FROM THE EDITOR

Anthony A. Caldamone, M.D.

Prenatally detected abnormalities continue to represent an increasing percentage of patients that we see in our offices as pediatric urologists. Technology and expertise have pushed the envelope beyond hydronephrosis and into other structural genitourinary abnormalities. This issue of the dialogues is composed of papers that were presented at the spring and fall meetings of the Society of Fetal Urology in 2007. You will be impressed by the vast array of congenital abnormalities that are discussed. Based on these excellent case reports, it is obvious that in most instances knowledge of the prenatal abnormality is helpful in planning a postnatal management strategy for these babies. Preparation of the parents, in particular, is a very important part of the prenatal assessment and plan.

The cases presented here represent an intellectually challenging spectrum of prenatal abnormalities. As an aside included in this issue is a point counter – point discussion between Hans Pohl and Earl Cheng regarding PIC cystography. This was moderated by Jack Elder at the fall meeting of the SFU. It represents one of the most controversial areas that we as pediatric urologists deal with on a regular basis.

My congratulations and thanks to the Society for Fetal Urology for once again providing an excellent scientific issue. I have enjoyed the association between the Dialogues in Pediatric Urology and the Society for Fetal Urology over the last three years.
Special Edition: Society for Fetal Urology - Year in Review

Index of Papers in this Issue

38th Annual Meeting, May 18, 2007, Anaheim

Prolapsing Ectopic Ureter Mimicking Posterior Urethral Valves .................................................. Page 3
Michael S. Gomez M.D., Dennis Liu M.D.
University of Toledo Department of Urology

Prenatally Diagnosed Horseshoe Kidney with Proximal Ureteral Stricture in the Absence of Ureteropelvic Junction Obstruction .................................................. Page 4
Pravin K. Rao, M.D., Jeffrey S. Palmer, M.D.
Glickman Urological and Kidney Institute

BEST PAPER - Anencephalic Renal Donor Transplantation to a Monoamniotic Monochorionic Twin with Bilateral Renal Agenesis .................................................. Page 4
Stephen D. Confer M.D., Nasir B. Chaudry M.D., E.N. Shea Samara M.D., Charles P. Mirabile M.D., Puneet Sindhwani M.D., E. N. Scott Samara M.D., FACS, Bradley P. Kropp M.D., FAAP
University of Oklahoma Health Sciences Center, Department of Urology, Section of Pediatric Urology, Children’s Hospital of Oklahoma

A Rare Case of Gartner's Duct Ectopic Ureter....... Page 7
Andy Y. Chang, M.D., Children’s Hospital Los Angeles Pasquale Casale, M.D., Alexander Kutikov, M.D.,
Douglas A. Canning, M.D., Children’s Hospital of Philadelphia

Fusiform Megalourethra and Oligohydramnios...... Page 8
Ranjiv Mathews, M.D., Amanda North, M.D., Jude Crino, M.D.
The Johns Hopkins Medical Institutions

39th Annual Meeting, October 26, 2007, San Francisco

PIC Cystogram Debate

Positional Instillation of Contrast Cystography (PICC): A Point-Counterpoint Debate? ......................... Page 9
Jack S. Elder, M.D.
Vattikuti Urology Institute, Children’s Hospital of Michigan

PIC Cystography: Is There a Role?...................... Page 9
Earl Y. Cheng, M.D., Associate Professor of Urology
Children’s Memorial Hospital,
The Feinberg School of Medicine at Northwestern University

PIC Cystography: An Argument for a Selective Approach to the Diagnosis of VUR ............................ Page 11
Hans G. Pohl, M.D., FAAP, Assistant Professor, Urology and Pediatrics, George Washington University, School of Medicine,
Children’s National Medical Center

An Unusual Fetal Presentation of an Abdominal Wall Defect in a Dichorionic-Diamniotic Twin ............... Page 12
William Tu, M.D., Ph.D., Lu Anne Dinglasan B.S.,
Jane Chueh M.D. William Kennedy M.D.
Departments of Urology and Obstetrics and Gynecology
Stanford University School of Medicine

Antenatal Imaging of the Ectopic Ureter Allows for Early Reconstructive Surgery and Maximizes Recoverable Function .................................................. Page 12
John G. Van Savage, M.D., FAAP, Regional Urology

Bladder Exstrophy Variants: An Individualized Approach .................................................. Page 13
Douglas B. Clayton, M.D., David M. Kitchens, M.D., David B. Joseph, M.D., C. D. Anthony Herndon, M.D.
Section of Pediatric Urology, University of Alabama at Birmingham

Management of Distal Ureteral Stricture by Refluxing Ureteral Reimplant ........................................ Page 14
Joshua J. Meeks M.D., Ph.D., Elizabeth B. Yerkes, M.D., Bruce W. Lindgren, M.D., Jennifer A. Hagerty, D.O.
Division of Urology, Children’s Memorial Hospital

Postpartum Hemorrhagic Renal Rupture in a Patient with Massive Prenatal Hydronephrosis ................ Page 15
Eric D Nelson M.D., Job K Chacko M.D., Philip Shlossman M.D.,
T. Ernesto Figueroa M.D., AI duPont Hospital for Children, Thomas Jefferson University, Christiana Care Health System

Midline Scrotal Mass in a Six Year-Old.................. Page 16
Kate H. Kraft, M.D., Wolfgang H. Cervinka, M.D., Andrew J. Kirsch, M.D.
Emory University Department of Urology

Collateral Urethral Duplication ............................ Page 16
Christina B. Ching, M.D., Jeffrey S. Palmer, M.D., FAAP, FACS
Glickman Urological and Kidney Institute
Cleveland Clinic Children’s Hospital

Congenital Megalourethra Associated with Bilateral Hydroureteronephrosis and Renal Dysplasia ........ Page 17
Dena L. Walsh M.D., John C. Thomas M.D., John C. Pope, IV M.D.,
Mark C. Adams M.D., John W. Brock, III M.D.
Division of Pediatric Urology, Department of Pediatric Urology,
Vanderbilt University Medical Center

Aphallia with Urethrocystic Fistula and End-Stage Renal Disease: A Case Report and Review of Literature .. Page 18
Heidi A. Stepnansy, M.D., John M. Gatti, M.D., Romano T. Demarco,
M.D., J. Patrick Murphy, M.D., Department of Surgery, Children’s Mercy Hospital and the Department of Urology, University of Kansas

Megalourethra, Urethral Stricture Disease, Urachal Diver-
ticulum, Bilateral Ureteropelvic Junction Obstruction,
Obstructive Megaureter and Absent Rectus Musculature: A Prune Belly Variant? ............................... Page 18
Douglas W. Sturm, M.D., Patrick E. Davol, M.D., Joel M. Sumfest, M.D.
Geisinger Medical Center

BEST PAPER - Cloacal Exstrophy in Two Separate Cases of Monoamniotic, Monochorionic Twins ........ Page 19
Sarah Marietti, M.D., Karen Driscoll, M.D., Jeffrey Spencer, M.D.,
Joshua Copel, M.D., Christine Kim, M.D., and Fernando Ferrer,
M.D. / University of Connecticut Health Center Departments of Urology and Maternal Fetal Medicine, Yale School of Medicine Department of Maternal Fetal Medicine, Connecticut Children’s Medical Center Department of Urology
Prolapsing Ectopic Ureter Mimicking Posterior Urethral Valves
Michael S. Gomez M.D., Dennis Liu M.D., University of Toledo Department of Urology, Toledo, Ohio

Introduction
Ultrasound findings of hydroureronephrosis and a thick-walled bladder in combination with VCUG findings of a dilated, elongated prostatic urethra with unilateral vesicoureteral reflux (VUR) are common findings in a neonate with posterior urethra valves (PUV). We present an unusual case of a boy with an ectopic prolapsing ureter whose radiographic findings mimicked PUV.

Case Report
A urologic consultation was obtained on a one-day-old male neonate with left antenatally detected hydronephrosis. Postnatal ultrasound showed SFU Grade 4 left hydronephrosis with hydroureter and a thick-walled bladder. A VCUG was obtained and demonstrated Grade 5 left VUR with an elongated, dilated prostatic urethra. A presumed diagnosis of PUV is made. Cystoscopy was performed and instead, a prolapsing ectopic left ureter entering the prostatic urethra was identified. The ureteral orifice was clearly seen in the prostatic urethra. While observing the ureteral orifice a membrane began to enlarge from the orifice and eventually obstruct the urethra. The membrane then decompressed and retracted back into the prostatic urethra relieving any obstruction. This membrane was subsequently incised endoscopically.

Discussion
This case demonstrates an unusual presentation of an ectopic ureter mimicking the findings of PUV. The peristalsis of the obstructed ureter likely led to intermittent ball-valving and obstruction of the prostatic urethra in utero. This mechanism likely accounts for the findings of a thick-walled bladder, unilateral VUR and hydronephrosis that are also commonly seen in the VURD variant of PUV. Due to the common pathophysiology involved, close long-term monitoring of this neonate for clinical sequela similar to PUV must be instituted.

In this case, we have a ureter which bypasses the bladder and enters the prostatic urethra directly. This was confirmed by our cystoscopic findings. Associated with this ureter is an overlying membrane which prolapsed causing subsequent obstruction.

References

Society for Fetal Urology - Year in Review
(continued from page one)

The intent of this statement would be to characterize our current position on “Prenatal Hydronephrosis” and identify future areas of research that may be addressed through recruitment of patients in the prenatal hydronephrosis registry.

Our meeting attendance and membership both continue to grow. As always we encourage suggestions and comments from our members concerning all Society for Fetal Urology issues. Recruitment of new members or participation in the biannual meetings is also encouraged.

See you in Orlando!

Christopher S. Cooper, President
C.D. Anthony Herndon, Secretary/Treasurer
Prenatally Diagnosed Horseshoe Kidney with Proximal Ureteral Stricture in the Absence of Ureteropelvic Junction Obstruction

Pravin K. Rao, M.D., Jeffrey S. Palmer, M.D., Glickman Urological and Kidney Institute, Cleveland, Ohio

Introduction

Horseshoe kidney (HSK) is associated with numerous genitourinary and extrarenal abnormalities. Associated urogenital abnormalities that cause prenatal hydronephrosis include vesicoureteral reflux (VUR) and ureteropelvic junction obstruction (UPJO). A case of prenatally detected HSK and hydronephrosis with unsuspected proximal ureteral stricture in the absence of UPJO or VUR is described.

Case Report

HSK and left hydronephrosis were detected during 18-week prenatal ultrasonogram. A full-term delivery was unremarkable, and antibiotic prophylaxis was started. Postnatal renal ultrasonogram at two weeks of age confirmed severe left hydronephrosis with cortical thinning in the absence of ureteral dilation, suggesting UPJO. A voiding cystourethrogram did not demonstrate VUR.

The patient was lost to follow-up until the age of 13 months, and repeat renal sonogram revealed findings similar to the previous study. MAG3 diuretic renal scan demonstrated a differential function of 52% and 48% of the right and left kidneys, respectively, with failure to clear contrast after administration of furosemide of the latter kidney. These two radiographic tests were consistent with a left UPJO and the patient was scheduled for a left open pyeloplasty.

At the time of surgery, a retrograde pyelogram showed proximal ureteral narrowing without distal obstruction. Intraoperatively, proximal ureteral obstruction with severe kinking and narrowing of the ureter was observed in the absence of UPJO. The region was not associated with the renal isthmus or a crossing vessel. The obstructed ureteral segment was excised, and a spatulated ureteroureterostomy was performed. Postoperative renal ultrasonogram was consistent with resolution of the obstruction.

Discussion

HSK has a strong association with VUR and UPJO, both of which can cause prenatal hydronephrosis. In this case, all preoperative studies suggested UPJO of a HSK, but the intraoperative retrograde pyelogram and dissection revealed proximal ureteral stricture in the absence of UPJO. This case also demonstrates the utility of a retrograde pyelogram, which is controversial in the setting of suspected UPJO, to identify ureteral anomalies not identified on preoperative testing.

References


Anencephalic Renal Donor Transplantation to a Monoamniotic Monochorionic Twin with Bilateral Renal Agenesis

Stephen D. Confer M.D., Nasir B. Chaudry M.D., E.N. Shea Samara M.D., Charles P. Mirabile M.D., Puneet Sindhwani M.D., E. N. Scott Samara M.D., FACS, Bradley P. Kropp M.D., FAAP, University of Oklahoma Health Sciences Center, Department of Urology, Section of Pediatric Urology, Children’s Hospital of Oklahoma, Oklahoma City, Oklahoma

Introduction

We present a case of monoamniotic monochorionic twins, one with bilateral renal agenesis and one with anencephaly identified at 21 weeks gestation. Both conditions are considered incompatible with life. There have been 6 reported cases of fetuses with bilateral agenesis surviving to birth. All 6 cases involved a monoamniotic twin providing normal amniotic fluid indices and, therefore, normal pulmonary development. No cases of survival beyond infancy have been reported due to complications of renal replacement therapies. In the case of an anencephalic twin with normal kidneys, an option for donor transplantation as this would be a perfect match and immunosuppression would not be required. However, the use of anencephalic infants as organ donors remains controversial. Prenatal management consisted of multidisciplinary counseling including multiple legal and ethical meetings regarding possible surgical intervention.

Case Report

A 23-year-old female G1PO with 23 week monoamniotic monochorionic twins is presented, one with bilateral renal agenesis and one with anencephaly by report of the 21 week ultrasound. Fetal weight was reported appropriate for gestational age and amniotic fluid indices were within normal limits. No other identification of syndromes is identified by multiple prenatal ultrasounds. Prenatal history is only remarkable for mother being a silent carrier for cystic fibrosis mutation 1898 + 16. Prenatal counseling occurred with parents requesting donor transplantation to child with bilateral renal agenesis. A multidisciplinary decision was made to proceed with the Cesarean section (C-section) followed by unilateral donor nephrectomy from the anencephalic twin ‘A’ with unilateral transplant into twin ‘B’. A lengthy (continued on next page)
discussion ensued with the parents about how this had never been done and the success of such a proposal is unknown. The parents understood associated alternatives, risks, and benefits and ethical/legal considerations were satisfied. Transplant, obstetrics, radiologic, and neonatal intensivist teams coordinated elective C-section at Children’s Hospital of Oklahoma. On the morning of C-section, a neonatal intensive care unit (NICU) bed, including appropriate personnel, was assembled in the recovery area. Radiologist and ultrasonography with technician was ready at time of C-section for evaluation of recipient. The C-section was performed in the Children’s hospital by the obstetric team.

Twin B was received with Apgar scores of 9 and 9 and a normal physical examination. After family visitation, complete postnatal ultrasound evaluation was performed, which revealed only bilateral renal agenesis without evidence of bladder. Retrograde urethrogram demonstrated urethral atresia. The scenario had already been discussed preoperatively, and the patient was prepared by anesthesia for transplantation.

Twin A was confirmed to have anencephaly upon C-section. Spontaneous breathing was poor and intubation was required. Additionally, a section of umbilical and placental vessels were harvested and placed in saline for repair of anticipated aortic defect. After family visitation, Twin A underwent living renal donation in a second operative suite. Upon dissection to the renal vessels, the donor had bilateral duplicated renal arteries (Figure 1). The left kidney was selected for length of vessels and removed without complication. A 5mm aortic cuff was harvested, and the placental vein patch was used to repair the aortic defect. The anencephalic twin survived the donor nephrectomy and was returned to his family for a peaceful, natural death after extubation.

Meanwhile, Figure 1: Bilateral duplication of renal arteries identified after dissection down to the hilum.

the kidney was transferred to a third operative suite. The transplant kidney was directly anastomosed to the recipient abdominal aorta and vena cava with demonstration of good perfusion. Upon surgical inspection, we were unable to identify a urinary bladder, therefore, a cutaneous ureterostomy was placed in the right lower quadrant and diverted with a 5F feeding tube. Peritoneal dialysis catheter was also placed at the time of abdominal closure. The procedure was well tolerated, and Twin B was transferred to the NICU.

The initial post-operative course of Twin B demonstrated acute tubular necrosis and peritoneal dialysis (PD) was required. Development of grade 1 hydronephrosis was noted on day 4 ultrasound with resultant stent manipulation and urine production of 4cc that day. Urine output of 1-2cc per kilogram per hour continued after day 5. Fluid shift delayed extubation until day 11. Post operative hypertension was managed with standard 2 drug therapy. This hypertension was most likely due to arterial anastomosis narrowing or the very small caliber of duplicated renal arteries with associated aortic patch (Figure 2). An ACE inhibitor was introduced for hypertension on day 11 to improve hypertension control. Urine production ceased and renal failure ensued. Ultrasonography revealed high pressure at the renal artery consistent with renal artery stenosis. Reintubation was required on day 15 due to fluid status and labored respirations with desaturation. Severe peripheral and pulmonary edema followed and, eventually, PD was re instituted. Pulmonary condition worsened requiring oscillatory ventilation on day 17. Urine output resumed on day 18 and normalized on day 21. PD was stopped on day 20. Cr fell to 0.5 by day 45 with resolution of peripheral edema. Pulmonary status did not recover despite antibiotics and multiple efforts at extubation by NICU team. The neonate died of pseudomonas pneumonia and overwhelming sepsis on day of life 56.

Discussion

Survival of Neonates with Bilateral Renal Agenesis

Klinger et al. reviewed 5 similar cases involving twins with bilateral renal agenesis, and all showed signs of VATER syndrome. All 5 subsequently died within the first 2 weeks of life. In each case, a twin provided normal amniotic fluid levels. Only 1 out of the 5 neonates received aggressive treatment in the form of hemodialysis. However, the neonate developed sepsis and expired.\(^1\)

In 2004 Perez-Brayfield et al. reported a case of monoamniotic twins: one presenting with bilateral renal agenesis with no associated VATER symptoms, and the other twin was found to be normal. CT scan of this child showed absent kidneys with a small, non-distended bladder. The child was managed with peritoneal dialysis and subsequently died 2 months later of peritoneal complications.\(^3\)

(continued on next page)
Anencephalic Donors - Data Review

With this current case a kidney transplant would be necessary for long term survivals, however, the ability to receive an isogenic renal graft would only be available for the first few days of life. The lifespan for an anencephalic infant varies from minutes to a few days depending on the level of brainstem function. The historic literature supports the use of ‘en boc’ kidneys from anencephalic donors as a viable source for renal transplant. A case review by Gomez-Campdera et al. done in 1990 showed a 12 month graft survival rate of 60%. These grafts, however, were transplanted in recipients whose ages ranged from 8 to 50 years. There are no published records of a neonate surviving an anencephalic kidney A transplant.

Anencephalic Donors - Ethical and Legal

Historic discussions and publications document the need for lifesaving organ donations from anencephalic infants. Official statements from the American Academy of Pediatrics (1992) affirmed that anencephalic infants were not appropriate organ donors if this organ donation would shorten the life expectancy of the anencephalic child and rejected arguments to modify medical criteria of brain death. In this case, legal and ethical consultations determined that the anencephalic child could provide a unilateral kidney as a living donation. As parents of twins, a conflict of interest exists when the promotion of life of one twin stands over the second. The anencephalic twin had an appointed guardian ad litem to ensure protection of interests as recommended by legal and ethics committee members.

Neonatal Transplantation

There is 1 reported case of an attempted neonatal renal transplant from an anencephalic fetus. In 1970 LaPlante, Kaufman, Goodwin, et al. published findings of an attempted renal transplant for a 6-day-old infant with an ‘en bloc’ renal graft from an anencephalic donor. This case involved ESRD secondary to severe congenital obstructive uropathy secondary to posterior urethral valves. On day 6 of life, sleeves of the donor aorta and vena cava with their renal vessels intact were interposed into the recipient’s aorta and vena cava. Although the grafts were found to be patent, the renal arteries never perfused and the kidneys were immediately removed. The authors concluded that this represented a technical failure. The child died 3 months later secondary to hyperkalemia.

Renal Transplant in Pediatric Population

In 2000, Millian et al. showed a 90% survival rate of kidney allografts in children less than 15 kg, however, the mean weight of transplantation was 11.2 kg. They report no graft loss due to vascular thrombosis, primary non-functional, technical error, or acute rejection. One graft was lost secondary to potassium overdose at day 1, two grafts were lost secondary to sepsis after 2 years, and one graft was lost due to chronic rejection. While pediatric transplantation success is well described, there is a paucity of data regarding success of neonatal transplantation.

Conclusion

Although the clinical outcome was death, this rare case represents a technical success of a premature neonatal living related renal transplant. Additionally, transplant surgeons should be alerted to the potential for unilateral donor nephrectomy and aortic reconstruction procedure allowing anencephalic neonates to serve ethically and legally as living donors for renal transplantation. With regard to the rare presentation of this case, we predict future success with transplantation in this rare clinical scenario.

References


www.spuonline.org

The Society for Pediatric Urology

57th ANNUAL MEETING

May 17, 2008 Orlando, Florida

There is no registration fee for the SPU Annual Meeting. However, REGISTRATION IS REQUIRED to process your CME credits. The American Urological Association, Inc. designates this Continuing Medical Education activity for a maximum of 8.5 credit hours in Category I of the Physician’s Recognition Award of the American Medical Association and the American Urological Association’s Certificate of Continuing Medical Education.

Check out the complete program and register online at www.spuonline.org!
A Rare Case of Gartner’s Duct Ectopic Ureter

Andy Y. Chang, M.D., Children’s Hospital Los Angeles
Pasquale Casale, M.D., Alexander Kutikov, M.D., Douglas A. Canning, M.D., Children’s Hospital of Philadelphia, Philadelphia, Pennsylvania

Introduction

We present a case of a single system ectopic ureter inserting into a Gartner’s duct cyst.

Case

A 1 month old baby girl presented to clinic with a history of left cystic kidney discovered at 16 weeks gestation. A repeat ultrasound revealed a right kidney measuring 5.7 cm with mild pelviectasis of 0.5 cm. The left kidney was 4.9 cm with multiple cysts and minimal blood flow. A large cystic structure consistent with a ureterocele was also present. VCUG at age 7 weeks revealed a 35 ml bladder without VUR and a normal urethra. Again noted was a mass at the base of the bladder consistent with a ureterocele. Mercaptoacetylglycine renal scan showed normal right but nonfunctioning left kidney.

A follow-up exam a month later revealed a protruding mass apparently originating from the urethra. The baby girl seemed to “strain” with urination. She was taken to the operating room several days later for incision of the ureterocele. Intra-operatively, we found an ectopic left ureter inserting into a Gartner’s duct cyst, confirmed by ureteroscopy after incision of the cyst.

Repeat ultrasound 2 months later showed resolution of the right pelviectasis and involution of the left multicystic dysplastic kidney.

Discussion

In the strictest term, ureteral ectopia is defined as an orifice that is not at the orthotopic, cranialateral position on the trigone. The incidence is estimated at 1 in 1900 with a predominance towards girls over boys of 2.9 to 1.1. Eighty percent of ectopic ureters are associated duplicated systems and more than 80% of girls with ectopic ureters have duplicated systems.2 Single ectopic ureters are more commonly found in males (75%) though this does not indicate that the majority of males with ectopic ureters have single systems.3

Ectopic ureters may drain anywhere along the GU tract, with fewer than 1% draining into Gartner’s duct, a vestigial Wolffian duct structure.4 The existence of this developmental anomaly can be better understood with the help of embryology. In the female embryo, the Wolffian ducts are reabsorbed into the Mullerian tract leaving possible residual Wolffian duct remnants, Gartner’s duct cysts, located on the anterolateral vaginal wall down to the hymen. The ureters also develop from outgrowths of the Wolffian ducts, the ureteral buds. Thus, failure of the ureteral buds to separate from the Wolffian ducts can result in the ectopic ureters terminating in the Gartner’s duct cysts. And as predicted by Mackie and Stephens, these ectopic ureters are associated with severely dysplastic renal units4, as seen in our patient.

Conclusion

Ectopic ureters ending in Gartner’s duct cyst are extremely rare and often associated with renal dysplasia or dysgenesis. Although this embryologic maldevelopment is rare, it should be on one’s differential diagnosis for girls with ectopic ureters. When encountered, surgical intervention is recommended.

References

Fusiform Megalourethra and Oligohydramnios
Ranjiv Mathews, M.D., Amanda North, M.D., Jude Crino, M.D., The Johns Hopkins Medical Institutions, Baltimore, Maryland

Introduction
Megalourethra is a rare anomaly and is rarely diagnosed antenatally on ultrasonography. Fusiform megalourethra is associated with deficiency of both the corpus spongiosum and corpora cavernosa. There is a high infant mortality secondary to obstruction and renal failure. The infant presented in this report has survived with early institution of dialysis and is being considered for renal replacement and reconstruction.

Case Presentation
A 39 year old Gravida 5 Para 4-0-0-4 mother presented at 22 weeks gestation following a sonogram at an outside institution indicating the presence of a distended fetal bladder, dilated ureters bilaterally and fetal pericardial effusion. Repeat ultrasonography indicated the presence of bilateral hydronephrotic echogenic kidneys, ureteral dilation, distended fetal bladder and posterior urethra as well as a dilated penile urethra (Figure 1a, b, c). Vesicocentesis indicated the presence of decreased urine sodium (101 meq/l), urine chloride (87 meq/l) and osmolality of 218 mOsm/kg. Karyotype was noted to be 46XY. On ultrasonographic follow-up, oligohydramnios was noted by 29 weeks but fetal growth continued to remain normal. Delivery was induced at 37 weeks 2 days and birth weight was 2860 g. Apgar scores were noted to be 5 at 1 min and 7 at 5 mins. The baby was initially intubated in the delivery room but was able to be extubated on day one.

On examination at birth the baby was noted to have a small thorax, abdominal distension, a flaccid penis (Figure 2) with no evidence of palpable corpora and a single band indicating the location of the urethra. A urethral catheter was placed with some difficulty, however, urinary output was low and the creatinine continued to rise. A peritoneal dialysis catheter was placed on day 9 of life and the urethra was endoscoped (Figure 3 a,b). Additionally a vesicostomy was performed to provide bladder drainage. Dialysis was begun on day 13. The infant remains on dialysis in renal failure. He has had one episode of sepsis, but is continuing to increase in weight. Plans are being made for reconstruction of the penis and also for potential renal replacement.

Discussion
Megalourethra is a rare congenital anomaly that is caused by deficiency of the erectile tissue in the penis. Stephens and Fortune postulated that this deficiency may be caused by delay in canalization of the urethra in the glans. Another suggested mechanism is failure of mesodermal differentiation and secondary lack of support of the urethral epithelium. Megalourethra has been classified as scaphoid or fusiform depending on the whether the corpus cavernosum is involved, however, there is a significant spectrum of disease. Both forms of megalourethra are associated with upper tract malformations, so evaluation of the upper tracts is mandatory.

Ultrasound diagnosis has been reported, however, none of the prior reported cases of fusiform megalourethra has been associated with a surviving patient. Infants born with renal impairment require early institution of dialysis to support life. Reconstruction of the penis in patients with scaphoid forms was described by Nesbitt. Fusiform variants with little or no corporal tissue have few options for functional surgical reconstruction.

Early ultrasonographic diagnosis permits parental counseling and consideration of terminating pregnancy in the fetus with multiple anomalies.

References
According to the Urologic Diseases in America project, 2.4% to 2.8% of children develop a urinary tract infection (UTI) each year and result in 1.1 million annual office visits. Estimated costs for the inpatient treatment of clinical pyelonephritis are $180,000,000 each year.

When discussing the management of UTIs, it is important to be specific about the type of UTI. When a child has a febrile UTI or experiences significant abdominal or flank pain, it is termed “clinical pyelonephritis”. If an imaging study such as a DMSA scan or CT scan demonstrates acute renal cortical involvement, then it is termed acute pyelonephritis. This condition occurs most commonly with grades III-V vesicoureteral reflux (VUR), but certainly can occur in the absence of demonstrable VUR. Areas of involvement with acute pyelonephritis have a 50% risk of becoming renal cortical scars. If a child with a febrile UTI does not show evidence of acute renal cortical involvement, then infection is termed “acute pyelitis”. These children may be as ill as children with acute pyelonephritis, but will not develop renal scarring from that particular infection.

Recurrent UTIs are common. In infants, the rate of recurrence is 18% - 26% and is higher in girls than boys. In girls > 1 year of age, the risk of recurrence is proportional to the number of previous UTIs.

In the Hoberman study of children < 2 years of age with their first febrile UTI, 61% had a positive DMSA renal scan. Overall, 39% had VUR, and 96% had grades I, II or III. Consequently, more than half of children with clinical pyelonephritis did not have VUR on contrast cystography. Similar findings have been reported by many investigators over the years.

The pathogenesis of reflux negative febrile UTI has been attributed to bacterial virulence factors. However, other contributing factors are possible also. VUR on conventional imaging can be intermittent. For example, in children with low grade VUR, 15% to 20% with a negative VCUG will have a positive VCUG one year later if the study is repeated. It is likely that some of children with clinical pyelonephritis and a negative VCUG have intermittent VUR. However, what about the rest? Do some children have a ureterovesical junction (UVJ) that is borderline incompetent, one that hydrodistends on cystoscopy but does not reflux on VCUG, that makes them at increased risk for clinical pyelonephritis? In other words, are hydrodistending UVJs more likely to allow bacterial ascent than those that do not hydrodistend?

Data from the early PICC studies suggest that we reconsider our traditional concepts of the pathogenesis of upper tract infection and be willing to evaluate and treat some children for VUR when reflux has not been documented by conventional imaging. However, by advocating PICC, are we failing to take into account other likely contributing factors, such as voiding dysfunction and the possible effect of cystoscopy. Two experts in the field of UTI, Dr. Earl Cheng and Dr. Hans Pohl, will present a point-counterpoint discussion in what I think is one of the most controversial and polarizing topics in pediatric urology.

References

PIC Cystography: Is There a Role?

Earl Y. Cheng, M.D., Associate Professor of Urology, Children’s Memorial Hospital
The Feinberg School of Medicine at Northwestern University, Chicago, Illinois

This case is certainly not uncommon since up to 50% of children with febrile UTIs will be found to have normal radiographic studies. Many of these children without radiographic evidence of reflux will go on to have recurrent febrile UTIs. When this occurs, there are two questions that arise: 1) Why is the patient having recurrent febrile UTIs? and 2) What’s the next step? There may be many reasons why the infections are occurring including the presence of various host and bacterial virulence factors, as well as possible other contributing factors. As stated, infection may be intermittent. Some children may have a UVJ that is borderline incompetent, one that hydrodistends on cystoscopy but does not reflux on VCUG. These children may be at increased risk for clinical pyelonephritis.

Following early observations that demonstrated that children with recurrent febrile UTIs and negative VCUGs were frequently found to be PICC positive for reflux, several studies have been performed to evaluate the validity and utility of this study.

(continued on next page)
as inadequately treated dysfunctional elimination syndrome. Nevertheless, it is widely accepted that it is also possible that this type of patient may have occult reflux that can allow onset of a lower tract infection to an upper tract infection that is febrile in nature. If so, identification and treatment of this form of occult reflux should result in a decrease in recurrent febrile UTIs. The PIC cystogram represents a relatively simple way to identify this type of reflux that may be clinically significant. The evolution of this test was based upon previous observations that many of these children were found to have patulous orifices that easily hydrosidtend when they were evaluated endoscopically.

The protocol for the PIC cystogram has been previously described. Briefly, it involves placing the beak of the cystoscope at the level of the orifice and instilling contrast under gravity at a height of 1 meter. This is the height that the contrast is hung for a conventional VCUG. Ongoing studies at our institution have demonstrated that when the PIC cystogram is performed in this fashion, the pressure at the orifice does not exceed 20 cm. water pressure.

Following early observations that demonstrated that children with recurrent febrile UTIs and negative VCUGs were frequently found to be PIC positive for reflux, several studies have been performed to evaluate the validity and utility of this study. In the first pilot study, Rubenstein et al evaluated a control group (n=15) and study group (n=30). The control group consisted of patients that had no history of febrile infections and were known to have a normal ultrasound and VCUG (i.e., patients with dysfunctional voiding and recurrent cystitis or patients with UPJ obstruction). The study group consisted of patients with recurrent febrile infections that had a normal ultrasound and VCUG. All patients underwent PIC cystography. None of the patients in the control group were found to be PIC positive for reflux while all 30 patients in the study group were found to have reflux. The finding of a 0% incidence of PIC positive reflux in the control group is a very important one since this demonstrates that a patients with a normal orifice will not reflux with PIC cystography and that it is only those patients that are having clinical problems with reflux that have PIC positive reflux. Edmonson et al performed a f/u study at 4 separate institutions to ensure that these findings were valid and not institution specific. This study demonstrated that over 80% of patients with febrile UTIs and negative radiographic studies were found to be PIC positive for reflux and that there was a strong correlation between those orifices that refluxed and those that hydrosidtended.

Following review of these initial studies, many would comment “So what? Just because a patient has PIC positive reflux doesn’t mean that this reflux is clinically significant.” This is a very valid and important question to raise. To evaluate whether PIC positive reflux is clinically significant, a recent multi-institutional study was performed. The hypothesis was straightforward: If this type of reflux is clinically significant (and not just an artifactual radiographic finding), then patients that are found to be PIC positive for reflux and are treated surgically should have a lower rate of febrile UTIs post surgery. The study evaluated 87 patients that had surgical correction of PIC positive reflux (85 endoscopic and 2 open reimplantation). The number of febrile UTI/child/month decreased from 0.17 pre-op to 0.01, which is highly statistically significant. This strongly suggests that PIC positive reflux is indeed clinically significant. However, to further definitively prove this, a prospective randomized trial is now underway in which patients that have PIC positive reflux are being randomized into 2 study groups: no treatment observation (no antibiotics or surgery) and treatment (endoscopic or open surgical correction of reflux).

Those individuals that argue that the PIC cystogram has no place in the evaluation of children with febrile UTIs will point out that there is no data to look at scarring on the kidney and that there are other reasons for ascending infections in children. Also, febrile UTIs do not necessarily translate into true renal parenchymal infections. All of these points are true. In addition, many have also argued that we shouldn’t be looking for occult reflux when we aren’t even sure if we should be treating low grade reflux in symptomatic children. This may also be true. However, if we attempt to simplify the question, we hopefully can come to more of a consensus. Thus, if one asks the question: In patients that have recurrent febrile UTIs and a negative VCUG, can the PIC cystogram identify clinically significant reflux? The preponderance of available data strongly suggest that the answer is: YES. More importantly, the majority of patients that are found to be PIC positive for reflux will benefit clinically with a reduced risk for recurrent febrile UTI when this reflux is surgically corrected. While we may not know whether this type of occult reflux has the potential for renal scarring, this isn’t the question that we are asking at this time, although it does need to be evaluated in the future. Rather, we are simply trying to identify a subset of difficult patients that have occult reflux that can be treated and in turn reduce the morbidity of future febrile UTIs. Even though many of these patients may not have significant renal scarring or be at risk for renal damage, the clinical benefit to these patients should not be downplayed. The ability to identify a causative factor that can be treated surgically and reduce or eliminate future febrile infections is very pleasing and beneficial to both patients and their families.

I am the first to admit that when I first heard of the idea of the PIC cystogram. I had my doubts. In my own practice, these types of patients that may benefit from a PIC cystogram are not that frequent. My own personal bias is that the reason that they aren’t very frequent in my practice is that I tend to be very aggressive in identifying and treating dysfunctional voiding and constipation. However, I still have had a few patients over the past few years that have had negative radiographic studies and recurrent febrile UTIs despite aggressive management of their dysfunctional elimination. All of these patients were found to have PIC positive reflux. They were all treated with endoscopic subureteric injection therapy and none of them have gone on to have recurrent febrile infections.

Hopefully, future studies will elucidate the exact pathophysiology of PIC positive reflux and its true effects on the kidney. Until then, I would strongly urge those of you that have your doubts to put away any biases that you may have and strongly consider the use of the PIC cystogram in the future and also consider enrollment of patients in the prospective randomized trial.

References


We cannot deny the positive impact that has been made on the incidence of severe acquired renal cortical scarring through the early diagnosis of vesicoureteral reflux (VUR) and the prevention of recurrent acute pyelonephritis (APN) through prophylactic antibiotics and/or surgical intervention, as indicated. The standard evaluation of fUTI in children has long included ultrasonography and VCUG. Using this approach, the prevalence of VUR in children presenting with fUTI varies between 25-50% (mean, 35%) depending on the timing of the VCUG and the number of filling-voiding cycles performed. Using DMSA renal scans to document the presence of APN, Majd and Rushton performed a VCUG during hospitalization, finding VUR in only 37% of patients with renal cortical abnormalities. Other studies have shown that when a VCUG is delayed for 4 – 6 weeks following presentation for fUTI, as many as 28.5% of children with VUR will escape diagnosis. In addition, it has been reported that grades III and higher VUR can be missed in up to one-third of patients undergoing only one filling cycle, generally performing two filling cycles increases the yield of identifying VUR, that is generally of low to moderate grade. While it is true that VUR can elude diagnosis depending on the timing and technique used during VCUG, it is illogical to argue that that VUR which can only be demonstrated by positioning a stream of contrast at the ureteral orifice is not iatrogenic. Uropathogenic bacteria that infect the upper tracts of children with presumed PIC-positive VUR did not have the benefit of a cystoscope to gain access to the upper urinary tract. Even so, when PIC cystography is performed the VUR that is identified is generally of low grade and no child is offered the opportunity to undergo long-term follow-up in order to assess the impact of maturation and/or intervention for voiding dysfunction before surgery is performed. Assuming that occult VUR does indeed exist, what is the evidence in support that all VUR, even that which presents with UTI, requires surgical intervention? Cooper et al reported their observations on 51 children whose prophylaxis was discontinued despite persistence of VUR. During a mean follow-up of 3.7 years, VUR resolved in 19.6% of the children. A UTI occurred in 11.8% (6/51) following cessation of prophylaxis had another UTI, prompting reimplantation in 5 of the 6 children, but in no child was acquired renal cortical scarring documented. Clearly there is a subset of children at risk for recurrent UTI’s and renal scarring, both with and without VUR, which should be the focus of intervention, while the majority of children may do well without antibiotics or surgery for VUR.

Recently, several studies have advocated the concept of a “top down” approach to the evaluation of children with UTIs. This selective approach focuses on renal status rather than the presence or absence of VUR. The goal of this approach is to identify only those children with clinically significant VUR, defined as those at risk for renal scarring or recurrent febrile UTIs. It begins with a DMSA scan, reserving evaluation with a VCUG only for those who have abnormal DMSA findings or subsequent recurrent UTIs. This approach is supported by prior clinical studies that have reported abnormal DMSA findings in 80-90% of children with VUR and febrile UTIs, including almost all with Grade III or higher VUR. A recent prospective study analyzed this approach in 290 patients (79% with fever ≥38.5C). DMSA scans performed within 2 weeks of the infection demonstrated abnormalities in 51% of patients, including 26/27 (96%) of those with grade III or higher VUR. Using this approach, the authors conclude that, if only those with positive DMSA scans were evaluated with cystography, VCUGs would have been avoided in half of their patients. Not only would this reduce the number of children being evaluated with VCUGs, it would also avoid treatment of children with clinically insignificant VUR. It is obvious that the debate regarding the optimal management of VUR and UTIs continues, as it should, since treating every child as if they were a “nail” with one or another therapeutic “hammer” does a disservice to our patients.

References
An Unusual Fetal Presentation of an Abdominal Wall Defect in a Dichorionic-Diamniotic Twin

William Tu, M.D., Ph.D.1, Lu Anne Dinglasan B.S.1, Jane Chueh M.D.2, William Kennedy M.D.1
Departments of Urology1 and Obstetrics and Gynecology2, Stanford University School of Medicine, Stanford, California

Introduction

Bladder exstrophy is a rare congenital malformation from incomplete closure of the lower anterior abdominal wall. Failed mesenchymal cell migration between the cloaca and abdominal wall ectoderm leads to absence of connective tissue and muscles. The diagnosis must be differentiated from the more common anterior wall defects.

Case Report

A 38-year-old, gravida 8, para 1-0-6-1, women presented with a dichorionic-diamniotic twin intrauterine pregnancy with discordant growth. Twin A appeared normal but twin B was 37% smaller. Ultrasound at a late gestational age of 32 weeks on twin B showed normal amniotic fluid and a 2-3 cm diameter lower abdominal mass with an entering vessel and extracorporeal liver suspicious for an omphalocele (Figure 1). The patient was counseled on abdominal wall defects. At 35 6/7 weeks, the patient underwent cesarean section delivery. Examination of newborn B showed a low set umbilicus, diastatic pubis, epispidias, and large everted bladder consistent with bladder exstrophy (Figure 2).

Discussion

Congenital anterior abdominal wall defects are diagnosed initially on perinatal ultrasound. Anatomic evaluation is limited in twin pregnancies and is optimal at an earlier gestational age of 20 weeks. Unlike bladder or cloacal exstrophy, omphalocele and gastroschisis are associated with a filling and emptying bladder. A persistent urachus may be misinterpreted as a bladder separate from the lower abdominal mass. Umbilical arteries running inferior to the mass along with an umbilical vein running into the mass suggest an omphalocele. Umbilical arteries running alongside the mass indicate a bladder exstrophy. Ultrasound diagnosis of congenital defects is important for perinatal counseling.

Antenatal Imaging of the Ectopic Ureter Allows for Early Reconstructive Surgery and Maximizes Recoverable Function

John G. Van Savage, M.D., FAAP, Regional Urology, Shreveport, Louisiana

Introduction

The upper pole ectopic ureter with complete ureteral duplication is frequently difficult to diagnose preoperatively despite appropriate imaging studies. Suspicion antenatally usually leads to a rapid radiologic evaluation postnatally with early surgical reconstruction. We present our experience of ectopic ureters with duplicated systems suspected antenatally and confirmed postnatally.

Cases

Five female neonates presented with a history of antenatal hydronephrosis. Renal sonography, VCUG and diuretic renography documented 4 upper pole megaureters and 1 upper pole refluxing ectopic ureter. Upper pole renal function was preserved in each case. Cystoscopy at the time of surgery confirmed complete ureteral duplication and an upper pole ectopic ureter in each case. Retrograde uroteropyelography of both upper and lower poles moieties defined the anatomy very well (Figure 1). An extravesical common sheath tapered ureteroneocystostomy with division and reanastomosis of the upper pole ureter was performed. There were no complications. Followup imaging studies revealed decompression of all ureters and preservation of upper pole renal function. Mean follow-up was 2 years.

(continued on next page)
Discussion

Early recognition and radiologic evaluation of antenatal hydronephrosis can lead to the suspicion of the ectopic ureter but usually not a definitive diagnosis. Historical series indicate rates for upper pole nephrectomy for the nonfunctioning upper pole ectopic ureter as high as 100% in patients without the benefit of antenatal sonography. These children presented at a mean age of 5 and none of them had preserved upper pole renal function with their ectopic upper pole ureter. In this small series we avoided the morbidity of the usual presentation of pyelonephritis with loss of upper pole renal function. We were able to preserve all of the upper pole moieties diagnosed with an antenatal suspicion through early reconstructive surgery. As MRI improves, it is possible that it may be able to better delineate the site of insertion of a hydronephrotic upper pole ectopic ureter preoperatively without sedation in the future.

References


Bladder Exstrophy Variants: An Individualized Approach

Douglass B. Clayton, M.D., David M. Kitchens, M.D., David B. Joseph, M.D., C. D. Anthony Herndon, M.D.
Section of Pediatric Urology, University of Alabama at Birmingham, Birmingham, Alabama

Introduction

Classic exstrophy is a rare congenital malformation consisting of an exstrophic bladder plate, epispadiac penis in boys and bifid clitoris in girls, divergent rectus muscles and a widened pubic symphysis. Moreover, variants of the classic bladder exstrophy and their management are even more rare. We present two cases of bladder exstrophy variants and their subsequent management.

Cases

MS is a male patient who presented to our institution with an early second trimester prenatal ultrasound that revealed a left multicystic dysplastic kidney and concomitant right hydronephrosis with normal amniotic fluid volume. A followup ultrasound at approximately 30 weeks revealed a bladder abnormality suspicious for classic bladder extrophy, and at this time we offered prenatal exstrophy counseling. The patient was delivered at term and subsequently transferred to our institution. Newborn physical exam revealed an abdominal wall defect covered by a layer of loose overlying skin with a small inferior orifice noted to be draining urine (Figure 1). Other pertinent findings included bilateral undescended testicles, imperforate anus, and a dysplastic right thumb. Cystoscopy via the inferior urine draining orifice on DOL 2 did not identify any recognizable structures. Pelvic radiography demonstrated 5.5 cm of pubic diastasis. Subsequent management included surgical exploration of the abdominal wall via a midline, clam-shell type incision through the loose overlying skin. This revealed an exstrophic bladder plate and epispadic penis; thus, confirming the diagnosis of covered bladder extrophy. The patient then underwent a single-stage bladder extrophy closure with performance of pelvic osteotomies.

SH is a two day old female patient born full term at an outside facility. The initial newborn examination revealed a mass on the ante-
rior abdominal wall prompting neonatal ICU admission and subsequent transfer to our facility. Our initial evaluation revealed an anterior abdominal wall defect with an extrophic bladder plate lacking any obvious urine output or ureteral orifices (Figure 2). The clitoris was also noted to be intact. The VCUG and vaginogram demonstrated two separate structures, an intact posterior bladder without reflux (Figure 3A) and a separate vagina with a normal cervix (Figure 3B). Upon maximal filling with contrast, the posterior bladder herniated through the abdominal wall defect beneath the approximate location of the exstrophic bladder plate. Pelvic radiography revealed 3.3 cm of pubic diastasis. Cystoscopy was later performed which demonstrated a coapted urethra and normal bilateral ureteral orifices. Thus, the patient was subsequently diagnosed with duplicate bladder exstrophy. At 8 months of life, the patient underwent excision of this exstrophic plate, abdominoplasty and umbilicoplasty. The role of pelvic osteotomies and uterine suspension were discussed with the parents, but neither of these adjunctive procedures were performed at the time of repair.

Discussion

Classic bladder exstrophy is only seen in 1 in 40,000 live births and is predominately found in males. The recognized variants of classic bladder exstrophy are up to 10 times as rare. From a database of over 800 exstrophy-epispadias patients, Lowentritt et al in 2005 identified 19 patients with variants of the classic bladder exstrophy and proposed four major categories of these variants. These categories include pseudoexstrophy, superior vesical fissure, duplicate bladder exstrophy and covered bladder exstrophy. The same musculoskeletal defects of classic bladder exstrophy are consistently seen in each of these categories, but the patients vary based on the severity and appearance of the genitourinary anomalies. Due to the rarity of these variants, the management of each patient must be individualized. However, the tenets of repair remain similar to the classic bladder exstrophy patient. These tenets include closure of the genitourinary anomalies and tension-free approximation of the musculoskeletal defects with or without the use of pelvic osteotomies.1-3-4

References


Management of Distal Ureteral Stricture by Refluxing Ureteral Reimplant

Joshua J. Meeks M.D., Ph.D., Elizabeth B. Yerkes, M.D., Bruce W. Lindgren, M.D., Jennifer A. Hagerty, D.O., Division of Urology, Children’s Memorial Hospital, Chicago, Illinois

Introduction

While antenatal hydronephrosis has become a common finding on screening prenatal ultrasound, the resultant diagnosis of obstructing ureteral stricture is very rare.1 Traditional treatment options include percutaneous nephrostomy, cutaneous urostomy and ureteral tapering with reimplant. We describe two patients with ureteral strictures that were operatively managed with refluxing ureteral reimplants.

Case Reports

Patient 1 was born at 29 weeks with prenatal right hydronephrosis and left multicystic dysplastic kidney (MCDK). Serum creatinine peaked at 3.5 mg/dL. Antegrade and retrograde nephrostograms identified a long distal ureteral stricture near the bladder (Figure and inset). Patient 2 was a full-term female with prenatal hydronephrosis. Postnatal imaging demonstrated severe right hydronephrosis, left MCDK with right reflux identified on VCUG. Cystoscopy and retrograde pyelograms revealed bilateral ectopic ureters and a right distal ureteral stricture. Both patients underwent temporizing placement of percutaneous nephrostomies prior to surgery. Yet, internalization via ureteral stent was not possible due to narrow ureteral stricture (Patient 1) and ectopically located ureter at the urethra (Patient 2). In order to allow internal drainage of urine extravesical refluxing ureteral reimplant was performed at 83 and 17 days, respectively. Both patients tolerated the procedure well. Renal function has remained stable and both are without infection.

(continued on next page)
Discussion

Congenital ureteral strictures occur rarely and may be difficult both to diagnose and manage. The patients described had prenatal diagnosis of hydronephrosis and contralateral MCDK. In addition to internal drainage, reimplantation will allow developmental cycling of the urinary bladder. This procedure may be a temporizing or definitive procedure in these patients. While refluxing reimplant has been performed for obstructed megaureters, we describe the first application of this operation to ureteral strictures in patients with solitary kidneys.

References

Postpartum Hemorrhagic Renal Rupture in a Patient with Massive Prenatal Hydronephrosis

Eric D Nelson M.D.1, Job K Chacko M.D.1, Philip Shlossman M.D.2, T. Ernesto Figueroa M.D.1

1AI duPont Hospital for Children, Thomas Jefferson University, Wilmington DE, 2Christiana Care Health System, Wilmington Delaware

Introduction

Massive prenatal hydronephrosis is commonly caused by a ureteropelvic junction (UPJ) obstruction. Urinary ascites and urinomas are rare but documented sequelae of such obstruction, but to our knowledge, significant associated hemorrhage has not been reported. We present a case of massive prenatal hydronephrosis with hemorrhagic rupture of the UPJ in the neonatal period.

Case Report

A 29 year old female underwent routine ultrasound at 27 weeks. Images showed a male with bilateral hydronephrosis (30mm left renal pelvis and 11mm right renal pelvis). Repeat ultrasound at 34 weeks showed hydronephrosis on the left to 6.0 cm (Figure 1).

The patient was born at 38 weeks via C-section after a failed induction. Gross hematuria at first void was noted, and within 24 hours the hemoglobin decreased from 11.2 to 8.0 requiring three transfusions. Initial ultrasound showed free fluid within the abdomen and significant left hydronephrosis with clot within the renal pelvis. CT scan confirmed hemorrhagic rupture of UPJ (Figure 2) and a nephrostomy tube was placed.

Subsequent antegrade nephrostogram was consistent with a UPJ obstruction. A renal scan showed a split function of 40% on this affected side. He then underwent an open dismembered pyeloplasty and is doing well after stent removal.

Discussion

To our knowledge, this is the first report of ureteropelvic junction rupture associated with significant hemorrhage. In review, many questions are raised, including the prenatal management of such massive hydronephrosis. This degree of dilation usually indicates an obstruction and many would argue that this kidney would likely have limited function. Prenatal intervention is usually only reserved for cases when there is mass effect on adjacent major structures. It is conceivable that aspiration may have decreased the tension of the urinary system and made the possibility of hemorrhage less likely. Because this is only a temporary solution, delivery would have needed to be induced shortly after.

A second potential criticism is the timing and mode of delivery. Some may argue for an earlier delivery and a C-section to decrease the risk of rupture. Multiple cases have been reported in adults with an undiagnosed UPJ obstruction after blunt trauma. C-section would likely be a less traumatic mode of delivery but as illustrated in this case, it still occurred.

Postnatally, one could argue not to intervene at all. The patient responded well to transfusions and subsequently remained hemodynamically stable. If we had treated this patient as a simple blunt injury trauma, recent literature supports conservative management. However, the major difference in this case is that there was an underlying UPJ obstruction with unknown kidney function. We chose an aggressive route and performed percutaneous drainage. Admittedly, there are risks for the child and we could have altered any tamponade effect of the perinephric hematoma. The nephrostomy tube had the advantage of allowing us to monitor differential urine output as well as easily perform an antegrade study. In this patient, the nephrostomy tube turned out to be an excellent adjunct.

(continued on next page)
It is interesting to find that the kidney contributed 40% of the function even after having such severe prenatal obstruction and neonatal trauma. Although we will unlikely again see a similar presentation, we will now give these apparently severely obstructed kidneys the “benefit of the doubt.”

**Midline Scrotal Mass in a Six-Year-Old**

**Introduction**

We present an unusual case of a midline scrotal mass in a six-year-old boy.

**Case Report**

A six-year-old boy presented with a midline scrotal mass that had developed over several months. He was otherwise asymptomatic. The patient had no additional medical or surgical history. On physical examination, the patient was a circumcised male with normal testes descended bilaterally. A well-encapsulated round smooth lesion approximately the same size as the testes was located in the midline of the scrotum. A scrotal ultrasound demonstrated a septated hypoechoic lesion surrounded by an echogenic rim. A VCUG revealed a blind-ending structure from the bulbular urethra filling with contrast and projecting into the scrotum. The patient was taken to the operating room for excision. A perineal incision was made and the mass was dissected down to a small neck that appeared connected to the bulbular urethra. A small incision was made in the lesion, draining serous fluid. We attempted to intubate the neck of the lesion with a glidewire, but no fistulous tract contiguous with the urethra was appreciated on urethroscopy. After excision, gross pathology revealed a cyst lined with verrucous mucosa. Micropathology confirmed the presence of skeletal and smooth muscle, nerve fibers, and glandular components. The lumen was lined with keratinizing squamous epithelium. Final pathology could not confirm a definitive diagnosis.

**Discussion**

Our differential diagnosis included an epidermal inclusion cyst with a fistulous connection to the urethra, an ejaculatory duct cyst, or a Cowper’s duct cyst. Epidermal inclusion cysts may occur anywhere along the median raphe and can mimic polorchidism. They contain keratinous material but are typically well-encapsulated. Ejaculatory duct cysts have been described as a cause of epididymoorchitis in prepubertal children. Our patient’s pathology, however, was not consistent with this diagnosis. Cowper’s duct syringoceles in children likely results from a congenital retention cyst and may be either closed or open. In the open subtype, urine can reflux into the syringocele and result in post-void dribbling among other urinary symptoms. While a definitive diagnosis has not been confirmed, this unusual case highlights several points in considering the differential diagnosis.

**References**


**Collateral Urethral Duplication**

**Introduction**

Urethral duplication is a rare anomaly. Most urethral duplications occur in the same sagittal plane while those in the same horizontal plane are extremely uncommon. There have been 13 cases of collateral duplication in male patients previously described in the literature, most of which have other associated congenital anomalies. We present the third description of collateral urethral duplication without other associated anomalies in a male child.

**Case History**

An 8 month old male presented with two granular openings noted at birth. One meatus is midline and the other is left of midline. The patient voids only through his midline opening. There is no history of urinary tract infections. No other significant abnormalities are noted on physical exam. At 20 months of age, the patient underwent cystourethroscopy with retrograde urethrogram of both meatal openings demonstrating a normal caliber midline urethra with a smaller caliber lateral urethra. The lateral urethra narrowed to a pin-point opening just proximal to the external urinary sphincter and distal to the verumontanum.

(continued on next page)
Discussion

There have been approximately 200 cases of urethral duplication reported in the literature with even fewer describing duplication in the collaterals plane. The embryology of urethral duplication is not well established with multiple theories including ischemia, abnormal mullerian duct termination, error of septation, and irregularity or partial failure of mesodermal midline fusion. It is evident that whatever embryologic explanation exists has a global impact accounting for the high frequency of other congenital anomalies observed in conjunction with urethral duplication. Our case is an unusual variant of an already uncommon form of urethral duplication. Only two other cases of patients with collaterals urethral duplication without other associated anomalies have been reported.1, 4

References

Megalourethra Associated with Bilateral Hydroureteronephrosis and Renal Dysplasia

Dena L. Walsh M.D., John C. Thomas M.D., John C. Pope, IV M.D., Mark C. Adams M.D., John W. Brock, III M.D.
Division of Pediatric Urology, Department of Pediatric Urology, Vanderbilt University Medical Center, Nashville, Tennessee

Introduction

We present a case of antenatally detected megalourethra.

Case Report

U/S of a 20 6/7 wks EGA fetus showed bilateral hydroureteronephrosis, thick-walled bladder and a cystic structure connected to the perineum between the fetal legs without identification of genitalia. AFI was normal and amniocentesis showed a 46XY male. U/S at 25 6/7 wks EGA showed an increase in bilateral hydroureteronephrosis and abnormal echogenicity of the kidneys. Male genitalia were identified and the cystic structure appeared connected to the penis and intermittently distended with fluid. U/S at 30 1/7 wks EGA failed to show the cystic structure but a normal penis and scrotum were identified. The child was delivered at 37 wks EGA. On exam the child had a floppy phallus, large meatus and full foreskin. Postnatal U/S showed bilateral renal dysplasia and right hydroureteronephrosis. VCUG revealed a scaphoid-type megalourethra. Creatinine peaked at 3.1 and since a catheter could not be placed the child underwent a vesicostomy.

Discussion

Megalourethra is classified into two types.1 The fusiform type is a deficiency of the corpus cavernosum and spongiosum with circumferential dilation of the penis. The scaphoid variant is a deficiency of the spongiosum with ventral bulging of the urethra.

Megalourethra has been associated with genitourinary and non-genitourinary anomalies.2 Mortality rates of 13% - 66% have been reported in the scaphoid and fusiform variants, respectively, usually due to non-genitourinary malformations.3, 4

Prenatal sonographic findings that suggest megalourethra include a blind ending cystic structure between the legs or elongation and dilation of the penile urethra. Color doppler is useful for ruling out the presence of a loop cyst of the umbilical cord.

Although, detailed examination of the genitalia and perineum during a prenatal ultrasound can aid in diagnosis, megalourethra is often missed due to the rarity of this condition.

References

Cystoscopic view of wire within accessory urethra communicating with midline urethra.

Figure 1 – Ultrasound demonstrating a cystic structure connected to the perineum and located between the fetal legs, measuring approximately 1.9 x 1.0 cm.

Figure 2 – VCUG demonstrating a scaphoid megalourethra.

Figure 2 – VCUG demonstrating a scaphoid megalourethra.
Aphallia with Urethrorectal Fistula and End-Stage Renal Disease: A Case Report and Review of Literature
Heidi A. Stephany, M.D., John M. Gatti, M.D., Romano T. Demarco, M.D., J. Patrick Murphy, M.D., Department of Surgery, Children’s Mercy Hospital, Kansas City, MO and the Department of Urology, University of Kansas, Kansas City, Kansas

Introduction
Penile agenesis is a rare disorder resulting from failure of development of the genital tubercle. The karyotype is almost always 46 XY and the urethra often empties into the rectum or is seen at the anal verge. Most of the literature supports reassignment with feminizing genitoplasty and bilateral orchietomy.1

Case Report
T.S. was evaluated at 2 days of age for aphallia and urethrorectal fistula. Karyotype is 46 XY, with testosterone level 320ng/dL his first week of life. Multiple cysts were seen bilaterally on ultrasound and grade IV reflux was seen on cystogram with the posterior urethra connecting high to the rectum. The patient’s renal function continued to decline and peritoneal dialysis was inevitable. After urodynamics demonstrated a compliant bladder with normal capacity and ability to store at safe pressures, it was decided to close his vesicostomy and allow him to void through his urethrovaginal fistula. He was unable to void spontaneously and subsequently underwent repair of his reflux and continent reconstruction with a ureteral Mitrofanoff catheterizable stoma. Peritoneal dialysis was initiated.

Discussion
Penile agenesis is a rare disorder resulting from failure of development of the genital tubercle. Approximately 80 cases have been reported to date. Gender assignment is an issue in all patients and most reported cases underwent female gender reassignment.2 Recent literature challenges this consensus as it may lead to gender dysphoria as a result of prenatal and postnatal testosterone effects on the brain and sexual development.3 The difficulty arises with creation of a cosmetically appealing and functional phallus. De Castro et al. performed preliminary penile reconstruction during childhood in 4 patients and had good cosmetic results in all patients with only one requiring perineal urethrostomy. They concluded patients with XY karyotype and corresponding hormone production should be raised as males. After appropriate counseling, the parents chose to raise our patient as a male, however, this particular case presented a significant challenge with creating a urinary diversion compatible with minimizing the risk of peritonitis in a child with renal failure on peritoneal dialysis. Warady et al. reported 491 episodes of peritonitis with 5.5% associated with cutaneous incontinent urinary stomas.4 Perineal urethrostomy is an alternative option in this case to decrease the risk of peritonitis, with creation of a phallus during adolescence. This was not chosen because the high urethral insertion would have required an abdominal approach violating the peritoneum in the setting of peritoneal dialysis dependence. Additionally, our patient has presumed detrusor sphincter dyssynergia and would still require catheterization due to his inability to void spontaneously. At this stage, the patient is continent, his stoma is easily catheterized and penile reconstruction and renal transplantation are likely in his future.

References

Megalourethra, Urethral Stricture Disease, Urachal Diverticulum, Bilateral Ureteropelvic Junction Obstruction, Obstructive Megaureter and Absent Rectus Musculature: A Prune Belly Variant?
Douglas W. Storm, M.D., Patrick E. Davol, M.D., Joel M. Sunfest, M.D., Geisinger Medical Center, Danville, Pennsylvania

Introduction
The prune belly syndrome is classically described as a symptom complex consisting of absent abdominal wall musculature, bilateral intra-abdominal testes and various anomalies of the urinary tract. Multiple genitourinary abnormalities, in various forms of severity, have been associated with this syndrome.1,2 We describe a baby boy who was found to have multiple genitourinary anomalies, as well as absent abdominal wall musculature, as a potential prune belly variant.

Case Report
This boy was born at 36 1/7 weeks gestation. Serial prenatal ultrasounds had demonstrated worsening bilateral pyelectasis and polyhydranions (Amniotic fluid index - 28). Evaluation at the time of birth demonstrated a distended, soft abdomen with palpably enlarged kidneys bilaterally. The patient was also found to have an enlarged penis which ballooned ventrally with voiding, consistent with a megalourethra. Both testicles were descended. Electrolytes demonstrated a mild metabolic acidosis. Serum creatinine at birth was 1.3 ng/ml (maternal creatinine 0.9 ng/ml) which decreased to 1.0 ng/ml by day of life six. A renal ultrasound demonstrated bilateral severe pelviectasis and minimally dilated ureters. A voiding cystourethrogram (VCUG) was attempted; however, we were unable to pass a catheter into the urinary bladder. Therefore, cystoscopy was performed, demonstrating dense circumferential strictures at the penoscrotal junction and bulbar urethra which were bluntly dilated. The bladder was found to be severely trabeculated and a urachal diverticulum was also noted. No vesicoureteral reflux was noted on the subsequent VCUG. Endocrine and genotyped analysis were normal.

Further analysis via serial ultrasounds and a MRI urogram detected bilateral ureteropelvic junction obstructions (UPJO) and a left obstructed megaureter. The UPJOS were managed via staged open pyeloplasties. A left tapered ureteral reimplant and partial cystectomy, for removal of the urachal remnant, was performed at 23 months of age. At this time the patient was found to have a thin abdominal wall with absent rectus musculature, consistent with prune belly syndrome. Follow up cystoscopy has demonstrated stable, wide caliber urethral stricture disease. The patient continues to void with a satisfactory stream and without urinary tract infections.

Discussion
The prune belly syndrome is classically associated with absent abdominal musculature, bilateral cryptorchidism and various associated genitourinary malformations.1,2 We present this child as a possible prune belly variant. While he was found to have multiple genitourinary anomalies and an absent rectus muscle, consistent with the prune belly syndrome, he did have bilateral descended testes. Bilateral cryptorchidism has been described as a consistent characteristic of the prune belly syndrome.2 However, we know that in patients with this syndrome there may be varying degrees of abdominal wall deficiencies and a constellation of associated genitourinary abnormalities. Perhaps, as this child’s findings suggest, there may also be a prune belly variant consisting of various genitourinary abnormalities and absent abdominal muscles, but with descended testes.

References
Introduction
Cloacal exstrophy is an extremely rare condition with an incidence of 1/200,000 to 1/400,000 live births.1 It is currently believed that cloacal exstrophy results from a persistent cloacal membrane leading to herniation of the anterior abdominal wall during cloacal embryogenesis.2 The timing of the rupture of this membrane leads to cloacal (prior to 8 weeks gestation) or bladder exstrophy (after 8 weeks gestation).3 Monoamniotic, monochorionic (mono-mono) twinning is also very rare, representing less than 1% of twins born.4 Mono-mono twins are at an increased risk of perinatal mortality secondary to vascular compromise from cord entanglement5. We present our experience with two separate sets of mono-mono twins in which one twin is affected with cloacal exstrophy.

Case 1 - Twin A is a 46XY phenotypic male, born at 32 weeks via cesarean section to a 21-year-old G2P2 mother. Twin B is healthy with no anomalies. Twin A was diagnosed by prenatal ultrasound with cloacal exstrophy, omphalocele, and malformation of the right lower extremity. The mother was transferred to a tertiary care facility at 32 weeks to undergo scheduled cesarean section and consultation by appropriate multispecialties. The treating urologist was present at the birth, the cord was tied off and the baby was immediately resuscitated and intubated for respiratory distress. The exposed cloaca was moistened and covered with a hydrophilic dressing.

Twin A was found to have sacral agenesis, spinal cord tethering and normal kidneys by ultrasound. After medical stabilization of the baby and involvement of appropriate specialty consultants, the baby was taken to the operating room on day 2 of life, where the omphalocele was dissected free from liver and small bowel and the superior fascia was closed. The cecal plate was then divided off the bladder halves, tubularized and anastomosed to the hindgut. The hindgut was brought out to the skin as an ostomy. Because the bladder halves were of insufficient size and the pubic diastasis was measured at greater than 8 cm, the bladder was left open, moistened and covered with a hydrophilic dressing. The baby recovered from the procedure and after adequate feeding was discharged on hospital day number forty-three.

Case 2 - Twin A is a 46XX baby born at 32 weeks via cesarean section to a 28-year-old G8P9 mother. Twin B is healthy with no anomalies. Twin A was diagnosed via prenatal ultrasound with an omphalocele, ventricular septal defect, and right-sided hydropneumothorax. The mother was followed in prenatal clinic by pediatric surgery for this initial diagnosis. At 32 weeks gestation, the mother was transferred to a tertiary care facility to undergo a scheduled cesarean section. Upon delivery, the baby was noted to have a very large cloacal exstrophy and omphalocele. Following immediate resuscitation and intubation for increased work of breathing, the cloaca was moistened and covered with a hydrophilic dressing.

The infant was found to have a multicystic, dysplastic left kidney and a hydropneophrotic, dysplastic right kidney by renal ultrasound. A percutaneous nephrostomy tube was placed on day 2 of life and antegrade nephrostogram revealed a patent right ureter. Her creatinine nadired at 1.0 mg/dL on day thirteen of life. Cardiothoracic surgery was consulted to address multiple cardiac issues including patent ductus arteriosus (PDA), pulmonary stenosis, atrial septal defect and left rotated heart. On day nine of life the PDA was ligated. After extensive consultation, on multiple occasions, with the patient family, appropriate specialists and the ethics committee, the decision was made to progress with surgery.

The infant was taken to the operating room on day 14 of life where she was found to have a large abdominal wall defect, small bladder halves, large pelvic diastasis, blind ending duplicated hindgut, right-sided uterus with tube and ovary, left-sided ovary and tube, ileal atresia and cecal plate within the two bladder halves. A right ureteral orifice was located and a retrograde 3 Fr. feeding tube was passed to the renal pelvis. The large omphalocele was dissected off the liver, the atretic ileum was resected and the cecal plate was dissected free of bladder halves, tubularized and anastomosed to the hindgut. The hindgut was brought out to the skin as an ostomy. The left multicystic, dysplastic kidney was removed to create more intra-abdominal space, however, the defect was too large for primary closure. Accordingly, the abdominal contents were covered with an acellular dermal matrix and the bladder halves were moistened and covered with a hydrophilic dressing.

The post-operative course was complicated by necrotic bowel, chylothorax requiring multiple chest tubes, malnutrition and high ileostomy outputs. Both her nephrostomy tube and retrograde stent were removed and her renal function remained stable without hydrenephrosis. At the time of this writing, the baby is 60 days post-op and remains admitted to the tertiary care facility.

Discussion
Monoamniotic, monochorionic twins are a unique subset of twins in that they share a common amniotic sac. Their unique environment makes them susceptible to vascular accidents due to cord entanglement. The perinatal mortality reaches upwards of fifty percent5. Mono-mono twins also have an increased incidence of congenital abnormalities. Recent studies have revealed that attempting to carry these babies to term can increase their risk of death. Alternatively, after 32 weeks, steroids can be given, and after adequate lung maturation has been verified, the twins can be delivered via scheduled cesarean section.

Mono-mono twins are, therefore, a very unique subset of cloacal exstrophy patients secondary to their immaturity and small size. The standard approach to cloacal exstrophy consists of medical stabilization and early operative management with either staged closure or complete primary repair6-7. However, these mono-mono twins are much more difficult to manage secondary to their small size, likelihood of associated abnormalities and low tolerance of long operative procedures. This small subset of cloacal exstrophy patients may be more well-suited for a staged repair including removal of omphalocele and conversion to classic bladder exstrophy during the first operative procedure.

References

Awarded Best Paper at the Fall Meeting

Cloacal Exstrophy in Two Separate Cases of Monoamniotic, Monochorionic Twins
Sarah Marietti, M.D.1, Karen Driscoll, M.D.1, Jeffrey Spencer, M.D.2, Joshua Copel, M.D.3, Christine Kim, M.D.4, and Fernando Ferrer, M.D.1
1University of Connecticut Health Center Departments of Urology and Maternal Fetal Medicine, 2Yale School of Medicine Department of Maternal Fetal Medicine, 3Connecticut Children’s Medical Center Department of Urology, 4New Haven & Farmington, Connecticut

19
SPECIAL EDITION:
SOCIETY FOR FETAL UROLOGY YEAR IN REVIEW

This issue includes selected case reports and special presentations from the Spring and Fall 2007 Biannual Meetings of the Society for Fetal Urology.