With perinatal ultrasonography (US), urologists who primarily treat adults should be familiar with the principles of evaluation and management options for adult, pediatric, and fetal populations as they may be asked to consult on a newborn or fetus with a dilated ureter. See “Embryology,” page 45, for a brief review of ureteral development.

A variety of terms have been applied to ureters of abnormal caliber, including megaloureter, megaureter, dilatation of the ureter, and widened ureter. King has suggested the term megaureter to describe any ureter that is found to be dilated.1 This broad term is defined further by 3 etiologic categories: obstructed megaureter, refluxing megaureter, and nonrefluxing nonobstructed megaureter (Table 1). Each category is further classified into primary and secondary subgroupings. This article focuses on diagnosis and management of primary obstructed megaureter.

ETIOLOGY AND PRESENTATION

Primary obstructed megaureter occurs in both pediatric and adult populations.2 Rather than being caused by a lumenal obstruction, the condition is due to an intrinsic abnormality or an adynamic segment of the distal ureter that leads to a functional obstruction. In the past, the functional obstruction was thought to be similar to that seen in Hirschprung’s disease of the colon. However, this has been disproven as excised samples demonstrated the presence of neural plexuses.3,4

Approximately two thirds of reported patients are males; however, 1 series displayed a female predominance.5 The abnormality is unilateral in approximately two thirds of cases.5 The dilation is due to an intrinsic abnormality or an adynamic segment of the distal ureter that leads to a functional obstruction. In the past, the functional obstruction was thought to be similar to that seen in Hirschprung’s disease of the colon. However, this has been disproven as excised samples demonstrated the presence of neural plexuses.3,4

While dilated ureters are usually identified in adults when they present with symptomatic complaints, in children, identification is most likely due to aggressive screening with perinatal ultrasonography (US). Urologists who primarily treat adults should be familiar with the principles of evaluation and management options for adult, pediatric, and fetal populations as they may be asked to consult on a newborn or fetus with a dilated ureter.

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The presence of other anomalies, such as contralateral ureteropelvic junction (UPJ) obstruction, renal agenesis or ectopia, and ureteral duplication associated with primary obstructed megaureters, may be seen in up to 40% of affected patients. In adults, primary obstructed megaureter is usually detected when patients present with pain or other symptoms from urinary tract infection (UTI), calculi, or decreased renal function. In mild cases, however, patients are typically asymptomatic, and the condition is found during an unrelated workup.

In the pediatric population, routine perinatal US has dramatically increased the likelihood of detection of the dilated ureter. The megaureter can be dilated up to 3 cm and thus is easily identified on US as a hypoechoic cystic-appearing mass in the retroperitoneum that may extend from the UPJ to the ureterovesical junction (UVJ). In a 5-year series in which US was performed on 3,856 fetuses after 28 weeks of gestation, the prevalence of primary megaureter at the level of the UVJ was approximately 1 in 2,000.

**GRADING**

Several different grading systems have been promulgated, but none is useful in predicting which patients will require surgery and which will benefit from observation. The earliest classification system, devised in 1978 by Pfister and Hendren, categorizes the development of the ureter begins around the fifth week of gestation. A diverticulum arises from the posteriomedial aspect of the lower portion of the bilateral mesonephric ducts. It then elongates posteriorly to meet the metanephric blastema, thus inducing nephrogenesis. The tip of the ureteric bud dilates to form the collecting system from the ureterovesical junction (UVJ) to the level of the collecting duct. During the sixth through the ninth weeks, the embryonic kidney ascends from its pelvic position.

During the ascent of the kidneys, the ureters elongate. The lumen of the forming ureter develops from the midureter cranially and caudally until the eighth week. The urogenital sinus remains separated from the ureteric lumen by a membrane. This membrane, known as the Chwalle’s membrane, disappears by the middle of the eighth week. At approximately the ninth week of gestation, muscularization is induced by the passing of the first excreted urine. At 18 weeks, normal physiologic narrowing can be discerned at the ureteropelvic junction (UPJ) and the UVJ. It is at this time that dilation of the ureter may be observed. After 19 weeks, the ureter continues to grow; however, the normal ureteral diameter in the fetal population rarely exceeds 5 mm.

**REFERENCE**

primary obstructive megaureter in children and adults according to the degree of dilatation of the proximal ureter. In grade 1, the dilatation is limited to the distal ureteral segment. In grade 2, the dilation extends into the proximal ureter and there may be mild caliectasis. In grade 3, the entire ureter is dilated proximal to the adynamic segment and there is moderate to severe caliectasis. One drawback to Pfister and Hendren’s grading system is that excretory urography, which is rarely necessary today, is required.

The most commonly used classification for neonates, which is now standard in adults, was devised by the Society for Fetal Urology (SFU). It assigns a grade from 0 to 4 based on the degree of upper urinary tract dilation. In grade 0, the central renal complex is intact. In grade 1, there is mild splitting of the renal complex. In grade 2, the pelvis is dilated but the calyces are not dilated. In grade 3, the pelvis is markedly split and the calyces are uniformly dilated but the renal parenchyma is normal. Grade 4 shares the characteristics of grade 3, but there is thinning of the renal parenchyma.

EVALUATION OF PRENATALLY DIAGNOSED MEGAURETER

While US is highly sensitive for the detection of a dilated ureter, it lacks specificity for diseases that will actually require intervention. Classification of megaureters based on the etiology of the dilation allows the clinician to better inform patients about the treatment options and their risks and benefits. Thus, at our institution, we recommend that newborns with prenatally diagnosed megaureters undergo US and voiding cystourethrography (VCUG) within 48 hours of birth. If reflux is ruled out on VCUG, we proceed to DTPA diuresis renography to better evaluate renal function and to determine the degree of ureteral obstruction (Figure 1).

MANAGEMENT IN ADULTS

Adults with primary obstructed megaureter typically present with pain, infection, or hematuria. Radiologic evaluation (Figure 1) usually reveals pathology in the distal ureter. Therefore, aggressive surgical management has been recommended for adults.

Hemal and colleagues identified 53 of 55 adult primary obstructed megaureters on the basis of studies obtained to evaluate symptoms. While the authors advocated surgical management in most instances, they found it unrewarding in patients with bilateral disease who have advanced renal failure. Of 5 patients with bilateral primary obstructed megaureter and uremia, 3 underwent ureteral reimplantation. Of that group, only 1 improved with adequate drainage, and the other 2 patients died.

MANAGEMENT IN CHILDREN

While surgery is the first-line therapy for primary obstructed megaureter in adults, the approach in children is evolving. Several studies have compared surgical outcomes with conservative management, but all are retrospective and involve a considerable selection

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**TABLE 1**

**Differential diagnosis of megaureter**

**OBLUCUTED MEGAURETER**

**Primary:** This condition is due to an intrinsic abnormality or an adynamic segment of the distal ureter that leads to a functional obstruction.

**Secondary:** Any cause of ureteral obstruction not due to an adynamic segment. Etiologies include congenital lesions (ureteropelvic junction obstruction, ectopic ureteroceles), inflammatory conditions (tuberculosis, schistosomiasis, Crohn’s disease, pelvic inflammatory disease, pelvic abscess), trauma, tumors, lower urinary tract conditions, neurogenic bladder, benign prostatic hypertrophy, pelvic lipomatosis, and urethral obstruction.

**REFLUXING MEGAURETER**

**Primary:** This condition occurs most often in children and describes a wide ureter secondary to vesicoureteral reflux (VUR), when an abnormality of the ureteral orifice (or tunnel length) is the cause of the reflux.

**Secondary:** Reflux due to high bladder pressures from a neurogenic bladder or bladder outlet obstruction.

**NONREFLUXING, NONOBLUCUTED MEGAURETER**

**Primary:** Dilation not due to obstruction or reflux (prune-belly syndrome.)

**Secondary:** In this condition, ureters remain dilated after the correction of initial pathology.
bias. Nevertheless, these studies document a shift from the traditional approach of surgical management to the current trend of surveillance.

Between 1981 and 1987, Keating and colleagues assessed 44 renal units in 35 neonates with primary obstructed megaureter.7 Infants with ureters dilated down to the UVJ with varying degrees of hydronephrosis were included in the study. Infants with secondary obstructed megaureters were eliminated from the analysis. Antenatal diagnosis was made by US in 23 of 44 units. Of the 23 units, 87% were managed nonoperatively. Diagnoses for the remaining 21 units were made on the basis of symptomatic complaints due to UTI or the presence of a flank mass, or they were incidentally discovered. Only 12 of the 21 symptomatic units were managed conservatively. Subsequently, surgery was performed on 2 of the 12 conservatively managed units because of increased obstruction on DTPA diuresis renogram and increased dilation on US. The authors suggested that a decision to manage asymptomatic patients conservatively should be based on an estimate of absolute renal function as determined by diuresis renography.7

In 1994, Baskin and associates9 provided a long-term follow up of Keating and colleagues’7 carefully selected group and found that 10 of the original 35 neonates with 44 renal units ultimately underwent surgery. The remaining 25 were observed and managed with serial urinary tract imaging using DTPA diuresis renography, intravenous pyelography (IVP), and/or renal US. Seventeen of the 25 were diagnosed antenatally, 2 were identified due to infection, and 6 incidentally diagnosed. Mean follow-up was 7.3 years for 24 patients. One patient was lost to follow-up after 1.5 years. The conservatively managed patients demonstrated no decline of renal function on DTPA diuresis renography during the observation period. The authors concluded that conservatively managed patients should be monitored closely as indications for surgical repair may arise.9

McLellan and co-workers evaluated the records of 54 newborns who were prenatally diagnosed with primary obstructed megaureter from 1993 to 1998.10 Median follow-up was 25.8 months. A total of 69 units were confirmed postnatally using various imaging modalities. Antibiotic prophylaxis was continued until the children were between the ages of 9 and 12 months, depending on physician’s preference. No child had a culture-documented UTI.

Resolution, defined as a decrease in hydronephrosis to SFU grade 1 without hydroureter or minimal residual hydroureter, occurred in 39 (72%) patients. Five patients (9%) had no resolution during the surveillance period, and 10 (19%) underwent surgery. The presenting grade of hydronephrosis appeared to be an important predictor of the resolution rate. SFU hydronephrosis grades 1 to 3 were
more likely to resolve within 12 to 36 months. Increasing or severe hydronephrosis, decreasing renal function, and/or retrovesical ureteral diameter greater than 1 cm seemed to correlate with the need for surgical repair.

Multiple reports support conservative management for primary obstructed megaureter detected in asymptomatic neonates. We follow these patients with serial US and DTPA diuresis renography if increased dilation is observed on US (Figure 2). Once vesicoureteral reflux has been excluded by VCUG, antibiotic prophylaxis can be discontinued.

**INDICATIONS FOR SURGERY**

The absolute indications for surgical intervention have yet to be determined. Indications for surgery suggested by Simoni and associates include significant impairment of urine flow on renal scan, worsening renal function during observation, and recurrent UTI in spite of adequate antibiotic prophylaxis.

Stehr and colleagues proposed 3 indications for surgery using US, VCUG, IVP, and MAG-3 renal scan: initial impaired renal function with an obstructive pattern, normal function and at least an equivocal urinary drainage pattern with no improvement, or deterioration of the urinary drainage and/or function during follow-up. Using these criteria, only 5 (9.6%) of 42 patients were managed surgically.

Liu and co-workers studied 67 units with pathology at the UVJ. Eleven (17%) patients failed conservative therapy and required surgical repair—3 due to breakthrough infections and 8 because of deteriorating function. The remaining 56 (83%) patients were managed conservatively by periodic followup with US and DTPA diuresis renography.

**SURGICAL METHODS OF REPAIR**

In uncomplicated nondilated ureters, reported success rates for the various open reimplantation techniques are well over 95% in children 1 year of age or older. However, for large dilated ureters requiring plication or tapering in addition to the usual intravesical reimplant, no conclusive data have been published.

Glassberg and associates reported a 99% success rate with tapering using a transverse ureteral advancement technique of ureteroneocystostomy (Cohen reimplant) in 7 primary obstructed megaureters. They noted that megaureters measuring 8 to 12 mm in width, regardless of etiology, can be reimplanted successfully without tapering.

Hospitalization following reimplantation usually only requires an overnight stay. A double-pigtail ureteral stent is routinely placed and is removed at 1 month. Ureteral reimplantation in the neonatal period can be difficult due to the discrepancy in size between the megaureter and the small neonatal bladder.

Should surgery become necessary in the neonatal period, we recommend that a cutaneous ureterostomy be performed. This procedure is followed in the first year of life by open intravesical reimplantation with or without the tapering. The need for surgery is based on initial poor renal function tests, a 10% reduction in serial renal function tests, worsening serial US findings, and/or the presence of breakthrough infections.

**CONCLUSION**

The most important aspect in the management of the dilated ureter is identification of patients who will need and benefit from surgical repair. Historically, patients with dilated ureters presented with symptomatic complaints, and diagnosis preceded surgical correction. Today, antenatal US screening identifies most cases in children. The antenatal diagnosis of a dilated ureter with or without hydronephrosis warrants postnatal confirmation by US and institution of antibiotic prophylaxis. Ominous signs include bilaterally dilated systems, anuria, maternal oligohydramnios, or poor renal function, and necessitate more aggressive evaluation within the first 48 hours of life.

Most patients with primary obstructed megaureter detected antenatally can be followed conservatively with US. DTPA diuresis renography may be performed after identification of increased dilation on US. Following antenatal diagnosis of primary obstructed megaureter, 72% resolve spontaneously, 19% will need operative repair, and 9% will have persistent dilation on imaging without symptoms or deterioration of func-
Predictors for the need for surgical management include severe or worsening hydronephrosis, ureteral dilation greater than 1 cm in diameter, and worsening renal function. While aggressive surgical management is usually recommended for adults with primary obstructed megaureter, parents of children with the condition should be reassured that it is likely to resolve spontaneously. However, surgical management, when indicated, is usually successful.

REFERENCES