Society for Fetal Urology:
2015 Meeting in Review

FROM THE EDITOR

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For this Edition I have the pleasure and privilege of writing the Editorial introduction to the 2015 Society of Fetal Urology case presentation edition as both Editor of DPU and as President of the SFU. The SFU has a long and mutually beneficial relationship to Dialogues, beginning well before my role in either organization. The relationship is very natural, as both are focused on sharing experiences and information in a digestible and conversational format.

Case presentations related to fetal or perinatal urology or with an embryologic focus have been a tradition with the Society for Fetal Urology. These sessions have been an important moment for many students, residents and fellows to have the opportunity to present an interesting case, to field questions and to reap the clinical and academic benefits of the discussions.

This Edition includes the excellent case presentations from the 2015 Spring SFU meeting, which was held in New Orleans in conjunction with the SPU-AUA meeting. My sincere thanks to Jeff Campbell for gathering and editing the case submissions. John Kryger chaired the SFU portion of the Spring meeting and the evening case session.

In addition to the moderated podium session on Perinatal Urology, the program included an outstanding panel on bladder extrophy, looking at it embryologically, surgically and from a patient perspective. The panel consisted of individual expertise on Embryology of Bladder Extrophy (Michael E. Mitchell, MD), Sexuality and Fertility Issues Among Exstrophy Patients (Barry P. Duel, MD), Phallic Reconstruction in Exstrophy Males (Bradley P. Kropp, MD) and Role of Multi-institutional Consortium in Future of Exstrophy Care (John Kryger, MD).

This meeting will be the only SFU meeting for 2015, as we will not have independent programming at the ESPU and Joint Societies meeting in Prague in the Fall. We are delighted to be one several societies collaborating in the academic planning and abstract selection for this landmark meeting, and we look forward to learning alongside colleagues from around the globe.

The next case presentation session will be held on the eve of the SFU session in conjunction with SPU-AUA in San Diego. Join us to partake in the interesting cases and to encourage participation of the next generation of pediatric urologists! Already seen it all? Come on by to participate in the dialogue.
Urinary Ascites Secondary to Bladder Perforation in a Neonate with Bladder Outlet Obstruction

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Case Presentation
A 19-year-old female, delivered a 26-week-old male by emergency caesarian section secondary to placenta previa and vaginal bleeding. The patient weighed 1 kg and was admitted to NICU. Umbilical artery and vein catheters were placed.

On day of life (DOL) 2, the patient had a BUN of 35 mg/dL and creatinine of 1.1 mg/dL. The physical exam was within normal limits. On DOL 8, the BUN and creatinine had risen to 62 and 1.6 mg/dL, respectively. Urine output was recorded as normal. On DOL 9 the patient became oliguric, with a distended abdomen and a creatinine of 3.9 mg/dL. An abdominal ultrasound (US) demonstrated bilateral severe hydronephrosis, a decompressed thick-walled bladder, and marked ascites. Paracentesis revealed a BUN 134 mg/dL and a creatinine of 6 mg/dL.

The patient was transferred to our facility for further management. On arrival, the creatinine was 4.1 mg/dL. The patient was continued on ventilatory support and a 5 French feeding tube was placed per urethra into the bladder, with a small amount of urine drained. An abdominal US demonstrated echogenic kidneys with mild to moderate hydronephrosis, a decompressed thick-walled bladder, and marked urinary ascites. On DOL 11, peritoneal dialysis was initiated. Urine output slowly increased to 4 mL/kg/h and by DOL 14 the creatinine had decreased to 1.3mg/dL. An abdominal US demonstrated stable hydronephrosis and thick-walled bladder, with a significant decrease in the ascites. On DOL 21, a VCUG demonstrated right dilating vesicoureteral reflux (VUR) and an intraperitoneal bladder perforation. The VCUG was aborted after instilling 25 mL of contrast.

The patient was taken to the operating room. An extraperitoneal exploration revealed a 5 x 5 mm mature perforation at the dome of a thick-walled bladder (Figure 1). The perforation was closed in two layers, and a cutaneous vesicostomy was created. Postoperatively, the creatinine decreased to 0.6 mg/dL. On DOL 31, a cystogram through the vesicostomy demonstrated a dilated posterior urethra consistent with a history of posterior urethral valve (PUV), and right grade 4 VUR (Figure 2).

One month post vesicostomy, the patient returned to the OR for closure of the vesicostomy and laser valve ablation. Two weeks postoperatively, an abdominal US demonstrated stable left hydronephrosis and decreased bladder wall thickness, and the creatinine remained stable at 0.6mg/dL.

Discussion
PUV is the most common cause of urinary ascites in the neonate. The ascites most commonly result from the rupture of a calyceal fornix secondary to increased intrarenal pressure.

Perforation of the urinary bladder with urinary ascites in the newborn is a rare clinical entity1. Etiologies include bladder outlet obstruction (PUV), urethral atresia, neurogenic bladder, traumatic perforation from umbilical artery or bladder catheterization, or, rarely, spontaneous rupture without an identifiable cause2, 3.

Management of intraperitoneal bladder perforation is based on etiology and condition of the neonate. Most advocate surgical repair, although, some recommend conservative management, with placement of a urethral catheter for 10 to 14 days .1, 3 Prognosis depends on early diagnosis/treatment and adequate urinary drainage. Our case was managed surgically, and was successfully treated with vesicostomy and laser valve ablation, with a nadir creatinine of 0.6mg/dL at the time of discharge.

References
Urologic Considerations in the Separation of Conjoined Twins


Case Presentation

A 24-year-old G1P2 female presented to the Fetal Center at 23 weeks gestation for evaluation of a fetal anomaly on ultrasound. MRI was obtained at 29 weeks gestation revealing female thoracoamphalopagus (fusion at sternum and abdomen) conjoined twins. Urologic findings were similar for each twin; two kidneys, one of which demonstrated hydronephrosis, with ectopic insertion of the ureter into the bladder (Figure 1).

Postnatal physical examination demonstrated thoracoamphalopagus conjoined twins, a perineum that was fused and rotated 90 degrees, two separate clitorises and urogenital (UG) sinuses, and two separate anuses. However, because of rotation, it was not clear which UG sinus belonged to which twin. Postnatal radiography included abdominal ultrasound, VCUG, and MRI. MRI demonstrated Twin A having moderate left hydronephrosis with ectopic insertion of the ureter into the bladder neck, and Twin B having moderate right hydronephrosis with ectopic insertion into the bladder neck or lower. As with the prenatal MRI, it was unclear which bladder the dilated ectopically inserting ureters inserted into. Two sets of non-obstructed ureters didelphys and two normal ovaries were visualized. A VCUG obtained at 7 months of age demonstrated no reflux in Twin A's genitourinary (GU) tract, however there was reflux into Twin B's right ectopic ureter.

Separation occurred at 10 months of age and took 18 hours. Total OR time for Twin’s A and B were 23 hours and 26 hour, respectively. Separation was approached from cephalad to caudal. The bowel was separate, except for minimal rectal fusion. GU findings included the following: Twin A’s non-dilated right ureter crossed to twin B’s bladder and Twin A had a dilated ectopically inserting left ureter. Twin A underwent bilateral stented extravesical ureteral reimplantation (Fig 2). Twin B’s right ureter inserted ectopically into Twin A’s bladder neck and was dilated and tortuous. Twin B underwent a right stented extravesical ureteral reimplantation. Each twin was left with a whole bladder and one uterus didelphys. The pelvis was closed primarily after pelvic osteotomies and placement of external fixators by orthopedic surgery. Both twins have done well and have been discharged home.

Discussion

The incidence of conjoined twins is 1/50,000-100,000 live births with a 3:1 F:M ratio. The separation of conjoined twins necessitates a multidisciplinary approach including surgical, anesthesia, nursing, and critical care teams. Intraoperatively, the pediatric urologist should be prepared for anatomic variability from the preoperative imaging. In our patients, despite extensive review of preoperative images, including 3D models, it was still not obvious which bladder each of the ectopic, dilated ureters inserted into. However, intraoperatively, we could evaluate the entire GU tract and place probes in the perineal openings, resulting in very apparent anatomy and facilitating decisions regarding ureteral division, reimplantation and perineal dissection. Another urologic consideration for conjoined twins with two bladders includes transureteroureterostomy if reimplantation is not feasible. In twins with a single bladder or two inseparable bladders, the surgeon must decide how to divide the bladder as continence outcomes may be affected. The appendix should also be preserved for future GU reconstruction.

References


Figure 1

Figure 2
Malignant Glioma of the Sacral Spinal Cord causing Newborn Bladder Outlet Obstruction

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Case Presentation:
A 44-year-old female underwent prenatal ultrasonography at 40 weeks gestational age that demonstrated anhydraminos and fetal ascites. Prior ultrasounds revealed normal fetal urinary tract anatomy with normal amniotic fluid indices. The patient underwent emergency Cesarean section at 40 weeks 2 days gestation. The female infant was noted to have massive abdominal distention. A 6 French Foley catheter was inserted for optimal bladder drainage in light of suspected urinary ascites. Abdominal ultrasound demonstrated massive ascites with normal intra-abdominal organs, no hydronephrosis, and a decompressed bladder. Abdominal paracentesis was performed twice with removal of 125 cc of clear amber fluid. The blood urea nitrogen (BUN) and creatinine levels from the paracentesis were 13 and 0.4, respectively. When compared to serum levels of 17 and 0.55, the ascites was found to be consistent with serum. Two failed voiding trials necessitated replacement of the Foley catheter. At 7 days of age, cystography demonstrated no evidence of vesicoureteral reflux, bladder diverticula, or bladder rupture (Figure 1). Cystoscopy and bilateral retrograde pyelography revealed normal bladder anatomy with no evidence of anatomical urethral obstruction or calyceal rupture. Magnetic resonance imaging (MRI) of the abdomen revealed a lobulated appearing mass involving the conus medullaris and massive ascites. MRI of the brain and spinal cord revealed the spinal cord terminating at the level of L2 with an expansile, heterogeneous mass involving the conus medullaris (Figure 2).

At 22 days of age, pediatric neurosurgery performed an L1-L2 osteoplastic laminectomy with excisional biopsy of 70% of the conus medullaris mass. Pathology revealed malignant glioma. The Foley catheter was removed on postoperative day 12 and spontaneous voiding occurred. Postvoid residuals revealed excellent bladder emptying. A repeat spine MRI demonstrated that the mass was largely unchanged. Pediatric hematology and oncology consultation, along with further input from pediatric neurosurgery, was obtained from our institution and an outside high-volume pediatric center. Both agreed that the tumor was highly aggressive and would not respond well to chemotherapy or high-dose radiation therapy, especially when gross total resection was not an option. Prior to discharge, renal and bladder ultrasound revealed mild fullness of the collecting systems (bilateral) and a non-distended bladder. The patient was seen at 7 weeks of age and noted to have normal motor function along with continued spontaneous voiding. Bladder ultrasound verified excellent bladder emptying.

Discussion
Neonatal bladder outlet obstruction is an uncommon entity with differing etiologies in male and female infants. The differential diagnosis in the male includes posterior and anterior urethral valves, fibroepithelial polyps, urethral atresia, presacral teratoma, Prune Belly syndrome, and myelomeningocele. In female infants, the differential diagnosis also includes ureterocele, imperforate hymen, cloacal abnormalities, Bartholin’s duct cyst, and megacystis microcolon. In this case, urinary retention was not due to anatomical urethral obstruction nor extrinsic obstruction from an extravesical mass. This is a rare instance of urinary retention secondary to a malignant lesion of the conus medullaris. An unusual aspect of this case is the antenatal sudden onset of urinary retention with anhydraminos and abdominal ascites.

References
Crossed Fused Renal Ectopia with Orthotopic Multicystic Dysplastic Kidney

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Case Presentation

A 27-year-old G1P1 female was referred to our center following a 30 weeks gestational age (GA) prenatal ultrasound, which revealed a dysplastic kidney on the right and multicystic dysplastic kidney (MCDK) on the left. The bladder was normal in appearance, and the amniotic fluid index (AFI) was within normal limits.

A fetal MRI was subsequently obtained at 31 weeks GA, more clearly defining the renal anomalies as right to left crossed fused renal ectopia, with a multicystic dysplastic upper moiety (left kidney) with an associated ectopic ureter to the seminal vesicle, and malrotation of the inferior moiety (right kidney). (Figure 1)

The patient was delivered at 38 weeks GA without complication. On day of life 10, a renal ultrasound was obtained, corroborating the findings on the fetal MRI. A voiding cystourethrogram (VCUG) was also obtained and found to be within normal limits.

At 2 months of age, the patient had progressed well, without infection. Repeat renal ultrasonography demonstrated stable findings. A MAG 3 renal scan was also obtained, demonstrating a solitary functioning kidney on the left. More specifically, the patient was found to have a photopenic area superior to the functional renal unit consistent with an orthotopic (left) MCDK. (Figure 2) Accordingly, arrangements have been made to have the patient undergo reevaluation with a renal ultrasound in 3 months.

Discussion

Crossed renal ectopia (CRE) with fusion and MCDK are relatively common entities, present in approximately 1 in 1000-4000 live births. While there are several reports of crossed fused renal ectopia with MCDK affecting the ectopic kidney, MCDK affecting the orthotopic kidney is exceedingly rare, accounting for a few published case reports. We present a rare case of crossed fused renal ectopia with MCDK affecting the orthotopic kidney. To our knowledge, this is the first description associated with an ectopic ureter to the seminal vesicle.

The etiology of CRE and MCDK is not particularly well understood. That having been said, these entities are thought to result from aberrant renal development during early embryogenesis, likely related to an altered interaction between the ureteral bud and metanephric blastema. In CRE, this alteration disrupts normal renal ascent and arrests rotation of the crossed moiety while usually maintaining normal ureteral implantation into the bladder. Overall, MCDKs are benign, with 40% involuting spontaneously; likewise, involution of MCDK in CRE has been described in at least one case report.

Our case also illustrates the difficult task of accurate prenatal diagnosis of rare, complex renal anomalies and the utility of fetal MRI in this regard. In this case, the findings on prenatal ultrasonography were profoundly different than those on fetal MRI and postnatal ultrasonography. Fetal MRI provides a more detailed anatomic evaluation of complicated genitourinary anatomy, thus allowing for patient-specific postnatal planning and, hopefully, decreased parental anxiety.

References


Figure 1 - Fetal MRI 31 weeks GA

Figure 2 - MAG 3 Renal Scan - 2 months of age
Polypoid Cystitis: A Rare But Treatable Cause of Pediatric Lower Urinary Tract Symptoms

Esther Han, D.O. and Kristina D. Suson, M.D.

Case Presentation

A 2-year-old boy presented to the emergency department with intermittent gross hematuria. Renal bladder ultrasound demonstrated a possible bladder mass. The boy was referred to urology. He had no previous urologic history, but recently began complaining of straining and dysuria on urination. In clinic, he was noted to have meatal stenosis; repeat ultrasound revealed an unchanged 1 cm mass that did not move with varied position and exhibited vascular flow.

Meatoplasty was performed to enable scope placement. Cystoscopy revealed a smooth-walled, pedunculated mass arising from the prostate. It occluded the bladder neck in a ball-valving fashion. Transurethral resection was completed by amputating the tumor at its base. The urethra barely accommodated removal of the mass en bloc.

The tumor specimen consisted of a 1.1 x 0.7 x 0.6 cm polypoid pink-red, mucosal-covered tissue (Figure 1). Microscopic examination revealed multiple papillary fronds showing downward growth of the epithelium, presence of acute inflammatory infiltrate in the surface epithelium, Brunn’s nests and subepithelial stroma, accompanied by capillary proliferation in the stroma (Figure 2). The final pathologic diagnosis was polypoid cystitis.

On follow-up, the boy was free of pain and voided with a strong, solid stream.

Discussion

Although rare, bladder masses should be included in the differential diagnosis of children presenting with acute onset of hematuria and lower urinary tract symptoms. This boy was ultimately diagnosed with polypoid cystitis. Papillary, polypoid, and bullous cystitis are placed in a continuous spectrum with the same pathologic findings, differing only in the amount of stromal edema and the size of the base1. The presenting symptoms of these tumors include hematuria, obstruction, or inflammation, depending on the type and location of the lesion2. Particularly in the event of obstruction, excision of the lesion before complications occur is of utmost importance.

Although found in up to 80% of the indwelling catheter population, polypoid cystitis is rare in adults without a history of chronic catheterization3. Even more unusual is its discovery in the pediatric population, as a literature review produced only three such patients. Polypoid cystitis was found in a 20-month-old at the time of repairing extensive congenital urinary tract anomalies3, a 10-year-old who presented with gross hematuria and a history of suprapubic trauma2, and a 13-year-old with a 6 month history of intermittent, painless gross hematuria at the end of micturition.

Polypoid cystitis has been associated with urinary tract obstruction in the literature, and meatal stenosis was noted in our patient. To the authors’ knowledge, however, there have been no reported connections of polypoid cystitis to meatal stenosis.

While evaluation of the child with gross hematuria and lower urinary tract symptoms infrequently reveals a correctable cause, prompt ultrasound evaluation and endourologic resection of a mass intermittently obstructing the bladder outlet may result in complete resolution of symptoms.

References

Case Presentation

Prenatal ultrasound for a 35-year-old G1P0 female at 18 weeks estimated gestational age revealed echogenic, hydronephrotic kidneys with a distended, thick-walled bladder, and the classic keyhole bladder neck suggestive of lower urinary tract obstruction (LUTO) (Figure 1). Fetal anatomic survey was otherwise unremarkable. Amniocentesis revealed a normal 46,XY fetal karyotype, normal alpha-fetoprotein and acetylcholinesterase levels. Vesiocentesis revealed normal levels of sodium, chloride, calcium, and osmolality. Amniotic fluid infusion was performed at 20 weeks to provide space for vesicoamniotic shunt (VAS) placement. Fetal presentation resulted in difficulty targeting the bladder through the suprapubic region, but a VAS was successfully placed. At 23 weeks, the bladder was decompressed, suggesting that the VAS was functioning properly. Unfortunately, the mother developed preterm premature rupture of the membranes (PPROM). The amniotic indices between weeks 24 and 27 were at the low end of normal. Circumstances were further complicated by fetal dislodgment of the stent and ensuing fetal ascites, which could not be managed by secondary stent placement, likely eliminated any initial benefit from VAS placement. The right upper quadrant insertion site, in this case, led to intestinal evisceration. As such, the question remains as to whether there is a benefit to percutaneous shunting for LUTO. The PLUTO trial was the most notable effort to answer this question. This randomized trial evaluated VAS placement versus conservative management, with the primary outcome being fetal survival at 28 days. While the study was closed early because of poor recruitment, and data analysis was confounded by significant crossover between the two arms of the trial, the study found a non-significant trend towards improved survival in the intervention arm. (3) Interestingly, only two of the ten survivors at two years had normal functioning kidneys. Though VAS placement has the potential to improve fetal outcomes, many factors, including proper positioning and adequate drainage, need to occur improve the rate of success.

Discussion

VAS is a method by which the fetal bladder is drained into the amniotic sac in the setting of LUTO. The goal is to decrease renal parenchymal damage and to improve pulmonary development. Many complications have been reported after VAS placement, including dislodgement, PPROM, preterm delivery, and injury to the fetus, such as trauma to the abdominal wall or extremity (1, 2). In our case, routine prenatal evaluation led to detection of fetal LUTO. Urine electrolytes suggested that the fetus was a suitable candidate for VAS placement. While insertion of the VAS was uncomplicated, the subsequent fetal dislodgment of the stent and ensuing fetal ascites, which could not be

References


Figure 2. Upon delivery, Potter facies, respiratory distress, and evisceration of the appendix and cecum are evident.
Case Presentation

We present a female neonate born at 37 weeks gestation, whose mother was seen for prenatal urologic consultation at 24 weeks gestation for prenatal ultrasounds demonstrating a presumed omphalocele and bladder extrophy, which, after further review, were concerning for cloacal extrophy.

The patient was born by uncomplicated cesarean section, which was performed prior to reaching full term due to polyhydramnios and intrauterine growth restriction.

Newborn examination revealed an omphalocele and exposed bladder mucosa just inferior, abutting the sac. Pressure applied to the lower abdomen demonstrated efflux of urine from the exstrophied bladder tissue. External genitalia appeared normal, with an identifiable clitoris and separate orifices assumed to be the urethra and vagina. The anus appeared normal and patent and there was no evidence of pubic diastasis. Urine was also noted to pass from the perineum.

An ultrasound of the abdomen performed on the first day of life demonstrated bilateral Society for Fetal Urology (SFU) grade 3 hydrenephrosis, non-visualized ureters, and an anterior fluid-filled structure in the midline of the pelvis suggestive of a partially distended bladder. Also visualized were two parallel, fluid-filled cystic structures posterior to the presumed bladder, which were thought to represent hydrocolpos, hydrometrocolpos, or, possibly, a fluid-filled urogenital sinus.

The patient was brought to surgery in conjunction with pediatric general surgery on day of life two for cystourethroscopy, vaginoscopy, closure of the omphalocele, and repair of the bladder extrophy.

Operative findings included a normal appearing urethra with good coaptation of the bladder neck and a normal appearing bladder with a capacity of nearly 100 mL. There were two ureteral orifices in orthotopic positions. The extrophied bladder mucosa communicated with the bladder lumen through a connecting tract, which was identified by probing with a small feeding tube. This confirmed the diagnosis of superior vesical fissure. There was a normal appearing vaginal introitus. However, vaginoscopy revealed a nearly complete vaginal duplication terminating in two separate cervices, indicating uterine duplication.

After excision of the omphalocele sac, the extrophied bladder tissue was dissected down to a narrow fistula, which was excised. The resultant defect in the bladder was closed in two layers. The abdominal wall was then closed. A Foley catheter was left in situ at the conclusion of the case to allow for bladder decompression.

A voiding cystourethrogram performed on post-operative day seven demonstrated a normal appearing bladder contour and no vesicoureteral reflux. After removal of the catheter, the patient was able to void normally. The patient will follow up as an outpatient for further monitoring and for discussion of plans to address the vaginal duplication.

Discussion

Superior vesical fissure is a rare variant of bladder extrophy, and although the incidence is not well described, one large series with various presentations of bladder extrophy reported one case of superior vesical fissure in 72 patients. Müllerian duplication has been described in cases of duplicate bladder extrophy but, to our knowledge, our patient represents the only reported case of Müllerian duplication in the setting of superior vesical fissure.

References


Figure 1
Ultrasound of the midline lower pelvis shows hydrocolpos with a midline longitudinal vaginal septum that was later seen on vaginoscopy.

Figure 2
Exstrophy of the bladder mucosa associated with the resected omphalocele sac. The fistulous tract to the bladder proper is being cannulated with a feeding tube.
Antenatal Diagnosis of Bladder Exstrophy with Vaginal Duplication

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Case Presentation

A 35-year-old G5P3A1 female, who resided in rural LA, was referred to Maternal Fetal Medicine (MFM) for advanced maternal age and a past history of prolonged labors. She underwent obstetrical screening ultrasound at 20 weeks gestational age (GA), which demonstrated findings concerning for a bladder abnormality in the fetus, more specifically, difficulty identifying the urinary bladder. The diagnosis of bladder exstrophy was confirmed with magnetic resonance imaging (MRI) performed at 24 weeks GA, which demonstrated an intra-abdominal soft tissue mass with no evidence of a urinary bladder, as well as bulging of the abdominal wall just below the level of the umbilical cord insertion. The pregnancy was regularly monitored by her local MFM, and the patient was referred for urologic consultation during her antenatal course. Vaginal delivery was induced at term in our medical complex, and postnatal physical examination revealed classic bladder exstrophy, with a generous bladder plate and a bifid clitoris. Postnatal renal ultrasound identified unilateral pelvicaliectasis.

On day of life 2, the patient underwent a single-stage primary closure of exstrophy at Children’s Hospital of New Orleans. Intraoperatively, the patient was noted to have Müllerian duplication, with two discrete vaginas (Image 1). A 2 cm V-shaped segment of the vaginal septum was excised and the anterior and posterior mucosal edges were reaproximated. Bilateral cross-trigonal ureteral reimplantation was performed, relocating each ureteral orifice further cranial from the bladder neck. After closure of the bladder plate, the bladder neck was reinforced using paravesical tissue. The urethrovaginal complex was left en bloc and advanced posteriorly in Y-V plasty fashion. The pubic diastasis measured 3.5 cm. The symphysis reduction was performed by the orthopaedic surgery team using Buntline hitches placed with non-absorbable suture. Good symphyseal approximation was achieved without the need for osteotomies. We preserved the umbilical stump for cosmesis. Postoperatively, the patient was maintained in a unique bobsled orthosis adapted specifically for bilateral lower extremity traction (Image 2). The hospital course was uneventful and the bobsled orthosis facilitated care at home.

Discussion

Reconstruction of bladder exstrophy in the neonate is a challenging surgical endeavor, and there is controversy about the extent and timing of the different procedures incorporated into the initial repair. Fetal diagnosis of exstrophy allows for early detection, counseling, and prenatal planning. Variants of the exstrophy/epispadias complex, such as concurrent Müllerian abnormalities, which occur in only a small subset of female exstrophy patients, increase the complexity of the repair.

The female neonate with exstrophy should be carefully examined to identify any Mullerian abnormalities. Bladder exstrophy with duplicate vagina can be safely managed with incision of the vaginal septum at the time of primary exstrophy closure, as the ectopic anterior position of the vagina facilitates this maneuver, particularly prior to closure of the pelvis. Antenatal diagnosis of bladder exstrophy affords family counseling, optimal surgical planning, and multidisciplinary coordination for this extensive reconstructive procedure.
Acute and Delayed Presentation of Bilateral Meconium Hydroceles in a Neonate

JR Liggit, Dana Point, Richard Vaughan, Osama Al-Omar

Case Presentation

A 3-day-old male was delivered at 36 weeks gestation with a twin sister via uncomplicated vaginal delivery. He was noted to have a significantly enlarged scrotum, appearing to be fluid filled on palpation with no erythema or apparent tenderness. The scrotum did not transilluminate and an ultrasound demonstrated appropriately sized testicles with normal echotexture, and vascularity on color-flow Doppler. Numerous bowel loops and fluid were also reported to be within the scrotal sac (Figure 1).

Bilateral inguinal hernia repair was conducted on day four. Intraoperative findings demonstrated a right hydrocele sac, which when opened, revealed a large amount of green, gelatinous material with no bowel identified (Figure 1). This material was evacuated and sent to pathology. Orchiopexy was also performed at this time. Pathological specimen was consistent with meconium hydrocele and was negative for malignant cells. The post-operative course was uneventful and the patient was discharged on post-operative day one.

At one and two month follow-ups, soft tissue masses were palpable in the scrotum bilaterally. Ultrasoundography at two-month follow-up revealed normal testicles and complex fluid with echogenic foci in the left hemiscrotum. A right hydrocele was present with minimal echogenic foci within the hydrocele and an overall decrease in size (Figure 2). Bilateral scrotal exploration was performed for evaluation of bilateral meconium hydroceles, revealing a left sided meconium hydrocele and a right sided meconium mass. Left meconium hydrocelectomy with orchiopexy and excision of right meconium mass were performed at 4 months. Pathology reported fibromembranous tissue with meconium staining consistent with meconium hydrocele and the right mass as calcified and mucinous material consistent with meconium mass. At the five month follow-up from the second procedure, the patient was doing well, with normal palpable testicles bilaterally and some residual excess scrotal skin (Figure 3).

Discussion

Meconium hydroceles are rare entities presenting as palpable, soft scrotal masses at birth. They can manifest later as calcified, tumor-like lesions. These hydroceles are a result of in utero perforation of the gastrointestinal tract with meconium spillage into the peritoneum. The meconium subsequently travels through the patent processus vaginalis resulting in a scrotal or labial hydrocele. While in 50% of cases mechanical obstructive lesions can be found, at times the gastrointestinal perforation heals prenatally without gastrointestinal complications (Tai). Initial presentation can be confused with torsion or a hernia. Isolated meconium hydroceles rarely result in complications and small ones can be observed, however given rarity of presentation and possible differential diagnoses, most patients will require exploration (Kizer).

The natural course of the meconium in hydrocele is to harden over the next 3-4 weeks due to calcification and fibrosis. Delayed presentation as calcified masses may be associated with meconium peri-orchitis or can be confused with possible testicular or cord tumors, and therefore should be removed (Tai).

References

Perinatally Detected VUR with Reflux Nephropathy May Cause Hypertension in Infancy

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Case Presentation

A male fetus was monitored by perinatology for bilateral hydronephrosis on 28-week gestational ultrasound. At 36 weeks gestation, ultrasonography showed bilateral renal pelviectasis with AP diameters measuring 12.6 mm on the left and 9.6 mm on the right. Postnatal renal ultrasound confirmed the bilateral hydronephrosis, and voiding cystourethrogram (VCUG) demonstrated left grade IV vesicoureteral reflux (VUR) and right grade III VUR [Figure 1]. Prophylactic antibiotics were instituted and the boy did not experience any urinary tract infections (UTI) or febrile episodes.

At 22 months of age, he was admitted to the hospital with elevated blood pressure and metabolic acidosis. Repeat VCUG demonstrated persistent bilateral VUR and a MAG-3 scan showed left renal function of 40% and right renal function of 60% with normal drainage curves. Initially, the child’s blood pressure was difficult to control and ultimately 2 oral antihypertensive medications were required. CT angiography was obtained to exclude a renovascular etiology of his hypertension and this revealed no evidence of hydronephrosis with normal renal vasculature bilaterally, but obvious left upper and lower pole renal cortical thinning [Figure 2].

At age two, the parents opted for the child to undergo bilateral ureteral reimplantation for definitive surgical management of his VUR, due to concerns about the expected lower rate of resolution of his VUR and the risk that future episodes of pyelonephritis could potentially cause renal scarring and exacerbate his hypertension. Recovery from surgery was uneventful and a postoperative renal ultrasound demonstrated resolution of his hydronephrosis. A repeat VCUG confirmed the absence of VUR. The child has been maintained on Norvasc and Enalapril and at one year postoperatively, his blood pressure has been well controlled.

Discussion

This case illustrates the potential for antenatally detected, bilateral VUR with congenital reflux nephropathy to cause hypertension during infancy, in the absence of a urinary tract infection. Our patient demonstrated evidence of unilateral reduction of renal function, with both upper and lower pole cortical hypoplasia of his left kidney. Renal segmental hypoplasia (RSH), or Ask-Upmark kidney is a rare condition in which a portion of the renal parenchyma is hypoplastic with thickened vessels and thyroidisation of the tubules1. The exact etiology of RSH is debated, but can be attributed to VUR or aberrant renal vasculature, leading to congenital hypoplasia and excessive renin secretion by the abnormal kidney causing hypertension2,3. For a poorly functioning kidney, nephrectomy is an option and for localized RSH, partial nephrectomy can normalize the blood pressure1-3. This would not be appropriate in our patient’s scenario, as he demonstrated good ipsilateral renal function and more diffuse RSH, which prompted correction of his VUR. While he still has hypertension, it is under better control since his initial diagnosis. Absence of any VUR should greatly reduce the risk of any further UTI and inflammatory renal scarring, and avoid worsening of his hypertension.

References
Cardiomyopathy in Infantile Wilms Tumor without Prenatal Care
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Case Presentation
A 7 month old boy was evaluated in our emergency department after his foster parents noted an enlarging abdominal mass in the setting of an upper respiratory infection. His birth and family histories are unknown other than a lack of prenatal care. On work up, he was found to be in hypertensive crisis with cardiomegaly. Imaging revealed a 10 x 9.2 x 9.3 cm heterogeneous mass arising from the right kidney with an intra-caval tumor thrombus, concern for a contralateral renal mass, and a suspicious pulmonary lesion. The patient was admitted to the cardiac intensive care unit for control his hypertension with milrinone and nicardipine drips, with subsequent transition to oral captopril and amiodipine. Once stabilized from a cardiac standpoint, given his young age, evidence of a tumor thrombus, bilateral disease, and concern for atypical tumor histology, he underwent a CT guided percutaneous biopsy of the right renal mass, which confirmed Wilms tumor (WT). Given Stage V disease, he was started on vincristine and dactinomycin, withholding the conventional doxorubicin given his compromised cardiac function. Post-chemotherapy, pre-surgical imaging demonstrated resolution of the tumor thrombus, but limited volumetric response of the primary renal tumors. The patient underwent bilateral partial nephrectomies 6 weeks after diagnosis. Pathology revealed a completely resected, favorable histology, Stage I WT on the right and a nephrogenic rest on the left. The lung wedge biopsy revealed mycobacterium avian complex, but no tumor. The patient is undergoing standard adjuvant vincristine and dactinomycin therapy, with the addition of right-sided flank radiation due to the renal biopsy.

Discussion
Wilms tumor (WT) is the most common renal malignancy in children, with an incidence of 1/10,000 before the age of 15 years. Three-quarters of patients with WT are diagnosed between 3 and 5 years of age, and less than 10% are diagnosed before 6 months of life (1). However, up to 15-25% of these youngest patients are noted to have a renal mass on prenatal imaging1. In terms of presentation, most pediatric renal masses are diagnosed after a palpable mass prompts ultrasonography and subsequent CT or MRI. Hypertension can be present in up to 55% of children with renal tumors2.

Hypertension-induced cardiomyopathy is a rare sequela of WT limited to case reports. In this case, appropriate prenatal care including prenatal imaging may have enabled a prenatal diagnosis and prevented late sequelae of the WT. The effects of the hypertension and subsequent heart failure are likely related to secretion of the vasoactive enzymes renin and angiotensin II 2,3. Hyperreninemia can be present in WT due to renal artery compression or direct secretion from the tumor2. Previous studies have shown the effects of angiotensin II on cardiac remodeling, which include hypertrophy3. Our patient’s hypertensive cardiomyopathy was likely due to the chronic effects of his renal tumor. Now that his WT has been removed, we have observed a gradual improvement in his hypertension and cardiac function, with a reduction in his antihypertensive medications. However, he will require ongoing cardiology follow up.

References

Figure 1

Figure 2

Figure 2: Right Renal Mass

Figure 1a: Right Renal Mass and Cardiomyopathy

Figure 1b: Left Renal Mass
Neonatal Torsion: What’s a Doctor to Do?
Rebecca S. Zee and C.D. Anthony Herndon, University of Virginia Department of Urology

Case Presentation
B.G. presented to the Pediatric Urology service on day of life two. He was delivered at term with APGARS of 8 and 10 at 1 and 5 minutes, respectively. Recurrent fetal decelerations were noted prompting vacuum-assisted vaginal delivery. His mother is a healthy non-smoker free of gestational diabetes.

The newborn exam was unremarkable except for a left-sided hydrocele prompting a scrotal ultrasound (US). The left testis was homogeneous, with a normal Doppler signal and a hydrocele. The right testis was heterogeneous with questionable decreased Doppler signal. The hydrocele was heterogeneous, representing debris (Image 1). The reading was consistent with possible intermittent torsion.

On evaluation by Urology, the child was asymptomatic, non-acute, and the scrotal exam revealed a left non-communicating hydrocele with a non-palpable testis. The right testis was non-tender but concerning for a somewhat thickened tunica vaginalis. The scrotal skin was free of induration or erythema. The child was observed with serial US. Over the next 24 hours, he remained asymptomatic. An interval US revealed increased thickening of the tunica vaginalis, parenchymal heterogeneity, and questionable decreased blood flow compared to 12 hours prior. His physical exam was unchanged aside from increased thickening of the tunica vaginalis.

Based on the above, an informed discussion was undertaken with the family and we proceeded to perform surgical exploration. A scrotal incision was made and the right hemi-scrotum was entered. There was no evidence of extravaginal torsion; however, a clear bottleneck defect was present, suggestive of prior torsion. The thickened tunica vaginalis was incised revealing a tense hematocele (Image 2). There was no evidence of intravaginal torsion. An intraoperative cost/benefit analysis was made and we performed bilateral septopexy to mitigate the possibility of future torsion. The intraoperative findings were attributed to either traumatic delivery or intermittent extravaginal torsion. The patient was evaluated one month post-operatively with returned blood flow to the right testis in comparison to the left and decreased heterogeneity of the right testis.

Discussion
Broderick et al. performed a survey of pediatric urologists to assess practice patterns in regards to neonatal torsion¹. This case most resembles that of a postnatal torsion. Among pediatric urologists surveyed, there was agreement for management of postnatal torsion, with 93% of respondents choosing immediate surgical exploration; ninety-six percent of respondents would perform contralateral orchiopexy.

This case illustrates neonatal torsion in which the clinical picture is not clear at presentation. Classically, a postnatal torsion presents with a firm, tender testis, which was not consistent with the physical findings in this neonate. Aside from the non-tender induration of the tunica vaginalis, the exam was benign. The intraoperative finding of a tense hematocele raises the question of whether the hematocele was contributing to a double compartment syndrome in which edema of the testis within the tunica albuginea and fluid contained within the thickened tunica vaginalis synergystically compromised flow to the testis. Ultimately, the outcome was favorable with successful salvage of the affected testis and preservation of the left testis after bilateral septopexy.

References

Image 2
Right testis delivered through scrotal incision with bottleneck defect observed and no evidence of extravaginal torsion. The thickened tunica vaginalis was opened releasing a tense hematocele surrounding a hyperemic testis.