



Society for Fetal Urology

International Maternal/Fetal Organization

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33rd Biannual Meeting
October 8, 2004
Hotel Nikko San Francisco
Meeting details inside, and
at www.fetalurology.org

Letter from the President

The spring meeting was a great success thanks to excellent lectures, case presentations, and audience participation. Dr. Peter Lee, pediatric endocrinologist from Penn State University College of Medicine gave a talk entitled "Epidemiology of Cryptorchidism" and Dr. Doug Hussman, pediatric urologist from the Mayo Clinic in Rochester presented "Cryptorchidism: Etiology, Neoplasia, Testicular Microlithiasis, and Post-Pubertal Presentation."

In his study, Dr. Lee noted a 93.2%, 89.7%, and 65.3% fertility rate among control patients, patients with unilateral undescended testicle (UDT), and those with bilateral UDT, respectively. There was also no statistically significant difference between men with a history of unilateral cryptorchidism and controls. A statistical difference with respect to paternity could not be found relative to preoperative testicular location or size. There was no difference between control and uni- or bilateral UDT with respect to FSH, LH, or testosterone, but the control group did have a significantly higher inhibin B level. Inhibin B levels may be more sensitive at detecting compromised testicular function than paternity data. As anticipated, those with higher FSH levels had lower sperm density and those with higher inhibin B levels had higher sperm density. The optimal timing of surgery was recommended at 6-9 months of age. Data supporting this includes lower inhibin B levels and higher FSH levels in children who undergo orchidopexy later in life as well as decreased testosterone levels.

Dr. Hussman's lecture described gubernacular outgrowth at 15-23 weeks gestation, which establishes the inguinal canal. Transinguinal descent occurs between 24 and 28 weeks. Failure of gubernacular outgrowth may result from lack of descendin/gubernaculotropin, or a receptor problem in the gubernaculum and lead to an undescended testicle. Other factors related to the etiology of UDT may include abnormalities in the hypothalamic-pituitary-testicular axis, blockade of the inguinal ring, and abnormalities of abdominal pressure or the spinal cord.

The lifetime risk of testicular cancer in the general U.S. population is 0.4%. There is an increased risk (4- to 7-fold or 3-5% lifetime risk) of testicular cancer with a UDT, assuming no genital or karyotypic abnormalities. Previous studies suggesting a 20-fold increased risk were flawed by inclusion of patients with abnormal karyotypes and ambiguous genitalia. Several studies suggest cancer risk normalizes if an orchidopexy is done before 2 years of age; however, other studies show no change.

There is a lack of data proving that orchidopexy leads to earlier cancer detection. In fact, 25% of patients with a history of UDT have metastases at the time of diagnosis—the same percentage as those with a non-pexed UDT. Orchidopexy also changes lymphatic drainage and 50% of those with metastases have inguinal metastases. Testicular biopsy is recommended in those undergoing orchidopexy over 11 years of age, or those with abnormal karyotypes, prune belly syndrome or proximal hypospadias/ambiguous genitalia. Five percent of these patients have carcinoma in situ which warrants removal.

Secretary/Treasurer's Report

The SFU administrative office is now located at the University of Iowa. We would like to welcome Ms. Kris Greiner as our Administrative Coordinator. Kris has worked as a medical editor for the University of Iowa Department of Urology for 13 years and has also worked with several urology organizations in an administrative capacity. She will be happy to assist SFU members with any inquiries regarding membership, meetings, publications, web site, and SFU business. You can reach her by phone, at 319/353-7871 or email, kristina-greiner@uiowa.edu.

New SFU members: We would like to welcome Hans Pohl and Walid Farhat, full membership, and Renato Ximenes, associate membership.

The SFU website has been updated! Please check it out at www.fetalurology.org. Special thanks to Tony Herndon, Web Site Administrator, and webmaster Jeff Keeton for their hard work on redesigning our web pages. If you have any suggestions for the site, please send your comments to Kris Greiner (kristina-greiner@uiowa.edu).

The **Fall 2004 meeting** will be held October 8, 2004, at the Hotel Nikko San Francisco (same site as the Spring 2004 meeting). Bob Nguyen and Jennifer Abidari will serve as Course Co-chairs. The topic for this meeting is Prenatal Hydronephrosis/UPJ Obstruction. Our guest speaker and panel moderator will be Stephen A. Koff, Chief of Urology at Columbus Children's Hospital, Columbus, Ohio. A large portion of this meeting will be interactive, utilizing an audience response system provided as an educational grant in the form of a gift-in-kind by Cook Medical. Registration information and call for abstracts for this full-day meeting are located on the last two pages of this newsletter, or on our web site, at www.fetalurology.org. Space will be reserved in a special section of *The Journal of Urology* for presentations. 5.75 AMA PRA category 1 credits are available for this meeting. *A cocktail reception sponsored by Q-Med Scandinavia will be held at Harry Denton's Starlight Room, located at the top of the Sir Francis Drake Hotel, following the meeting, from 6-8 p.m.*

The **Spring 2005 meeting** will be held in conjunction with the AUA, on Friday, May 20, in San Antonio, Texas. The topic for this meeting will be Cystic Disease of the Kidney. Marcos Perez-Brayfield will serve as Course Chair. Registration information and call for abstracts will be posted at a later date on the web site and in the winter 2005 newsletter.

Case reports presented at all meetings will now be published. Reports presented at the spring meetings will appear in *Urology* and those from the fall meetings will appear in *The Journal of Urology*.

IMPORTANT NOTICE TO MEMBERS

We are in the process of updating our membership files and creating an SFU listserve database for email announcements. Please email your complete, current, contact information, including email address, phone and fax numbers, and postal addresses to our Administrative Coordinator, Kris Greiner, at: kristina-greiner@uiowa.edu.

Crossed Fused Ectopic Multicystic Dysplastic Kidney with Associated Ureterocele

Darcie A. Kiddoo, Richard Bellah, Michael C. Carr, Children's Hospital of Philadelphia

Our report is an interesting case of an extremely rare congenital anomaly. It also demonstrates how the MRI is a noninvasive investigation which can provide an accurate urologic diagnosis in utero.

A 44-year-old G2, P1 Caucasian female presented after an antenatal ultrasound performed for advanced maternal age at 18 weeks gestational age revealed what was initially thought to be a left pelvic kidney with mild obstruction. The diagnosis, however, was uncertain and the patient was referred to the Center for Fetal Diagnosis and Treatment at the Children's Hospital of Philadelphia. A fetal echocardiogram, ultrasound and MRI were performed at 21 weeks gestational age, documenting a mid-thoracic vertebral anomaly—likely hemivertebra and a crossed fused ectopic multicystic dysplastic left kidney with normal amniotic fluid volume. The diagnosis was made based on the MRI. Ultrasounds were repeated at 32 weeks and postnatally with the same findings. A VCUG and renal scan performed at 2 months revealed a left ureterocele and normally functioning right kidney with a non-functioning left kidney. The child had been placed on amoxicillin prophylaxis at birth and a repeat ultrasound performed at 9 months of age documented a right kidney measuring 7.3 cm, a multicystic crossed fused ectopic kidney measuring 3.5 x 2.3 x 2.9 cm, and a left ureterocele. Follow-up ultrasounds have been scheduled to document involution of the left kidney.

Anterior Urethral Diverticulum and Non-Neurogenic Neurogenic Bladder

Marcos R. Perez-Brayfield and Andrew J. Kirsch, Children's Healthcare of Atlanta

We present a unique and challenging case of both non-neurogenic neurogenic bladder and anterior bladder diverticulum/valve complex and its possible treatment options.

A 2-year-old previously healthy male presented with a history of painless intermittent hematuria and several episodes of urinary retention up to 36 hours. The patient strains to void and refers mild discomfort after 36 hours of retention. He had experienced no difficulties with urinary tract infections or constipation. His physical exam showed a palpable bladder, but was otherwise normal. An ultrasound from 1 year previous to his current complaints showed no genitourinary abnormalities. Current workup with RBUS and voiding cystourethrogram showed an anterior urethral diverticulum/valve complex and left grade III reflux. Laboratory values, including creatinine, were negative. The patient continued with the episodes of urinary retention in spite of conservative management with alpha blockers and aggressive bowel management. Urodynamics evaluation revealed findings consistent with a non-neurogenic neurogenic bladder.

Vanishing Testes in Fraternal Twins Conceived by ICSI

Jennifer M. Abidari, Stanford University Medical Center Department of Urology

Cases of familial testicular torsion are rarely described and only 3 cases of testicular torsion among twins can be found in the literature. Fraternal twins presented at age 2 with a history of undescended left testis since birth. Conception had been achieved by ICSI and pregnancy was otherwise uncomplicated with vaginal delivery at 37 weeks. Family history was significant for a paternal nephew with a history of cryptorchidism and a grandmother with an unidentified pituitary disorder. Twin A had a history of a palpable left testis at birth which was not felt at 6 months of age. Twin B was noted to have an absent left testis at birth. At preoperative physical examination Twin B exhibited right testicular hypertrophy, but Twin A did not.

Diagnostic laparoscopy was performed and both twins were found to have markedly atretic vas and vessels proximal to a closed left inguinal ring, giving the appearance of an intra-abdominally vanished testis. Subsequent to laparoscopy, scrotal exploration was performed and an intra-scrotal testicular nubbin was found in both cases. In Twin A pathologic microscopic examination revealed fibrovascular tissue, vasal remnants and dystrophic calcification. In Twin B fibrovascular tissue and focal vasal remnants were identified. On follow-up examination, Twin A continued to have a normally sized testicle and Twin B continued to exhibit hypertrophy of the solitary right testicle. Whether ICSI might cause changes in the hormonal milieu of adjacent tissues, thus increasing the chance for a torsion event, is speculative at best and this case may represent an isolated event.

Melanotic Neuroectodermal Tumor of Infancy with Extratesticular Spread

Jennifer M. Abidari, Stanford University Medical Center Department of Urology

Melanotic neuroectodermal tumor of infancy is an uncommon tumor generally presenting in the maxilla of infants under the age of 1 year. The epididymis is its most common extra-cranial location. We report the findings in an infant with marked spread not only in the epididymis, but in the testis and surrounding soft tissue. A 7-month-old male presented with a firm, nontender nodular mass in the right hemi-scrotum that had been slowly enlarging over several months. The patient was an otherwise healthy infant delivered at term and physical exam was otherwise unremarkable. The inguinal region was uninvolved, but the right scrotal mass displaced the left testis superiorly. A scrotal ultrasound demonstrated a large right hydrocele and a heterogeneous, irregular mass in the testes and epididymis. A central area of lower echogenicity extended peripherally in the superior pole surrounded by a rim of normal echotexture. Blood flow was demonstrated in the testis. Serum alpha-fetoprotein level was 16.1 ng/ml (normal for age and sex) and quantitative β -hCG was <2.

The patient underwent right radical orchiectomy in the standard fashion. The specimen was bivalved in the operating room, revealing the inside of the tunica vaginalis to be peppered with pinhead-sized sooty spots, and for most of the testicular parenchyma and epididymis to be grossly replaced by a gray-black mass measuring 4 x 2 x 1 cm. Pathologic examination showed the tumor to be non-circumscribed and infiltrating both the epididymis and the testis with microscopic involvement of the tunica vaginalis, the surrounding soft tissue and non-striated dartos muscle. The spermatic cord and surrounding tissues were free of tumor. A biphasic cell population of small nests of primitive blue cells resembling neuroblasts and larger cells in a gland-like or tubular pattern with stippled intra-cytoplasmic melanin was present. The cells were embedded in a markedly desmoplastic stroma. CT scan showed no cranial, thoracic, abdominal or bony disease. One year postoperatively the patient was free of recurrence.

New Features of Prenatal Imaging of the Exstrophy Complex

Seth A. Alpert¹, John D. Edmondson¹, Max Maizels¹, Earl Y. Cheng¹, William E. Kaplan¹, Norman A. Ginsberg², Daniel W. Gauthier³, Christopher Chicoskie², Jacques S. Abramowicz⁴

¹Children's Memorial Hospital, Chicago, IL, ²Feinberg School of Medicine, Northwestern University, Chicago, IL, ³Hinsdale Hospital, Hinsdale, IL, ⁴University of Chicago Hospitals, Chicago, IL

We report two cases which show new aspects of fetal diagnosis of the exstrophy complex.

Case 1 A 36-year-old G5 P3 at 19 weeks gestational age (GA) was seen by her obstetrician for routine prenatal ultrasound. Her medical history was negative other than endometriosis. The ultrasound revealed a male fetus, multiple cysts of the umbilical cord and no visualization of the bladder. The pregnancy proceeded normally until 30 weeks GA when intrauterine fetal demise occurred. Previously, Doppler signals had revealed good vascular flow in the cord around the cysts. After delivery of the fetus, bladder exstrophy was confirmed on external exam. Kinking of the umbilical cord around the cysts was thought to be the cause of fetal demise. In this case, multiple umbilical cysts in a fetus with bladder exstrophy were thought to have caused intrauterine fetal demise from kinking of the cord. No previous cases in the literature have documented an association between umbilical cord cysts and bladder exstrophy.

Case 2 A 26-year-old G1 P0 underwent prenatal ultrasound at 16 weeks GA after successful in-vitro fertilization. A twin pregnancy was imaged. Twin A was visually normal; Twin B showed a large omphalocele and no bladder, suggesting a diagnosis of cloacal exstrophy. Level 2 ultrasound was performed at 25 weeks GA, but there was suboptimal visualization of the fetus due to the patient's obese habitus; therefore, an MRI was performed for serial imaging of the pregnancy. The MRI confirmed the findings of the earlier ultrasound; however, it did not change the overall outcome or clinical management in this case. Both babies were born by cesarean section at 37 weeks GA, and Twin B underwent successful surgical reconstruction on day 2 of life. Subsequently, Twin B had repair of a spinal myelocystocele at 4 months of age and both twins are presently thriving. In this case, MRI was used to confirm the diagnosis of cloacal exstrophy that had been suspected on a previous ultrasound; however, its use did not alter the diagnosis or management in this case.

33rd Biannual Meeting Information

Overview

The 33rd Biannual Meeting will be held October 8, 2004, at the Hotel Nikko San Francisco. This is the same venue as the Spring 2004 meeting. Registration begins at 8 a.m.; meeting 9 a.m. to 4 p.m.

This conference will focus on the management of prenatally detected hydronephrosis. It is estimated that approximately 70,000 newborns per year are found to have hydronephrosis detected during a prenatal ultrasound evaluation. Clinical experience indicates that in the majority of cases the hydronephrosis does not have any clinical significance. However, in approximately 10% of cases the hydronephrosis represents urinary obstruction or vesicoureteral reflux that can cause significant morbidity and mortality. It remains controversial as to what are the criteria to use in determining which patients to work up postnatally. In addition, there is no consensus as to how to work up these patients. We will present specific case scenario to highlight some principles in the management of prenatally detected hydronephrosis, and a combination of lectures from the experts in the field and audience interaction. At the completion of this activity, participants should be able to discuss guiding principles in the management of prenatally detected hydronephrosis and UPJ obstruction.

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 California, San Francisco School of Medicine (USCF) and the Society for Fetal Urology. UCSF is accredited by the ACCME to provide medical education for physicians.

USCF designates this educational activity for a maximum of 5.75 AMA Physician's Recognition Award category 1 credits. Each physician should claim only those credits that he/she actually spent in the activity.

Course Co-chairs/Speaker

Bob Nguyen, M.D., Assistant Professor of Surgery, Department of Urology, Harvard Medical School, and Jennifer Abidari, M.D., Assistant Professor of Urology, Stanford University School of Medicine, will serve as Course Co-chairs. Our guest speaker will be Stephen A. Koff, M.D., Chief of Urology at Columbus Children's Hospital, Columbus, Ohio.

Call for Abstracts

If you would like to submit an abstract for consideration for presentation at the meeting, please go to our web site, at www.fetalurology.org, and click on "Submit Abstracts." Feel free to contact our Administrative Coordinator, Kris Greiner, at kristina-greiner@uiowa.edu or (phone) 319/353-7871 if you encounter any difficulties using the online form.

Register to Attend

Online

Go to www.fetalurology.org and click on "Register for Fall Meeting." You will be taken to the online registration form within the UCSF Office of CME web site.

By Mail

Please fill in the registration form, enter the VISA/MasterCard information, or attach a check (payable to U.C. Regents), and mail to:

UCSF Office of CME
P.O. Box 45368
San Francisco, CA 95145-0368

By Fax

Please fill in the form, enter the VISA/MasterCard information, and fax to: 415/502-1795.

By Phone

Please call the UCSF Office of CME Registration at 415/476-5808 and have your VISA/MasterCard ready.

33rd Biannual Meeting Registration Form

University of California, San Francisco School of Medicine and the Society for Fetal Urology present



Society for Fetal Urology 33rd Biannual Fall Meeting

Course MMJ05002

October 8, 2004

Check-in begins 8 a.m., Meeting 9 a.m. — 4 p.m.
Hotel Nikko San Francisco, Golden Gate Room, 25th Floor



Name _____
Last First MI Degree

Date of Birth ____ / ____ / ____ Specialty _____
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Thank you

Please join us for a

Cocktail Reception

Friday, October 8

6-8 p.m.

Harry Denton's Starlight Room
(at the Sir Francis Drake Hotel)

*Special thanks to Q-Med Scandinavia
for sponsoring this event!*

Special thanks to

Cook Medical

*for providing an educational grant in
support of our 33rd Biannual Meeting.*

Society for Fetal Urology

Summer 2004

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Address Correction Requested

Join us for our 33rd Biannual Meeting
The Hotel Nikko San Francisco, October 8, 2004
Meeting details inside, or at www.fetalurology.org