FROM THE GUEST EDITOR

Douglas W. Storm, MD

As highlighted below by Drs. Giel and Liu, it was quite a year for the SFU. We would like to thank all those who participated in the virtual “Cases and Cocktails” session in September as well as the presenters in the inaugural “Cases and Conundrums” session held during the in person (YEAH!!) Fall Congress in December. We would like to also thank all the audience members from both sessions for their participation and perceptive questions. All of the cases were expertly presented, with each making us consider embryology as well as the different management options and ethical dilemmas.

Inside you will find a synopsis from the presenters who kindly submitted their case reports from both the September and December sessions. I would like to thank them for their willingness to contribute to this edition of Dialogues in Pediatric Urology, so that each of us can learn from their experiences and expertise. These rare and interesting cases contribute to making the SFU unique and help create this distinctive edition of the DPU.

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Although many of us had high hopes for getting back to “normal” this past year, 2021 has continued to remind us that the unexpected has become the expected. While an in-person AUA was not possible, with the hard work of Doug Storm and Candace Granberg, the SFU was able to host a second edition of the “Cases and Cocktails” virtual webinar on Sept. 9th. The meeting was attended virtually by 73 participants who enjoyed nine excellent presentations. The top three presentations, which will be published in an upcoming Gold Journal, were:

1. **Collateral Damage from Fetal Interventions: Increasing the Complexity of Complex Patients**  
   Niki N. Parikh M.D., M.B.A., M.S.B.A.; Patricio C. Gargollo M.D.; Candace F. Granberg M.D

2. **Bilateral Ureterovesical Junction Obstruction Resulting in Anuria in a Three-Week-Old Infant**  
   Joseph A. Baiocco MD, Neha R. Malhotra MD, Andrew Tam MD, Fernando A. Ferrer MD

3. **Persistent Anhydramnios and LUTO in a Patient with a Vesicoamniotic Shunt**  
   Arthi Hannallah MD, Zoe G. Baker PhD MPH, Andy Y. Chang MD, Joan S. Ko MD

After nearly two years of virtual meetings, we were fortunate enough to finally meet in person for the Fall Congress in December. While enjoying the warmth of Miami, we were treated to an excellent program organized by Marcos Machado. The program started with a panel featuring Drs. Rebeca Caridad Martinez and Cherie Foster that focused on the COVID-19 experience from the OB-GYN’s and neonatology’s perspectives. Dr. Rubin Quintero then gave a fantastic talk reflecting on the past and current status of vesico-amniotic shunts.

This was followed by a panel discussion on the status of prenatal diagnosis and intervention in South America, featuring the experiences of Drs. Miguel Castellan from Argentina, Marcos Machado from Brazil, and Francisco Reed from Chile. The SFU portion of the Congress concluded with an outstanding Cases and Conundrums interactive session moderated by Dr. Marie-Klaire Farrugia, in which challenging cases were submitted by SFU members or fellows and were presented and discussed by an expert panel consisting of Drs. Candace Granberg, Micah Jacobs, Doug Storm, and Vijaya Vemulakonda. The top three cases which will also be highlighted in an upcoming edition of the Gold Journal were:

1. **Prenatally Diagnosed Posterior Urethral Valves: Ethical Dilemmas of Fetal Intervention**  
   Drs. Kristen Meier, Margret Bock, Jacqueline Glover, Nicholas Behrendt, Regina Reynolds, Mariana Meyers, and Vijaya Vemulakonda.

2. **To Divert or Not?**  
   Drs. Arun Kelay, Caroline Shaw, Christopher Lees, Marie-Klaire Farrugia, and Abraham Cherian.

3. **Abdominal Evisceration: A Rare Complication After in Utero Vesicoamniotic Shunt Placement**  
   Drs. Cinthia Galvez, Daniel Nassau, Yisel Babstro, Matt Swirsky, Miguel Castellan, Andrew Labbie, and Rafael Gosalbez.

As we look back at 2021, we can take pride in the steps we have taken towards returning to normalcy. The Society continues to grow in its membership and looks forward to welcoming even more members in the upcoming year. We encourage all those interested to apply! In the upcoming year, we look forward to hosting our traditional Interesting Case presentations as part of the Spring SPU/AUA meeting and another excellent annual meeting in the Fall chaired by Dr. Elizabeth Malm-Buatsi.

Hopefully we will see everyone in New Orleans!
Discovery of Cloacal Anomaly in a Patient with Presumed Rectovestibular Fistula

Abstract

We present a case of a newborn female with a cloacal anomaly. Due to a difficult physical exam and indeterminate imaging, she was presumed to have a rectovestibular fistula (RVF). Only by cystoscopy were we able to correctly identify a cloacal anomaly with multiple blind-ending pits. This case demonstrated the importance of cystoscopy in the evaluation and diagnosis in pediatric patients with complicated external genital anatomy.

Introduction

Cloacal anomaly is a rare congenital malformation seen in about 1 in 50,000 live births. It is often correlated with VACTERL and 14% of cloacal anomalies have renal ectopia. External anatomy can be deceiving and often imaging can be unreliable, which can lead to misdiagnosis. Increased costs, increased resource use, over testing, and incomplete surgical treatment can result from misdiagnosis. Our case of a newborn with VACTERL association, crossed-fused renal ectopia (CFRE), and cloacal anomaly presumed to be RVF demonstrates the difficulty in establishing a correct diagnosis in the setting of complicated external genital anatomy.

Case

A female with tethered spinal cord, persistent left superior vena cava, patent ductus arteriosus, atrial septal defect, patent foramen ovale, imperforate anus with possible RVF, VACTERL association, left to right crossed-fused lower pole renal ectopia, and duplicated left collecting system was born at 37w5d and was admitted directly to the NICU for respiratory distress.
**Discovery of Cloacal Anomaly** *(continued from previous page)*

**Discussion**

This case demonstrates the importance of cystoscopy in the evaluation of challenging external genital anatomy. In the setting of difficult anatomy and unclear imaging, the patient was initially diagnosed with RVF. However, cystoscopy was able to elucidate the correct diagnosis of cloacal anomaly and establish that the presumed RVF was a blind ending pit. This finding thus allowed the appropriate treatment plan. Ashour et al. found that cystoscopy is more accurate in the diagnosis of cloacal anomaly compared to contrast imaging studies with 75% and 50% accuracy respectively. The result of misdiagnosis can lead to increased costs, increased resource use, and over testing. In addition, a misdiagnosis in this case may have resulted in repair of the rectal component without addressing the entire problem. It is possible that cystoscopy used earlier may prevent excessive and costly testing; an interesting area of further research and cost-effective analysis.

**Conclusion**

This is an interesting case of cloacal anomaly associated with VACTERL and CFRE. The importance of cystoscopy is highlighted here as the patient was initially presumed to have a RVF based on a difficult physical exam and unclear imaging, but was ultimately diagnosed with a cloacal anomaly after cystoscopy.

**Bibliography**


**Figure 2.**

- b. View from cloacal opening. Urethra at 1 o’clock position, vagina at 7 o’clock position.
- c. Bladder neck.
- d. View at the distal vagina just proximal to the cloaca. Rectovaginal fistula opening on picture left. Cervix and uterine opening in the middle. Blind ending pit on the picture right.
- e. View within the rectum with stool.
Ectopic Ureterocele Obstructing the Bladder Outlet

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Introduction

A ureterocele is a cystic dilatation of the distal ureter, which may be orthotopic or ectopic; associated with a single or duplex system; and may or may not be associated with vesicoureteral reflux (VUR). A ceceuroterocele is a type of ectopic ureterocele, which was first described by Dr. Douglas Stephens in 1971 as a “a bizarre form of vesico-urethral ureterocele,” with a wide incompetent ureteral orifice in the bladder and a long “tongue” extending submucosally beyond the bladder neck (BN) to the level of the urethra.1

Case Report

A pregnant woman presented at 23 weeks of gestation for a prenatal ultrasound, demonstrating a male fetus with a right multicystic dysplastic kidney (MCDK), right hydroureter, and right ureterocele, with a normal appearing left kidney and normal amniotic fluid index. Based on these findings, we (as pediatric urology consultants) recommended postnatal antibiotic prophylaxis as well as a renal-bladder ultrasound (RBUS), voiding cystourethrogram (VCUG), and circumcision. Labor was induced after oligohydramnios was detected and the child was born at 38 weeks and 2 days. A day of life (DOL) 1 RBUS newly demonstrated a possible cecoureterocele (Figure 1). On DOL 2, a VCUG found no VUR and demonstrated a ureterocele at the bladder base, a dilated BN and posterior urethra (PU), and a possible filling defect near the PU, suggesting either a ceceuroterocele or eversion of a ureterocele into the BN and PU (Figure 2). That day, a DMSA renal scan confirmed a nonfunctional right kidney. Cystoscopic evaluation on DOL 3 was notable for bladder trabeculation, a patulous ectopic right ureteral orifice, and a large ureterocele in the prostatic urethra, which was incised and opened widely until adequate decompression was visualized. A Foley catheter was left in place postoperatively.

Postoperatively, the patient was polyuric. The Foley catheter was removed on postoperative day 3 (DOL 6). By DOL 7, urine output had normalized and serum creatinine had fallen from 1.4mg/dL to 0.5mg/dL. The patient was discharged home on antibiotic prophylaxis. At 5 weeks old, a RBUS showed resolution of the right hydroureter. At 5 months, a VCUG showed resolution of the filling defect that we had confirmed to be a cecoureterocele; a stably dilated and irregular BN and PU; and grade 1 VUR on the affected side (Figure 3); antibiotic prophylaxis was then discontinued. Now 8 months old, he has had no urinary issues or infections, and a RBUS shows no upper tract dilatation bilaterally. He will repeat a VCUG at 1 year.

Discussion

In life, the first presentation of a cecoureterocele may be lower urinary tract symptoms and/or infection,1 urinary incontinence,3 or urinary retention.3 In retrospect, the oligohydramnios this patient developed at early term may have been a manifestation of bladder outlet obstruction. Retention may be attributable to the ureterocele filling and causing obstruction during voiding; or it may be that, as a result of the muscular defect created by the cecoureterocele, a diverticulum develope-
Ectopic Ureterocele (continued from previous page)

ops in the floor of the bladder, causing secondary obstruction of the bladder outlet during voiding.\(^3\) Furthermore, cecoureteroceles and other ectopic ureteroceles may present with a prolapsing mass at the urethral meatus that obstructs urine.\(^3,5\) Incontinence, on the other hand, is most likely attributable to associated BN distortion.

Confirming the diagnosis of cecoureterocele in life begins with a RBUS, VCUG – to better visualize the anatomy and assess for VUR – and nuclear renal scan to assess the function of the affected kidney. Diagnosis with RBUS may be challenged by difficulty visualizing the extension of the ureterocele into the BN. On VCUG, this should be visualized as contrast reflexes and fills the tongue of the ureterocele, but may be obscured by contrast in the bladder and urethra.\(^4\) Therefore, cystoscopy may ultimately be necessary to make the diagnosis.

Transurethral incision involves incising an obstructing leaflet in a manner akin to ablation of posterior urethral valves. It is critical to ensure that residual tissue extending into the urethra does not cause obstruction, while also taking care to preserve continence when working in this area. If performing definitive reconstruction with excision and reimplantation, it is important to address any potentially obstructing tissue flap in the cecoureterocele’s distal aspect, whether by resection or fulguration. Finally, if a significant BN or urethral defect is appreciated, reconstruction should be considered.

References


Figure 2: Day of life 2 voiding cystourethrogram. Left: oblique; right: anterior posterior.

Figure 3: Voiding cystourethrogram (oblique) at 5 months of age.
Fetal Bladder Rupture Due to Maternal Sedative and Narcotic Administration for COVID-19 Pneumonia

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Case Report

A 22-year-old pregnant multiparous woman presented to the emergency department in acute respiratory distress at 25 weeks of gestation (WG). She was diagnosed with COVID-19 pneumonia and intubated shortly after arrival. She concurrently had septic shock from separate bacteremia and urinary tract infections requiring vasopressor support and broad-spectrum antibiotics.

Patient’s gestational history was unremarkable with normal first trimester anatomic scans and routine prenatal care. Given the mother’s critical condition, elective caesarean delivery was performed at 26 WG to initiate maternal extracorporeal membranous oxygenation. Throughout her hospitalization, the mother received narcotics, sedatives, and neuromuscular blockade agents including fentanyl, ketamine, midazolam, and rocuronium.

At birth, baby weighed 910 grams with Apgar scores of 3 and 7 at 1 and 5 minutes respectively. She was intubated and transferred to the neonatal intensive care unit (NICU), started on mechanical ventilation, and administered surfactant for neonatal respiratory distress syndrome (RDS). IV fluids and empiric antimicrobial therapy were initiated. Babygram was remarkable for RDS and abdominal ascites. Initial blood work showed elevated serum creatinine of 0.94 mg/dL. COVID-19 testing, blood, and urine cultures were negative.

At 8 hours of life, an indwelling urinary catheter was placed for worsening abdominal distention and failure to void. Large volume blood-tinged fluid was drained, attributed to bladder over-distention and microvascular rupture. Subsequent renal and bladder ultrasound (RBUS) showed mild bilateral pyelectasis, decompressed bladder around the catheter, and minimal peritoneal fluid.

On day of life (DOL) 4, the catheter was removed, but replaced after 5 hours of anuria and urology consulted. Imaging, history, and physical exam suggested no lower urinary tract obstruction or neurogenic bladder, and repeat trial of void was recommended, which she again failed. After catheter replacement, repeat RBUS demonstrated no hydroureteronephrosis, decompressed bladder and increased free fluid in the pelvis. Bedside X-ray cystogram showed contrast outlining loops of bowel consistent with intraperitoneal bladder perforation (Fig. 1). The catheter was recommended to remain a minimum of two weeks followed by repeat cystography.

Following an interval 48 hours, baby became critically ill from recurrent pulmonary hemorrhage requiring increased cardiorespiratory support, fluid resuscitation, blood product transfusions, and stress dose steroids. She experienced generalized anasarca, but maintained adequate urine output. Three days later, abrupt oliguria and increased abdominal girth prompted repeat RBUS. The urinary catheter tip was identified in the pouch of Douglas and repositioned under ultrasound guidance. Repeat cystogram after two weeks demonstrated extraperitoneal posterior bladder extravasation, likely into the vagina and/or posterior extravesical space, and adequate bladder filling and capacity (Fig. 2a). The catheter remained for an additional week. Cystography subsequently illustrated a tract between the bladder and vagina with immediate vaginal leakage of contrast, consistent with a vesicovaginal fistula (VVF) (Fig. 2b). No additional defects were noted, and the catheter was removed as the intra-abdominal component of the bladder rupture had resolved. Baby was weaned to room air, treated for pathology related to prematurity, and maintained adequate urine output throughout her stay. Serum electrolytes including creatinine normalized. She was discharged to her guardian at approximately 15 weeks of age. At 8 months, cystoscopy and cystogram demonstrated resolution of VVF.

Discussion

Isolated fetal and neonatal ascites is very uncommon. Barring iatrogenic injury, it is typically associated with structural abnormalities including gastrointestinal and urinary defects. Urinary ascites is usually attributed to urinary obstruction, particularly males with posterior urethral valves. We report a rare instance of postnatal presentation of
fetal bladder rupture in a female without evidence of anatomic obstruction. The suspected etiology is transplacental transmission of adult-dose narcotics and sedatives inducing retention, with subsequent intraperitoneal rupture. Parenteral opioids may inhibit spinal opioid receptors causing bladder relaxation and urinary retention, particularly in premature infants.\textsuperscript{4,5} Prematurity may also have contributed to rupture and the unexpected VVF as immaturity of the musculature increases compliance of urothelium, leading to excessive dilatation of the urinary tract and tissue necrosis from the catheter.\textsuperscript{4}

Treatment of pregnant patients requires consideration of both mother and fetus. Sedatives such as narcotic medications should be minimized to reduce morbidity to both parties, particularly within the first and second trimester. Intrauterine fetal bladder rupture in the setting of maternal medication administration is rare, but conservative measures can be employed successfully in the presence of clinical stability.

References


Fig. 2. Repeat X-ray cystogram. a) 4 weeks of life, oblique view showing concern for extravasation of contrast from the posterior bladder. b) 5 weeks of life, oblique view showing vesicovaginal fistula.
Introduction

Fetal urinary tract surgery is an important branch of maternal fetal medicine that aims to give neonates an improved chance of survival and potentially alter the trajectory of obstructive processes post-natally. The role of fetal surgeons ranges from vesicoamniotic shunt placement for lower urinary tract obstruction to fetal cystoscopy for posterior urethral valve ablation. Despite numerous technological advances in the field, complications can occur. Here, we describe a case of inadvertent collateral injuries from trocar placement for vesicoamniotic shunts.

Case Presentation

A 25-year-old G1P0 with a past medical history of type 1 diabetes mellitus, history of traumatic brain injury, and obesity as well as a family history of cystic fibrosis presented with a male fetus who was noted to have with a 15-week ultrasound demonstrating a distended bladder with oligohydramnios. He underwent fetal bladder aspiration and vesicoamniotic shunt placement at 16 weeks. The first attempted shunt was noted to reside in the peritoneal cavity, so a second shunt was successfully placed in the bladder. Continued bladder distension was noted at 17 weeks, and a third vesicoamniotic shunt placement was attempted and successful. Around this time, fetal cysts were discovered on ultrasound. Preterm premature rupture of membranes occurred at 23 4/7 weeks gestation and mother was placed on bedrest and steroid treatments.

Cesarean delivery was performed at 35 weeks due to fetal decelerations. Examination at time of delivery showed eviscerated bowel through a right lower quadrant shunt site, bilateral undescended testicles, hypoxic respiratory failure, congenital malformation of the ear, and club foot. He was noted to be in renal failure with a creatinine of 1.60. X-ray revealed two shunts, with one exiting the abdomen (Figure 1). He was taken to the operating room for exploratory laparotomy. The vesicoamniotic shunt was noted to be exiting the left abdominal wall with a large amount of matted, inflamed intestine and the left testicle, which was slightly dusky (Figure 2). The right testicle was nonpalpable. An incomplete prepuce with orthotopic urethral meatus

Figure 1

Figure 2
The Road to Transplant (continued from previous page)

of small caliber was noted. Feeding tubes as small as 3.5 French were attempted to be placed, however resistance was encountered in the proximal urethra. These findings were consistent with near complete urethral atresia. Three vesicoamniotic shunts were extracted with one noted to be in the bladder. A cutaneous vesicostomy was created and the bowel was able to be reduced into the abdomen with no resection required.

On day of life 1, a renal ultrasound revealed bilateral renal dysplasia with marked atrophy of the left kidney. The baby had normal urine output; however creatinine rose to 2.6 He required a 5-week NICU stay and was discharged with a NG tube in place for feeding. At age 4 months, he underwent G-tube placement and rigid cystoscopy via vesicostomy, which confirmed findings of complete proximal urethral atresia. A full genetic work-up was conducted with no underlying etiologies discovered. The patient ultimately progressed to stage V CKD and underwent a living unrelated donor kidney transplant at 17 months without requiring dialysis in the interim. Intraoperatively, the right testis was discovered intra-abdominally and first-stage Fowler-Stephens orchiopexy was performed. The patient is currently doing well and thriving.

Discussion

Currently, there are no established guidelines for fetal urology and vesicoamniotic shunt placement. There is no limit on the number of vesicoamniotic shunt attempts that can and should be performed. This patient had three shunts placed with only one shunt in the correct position within the bladder. Shunt migration/malposition continues to be a difficult complication of fetal surgery, and as documented, placement of trocars can have associated collateral injury.

Conclusion

While fetal surgery can be beneficial and lifesaving, it can have associated complications, thus in-depth discussion of potential risks with family must occur. In these situations, early recognition and intervention by a multidisciplinary team is essential in management of these complex cases.

Prune Belly Syndrome with Urethral Atresia and Anorectal Malformation

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Introduction

Prune Belly Syndrome (PBS) is a rare congenital condition defined as the triad of the absence of abdominal musculature, bilateral intra-abdominal testicles, and urinary tract anomalies. PBS is often diagnosed prenatally, is estimated to occur in 1/29,000-45,000 live births and may have a sex linked autosomal-recessive inheritance, although most commonly sporadic. In addition to the triad of clinical characteristics in PBS, other organ systems may have associated abnormalities, including the gastrointestinal system in about 24% of cases. Despite the high frequency of gastrointestinal anomalies in PBS, anorectal malformations (ARM) are rare. In PBS, regardless of concurrent comorbidities, the urinary tract anomalies and the extent of renal dysplasia as the most important detriment of long-term survival and is highly variable. Bladder outlet obstruction, typically due to urethral atresia or megalourethra can be lethal prenatally without a patent urachus. We present a case of male born with PBS, with urethral atresia and an ARM.

Case Presentation

The patient (Twin B) is a male born via C-section at 33 weeks to a 27-year-old woman G1P0 with preeclampsia. Twin A was a normal female. He was discovered to have bilateral hydroureteronephrosis and a distended bladder on sonography at 12-weeks gestation. Vescicocentesis suggested a favorable prognosis and the parents elected to proceed with vesicoamniotic shunt (VAS) placement. At 18-weeks gestation, vesicoinfusion and VAS placement was attempted but the procedure was complicated by urinary ascites, so the shunt was left between the peritoneum and the amniotic cavity. At 19-weeks gestation, VAS was successfully placed with bladder decompression and reconstitution of amniotic fluid.

At birth, he was noted to have a Prune Belly configuration with bilateral undescended testes, a normal phallus and an imperforate anus. He was anuric after birth without urine per urethra or VAS. A urethral catheter was unable to be placed for a VCUG and retrograde urethrogram performed confirmed urethral atresia (Figure 1). Shunt removal, vesicosotomy, sigmoid colostomy and a Ladd’s procedure without appendectomy was performed on the first day of life. Further workup revealed moderate right hydronephrosis, left vesicoureteral reflux, a recto-bladder neck fistula, sacral hypoplasia without cord tethering, patent foramen ovale and resolved patent ductus arteriosus.

Within the first month of life, while in the NICU, he suffered E. Coli bacteremia presumably from a urinary tract infection (UTI). At 5 months of age, cystoscopy revealed a pinhole sized opening in the bulbourethra and a marked decrease in renal function. After successful cannulation, progressive augmentation by dilating the urethra anterior (PADUA) was attempted. PADUA was performed with 5 procedures over a two-week period with dilation from 3Fr up to 10.5F but he was unable to void after catheter removal (Figure 2), and a vesicostomy was recreated.

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Prune Belly Syndrome (continued from previous page)

A laparoscopic posterior sagittal anorectoplasty with rectovesical fistula excision and colostomy closure was performed at ages 7 and 9 months, respectively. At 15 months, a Montfort abdominoplasty with left vesicoureteral reimplant, right ureteroureterostomy for a mid-ureteral stricture, bilateral abdominal orchiopexy and an appendiceal Mitrofanoff was performed. He is now 6 months post-op. Although he recently underwent a stoma revision for skin stenosis at the Mitrofanoff site, he has continued to catheterize without any issues and has not developed any urinary tract infections. He continues to be unable to void via the urethra.

Summary

This Twin B male was born with PBS, urethral atresia and imperforate anus with a rectovesical fistula. He failed the PADUA procedure and ultimately required an appendiceal Mitrofanoff for bladder management. To our knowledge, this is one of few reports of imperforate anus associated with PBS. Morgan et al. reported two similar infants who ultimately died shortly after delivery; however, unlike the patient described above, no fistula was identified between the genitourinary and gastrointestinal tracts on autopsy. Mahajan and colleagues also report a case with the same clinical picture but the infant died after developing fatal septicemia from necrotizing gastritis with gastric perforation after colostomy.

PBS in combination with imperforate anus is rare, and likely carries a poor prognosis. Although PBS with urethral atresia can present challenging management dilemmas for management of the genitourinary tract, it is currently unknown how concurrent ARM may further worsen renal outcomes.

References


Figure 1.
VCUG performed at birth demonstrating urethral atresia.

Figure 2.
Retrograde urethrogram after PADUA procedure demonstrating a patent but stenotic urethra. Ultimately the patient was unable to void and required a Mitrofanoff appendicovesicostomy.