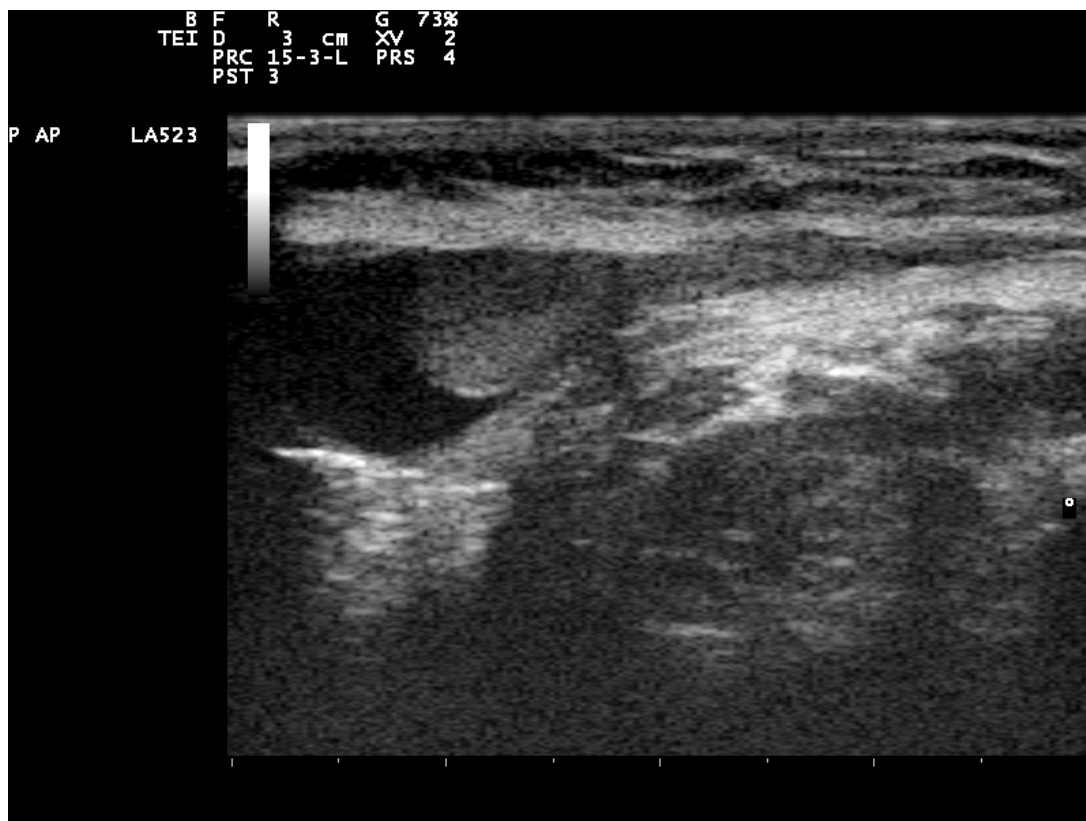


Undescended Testis (Cryptorchidism) in Children — Definition and Surgery

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Cryptorchidism or undescended testis is the most common disorder of the sexual differentiation, seen in 2–4% of male infants at birth. The exact cause of the cryptorchidism is still not known. Early treatment of this condition is necessary to avoid complications, such as infertility, testicular malignancy, torsion, and/or an inguinal hernia in later life. The treatment is essentially surgical which involves the re-positioning of the testis within the scrotal sac, also known as an orchidopexy. The currently recommended timing for an orchidopexy is between 6 and 12 months of life. In this article, epidemiology, etiology, pathogenesis, diagnosis, differential diagnosis, evaluation, complications and the treatment of undescended testis in children are described.



Definition

Cryptorchidism can be **defined as hidden testis or a testis which is not within the scrotum** and cannot be manipulated into the scrotum without pain. Cryptorchid testis can be absent, but most of them are undescended (UDT). Testis may be absent due to

agenesis or intrauterine vascular compromise (e.g. torsion). A true or congenital UDT is one which had not previously descended along its normal path.

Ectopic testes (accounts for < 1% of UDT) descend normally, but then are diverted to an aberrant position such as suprapubic region, perineum, femoral canal, superficial inguinal pouch, or contralateral scrotum. Retractable testes are suprascrotal testes which are often misdiagnosed as UDT. Due to brisk cremasteric reflex, it becomes difficult to manipulate testis into the scrotum; although, it can be pulled into the scrotum and remain there if cremasteric reflex is overcome.

Acquired or ascending UDT, recognized more frequently now, is defined as UDT in which a previous scrotal position was documented at least once (Hack WW, *et al* 2003). Usually, testes are present in a scrotal position at birth, but, in early childhood (between 4–10 yrs), the testes “ascend” out of the scrotum. Secondary cryptorchidism can be seen as a complication of inguinal hernia repair in up to 1–2% of the patients, commonly neonates and young infants.

Epidemiology of Cryptorchidism

Cryptorchidism is more common in premature infants as testicular descent occurs late in gestation; therefore, up to 30% of premature male newborns have UDT at birth in comparison to 2-5% of full-term infants. Most of the UDT descend spontaneously so that, by the age of one year, the prevalence decreases to < 1%. Spontaneous descent is rare after six months; therefore, the optimal time for surgery is six months of age.

Geographical variation in its prevalence is seen and it varies from 4.3–4.9% at birth to 1–1.5% at 3 months to 0.8–2.5% at 9 months of age; however, the exact reason of this variation is unknown but may be due to genetics or environmental factors. Risk ratios for cryptorchidism are 10.1 in twins, 3.5 in brothers, and 2.3 in offspring of fathers who had a UDT. Cryptorchidism is usually unilateral with a left-sided predominance; however, bilateral UDT is seen in 10% of cases.

Most of the UDT is palpable and common locations are suprascrotal (outside the external ring), inguinal canal, and abdomen. Approximately 20% of boys have at least one non-palpable testis which may be intra-abdominal, atrophic, or absent.

Etiology of Cryptorchidism

These include prematurity, low birth weight, small for gestational age, twin pregnancy, breech presentation, and maternal factors (elderly mothers, obesity, diabetes, and estrogen exposure during pregnancy) and family history of cryptorchidism. Genetic susceptibility also exists, but this is likely to be polygenic and multifactorial.

Exposure to endocrine-disrupting chemicals may contribute to cryptorchidism and may account for the increasing incidence rate of cryptorchidism seen in some regions. Synthetic chemicals identified as endocrine disruptors include phthalates, pesticides, brominated flame retardants, diethylstilbestrol, and dioxins

Although its exact mechanism of action is unclear, the gubernaculum has significant importance in undescended testes. In patients with cryptorchidism, the gubernaculum is not firmly attached to the scrotum, and the testis is not pulled into the scrotum.

Both hormonal and mechanical factors appear to mediate the aid of the gubernaculum and descent of the testis. The genitofemoral nerve may also aid in descent and

gubernacular differentiation, which may be mediated by calcitonin gene-related peptide.

Intra-abdominal pressure also appears to play a role in testicular descent. Conditions associated with decreased pressure include [prune belly syndrome](#), [cloacal exstrophy](#), [omphalocele](#), and gastroschisis, among other various syndromes. Each is associated with an increased risk of undescended testes.

The effect of decreased intra-abdominal pressure is most significant during transinguinal migration to the scrotum, probably in conjunction with androgens and a patent processus vaginalis.

Associated Anomalies

Associated anomalies and conditions may include the following:

- Patent processus vaginalis
- Abnormal epididymis
- [Cerebral palsy](#)
- Mental retardation
- [Wilms tumor](#)
- Abdominal wall defects (eg, gastroschisis, omphalocele, prune belly syndrome)
- [Hypospadias](#)

In general, ductal abnormalities, hernias (patent processus vaginalis), and testicular maldevelopment are more common in patients with abdominal testes. Overall, 32-79% of undescended testes are associated with some type of epididymal abnormality.

Cryptorchidism is an associated finding in many clinical syndromes e.g. Kallmann syndrome, Noonan syndrome, trisomy 13, trisomy 18, Prader-Willi syndrome, and Laurence-Moon-Biedl syndrome and abdominal wall defects, neural tube defects etc. Co-existing cryptorchidism and hypospadias increase the risk of sex development disorders, such as mixed gonadal dysgenesis.

Pathogenesis of Cryptorchidism

The development of testis begins at 7–8 wks of gestation. At 10–11 wks, the Wolffian (mesonephric) duct is differentiated into the epididymis, vas deferens, seminal vesicle, and ejaculatory duct in the presence of testosterone. The intra-abdominal descent of testis is androgen-independent.

By the 28th week of gestation, testis starts descending through the inguinal canal. Various hormonal (a müllerian-inhibiting substance (MIS), testosterone, dihydrotestosterone) and mechanical factors (gubernaculum, intra-abdominal pressure, genitofemoral nerve) help to regulate this process.

At 32–36 wks, gubernaculum anchors the testis at the internal inguinal ring and guide the testis into the scrotum by distending the inguinal canal. Alterations in any of the above-mentioned factors may lead to UDT e.g. gonadotropin deficiency, decreased MIS, and increased estradiol expression in the placenta, changes in abdominal pressure, and gubernacular regression. The role of molecular factors e.g. mutations in insulin-like factor-3 is also being studied.

Differential Diagnosis of Cryptorchidism

Differential diagnosis includes retractile, ectopic, or absent testis. Retractable testes can easily be ruled out with a thorough history and examination; however, it is difficult to differentiate retractile from ectopic testis. A phenotypically male newborn with bilateral non-palpable testes should be evaluated to rule out anorchia or a genetic female with congenital adrenal hyperplasia (CAH), or androgen receptor disorders, or disorders of sex development. Surgical exploration often differentiates testicular agenesis and intrauterine torsion as in agenesis. All testicular structures are absent whereas their remnants can be seen in torsion.

Evaluation

History

Try finding the family history of infertility or genital anomalies, and unexplained neonatal death or history of endocrine problems during pregnancy. Try to elucidate the presence of testes in the scrotum during the neonatal period.

Physical examination

To look for the features of congenital malformation syndromes.

Genital examination

Look for any abnormality, such as hypospadias, hypoplastic or hemiscrotum and inguinal fullness. In infants and young children, the testicular examination should be done with two hands with one hand near the anterior superior iliac spine (ASIS) and other on the scrotum. The first hand is swept from ASIS along the inguinal canal towards the scrotum. A true undescended or ectopic inguinal testis slides under the examiner's fingers, while low ectopic or retractile testis is felt by the second hand as testis is milked into the scrotum.

If the testis is non-palpable, a "soap test" is often useful; soap applied to the inguinal canal and hand significantly reduces friction and facilitates identification of inguinal testis.

When, after pulling the testis into the scrotum, it is held for one minute to fatigue cremasteric muscle. After this maneuver, retractile testis remains in the scrotum, whereas ectopic testis immediately comes out of the scrotum. Pediatric urologist opinion should be taken to differentiate retractile and true UDT or in the presence of palpable, but probably atrophic testis in the scrotum.

The examiner should note the position of the testis, its consistency, and size of both testes. If the testis is not palpated in inguinal canal or scrotum, or in its usual ectopic sites, evaluation should be done for non-palpable testis.

Radiologic evaluation

It is usually not warranted in the evaluation of non-palpable testes due to the lack of sensitivity and specificity. However, ultrasound may be useful 1) in male newborns with bilateral non-palpable testes to identify the gonads (testis or uterus); and 2) in case of difficulty to feel intracanalicular testes, such as in obese boys.

Laboratory evaluation

In phenotypically male newborns with bilateral non-palpable testes, the life-threatening condition CAH must be excluded by the following investigations: ultrasound pelvis, karyotype, and measurement of electrolytes, LH, FSH, testosterone, MIS, and adrenal hormones and metabolites (e.g. 17-hydroxyprogesterone).

Hormonal evaluation should be done in older children with bilateral non-palpable testes and elevated gonadotropin levels, absent MIS levels and a negative human chorionic gonadotropin (hCG) stimulation test may suggest testicular absence in them. Boys with anorchidism do not respond to hCG; whereas, boys with bilateral cryptorchidism respond with increased testosterone production.

Complications of Cryptorchidism

The complications include testicular malignancy, infertility, testicular torsion, inguinal hernia and psychologic effects of an empty scrotum. Retractable and absent testicles do not increase these risks. Ectopic testes are prone to trauma, leading to decreased spermatogenesis; however, the risk of malignancy is not increased.

Testicular cancer (high yield for USMLE)

Males with UDT have 4 to 10 times more risk of malignancy than the general population with the peak age of 15–45 yrs. Approximately, 1 in 80 patients with unilateral and 1 in 40 in bilateral UDT has a risk of developing a malignancy. Approximately, 10% of the patients with testicular tumors have a history of UDT. The risk of developing malignancy is four times less in inguinal than with intra-abdominal testes.

The most common tumor is seminoma (65%); however, after surgery, only 30% of testis tumors are represented by seminoma. The role of early orchiopexy to reduce the risk of testicular malignancy is controversial; however, chances of development of cancers are rare if orchiopexy has been performed before 2 years of age. The contralateral scrotal testis is not at an increased risk for malignancy. Early surgery also permits the earlier detection of testicular masses, improves fertility, and prevents torsion.

Infertility (high yield for USMLE)

These patients have an increased incidence of infertility along with lower sperm counts, and sperms of poorer quality due to the adverse effect of temperature on spermatogenesis. Intraabdominal and intracanalicular UDT are affected similarly. The degree of germ-cell dysfunction is correlated with the duration of the suprascrotal location of the testes and whether one or both testes are affected. In contrast to malignancy, there is a clear advantage to performing early orchiopexy for the protection of fertility.

After treatment, 50–65% of the patients with bilateral UDT and 85% of patients with unilateral UDT are fertile. An orchidopexy should be performed before 1 year of age to minimize infertility (Chung E, *et al*, 2011).

Testicular torsion

The incidence of torsion is 10 times higher in UDT than in normal scrotal testes. It is more common in neonatal age and after puberty; although it can occur at any age. Increased weight and distorted architecture of the organ may be the cause of the torsion. Torsion of an intra-abdominal testis can present as an acute abdomen. Early diagnosis and surgical

treatment can prevent this complication.

An inguinal hernia

Up to 90% of the UDT have associated inguinal hernia. In untreated cases, it can present with complications e.g. incarceration or strangulation. In symptomatic cases, complete repair with orchiopexy should be done at presentation; otherwise, an orchiopexy should be combined with hernia repair.

Treatment of Cryptorchidism

Treatment can be surgical, hormonal, or a combination of both.

American Urological Association Guidelines 2014 (Kolon TF, *et al*, 2014):

- Testes should be examined for their position at every well-baby visit.
- Children with cryptorchidism should be referred by the age of 6 months for further evaluation and management.
- Immediate consultation should be taken in all phenotypic male newborns with bilateral non-palpable testes to rule out disorders of sex development.
- USG or other imaging should not be done prior to the referral to a specialist.
- Orchidopexy should be done in case of the failure to descend spontaneously by 6 months of age.
- Currently, hormonal therapy has no role in the management of cryptorchidism.
- Surgical exploration should be done in pre-pubertal boys with non-palpable testis and, if indicated, an abdominal orchidopexy should also be performed.
- Patients and their parents should be counseled about the risk of infertility and malignancy.

Surgical

The congenital UDT should be treated surgically by 9-15 months of age; even at 6 months as the spontaneous descent is unlikely after the age of 4 months.

Orchiopexy

Surgery of choice for palpable UDT is inguinal orchiopexy, in which testis and spermatic cords are brought into the scrotum and sutured in place along with the correction of an inguinal hernia.

The procedure is typically performed on an outpatient basis with a success rate of 98%.

For laparoscopy-assisted orchiopexy (LAO), mobilize the testicular vessels laparoscopically up to the renal level to avoid tension for a classic open inguinal orchiopexy. Increased magnification aids in dissection. Make abdominal port incisions and an open inguinal incision. The complications of orchiopexy are rare and include testicular atrophy (most significant), the ascent of the testis, infection, and bleeding.

Complications

Complications of an orchiopexy are as follows:

- Inadequate testis position occurs in up to 10% of patients and is due to incomplete retroperitoneal dissection. It is usually corrected with a second

procedure.

- Testicular atrophy due to devascularization during dissection of the cord occurs in approximately 5% of patients. Orchiectomy may be indicated to treat cancer, with subsequent prosthesis placement if requested.
- Accidental division of the vas deferens occurs in 1-2% of patients. Immediate or post-pubertal microvascular repair may be used.
- Epididymo-orchitis is uncommon and may be treated with antibiotics.
- Scrotal swelling may occur and is usually secondary to edema. If progressive, it may be due to bleeding and should be explored. A later presentation of swelling may be secondary to a [hydrocele](#), which, if large, requires transscrotal repair.

Complications of laparoscopy include the following:

- Preperitoneal emphysema may develop secondary to poor needle or trocar placement and insufflation.
- Hypercarbia may occur with pneumoperitoneum. This can be counteracted by increasing minute ventilation and keeping insufflation pressures at less than 12 mm Hg.
- The puncture of a viscus with the Veress needle is not harmful unless insufflated or dilated by a trocar.
- The puncture of a major abdominal vessel is a life-threatening complication. The needle or sheath should be left in place for a tamponade, and an emergency open laparotomy is performed. (Almost all the above potentially life-threatening vascular and enteric complications can be avoided by using an open technique for access.)
- Injury to the inferior epigastric vessels may also occur with trocar placement. Hemostasis can be achieved via direct cautery or via suture/clip ligation from the external surface or laparoscopically.
- Bladder puncture may occur because the bladder is intra-abdominal in younger children. Care must be taken when placing lower abdominal and scrotal trocars. Repair can be achieved laparoscopically or through a small suprapubic incision. A ureteral injury requires stenting.

Exploration

Surgery for the non-palpable testis is both diagnostic and potentially therapeutic which can be either an open inguinal approach or laparoscopy, the latter being preferred. Surgical exploration determines the presence and location of the testis. If the testis is found viable, positioning and fixing within the scrotum is attempted, but complete removal is done if it is non-viable.

About 50% of boys have testis in the abdominal or high inguinal canal, while the remaining have absent testis. Vanishing testes is a term given to atrophic remnants found in the scrotum or inguinal canal in cases with absent testis. In the majority of the cases, an orchidopexy is done for intra-abdominal testis (near internal inguinal ring) and is successful; however, a two-stage orchidopexy is sometimes needed in high abdominal testes. An orchiectomy is reserved for the difficult cases or if the testis is atrophic.

To alleviate undesirable psychologic effects due to absent gonads in the scrotum in older children and adolescents, testicular prostheses can be implanted inside the scrotum. The FDA has approved a saline testicular implant; although, solid silicone "carving block" implants are also being used. In boys with anorchia, testicular prostheses should be

placed early in childhood.

Hormonal

Hormonal treatment, such as hCG and gonadotropin-releasing hormone (GnRH), is used infrequently. These may stimulate testicular descent by stimulating Leydig cell production of testosterone. In the US, hCG is the only hormone approved for the treatment of cryptorchidism. It is administered intramuscularly, 1500 to 2500 units twice-weekly for four weeks. Initial treatment with GnRH may deserve some consideration because it is administered as a spray rather than an injection.

The recognized adverse effects of increased androgens, including increased penile or testicular size, scrotal erythema, or erections, seem to be less with GnRH than with hCG.

Either of these hormones is not found to be very effective with reported efficacy of 5 to 20% and a relapse rate of 25%; however, it may improve fertility in cryptorchid boys. The decision to use hormonal treatment depends on the pre-treatment location of the testis.

Review Questions

The correct answers can be found below the references.

1. A 6-month old boy was brought in by the parents when the mother noticed that the left side of the scrotal sac is empty. On examination, you were able to pull the testis down to the scrotal sac which remained there. How will you manage this child?

- A. Scrotal orchidopexy
- B. Re-assurance and re-evaluation
- C. Refer to pediatric urologist
- D. USG pelvis

2. A 2-month-old male baby was brought in with complaints of an empty scrotal sac. On genital examination, some mass was felt below the examining finger. How will you manage this case?

- A. Nothing to do, reassure the parents
- B. Order USG pelvis
- C. hCG therapy
- D. Re-evaluate at 6 month

3. A phenotypically male newborn was found to have bilateral non-palpable testes. Which of the disorder should be ruled out immediately?

- A. Congenital Adrenal Hyperplasia
- B. Vanishing Testis
- C. Testicular Torsion
- D. Retractable Testis

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Correct answers: 1B; 2D; 3A

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Notes