Society for Fetal Urology: 2020 Meeting in Review

FROM THE GUEST EDITOR

Douglas W. Storm, MD

To say that 2020 has been a challenge, may be the understatement of the century. In the field of medicine, we as practitioners have had to rise to the occasion, to care for our patients, manage our practices and continue our research endeavors while also helping our children learn virtually and protecting our families and loved ones, in the face of a once in a lifetime pandemic. When looking at it from this perspective, it is nothing short of remarkable that The Societies for Pediatric Urology were able to pull off the SPU Live event on Saturday, June 27, 2020. Part of the SPU Live event included the Society for Fetal Urology portion of the meeting and I am excited that the Dialogues in Pediatric Urology staff asked me to guest edit this edition, to highlight the SFU portions of the June meeting and the happenings from the organization within the last crazy year.

As part of the SPU Live event, the Society for Fetal Urology was able to kick off the weekend with its virtual “Cases and Cocktails” webinar. I would like to thank all of the presenters in this virtual webinar for their flexibility in participating in this virtual event and for providing excellent, thought provoking, interesting cases. In this edition of Dialogues in Pediatric Urology, the webinar participants present their excellent case reports. In case you were not part of the audience during this webinar, I would invite you to visit the SPU website for a full recording. This can be found at: http://www.sfu-urology.org/sfu-cases-cocktails-virtual-webinar-june-26-2020-600-cst/

During the actual SPU Live event, Tony Herndon and Luis Braga graciously included the panel discussions “What is the Best Postnatal Surgical Intervention for Posterior Urethral Valves” and “Management of the Ectopic Upper Pole Obstructed Duplicated Ureter” as well as the presentation by Dr. Linda Baker on “The Modern Management of Prune Belly Syndrome. These talks were originally planned as part of SFU portion of the Society of Pediatric Urology Meeting, when it was scheduled as an in-person event in Washington D.C. I would like to thank all of the presenters for their flexibility and willingness to participate and provide such insightful discussion, as this transformed into a virtual event. In this edition of Dialogues in Pediatric Urology, the moderators of the SFU panel discussions, Tony Herndon, M.D. and Paul Austin, M.D., have generously provided a synopsis of their individual panels. Again, I would like to thank Tony and Paul for their willingness to moderate these virtual sessions and for participating in this special SFU Edition of DPU. If you missed out on these presentations, I invite you to view a recording on the SPU website: https://spuonline.org/meeting/abstracts/2020-Live.cgi

I am also excited to include in this SFU edition of Dialogues in Pediatric Urology a discussion with Tony Herndon, M.D., regarding the Hydronephrosis registry. The Hydronephrosis registry has become synonymous with the Society for Fetal Urology and I am hopeful that you will enjoy (as much I did) the discussion that I was privileged to have with Tony. I would like to thank Tony for taking the time to have this conversation, providing insight into the history and future of this important database.

I would also like to thank the leadership of the Society for Fetal Urology for providing me the opportunity to organize the SFU portion of the Societies for Pediatric Urology annual meeting. The support that the leadership of the SFU provided me in the planning and implementation of the meeting was invaluable. Although none of us could have foreseen the challenges that 2020 would present, I think in the end, we achieved an informative, stimulating virtual event. This could not have been accomplished without the hard work of the panelists, moderators and presenters that participated in the SFU portion of SPU Live and the Cases and Cocktails webinar and I am eternally grateful to them.

I look forward to the time where we can again be in a conference room with one another, without 6 feet between each of us, listening to these illuminating presentations in person. Until that time arrives, I wish you all nothing but the best. Stay healthy and safe.
Welcome to the Society for Fetal Urology Year in Review. 2020 continues to throw the unexpected our way but we have been nimble and rolling with the punches. We can proudly say that we were able to move forward with the SFU Spring Meeting in June thanks to the hard work and dedication by SFU meeting chair, Dr. Douglas Storm. We kicked off the meeting with our traditional case presentations in the “Cases and Cocktails” Virtual Webinar Event on the evening of Friday, June 26th with an impressive live attendance of 161 viewers. Ten excellent presentations were given and three were selected as the top cases including:

1. “Bladder Duplication in a Setting of VACTERL Association” by Cinthia Galvez, Andrew Ransford, Rafael Gosalbez, and Alireza Alam
2. “Should 46 XY DSD Patients Receive a Follow Up Serum Electrolyte Screen in the 1st Months of Life?” by Angelena Edwards, Niccolo Passoni, and Linda Baker
3. “Prenatally Diagnosed Solid Renal Mass” by Sarah A. Holzman, Josephine HaDuong, and Antoine E. Khoury. Look out for the publication of these top three aforementioned presentations as case reports in the Urology Gold Journal!

The following day the SFU educational program focused on the postnatal management of prenatally-detected, congenital, urologic conditions. There were two live panel discussions including “What is the Best Postnatal Surgical Intervention for Posterior Urethral Valves?” moderated by Dr. Paul Austin with panelists Drs. Jonathan Ellison and William R. DeFoor and the second on “The Management of the Ectopic Upper Pole, Obstructed, Duplicated Ureter” moderated by Dr. Tony Herndon with panelists Drs. Daryl Mcleod, Kate H. Kraft, Jason P. Van Batavia, Rosalia Misseri, and Vijaya Vemulakonda. The invited SFU lectureship was given by Dr. Linda Baker on “The Modern Management of Prune Belly Syndrome”.

On deck is our next meeting in 2021 in Las Vegas chaired by Dr. Marcos Machado. We know that his energy, positivity, and creativity are sure to bring a stimulating and thoughtful meeting our way. While it is hard to imagine where we will all be a half a year from now, we hope that the coming months bring health, safety, strength, and resilience to us all.

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What is the Best Postnatal Surgical Intervention for Posterior Urethral Valves?

The Societies for Pediatric Urology featured an exciting session at the SPU Live meeting on Saturday, June 27, 2020 that discussed the topic of “What is the best postnatal surgical intervention for posterior urethral valves (PUV).” The live panel discussion entailed an introduction by Dr. Paul F. Austin from Texas Children’s Hospital, Houston, TX followed by engaging presentations by Dr. Jonathan Ellison from Children’s Hospital of Wisconsin, Milwaukee, WI and Dr. William R. DeFoor, Jr. from Cincinnati Children’s Hospital, Cincinnati OH. Dr. Ellison discussed ablation of PUV whereas Dr. DeFoor discussed the role of vescicostomy with PUV.

To set the table for the PUV panelists, Dr. Austin provided an overview of the anatomical features of PUVs with 95% of PUV cases being type 1 that involve tissue folds that emanate from the verumontanum toward the posterior urethra distally and the remaining 5% are type 3 that are diaphragmatic in configuration. Additionally, Dr. Austin discussed the implications with PUV and lower urinary tract obstruction (LUTO) on the response of the bladder smooth muscle. Specifically, that PUV and LUTO results in tension-induced bladder smooth muscle hyperplasia and hypertrophy that leads to an alteration in bladder wall compliance that ultimately poses a risk for hydronephrosis, vesicoureteral reflux, renal damage and renal failure.

Early ablation of PUV

Dr. Ellison reviewed the historical aspects of endoscopic valve ablation. The first reported description of PUV ablation was by Hugh Hampton Young in 1929. The 1970’s and 80’s was the time of improved instrumentation and the 1990’s provided intermediate outcomes for follow up. More recently, endoscopic PUV ablation has focused on using alternative energy sources, fetal valve ablation and definition of “success”.

The goals of endoscopic valve ablation are to eliminate obstructive tissue and maintain bladder integrity. Essentially, endoscopic valve ablation allows early bladder cycling and anatomical normalization. In animal studies of fetal bladder outlet obstruction, there appears to be less fibrotic changes without the typical collagen deposition and this unique observation in fetal tissues may provide an opportunity to intervene before the development of the typical postnatal irreversible fibrotic changes that occurs with LUTO.

A review of different techniques of valve ablation showed a higher association of urethral stricture with using a diathermy hook to cauterize/cut the valves as opposed to a cold knife and Dr. Ellison offered the advice that “cold is better and hot is not”. Interestingly, residual valves may occur in 10% of PUV ablations and there should be a low threshold to re-evaluate the urethra with cystoscopy.

Patients that undergo endoscopic valve ablation typically have lower severity of hydronephrosis and lower grade of vesicoureteral reflux in comparison to those individuals managed with urinary diversion. Additionally, the patients with valve ablation alone tend to have delayed progression of chronic kidney disease (CKD) compared to patients that have urinary diversions with valve ablation. The difference of CKD outcomes with valve ablation from urinary diversions may reflect a more favorable, selected patient population.

Role of urinary diversion in neonatal bladder outlet obstruction

Dr. DeFoor presented two interesting prenatal cases to illustrate the primary postnatal PUV management goals: 1) Immediate bladder drainage, 2) Supportive NICU care, 3) Monitor upper tract/renal function with nephrology consultation and 4) Minimize UTIs.

Although bladder function is important, it was stressed that renal function is more important. Benefits of bladder cycling, clean intermittent catheterization (CIC) and anti-cholinergic medications were discussed but in the worst-case scenario, these interventions may not adequately manage the bladder and protect the kidneys and thus urinary diversion is required. Urinary diversion can be achieved with a cutaneous vescicostomy or a cutaneous ureterostomy. Cutaneous vescicostomy is generally the initial diversion of choice and indications for cutaneous vescicostomy include the following: 1) Infant is too small to safely allow passage of a cystoscope for valve ablation, 2) Inability to perform CIC when indicated by either urethral anomalies or family/social concerns. The rare indication for cutaneous ureterostomy includes worsening hydrourerteronephrosis and/or renal function despite maximal bladder drainage. This observation may be seen with secondary ureterovesical junction obstruction in the setting of a thickened bladder wall and a dilated, tortuous, high-capacitance ureter. A cutaneous ureterostomy will subsequently achieve low-pressure drainage of the kidney(s) and optimize renal function.

Dr. DeFoor acknowledged some concerns with supravesical diversions including no long-term data regarding preservation of renal function in most contemporary studies. Ureterostomy may delay the time needed for dialysis but there is a need for additional major surgery for reimplantation and undiversion. There is also the concern for defunctionalization of the bladder with loss of bladder cycling. Outcome studies are hampered by intrinsic selection bias as supravesical diversion is reserved for the most severe cases. An additional challenge is comparison of bladder function with urodynamic data in cohorts of patients with evolving bladder dynamics as they age.

Closing

In summary, the panel on postnatal surgical management of PUV provided compelling rationale and indications for the treatment selection in PUV patients. Both panelists identified the current need for a variety of interventions in managing these complex, heterogeneous group of patients and the need for continued bladder and renal observation.

To listen to a recording from the panel, we invite you to visit: https://spuonline.org/meeting/abstracts/2020-Live.cgi
Management of Ectopic Upper Pole Ureter

It was with great pleasure that I was asked to moderate the SFU panel “Management of the Ectopic Upper Pole Ureter” for the SPU Live event. Going into the programming, we felt that having live presentations with discussion certainly would carry some risks, but would equally provide a familiar platform to an in-person meeting. The session ran free of any technical issues and I thoroughly enjoyed working with this group.

The index case was a 3-year-old girl, whom I had observed since birth, with a dilated upper pole ectopic ureter. Over the course of time, she had resolved her lower pole vesicoureteral reflux (VUR) and was managed off of prophylactic antibiotics. After she was toilet trained, her mother noted that she experienced anticipated continual incontinence. Incidentally, she had a twin sister with similar complaints and anatomy.

Leading off the discussion was Darryl McLeod, MD who advocated for upper to lower ureteroureterostomy and felt this was the best approach, given the current level of evidence in the literature. Concerns for de-novo Yo-Yo reflux and issues with the ectopic stump were discounted, although admittingly they do occur. He felt the balance of literature supported that the function of the upper pole had little consequence and the procedure is well suited for all surgical approaches - from open surgery to robot-assisted laparoscopy. Personally, he performs a HIDES approach when managing these patients.

Kate Kraft, MD took the position of upper pole nephrectomy and reviewed the literature to support her position. She felt this approach was more definitive and could avoid lower tract surgery, given the ideal patient. With the addition of robot-assisted techniques, the upper pole nephrectomy has become less morbid as compared to the historical open approach. Although concerns exist for lower pole injury, the literature does not bare this to be a major concern. In addition, there exists the risk of bleeding, which also appears to be quite low. She concluded that given the ideal patient, the upper pole vasculature should not be well developed, to the extent that contemporary energy devices should alleviate any major concerns with bleeding. Finally, she reported multiple series that affirmed preservation of lower pole function.

Jason Van Batavia, MD was tasked with the most difficult approach of ureteral clipping. His presentation was very non-biased, aside from the discloser that he went straight to the creator of “Clip It and Forget It” - Armando Lorenzo, MD (also known as “Messi”) - to gather information to support his position. He gave a nice historical perspective, that gave insight to the evolution of this approach from inadvertent injuries in the 1900’s to more intentional use by the transplant surgeons in the later part of the 20th century. Given the lack of symptomatic post-operative complaints in the large transplant population, he felt that it was reasonable to apply this approach to our pediatric patients with an ectopic upper pole ureter, given the ideal patient. (Full disclosure, he did ask forgiveness from Dr. Canning at this point!!). In conclusion, Jason felt that ureteral clipping provided a shorter operative time, with day surgery as an option, minimal risk to lower pole injury, historically has a low complication rate and most importantly does not burn any bridges. He did concede that follow up of less than 2 years was a little too short to declare this as the best approach, given that many of these patients are females and at higher life time risk of UTI.

Rosalia Misseri, MD declared herself the senior stateswoman of the group and presented a goal-oriented approach that prioritized minimizing the development of UTI and optimizing continence. She felt strongly that in the presence of lower pole VUR that a lower tract approach would achieve both goals by addressing the ectopic ureter as well as correcting VUR. This approach really has minimal risk, avoids a visible scar, avoids any potential renal vascular injury and can be definitive in most patients. In addition, it is the approach that offers the advantage of almost completely excising the distal portion of the ectopic ureter which minimizes the existence of a residual ectopic ureteral stump.

Finally, Vijaya Vemulakonda, MD took the non-operative approach and carefully crafted an argument in support of this position. The inherent risk of any surgical procedure must be examined through the lens of its potential complication, which she outlined nicely - including ureteral stricture after upper pole tapering, urine leak following upper ureteral surgery, creation of de-novo VUR and the risk of prolonged anesthesia. She closed her argument as an extension of the evolution from open to minimally invasive surgery, to the option of active surveillance shared by our adult colleagues.

Ultimately, the index patient was managed with a combination of approaches, including active surveillance for the first three years of her life. Despite fairly significant dilation of the upper pole ureter, we elected to defer surgery until clear signs of an ectopic ureter were present.

C.D. Anthony Herndon, MD
The Hydronephrosis Registry Turns 21: A Sit-Down Conversation with Tony Herndon

I was fortunate enough to catch-up with Tony Herndon to discuss the Hydronephrosis Registry, which turned 21 this year. As the creator of the registry, Tony has the unique perspective of understanding how to construct this irreplaceable registry, keep it afloat all these years, seeing it change and having a vision for the role it will play in the future. I would like to thank Tony for taking the time out of his busy schedule to answer my questions. Below, is a transcript of our conversation, which I think would make a great podcast! I hope that you will find this as informative and perceptive as I did. Enjoy.

Doug: What prompted you to develop the hydronephrosis registry?

Tony: One of my mentors recommended that I do research in an area that had a big impact. Prenatal hydronephrosis is common and if you could identify areas that would benefit form change, then it would have a huge impact. Given that we follow these patients longitudinally it made sense to me and fit well with a registry.

Doug: Before the hydronephrosis registry started, had there been other pediatric urology registries?

Tony: There were two registries that were running. The MCDK and Testis Tumor registry.

Doug: When did the Hydronephrosis Registry begin? When did you enroll your first patient? And what institutions were originally included in the registry?

Tony: There were initially three centers involved: University of Connecticut, Johns Hopkins and Baylor. I was a senior resident in 1999 rotating on Pediatric Urology at the University of Connecticut, when it started on paper. I emailed “Follow-Up sheets” (for data collection) to Linda Baker and Tom Kolon and we followed patients initially with prenatally detected vesicoureteral reflux (VUR). We published the first multi-center study, which confirmed that a majority of prenatal VUR was in boys and a significant portion had renal dysplasia on DMSA imaging in the absence of UTI. Equally, we identified that circumcision status had a significant impact on UTI.

Doug: What were your original goals/aims with the registry?

Tony: After the initial success of this manuscript and I moved up the ranks in the Society of Fetal Urology (SFU), the registry was incorporated into the SFU. The main goal was to give the SFU an identity and it only made sense to have this married to a longitudinal registry. This was about 2005 and a similar concept was being discussed in Europe, but never got off the ground.

The initial goal of the registry was to digitally warehouse data, to serve as a clinical repository to which one could access the data - to test any hypothesis with respect to patient outcomes and management. We set up very loose ground rules with respect to intellectual property and whom would present at meetings.

Doug: How many publications have come from the data within the registry?

Tony: We have had a number of SFU presentations and 3 peer-reviewed long term publications. Just this year, we have broken through, and had a prize finalist at the 2020 American Urological Association meeting and have another excellent abstract that was accepted for the combined European Society of Pediatric Urology/Society of Pediatric Urology meeting for April 2021.

Doug: How has the registry changed over time? Certainly, it has changed by adding more institutional members, but what other changes have occurred? How has data input/collection changed? How has the data team changed over time?

Tony: The short answer is that I have done what I could do - without extramural funding. There are a number of things that have made this, in what I see, as a successful endeavor. Primarily, my stubbornness or persistence in pushing this day in and day out. Incorporating the registry into the SFU was integral, as it made it less about me and more about the organization, and it also gave the SFU an identity. With such a small number of pediatric urologists and at least 4 organizations, I felt this was an important step in maturation of the SFU and assurance of success, because it was equally beneficial. The second component was intramural funding, which I have been fortunate to obtain at all three of my stops along my career path.

With respect to the data team, version 1.0 was in Birmingham and with most things, “timing” creates opportunities that may otherwise not be available. One of my marathon training partners ran a “Trauma Registry” and he was able to replicate this on an Access database. At the time, we had 3 institutions which included the University of Connecticut, the University of Alabama at Birmingham, and the University of Iowa. Upon my arrival to the University of Virginia (UVA), we transitioned to a new team and version 2.0 was launched with the same institutions and the addition of UVA and the University of Wisconsin. Finally, upon my transition to Richmond, version 3.0 was launched with the role out of our REDCap database, which allowed access on a much broader level. The previous internet version was quite cumbersome.

I do feel compelled to mention that Kathy Herbst has been the backbone behind the scenes for this registry, and I am not sure many people realize this. She has been selfless in willingness to spend countless days and weeks cleaning the data and educating our team to assure that we get this right. I cannot think of many people that I have been around in my career that are as committed to the process as Kathy.

Doug: For those thinking about starting a registry, what advice would you provide them?

Tony: Call Kathy!!!!!!!!!.....Seriously, pick something that is widely applicable. Secure funding and an infrastructure that includes a data manager and someone with knowledge in the space of Population Health.

Doug: If there was something that you could have changed at the inception of the hydronephrosis registry what would it be?

Tony: To be honest, I am happy with how things are playing out. The pivotal step was the incorporation of the McMaster registry and (continued on next page)
Hydronephrosis Registry Turns 21 (continued from previous page)

brining our friends from Children’s Hospital of Orange County on board. This did a couple of things. First, it increased our numbers, but more importantly, it made our population management more diverse. This pushed us over the top with respect to meaningful multi-center data.

Doug: What are the biggest lessons that you have learned about running a registry?

Tony: Patience!!!!.....This in combination with adopting a Stoic philosophy has served me well recently. The obstacle is the path certainly rings true.

The ground rules have always allowed each institution to use their own data and my data manager free of charge. In my mind, the “carrot” works better than the “stick”. Not many folks have the time or energy to push folks to do things. On the other hand, if you are self-motivated to enter data, then it will run on its own.

The most difficult aspect of recruitment of centers into the registry is data entry. We have had a number of centers “knock at the door” - only to later “ditch it”, to use a colloquialism, because they did not have time to enter data. Certainly, the availability of REDCap should help this, but at the end of the day, the data needs to be entered. We are working towards adopting a seamless transfer from the electronic medical record, but currently we are not quite there yet.

Doug: To date, what has the biggest achievement of the registry been?

Tony: The recent publications on UTI risk and selective lower urinary tract imaging have been exciting, as well as the work on defining which patients benefit from prophylactic antibiotics.

Doug: What do you hope the lasting impact of the registry will be?

Tony: Twofold, one of which I have mentioned before. My vision would be that the hydronephrosis registry becomes the identity of the SFU and that will be its legacy. From a clinical perspective, we have a little way to go, but using this instrument as a means to be incorporated in a shared decision-making platform for families, would be an exciting prospect. This would provide families, patients and practitioners real time data. We would need the big guns, like Bill Gates to facilitate this, however.

Doug: If others want to become involved with the registry, what is the best way to do that?

Tony: Very easy to answer.

Email me at claude.herndon@vcuhealth.org

We have a very streamlined cookie-cutter process and part of a shared IRB process which should facilitate things quite a bit.

Doug: Tony, thank you for your time and for all the work that you have put into this registry. Your persistence and diligence have led to exciting, valuable outcome data and your hard work and dedication stands as a role model for the pediatric urology community. We look forward to the continued fruits that come from this database.

Bladder Exstrophy with Normal Appearing Phallus and Duplicated Urethra: Variants in EEC

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Introduction

Both bladder exstrophy and urethral duplication are rare congenital anomalies with bladder exstrophy occurring in 2-3 per 100,000 live births and less than 200 cases of urethral duplication having been reported in the literature.1 We present a unique case of a bladder exstrophy variant with normal appearing phallus and duplicated urethra.

Case Report

An 8-year-old boy presented to an international bladder exstrophy workshop with a bladder exstrophy variant. He had not undergone prior repair given limited access to medical care. On exam he had an exposed, easily depressible exstrophic bladder plate, with considerable squamous change and no umbilicus. His phallus was normal in size and appearance with a small protuberance of tissue at the penopubic junction that was suspicious for a second, dorsal urethra containing no lumen (Figure 1). He reported continuous urinary incontinence and normal, straight erections.

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Prior to bladder closure, renal ultrasound was normal and renal scan showed normal and symmetric renal function. Lab work including creatinine and electrolytes was within normal limits. Under anesthesia, we performed antegrade cystoscopy from the exstrophic bladder that revealed a competent appearing bladder neck leading to a normal posterior urethra, which was blind ending at the level of the membranous urethra. Retrograde cystoscopy was also performed via the glanular, orthotopic urethra which showed a narrow segment that ended approximately 1.5 cm proximal to the meatus.

He underwent bladder closure, anterior osteotomies, perineal urethrostomy and umbilicoplasty. The bladder was first dissected and freed circumferentially from the surrounding skin and fascia. Next the penis was degloved, initially leaving the suspected dorsal urethra intact (Figure 2). Further dissection revealed an obliterated dorsal accessory urethra with surrounding spongiosum tissue. We could not cannulate the lumen of this dorsal urethra but it appeared to connect with the anterior wall of the bladder between the proximal corporal bodies with a separate trajectory from the more normally located ventral urethra. We removed the dorsal accessory urethra and closed the bladder. Finally, we created a perineal urethrostomy with the assistance of a transverse preputial island tube to bridge the distance between the blind ending ventral membranous urethra to the perineal skin just posterior to the scrotum. The bladder, pubis and osteotomies were then closed and umbilicoplasty performed.

Prior to discharge, a cystogram showed the bladder to be well-healed and he was able to void via his perineal meatus with a strong stream and dry intervals of 5-10 minutes between voids. At the time of last follow up, approximately 5 months post-operatively, he continued to do well with no episodes of urinary tract infection and volitional voiding with dry intervals of 15-30 minutes during the day.

Discussion

Bladder exstrophy variants are rare conditions and only 8 cases of bladder exstrophy with duplicated urethra but not duplicated bladder have been previously reported. The largest case series published by Pippi Salle was 5 cases, 4 of whom were not recognized to have urethral duplication until after their initial closure. The unifying characteristics of these cases include a well-developed phallus with conical glans as opposed to the flat, epispadiac, glans and phallus typically seen in exstrophy patients, as well as a bladder plate that is deeply situated in the pelvis. As a rule, the ventral urethra contains the verumontanum and a competent bladder neck. These cases can be managed by simply closing the bladder and pelvis and excising the dorsal accessory urethra and because of the intact bladder neck within the ventral urethra, continence outcomes can be expected to be excellent.

Our case is unique in that our patient did not undergo treatment for his bladder exstrophy until 8 years at which point it was easier to recognize the urethral duplication than it might have been in a neonate. Additionally, because his intact ventral urethra did not pass urine for the first 8 years of life since his bladder plate remained open, a large portion of the urethra was stenotic—therefore requiring urethral reconstruction to a perineal urethrostomy with augmentation by a preputial island flap. Though rare, it is important for the pediatric urologist to be cognizant of exstrophy variants, such as duplicated urethra, as they require a high index of suspicion for diagnosis pre-operatively and alter both surgical management and expected outcomes.

References


Figure 2. Penile degloving and dissection identifying accessory dorsal urethra with spongiosal tissue (arrow indicates accessory dorsal urethra)
LUMBAR AND (P)LUTO: A RARE COMBINATION
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Introduction
LUMBAR syndrome is a rare clinical spectrum of congenital anomalies that occur in the presence of infantile hemangiomas (IH). More specifically, findings include lumbosacral hemangiomas, urogenital anomalies, myelopathy, bony deformities, anorectal and arterial anomalies, and renal anomalies.1 Herein, we present a case of a male with rare findings of prune belly and lower urinary tract obstruction [P(LUTO)] in the setting of LUMBAR syndrome.

Case Report
A 30 year old G2P1 woman was found to have a singleton male gestation with severe megacystis, left ureteral dilation, and bilateral hyperechoic kidneys on first trimester ultrasound (Figure 1). Given the implications of LUTO, the patient underwent vesicoamniotic shunt placement at 16 weeks. Additional shunts were placed at 19 and 20 weeks with simultaneous amnioinfusion due to shunt dislodgement and anhydramnios. Fetal MRI revealed bilateral hydroureteronephrosis, severe oligohydramnios, and a tapered rectum with dilated distal colon. Chromosomal analysis revealed a 12q14.1 microdeletion that was shared with the mother and unrelated to the presence of LUTO.

Following C-section at 34w6d due to fetal distress, baby was found to have abdominal wall laxity, distal hypospadias, undescended testes, imperforate anus, and a right buttock hemangioma (Figure 2). These findings were concerning for Prune Belly Syndrome with possible VACTERL overlap. The remaining vesicoamniotic shunt was not seen on exam, and no urine output was appreciated. Spinal ultrasound on day of life (DOL) 1 revealed a tethered cord. On DOL2, at the time of colostomy creation, attempted cystoscopy demonstrated near complete urethral atresia. A vesicostomy was created in addition to a left cutaneous ureterostomy due to significant left hydroureteronephrosis on postnatal ultrasound. CT cystogram revealed a retained vesicoamniotic shunt within the bladder, high grade right VUR, and a fistulous tract from the bladder neck/posterior urethra to the rectum. The shunt was ultimately retrieved cystoscopically via the vesicostomy at the time of central line exchange.

For the first 9 weeks of life, his kidney function remained stable and he did not require dialysis, however his course ultimately became complicated by ventilator dependent respiratory failure requiring tracheostomy placement. He developed further respiratory compromise in the setting of his pulmonary hypoplasia and pulmonary hypertension. Despite progression of renal failure, he was not a candidate for renal transplantation given the presence of significant pulmonary disease. After multidisciplinary discussions between pediatric nephrology, urology, general surgery, dermatology, and palliative care, the family ultimately decided to pursue comfort care and the patient passed peacefully at home at the age of 5 months.

Discussion
The pathogenesis of LUMBAR syndrome remains unclear. To date, 54 cases have been reported in the literature with an overall female predominance.1,2 A multi-institutional review of 53 cases by Iacobas et al. discussed risk of underlying anomalies based on four regional distributions of hemangioma: (A) lumbar, (B) sacral, (C) perineum/genital, (D) lower extremity.1 Of the cases with urogenital and renal anomalies, all had IH within region B. Overall, urogenital and renal anomalies were seen in 32% and 28% of cases, respectively, and solitary kidney was the most common renal anomaly observed.

Lower urinary tract obstruction is a rare finding in the setting of LUMBAR syndrome and contributes to significant morbidity and mortality. Prenatal management varies based on the degree of obstruction and amniotic fluid measurements, and typically involves vesicoamniotic shunt placement with or without amniocentesis. Delayed recognition can have serious implications in both pulmonary development and upper tract deterioration. Despite efforts to alleviate the obstruction either pre- or postnatally, a percentage of these patients ultimately go on to receive renal transplantation. Of children undergoing renal transplantation for a urologic condition, nearly 26% have primary problem of congenital obstructive uropathy.3 Management of the urologic anomalies seen in LUMBAR syndrome is further affected by the presence of underlying myelopathy, which occurs in 80% of cases most commonly

Figure 1: Prenatal ultrasound demonstrating megacystis (red arrow) and left ureteral dilatation (yellow arrow).
**Lumbar and (P)LUTO (continued from previous page)**

due to tethered cord. Therefore, evaluation for neurogenic bladder should be carefully considered prior to planning future reconstruction of urogenital anomalies or renal transplantation.

While overlap with other congenital syndromes exists, suspicion for LUMBAR should be raised in the setting of lower body hemangiomas. Early recognition and intervention by a multidisciplinary team is essential in managing this complex condition. Additionally, setting appropriate expectations for families is equally as important given the wide variability of LUMBAR syndrome.

**Figure 2: Abdominal (left) and perineal (right) exam findings, significant for abdominal wall laxity, right buttock and lower extremity hemangioma, empty scrotum, dorsal hooded prepuce with distal hypospadias, and imperforate anus.**

**References**


Dialogues in Pediatric Urology

**Time is Nephrons: The Importance of Prenatal Imaging in Detection and Timely Intervention to Protect Renal Function in Patient’s with Posterior Urethral Valves**

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**Introduction**

With an incidence of 1 in 5000 male pregnancies, posterior urethral valves (PUVs) are the leading cause of congenital bladder outlet obstruction.\(^1\) Despite the high frequency of this diagnosis, only 35% of cases are detected prenatally, limiting the possibility for prenatal or prompt postnatal intervention and preservation of renal function.\(^2\) Changes in renal development begin as early as 14 weeks gestation, yet prenatal screening US doesn’t occur routinely until 20 weeks gestation.\(^3\) There are 3 main time frames for intervention in management of PUVs: in utero, early postnatal and late (greater than 1 year of age). In utero intervention is typically achieved with vesicoamniotic shunting and is performed in specially selected cases. Early postnatal intervention with vesicostomy or PUV ablation has been shown to have a significant improvement in long term renal outcomes when compared to late intervention.\(^4\) Early identification on prenatal imaging and coordinated postnatal intervention are paramount to preserving renal function in males with PUVs.

**Case Report**

A 27-year-old G2P0 female, pregnant with a male fetus, was found to have bilateral SFU grade 4 hydronephrosis with mild parenchymal thinning on 25-week ultrasound (US). The bladder was moderately distended, without a keyhole sign. Amniotic fluid index (AFI) was normal at 11. She subsequently underwent further evaluation with a fetal MRI (Figure 1) which confirmed bilateral SFU grade 4 hydronephrosis and hydroureter. US at 32 weeks gestational age showed persistent bilateral SFU grade 4 hydronephrosis with hydroureter and worsening parenchymal thinning. The bladder was markedly distended with wall thickening. AFI was 2.5, consistent with oligohydramnios. The decision was made to deliver the baby at 34 weeks with immediate transfer to Children’s National Medical Center after delivery. A catheter was placed shortly after birth and initial postnatal creatinine was 2.6. A voiding cystourethrogram (VCUG) (Figure 2) was obtained showing a PUV and bilateral grade 5 vesicoureteral reflux (VUR). Patient was taken to the operating room at 1 week of age for a transurethral valve ablation, but the urethra was too small to accommodate an infant resectoscope. Therefore, a vesicostomy was performed. The creatinine down trended, with the most recent creatinine of 0.69 at 8 months old.

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Discussion
Despite the presence of severe bilateral grade 5 VUR, our patient with a PUV had significant improvement in renal function with prompt in utero diagnosis and early postnatal intervention.

There are conflicting reports regarding whether early diagnosis in utero portends a poor outcome. Presumably severe bladder outlet obstruction would be easier to diagnose early in gestation with more reliable radiologic findings, therefore, those patients might tend to do worse. Additionally, some assume that patients who present late postnatally have a lesser degree of obstruction, which is why it was not noted on prenatal imaging.

However, more recent reports suggest that later diagnosis and intervention lead to a greater likelihood of chronic kidney disease. One study quotes that at 10 years post-diagnosis, 30% of those with early intervention had chronic kidney disease versus 41% of those with late diagnosis.5 A multi-center study looking at 315 patients with PUVs, separated into 2 groups (antenatal versus late diagnosis) with a mean follow up of 5.5 years showed that CKD developed in 19% of the antenatal group and 40% in late diagnosis group.4 Serum creatinine nadir (0.6 versus 0.8, respectively) and creatinine at final follow up (0.9 versus 1.7, respectively) was also significantly different.4 Therefore, it appears that prenatal recognition and monitoring is crucial to allow for timely intervention postnatally.

Our patient had early prenatal recognition of outlet obstruction and immediate postnatal diagnosis of PUV which lead to early intervention. The patient was delivered early with planned transfer to a tertiary care center in order to achieve a timely procedure. Though his urinary tract did sustain damage due to obstruction in utero, prompt intervention allowed him to regain significant renal function and unobstructive bladder function as well.

References

FROM THE EDITOR

Dear Friends and Colleagues, both near and far:

This excellent SFU Edition speaks for itself, so there was little room for comments from the Editor.

However, when Doug Storm shared these screenshots from the meeting, I was reminded of how remarkable the pediatric urology community has been in sharing ideas and expertise through various virtual educational activities during this very strange year of 2020. Kudos to each of you who has bravely appeared for a Zoom lecture or panel before an invisible and eerily silent local, national or international audience!

SFU Case Presentations and Cocktails, COVID edition

I can’t wait to close my laptop once in a while and to see you all again in person sometime in the not-so-distant future. Until then, wishing you a meaningful and healthy 2021!

Elizabeth

P.S.: Should things not turn around as quickly as we hope, please refer to Tony Herndon’s application of the philosophy Stoicism (see SFU registry interview with Dr. Herndon above) to quietly accept this turn of events and move forward productively!

Time is Nephrons (continued from previous page)

Ectopic ureter panelists share wisdom virtually

FROM THE EDITOR

Elizabeth B. Yerkes, MD

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Dialogues in Pediatric Urology

Prostatic Utricular Cyst with Ectopic Ureter Presenting as Anuria in a Newborn

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Introduction
Prostatic utricular cysts are rare and present a challenge in diagnosis and management. We report a case of a prostatic utricular cyst with an ectopically inserting ureter found on workup for anuria in a newborn.

Case Report
A two-day old male who was born at 34 weeks and 1 day via cesarean section due to premature rupture of membranes, chorioamnionitis, oligohydramnios and breech presentation, presented with anuria. Antenatal history was notable for a right dysplastic kidney and normal left kidney detected at 21 weeks gestation on a comprehensive ultrasound. Subsequent surveillance ultrasounds demonstrated normal amniotic fluid levels. Family history was notable for an absent left kidney in the patient’s mother and multicystic kidney disease in the maternal grandfather.

On presentation, serum creatinine was elevated at 2.2 and a renal-bladder ultrasound was notable for bilateral multicystic kidneys without evidence of hydronephrosis, and a distended, fluid-filled structure believed to be the bladder (Figure 1). Urethral catheterization was attempted, but there was no return of urine and the catheter was unable to be visualized within the bladder on ultrasound. Urgent ultrasound-guided bedside aspiration was performed for decompression. Subsequently, a foley was placed and a voiding cystourethrogram was obtained that showed no evidence of outlet obstruction. However, it did reveal a large, midline cystic mass with mass effect on the bladder, approximately 80 mL in volume. This was also visualized on ultrasound.

The patient’s renal function continued to deteriorate and a peritoneal dialysis catheter was emergently placed on day of life 3. During catheter placement, the cystic structure was biopsied and drained. Pathology revealed the mass was of Mullerian origin, suggestive of a prostatic utricular cyst. Despite drainage, the cystic structure recurred and an MRI was obtained to better elucidate the patient’s anatomy (Figure 2). MRI revealed bilateral dysplastic kidneys, a large prostatic utricular cyst draining an ectopic left ureter, a right-sided ureterocele, a small contracted bladder, and streak gonads.

Percutaneous drainage of the left collecting system was unsuccessful, but a drain was successfully placed in the utricular cyst. Retrograde studies revealed no passage of contrast into the ectopic ureter or urethra. Several additional attempts were made to decompress his urinary system. First, the right-sided ureterocele was incised endoscopically, and the utricular cyst was unroofed transurethrally. The utricular cyst was then drained percutaneously but recurred. Next, the cyst was externalized though the creation of a stoma, and a percutaneous nephrostomy tube was placed into the left renal collecting system. Finally, the utricular cyst was excised, and a left ureteral reimplant was attempted but was unsuccessful due to the hypoplastic ureter. The patient’s renal function continued to deteriorate, and he is currently on dialysis awaiting transplant.

Discussion
In evaluating patients who present with lower urogenital tract cysts, location of the cyst allows for narrowing of the differential. Extraprostatic cysts include those of the seminal vesicle, Cowper’s duct, and vas deferens. Intraprostatic cysts can be further classified as lateral, paramedian, and median, with median prostatic cysts including Mullerian duct cysts or prostatic utricular cysts.1

Figure 1. Renal bladder ultrasound obtained on day of life 2 demonstrating (a) right dysplastic kidney, (b) left dysplastic kidney, and (c) fluid filled structure, measured at 82 mL, believed to be the bladder

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Prostatic Utricular Cyst  (continued from previous page)

Prostatic utricular cysts occur in up to 5% of the population, with higher incidences in individuals with other genitourinary abnormalities including hypospadias, disorders of sexual development, cryptorchidism, and ipsilateral renal agenesis.\(^2\) They occur due to a failure of complete regression of the Mullerian duct, which occurs during the first trimester in response to Mullerian Inhibiting Factor. Although many individuals will remain asymptomatic, patients may present with recurrent urinary tract infections, epididymitis, incontinence, or hematospermia. There is also a risk of infertility and neoplastic degeneration.\(^1\)

Due to the long-term sequelae and complications associated with prostatic utricular cysts, surgical excision may be performed. Several surgical techniques have been used, including extravesical, transvesical, perineal, and transrectal approaches.\(^3\) Each of these approaches has variable success rates and limitations. More recently, endoscopic management with simultaneous cystoscopy of the utricular cyst has been utilized, allowing for easy laparoscopic identification of the utricular cyst while avoiding the limitations of other surgical approaches.

References

Figure 2. MRI with (a) coronal view demonstrating bilateral dysplastic kidneys and (b) prostatic utricular cyst
An Argument for Obstruction: In utero Multicystic Dysplastic Kidney with Associated Ureterocele

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Introduction
Multicystic dysplastic kidney (MCDK) is a form of renal dysgenesis which results in a non-functioning kidney. While not harmful in the setting of a normal contralateral kidney, this anatomic abnormality may be associated with a variety of other urinary tract findings. Rarely, MCKD is seen in the setting of hydronephrosis and more infrequently, with a concurrent ureterocele. These findings in particular may shed light on the etiology of this abnormality.

Case Report
A 31-year-old female was referred for anomalous renal findings noted on a 24-week prenatal ultrasound. This revealed numerous non-communicating cysts of the right kidney, concerning for multicystic dysplastic kidney. Additionally, a right sided ureterocele and hydronephrosis were noted.

The mother’s medical diagnoses included depression and she was an active smoker at 1.5 packs per day. She is G0P0 with no family history of congenital renal abnormalities. The pregnancy had been complicated by intrauterine growth restriction, though amniotic fluid levels remained normal throughout. The patient was born at 32-weeks gestation via emergency C-section for suspected placental abruption. After a brief stay in the NICU the patient with discharged home with no complications. At 4 weeks of life the patient was seen for follow-up in the pediatric urology clinic. A repeat ultrasound again demonstrated right sided MCDK with associated right sided ureterocele (Figures 1 and 2). A voiding cystourethrogram was performed with no vesicoureteral reflux detected bilaterally.

Discussion
MCDK is the finding of numerous non-communicating renal cysts which ultimately results in a non-functioning kidney due to lack of normal parenchyma. Two main theories exist as to why this abnormality develops. The ureteric bud theory suggests this is due to an abnormal interaction between the ureteric bud and the metanephric tissue (1). Alternatively, the obstruction theory proposes that an atretic ureter or renal pelvis induces an extreme form of obstructive hydronephrosis which ultimately results in development of MCDK (2). This has been supported by the findings of normal renal parenchyma between the cysts in studies of fetal kidneys (3). This suggest that the metanephric tissue had in fact undergone induction of by the ureteric bud at some point in development. It is the back up of urine, commonly due to an atretic ureteral segment, which leads to cystic degeneration. In some cases, the obstructive cause may not be related to the ureter itself, but to the presence of a ureterocele, as seen in this case. The ureterocele causes obstruction which ultimately leads to cyst formation in the developing kidney. The radiographic findings demonstrated here provide support for the obstructive theory as the cause of MCDK development as we see a cystic renal unit with presence of hydronephrosis and a ureterocele. Alternatively, there exists the possibly of convergent etiologies with multiple pathways resulting in this singular abnormality. The different anatomic variations in each case may well be a clue in the underlying mechanism of disease in that specific renal unit.

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