Editorial

Fetal Hydronephrosis

As real-time ultrasonography became more accurate and widespread (approximately 15 years ago), obstetricians began to use it widely for obstetric indications. It quickly became apparent that many urologic conditions could be presumptively diagnosed antenatally. Absence of amniotic fluid was often associated with agenesis of the fetal kidneys, and bilateral cystic dysplasia or severe obstruction could reduce amniotic fluid formation from fetal urine. Whenever the urinary tract was dilated, an obstruction could be sought immediately after birth. McCrory and associates' and also Mayor and colleagues, who reviewed the topic of bilateral renal obstruction (or obstruction of a solitary kidney) in infants in 1971 and 1975, respectively, concluded that renal function improved most when obstruction was relieved before 6 months or 1 year of age, respectively. These early but compelling clinical studies showed that obstruction should be relieved at the earliest possible age consistent with safe anesthesia. Fortunately, by 1980 or 1982, advances in monitoring began to make general anesthesia safe at any age. Thus, physicians were positioned to take advantage of intrauterine "diagnoses" about the time of widespread availability of fetal ultrasonography. In their review of fetal urinary tract anomalies detected by ultrasonography, presented in the current issue of the Mayo Clinic Proceedings (pages 526 to 531), Gloor and coworkers report that isolated hydronephrosis was the most common abnormality.

Urethral Valves Versus Prune-Belly Syndrome.—Severe bilateral hydronephrosis and a bladder that does not empty on successive examinations suggest urethral valves or the prune-belly syndrome in boys and urethral atresia or a ureterocele obstructing the bladder outlet in girls. Antenatal diagnosis has considerably changed the prognosis for boys with urethral valves. Before antenatal ultrasonography was available, most boys with severe perinatal obstruction from valves initially had neonatal ascites, an abdominal (bladder or kidney) mass, inability to void, sepsis, or increasing azotemia. The mortality rate during the first month of life was 38 to 70% despite temporary urinary diversion by vesicostomy or loop ureterostomy. The current mortality rate in such patients, whose condition can now be presumptively diagnosed antenatally, is 3 to 5%, although many with mild degrees of hydronephrosis are in this latter group. Even though most patients are now treated by valve resection only, the outlook is not invariably good. Severe bladder obstruction can result in scarring and deposition of collagen in the bladder wall, making the bladder noncompliant. This condition can lead to high storage pressures and gradual late-onset hydronephrosis as well as residual urine and urinary tract infections. Alternatively, the kidneys may remain so damaged that they cannot grow as the child grows. In a study of 20 boys who had had valves and severe hydronephrosis during the second trimester, Thomas found that 10 had died or had renal failure 10 years later. This outcome contrasts with that in a group of boys in whom severe hydronephrosis had occurred during the last trimester. In these patients, the outcome was good with valve resection only, and the kidney function improved over time.

Thus, severe obstruction from probable posterior urethral valves remains the major reason to consider in utero decompression when oligohydramnios is also present. Affected male fetuses often develop successfully to term but have azotemia after birth and may still have deterioration to renal failure. One reason that fetal decompression is seldom used is that hydroureteronephrosis due to valves is impossible to distinguish from the prune-belly syndrome before birth, and intrauterine decompression does not seem to help those patients with prune-belly syndrome. They may have had high-grade urethral obstruction early during pregnancy because of prolapsed mucosa from the posterior urethra, but such obstruction is relieved as the fetus grows. At birth, few such patients have urethral obstruction, although the urinary tract is often severely damaged. Similarly, one cannot intervene early enough to prevent renal dysplasia in the developing kidney, which may be attributable to ureteral or intravesical obstruction.

Reflux.—Vesicoureteral reflux is one of the most common congenital anatomic abnormalities of the urinary tract, but such reflux is severe enough to cause hydronephrosis in only a minority of fetuses. In one review, 17% of cases of fetal hydronephrosis were considered due to reflux on evaluation after birth. Of note, however, 48% of babies with fetal hydronephrosis that persists at birth have some reflux, which is also present in 25% of babies in whom the fetal hydronephrosis has resolved. The significance of mild reflux is that urinary infection, which generally begins as a bladder infection, has a pathway to the kidney. Bacteria may be "injected" into the kidney at high pressure, especially when the ureter is dilated and cannot coapt to dampen the pressure generated by the bladder. In this situation, most compound or confluent papilla will allow reflux; thus, bacteria gain entry into the parenchyma and predispose all such renal segments to scarring after a single infection. This "big bang" is not quite comprehensive, inasmuch as new renal scars can occur in already scarred kidneys, but the detection of fetal hydronephrosis facilitates prevention of most such

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scars. If the fetus has or had hydronephrosis, all such babies should receive antibiotic prophylaxis (usually amoxicillin) immediately after birth until cystography can be performed to exclude reflux. If reflux is present, prophylactic use of antibiotics is usually continued until 6 to 8 weeks of age and then replaced with sulfisoxazole. If no infections occur, no scars will occur.

Pyelocaliectasis.—Pyelocaliectasis accounts for perhaps 65% of cases of fetal hydronephrosis. Such dilatation should not be equated to obstruction of the ureteropelvic junction, which is a diagnosis that must be made postnatally and is sometimes difficult. Pyelocaliectasis probably occurs as the kidneys ascend and induce new blood vessels. When the older, more inferior, vessels persist, they may pinch or angulate the ureter and result in hydronephrosis, which resolves when the offending vessels or bands are reabsorbed. Fully half the cases of fetal pyelocaliectasis resolve in this way. If vessel reabsorption occurs at a later time or stage, the pyelocaliectasis may persist, but obstruction may still be absent.

Recommended Management.—After birth, cystography is done to rule out reflux. Renal scanning (with use of diethylenetriamine pentaacetic acid or mercaptoacetyltriglycine) is usually performed at 1 to 4 weeks of age to assess renal drainage.8 Obstruction can be relieved when it can be diagnosed on the basis of a prominent palpable kidney or obstructed drainage pattern, but a considerable diversity of opinion currently exists about the criteria for diagnosis.7,8 Most clinicians believe that the dilated side should exhibit reduced function as well as a prolonged drainage curve,9,10 but a high proportion of patients with good ipsilateral function and evidence of obstruction on renography require surgical intervention after years of follow-up because of decreasing kidney function.

Those patients with equivocal drainage curves may have improvement, but obstruction occurs at the rate of 3 to 5% per year.11,12 Obstruction of the ureteropelvic junction is almost always congenital, but obstruction may not become severe until late childhood or beyond.13 The only other known cause of obstruction of the ureteropelvic junction is trauma and urinary extravasation.

Fetal ultrasonography facilitates the diagnosis of urethral valves soon after birth and helps prevent most renal scarring from urinary tract infection in babies with reflux. Better techniques are needed to diagnose or exclude ureteral obstruction during the perinatal period to achieve optimal renal salvage.14 Nonetheless, patients at risk are easy to identify and can be subjected to close follow-up.

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REFERENCES