Welcome to the inaugural issue of the Society for Fetal Urology Year in Review. Here we present select case reports and special guest presentations from our 2005 biannual meetings. Our spring 2005 meeting was held in conjunction with the AUA Annual Meeting in San Antonio and the topic was Fetal Cystic Kidney Disease. Our fall 2005 meeting took place in conjunction with the AAP Annual Conference in Washington, DC and the topic was Fetal Bladder Outlet Obstruction. The SFU membership continues to grow, as does our meeting attendance.

The Society for Fetal Urology has developed an Antenatal Hydronephrosis Database, under the direction of Dr. Tony Herndon, University of Alabama-Birmingham. The initial data-gathering phase of implementation is underway and we encourage all SFU members to consider participation. Dr. Herndon may be contacted by email at Anthony.Herndon@ccc.uab.edu for further details. He can also provide a copy of his IRB-approved form. Subsequent phases of this project are anticipated to involve data analysis regarding evaluation, management, and outcome of fetal hydronephrosis.

Another action item of the Society for Fetal Urology is to increase education and communication with obstetricians and maternal-fetal medicine specialists. We look forward to their participation in the Antenatal Hydronephrosis Database. We also anticipate an increased frequency of maternal-fetal specialist lectures at our meetings.

As always, we look forward to hearing suggestions and comments from our members regarding meeting topics and our organization’s efforts to educate health care professionals about fetal urology issues. We also encourage potential members to consider joining us in our endeavors. New members are considered at each of our biannual meetings.

Mark P. Cain, President
Christopher S. Cooper, Secretary/Treasurer

Society for Fetal Urology Year in Review
Spring 2006
36th Biannual Meeting
Friday May 19, 2006
Georgia World Congress Center,
Atlanta, Georgia
Congenital Adrenal Hyperplasia:
Prenatal Diagnosis and Treatment,
and Psychosexual Evaluation

Walid A. Farhat, MD, Course Chair
Guest Speakers:
Kenneth J. Zucker, PhD, C. Psych.
Psychologist-in-Chief, Toronto Centre for Addiction and Mental Health / Head, Gender Identity Service, Child, Youth and Family Program, CAMH
Maria I. New, MD
Professor, Pediatrics and Human Genetics, Mt. Sinai / Director, Adrenal Steroid Disorders Program, Mt Sinai / Founder, Weill/Cornell College of Medicine Children’s Clinical Research Center / Founder, Maria I. New Children’s Hormone Foundation

Please visit our website for registration and abstract submission information: www.fetalurology.org.

FROM THE EDITOR

Anthony A. Caldamone

This issue represents a joint venture between the Society for Pediatric Urology and the Society for Fetal Urology. The Society for Fetal Urology has distinguished itself as an academic organization with the purpose of improving the care of children with congenital urological abnormalities. Its biannual meetings are attended in force and they cover all genitourinary aspects of the fetus. We enthusiastically welcome this association and are happy to provide a forum for the documentation of the highly academic nature of SFU meetings. I applaud the leadership of the SFU and thank the contributors for their excellent manuscripts.
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**Winner of the Excellence and Innovation in Case Presentation Award**

**Glomerulocystic Disease in a Duplicated Kidney**

**Introduction:** We present a unique case of segmental glomerulocystic disease, discovered prenatally, affecting half of a duplicated renal moiety, and with an excellent clinical outcome.

**Case Report:** The 38-week ultrasound was normal save for an 8 cm x 5 cm x 9 cm microcystic mass in the right renal fossa. Birth was uncomplicated, and the neonate was otherwise healthy. There was no family history of any renal or cystic disease. Postnatal ultrasound and CT scan confirmed the lesion and was suspicious for a central area of normal parenchyma. This was managed conservatively at an outside institution and the child was referred to Riley Hospital for Children at age 2.

Cystoscopy revealed 2 ipsilateral ureteric orifices, with retrograde pyelograms confirming separate renal moieties. The upper pole was rotated and laterally displaced. The patient underwent surgical exploration whereupon the cystic mass was found as the entire upper pole moiety of a duplicated system, enveloping a distinct lower pole. The 516 gm mass was removed with preservation of the normal lower pole. The child recovered without incident, and there has been no evidence of further cystic disease anywhere in the body.

Pathology was consistent with glomerulocystic disease, with characteristic cystic dilations within Bowman’s space and associated collapsed glomerular tufts. The cysts completely replaced the subcapsular tissue, and the associated tubules were described as immature.

**Discussion:** Glomerulocystic disease was first described by Taxy and Filmer in 1976, and comprises a heterogeneous group of disorders, all with the pathognomonic cystic dilations of Bowman’s space [1]. The disorders are loosely classified as syndromic, familial, and sporadic. The syndromic patients usually have systemic disorders, including tuberous sclerosis, Trisomy 13, Zellweger’s cerebral-renal-hepatic syndrome, and oral-facial-digital syndrome. Approximately 50% of described cases are classified as familial glomerulocystic disease. These do resemble autosomal dominant polycystic kidney disease, and many of these patients’ relatives do have more typical findings. The glomerular cysts separate the entities, and it tends to occur in younger patients.

Sporadic glomerulocystic disease usually occurs in infants and young children, and is commonly bilateral. Although prognosis is somewhat variable, long-term survivors are uncommon. This is especially true with previous reports of prenatally detected glomerulocystic disease, where all cases have suffered early deaths. The suspected pathogenesis involves obstruction of the glomerulo-tubular junction, and cases associated with hepatoblastoma have examined the role of hepatocyte nuclear factor-1-beta.

Therefore, our case presents several unique aspects. Prenatal diagnosis is rare, especially with such an excellent outcome. Also, instead of a global defect, we present the first case of glomerulocystic disease in a duplicated kidney. In keeping with a sporadic mutation, all other organ systems appear normal and the child has normal renal function.

**References**


**An Atypically Presenting Ureterocele**

**Introduction:** We illustrate a case of antenatal hydronephrosis and oligohydramnios that eventually required renal transplant despite early, aggressive intervention.

**Case Report:** A vesicoamniotic shunt was placed at 21 weeks gestation in a female fetus with antenatal right hydronephrosis and oligohydramnios. Despite this intervention, oligohydramnios recurred at 36 weeks and prompted an induced vaginal delivery. Early postnatal ultrasound and CT scan confirmed multiple right renal cysts with poor corticomедulary differentiation, mild left renal pelvic fullness, but no ureterocele. A voiding cystourethrogram (VCUG) showed a right-sided bladder diverticulum and grade V reflux. Renal scan noted a non-functioning right kidney, and delayed left renal perfusion, with no left ureteral obstruction. We repeated the VCUG at 11 months of age following a febrile urinary tract infection (UTI) that was associated with acute urinary retention. Cystoscopy revealed a duplicated right collecting system with a large, intermittently obstructing ectopic ureterocele, which we incised. Urinary retention recurred one month later. We then performed a right nephroureterectomy, excision of ureterocele, reconstruction of the bladder neck, and left ureteroneocystostomy. Her renal function and mild right hydronephrosis remained stable for several years. Her renal function eventually deteriorated, leading to a living-related renal transplant. Following her transplant, she had two more UTIs with fever. We performed a repeat VCUG that showed grade III vesicoureteral reflux. We reimplanted her allograft ureter. She has had no further infection since her reimplant.

**Discussion:** In this case, early gestational bladder outlet obstruction resulted in the need for renal transplant despite early, aggressive interventions. Delayed diagnosis and treatment of an atypically presenting ureterocele may have contributed to the eventual renal failure. Surgical correction of the ureterocele resulted in stable renal function for a period of years before transplant was required.

This case illustrates the natural course of bladder outlet obstruction in a baby girl occurring early in gestation secondary to an atypically presenting ureterocele. The patient eventually required renal transplant despite appropriate intervention, treatment, and close follow-up. We also emphasize the role of a non-refluxing renal transplant ureteroneocystostomy in the abnormal bladder.
Segmental Multicystic Dysplastic Kidney with Ipsilateral UPJ Obstruction
Adrienne J. K. Carmack, Miguel Castellan, Marcos Perez-Brayfield, Rafael Gosalbez, University of Miami Dept of Urology

Introduction: Multicystic dysplastic kidney (MCDK) and ureteropelvic junction (UPJ) obstruction are two common causes of neonatal abdominal masses. The segmental form of MCDK is exceedingly rare, however, with only 25 cases reported [1-4]. Only one case of UPJ obstruction with hydronephrosis and segmental multicystic dysplasia in a kidney with a single collecting system has been reported [3]. We report the second case, which was treated with resection of the dysplastic portion of the kidney and a dismembered pyeloplasty with preservation of the lower pole.

Case Report: A two-month-old girl presented for follow-up of prenatal hydronephrosis. Imaging studies were consistent with a right duplex system with a dysplastic, nonfunctioning upper pole and lower pole ureteropelvic junction obstruction. We proceeded with removal of the upper pole and pyeloplasty, and were surprised to find a single collecting system with a cystic, dysplastic upper pole segment and the absence of an upper pole pelvis or ureter (Figure). The rare diagnosis of a segmental multicystic dysplastic kidney with ipsilateral ureteropelvic junction obstruction was made.

Discussion: Several theories have been proposed for the etiology of MCDK, including ureteral atresia, pelvoinfundibular atresia, faulty development of the blood supply, inadequate branching of the ureteral bud, and inhibition of nephrogenesis by an unknown substance [2,5]. In this case, the embryologic insults occurred at two distinct time points as evidenced by the multicystic dysplasia, which results from an early insult, and the severe hydronephrosis with normal surrounding parenchyma resulting from the UPJ obstruction, presumably occurring much later in gestation. Initially, only a portion of the ureteric bud-mesenchymal complex was affected, while a later insult to the entire ureter occurred. The contralateral kidney was spared. Whether the etiologic agent of the multicystic dysplasia and the UPJ obstruction are causally related or coincidental is uncertain. The combination of an early insult upon only a section of the ureteral bud-mesenchymal complex, the late development of a ureteral obstruction, and the presence of an unaffected contralateral ureter nicely demonstrates the range of severity and the importance of timing in the development of in utero obstructive renal anomalies.

References

Primary Obstructed Megaureter with Contralateral Cystic Dysplasia and Perinephric Fluid Collection
Adam G. Baseman, Leah P. McMann, Andrew J. Kirsch, Edwin A. Smith
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Case Report: A prenatal ultrasound revealed right-sided renal cystic dysplasia and left-sided hydroureronephrosis in an otherwise normal-appearing male fetus. There was a normal amount of amniotic fluid. The infant did well initially after an uncomplicated delivery at an outside hospital, with appropriate weight gain and development, and was referred for evaluation at four weeks of age. His serum creatinine was 0.5 mg/dl. An ultrasound at that time confirmed persistence of left hydroureronephrosis and right cystic kidney. A magnetic resonance urogram was obtained to delineate renal function and define ureteral anatomy (Figure). The right kidney was small and had poor corticomedullary differentiation with numerous cysts. In addition, there appeared to be perinephric urinary extravasation on post-gadolinium imaging. The left kidney demonstrated hydroureronephrosis with dilation to the level of the ureterovesical junction. The differential renal function was calculated at 61% on the left and 39% on the right. The patient presented shortly after the study in acute renal failure with a serum creatinine of 4.0 mg/dl and a potassium of 8.2 mEq/L. After stabilization in the pediatric intensive care unit, he was managed with percutaneous drainage of the left kidney with a rapid return to baseline renal function. He was taken to the operating room for definitive management at eight weeks of age.

Cystoscopy at that time revealed an orthotopic single left ureteral orifice; no right ureteral orifice was visualized. A tapered ureteral reimplant was performed with a ureteral stent left in place. The patient did well without complications following this procedure. His creatinine after stent removal remained at baseline. Postoperative magnetic resonance urogram demonstrated mild persistent left-sided hydroureronephrosis with an improvement in renal transit time down to 4 minutes, 2 seconds and overall improved renal function.

Discussion: Kidneys that are obstructed during development may demonstrate perfusion and minimal excretion that can be interpreted as function on renal scan or MRI without affording any meaningful contribution to renal function. Additionally, the intermingling of cystic and solid elements in the right kidney places it on the continuum of multicystic dysplastic kidney disease even though some function was demonstrated on radiographic imaging.
Renal Artery and Vein Thrombosis Masquerading as a Mesoblastic Nephroma

Lincoln T. Olsen, John M. Gatti, John P. Murphy
The Children’s Mercy Hospital, Department of Surgery and University of Kansas, Department of Urology

Introduction: We present an uncommon case of prenatal renal vein thrombosis (RVT) and renal artery stenosis presenting as a renal tumor which was managed successfully by nephrectomy.

Case Report: A 1-day-old female whose 33-week gestation was complicated by pre-eclampsia and emergent caesarean section due to a nonreactive fetal stress test was born with a right-sided palpable abdominal mass. She developed hypertension, microscopic hematuria, and thrombocytopenia. Abdominal CT scan showed a large infiltrating renal mass measuring 15 cm in largest diameter with poor contrast enhancement, and a normal contralateral kidney (Figure). The renal pedicle was not visualized on CT. This mass was thought to represent a congenital mesoblastic nephroma (CMN). At 3 days of age the patient was taken to the operating room and a right nephrectomy was performed. At the time of operation few vascular structures or renal pedicle elements were identified. Gross analysis suggested infarction of the kidney with diffuse hemorrhage and necrosis. Microscopic sectioning revealed extensive hemorrhagic necrosis without evidence of neoplasm. The main renal vein and intrarenal veins showed extensive thrombosis and the renal artery demonstrated fibrointimal proliferation with luminal narrowing. Postoperatively, the patient developed portal vein thrombosis. She was maintained on low molecular weight heparin anticoagulation for 3 months after her hospital stay. Her work-up for a hypercoaguable state was negative.

The patient is now 18 months old and has done well. She has developed clinical stigmata of neurofibromatosis but a definitive diagnosis has not yet been made. She is normotensive and renal ultrasound surveillance reveals a normal left kidney.

Discussion: CMN is the most common renal tumor in infants. This tumor typically replaces the entire kidney with a homogeneous, round mass. This patient presented with palpable abdominal mass, hypertension and CT scan suggesting a large solid lesion. Unfortunately, imaging cannot reliably distinguish CMN from other renal mass lesions. RVT is an uncommon occurrence (1/50,000) [1]. Typical presentation RVT is renal enlargement, hematuria, hypertension, and thrombocytopenia. Genetic risk factors that predispose to RVT are factor II and V abnormalities as well as elevated lipoprotein A levels or protein C or antithrombin deficiency [2], none of which were found in this patient. Environmental factors that contributing to RVT formation are sepsis, fetal asphyxia, maternal steroid treatment, and gestational diabetes [2]. Treatment of RVT is based on anticoagulation or thrombolytics; however, 90% of patients treated with anticoagulation have renal atrophy at 4 years of follow-up [2]. Nephrectomy is sometimes indicated for control of hypertension [3]. Perinatal RVT can be managed conservatively, but few reports have good long-term follow-up on renal function. A single case of simultaneous renal artery stenosis and renal vein thrombosis treated successfully with nephrectomy was found in the literature [4]. No association of congenital renal vein thrombosis with neurofibromatosis could be found in the literature; however, neurofibromatosis type I is associated with renal artery stenosis.

References

Coincident Prune Belly Syndrome and Posterior Urethral Valves

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Case Report: A one-month-old white male, born at 36 weeks gestational age with prune belly syndrome (PBS) and a patent urachus was admitted to the hospital with renal insufficiency and a urinary tract infection. He occasionally dribbled urine out of his penis but most of his urine drained from his urachus. His prenatal course had been complicated by oligohydramnios and a prenatal ultrasound showed bilateral hydrourephrosis. His physical examination was notable for bilaterally undescended testes, and lax abdominal musculature. A renal ultrasound (US) and voiding cystourethrogram (VCUG) were obtained. The US showed severe bilateral hydronephrosis and dilated tortuous ureters. The VCUG showed bilateral vesicoureteral reflex with dilated ureters as well as a dilated bladder neck and posterior urethra. A persistent narrowing of the urethra, most consistent with posterior urethral valves, was also noted (Figure). The patient was taken to the operating room where cystourethreoscopy was performed and annular Young’s type III posterior urethral valves were encountered just distal to the verumontanum. The valves were ablated endoscopically and postoperative credé maneuvers were performed. The patient has done well postoperatively and has reached a normal level of renal function.

(continued on next page)
never demonstrated a forceful urinary stream from the penis. Catheter drainage was maintained for 24 hours. Once the catheter was removed the patient was noted to drain most of his urine through his penis. On one month follow-up he continued to have a forceful urinary stream. He is currently scheduled for bilateral laproscopic orchiopexy and closure of his patent urachus.

**Discussion:** Prune belly syndrome is a rare (1/29,000-40,000) combination of lax abdominal musculature, bilateral cryptorchidism, and dysplastic kidneys/ureters. The etiology of PBS is unclear but the two most prominent theories relate to transient urethral obstruction or a primary mesenchymal defect. Posterior urethral valves are one possible cause of urethral obstruction. However, posterior urethral valves in viable prune-belly patients are extremely uncommon. One postmortem study found that up to 25% of prune-belly patients have posterior urethral valves at autopsy. However, none of these patients survived beyond 31 weeks of gestational age [1]. Manivel et al. identified PUV endoscopically in 4 of 29 patients presenting with PBS [2]. However, the authors admit that the nature of the obstructive lesion was uncertain in those cases. They noted that the obstructive lesion very well could have been hypoplastic prostatic tissue telescoping into the posterior urethra, or so-called type IV PUV. We found a single report of a viable infant with coincident PBS and PUV based on clinical and radiologic findings; however, the diagnosis was never confirmed endoscopically [3]. We postulate that the reason for our patient’s survival with coincident PBS and PUV beyond gestation is likely due to his patent urachus acting as a pop-off valve and preventing fulminate renal failure from the combined insult of a hypocontractile bladder working against a urethral obstruction. Although concomitant PUV and PBS is uncommon, it must be considered in the differential diagnosis in PBS patients presenting with early renal insufficiency.

**References**

Severe Congenital Midureteral Dilatation

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

Discussion: Congenital midureteral stricture is an uncommon cause of antenatal hydronephrosis and no standardized terminology has been proposed. We were unable to find a similar case reported in the literature showing a severe midureteral dilatation with minimal proximal ureteral dilatation and normal distal ureteral diameter [1-3].

Intrauterine ureteritis, extrinsic mechanical compression by blood vessels and inadequate intrauterine ureteral recanalization have been implicated in the etiology of this rare congenital entity [4]. Embryologically, between 37 and 40 days’ gestation, the lumen of the ureter is tran-
The Developing Bladder of Early Childhood (Normal and Pathologic)

Michael E. Mitchell

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The purpose of this presentation is to develop the concept that the bladder in children develops in utero and after birth and that this developmental process is dependent primarily on physical factors (motion and stress forces). Furthermore, we would propose that in utero pathologic development can be reversed if the causative pathologic stress is corrected during neonatal bladder development.

Growth and development, even the evolution of organ systems, are dependent on the response to local environmental stress. For example, in the development of the chick, the environment and, therefore, the developmental processes inside the egg, differs markedly from that of the chick after hatching from the egg, primarily because the environment is radically different. Similarly, human development in utero is dictated by an environment completely different from life outside the womb. Furthermore, these normal stresses and environment drive normal development. However, if stress factors are abnormal, pathologic development may result. The real question is, “can such an abnormally developed bladder change if the physical environment is normalized?”

Phases of Bladder Development

Let us postulate that normally the bladder has three rather unique phases of development dependent on the demands of the local environment. These would be: 1) in utero, a small bladder that has high pressure that is in early developmental phases and differentiation, 2) after birth and the first few months of life, a period of change from in utero structure and growth, and 3) the “mature bladder” which is large and compliant and which facilitates urinary continence and normal voiding. Using polarizing light microscopy Kim, et al. has shown that bladders develop and change in utero and have predictably different histologic characteristics with development[1]. For example, the morphology of the 21 week old bladder wall structure is rather disorganized, however, that of the 38 week old is much more identifiable yet is different from the adult bladder. Furthermore, the physiology (dynamics) of the neonatal bladder is very different than that of the “adult bladder” as has been shown by a number of authors[2-4]. The neonatal bladder has premature voiding contractions, tends to have high voiding pressures and detrusor sphincter discoordination. It has also been shown that with time the bladder capacity will increase and over-activity resolves. These changes are accelerated in the first few months after birth. During this phase shortly after birth, the frequency ofvoidings decreases and bladder capacity increases. It has also been reported that infants do not empty consistently with every voiding, but by age three years bladder emptying with voiding tends to be complete [5-6].

Pathologic States in Early Bladder Development

Pathologic conditions in early development and during the early phases of bladder development are clinically manifest. These can be categorized into three general pathologic states: 1) No urine production, as seen in renal aplasia which results in a small hypoplastic bladder and small thread-like ureters, 2) In utero urinary diversion, as seen in bladder exstrophy/epispadias and bilateral single ectopic ureters, both result in abnormal bladder and urethra development, but have potential for normal ureteric development, 3) Obstruction in utero, as seen in patients with posterior urethral valves, perhaps in Prune Belly Syndrome, and possibly some cloaca malformations in the female. These result in abnormal pressure work by the bladder in utero leading to a trabeculated hypertrophied bladder, hydroureteronephrosis and potential for voiding dysfunction.

The following discussion relates to some clinical observations over the years relating to these pathologic states. State one (no urine ever) can be considered the extreme of state two (diverted). However, in state two, unlike state one, the ureters tend to be normally developed because of the normal production of urine with the physical stimulus of flow and peristalsis resulting in normal development of the ureters. The spectrum of pathology of bladder plate development in state two probably depends on the timing of the original insult (e.g., exstrophy versus cloacal exstrophy). The pathology of state three is almost the opposite of state one and two. State three relates to development in an obstructed state resulting in hypertrophic and hyperplastic changes in the bladder and ureters related increased pressure/volume work. The spectrum of pathology depends on timing of the obstructive process (e.g., cloaca and possibly Prune Belly Syndrome would be associated with the early obstruction vs. classic urethral valves with later obstruction) and degree of obstruction (e.g., “bad” valves versus “mild” valves).

That a newborn with in utero obstruction (state 3) has an abnormal bladder is well accepted by most clinicians. The nature of the bladder wall pathology, furthermore, has been defined well by a number of studies. For example, Kim et al. has observed increased muscle thickness and collagen encasement of muscle bundles in patients with in utero bladder obstruction[7]. They also reported an increase in thick/thin collagen ratio and elastic fibers. Experimentally, in an obstructed neonatal rabbit model, we noted similar histologic changes[8]. Clinically, we have observed that patients with posterior urethral valves with severe bladder changes at birth still have great potential for resolution of the above pathologies simply by correcting the outlet obstruction of the bladder with neonatal valve ablation [9]. Apparently, reducing pressure work of the bladder results in dramatic changes. This is evident by the improved appearance of the bladder (trabeculations and diverticula revert to smooth bladder wall after one year). Furthermore, positive changes in bladder function were measured by significant resolution of vesicoureteral reflux and improved dynamics of the bladder. It is interesting that 92% of early valve ablation patients were potty trained by age four (this was not the case in patients who were diverted). We concluded from these observations that early ablation of valves permitted normalization of bladder function and proximal urinary diversion in boys with valves may ultimately jeopardize the potential for positive “healing” changes in these patients. Apparently, the problem with early diversion is that the pathologic state of in utero obstruction was preserved by diversion after birth, resulting in the loss of the potential for healing in the neonatal phase (phase 2) of bladder development. Patients diverted early in our study tended to have a high potential to develop the “Valve Bladder Syndrome” when undiverted.

Similarly, in the diverted model (state one and two) the neonatal bladder is unquestionably abnormal. The interesting finding is that the patients with exstrophy seem to have a “disorganized and very immature” bladder structure as defined by the early abnormal architecture of the developing bladder in utero [1]. It seems as if the bladder were “frozen” early in utero (phase one) in bladder development. Clinically, neonatal repair (complete primary repair of exstrophy or CPRE) seems to enable normal filling and emptying. In these patients we have observed a rapid improvement of bladder function and anticipate that need (continued on next page)
Prenatal Diagnosis of Megacystis-Microcolon-Intestinal Hypoperistalsis Syndrome

**Introduction:** Megacystis-microcolon-intestinal hypoperistalsis syndrome (MMIHS) is a rare congenital disorder causing a functional/anatomic obstruction of the urinary and gastrointestinal tract. The majority of patients affected die within the first year. Elements to facilitate prenatal diagnosis and postnatal management are reviewed.

**Case Report:** Two female fetuses (denoted as “A” and “B”) were identified in the second trimester with distended bladders, severe hydroureronephrosis and normal amniotic fluid (Figure). Both were precipitously delivered via caesarean section at 30 (A) and 32 (B) weeks due to premature labor with fetal decelerations. Postnatal ultrasound verified dilation of the urinary tract. Each had persistent elevation of serum creatinine to 1.2 mg/dL (A) and 1.1 mg/dL (B) after the first 24 hours of life. Urethral catheter drainage was implemented immediately with eventual decrease in creatinine to 0.6 mg/dL (A) and 0.3 mg/dL (B). Baby A developed an acute abdomen on day 2 of life, requiring exploratory laparotomy, indicating a pencil-thin colon with perforation of the stomach. A vesicostomy was performed to manage the urinary tract. Baby B had cardiac and pulmonary anomalies, which delayed attempts at feeding. Once feedings were attempted, Baby A had recurrent emesis. Barium swallow with small bowel follow-through indicated a malrotation with markedly decreased peristalsis. Exploratory laparotomy was performed to correct the malrotation. The functional bladder obstruction was managed with clean-intermittent catheterization. Each required long-term intravenous parenteral nutrition (TPN) and Baby A is awaiting bowel transplantation.

**Discussion:** Fewer than 100 cases have been reported in the literature concerning MMIHS. The majority of patients on prenatal ultrasound demonstrate an enlarged bladder with hydronephrosis. Amniotic fluid is usually normal to slightly increased. This predominantly affects female patients in a ratio of 4:1. Death during the first year of life is secondary to intestinal obstruction and sequelae of chronic TPN. A high index of suspicion must exist for the female fetus with an enlarged bladder and hydroureronephrosis for MMIHS.

**References**


**Prenatal ultrasound of fetus B with dilated bladder.**
In Utero Spontaneous Intraperitoneal Bladder Rupture Presenting as Urinary Ascites in Two Newborn Females

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Introduction: Antenatal urinary ascites is most commonly due to urethral obstruction from posterior urethral valves and consequently occurs primarily in males. Spontaneous in utero bladder rupture in an otherwise normal female is exceedingly rare. We report two female infants born with massive urinary ascites from a spontaneous in utero intraperitoneal bladder rupture. Both had otherwise normal genitourinary tracts and were managed nonoperatively.

Case 1. A female infant was born at 35 weeks gestational age to a 32-year-old G1 with gestational diabetes but an otherwise uncomplicated pregnancy. The infant was noted to have massive ascites at delivery. Three antenatal ultrasounds (US), one in the third trimester, showed no abnormalities and no problems with amniotic fluid volume. In her third trimester, the mother developed intermittent abdominal pain, prompting hospitalization. Upon admission, she was noted to be severely anemic, requiring 4 units of packed red blood cells. Fetal nonstress testing was normal and reactive. An MRI scan showed blood in the mother’s abdomen, but there were no abnormalities with the fetus. The mother remained on bedrest for 10 days and was hemodynamically stable until recurrence of abdominal pain. A fetal US was performed, this time showing oligohydramnios and massive fetal ascites. The mother underwent emergent caesarean section, where placenta percreta was noted intraoperatively. After delivery, the infant was intubated for respiratory distress and transferred to our institution for further evaluation and management.

Upon arrival, the infant underwent immediate paracentesis of 50 cc of dark yellow fluid. A foley catheter was inserted into the bladder and a 21-gauge angiocath was used as a peritoneal drain. The peritoneal fluid creatinine was 2.9 mg/dL (serum creatinine, 1.3 mg/dL), protein was < 2.5 g/dL, and culture showed no growth. Radiographic evaluation consisted of an abdominal plain film consistent with ascites, and a renal/bladder US showing abdominal ascites, kidneys with normal echotexture and without hydropnephrosis, and a decompressed bladder (Figure). A voiding cystourethrogram (VCUG) showed a small intraperitoneal rupture at the dome of the bladder.

The infant was managed with a foley catheter and peritoneal drain for two weeks. She was extubated on day of life 4 and immediately began to feed and have normal bowel movements. A cystogram performed after 2 weeks of drainage showed no evidence of leak. The foley catheter and peritoneal drain were removed and she voided normally. An US performed 3 days later showed no reaccumulation of ascites and she was discharged home.

Case 2. The second female infant was born at 34 weeks gestational age to a 32-year-old G2, whose pregnancy was complicated by preterm labor and hypertension at 32 weeks. Three antenatal US up to 32 weeks gestational age were normal. On the day prior to delivery, routine fetal nonstress testing was nonreactive. An US showed new onset fetal ascites and an emergency caesarean section was performed. The infant was noted on delivery to have massive ascites and developed respiratory distress, requiring intubation 3 minutes after birth. Paracentesis was performed in the delivery room with a 22-gauge angiocath, aspirating 120 cc of clear yellow fluid. The peritoneal drain and a foley catheter were left to drainage. She was transferred to our institution on day of life 3 for further evaluation and management.

Upon arrival, fluid from the peritoneal drain was analyzed. The fluid creatinine was 3.3 mg/dL (serum creatinine, 1.6 mg/dL) and fluid protein was <3.0 g/dL. A renal US showed normal kidneys and VCUG showed a small bladder rupture on the posterior wall outlining loops of bowel, consistent with an intraperitoneal bladder rupture. After two weeks of catheter drainage, a repeat VCUG showed no further leak. The catheter was removed, followed by removal of the peritoneal drain 4 days later. Serum creatinine on discharge was 0.6 mg/dL. Follow-up US one month later showed normal kidneys without hydronephrosis or reaccumulation of ascites.

Discussion: Antenatal urinary ascites is usually due to urethral obstruction from posterior urethral valves. Bladder rupture in the neonatal period can also present as urinary ascites in the newborn [1]. In utero bladder rupture in females with normal genitourinary tracts is extremely rare. The etiology of the in utero bladder ruptures in our two patients is unknown. To our knowledge, there has been only one other report of a female with a normal urinary tract born with ascites from a spontaneous in utero bladder rupture [2]. She was also managed with paracentesis and a urethral catheter. Although traditional management of intraperitoneal bladder rupture is operative repair, there are reports of successful nonoperative management of isolated intraperitoneal bladder ruptures in children using bladder and peritoneal drainage [3]. We present the rare occurrence of two females with normal genitourinary tracts who were born with urinary ascites following spontaneous in utero bladder rupture. Both were successfully managed nonoperatively.

References
2. McDonald, J. and Murphy, A.V.: Neonatal ascites from spontaneous rupture of the bladder. Archives of Dis in Childhood, 50: 956, 1975

Case 1. Postnatal ultrasound showing decompressed bladder with abdominal ascites. UT: uterus.
Unexpected Pyoureteronephrosis in Neonates with Ureteroceles

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Introduction: Surgical management of ureteroceles has traditionally been dictated by symptomatic presentation, such as UTI. However, due to the increased use of prenatal ultrasounds, the diagnoses of asymptomatic or “incidental” ureteroceles have increased. Thus, recent studies have begun advocating nonoperative or delayed surgical management in select cases of neonatal ureteroceles [1-3]. We present our recent experience with three cases of neonates with ureteroceles who unexpectedly presented for emergency surgery with pyoureteronephrosis despite prophylactic antibiotics.

Case Reports: The three cases consisted of female neonates with a median age of presentation of 16 days (range 14-30 days). One of three was diagnosed prenatally with a ureterocele while in the other neonates, ureteroceles were found on postnatal ultrasound (Table below). All three neonates had ureteroceles associated with duplicated collecting systems. Two had unilateral right-sided duplicated systems while the last child had bilateral duplicated collecting systems and bilateral ureteroceles (Figure). All three were taking prophylactic dose amoxicillin before ultrasound testing was completed. Two cases presented with fevers and negative urine cultures while the last presented with a positive urine culture without clinical symptoms.

After ultrasounds showed pyoureteronephrosis, cystoscopy and transurethral incision of ureteroceles were performed. All three patients were found to have extravesical ureteroceles (submucosal extension into the urethra) and, upon incision, all drained grossly purulent urine. No complications were encountered during or after surgery, and all three were discharged home on postoperative day 1 with a therapeutic course of organism-specific antibiotics.

Discussion: The optimal management of ureteroceles remains controversial. Widespread use of prenatal ultrasound has increased the diagnosis of ureteroceles from 2% to 28% [4]. Whether all of these ureteroceles require surgery remains in question. Less than 25% of neonates diagnosed with ureteroceles present with symptomatic UTI [1]. In a prospective randomized trial, there was no statistical difference in UTI rates between neonates treated with early surgical intervention (less than 2 weeks of age) and those treated conservatively with prophylactic antibiotics until delayed surgical repair (9% versus 8%, respectively) [1]. Recent studies have identified two subsets of patients with ureteroceles who may be candidates for observational management: neonates without high-grade obstruction of functioning upper pole moieties, and neonates with ureteroceles associated with a multicystic dysplastic or nonfunctional upper pole moiety [2,3].

However, as demonstrated in our cases, there is the inherent danger of neonatal sepsis with ureteroceles treated expectantly despite prophylactic antibiotics. Prophylactic antibiotics have been shown to be insufficient for preventing UTI in neonates with ureteroceles [5]. Our three cases of pyoureteronephrosis reflect our experience over the past 2 years (27 cases) for an incidence of 11.1%. Similarly, Weiner has also seen pyoureteronephrosis in 1 of 9 (11.1%) neonates treated for ureteroceles [Weiner JS, personal communication].

The three cases presented in this report reinforce that early surgery is still required in select patients. Furthermore, we hope this report serves as a foundation for further studies that will clarify which patients would benefit from early surgical intervention.

References

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Ultrasound images from patient MW. A) Duplex kidney with hydronephrotic upper pole with echogenic debris suggestive of pus. B) Bilateral ureteroceles with echogenic debris suggestive of pus.

Ultrasound images from patient MW. A) Duplex kidney with hydronephrotic upper pole with echogenic debris suggestive of pus. B) Bilateral ureteroceles with echogenic debris suggestive of pus.
Ureterocele Causing Bladder Outlet Obstruction in a Female Infant

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**Introduction:** We present a case of a female infant with prenatal hydronephrosis who was found postnatally to have bladder outlet obstruction secondary to a left upper pole ureterocele. Bladder outlet obstruction is extremely uncommon in female newborns and has only rarely been reported to result from a ureterocele.

**Case Report:** Our patient was followed prenatally for bilateral hydronephrosis and maintained a normal amniotic fluid index. Postnatally, she was found to have bilateral duplex systems, with a left upper pole ureterocele and right grade V lower pole vescoureteral reflux. A voiding cystourethrogram performed on day of life 7 revealed a markedly trabeculated bladder with a ureterocele at the bladder neck (Figure). She underwent ureterocele puncture at 11 days of life with resolution of the bladder outlet obstruction. Because of breakthrough urinary tract infection with persistent severe reflux on the contralateral side, bilateral ureteral reimplantation with excision of the left ureterocele and right lower to upper end-to-side ureteroureterostomy was performed at age 7 1/2 months.

**Discussion:** The first report of a ureterocele causing prenatal bladder outlet obstruction was published by Gloor et al. in 1996 [1]. Seven additional cases have been reported, 4 of which were treated in utero [2-7]. Two of the 4 patients treated postnatally ultimately required nephroureterectomy after 4 and 13 months of follow-up [2], as did 1 of the 4 treated prenatally [5]. The other 2 patients treated postnatally who did not have nephrectomy had follow-up of only 18 days and 5 months [1,3]. Follow-up in the patients treated prenatally who did not undergo nephrectomy was less than 2 months in 2 [6,7] and 5 months in 1 [4].

Ureteroceles causing prenatal bladder outlet obstruction are rare and have been reported to cause significant urologic morbidity. However, no reports of pulmonary or other complications have been published. This raises the question of whether there is really a benefit to intervening on these patients prenatally. Prenatal intervention can have serious complications and should only be performed after thoughtful consideration and meeting the criteria of the SFU. Reports of long-term outcomes in patients treated both prenatally and postnatally are needed.

**References**

Asynchronous Bilateral Renal Venous Thrombosis with Subsequent Development of Cystic Kidneys

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**Introduction:** We discuss a case of asynchronous bilateral renal venous thrombosis.

**Case Report:** A 6-day-old, 34-week fraternal twin boy was delivered via Cesarean section to a mother with a history of Group B streptococcal infection, pregnancy induced hypertension, and Grave’s disease. A left flank mass was palpated at birth and abdominal ultrasound findings were consistent with left renal venous thrombosis (RVT). Hematuria and thrombocytopenia of 43,000/cmm were subsequently observed. The serum creatinine concentration (S Cr) decreased from 1.5 to 0.8 mg/dL with intravenous volume expansion. However, on the day of transfer to our institution, the infant had repeat gross hematuria and a new right flank mass. The S Cr was 2.3 mg/dL and the platelet count was 62,000/cmm. A repeat ultrasound revealed bilateral renal venous thrombosis and a left adrenal hemorrhage. MRI showed multiple areas of intracerebral bleeding. The S Cr peaked at 4.7 mg/dL on day 8 of admission to our institution and began to normalize with intravenous fluids and supportive care.

The patient’s S Cr nadir was 0.7 mg/dL at 8 months of age. His most recent estimated glomerular filtration rate was 67 ml/min/1.73m². A renal ultrasound at 8 months of age showed bilateral corticomedullary cysts. He has since become hypertensive and is being treated with enalapril.

**Discussion:** Renal venous thrombosis usually occurs in neonates. Renal vein thrombosis can occur in older children, usually in association with nephrotic syndrome. The development of asynchronous bilateral renal venous thrombosis is unusual. In our patient, evaluations of known risk factors for RVT revealed a low level of antithrombin antigen at 58% (normal 82–136%) during hospitalization. Whether the decreased level was the result or cause of the disease process is unknown. Our patient also had a persistently unexplained elevation of the hemoglobin concentration, at 14.9 g/dL. The triad of flank mass, gross hematuria, and thrombocytopenia was present at both RVT occurrences. Treatment consisted of intravenous fluids. Thrombolytic treatment was contraindicated because of the cerebral hemorrhages. There are no reported deaths attributable to RVT regardless of management. However, about 30% of children may have progressive renal failure and usually late-onset hypertension. Our patient’s slightly elevated S Cr, the development of hypertension, and the ultrasound findings are poor prognostic signs for progressive renal dysfunction. The precise histopathological characterization of the cystic kidney disease in the context of renal venous thrombosis remains unexplained. Most of these patients have small contracted kidneys but cysts have, to our knowledge, not been reported in this context.
Prenatal Evaluation and Treatment of Fetal Lower Urinary Tract Obstruction

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Fetal lower urinary tract obstruction (LUTO) due to obstruction of the proximal urethra leads to progressive oligohydramnios, physical deformations, pulmonary hypoplasia, and renal fibrocystic dysplasia. When LUTO is associated with anhydramnios prior to 22 weeks gestation, it should be considered a lethal congenital anomaly. Experimental work using a number of animal models has suggested that early prenatal treatment using diverting shunts may prevent pulmonary hypoplasia and potentially preserve renal function. However, interventions in the setting of existing irreversible renal dysplasia would obviously not be beneficial, and, therefore, patient selection for intervention is critical. The present approach to prenatal evaluation of fetuses with LUTO involves detailed high-resolution sonographic evaluation to rule out associated anomalies and fetal karyotype analysis given the 6 to 8% incidence of aneuploidy in this population and to confirm male karyotype, and renal function evaluation using serial urine chemistries from fine needle bladder drainage. Sonographic evaluation is hampered by the absence of amniotic fluid and, therefore, we routinely perform amniinfusion to reestablish normal amniotic fluid volume and the fluid-tissue interface allowing better sonographic imaging. Karyotype analysis is also important to identify female fetuses, as they have not been shown to benefit from shunting given the different etiologies for obstruction, and, therefore, should not be offered this intervention. Fetal urine is analyzed for a combination of electrolyte and protein markers that have been shown histologically to correlate with the degree of fibrocystic damage in the kidneys between 16 and 24 weeks gestational age. It is important to observe a pattern and decreasing hypertonicity with each serial bladder drainage, and sodium values of < 90 mEq/L, calcium < 80 mMol/L, calcium < 7 mg/dL, osmolality < 180 mOsm/L, beta-2 microglobulin < 6mg/L, and total protein <20 mg/dL have been shown to be associated with minimal renal histologic changes, and potential for renal salvage with successful shunt intervention.

Fetuses between 16 and 24 weeks gestation that have no associated anomalies that might impact on postnatal survival, have a normal male karyotype, and urine profile studies suggesting the potential for renal salvage represent appropriate candidates to counsel about in utero therapy. Traditionally, therapy has involved placement of a double-pigtail vesicoamniotic shunt into the distended, obstructed bladder. Unfortunately, shunt displacement is common and occurs in 40% of cases. This is either due to improper placement of the shunt within the bladder, or to physical displacement due to the intra-amniotic segment becoming entangled in the fetal extremities and subsequently pulled out of the bladder. In such cases, the shunt can be replaced once the defect in the bladder has closed and the bladder re-enlarged providing an adequate target, which usually takes 10 to 14 days after initial shunt displacement.

In our experience with intervention in carefully selected cases, neonatal survival has reached 90% primarily due to the prevention of pulmonary hypoplasia by prenatally reestablishing the amniotic fluid space during the critical developmental period of lung growth and differentiation. Neonatal renal function at 3-4 months of age correlate with prenatal urine values, and subsequent worsening of renal status in many cases seems to be related to urinary tract dysfunction, reflux, recurrent infections, and complications of surgical procedures. In a recent publication [1] we were able to obtain long-term follow-up in 23 children treated prenatally with vesicoamniotic shunts. The mean age of delivery was 35 weeks, mean birth weight was 2573 gms, and mean interval from shunt placement to delivery was 84.4 days. Two of these infants died shortly after birth from complications of respiratory insufficiency. The mean age at follow-up in survivors was 5.8 years. All children had postnatal confirmation of the etiology of their obstruction, with 39% having posterior urethral valves, 22% with urethral atresia, and 39% with Triad syndrome due to the mid-urethral hypoplasia. Fifty percent of these children were less than the 25th percentile for age in their height and weight. Over half were on special diets to promote growth. Fifty-five percent had normal pulmonary function, while 40% had asthma, 28% had recurrent pulmonary infections and 10% were described as having sleep apnea. Acceptable renal function was defined as a creatinine clearance of greater than 70 ml/min. Using this definition, 45% of children had acceptable renal function, 22% had mild renal insufficiency, and 33% had end-stage renal disease requiring dialysis and subsequently undergone transplantation. Two thirds of the children were able to spontaneously void, while the remainder required intermittent catheterization or were catheter dependent. Based on these results, families carrying a pregnancy with fetal LUTO and anhydramnios can be counseled that if they meet selection criteria, successful prenatal shunt placement has resulted in a 90% survival in this otherwise lethal disorder, with nearly half of children having normal renal function and two thirds having normal bladder function. We also found that there was a correlation between the type of obstruction and subsequent renal and bladder function. We found that 50% of children with urethral atresia had been transplanted at an early age, and 75% had normal bladder function. Forty-three percent of children with Triad syndrome had been transplanted, and 43% had normal bladder function. More surprisingly, only 14% of children with posterior urethral valves had been transplanted, and 72% had normal bladder function.

The primary limitation to prenatal shunt therapy is the high incidence of shunt displacement. To circumvent the physical limitations of shunt therapy, we are investigating the use of fetal microcystourethroscopy to define the etiology of obstruction and allow more etiology-specific counseling, and to identify cases of posterior urethral valves that may be treated primarily with in utero laser resection. Such surgical intervention carries the potential to not only destroy the source of obstruction, but may also allow the bladder to cycle and potentially improve postnatal bladder development and function.

References
Antenatally Diagnosed Bilateral Ureteropelvic Junction Obstructions Presenting with Sudden Anuria

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Introduction: We discuss a female neonate with antenatally diagnosed bilateral hydronephrosis and appropriate postnatal follow-up, who presented at six weeks of life with sudden onset of anuria secondary to bilateral ureteropelvic junction (UPJ) obstructions.

Case Report: Bilateral hydronephrosis was detected in one female neonate of a twin gestation at 20 weeks to a 34-year-old P2G1 mother. Hydronephrosis increased slightly during the course of pregnancy with maximal dilation to 18 mm on the right side and 9 mm on the left side. No oligohydramnios or bladder distension was noted. Upon delivery at 36 weeks the physical examination, blood urea nitrogen (BUN), creatinine (Cr), and a VCUG were normal. Postnatal ultrasound on day 3 revealed SFU grade 1 on the left side and grade 3-4 on the right side, consistent with moderate bilateral UPJ obstructions. A renal scan was scheduled and the infant was maintained on antibiotic prophylaxis.

At 6 weeks of life she presented to the emergency treatment center at her local hospital with abdominal distention. Anuria was noted, and ultrasound confirmed worsening bilateral pelvicaliectasis. Creatinine and K+ were elevated to 2.0 mg/dL and 8mEq/L, respectively. Following transfer to our institution, emergent bilateral percutaneous nephrostomies were performed one week later with intraoperative verification of narrowed segments at both UPJs. After removal of the indwelling ureteral stents 6 weeks postoperatively, the patient remains asymptomatic with normal Cr and markedly improved ultrasound imaging 8 months after surgery.

Discussion: This is a highly unusual case of bilateral symptomatic UPJ obstruction in a neonate. Even with severe hydronephrosis many kidneys show substantial improvement in morphology and function remains stable [1,2]. A general algorithm for the management of postnatal hydronephrosis has evolved and seems to be appropriate and safe in preserving renal function [3]. But this case offers several points to consider. First, most patients with prenatally diagnosed hydronephrosis are assumed to be asymptomatic. The dramatic presentation at the emergency treatment center came about because of pronounced abdominal distention. The question then arises: how many patients assumed to be asymptomatic might be suffering from intermittent symptoms? Second, traditional thinking has suggested that in neonates, intrinsic narrowing is the primary pathogenesis, and a kinking UPJ obstruction is more likely to be seen in the older patient. This case highlights that kinking UPJ obstructions on an intermittent basis may occur in the neonatal setting. Finally, management algorithms have been developed in situations where unilateral hydronephrosis predominates, but for bilateral cases, more frequent observation seems warranted. Documenting rare cases such as the one presented here are important for development of safer approaches for the management of antenatally diagnosed hydronephrosis. While most of these cases remain asymptomatic, our case serves as a reminder that newborn hydronephrosis initially managed nonoperatively. J Urol 2002; 168: 1118-1120.

References

Pre- and Postnatal Management of a Unilateral Ureterocele Associated with Bilateral Hydronephrosis and Oligohydramnios

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Introduction: Approximately 75% of ureteroceles are diagnosed on prenatal ultrasound [1]. Often, the fetus is managed expectantly, with incision of the ureterocele after birth. When antenatal testing suggests that the condition is causing severe bilateral obstruction, fetal decompression of the ureterocele via laser, puncture, or placement of a vesicoamniotic shunt has been implemented [2-4]. Another option is preterm delivery. Five criteria have been suggested for antenatal intervention: 1) prolonged oligohydramnios, 2) renal cortical cysts, 3) urinary Na >100 mEq/L, Cl >90 mEq/L and Uosm>210 mOsm/L, or 4) reduced lung area or thoracic or abdominal circumference; 5) gestational age <32 weeks [5]. We detail a case of antenatal and postnatal management of a patient with a left ureterocele causing bilateral hydronephrosis.

Case Report: A 28-week gestational fetus was noted to have a large unilateral ureterocele, duplicated right hydrourter, and left hydronephrosis on routine ultrasonographic (US) evaluation (Figure). Although a (continued on next page)
US 10 days prior revealed normohydramnios, the fetus had progressively become oligo-, then anhydramiotic. The mother, a 32-year-old G1, opted for vesicocentesis. Multiple punctures were made in the ureterocele under US guidance. The fetus and mother tolerated the procedure well and had normalization of the amnio-fetal index and decompression of the ureterocele until 37 weeks, when she delivered vaginally without complication. A full description of this procedure and immediate postnatal results has been published previously [6].

Birth weight was 3,281 grams and Apgar scores were 8 and 9. US done immediately after delivery demonstrated left lower pole hydronephrosis, a decompressed ureterocele, and right hydronephrosis. The patient voided on the first day of life. Antibiotic prophylaxis was started immediately. She was discharged home on day of life 2 without incident.

The patient was seen in follow-up at 1 week of life. US at that time demonstrated similar findings as one week prior. VCUG demonstrated grade V reflux into the lower pole moiety on the left. No right-sided reflux was noted. At 5 weeks, serum creatinine (SCR) was 0.6 mg/dl and a diuretic renal scan demonstrated left-sided differential function of 28% with a T1/2 of 39.5 minutes. The right side demonstrated a normal washout curve despite hydronephrosis seen on US. The patient was maintained on antibiotic prophylaxis and closely followed with SCr, renal US and VCUGs for the next 18 months. No breakthrough infections occurred during this period.

At age 19 months, a VCUG demonstrated persistent grade V reflux in the left lower pole. Renal US revealed residual right collecting system fullness, left lower pole hydronephrosis and no upper pole hydronephrosis. The collapsed ureterocele was evident in the bladder. A diuretic renal scan demonstrated a differential function on the left of 26% with no obstruction on either side. Given the failure of the reflux to improve, surgery was recommended at this time. At 23 months, the patient underwent excision of ureterocele and left ureteral reimplantation. The patient was discharged home on postoperative day 2 without complication. Follow-up is ongoing.

References

Isolated Ascites in a Female Fetus with Congenital Adrenal Hyperplasia

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Case Report: A 36-year-old woman at 22 weeks’ gestation was found to have a female fetus with significant ascites. Amniotic fluid was normal and TORCH antibodies were negative. Fetal anatomy was unremarkable. Ascites was confirmed at 31 weeks with significant elevation of the diaphragm, prompting elective C-section. A 2285 gm female newborn was delivered. Initial examination revealed a massively distended abdomen which was compromising respiratory function and required mechanical ventilation. 2400 ml of ascitic fluid was drained from the abdominal cavity. A distended bladder became evident on day of life 2, with progressive elevation of serum creatinine (SCR) levels. A Foley catheter was positioned without difficulty and ultrasound demonstrated bilateral hydronechoic kidneys with ureteral dilatation. Over the following days, an incremental diuresis was observed with progressive normalization of SCr levels, resolution of ureteral dilatation and no reaccumulation of ascites. Extubation occurred on day of life 20. An attempt to remove the bladder catheter failed since the bladder became distended again, and the catheter had to be reinserted. A voiding cystogram was then performed, which demonstrated a short (1 cm) urogenital sinus (UGS) with the catheter at the junction between the bladder and vagina. UGS was confirmed with cystoscopy, and vesicostomy was carried out during the same operative session. Serum 17-OH progesterone, testosterone and testosterone precursors were all elevated and the diagnosis of congenital adrenal hyperplasia was established. Urinary electrolytes were within normal limits. The baby is presently on corticosteroid treatment awaiting surgical reconstruction.

Discussion: These findings suggest that fetal ascites might be of urinary origin, with passage of urine from vagina into peritoneum via fallopian tubes. Even in the presence of normal fetal anatomy, persistent ascites may be the expression of UGS/cloacal disorders and such issue should be incorporated into antenatal counselling.