

Undescended Testicles, Retractable Testicles, and Testicular Torsion

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Key Points

- Testicles should be palpable in the scrotum by 6 months of age.
- When testicles are not palpable, are unable to be brought to the scrotum, or do not remain in the scrotum by 6 months of age, a referral to a pediatric urologist is recommended for evaluation of an undescended testicle.
- Retractable testicles can be brought down to the scrotum and will remain there. If there is difficulty with bringing the testicles to the scrotum, a referral to a pediatric urologist is recommended.
- Testicular torsion outside of the perinatal period is a surgical emergency and emergent pediatric urology consultation is recommended.
- Perinatal testicular torsion presents with a painless firm testicle noted right after birth. A pediatric urology consultation is recommended.

Undescended Testicle

Definition

- Testicle is not located in the scrotum and classified based on location
 - Intra-abdominal (non-palpable)
 - Inguinal canal
 - Superficial inguinal pouch

- Upper scrotum
- Ectopic (rarely)
- Incidence
 - Term newborn 3%; at 1 year 0.8%
 - Pre-term newborn <37 weeks 30%; at 1 year 10%
 - Twenty percent (20%) of undescended testicles (UDTs) are non-palpable
 - More common on the right side (2:1)
 - Monorchid or anorchid occurs 33% in child presenting with non palpable testicles.¹
 - Occurs because of in-utero torsion or vascular event during development or descent
 - Bilateral anorchia estimated to occur 1 of every 20,000 boys
 - Three times more likely with family history of UDT

Assessment

- Birth history
 - Term or preterm baby
 - Easiest to detect in the newborn period when cremasteric reflex is weak and absence of large amounts of fat
- Majority of testicles will not descend after 6-9 months of age.
 - Testosterone surge at 2-4 months augments descent of UDT; 70-77% of UDTs will descend spontaneously in first 3 months²
 - Seven percent (7%) of UDTs will descend spontaneously after 6 months^{2,3}
- Malignancy risk
 - Increased risk of malignancy compared to general population
 - 1:1000 to 1:2500 vs. 1:100,000⁴
 - Three times an increased risk compared to general population⁵
 - Twenty percent (20%) of testicular cancers in men with UDTs occurs in contra-lateral descended testicle.
 - Orchiopexy does not change the risk of cancer, allows for self-examination.
- Fertility potential
 - Paternity rates lower for men with history of bilateral UDTs (62%) compared to men with unilateral UDT (89.5%) and general population (94%)⁶
 - Timing of orchiopexy helps protect fertility; testicular biopsy has shown decreasing germ cell density starting at 1 year of age.⁷
 - Location of UDT does not significantly affect paternity; although intra-abdominal UDT was borderline significant ($p = 0.06$) (90% all UDT; 83.3% intra-abdominal)⁸

Diagnosis

- Prior history of testicles in scrotum at birth or during first year of life
- Parental reporting of visualizing the testicles in the scrotum during baths
- Examination in supine frog-leg position with warm hands
 - Start at anterior superior iliac spine and move down canal toward scrotum.

- Once testis palpated, bring to scrotum and hold for short time to fatigue the cremasteric muscles and then release.
- An UDT will return to pre-scrotal position, a retractile testicle will remain in scrotum.
- Imaging
 - Ultrasound, CT, and MRI not recommended for non-palpable testicles.^{9,10}
 - Ultrasound may be indicated in the obese child
- Laboratory testing
 - Not recommended for unilateral UDT or in boys with bilateral UDTs when testes were once palpable
 - If bilateral non-palpable testicle and phenotypically male work up for disorders of sexual differentiation should be performed¹¹
 - Karyotype, serum electrolytes, and hormonal profile (17-hydroxyprogesterone, luteinizing hormone, follicle-stimulating hormone, testosterone, and androstenedione)
 - Consultation with pediatric endocrinology and urology
 - If bilateral non-palpable testicle and no congenital adrenal hyperplasia; consider workup for bilateral vanishing testes versus bilateral intra-abdominal testes (20x more likely)¹¹
 - Consider müllerian inhibiting substance or anti-müllerian substance
 - Inhibin B, luteinizing hormone (LH), follicle-stimulating hormone (FSH) and testosterone determines if testes present
 - If >3 months consider human chorionic gonadotropin (hCG) stimulation test (100 IU/kg) to determine if testes are present - rise in testosterone; along with FSH and LH which should be elevated in anorchia
 - If unilateral UDT and severe proximal hypospadias consider possibility of disorders of sexual differentiation.¹¹

Management

- If there is difficulty in palpating the testicle at 4-6 months of age, refer to pediatric urologist for orchiopexy within first year.¹¹
- Orchiopexy as an outpatient surgical procedure.
 - Mainstay in the United States
 - For an UDT located in the groin, an inguinal incision is made to locate the testicle, the hernia sac is freed from the testicle, vessels and vas deferens, and then the testicle is positioned in the scrotum through a separate incision.
 - This can also be done through a scrotal incision.
 - Success rate for inguinal testicles is 85-91%.¹²
 - For a non-palpable testicle either a groin exploration or diagnostic laparoscopy is performed to locate the testicle, based on surgeon preference, and the testicle is brought down to the scrotum.
 - For high intra-abdominal testicles, alternative maneuvers may be used, such as ligation of the testicular vessels (one stage or two-staged orchiopexy) to gain additional length on the testicle.

- Success rate is 60-80%.¹³
- Hormonal therapy
 - Not recommended by the AUA.¹¹
 - More common in Europe
 - hCG is analog to LH; mechanism for testicular descent is unknown
 - Dosage:
 - 250 IU/dose in young infants
 - 500 IU/dose up to 6 years old
 - 1000 IU/dose >6 years
 - Maximum dose of 15,000 IU (closure of epiphyseal plate)
 - Twice a week for 5 weeks (International Health Foundation)
 - Success rate is 6-21% in randomized, blinded studies.
 - May be indicated in setting of bilateral non-palpable testicles to confirm testicular tissue
- Gonadotropin-releasing hormone (GnRH) analog to simulate pituitary to release LH and FSH
 - Available as a spray
 - Only approved to treat UDT in Europe

Retractile Testicles

Definition

- Movement of the testicle from the scrotum to the suprascrotal position
- Affected by the strength and contraction of the cremasteric reflex
 - Affected by temperature, stimulation of superficial branch of the genitofemoral nerves, emotional anxiety
 - Varies with age
 - Reflex is weak in the neonate (testicles are larger)
 - Strongest in 5-10 years
 - Post pubertal (testicles are larger)
- Can lead to ascending or acquired UDT
 - Hyperactive cremasteric reflex, incomplete absorption of patent process vaginalis, or low-lying UDT
 - Three to thirty-three percent (3 -33%) of retractile testicles become UDT^{11,14,15}
 - Greatest risk is in boys <7 years of age

Assessment

- Parental history of prior testicular location
- History of identifying testicle during baths or showers

Diagnosis

- Examination in supine, frog-leg position with warm hands
- A retractile testicle will remain in the scrotum once it is brought down and the cremaster

muscle is fatigued

Management

- Yearly exam to confirm testicles are palpable in the scrotum¹¹
- If difficult to palpate, refer to pediatric urologist
- Have parents evaluate and report if the testicles are visible in the bath

Testicular Torsion

Definition

- Normal anatomy of the testicle relies on tunica vaginalis covering the anterior surface of the testicle, epididymis; attachment of the gubernaculum (inferiorly) and scrotal wall posteriorly preventing twisting around vascular mesentery
- Lack of normal attachments can lead to twisting
- Bimodal distribution
 - Perinatal
 - Ten percent (10%)
 - Extra-vaginal, twisting of entire spermatic cord
 - Pubertal
 - Ninety percent (90%)
 - Intra-vaginal, abnormal fixation of testicle/epididymis leads to twisting within the tunica vaginalis; bell clapper deformity
 - Increasing testicular mass increases chance of torsion
 - Intermittent torsion, spontaneous twisting and untwisting of testicle

Assessment

Perinatal torsion

- Seventy percent (70%) occurs prenatally.
- Thirty percent (30%) occurs in previous normal testicle.¹⁶
- Painless swelling and scrotal discoloration/induration

Pubertal torsion

- History and physical exam critical
- Acute onset of localized severe scrotal pain is 89%.¹⁷
- Associated nausea and vomiting is 39%.
- Prior history of ipsilateral testicular pain is 36%.
- Associated trauma and recent exercise 4% and 10%, respectively
- Dysuria and urgency is <5%
- Prior history of orchiopexy does not rule out testicular torsion.
- On exam, high riding firm tender testicle, abnormal transverse orientation of testicle, lack of cremasteric reflex
- Late findings include hydrocele, scrotal wall edema

Torsion of appendix testis or epididymis

- Comparatively, torsion of appendix testis or epididymis usually has a gradual onset; localized pain to superior portion of testicle initially; nausea, vomiting and abdominal pain usually absent

Diagnosis

- Clinical history may help differentiate between testicular torsion and torsion of appendix testis/epididymis.
- Scrotal ultrasound maybe obtained to confirm absence of testicular torsion to avoid surgical exploration; if ultrasound demonstrates normal blood flow then testicular torsion is very unlikely.¹⁸
- Testicular atrophy can occur as early as 4 hours if the degree of torsion is $>360^\circ$, but generally occurs after 8 hours.

Management

Perinatal torsion

- Treatment is controversial; rare to salvage a unilateral neonatal torsion
- In some instances delayed orchiectomy and contra-lateral orchiopexy is performed.

Pubertal torsion

- If clinical concern for testicular torsion, emergent pediatric urology consultation indicated for possible surgical exploration
- If detorsion is attempted, success would be defined by immediate resolution of pain.
 - Testis should be turned medial to lateral
 - If unsuccessful, can attempt lateral to medial
 - Should have immediate relief of pain
- Scrotal exploration with possible ipsilateral orchiopexy vs. orchiectomy of torsed testicle and contralateral orchiopexy

This guideline was developed to improve health care access in Arkansas and to aid health care providers in making decisions about appropriate patient care. The needs of the individual patient, resources available, and limitations unique to the institution or type of practice may warrant variations.

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