SPECIAL EDITION: SOCIETY FOR FETAL UROLOGY YEAR IN REVIEW

The Society for Fetal Urology had another highly successful year in 2006. Our spring meeting was held in conjunction with the AUA Annual Meeting in Atlanta and the topic was Congenital Adrenal Hyperplasia: Prenatal Diagnosis and Treatment and Psychosexual Evaluation. This meeting brought to light not only the epidemiology and pathophysiology of this disorder, but the psychological issues facing patients, their families, and treating physicians. Our fall 2006 meeting, also held in Atlanta, took place in conjunction with the AAP Annual Conference, where the topic of ureteroceles was presented.

Spring 2007 Meeting

Our spring 2007 meeting, to be held in Anaheim, California, on Friday, May 18, in conjunction with the AUA Annual Meeting, will focus on antenatal MMC. Dr. John Brock and Dr. Katharine Wenstrom, both of Vanderbilt Medical Center, will present the latest on diagnostic modalities, current status of fetal surgery, and postnatal genitourinary and overall outcomes.

No pre-registration is needed for the spring 2007 meeting. Simply register at an AUA booth and present your AUA access card at the SFU meeting. Information regarding abstracts and further meeting details can be found on our website, www.fetalurology.org.

Antenatal Hydronephrosis Registry

The SFU Antenatal Hydronephrosis Registry is up and running with four participating centers that include Chicago, Oklahoma, Memphis and Alabama. Currently, several other institutions are pending IRB approval for their participation. Dr. Tony Herndon has secured funding for this database for a 5-year period. All documents required for IRB approval can be downloaded from the SFU web site. In addition, a complete grading system key for the SFU grading system can be found on the web site as well. For more information contact Dr. Herndon at anthony.herndon@ccc.uab.edu.

We are again proud to present our 2006 meeting presentations in the Dialogues in Pediatric Urology, and we thank Dr. Tony Caldamone and Lorraine O’Grady for their special assistance.

FROM THE EDITOR

Anthony A. Caldamone

The SFU continues to push new frontiers in pediatric urology. I had the opportunity to attend the fall meeting and what I found was a highly energized and enthusiastic group of pediatric urologists and other related sub-specialists. This Society provides a forum for the exchange of ideas not only among pediatric urologists, but across many disciplines which are engaged in the care of anomalies detected prenatally. This issue has an interesting array of presentations. When taken globally, however, it begs the question as to whether prenatal diagnosis alters the diseases that we would identify postnatally presenting symptomatically. The answer probably differs depending on the anomaly and over time the answers may change.

I congratulate the SFU on their continued enthusiasm and its ability to keep our specialty engaged in fetal medicine. I enjoy the continued association of the SFU with Dialogues in Pediatric Urology.
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Society for Fetal Urology
Spring 2007 38th Biannual Meeting
Friday May 18, 2007 / Anaheim Marriott / Anaheim, California

TOPIC: ANTENATAL MMC
Jeffrey B. Campbell, MD, Course Chair

GUEST SPEAKERS:
John W. Brock, III, MD
Professor of Pediatrics and Urology
Director of Pediatric Urology Fellowship
Vanderbilt University School of Medicine

Katharine D. Wenstrom, MD
Professor of Human Genetics and Obstetrics and Gynecology
Vanderbilt University School of Medicine

Visit the SFU Website www.fetalurology.org
Segmental Multicystic Dysplasia in a Horseshoe Kidney Mimicking Severe Contralateral Congenital Hydronephrosis

Juan C. Prieto, Miguel Castellan, Andrew Labbie, Rafael Gosalbez, Marcos-Perez-Brayfield
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and Department of Urology, University of Miami

Introduction

We present a unique case of segmental multicystic dysplasia in a horseshoe kidney diagnosed prenatally as right-sided hydronephrosis and left multicystic dysplastic kidney.

Case Report

A one-day-old male (37 weeks gestational age) was born to a 27-year-old female G1P1 (C-section). Serial prenatal sonography demonstrated bilateral hydronephrosis, renal cystic disease, distended bladder, and a normal volume of amniotic fluid. Physical examination performed immediately after birth revealed a healthy appearing male with an imperforate anus. A right upper quadrant mass was noted. Initial creatinine was 1.0 which subsequently decreased to 0.4 by one week of age. Initial renal and bladder ultrasonography revealed right moderate hydronephrosis with a presumed large extrarenal pelvis and hydroureter associated with a left multicystic dysplastic kidney (MCDK). A VCUG demonstrated a normal bladder contour and capacity without evidence of vesicoureteral reflux and a normal urethra. CT scan and MRI of the abdomen were consistent with moderate right-sided hydronephrosis associated with a left MCDK crossing the midline. Due to difficulties with feeding and right-sided hydronephrosis secondary to external mechanical compression, the patient was taken to the operative room for left nephrectomy. A left segmental multicystic dysplastic moiety of the horseshoe kidney was encountered. The former suggestive image of right extrarenal pelvis was a large cyst that was part of the left dysplastic kidney. A left nephrectomy was performed uneventfully.

Discussion

Horseshoe kidney is the most common fusion anomaly of the kidney. There are associated anomalies in approximately 25% to 50% of the cases. MCDK occurs in 1:4300 live births. The presence of a MCDK in a horseshoe kidney is an extremely rare event and only approximately eighteen cases have been reported in the literature. This case represents a challenge in terms of the diagnosis emphasizing the preponderance of the prenatal diagnosis in pediatric urology. The use of radiological tools such as CT scan and MRI of the abdomen should be considered when facing complex urological entities as in our particular case. Conservative management is the initial therapeutic approach in MCDK; however, most of the cases of MCDK associated with horseshoe kidney have undergone surgical correction.

References

Perinatal Diagnosis and Management of Congenital Mesoblastic Nephroma and Neuroblastoma in Neonates

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Introduction
Perinatally diagnosed pediatric tumors are rare and are always a concern for malignancy. We report 2 cases of pediatric tumors, one diagnosed prenatally and the other diagnosed postnatally.

Case Reports
Case 1: An 18-year-old prima gravida woman was found at 30 weeks’ gestation on prenatal ultrasound (US) to have polyhydraminos and a fetus with a right renal mass. MRI demonstrated a right renal mass measuring 8.5 x 7.2 x 6.6 cm and a contralateral normal kidney (Figure). The baby (2470g) was delivered via emergent C-section at 31 weeks due to PROM and preterm labor with normal APGAR scores. Physical exam was unremarkable except for a large right flank mass and hypertension. Postnatal US showed an increase in the size of the prenatally observed mass and a contralateral left grade II to III hydronephrosis. VCUG was negative. The patient underwent an uneventful and successful open radical nephrectomy one month after birth. Hypertension resolved postoperatively. Surgical pathology demonstrated mesoblastic nephroma.

Case 2: A 6-week-old male infant was diagnosed prenatally with a solitary right kidney in an otherwise unremarkable pregnancy and delivery. Physical examination on presentation was unremarkable. Postnatal US demonstrated a right solitary kidney with grade II to III hydronephrosis and a solid echogenic mass (3.3 x 2.6 x 2.9 cm) in the left renal fossa without evidence of medullary cortical differentiation. VCUG was negative. CT scan confirmed the finding and an uneventful open radical nephrectomy was performed. Surgical pathology showed neuroblastoma of the adrenal gland. Patient was referred to pediatric oncology. Hydronephrosis of the right kidney was resolved at 2 months’ follow-up.

Discussion
The perinatal finding of a retroperitoneal mass is always concerning for malignancy for physicians and parents. Imaging with US and MRI plays an important role in the clinical decision making regarding location and whether emergent removal is indicated.

Congenital mesoblastic nephroma (CMN) is the most common neonatal renal tumor. Prenatal diagnosis of CMN with US was previously reported but US features cannot reliably differentiate CMN from neonatal Wilms’ tumor. However, Wilms’ tumor is very rare in the first year of life and there are extremely few reports of confirmed prenatal Wilms’.

There is no particular MRI finding that confirms CMN but as Irsutti et al. showed, MRI can provide valuable information about the origin and nature of a fetal abdominal mass which will aid in management as well as counseling the parents. The MRI findings were suggestive of CMN, therefore, the decision was made to proceed with the pregnancy and delay emergent intervention. With complete resection of CMN, patients usually have excellent survival which was the case in our patient.

Neuroblastoma is the most common extracranial solid tumor in children and accounts for 8% to 10% of all cancers in children. Prenatal diagnosis of neuroblastoma has been reported but there is only one report of neuroblastoma occurring in the presence of a solitary kidney in a child. We report a case of a prenatally diagnosed solitary kidney with an empty contralateral renal fossa, which was found to contain a mass on postnatal follow up US. This illustrates the need for postnatal follow up to confirm an absent kidney and to rule out other congenital malformations of the GU tract as well as potential malignancies. Unlike CMN, neuroblastoma needs immediate surgical and medical treatment. In our case postnatal US identified a malignant tumor early that would have otherwise been delayed in diagnosis.

Conclusions
Prenatally diagnosed tumors although rare are increasing due to increasing prenatal ultrasound screenings. With a prenatal diagnosis of a mass, MRI can aid with delineating the relationship of the mass to the surrounding structures and sometimes the origin of the tumor. This can help in making the accurate diagnosis as well as counseling the future parents. Prenatal diagnosis of a solitary kidney should include careful examination of the contralateral side with appropriate imaging studies to confirm the diagnosis and rule out other malignancies.

References

Case 1 - Sequential axial MRI demonstrating mass originating from the right kidney.
Ambiguous Genitalia Presenting in a 7-Year-Old Child

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Introduction
We discuss a case of ambiguous genitalia presenting late in childhood.

Case Report
A 7-year-old phenotypic female presented to clinic with a complaint of clitoromegaly. Her birth history was unremarkable and she had no history of ambiguous genitalia. She was recently found to have clitoromegaly after immigrating to America from Eritrea, Africa, eight months prior. Any prior surgery was denied. Family history was unremarkable.

Physical examination revealed a moderate amount of isolated clitoromegaly, though the child was only moderately cooperative. The urethral meatus, hymenal ring, and anal position appeared normal. Her karyotype was 46,XX. Laboratory values and pelvic and adrenal ultrasonics were all normal. A pelvic MRI revealed an ovoid high T2 signal in a midline, phallic structure and a prepubertal vagina and uterus.

Evaluation under anesthesia revealed a soft, rubbery mass superior to a normal-sized clitoris palpable under a closed clitoral hood. Urethroscopy and vaginoscopy showed normal female anatomy. Clitoral mass exploration revealed an epidermal inclusion cyst with absent glans clitoris but intact corporal bodies.

Upon further questioning, the mother stated that the child had previously undergone a “female circumcision” (Type II- clitoridectomy) in Eritrea, which was denied preoperatively by both parents.

Discussion
“Female circumcision,” otherwise known as female genital mutilation or cutting, is a common practice exercised in Africa, the Middle East, and, rarely, Asia. An estimated 130 million women have been subjected to or are at risk of this, 130,000 of whom are in the United States. The World Health Organization (WHO) has 4 classifications, types I – IV. Immediate complications are due to bleeding or infection, which are most common immediately after the procedure. Long-term complications are usually due to type III procedures (infiltration-closure of the introitus) and include pain, chronic UTIs, infertility, sexual dysfunction, and obstetrical and fetal complications. Treatment is deinfibulation—genitoplasty to create a normal sized introitus.

Adrenal Hemorrhage in Newborns: Diagnosis, Parent Counseling and Management in Two Cases

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Introduction
Diagnosis of adrenal pathologies is becoming more frequent due to the increasing use of pre- and postnatal ultrasound examinations. We present two cases of adrenal hemorrhage in the newborn with conservative management through serial radiologic imaging.

Case Report
Case 1 presents a 3-month-old female diagnosed antenatally with a right upper quadrant cystic mass. Prenatal ultrasound at 34 weeks revealed a 2.7 x 2.8 cm cystic mass in the area of the right kidney. A repeat ultrasound was obtained at 38 weeks with a noted increase in the size of...
the mass measuring 3.9 x 4.8 cm, however, the mass was clearly identified as suprarenal in origin. The child was delivered without complication and physical exam, VCUG, and urine catecholamines were all unremarkable. A repeat US after birth revealed a stable 3.7 x 4.6 cm adrenal mass and at this time an adrenal hemorrhage was suspected. Serial ultrasounds were then obtained at 3, 4, and 8 months with noted involution of the mass on all imaging. A final ultrasound was obtained at 14 months and complete resolution of the adrenal mass was noted.

Case 2 presents a 2-month-old female who underwent sibling screening for reflux. The child’s parents reported normal development, and physical exam and VCUG were normal. The renal ultrasound revealed a right-sided mass with questionable origin of intrarenal versus suprarenal. A CT scan was obtained and a 4.8 x 3.1 cm mass was seen on the superior pole of the kidney. A partial nephrectomy was initially planned, however, a repeat CT was obtained at 3 months and the mass was suprarenal in origin and measured 3.0 x 1.7 cm. At this time adrenal hemorrhage was suspected with negative urine catecholamines. Serial US was again implemented with noted involution of the adrenal mass at 5, 7, and 10 months.

Discussion

The pathogenesis of adrenal hemorrhage in newborns is unknown, but is estimated to occur in 1-2% of healthy infants. Predisposing factors include: prolonged labor, birth trauma, large birth weight, asphyxia, sepsis, or a complication of a systemic coagulopathy. Radiographic assessment of a suspected adrenal hemorrhage with ultrasound is usually sufficient, but CT scan and MRI can be beneficial in the evaluation of an adrenal process. When dealing with a suprarenal mass a neuroblastoma is always a concern; however, negative urine catecholamines and involution of the mass on serial US confirms a benign process. Serial imaging is recommended for a suspected adrenal hemorrhage with surgical intervention reserved for cases of hemorrhagic shock or progressive processes.

These two cases illustrate antenatal and postnatal discovery of adrenal hemorrhage in newborns. Both demonstrate the importance of serial imaging to rule out a malignant process. With negative urine catecholamines and noted involution of the adrenal mass patients can be managed conservatively without surgical intervention.

References

With the Society for Fetal Urology Fall 2006 Annual Meeting program reviewing alternative treatment modalities for ureterocele surgical management, Drs. Tom Kolon and Ferd Ferrer thought that a survey of the membership would be interesting and, possibly, demonstrate a common thread in evaluation, medical and surgical management, and subsequent care of patients with ureteroceles.

To this end, a survey was developed with 13 questions primarily dealing with ureterocele management in those patients with antenatal diagnosis. Demographic questions were also included to formulate the geographic, age, and educational features of the members who participated in the survey.

The questionnaire was sent via e-mail to 247 urologists from the Society for Fetal Urology membership list and was in a SurveyMonkey format. Basic statistical analysis was performed using the SurveyMonkey program as well as ABstat (ver. 1.93 copyright, Anderson-Bell Corp., 1996). Responses were received from 132 members, about 53% of the membership. Due to technical difficulties on initial survey, a second survey was submitted to the membership and the two surveys were combined to achieve the 132 member response without duplicating the information received.

The demographics of the memberships showed that most of the respondents were between 40 and 50 years old (54 members or 41% of total). The 30-40 year and 50-60 year groups represented 35 and 32 members, respectively. Eighty-seven percent of the respondents were fellowship trained from 1980 to the present, representing 115 members. Geographic distribution using the American Urological Association Section demarcation had the participating members fairly equally representing all of the regions. Most responses came from members in the North Central (28), Southeastern (24) and Western (22) regions.

The first question queried understanding of the embryologic origin of ureteroceles and most of the membership (74%) felt that ureteroceles represented incomplete regression of Chwalle’s membrane. A smaller portion of the group (13 members) was unsure that the derivation of ureteroceles was fully known.

Use of antibiotic prophylaxis in patients with ureteroceles was asked in two questions: generally, when is it used and, if used, how long is it used. About 83% of respondents used antibiotic prophylaxis in all neonates with prenatally detected ureteroceles. Some members qualified their use of antibiotics as prophylaxis to only those with obstructed or refluxing segments, based on orthotopic or ectopic ureterocele location and in those with significant hydronephrosis or vesicoureteral reflux. Concerning duration of prophylaxis, 76 members, representing 58% of respondents, used antibiotic prophylaxis only until vesicoureteral reflux resolution. The responses for this question were varied from only until surgical correction (28 members) to 3 members who did not use prophylaxis at all.

Ultrasound imaging was used by all respondents in the initial evaluation of patients with ureteroceles, but only 97 members would use ultrasound after surgery. Most members obtained voiding cystography (VCUG) with the initial evaluation (130) and 108 members would also get a VCUG after surgical treatment. Most members would obtain an initial renal functional radiographic study, either nuclear renography with DTPA, MAG3 or DMSA as testing agents, or computerized tomography or magnetic resonance imaging. With 145 studies obtained by members, it appears that several members obtain more than one study on their patients. Postoperatively only 33 members obtain renal function tests.

Despite discussion of aggressive surgical management of ureteroceles in the literature, 40% of the membership believed in a “watchful waiting” approach in most patients. Generally, endoscopic treatment of ureteroceles, representing 41% of respondents, was utilized initially by those who treated children with antenatally diagnosed ureteroceles without infection or vesicoureteral reflux. Generally, the members responding did not feel that in utero or fetal puncture of ureteroceles was appropriate management of antenatally detected ureteroceles (54% believing that puncture should never be performed and 44% believing that in rare cases should antenatal puncture be performed.)

Controversy arose in the surgical management, timing, and procedures of choice. Although 53% of the members would wait until the child was 2-3 months old, 27% would perform some sort of surgical procedure “immediately” (less than or at 2 weeks of life). Procedure preference for those kidneys with poor or no function on an upper pole feeding a ureterocele was most controversial. Although 73% would remove the upper pole segment with either an open surgical (58) or laparoscopic (38) procedure, 27% or 36 members would not remove any segments, regardless of the function.

All members performed some type of endoscopic procedure (puncture or incision) but 58% admitted that they frequently had to perform some other surgical procedure for vesicoureteral reflux after the initial endoscopic procedure. Thirty-five percent rarely or never performed a second procedure. If a second procedure was needed for vesicoureteral reflux, 61% would perform ureteral reimplantation and ureterocele reconstruction but 25% would not perform any further surgery unless infection or high grade vesicoureteral reflux resulted after the endoscopic surgery. No questions concerning technique of ureterocele management were asked to better define its reconstruction at the time of initial survey.

In compiling the responses from the questionnaire and on written explanations of answers submitted, it was apparent that there is no consensus about surgical approach, if any is done, and prolonged prophylaxis use. Many of the written answers were thought provoking and stimulating enough to consider either a subsequent survey or an expanded survey of the membership, as there is significant debate as to appropriate management, or possibly over-management, of children with ureteroceles.
Watchful Waiting for Antenatally Detected Ureteroceles

Michael Leonard, Professor of Surgery and Pediatrics, University of Ottawa Chief of Pediatric Urology, Children's Hospital of Eastern Ontario

Currently, the majority of patients with duplex system ureteroceles are being detected by antenatal sonography.\textsuperscript{1} The natural history of ureteroceles detected in this manner is largely unknown, and may in some cases be innocuous. A paradigm which mimics this concept is well known to all pediatric urologists – that of antenatally detected pyelocaliectasis or indeterminate neonatal hydronephrosis (INH). Amongst patients with INH, approximately 25% will require surgery over 3-10 years follow-up.\textsuperscript{2,3} Only four studies in the English literature have examined the approach of watchful waiting for antenatally detected ureteroceles.\textsuperscript{4,7}

Data relating only to antenatally detected duplex system ectopic ureteroceles was extracted from these publications. In exception to this criterion, 3 patients who presented postnatally with UTI were included in the analysis, as they were not readily identified for exclusion. Observation was offered to patients who had either no upper moiety function or function with adequate drainage as determined by IVP or renal scan. The presence of ipsilateral lower pole vesicoureteral reflux (VUR) $\leq$ IV/V or contralateral VUR was not considered a contraindication to observation. However, the ureterocele could not cause contralateral renal and/or bladder outlet obstruction. This resulted in a total of 35 patients observed over a period of 1-12.8 years. All patients were placed on antibiotic prophylaxis until they were either toilet trained or VUR had resolved. Of patients on observation, 0-27% developed UTIs during follow-up. Breakthrough infections more commonly occurred in patients with concurrent VUR. These events usually heralded the need for surgical intervention. Resolution of hydronephrosis was seen in 43-67% of patients, improved in 0-43% and was stable in 14-57%. In only one patient was the hydronephrosis progressive, requiring surgical intervention. VUR resolved in 38-71% of cases, even reflux of high grade (IV/V). Contralateral VUR resolved in all cases.

In general, observation of patients with duplex system ureteroceles raises questions about safety. The data shown above should reassure most clinicians that complications are uncommon, and many patients do well. Those who fail due to breakthrough UTI or progressive hydronephrosis would be the potential for loss of relative renal function (RRF). It should be noted that none of the quoted studies addressed preservation of RRF in the upper pole moiety over time. However, we documented stability of upper pole parenchymal appearance by renal ultrasound in our small cohort.\textsuperscript{7} In fact, the amount of RRF at risk is minimal, given the severe irreversible histological changes seen in 70% of hemi-nephrectomy specimens.\textsuperscript{8} Moreover, when comparing the average change in RRF in patients undergoing upper pole sparing procedures (ureteropelvectomy) to upper pole hemi-nephrectomy, Vates found no difference in ipsilateral change in RRF.\textsuperscript{9}

Finally, in one series after upper pole heminephrectomy, there was a loss of RRF in 60% of cases.\textsuperscript{10} Clearly observation could be no worse.

What about the occurrence of hypertension in patients in whom a potentially dysplastic upper pole moiety is left in situ? It should be stated upfront that patients who have undergone ureterocoele incision without any further intervention would carry the same risk. Levy documented no increased risk for hypertension when comparing a group of patients who underwent upper pole salvage procedures vs. those who underwent heminephrectomy over a median follow-up of 15 years.\textsuperscript{11} Likewise, Gran et al. reported no incidents of hypertension over 62 months in a group of patients treated with lower tract reconstruction in the setting of duplex system ureteroceles.\textsuperscript{12} Thus, although follow-up is not over a lifetime, it seems the risk of hypertension caused by the dysplastic upper pole is insignificant. Additionally, would these dysplastic upper poles lead to increased risk of tumour formation? Although it has been documented that metanephric blastema can be present in upper pole hemi-nephrectomy specimens\textsuperscript{11}, to my knowledge no malignant tumours have been reported in upper pole moieties associated with ureteroceles. Finally, there have been sporadic reports of stones in duplex system ureteroceles.\textsuperscript{14} However, given that all patients selected for observation either have no function of the upper pole moiety or adequate drainage, and that most moieties show diminished hydronephrosis on follow-up, stone occurrence should be rare.

Aside from the criteria outlined above, there are ways we can better select patients for the option of observation? The only study which attempted to address this out of the four quoted was that of Han et al.\textsuperscript{4} In this study MAG3 ioxix renography was performed in all patients. Amongst the group who did well with observation (9/13), there were three with no function of the upper pole moiety. In the remaining 6/13, mean upper pole to lower pole moiety relative renal function was 40.8% and median upper pole $t_{1/2}$ was 5.3 minutes. When considering the group that failed observation (4/13) the mean upper pole to lower pole moiety relative renal function was 24.3% and the median upper pole $t_{1/2}$ was 12.5 minutes. Although these findings would suggest that diminished upper pole to lower pole moiety relative renal function and prolonged upper pole $t_{1/2}$ would be associated with failure on observation, the findings were not statistically significant. Given the small patient numbers, one would have to question the power of this study.

In summary, these findings suggest there is a role for observation in carefully selected and followed patients with antenatally detected duplex system ectopic ureteroceles. Prophylactic antibiotic use would seem prudent, especially in those with concurrent VUR. Indications for surgical intervention would comprise breakthrough UTI and/or increasing hydronephrosis. The tools to help us select which patients would be optimum candidates for observation have yet to be honed, but hopefully with co-operative multi-centre studies they may come to light. As with any non-interventional protocol in infants and children, long-term follow-up remains the onus of the urologist.

References

**Winner of BEST CASE PRESENTATION**

**A Cystic Sacrococcygeal Teratoma Mimicking Posterior Urethral Valves**

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**Introduction**

We present an interesting case of a cystic sacrococcygeal teratoma mimicking posterior urethral valves prenatally.

**Case Report**

Serial prenatal ultrasounds of a second trimester male fetus were felt to reveal steadily increasing bladder distention without hydronephrosis or diminution of amniotic fluid volume. At 30 weeks, bladder distention reached 7cm and new onset right grade II hydronephrosis was noted. Dilation of the inferior portion of the bladder was suspicious for a “keyhole sign.” Repeat ultrasound 2 weeks later revealed no changes. The infant was delivered at 33 weeks gestation due to spontaneous premature rupture of the membranes. On day one of life, a plain film revealed displacement of normal caliber bowel loops to the upper abdomen bilaterally with a ground-glass appearing soft-tissue density in the central and lower abdomen, consistent with hydronephrosis, bladder distension, and/or urinary ascites. Renal ultrasound revealed normal kidneys and an empty bladder which was displaced anteriorly by a large complex cystic mass extending from the lower sacrum to the upper abdomen. VCUG confirmed anterior displacement of a normal appearing bladder with a normal caliber urethra. An MRI revealed a 9x7x8cm cystic mass with a single septation arising from the posterior aspect of the coccyx, concerning for either a sacrococcygeal teratoma or an anterior meningocele. Exploratory laparotomy with coccygectomy several days later confirmed a type 4 cystic sacrococcygeal teratoma.

**Discussion**

This case highlights how a non-urologic entity can mimic posterior urethral valves in the prenatal setting. When presented with a large fluid-filled mass in the pelvis in the absence of oligohydramnios or marked hydronephrosis, cystic sacrococcygeal teratoma should be considered.

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**Bilateral Antenatal Hydronephrosis and a Dilated Bladder: A Postnatal Diagnostic Dilemma**

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**Introduction**

A fetus was diagnosed with moderate bilateral hydronephrosis and a dilated bladder. A tentative diagnosis of posterior urethral valves (PUV) was made. Throughout gestation, amniotic fluid levels were normal, but hydronephrosis worsened and hydroureter appeared later in pregnancy.

**Case Report**

The fetus was delivered at 39 weeks of gestation with good APGAR scores. The bladder contained 70cc of urine, and urine could be expelled with Crede maneuvers. A urethral catheter was placed, a circumcision was performed, and antibiotic prophylaxis was started. The patient had good urine output and his creatinine nadir was 0.6. Sonography showed improved hydronephrosis after urethral catheter placement. Initial VCUG showed bilateral vesicoureteral reflux (VUR) and a trabeculated bladder, but the posterior urethra could not be seen (Figure 1). A spinal ultrasound showed abnormalities, but spinal MRI was negative. Clean intermittent catheterization was started. Urodynamics, including VCUG performed on day of life 16 showed a normal bladder and posterior urethra, as well as bilateral grade 5 VUR.

**Discussion**

PUV is on the differential diagnosis for any male fetus with bilateral hydronephrosis and a dilated bladder. Other potential diagnoses include bilateral VUR, neurogenic bladder, and megacystis with megaureter. Inability to identify the posterior urethra antenatally on ultrasound and postnatally on VCUG can complicate making the diagnosis. False positive results from spinal sonography can further confuse the diagnosis.

**References**


*Figure 1 - Neonatal VCUG demonstrating bilateral VUR and a trabeculated bladder.*
Ten-Year-Old Female with Incontinence with Crossed-Fused Ectopic Kidney and Ectopic Ureter to the Vagina

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Introduction
Crossed fused renal ectopia is nearly always associated with a normally placed ureteral orifice located within the contralateral trigone.

Case Report
A 10-year-old female presented with constant urinary incontinence. At age 5, an evaluation including MRI, cystoscopy, and vaginoscopy revealed a solitary malrotated left kidney. At age 10 years, she underwent repeat cystoscopy and retrograde pyelography with the same result. A small orifice was noted on vaginoscopy in the right lateral wall of the vagina. Attempts to cannulate this were unsuccessful. A flush vaginogram demonstrated an ectopic ureteral insertion to the vagina (Figure 1). A gadolinium magnetic resonance urogram confirmed the presence of a crossed fused ectopic right kidney with some relative function and an ectopic ureteral insertion to the vagina (Figure 2). The patient subsequently underwent open extravasal ureteral reimplantation of the ectopic ureter and remains completely continent postoperatively.

Discussion
Only one case in the literature has reported solitary crossed renal ectopia and a single-system vaginal ectopic ureter, which was managed with nephroureterectomy.1 This case highlights the use of flush vaginography for performing retrograde pyelography in the setting of ectopic ureteral insertion. This case illustrates the superiority and utility of today’s MRU imaging in the evaluation of ectopic ureters and poorly functioning renal moieties.

References

Figure 1 - Retrograde vaginogram demonstrating an ectopic ureteral insertion.

Figure 2 – Gadolinium MR demonstrating crossed fused ectopic right kidney.

Unusual Delayed Presentation of Ureteropelvic Junction Obstruction in Patient with Prenatal Hydronephrosis and “Non-Obstructive” Postnatal Work-Up

Jeremy B. Wiygul, John S. Wiener
Division of Urology, Duke University Medical Center, Durham, North Carolina

Introduction
Antenatal hydronephrosis is a common problem faced by pediatric urologists; however, its natural history is poorly understood. We present a case of antenatal hydronephrosis with a non-obstructive postnatal evaluation presenting many years later as a traumatic renal pelvic rupture.

Case Report
A male fetus was diagnosed with prenatal bilateral hydronephrosis, and postnatal ultrasonography confirmed left hydroureteronephrosis. Voiding cystourethrogram revealed no reflux, and lasix renal scan showed equal function bilaterally with delayed but nonobstructive washout on the left. Sonography at nine months of age showed improving left hydronephrosis with good interval renal growth; however, the patient did not return for any further scheduled follow-up appointments. At 10 years of age, the patient presented with a 12-hour history of left flank pain associated with nausea, vomiting, and gross hematuria after being struck during a football game. Computed tomography of the abdomen revealed moderate to severe left pelviccaliectasis with possible renal pelvic rupture, and the patient was taken to surgery. Cystoscopy and retrograde pyelogram revealed a large filling defect in the left renal pelvis with extravasation. During flank exploration, a 6 cm rent in the renal pelvis was identified, along with ureteropelvic junction (UPJ) obstruction secondary to a crossing vessel. Repair of the pelvic tear and dismembered pyeloplasty were performed.

Discussion
Traumatic renal pelvic rupture is a rare clinical entity, with only a handful of case reports.1,2 It is thought that UPJ obstruction predisposes to this injury, due to increased shearing forces in the dilated renal pelvis.3 Interestingly, this patient presented with renal pelvic rupture as a delayed complication of prenatal hydronephrosis despite “non-obstructive” findings on postnatal workup. This suggests that the degree of obstruction in children is a dynamic process, necessitating long-term follow-up in these patients.

References
Familial Ureteral Anomalies: A Pedigree Analysis

Patrick Davol, Alice Cavanaugh, Larry Rothblum, Joel Sumfest
Geisinger Medical Center, Danville, Pennsylvania

Introduction

The genetic basis of renal and ureteral development is poorly understood, and few reports in the literature have identified inherited genitourinary anomalies. Possible modes of inheritance include autosomal dominant with variable penetrance (most common), X-linked recessive, polygenic, and multi-factorial. We report a familial syndrome of ureteral and renal anomalies found in a family spanning four generations, and present a plan for studying the genetic inheritance of these familial ureteral anomalies (FUA) using single-nucleotide polymorphism (SNP) analysis.

Case Study

Data were collected on a family originally identified when a patient in the fourth generation presented with a ureterocele. When further questioning identified a strong family history for congenital anomalies of the urinary tract (CAKUT), IRB approval was obtained for further study of the family. All available family members were questioned for a history of genitourinary anomalies, renal impairment, urinary tract infections or surgery. All consenting patients underwent screening renal ultrasound if no prior imaging work-up had been performed. DNA was collected via either a cheek swab or blood sample in anticipation of future SNP analysis.

Results

Forty-two family members spanning four generations were identified, including 27 family members who were alive and available for screening. Affected members with anomalies, including partial and complete ureteral duplication, renal agenesis, vesico-ureteral reflux, ureterocele, and UPJ obstruction were identified in each generation. A family tree is depicted in Figure 1.

Conclusion

Congenital, heterogenous anomalies were identified in generations of this family. These CAKUTs are consistent with the Mackie-Stephens hypothesis for abnormalities associated with development of the ureteral bud and metanephric blastema. An autosomal dominant mode of inheritance, possibly with variable penetrance has been previously proposed as a common mode of transmission for CAKUT abnormalities. However, the mode of inheritance in our report is not entirely consistent with this, as 6 of 6 available family members in the fourth generation are affected. With the help of information made available by the SNP consortium, we hope to further characterize the genetic basis and inheritance of the abnormalities observed in this family by performing an SNP analysis on DNA collected from consenting family members. Results from this analysis have the potential to further elucidate the genetic basis of these clinically significant developmental abnormalities.

References

Prenatally-diagnosed UPJ Obstruction with Unappreciated Ipsilateral Ectopic Ureter in a Functionally Solitary Kidney

Introduction

Ureteropelvic junction obstruction (UPJO) associated with a secondary distal obstruction is rare, and the presence of one may complicate the diagnosis of the other.1 We present a case of prenatally-detected UPJO with an unappreciated ipsilateral ectopic ureter in a solitary functioning dysplastic kidney.

Case Report

Bilateral hydronephrosis was detected on prenatal ultrasound (US) at 17 and 21 weeks in a 30-year-old primagravida. A male infant was delivered at 37.5 weeks.

US on day 6 demonstrated severe left hydronephrosis consistent with UPJO and bilateral renal dysplasia, worse on the right. VCUG was normal. Significant renal impairment was observed shortly after birth, and left pyeloplasty was performed on day 10. A 4 cm segment of stenotic proximal left ureter was found and resected at the time of pyeloplasty. Resistance was encountered with attempted passage of the stent into the bladder. A ureteral stent across the anastomosis and percutaneous nephrostomy tube were left in place. Though improvements were seen in creatinine, electrolytes, and acidosis, renal function was not fully recovered.

Postoperative nephrostogram demonstrated good result from the pyeloplasty and a previously-unappreciated ectopic left ureter inserting into the bulbous urethra (Figure 1). Cystoscopy revealed a left ureteral orifice just distal to the bladder neck, and retrograde pyelogram confirmed prior findings. Renal scan demonstrated 83% function on the left. The patient tolerated nephrostomy tube clamping without change in creatinine or hydronephrosis, and the tube was removed. At one year of age, the patient underwent left ureteral reimplantation. He did well postoperatively, but due to dysplasia, renal function deteriorated, and the patient ultimately underwent kidney transplant.

Discussion

UPJO is the most common cause of hydronephrosis detected by prenatal US.2,3 Additional ureteral abnormalities may be present in a small percentage of cases. This may be more likely to occur when renal dysplasia is present given the embryologic relationship between ureteral budding and metanephric differentiation.4 Diagnosis may be problematic when multiple ureteral anomalies occur simultaneously. While ureterography is not routinely performed in the evaluation of hydronephrosis, it should be obtained in the setting of renal dysplasia and UPJO, as complex ureteral anomalies may be more likely.

Figure 1 – Post-pyeloplasty nephrostogram demonstrating UPJ and ectopic insertion of the left ureter.

References


3-Month-Old Infant with True Hermaphroditism: One Ovary and One Torsed Testis

Introduction

Gender assignment in patients with intersex conditions is one of the most difficult scenarios pediatric urologists face. True hermaphroditism, the rarest type of ambiguous genitalia, poses a particular problem because of the presence of both ovarian and testicular tissue, and, hence, highly variable formation of internal and external genitalia. Gender assignment depends mainly on potential function (i.e., voiding, sexuality, fertility) and social considerations (e.g., patient/parent wishes). Not surprisingly, the literature reflects this complexity by supporting both male and female gender assignment in cases of true hermaphroditism.1,3 We present a case of a true hermaphrodite who was considered male due to prenatal exposure to testosterone but experienced a perinatal testicular torsion.

Case Report

A 3-month old infant was seen for ambiguous genitalia. At the community hospital, congenital adrenal hyperplasia was ruled out; testosterone shortly after birth was 150 ng/dL. Physical exam revealed penoscrotal hypospadias, a small penis, and penoscrotal transposition with non-pigmented labioscrotal folds. The left hemiscrotum was hypoplastic with a small, firm palpable gonad. The right hemiscrotum was also hypoplastic and a hernia with questionable gonad was palpable within the superficial inguinal pouch. A scrotal sonogram suggested a torsed left testis and right inguinal hernia. Müllerian structures were not seen sonographically.

(continued on next page)
The patient underwent endoscopy, diagnostic laparoscopy, right inguinal hernia repair, right gonad biopsy, and left orchiectomy. The right inguinal hernia contained a vas deferens, gonad, and müllarian structures, consistent with primitive ovary and hemi-uterus. Exploration of the left hemiscrotum demonstrated a nonviable testis that was subsequently removed. Repeat hormone profile at 5 months revealed a testosterone <10 ng/dL with elevated FSH (250 mIU/mL) and LH (34.3 mIU/mL). HCG stimulation resulted in no additional rise in serum testosterone. Additionally, karyotype revealed the patient to be 45XO, 46XY.

Endocrine, psychiatry, urology, and genetic counseling provided a gamut of suggestions ranging from observation to male or female gender assignment. To assist in the decision making process the child received short-term testosterone stimulation with consequent appropriate genital growth. This positive response to testosterone not only indicated an intact 5α-reductase/androgen receptor system but also male imprinting during fetal life; the child was assigned the male sex. In the near future, penoscrotal hypospadias repair, removal of the ovary and müllerian structures are planned. Testosterone replacement will be initiated at puberty age.

Discussion

Sex determination in patients with intersex conditions is very complex and generally accomplished by a multidisciplinary team approach. Early gender assignment is currently favored by parents and health care professionals alike. Growing evidence suggests that early hormone exposure may imprint the human brain and shape our sexual identity. This patient is a true hermaphrodite who was exposed to prenatal testosterone; however, the testis was torsed perinatally. With a functioning testis, this child would have been undoubtedly assigned the male sex. Short-term testosterone treatment helped in the decision making process by assessing genital growth in response to testosterone. This case poses a unique dilemma, which to our knowledge has not been previously reported.

References


Solitary Kidney Ectopic to Cloacal Anomaly

Blair St.Martin, Gordon Lees, Darcie A. Kiddoo, Stollery Children’s Hospital, Edmonton, Canada

Introduction

We present a unique case of a child born with a solitary kidney ectopic to a cloaca.

Case Report

A 37-year-old woman presented with an antenatal fetal ultrasound diagnosis of a solitary right kidney and hydroureteronephrosis. Initial physical exam after birth revealed a small dimple in the region of the meatus, a large opening in the region of the vagina, and a tiny fistula at its inferior aspect with no anal opening (Figure 1). Within the first day of life, a colostomy and mucous fistula were created. MRI revealed a tethered cord at L5, syringohydromyelia, and a solitary right hydroureteric kidney.

At 20 days of life, on cystoscopy through the single opening, there were no ureteric orifices in a small but relatively normal bladder. A blind ending pouch was found distal and to the left of the bladder neck. The right ureteric orifice was seen distal and to the right of the bladder neck. At 5 months of age, posterior sagittal anorectoplasty, closure of fistula and implantation of the ectopic ureter into bladder was performed. A renal scan at 7 months of age demonstrated uniform uptake in the single renal unit. At 8 months, during closure of colostomy, examination of pelvic structures identified ovaries, a rudimentary left uterine horn, and absence of a vagina or right uterine horn. Urodynamic studies at 10 months of age indicated reasonable bladder compliance, right vesicoureteral reflux, and capacity of 47cc.

Discussion

Solitary kidneys are well documented in the literature and renal anomalies are commonly seen with cloacas. This puts children with cloacal anomalies at increased risk for subsequent renal failure. To our knowledge there are no reports of a solitary kidney with an ectopic ureter to a cloaca. The spinal pathology also complicates the situation and increases the potential risks to the genitourinary tract. The hope for bladder development stimulated ureteral reimplantation in this patient, however, close follow-up is planned to ensure this does not result in renal deterioration.

References

A Single System Ectopic Ureter to the Rectum Subtending a Solitary Kidney and Bladder Agenesis in a Newborn Male

Christopher J. Weight1, Deepa Chand2, Jonathan H. Ross1
Glickman Urologic Institute1, Cleveland Clinic Foundation, Cleveland, Ohio
Section of Pediatric Nephrology2, The Childrens Hospital, Cleveland Clinic Foundation, Cleveland, Ohio

Introduction
Ectopic ureters are reported to occur in less than 1% of live births. In vanishingly rare cases, an ectopic ureter to the rectum has been described. Only 5 cases can be found in the literature1,2. Here we report the sixth case of ectopic ureter to the rectum and only the third diagnosed in a living neonate.

Bladder agenesis is also a rare entity and has been reported in fewer than 60 patients3,4; the vast majority of these patients are females with associated severe anomalies and in utero death. This case represents the nineteenth live birth with bladder agenesis and only the second case in a viable male.

Case Report
A newborn male was transferred on day of life four after spontaneous vaginal delivery at 36 weeks, from an outside hospital, with acute renal failure manifested by anuria. Apgar scores were 5 and 1 at one and five minutes, respectively, and he was intubated and placed in the neonatal ICU. Physical exam identified normal external male genitalia with bilateral descended testicles and anal stenosis. Creatinine was elevated (3.4 mg/dL), and abdominal ultrasound demonstrated a solitary dysplastic left kidney with no hydronephrosis. The bladder could not be identified. Attempts to place a urethral catheter were unsuccessful. The patient was having loose, watery stools and a creatinine measurement of the stool was 11 mg/dL, confirming urine output via the rectum.

Due to worsening renal function manifested by hyperkalemia, refractory acidosis and uremia, a peritoneal dialysis catheter was placed on day of life 10. Anal dilation and urethroscopy were performed at the same time. The meatus was dilated to accommodate a 7.5 French cystoscope. The distal urethra was normal in appearance until the junction of the bulbar and proximal penile urethra, at which point, it narrowed to a pinpoint. A 0.25 French guidewire could only be passed one millimeter beyond this point. Proctoscopy was performed, but no connection to the urinary tract could be discerned.

The patient had multiple other anomalies and characterization of the urinary tract could not be safely carried out until 3.5 months of age. At that time, ureter, MRI and open exploration failed to identify a bladder. The MRI with contrast confirmed a serpiginous ureter with focal areas of cystic dilation that coursed into the pelvis where it entered the anterior wall of the rectum (Figure 1). At the age of 7 months, the patient developed overwhelming line sepsis and expired. An autopsy was not performed due to family preference.

Discussion
The management of an ectopic ureter to the rectum and bladder agenesis depends, in part, on the associated anomalies. Most associated kidneys will ultimately be removed for dysplasia, hypertension or infection. Successful treatment and long term prognosis remains poor because of the associated abnormalities. Our experience suggests that, in the absence of hydronephrosis, the kidney may be left in place to facilitate fluid management during dialysis and/or to await a time when the kidney may be removed with lower anesthetic risk. The use of antibiotic prophylaxis for urinary tract infection in these cases seems prudent.

References
Imaging of Ureteroceles using Volumetric MR Urography

Paul Kokorowski, Hollie Jackson, Rex Moats, Lee Schiel, Dawn Berkeley, Theresa Dunway, Brian Hardy and Roger DeFilippo
Division of Urology and Department of Radiology, Children’s Hospital Los Angeles
Keck School of Medicine of the University of Southern California

Introduction

MRI is rapidly gaining popularity as an emerging imaging modality in pediatric urology. We present a case which highlights a novel imaging modality based on 3D volumetric acquisition and post-processing, which provides unprecedented detail when imaging ureteroceles.

Case Report

A 25-year-old female with intrauterine pregnancy at 28 weeks underwent prenatal ultrasonography demonstrating right greater than left hydronephrosis. Postnatal studies were performed including renal ultrasound and voiding cystourethrogram, confirming the diagnosis of a ureterocele involving the upper pole moiety of a duplex system without reflux.

MR urogram demonstrated the associated anatomy with thin parenchyma and markedly diminished to absent function. 3D volumetric study provided an instantaneous reconstruction of this anatomy with precise detail, regarding the insertion of the ureterocele and all surrounding structures. The patient underwent upper pole partial nephrectomy with resection of approximately 11 cm of a dilated ureter at four months of age. Pathologic examination revealed extensive fibrosis with chronic inflammation. Postoperative imaging demonstrated a marked reduction in the size of the ureterocele.

Discussion

Ureteroceles are commonly encountered during prenatal ultrasonography or work-up of urinary tract infection in children. Voiding cystourethrogram are commonly used to detect reflux often associated with these conditions. Some difficulty is often encountered when distinguishing ectopic ureters from ureteroceles.

Magnetic resonance imaging is quickly emerging as a powerful imaging tool in pediatric urology. Key benefits include the superior anatomic detail provided without the need for radiation exposure. While many protocols still require sedation in young children and infants, technological enhancements and improvements are allowing sedation-free acquisition as well. These benefits, along with the potential for functional assessment may allow MR urography the ability to replace multiple other studies in the future.

When imaging dilated ureters it is often difficult to clearly visualize their insertion into the bladder. Other investigators have determined that the ureterovesical junction is best visualized in 60 and 90 oblique planes. These views are commonly available with intravenous urography requiring nephrotoxic contrast agents and radiation or with maximal intensity projections (MIP) from T2 weighted MR reconstructions. In our case, we are able to demonstrate the anatomy in any conceivable orthogonal oblique plane without loss of detail or the need for radiation or MIP reconstruction. This is made possible through volumetric 3D acquisition and a novel viewer with near instantaneous conversion of data into an image that can be manipulated in every imaginable plane. Unlike MIPs, the T1 gadolinium enhanced 3D volumetric protocol allows assessment of surrounding structures and qualitative assessment of function.

References

Antenatally Detected Ureterocele Associated with a Segmental Multicystic Dysplastic Kidney

Andy Y. Chang, Thomas F. Kolon, and Douglas A. Canning
Children’s Hospital of Philadelphia, Division of Urology

Introduction
We describe the prenatal diagnosis and management of ureterocele and associated dysplastic renal segment.

Case Report
We diagnosed a female fetus of 27 weeks gestation with left upper pole hydronephrosis and a ureterocele on routine ultrasound. The fetus was followed with serial ultrasounds until birth. A postnatal renal bladder ultrasound showed a small ureterocele, no left upper pole hydroureter, and left upper pole dilation with little parenchyma. A voiding cystourethrograph (VCUG) showed a small filling defect in the bladder, but no vesicoureteral reflux (VUR). Prophylactic antibiotics were continued until a renal scan showed good drainage from both kidneys and no function to the left upper pole at 2 months of age. Serial ultrasounds every 6 months always had a stable, small ureterocele with eventual disappearance of the left upper pole dysplastic segment at 12 months of age. The most recent ultrasound indicated stable renal asymmetry of 9.1 cm (right) and 7.6 cm (left) with no parenchymal abnormality and a left ureterocele without ureterectasis.

Discussion
Upper pole segmental multicystic dysplastic kidney (MCDK) is a rare entity. Corrales and Elder reported a prevalence of 4% (3/68) in their series of patients with MCDK.1 Of the 13 segmental MCDK patients cited, 38.5% had associated ureteroceles.1 Coplen and Austin found 4 out of 8 MCDK patients to have duplicated systems with all MCDKs resolving spontaneously within 18 months.2 Furthermore, VUR is the most common associated urologic anomaly found in patients with MCDK.3 Thus, for our patient to have a ureterocele and no VUR with MCDK is very rare. With spontaneous MCDK involution, surgical intervention was never necessary.

To understand why this combination is so rare, we refer to lessons in embryology and genetics. First, complete ureteral duplication can be explained by the Weigert-Meyer rule or abnormal gene activity.3,4 Next, the existence of a ureterocele has multiple plausible theories, including persistent Chwalle’s membrane and abnormal ureteral muscular development.3 Finally, renal dysplasia maybe elucidated by Mackie-Stephens bud theory3, the work of Peters et al5 on early embryonic ureteral obstruction, or genetic abnormalities.4

Our patient highlights the natural history of segmental MCDK associated with an ipsilateral ureterocele. No surgery is required in this setting.

References
Errata

Due to a printing error in the December issue (Vol. 27, No. 6), several photos were incorrect.

On page 4, Figures 3 - 6, the arrows were omitted.

On page 6, Figures 2A and 2B, these are the correct images.

Figure 2A. Buried penis s/p prior circumcision.

Figure 2B. Post circumcision revision after 3, 6 and 9 o’clock fixation.

A corrected version is available on the website (www.spuonline.org) and can be downloaded and printed.
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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 2006 Biannual Meetings of the Society for Fetal Urology.