SPECIAL EDITION:  
Society for Fetal Urology:  
Year in Review

FROM THE GUEST EDITOR  
Jeffrey B. Campbell, M.D., Secretary/Treasurer

In 2011, the Society for Fetal Urology (SFU) held its 46th Biannual Meeting in conjunction with the AUA Annual Meeting in Washington, DC. Dr. John Thomas (Division of Pediatric Urology, Monroe Carell Jr. Children’s Hospital at Vanderbilt) put together an outstanding program entitled Antenatal Hydronephrosis: Why it Happens, What it Means, and Why We Manage It. The program included a lecture by Dr. Robert Chevalier on renal embryology, with a focus on the effects of hydronephrosis on the developing kidney, and a panel discussion entitled Postnatal Management of Antenataly-Detected Hydronephrosis: A Case by Case Discussion. There were also a number of abstract presentations, including case reports, case series, and original research.

The 47th Biannual Meeting was held in conjunction with the American Academy of Pediatrics National Conference and Exhibition in Boston, MA. The program was entitled Neurogenic Bladder: Infancy to Adulthood and was chaired by Dr. Peter Metcalfe (Pediatric Urology, Stollery Children’s Hospital). We were treated to a number of outstanding guest lectures (General Overview of Basic Science and Translational Research of the Bladder – Dr. Stacy Tanaka, Evidence-based Assessment and Management of the Newborn with Myelodysplasia – Dr. Warren Snodgrass, and Progression of the Neuropathic Bladder from Infancy to Adulthood: Is 40 cm of Pressure on the UDS Really the Key in Preventing Upper Tract Deterioration? – Dr. Douglas A. Husmann) and abstract presentations.

Many of the abstracts presented at the biannual meetings are included in this publication. The SFU remains indebted to Dr. Tony Caldamone, Dialogues in Pediatric Urology, and the Society for Pediatric Urology for their continuing support.

As we look ahead to 2012, 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.

The Society for Fetal Urology (SFU) will hold its 48th Biannual Meeting in conjunction with the AUA Annual Meeting on Friday, May 18, 2012. Please note that this is a full-day meeting.

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FROM THE EDITOR  
Anthony A. Caldamone, M.D.

The Society for Fetal Urology continues to cull out interesting cases that are presented as case reports at their semi-annual meeting. The Dialogues in Pediatric Urology Editorial Board is fortunate to have an ongoing affiliation with the SFU to publish these case presentations.

As one can see from the contents of this issue, the subject matter of these cases is quite diverse, each with its own lesson. I applaud both the SFU and the authors of these contributions for continuing to further our knowledge in fetal urology, where at times we still struggle to even figure out what the right questions are.
Antenatally Detected Adrenal Cysts: Diagnosis and Management

Paul R. Bowlin, M.D., Jeffrey B. Campbell, M.D.
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Introduction
We present two cases of antenatally detected adrenal cysts. Guidelines for diagnosis and subsequent management are discussed.

Case Report
The first patient is a female with a history of antenatally detected hydronephrosis on the right. Initial postnatal renal ultrasonography (RUS) demonstrates bilateral grade 1 hydronephrosis, and a 9 mm adrenal cyst on the right (Figure 1). Urine homovanillic acid (HVA) and vanillylmandelic acid (VMA) were subsequently obtained and found to be within normal limits. A follow up RUS at 1 month of age showed complete resolution of the cyst.

The second patient is a male with a history of an antenatally detected cystic adrenal mass on the left. Serial antenatal RUS demonstrated the mass to be stable in size. Initial postnatal RUS demonstrated a cystic adrenal mass (Figure 2). Urine HVA and VMA were subsequently obtained and found to be within normal limits. A follow up RUS at 1 month of age showed complete resolution of the cyst.

Discussion
The differential diagnosis of an antenatally detected adrenal cyst includes a simple adrenal cyst, an adrenal hemorrhage, a cystic neuroblastoma, and a retroperitoneal lymphatic malformation. Postnatal management typically includes obtaining urine HVA and VMA levels to rule out neuroblastoma, and serial RUS. Some studies also suggest obtaining a metaiodobenzylguanidine (MIBG) scan. In most cases, these lesions resolve spontaneously without sequelae. Surgery is typically reserved for masses that increase in size or persist for greater than 6 months. Collaboration with oncology is important as many of these patients may be eligible for enrollment in a clinical trial.

Reference
Fetal Abdominal Ascites from Intraabdominal Bladder Rupture Secondary to Posterior Urethral Valves: Diagnostic Challenges

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Introduction

Though rare, fetal ascites is described sonographically and has a broad differential diagnosis. We present a case of fetal ascites wherein confounding data delayed the diagnosis of urinary ascites secondary to posterior urethral valves (PUV).

Case Report

A 14 week prenatal ultrasound (US) identified normal male-female twins. US at 20 weeks identified severe hydrops in the male. At week 29, C-section was performed at OSH for breech presentation and decreasing cardiac function. Apgars were 1, 6, and 6. The male newborn had normal facies, systolic murmur, rib flaring, massive abdominal distension, normal GU exam and no anasarca. After intubation for respiratory distress from abdominal distention, 550 cc were removed via paracentesis. Echocardiography identified an ASD and PDA with R to L shunt but good function. Ultrasound on first day of life (DOL1) demonstrated abdominal ascites, normal kidneys(4.2cm) without hydrourerteronephrosis, and decompressed bladder by catheter. Despite catheterization, he was anuric, with minimal transient response to loop diuretic and renal-dose dopamine. He required paracentesis twice to permit ventilation prior to transfer. Labs excluded chylous or bilious ascites; cultures were sterile with additional serum and ascites labs in Table 1.

On DOL8 the patient was transferred for dialysis (Fig1). Urology recommended repeat paracentesis and VCUG, which identified a dilated posterior urethra with intraperitoneal bladder rupture. Small size (1.25kg) precluded attempt at cystoscopy or valve incision. Vescicostomy with drainage of 300cc urinary ascites via vescicostomy incision was performed. No clear area of bladder rupture was identified. Serum creatinine decreased from 4.9 to 0.3 and he was extubated.

Discussion

The causes of fetal and neonatal ascites include biliary, urinary, chylous, iatrogenic, cardiac, infectious, pancreatic and heptocellular etiologies.1 In complex cases, high suspicion for PUV must be maintained as 30% of neonatal ascites is caused by obstructive uropathy though spontaneous bladder rupture is reported less than 20 times since 1985.2 Paracentesis creatinine values may be misleading and must be interpreted with caution. Confusion arose as the initial ascites creatinine was less than the serum creatinine; subsequent taps showed minimal elevation of ascites over serum creatinine. This is postulated to occur because of autodialysis,3 wherein the peritoneal urine contains high potassium, urea, and creatinine with low sodium. Because of high ascites osmolality, water shifts from plasma to peritoneum. Sodium also enters the peritoneum, causing a relative hyponatremia. In turn, the renin-angiotensin-aldosterone axis is activated, leading to lower sodium concentration and higher potassium excretion in the ascites fluid. The high levels of potassium, urea, and creatinine in the ascites cause a solute migration into the vasculature, equilibrating the serum-ascites creatinine gradient. We advocate VCUG in cases of neonatal ascites of unclear etiology, especially in the face of confounding laboratory values and sonographically normal or near normal urinary tract.

Table 1: Serum and Ascites Fluid Laboratory Values

<table>
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<th>DOL</th>
<th>Serum Na</th>
<th>Serum BUN</th>
<th>Serum Creatinine</th>
<th>Paracentesis Ascites Volume</th>
<th>Ascites BUN</th>
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</table>

Figure 1: Ex-29 week male infant on DOL8 with massive urinary ascites from intrauterine bladder rupture induced by posterior urethral valves. Vescicostomy with intraoperative drainage of 300 cc of urinary ascites was performed as transurethral resection of valves was not feasible due to small size.

References

Ectopic Ureter in a Patient with Ehlers-Danlos Syndrome: Surgical Considerations

Paul R. Bowlin, M.D., Jeffrey B. Campbell, M.D. Children’s Hospital Colorado, Aurora, CO

Introduction
We present a case of antenatally detected hydroureteronephrosis (HUN) secondary to an ectopic ureter in a patient with Ehlers-Danlos syndrome.

Case Report
The patient is a male with a history of antenatally detected HUN on the right. Initial postnatal renal ultrasonography (RUS) demonstrated grade 3-4 HUN. A voiding cystourethrogram (VCUG) was subsequently obtained and found to be within normal limits. At 6 weeks of age, a RUS was repeated and a diuretic nuclear renal scan was obtained. The RUS demonstrated grade 3-4 HUN on the right. The renal scan demonstrated impaired drainage of the right kidney, without an appreciable improvement in drainage following the administration of Lasix, and split renal function of 49/51 favoring the left kidney. A magnetic resonance urogram (MRU) was subsequently obtained, demonstrating an ectopic ureter on the right. At 3 months of age, the patient underwent a right loop cutaneous ureterostomy. At the time of surgery, the ureter was noted to be dilated and floppy. A RUS was obtained 1 month postoperatively, demonstrating grade 4 HUN on the right. In addition, the patient’s parents noted that there had not been much drainage across the ureterostomy. During further discussion with the patient’s parents, it was learned that a family member had recently been diagnosed with Ehlers-Danlos syndrome. Given the poor drainage, a red rubber catheter was placed into the afferent limb of the ureterostomy. The catheter was exchanged weekly, with a follow up RUS demonstrating an improvement in the HUN. Catheterization was subsequently discontinued, with improved drainage across the ureterostomy and stable findings on RUS. The patient was subsequently diagnosed with Ehlers-Danlos syndrome. He is currently scheduled to undergo a right urinary undiversion and ureteroneocystostomy, with excisional tailoring.

Discussion
Ehlers-Danlos syndrome results from a defect in the synthesis/structure of collagen. There are 10 recognized subtypes, with variable patterns of inheritance; most are inherited in an autosomal dominant fashion. There is little known about the implications of Ehlers-Danlos and other collagen diseases on the structure and function of the ureter. There are several reports of bladder diverticula associated with the syndrome, but limited literature addressing any ureteral involvement. It is likely that this condition has effects on the elasticity and contractility of the ureter, which is liable to affect the ease and success of reconstructive procedures involving the ureter.

References

Dysfunctional Voiding in Two Sets of Twins
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Introduction
Posterior urethral valves (PUV) account for 10% of urinary obstructions diagnosed in utero. Rare concordant and discordant presentations of PUV in twins have been described in case reports. We hypothesize that the twin gestation may complicate diagnosis of this entity and could lead to a delay in treatment.

Case Report
We describe a case series of two families in which there was a discordant presentation: one twin of each gestation was eventually diagnosed with PUV. An 8-year-old dizygotic twin boy was evaluated for dysfunctional voiding following a febrile UTI. Workup subsequently revealed PUV. His brother had been evaluated soon after their birth for a history of prenatal hydronephrosis, and was later discharged from clinic with a normal workup. A second twin boy from a different family presented with voiding complaints at age 4. Workup subsequently identified PUV. His brother had no similar symptoms.

Discussion
Discordant disease in twins has been a source of medical dilemmas. A delay in diagnosis of PUV in twin gestations could result from failed sensitivity of prenatal ultrasound or from postnatal evaluation of the incorrect twin. Screening of each twin should be considered in cases of discordant prenatal hydronephrosis and when the diagnosis of PUV is made in one of the twins.

References
Vesicoamniotic Shunt Migration in a Male Fetus with Posterior Urethral Valve

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²Connecticut Children’s Medical Center Division of Urology, Hartford, CT

Introduction

Lower urinary tract obstruction in a fetus, including posterior urethral valves, can lead to abnormal renal development, oligohydramnios, pulmonary hypoplasia, and postural anomalies. Perinatal morbidity and mortality can be profound.¹ The diagnosis is now commonly made by prenatal sonography, however, the therapeutic benefit of antenatal intervention is yet unknown.

Bladder drainage by vesicoamniotic shunting is an attractive concept to relieve obstruction, thereby avoiding renal parenchymal damage and subsequent adverse pulmonary development.²³ This procedure carries significant risk to mother and fetus. A complete understanding of the risks and complications accompany its use in current practice.

Case Report

A male fetus with imaging findings suggestive of posterior urethral valves underwent vesicoamniotic shunt placement at 28 weeks gestational age by an outside institution. The mother was a healthy 30-year-old G1P0. Fetal urinary electrolytes appeared favorable.

Records indicate a vesicoamniotic shunt was satisfactorily placed, however, repeat sonography revealed migration of the shunt to a vesicoperitoneal position. The patient subsequently developed oligohydramnios with an amniotic fluid index of 1.9. A peritoneoamniotic shunt was placed at 30 weeks to decompress urinary ascites.

The fetus was delivered by emergent cesarean section at 32 1/7 weeks secondary to fetal distress. There was no visible shunt on the abdominal wall and abdominal contents were seen protruding through a ventral hernia site. The baby was intubated shortly after delivery due to poor ventilatory effort. Imaging revealed urinary ascites, bilateral SFU grade 3 hydronephrosis, and a double pigtail catheter from the bladder to the pelvis. A 3.5fr urethral catheter was placed without significant return of urine.

In order to avoid electrolyte abnormalities and ventilatory complications from urinary ascites, the patient was taken for urgent operative intervention. The patient’s urethra was too small to accommodate endoscopic valve ablation or shunt removal. He underwent cutaneous vesicostomy, vesico-peritoneal shunt removal, exploratory laparotomy and ventral herniorrhaphy.

Postoperatively, the baby’s creatinine trended down from 1.6 to 0.3mg/dL, and has remained normal in follow up. At 7 months of age, the patient underwent endoscopic ablation of a type I PUV and closure of the vesicostomy.

Discussion

The complication of shunt migration has been reported.⁴ Shunt dislodgement or occlusion appears to be the most common complication, occurring in about 1/3 of cases.⁵ Reported risks to the mother include preterm labor, premature rupture of membranes, and chorioamnionitis. Potential morbidity to the fetus includes traumatic injuries such as hernia and bladder rupture. Finally, death in association with shunt procedures has been attributed to preterm delivery and chorioamnionitis. Overall complication rates from antenatal intervention may occur in 50% of cases.⁴⁵

The future of antenatal intervention for lower urinary tract obstruction remains to be seen. While a 2009 Cochrane review suggests a benefit in perinatal mortality, it does not address renal function and randomized controlled trials are lacking.⁵ As our understanding of the potential benefits of the procedure evolve, the risks must also be understood. We present this patient to discuss the varied outcomes of vesicoamniotic shunting for posterior urethral valve.

References

Posterior Urethral Valves Masquerading as Triad Syndrome on Antenatal Imaging

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Introduction
Evidence of lower urinary tract obstruction on antenatal imaging of a fetus can include hydronephrosis, hydroureter, distended bladder, prominent posterior urethra and ascites. The differential diagnosis of lower urinary tract obstruction detected with antenatal imaging includes urethral atresia, posterior or anterior urethral valves and Triad syndrome. We present a case of lower urinary tract obstruction secondary to posterior urethral valves that posed as Triad Syndrome on antenatal imaging.

Case Presentation
A newborn boy born at 34 weeks GA was found to have severe bilateral hydroureteronephrosis and severe ascites on 24 week GA ultrasound. Prior antenatal ultrasonography at 20 weeks GA was normal (Figure 1). A fetal MRI was performed at 26 weeks GA, demonstrating a dilated posterior urethra, a dilated and thickened bladder, severe bilateral hydroureteronephrosis, bilateral enlarged kidneys, significant ascites, abdominal distension and mild oligohydramnios (Figure 2). The fetus was diagnosed with a possible variant form of Triad Syndrome, given the lack of keyhole sign on antenatal ultrasonography and the normal exam at 20 weeks GA. The fetus underwent percutaneous drainage of the bladder and ascites at 26 5/7 weeks GA, and the ascitic fluid was confirmed to be urine (Figure 3).

Subsequent antenatal imaging demonstrated no recurrence of ascites and a normal amniotic fluid index. The patient was born at 38 and 5/7 weeks by normal spontaneous vaginal delivery. Physical exam revealed a normal 4335-gram male infant with normal genitalia who voided spontaneously. Postnatal ultrasonography confirmed severe bilateral hydroureteronephrosis with thickened bladder and a dilated posterior urethra, without a classic keyhole sign. VCUG revealed posterior urethral valves. The patient underwent endoscopic ablation of posterior urethral valves on DOL #1. His creatinine increased to 1.9 mg/dL on DOL #3 then steadily fell to a nadir of 0.4 mg/dL.

Discussion
The etiology of lower urinary tract obstruction on antenatal imaging may be difficult to define. We present herein a case of lower urinary tract obstruction evident at 24 weeks GA after a normal antenatal ultrasound at 20 weeks GA. Common antenatal findings in Triad Syndrome include a relatively normal amniotic fluid volume until 20-24 weeks GA. Development of the fetal pelvis thereafter may cause an angular relationship between bladder neck and urethra as it courses under the pubic bone. This may cause kinking of an already hypoplastic urethra resulting in obstruction.

Massive ascites and oligohydramnios responded to a single percutaneous drainage procedure with normal amniotic fluid parameters subsequently. This favorable outcome occurred without vesicoamniotic shunting. The diagnosis of posterior urethral valves was made postnatally and he was treated appropriately.

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Multicystic Dysplastic Kidney in the Bladder Wall Causing Voiding Dysfunction

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Introduction

We present a case of voiding dysfunction secondary to apparent compressive effect of a multicystic dysplastic kidney (MCDK) in the detrusor wall. This represents an unusual manifestation of a dysplastic kidney most likely due to both severe obstruction and an anomaly of ascent early in renal embryogenesis. At 4 weeks gestational age, the ureteric bud comes into contact with the metanephric mesenchyme.1 Normal renal ascent commences at 6 weeks gestational age.1 In our case, there was both a failure of ascent and high grade obstruction resulting in MCDK, which became incorporated into the detrusor wall, encroaching upon the lumen of the bladder itself. MCDK has low malignant potential, and usually does not require treatment.2

Case Presentation

A 6-year-old male presented with urinary frequency and urge incontinence. He underwent cystometrics, which demonstrated detrusor overactivity and a small bladder capacity for age (120-170 mL, expected 240 mL). He was treated with anticholinergic medications without improvement. CT scan revealed a MCDK located in the wall of the bladder, which significantly encroached upon luminal capacity (Figure 1). Holmium laser puncture of the MCDK was performed endoscopically to decompress the cysts, with intention to increase his bladder capacity. His voiding symptoms resolved postoperatively, and his voided bladder volume improved to 220 ml.

At two-year follow-up he has no voiding dysfunction off anticholinergic medication. Also, the MCDK is no longer visible on ultrasound.

Discussion

A plausible embryologic explanation can be offered for the unusual ectopic location of this MCDK. Hypothetically the kidney may have been incorporated into the bladder wall during formation of the trigone as the common excretory duct, mesonephric duct, and ureteric bud evaginates. MCDK is commonly attributed to either ureteral atresia/high grade obstruction during very early renal embryogenesis or from abnormal interaction between the ureteric bud and metanephric mesenchyme.2-3 It is conceivable that the ureteric bud failed to canalize or had abnormal embryologic fusion resulting in cystic malformation of the kidney. We believe that endoscopic puncture was the best approach to this case, as laparoscopic nephrectomy may have created a larger defect in the bladder wall.

References

Long Term Bladder Dysfunction as a Result of In Utero Outlet Obstruction in Females

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Introduction

With improvements in prenatal imaging, most ureteroceles are now diagnosed in utero. Historically, ureteroceles were not surgically repaired until clinical manifestations, such as recurrent urinary tract infection or bladder dysfunction, occurred. However, as our understanding of the potential negative long term sequelae of bladder outlet obstruction improves, early surgical intervention may be warranted for large obstructing ureteroceles. Controversy exists surrounding appropriate timing and optimal treatment.

Case Report

A female fetus at 32 weeks with a normal amniotic fluid index was noted on prenatal sonogram to have bilateral hydroureteronephrosis and a thickened bladder wall suggestive of bladder outlet obstruction. The right kidney was difficult to visualize. Serial sonograms did not significantly change and she was born at term with a normal genitourinary and back exam. Postnatal sonogram revealed bilateral duplication, hydroureteronephrosis, and a small right kidney. There was no evidence of vesicoureteral reflux (VUR) on voiding cystourethrogram (VCUG), but a large right ureterocele and trabeculated bladder were identified. Incision of the ureterocele occurred on day of life five and postoperative imaging demonstrated resolution of the ureterocele, persistent hydroureteronephrosis and new right grade 2 VUR.

She was observed on prophylactic antibiotics and serial sonography showed improved hydronephrosis. However, a second incision of the ureterocele was performed after a febrile UTI at 3.5 months of age. Subsequent ureterocele excision was performed at 5 months due to recurrent infection and concern for incomplete bladder emptying. She remained stable and infection free for 48 months, during which time urodynamics showed minimally elevated filling pressures and nuclear renography confirmed a non-functioning right kidney.

With initiation of toilet training she again developed recurrent febrile UTIs ultimately resulting in a minimally invasive right nephroureterectomy. Five months after nephroureterectomy she presented with left pyelonephritis and acute renal failure. Video urodynamics demonstrated a poorly compliant bladder with limited capacity and high storage pressures. Despite numerous attempts with experienced nursing staff, she was unable to tolerate catheterization per urethra. Out of concern that she would also not catheterize per accessory channel, the child underwent cutaneous vesicostomy. Post-vesicostomy imaging reveals improvement in her hydronephrosis.

Discussion

There is evidence suggesting no difference in rates of UTI or progression of hydronephrosis in those treated with immediate endoscopic decompression when compared to delayed open repair, while other data show that early decompression will help preserve upper tract function. Still others argue that relief of obstruction may recover some function but often leads to new VUR and ongoing breakthrough UTIs, thus intervention is advocated only in the setting of urosepsis, pyonephrosis, or complete outlet obstruction. Finally, there are reports of successful in utero decompression of ureteroceles aimed at avoiding the long term complications of obstruction.

We present the case of a female child with bilateral antenatally detected hydroureteronephrosis secondary to large obstructing ureterocele. Much like a male with posterior urethral valves, neonatal obstruction from ureterocele can have devastating long-term consequences on bladder function; the timing of surgery as well as the initial method of repair remains controversial.

References

Difference in the Second and Third Trimester Ultrasound of Fetal Hydronephrosis: Predict for Postnatal Surgery

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Introduction

Although pyelectasis can be easily identified in the second trimester by prenatal ultrasound, the ability of prenatal ultrasound to predict those fetuses that will go on to demonstrate significant renal or urinary tract pathology is poor, and there is little agreement on the best follow up. There is also little data in the literature regarding the impact of prenatally change pyelectasis and postnatal outcomes.

We sought to determine whether the difference in the second and third trimester hydronephrosis in the fetus can predict the postnatal surgery.

Methods and Materials

This is a retrospective study of fetuses received prenatal screening at the Perinatal-Pediatric Urology Clinic (PPUC)/University of Connecticut Health Center-John Dempsey Hospital and had their follow up care at Connecticut Children’s Medical Center Department of Pediatric Urology between January 2007 and January 2010. Of these 96 fetuses, 33 (34%) were active enrollees in the Society for Fetal Urology’s (SFU) Hydronephrosis Registry. Renal pelvis anterior-posterior diameter (APD) was measured in a transverse midabdominal fetal scan plane showing the fetal kidneys with the technique described by Corteville et al.1 In patients who had multiple exams, the largest APD in each trimester was used for our analysis. If hydronephrosis was bilateral, the larger APD was used in analysis. The difference between the largest APD in the third and second trimester was calculated as Δ APD. All infants were had follow up with pediatric urologist at Connecticut Children’s Medical Center. Sensitivities and specificities for various cut-off levels were calculated and graphically displayed using receiver–operating characteristic (ROC) curves.

Results

There were 96 fetuses referred to PPUC during the period of the study which 53 had hydronephrosis and postnatal follow up. Mean postnatal follow up time for neonates was 413 days (SD± 275 days). The ROC curves for the Δ APD to predict surgery is displayed in Figure 1. The AUC for the Δ APD was 0.862. The Δ APD threshold that best predicted postnatal surgery was 17.8 mm (sensitivity 66.7%, false positive rate 2.3% and likelihood ratio of 29).

Discussion

Parents of fetuses with antenatal hydronephrosis are particularly interested in whether their children will need further investigation and treatment after birth. Our study focused on these parental concerns by generating additional information that may be useful when counseling parents. Our group previously reported that neonatal nephro-uropathy requiring surgery is best predicted by a third trimester renal APD threshold of 15 mm². Now, we can advise parents that it is unlikely their infant will require surgery if Δ APD is less than 17.8 mm.

References


Figure 1. Δ APD to Predict Pediatric Surgery.
Duplicated Renal Collecting Systems: When “The Rules Don’t Apply”

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Introduction
The classical presentation for a duplicated collecting system is a poorly functioning dilated superior moiety and refluxing inferior moiety.1 We present two patients with abnormalities of renal development in whom these “rules do not apply.”

Case Reports
Patient 1 is a male infant with a right duplicated system with moderate inferior moiety hydronephrosis without hydroureter. VCUG revealed grade 2 and 5 VUR into the superior and inferior moieties respectively. A radionuclide scan revealed no function of the right inferior moiety. At 7 months of age, the patient underwent a robotic assisted laparoscopic inferior moiety nephroureterectomy and superior moiety extravesical ureteroneocystotomy. VCUG and renal ultrasound at 11 months of age revealed no VUR and a normal appearing right superior moiety.

Patient 2 is a two-month-old male with a duplicated right renal system with a markedly hydronephrotic inferior moiety and a dilated ureter. VCUG revealed a right bladder diverticulum, but no VUR or ureterocele. Radionuclide scan revealed a nonfunctioning inferior moiety. At 18 weeks, the patient underwent a right inferior moiety nephrectomy, revealing no inferior moiety ureter and the dilated ureter to be of the superior moiety. Postoperative imaging showed the remaining superior moiety with moderate hydroureteronephrosis to the level of the bladder and grade 4 VUR. At one year, the patient underwent a right intravesical cross-trigonal ureteroneocystotomy.

Discussion
In duplicated systems, “the rule” is that the superior moiety is the nonfunctioning segment, typically secondary to obstruction. These two patients had nonfunctioning, hydronephrotic inferior moieties: Patient 1 with massive inferior moiety VUR, and patient 2 with complete inferior moiety UPJ obstruction. Patient 2 also had a hydronephrotic superior moiety due in part to reflux, but also secondary obstruction from the massively dilated inferior moiety. These two cases illustrate that “the rules” of “the nonfunctioning segment is the superior moiety” and “superior moiety dilation is due to obstruction” do not always apply.

References