2014 was a momentous year for the Society for Fetal Urology (SFU). The SFU participated in drafting a manuscript entitled *Multidisciplinary Consensus on the Classification of Prenatal and Postnatal Urinary Tract Dilation (UTD Classification System)*. J Pediatr Urol (2014) 10, 982-999.

Under the leadership of Dr. Hiep T. Nguyen, representatives from a number of societies with an interest in the pre- and postnatal evaluation and management of patients with antenatally-detected urinary tract dilation (UTD) assembled at the AUA Headquarters in Linthicum, MD to establish/propose 1) a unified description of UTD that could be applied both pre- and postnatally and 2) a classification system for the pre- and postnatal evaluation of patients with UTD based on sonographic criteria.

The UTD Classification System was presented at the Joint Session with the SFU, held in conjunction with the Society for Pediatric Urology Annual Meeting in Orlando, FL, by Dr. Nguyen. In addition, Dr. C. D. Anthony Herndon (University of Virginia) put together an outstanding panel discussion entitled *Management of Ureteroceles and Duplication Anomalies*. The panelists were Drs. Sean T. Corbett, Michael P. Leonard, and Aseem R. Shukla.

The fall meeting was held in conjunction with the Pediatric Urology Fall Congress in Miami, FL. The program was entitled *Primary Obstructive Megaureter: Surgical Options and Long-term Follow-up* and was chaired by Dr. Luis Braga (McMaster University). We were treated to a number of outstanding guest lectures (listed) and abstract presentations, including case reports, case series, and original research.

- Tapered Ureteral Reimplantation for Primary Obstructive Megaureter: Intra- or Extra-vesical Technique; Pramod P. Reddy, MD
- Endoscopic and Robotic Options for Management of Primary;Obstructive Megaureter: Stenting vs Robotic Tapered Ureteral Reimplantation; Pasquale Casale, MD
- Refluxing Ureteral Reimplantation for Primary Obstructive Megaureter: A Temporary or Definitive Solution?; Martin Kaefer, MD
- Primary Megaureter: Long-term Follow-up and Complications; Anthony A. Caldamone, MD
- British Association of Pediatric Urologists (BAPS) Consensus on Management of the Primary Obstructive Megaureter; Marie-Klaire Farrugia MD

Many of the abstracts presented at the fall meeting are included in this publication. The SFU remains indebted to Dialogues in Pediatric Urology and the Society for Pediatric Urology for their continuing support of our group.

As we look ahead to 2015, we anticipate continued growth, productive collaborations, and stimulating exchanges as we seek to improve the care of children affected with congenital genitourinary anomalies. As always, we welcome comments or suggestions regarding current or future projects, meeting topics, and the like.

Additional information can be found on the SFU website (www.sfu-urology.org).
Severe Renal Functional Loss in a Patient with Recurrent Hydronephrosis Following Improvement of Mild Antenatal Hydronephrosis

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Case Presentation

A 3-year-old male with a history of resolved left-sided antenatal hydronephrosis presented to the pediatric urology clinic with left flank pain and vomiting. Past medical history included antenatal hydronephrosis, meconium aspiration requiring NICU admission, intussusception requiring partial colectomy and colostomy formation at 5 months of age, with reversal 2 months later. He was followed postnatally by nephrology for Society for Fetal Urology (SFU) grade 1 left hydronephrosis [Figure 1a] with a negative voiding cystourethrogram. He had one urinary tract infection at 3 months of age. Although repeat renal ultrasound (RUS) revealed slight progression of hydronephrosis at 7 months, a diuretic renogram at that time showed symmetric differential function and spontaneous drainage following diuretic administration. Renal ultrasound at 15 months showed improvement with minimal residual hydronephrosis. [Figure 1b] The patient had no discernible symptoms or further infections and was discharged from follow-up at this time.

Upon presentation to the pediatric urology clinic at 36 months of age, his mother noted a 4-month history of vague episodic flank pain and vomiting. These symptoms prompted a RUS demonstrating SFU grade 4 left-sided hydronephrosis. [Figure 1c]. A diuretic renogram showed 10.6% split function of the left kidney and no washout of radiotracer. After the family was counseled on management options including nephrectomy, he underwent robotic-assisted laparoscopic pyeloplasty one month later. A crossing vessel was not found intraoperatively. At 15 month post-operative follow-up, he remained symptom free with a RUS showing complete resolution and 14% differential function on renal scan. [Figure 2]

Discussion

Most cases of antenatal hydronephrosis will spontaneously resolve, however the optimal algorithm for how to follow these patients is unclear. Recommendations range from continuing serial ultrasounds until patients are able to communicate, to cessation of surveillance once the hydronephrosis resolves with appropriate parental education about re-evaluation if the patient becomes symptomatic.1,2

Our case illustrates a rare event in which mild resolved hydronephrosis recurred and progressed to symptomatic severe hydronephrosis, and a documented significant loss of renal function. A retrospective review of 344 cases found 4 patients (1%) who recurred with symptoms at a mean of 40 months of age, however each patient in their series had differential renal function greater than 45%.1 In contrast, our patient with initial SFU grade 1 hydronephrosis subsequently progressed to severe hydronephrosis with only 10% differential renal function at 36 months of age after approximately 4 months of symptoms. This case report raises many questions including whether routine surveillance for patients with resolved hydronephrosis should be more stringent, whether initial grade should play a role in length of postnatal surveillance, and if parental education is in fact enough to avoid future cases of renal function loss. As such, we advocate following any grade of hydronephrosis until the patient can reliably vocalize symptoms.

References

An Unusual Presentation of Anterior Urethral Valves

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Case Presentation
A newborn male was born at 33 weeks due to pre-term labor. The neonate had been followed prenatally with serial ultrasounds, which demonstrated severe bilateral hydronephrosis, a dilated proximal urethra, and a thick-walled bladder (Figure 1). The mother’s amniotic fluid index was normal throughout pregnancy until a significant decrease occurred several days prior to birth at which time significant fetal ascites was also seen.

Once delivered the neonate was limp with no respiratory effort and was immediately intubated. His abdomen was massively distended and an 8 Fr bladder catheter was placed. Emergency paracentesis was performed and 150 mL of clear, yellow fluid confirmed to be urine by chemistry evaluation was aspirated. A renal ultrasound performed on day of life 0 demonstrated prominent bladder wall thickening, ascites, and improved but persistent mild bilateral hydronephrosis (Figure 2). A VCUG was performed once the child was stable and demonstrated a discrete segment of narrowing in the proximal penile urethra without evidence of a diverticulum or significant proximal urethral dilation. No VUR or extravasation of contrast was seen. Cystourethroscopy was subsequently performed and confirmed an anterior urethral valve at the penoscrotal junction, which was successfully ablated with a pediatric resectoscope. Following ablation, the child has done well on follow-up with resolution of his hydronephrosis and has normal renal and bladder function.

Discussion
Congenital obstruction of the anterior urethra is rare and occurs much less frequently than posterior urethral valves (PUV).1 Congenital anterior urethral obstruction presents as two distinct findings: either as a discrete obstructive fold or valve (anterior urethral valve – AUV), or as a diverticulum (anterior urethral diverticulum – AUD) which fills with voiding and obstructs the lumen of the urethra. Similar to PUV, the clinical presentation of a boy with AUV or AUD can vary widely. However, the degree of urethral obstruction in patients with AUV or AUD is thought to be less severe as compared to PUV, which likely plays a role in the generally accepted better prognosis in boys with anterior urethral obstruction.2

It is well known that PUV or urethral atresia can cause significant bladder outlet obstruction which in some cases leads to bladder and/or fornical rupture with resultant urinary ascites in the fetus. However, severe antenatal bladder outlet obstruction is rarely seen in boys with AUV, and to our knowledge has been reported only once previously.3 Following diagnosis of the AUV in our patient and appropriate endoscopic treatment, both his renal and bladder function have been normal on follow-up. Our experience mirrors the outcome of the patient in the previously published case report.

In conclusion, it is important that adequate evaluation of the entire urethra be performed in newborn males with suspected bladder outlet obstruction so that an obstructive lesion in the anterior urethra is not missed.

References
High-Volume Urinary Retention in a Two Month Old Boy: An Unexpected Sequela of a Large Urachal Remnant

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Case Presentation

If the urachus fails to obliterate during development, cystic structures can form at any point along its course. Of the four described anomalies, patent urachus is the most common (50% incidence), followed by urachal cyst (30%), urachal sinus (15%), and urachal diverticula (5%). The most common presenting complaint is umbilical discharge, followed by abdominal mass.

In this case, Pediatric Urology was consulted for a male fetus with a “large bilobed cystic mass in the anterior aspect of the abdomen,” detected on routine ultrasound at week 18 of pregnancy. Further imaging at 33 weeks revealed a 8 x 6 x 7 cm mass superior to and communicating with the bladder. The kidneys and external genitalia were normal-appearing, and the ureters could not be visualized (figure 1).

The baby was born at 37 weeks after an uncomplicated delivery and voided 6 hours later. Postnatal renal-bladder ultrasound and VCUG revealed a patent urachal remnant with a large urachal cyst communicating with both the bladder and umbilicus. There was no VUR or urethral abnormality, though it was noted that the urachal remnant did not empty well with voiding. Circumcision was performed and amoxicillin prophylaxis initiated. It was planned that the child would undergo excision at 6 months of age.

Following discharge from the hospital, the child initially did well. However, at 8 weeks of age, he had increasing abdominal girth with poor PO intake. Investigation revealed closure of his urachal sinus and substantial interval growth of his communicating urachal cyst, measuring 13 x 10 x 11 cm on ultrasound, essentially filling the entire abdominal cavity. Urethral catheter placement drained 500 mL of urine.

Over the subsequent days, the catheter fell out at home. The child once again developed high-volume urinary retention. Ultimately given his symptomatic retention, he underwent partial cystectomy and excision of urachal remnant at three months of age, one week after his second retention episode. The child had an uneventful post-operative course. His catheter was removed postoperative day 5, and he has voided without difficulty since then. Follow-up ultrasound is pending.

Discussion

Urinary retention in infants is rare, and we are unaware of prior reports of retention due to a high-volume urachal remnant in a child. It is certainly plausible that urine was trapped in the narrow-necked cyst during voiding, which would then refill the bladder, leading to a cycle of chronically elevated bladder volumes and ineffective emptying. This case sheds light on instances in which prompt surgical intervention on an urachal remnant is warranted, even in very young infants.

References

Ectopically Inserting Obstructed Megaureter: Application and Utility of Magnetic Resonance Urography

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Case Presentation
An asymptomatic 4-week-old male infant presented to the pediatric urology clinic with hydronephrosis, initially diagnosed on prenatal ultrasound. He was born full term and healthy, with an unremarkable birth history. A renal ultrasound (RUS) was performed at 5 weeks, which revealed Society for Fetal Urology (SFU) grade 4 right-sided hydronephrosis and a severely dilated (19 mm) right ureter. The insertion point of the dilated right ureter was not apparent on ultrasound. A subsequent voiding cystourethrogram revealed no evidence of vesicoureteral reflux. A technetium-99m mercaptoacetyltriglycine (MAG3) renogram with Lasix was then performed, which revealed a normal left kidney and a dilated, poorly draining right collecting system. To better assess the anatomy a magnetic resonance urogram (MRU) was performed, which revealed a dilated ectopic right ureter traveling posterior to the trigone with insertion into the right seminal vesicle (Figure 1). Interim management options prior to possible reimplantation, including cutaneous ureterostomy to relieve obstruction and serial RUS to monitor for renal parenchymal loss, were offered. The parents elected to monitor with RUS with potential surgical intervention at a later date.

Discussion
Primary obstructed megaureters typically represent a functional obstruction, and are thought to have an adynamic, aperistaltic, juxtavesicular section. Histologically, there is increased collagen deposition and hypertrophy of outer non-peristaltic muscle fibers. Ectopic ureteral insertion may also result in obstructed megaureter and abnormal insertion in male infants involves the seminal vesicle in up to one third of cases. Here we present a case of a male newborn with primary obstructing megaureter resulting from ectopic ureteral insertion into the right seminal vesicle, identified by magnetic resonance urogram.

The majority of obstructing megaureters are diagnosed prenatally and many resolve spontaneously with time. Observation is often favored over surgery, as over 70% of obstructed megaureters resolve spontaneously within 2 years. In addition, ureteral reimplantation in this population can be extremely difficult owing, in part, to patient size and the close proximity of adjacent structures. Our case of an ectopically inserted ureter is clearly unlikely to resolve without surgical correction. Identifying these cases early may help in planning earlier intervention or closer follow-up. We and others have found the level of anatomic detail from MRI studies extremely useful in evaluation of select cases. Modern MRI techniques are fast and safe, avoid radiation, and are arguably the best study to define both the collecting system and soft tissue of the pelvis. We now use MRU to identify patients that may benefit from early intervention aimed at preserving renal function. Overall, MRI imaging will undoubtedly prove useful in planning a variety of challenging operations in the pediatric population.

References
A Case of Perineal Lipoma, Penoscrotal Transposition, Accessory Scrotum, and Imperforate Anus

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Case Presentation

We report the case of a neonate who presented with a congenital perineal mass with radiographic characteristics consistent with lipoma. This patient exhibited urogenital abnormalities including right ectopic scrotum (Figure 1a), a small accessory scrotum (Figure 1b), penoscrotal transposition, glanular hypospadias, hooded foreskin, and penile torsion. The patient also had imperforate anus, with a rectocutaneous fistula located in the midline just posterior to the lipoma. This patient was evaluated with radiographic studies including renal ultrasound and VCUG, which did not demonstrate renal abnormalities or communication of the rectal fistula with the urinary tract. The patient then underwent a planned surgical intervention at approximately 6 months of age with both urology and pediatric surgery. First, the pediatric surgery team performed a posterior sagittal anorectoplasty to move the anus posteriorly into a more orthotopic location. Peña stimulation was used to locate the sphincter complex and place the anus centrally, with good anal wink achieved on all sides at the conclusion of this portion of the procedure. The urology team then performed a complex scrotoplasty and right ectopic scrotal free flap reconstruction along with resection of the perineal lesion and meatal advancement and glanuloplasty for the patient’s hypospadias. This patient achieved an excellent cosmetic result (Figure 2a). Final pathology of the mass was mature fibrolipoma. At his 6 week follow up clinic visit, the patient had well healed incision sites, an orthotopic urethral meatus, and only slight penile torsion with some penile concealment due to his prominent suprapubic fat pad as remaining urologic abnormalities (Figure 2b).

Discussion

Congenital perineal lipomas are rare benign lesions that have been associated with a host of urogenital abnormalities (1) (hypospadias, penoscrotal transposition, ectopic scrotum, accessory scrotum), renal anomalies (renal agenesis, collecting system duplication), and anorectal malformations (2) (imperforate anus, rectovesical fistula, rectourethral fistula). These lesions can impinge upon the sphincter complex during development, attenuating eventual functional potential, and also distort the external genitalia, complicating surgical correction. These patients most typically present as neonates, due to the outwardly obvious urogenital and anal defects. Cases of congenital perineal lipoma associated with ectopic scrotum or with accessory scrotum are exceedingly rare, with case numbers in the literature reported as 23 and 34 cases respectively (1). Although the incidence of congenital perineal lipoma is low, the associated urogenital and anorectal malformations often make the treatment of these lesions complex, thereby benefiting from careful preoperative planning and a multidisciplinary approach.

References

Spontaneous Bladder Rupture in a Premature Female Neonate:  
A Case Report

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Case Presentation

34 hour female was born at 29 weeks 6 days gestation via emergent caesarean section to a 23 year old G4P1 mother at an outside institution. The pregnancy was complicated by severe mycoplasma multilobar pneumonia requiring ICU admission, intubation and administration of alprazolam, morphine, and fentanyl. The baby was edematous at birth. An abdominal ultrasound demonstrated ascitic fluid in all four abdominal quadrants and nonvisualization of the bladder, which raised concern for bladder rupture.

Upon admission an abdominal x-ray showed diffuse increased opacity peripherally surrounding centralized bowel loops, consistent with ascites. A subsequent abdominal ultrasound showed diffuse abdominal and pelvic ascites and a cystogram (figure 1) demonstrated a defect within the mid left urinary bladder wall. A Foley catheter was placed and a voiding cystourethrogram was obtained, demonstrating intraperitoneal perforation in the bladder in the inferior left bladder wall. Over the following 24 hours the baby became oliguric and azotemic with a creatinine of 2.2 mg%.

Following cystoscopy exploratory laparotomy and bladder exploration was performed, revealing a 7 mm bladder rupture on the left side of the bladder. In addition, a large volume of urinary ascites was drained. A 2-layer bladder closure was performed. Serum creatinine normalized by postoperative day (POD) 1, and a postop VCUG (figure 2) at one week was normal.

Discussion

Neonatal ascites in a female neonate is a rare condition. In boys the most common cause is posterior urethral valves. To our knowledge, this is the fourth reported case of female intrauterine bladder rupture. In the absence of an obvious cause, it is likely that transplacental exposure to medication led to fetal urinary retention and subsequent intrauterine bladder rupture. Several maternal medications used in this case, alprazolam, morphine, and fentanyl have been noted to cause urinary retention1,2. Opioids block parasympathetic nerves decreasing the sensation of fullness and stimulate sympathetic nerves increasing bladder neck tone resulting in urinary retention. Benzodiazepines have been speculated to cause retention via their muscle relaxant properties1,3. Another potential cause is spontaneous rupture of a congenital bladder diverticulum. Urinary ascites is rare, but prompt diagnosis is paramount.

References


Figure 1
Cystogram. Intraperitoneal perforation in the bladder with the site of perforation in the inferior left bladder wall.

Figure 2
Postoperative VCUG. Postsurgical defect of the left inferior bladder wall. No contrast extravasation to suggest perforation.
Case Report: Temporary Double-J Ureteral Stenting for Management of Nonrefluxing, Obstructive Megaureter

Case Presentation
The patient is a 28-day-old male referred to Arkansas Children’s Hospital for prenatally observed hydronephrosis confirmed to have right grade 3 and left grade 2 hydronephrosis with bilateral hydroureter (1.2 cm right and 0.8 cm left) on postnatal ultrasound. Voiding cystourethrogram demonstrated no vesicoureteral reflux. Prophylactic antibiotics were started. Renal ultrasound at 3 months revealed worsening right hydroureteronephrosis to grade 4 with increased hydroureter of 1.8 cm (Figure 1). Diuretic renogram exhibited symmetric differential function but right hydroureteronephrosis with delayed washout. Right ureterostomy with second stage ureteral reimplantation and circumcision were recommended although the family communicated interest in consideration of alternative approaches without open surgery or circumcision if possible.

Cystoscopy and bilateral retrograde pyelogram were performed revealing right hydroureteronephrosis with an aperistaltic segment just proximal to the right ureterovesical junction. After counseling the family intraoperatively, temporary right ureteral stent placement was performed instead of ureterostomy. The postoperative evaluation in 3 months revealed improvement of bilateral hydroureteronephrosis from grade 4 to grade 3 on the right and from grade 2 to grade 1 on the left. The diameter of the right and left ureters decreased in size to 1.0 cm and 0.6 cm, respectively. The patient underwent right ureteral stent exchange approximately 5 months after his initial procedure and then removal of the stent 4 months later.

Repeat ultrasound one month later revealed mildly increased right hydroureter to 1.5 cm with continued improvement in his right kidney to grade 2 hydronephrosis (Figure 2). His postoperative renogram showed stable differential function with improved washout bilaterally. The patient is now 4 years of age and continues to progress clinically without urinary tract infection or other urologic problems.

Discussion
Megaureters are ureters that exceed 0.7 cm in diameter with multiple subtypes. We present a case of a non-refluxing, obstructive megaureter, which has traditionally been managed with ureteric tapering and ureteral reimplantation. Early ureterostomy in small infants with subsequent ureteral reimplantation is another approach. In 1984, a minimally invasive technique for obstructive ureters was initially described where a double-J indwelling stent is placed. Multiple small series have supported this to be a reasonable approach.

This case report supports the validity of this management strategy and demonstrates an example of a patient with a non-refluxing, obstructive megaureter who benefited from this less invasive approach by avoidance of an open procedure.

References

Figure 1
Renal and Ureter Ultrasound Prior to Double-J Stenting a) Hydronephrosis Grade 4 of the right kidney  b) Right and left ureters measuring approximately 1.8 cm and 1.0 cm, respectively.

Figure 2
Renal and Ureter Ultrasound 1 Month after Double-J Stenting a) Hydronephrosis Grade 2 of the right kidney  b) Right and left ureters measuring approximately 1.5 cm and 0.5 cm, respectively.