Letter from the President

Our 34th Biannual Meeting, held May 20 in San Antonio, was a great success, with over 80 attendees. The topic was polycystic kidney disease. Program chair was Dr. Marcos Perez-Brayfield from the University of Miami Department of Urology. Guest speakers included Dr. Jack Elder from Rainbow Babies and Children’s Hospital in Cleveland, Ohio, and Dr. Lisa Guay-Woodford from the Division of Genetic and Translational Medicine at the University of Alabama-Birmingham. Lectures and case reports were well received and sparked lively debates.

In his lecture entitled “Multicystic Dysplastic Kidney: Much Ado About Nothing?,” Dr. Elder noted that the incidence of MCDK is 1:4,300 with the majority detected by antenatal sonography. About 2/3 involve the left kidney and about 2/3 are in males. Contralateral reflux occurs in 18-43% as well as contralateral ureteropelvic junction obstruction in 3-12%. Although inheritance is autosomal dominant in some families, most families do not show a definable inheritance pattern, and therefore screening is not recommended.

Initial evaluation should include history and physical, serum creatinine, renal ultrasound, VCU, and DMSA scan. Some in utero diagnoses of MCDK completely regress and result in the appearance of renal agenesis postnatally. Dr. Elder noted that a report to the AAP MCDK Registry demonstrated that at 5 years of follow-up, 37% of multicystic dysplastic kidneys had no change, with only 1.3% becoming larger. The remaining kidneys either became smaller or were not identifiable. Contralateral renal growth appears normal. The incidence of hypertension is 1:200 and the incidence of Wilms’ tumor in MCDK is estimated at 1:1600. Cyst progression was not associated with clinical outcome and screening for Wilms’ tumor appears to be effective only for early detection if an ultrasound was done less than every four months, and therefore is probably not helpful.

Dr. Elder recommends either nephrectomy at about six months of age or follow-up with a pediatrician for annual blood pressure assessment. One issue supporting nephrectomy over observation is that insurance companies are more likely to offer standard insurance rates to a person with an absent kidney as opposed to someone with a multicystic dysplastic kidney. Dr. Elder’s experience with outpatient nephrectomies done through a 2.5 cm incision began in the early 1990s. Most children were under 1 year of age and all kidneys were over 3 cm. He noted that the scar does grow with the child and he now recommends laparoscopic nephrectomy.

Dr. Lisa Guay-Woodford, presenting “ARPKD: Survival Myth, Genetic Insight, and Therapeutic Promise,” discussed the characteristics of autosomal recessive polycystic kidney disease, noting the incidence to be about 1:20,000. Most cases are detected in the perinatal period, though some don’t present until adulthood. Cysts occur in collecting ducts and tend to be 1-2 mm. The liver is invariably involved and there is an element of hepatic fibrosis. Mortality is high, approximating 30%, in the perinatal period. If a child lives past the first several months of life, however, they are likely to survive. Dr. Guay-Woodford stressed that this is not a uniformly fatal disease. Associated clinical problems include hypertension, hyponatraemia, chronic renal insufficiency, and portal hypertension, as well as liver and renal failure. Dr. Guay-Woodford noted that there is significant intrafamilial variation between siblings with the disease. The genetic defect most often detected is a mutation occurring on chromosome 6p-12 at the PKHD1 locus. She pointed out that there are about 500,000 base pairs at this locus, and multiple proteins are made, which makes genetic testing very difficult since 40% of mutations are single amino acid substitutions. Despite this, there is some genetic testing being performed at the University of Alabama and a few other centers.

Finally, we enjoyed case presentations by both current pediatric urology fellows and senior SFU members. The award for best case presentation went to Dr. Peter Metcalf, Fellow from Indiana University, for his case entitled “Prenatally Diagnosed Renal Cystic Mass.”
Welcome New SFU Members: Dr. Christian Radmayr (Innsbruck University, Austria), Dr. Abdurrahman Önen (Dicle University, Turkey), Goedele Maria Beckers (Vumc Amsterdam, The Netherlands), Dr. Louis J. Wojcik (Upstate Pediatric Urology, Greenville, SC), Dr. Jeffrey A. Campbell (University of Oklahoma).

The SFU Fall 2005 meeting will be held October 7, 2005, at the Four Points Sheraton Downtown, Washington, D.C., in conjunction with the AAP Section on Urology Annual Meeting. The meeting topic is In Utero Bladder Outlet Obstruction. This program is sponsored jointly by the Society for Fetal Urology and the University of Iowa Roy J. and Lucille A. Carver College of Medicine. CME credits will be offered.

Program Chair
Anthony Herndon, M.D., Assistant Professor, University of Alabama-Birmingham

Invited Speakers
Mark P. Johnson, M.D., Children’s Hospital of Philadelphia, University of Pennsylvania School of Medicine, Philadelphia, PA

Michael E. Mitchell, M.D., Children’s Hospital & Regional Medical Center, University of Washington, Seattle, WA

Craig A. Peters, M.D., Children’s Hospital Boston, Harvard Medical School, Boston MA

Full details, registration and call for abstracts for this full-day meeting are located on pages 5 and 6, and on our web site, at www.fetalurology.org.

PROPOSED AMENDMENT TO SFU BYLAWS: A vote will be held at the fall meeting to determine a change in the bylaws. Article III (Membership), Full Membership Requirements, Sections 4-5, currently state: 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 multicenter studies, clinical case presentations, or presentation of ongoing laboratory research at biannual meetings. The membership will be asked to vote for removal or continued inclusion of these membership requirements. Simple majority will determine the outcome.

The Spring 2006 meeting will be held in conjunction with the American Urological Association Annual Meeting on Friday, May 19, in Atlanta, Georgia. The topic for this meeting will be Congenital Adrenal Hyperplasia: Prenatal Diagnosis and Treatment, and Psychosexual Evaluation. Dr. Walid Farhat will serve as Course Chair for this half-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 winter 2006 newsletter.

Meeting case presentations will now be published in Dialogues in Pediatric Urology. The SFU has arranged with the Society for Pediatric Urology and the DPU for presentations to be published in a special annual supplement to DPU, as well as online publication. No registration or fee is required to access this journal online. The SFU President will serve as guest editor for the special annual issues.

Notice to Pediatric Urology Fellows: We need your contact information! We are currently working with the SPU to assemble and maintain a contact database of pediatric urology fellows. All current fellows are eligible for free membership in the SFU and free registration at all SFU biannual meetings. If you are currently a pediatric urology fellow, or are a faculty member overseeing a fellow, please send names, all contact information, and year of completion of fellowship to Kris Greiner, at kristina-greiner@uiowa.edu.

Renal glomerulocystic disease is an uncommon and sporadic disorder characterized by cystic dilation of Bowman’s capsule. This rare entity is more common in infants and young children, but to our knowledge has never been diagnosed antenatally. We present a case of a prenatally diagnosed renal mass, removed from a duplicated system. Pathology revealed a 16x10x6 cm mass, with microscopic cysts as per a typical glomerulocystic pattern.

Primary Obstructed Megaureter with Contralateral Cystic Dysplasia and Perinephric Fluid Collection. Adam G. Baseman, Leah P. McMann, Andrew J. Kirsch, Edwin A. Smith, *Children’s Healthcare of Atlanta*

A prenatal ultrasound revealed right-sided cystic dysplasia and left-sided hydroureteronephrosis in an otherwise normal-appearing male fetus. There was a normal amount of amniotic fluid and the mother had an uncomplicated delivery at term. The infant did well initially, with appropriate weight gain and development and was referred for evaluation at four weeks of age. A magnetic resonance urogram was obtained to delineate renal function and define ureteral anatomy. The right kidney was small and had poor corticomedullary differentiation with numerous cysts. There also appeared to be perinephric urinary extravasation on post-gadolinium imaging. The left kidney demonstrated hydroureteronephrosis with dilation to the level of the ureterovesical junction. The differential renal function was calculated at 61% on the left and 39% on the right. The infant was managed with percutaneous drainage of the left kidney with rapid return to baseline renal function. He has subsequently undergone tapered ureteral reimplant and has done well, without complications. His creatinine after stent removal remained at baseline.

An Atypical Presentation of a Ureterocele. Andy Chang, Stephen Zderic, Douglas Canning, *Children’s Hospital of Philadelphia*

We illustrate a case of antenatal hydronephrosis and oligohydramnios that eventually required renal transplant despite early, aggressive intervention. An infant girl was diagnosed with right hydronephrosis and oligohydramnios. Her mother’s fetal team placed a vesicoamniotic shunt at 21 weeks’ gestation. However, oligohydramnios recurred at 36 weeks’ gestation and prompted an induced vaginal delivery. Early postnatal ultrasound revealed multiple right renal cysts with poor corticomedullary differentiation, mild left renal pelvic fullness, but no ureterocele. VCUG showed a right-sided bladder diverticulum and grade V reflux. Renal scan noted a nonfunctioning right kidney and delayed left renal perfusion, with no left ureteral obstruction. VCUG was repeated at 11 months following a febrile UTI associated with acute urinary retention. Cystoscopy revealed a duplicated right collecting system with a large intermittently obstructing ectopic ureterocele, which was incised. Urinary retention recurred one month later. We then performed a right nephroureterectomy, excision of the ureterocele, reconstruction of the bladder neck, and left ureteroneocystostomy. Renal function eventually deteriorated, leading to a living-related renal transplant. Following transplant, two more febrile UTIs occurred. VCUG at that time showed grade III vesicoureteral reflux and we subsequently reimplanted the allograft ureter. The patient has had no further infections since reimplant.

Fetal Presentation of Denys-Drash Syndrome. James M. Elmore, Thomas L. Slabaugh, Edwin A. Smith, *Children’s Healthcare of Atlanta*

Denys-Drash syndrome (DDS) is a rare disorder consisting of the triad of congenital nephropathy, Wilms tumor, and intersex disorder, resulting from mutations in the Wilms tumor suppressor gene (WT1). We present an infant diagnosed with DDS after fetal ultrasound revealed oligohydramnios. The newborn’s physical examination was remarkable for ambiguous genitalia and bilaterally undescended testes. Laboratory evaluation revealed a 46XY karyotype and a creatinine of 1.3. A VCUG and renal sonogram were performed soon after birth. Following complete work-up, the infant was taken to the operating room for laparoscopic exploration and testis biopsies. Pathology was consisted with bilateral streak gonads. Renal biopsies were performed at the time of the gonadectomies and showed diffuse mesangial sclerosis. The infant quickly progressed to ESRD and on DOL 29 underwent bilateral nephrectomies. He currently remains on dialysis awaiting renal transplantation.
A 33-week gestation female was born with a right-sided palpable abdominal mass and developed hypertension, hematuria and thrombocytopenia. Abdominal CT scan showed a large infiltrating renal mass measuring 15 cm at largest diameter with poor contrast enhancement. The contralateral kidney was normal. CT images were thought to represent congenital mesoblastic nephroma. At 3 days of age the patient was taken to the operating room and a right nephrectomy was performed. Gross analysis suggested infarction of the kidney. Microscopic sectioning revealed extensive hemorrhagic necrosis. No neoplastic process could be identified. The main renal vein and intrarenal veins showed extensive thrombosis and the renal artery was found to have partial fibrointimal proliferation with luminal narrowing. Postoperatively, the patient developed a small bowel obstruction which was surgically explored. Malrotation was found as was a portal vein thrombosis. She was maintained on Lovenox anticoagulation for three months after her hospital stay. Work-up for hypercoaguable state was negative. She is currently over one year old and has done well, and though she has some clinical stigmata of neurofibromatosis, a definitive diagnosis has not been made. She is normotensive and renal ultrasound reveals a normal left kidney.

A one-month-old male with prune belly syndrome and patent urachus was admitted for renal insufficiency and a urinary tract infection. His mother reported that he occasionally dribbled urine out of his penis but that most drained from his urachus. A renal ultrasound and voiding cistouretrogram showed bilateral vesicoureteral reflux with severely dilated ureters bilaterally, as well as a dilated bladder neck, but a persistent narrowing of the urethra more consistent with posterior urethral valves. We subsequently performed cistouretroscopy and annular Young’s type 3 posterior urethral valves were encountered just distal to the vermontanum. The valves were ablated at the 12, 7, and 5 o’clock positions. Catheter drainage was maintained for 24 hours. After catheter removal, the patient was noted to drain most urine through his penis. At one month follow-up he continued to have a forceful urinary stream. Our patient’s survival beyond gestation is likely due to his patent urachus that acted as a pop-off valve and prevented fulminate renal failure. He is currently scheduled for bilateral laparoscopic orchiopexy and closure of his patent urachus.

A two-month-old child presented for evaluation of prenatal hydronephrosis. Imaging studies were consistent with a right duplex system with an obstructed, nonfunctioning upper pole and lower pole ureteropelvic junction obstruction. Removal of the upper pole and pyeloplasty were planned, but when the kidney was explored, we were surprised to find a single collecting system with a cystic, dysplastic upper pole segment. The rare diagnosis of a segmental multicystic dysplastic kidney with an ipsilateral ureteropelvic junction obstruction was made.

Over the past decade we have used the Kidney Internal Splint-Stent (KISS) catheter for internal/external urinary diversion after pyeloplasty. Between January 2001 and February 2005 there were 78 pediatric pyeloplasties in 38 children under 6 mos. of age, and done for UPJ obstruction employing a KISS catheter for postoperative drainage. The catheter has been recently modified by a manufactured update which accentuates the curl in the catheter. This facilitates hairpin turns at the kidney pelvis to exit the flank. Additionally, an “on the table” modification extends the trough proximally to better assure consistency in drainage. Both KISS catheter and drain are passed out of the flank using a 16-gauge angiocath as a pass-through rather than a stab incision. The catheter has served reliable drainage and is easily removed in the awake, non-sedated child in the office setting. In this age group there have been no untoward experiences. Empiric observations show the catheter minimizes flank urine drainage and shortens the postoperative interval needed to observe reduction in hydronephrosis.
Overview: The 35th Biannual Meeting will be held October 7, 2005 at the Four Points Sheraton Downtown, 1201 K St. NW, Washington, D.C. Check-in/registration begins at 7 a.m.; meeting 7:45 a.m.—4 p.m. This meeting is jointly sponsored by the Society for Fetal Urology and the University of Iowa Roy J. and Lucille A. Carver College of Medicine.

Intended Audience: Pediatric urologists and maternal/fetal medicine specialists with a particular interest in fetal urologic conditions.

Purpose: This conference will focus on the diagnosis and management of fetal bladder outlet obstruction. The latest information will be disseminated through lectures by world-renowned specialists and case presentations, about in utero bladder outlet obstruction, in order to further the knowledge by those practicing in the field of fetal urology.

Educational Objectives: Upon completion of this conference, the participant should be able to:
• Describe the current evaluation and treatment methods for fetal bladder outlet obstruction
• Discuss the common causes of fetal bladder outlet obstruction
• Explain normal and abnormal postnatal bladder development
• Describe potential outcomes of fetal bladder outlet obstruction

Registration Fee: The fee is $150 for physicians and allied health care professionals. Residents and Fellows in training will be allowed to attend free of charge. Fee includes registration, instruction, educational materials, refreshment breaks, and lunch. While registration is open until the start of the meeting, we encourage early registration to enable us to provide the best possible service to participants.

Accreditation: This activity has been planned and implemented in accordance with the Essential Areas and Policies of the Accreditation Council for Continuing Medical Education (ACCME) through the joint sponsorship of the University of Iowa Roy J. and Lucille A. Carver College of Medicine and The Society for Fetal Urology. The UI Carver College of Medicine is accredited by the ACCME to provide continuing medical education for physicians.

Continuing Medical Education Credit: The University of Iowa Carver College of Medicine designates this educational activity for a maximum of 6.25 category 1 credits toward the AMA Physician’s Recognition Award. Each physician should claim only those credits that he/she actually spent in the activity. All registered attendees will receive a certificate of participation.

Determination of educational content for this program and the selection of speakers are responsibilities of the program director. Firms providing financial support did not have input in these areas.

Guest Faculty
Program Chair
Anthony Herndon, M.D., Assistant Professor, University of Alabama-Birmingham

Invited Speakers
Mark P. Johnson, M.D., Children’s Hospital of Philadelphia, University of Pennsylvania School of Medicine, Philadelphia, PA
Michael E. Mitchell, M.D., Children’s Hospital & Regional Medical Center, University of Washington, Seattle, WA
Craig A. Peters, M.D., Children’s Hospital Boston, Harvard Medical School, Boston MA

University of Iowa Carver College of Medicine Faculty
Christopher S. Cooper, M.D., Associate Professor, Department of Urology, Society for Fetal Urology Secretary/Treasurer

Disclosure Policy: Everyone in a position to control the content of this educational activity will disclose to the CME provider and to attendees all relevant financial relationships with any commercial interest. They will also disclose if any pharmaceuticals or medical procedures and devices discussed are investigational or unapproved for use by the U.S. Food and Drug Administration (FDA).

Disability Statement: The University of Iowa prohibits discrimination in employment and in its educational programs and activities on the basis of race, national origin, color, creed, religion, sex, age, disability, veteran status, sexual orientation, gender identity, or associational preference. The University also affirms its commitment to providing equal opportunities and equal access to University facilities. For additional information on nondiscrimination policies, contact the Coordinator of Title IX, Section 504, and the ADA in the Office of Affirmative Action, (319) 335-0705 (voice) or (319) 335-0697 (text), The University of Iowa, 202 Jessup Hall, Iowa City, Iowa 52242-1316. Individuals with disabilities are encouraged to attend all University of Iowa sponsored events. If you are a person with a disability who requires an accommodation in order to participate in this program, please contact the Continuing Medical Education Division in advance at 319/335-8599.
Abstract submissions will be accepted online at www.fetalurology.org (click on “Submit Abstracts”). Notifications will be sent via email to the corresponding author no later than September 23. Upon acceptance, the corresponding author will be sent instructions for preparation of the presentation.

A disclosure form must accompany all abstract submissions. The disclosure form is available as a Word download on the “Submit Abstracts” web page.

Please FAX the completed and signed disclosure form to: 319-356-3900.

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.

Checklist

• Corresponding author full name, email, mailing address, phone and fax

• All coauthor full names and affiliations

• Abstract title

• Abstract body: must be no longer than 2,800 characters
  
  • Do not submit a full case report. Full case reports will be solicited after the meeting, to be submitted to Dialogues in Pediatric Urology for publication.

  • Abstracts are not structured, but authors should refrain from general statements such as “treatment options will be discussed.”

• Disclosure form
Society for Fetal Urology 35th Biannual Meeting
Friday, October 7, 2005
Check-in begins 7 a.m., Meeting 7:45 a.m.—4 p.m.
Four Points Sheraton Downtown, Washington, D.C.

Name _________________________________________________________
Last First MI

Address 1_______________________________________________________________________
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Email Address __________________________________________________________________

Registration Fee:
Physicians, Allied Health Care Professionals $150
Residents, Fellows $0

Cancellation Policy
Registrations must be cancelled by September 23. No refunds will be given after this date.

Method of Payment
I enclose a check made payable to Society for Fetal Urology in the amount of $ _____________

Please charge my: □ VISA □ MasterCard $ _____________ (total charge)

Account # ________________________________________________ Exp. Date _____________
Signature ___________________________________________________________________________

Four Ways to Register

Online
Go to www.fetalurology.org and click on “Register for Fall Meeting.”

By Mail
Mail form with payment information to:
Society for Fetal Urology
University of Iowa Department of Urology
200 Hawkins Drive, 3 RCP
Iowa City, IA 52242-1089

By Fax
Fax form to the SFU Administrative Office at: 319-356-3900 (for credit card payment and registration exempt attendees only).

By Phone
Please call the SFU Administrative Office at 319-353-7871 and have credit card information ready.
Special Invitation to Fellows!

American Academy of Pediatrics Section on Urology Fellows’ Luncheon

Special Presentations by
Alan B. Retik, MD, FAAP, and David H. Ewalt, MD, FAAP

“Finding a Job in the Academic or Private World”

Four Points Sheraton Downtown
11:30 a.m.—1:00 p.m.

Join us for our 35th Biannual Meeting
Friday, October 7, 2005
Four Points Sheraton Downtown, Washington, D.C.
Meeting details inside, and at www.fetalurology.org