Society for Fetal Urology: 
2016 Meeting in Review

FROM THE GUEST EDITOR
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We had another great meeting in San Diego. The topic of this year’s meeting was posterior urethral valves. The meeting was really well attended and there was an abundance of cases submitted for presentation. Unfortunately due to time constraints we had to limit the cases accepted to 18.

As usual the breadth of cases presented covered a wide array of topics including sagittal diphallia, OHVIRA syndrome, bladder exstrophy, and congenital mesoblastic nephroma. Coupled with the case presentations we also had a robust group of abstracts that varied as well from posterior urethral valves, prenatal hydronephrosis, ureteropelvic junction obstruction, and vesicoureteral reflux. In keeping with the theme of posterior urethral valves and the popular question of how best to grade hydronephrosis we had a fantastic and accomplished group of speakers present. The speakers included Alan Bailie from Belfast who presented an excellent talk on the BAPU endorsement of the consensus statement on hydronephrosis, Curtis Sheldon from Cincinnati who presented on the management of posterior urethral valves, and finally, Andrew Hull from California who presented on prenatal imaging and management from the MFM perspective.

Overall the meeting was very successful and built up excitement for the Fall meeting.

Topics Included:

- LUMBAR Syndrome: A Rare and Complex Variant of Bladder Exstrophy
- Sagittal True Diphallia
- Urogenital Sinus with Oligohydramnios: A Stepwise Approach to Evaluation and Management
- Bilateral Prenatal Hydronephrosis: An Interesting Case of Posterior Urethral Valves in the Setting of Currarino Syndrome
LUMBAR Syndrome: A Rare and Complex Variant of Bladder Exstrophy

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Case Presentation

A newborn girl with prenatally diagnosed solitary kidney was found to have bladder exstrophy on initial exam. She was further noted to have subtle infantile hemangioma (IH) on the left leg, omphalocele, anteriorly displaced anus, and agenesis of the left external genitalia (Figure 1). Ultrasound on day of life (DOL) 1 demonstrated a solitary right kidney and presence of a uterus. The pediatric urology, general surgery, orthopedic surgery, and dermatology teams were consulted. Given the constellation of findings, there was concern for LUMBAR syndrome which is characterized by Lower body hemangioma, Uro-genital anomalies, Ulceration, Myelopathy, Bony deformities, Anorectal malformations, Arterial anomalies, and Renal anomalies. An MRI of the spine, abdomen, pelvis, and left limb was obtained to evaluate for myelopathies and vascular anomalies that would complicate surgical correction of the exstrophy and pelvic diastasis. In addition to the previously discussed clinical findings, the MRI demonstrated uterine didelphys, a lipomyelocele and tethered cord. No arteriovenous malformations were seen in the pelvis or thighs. The baby underwent omphalocele closure on DOL 3. Due to location and extent of the infantile hemangioma and spinal cord anomalies, bladder exstrophy closure was delayed. The bladder plate was managed with Vaseline and non-stick dressing.

During the first month of life, the infantile hemangioma (IH) blossomed into bright red superficial hemangiomas with ulcerations that involved the left leg, external genitalia, and lower back (Figure 1). Dermatology treated the hemangiomas with oral propranolol and topical timolol. The ulcerations improved and the hemangiomas stabilized. The baby remained free of neurologic symptoms. Her solitary kidney was followed at three month intervals with ultrasound and was noted to have improving hydronephrosis. At age 7 months, she underwent L5-S1 laminectomy and spinal cord untethering. To allow for recovery from untethering and resolution of spinal shock, operative repair of the bladder exstrophy as well as osteotomies was delayed until approximately age 11 months of age. Follow-up at 6 months following repair is shown in figure 1.

Discussion

LUMBAR syndrome is a rare entity with less than 60 cases reported in the literature.¹ The pathogenesis is currently unknown. It represents a spectrum of disease, but universally presents with infant-age issues.
tile hemangiomas. Myelopathy, most commonly tethered cord, is found in 80% of infants with LUMBAR syndrome and necessitates imaging which is best accomplished with MRI. Urogenital abnormalities including bladder extrophy and ambiguous genitalia are found in about one third of infants and almost 40% will have renal anomalies including solitary kidney, hypoplasia, or hydronephrosis. As demonstrated in figure 1, the infantile hemangiomas proliferate rapidly during the first year of life and can make reconstructive genitourinary surgery challenging. Delaying surgical therapy until hemangiomas can be reduced has been reported in the literature and we employed this approach in our case. Infants born with LUMBAR syndrome are medically complex and require imaging and optimization of hemangiomas prior to proceeding with surgical intervention. This case highlights the need for multidisciplinary care of infants born with LUMBAR syndrome.

References

Sagittal True Diphallia
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Case Presentation
The patient was born at 34 weeks gestation with a prenatal diagnosis of a left multicystic dysplastic kidney (MCDK). On examination, the patient was noted to have an incomplete (dorsal hood) prepuce, sagittal diphallia, with a dominant dorsal glans and urethral polyp protruding through the urethral meatus and a non-dominant ventral glans with a small caliber subcoronal hypospadiac urethral meatus, and descended testes (Figure 1). A karyotype was obtained, demonstrating 46,XY.

Renal ultrasonography confirmed a left MCDK. Voiding cystourethrography (VCUG) demonstrated sagittal duplication of the anterior urethra. Pelvic magnetic resonance imaging (MRI) demonstrated complete duplication of the corpora cavernosa and spongiosa, as well as sagittal duplication of the anterior urethra (Figure 2). A lumbosacral MRI was obtained, demonstrating a sixth lumbar vertebra.

After a multidisciplinary review of the case and discussion with the patient’s family, the decision was made to proceed with cystourethroscopy to excise the urethral polyp and further delineate the urethral anatomy. At five months of age, cystourethroscopy was performed, and the patient’s urethral polyp was excised. The bifurcation of the urethra was confirmed to be at the mid-bulbar urethra, and the ventral urethra was noted to be hypoplastic. At ten months of age, surgical reconstruction of the phallus was performed. The ventral phallus and corpus spongiosum/urethra were excised. The urethrotomy was closed in two layers. The ventral corpora were approximated in the midline over the repair, and a glanuloplasty and circumcision were performed (Figure 3).

At the most recent follow-up (eleven months postop), the patient was voiding normally and having straight erections, and the family was pleased with the cosmetic result.

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Diphallia, or penile duplication, is an extremely rare congenital anomaly, with an incidence of 1 per 5.5 million live births. The embryology is poorly understood. Penile duplication is classified as true diphallia and bifid phallus, with each classification subdivided into complete or partial based on size or point of duplication, respectively. In true diphallia, there is complete duplication of the corpora cavernosa and spongiosa, while in (complete) bifid phallus each phallus has a single corpus cavernosum.¹

During the initial evaluation of a patient with diphallia, a detailed physical examination aids in classification and assists in surgical planning. Further delineation of the corporal and urethral anatomy with diagnostic imaging is essential. As in our case, many reports recommend utilizing a VCUG to assess the urethral anatomy and MRI to evaluate soft tissue. Given its association with other congenital anomalies, including urinary tract duplication, anorectal malformation, and vertebral anomalies, a detailed systemic evaluation is warranted as well.²

Surgical management poses functional, aesthetic, and ethical issues. Surgical correction can be challenging, and must be individualized based on degree of duplication, imaging studies, and severity of concomitant anomalies. The goals of surgery are to preserve function, minimize complications, and provide a good cosmetic result. In general, most recommend excision of the less developed phallus with or without urethral reconstruction.³

To our knowledge, this is the first reported case of partial true diphallia associated with a urethral polyp and a MCDK.

References
Urogenital Sinus with Oligohydramnios: A Stepwise Approach to Evaluation and Management

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Case Presentation

Patient S.P. presented to the pediatric urology service on day of life one. She was identified prenatally with oligohydramnios and delivered at an outside hospital by C-section at 30 4/7 weeks. She had an enlarging fluid-filled abdominal mass and ascites which was compressing the thoracic cavity (Figure 1). APGARS were 1 and 6 at 5 and 10 minutes, respectively. She was intubated, a Foley catheter was placed with urine return, and she was emergently transferred to our hospital for specialist care. She did not receive prenatal urologic counseling.

On arrival she had a massively distended, tense abdomen and a single opening at the introitus. Adjustment of the Foley catheter resulted in 20 mL UOP and a slight decrease in abdominal distention. Renal ultrasound (RUS) demonstrated P2 urinary tract dilation (UTD) bilaterally (Figure 2). She was started on amoxicillin prophylaxis. A voiding cystourethrogram (VCUG) revealed a urogenital (UG) sinus (Figure 3A). Voiding resulted in filling of the vagina and extravasation into the peritoneum; presumed from the fallopian tubes (Figure 3B). There was no vesicoureteral reflux.

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For several weeks, she was maintained on Foley catheter drainage; however, given increased risk of UTI, she was started on clean intermittent catheterization (CIC). Despite CIC, she preferentially drained into and distended her vagina. Multiple attempts to catheterize the vagina to establish drainage were unsuccessful due to the acute angle of insertion of the vagina at the confluence.

At 6 weeks of age she underwent cystourethroscopy and vesicostomy. Cystourethroscopy revealed a high confluence urogenital sinus with an incompetent bladder neck and an atretic communication with the vagina. This communication was sequentially dilated to #8 French. Although the vesicostomy drained well, a follow-up ultrasound revealed persistent massive dilation of the vagina. Therefore, she returned to the OR to undergo vaginostomy 2 months after vesicostomy. During the case, several hundred milliliters of turbid, foul-smelling fluid were aspirated from the vagina. Follow-up ultrasound confirmed marked improvement in vaginal distension. At six months, she underwent vaginostomy and vesicostomy takedown in addition to reconstruction with vaginal pull-through via anterior sagittal transanal rectal (ASTRA) approach. She was discharged home on post-operative day 3 with Foley drainage maintained for 1 week post-operatively. At 9 month follow-up she is doing well. Ultrasound demonstrates no evidence of vaginal distension, the bladder is decompressed without evidence of UTD.

Discussion
This case illustrates management of a urogenital sinus recognized prenatally as a fluid filled cavity which highlights the need for a multidisciplinary approach. After birth we recommend prophylactic antibiotics, catheter drainage, and imaging including RUS, and VCUG. Bladder drainage and, if necessary, vaginal drainage should be established until the patient is old enough to undergo reconstruction. Cystoscopy should be performed prior to surgical repair to measure the length of the common channel as well as the distance between the bladder neck and vagina to determine the operative approach [a]. High UG sinus repair can be successfully performed using the ASTRA approach [b]. Timing of UG sinus repair is controversial. We prefer to perform reconstruction at 6 months of age.

References
Bilateral Prenatal Hydronephrosis: An Interesting Case of Posterior Urethral Valves in the Setting of Currarino Syndrome

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Case Presentation

The patient presented as a newborn male born at 37 weeks gestation. Prenatal ultrasound at 17 weeks revealed a focus of echogenic bowel within the pelvis. This was confirmed on 30 week ultrasound, at which time bilateral renal pelvic dilation was identified. Growth parameters and fluid indices were normal. Ultrasound at 32 weeks revealed an interval increase in the degree of bilateral renal pelvic dilation, with anterior-posterior diameters measuring 1.2-cm bilaterally.

The patient was followed with serial ultrasounds. Final prenatal ultrasound at 37 weeks revealed renal pelvis diameters of 1.6-cm on the right and 1.4-cm on the left, along with new oligohydramnios. Labor was induced, and the patient was born via cesarean section. Physical examination at birth was notable for a palpable bladder, imperforate anus, and 2-cm soft tissue mass overlying the sacrum.

A 5Fr pediatric feeding tube was placed per urethra. Renal ultrasound (RUS) obtained on day of life one showed bilateral SFU Grade 4 hydronephrosis with ureterectasis and a thick-walled bladder. Voiding cystourethrogram (VCUG) revealed a dilated posterior urethra and marked bladder trabeculation suggestive of posterior urethral valves (Figure 1). Serum creatinine was 0.9 ng/dL. Due to the sacral mass and imperforate anus, an MRI of the pelvis was obtained. This showed a 3.5-cm heterogenous cystic presacral mass with extension into the distal spinal cord, consistent with sacrococcygeal teratoma, as well as dilated loops of bowel and a blind-ending rectum (Figure 2).

The patient underwent teratoma resection, anorectoplasty, and diverting loop colostomy on day of life two. Given the small caliber of the urethra and concern for spinal shock following resection of the teratoma, the decision was made to perform a vesicostomy. The serum creatinine reached a postoperative nadir of 0.3 ng/dL. Genetic testing revealed partial deletion of exon 1 in the MNX1 gene, consistent with a diagnosis of Currarino syndrome. At 12-months of age, the patient underwent colostomy takedown and at 15-months of age he was taken to the operating room for posterior urethral valve ablation and vesicostomy closure. At 38-month follow-up, RUS showed stable pelvocalyctasis. Formal urodynamics revealed an enlarged bladder capacity, detrusor overactivity, mild terminal non-compliance, and complete bladder emptying. He is currently managed with oral anticholinergics, and his serum creatinine remains stable at 0.3 ng/dL.

Discussion

Currarino syndrome is a triad consisting of an abnormality of the bony sacrum, an anorectal malformation, and a presacral mass.[1] Several associated urogenital abnormalities including horseshoe kidney, vesicoureteral reflex, and neurogenic bladder have been reported to coexist in these patients.[2, 3] To our knowledge this represents the first case report describing bladder outlet obstruction from posterior urethral valves in a child with Currarino syndrome. Reported rates of urinary sequelae secondary to congenital abnormalities or side effects of treatment vary, but may be as high as 30%.[3] Therefore, surveillance may be warranted as many of these patients are at increased risk for urologic complications.

References