

# Common Pediatric Scrotal Conditions

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## Last Updated:

Friday, February 24, 2023

## 1. Introduction

This chapter will introduce common scrotal anomalies that pediatric urologists will evaluate in their practice. The goal is to introduce the most common conditions seen in the outpatient setting as well as raise attention to common differential diagnoses for the acute scrotum.

### 1.1 Key Words:

**Testicular torsion, Torsion of appendix testis, Epididymitis, Orchitis, Epididymal cyst, Hernia, Hydrocele, Varicocele**

## 2. Testicular torsion

Testicular torsion is an emergent surgical condition caused by lack of blood flow to the testicle secondary to a twist in the spermatic cord. Males are affected in a bimodal distribution with peaks during the neonatal period and again around puberty. In the perinatal period, the entire spermatic cord, testis and tunica vaginalis twist as a unit within the scrotum and this is referred to as extravaginal torsion (**Figure 1**). In contrast, older children who experience testicular torsion often have an anatomic variation known as the ‘bell-clapper’ deformity, in which there is a high insertion of the tunica vaginalis on the spermatic cord. The tunica vaginalis completely encircles the testis, epididymis and distal spermatic cord, rather than attaching in the normal, posterolateral location within the cord. In this type, the testis and the cord twist within the tunica vaginalis and this has been termed intravaginal torsion (**Figure 2**). Streamlined clinical evaluation and time-critical re-establishment of blood flow to the testis are keys to minimize the extent of ischemic damage.

Classically, testicular torsion presents with acute onset of unilateral scrotal pain. As time is of the essence, accuracy of diagnosis is germane to scoring systems such as the Testicular Workup for Ischemia and Suspected Torsion (TWIST). This model seeks to determine the likelihood of torsion based on the additional presenting symptoms. Points are assigned based on findings of: testicular swelling (two points), a hard testicle (two points), an absent cremasteric reflex (one point), nausea and/or vomiting (one point), and a high-riding testis (one point).<sup>1</sup> This scoring system has been prospectively validated, with one group delineating that a score of seven or higher was associated

with a high-risk of having torsion with 100% specificity and 100% positive predictive value.<sup>2</sup> Neonatal torsion is typically noticed on physical exam after birth, as boys present with a firm, enlarged testicle, often with discoloration of the hemiscrotum (**Figure 3**).

Testicular torsion is primarily a clinical diagnosis. If history and physical exam strongly suggest testicular torsion, no further workup is necessary and scrotal exploration should be pursued without delay. In cases where the diagnosis is unclear, color Doppler ultrasound can be obtained. Torsion is diagnosed by a lack of blood flow within the testis (**Figure 4**), and sensitivity and specificity of ultrasound in diagnosing torsion is 94% and 98%, respectively.<sup>3</sup> Ultrasound may visualize the “whirlpool sign,” a spiral/whorl-shaped area along the spermatic cord that signifies a twist of the cord structures. In one study, the whirlpool sign was present in 96% of children who had torsion on scrotal exploration,<sup>4</sup> while a meta-analysis documented 73% sensitivity and 99% specificity.<sup>5</sup>

Testicular salvage (excluding perinatal torsion) is more likely to be achieved when scrotal exploration and detorsion are performed early after loss of flow (acute onset of symptoms). A systematic review of 30 articles including over 2,000 patients cites a testicular salvage rate of 97.2% within six hours of symptom onset, and dropping with each six-hour interval thereafter, to a rate of only 24.4% at 25-48 hours and only 7.4% at > 48 hours.<sup>6</sup> Despite those testicles “successfully” salvaged (detorsed and paxed), long-term viability has been reported around 50%.<sup>7</sup> Detorsion of the testis, with subsequent reperfusion and edema, has been described as a type of compartment syndrome that can lead to additional parenchymal injury. As such, some have advocated for incision of the tunica albuginea and placement of a tunica vaginalis flap to decompress the compartment with sustained release of pressure. Chu et al reported on a retrospective matched cohort study comparing those with incision and flap versus orchiectomy alone. These authors noted that in patients undergoing incision and tunica vaginalis flap, the postoperative viability was 95% in those with ischemia times < 24 hours compared to only 67% with ischemia times > 24 hours. Moreover, atrophy rates were high in these groups, occurring in 67% of those with less than 24 hours of ischemia time versus 85% of those with more than 24 hours of ischemia.<sup>8</sup> Nonetheless, the tunica vaginalis flap was considered a valid alternative to orchiectomy.

Perinatal torsion has more dismal outcomes, with salvage rates ranging from approximately 9-22%.<sup>9</sup> One meta-analysis of 196 neonates found an overall salvage rate of only 7%, with 4% of explorations potentially preventing asynchronous torsion.<sup>10</sup> Risks of anesthesia in a neonate must be considered. The experience of the entire team is an important safety consideration prior to proceeding to the operating room in these specific cases. A recent survey of pediatric urologists demonstrated that management remains controversial with respect to the decision to surgically intervene in these cases or not, and if so, whether to do so with immediate vs. urgent vs. elective surgery; however, a suspected postnatal testicular torsion event warrants emergent surgical exploration, as does bilateral testicular torsion in a neonate.<sup>11</sup>



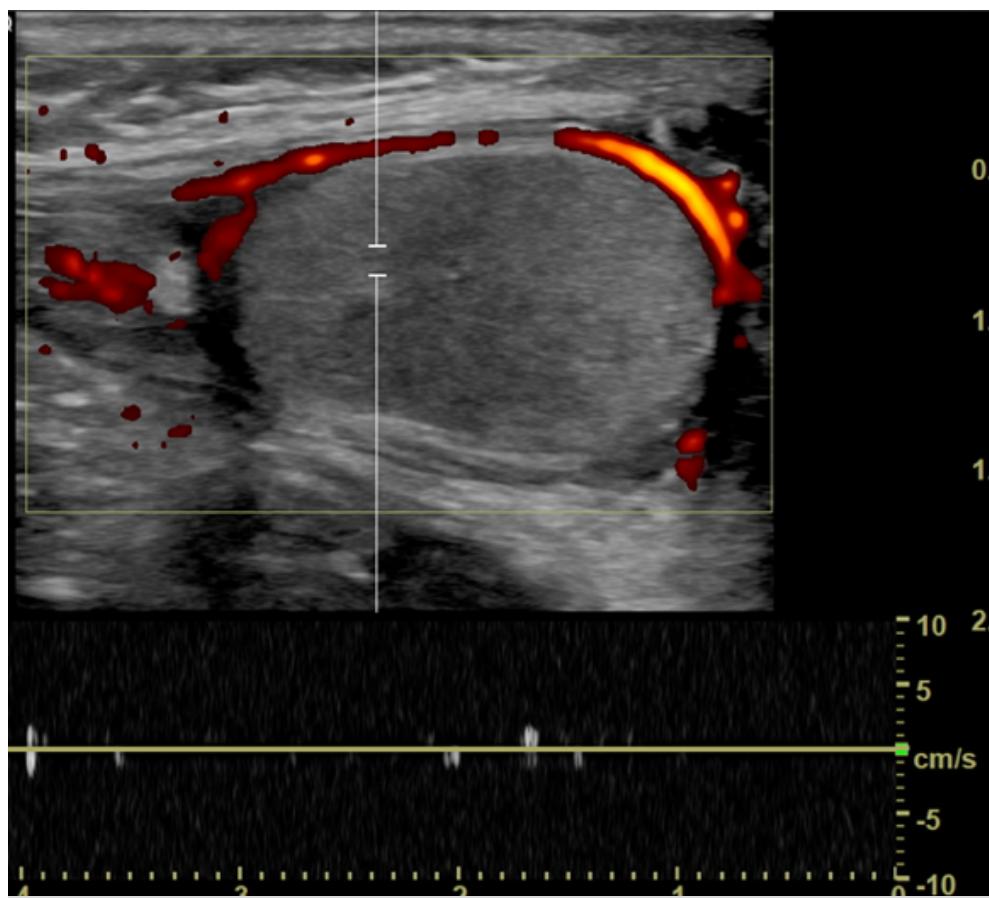
**Figure 1:** Extravaginal torsion at surgical exploration.



**Figure 2:** Intravaginal torsion at surgical exploration.



**Figure 3:** Discoloration of scrotal skin secondary to perinatal testicular torsion.



**Figure 4:** Color Doppler showing absence of flow within a torsed testicle.

### 3. Torsion of appendix testis or appendix epididymis

The appendix testis is a vestigial structure (cranial end of the Mullerian remnant) located on the superior pole of the testis. The appendix epididymis persists as the cranial end of the Wolffian remnant, attached to the head of the epididymis. Either of these can twist upon its attachment (**Figure 5**), leading to gradual onset of pain, as opposed to the acute onset of pain typically encountered in testicular torsion. Pain symptoms often proceed through a crescendo decrescendo over several days. Children with a torsed appendage are usually younger than those with testicular torsion, with age < 12 years reported as a useful predictor of a torsed appendage.<sup>12,13</sup> The cremasteric reflex is typically preserved with torsion of an appendage. A firm, tender, discrete nodule may be palpable in the superior aspect of the testis, and in some cases the hemorrhagic appendage has a bluish discoloration, visible through the skin as a “blue dot sign.”

As this is sometimes difficult to discern from testicular torsion, scrotal ultrasound may be performed during evaluation. The torsed appendage may appear as a hypoechoic mass in the superior pole, and with color Doppler there may be decreased or absent blood flow within the mass itself, with surrounding hyperemia extending throughout the epididymis consistent with ultrasound findings commonly seen in epididymitis. As such, urinalysis should be obtained to rule out infection. If negative, conservative treatment should be undertaken to include scrotal rest (restriction of physical activity) and NSAIDs. Depending on the timing of presentation during the course, patients may need to be counseled to expect that symptoms may get worse before they get better. Improvement is generally seen in approximately a week.



**Figure 5:** Torsed appendix testis.

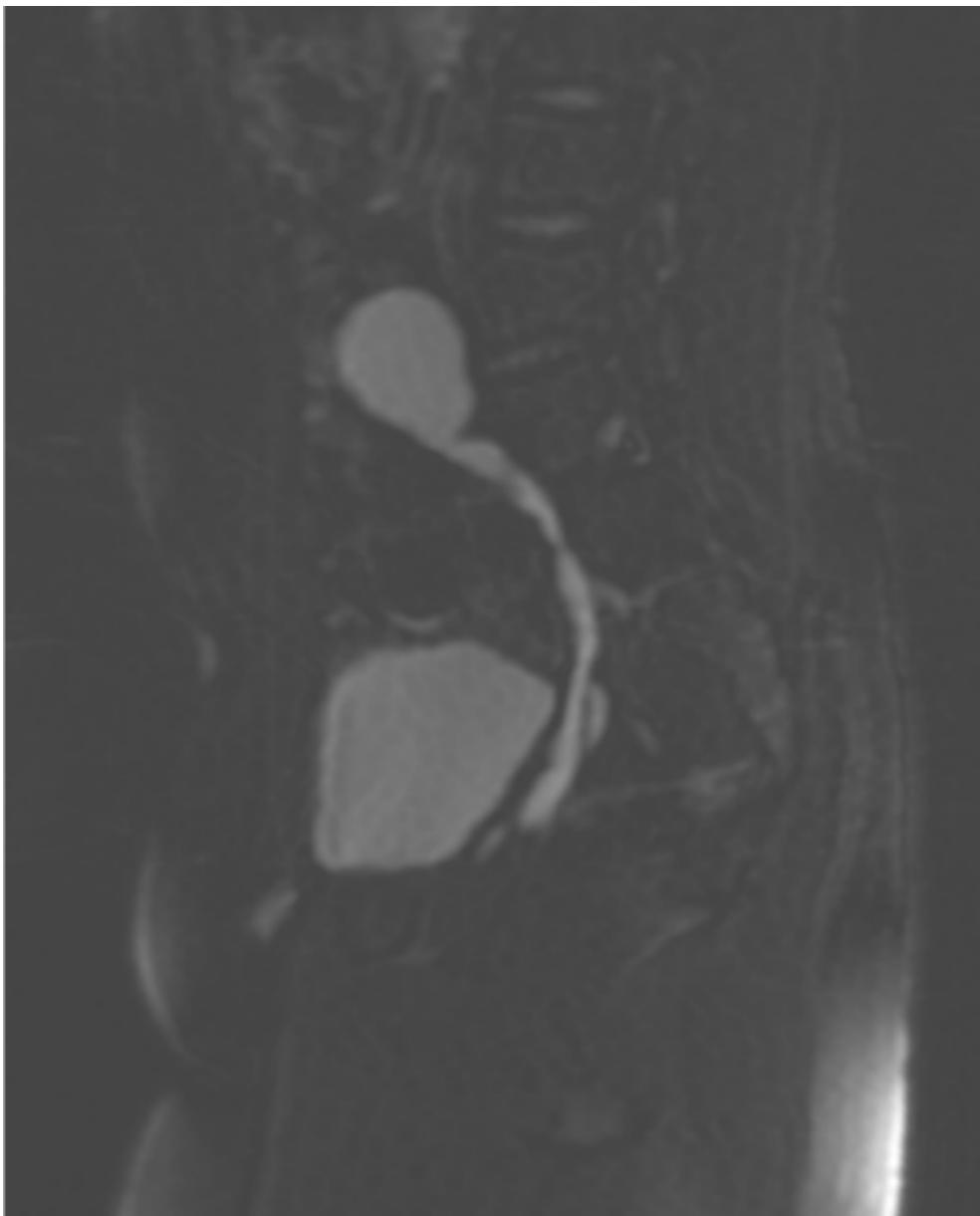
### 4. Epididymitis / orchitis

Epididymitis (inflammation of the epididymis) and orchitis (inflammation of the testicle) in the pediatric

population may be due to infectious or noninfectious causes. In prepubertal boys, infectious etiologies are rare, and the cause is often a torsed appendix testis/epididymis; in sexually active postpubertal young men, infectious etiologies including sexually transmitted diseases are much more common. Urinalysis and culture must be obtained, and antibiotics prescribed if infection is suspected based on test results. Bacterial sensitivities are required as empiric antibiotic therapy may not cover the responsible bacteria.<sup>14</sup> If urine culture is positive further workup should be performed, including a renal/bladder ultrasound and consideration for a voiding cystourethrogram. Infants and children with recurrent episodes of epididymitis, or those with a history of any genitourinary anomaly should undergo evaluation for structural abnormalities, such as a prostatic utricle, posterior urethral valves, or ectopic ureteral insertion into the vas deferens, epididymis, or seminal vesicle (**Figure 6** and **Figure 7**).<sup>15,16</sup>

Nonbacterial epididymitis may be “chemical” in nature, with inflammation secondary to uncoordinated voiding against a closed sphincter, causing reflux of urine through the ejaculatory ducts into the vas deferens. This may be evidenced in one’s history by holding maneuvers, infrequent voiding, urinary urgency, and/or daytime urinary incontinence; these children should be placed on a timed voiding schedule. In the acute setting, analgesics, NSAIDs, scrotal rest, and a tincture of time are warranted. Some patients may benefit from alpha blockers to address detrusor sphincter dyssynergia that can exist.

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**Figure 6:** Ectopic insertion (arrow) of nonfunctioning atrophic kidney (asterisk) into the seminal vesicle in an 8-year-old boy with recurrent epididymitis.



**Figure 7:** Ectopic insertion (arrow) of nonfunctioning atrophic

kidney (asterisk) into the seminal vesicle in an 8-year-old boy with recurrent epididymitis.

## 5. Acute idiopathic scrotal edema

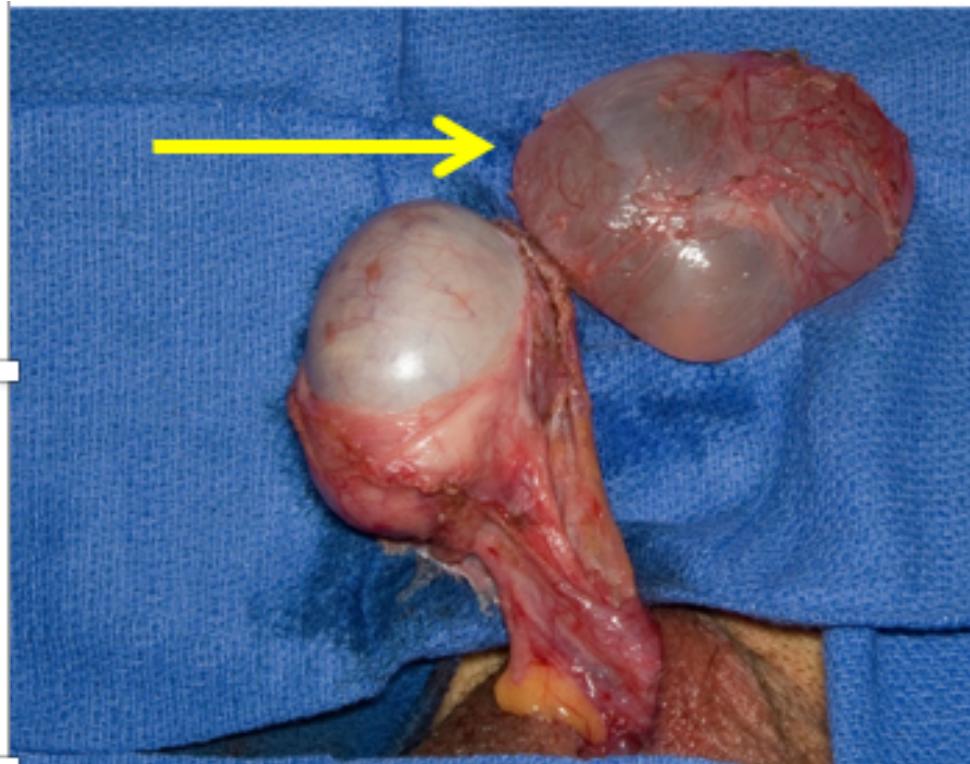
Acute onset of scrotal edema in children, with or without pain or tenderness, is frequently idiopathic in nature and treated conservatively with limiting physical activity, scrotal elevation, and if needed, analgesics. If the testes cannot be easily palpated, scrotal color Doppler ultrasound should be performed to rule out testicular mass or other intrascrotal pathology.

Henoch-Schonlein purpura is an IgA-driven systemic vasculitis in children presenting with non-thrombocytopenic purpura, arthralgias, gastrointestinal bleeding and abdominal pain. Genitourinary organ involvement is common, including acute onset of scrotal swelling, erythema and pain. This entity is self-limited and managed supportively and may include NSAIDs or steroids to alleviate pain.<sup>17</sup>

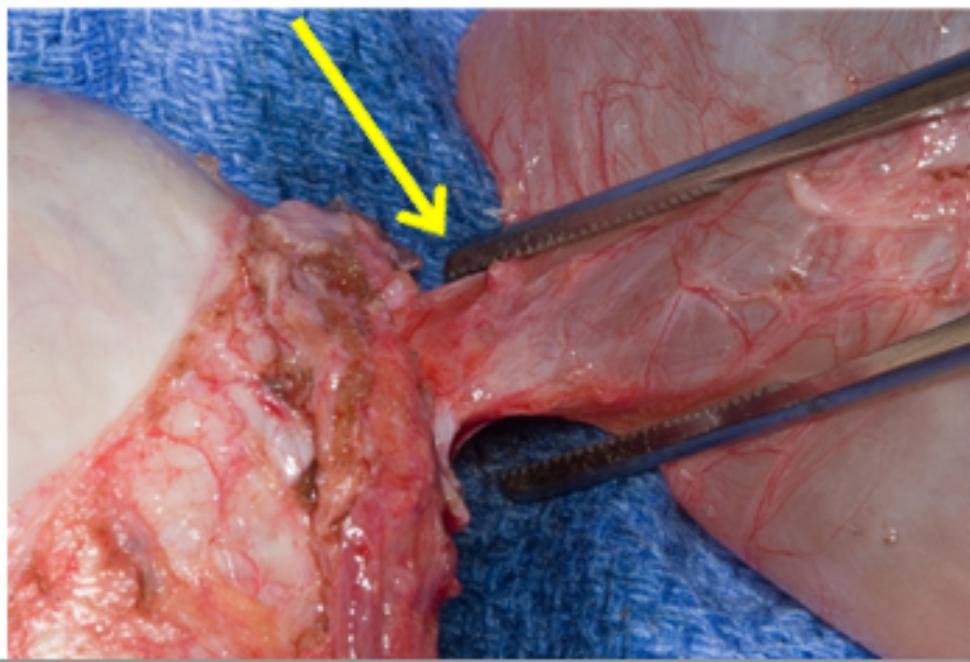
Acute idiopathic scrotal edema may be the presenting symptom of a systemic disease process. Barrick et al reported on a series of patients with penile and scrotal swelling who were subsequently diagnosed to have the cutaneous manifestation of Crohn's disease (CD), or metastatic Crohn's disease. In their cohort, 88% had cutaneous symptoms before their diagnosis of Crohn's, with gastrointestinal symptoms variably present. Skin biopsy was diagnostic of CD in 100%, and treatment involves that of the underlying disease.<sup>18</sup>

## 6. Epididymal cysts

Epididymal cysts, also known as spermatoceles, in the pediatric population may be found incidentally on ultrasound or present as an enlarging and/or painful mass on physical exam. Conservative management in the form of observation is recommended if the spermatocele is small and/or asymptomatic especially since 30% of males after puberty will have small epididymal cysts. One study reported complete involution in 29% of affected individuals over a median 11.2 months (1-37 months), and a decrease in cyst size in an additional 41% of these children.<sup>19</sup> Surgical excision of the cyst (**Figure 8** and **Figure 9**) through a transscrotal approach may be pursued in those with an enlarging cyst or those with symptoms of pain, understanding that pain may persist post-operatively. Complete resection of the cyst and ligation of the stalk is necessary to prevent recurrence. Patients and families should be counseled specifically on the risk of iatrogenic infertility as a complication of surgical intervention in these cases.



**Figure 8:** Epididymal cyst arising from the head of the epididymis.



**Figure 9:** Dissection of the cyst is carried out with the cyst intact, isolating the neck which is tied off, with the cyst then removed.

## 7. Scrotal median raphe cysts

Cysts that appear along the median raphe extending from the perineum to the urethral meatus are typically noticed at birth or go unrecognized until later in childhood (**Figure 10**). They are thought to form as aberrant cells which are trapped during fusion of the urethral folds, or later in development as

outgrowths of epithelium after primary closure.<sup>20</sup> These lesions are typically asymptomatic in childhood, may grow in size with age, and become symptomatic if infected. Simple excision with primary closure is recommended (as opposed to aspiration or marsupialization) to avoid post-operative complications.



**Figure 10:** Scrotal median raphe cysts noted in a newborn.

## 8. Hernia/hydrocele

A patent processus vaginalis that persists beyond infancy can present clinically with an enlarged scrotum (bilateral) or hemiscrotum (**Figure 11**), with an overall incidence of approximately 1-4% but up to 30% in premature infants.<sup>21</sup> Fluid in this space is termed a hydrocele. A history of fluctuation in the size of the scrotum is consistent with dynamic fluid shifts seen in a communicating hydrocele, while a static enlargement of the scrotum generally corresponds to fluid that is trapped within the scrotum behind a closed processus vaginalis (non-communicating hydrocele). In newborns, the latter typically resolves spontaneously and does not warrant surgical intervention; observation is recommended, and timing of recommended repair is debatable as many may still resolve 24-36 months after birth.<sup>22</sup> Fluid may also be trapped along the spermatic cord in what is termed a cord hydrocele. Hydroceles that occur after minor trauma may resolve spontaneously and a trial of observation for several weeks is advised.

In addition to peritoneal fluid, intraabdominal structures may herniate through the inguinal canal. If

visceral herniation is suspected, repair should be performed in infancy to prevent incarceration or strangulation which may result in obstruction and necrosis of bowel. Ultrasound may be performed to document structures within the inguinal canal and scrotum, particularly if the testes are not palpable. The surgical approach to the pediatric patent processus vaginalis has historically been through an open inguinal incision to identify and tie off the proximal end of the processus vaginalis at the level of the internal inguinal ring; this has been termed a “high ligation.” Laparoscopic techniques in children have been introduced more recently, and have evolved from a standard 3-port laparoscopic hernia repair to single-site laparoscopy and percutaneous, needlescopic approaches. In a review of 90 studies of open and laparoscopic hernia repairs, there was no significant difference in operative time in open vs laparoscopic approaches for unilateral repair, but time was significantly shorter with laparoscopy for bilateral repair (46.1 min vs 30.9 min, p=0.01). Recurrence rates were similar between the two techniques (0-6% for open, 0-5.5% for laparoscopic, p=0.66) but complication rates (wound infection, testicular atrophy, hydrocele) were statistically significantly lower in the laparoscopic group (2.7% for open, 0.9% for laparoscopic, p=0.001).<sup>23</sup> Bladder injury has also been described in the setting of pediatric inguinal hernia repair. These complications can be severe and, in many cases, require additional surgery and adversely affect bladder capacity. Surgeons should be aware of these potential “bladder ears” that may protrude through the internal inguinal ring and be encountered during dissection of hernias.<sup>24</sup>

Contralateral exploration at the time of unilateral hernia/hydrocele repair is controversial as it is not without risk. Moreover, a patent internal ring does not commonly result in a clinically apparent hernia/hydrocele. The risk of developing a metachronous contralateral hernia after unilateral repair is reported at approximately 7%.<sup>25</sup> The risk of developing a metachronous contralateral hernia after negative laparoscopic evaluation of the contralateral ring for a patent processus vaginalis is reported at 1.31%.<sup>26</sup>



**Figure 11:** Communicating right hydrocele in a 1-month old baby.

## 9. Varicocele

A varicocele is an abnormal dilation of the pampiniform venous plexus of the testicle. Its etiology is primarily related to the drainage pattern of the testicular vein and the associated hydrostatic pressure within this venous plexus. A significant majority of varicoceles are left-sided. This predisposition is thought to be due to differences in the venous insertion between the left and right testicular veins (right-angle vs. oblique angle, respectively), the relative greater length of the left testicular vein compared to the right, and the decreased flow of the left renal vein when compared to that of the inferior vena cava. These factors increase the relative hydrostatic pressure within the left testicular vein and its associated pampiniform plexus.<sup>27</sup> A varicocele results when valves within the testicular vein become incompetent in response to this increased pressure. Extrinsic compression of the testicular vein within the retroperitoneum, possibly related to a mass, is another etiology of varicocele.

(although this is much less commonly encountered).

Varicoceles are rare in the pre-adolescent population. Prevalence increases with pubertal development and, as adolescence progresses, approaches the typical rate seen within adults (15%).<sup>28</sup> Patients may be asymptomatic or may present with pain and/or infertility. The pain is classically described as a dull ache within the affected testicle. The inability to identify infertility in the pediatric aged population has resulted in significant debate regarding optimal management of pediatric varicoceles.

Physical exam should be performed with the patient both standing and supine. A typical grading scheme is represented by Grade 1 (varicocele palpable only with the patient standing and performing a Valsalva maneuver), Grade 2 (varicocele palpable with the patient standing and breathing normally), Grade 3 (varicocele is visible through the scrotal skin when standing). Investigation for a retroperitoneal mass should be considered for a varicocele that remains palpable when the patient is supine and for an isolated right sided varicocele.

The significant majority of children with a varicocele are asymptomatic. The evaluation has therefore evolved to focus on identification of factors that may predict infertility. Factors that have been studied include varicocele grade, testicular volume by orchidometry, testicular volume by ultrasound, testicular volume differential/atrophy index, and peak retrograde flow.<sup>29,30,31,32,33,34</sup> Various endocrine markers have also been investigated, including gonadotropin releasing hormone stimulation, inhibin B, and anti-Mullerian hormone, among others; but there is currently no clear role for hormonal evaluation in this population.<sup>35,36,37</sup> Semen analysis also offers another potential lens through which to view the associated risk of a varicocele.<sup>38</sup> Studies of this diagnosis in this specific aged population are difficult due to the large lead-time between diagnosis of the varicocele and any future demonstrated infertility. As such, management has failed to consolidate around a standard evaluation of these patients and much debate has ensued over disparate results between researchers' results in this area. While no clear consensus exists, pain and/or a differential testicular volume of 15% to 20% have been used by many institutions as indications for treatment, or at least as a cause for increased concern.

Ultimately, paternity is the gold-standard outcome measure for the asymptomatic adolescent with a varicocele. Few studies have adequately examined this question. Bogaert et al. from Belgium surveyed 661 men, of whom 361 responded to the survey, and of whom 158 desired paternity. All of these men surveyed had a varicocele identified during puberty with 372 undergoing treatment and 289 electing surveillance/observation. Of those desiring paternity, no significant differences in achieving paternity were seen between these groups. Paternity was achieved in 85% of men who were observed and 78% of men who underwent active treatment earlier in life.<sup>39</sup>

Management strategies range from surveillance to surgical intervention. Chu et al found that 47% of Tanner V boys with an untreated varicocele and with an initial total motile count (TMC) of < 20 million, would demonstrate improvements in TMC to normal levels (above 20 million) with observation alone. These data support the hypothesis that there is at least a subset of these patients

who can be safely surveilled.<sup>40</sup> When surgical intervention is elected there are several approaches that have been described: open (Palomo technique), laparoscopic, microsurgical (inguinal or subinguinal), and endovascular. The open technique typically involves ligating the testicular artery and internal spermatic vein, comfortably cephalad to the vas deferens and deferential artery (an artery preserving modification has also been described). Feber reported on outcomes in 233 adolescent boys who underwent the open Palomo repair. Post-operative hydrocele was identified in 29% and a persistent varicocele was found in 3.9%.<sup>41</sup> The laparoscopic approach is modeled after the open approach and has demonstrated similar results as its open ancestor. Modifications have led to a reduction in the post-operative hydrocele rate in some series.<sup>42</sup> As with the open approach, concern remains regarding potential testicular atrophy secondary to testicular artery ligation. The microsurgical approach is generally recognized as the preferred approach in adults; in children, this procedure has similarly been associated with low rates (below 3%) for both secondary hydrocele and recurrent/persistent varicocele.<sup>43</sup> In this approach, the testicular artery can be prospectively identified with the aid of magnification and with use of a doppler probe, thus lowering concern for testicular atrophy. Recurrence rates following endovascular treatment have been high but seem to be improving as technique evolves. This approach provides a good alternative for recurrent varicoceles following any of the aforementioned surgical interventions.

## 10. Other scrotal conditions

The differential diagnosis for scrotal complaints would not be complete without reference to **scrotal and testicular trauma** and **testicular mass/malignancy**. These topics are addressed in more detail elsewhere within the AUA core curriculum.

## Presentations

Common Pediatric Scrotal Conditions Presentation 1

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