



# Society for Fetal Urology

## International Maternal/Fetal Organization

Volume 13, Number 1, Winter 2006

### Letter from the President

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Our 35<sup>th</sup> Biannual Meeting, held October 7, 2005, in Washington DC, was a great success. Once again, our attendance continued to grow, and we enjoyed the largest number of case presentations to date.

Dr. Mark Johnson, Children's Hospital of Philadelphia, presented "Prenatal Evaluation and Treatment for Fetal Lower Urinary Tract Obstruction." He noted when there is oligohydramnios and a need for fetal intervention, the renal evaluation may be assessed by serial bladder drainage with sequential fetal urine analysis. Poor prognosis would be indicated by sodium > 100, chloride > 90, osmolality > 200 or calcium > 8 mg/dl. Additionally,  $\alpha_2$  microglobulin > 10 mg/l or total protein > 40 mg/l without improvement of sequential urinalyses also suggest poor prognosis for renal function. Dr. Johnson reviewed the ultrasonographic appearance of the bladder after intervention. He noted that a large symmetrically thick-walled bladder was often consistent with urethral atresia, whereas an elongated-appearing bladder was consistent with posterior urethral valves. If there was a bilobed "snowman" type appearance to the bladder, this was more consistent with Triad syndrome.

Ideal placement of a vesicoamniotic shunt is between the umbilicus and pubis, which can be difficult to achieve because of the small distance between the two structures. One complication that may occur is dislodgement of the shunt by the fetus. Alternatively, there is a 1 mm fetal cystoscope available through which a diode laser may be used for valve ablation. The angle between the bladder neck and urethra does increase with gestation, so earlier intervention is technically easier to negotiate.

Dr. Craig Peters, Children's Hospital of Boston, presented "Fetal Bladder Outlet Obstruction: Translating Science to the Patient." He discussed reasons for post-obstructive bladder dysfunction, including hypertonicity as well as myogenic failure and hyperactivity. The major areas of investigation at this time include extracellular matrix homeostasis, smooth muscle activity function and dysfunction, smooth muscle cell growth regulation; and neurologic abnormalities. With fetal bladder obstruction, the bladder grows in response to the obstruction both in the smooth muscle component as well as the extracellular matrix component. There is no significant change in the ratios between the smooth muscle and extracellular matrix; however, collagen is increased. This is thought to potentially be due to a down-regulation of proteolysis as opposed to an increase in gene expression of collagen. With changes in the bladder wall occurring secondary to outlet obstruction, the bladder wall tension is increased which can subsequently lead to a vicious cycle with more pathologic changes taking place within the bladder wall, leading to further increase in wall tension. Sites of potential targets for therapeutic intervention include smooth muscle cells, the extracellular matrix, or the nerves.

Another region for investigation includes better prognostic markers. These may be urinary enzymes or proteins or be obtained by tissue sampling, such as cytology or biopsy. Current treatments may focus on changing the mechanical dynamics of the obstruction such as valve ablation, vesicostomy, clean intermittent catheterization, or overnight catheterization. Future treatments may include gene therapy. Currently, there are potential pharmacological treatments which are used such as anticholinergic agents, but other agents that may be useful include ace inhibitors, nitric oxide blockade, TIMP inhibitors (doxycycline), or blockade of growth factors. Advancement in treatment requires future translation of these scientifically-proven mechanisms into clinical trials and therapy.

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36<sup>th</sup> Biannual Meeting  
May 19, 2006  
1-5 p.m.

Georgia World  
Congress Center,  
Atlanta, GA

Meeting details inside, and  
at [www.fetalurology.org](http://www.fetalurology.org)

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The final lecture, by Dr. Michael Mitchell, University of Washington, was entitled, "The Changing Bladder of Early Childhood." He noted that development of the bladder, both in utero and after birth, was dependent on physical forces, emphasizing that everything responds to local environmental stress. The phases of normal bladder development were categorized as: 1) in utero, where there is a small bladder with high pressure; 2) first year of life, when there is a transition to an adult-type bladder; and 3) over one year of age, when there is a large, compliant bladder. In neonates, there is often a normal detrusor sphincter dyssynergia which leads to increased bladder capacity. Infants do empty their bladders, but not routinely after every void until they are at least three years of age.

Part of the changes in the bladder are related to the timing of the insult as well as the amount of pressure work the bladder is made to perform. Dr. Mitchell concluded that most bladder pathology in children is reversible. The key features of treatment should be to retain urine going through the bladder and get the bladder functioning and cycling as early as possible. Along these lines, epispadias should also be treated like exstrophy and managed around the time of birth.

## Secretary/Treasurer's Report

**Welcome New SFU Members:** Dr. Luis Baez-Trinidad (University of Puerto Rico), Dr. Seth Alpert (Columbus Children's Hospital, Ohio), Dr. Paul Noh (duPont Hospital for Children, DE), Dr. R. Guy Hudson (Oregon Health and Science University), Dr. Kazuyuki Nishinaka (University of Minnesota), and Dr. Osama AL-Omar (Wayne Staet University).

The **SFU Spring 2006 meeting** will be held May 19, 2006, at the Georgia World Congress Center, Atlanta, GA, in conjunction with the AUA Annual Meeting. The meeting topic is *Congenital Adrenal Hyperplasia: Prenatal Diagnosis and Treatment, and Psychosexual Development*. CME credits will be offered through the AUA. **Full details, registration and call for abstracts** for this full-day meeting are located on page 3, and on our web site, at [www.fetalurology.org](http://www.fetalurology.org).

The **Fall 2006 meeting** will be held in conjunction with the AAP Section on Urology Annual Meeting on Friday, October 6, in Atlanta, GA. The topic for this meeting will be Ureterocele. Dr. Tom Kolon will serve as Course Chair for this full-day meeting. CME credits will be offered. Registration information and call for abstracts will be posted at a later date on the web site and in the summer 2006 newsletter.

The inaugural issue of the SFU *Dialogues in Pediatric Urology* special issue has been completed. Copies of this issue will be emailed shortly to all SFU members, and print copies will be available at the spring meeting. Special thanks to Dr. Tony Caldamone for all of his work and support in creating this special SFU edition.

A vote was taken at the fall 2005 meeting for a **change in Bylaws**. Article III (Membership), Full Membership Requirements, Sections 4-5, had stated: *4) Members are expected to attend at least one of the two biannual meetings. 5) Continued membership in the Society will require each member to participate in either registration of new patient cases in ongoing multi-center studies, clinical case presentations, or presentation of ongoing laboratory research at biannual meetings.* By unanimous member vote, these sections have been removed from the Bylaws. A copy of the Bylaws can be found on the SFU website, at [www.fetalurology.org](http://www.fetalurology.org).

Dr. Mark Johnson, Children's Hospital of Philadelphia, is currently collaborating with maternal-fetal medicine specialists and pediatric urologists to establish the **North American Fetal Treatment Network**. This would consist of 50-70 centers and will require the Society for Fetal Medicine and other organizations to approve. If established, Dr. Tony Herndon will serve as SFU liaison. The SFU will continue its mission to improve communications with maternal-fetal medicine specialists.

## 36<sup>th</sup> Biannual Meeting Information

### Overview

The 36<sup>th</sup> Biannual Meeting will be held May 19, 2006, at the Georgia World Congress Center, Atlanta, GA, in conjunction with the AUA Annual Meeting. Check-in/registration begins at 12 p.m.; meeting 1—5 p.m. CME credits will be offered through the AUA.

Please check in at the AUA registration station to receive your AUA access card.

### **Program Topic: Congenital Adrenal Hyperplasia: Prenatal Diagnosis and Treatment, and Psychosexual Development**

**Intended Audience:** Pediatric urologists and allied health care professionals with an interest in the diagnosis and treatment of prenatal congenital adrenal hyperplasia.

**Prerequisites:** None.

**Statement of Need:** Pediatric urologists are frequently consulted for prenatal and perinatal cases to evaluate and manage ambiguous genitalia associated with congenital adrenal hyperplasia.

### **Educational Objectives**

Upon completion of this conference, the participant should be able to:

- Identify the diagnostic tests that need to be requested for pregnant women who may be carriers of patients with congenital adrenal hyperplasia.
- Interpret the results of prenatal intervention.
- Predict the outcome of prenatal intervention and analyze the data to be able to counsel parents in terms of possible outcome.
- Assess the prenatal diagnostic tests done for congenital adrenal hyperplasia
- Formulate and organize a plan for assessing parents or pregnant women who may harbor or carry patients with congenital adrenal hyperplasia.
- Recommend a form of diagnostic modality.
- Understand the different psychosexual assessment tests and the age at which intervention may need to be indicated.

### **Program Chair**

Walid A. Farhat, M.D., Hospital for Sick Children, Toronto

### **Invited Speakers**

#### **Kenneth J. Zucker, PhD, C Psych**

- Psychologist-in-Chief, Toronto Centre for Addiction and Mental Health (CAMH)
- Head, Gender Identity Service, Child, Youth and Family Program, CAMH

#### **Maria I. New, MD**

- Professor, Pediatrics and Human Genetics, Mt. Sinai
- Director, Adrenal Steroid Disorders Program, Mt. Sinai
- Founder, Weill/Cornell College of Medicine Children's Clinical Research Center
- Founder, Maria I. New Children's Hormone Foundation

## **Accreditation**

The American Urological Association Education and Research, Inc., is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to provide continuing medical education for physicians.

The American Urological Association Education and Research, Inc., takes responsibility for the content, quality, and scientific integrity of this CME activity.

## **Sponsorship**

This continuing medical education activity has been planned and implemented in accordance with the Essential Areas and Policies of the Accreditation Council for Continuing Medical Education (ACCME) and is sponsored by the American Urological Association Education and Research, Inc.

## **Credit Designation**

The American Urological Association designates this educational activity for a maximum of 3.5 *AMA PRA Category 1 Credit(s)*<sup>™</sup>. Physicians should only claim credit commensurate with the extent of their participation in the activity.

## **AUA Disclosure Policy**

As a provider accredited by the Accreditation Council for Continuing Medical Education (ACCME), the American Urological Association Education and Research, Inc., must insure balance, independence, objectivity and scientific rigor in all its activities.

All faculty participating in an educational activity provided by the American Urological Association Education and Research, Inc. are required to disclose to the audience any relevant financial relationships with any commercial interest to the provider. The intent of this disclosure is not to prevent faculty with relevant financial relationships from serving as faculty, but rather to provide members of the audience with information on which they can make their own judgments. The American Urological Association Education and Research, Inc. must resolve any conflicts of interest prior to the commencement of the educational activity. It remains for the audience to determine if the faculty's relationships may influence the educational content with regard to exposition or conclusion. When unlabeled or unapproved uses are discussed, these are also indicated.

## 36<sup>th</sup> Biannual Meeting Call for Abstracts

**Abstract submissions will be accepted online at [www.fetalurology.org](http://www.fetalurology.org).** Members are encouraged to submit interesting and unique case reports. Preference is given to cases related to the meeting topic, but all reports related to fetal and perinatal urologic topics will be considered. Case presentations should be no longer than 10 minutes. The guest speakers and program chair will judge the presentations and select the “Excellence and Innovation in Case Presentation” award.

Notifications will be sent via email to the corresponding author no later than May 1. Upon acceptance, the corresponding author will be sent instructions for preparation of the meeting presentation.

All abstracts accepted for presentation will be published as full case reports, along with our fall 2006 meeting reports, in a special supplemental issue of *Dialogues in Pediatric Urology* in early 2007. After the meeting, all presenters will be required to submit a full case report. Instructions for submission will be sent to presenters upon acceptance of their abstract.

Please follow all submission directions carefully. Provide a brief Introduction, Case Report, and brief Discussion. The abstract should total no more than 250 words, not including title and authors. Contact our administrative office with questions, by phone at 319-353-7871, or by email ([kristina-greiner@uiowa.edu](mailto:kristina-greiner@uiowa.edu)).

**Society for Fetal Urology Members are cordially invited to an after-meeting**

*Cocktail Reception*

sponsored by Q-Med Scandinavia

May 19, 2006

5—7 p.m.

Georgia World Congress Center, Room A316

**Antenatal Intervention for Bladder Neck Obstruction in a Twin Pregnancy.** Christopher M. Whelan, L. Scherba, Patrick H. McKenna, *Division of Urology, Southern Illinois University School of Medicine.*

*Winner of the Excellence and Innovation in Case Presentation Award.*

We report a prenatal intervention on a twin pregnancy with discordant lower urinary tract obstruction. A 19-year-old G3P0 was diagnosed with a twin pregnancy at 16 weeks. Ultrasound revealed two male fetuses in two sacs consistent with a diamniotic pregnancy. While no anomalies were identified in baby A, baby B was found to have severe oligohydramnios, mild hydronephrosis, and echogenic kidneys. Baby B's bladder was markedly enlarged with the dome reaching the diaphragm. At 20 weeks gestation, 92 ml of fluid was percutaneously obtained from baby B's bladder. The sample contained microalbumin of 3.3 mg/dL and sodium of 91 mmol/L. Chromosomal analysis revealed a normal male karyotype. At 23 weeks, the mother consented to percutaneous placement of a pigtail catheter into the bladder of baby B. Following this, the bladder never redeveloped significant urine. Follow-up ultrasound showed that membranes between baby A and B were divided during placement of the stent. Baby B was bathed in the fluid surrounding baby A, establishing a normal amniotic fluid volume. The pregnancy progressed until 33 weeks where an emergency cesarean was performed for pre-term labor. Baby B was delivered limp, dusky and bradycardic. He was intubated, and expired the following day from respiratory failure. Baby A was born healthy and had an unremarkable postnatal course. Autopsy revealed bilateral dysplastic kidneys with dilated ureters and bladder. Proximal urethral atresia was identified with the absence of posterior urethral valves. Examination of the lungs revealed pulmonary hypoplasia with diffuse alveolar damage.

**Postnatal Outcome: A Patient with Severe Prune-Belly Syndrome Treated with 32 Vesicocentesis and Amnioinfusion Procedures.** Vincenzo Galati, Stephen Confer, Dominic Frimberger, James Beeson, Bradley Kropp, *Department of Urology, University of Oklahoma, Division of Pediatric Urology, Children's Hospital of Oklahoma.*

Patients with category I prune-belly syndrome rarely survive beyond the first days of life. We present a case of prune-belly syndrome that initially presented with oligohydramnios and megacystis at 16 weeks of gestation. A follow-up ultrasound at 17 weeks, 2 days demonstrated poor prognostic findings indicative of impending fetal demise. Serial vesicocentesis procedures and subsequent analysis of the fetal urine revealed a normal male karyotype but poor urine electrolytes, and unfavorable  $\alpha$ -2 microglobulin levels. Fetal intervention was performed with 32 vesicocentesis and amnioinfusion procedures for obstructive uropathy and severe oligohydramnios. Despite the severe oligohydramnios, bilaterally dysplastic kidneys, poor urine electrolytes, and unfavorable  $\alpha$ -2 microglobulin levels, the birth resulted in a viable infant. To our knowledge this is the first reported case of severe prune-belly syndrome treated with numerous amnioinfusion and vesicocentesis procedures with a resultant favorable outcome.

**In Utero Spontaneous Intraperitoneal Bladder Rupture Presenting as Urinary Ascites in Two Newborn Females.** Leah P. McMann, Bruce H. Broecker, Francine D. Dykes, Andrew J. Kirsch, Edwin A. Smith, *Children's Healthcare of Atlanta.*

In utero urinary ascites is almost always due to posterior urethral valves. Spontaneous bladder rupture in a newborn female with a normal urinary tract is rare. We present two cases of female infants born at 34 and 35 weeks, respectively, with urinary ascites from intraperitoneal bladder rupture and a history of normal third trimester prenatal ultrasounds. Postnatal evaluation consisted of renal bladder ultrasounds and cystourethrograms, which were normal except for the bladder perforation. Both cases of intraperitoneal bladder rupture were treated with foley catheter drainage. There has been only one other report in the literature of a female infant with a normal urinary tract born with ascites from a spontaneous intraperitoneal bladder rupture.

**Antenatally Diagnosed Bilateral Ureteropelvic Junction Obstructions Presenting with Sudden Anuria.** Jenny Lassmann, Thomas F. Kolon, Stephen A. Zderic, *Children's Hospital of Philadelphia.*

We discuss a female neonate with antenatally diagnosed bilateral hydronephrosis and appropriate postnatal follow-up, who presented at six weeks of life with sudden onset of anuria secondary to bilateral ureteropelvic junction (UPJ) obstructions. Bilateral hydronephrosis was detected in one female neonate of a twin gestation at 20 weeks to a 34-year-old P2G1 mother. Hydronephrosis increased slightly during the course of pregnancy with maximal dilation to 18 mm on the right side and 9 mm on the left side. No oligohydramnion or bladder distension was noted. Upon delivery at 36 weeks the physical examination, blood urea nitrogen (BUN), creatinine (Cr), and a VCUG were normal. Postnatal ultrasound on day 3 revealed SFU grade 1 on the left side and grade 3-4 on the right side, consistent with moderate bilateral UPJ obstructions. A renal scan was scheduled and the infant was maintained on antibiotic prophylaxis. At 6 weeks of life she presented to the emergency treatment center at her local hospital with abdominal distention. Anuria was noted, and ultrasound confirmed worsening bilateral pelvicaliectasis. Cr and  $K^+$  were elevated to 2.0 mg/dL and 8mEq/L, respectively. Following transfer to our institution, emergent bilateral percutaneous nephrostomies were placed and no antegrade flow was observed. Repeat nephrostograms 24 hours after decompression of the system revealed antegrade flow of contrast across the narrowed UPJs. Urine excretion, Cr and  $K^+$  normalized over the following few days. Bilateral dismembered pyeloplasties were performed one week later with intraoperative verification of narrowed segments at both UPJs. After removal of the indwelling ureteral stents 6 weeks postoperatively, the patient remains asymptomatic with normal Cr and markedly improved ultrasound imaging 8 months after surgery.

**Surgical Management of a Left-Sided Duplicated Collecting System and Ureterocele Pre- and Postnatally.** Hadley Wood, Jonathan Ross, *Glickman Urological Institute, Cleveland Clinic Foundation,* Jack Elder, *Rainbow Babies and Children's Hospital.*

We present a case of bilateral obstruction due to a unilateral ureterocele associated with anhydramnios, successfully managed with in utero ureterocele puncture. A 28-week GA female fetus was noted to have a large left-sided ureterocele, hydronephrosis in both moieties of a duplicated right collecting system, and left sided hydronephrosis on routine US. Although US 10 days prior revealed normohydramnios, the fetus had progressively become oligo-, then anhydramiotic. The mother, a 32 year-old G1 opted for vesicocentesis. The procedure was tolerated well and normalization of the amnio-fetal index and flattening of the ureterocele was noted until 37 weeks, at which time the fetus was delivered vaginally without complication. US done immediately after delivery demonstrated left lower pole hydronephrosis, a decompressed ureterocele, and right hydronephrosis. The patient was immediately started on antibiotic prophylaxis and voided on the first day of life. US at 1 week of life demonstrated findings similar to 1 week prior. VCUG demonstrated grade V reflux into the lower pole moiety on the left and no reflux on the right. By 5 weeks of life, Cr= 0.6 mg/dl and a diuretic renal scan demonstrated left-sided differential function of 28% with a  $T_{1/2}$  = 39.5 minutes. The patient was followed with serum creatinine, renal ultrasounds and VCUGs for the next 18 months. She experienced no breakthrough infections during this period. Her right hydronephrosis slowly improved during this period suggesting that it was initially due to bladder outlet obstruction by the ureterocele. At age 19 months, a VCUG again demonstrated grade V lower pole reflux and a left kidney that was smaller in size when compared with prior. A diuretic renal scan demonstrated a differential function on the left of 26% with  $T_{1/2}$  of 6.3 and 6.1 minutes on the left and right, respectively. Given the failure of the reflux to improve, surgery was recommended. At 23 months, the patient underwent an uncomplicated excision of ureterocele and left ureteral reimplantation.

**Ureterocele Associated with Prolonged Inability to Void after Incision.** Heather Wallace, John Wiener, *University of Mississippi Medical Center.*

Fetal sonogram at 21 weeks gestation revealed left hydronephrosis in a male fetus with renal pelvis diameters of 21 mm on the left and 5.1 mm on the right and a single, thin septation within the bladder. Amniotic fluid volume was within normal limits. A presumed diagnosis of left ureteropelvic junction obstruction was made. Pediatric urology was not consulted. Initial post-natal ultrasound revealed severe left hydronephrosis, a normal right kidney, and a large ureterocele. VCUG confirmed the presence of a very large ureterocele. No spontaneous voiding was prompted after filling the bladder with 60 cc of contrast. Cystoscopy with incision of the ureterocele was performed on day of life 2. After the procedure, the patient still did not void spontaneously and CIC was initiated. Catheterized volumes of up to 210 cc were obtained. An ultrasound of the bladder confirmed almost complete decompression of the ureterocele. The neonate was discharged to home on CIC.

Initially, the patient voided only small amounts; however, over several weeks, voided volumes began to exceed catheterized volumes. At six weeks' follow-up, renal ultrasound showed dramatically improved left hydronephrosis with a small residual ureterocele. VCUG revealed that bladder capacity had returned to normal. CIC was discontinued since post-void volumes were <3 cc. The voiding dysfunction in this case appears to be secondary to a myogenic cause rather than obstruction, as improvement was gradual after the ureterocele was decompressed. Persistence of obstruction by the ureterocele would be a less likely explanation.

**Ureterocele Causing Bladder Outlet Obstruction in a Female Infant.** Adrienne J. K. Carmack, Andrew Labbie, *Miami Children's Hospital.*

We present a case of a female infant with prenatal hydronephrosis who was found postnatally to have bladder outlet obstruction secondary to a left upper pole ureterocele. Bladder outlet obstruction is extremely uncommon in female newborns and has only rarely been reported to result from a ureterocele. Our patient was followed prenatally for bilateral hydronephrosis and maintained a normal amniotic fluid index. Postnatally, she was found to have bilateral duplex systems, with a left upper pole ureterocele and right grade V lower pole vesicoureteral reflux. A voiding cystourethrogram performed on day of life 7 revealed a markedly trabeculated bladder with a ureterocele at the bladder neck. She underwent ureterocele puncture at 11 days of life with resolution of the bladder outlet obstruction. Because of breakthrough urinary tract infection with persistent severe reflux on the contralateral side, bilateral ureteral reimplantation with excision of the left ureterocele and right lower to upper end-to-side ureteroureterostomy was performed at age 7 1/2 months. Ureteroceles causing prenatal bladder outlet obstruction are rare and have been reported to cause significant urologic morbidity. However, no reports of pulmonary or other complications have been published. This raises the question of whether there is really a benefit to intervening on these patients prenatally. Prenatal intervention can have serious complications and should only be performed after thoughtful consideration and meeting the criteria of the SFU. Reports of long-term outcomes in patients treated both prenatally and postnatally are needed.

**Inguinal Bladder Herniation in a Child.** Scott Manatt, Faridali Ramji, Ann Kuhn, Jeffrey Campbell, Dominic Frimberger, *University of Oklahoma Health Sciences Center.*

Bladder herniation into the inguinal canal is a rare event, but as many as 4% of inguinal hernias may involve the bladder, usually in the form of a sliding hernia. It mostly occurs in men with a history of intravesical obstruction. The increased intravesical pressure causes bladder hypertrophy forcing the bladder through a weakness of the supporting musculature. Preoperative diagnosis using ultrasound or CT is possible, however bladder hernias are usually an incidental finding during inguinal hernia repair. While inguinal hernias are common in children there has been no report to date of a pediatric bladder herniation. We report about the initial presentation and management of a bladder hernia in a 6-month-old infant.

**Isolated Ascites in a Female Fetus with Congenital Adrenal Hyperplasia.** A. Zaccara, I. Capolupo, B. Iacobelli, M. Cappa, C. Corchia, P. Bagolan, *Bambino Gesù' Children's Hospital, Rome, Italy.*

A 36-year-old woman at 22 weeks gestation was found to have a female fetus with significant ascites. Amniotic fluid was normal and TORCH antibodies were negative. Fetal anatomy was unremarkable. Ascites was confirmed at 31 weeks g.a. with significant elevation of diaphragm, prompting elective C-section. A 2285 female newborn was delivered: initial examination revealed a massively distended abdomen which was compromising respiratory function and required mechanical ventilation. 2400 ml of ascitic fluid was drained from the abdominal cavity. A distended bladder became evident on day of life 2 with progressive elevation of serum creatinine levels. A Foley catheter was positioned without difficulty and US demonstrated bilateral hyperechoic kidneys with ureteral dilatation. Over the following days an incremental diuresis was observed with progressive normalization of SCr levels, resolution of ureteral dilatation and no ascites reaccumulation. Extubation occurred on day of life 20. An attempt to remove the bladder catheter failed since the bladder became again distended and the catheter had to be reinserted. A voiding cystogram was then performed which demonstrated a short (1 cm) urogenital sinus (UGS) with the catheter at the junction between bladder and vagina. UGS was confirmed with a cystoscopy. Vesicostomy was carried out during the same operative session.

Serum 17-OHP, testosterone and testosterone precursors were all elevated and the diagnosis of congenital adrenal hyperplasia was established. Urinary electrolytes were within normal limits. The baby is presently on corticosteroid treatment awaiting surgical reconstruction. These findings suggest that fetal ascites might be of urinary origin, with passage of urine from vagina into the peritoneum via the fallopian tubes. Even in the presence of normal fetal anatomy, persistent ascites may be the expression of UGS/cloacal disorders and such an issue should be incorporated into antenatal counselling.

**Cloacal Anomaly and Female Pseudohermaphroditism.** Paul A. Merguerian, Laurie Latchaw, *Dartmouth-Hitchcock Medical Center.*

A 27-year-old female G1P0 had a reported normal prenatal ultrasound at 18 weeks' gestation with female fetus. Due to hypertension and proteinuria, a repeat ultrasound was performed at 32 weeks' gestation. This showed enlarged cystic structures in the lower abdomen with bilateral pelvicaliectasis and oligohydramnios. An MRI was then performed that showed marked bilateral hydronephrosis with oligohydramnios and no structure that could be identified as bladder. There were multiple dilated cystic spaces filling the pelvis and extending into the abdomen. Fetal thorax appeared small. There was also maternal massively enlarged ovaries consistent with hyperreactio luteinalis. Due to maternal pre-eclampsia, labor was induced at 32 weeks' gestation. Ambiguous genitalia were noticed with enlarged phallus and pinpoint "urethral" meatus of the phallus. Labial folds were fused with imperforate cloaca. The abdomen was markedly distended with multiple masses palpated measuring 8-10 cm in diameter. The patient was intubated and could be easily ventilated. Chromosomal studies revealed 46XX. 17 hydroxyprogesterone levels were normal. The patient was taken to the operating room on day 1. Two large encapsulated urinomas were identified and drained. The patient was also found to have a distended, fluid-filled vagina and uterus. The bladder was also distended. A diverting transverse colostomy was performed, as was a vesicostomy. The patient is currently stable and awaiting final reconstruction.

**Severe Congenital Midureteral Dilatation.** Juan C. Prieto, Miguel Castellan, Andrew Labbie, Rafael Gosalbez, Marcos Perez-Brayfield, *Miami Children's Hospital and Jackson Memorial Hospital.*

A 3-week-old female was referred for evaluation of antenatally diagnosed left-sided hydronephrosis with a normal contralateral kidney without oligohydramnios. She was born prematurely at 35 weeks' gestation with Turner's syndrome. The initial renal and bladder ultrasound revealed a left-sided grade 3-4 hydronephrosis as well as a left abdominal mass of 5.25 x 4.75 cm containing fluid material. A VCUG demonstrated a right lateral displacement of the bladder secondary to the fluid mass, but no evidence of vesicoureteral reflux. A MAG-3 renal scan showed 39% function in the left kidney with a delayed excretion of the radiotracer. An MRI scan of the abdomen and pelvis showed left hydronephrosis and a large fluid-filled mass occupying almost the entire circumference of the lower abdomen. At 4.5 weeks of age, the patient was brought to the operating room for correction of what was presumed to be either a left ureteropelvic junction (UPJ) obstruction or a duplex collecting system with an upper pole ectopic ureter. A left retrograde pyelogram showed a normal distal ureter connected to what was believed to be a large left renal pelvis. A left flank incision was performed. A severe midureteral dilatation was found associated with minimal proximal ureteral dilatation and normal distal ureteral diameter. The dilated midureter was excised and an end-to-end ureteroureterostomy was performed. Pathological evaluation revealed ureter with mild focal chronic inflammation and muscular hypertrophy. Follow-up ultrasound at 6 months after surgery demonstrated improvement in the left-sided hydronephrosis and complete resolution of the hydroureter.

**Prenatal Diagnosis of Megacystis-Microcolon-Intestinal Hypoperistalsis Syndrome.** Nicholas M. Holmes, *Naval Medical Center, San Diego.*

Megacystis-Microcolon-Intestinal Hypoperistalsis Syndrome (MMIHS) is a rare congenital disorder causing a functional/anatomic obstruction of the urinary and gastrointestinal tract. Less than 100 cases have been reported in the medical literature. Two cases at Naval Medical Center San Diego were identified in 2000 (Baby A) and 2004 (Baby B) via prenatal ultrasound. Both fetuses were female and diagnosed with large distended bladders during the second trimester. Also, the fetuses were noted to have severe hydroureteronephrosis and normal amniotic fluid. Both were precipitously delivered via caesarean section at 30 (Baby A) and 32 (Baby B) weeks due to premature labor with fetal decelerations. Postnatal ultrasound verified dilation of the urinary tract and VCUG indicated no reflux. Each had persistent elevation of serum creatinine to 1.2 mg/dL (Baby A) and 1.1 mg/dL (Baby B) after the first 24 hours of life. Urethral catheter drainage was implemented immediately with eventual decrease in creatinine to 0.6 mg/dL (Baby A) and 0.3 mg/dL (Baby B) over the course of the first week of life. Baby A developed an acute abdomen on day 2 of life, requiring exploratory laparotomy, which indicated a pencil-thin colon with perforation of the stomach. A vesicostomy was performed at the time of laparotomy to manage the urinary tract. Baby B had severe cardiac and pulmonary anomalies, which delayed vesicostomy creation and enteral feedings. Once feedings were attempted, Baby B had recurrent emesis. Barium swallow with small bowel follow-through indicated a malrotation with markedly decreased peristalsis. Exploratory laparotomy was performed to correct the malrotation. The parents of Baby B did not desire vesicostomy creation. The functional bladder obstruction was managed with clean-intermittent catheterization and she has continued to remain infection free with normal serum creatinine with stable upper tract dilation. Each has required long-term intravenous parenteral nutrition and Baby A is awaiting bowel transplantation.

**Asynchronous Bilateral Renal Venous Thrombosis with Subsequent Development of Cystic Kidneys.** Andy Y. Chang, C. William Schwab, II, Bernard Kaplan, and Thomas F. Kolon, *Children's Hospital of Philadelphia, and the University of Pennsylvania.*

A 6-day-old, 34-week fraternal twin boy was delivered via cesarean section to a mother with a history of Group B streptococcal infection, pregnancy-induced hypertension, and Grave's disease. A left flank mass was palpated at birth and abdominal US findings were consistent with left renal venous thrombosis (RVT). Hematuria and thrombocytopenia of 43,000/cmm were subsequently observed. The serum creatinine concentration (SCr) decreased from 1.5 to 0.8 mg/dL with intravenous volume expansion. However, on the day of transfer to our institution, the infant had repeat gross hematuria and a new right flank mass. SCr was 2.3 mg/dL and platelet count was 62,000/cmm. A repeat ultrasound revealed bilateral renal venous thrombosis and a left adrenal hemorrhage. MRI showed multiple areas of intracerebral bleeding. SCr peaked at 4.7 mg/dL on day 8 of admission to our institution and began to normalize with intravenous fluids and supportive care. The patient's SCr nadir was 0.7 mg/dL at 8 months of age. His most recent estimated glomerular filtration rate was 67 ml/min/1.73m<sup>2</sup>. A renal ultrasound at 8 months of age showed bilateral corticomedullary cysts. He has since become hypertensive and is being treated with enalapril.

**Unexpected Pyoureteronephrosis in Neonates with Ureterocele.** Dennis Liu, Max Maizels, Christopher Talbot, Elizabeth Yerkes, Earl Cheng, Antonio Chaviano, William Kaplan, *Children's Memorial Hospital, Northwestern University.*

We present our recent experience with three cases of neonates with ureterocele who unexpectedly presented for emergency surgery with pyoureteronephrosis despite prophylactic antibiotics. The three cases consisted of female neonates with a median age of presentation of 16 days (range 14-30 days). One of three was diagnosed prenatally with ureterocele while in the other neonates, ureterocele was found on postnatal ultrasound. All three neonates had ureterocele associated with duplicated collecting systems. Two had unilateral right-sided duplicated systems while the last child had bilateral duplicated collecting systems and bilateral ureterocele. All three were taking prophylactic dose amoxicillin before ultrasound testing was completed. Two cases presented with fevers and negative urine cultures while the last presented with a positive urine culture without clinical symptoms. After ultrasounds showed pyoureteronephrosis, cystoscopy and transurethral incision of ureterocele were performed. All three patients were found to have extravesical ureterocele (submucosal extension into the urethra) and, upon incision, all drained grossly purulent urine. No complications were encountered during or after surgery, and all three were discharged home on postoperative day 1 with a therapeutic course of organism-specific antibiotics. As demonstrated in our cases, there is the inherent danger of neonatal sepsis with ureterocele treated expectantly despite prophylactic antibiotics. These cases reinforce that early surgery is still required in select patients. We hope this report serves as a foundation for further studies that will clarify which patients would benefit from early surgical intervention.

**Postnatal Outcomes: Megacystis with an Anterior Urethral Valve Identified by Prenatal Screening.** Stephen D. Confer, Vincenzo Galati, Bradley P. Kropp, *Children's Hospital of Oklahoma.*

Obstructive uropathy is readily identified by prenatal ultrasound as early as the 10<sup>th</sup> week. A case of prenatal bladder outlet obstruction due to an anterior urethral valve causing megacystis is reported. Neonatal imaging revealed dilation with normal amniotic fluid levels. Fetal intervention with vesicocentesis and amnioinfusion was not performed. VCUG performed on the neonate demonstrated a large bladder without reflux. Cystourethroscopy was done, revealing a diverticulum in the mid-penile urethra with a narrowing just distal, consistent with an anterior urethral valve. Incision of the anterior urethral valve was subsequently performed. We present what we believe to be the first case of megacystis coexisting with an anterior urethral valve accurately diagnosed prenatally.