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FROM THE EDITOR

Anthony A. Caldamone, M.D.

The Dialogues is pleased to continue its affiliation with the Society for Fetal Urology. This organization has provided academic leadership and direction for all of us in pediatric urology. We are all continuing to see a large number of patients who present prenatally and the Society for Fetal Urology has continued provided us with established guidelines in managing these patients both prenatally and postnatally.

The reports presented here come from the Fall meeting of the Society for Fetal Urology, which was held in conjunction with the American Academy of Pediatrics in San Francisco. Each of these contributions is worth reading as each has an important take home message.

My thanks to Jeff Campbell for the coordination of this issue of the Dialogues and to the Society for Fetal Urology for a continued mutually beneficial association.

SPECIAL EDITION:  

SOCIETY FOR FETAL UROLOGY YEAR IN REVIEW

Jeffrey B. Campbell, M.D., SFU Secretary/Treasurer

In 2010, the Society for Fetal Urology (SFU) held its 44th Biannual Meeting in conjunction with the 1st World Congress of Pediatric Urology in San Francisco, CA. Dr. John W. Brock, III gave an excellent Mini State of the Art Lecture entitled Impact of Prenatal Diagnosis on the Practice of Pediatric Urology, and Drs. Tony Herndon and Ibrahim Mokhless moderated an informative and thought-provoking Moderated Poster Session entitled Perinatal Urology.

The 45th Biannual Meeting was held in conjunction with the American Academy of Pediatrics National Conference and Exhibition in San Francisco, CA. The program was entitled Fetal Therapeutics and was chaired by Dr. Carlos R. Estrada. We were treated to a number of outstanding guest lectures (Fetal Intervention – Dr. Hammin Lee, Minimally Invasive Fetal Intervention – Dr. Hiep T. Nguyen, MOMS Trial Update – Dr. Michael Carr, Amniotic Fluid Stem Cells – Dr. Roger De Filippo, and Embryonic Stem Cells – Dr. Richard N. Yu) and abstract presentations. Many of the abstracts presented at this meeting 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.

In 2010, the SFU entered into an affiliation with the Journal of Pediatric Urology (www.jpurol.com). The first subsection of Fetal Urology was published in June, and included The SFU Consensus Statement on the Evaluation and Management of Antenatal Hydronephrosis. 2010 also marked the end of Dr. Tony Herndon’s 3-year term as Secretary/Treasurer of the SFU. Tony has been instrumental in the growth and development of the SFU, and has been the driving force behind the SFU Hydronephrosis Registry. We now have 8 sites with IRB approval, and have enrolled over 380 patients. Additional information regarding the registry can be found on the SFU website (www.sfu-urology.org).

As we look ahead to 2011, 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 SFU will hold its 46th Biannual Meeting in conjunction with the AUA Annual Meeting on Friday, May 13, 2011 in Washington, D.C. Dr. John Thomas has put together an outstanding program entitled Antenatal Hydronephrosis: Why It Happens, What It Means, and Why We Manage It. The program will include a lecture on renal embryology, with a focus on the effects of hydronephrosis on the developing kidney, and a panel discussion entitled Postnatal Management of Antenatally-Detected Hydronephrosis: A Case by Case Discussion. There will also be a number of abstract presentations, to include case reports, case series, and original research. Additional information can be found on the SFU website. See you in D.C.!
Special Edition: Society for Fetal Urology
45th Bi-Annual Fall Meeting, October 2010 / San Francisco, California

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Open Fetal Vesicostomy for Management of Bladder Outlet Obstruction in Posterior Urethral Valves: Two Cases

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Introduction
Prenatally detected hydronephrosis caused by posterior urethral valves (PUV) can be a source of morbidity and mortality in infants. Open fetal intervention is offered through the Fetal Care Center at Cincinnati Children’s Hospital Medical Center to male infants who exhibit evidence of bladder outlet obstruction in association with severe oligohydramnios and favorable urine electrolytes and who otherwise be unable to survive to term without intervention. In fetuses with unilateral renal cystic changes, this option is discussed on a case-by-case basis. We discuss our experience with open fetal vesicostomy in two patients.

Case Reports
Patient A is a male fetus in whom bilateral hydronephrosis and megacystis was identified on prenatal ultrasound. Fetal MRI demonstrated anhydramnios and findings consistent with PUV. Serial fetal urine electrolytes were favorable. After discussing possible treatment options the mother elected to undergo open fetal vesicostomy. He was subsequently born at 35 weeks and required initial intubation. By day of life three, he had a serum creatinine of 3.4 which continued to rise. Ultrasound demonstrated bilateral dysplastic kidneys. Renal function deteriorated and the patient required dialysis. The patient was eventually weaned off the ventilator. An attempt at vesicostomy closure failed due to difficulty catheterizing the urethra and the vesicostomy was reopened. This patient’s renal function continued to deteriorate and he has since received a kidney transplant at 24 months. A vesicostomy is still required for bladder management at this time with future plans for urinary reconstruction.

Patient B is a male fetus with prenatal bilateral hydronephrosis in whom severe oligohydramnios was discovered at 20 weeks. Fetal MRI revealed findings consistent with PUV and left VURD syndrome. Serial fetal urine electrolytes were favorable. Because the prognosis with VURD syndrome is better, open vesicostomy was included as an option for intervention. The mother underwent fetal vesicostomy at 22 weeks. The baby was delivered at 29 weeks. He did not require intubation and was found to have a baseline serum creatinine of 0.3. Postnatal workup demonstrated a left dysplastic kidney with grade 5 VUR. Vesicostomy was closed at 2 months and left nephroureterectomy was performed at 18 months. The patient is now 3 years old and has normal renal function (GFR 96ml/min.).

Discussion
Fetal intervention in prenatally diagnosed bladder outlet obstruction due to PUV is performed in fetuses who otherwise would not survive in utero. Open fetal vesicostomy can be an effective intervention in this population. In these two cases, open fetal vesicostomy was the intervention utilized to prevent fetal demise. Fetal vesicostomy allows for the accumulation of amniotic fluid for lung maturation and provides bladder decompression to decrease pressures in the bladder and upper tracts to prevent further renal injury. Open fetal surgery comes with high risks, but is thought to be more effective than vesico-amniotic shunting at reducing bladder pressures. Treatment options for this population of patients have variable outcomes. Today it remains unclear whether the effect of the vesicostomy on bladder and upper tract pressures will be able to ultimately preserve renal function.

A Description of an Experimental Vesicoamniotic Shunt Causing Ureteral Obstruction and Requiring Open Excision

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Introduction
We describe a newborn boy with a novel vesicoamniotic shunt (VAS) causing left ureteral compression and requiring surgical excision.

Case Report
Pediatric urology was consulted on a newborn male born at 35 and 1/7 weeks gestational age. The baby received prenatal care at an outside institution and a VAS was placed at 17 weeks gestation for presumed posterior urethral valves. On initial examination, the baby’s abdomen was “prune-like” with an ostomy bag over the stent in the left lower quadrant; normal urine output was noted. The newborn ultrasound showed mild right pelvocaliectasis and massive left hydronephrosis with hydroureter. The baby was taken to the operating room on day of life (DOL) #2 for valve ablation and stent removal. Cystoscopy revealed a narrowed bulbar urethra with a Type 3 posterior urethral valve. The bladder was trabeculated and ureteral orifices were not seen. On the left postero-lateral aspect of the bladder, an unknown metal mesh cage-like device was seen embedded within the wall (Fig 1a). The procedure was aborted and a foley was placed. We later determined that the foreign body was an experimental device implanted with the stent to prevent dislodgement (Fig 1b). Because of concern for left ureteral obstruction, the patient returned to the operating room on DOL #11. The mesh was found interdigitated with the detrusor and skin (Fig 1c) and the left ureter was compressed. The device was excised, the bladder was closed, and a foley and ureteral stent were placed for three weeks. He currently displays a neurogenic voiding pattern and is on clean intermittent catheterization.

(continued on next page)
Discussion

Vesicoamniotic shunts are commonly dislodged in utero. Quintero, et al. has reported a technique in four patients in which a ureteral stent is sutured to a commercially available atrial septal device. The double-barrel configuration is designed to prevent dislodgement. In this case, the device did indeed ensure stent retention. Unexpectedly, the detrusor and skin became interdigitated with the mesh and the left ureter was compressed requiring open excision in the newborn period. This case underscores the importance of continuity of care as well as physician communication.

References


A Congenital Segmental Giant Megaureter Presenting as a Fetal Abdominal Mass: Preoperative Diagnosis is Feasible

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Introduction

Congenital segmental giant megaureter (CSGM) is an extremely rare anomaly, with less than 10 cases in the literature. We report a male case of CSGM that presented as a 5 cm fetal abdominal mass detected by prenatal ultrasound.

Case Report

A newborn male was the product of a 39 week gestation with a birthweight of 4648g. Prenatal MRI diagnosis was UPJO with large extrarenal pelvis, but postnatal MRI, VCUG and DMSA scan established the diagnosis of CSGM. Diuretic renogram did not demonstrate functional obstruction (Fig 1), and an elective ureteroureterostomy was performed at 12 months. Bilateral orchidopexies were performed simultaneously. He had bilateral VUR with ectopic ureteral openings to the bladder neck, which were corrected at 18 month. Intraoperatively, bilateral vasa deferentia were found to drain into the ureters (Fig 2). In addition to the GU anomalies, he also presented with neonatal hypoglycemia, segmental aniridia, all suggesting a lesion in chromosome 11, although no apparent genetic anomaly was demonstrated.

Discussion

Although extremely rare, CSGM can be diagnosed preoperatively by a series of modern imaging techniques, and should be included in differential diagnosis of fetal urinary tract dilatation.

References

Non-Identical Twins Associated with Identical Karyotype: An Unusual Case of Chimerism

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Introduction

Chimerism is a rare condition resulting from the fusion of two zygotes at an early stage of development giving rise to separate cell lines within an individual. 46, XX/46, XY can occur from the fusion of two separately fertilized ova during in-vitro fertilization or less commonly during natural pregnancy. Most cases of 46, XX/46, XY are diagnosed early during workup for ambiguous genitalia, however, both normal male and female phenotype have also been reported.

Case Report

Male and female baby twins were referred to urology for a possible intersex workup. The pregnancy was a result of IVF where two fertilized embryos were transferred. Early prenatal ultrasounds performed at 6, 8, and 10 weeks gestation showed one chorion and two amnions, which appeared consistent with twin gestation from a single embryo. Follow-up 20 week ultrasound revealed a male and female fetus. Subsequent blood karyotype analysis revealed identical 46, XX/46, XY (33% and 67% of isolated cells respectively) in both the male and female baby. Examination after birth revealed healthy phenotypically normal male and female babies. Initially these patients were thought to be tetragemetic or true chimeras, however, the possibility of this occurring in two individuals is extremely rare. For this reason, a repeat karyotype analysis from buccal mucosa was obtained that revealed 46 XX in 100% of cells in the female baby and 46 XY in 100% of cells from the male baby.

Discussion

The actual incidence of chimerism is unknown although the condition is assumed to be rare. Up to 20% of true hermaphrodites have been found to be chimeras in previously published reports.1 The most common presentation of is an infant or young child with ambiguous genitalia.123 Phenotypically normal male and female children are unusual and their optimal postnatal workup and management is not well defined. In this particular case both children are hematopoetic chimeras’s as a result of exchange of blood cells via the placenta in utero.

This case brings to light the potential situation that may arise more frequently with increased use of IVF. In almost all cases, monochorionic diamnionic twins are the result of a single embryo that splits soon after fertilization. Although very rare, case reports exist describing monochorionic diamnionic dizygotic twins arising from IVF. The possibility of this scenario exists due to assisted hatching techniques that thin or disrupt the zona pellucida. This increases the likelihood of fusion of the embryo’s leading to a single chorion. Exchange of blood in utero leads to hematopoetic chimerism that likely persists throughout adulthood. Tetragametic chimera should be distinguished from hematopoetic chimera to aid in counseling. Tetragametic chimeras are likely at higher risk for gonadoblastoma and should undergo gonadal biopsy while hematopoetic chimera likely can be observed safely.

References

Intravesical Ureterocele Associated with Renal Duplication Causing Bladder Outlet Obstruction and Contralateral Ureterovesical Junction Obstruction in a Neonate

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Introduction

Ureteroceles, particularly ectopic ureteroceles, have been known to cause bladder outlet obstruction (BOO), but to our knowledge not contralateral ureterovesical junction (UVJ) obstruction.1,2,3,4 We report a case of an orthotopic ureterocele causing BOO as well as contralateral UVJ obstruction and renal failure in a neonate.

Case Report

A newborn boy born at 32 weeks EGA was prenatally found to have bilateral hydronephrosis. Postnatal ultrasound demonstrated a left renal duplication anomaly with a left upper pole cystic moiety draining into a massive intravesical ureterocele. The left lower renal moiety and right kidney demonstrated increased echogenicity. Postnatally he developed urinary retention treated initially with urethral catheterization and subsequent suprapubic cystostomy tube placement. Voiding cystourethrography ruled out posterior urethral valves and/or reflux. Repeat renal ultrasound demonstrated increased right hydronephrosis. Renal scan showed obstruction on the right side and minimal function on the left. He underwent right percutaneous nephrostomy on DOL 20, and antegrade nephrostogram demonstrated complete obstruction at the level of the right UVJ (Figure 1). Creatinine subsequently stabilized (0.8 mg/dL) with the nephrostomy in place and the infant was managed with clean intermittent catheterization after cystostomy was discontinued. At six months of age he underwent cystoscopy/transurethral incision (TUI) of a massive intravesical ureterocele. Antegrade nephrostogram revealed no flow of contrast into the bladder before TUI, but excellent flow through the right UVJ post-procedure (Figure 2). His nephrostomy tube was discontinued one week later and the infant was initially able to void per diaper.

One month later ultrasound demonstrated severe right hydronephrosis, requiring percutaneous nephrostomy tube placement. Cystoscopy revealed a decompressed ureterocele, which prolapsed into the posterior urethra causing BOO and recurrent right UVJ obstruction. He underwent open cystotomy and unroofing of the ureterocele. Free flow of contrast was seen through the right UVJ post-procedure. Subsequent cystography demonstrated marked vesicoureteral reflux (VUR) into both moieties on the left.

He ultimately underwent left nephroureterectomy at nine months of age secondary to marked left VUR and minimal left renal function. The patient is doing well post-operatively with a nadir creatinine of 0.4 mg/dL.

Discussion

It is quite plausible that a massive left ureterocele caused extrinsic obstruction to the contralateral UVJ and BOO. This “dual hit” adversely affected renal embryogenesis with resultant echogenic kidneys and impaired renal function. If the BOO was a significant component during early renal development, then antenatal intervention would likely have been ineffective.

References
Prenatal Discovery of Renal Vein and Inferior Vena Cava Thrombosis

Brian M. Whitley, M.D., Sherry S. Ross, M.D., John S. Wiener, M.D.
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Introduction

This case report documents the prenatal findings and management of a right renal vein thrombosis in a 27-week-old fetus, as well as the postnatal course. Although neonatal renal vein thrombosis is well described in the literature, few case reports document prenatal findings, diagnosis, course, and outcome of prenatal renal vein thrombosis.

Case Presentation

A 45 year old G6P4 female presented with preterm labor at 23 weeks gestation and was admitted for tocolysis. A previous screening ultrasound at 20 weeks of pregnancy showed no evidence of abnormality in the male fetus. At 27 weeks a repeat sonogram revealed bilateral pleural effusions, ascites, a severely enlarged right kidney without hydronephrosis and echogenicity of the inferior vena cava (IVC). Follow-up ultrasounds at 28 and 29 weeks demonstrated resolution of ascites and effusions, however, the enlarged right kidney and echogenic IVC persisted. The pregnancy continued until 31 weeks, when the mother developed rupture of membranes and proceeded to vaginal delivery. Postnatal ultrasound on day of life 1 demonstrated a right kidney measuring 4.3cm with poor corticomedullary differentiation and a right renal vein thrombus extending into the IVC and right renal pelvis. Initial infant thrombophilia labs were negative. There was no history of hypercoagulability disorder in the mother or family. The mother declined additional hypercoagulability testing and genetic testing of her infant. Thrombolytic therapy was avoided due to prematurity of the infant and risk of intraventricular hemorrhage. Creatinine rose to 1.1mg/dL during this first week of life but quickly normalized thereafter. Follow-up ultrasounds on day of life 7 and 22 revealed resolution of the right renal vein thrombosis with improvement in right corticomedullary differentiation but persistence of calcified IVC thrombus. At 3 month follow-up, a MAG-3 renal scan demonstrated a differential function of 68% of the right kidney. There was no evidence of hypertension, and subsequent infant development has been normal.

Discussion

Prenatal renal vein thrombosis is a rarely diagnosed clinical entity. The typical course is either progression to renal atrophy or resolution via thrombus recanalization or development of collateral circulation. In this case, while the renal vein thrombus is not clearly demonstrated on prenatal sonography it is suspected due to IVC clot. Serial prenatal imaging ensured fetal progression and a safe delivery. Postnatal imaging provided a definitive diagnosis of a renal vein thrombus and allowed monitoring of the renal moiety. Unlike typical cases, expectant management resulted in resolution of the thrombus and recovery of differential renal function in the affected kidney.

The etiology of the renal vein thrombus remains unclear. Historically, thrombus formation has been attributed to fetal hypercoagulability or fetal insult such as infection or hypoxia. In this case, there were no known maternal illnesses or fetal insults although no prenatal serologies were performed and hypercoagulability evaluation was limited. Overall, this case demonstrates an excellent outcome with expectant management of a rare clinical event, which resulted in normal differentiation and differential renal function of the affected kidney.

2011 SFU Spring Meeting

The 46th Biannual Meeting of the Society for Fetal Urology

Friday, May 13, 2011
1:00 pm - 5:00 pm

Grand Hyatt Washington
1000 H. Street NW • Washington, DC
**Congenital Hypoplastic Urethra with a Vesicovaginal Fistula in a Girl**

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**Introduction**

Congenital urethral fistula in a girl with a vesicovaginal fistula is extremely rare. Here we report a patient with this condition and our management.

**Case Report**

A 4-year-old girl presents with incontinence and recurrent UTIs. Her mother reports that the child has normal sensation of filling and desire to void but has urgency, frequency and urge incontinence. Examination reveals clitoromegaly that is circumferentially hooded, an absent urethral meatus but normal vagina and labia.

Ultrasound reveals a large-for-age bladder capacity with bladder outflow obstruction with overflow. Vaginoscopy reveals a normal cervix and a hysterosgram shows normal uterus and fallopian tubes. A fistulous communication to the bladder could be seen in the anterior vaginal wall. The tip of the clitoris has a meatus through which a wire could be passed but was blind ending.

Cystotomy was performed through a midline incision. Antegrade cystoscopy reveals two channels out of the bladder. The posterior channel was a vaginal fistula to the anterior vaginal wall. A wire that was passed through the anterior channel exited through the meatus in the tip of the clitoris. We were hopeful to augment the normal urethra, and to achieve this we gained access to the entire length of the urethra by dividing the pubic symphysis. Unfortunately, the urethra was very hypoplastic without any muscular support and fibrotic, and hence, we had to abandon urethral reconstruction. We repaired the fistula with omental interposition, closed the bladder neck and provided a mitrofanoff channel for catheterization. Clitoroplasty was also performed.

**Discussion**

Congenital Y type fistulas in girls are extremely uncommon and in this case could have been secondary to a hypoplastic urethra. Urethral duplication is not considered in this case since all cases reported in the literature on girls with urethral duplication described them with an orthotopic urethral meatus and a secondary channel. It is also interesting to note that the hypoplastic urethra ended at the tip of the clitoris and the presence of clitoromegaly in the absence of masculinizing syndromes in this child. These constellations of findings do not fit into any syndromic pattern and hence an interesting case report.

**Isolated Distal Vaginal Agenesis Masquerading as Recurrent Urinary Infections in an Adolescent Female**

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**Introduction**

Vaginal agenesis is the failure of this vaginal plate to canalize. Embryologically, in the female genetic phenotype, müllerian ducts form around the 6th week of fetal development and give rise to the proximal 2/3rd of the vagina, cervix, and uterus. Around the 10th week, the müllerian ducts contact a thickened portion of the urogenital sinus called the sinovaginal bulb that ultimately becomes the distal 1/3 of the vagina. This vagina finishes canalization during the 3rd to 5th month of embryological development.1-3 Vaginal agenesis comes as a spectrum from partial to complete and is mostly associated with a variety of müllerian abnormalities.4 Isolated vaginal agenesis distal to a normal cervix and uterus is exceedingly rare with fewer than 3 case reports in the literature.5,6

**Case Report**

A previously healthy 12-year-old girl was referred to pediatric urology reportedly with recent onset of recurrent urinary tract infections (UTI), microscopic hematuria, intermittent abdominal discomfort and dysuria. Her prior urologic history is unremarkable; she is continent, and has regular bowel habits. She had not yet started menstruation. She was a healthy appearing adolescent female with a normal abdominal examination. Her genitourinary exam revealed a Tanner stage 4 female with severe labial adhesions such that neither her urethral meatus nor vaginal introitus could be visualized. An ultrasound revealed an absent left kidney with compensatory hypertrophy of the right renal unit without hydroureteronephrosis. The urinary bladder was unremarkable; however the vagina was distended with echogenic debris representing hematocolpos (Figure 1). The patient underwent an examination under anesthesia, cysto-vaginoscopy, (continued on next page)
and lysis of labial adhesions. When the labial adhesions were separated, a normal urethral and anus were noted, yet no opening in the perineum representing a vaginal communication was present. Cystourethroscopy revealed normal urethra, without evidence of vaginal communication. A digital rectal examination revealed a smooth external mass anterior to the rectal wall approximately 5cm proximal to the anal verge.

An MRI revealed a complex malformation with an absent distal 1/3rd of the vagina along with hydrometrocolpos (Figure 2). A solitary orthotopic cervix was noted, normal left ovary and right ovary were unidentified. Due to the paucity of any vaginal tissue between the distal urethra and anus, she was diagnosed with distal vaginal agenesis.

Operative intervention was undertaken in which the hematocolpos was drained and distal vaginoplasty were completed through a perineal incision. The patient was discharged home with a patent vagina to start vaginal dilation 2 weeks postoperatively.

The patient had considerable psychological difficulties regarding genital reconstruction, starting menstruation, and need for vaginal dilations. During follow-up, she improved her compliance with extensive counseling and family assistance. The patient began menstruating immediately, and at 4 month follow-up, the vaginal introitus remains patent and the patient is back to baseline activities.

Discussion

Isolated distal vaginal agenesis is exceedingly rare yet the consequences of this condition can be both physically and psychologically severe. In this case, the labial adhesions were so severe that the urethral meatus was hidden and the perineal anatomy was profoundly distorted on direct inspection. A focused genitourinary exam performed as a first step in the workup of a child with new onset urologic complaint will rule out many congenital and surgically correctable abnormalities.

The incidence of obstructive Müllerian anomalies is estimated to be around 0.1%. While distal vaginal agenesis is a small subset, it is a surgically correctable birth defect. Several techniques have been described to repair this spectrum of vaginal malformations; the most important aspect in the reconstructive axis is the delineation of the patient’s internal and external female reproductive anatomy. MRI has become the gold standard preoperative assessment tool for internal female pelvic anatomy. In addition to operative planning, knowing the reproductive anatomy is essential to patient counseling with regards to prognosis and future sexual development.

The psychological aspect of the disorder comes with presentation at a precocious time in adolescent development. A young woman must now confront her sexuality, menstruation, and the need for surgical reconstruction and routine vaginal dilations to become functionally and sexually normal. The psychological barriers abound, necessitating good parent and child education with psychological support. This aspect reemphasizes the advantages of earlier diagnosis and treatment, thereby minimizing the psychological aspects around puberty.

While rare, this case of vaginal agenesis reinforces a most basic lesson that is time tested by all physicians: good clinical acumen along with a proper physical exam is the cornerstone of every clinical encounter.

References

Tubularization of the Urethra for Repair of an Incompetent or Absent Urethra

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Introduction
Repair of the incompetent or absent urethra in females is a surgical dilemma. Several procedures have been described which require an abdominal approach and which can be associated with specific problems. We describe a novel technique to create a continent, competent urethra and bladder neck using a completely perineal approach.

Case
A 5-year-old female with vaginal agenesis and absence of the urethra presented with total incontinence. She initially underwent a period of observation without resolution of her symptoms and by the age of 5 she remained completely incontinent. Preoperatively, urodynamic testing was performed which confirmed continuous urinary incontinence starting at 35 cc of water. The bladder neck was reconstructed using a novel, exclusively perineal approach with tubularization, elongation and fixation of the basically absent urethra to create a more physiologic urethra and a continent bladder neck (Figure 1). Six months after surgery the patient is doing well and has only minimal stress urinary incontinence which is managed with a single pad daily.

Discussion
Repair of the incompetent or absent urethra and bladder neck in the female is a surgical dilemma because of the close proximity of the bladder neck to the female perineum. Most surgical techniques involve an abdominal approach and each has its drawbacks. We describe a novel technique to tubularize, elongate and fix the short, incompetent or absent female urethra which creates a competent bladder neck and continent urethra; all through a perineal approach. (Figure 1) This technique was originally described by Mr. Philip Ransley in a personal communication regarding repair of the female epispadias but works equally well in cases such as this. Seven of our patients with bladder neck incompetence or female epispadias have undergone this procedure. Three patients are completely continent, 2 are too young to evaluate (pre-toilet training) and 2 require a single pad per day to manage minimal stress incontinence at a maximum of 8 years of follow-up.

Figure 1: Photographs from this patient’s surgery and diagrams from the original communication from Mr. Philip Ransley showing the repair of the incompetent or absent urethra. A) Preoperative view. The arrow indicates the urethral opening and bladder neck. The vagina is congenitally absent. B) Stay sutures are placed on the urethra to place it on stretch. C) The dissection is carried around the urethral “plate” to create a strip of urethra that can be tubularized. D) The urethra is tubularized over an 8 French catheter. Generally 2-3 cm of usable urethral length can be achieved. E) The mid urethra is fixed as far posterior as possible to create angulation and a mechanism for continence. F) The distal urethra is brought forward as much as possible and fixed to the perineal skin to create a normal meatus. G) Postoperative appearance of the reconstructed urethra.
Pyeloplasty in a Preterm Infant

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Introduction
The management of ureteropelvic junction obstruction (UPJO) in the neonate has been argued. Observational and surgical approaches have been advocated, with proponents of each demonstrating successful outcomes. Pyeloplasty has been shown to be safe and effective in very young infants. We present the case of a premature infant with a UPJO in a functionally solitary kidney.

Case Report
AA was followed throughout gestation for a history of antenatal hydronephrosis. He was delivered by Caesarean section at 35 weeks secondary to oligohydramnios (amniotic fluid index 3.9 cm, deepest pocket 2.1 cm). Prenatal ultrasound suggested bilateral cystic dysplastic kidneys with right sided obstruction. On day of life (DOL) 1, he voided and his creatinine was 1.3 mg/dL. The initial renal/bladder ultrasound was consistent with prenatal imaging. There were no respiratory issues. A voiding cystourethrogram was negative for reflux or other abnormality. Over the next several days creatinine continued to climb to a level of 2.5 mg/dL by DOL 5. Serial ultrasounds demonstrated progressive worsening of the right sided hydronephrosis. A percutaneous nephrostomy (PCN) tube was placed. A controlled post-obstructive diuresis and steady improvement in the creatinine followed. An antegrade nephrostogram was performed and clearly demonstrated a right UPJO. On DOL 12 a right pyeloplasty and circumcision were performed. The renal parenchyma contained cysts of varying size. A ureteral splint and new nephrostomy tube were left in place and sequentially removed in follow-up. At the time of surgery the initial nephrostomy tube was seen traversing through-and-through the kidney, making the procedure more difficult to complete secondary to urinary extravasation with a surrounding inflammatory response (Figure 1). Creatinine reached a nadir of 0.9 mg/dL, post-operatively. Ultrasound appearance of the kidneys has been stable.

Discussion
Obstruction in a solitary kidney is generally regarded as an indication for emergent treatment. In neonates, this may initially be managed with PCN. While this option is less invasive than open surgery, tube placement and management in the neonate can be difficult. In the case of UPJO in premature infants, the optimal timing of reconstruction remains to be elucidated.

References
An Unusual Variant of Covered Bladder Exstrophy

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Introduction
Bladder exstrophy has an incidence of 3.3 per 100,000 live births, but covered bladder exstrophy is a rare variant of the exstrophy-epispadias complex not commonly reported in the literature.1,2,3 The most accepted theory for the embryologic cause of the exstrophy-epispadias complex was postulated by Marshall and Muecke, which states that an abnormal cloacal membrane prevents proper midline development by preventing the mesenchymal tissue from migrating medially to properly close the lower abdominal wall.4,5

Case Report
The patient is a normal birth weight infant born at 37 weeks gestation whose 20-year-old mother was followed by the high-risk obstetrics clinic for multiple fetal anomalies identified prenatally. Prenatal ultrasounds and an MRI, performed at 30 weeks 5 days gestation, demonstrated hydrocephalus, tethered cord with sacral dysraphism, and imperforate anus. Prenatal karyotype demonstrated a 46, XY genotype. No abnormal urinary findings were suspected prenatally since the bladder was closed and no hydronephrosis was present.

After delivery, the patient was found to have a low set umbilicus and a dorsal hood deformity where stool and urine drained. Urine was noted from the tip of his penis as well. Initial examination revealed bilateral descended testes and diastasis of his pubic symphysis approximately 2 cm. A mucous membrane was noted over the dome of his bladder extending from the low set umbilicus to the pubic diastasis. Bilateral inguinal hernias were palpated. Retroperitoneal ultrasounds demonstrated kidneys of normal size, position, and echogenicity, with right grade 1 hydronephrosis. A voiding cystourethrogram revealed that the two openings were connected in the vicinity of the bladder neck. In addition, left vesicoureteral reflux was identified and a fistula was found between the urinary system and a dilated hindgut.

Discussion
Covered bladder exstrophy is a rare variant of the exstrophy-epispadias complex, which may not be obvious in the prenatal work-up. It is important to think of the possibility of covered bladder exstrophy in cases with suggestive features so that the parents and physicians may have time to prepare for the necessary treatment of the child. In addition, this case provides an example of a patient with imperforate anus and covered bladder exstrophy, a variant not classically associated with bladder exstrophy or cited in the literature.

References
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Duplicated Bladder Exstrophy with Posterior Urethral Valves

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The patient was taken to the operating room by the general surgery and urology teams with plans for a cystoscopy and diverting colostomy to be performed. Examination of his urethra revealed a patulous bladder neck opening and an enteric fistula in the posterior bladder wall at the trigone in between the ureteral orifices. Exam of the fistulous opening in the dorsal hood of the foreskin revealed that it communicated with the anterior bladder with an approximately 2 cm defect observed. A diverting colostomy with mucous fistula was performed along with a Ladd procedure for an incidentally found malrotation. A staged repair of his bladder exstrophy with closure of the enterovesicular fistula and osteotomies is planned at six months of age.

Introduction
Bladder exstrophy is a rare condition with an incidence from 1:10,000 to 1:50,000.1 There are four classifications of bladder exstrophy including: classical exstrophy, cloacal exstrophy, isolated epispadias, and exstrophy variants. Sub-classifications of exstrophy variants include covered exstrophy, pseudoexstrophy, duplicate exstrophy, and superior vesical fissure.2 These variants all have the presence of a musculoskeletal defect associated with classic exstrophy but no significant defect of the urinary tract. We report a case of a boy with the unusual combination of duplicated bladder exstrophy and posterior urethral valves.

Case Report
A four-month-old boy presented after a history of antenatal hydronephrosis and irregularities on abdominal ultrasound performed at four days of life. He was referred for these abnormalities and a “skin tag” on the abdominal wall by their local primary care physician. The patient had a normal physical exam except that midway between the umbilicus and the pubic symphysis he had what appeared to be pedunculated bladder mucosal mass.

Radiology studies were performed including renal ultrasound and voiding cystourethrogram. On US the right kidney was normal in size and contour, the left kidney showed grade IV hydronephrosis and an extrarenal pelvis measuring 10mm. The left ureter was dilated from the renal pelvis to the level of insertion on the bladder. The bladder was mildly trabeculated with a thickness of 3mm. Additionally, in the midline there was a soft tissue mass above the fascia but below the skin measuring 1.5 x 5 cm. VCUG demonstrated no reflux, nor any

(continued on next page)
communication between the bladder and the anterior soft tissue mass. There was significant prostatic urethral dilation, sharp urethral angulation, and a urethral filling defect consistent with urethral valves.

The patient underwent cystoscopy and excision of the bladder mass. Cystoscopy confirmed type 1 annular posterior urethral valves, which were ablated, and the anterior mucosalized mass was explored and excised completely. The mass was found to attach to the anterior bladder wall; however it did not have any luminal connection with the underlying bladder. Final pathology of the mass showed urothelial epithelium surrounding bundles of smooth muscle consistent with duplicated exstrophy.

**Discussion**

Duplicated exstrophy is a sub-classification of exstrophy-epispadias complex. It is theorized to be caused by a persistent cloacal membrane which then ruptures later in development and a portion remains isolated on the surface of the abdominal wall. Posterior urethral valves are theorized to be caused by abnormal migration of mesonephric ducts with resulting midline fusion. Posterior urethral valves are believed to occur between the embryologic 4-10 weeks of development. There is no consensus when duplicated exstrophy would arise, however it is thought to occur later in development than urethral valves. Both conditions could be related to inappropriate tissue interaction or gene regulation leading to abnormal development. More likely, this may represent two separate processes occurring simultaneously. There have been case reports regarding duplicated exstrophy and associated urogenital abnormalities. To our knowledge this is the first case report of covered exstrophy and concurrent posterior urethral valves.

**References**


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**Bladder Exstrophy with Anorectal Malformation: A Case Series of 3 Patients**

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**Introduction**

Bladder exstrophy (BE) represents a distinct presentation of the rare urogenital malformation referred to as the exstrophy/epispadias complex (EEC). Incidence of BE is estimated to be between 1:10,000 and 1:50,000 live births. Other variants along the continuum of the EEC, particularly cloacal exstrophy (CE), often are associated with significant gastrointestinal anomalies. Historically, similar gastrointestinal anomalies are not found in cases of BE. We report a series of 3 patients with BE found to have concurrent anorectal malformation.

**Case Series**

Patient 1 presented following C-section delivery at 37 weeks gestation with BE, imperforate anus, omphalocele, left hydrenephrosis with megaureter, left cryptorchidism, and epispadias. A perineal fistula was initially dilated with Hagar dilators until age 2 weeks at which point he underwent perineal anoplasty at time of exstrophy repair with ostotomies, repair of omphalocele, ureteral reimplant, orchioepxy, and umbilicoplasty. The patient initially did well, but later developed prolapse of his repair and eventually required repeat closure with external fixation at 12 months of age.

Patient 2 presented shortly after birth with BE, imperforate anus with a fistula to the bladder plate, colonic duplication superior to the bladder, epispadias, cryptorchidism, omphalocele, and a dysplastic left kidney. He underwent initial closure of omphalocele and diverting colostomy, and recently underwent extensive reconstruction including takedown of colostomy with temporary ileostomy, anorectal pull-through, exstrophy closure with creation of Mitrofanoff catheterizable stoma, ostotomies with external fixation, left nephrectomy, orchiopexy, and ureteral reimplantation.

Patient 3 presented on day 1 of life following delivery at 38 weeks gestation. In addition to trisomy 21, patient was found to have BE, imperforate anus with anterior anal displacement, and epispadias. He has undergone exstrophy repair with repair of dorsal curvature and epispadias, and has responded well to anal dilation thus far.

**Discussion**

The exstrophy/epispadias complex presents a need for extensive reconstructive repair, further complicated by concurrent gastrointestinal anomalies. The association of CE and anorectal anomalies is well described in the literature; however, description of the association of BE and anorectal malformation has been limited, or identified as part of the OEIS Complex (Omphalocele-Exstrophy-Imperforate Anus-Spinal Defects). Proposed mechanisms for BE include abnormal mesodermal migration or cloacal membrane abnormality that leads to displacement of medial structures and membrane rupture. Severity of the exstrophy is correlated with the location and timing of the cloacal membrane defect. Anal and rectal tissues are thought to develop from the dorsal portion of the cloacal cavity, and potential field change defects may lead to abnormal development and combined exstrophy and anorectal anomalies. Our series highlights the rare association of bladder exstrophy with anorectal malformation, as well as the subsequent need for a multidisciplinary approach to care and surgical reconstruction.

**References**

Juvenile Granulosa Theca Cell Tumor of the Testis: A Case Series

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Introduction
Stromal testes tumors make up just less than 10% of testicular neoplasms and include Leydig cell, Sertoli cell, undifferentiated sex cord-stromal (SCST), and juvenile granulosa theca cell (JGTC).¹ Although JGCT is the most common of these accounting for 1.2% of all tumors in the National Prepubertal Testis Tumor Registry, only 11 cases had been reported, as of October 2002.² It is important to differentiate JGTC from other testicular neoplasms such as gonadoblastomas and yolk sac tumors.³ This can be accomplished using microscopic as well as immunohistochemical special staining.¹-⁵ The usefulness of tumor markers in the neonate population remains to be determined.⁴ Partial orchiectomy and inguinal orchiectomy have been used to treat JGTC and there have been no reported cases of metastasis or recurrence.⁵ We report three cases of neonatal testis tumors ultimately diagnosed as juvenile granulose theca cell tumor and treated with inguinal orchiectomy.

Case 1
A 12-day-old neonate presented with swollen left testis. Ultrasound revealed cystic lesions with moderate blood flow. Tumor markers were normal/ A left inguinal orchiectomy was performed. Microscopic analysis showed a well-confined, intra-testicular neoplasm composed of variable sized cystic follicles lined by granulosa cells and separated by fibrous septa. Histological staining of the lesion was positive for inhibin. The eosinophilic fluid of the follicles was positive for mucicarmine. Staining was negative for placental alkaline phosphatase (PLAP).

Case 2
A 3-week-old diagnosed with left undescended testis at birth underwent evaluation. Ultrasound revealed an enlarged, echogenic, intra-abdominal testes. AFP was 8274. HCG was <1. A left laparoscopic orchiectomy was performed. Microscopic analysis revealed a well-circumscribed intra-testicular lesion with variable sized cystic follicles surrounded by areas of cells in sheets, nodules, and irregular clusters. Histologic staining was positive for inhibin. The eosinophilic fluid of the follicles was positive for mucicarmine. Staining was negative for PLAP.

Case 3
A 6-day-old neonate presented with a left testicular mass. Ultrasound revealed a heterogeneous, well-vascularized, cystic mass. Tumor markers were normal. A left inguinal orchiectomy was performed. Microscopic analysis showed a well-confined, intra-testicular neoplasm composed of variable sized cystic follicles lined by granulosa cells and separated by fibrous septa. Histological staining of the lesion was positive for inhibin. The eosinophilic fluid of the follicles was positive for mucicarmine. Staining was negative for PLAP.

JGTC tumors of the neonatal testis are rare with only a handful of cases. We present 3 cases of testes tumors treated with surgical excision. Partial orchiectomies were not performed in any case because the lesion appeared to occupy the entire testis on ultrasound. All specimens were intra-departmentally reviewed by all pathologists at our institution and ultimately diagnosed as JGTC tumor. Follow-up has been less than one year with no evidence of recurrence or metastasis. These will help expand the limited body of literature involving these rare tumors.

References
Triorchidism or Complete Separation of Vas Deferens with Congenital Torsion

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Introduction

Both polyorchidism and complete separation of the vas deferens and testicle are rarely reported congenital anomalies. We present the case of an infant with a non-palpable left testis who underwent diagnostic laparoscopy and was found to have maldescent and torsion of a supernumerary abdominal testis and ipsilateral main testis versus a complete separation of vas deferens with ipsilateral congenital testicular torsion. In either case, this represents an unusual presentation of a rare testicular anomaly.

Case Report

An 8-month-old male who presented for evaluation of a non-palpable left testis underwent diagnostic laparoscopy. The right testicle was normally descended with normal appearing spermatic vessels and vas deferens and a closed internal ring. The left spermatic vessels and vas deferens were noted to enter a closed internal inguinal ring (Figure 1). A divergent branch from the left spermatic vessels entered an intra-abdominal hemosiderin laden testicular remnant without associated vas deferens (Figure 1). The left internal ring was opened and the left spermatic vessels and vas deferens ended into an inguinal atrophic testicular remnant. On histology, the intra-abdominal left testicular remnant demonstrated fibrous tissue with calcifications and hemosiderin deposition consistent with testicular atrophy. The left inguinal testicular remnant demonstrated benign fibroconnective tissue and vas deferens.

Discussion

Polyorchidism is a rare genitourinary anomaly of unknown embryological origin with less than 200 cases reported in the literature. In the largest review to date, the average age of presentation is 17 years with the majority of supernumerary testes left-sided (65%), either scrotal or inguinal in location (89%) and drained by a vas deferens (64%).1 In this same review, an estimated 52% were found to have age-appropriate testicular tissue and spermatogenesis. Intra-abdominal location (9%), lack of draining vas deferens (18%) and associated torsion (15%) were all rare features. The present case would represent not only an exceptionally young age of presentation for polyorchidism but is unusual in the intra-abdominal location of the supernumerary testis, the absence of connecting vas deferens and the presence of maldescent and atrophy (presumed congenital torsion) of both the main testis and supernumerary testis. Complete separation of vas deferens and testicle with congenital ipsilateral torsion, however, cannot be excluded. Testicular fusion anomalies have been reported to occur in 36% of cases of cryptorchidism, however, cases of complete separation have only rarely been reported and are thought to represent just 1.2-5% of cases.2, 3

Management was ultimately guided by considerations of the location and functional status of the testes as well as the risk for malignancy. All reported cases of complete separation of vas deferens and testicle have been associated with cryptorchidism,2 which is a known risk factor for malignancy. Of cases of polyorchidism 1-7% are associated with malignancy, however, cryptorchidism is believed to be the most important risk factor in such cases.1 In the present case, the non-functional appearance of both supernumerary and main testis and testicle have been associated with cryptorchidism,2 which is a known risk factor for malignancy. Of cases of polyorchidism 1-7% are associated with malignancy, however, cryptorchidism is believed to be the most important risk factor in such cases.1 In the present case, the non-functional appearance of both supernumerary and main testis and testicle have been associated with cryptorchidism,2 which is a known risk factor for malignancy. Of cases of polyorchidism 1-7% are associated with malignancy, however, cryptorchidism is believed to be the most important risk factor in such cases.1 In the present case, the non-functional appearance of both supernumerary and main testis and testicle have been associated with cryptorchidism,2 which is a known risk factor for malignancy. Of cases of polyorchidism 1-7% are associated with malignancy, however, cryptorchidism is believed to be the most important risk factor in such cases.1 In the present case, the non-functional appearance of both supernumerary and main testis and testicle have been associated with cryptorchidism,2 which is a known risk factor for malignancy. Of cases of polyorchidism 1-7% are associated with malignancy, however, cryptorchidism is believed to be the most important risk factor in such cases.1 In the present case, the non-functional appearance of both supernumerary and main testis and testicle have been associated with cryptorchidism,2 which is a known risk factor for malignancy. Of cases of polyorchidism 1-7% are associated with malignancy, however, cryptorchidism is believed to be the most important risk factor in such cases.1 In the present case, the non-functional appearance of both supernumerary and main testis and testicle have been associated with cryptorchidism,2 which is a known risk factor for malignancy. Of cases of polyorchidism 1-7% are associated with malignancy, however, cryptorchidism is believed to be the most important risk factor in such cases.1 In the present case, the non-functional appearance of both supernumerary and main testis and testicle have been associated with cryptorchidism,2 which is a known risk factor for malignancy. Of cases of polyorchidism 1-7% are associated with malignancy, however, cryptorchidism is believed to be the most important risk factor in such cases.1

References


FIGURE 1: Laparoscopic image of left intra-abdominal testis and spermatic vessel branch (*) and main spermatic vessels and vas deferens entering closed left internal ring (**).