Hiep “Bob” Nguyen, M.D., President, Society for Fetal Urology

In 2008, The Society for Fetal Urology held its 41st biannual meeting. In May, the spring meeting was in Orlando, Florida and was chaired by Dr. Richard S. Lee. The program was focused on the bioethics of prenatal counseling. Dr. David Diamond from Children’s Hospital, Boston, provided insights on why medical ethics are particularly important to the surgeon and on the opportunities available in ethics training for the surgeon. Dr. Walter Robinson from IWK Health Centre, Halifax, Nova Scotia discussed the bioethics of prenatal counseling and role of the ethics committee.

In October our society participated in a joint meeting with the AAP Section on Urology, The Pediatric Urology Nurse Specialists and the International Children’s Continence Society in Boston, Massachusetts. Working with these organizations, Dr. Aseem Shulka, the program chairman, assembled a program focused on the developmental aspects of continence and the potential future treatment for incontinence. Dr. Antoine Khoury from the University of California, Irvine Medical Center and the Children’s Hospital of Orange County reviewed the Valve Bladder Syndrome and incontinence in adolescents with posterior urethral valves. Dr. George Daley from Howard Hughes Medical Institute and Harvard Medical School provided the state of art lecture on stem cell research. Dr. Laurence Baskin from the University of California, San Francisco Medical Center presented our current understanding of the anatomy and development of the bladder neck and external sphincter.

The attendance for our biannual meeting last year was at a record level and our memberships continues to grow exponentially. It is our mission to promote fetal and prenatal urology, appropriate practice, education as well as exchanges between practitioners involved in the treatment of hydronephrosis. To that end, our two current major projects are the Prenatal Hydronephrosis Registry and the consensus statement for “Prenatal Hydronephrosis.” Three years since its inception, the registry is accruing a greater number of patients for the antenatal and postnatal evaluation of children with prenatally diagnosed hydronephrosis. Currently, we are actively recruiting for children with SFU Grade 3 prenatal hydronephrosis to evaluate the natural history of these patients. It is well recognized that there is much variability in the prenatal and postnatal management of children with prenatally diagnosed hydronephrosis. With a consensus statement, our goal is to define our current recommendations on the management of these patients and to identify further areas of research that may be addressed through recruitment of patients in the Prenatal Hydronephrosis Registry.

It would not be possible to accomplish the mission of our society without the active involvement and input from our members. As always we encourage any suggestions and comments with regards to our current projects, meeting topics and any other fetal urological issues. Our 42nd Biannual Meeting will begin on April 24th in Chicago. This year’s program chairman, Dr. John Gatti has assembled a panel of experts to discuss the components of study design and its importance in furthering our understanding of fetal/neonatal urology. From this program, we hope to provide the participants with a framework for conducting evidence/outcome based research and for interpreting the quality of studies published in the literature.

Hope to see you in Chicago. See if you can bring a colleague new to SFU along to the meeting.
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Anuria and Urinary Ascites in an Infant with Antenatal Unilateral Hydronephrosis and Normal Contralateral Kidney

Daniel A. Hirselj, M.D.1, Paul M. Zmaj, M.D.2, Casimir F. Firlit, M.D., Ph.D.2, 1Section of Urology, Nationwide Children’s Hospital, Columbus Ohio1 and 2Division of Pediatric Urology, Cardinal Glennon Children’s Medical Center, Saint Louis Missouri

Introduction

Hydronephrosis is a common fetal ultrasonographic diagnosis. Postnatal screening is recommended in many, though the appropriate timing of evaluation is debated. Urinary ascites in an uncommon, life-threatening condition that is most commonly seen in the neonatal population associated with lower and upper urinary tract obstruction. We present a case of a 4-week old infant with antenatal unilateral hydronephrosis presenting with anuria and urinary ascites and found to have obstruction of his contralateral kidney.

Case Presentation

A 35-week premature male with antenatal unilateral hydronephrosis underwent postnatal ultrasound on his 10th day of life. Imaging demonstrated severe left hydronephrosis and minimal right hydronephrosis (Figure 1a,b). Postnatal VCUG was normal and serum creatinine at that time was 0.6 mg/dL.

He presented to the emergency department on DOL #28 with 20 hours of anuria, lethargy, and poor feeding. Examination revealed abdominal distension and absent bowel sounds. Serum chemistries showed azotemia (Cr:1.9 mg/dL) and hyperkalemia (7.3 mg/dL). Urethral catheter was placed without urine return. Abdominal ultrasound demonstrated stable left hydronephrosis with worsening right hydronephrosis, right perirenal fluid, intraperitoneal fluid and right pleural effusion (Figure 1 c,d).

The patient was taken to OR, and retrograde pyelography identified right upper pole fornical rupture with urinary extravasation. Exploration, peritoneal lavage, and pyeloplasty with nephroureteral stent were performed. Azotemia and hyperkalemia resolved within 24 hours. Subsequent evaluation confirmed drainage from his right kidney with obstruction and poor function contralaterally.

Discussion

Postnatal evaluation of antenatal hydronephrosis is recommended in many children. The appropriate timing of evaluation, however, is not agreed upon.1,2 Most agree that low grades of hydronephrosis do not require follow-up.1,2,3 Others have noted significant urologic pathology despite clinically non-significant hydronephrosis in the neonatal period.4 Despite low-grade hydronephrosis, our patient presented with obstruction of this renal unit and urinary ascites. Prompt evaluation, imaging and treatment successfully corrected this life-threatening problem.

References


Figure 1A: Ultrasound at DOL #10 demonstrating severe left hydronephrosis
Figure 1B: Ultrasound at DOL #10 demonstrating minimal right pyelecstasy
Figure 1C: Ultrasound at DOL #28 demonstrating persistent severe left hydronephrosis
Figure 1D: Ultrasound at DOL #28 demonstrating right hydronephrosis with perirenal fluid collection
Midline Perineal Mass in Newborn with Penoscrotal Hypospadias

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Introduction

Although the etiology of hypospadias is unknown, cases of genital malformation in the presence of a perineal mass have been reported. We present an unusual case of a midline perineal mass in a newborn male with severe hypospadias.

Case Report

A newborn patient followed prenatally for ambiguous genitalia presented at one day of age with a midline perineal mass. Physical examination revealed perineal hypospadias, a bifid scrotum, and a serpiginous soft tissue mass extending between the meatus and anus (Figure). The patient had normal anal position and configuration and had passed meconium. Additionally, MR urography revealed a dysplastic left kidney. Attempts to pass a catheter for VCUG were unsuccessful. The patient was discharged and returned two weeks later with bleeding and fibrinous exudate at the site of the mass. Intraoperative cystoscopy, VCUG, and contrast enema did not reveal a confluence between the mass and either the genitourinary tract or gastrointestinal tract. The mass was excised, and the perineal wound was closed primarily. Final pathology revealed ectopic bowel. The patient had an uneventful postoperative course and excellent cosmetic result. He underwent successful hypospadias repair six months later.

Discussion

Genital malformations in association with aberrant midline perineal structures have previously been described.1 In a review of 23 cases of accessory scrotum, 19 were associated with perineal hamartoma or lipoma.2 Presence of a perineal lesion may have an impact on normal urethral and scrotal development by interfering with the midline fusion of the urethral folds and genital swellings. Intestinal duplication is exceedingly rare with reports in 1:18,000 live births. Typical features include a hollow structure lined with gastrointestinal epithelium that contains a layer of smooth muscle contiguous with that of the gastrointestinal tract. The most common forms of intestinal duplication arise from the ileum (30%) or ileocecal valve (30%), but all regions of the gastrointestinal tract from the base of the tongue to the anus may be involved.3,4 To our knowledge, this is the first reported case of ectopic extra-abdominal bowel presenting as a perineal mass with associated perineal hypospadias and bifid scrotum.

References


Eagle-Barrett Syndrome Associated with Urethral Atresia and a Patent Urachus

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Introduction

Eagle-Barrett syndrome is a rare condition that consists of the classic triad of abdominal wall musculature defect, genitourinary abnormalities and undescended testes. The syndrome has an incidence of 1/30,000 to 1/40,000 live births.1 We describe our patient born with Eagle-Barrett syndrome, a large patent urachus and urethral atresia.

Case Report

A 16-year-old mother presented to the office after a prenatal ultrasound performed at 16 weeks gestation revealed an extremely enlarged bladder and low amniotic fluid levels (AFL). Amniocentesis was attempted, and an incidental aspiration of bladder fluid revealed a 46XY karyotype. At 18 weeks gestation the AFL had normalized. Serial ultrasounds, performed between 19 and 26 weeks revealed bilateral hydronephrosis and an enlarged bladder. At 28 weeks, ultrasound demonstrated severe fetal ascites, but normal AFL. Fetal paracentesis was performed along with serial vesicocentesis in order to evaluate fetal renal function. The first paracentesis removed only 13 ml. The bladder was aspirated on three consecutive days for volumes of 165 ml, 87 ml, and 110 ml. The urine sodium was 58 mEq/L and osmolarity was 123 mosm/kg, predicting adequate renal function. At week 29, severe ascites had reaccumulated and was compressing the fetal diaphragm, thus paracentesis was repeated for 605 ml. At week 30, the fetus developed decelerations and low AFL were demonstrated by ultrasound. The baby was delivered at 30 weeks by cesarean section.

Upon delivery, the baby had an extremely large, floppy abdominal wall, bilateral undescended testes, and a defect at the base of the umbilicus, consistent with a patent urachus. Attempts to place a urethral catheter were unsuccessful. The baby was taken to the operating room for an antegrade and retrograde urethrogram and cystoscopy. Attempts at retrograde cystoscopy were unsuccessful. Antegrade cystoscopy via urachus revealed an atretic urethra. However, on retrograde urethrogram, a very small connection was identified between the urethra (continued on next page)
Is Reducing the Renal Pelvis Necessary During Pyeloplasty?

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Introduction

The Anderson-Hynes dismembered pyeloplasty for repair of ureteropelvic junction obstruction (UPJO), which is considered the gold standard to which other procedures are compared, was described in 1949. Although the original manuscript describes surgical reduction of the size of the renal pelvis, it was not until the early 1990s that the rationale for this practice became popular. At that time, many studies demonstrated histologic changes in the renal pelvis consisting among other things of deposition of excess collagen between the smooth muscle fascicles and general disorganization at the UPJ. Therefore, it was reasoned that removal of the diseased “pelvis” together with the UPJ at the time of dismembered pyeloplasty would yield better results. However, 3 recent studies seem to add controversy and go counter this argument.

Case Reports

In 2001, Diamond and Nguyen described a dismembered vertical flap pyeloplasty without surgically reducing the size of the renal pelvis. Similarly, Palmer et al in 2005 (cuff pyeloplasty) and Mesrobian (bypass pyeloplasty) independently described 2 different procedures for the repair of the obstructed UPJ associated with a high inserting ureter. They both preserved the redundant renal pelvis. The cumulative experience of these 3 series reports success in 30 out of 30 repairs with resolution or diminution in the degree of hydronephrosis.

Discussion

Congenital ureteropelvic junction obstruction is the most common renal anomaly detected by virtue of maternal sonography. The grade of hydronephrosis is variable but even with high grade disease, renal function may be well preserved (to the extent that we can evaluate it with current methods). Renal function is thought to be mediated in large part by the associated hydropnephrosis, which insures a low intrarenal pelvic pressure. Therefore, the redundant renal pelvis is protective and removing it would deprive the post-pyeloplasty kidney from its benefit at a time when it may need it most. A temporary indwelling stent may not be sufficient to compensate for the absence of a low pressure renal pelvis. In fact, it may take up to 3 to 5 months for repaired UPJ to recover near normal peristalsis according to 2 relatively recent experimental studies.

Therefore, excision of large portions of the renal pelvis during pyeloplasty for UPJO may not be desirable. In addition, when the renal pelvis is not surgically reduced, it appears that it may over time reduce itself as described in the above 3 different pyeloplasty procedures.

References

Prenatal Diagnosis of Bilateral Segmental Multicystic Dysplastic Kidneys

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Introduction
Bilateral renal multicystic dysplasia is usually fatal in utero and provides challenges in prenatal counseling. We present a case of prenatally diagnosed bilateral renal multicystic dysplasia with minimal normal tissue. At one year of life there has been almost complete involution of the cysts with compensatory hypertrophy of normal parenchyma.

Case Report
A G3P2 mother at 22 weeks gestation referred for bilateral, cystic, enlarged hyperechoic kidneys on prenatal ultrasonography without hydronephrosis or associated abnormalities. The parents were previously counseled to abort the fetus. Amniotic fluid levels remained stable and the parents continued with observation.

Postnatal ultrasound consistent with bilateral, segmental multicystic dysplastic kidneys with small amounts of spared tissue bilaterally. Cystourethrogram did not elicit vesicoureteral reflux. Child remained stable and had resolution of almost all cysts within one year with compensatory hypertrophy of normal parenchyma. Renal function and blood pressure remain normal. A midshaft hypospadias was present and surgically corrected.

Discussion
Multicystic dysplastic kidney (MCDK) is a congenital disorder with a prevalence of approximately 1:4000 live births. It is almost entirely unilateral and characterized as an obstructive process with multiple cyst formations with little to no functional parenchyma. Bilateral cases are usually incompatible with life and only rare cases exist.1 Detection of MCDK on prenatal ultrasound is up to 97% for unilateral disease and 91% for bilateral renal agenesis/dysplasia.2

Segmental MCDK is a rare variant occurring when only a portion of the kidney is involved with cystic changes. Typically it presents in the upper pole of a duplicated system.3,4 Diagnosis and management is extremely difficult without other associated abnormalities and in bilateral cases.

One series of segmental MCDK presented six kidneys, one child with bilateral disease, of which four underwent spontaneous complete or partial involution.3 Antenatal counseling is critical in situations of oligohydramnios and bilateral renal cystic disease as this is unequivocally fatal.1

We present a case of bilateral segmental MCDK without known associated renal anomalies. This may represent a variant of MCDK or a separate process in nephrogenesis. Further investigation into other patients with similar abnormalities may provide insight into the disease process and potential outcomes for patients with similar presentations.

References

Figure 1. (Top row) Sagital views of the right and left kidneys on prenatal ultrasound at 22 weeks gestation by linear transducer. Right 4.67cm, Left 4.15cm. (Bottom row) Sagital views of right (lower left corner) and left kidneys (lower right corner) at 10 months of age. Right 5.65cm, Left 6.66 cm.
Evolving Pattern of Bladder Dysfunction in Boys with Posterior Urethral Valves (PUV)

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Introduction

Posterior urethral valves (PUV) are the most common congenital malformation of the lower urinary tract in boys with a reported incidence from 1:3000 to 1:8000 live male births.1-3,4 The mortality of neonates with this congenital malformation has decreased over years due to improved diagnostic and therapeutic strategies. However long term consequences in the form of impaired renal function and bladder dysfunction have become new challenges. Bladder dysfunctions are reported in 50-80% of patients with posterior urethral valves.5,6,7,8 The types of bladder dysfunction described include detrusor overactivity, decreased bladder compliance and detrusor underactivity.7,9,10,11 It is unclear whether boys with PUV demonstrate various types of bladder dysfunctions or display a changing pattern of dysfunction over time.8,10,11

Material

Among 82 boys treated for posterior urethral valves at a single centre over 21 years (1986-2007) 62 boys were available for follow-up. The mean age at diagnosis was 2.8 years (SD±3.53 years) with 68% presenting in the first year of life. The mean age at analysis was 8.6 years (range 1-15). The studied cohort was divided into a younger subgroup of 39 (62.9%) patients who were under 10 years of age (mean age 6.44± years; range 2-8) and an older subgroup of 23 (37.1%) boys over 10 years age (mean age 15.8± years; range 10-20). The severity of the initial manifestation (initial ultrasound findings, presence and grade of vesico-ureteral reflux and primary surgical intervention) was similar for both subgroups (Table 1).

<table>
<thead>
<tr>
<th>Type of Primary Surgical Intervention</th>
<th>Younger Group</th>
<th>Older Group</th>
<th>Chi-square</th>
<th>P value</th>
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<tr>
<td>Loss of cortico-medullary differentiation in initial ultrasound examination</td>
<td>16 (41.02%)</td>
<td>5 (21.7%)</td>
<td>2.57</td>
<td>0.275</td>
</tr>
<tr>
<td>- without</td>
<td>5 (12.8%)</td>
<td>3 (13.04%)</td>
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</tr>
<tr>
<td>- unilateral</td>
<td>18 (46.10%)</td>
<td>15 (65.21%)</td>
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<td></td>
</tr>
<tr>
<td>- bilateral</td>
<td>2 (5.12%)</td>
<td>1 (4.16%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bilateral high-grade (3-5) VUR</td>
<td>22 (56.41%)</td>
<td>12 (52.17%)</td>
<td>2.8</td>
<td>0.758</td>
</tr>
<tr>
<td>- absent</td>
<td>17 (43.58%)</td>
<td>11 (47.83%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>- present</td>
<td>5 (12.8%)</td>
<td>1 (4.16%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Type of posterior valve ablation</td>
<td>18 (46.15%)</td>
<td>15 (65.22%)</td>
<td>4.1</td>
<td>0.036</td>
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<td>- valve ablation</td>
<td>9 (23.08%)</td>
<td>1 (4.35%)</td>
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<tr>
<td>- primary valve ablation</td>
<td>7 (18.46%)</td>
<td>8 (33.33%)</td>
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</tr>
</tbody>
</table>

Method

The bladder function of boys with PUV in both age groups was evaluated by means of a bladder diary (according to recommendations of Standardization Committee of International Children's Continence Society) and an urodynamic examination.12

Bladder diary

Normal voiding frequency was considered to be 4 to 7 micturitions per day with adequate fluid administration. A decreased frequency was defined as 3 or less micturitions per day and an increased frequency as over or equal to 8. The maximal voiding volume was noted as bladder capacity (BC), which was compared to expected bladder capacity (EBC) for age. EBC was calculated for boys up to the age of 12 according to the formula: EBC (ml) = [30+ (age x 30)]. A bladder volume of 390ml was recognized as expected bladder capacity for boys over 12 years of age.13 Decreased bladder capacity was classified as below 65% of EBC and increased BC as higher than 150% of EBC.12

Urodynamic examination

Maximal urine flow, average urine flow, the shape of the uroflow curve and voided volume were noted during uroflowmetry. Post-void residual urine volume was assessed by ultrasound. Bladder capacity was calculated as the sum of voided volume and residual volume after micturition. Normal maximal urine flow was defined as (Qmax)2 > (V bladder). Uroflowmetry was performed in cooperative patients.12,14 The shape of urine flow was classified into four categories: bell, plateau and tower shaped curves and staccato voiding. Residual urine greater than 10% of bladder capacity or over 20 ml was assessed as abnormal.3,12

For cystometry the bladder was filled with 0.9% saline dilution at a filling rate of 10% expected bladder volume per minute. Measurements of bladder pressure were made by a double lumen catheter inserted transurethrally and abdominal pressure by a transrectal catheter. The activity of pelvic floor muscles was registered by two superficial electrodes placed in the anal region.

Intrabladder pressure, intraabdominal pressure, detrusor pressure, maximal cystometric bladder capacity, bladder compliance, EMG of pelvic floor muscles was measured or calculated during the filling and voiding phases of cystometry.

Detrusor overactivity was defined as an involuntary increase of detrusor pressure over 15 cm H2O above baseline.12,15 Decreased compliance was defined as compliance equal or smaller than 10 ml/cm H2O or a change in pressure during filling greater than 15 cm H2O from beginning to expected bladder capacity for age.12,16 Bladder pressure during voiding was classified as decreased when maximal detrusor pressure was lower than 50 cm H2O, normal for values between 50 cm and 90 cm H2O and increased when values exceeded 90 cm H2O.8 Dysfunctional voiding was recognized when pelvic floor muscle activity was recorded by EMG during the voiding phase.12

Statistical analysis was performed using STATISTICA 7.1 version (StatSoft Inc, Tulsa, Oakland). For analysis of differences between groups chi square test was used with p<0.05 considered statistically significant.

Results

Urodynamic abnormalities were present in 89% (55/62) of boys with PUV. Increased bladder capacity compared to expected BC for age was observed in 16/62 (25.8%) patients and decreased BC in 7/62 (11.2%). Abnormalities of voiding frequency were detected in 34% (20/62) patients: 17 (28.9%) boys showed increased and 3 (5.1%) decreased voiding frequency. There were no significant differences in bladder capacity (p=0.669) or voiding frequency (p=0.593) abnormalities between the younger and older subgroups.
Abnormal uroflowmetry curves were detected in 63% (36/57) of the examined patients (plateau-shaped - 45.6%, tower-shaped - 5.2% and staccato voiding - 12.28%). Maximal urine flow was decreased in 49% (28/57) of patients. There were no statistical differences between age groups in the proportion of different flow curves (p=0.154) or decreased urine flow rates (p=0.49). Residual urine volume greater than 10% of BC was detected in 25% and higher than 20ml in 54% of boys. The proportion of boys with insufficient bladder emptying was significantly higher in older patients (73%) than the younger ones (41%) only for the latter criterion (p=0.014). Uroflowmetry increased voiding volumes were detected in 18/57 (31.6%) and decreased in 11/57 (19.2%) boys with PUV (Table 2).

Table 2 Uroflowmetry findings in boys with posterior urethral valves according to age.

<table>
<thead>
<tr>
<th>Flow curve shape</th>
<th>Boys &lt; 10 yrs (mean age 6.2 years)</th>
<th>Boys &gt;10 years (mean age 15.8 years)</th>
<th>Chi-square</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>16 (47.06%)</td>
<td>13 (56.52%)</td>
<td>0.49</td>
<td>0.48</td>
</tr>
<tr>
<td>Decreased</td>
<td>18 (53.94%)</td>
<td>10 (43.48%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Post-void residual urine</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&gt; 20ml</td>
<td>14 (41.18%)</td>
<td>17 (73.91%)</td>
<td>5.9</td>
<td>0.015</td>
</tr>
<tr>
<td>&lt; 20ml</td>
<td>20 (58.82%)</td>
<td>6 (26.09%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&gt;10% BC</td>
<td>10 (29.41%)</td>
<td>4 (17.31%)</td>
<td>1.1</td>
<td>0.301</td>
</tr>
<tr>
<td>&lt;10% BC</td>
<td>24 (70.59%)</td>
<td>19 (82.69%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bladder capacity</td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Decreased</td>
<td>8(23.53%)</td>
<td>3(13.04%)</td>
<td>0.980</td>
<td>0.612</td>
</tr>
<tr>
<td>Normal</td>
<td>16(47.06%)</td>
<td>12(52.17%)</td>
<td></td>
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<tr>
<td>Increased</td>
<td>10(29.41%)</td>
<td>8(34.78%)</td>
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</tbody>
</table>

A change of bladder pressure during filling greater than 15 cm H₂O was detected in 27 (43.5%) patients and a change in detrusor pressure during change of bladder volume smaller than 10ml/ cm H₂O was detected in 27 (45.1%). Decreased compliance occurred more frequently in the younger group of patients for both of the respective parameters analyzed (p<0.0001, p<0.002). Detrusor overactivity was seen in 23 patients with PUV. This abnormality was more frequently observed in the younger age group (53% v. 8%), (p=0.00038). Maximal voiding pressures were decreased in 12 (19.35%) patients and increased in 22 (35.45%). Detrusor pressures over 90cm H₂O during voiding were observed more frequently in younger (48.72%) than older boys (13.04%) (p<0.002). In older patients the proportion of detrusor underactivity (39.13% v. 7.69%) was significantly higher. Dysfunctional voiding was present in 29 (46.7%) patients with PUV, predominantly in the younger subgroup of patients (56%) (p<0.05). Straining during voiding was detected in 32 (51.6%) patients. This occurred more frequently in older children (65% v. 43%) but the difference was not significant (p=0.28). The detailed urodynamic findings are presented in Table 3.

Table 3 Cystometry findings in boys with PUV according to age.

Table:<br>Table 3 Cystometry findings in boys with PUV according to age.

<table>
<thead>
<tr>
<th>Bladder compliance</th>
<th>Boys &lt; 10 yrs (mean age 6.2 years)</th>
<th>Boys &gt;10 years (mean age 15.8 years)</th>
<th>Chi-square</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Overactive</td>
<td>21(53.85%)</td>
<td>2(8.70%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Decreased</td>
<td>3 (7.69%)</td>
<td>9 (39.13%)</td>
<td>12.6347</td>
<td>0.00181</td>
</tr>
<tr>
<td>Normal</td>
<td>17 (40.9%)</td>
<td>11 (47.83%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Increased</td>
<td>19(48.72%)</td>
<td>3 (13.04%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dysfunctional voiding</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Present</td>
<td>22(56.41%)</td>
<td>7(30.43%)</td>
<td>3.921021</td>
<td>0.04769</td>
</tr>
<tr>
<td>Absent</td>
<td>17(43.6%)</td>
<td>16(69.57%)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Straining</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Present</td>
<td>17 (43.5%)</td>
<td>15 (65.22%)</td>
<td>2.709764</td>
<td>0.09974</td>
</tr>
<tr>
<td>Absent</td>
<td>22 (56.41%)</td>
<td>8(34.78%)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Discussion<br>Bladder dysfunction was detected by urodynamic studies in the majority of boys with PUV (89%). Voiding abnormalities in the form of increased voiding frequency and wetting accompanied decreased bladder capacity and detrusor overactivity. Incomplete bladder emptying was frequent in boys with increased bladder volume and low detrusor pressure. A comparison of uroflowmetry findings in younger and older boys with PUV revealed a significantly higher proportion of post-void residual urine in patients > 10 years age but only when more restrictive criteria were used (post void urine volume greater than 20ml). There were no statistically significant differences when the post-void residual urine was calculated as higher than 10% of voided volume. Previous publications have noted no differences between age groups when significant residual volume was defined as 10% or even 20% of voided volume. It seems therefore that straining enables boys with PUV to empty over 90% of their bladder volume, but evacuation is not complete, leaving a significant amount of post void residual urine.

The comparison of cystometry findings in younger and older boys with PUV revealed significant differences in a number of abnormalities observed. Decreased compliance occurred more frequently in the younger than the older age group (64% v. 13%), an observation re-
ported also by Holmdahl et al. who examined bladder compliance in boys with PUV in different age groups. This finding is probably caused by reactive hypertrophy of the detrusor muscle to outlet obstruction with a resulting increase in collagen fibers in the bladder wall. This leads to a loss of elasticity of the bladder which cannot increase its capacity without an increase of pressure. Similar changes have been described in an animal model of bladder outlet obstruction. Younger patients also demonstrated detrusor overactivity more frequently, a finding reported previously by other groups.  

In our opinion during childhood the PUV bladder begins to lose its capabilities of adequate contraction with the gradual increase in urine output and continuous stretching of the abnormal bladder wall. This is reflected in the urodynamic findings of older boys in whom the proportion of detrusor overactivity and low compliant bladders is much lower. The observation of an age dependent pattern of maximum voiding pressure confirms this theory. 48% of boys under 10 years of age demonstrate maximum voiding pressure over 90 cm H2O and a minority have maximal voiding pressure below 50 cm H2O (7%). This proportion reverses in older boys with only 13% of patients generating voiding pressures over 90 cm H2O whereas 40% demonstrate decreased maximum voiding pressure. Our findings support the publications of Holmdahl and De Genaro who suggest that an evolution of bladder dysfunction occurs in patients with PUV. Initially low complaint bladders with detrusor overactivity decompensate with time and become abnormal large, low pressure containers which cannot empty effectively.

Conclusions

Bladder dysfunction is an integral component of a complex posterior urethral valve syndrome. The pattern of bladder dysfunction evolves during childhood from a low complaint and overactive bladder to a decompensated, underactive bladder. Regular urodynamic follow-up is essential for patients with PUV for adequate recognition and treatment of these changing bladder dysfunctions.

References

hold larger volumes. It is believed that adolescents with valve bladders have poor sensation, thus tolerate large volumes, and void infrequently and incompletely. This voiding inefficiency is multifactorial. The myogenic detrusor failure in combination with high-grade reflux and bladder outlet dysfunction results in incomplete voiding. The high post-void residual volume reduces the functional bladder capacity, such that storage occurs in the high-pressure zone of the filling curve (a right shift on the urodynamic curve).

Long-term Data from Sick Kids in Toronto

A review of the uroflow and urodynamic data in the post-pubertal valve population in Toronto was completed to determine the long-term evolution of bladder capacity, and the impact of primary ablation vs. diversion on bladder capacity, and the need for bladder augmentation.

We reviewed 78 patients with PUV born between 1985-1995. Twelve patients underwent bladder augmentation. Of the 66 patients with native bladders, 39 patients had multiple (>3) bladder capacity measurements by uroflowmetry or urodynamic studies. Median follow-up was 15 years, ranging from 10 to 18 years.

We found that although most children initially have smaller bladder capacities, after age 11, 85% develop a large bladder. This evolution of bladder capacity over time is demonstrated in Figure 1. Only 15% (6/39) of the patients had a bladder capacity that was smaller than expected at their last visit. Of these 6 boys, 3 were initially diverted by vesicostomy and 3 underwent primary valve ablation.

We compared patients who underwent primary ablation versus diversion (with subsequent undiversion without augmentation) and observed no difference in bladder capacity at the last follow-up visit (Figure 2). Temporary cutaneous vesicostomy has a specific role in the primary management of 5-10% of newborns with valves. However, it is important to note that diversion has no impact on renal dysfunction due to dysplasia and scarring. Diversion reverses detrusor hypertrophy but does not immediately alter the changes in the extracellular matrix. These changes remodel over time particularly when the vesicostomy is closed and the bladder begins to cycle again. On a longer-term basis, temporary diversion in a non-infected bladder does not appear to have a negative impact on ultimate bladder storage.

The severity of obstruction at birth and its impact on the bladder determined the need for bladder augmentation. The 12 patients who were ultimately augmented were all on the severe end of the spectrum and were all initially diverted. Seven of these underwent high diversion (bilateral ureterostomy/pyelostomy) and 5 vesicostomy.

Voiding efficiency was measured by uroflowmetry combined with ultrasound estimation of post-void residual. The majority of children were able to empty efficiently by adherence to timed voiding and double voiding. Self-catheterization with or without overnight catheterization was prescribed in 8/66 (12%) of patients with native bladders.

The role of and need for anticholinergic therapy at puberty should be reevaluated in view of these data demonstrating the expansion of bladder capacity. Confirming complete bladder emptying (by uroflowmetry and measurement of postvoid residual by ultrasound), may effectively improve functional bladder capacity without the need for anticholinergics.

In summary, initial dysfunction in the valve bladder results from a combination of factors including the degree of obstruction, high-pressure storage, incomplete emptying and loss of urinary concentrating ability. With careful management the majority of PUV patients achieve excellent bladder storage such that by puberty, 85% of these patients have a larger capacity than expected for age. Most are able to empty to completion with only 12% requiring self-catheterization.

The management of newborns with PUV requires long-term commitment by the treating team and the family with particular attention and reinforcement of voiding habits at puberty and adolescence.

References

A Prenatally Detected Bladder Mass
Sherry Sedberry-Ross, MD1, Compton Benjamin, MD2; and Hans G. Pohl, MD, FAAP1 1Children’s National Medical Center, 2 Department of Urology George Washington University, Washington, DC

Introduction
Prenatal sonography has facilitated the detection of genitourinary abnormalities, which account for over half of all fetal malformations.1 Abnormalities of the bladder are rare and commonly include ureteroceles, bladder diverticula, exstrophy, outlet obstruction or prune belly syndrome. We present a case of a prenatally detected bladder mass, including management, treatment and diagnosis.

Case Report
A 35 year old G7P5 woman presented for fetal evaluation after a standard 20 week gestation prenatal sonogram identified an elongated, intraluminal bladder mass at the dome of the bladder (Figure 1). Since no other abnormalities were found, full term delivery was recommended. The mass was confirmed on post-natal sonography. Cystoscopy demonstrated a pedunculated, benign-appearing mass at the bladder dome (Figure 2). The polyp was too large for endoscopic resection so it was excised through a small cystotomy. Pathological examination confirmed a 1.5 x 0.7 x 0.4 cm fibroepithelial polyp.

Discussion
Fibroepithelial polyps of the genitourinary tract are rare. They commonly present with hematuria and typically occur at the ureteropelvic junction, the upper third of the ureter and the posterior urethra.2 To our knowledge, this is the first case of a prenatally detected congenital fibroepithelial polyp of the bladder dome. When a smooth, polypoid filling defect in the urinary tract is identified, the diagnosis of a fibroepithelial polyp should be entertained. Given their benign nature, fibroepithelial polyps should be addressed by the least invasive means possible, unless size or location dictates otherwise.

References
Ureteroceles Presenting as Perineal Mass

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Introduction

A ureterocele is a cystic dilatation of the terminal ureter, associated with a single or duplicated ureter. The ureteral orifice may be located either in the bladder (intravesical form), or distal to the bladder neck (ectopic form). Ureteroceles occur more commonly in females than males (4:1 ratio), and almost exclusively in caucasians.1

In the past, febrile urinary tract infection (UTI) was the most common presentation of a ureterocele. Currently, most cases are diagnosed prenatally on ultrasound imaging. The use of magnetic resonance imaging for fetal diagnosis of ureterocele in select cases has also been described.2,6

The incidence of ureteroceles is estimated to be 1 in 500 births. Cecoureteroceles are a subset of ureteroceles, characterized by a tongue-like extension of the ureterocele into the mucosa of the bladder or urethra. The exact incidence of cecoureteroceles is unknown. We report a variation of a cecoureterocele presenting prenatally as a perineal mass.

Case Report

A 26-year-old caucasian G2P2 woman was referred for evaluation after prenatal ultrasound at 22 weeks of gestation showed bilateral hydronephrosis, a ureterocele, and a perineal mass (Figures 1A-C). Amniotic fluid index was normal. The patient was managed conservatively, and followed with weekly ultrasounds until term delivery. Serial ultrasounds showed no significant changes.

Physical examination of the female newborn revealed an interlabial perineal mass (Figure 2). Postnatal ultrasound revealed a “dumbbell-type” ureterocele, consisting of an intravesical portion, a tract extending parallel to the posterior wall of the urethra, and a perineal portion (Figures 3A-C). Given the radiologic findings, the perineal mass was considered the extravesical portion of the ureterocele. The urethral meatus was identified anterior to the perineal mass (Figure 4). The vaginal introitus was normal. A urethral catheter was placed for drainage of the bladder. Bedside incision of the perineal mass was performed, with resultant decompression. Follow-up renal-bladder ultrasound on the following day confirmed decompression of the intravesical portion of the ureterocele as well, but demonstrated persistent bilateral hydronephrosis. Prophylactic antibiotics were initiated. A VCUG was also performed, which showed no evidence of vesicoureteral reflux (Figure 5).
The patient was seen in the office two months later. She had no UTIs in the interim. On exam, there were no palpable abdominal masses, and the perineum was normal except for some residual mucosa from the ureterocele. An ultrasound at this time showed a decompressed ureterocele, bilateral grade II hydronephrosis, and a renal size discrepancy owing to a duplicated right collecting system with a dysplastic right upper pole (Figure 6). Voiding cystourethrogram (VCUG) demonstrated grade I reflux into the right upper pole ureter (Figure 7). Prophylactic antibiotics were continued.

At four months of age, the patient had a febrile UTI requiring hospital admission. Physical examination and renal-bladder ultrasound were consistent with left pyelonephritis. DMSA scan performed after resolution of infection demonstrated a small right kidney with 20% differential function on the right side, compared to 80% on the left. As the degree of hydronephrosis on ultrasound had not changed, management with prophylactic antibiotics was continued.

The patient did not have any further UTIs. An ultrasound done at one year of age showed a decompressed ureterocele and interim resolution of hydronephrosis. The upper pole moiety of the right kidney was not visible. VCUG performed six months later also showed resolution of vesicoureteral reflux. Prophylactic antibiotics were discontinued. The patient has since been followed on an annual basis. She is now four years old and continues to be well, with day and night-time continence, and without UTIs.

**Discussion**

We describe the case of an ectopic ureterocele detected on prenatal ultrasound that presented at birth as a perineal mass. Differential diagnosis included prolapsed ureterocele as well as cecoureterocele. The location of the urethral meatus anterior to the perineal mass excludes the possibility of prolapse. Our finding of a dumb-bell type ureterocele is, therefore, most consistent with a variation of a cecoureterocele, and has, to our knowledge, not been previously described.

Management of duplicated ectopic ureteroceles is complex, as they are associated with obstruction, primary and iatrogenic reflux, and an increased need for reconstructive surgery compared to single system ureteroceles and duplicated intravesical ureteroceles.3,5

Bladder outlet obstruction secondary to ureterocele is a major predisposition to infection, and should be relieved by incision of the ureterocele. In our case, incision of the perineal portion of the ureterocele was sufficient to relieve obstruction. Based on a review of the recent literature by Coplen, 70% of patients who undergo incision of an ectopic ureterocele require a subsequent open operation.1 However, it is the presence and severity of vesicoureteral reflux that drives the need for a second operation, and in our patient reflux resolved spontaneously. Decompression of an ectopic ureterocele in neonates may simplify later reconstruction.1 Evidence indicates that a lower tract approach, for those requiring secondary surgery, may suffice with minimal morbidity from the retained dysplastic upper pole.

**Conclusion**

We have described the presentation of a dumb-bell ureterocele associated with a duplicated system that presented prenatally as a perineal mass. The suburethral extension of the ureterocele, continuous with the intravesical and extravesical portions, distinguish this from a prolapsed ureterocele. The dumb-bell ureterocele is, in fact, a rare variation of a cecoureterocele, which, to our knowledge, has not been previously described. Treatment options include endoscopic incision, excision with ureteral reimplantation, and partial nephrectomy. Choice of treatment is driven by degree of obstruction and severity of vesicoureteral reflux of the urinary tract.

**References**

Two Cases of Cloacal Exstrophy without Prenatal Diagnosis

Blake Palmer MD1, Christopher Roth MD1, John Stanley MD2, Dominic Frimberger MD1, and Bradley Kropp MD1
1University of Oklahoma, Department of Urology, Section of Pediatric Urology, Oklahoma City, Oklahoma, 2Perinatal Center of Oklahoma, Mercy Hospital, Oklahoma City, Oklahoma

Introduction

Cloacal exstrophy is one of the most complex and rare congenital anomalies encountered by the pediatric urologist. The incidence is estimated at 1 in 200,000 to 400,000 live births1. This entity has been described as the most severe form of the epispadias-exstrophy complex, and as a part of the Omphalocele-Exstrophy-Imperforate anus-Sacral defects (OEIS) complex when described with its commonly associated anomalies.2 The etiology is felt to be heterogeneous2 though possibly develops as a result of premature rupture of the cloacal membrane.3 Descriptions of cloacal exstrophy have existed for hundreds of years but the first long-term survivor was not documented until 1960.4 Over the past few decades, management of cloacal exstrophy patients has advanced beyond survival to improving quality of life as the expected survival rates approach 100%.5

In 1998, Austin et al retrospectively reviewed prenatal ultrasounds of patients with cloacal exstrophy and established ultrasound criteria for the diagnosis of cloacal exstrophy in the prenatal period. The major criteria were nonvisualization of the fetal bladder, large midline infraumbilical anterior wall defect or cystic anterior wall structure, omphalocele, and myelomeningocele.1 These criteria were based upon the frequency of findings in their patients and reports in the literature. However, ten years from this description the reliable prenatal diagnosis of cloacal exstrophy remains challenging.6

Newborns with cloacal exstrophy require immediate evaluation and intervention by a multi-specialty team to properly manage this complex disorder optimally from a medical, social and ethical standpoint.

Materials and Methods

We identified 2 consecutive newborns identified in the postnatal period with cloacal exstrophy at our institution from March 2007 to July 2008. A retrospective chart review was completed to assess presentation, diagnosis and outcomes including review of prenatal ultrasound images.

Results

The diagnostic of cloacal exstrophy in the prenatal time period can be difficult due to its infrequent occurrence and variance in presentation.

Case 1. Baby G was followed during the prenatal period with serial ultrasounds secondary to identification of an anterior abdominal wall defect at a second trimester routine ultrasound. The defect was felt to be an omphalocele and no other significant anomalies were appreciated with a normal 3 vessel umbilical cord. The fetal bladder, however, was never identified.

After an uncomplicated delivery at 34 weeks the newborn exam revealed an omphalocele with an inverted perineal presentation of cloacal exstrophy with bilateral descended and palpable testicles in a well developed widely bifid scrotum on either side of the exstrophy defect. The exstrophy defect was inferior to the bifid corporeal bodies that were connected superiorly by paraexstrophy tissue in the midline. Postnatal workup revealed a tethered cord, unilateral grade 1 hydronephrosis and chromosomal analysis revealed 46 XY.
with grade 4 hydronephrosis, and the patient was identified to have a tethered cord. The baby also had unilateral cleft foot and polydactyl.

**Discussion**

Any prenatal ultrasound finding of an anterior abdominal wall defect, omphalocele, cystic or irregular mass should increase the suspicion for associated anomalies. This makes the visualization of the fetal bladder imperative. The spine, heart, head, and limbs should also be examined with closer attention as the possibility of an associated anomaly outside of the lower genitourinary system is also increased.7-9

In the case of Baby G the bladder was not visualized on prenatal ultrasound. An irregular anterior abdominal wall defect or cystic structure was identified as an omphalocele. After retrospective review of the prenatal ultrasound images the insertion of the umbilical cord is abnormal related to the omphalocele but there is a mass infraumbilical that although difficult even in retrospect can be identified to be independent of the umbilical insertion. Three of 4 major criteria set by Austin et al were identified but not fully appreciated in this case. The fetal bladder was not visualized in this case. The fetal bladder normally cycles every 50-155 minutes1. Diligence is required to correctly identify the fetal bladder in all cases of an anterior abdominal wall defect to rule out further anomalies, but as seen here correctly identifying the bladder can still be challenging.

In the case of Baby S, prenatal ultrasounds showed a large midline infraumbilical anterior wall defect. This was felt to be related to an omphalocele but in retrospect this mass was significant more complex, below and not associated with the umbilical cord insertion and more irregular than expected than with an isolated omphalocele. A hypoechoic structure was identified as the “bladder” on prenatal ultrasound although in retrospect this likely represented a hydronephrotic pelvic kidney. This was further complicated when the anterior abdominal mass was misidentified as a “scrotum” when the baby was viewed from a perineal angle. This also led to the misidentification of the fetus as male.

Both of these cases were identified to have an omphalocele prenatally. Omphalocele is a rare anterior wall defect with an incidence of approximately 1 in 5000 live births and 50% are associated with chromosomal abnormalities or as part of a syndrome.7 Cloacal extrophy patients have an associated omphalocele in 77-100% of cases.1,5 The recommended workup for an omphalocele diagnosis would involve an amniocentesis and prenatal karyotype determination7, although in both of these cases the parents declined.

The location of the anterior abdominal wall defect in relation to the umbilical cord insertion can be used to determine the likely etiology of the defect. If the defect is in the midline and the cord inserts into the defect the considerations should include omphalocele and Pen talogy of Cantrell. When the defect is lateral to the cord limb-body wall complex and amniotic band syndromes are of concern. If the defect is laterally displaced to the right of the defect gastrochisis should be considered. An infraumbilical defect is usually present with bladder or cloacal extrophy.9

In those patients with whom a definitive diagnosis of cloacal extrophy cannot be made by serial prenatal ultrasonography or in whom there is suspicion of significant anomalies further imaging may benefit the diagnostic process. There are a number of reports of fetal MRI used to diagnosis and depict the phenotype of genitourinary abnormalities, including bladder extrophy, cloacal defects, and OEIS complex.10-15 The reported benefits of fetal MR over ultrasound include greater spatial resolution, high T2-contrast between fluid and solid tissue, large field of view, and eliminating maternal bowel gas and pelvic bony structures that can obscure fetal anatomy.15 However, the use of fetal MRI has not been widely established or proven to benefit diagnosis and outcomes at this time for genitourinary anomalies. It is a potential area for further research.

Earlier diagnosis allows for accurate and thorough prenatal counseling with the proper specialists. Amniocentesis can be preformed to establish genetic sex prenatally, and delivery can be arranged at an institution capable of managing this complicated anomaly in an experienced, sensitive, and efficacious manner. The improved survival rates (continued on next page)
for newborns with cloacal extrophy are widely attributed to better neonatal management which is not available at every medical center. Immediate decisions and support psychologically, socially and ethically in terms of the challenging questions of gender assignment are best handled by multispecialty centers experienced in these difficult cases. The immediate surgical management of the difficult situation of ambiguous genitalia and gender assignment decisions in a timely fashion. That can limit the added psychological and social stress of the parents and family dealing with uncertain gender identity on top of the other multi-organ system anomalies. As outlined in Lund and Hendren, the importance of these patients being delivered and managed at a center with experience in this complex multi-specialty diagnosis cannot be dismissed.3

Conclusions
Although the prenatal diagnosis of such a rare congenital anomaly may be difficult, there should be multiple findings that increase the suspicion for other possible, more severe associated anomalies. Any prenatal ultrasound finding of an infra-umbilical anterior abdominal wall defect, omphalocele, cystic or irregular mass should increase suspicion, and visualization of the fetal bladder should be confirmed. Earlier diagnosis allows for accurate and thorough prenatal counseling with the proper specialists. Amniocentesis can be preformed to establish genetic sex prenatally, and delivery can be arranged at an institution capable of managing this complicated anomaly in an experienced, sensitive, and efficacious manner.

References

Ectopic Intestine in the Vulva of a Newborn with Complete Duplication of Bladder and Uterus
Heather McCaffrey, M.D.1, Alek Mishail MD.1, Robert Wasnick M.D.1, Moneer Hanna, M.D.2, 1 Division of Urology, SUNY at Stony Brook, Stony Brook, New York; 2New York Presbyterian-Cornell Medical Center, New York, New York

Introduction
Visceral sequestration is a rare condition in which intestinal segments are found in association with anterior abdominal wall and genital area. We report a case of a neonate presenting with a vulvar mass containing extra-abdominal colonic tissue associated with complete duplication of the bladder and uterus.

Case Report
A urological evaluation was requested for a newborn with a protruding vulvar mass. A 34 year old female (G2P2) underwent Caesarean section at 34 weeks due to suspected fetal ascites. Prenatal ultrasound evaluation had revealed no hydronephrosis or amniotic fluid abnormalities. Both parents were healthy and had no family history of congenital disorders.

Genital exam revealed a tubular soft tissue mass arising from the superior aspect of the vulva as well as a protruding vaginal membrane. The external genitalia included duplicated clitoris and a single vaginal opening on the right and a urethra on the left. The anterior abdominal wall, umbilicus, and anus appeared normal. Pelvic ultrasound and MRI revealed uterus didelphys with hydromucocolpos of the left uterine component, and complete bladder duplication without other congenital abnormalities. The patient underwent percutaneous vaginal drainage of the hydromucocolpos at 7 days of age. Cystoscopy and EUS revealed complete bladder and urethral duplication with grade IV reflux.
She underwent complete excision of the genital mass at one year of age. It was covered by exteriorized mucosa and extended 1 cm deep in the peripubic area without communication with the abdominal cavity. The mass was between the two urethras. Pathologic evaluation revealed well formed blind ending colonic tissue with fecal material in the lumen. We are waiting until the child is older to evaluate continence of the two bladder sphincteric mechanisms.

Discussion
Genital masses containing sequestered bowel segments are extremely rare. This anomaly has been associated with abnormal development of cloacal membrane and extrophy/epispadias complex. It has been suggested that colonic sequestration may result from hindgut duplication. If this is true, then our case represents a variation of a duplication anomaly involving the development of cloacal membrane and the hindgut. The finding of fecal material in the lumen suggests past continuity with the GI tract. The exact embryological mechanism for such anomaly is yet to be elucidated.
Management of Keloid Scar Formation in Two Stage Penoscrotal Hypospadias Repair

Thomas Forest, M.D.¹, Jason Romero, M.D.² and Robert Wasnick, M.D. F.A.A.P., F.A.C.S.³, ¹Pediatric Urology Clinic, Lafayette, Louisiana, ²Dermatology and Skin Surgery, Shreveport, Louisiana, ³Division of Urology, S.U.N.Y. at Stony Brook, New York

Introduction
Post operative keloid formation can be an unexpected disconcerting finding. We present management of an unusual keloid formation in a two stage hypospadias repair using a series of triamcinolone acetonide 10 mg/ml injections and dorsal inlay buccal mucosal grafting.

Case Report
A 4 year old patient with penoscrotal hypospadias, penile scrotal transposition and severe chordee underwent the first of a two-stage hypospadias repair. The patient missed several post-op appointments and presented eight months later with a thick keloid collar at the native meatus site which fixated and limited the availability of the surrounding skin and tissues (Figure 1). Simple excision was not an option because it would leave no available tissue for tubularization in the second stage repair. The patient was treated with a series of three injections of triamcinolone acetonide10 mg/ml at approximately one month intervals in order to soften and preserve the periurethral skin and tissue. The keloid softened allowing a Thiersch-Duplay closure in the second stage operation (Figure 2). Additionally buccal mucosa dorsal inlay grafting was placed as a prophylactic measure as keloid formation of the neourethra could potentially narrow and compromise the lumen. The patient did develop such keloids involving the neourethra which responded to a single triamcinolone injection. A fistula that had developed at the site of the original keloid collar failed initial simple closure, but was successfully closed with tunica vaginalis flap placement. At two years post op from his final operation patient is voiding well without difficulties.

Discussion
Keloids have been treated with triamcinolone, 5-FU, silicone gel dressings, interferons, imiquinod, radiation, laser and surgical excision.¹,² Triamciolone acetonide is usually given in strengths of 10mg/ml, 20mg/ml or 40 mg/ml. The higher concentrations are sometimes associated with permanent hypopigmentation or atrophy of the surrounding skin.³ The steroid is long acting and typically given 3 to 6 weeks apart. Triamcinolone injection was an effective treatment for keloid formation complicating this hypospadias repair. Buccal mucosa grafting should be considered in patients who develop keloid undergoing hypospadias repair.

References
PRELIMINARY PROGRAM (subject to change)

6:30 am  Registration and Exhibit Hall Opens

7:00  Introduction and Welcome  
John W. Brock, MD, SPU President

7:45 – 8:45  
**Challenges in Pediatric Urology: The Epidemic of Childhood Obesity**  
David Roth, MD, Moderator
The Role of Obesity in Surgical Complications  
Sean Corbett, MD
Evaluation and Treatment of the Undescended Testicle in Obese Males  
Michael Hsieh, MD
Penile Surgery in Obese Males  
Ross Decter, MD
Obesity and Pediatric Stone Disease  
Lars Cisek, MD

8:45 – 9:30  
**Genitourinary Development and Biomarkers of Disease**  
Kirstan Meldrum, MD, Moderator
Bladder Development: Where We Are and Where We Are Going  
Chester Koh, MD
Knock-Out Technology and Genes That Direct Genitourinary Development  
Si Oottamasathien, MD
Urinary Biomarkers of Bladder Dysfunction and Renal Disease  
Richard Lee, MD

9:30-10:00  Break in Exhibit Hall

10:00-10:45  
**Organ-Preserving Strategies For Pediatric GU Tumors-Spare The Rod And Spoil The Child?**  
Jonathan Ross, MD, Moderator
Testis-sparing Surgery for Pediatric Testis Tumors  
Rama Jayanthi, MD
Organ-Sparing Approach For GU Rhabdomyosarcoma - To What End And At What Price?  
Fernando Ferrer, MD
Limited Versus Expanded Role For Nephron-Sparing Surgery In Wilms' Tumor (Point/Counterpoint)  
Mike Ritchey, MD vs. Patrick McKenna, MD

10:45-11:30  
**State of the Art Lecture: Past Dreams and Future Realities: Adventures in Hypospace**  
Howard Landa, MD

11:30 – 1:00  Lunch (on own)

1:00 – 1:45  
**Infant Surgery: What is “Minimum” Invasiveness**  
John Thomas, Moderator
Transitioning To Robotic Surgery In Infants: An Open Surgeon’s Perspective  
Jack Elder, MD
Benefits of Laparoscopy and Robotic Surgery In Infants  
Pasquale Casale, MD
Minimally Invasive Open Surgery in Infants  
Martin Koyle, MD

1:45 – 2:30  
**Meredith Campbell Lecture: Burnout in Academic Medicine – From Students to Deans**  
Steven Gabbe, MD

2:30 – 3:00  Break

3:00 – 3:15  Presentation of Graduating Fellows
Presentation of SPU Research Awards

3:15-4:30  
**Updates from our Colleagues**  
Edwin Smith, Moderator
Radiologic Issues: Image Gently: Applications for Pediatric Uroradiology  
Kate Feinstein, MD
To Sleep, Perchance to Die: Anesthesia, Neurotoxicity and the Pediatric Brain  
Stephen Hays, MD

4:30 – 5:15  
**The Cold Hard Facts: What Pediatric Urologists Do ….And What Should We Do About It**  
Barry Kogan, MD
Program Director Perspective  
Rick Rink, MD
Manpower Perspective  
Doug Husmann, MD
New Practitioner/Recent Fellow Perspective  
John Makari, MD

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SOCIETY FOR FETAL UROLOGY YEAR IN REVIEW

This issue includes selected case reports and special presentations from the Spring and Fall 2008 Biannual Meetings of the Society for Fetal Urology.