SPECIAL EDITION: Society for Fetal Urology: Year in Review - Part II

INSIDE THIS ISSUE:

- Ovotesticular Disorder of Sexual Development Secondary to Chimerism
- Posterior Urethral Valves: Is Early Delivery of Any Benefit?
- Juvenile Granulosa Cell Tumor in a Newborn with an Undescended Testicle
- Complete Sex Reversal in Disorders of Adrenal Steroidogenesis: Consideration for the Phenotypic Sex of Rearing
- Prenatal Detection of a Solid Renal Mass
- Bilateral Ureteropelvic Junction Obstruction in Crossed Renal Ectopia with Fusion
- Prenatal Diagnosis of a Paraurethral Cyst
- Clitoral Mass in a Prepubescent Female
- CEVL Responds to the AAP Report On Male Circumcision
- Serial Fetal Renal Aspirations for Prenatal Hydronephrosis: Failure due to Gastric Outlet Obstruction?
- An Atypical Presentation of Multicystic Dysplastic Kidney
- Pre-sacral Teratoma as the Cause of Neonatal Bladder Outlet Obstruction
- Vaginal Atresia in a Two Year Old Presenting with Urinary Retention
- Is There a Role for Fetal Intervention in the Management of Ureteroceles?
- Deforming Penoscrotal Lesions in a 9 Year Old
- An Uncommon Cause of Fetal Urethral Obstruction
Ovotesticular Disorder of Sexual Development Secondary to Chimerism

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Case Presentation
A 3 year-old child being raised male and recently adopted from overseas was referred for ambiguous genitalia. Examination revealed dorsal-hooded prepuce with penoscrotal hypospadias, ventral chordee, bifid scrotum and a palpable right gonad. He was also noted to have whorled areas of hyperpigmented skin over the back and lower extremities. Work-up included pelvic ultrasound, which demonstrated a uterus and right testicle with microlithiasis. Karyotype revealed a normal 46,XY male karyotype in all 50 cells examined. Follicle-stimulating hormone, luteinizing hormone and testosterone were all within normal range for a prepubescent male.

The patient underwent bilateral gonadal exploration, right testicular biopsy with orchidopexy, bilateral inguinal hernia repair, multiple skin biopsies and first stage hypospadias repair with transfer of preputial skin. The right gonad appeared to have both ovarian and testicular tissue, with the ovarian tissue located superiorly (figure 1). Testicular tissue was biopsied but preserved. The left ovary and fallopian tube were removed due to discordance with the sex of rearing.

Figure 1. Right gonadal exploration. Ovarian tissue located superiorly with testicular tissue inferiorly.

Six tissue samples including bilateral gonadal and skin biopsies were submitted for pathologic and chromosomal analysis. The left gonad revealed ovarian and fallopian tissue. The right gonad demonstrated both ovarian and testicular tissue (figure 2). All samples revealed two cell lines; 46,XY and 46,XX consistent with chimerism. The percentage of male versus female cells in each tissue sample varied.

The child is currently followed by Genetics and Endocrinology as well as Pediatric Urology. He will undergo a second stage hypospadias repair 6 months from his initial surgery.

Discussion
True hermaphroditism, now referred to as ovotesticular disorder of sexual development (ODSD), is a rare disorder of sexual differentiation in which the external genitalia are ambiguous, and the gonads have both testicular tissue with seminiferous tubules and ovarian tissue with primordial follicles.1 Approximately 10-30% of patients with ODSD have a 46,XX/46,XY karyotype, which can result from either mosaicism or chimerism. Chimerism is an uncommon etiology, and refers to the presence of two genetically distinct cell lines occurring in one individual as a result of fusion of two separate conceptions or dispermic fertilization. Chimerism is clinically distinct from mosaicism, which is the presence of two genetically distinct cell lines resulting from a single zygote.2

Management of ODSD requires a multi-disciplinary approach and is dependent upon age at diagnosis, genital development, internal structures and reproductive capacity as well as preference of well-informed parents. Discordant gonadal tissue should be removed.1 Incidence of gonadoblastoma is less than 5% in ovotestes, but remains a consideration in children with ODSD. Management of a histologically normal, scrotally positioned testis requires careful surveillance.1 Leaving the testis in situ allows for potential endocrine function at puberty. Future fertility is another issue facing males with ODSD. Immature testicular cryopreservation in prepubertal boys remains experimental and involves extraction of immature gamates for subsequent in vitro maturation. There may also be a role for intracytoplasmic sperm injection, successful pregnancy has been achieved with a chimeric 46,XX/46,XY male using this technique.3

References
Posterior Urethral Valves: Is Early Delivery of Any Benefit?

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Case Presentation

The patient is a product of a 37-week gestation pregnancy delivered by cesarean section with the pregnancy complicated by severe oligohydramnios and suspected posterior urethral valves. This patient was followed prenatally with ultrasounds (PNUS) through the obstetrics and gynecology department’s telemedicine program. The 20 week PNUS showed the fetus to have bilateral hydronephrosis, a thickened bladder wall, and normal amniotic fluid index (AFI). PNUS at 23 weeks showed ascites, decrease in degree of hydronephrosis, and normal AFI. By the 24 week PNUS, the ascites had subsided and severe hydronephrosis had resumed with normal AFI. The Urology team was consulted at 30 weeks and recommended repeat PNUS every two weeks to observe for changes in AFI and character of the fetal kidneys. The baby was delivered at 36 weeks due to severe oligohydramnios. Upon an uncomplicated cesarean section, the patient was transferred to Arkansas Children’s Hospital for emergent renal ultrasound, foley catheter placement, and voiding cystourethrogram (VCUG). Renal ultrasound revealed bilateral Grade 3 to 4 hydroureteronephrosis, and a thick, trabeculated bladder wall, and contrast voiding cystourethrogram confirmed posterior urethral valves. Cystoscopy with incision of type 1 posterior urethral valves was performed with an uncomplicated postoperative course. His creatinine peaked at 2.1 mg/dL by day of life 3 and improved to 1.7 mg/dL by day of discharge and a nadir of 0.5 mg/dL two months later.

Discussion

Posterior urethral valves (PUV) occurs in 1 in 8000-25000 live births, is the third most common antenatal genitourinary diagnosis, and accounts for 10% of all fetal uropathy.1) PUV can be a devastating anomaly with sequelae including pulmonary hypoplasia, dysplasia, renal tubular injury, and extensive bladder problems. Since the addition of prenatal ultrasonography, PUV diagnosis has been made in 1 in 1250 screenings, although prenatal diagnosis appears to have questionable impact on the prognosis of renal function. The classic ultrasound findings include bilateral hydroureteronephrosis, distended bladder, dilated posterior urethra, thickened bladder wall and the “keyhole” sign.1 Several perinatal interventions including vesicoamniontomic shunting, serial ultrasound-directed vesicocectomy, fetal cystoscopy and valve ablation have been discussed in the literature in animal models and humans. However, no consistent benefit was observed in the human fetuses to preserve normal renal function.1)

There is no data available indicating whether early delivery during third trimester could potentially be advantageous. Noe et al. reported one rare case for early delivery in a newborn who had bilateral UPJ obstruction and severe oligohydramnios delivered at 32 weeks with percutaneous nephrostomy drainage, bilateral pyeloplasties at one month, and normal creatinine at three months.3)

With significant efforts directed toward intervention during the second trimester and known long term impact upon bladder and renal function in the setting of PUV, we postulate that there may be potential benefit to bladder and renal function by early delivery in the third trimester. The question in this clinical situation is: would the benefits outweigh the risks of early delivery for the fetus? We recommend further consideration of this question in future study of PUV.

References

Juvenile Granulosa Cell Tumor in a Newborn with an Undescended Testicle

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Case Presentation

A two week old infant was referred for Urologic evaluation with right cryptorchidism. He was born at 39 WGA via cesarean section and the right testicle was not initially palpable. Interestingly, an antenatal CT was performed for maternal abdominal pain. Retrospective review did not review any testicular pathology. Ultrasound revealed a size discrepancy between the testicles with the right side measuring 70% larger than the left. He was treated with antibiotics for possible orchitis which failed to resolve the size discrepancy. At this time, his tumor markers were obtained: AFP (>35,350), beta human chorionic gonadotropin (â-hCG) (<1.2), leucocyte dehydrogenase (LDH) (301). Retrospective review of an antenatal CT scan did not demonstrate any testicular pathology.

At one month of age his right testicle had descended spontaneously. It continued to be larger than the left but without palpable mass. Repeat ultrasound revealed heterogeneity of the right testicle, but no definitive mass was noted. Serial ultrasounds continued to demonstrate the size discrepancy without a discrete mass. Serial AFP levels were obtained which failed to decline in the expected fashion.

Due to the size discrepancy between the testicles as well as failure of decline of the AFP level, there was concern about a testicular tumor. The patient then underwent inguinal exploration at 3 months of age. Intra-operatively, no discrete mass could be identified. A frozen specimen was performed during the procedure that was initially read as benign testicular tissue with focal tubule dilation. The specimen was further examined which revealed stromal proliferation with ductal dilation filled with mucinous material (Fig. 1). The specimen demonstrated positive staining for inhibin (Fig. 2) and CD99 and negative for AFP. The final pathologic diagnosis revealed PT1 juvenile granulosa cell tumor with negative surgical margins.

The patient was then followed clinically and the AFP values progressively declined.

**AFP Levels:**

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<td>3 months of age</td>
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<tr>
<td>18 months of age</td>
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**Discussion**

JGCT usually presents as a painless, cystic mass that is hormonally inactive. To date only about 50 cases have been described in the literature since its recognition in 1983. 90% of the cases have been diagnoses in children less than 1 year of age. Up to 25% of these patients also have an associated chromosomal abnormality. To date there have been no reports of metastases with JGCT. No follow up is currently recommended for these patients.

**References**

Complete Sex Reversal in Disorders of Adrenal Steroidogenesis: Consideration for the Phenotypic Sex of Rearing

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Case Presentation

Inborn errors of adrenal biosynthesis can present with severe metabolic abnormalities as well as significant variations in genital ambiguity. We present two cases of complete sex reversal due to mutations in adrenal hormone metabolism with opposite clinical presentations, in which both families chose the phenotypic sex of rearing. The first patient was a phenotypic male born at term without complication at an outside facility. The antenatal history was unremarkable and prenatal ultrasound identified the fetal sex as male. The neonatal genital exam demonstrated a phallic structure with foreskin, an orthotopic urethral meatus, scrotalized perineal skin, and bilateral non-palpable gonads (Fig. 1). A neonatal circumcision was performed, and the patient was discharged home. On routine neonatal screening, the patient had an elevated 17-hydroxyprogesterone, suggesting a diagnosis of congenital adrenal hyperplasia with complete virilization. The result of the karyotype was 46 XX. An ultrasound demonstrated the presence of Müllerian structures and enlarged adrenal glands, and a voiding cystourethrogram (VCUG) demonstrated a urogenital sinus. A MRI of the pelvis confirmed the presence of a fully developed phallus with corpora cavernosa and suggested the presence of a prostate.

The second patient was a phenotypic female born at term without complication to consanguineous parents of Middle Eastern descent. The patient was admitted to the NICU for meconium aspiration. On day of life 10, the newborn was found to have hyponatremia, hyperkalemia, and hypoglycemia. Adequate resuscitation was achieved with steroid administration. The neonatal genital exam was typical for a female—labial folds, an orthotopic urethral meatus, and a vaginal introitus were present (Fig. 2). However, gonads were palpable bilaterally within the inguinal canal. Subsequent labwork identified low testosterone, low 17-hydroxyprogesterone, low DHEA, low androstenedione, and elevated ACTH levels suggesting adrenal insufficiency. The result of the karyotype was 46 XY. An ultrasound confirmed the absence of Müllerian structures and the presence of bilateral gonads in the inguinal canal. There was no enlargement of the adrenal glands and no renal abnormalities. A genitogram showed a rudimentary vagina. Genetic testing revealed a homozygous variant of congenital lipoid adrenal disorder with steroidogenic acute regulatory protein (StAR) deficiency. Bilateral gonadectomy was performed at six months of age, and pathologic analysis confirmed the presence of only testicular tissue.

In both cases, there was a multi-disciplinary conference to discuss the sex of rearing including consideration of prenatal androgen imprinting, fertility potential, risk of gender identity disorder, and need for long-term hormone replacement and surgical intervention. Both families elected to maintain the phenotypic sex. The children are currently managed with steroid replacement and meeting appropriate developmental milestones.

Conclusion

Genotypic and phenotypic sex, societal pressure, personal and cultural beliefs, prenatal imprinting, and physician influence can all be contributing factors for families to determine sex assignment. These two cases support the consideration for the phenotypic sex of rearing in complete sex reversal due to congenital adrenal disorders.

References

Prenatal Detection of a Solid Renal Mass

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Case Presentation

A 31 year-old G3P2 healthy female presented to her obstetrician for prenatal care. A routine prenatal sonogram was performed at 21 weeks gestation. The kidneys and amniotic fluid level were normal, however, placenta previa was noted prompting subsequent prenatal imaging. Fetal sonography at 30 weeks revealed normal kidneys, normal amniotic fluid level, and a low lying placenta. At 36 weeks gestation, mild polyhydramnios (amniotic fluid index 27.1) and a new 5.7 x 5.2 x 5.1 cm right renal mass were visualized. This mass was noted to be heterogeneous, encapsulated, well-vascularized, and associated with displacement of normal renal parenchyma medially (Fig. 1). The right adrenal, left adrenal, and left kidney were normal.

Congenital mesoblastic nephroma was the presumed diagnosis. Continuation of pregnancy with careful monitoring and vaginal delivery, if desired and safe, was recommended. Additionally, newborn serum chemistry and blood pressure evaluation were recommended. Induction was performed at 39 weeks and a male infant was born via vaginal delivery.

Neonatal exam was normal except for a palpable right renal mass. Blood pressure was normal. Serum calcium level was slightly elevated at 11 mg/dL. The infant was transferred to our pediatric hospital on DOL #2 where repeat ultrasound revealed a 6.6 x 5.2 x 4.4cm well circumscribed, heterogeneous mass arising from the right kidney. The child was taken to the operating room on DOL#3 where he underwent an uncomplicated right open nephrectomy via a subcostal incision. Pathology demonstrated a firm, yellow renal mass with displacement of normal renal parenchyma medially (fig. 2). Serum calcium on POD #1 was 9.1 mg/dL. The patient was discharged home on POD #3 following an uncomplicated hospital stay. Pathology revealed congenital mesoblastic nephroma, classic type.

Discussion

Sonographic detection of renal masses in the fetus is unusual, especially given the low incidence of renal tumors in infants. Congenital mesoblastic nephroma represents only 2-4% of pediatric renal tumors, but it is the most common solid renal tumor in infants and it is the most common renal tumor diagnosed on prenatal sonography.1 The primary differential diagnosis for congenital mesoblastic nephroma are Wilms tumor and malignant renal rhabdoid tumor. The male to female ratio is 2:1. The three recognized cellular variants are classic (24%), cellular (66%) and mixed (10%).1 Survival is 100% for the classic variant and 85% for the cellular variant.

When evaluating the fetus with presumed congenital mesoblastic nephroma, it is important to recognize that the pregnancy can be complicated by polyhydramnios, hypertension, premature delivery, and hydrops fetalis.2 While the most common presentation postnatally is a palpable, unilateral abdominal mass, infants can present with hematuria, hypercalcemia, hypertension, and hyperreninemia.3 Treatment is nephrectomy with wide surgical margins as the tumor can be infiltrative.3 Thereafter, patients can be carefully followed for one year after surgery with repeat sonograms, especially when pathology reveals the cellular or mixed variant. Local recurrence is uncommon(<5%) and the tumor rarely can metastasize to the brain, bone, liver, lungs and heart.1

References

Bilateral Ureteropelvic Junction Obstruction in Crossed Renal Ectopia with Fusion

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Introduction
Crossed renal ectopia with fusion is a relatively rare congenital anomaly with obstructive uropathy more commonly associated with the inferior crossed renal unit. We present a rare case of bilateral ureteropelvic junction obstruction in a patient with a left to right crossed fusion anomaly. Despite early bilateral repair at 18 days of age, the renal cortex remains highly echogenic on ultrasound, which may bode poorly for long term recovery of renal function.

Case Presentation
A male born at 36 weeks gestational age (GA) was found to have an empty left renal fossa with right hydronephrosis and a cystic structure in the right pelvis on maternal prenatal ultrasound (US) done at 21 weeks GA. The fetus was noted to have a crossed fused renal ectopia (left to right) with bilateral severe hydronephrosis on follow-up fetal MRI at 24 weeks GA. Serial US imaging every two weeks revealed increased echotexture of both kidneys, with normal amniotic fluid indices (AFI).

Ultrasound imaging at 36 weeks GA demonstrated acute oligohydramnios (AFI <2.5cm) requiring urgent C-section. A male infant with birth weight of 2.55kg and APGAR scores of 7/8 was delivered, and he voided spontaneously within hours.

Serial US at two weeks of age showed worsening hydronephrosis of both the orthotopic right kidney and the lower ectopic left kidney, with cortical thinning bilaterally. VCUG confirmed a normal male urethra and no vesicoureteral reflux. Progressive hydronephrosis caused increased abdominal girth, tachypnea and feeding difficulty.

At 18 days of age, the infant underwent bilateral retrograde ureteropyelography, which revealed normal caliber ureters with severe bilateral hydronephrosis secondary to ureteropelvic junction obstruction. Dismembered pyeloplasty of both renal units was performed via single 2.5cm flank incision, with decompression of bilateral renal pelves.

Follow-up renal ultrasound, post stent removal, showed moderate hydronephrosis and persistent echogenicity of renal parenchyma. Postoperative serum creatinine peaked at 1.28 with nadir of 0.56 less than 2 weeks after pyeloplasty.

Discussion
While the cause of crossed renal ectopia with fusion remains unknown, the final configuration is related to the time at which the metanephric masses come in contact during cranial renal migration. Once fusion occurs, rotation of the renal units ceases, but the fused units continue to migrate cranially. Approximately ninety percent of crossed ectopic kidneys are fused with the contralateral kidney, and incidence has been estimated at 1 in 1000-2000 with male predominance and slight left to right predilection.

Although the incidence of hydronephrosis in simple renal ectopia is over 50%, that of crossed renal ectopia with fusion is less well defined. Autopsy incidence of crossed fused ectopia has been reported as high as 1 in 1000, with most being asymptomatic. This case represents a rare occurrence of bilateral obstructive uropathy in crossed fused ectopia. It further exemplifies that bilateral obstruction in early renal embryogenesis, despite early post-natal intervention, may have long-term adverse renal implications.

References
Case Presentation

A 36-year-old nulligravida with dichorionic twins conceived via in vitro fertilization was transferred to our institution at 29 weeks gestation with preterm labor and preeclampsia. Her care to date had been uncomplicated including normal anatomic surveys and normal monthly sonograms of both fetuses.

Upon admission an ultrasound demonstrated normal growth of both fetuses, one male (twin A) and one female (twin B). A 1.0 x 1.0 x 1.3 cm simple cyst was noted at the vaginal introitus of twin B with normal appearing labia bilaterally. There was no prominent vascular flow within the cyst or cyst wall noted on color Doppler evaluation. The cyst did not appear to communicate with the bladder, uterus or gastrointestinal (GI) tract, and no other anomaly or obstruction was visualized. The pregnancy was managed expectantly.

At 32 weeks gestation the cyst measured 2.0 x 1.6 x 1.9 cm without evidence of nearby communication to the bladder or anus (Fig. 1). Fetal growth of both twins was appropriate with a normal amniotic fluid volume.

At 33 weeks, labor was induced for severe preeclampsia with an uncomplicated vaginal delivery of cephalic/cephalic twins. Twin B (the female) weighed 1990 grams with Apgar scores of 5 and 8 at one and five minutes respectively. A cystic structure bulging from the vaginal introitus was seen without evidence of any other abnormality. The neonate was transferred to the neonatal intensive care unit secondary to prematurity.

On examination the labia and clitoral hood appeared normal, a normal stream of clear urine was observed. A thin walled cyst filled with clear fluid, that appeared to originate from the right side of the introitus but separate from the urethral meatus, was seen (Fig. 2). Ultrasound demonstrated normal appearing kidneys and a decompressed bladder without hydronephrosis or evidence of abnormal Müllerian dilation. The simple cyst was visualized in the midline and measured 1.5 x 1.2 cm.

The diagnosis of a paraurethral cyst arising from Skene’s gland. The cyst was managed conservatively with spontaneous rupture on the second day of life draining clear fluid with complete resolution. The infant was discharged from the hospital on day of life 13, and at one year has had no evidence of recurrence.

Discussion

The Skene’s gland produces a mucoid material that, if obstructed, may lead to development of a paraurethral cyst. The occurrence of this in the neonate is relatively rare. The exact mechanism of obstruction of the Skene’s gland remains unclear, and as shown here can happen early in utero.

The diagnosis of a paraurethral cyst should be considered when a pelvic cyst is noted on antenatal ultrasound. When the diagnosis is suspected, serial sonograms serve to confirm that obstruction or involvement of adjacent structures does not develop. MRI imaging may be considered to help define anatomic relationships. With acute neonatal obstruction, surgical management with needle drainage, marsupialization or complete excision of the cyst can be considered. Fortunately, associated obstruction is rare and most cysts spontaneously resolve over weeks, therefore expectant management is typically the preferred approach.

References


Figure 1: Left: Transverse view of the pelvic cyst on ultrasound at 32 weeks gestation showing the cyst (C) in relation to the labia (L). Right: Three-dimensional ultrasound of the pelvic cyst (C) in relation to normal appearing labia (L), thighs (Th), and anus (A) at 32 weeks gestation.

Figure 2: Appearance of the pelvic cyst at the vaginal introitus on the first day of life, noting a thin walled cyst with clear fluid behind and small blood vessels running across its surface.
Clitoral Mass in a Prepubescent Female

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Introduction

Acquired masses of the clitoris are rare. We present a case of a prepubescent female with a periclitoral mass. Pathology revealed a hemorrhagic pseudocyst. To our knowledge, this is the first reported case of a periclitoral pseudocyst.

Case Presentation

An eleven-year-old African American female presented with a three-day history of dull perineal pain and acute clitoral swelling. She denied fevers, abdominal pain, vaginal discharge, dysuria or hematuria. There is no history of voluntary or involuntary sexual activity, masturbation or genital trauma. Limited bedside examination revealed a Tanner Stage 1 female with a tender mass between the labia. There was no inguinal adenopathy. Laboratory data and urinalysis were unremarkable. A pelvic ultrasound revealed no adnexal masses with normal Doppler flow to both ovaries. Magnetic resonance imaging of the pelvis revealed a 3.8 cm x 2.9 cm x 2.1 cm well-circumscribed oval mass adjacent to the clitoris. There were no other concerning pelvic findings. Decision was made to perform an exam under anesthesia, cystoscopy, and vaginoscopy with possible resection. Intraoperatively, examination revealed a normal urethral meatus and vaginal introitus. The mass was adjacent to the right clitoral hood. Complete resection with enucleation of the mass was achieved with reconstruction of the clitoral hood. The neurovascular bundles of the clitoris and corpora cavernosa were preserved during dissection. The mass was supplied by a single artery, which was dissected and tied off. Cystourethroscopy and vaginoscopy were performed after resection which was unremarkable. Histological examination of the lesion revealed a hemorrhagic cyst without a true epithelial lining; this finding was consistent with a hemorrhagic pseudocyst. The patient was discharged on postoperative day one. On clinical follow-up, her cosmetic result was excellent and her pain had resolved.

Discussion

Cysts of the clitoris are rare. Differential diagnosis include epidermoid cysts, pilonidal cysts, paraurethral cysts, and dysontogenetic cysts of paramesonephric or mesonephric origin. Benign cysts should be differentiated from other causes of acquired clitoromegaly, which may be viewed in the setting of an endocrinopathy such as virilizing tumors of the ovaries and adrenal gland, polycystic ovarian syndrome, or exogenous androgen administration. This also may be mistaken for ambiguous genitalia. Non-hormonal causes of clitoromegaly include lymphoma, rhabdomyosarcoma, endodermal sinus tumor, leiomyoma, genital neurofibromatosis, and distant metastatic disease. History and physical examination are important in narrowing a clinician’s differential and assessing the need for an endocrinology evaluation. To our knowledge, this is the first case report of a periclitoral hemorrhagic pseudocyst. The inciting event is unclear and appears to be spontaneous based on her clinical history. Radiographic imaging ruled out a malignant process in the pelvis, or mullerian abnormalities. Goals of surgical management include total excision of the lesion for pathologic diagnosis, alleviation of patient discomfort, and preservation of clitoral sensation and future sexual function.

References


Figure 1: Clitoral mass noted on physical examination.

Figure 2: T2 MRI of the Pelvis. Coronal reconstruction demonstrates the location of the periclitoral mass (asterisk) in relationship to the bladder (B). Note leftward displacement of the clitoral bodies (arrow).
CEVL Responds to the AAP Report On Male Circumcision

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Case Presentation

In 2012, the American Academy of Pediatrics updated the organization’s Statement on Male Circumcision, recognizing that the benefits of circumcision outweighed the risks and recommended that circumcisions be offered to families who desire it. In anticipation of an increase in demand for neonatal circumcision, the AAP called for the unification of training and the establishment of proficiency standards. The CEVL™ (Computerized Enhanced Visual Learning) platform currently meets these challenges set forth by the AAP.

Discussion

CEVL™ was first introduced as an educational tool to teach pediatric orchiopexy to urology residents in 2006. Since then, CEVL™ has been shown to be an effective platform for teaching other open and endoscopic surgical procedures as well as teaching imaging and diagnostic skills. The CEVL™ teaching method centers on: accessibility to online curriculum, teaching surgical procedures in a step-wise fashion, incorporating the use of simulation models to obtain mastery of skills, and immediate feedback and remediation. These key concepts make CEVL™ the ideal teaching platform for neonatal circumcision.

CEVL™ for Circumcision began development in 2005 and has evolved in a multi-institutional collaborative fashion. The program is structured with an interactive online teaching module that educates the user in judging the suitability of the penis for circumcision (Fig 1). Furthermore, it teaches the performance of circumcision broken down into a series of components and provides video and animations to help familiarize the learner with the procedure. Next, CEVL™ provides a simulation model on which to practice. Real-time feedback and remediation is provided to the learner during simulation and proficiency is evaluated using a ROSe (Recording of Skills Evaluation) checklist (Fig 2). The second portion of the module consists of clinical application. Again, the components and steps of the procedure are reviewed. Video clips are provided to demonstrate the components of the circumcision as well as common bloopers and rescue strategies. The learner is then asked to show their readiness through an interactive quiz. After completion of the training module, the learner graduates to performing a newborn circumcision under supervision. Again, immediate feedback and remediation is provided to the learner. After completion of the program, it is expected that the learner achieves the knowledge and technical skills needed to safely and proficiently perform newborn circumcision. Modules using the GOMCO, Mogen, and Plastibell have been developed. For more information and a demonstration of CEVL™ for Circumcision, please visit: www.cevlforhealthcare.org.

To date, CEVL™ for Circumcision has been used successfully at 7 institutions to teach pediatric, obstetric, urology, and family practice residents. In a study of pediatric residents, CEVL™ for Circumcision was shown to improve residents’ proficiency scores from a mean of 85/100 to 97/100 after one month. All participants achieved scores consistent with mastery of skills at the completion of the study.

Conclusion

The CEVL™ platform for teaching newborns circumcision meets the needs as outlined by the AAP report on newborn circumcision. It provides a standardized training method that can be used by learners in multiple medical specialties and allows for the establishment of objective proficiency measures that may serve as a credentialing mechanism.

References

Serial Fetal Renal Aspirations for Prenatal Hydronephrosis: Failure due to Gastric Outlet Obstruction?

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Case Presentation

A male fetus was found to have massive right-sided hydronephrosis on second trimester screening ultrasound, with renal pelvis diameter 70 mm at 22 weeks gestation. The left kidney appeared normal and the amniotic fluid volume (AFV) was normal. Serial percutaneous aspirations of the kidney were performed in utero between 22 and 33 weeks, with removal of 200-240 mL of clear fluid at each attempt. Two of these aspirations resulted in complete decompression of the kidney. The fetus developed oligohydramnios at 33 weeks and ultrasound demonstrated a 12.4 x 8.1 x 9.9 cm hydronephrotic right kidney, right hydroureter, but a normal appearing left kidney (Fig 1). The infant was born by induction at 36 6/7 weeks and was intubated in the delivery room due to respiratory distress. Exam revealed a two-vessel umbilical cord, an imperforate anus, a bifid scrotum with descended testes, and a distended abdomen with a compressible right-sided mass. Postnatal ultrasound revealed severe right hydronephrosis with peripheral renal cysts, moderate right hydroureter, and a small left kidney with increased echogenicity, peripheral cysts and mild hydronephrosis (Fig 2). The bladder was only partially distended with normal wall thickness. A right percutaneous nephrostomy tube was placed to de-compress the kidney. The infant developed spontaneous bilateral pneumothoraces requiring chest tube placement. He ultimately expired from progressive respiratory failure within the first 24 hours of life.

Discussion

Evidence to support prenatal intervention in the setting of congenital hydronephrosis is currently lacking. The SFU guidelines recommend repeat ultrasound in cases of severe antenatal hydronephrosis and only suggest prenatal intervention on an individualized basis.¹ This is a case of fatal pulmonary hypoplasia despite normal AFV up to 33 weeks gestation and seemingly promising results of serial renal aspirations of hydronephrosis. It has been shown that the presence of amniotic fluid up to 27 weeks protects against profound pulmonary hypoplasia.² In this fetus with massive hydronephrosis, it is possible that the mass effect prevented adequate pulmonary development despite normal AFV. It has also previously been shown that massive hydronephrosis can cause fetal gastric obstruction, causing impaired clearance of amniotic fluid resulting in polyhydramnios.³ Thus, the renal function of this fetus may have been worse than presumed despite “normal AFV” in light of concomitant gastric outlet obstruction.

References

An Atypical Presentation of Multicystic Dysplastic Kidney

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Case Report
The patient is a normal birth weight product of a 38 week gestation, delivered by cesarean section. Pregnancy was complicated by mild polyhydramnios, maternal tobacco use, and a suspected right multicystic dysplastic kidney (MCDK). At birth, the patient was noted to have a palpable right abdominal mass. A renal ultrasound obtained following delivery revealed an enlarged right kidney with dysplastic renal parenchyma and multiple cysts of varying sizes including a large central cyst consistent with a renal pelvis (Fig 1). Renal function was within normal limits. Voiding cystourethrogram confirmed a normal bladder and urethra and an absence of vesicoureteral reflux. Urology was consulted to evaluate the MCDK. However, the renal ultrasound was suggestive of communication between the cortical cystic structures and a central hypoechoic area, resembling a dilated renal pelvis secondary to ureteropelvic junction obstruction (UPJO).

In order to evaluate for a poorly functional kidney with UPJO, a DMSA scan was obtained and demonstrated 16% function in the right kidney (Fig 2). In light of the degree of renal function observed, a percutaneous nephrostomy tube was placed. Output from the nephrostomy averaged 0.5 ml/kg/hr over the first week, but slowly decreased to zero after 2 weeks. A repeat renal ultrasound was obtained, again showing multiple cysts, but no definite collecting system was seen. Differential function remained stable on a repeat DMSA scan, with the right kidney contributing 16% function. At this point, the diagnosis of MCDK was felt to be most likely, so the nephrostomy tube was removed. He is currently being managed conservatively and has had no increased dilation of the renal cortical cystic areas with resolution of the central hypoechoic cystic structure.

Discussion
Multicystic dysplastic kidney (MCDK) is the most common cystic renal disease of infancy, involving 1:4300 live births, and is usually diagnosed by prenatal ultrasound.1 Findings consist of multiple cysts of varying sizes, a nonreniform shape, and no identifiable renal parenchyma. A less common variant, the hydronephrotic type, is characterized by a large central cystic structure and a more reniform shape.2 In cases where it is difficult to differentiate a hydronephrotic type multicystic kidney from a severe UPJO, nuclear scintigraphy is recommended, in which a MCDK will demonstrate minimal to no uptake.3 In this particular case, the presence of function on DMSA scan suggested a diagnosis of UPJO, but subsequent renal imaging confirmed MCDK. We are unaware of additional criteria or imaging modalities to aid in the diagnosis of MCDK. Had the diagnosis been more apparent, the morbidity and cost associated with temporary urinary diversion might have been avoided.

References
Pre-sacral Teratoma as the Cause of Neonatal Bladder Outlet Obstruction

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Case Report
An ex-36 week gestational age male was born following a prenatal course complicated by oligohydramnios beginning at 32 weeks. Ultrasound at 19 weeks was normal, though ultrasound at 36 weeks revealed a large cystic pelvic mass. Upon delivery, the patient experienced respiratory failure requiring intubation. Pediatric urology was consulted for presumed posterior urethral valves (PUV), and a urinary catheter was placed. Physical exam revealed a generally well developed male with normal external genitalia without evidence of abdominal or pelvic mass. Newborn ultrasound revealed hypoplastic lungs, echogenic kidneys with moderate bilateral hydronephrosis, and a large abdominal cystic structure measuring 5.3x2.1x3.7 cm in the upper abdomen.

This fluid-filled structure increased to 6.4x4.3x3.9 cm on day of life (DOL) 2, despite a persistently draining urinary catheter. Because of deteriorating respiratory status, percutaneous ultrasound guided drainage of the mass was performed. 45 milliliters of straw colored fluid were removed, and the mass was completely decompressed. Fluid analysis for creatinine and electrolytes were consistent with serum. Ultrasound on DOL 4 revealed re-accumulation of fluid in a 4.4x5.1x1.6 cm cystic pelvic mass, posterior to a now visualized bladder (Fig 1). Interventional radiology placed a drain in the structure on DOL 11, following further respiratory decompensation.

After the patient stabilized, voiding cystourethrogram revealed a normal posterior urethra, thickened bladder wall, and right grade II vesicoureteral reflux. MRI demonstrated a predominantly cystic pre-sacral teratoma (PST) (Fig 2). Alpha-fetoprotein (AFP) was measured at this time and was 27,693 ng/mL. The mass was resected via transabdominal and perineal approaches with pathology showing mature sacral teratoma.

The postoperative course was notable for urinary retention requiring clean intermittent catheterization (CIC). The patient was discharged to home on prophylactic antibiotics on DOL 72 after successful weaning of the ventilator. Creatinine at the time of discharge was 0.3 mg/dL, while alpha feto-protein normalized by 11 months postoperatively.

Discussion
While PUV are the most common cause of bladder outlet obstruction in newborn males,1 it is important to consider the diagnosis of PST. Sacrococcygeal teratomas (SCT) occur in 1 in 40,000 births. Mature tumors are benign and are cured by resection, while immature tumors may require multimodal treatment. Type IV SCT, pre-sacral teratoma, is entirely internal and thus difficult to diagnose on physical exam. Patients often present with abdominal distention, constipation, or urinary retention. Congenital PST can lead to significant neonatal morbidity. As in PUV, bladder outlet obstruction can cause oligohydramnios leading to hypoplastic lungs and respiratory compromise.2 The cardiovascular system may be affected as the tumor may cause vascular steal syndrome, leading to high output cardiac failure and hydrops fetalis.2 Furthermore, long term bladder dysfunction may result from obstructive changes during bladder development or from iatrogenic neurologic damage during surgical resection. Early treatment with anticholinergics or CIC improves outcomes.3

Given the complexity and potential morbidity of congenital PST, a high index of suspicion is required for diagnosis, and early involvement of multiple specialties are necessary for optimal patient care.

References

Figure 1: Abdominal ultrasound

Figure 2: Sagittal T2-weighted MRI.
Case Report
A 22-month-old otherwise healthy female was referred after presenting to an outside emergency room one week earlier with sudden onset of abdominal pain and emesis. CT scan was performed and read as an intra-abdominal mass and possible urachal cyst. Urine specimen revealed cloudy urine. Her symptoms subsided, and she was discharged on oral antibiotics. The patient was referred to our institution for further management of her “urachal cyst”. On initial evaluation, the patient’s abdomen was distended but without focal mass. Urologic exam was limited due to the patient’s discomfort. Review of the outside CT scan revealed bilateral hydronephrosis, a distended bladder, and a large cystic structure posterior to the bladder. Abdominal ultrasound demonstrated a multi-component cystic structure, with volumes of 400 mL, 70 mL, and 36 mL. Upon catheter placement for voiding cystourethrogram (VCUG), 400 mL of foul-smelling urine drained. Due to likely infection, the VCUG was canceled and the catheter left in place. Repeat ultrasonography revealed drainage of the largest cystic structure, now identified as bladder. The 70 mL structure was more clearly identified as vagina, and the 36 mL structure as uterus (Fig 1).

Subsequent magnetic resonance urogram suggested a diagnosis of vaginal atresia (VA) with a fluid-filled proximal vagina and uterus and 0.7 cm of tissue between the vaginal lumen and perineal skin (Fig 2).

After adequately treating her urinary tract infection, the patient was taken to the operating room. Examination under anesthesia revealed a solitary perineal opening consistent with urethral meatus. Cystourethroscopy was normal. Diagnosis of distal VA was confirmed, and flap vaginoplasty was performed. Viscous yellow fluid was returned upon entering the vaginal mucosa. Vaginoscopy revealed a normal vagina and cervix. Post operatively, the patient healed well and is managed with vaginal dilatation to prevent stenosis.

Discussion
During female embryologic development, the Müllerian ducts fuse to form the fallopian tubes, uterus, and proximal two-thirds of the vagina. An area of the posterior urogenital sinus thickens, forming the sinovaginal bulbs and eventually the distal 1/3 of the vagina. Vaginal developmental anomalies can occur and are classified as agenesis anomalies or anomalies in fusion. Vaginal agenesis is often associated with uterine, ovarian, renal, and skeletal anomalies. Anomalies in horizontal fusion lead to duplicated systems while anomalies in vertical fusion lead to transverse vaginal septa or VA.

Patients with VA often present at puberty with primary amenorrhea or abdominal pain secondary to hematometrocolpos. Our patient presented at a young age with urinary retention and hydronephrosis secondary to hydrometrocolpos. Patients generally experience good long-term surgical outcomes but require close follow-up through puberty and may require continued vaginal dilatation.

Developmental abnormalities of the vagina may lead to hydrometrocolpos and result in urologic sequelae including urinary retention and hydronephrosis. As with our patient with VA, symptoms are not necessarily present at birth but may develop acutely prior to the onset of menses. Early diagnosis and management prevents prolonged morbidity and is aided by careful physical examination and dedicated imaging studies.

References

Figure 1: Abdominal ultrasound reveals the drained bladder anterior to a fluid-filled uterus and vagina

Figure 2: Sagittal T2-weighted MRI demonstrates a distended bladder despite indwelling catheter, a distended vagina, and 0.7 cm of tissue between the distal end of the vagina and perineal skin
Is There a Role for Fetal Intervention in the Management of Ureteroceles?

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Introduction

Antenatal hydronephrosis is detected in 1-5% of all pregnancies. The most common etiology for this finding is transient hydronephrosis, with up to 5-7% of cases resulting from an ureterocele. In most cases, fetal intervention is not required. We present a case of a patient with prenatally detected ureteroceles and partial bladder outlet obstruction, and a review the literature regarding fetal intervention for ureteroceles.

Case Report

This 31-year-old female was found to have a fetus with bilateral severe hydronephrosis, an ectopic ureterocele, and a normal amniotic fluid index (AFI) on routine prenatal ultrasonography. She was referred at 34 weeks gestation to the Colorado Institute for Maternal and Fetal Health (CIMFH) at Children’s Hospital Colorado for further evaluation, which consisted of a prenatal ultrasound, a fetal MRI, and a multidisciplinary clinic visit.

The ultrasound confirmed the presence of bilateral severe hydronephrosis and a large ureterocele. The fetal MRI demonstrated duplex kidneys with upper pole > lower pole hydronephrosis, a distended bladder, and bilateral (L>R) ureteroceles (Fig 1). The fetus was further noted to have normal lung development and an AFI within normal limits. Following consultation with the CIMFH providers, the patient elected to deliver closer to home in Billings, MT.

The patient delivered a baby girl at forty weeks gestation via an uncomplicated, spontaneous vaginal delivery. The physical examination was within normal limits, and she voided spontaneously. Urinary tract infection (UTI) prophylaxis was initiated. A renal ultrasound and a voiding cystourethrogram (VCUG) were obtained on day of life (DOL) 2. The ultrasound was demonstrated duplex kidneys with upper pole > lower pole hydronephrosis and bilateral (L>R) ureteroceles (Fig 2). The VCUG demonstrated bilateral (L>R) ureteroceles and lower pole grade V vesicoureteral reflux (VUR) on the right.

On DOL 5, the patient underwent cystourethroscopy, with transurethral incision of the ureteroceles by Dr. Chavez (Billings, MT). The left ureterocele was incised with a 2.2 Fr Bugbee electrode. The right ureterocele was incised in a similar fashion via the decompressed left ureterocele (peek-a-boo procedure). The patient tolerated the procedure well and had an uneventful recovery.

A renal ultrasound was obtained three weeks postoperatively, demonstrating duplex kidneys with decompressed hydronephrosis and decompressed ureteroceles. A diuretic nuclear renal scan was subsequently obtained at two months of age, demonstrating equal function and drainage. Arrangements have been made to obtain a renal ultrasound and a VCUG at six months of age. In the meantime, the patient will remain on UTI prophylaxis.

Discussion

Although fetal intervention was not necessary in this case, one must consider when fetal intervention for an ureterocele may be of benefit. There are limited case reports in the literature describing various fetal interventions for obstructing ureteroceles (i.e., those ureteroceles associated with oligohydramnios or anhydramnios). The indications for fetal intervention and the optimal timing and technique have not yet been established.

References


Figure 1: Fetal MRI demonstrating duplex kidneys with upper pole > lower pole pelvicaliectasis (left) and a distended bladder with bilateral (L>R) ureteroceles (right).

Figure 2: Postnatal renal/bladder ultrasound demonstrating duplex kidneys with upper pole > lower pole pelvicaliectasis (above) and bilateral ureteroceles (below).
Deforming Penoscrotal Lesions in a 9 Year Old

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Case Presentation
A 9 year old boy presented with wart-like lesions and vesicles on his penis and scrotum that were present since newborn circumcision. The patient denied pain or discomfort. On examination, lesions covered his scrotum and part of his penis. The glans was edematous but non-tender. A differential diagnosis of genital warts and molluscum contagiosum was made. After evaluating the patient for sexual abuse, the family was instructed to apply imiquimod cream on the lesions and follow up in two months. On follow-up evaluation, the lesions were still present, and the patient was referred to dermatology where shave biopsy was performed. Histopathologic examination revealed lymphatic tissue, and a diagnosis of lymphangioma circumscriptum (LC) was made. After referral to pediatric urology, an incisional scrotal biopsy confirmed a microcystic lymphatic malformation. Further investigation with MRI of the abdomen, pelvis and lower extremities demonstrated subcutaneous soft tissue swelling of scrotum and distal half of the penis, and areas of microcysts on the scrotum (Fig 1). There was normal lymphatic absorption and drainage without obstruction in the lower extremities, abdomen or pelvis. A multidisciplinary team of pediatric urology, dermatology and interventional radiology convened and recommended photocoagulation of the lesions. To date, the patient has undergone two photocoagulation sessions using a Nd:YAG laser. Initial treatment saw an excellent response in the test area, with successful vaporization of lesions. The second treatment covered a larger surface of the inferior scrotum of approximately 2 cm x 4 cm. One week post treatment, his Primary Care Pediatrician noted significant improvement in the treated areas, with reduction in the number of lesions present in the treatment area, and vesicles in the few remaining clusters decreased in size and number (Fig 2).

Discussion
LC is a rare cutaneous lymphatic malformation characterized by localized, thin-walled vesicles. Genital LC occurs infrequently, and is often misdiagnosed as a sexually transmitted infection or molluscum contagiosum.1 It may be congenital or acquired following trauma, radiation or infection. Developmentally, deep cisterns form separate from the normal lymphatic system in the deep subcutaneous tissues. Lined by smooth muscle, the cisterns contracts and transmit pressure to ectopic lymphatics, creating the cutaneous manifestation.1 Clinical concerns include drainage, superinfection, cosmesis, and in sexually active patients, compromised erectile function.1,3 Superficial disease may be amenable to treatment with laser ablation, radiotherapy, cryotherapy, electrocautery, topical applications, and intense pulsed light source therapy.1 When deeper cisterns are present, surgical excision may be necessary. In order for surgery to be successful, all abnormal tissue must be removed. Surgery, even if curative, can be disfiguring, thus less invasive treatments should be considered.2 Extensive resection of penoscrotal skin requires creative uses of flaps and grafts to enable normal erectile and testicular function.3 A multidisciplinary approach aids in diagnosis and optimizes management of these rare patients.

References
An Uncommon Cause of Fetal Urethral Obstruction

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Case Presentation

A 35 year old female underwent ultrasonography at 28 weeks gestation documenting bilateral hydronephrosis, more pronounced on the right. A repeat study at 32 weeks gestation showed persistent bilateral hydronephrosis - grade 2 on the right and minimal on the left with a right renal pelvic diameter of 2.1 cm. She gave birth at term to a healthy boy. Neonatal evaluation demonstrated bilateral grade 4/5 vesicoureteral reflux on VCUG with a questionable urethral anomaly. The neonate was voiding without issue, with no evidence of urinary retention. Creatinine on 4 day of life was 0.5 mg%. A repeat VCUG at our institution was suggestive of a large pedunculated polyp arising from the verumontanum, prolapsing into the posterior urethra. In addition, bilateral grade 4/5 VUR was again noted with intermittent emptying. On neonatal cystoscopy, an obstructive pedunculated polyp was encountered in the posterior urethra, arising proximal to the verumontanum (Fig 1 and 2). The polyp was snared and placed on traction upwards to delineate the base. Using a holmium laser, the polyp was transected at the base of its stalk and extracted using a tipless nitinol basket. Post-operatively the creatinine was 0.1, after removal of the foley catheter, the patient voided without issue. The final pathology on the posterior urethral mass was a fibroepithelial polyp.

Conclusion

Fetal urethral obstruction may be caused by varying types of lesions, and frequently the definitive diagnosis is only made by neonatal imaging. Management depends on the etiology and severity of the obstruction. Neonatal fibroepithelial polyps of the urethra are rare lesions, usually arising from verumontanum. Malignant transformation has not been reported. Endoscopic excision has proven to be effective management.

References

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Figure 1

Figure 2