

UNDESCENDED TESTIS

Incomplete testicular descent is one of the most common malformations encountered in the male. Controversy remains regarding the aetiology, pathophysiology and optimal treatment. Testicular development and descent is a complex event and is effected by multiple factors, mechanical and hormonal, for its normal completion. Abnormalities in one or more of these can lead to an abnormal final gonadal location.

Definition

An undescended testis must be regarded as a testis that is not in the scrotum, was never there and cannot be brought down into the scrotum.

Embryology: Development and descent. Testicular development is determined at the time of conception by the presence of the SRY (sex-determining region on the short arm of the Y chromosome). Sexual differentiation begins at 7-8 weeks gestation when the developing testis arising from the genital ridge secretes testosterone (Leydig cells) and Mullerian inhibiting substance (MIS) (Sertoli cells). Testosterone allows the Wolffian ducts to develop into an epididymis, vas deferens and seminal vesicle, while MIS causes regression of Mullerian duct structures. Distal to the developing gonad the gubernaculum proliferates, connecting

the gonad to the developing inguinal region and future scrotum. Meanwhile the processus vaginalis testis, an outpouching of the peritoneum invades the gubernaculum. Testicular descent from its intra-abdominal position, through the inguinal canal takes place during the 8th intra-uterine month with concomitant increase in the length of the vas deferens and testicular vessels.

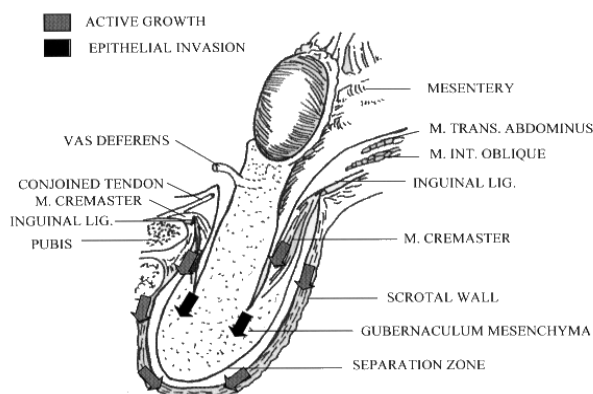
The left testis is said to precede the right in descent.

After descent the bulky gelatinous gubernaculum shrinks to thin fascial attachments.

Hormonal factors play a major role in descent, e.g. maternal chorionic gonadotrophic hormones and foetal interstitial cell activity with androgenic hormone production. Other hormones implicated are:

- Descendin. an androgen independent factor that provides gubernacular cellular growth and
- Estradiol. high levels in mothers may inhibit gubernacular growth.

Man is the only animal in which the testis descends before birth and with the chimpanzee are the only 2 in which the testis resides permanently in the scrotum. The current concept of the hormonal control of testicular descent is that transabdominal



descent is mediated by gubernaculum enlargement under the control of MIS. Inguinoscrotal migration is indirectly under testicular control through androgen secretion. Disturbance of the hypothalamo-pituitary testicular axis will interfere with the androgen dependent transinguinal descent to a variable extent.

Aetiology of incomplete descent

Failure of descent is likely to stem from two factors:

- Inherent defect in the testis, or failure of hormonal environment (vide supra)
- Mechanical defect preventing the otherwise normal testis from descending correctly, as genital development is usually normal. Occasionally a malpositioned testis is noted which is not really 'undescended', but has been hitched up out of the scrotum following herniotomy.

Side: R/L 55%/45%
Bilateral: 15-20%

Incidence

Preterm infant 21%
Full-term 2.7%
Age one year 1%

Family history 14%

The testis should be in the scrotum of a full-term baby. If not, then some spontaneous late descent may occur in two-thirds of full-term babies by 6 weeks of age, and in 3 months for premature infants. Minimal descent occurs after one year.

Histology of Undescended Testes

Normally age dependent morphological changes are seen from infancy to adulthood. These changes are caused by a congenital defect or by secondary alteration due to the malposition. Morphological changes are not seen within the first 12 months of life. Beginning with the second year histological changes involving all

structures of testicular parenchyma become apparent and become more abnormal as the malposition persists. The changes are present in bilateral and unilateral undescended testis, and in the latter situation, also in the contralateral scrotal testis. In summary there is decreased tubular size, decreased number of spermatogonia, atrophy of Leydig cells and interstitial fibrosis. Orchidopexy should be done early, before changes become irreversible and theoretically this should improve maturation after the testis is brought down.

Diagnosis

Undescended testis must be differentiated from a retractile testis where there is:

1. a history of a testis being felt or seen in the scrotum
2. a normally developed scrotum on that side
3. a testis of normal size that can be manipulated into the scrotum

75% of undescended testes referred for a specialist opinion are retractile

NOTE that both squatting and the 'chair test' where the knees are pulled up against the chest and the patient is examined relax the cremaster muscle allowing a retractile testis to be manipulated into the scrotum. The cremaster reflex is absent in the newborn and maximum at age 7-8 years.

Types

- a. Incomplete descent - arrested in its normal line of descent
- b. Ectopic - deviated from its normal line of descent after traversing the external ring.

Incomplete descent

- Abdominal - between the lower pole of the kidney and the internal ring

- Entrant inguinal - above the internal ring, but may enter the inguinal canal (impalpable)
- Inguinal - lying in the inguinal canal (usually palpable)
- Emergent inguinal - can be milked from the inguinal canal to emerge from the external ring when it becomes palpable.
- High scrotal - testis cannot be manipulated to the bottom of the scrotum, and lies at the entry to the scrotum.

Ectopic

- Superficial inguinal ectopic - lies in the superficial inguinal pouch lateral to the external ring
- Pubopenile - at the base of the penis
- Perineal - in the perineum outside the scrotum
- Femoral - in the upper thigh

Clinical examination

Should be conducted in a warm room having gained the confidence of the patient. Look first for development or abnormality of the genitalia. Warm hands. Gently bring the hand down from the ASIS towards the pubis to milk the testis out of the inguinal canal. If the testis is visible in the groin it is probably ectopic.

If the testis is difficult to feel or pick up between the fingers, it is probably in the inguinal canal.

The Impalpable Testis

When a testis is impalpable, it may be

- An incompletely descended testis lying in the abdomen or inguinal canal.
- Testicular agenesis
- 'Vanished' testis - vas and vessels are present, but the testis is absent or replaced by fibrous tissue. Probably due to a vascular accident to the testis, idiopathic infarction or antenatal torsion.

The 'Ascending' testis

An apparently normally descended testis in infancy but is later (\pm 4 yrs) noted to be incompletely descended. This is thought to be due to partial absorption of the patent processus vaginalis into the parietal peritoneum in the first years of life.

Treatment

Orchidopexy - that is mobilising the testis, its vascular supply with vas deferens and placing it in the scrotum usually in a sub-dartos pouch. Age at operation: approximately 2 years. In bilateral impalpable testis plus atrophic penis and scrotum - evidence of testicular tissue should be proven prior to operation (Testosterone levels with HCG Stimulation Test). Laparoscopy is an excellent method of identifying the presence or absence of an intra-abdominal testes when it is impalpable.

Reasons for operating

1. To improve fertility.
Unilateral undescend - 80% fertile (after orchidopexy)
Bilateral undescend - 30% fertile (after orchidopexy) ?less for intra-abdominal testis
2. Torsion is more likely in an undescended testis (10% torsion occur in UDT)
3. Prone to trauma (uncommon)
4. 70% associated with patent processus vaginalis therefore predisposes to hernia
5. Psychological
6. Malignant potential 20-50 times greater with abdominal testis: approximately 6-10 times greater in inguinal testis
Note in 20% of contralateral descended testes in unilateral undescend, dysplastic change shown. Therefore slight increased incidence in malignancy of the 'normal' testis.

Hormonal treatment

Either HCG or LHRH or both have been used. Little benefit is gained in truly undescended testes. Most testes, which have descended with hormonal therapy, have been high scrotal or retractile testes.

INGUINAL HERNIA

A hernia is defined as the protrusion of a viscus from the cavity in which it normally resides.

Indirect inguinal hernia is a hernia, which emerges through the internal inguinal ring within the cremasteric fascia extending down the inguinal canal for various distances.

If the hernia remains within the canal, it is incomplete. If it extends beyond the external ring, into the scrotum the hernia is complete.

Direct inguinal hernia.

This is a hernia presenting in the triangle of Hesselbach. There is disruption of the posterior wall of the inguinal canal medial outside the spermatic cord resulting from insufficiency of the internal oblique muscle and transversus abdominis muscle or aponeurosis in this area. Very uncommon in paediatric patients.

During embryonic development the processus vaginalis testis (canal of Nuck in a female), invades the gubernaculum. The processus vaginalis usually becomes obliterated. In a considerable number of infants failure of obliteration occurs in part or in toto, with a persistent peritoneal pouch. It is this sac of peritoneum, which is the congenital predisposition essential for the development of an indirect inguinal hernia in life. At the time of birth the sac is open in 80-94% of male infants. At the end of 2 years 56% remain open and in adults the patency rate varies between 15-35%. Once a hernia has occurred spontaneous resolution is not possible.

Incidence

Approx. 1-3%, (4% in premature births)

Male/female ratio : 6:1

60% right-sided presentation, 30% left-sided

10-15% bilateral presentation

The hernia most frequently comes to notice in the first year of life and particularly during the first three months. Direct and femoral hernias in children are extremely rare.

Clinical evidence: Bowel containing hernia

The symptoms and signs of inguinal hernia vary with the type, size, duration and presence of complications. In the groin of a child, a mass, which comes and goes with straining, must be a hernia. At times, it is necessary to base the diagnosis on a reliable history alone. Symptoms referable to hernia, eg. a feeding problem, pain, irritability, or colic, may be the first indication of a hernia.

On palpation

Thickening of the cord, a reducible tense fluctuant cylindrical or globular inguino-scrotal swelling with a blunt, rounded shape and sometimes with an expansile impulse when the abdominal pressure is increased. Ability to reduce the swelling frequently with a gurgling sensation. In infants the internal ring is palpable on rectal examination and this test may be used to confirm reduction of a hernia following an episode of irreducibility.

Congenital hydrocoele

Here, the fluid formed by the serosa of the peritoneal cavity trickles down a narrow patent processus vaginalis and collects in the tunica, around the testis. Clinically, one can get above it and it transilluminates. An encysted hydrocoele of the cord is where fluid is trapped within the tunica above the testis.

Fluid hernia

This implies the presence of a wider patency of an attenuated processus vaginalis and usually presents in the toddler with increasing scrotal swelling during the day, which settles overnight. Sometimes it is difficult to distinguish from a bowel containing hernia but it is usually confined to the scrotum, is a non-tender, transilluminable fluid collection around the testicle.

Exclude undescended testis in boys with a suspected hernia (6%).

Inguinal hernias in girls

Very similar to the male with a few exceptions.

1. Incidence of complications higher
2. 25% of hernias are sliding hernias, where the broad ligament, bladder or vascular pedicle make up part of the wall of the sac
3. The ovary and fallopian tube commonly (>75% at RXH) prolapse into the hernial sac. This may present as an 'irreducible' hernia, but without bowel symptoms. Torsion of the ovary in the sac may occur
4. Always the possibility of a testicular feminizing syndrome (Androgen insensitivity) viz: Incidence 1% of 'girls' with inguinal hernia

Hernias are usually bilateral and contain gonads

Phenotype - female

External genitalia - female, presenting with primary infertility if missed as children

On examination the vagina will be found to be short, the cervix, uterus, fallopian tubes rudimentary or absent, and the gonad - testicular on histology

Genotype of child - male with XY chromosomal pattern

CAUSE: due to enzymatic defect so that testosterone cannot exert its effect on the target organ with incomplete masculinization.

Management of androgen insensitivity

- Herniotomy - if hernia contains a gonad – biopsy
- Gonadectomy (controversial as to whether gonadectomy should be done in infancy)
- Provide endocrine function artificially at puberty

Complications

Irreducibility/Obstruction/Strangulation

Complications may be serious and the incidence is inversely related to age. The younger the child the more likely it is for the hernia to become complicated - 30% present with irreducibility.

1. Irreducibility may be the first manifestation of a hernia with 60% of them occurring within the first three months of life. This provides further strong support for the trend towards repairing hernias when they are diagnosed regardless of age.
2. Obstruction: Evidence of gastrointestinal tract involvement (cramping pain, bile stained vomiting and abdominal distension) together with groin mass.
3. Strangulation: this implies vascular compromise. This is clinically evident with an inguino-scrotal mass, which is tender. Eventually oedema and erythema of the overlying skin develops. Nausea, vomiting with abdominal distension is indicative of small bowel obstruction. Constitutional upset with fever, and shock may complicate the clinical situation.

Testicular ischaemia with potential necrosis of the testis is a further complicating factor. Testicular atrophy occurs in $\pm 10\%$ of testes following an episode of irreducibility.

Treatment

Uncomplicated- Herniotomy

All hernias should be repaired when they are diagnosed. The only contra-indication to immediate surgery irrespective of age, is concomitant disease or serious impairment of the patient's general condition. Only the symptomatic side is repaired in males. Surgery on the opposite side will depend on local evidence of patency of the processus vaginalis. Both sides are done in females as the incidence of a sac on the opposite side is 70% and there is no hazard of reproductive structure damage.

A congenital hydrocoele and hydrocoele of the cord not associated with a hernia will usually resolve by the age of 1 year; if not, then herniotomy.

Irreducible / Obstructed hernia

This needs urgent attention. The baby is sedated and a short attempt at reducing the hernia is made (<10 mins) using gentle, but never forceful pressure. If successful, elective herniotomy is then performed within 48 hours. If taxis fails, then immediate operation. There is a higher incidence of progression to emergency surgery in Cape Town (>50%) than in First World referral centres (<20%).

Strangulated hernia

Evidence of strangulation must be regarded as an emergency, the earliest sign being **oedema of the overlying skin**. Taxis should not be attempted. The infant requires urgent resuscitation and correction of electrolyte and metabolic abnormality.

As soon as the condition has been stabilised, proceed with operative relief of the obstruction, assessment of viability of the bowel and herniotomy.

ACUTE SCROTUM

Sudden onset of pain and swelling of the scrotal contents.

Differential diagnosis of an "acute scrotum"

1. Complicated hernia - excluded by clinical examination (inguino-scrotal swelling)
2. Testicular torsion (50%)
3. Epididymo-orchitis (30%)
4. Torsion of appendix of testis or epididymis (15%)
5. Idiopathic scrotal oedema (<2%)
6. Other rare causes

There is often a vague history of trauma - which should be ignored unless there is obvious evidence on clinical examination. It is extremely difficult to clinically differentiate testicular torsion from epididymo-orchitis. All "acute scrotums" should therefore be explored surgically without delay. In torsion, in addition to detorsion and fixation of the affected testis, the testis on the opposite side should also be explored and fixed, as the underlying anatomical defect of a high investment of the tunica vaginalis around the testis (so-called bell and clapper testes) usually affects both sides. If epididymo-orchitis is found a pus swab is taken for culture and antibiotic sensitivity. The testis may be very swollen and a releasing incision in the tunica albuginea will alleviate pain and protect the testis from atrophy. Post-operative underlying genito-urinary abnormality should be excluded by investigation in children <2yrs or with recurrent attacks.



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