullerian inhibiting substance (MIS) and testosterone respectively. Secreted MIS causes regression of the Mullerian structures. Testicular descent occurs as a result of a complex interaction of several factors namely testosterone, calcitonin G-related peptide, insulin the gubernaculum, the processus vaginalis and intra-abdominal pressure. Failure of any of these mechanisms may cause testicular non-descent or maldescent. When a testicle is not palpable, this may represent either an intra-abdominal testicle or vanishing testicle syndrome (testicular agenesis). A retractile testicle occurs as a result of a hyperactive cremasteric muscle contraction.

Diagnosis: clinical examination is key to establishing the correct diagnosis. It is important to differentiate between the true undescended testicle and the retractile testicle. In cases of retractile testicle, the testicle can easily be brought to the base of the scrotum and stays there without tension. Stimulating the cremasteric reflex by stroking the inner aspect of the thigh can induce the testis to move back up along the normal line of descent. A typical history suggestive of a retractile testis is one where the testicle is easily seen and felt in the scrotum when the boy is relaxed and in a warm bath. A careful search should be made for an ectopic testicle in the superficial inguinal pouch, perineum, medial aspect of the thigh or even base of the penis. In cases of bilateral impalpable testis, one must consider the possibility of an intersex condition, and the karyotype and hormonal profile should be characterized. If a unilateral undescended testicle is associated with a proximal hypospadias, an intersex condition should be considered and appropriate evaluation carried out, usually in conjunction with the paediatric endocrinologist.

Treatment: Huff and colleagues have characterized changes that take place within the testis in the first few years of life. An undescended testis undergoes structural changes within the first two years of life and may potentially affect fertility. However more recent evidence suggests that changes may occur much earlier within the first 6–12 months (John Hutson, BAPU consensus session September 2011). Furthermore it is more prone to trauma and torsion. The relative risk of malignancy in an undescended testis is 3.7–7.5 fold that of a normally descended testicle. Orchidopexy does not reduce this risk but makes the testis more amenable for self-examination.

A unilateral undescended palpable testis should be observed for the first 3 months of life to take advantage of the testosterone surge that may aid further descent. Current consensus is that if the testicle remains undescended after 3 months of age, it is unlikely to spontaneously descend and hence surgical intervention is warranted. Hutson and colleagues recommend surgery after 3 months of age in centres with facilities to carry out such procedures at this age (BAPU consensus session September 2011). In most specialist centres, surgery should be carried out at around 6 months under magnification where appropriate.

Laparoscopy is the treatment of choice for the unilateral impalpable testis. All patients are examined under a general anaesthetic and, if the testicle is palpable, an open orchidopexy is performed. If the testis is still impalpable, a diagnostic laparoscopy is performed. If the testicle is of good quality, it may be brought down by either a single-step or two-step orchidopexy according to the staged Fowler Stephen principle. There is very little role for groin exploration in isolation for an impalpable testicle. If the laparoscopy indicates blind-ending gonadal vessels and vas deferens in conjunction with an impalpable testis, the patient is declared to have vanishing testis syndrome and no further action is necessary. If the laparoscopy indicates viable gonadal vessels and vas exiting an open internal ring, the groin should be explored to confirm the presence or absence of viable testicular tissue. In cases where vas and vessels are seen to exit a closed internal ring, the author explores the groin and removes the testicular remnant which on histology shows no viable testicular tissue.

Outcome: boys with undescended testis may have diminished fertility potential in adulthood. Boys with unilateral undescended

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Conflict of interest: none declared.

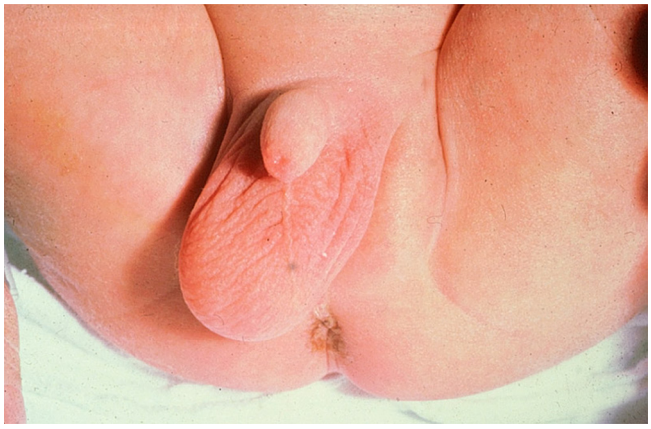

Figure 1 Picture depicting an empty hypoplastic looking hemiscrotum suggestive of an undescended testis *Courtesy of Dr Khizer Mansoor*.

testis have a better fertility outcome (considered to have normal fertility if the contralateral testis is normally descended) than those with bilateral undescended testis or intra-abdominal testis. Literature suggests a fertility rate of up to 80% for unilateral and 50–60% for bilateral undescended testes.

Ascending testis

Some boys may present later in infancy with a palpable undescended testicle. These have been noted to be normally positioned at birth in the scrotum at postnatal examination and further health checks. These testis are referred to as ascended testis as opposed to undescended testis. The ascending testis is as a result of differential growth of the boy as compared to the growth of the gubernaculum and cord structures which therefore fix the testis in a higher position relative to the scrotum. The ascended testis may represent the cohort of older children undergoing orchidopexy. The incidence of undescended testis may be as high as 32–50% in cases where a testis is significantly retractile. Testicular ascent may also be iatrogenic secondary to groin surgery after inguinal hernia repair. Orchidopexy is recommended and at the time of surgery it is common to find the obliterated processus vaginalis rather than a patent processus vaginalis.

Hydrocoele and hernia

The difference between a hydrocoele and hernia relates to the calibre of the patent processus vaginalis (PV) which develops in relation the descent of the testis (Figures 2 and 3). Failure of closure of the patent PV may lead to peritoneal fluid (hydrocoele) or intra-abdominal visceral (hernia) within a patent processus vaginalis.

Aetiology: the PV develops during the descent of the testis and acts as a conduit for the intra-abdominal fetal testis to pass to the scrotum. The natural history is for the PV to close spontaneously within the first two years' of life. A patent PV may however be present in up to 60% of boys in the first two months' of life following which there is a steady decline until 2 years of age. Boys with undescended testis (cryptorchidism) frequently have a patent PV. Hydrocoele and inguinal hernia are common in boys. About one in 50 boys will have an inguinal hernia and about 1 in 6 preterm male infants will have one.

Diagnosis: a patent PV should be regarded as a potential hernia. Consistent with the high incidence of a patent processus in the newborn, a hydrocoele may present in the neonatal period. Typically a hydrocoele presents as a painless scrotal swelling of variable size (Figure 2) and an inguinal hernia as an intermittent inguinal swelling (Figure 3). In older boys, a hydrocoele may manifest for the first time following a viral or gastroenteritic illness. A hydrocoele may give a bluish discolouration to the skin. The size may vary during the course of the day, being smallest first thing in the morning after a period of lying down.

Hydrocoeles are most often asymptomatic but occasionally children may complain of discomfort in the groin or scrotum. It is usually possible to 'get above' the scrotal swelling. Hydrocoeles are characteristically transilluminant although transillumination is not a very good diagnostic test in young boys and infants as fluid-filled obstructed loop of bowel may also give this appearance. In large tense hydrocoeles where the testis is impalpable, an ultrasound scan is advisable especially in infants where an abdomino-scrotal hydrocoele should be ruled out. If the patent processus undergoes partial obliteration, an encysted hydrocoele may develop along the line of the cord. This presents as a painless mobile cystic swelling separate from the testis but moves with the cord on gentle traction on the testis in a downward direction. A hydrocoele may present in unusual ways such as a meconium hydrocoele, with appendicitis, following intra-abdominal bleeding (haematocele).

A hernia presents as an intermittent lump in the groin and may descend down into the scrotum. In infants and especially premature babies, it may present as an incarcerated hernia in its first presentation. It is not possible to get above the swelling and in most cases can be reduced back into the abdomen. An incarcerated hernia may be painful and inflamed and may be associated with features of intestinal obstruction.

Treatment: the natural history of the processus is for it to undergo spontaneous obliteration by the age of 2 years. Surgery is indicated if the hydrocoele is present beyond this age. Surgery consists of high ligation of the processus and partial excision of the distal sac via a short groin incision. This is usually done as a day-case procedure. Abdomino-scrotal hydrocoeles can be excised with laparoscopic assistance with good outcomes. The need for contralateral groin exploration is controversial. In cases of hydrocoele secondary to ventriculoperitoneal shunts or peritoneal dialysis catheters, once the diagnosis has been made early bilateral ligation of the patent processus is recommended to prevent complications.

All inguinal herniae require surgical closure of the PV, the timing of the surgery dependant on the age of the child. Surgery is performed as soon as possible for children under 1 year of age due to the higher risk of incarceration. In premature babies on a neonatal unit, the author's preference is for the hernia to be repaired just before discharge of the child from the neonatal unit. Increasingly hernia repairs are being done laparoscopically with the advantage being that the contralateral internal ring can be