Inguinoscrotal conditions in children

1. Inguinal hernia: (very common)

Associations:

- Cystic fibrosis (15%)
- Hydrocephalus (VP shunts: ↑ intra-abdominal pressure → increased risk of bilaterality, incarceration, and recurrence).
- Peritoneal dialysis.

Others: undescended testes, abdominal wall defects, connective tissue disorders (Ehlers-Danlos syndrome), mucopolysaccharidosis (Hunter/ Hurler syndrome), ascites, congenital hip dislocation, meningomyelocele.

Pathophysiology: (check appendix 5 for full details)

- Descent of the testis is directed in the abdominal phase by insulin-like 3 protein (from Leydig cells), and in the second phase by androgens and release of calcitonin gene-related peptide (CGRP from the genitofemoral nerve), which also mediates closure of the patent processus vaginalis (PPV).
- Incidence of PPV in newborn is estimated to be 40-60% (1-5% will stay patent). **Failure of the PPV to close results in an indirect inguinal hernia.**

Epidemiology:

- 1-5% of all children with 5:1 M:F ratio).
- Premature infants: 10-30% (M:F 1:1)).
- 10% positive family history.
- ↑incidence in twins (esp. male twins).
- Right sided hernias are twice as common.
- Mean age at diagnosis 3.3 years.

Clinical presentation:

- Most are asymptomatic except for inguinal bulging upon straining.
- The diagnosis is clinical and depends on history and PEx. A good history is acceptable as an indication for operation.
- To demonstrate the hernia, have the child raise their head while supine or 'blow up a balloon' with a thumb in the mouth or stand the child upright.
- Cellphone picture documentation by parents has become commonplace.
- Radiologic diagnostic aids are not generally necessary or helpful.

DDx: retractile testis, lymphadenopathy, hydrocele, prepubertal fat

2. Hydrocele

- Noncommunicating, asymptomatic hydrocele in an infant can simply be followed up, 89% resolve by 1 year of age. Surgery can be done by 1 or 2 years of age if the hydrocele fails to resolve or if a clinical hernia is apparent.
- Communicating hydroceles (PPVs): 63% resolve completely without operation by nearly 1 year. Most surgeons repair hydroceles of the cord.
- Excision of the hydrocele sac is not necessary, the fluid should be evacuated, and the distal sac is opened widely. Large or thick sacs may be everted behind the cord (Bottle procedure).

*__Hydroceles in adolescents:__ management is different.
A complication of varicolectomy (often), an inguinal hernia, or an idiopathic hydrocele.

Incarceration: (12-17%)

Risk factors: younger age and maturity.

Signs and symptoms:

- Inconsolable infant with intermittent abdominal pain and vomiting.
- A tender and erythematous irreducible mass noted in the groin.
- Abdominal distention and bloody stools (late signs).
- Peritoneal signs (strangulation).

Reduction of incarcerated hernia:

- 90-95% of incarcerated hernias can be successfully reduced.
- Monitored conscious sedation, then firm and continuous pressure is applied around the incarceration.
- Questionable or incomplete reductions should be explored.
- Urgent operation is necessary if reduction fails.
- The presence of peritonitis or septic shock is an absolute **contraindication** to attempted reduction.
- *Reduction en masse*: the hernia contents are reduced into the peritoneal cavity, but the bowel remains incarcerated internally in the hernia sac.

Management:

1. **Open repair:** (IV fluids and hemodynamic stability is a priority before any surgery).

Inguinal crease incision, incise the external oblique aponeurosis to the internal inguinal ring, then the anteromedial hernial sac is grasped, the vas and the vessels are pushed away from the sac. The sac is clamped and divided. High ligation is performed after the sac is opened and inspected.
Mesh or prosthetic materials are almost never required in children (one exception is recurrent hernias in children with connective tissue disorders or mucopolysaccharidoses).

Sliding hernias are uncommon but are more frequent in females. (Fallopian tube, ovary, or bladder may constitute the medial wall of the sac). Appendix (Amyand's hernia) may form a sliding component on the right.

2. **Laparoscopic repair:**

- No difference in recurrence.
- Decreased incidence of metachronous hernia and time for bilateral repairs.
- Increased operative time for unilateral repair.
- Contralateral evaluation through laparoscopy when: premature, younger age, female, left-sided hernia.

Complications:

- Recurrence (<1%), higher in premature infants, incarcerated hernias, associated diseases (CT disorders, VPS).
- Injury to the spermatic cord or testis (rare).
- Infection 1-3%
- Hematoma.
- Persistent hydrocele.
- Chronic pain (uncommon in children).
- Loss of domain (due to large hernia).
- Iatrogenic cryptorchidism.

3. Undescended testes

A testis that has halted somewhere along the normal path of descent from abdomen to distal inguinal ring. Normal testicular descent relies on numerous factors (INSL-3, androgens, CGRP). Any deviation from the normal process can result in a cryptorchid.

Classification:

a. **Palpable** (65-75%) or **non-palpable** (Intra-abdominal, vanished, monorchia (intrauterine or perinatal torsion), Anorchia (if both testes are absent).
b. **'Peeping' testis:** when a previously palpable testis falls back into the abdomen through the open ring, or an intra-abdominal testis can be felt at the upper inguinal canal.
c. **Retractile testis:** is a normally descended testis that retracts into the inguinal canal due to cremasteric contraction. (Only observe and follow up).

d. **Ectopic UDT:** one that has deviated from the path of normal descent. Can be found in the inguinal region, perineum, femoral canal, penopubic area, contralateral hemiscrotum.
e. **Ascending/ acquired UDT:** a testis can no longer be brought down into the scrotum. Possible causes: Secondary to a retractile testis, change in position with growth of the child, iatrogenic after inguinal surgery.

Associated anomalies:

- PPV, epididymal abnormalities
- Specific syndromes with higher rates of UDT:
  - Prune-belly syndrome
  - Gastroschitis
  - Bladder exstrophy
  - Prader-Willi, Kallman, Noonan syndromes
  - Testicular dysgenesis
  - Androgen insensitivity syndromes.

Epidemiology:

- 3% of term male infants
- 33-45% of premature and/or birth weight <2.5 kg
- Most testes descend within the first 6-12 months, the incidence is down to 1% at 1 year of age.
- Testicular descent after 1 year is unlikely.

Diagnosis:

- History & Physical exam:
  - ✓ Retractile testis may be manipulated into the scrotum, once in scrotal position, it remains in place. Ipsilateral hemiscrotum is fully developed.
  - ✓ low UDT: may be manipulated into the scrotum, once in scrotal position it does not remain in place, ipsilateral hemiscrotum may be underdeveloped.
  - ✓ Experienced examiner has a higher sensitivity in locating UDT than does US, CT, or MRI.

Fertility:

- Some studies show that UDT and its contralateral descended testis are both histologically abnormal.
- Patients with a history of UDT showed subnormal semen analyses.
- Fertility is related to the position of the UDT (abdominal testes→ lower fertility).
- Unilateral UDT infertility rate is equivalent to normal population (10%), while bilateral UDT is 6x higher.
- Delayed orchiopexy at 3 years vs. 9 months resulted in impaired testicular catch-up growth in boys.

Risk of malignancy: 2-8 fold increased risk of malignancy

- Varies with location: 1% with inguinal testes, 5% with abdominal testes (mainly seminomas)
- Malignancies after successful orchiopexy (regardless of location) are nonseminomatous germ cell tumors.
- Among men with testicular cancer, up to 10% have a history of UDT.
- 'position theory', 'common cause' or 'testicular dysgenesis' theories.
- Orchiopexy facilitates subsequent testicular examination and cancer detection.

Management: Guidelines recommend that orchiopexy be performed by **12-18 months of age**

1. Reduces the risk malignancy, infertility, and torsion.
2. Facilitates testicular examination.
3. Improves endocrine function of the testis and creates a normal-appearing scrotum.
4. Hormonal treatment is controversial (buserelin LHRH agonist, low dose hCG)

4. Acute scrotum: Acute scrotal pain with or without swelling and erythema

- Most are nonemergent, but it's critical to differentiate between them and testicular torsion.
- Age at presentation is an important clue:
  Torsion of the appendix testis/ epididymis → prepubertal boys
  Testicular torsion → neonates and adolescents

DDx:

- torsion of the testis
- Torsion of the appendix testis/epididymis
- Epididymitis/orchitis
- Hernia/ hydrocele
- Trauma/ sexual abuse
- Tumor
- Idiopathic scrotal edema (dermatitis, insect bite)
- Cellulitis
- Vasculitis (Henoch-Schonlein purpura)

a. Testicular torsion: Results from twisting of the spermatic cord, which compromises the testicular vasculature and results in infarction. Probability of testicular salvage decline significantly beyond 6 hours.

Types:

1. **Intravaginal**: more common in children and adolescents, spermatic cord twists within the tunica vaginalis, 'bell clapper' deformity.
2. **Extravaginal:** occurs perinatally, spermatic cord twists proximal to the tunica vaginalis, the tunica and testis spin on the vascular pedicle.

Duration of torsion (hours)	Testicular salvage (%)
<6	85-97
6-12	55-85
12-24	20-80
>24	<10

Presentation:

- Sudden onset of severe unilateral testicular pain, can be intermittent (incomplete torsion with spontaneous detorsion).
- Lower thigh, or lower abdominal pain.
- Nausea and vomiting.

Prenatal torsion: hard, nontender scrotal mass noted at birth, with underlying dark skin, discoloration, and fixation of the skin to the mass.

Postnatal torsion: an acutely inflamed scrotum with erythema and tenderness. Requires emergent exploration with detorsion and bilateral fixation. (Normal scrotum at delivery).

Diagnosis:

Urinalysis revealing infectious epididymis/ orchitis doesn't rule out torsion.

- **Physical exam**: Enlarged testis, retracted up, transverse orientation, anteriorly located epididymis, severe generalized testicular tenderness, swelling and erythema, cremasteric reflex is often absent.
- **Ultrasound** with color flow doppler shows the decreased blood flow and coiling of the spermatic cord.

Management:

Manual detorsion: medial to lateral, 'open book' rotation. Not used anymore.

- **Exploration under GA**: detorsion, placement in warm moist sponges, and fixation.
- If the testis is clearly nonviable, it should be removed (orchidectomy) to avoid potential damage to the contralateral testis from the formation of antisperm antibodies.

b. Torsion of testicular appendages:

- Torsion of the appendix testis or appendix epididymis is the most common cause of an acute scrotum.
- Most commonly between ages 7 and 10 years.

Presentation:

- Sudden onset of pain and nausea
- Normal urinalysis
- Appendage can be palpated & focally tender
- 'blue dot' sign (ischemic)
- Edema and erythema

Rx: Self-limited, NSAIDs, restricted activity, and warm compress.

c. Epididymitis:

1. Bacterial epididymitis (rare in children)
   - Retrograde infection (from bladder and urethra)
   - Pain, swelling, and tenderness of the hemiscrotum.
   - A positive urinalysis and culture, or urethral swab in sexually active adolescents suggests the diagnosis.
   - Neisseria gonorrhea and Chlamydia are classically found in sexually active boys. While coliforms and Mycoplasma spp. are more likely in younger children.
   - Rx: abx

2. Viral epididymitis:
   - Mumps orchitis (rare)
   - Adenovirus, enterovirus, influenza, and parainfluenza.
   - Rx: supportive (self-limited)

d. Idiopathic scrotal edema: Scrotal swelling of unknown etiology (boys 5-9 years). Characterized by:

- Insidious onset of swelling and erythema that begins in the perineum/ inguinal region. And spreads to the hemiscrotum.
- Pruritis
- Testis is not tender (US shows normal testicular blood flow)

DDx: contact dermatitis, insect bites, minor trauma, cellulitis from an adjacent infection.

Rx: treat the underlying cause (antihistamines or topical corticosteroids).

e. Henoch-Schonlein Purpura

- A vasculitis syndrome that can involve the skin, joints, GI, and GU symptoms. Most commonly in boys younger than 7 years.
- S&S: scrotal spermatic cord pain, erythema, and swelling (in 1/3), skin purpura, joint pain, and hematuria.
- Doppler US: normal blood flow to the testis
- Rx: supportive measures, systemic corticosteroids

f. Testicular trauma: (Rare)

Dx: History (check for sexual abuse)
PEx: testis is swollen and tender, swelling and bruising of the scrotum
US: evaluate for rupture of the tunica albuginea

Rx: exploration +/- repair of the ruptured tunica albuginea

Appendix

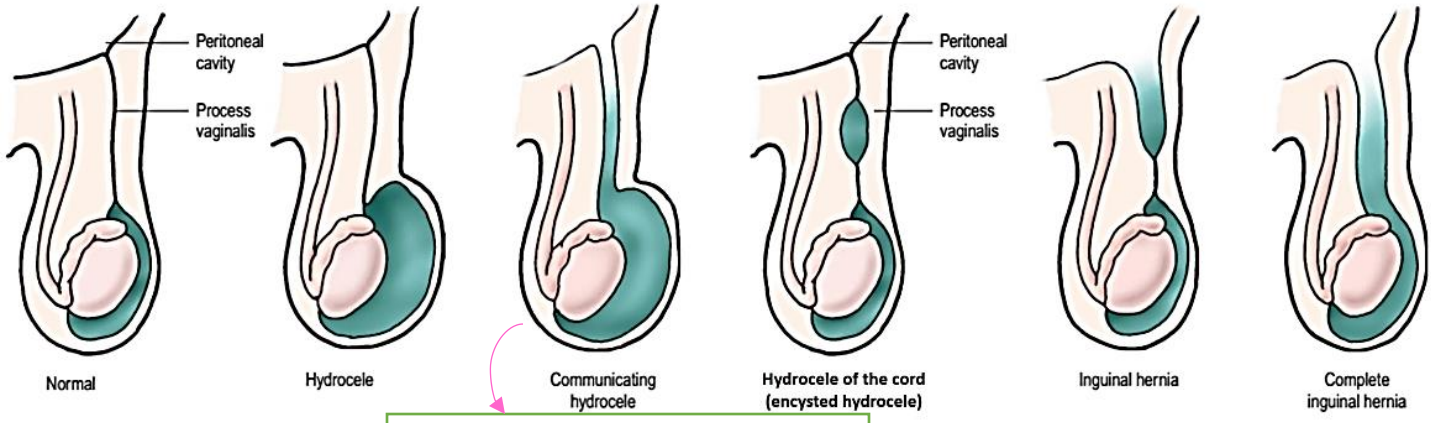
Embryology & anatomy: the **inguinal canal** is a six-sided cylinder:

1. Cephalad opening: internal inguinal ring
2. Caudal border: external inguinal ring
3. Cephalad aspect: internal oblique, transversus abdominis, and medial external oblique fibers
4. Anterior roof: external oblique aponeurosis
5. Inferior wall: inguinal ligament, lacunar ligament (medial 1/3), and iliopubic tract (lateral 1/3).
6. The floor: transversalis fascia and the 'conjoint tendon'

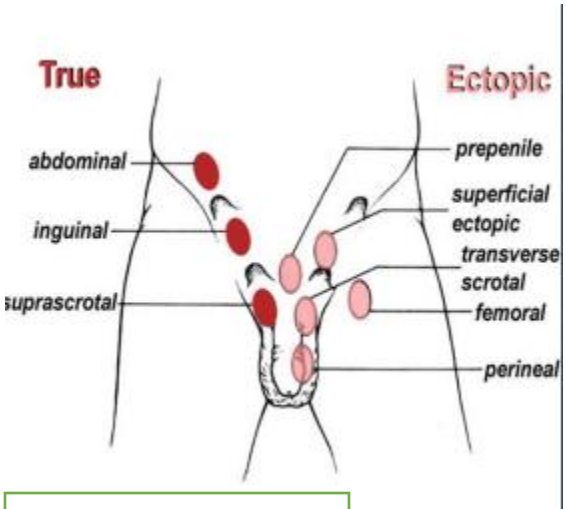
Contents: ilioinguinal nerve + spermatic cord or round ligament (in females).

- The gonads form on the anteromedial nephrogenic ridges in the retroperitoneum during the 5th week of gestation. They are attached to the scrotum by the gubernaculum in the male and to the labia via the round ligament in females.
- Processus vaginalis is a peritoneal diverticulum extending into the inguinal canal, anteromedial to the cord structures. It gradually obliterates, and the scrotal portion forms the tunica vaginalis. The female anlage of the processus vaginalis is the canal of Nuck (leads to the labia majora). Failure of the PPV to close results in an indirect inguinal hernia.

Inguinal hernias



PPV; can resolve on its own, more likely if not associated with other symptoms.

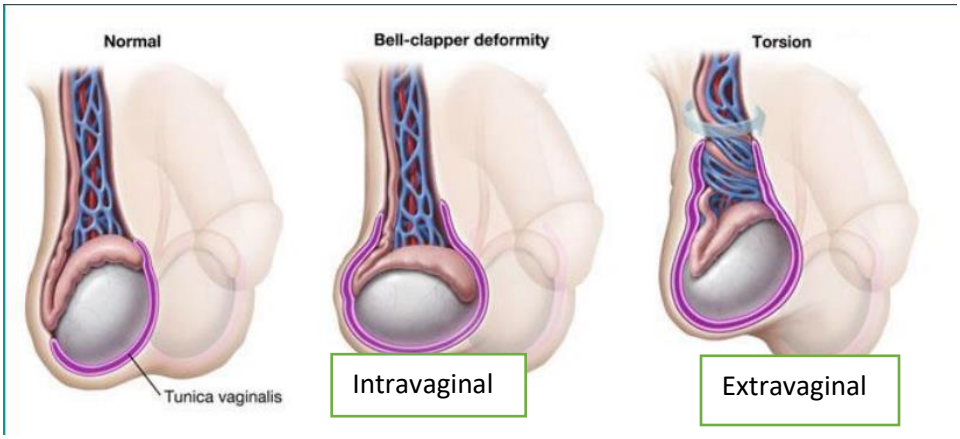


Ectopic UDT vs. true UDT

Acute scrotum: painful, red, swollen

NEVER reduce a complicated hernia; this will spread the inflammation and lead to sepsis

Types of testicular torsion



Intravaginal

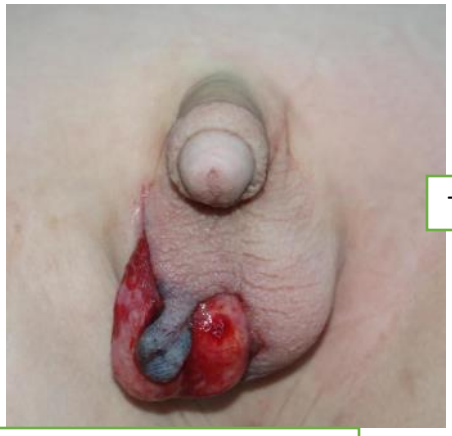
Extravaginal

Blue dot sign

Gangrenous appendage, with focal tenderness.

Ischemic appendage due to torsion

Erythematous scrotum, with bilateral edema



Exploration is needed to check for tunica albuginea rupture

Testicular trauma

Tunica vaginalis is opened; no hematoma, and no injury to the tunica albuginea.

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