

**Index terms:**

Posterior urethral valves  
Abdominal ultrasound  
Voiding cystourethrography  
Prenatal diagnosis of fetal abnormalities  
Urinoma  
Neonatal ascites



THIS EXHIBIT WAS DISPLAYED AT THE 71ST SCIENTIFIC ASSEMBLY AND ANNUAL MEETING OF THE RADIOLOGICAL SOCIETY OF NORTH AMERICA, NOVEMBER 17-22, 1985, CHICAGO, ILLINOIS. IT WAS RECOMMENDED FOR PUBLICATION BY THE PEDIATRIC RADIOLOGY AND ULTRASOUND PANELS AND WAS ACCEPTED FOR PUBLICATION AFTER PEER REVIEW AND REVISION ON FEBRUARY 18, 1986.

From the Departments of Radiology (\*) and Urology (†), Medical University of South Carolina, Charleston, Charleston.

Address reprint requests to R.I. Macpherson, M.D., Department of Radiology, Medical University of South Carolina, 171 Ashley Avenue, Charleston, SC 29425.

## Posterior urethral valves: An update and review

Roderick I. Macpherson, M.D.\*

Richard E. Leithiser, M.D.\*

L. Gordon, M.D.\*

W.R. Turner, M.D.†

*Here, in a comprehensive review of an important pediatric problem, the authors discuss the embryology, the clinical and radiological features, the complications and the management of posterior urethral valves.*

### Introduction

Posterior urethral valves are a common and serious cause of congenital bladder outlet obstruction in males (22). They can present in utero, in the neonate, later in childhood, and even in adults. Their pathologic effects vary in occurrence and magnitude. Because of this variability, these simple urethral obstructions can become diagnostic and management dilemmas.

In this article, we will review the current concepts of the morphology and embryology, and will present the clinical and radiological features of posterior urethral valves. Also, we will summarize their complications and the controversies surrounding their management.

### Materials and Method

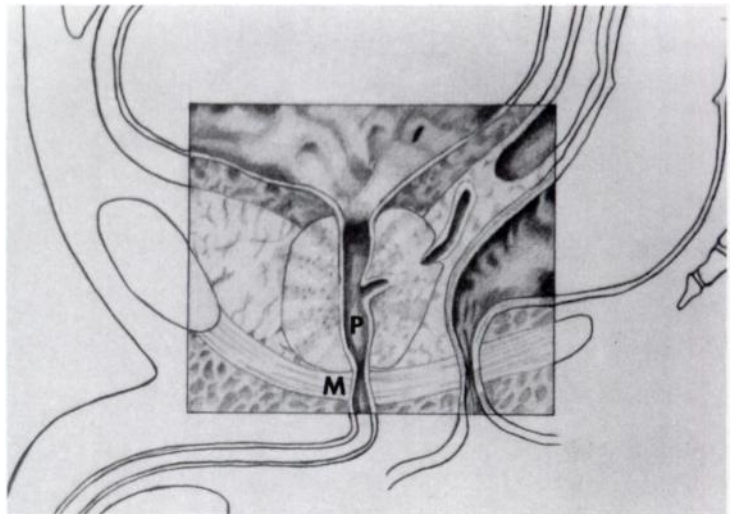
In the decade from 1975 to 1985, 47 males with posterior urethral valves were seen at the Medical University of South Carolina. Four were discovered during the course of prenatal obstetrical ultrasonography. Sixteen (34%) presented in the neonatal period and another 15 (32%) during the first year of life. Eleven (23%) were diagnosed between one and 16 years of age, while one was an adult.

The charts, radiographs and pathologic features of these 47 cases were reviewed in retrospect, and the clinical presentations, radiologic features, treatment and complications were tabulated. Using this information, along with illustrative examples, an update and review of the diagnostic and management problems concerned with posterior urethral valves was formulated.

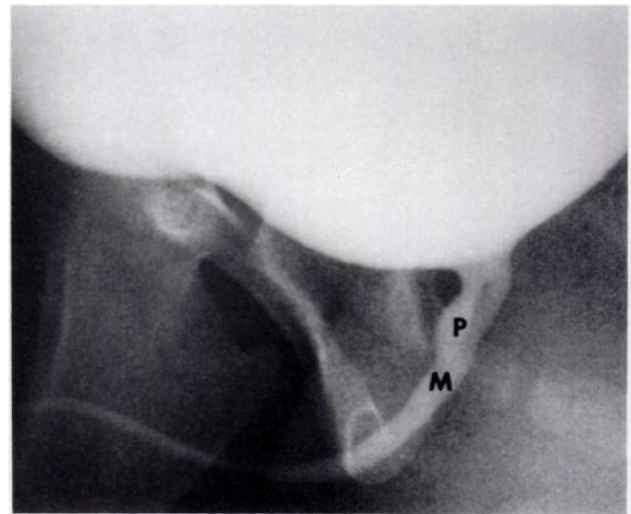
**Morphology**

**Figure 1**

**The male urethra** The male urethra is divided into five anatomic divisions. The glandular portion, the cavernous or penile portion, and the bulbous portion form the "anterior urethra." The prostatic portion, P, that part within the prostate gland, and the membranous portion, M, the short part traversing the pelvic floor, are the "posterior urethra." It is with the posterior urethra that we are mainly concerned here.



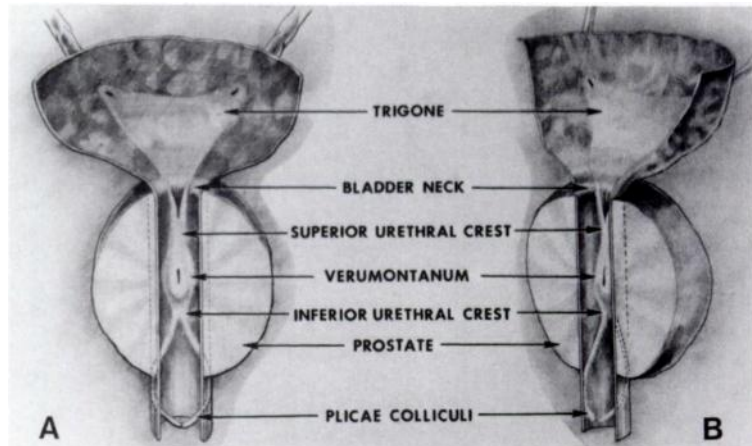
**1A**



**1B**

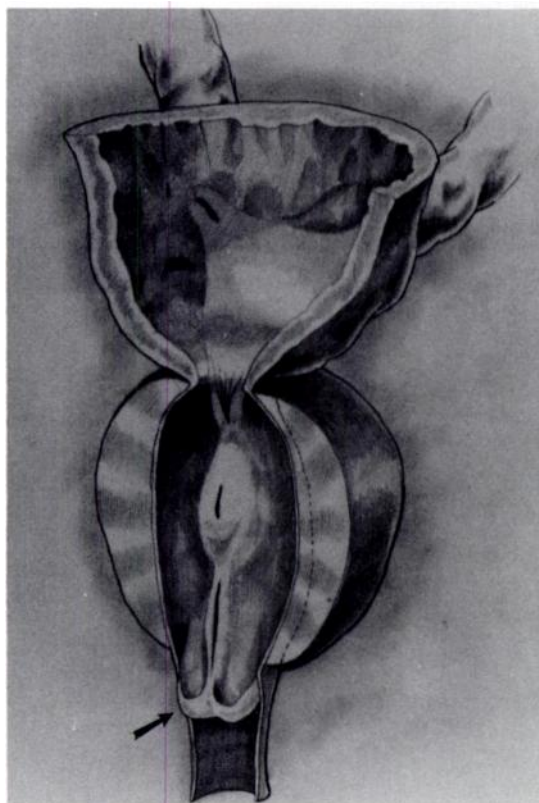
**Figure 2**

**The posterior urethra (21)** The important anatomic structures concerned in the morphology of posterior urethral valves are shown in these frontal (A) and oblique (B) diagrams of the bladder base and posterior urethra. The inferior urethral crest, the midline extension of the verumontanum, terminates in two to four plicae colliculi (or fins). These pass downward while encircling the urethra, ending anteriorly close to the midline at a more caudal level.



**A**

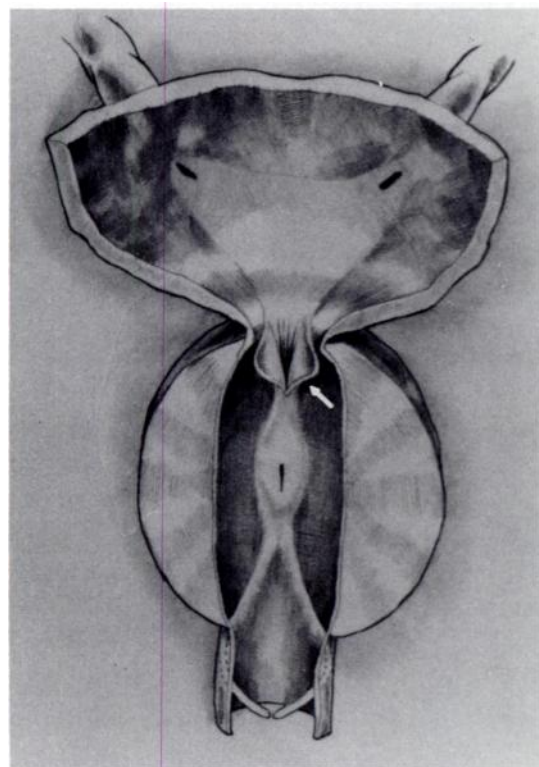
**B**



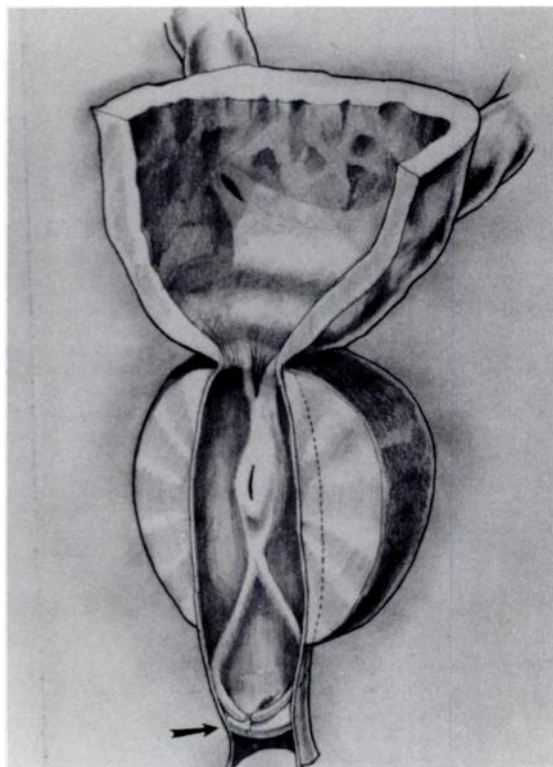
3A

**Figure 3**

**Types of posterior urethral valves** The original classification of the types of posterior urethral valves proposed by Young (27) in 1919 is still in use today. (A) Type 1 posterior urethral valves (arrow) are mucosal folds extending anteroinferiorly from the caudal aspect of the verumontanum, often fusing anteriorly at a lower level. They are derived from the plicae colliculi and constitute the vast majority of valves. (B) Type 2 posterior urethral valves (arrow) are mucosal folds extending anterosuperiorly from the verumontanum toward the bladder neck. A rare occurrence, they are probably an effect rather than a cause of bladder obstruction. (C) Type 3 posterior urethral valves (arrow) are disc-like membranes located below the verumontanum and unrelated to it. They constitute a small percentage of posterior urethral valves.

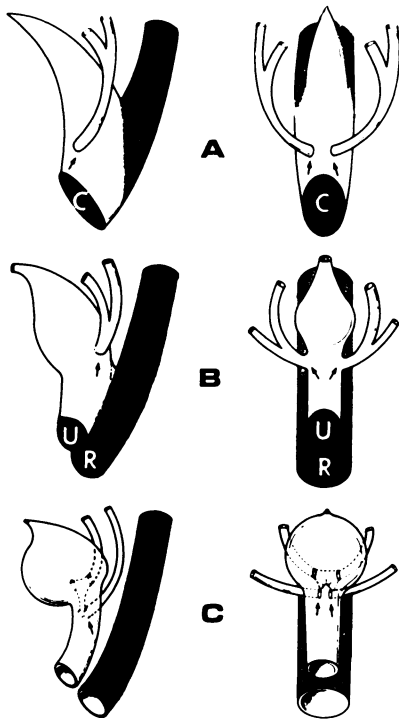


3B

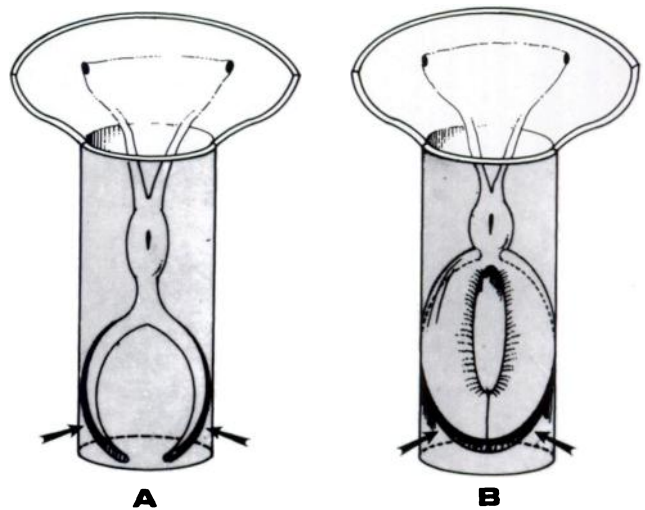


3C

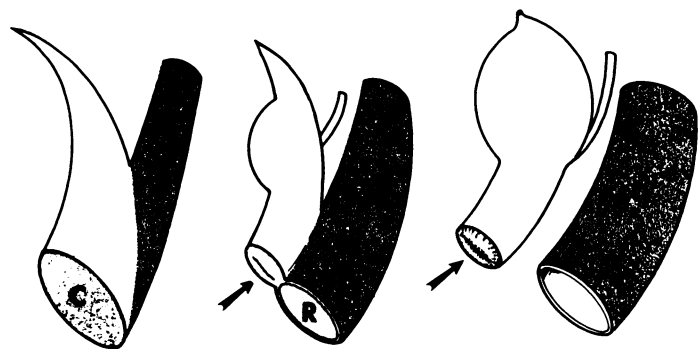
**Embryology**



**Figure 4**  
**Embryology of the posterior urethra (21)**  
 (A) In the early embryo, the orifices of the Wolffian ducts (arrows) are located anterolaterally on the cloaca close to the intact cloacal membrane, C. (B) As the cloaca is divided by the urorectal septum into the bladder and proximal urethra anteriorly, and the rectum posteriorly, the Wolffian duct orifices (arrows) migrate posterolaterally. Meanwhile, the cloacal membrane divides into the urogenital membrane, U, and the rectal membrane, R. (C) With the division of the cloaca completed, the Wolffian duct orifices now rest posteriorly on the urethra at the verumontanum (arrows) as the ejaculatory ducts. The ureters have inserted into the top of the trigone. Urogenital and rectal membranes are now perforated.



**Figure 5**  
**Embryology of Type 1 posterior urethral valves (21)**  
 (A) Normally, the vestiges of the migrating Wolffian duct orifices form the thin, pliable colliculi (arrows). (B) Type 1 valves occur when the Wolffian duct orifices originate too far anteriorly. Normal migration is altered and thick, rigid fused valve cusps (arrows) replace the plicae colliculi.



**Figure 6**  
**Embryology of Type 3 posterior urethral valves (21)** Type 3 posterior urethral valves are the result of faulty perforation of the urogenital membrane (arrow).

### Pathophysiology

The pathophysiology (Figure 7) and subsequent pathologic effects (Figures 3A, 3C and 18F) of posterior urethral valves must be understood in order for one to appreciate their clinical and radiologic features as well as the problems in their management (13).

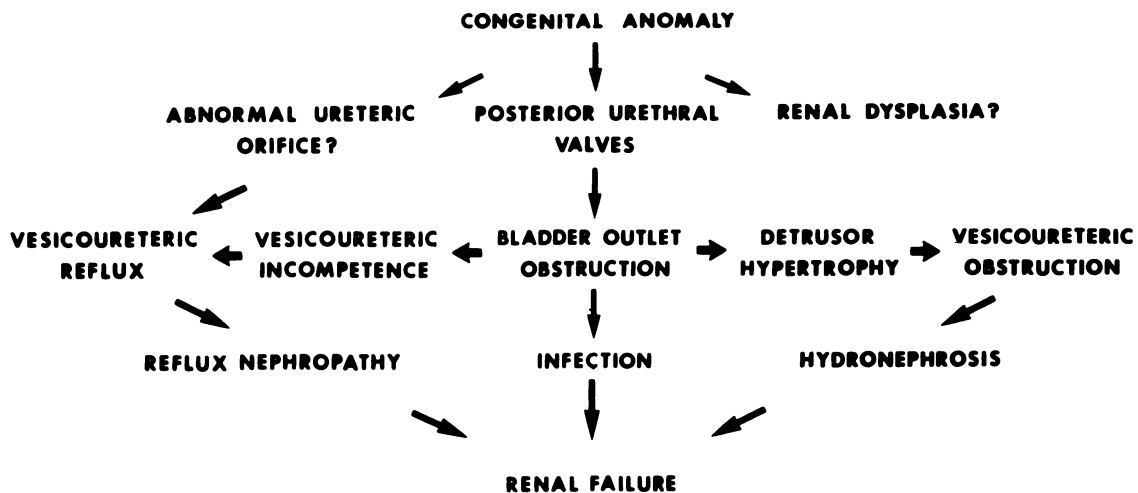
Posterior urethral valves are congenitally occurring thick folds of mucous membrane located in the posterior urethra distal to the verumontanum. They obstruct the outflow of urine from the bladder which responds to the increased resistance by detrusor hypertrophy. As a result, its walls become thickened with increased trabeculation, sacculation and diverticulum formation. The urethra proximal to the valves undergoes a fusiform dilatation while the bladder neck remains relatively narrow. There is some controversy as to whether or not this "bladder neck hypertrophy" is a secondary cause of obstruction (24). The superior mucosal plications or supramontane ridges also become more prominent in response to the more distal obstruction. These thickened folds are probably Young's Type 2 posterior urethral valves (Figure 3B), a result rather than a cause of urethral obstruction (13).

The hypertrophied detrusor and trigone may alter the vesicoureteric junction in two ways: one, to cause vesicoureteric incompetence with reflux; and the other, to cause vesicoureteric obstruction. Either way, massive hydroureteronephrosis can result. The back pressure in the pelvicalyceal system may lead to leakage of urine into the perirenal space (urinoma), peritoneal cavity (urine ascites), or pleural space (urothorax), with or without a demonstrable tear in the collecting system. Vesicoureteric reflux may occur as a result of a primary abnormality of the ureterovesical junction or as a result of the presence of a periureteric (Hutch) diverticulum (13). An early onset of vesicoureteric reflux in fetal life may interfere with normal renal development, resulting in renal dysplasia. These kidneys may be "hypodysplastic", "multicystic dysplastic", or both (9) (Figure 8). Although it has been suggested that posterior urethral valves, unilateral (usually left) vesicoureteric reflux, and renal dysplasia may be associated congenital anomalies within a syndrome (12), an intrauterine acquired etiology for the reflux and associated renal dysplasia seems more likely (10).

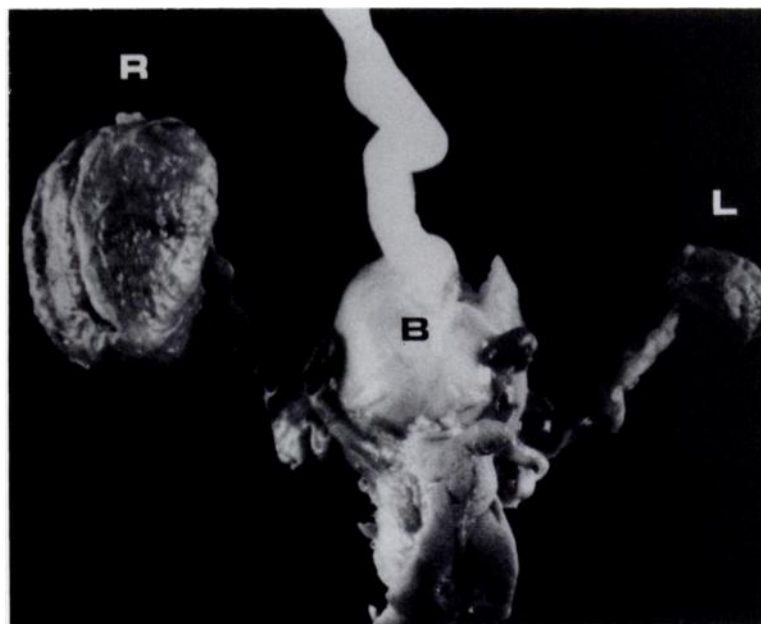
Vesicoureteric obstruction secondary to detrusor and trigonal hypertrophy can cause a massive hydroureteronephrosis (22). Persistence or progression of this dilatation following valve ablation with relief of the urethral obstruction may become a serious management problem. This is in large part due to the relentless ureteral obstruction caused by the thickened, non-compliant bladder, but the kidney (secondary nephrogenic diabetes) and ureters (dilated and aperistaltic) also contribute to this so called "valve bladder syndrome" (17).

Persistent hydroureteronephrosis, whether associated with vesicoureteric reflux or not, can progress through renal dysplasia, reflux nephropathy or chronic pyelonephritis to chronic renal failure.

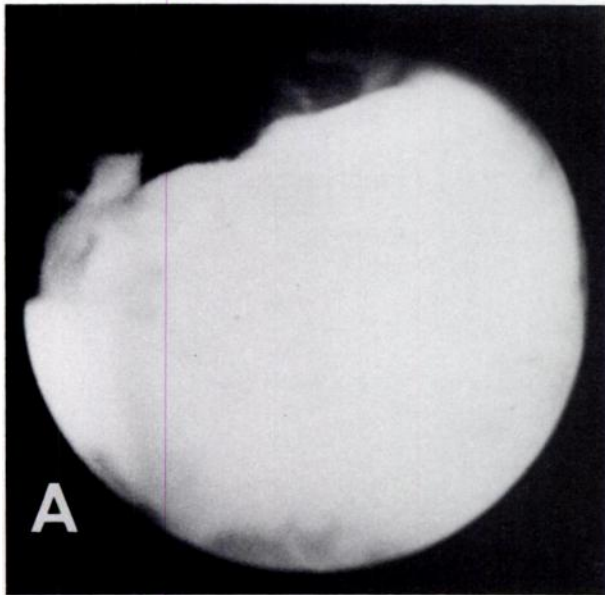
Pathophysiology



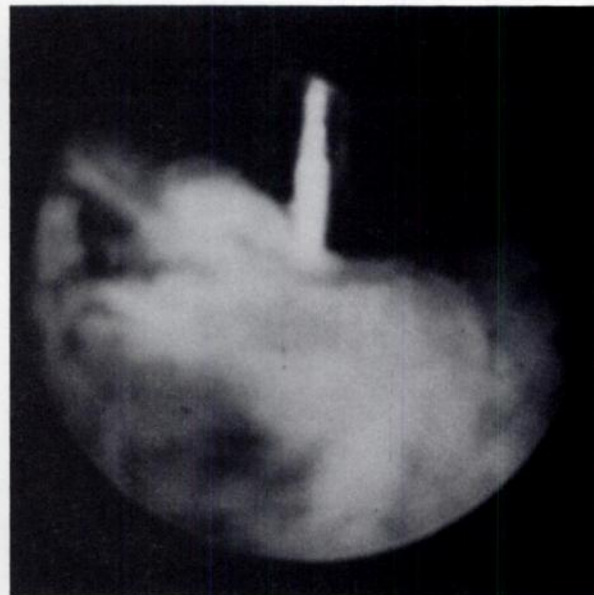
**Figure 7**  
Pathophysiology of posterior urethral valves



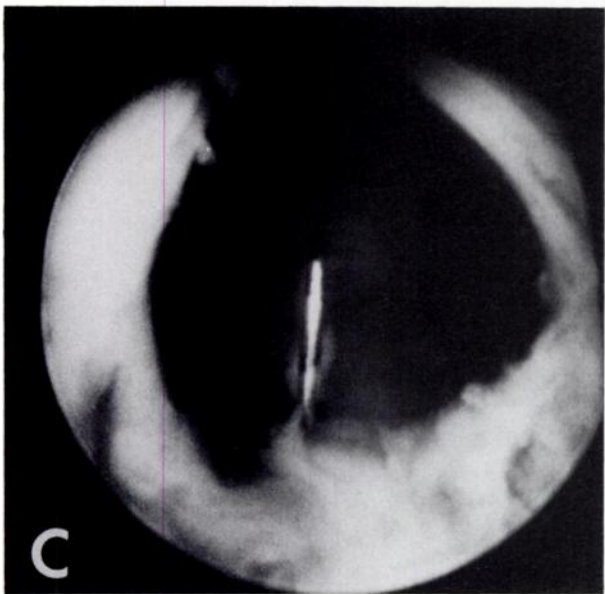
**Figure 8**  
**Renal dysplasia** This pathologic specimen is from a stillborn fetus with posterior urethral valves, a large bladder, B, and bilateral hydronephrosis. Both kidneys are dysplastic; the left, L, being "hypodysplastic" and the right, R, "multicystic dysplastic." This renal dysplasia is acquired in fetal life as the result of early onset of obstruction or vesicoureteric reflux or both, interfering with normal renal development.



9A



9B



9C

**Figure 9**  
**Cystoscopic findings** These photographs were made through a 13F resectoscope introduced into a six day old infant's urethra using a transperineal approach. (A) The fused intact valves seen through this scope must be differentiated from normal plicae colliculi. An opening anteriorly at 12 o'clock permits a glimpse of the posterior urethra proximal to the valves. (B) The resectoscope loop is engaging the left valve leaflet. (C) Ablation of the valves is completed and the posterior urethra beyond is now in full view.

## Clinical Features

The presenting symptoms and their incidence within our series are listed in Table I. These clinical manifestations fall mainly into two groups: those caused by obstruction and those caused by infection (20). The obstructive signs and symptoms are most prevalent in neonates, while symptoms resulting from infection tend to be more common in older children. Many of these patients have biochemical evidence of depressed renal function at the onset. Generally, the younger the child, the greater the magnitude of renal failure. In most, there is a rapid restoration of renal function following valve resection. Some, however, progress to chronic renal failure with all its stigmata, in spite of surgery. Glomerular filtration rate (GFR) is the renal function test that best correlates with recovery of renal function following valve ablation (20). If the preoperative GFR is less than 10 ml/min/1.75m<sup>2</sup>, improvement in renal function is not likely (1).

Presentation	In Our Series
Prenatal sonographic diagnosis	4
Neonatal urine leak	6
ascites, urothorax, urinoma	
Palpable kidneys and/or bladder in neonate	10
Obstructive symptoms	15
straining to void, dribbling, incontinence, etc.	
Urinary tract infection	17
fever, vomiting, jaundice, hematuria, etc.	
Failure to thrive	6
Neonatal pneumothorax or pneumomediastinum	4
Incidental finding at surgery	1
Unknown (diagnosis elsewhere)	3

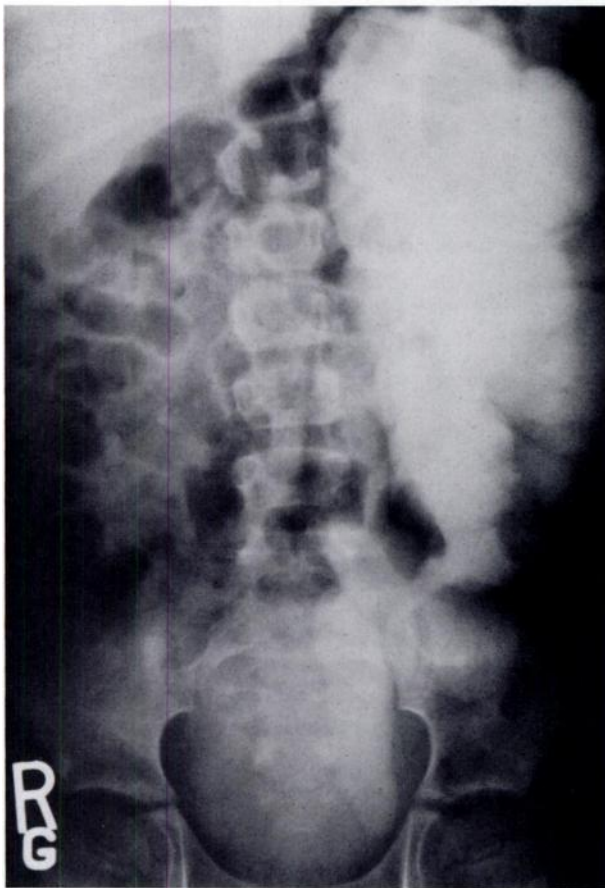
## Radiologic Investigation

The radiologic investigation of male patients suspected of posterior urethral valves is geared to demonstrate, not only the valves, but also the anatomic and functional alterations in the kidneys, ureters and bladder proximal to them.

Excretory pyelography (Figures 10 and 17A) is best utilized in older children with normal renal function. It can provide a good estimation of differential renal function, demonstrate the presence and degree of hydronephrosis, and pinpoint the level of obstruction. In infants and in older children with impaired renal function, excretory pyelography is less effective. In these patients, renal sonography (6) can demonstrate all the anatomic changes proximal to the urethral

obstruction (Figure 11). Also, it is the obstetrical use of sonography that is responsible for the prenatal diagnosis of posterior urethral valves (Figures 18 and 19).

Renal isotope studies, using technetium 99m labeled diethylenetriaminepentaacetic acid (99m Tc-DTPA) can provide the functional information lacking in renal sonograms. Further, diuretic radionuclide urography (Figure 12) can be used to distinguish between obstructive and non-obstructive hydronephrosis, a test that is particularly useful in postoperative patients. Urinomas (Figure 25D) and urine ascites (Figure 27D) can be identified through renal isotope studies using delayed images (16).



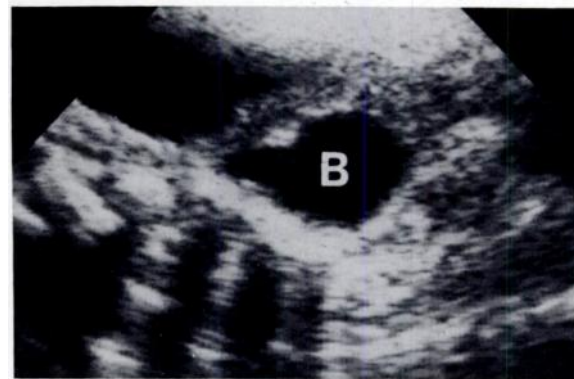
**Figure 10**  
**Excretory pyelography** This 7 year old boy had a long history of difficult micturition and of recurrent urinary tract infections. Excretory pyelography shows reduced renal function bilaterally with function on the right worse than on the left. There is massive hydronephrosis. The bladder distention indicates urethral obstruction. A urethral catheter could not be passed. Type 3 valves were found at cystoscopy.



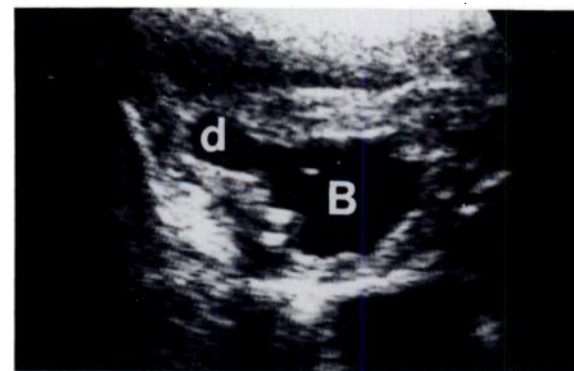
11A



11B



11C



11D

**Figure 11**  
**Sonography** A diagnosis of posterior urethral valves was made on the basis of the prenatal obstetrical sonographic study in this newborn boy. The postnatal sonogram confirms it. (A) A longitudinal scan of the left kidney, K, shows hydronephrosis. The thickness of the renal cortex can be evaluated. (B) A longitudinal scan shows the dilated tortuous left ureter, U. (C) A longitudinal scan shows the large, thick walled bladder, B. (D) A transverse scan of the bladder, B, shows multiple diverticula, d.

Computed tomography is not recommended for the investigation of children suspected of having posterior urethral valves. When performed in unsuspected cases (Figure 13), however, the characteristic hydroureteronephrosis and thickened, distended bladder can be demonstrated and a correct diagnosis established.

Voiding cystourethrography remains the definitive test for the demonstration of posterior valves (13). The retrograde catheterization of the bladder in children with the more commonly oc-

curing Type 1 valves is usually unimpeded. In those with Type 3 valves, on the other hand, catheterization may prove difficult (13). The bladder itself may have a large capacity with a thick irregular wall showing increased trabeculation, sacculation and multiple diverticula (Figures 14 and 15). Massive unilateral or bilateral vesicoureteric reflux (Figures 16A and 23) may be apparent and the presence of periureteric diverticula (Figure 15) in association with vesicoureteric reflux is not uncommon.



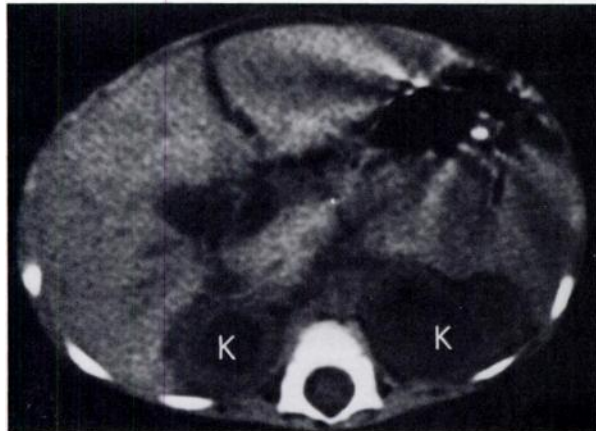
12A



12B

### Figure 12

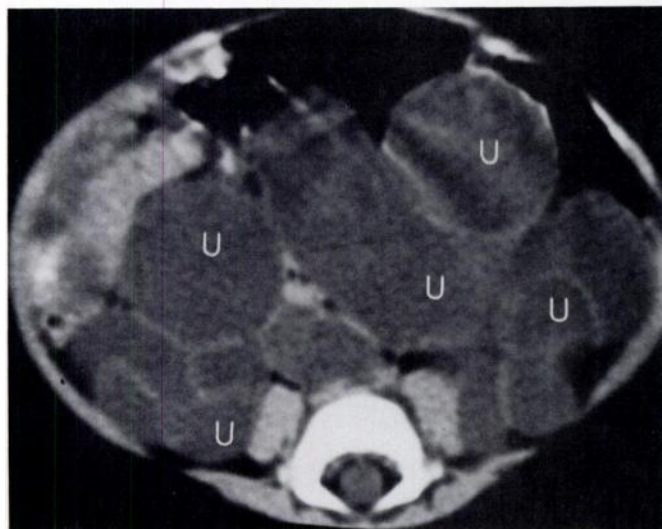
**Radioisotope studies (scintigraphy)** This 14 year old boy had posterior urethral valves resected at 10 months of age and was followed by annual  $^{99m}\text{Tc}$ -DTPA renal scans. (A) The early images showed bilateral hydronephrosis. At 20 minutes, when the renal pelvis and calyces were visualized, a diuresis was stimulated by furosemide, 0.3 mg/kg. (B) A later image, 20 minutes after the administration of the diuretic, shows persistent pelvicalyceal distention. The presence of radioactivity in both ureters and the bladder, however, suggests dilated, but unobstructed ureters.



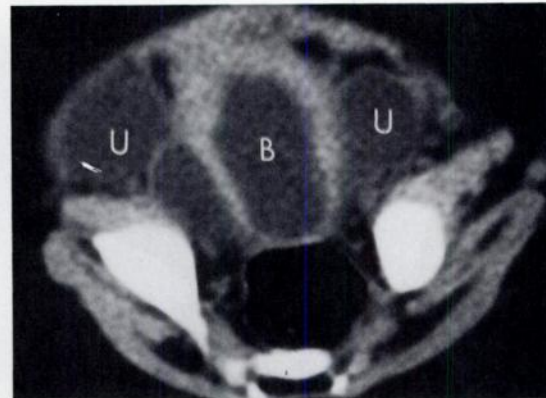
13A



13B



13C



13D

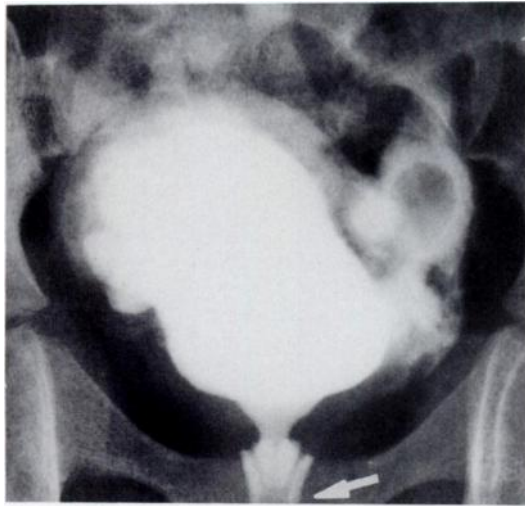
**Figure 13A-D**

**Computed tomography** These abdominal computed tomograms came from a referring hospital with a 10 month old infant who was being investigated for failure to thrive. Because of decreased renal function, no contrast medium was used. The hydronephrotic kidneys, K, and dilated tortuous ureters, U, were misinterpreted as "cystic kidneys". Note the marked thickening of the wall of the bladder, B.

During voiding, mild (Figure 17B) to marked (Figure 16B) fusiform dilatation of the posterior urethra will be seen. This dilatation terminates abruptly at the level of the valves. Occasionally, the valve cusps are perceptible as filling defects in the stream of contrast medium (Figure 15). An enlarged verumontanum (Figure 14), thickened plicae colliculi (Figure 26B), and prominent

superior mucosal plications (Figure 14) may be perceptible within the dilated posterior urethra. The bladder neck will appear relatively narrow (Figures 14 and 15).

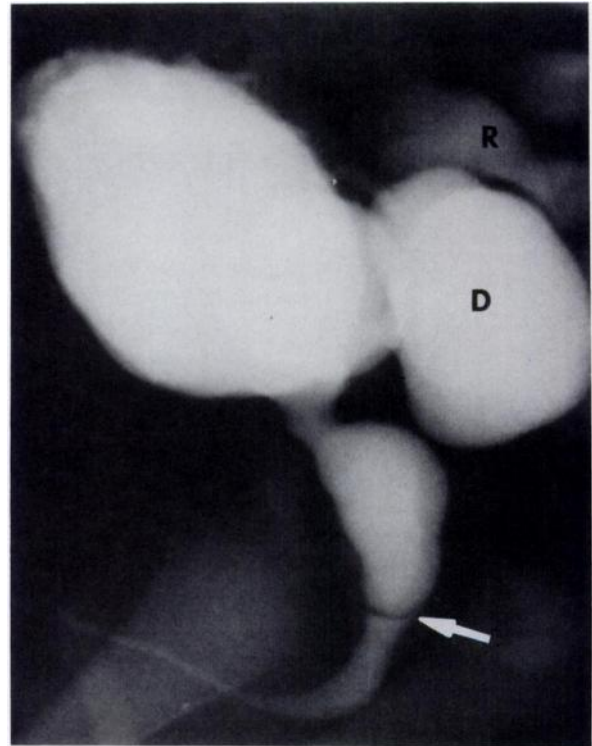
Following valve ablation, these radiologic features will revert to normal (Figure 34) in uncomplicated cases.



**Figure 14**

**Voiding cystourethrography: bladder findings**

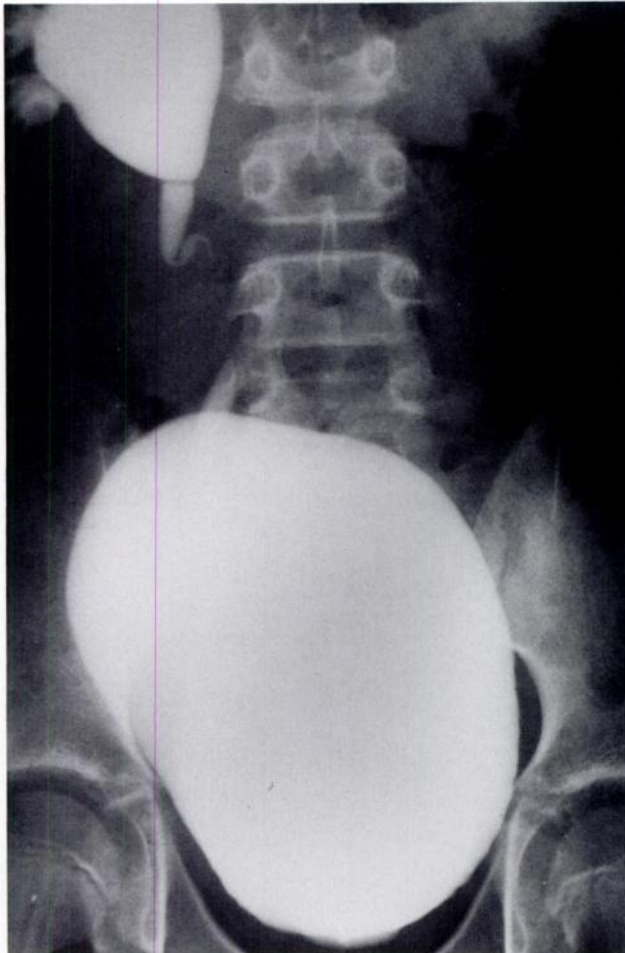
This 2 year old boy had a history of difficult micturition. This postvoiding radiograph from a cystourethrogram demonstrates many of the bladder findings of posterior urethral valves. There is a large postvoiding residuum in the bladder, and the bladder wall is thickened and irregular, showing multiple diverticula of varying sizes. Note the dilated posterior urethra, enlarged verumontanum (arrow) and superior mucosal plications along with relative narrowing of the bladder neck.



**Figure 15**

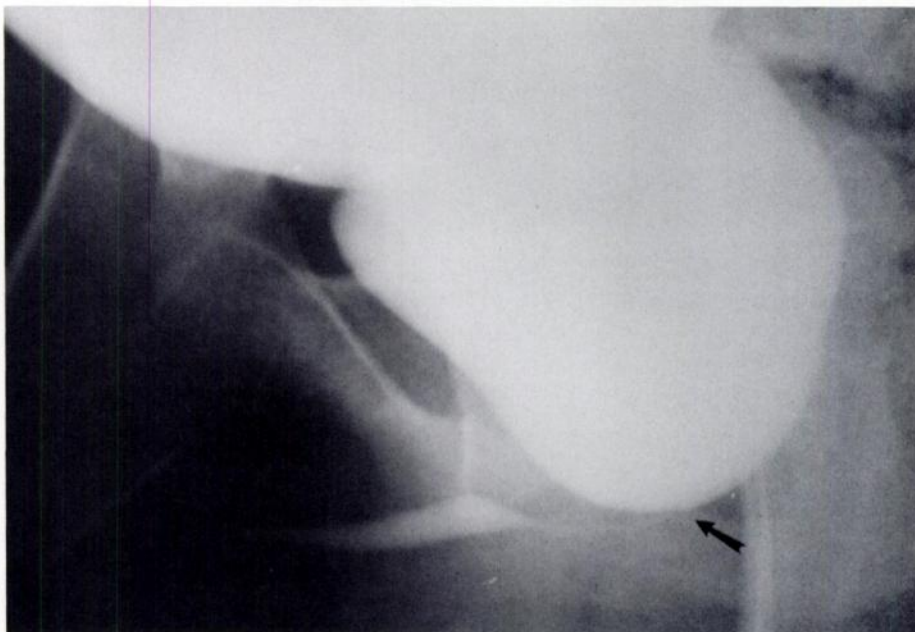
**Voiding cystourethrography: ureteral findings in a neonate**

The voiding radiograph of this newborn with posterior urethral valves shows the urethral obstruction (arrow), the dilated posterior urethra proximal to it, the relative bladder neck narrowing, bilateral periureteric diverticula, D, and vesicoureteric reflux, R. This constellation of findings is consistent with posterior urethral valves.

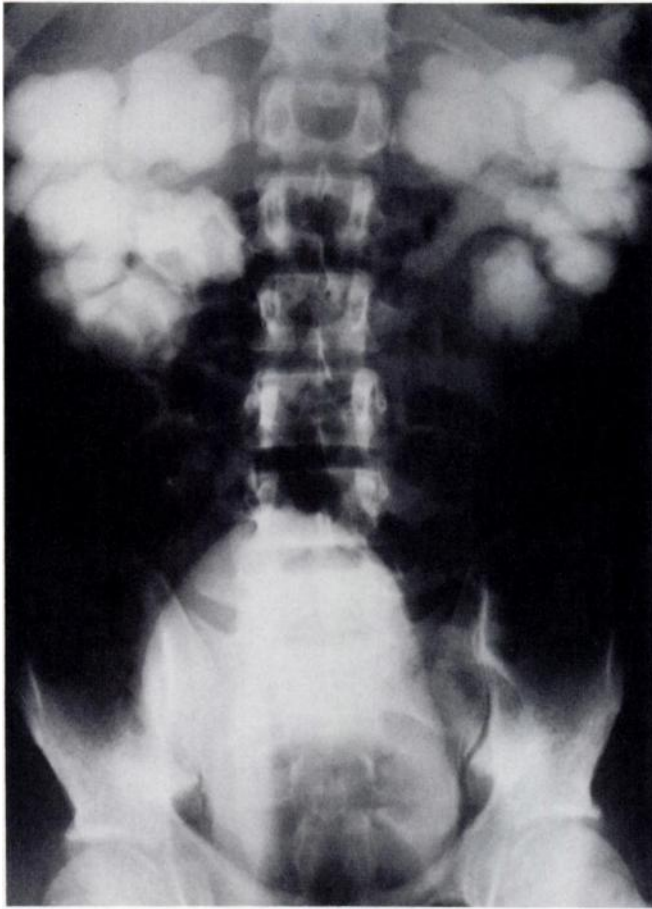


16A

**Figure 16**  
**Voiding cystourethrography in an older child** This 9 year old boy with a long history of difficult micturition was thought to have a neurogenic bladder. (A) A filling radiograph of the voiding cystourethrogram shows a large capacity bladder with right vesicoureteric reflux. (B) The voiding radiograph shows a massively dilated posterior urethra with an abrupt change of caliber at the site of posterior urethral valves (arrow). The diagnosis was confirmed at cystography and the valves were resected.



16B



17A



17B

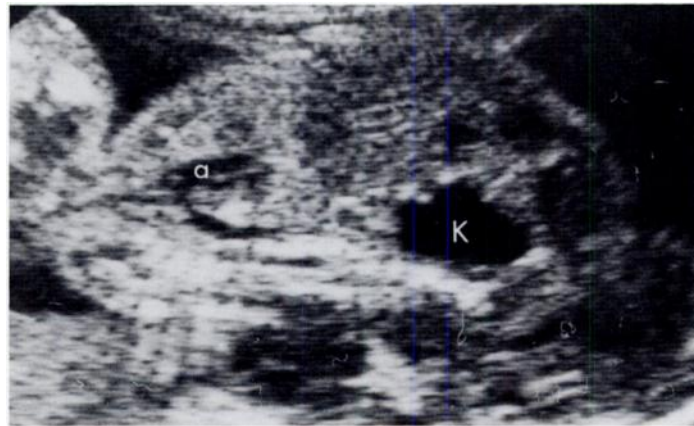
**Figure 17**

**Posterior urethral valves in an adult** This 26 year old man had a life long history of difficult micturition and recurrent urinary tract infection. (A) Excretory pyelography shows marked bilateral hydronephrosis and a large capacity bladder. (B) Voiding cystourethrography shows urethral obstruction (arrow) with modest dilatation of the posterior urethra. Posterior urethral valves were subsequently found and resected at cystoscopy.

### Prenatal Diagnosis

The increased use of sophisticated prenatal sonography in recent years has led to an increased recognition of congenital urinary tract anomalies (2 per 1,000 pregnancies) (19). The kidneys with their echolucent parenchyma and echodense renal sinuses can be identified at 14 weeks. The fetal bladder can be seen at about the same time and should partially empty once per hour. Normal ureters should not be seen. Therefore, sonographic evidence of bladder outlet obstruction can be perceptible relatively early in fetal life. The major hope for lowering the incidence and severity of chronic renal failure in children with posterior urethral valves lies in the antenatal diagnosis followed by either intrauterine surgery or early delivery of the fetus (20).

The fetus with bladder outlet obstruction may be difficult to evaluate because of associated oligohydramnios. Dilated renal pelvis (Figure 18A), enlarged tortuous ureters (Figure 19C), an overfilled, thick-walled bladder (Figures 18E, 19B and 19C), and, occasionally, a distended posterior urethra (Figure 18D) may be perceptible. Diffuse fetal ascites or localized perirenal fluid collections indicative of internal decompression may be present (2). The pelvicalyceal dilatation is often relatively mild, even in the face of massive bladder and ureteric distension (Figure 19) (9). The kidneys may be small and dysplastic (Figure 19D), with or without demonstrable cysts (9). Occasionally, a small thorax, due to oligohydramnios and subsequent pulmonary hypoplasia, is observed and is another indicator of bladder outlet obstruction.

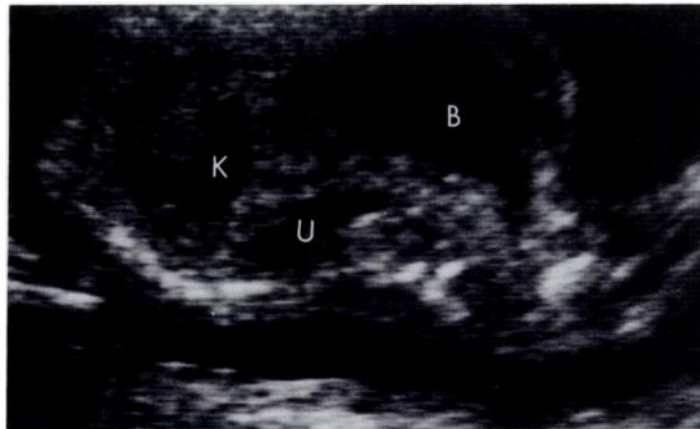


18A

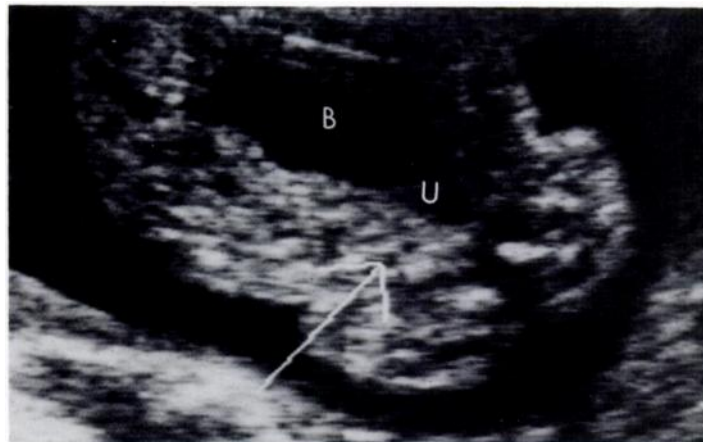


18B

**Figure 18A&B**  
**Prenatal diagnosis of posterior urethral valves** Prenatal obstetrical sonography demonstrates the diagnostic features of posterior urethral valves in this 22 week fetus. (A) An off-midline sagittal scan shows the aorta, a, and a hydronephrotic kidney, K. (B) Another sagittal scan shows a hydronephrotic kidney, K, and a distended bladder, B.



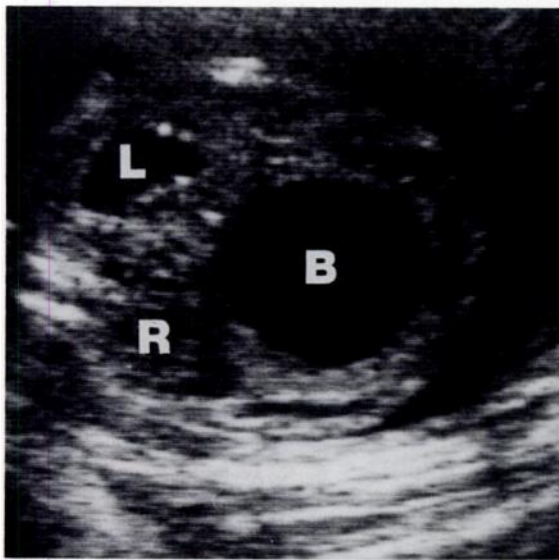
18C



18D

**Figure 18C&D**

(C) A further sagittal scan shows a hydronephrotic kidney, K, ureter, U, and distended bladder, B, with a dilated posterior urethra. (D) A transverse scan of the pelvis (with caudal angulation) shows the bladder, B, and dilated posterior urethra, U.



**Figure 18E&F**  
(E) A transverse scan of the abdomen shows the left, L, and right, R, hydronephrosis and a distended bladder, B. (F) At autopsy, hydronephrotic dysplastic kidneys and a dilated, thick walled bladder were found. The apparent intravesical "mass" is artifactual.

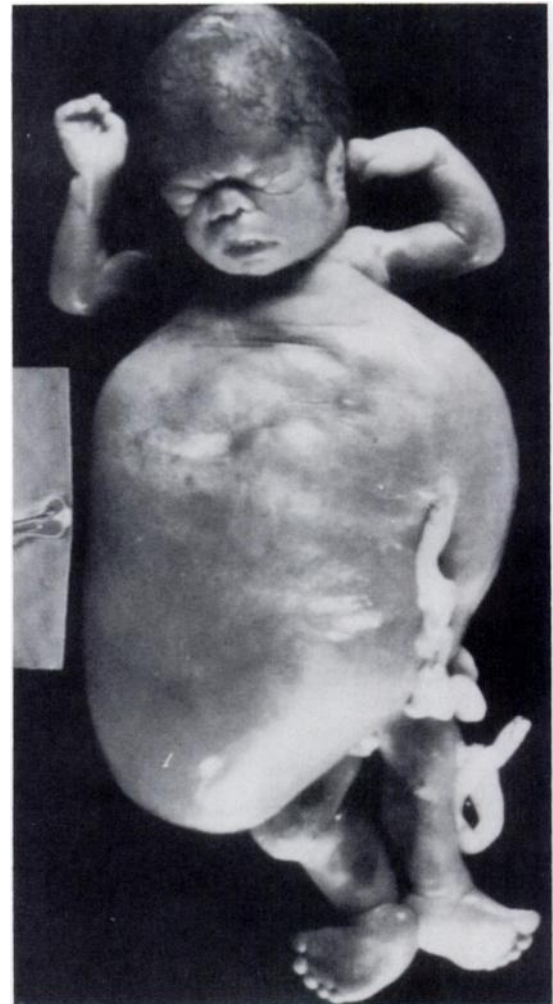
**18E**



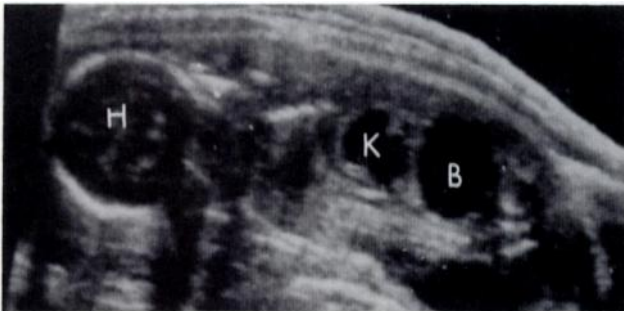
**18F**

**Figure 19A,B&C**

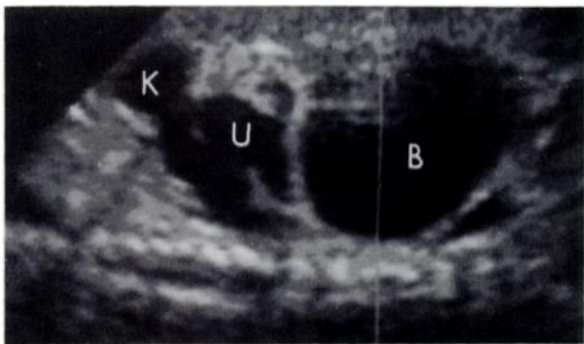
**Prenatal diagnosis of posterior urethral valves** The prenatal sonographic findings, postnatal radiologic findings and pathologic findings are correlated in the case of this stillborn infant with posterior urethral valves. (A) The infant had a markedly distended abdomen, but intact abdominal musculature. (B) A longitudinal prenatal sonogram shows the fetal head, H, a dilated renal pelvis, K, dilated bladder, B, and oligohydramnios. (C) A longitudinal prenatal sonogram of the abdomen shows a dilated renal pelvis, K, a dilated, tortuous ureter, U, and a distended bladder, B.



**19A**



**19B**



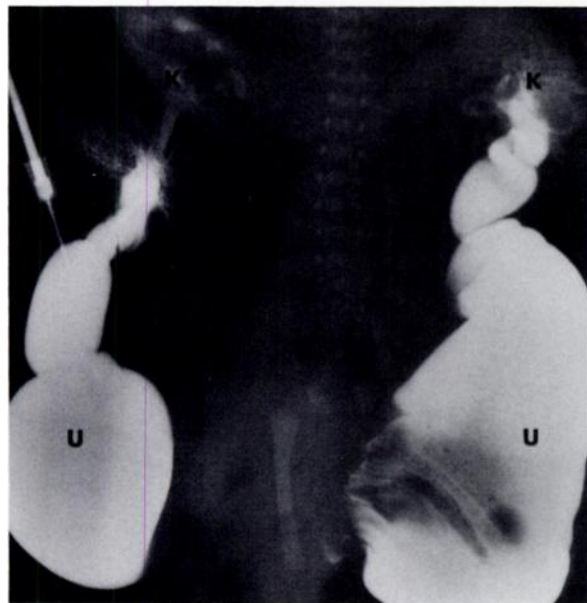
**19C**



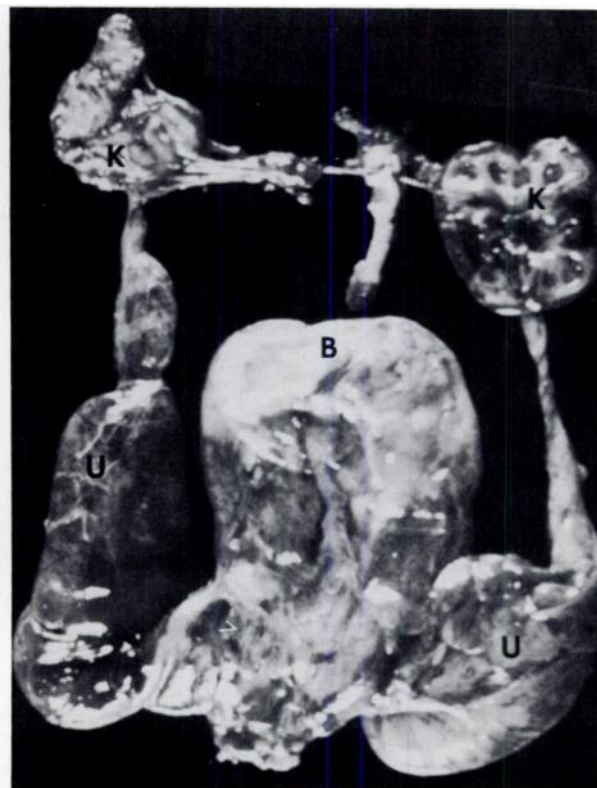
19D

Figure 19D,E&F

(D) A coronal prenatal sonogram shows small dysplastic, hydronephrotic kidneys, K, and massive hydroureters, U, bilaterally. (E) A postmortem contrast study correlates with the findings of the sonogram, showing the markedly dilated ureters, U, and the small dysplastic, yet hydronephrotic, kidneys, K. (F) The abnormalities suspected on the basis of sonography are confirmed by the gross pathology which shows marked bladder (B) enlargement, massive ureters, U, and dysplastic kidneys, K.



19E



19F

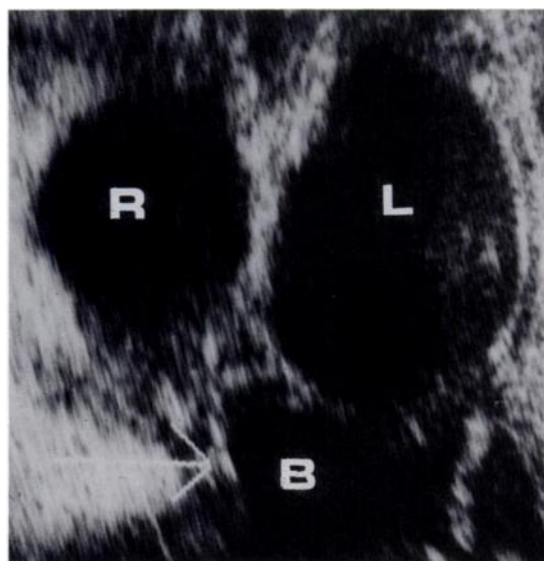
There are other causes of bilateral hydronephrosis that must be differentiated from posterior urethral valves. For example, transient hydro-ureteronephrosis and bladder distention may occur as a normal variant during fetal life (19). Bilateral, congenital, pelviureteric obstruction will show bilateral hydronephrosis with normal ureters and bladder (11) (Figure 20A). The "prune belly syndrome" can be indistinguishable from posterior urethral valves, displaying hydroureteronephrosis, a dilated bladder, and even a distended posterior urethra. Further, both diseases occur almost exclusively in males. It has been suggested that "prune belly syndrome" is a continuum of posterior urethral valves in a spectrum best called the "urethral obstruction malformation complex" (18).

There are certain megacystis-megaureter syndromes that can simulate the sonographic appearance of posterior urethral valves. One of

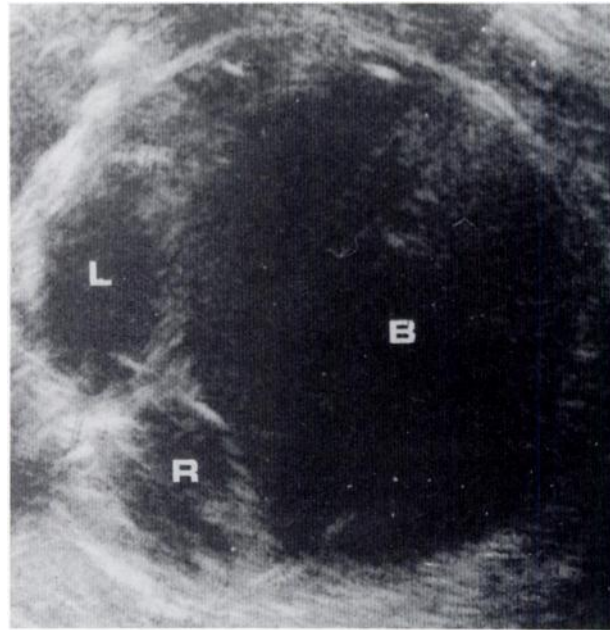
these, the "megacystis-microcolon-intestinal hypoperistalsis syndrome", will display a disproportionately distended bladder along with bilateral hydroureteronephrosis (Figure 20B). Because of the associated abnormal gastrointestinal motility and functional obstruction, these patients may have polyhydramnios rather than oligohydramnios (22). Further, this disease process is more common in females, so if intrauterine sex differentiation can be made with sonography, it could prove helpful in differentiating this syndrome from posterior urethral valves (11).

Other conditions that might enter the differential diagnosis of posterior urethral valves include multicystic kidneys (Figure 20C), polycystic kidneys and ovarian cysts (19). In addition to other sonographic differences, all of these conditions should have normal bladders.

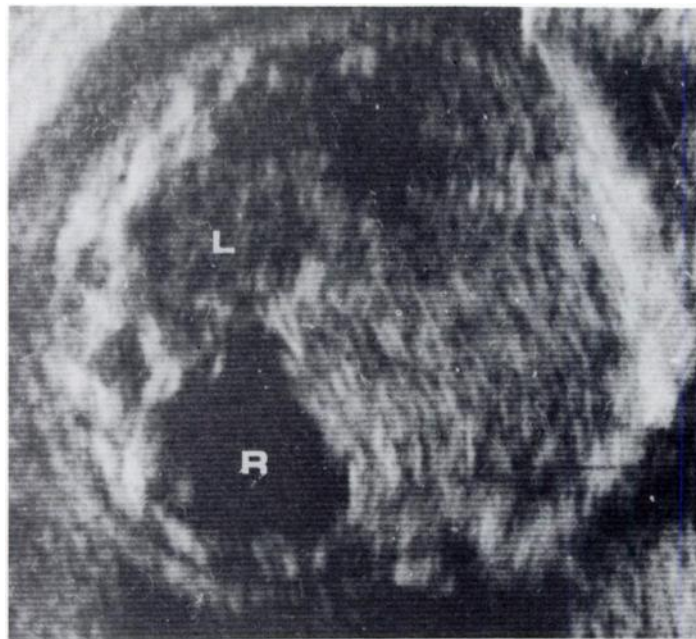
**Figure 20**  
**Differential diagnosis of posterior urethral valves on prenatal sonogram** (A) Bilateral ureteropelvic obstruction. This transverse scan shows left, L, and right, R, hydronephrosis with normal ureters and bladder. (B) Megacystis-microcolon-intestinal hypoperistalsis syndrome. This transverse B-mode, contact, prenatal maternal sonogram made at a gestational age of 33 weeks shows left, L, and right, R, hydronephrosis and a markedly distended bladder, B. These findings are similar to the findings of posterior urethral valves. Polyhydramnios was present, however. (C) Multicystic dysplastic kidney. This transverse sonogram shows a normal left kidney, L, and cystic replacement of the right kidney, R. The absence of ureteric and bladder dilatation are important differentiating features.



20A



20B



20C

**Urinomas**

Urinomas (uriniferous perirenal pseudocysts) are cystic masses formed by encapsulated extravasated urine (16). They tend to occur when continuing renal function is associated with a tear in the collecting system and distal obstruction. In all other age groups, trauma is the most common cause. In neonates, however, lower urinary tract obstruction, usually from posterior urethral valves, is more common. Urinomas can be regarded as "a blessing in disguise"; temporary decompression of a hydronephrotic system in this fashion can contribute to the long term preservation of renal function. Urinomas are usually demonstrated by excretory urography, voiding cystography (if reflux is present) or sonography, and they may be identified by renal scintigraphy as well (Figure 25).

There are four types of urinoma (Figure 21):

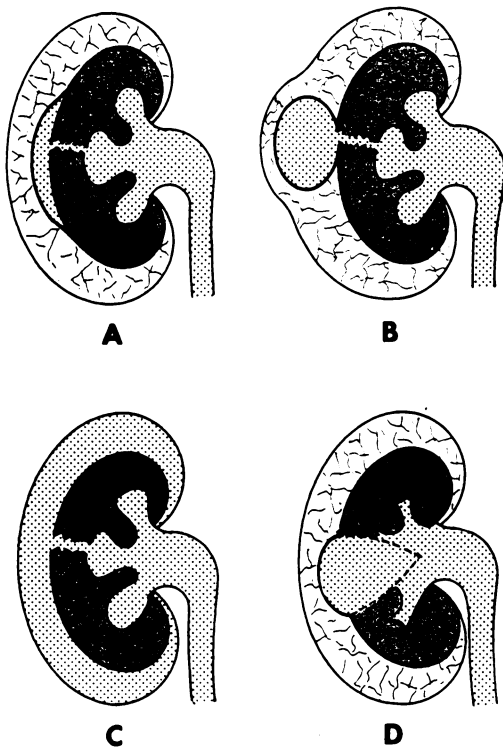
1, Subcapsular urinomas are extravasations of urine that localize between the intact renal capsule and the cortex (Figure 22). They typically

have a "C" or "J" shape depending on their location on the kidney and are frequently associated with urine ascites (1).

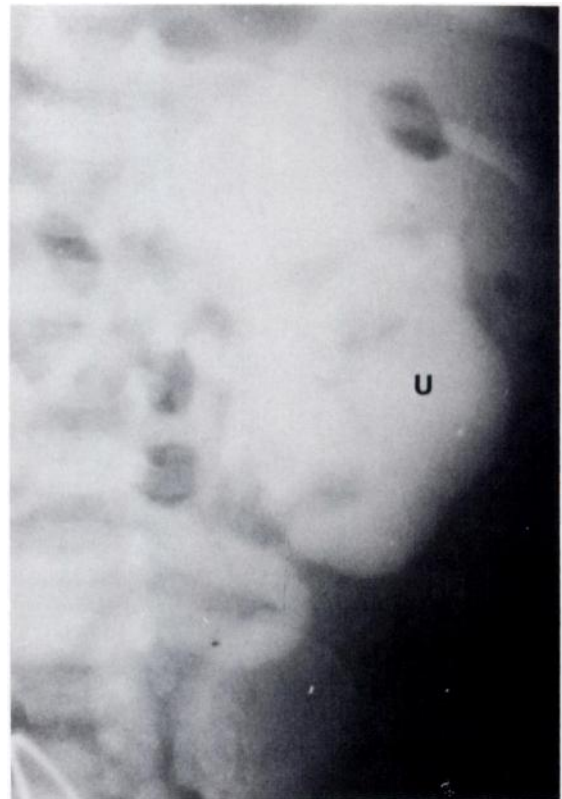
2, Localized perirenal urinomas (Figures 23 and 25) break through the renal capsule into the perirenal space where they wall off, forming localized, expanding cystic masses that can displace the affected kidney.

3, Diffuse perirenal urinomas occur when the extravasated urine fills the entire perirenal space surrounding the undisplaced, affected kidney (Figures 24 and 27C).

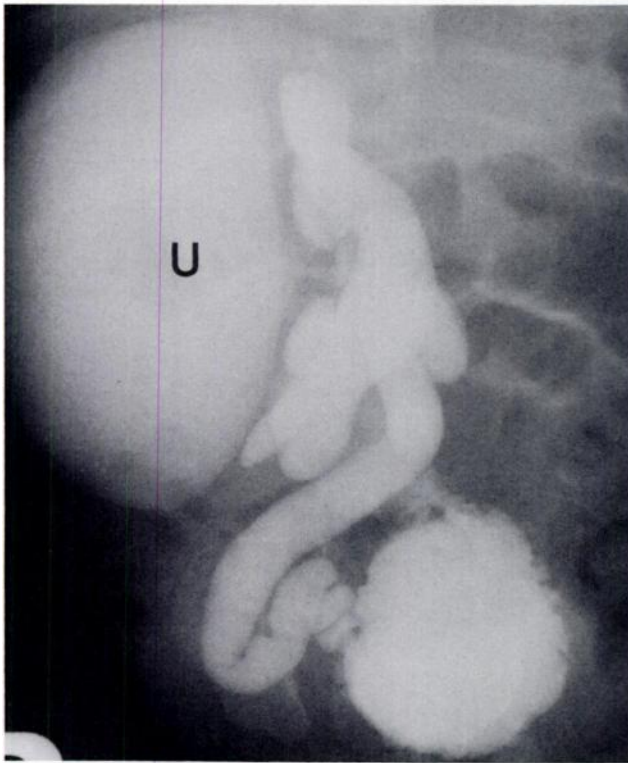
4, Intrarenal urinomas are usually the product of renal trauma and are encapsulated, expanding, intrarenal cystic masses that separate the fragments of a laceration.



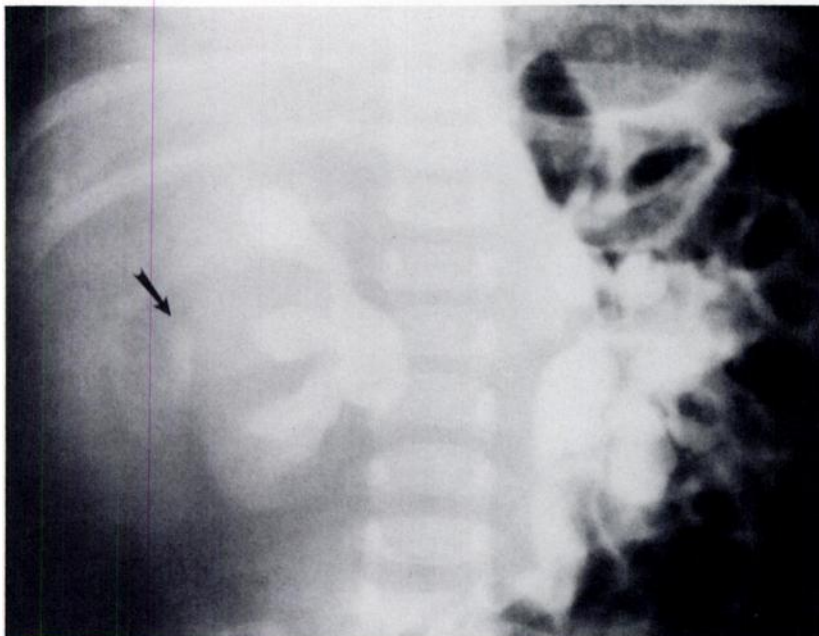
**Figure 21**  
**Types of urinoma** (A) Subcapsular urinoma (B) Localized perirenal urinoma (C) Diffuse perirenal urinoma (D) Intrarenal urinoma



**Figure 22**  
**Subcapsular urinoma** This three day old boy had ascites and continuous dribbling of urine. The typical "C" sign of subcapsular urinoma, U, was seen on excretory pyelography. Subsequently, posterior urethral valves were resected.



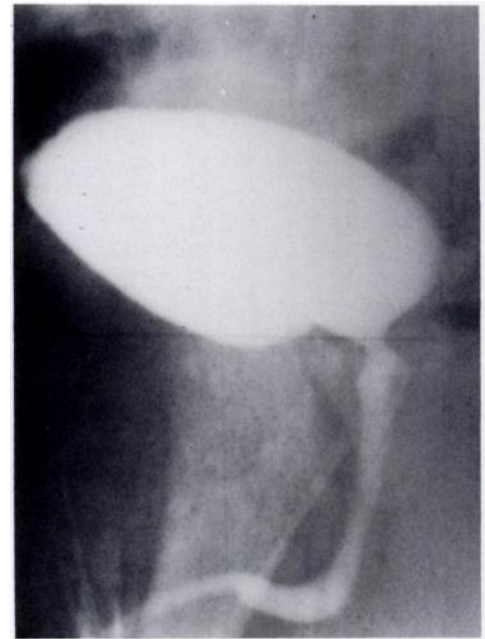
**Figure 23**  
**Localized perirenal urinoma** This three week old boy had ascites and a right abdominal mass. Voiding cystourethrography demonstrates right vesicoureteric reflux with extravasation into a large localized perirenal urinoma, U, which displaces the right kidney medially. The reflux and urinoma resolved rapidly after resection of posterior urethral valves.



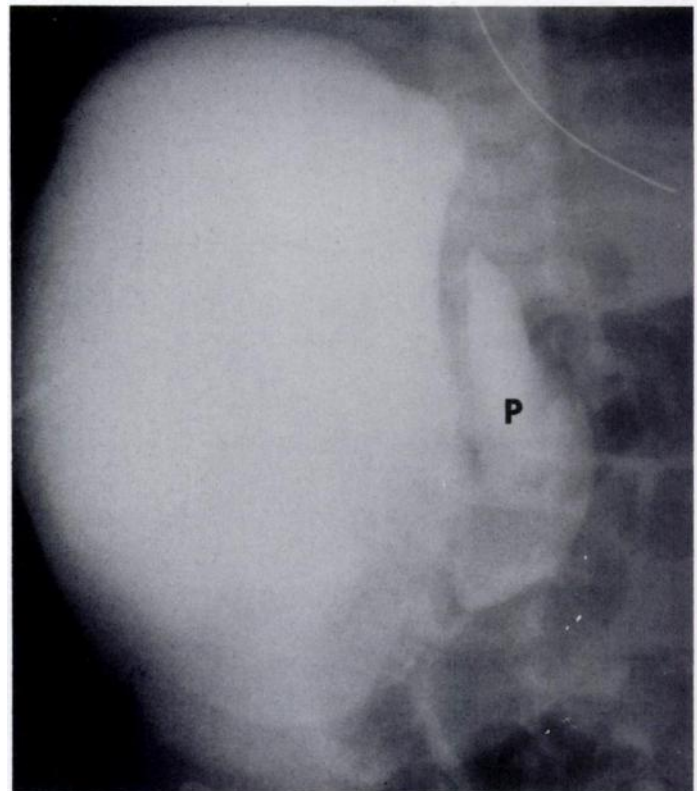
**Figure 24**  
**Diffuse perirenal urinoma** This three day old boy had ascites. Excretory pyelography showed extravasation of contrast medium (arrow) from the right collecting system into a large, diffuse perirenal urinoma surrounding the right kidney. The urinoma and posterior urethral valves were resected.



25A

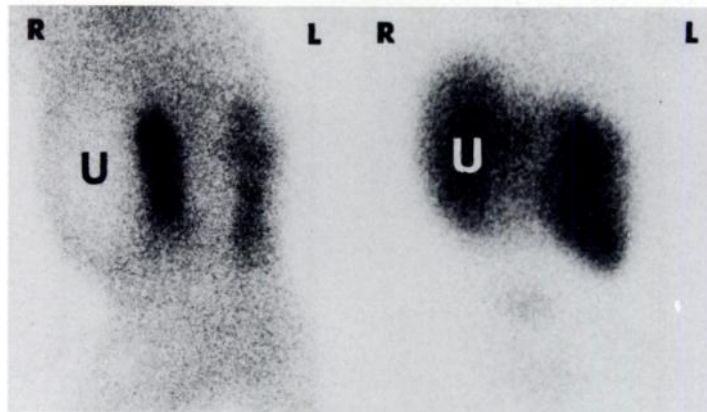


25B



25C

**Figure 25**  
**Urinoma associated with ureteric atresia** This 5 week old boy had a large right abdominal mass. He proved not to have posterior urethral valves, but his case is included as an excellent example of correlative imaging in the evaluation of a urinoma and the differential diagnosis of urinary tract obstruction. (A) Sonography shows an echolucent perirenal mass, U, displacing the hydronephrotic right kidney medially. (B) Voiding cystourethrography rules out posterior urethral valves. (C) Percutaneous puncture and opacification shows a cystic mass displacing and communicating with a blind-ending, dilated renal pelvis, P. (D) An early image from a <sup>99m</sup>Tc-DTPA renal scintigram reveals a photopenic area, U, lateral to the hydronephrotic right kidney. (E) On a delayed scintigram, this area shows increasing radioactivity confirming the presence of a communication between the pelvicalyceal system and the urinoma. There is pelviureteric obstruction on the left side, as well. At surgery, atresia of the proximal right ureter associated with rupture of a dilated calyx and a localized perirenal urinoma were found.



D

E

25D&E

**Urine Ascites**

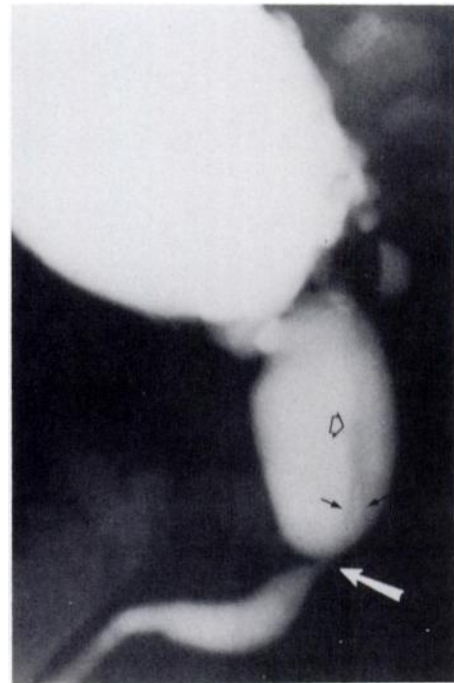
There are many potential causes of ascites in the newborn (Table II), but urine ascites, that is, the leakage of urine into the intraperitoneal space, is most common (26). It is usually the result of lower urinary tract obstruction from posterior urethral valves. The kidney is the most common site of rupture, but in many cases, no perforation can be

found. Often, urine ascites is associated with urinomas (Figure 27). The ascites begins in utero and is detectable on prenatal sonography (2). The associated abdominal distention can obstruct vaginal delivery or cause respiratory distress in the newborn.

TABLE II Causes of Neonatal Ascites	
Gastrointestinal	
Perforation of a hollow viscus; i.e., meconium peritonitis, etc.	
Inflammatory lesions; i.e., Meckel's diverticulitis, appendicitis, etc.	
Portohepatic	
Extrahepatic portal vein obstruction; i.e., compression by a mass, atresia of veins, etc.	
Intrahepatic portal vein obstruction: portal cirrhosis (neonatal hepatitis) biliary cirrhosis (biliary atresia)	
Genitourinary	
Urine ascites: lower urinary tract obstruction, i.e., valves, etc. upper urinary tract rupture, i.e., hydronephrosis	
Genital; i.e., ruptured ovarian cyst or hydrometrocolpos, etc.	
Miscellaneous	
Chylous ascites	
Immune hydrops fetalis	
Nonimmune hydrops fetalis (many causes)	
Congenital syphilis	
Idiopathic	



26A



26B

**Figure 26**

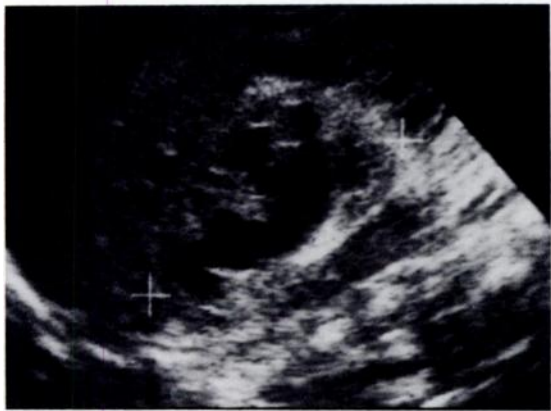
**Urine ascites associated with posterior urethral valves** This infant had abdominal distention at birth. (A) The abdominal radiograph shows the typical features of ascites; increased abdominal girth and bowel loops floating centrally. The diagnosis of ascites was confirmed by sonography. (B) Voiding cystourethrography shows typical posterior urethral valves (arrow). Note the visibility of the verumontanum (open arrow) and the valve cusps (small arrows) within the dilated posterior urethra.

**Figures 27A-E**

**Urine ascites with a perirenal urinoma** This newborn infant had a difficult vaginal delivery because of abdominal distention. (A) Abdominal sonography shows ascitic fluid, a, around the dome of the liver. (B) This right renal sonogram (longitudinal section) shows hydronephrosis. (C) A left renal sonogram (longitudinal section) shows a diffuse perirenal urinoma, u, surrounding a decompressed kidney. (D) An early image of a 99m Tc-DTPA renal scan shows bilateral renal function with leakage of the isotope into a diffuse left perirenal urinoma, U. (E) A 40 minute image from the renal scan shows the isotope moving from the urinoma, U, into the peritoneal cavity, A.



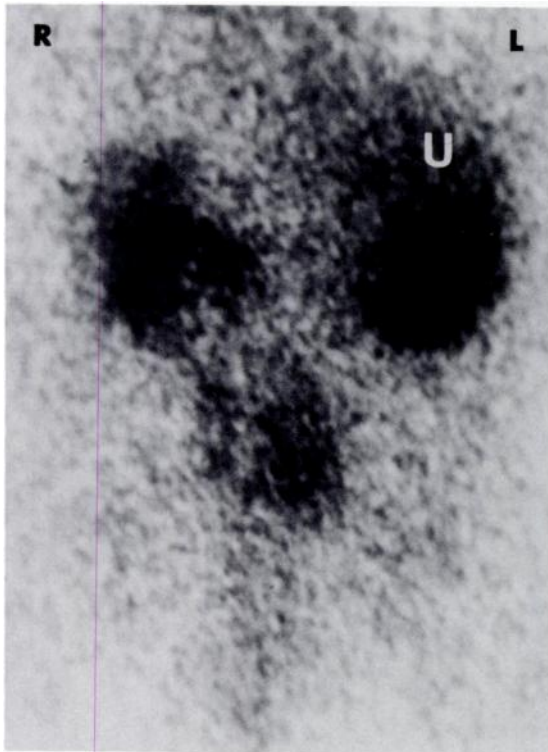
27A



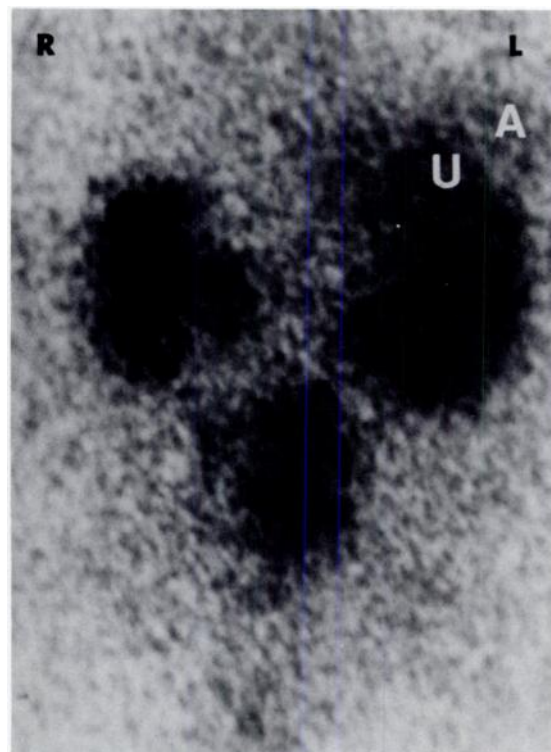
27B



27C



27D



27E



27F



27G

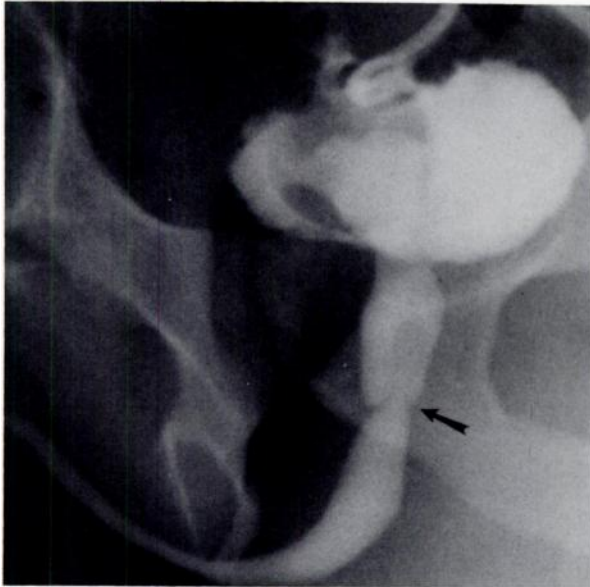
**Figures 27F&G**

(F) A voiding cystourethrogram shows a large bladder and posterior urethral valves. The contrast medium refluxing into the left ureter enters the left perirenal space, U, in this early image. (G) In a delayed radiograph, the contrast medium moved from the perirenal space, U, into the peritoneal cavity in the left flank (A). A diagnosis of posterior urethral valves with left vesicoureteric reflux, a diffuse left perirenal urinoma and urine ascites was made.

**Differential Diagnosis**

**TABLE III**  
Differential Diagnosis

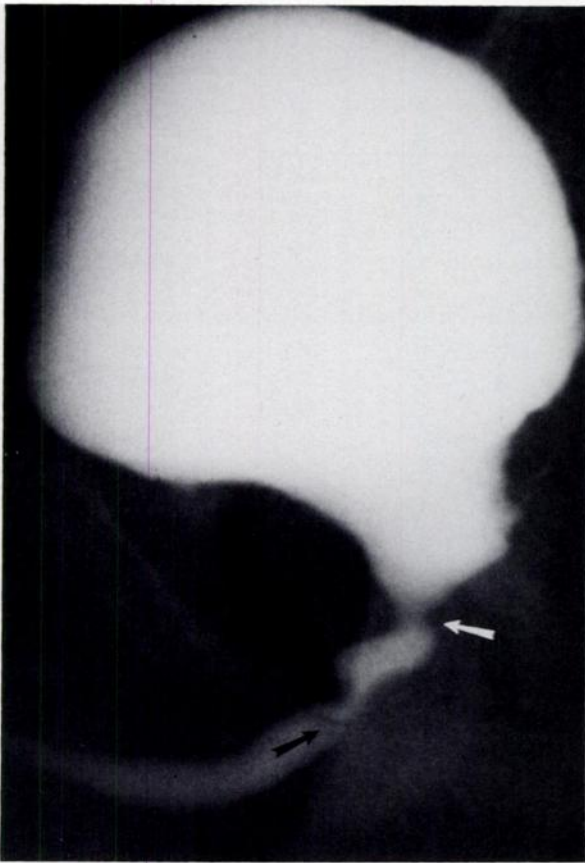
<p>A. Normal variations in cystourethrographic appearance of the posterior urethra</p> <ol style="list-style-type: none"> <li>1. Prominent plicae colliculi, i.e., nonobstructing valves (Figure 28A)</li> <li>2. Nonobstructive urethral ectasia (Figure 28C)</li> <li>3. Nonobstructive urethral "rings" or "kinks" (Figure 28B)</li> </ol>	<p>B. Other causes of urethral obstruction</p> <ol style="list-style-type: none"> <li>1. Anterior urethral valves (Figure 30)</li> <li>2. Urethral diverticula (Figure 31)</li> <li>3. Strictures; congenital or acquired (Figure 32)</li> <li>4. Hypertrophy of verumontanum (Figure 33)</li> <li>5. Urethral polyps and other neoplasms</li> <li>6. Meatal stenosis (Figure 29)</li> </ol>	<p>C. Other causes of bladder distention or hydronephrosis or both</p> <ol style="list-style-type: none"> <li>1. Megacystis-megaureter syndrome (Figure 20A)</li> <li>2. "Prune belly" syndrome</li> <li>3. Bilateral pelviureteric obstruction (Figure 20B)</li> <li>4. Bilateral vesicoureteric obstruction</li> <li>5. Multicystic dysplastic kidneys (Figure 20C)</li> <li>6. Neurogenic bladder</li> <li>7. Massive bilateral primary vesicoureteric reflux</li> </ol>
---	--	---



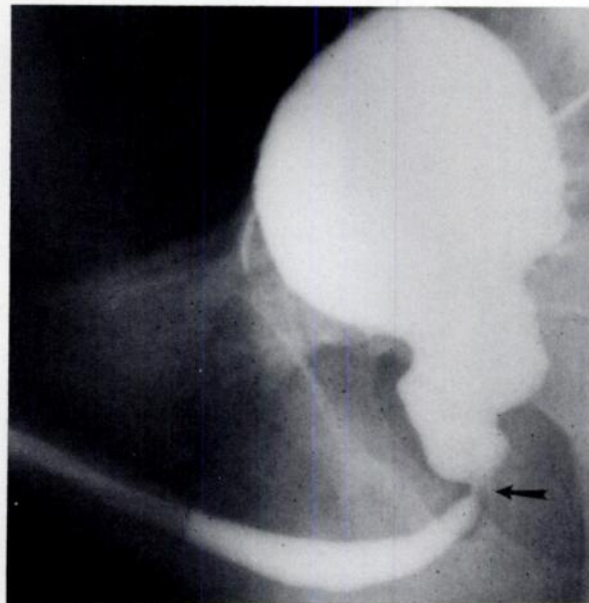
28A

**Figure 28**

**Normal variants** There are many variations in the appearance of the normal male urethra. None of those illustrated here showed an obstructive voiding pattern, and all were considered normal variants. (A) A large normal verumontanum and plicae colliculi (arrow) in an adult (B) Nonobstructing valves (plicae colliculi) (black arrow) and a "urethral ring" (white arrow) (C) Ectasia of the prostatic urethra with a relatively narrow membranous urethra (arrow)



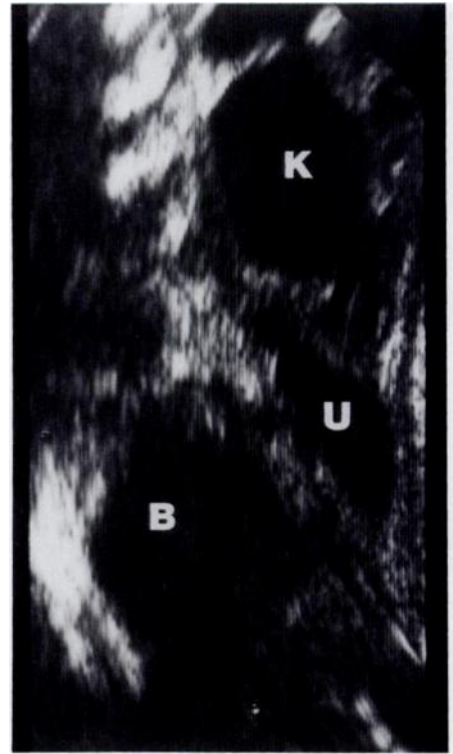
28B



28C



29A



29B

**Figure 29**  
**Meatal stenosis** Prenatal ultrasonic diagnoses of lower urinary tract obstruction and hydronephrosis were made on this infant. At birth, a catheter could not be passed into the bladder. (A) Cystography, via a supra-pubic catheter, shows a large bladder and bilateral vesicoureteric reflux. (B) The sonographic findings, with dilatation of the left renal pelvis, K, left ureter, U, and bladder, B, correlate with the cystographic findings. (C) Voiding cystourethrography reveals meatal stenosis (arrow) with a urethral diverticulum, d, just proximal to the hypospadiac meatus.



29C



30A



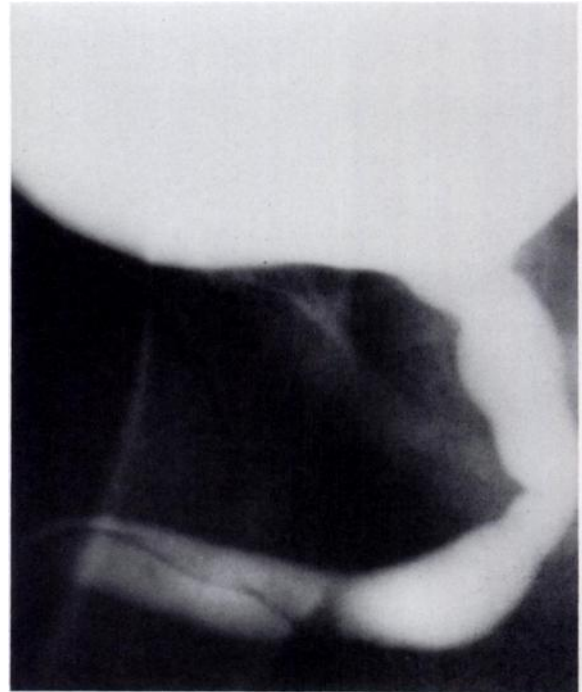
30B

**Figure 30**

**Anterior urethral valves (14)** This 9 year old boy, who had a history of micturition problems, was thought to have a neurogenic bladder. (A) The bladder is large and irregular, and there is right vesicoureteric reflux. (B) On voiding, however, anterior urethral valves (arrow) are demonstrated. Retrograde urethrography was normal. Anterior urethral valves are rare congenital obstructions usually located just proximal to the penoscrotal junction. They are believed to be abortive urethral duplications and are frequently associated with urethral diverticula.



31A



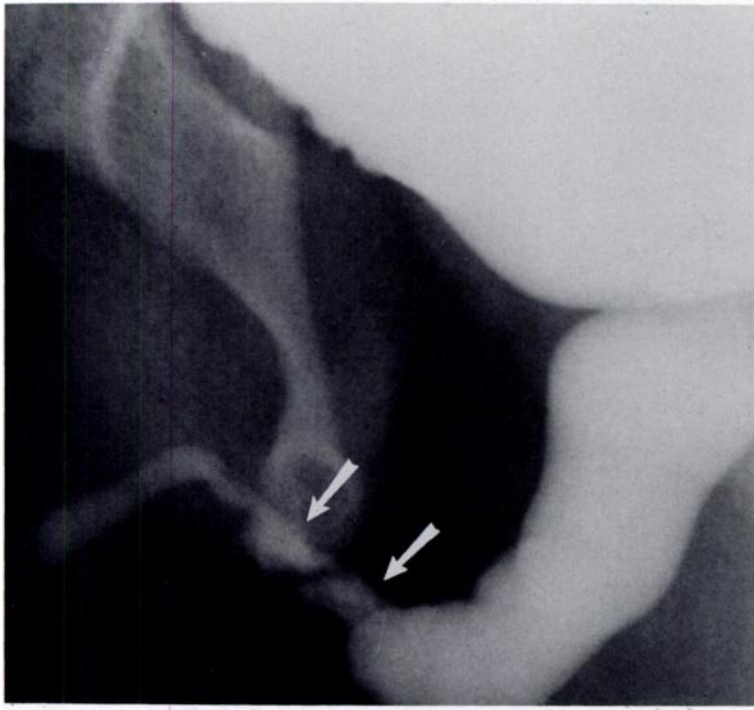
31B

**Figure 31**

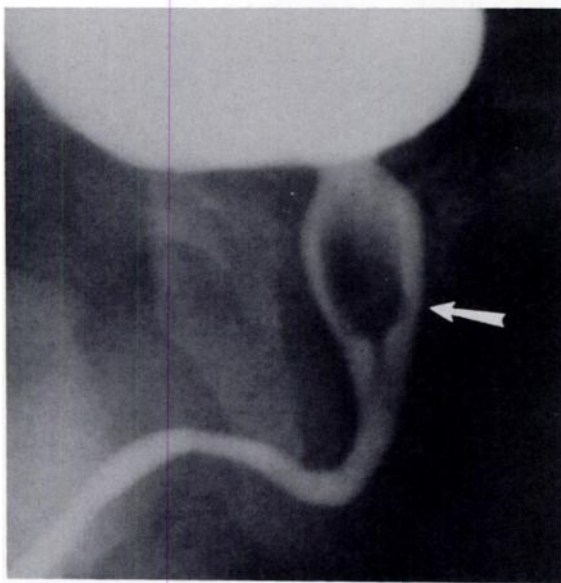
**Congenital urethral diverticulum (25)** A congenital sac-cular diverticulum of the bulbous urethra is shown here. (A) At the onset of voiding, the diverticulum begins to fill. (B) As voiding proceeds, the diverticulum enlarges. (C) The fully distended diverticulum obstructs the urethra. Diverticula of this type are frequently associated with anterior urethral valves.



31C

**Figure 32**

**Congenital urethral stricture (4)** This 10 year old boy had hematuria and dysuria. An irregular stricture (arrows) was seen in the bulbous urethra with dilatation of the posterior urethra. Urethral strictures in males can be congenital, inflammatory, traumatic or iatrogenic. The symptoms, site of occurrence, and appearance of the lesion in this case, correspond to those described in the rare cases of congenital stricture.

**Figure 33**

**Hypertrophy of verumontanum (15)** A very large verumontanum (arrow) was discovered during cystourethrography in this infant. There is no evidence of associated obstruction. Congenital hypertrophy of the verumontanum has been implicated as a cause of urethral obstruction. It can be difficult to differentiate from a congenital urethral polyp.

### Management

The treatment of posterior urethral valves can be a complex and controversial problem (7). Our series (Table IV) reflects some of the changes in the approach to treatment that have taken place over the past decade.

Primary valve ablation has proved most successful in older children with normal renal function (Figure 34). Earlier in the decade, a transperineal approach was used in five of our patients. In recent years, the transurethral approach has been employed exclusively. In addition, with the introduction of small resectoscopes (8), primary valve ablation is used in smaller infants than was possible earlier in the decade (7). Fourteen of the 21 children in our series who had primary valve ablations were presumably cured and required no further surgery. The other seven patients required one or more additional surgical procedures before improvement occurred.

Distal temporary diversion, that is, vesicostomy or cystostomy, has been used primarily in infants whose urethras were too small to accept the resectoscopes available at the time and in larger infants or children with significant renal failure.

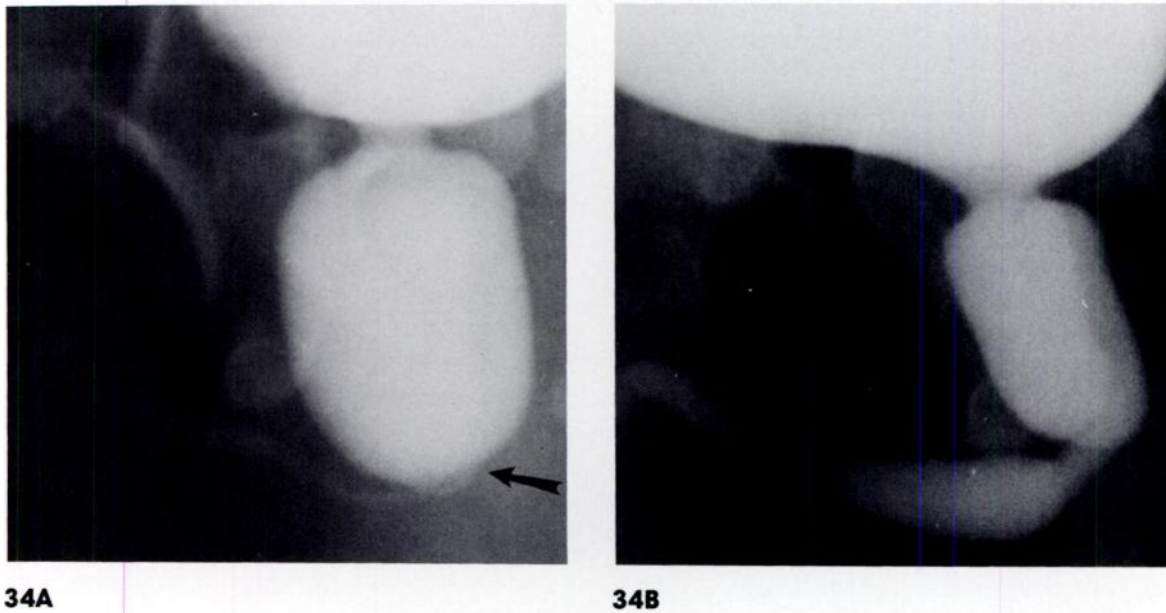
Later, when the child has grown sufficiently, the renal failure has resolved, or renal transplantation has been performed, the valves can be resected and the vesicostomy closed.

Proximal temporary diversions, that is ureterostomies or pyelostomies, are an alternative approach to diverting urine flow in small infants or larger children with renal failure. At an appropriate time, the valves can be resected and the proximal diversion taken down. Proximal diversion is also used as a treatment for markedly dilated and tortuous ureters that have not responded to valve ablation and vesicostomy. The majority of proximal diversions were performed early in the decade under study.

There are a number of complicated cases in which simple diversion and valve ablation are not enough to stop the progression of renal failure. These patients often have persistent vesicoureteric reflux or the "valve bladder syndrome" (17). They ultimately require combinations of surgical procedures and may eventually require renal transplantation.

TABLE IV

Treatment	In Our Series	Treatment	In Our Series
A. Primary valve ablation	21	D. Complicated cases	
with no further surgery	= 14	Multiple diversion procedures	6
B. Distal temporary diversion	10	Reimplantation of ureter	4
with secondary valve ablation only	= 4	Ileal conduit	3
with secondary valve ablation but		Dialysis	3
requiring further surgery	= 2	Nephrectomy	3
still awaiting surgery	= 4	Renal transplantation	4
C. Proximal temporary diversion	8		
with secondary valve ablation	= 7		
developed complications (see below)	= 7		



**Figure 34**  
**Valve ablation** This three year old boy had a history of incontinence. (A) The initial voiding cystourethrogram shows typical posterior urethral valves (arrows). A primary resection of the valves was performed. (B) Followup voiding cystourethrography, six months later, shows the urethra to be nearly normal in appearance. The child was then clinically asymptomatic.

**Complications**

The complications of posterior urethral valves and their incidence in our series are listed in Table V.

TABLE V Complications of Posterior Urethral Valves Following Treatment	
Complications	In Our Series
A. Urethral	
Stricture	4
Diverticulum	0
Incontinence	1
B. Bladder	
"Valve bladder" syndrome	8
Bladder diverticulum	4
Persistent vesicoureteric reflux	9
C. Renal	
Progressive renal failure	7
Renal dysplasia	3
Nephrogenic diabetes insipidus	0

Urethral strictures (Figure 35) following valve ablation are usually the result of instrumentation (3). They tend to occur when the resectoscope is too large for the infant's urethra. Alternatively, strictures may occur at the site of fulguration as a result of the heat injury associated with overzealous ablation. Urinary diversion may contribute to stricture formation by creating a "dry" urethra.

Incontinence following valve resection is a common complication. It is related to the marked distention of the prostatic urethra that occurs as a result of the valves (3). There are, however, iatrogenic factors that can contribute to it, such as, damage from instrumentation and from the now outdated practice of simultaneous bladder neck resection.

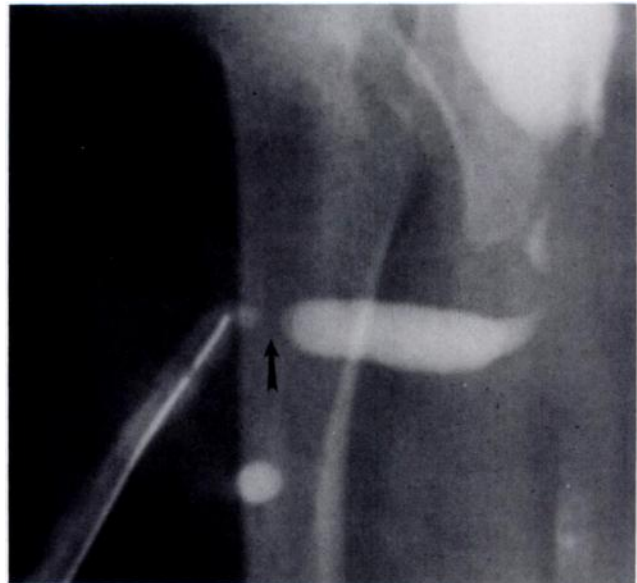
The "valve bladder syndrome" is suspected when upper tract dilatation unassociated with vesicoureteric reflux persists or progresses following correction of the urethral obstruction. In this instance, the thick, non-compliant bladder causes secondary obstruction at the vesicoureteric junction (17). The phenomenon of ureteric obstruction and dilatation being present when the bladder is full, and decreasing in severity when the bladder is empty, can be demonstrated by a renal radio-nuclide study (Figure 37) (8). This situation was seen in eight of our patients, four of whom went on to develop renal failure.

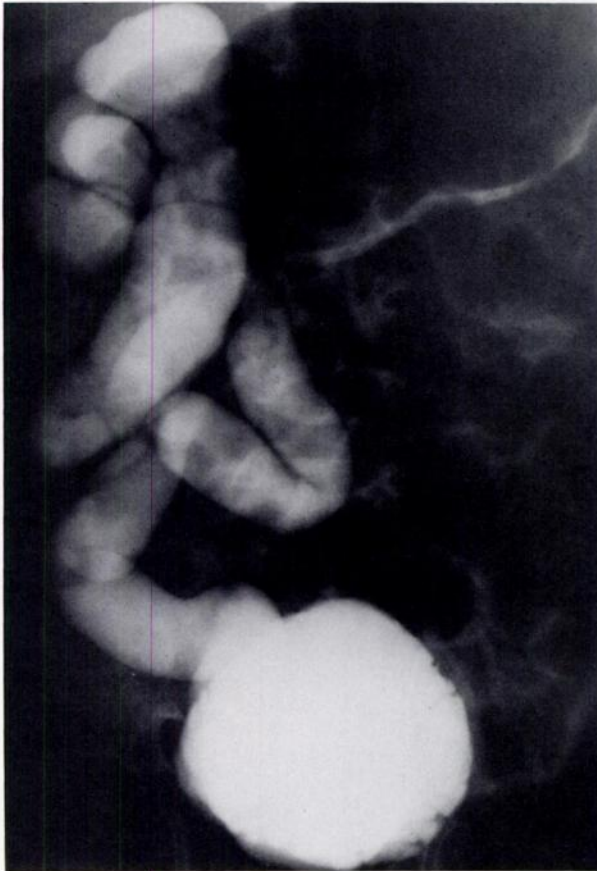
Bladder diverticula, most often the periureteric or Hutch variety (Figure 15), are commonly found at the time of their diagnosis in patients who have urethral valves (3). This location appears to be related to a detrusor weakness adjacent to the ureteric orifices. Usually, these diverticula are associated with ipsilateral vesicoureteric reflux, but they can obstruct the adjacent ureter.

The presence of vesicoureteric reflux at the time posterior urethra valves are diagnosed worsens the prognosis. Unilateral reflux is bad; bilateral reflux is much worse. Frequently, the reflux stops spontaneously following valve ablation. This is likely to occur if the associated kidney has reasonably good function. Ureters that continue to reflux following valve resection are usually associated with poorly functioning kidneys (7). In our series, there were 29 (36%) refluxing ureters at the time of diagnosis. Of these, there was persistent vesicoureteric reflux following valve ablation in ten (12%). All with persistent reflux were associated with poorly functioning, probably dysplastic kidneys at the time of diagnosis and all continued to deteriorate, eventually requiring nephrectomy (Figure 36), dialysis or renal transplantation. In our experience and that of others, antireflux surgery rarely alters the chain of events leading to renal failure (20).

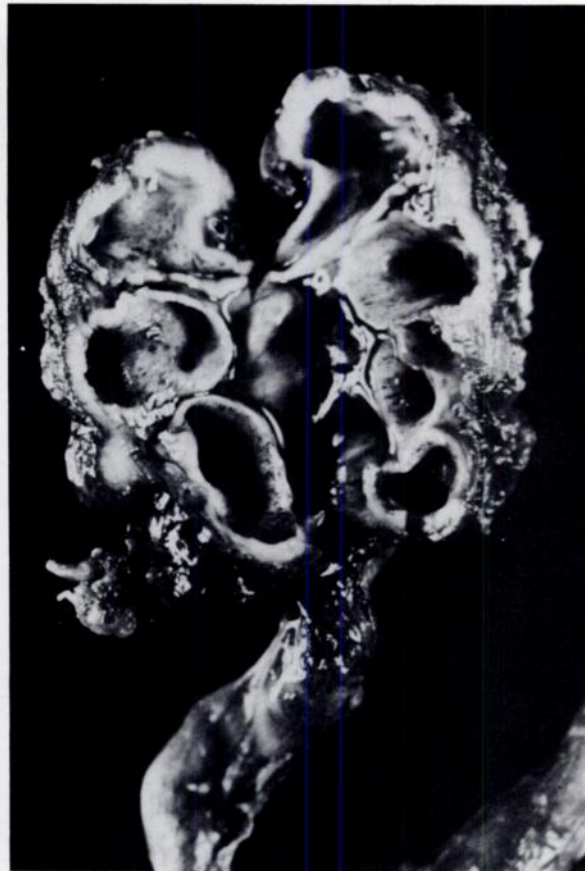
#### Figure 35

**Postoperative stricture** This 14 month old boy had posterior urethral valves diagnosed at another institution at birth. Vesicostomy and ureterostomies were established, and antegrade valve resection was attempted. A year later, a transurethral resection was performed. Now, two weeks later, he can no longer void via the urethra. This simultaneous antegrade and retrograde urethrogram shows the site and extent of the stricture in the anterior urethra (arrow). This was believed to be secondary to the instrumentation of valve resection.





36A



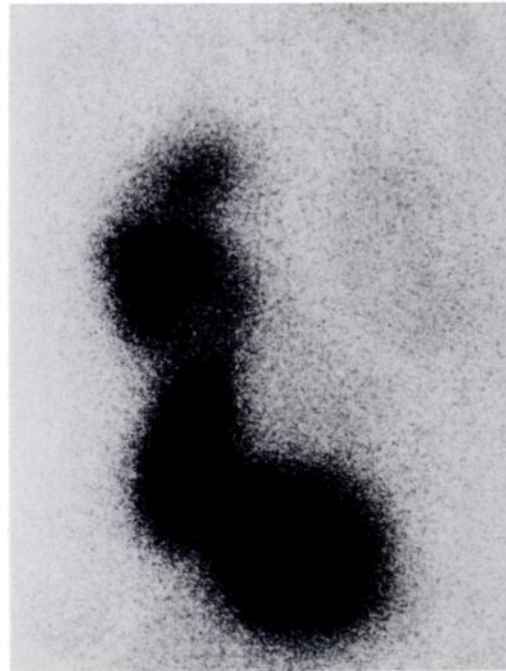
36B

**Figure 36**

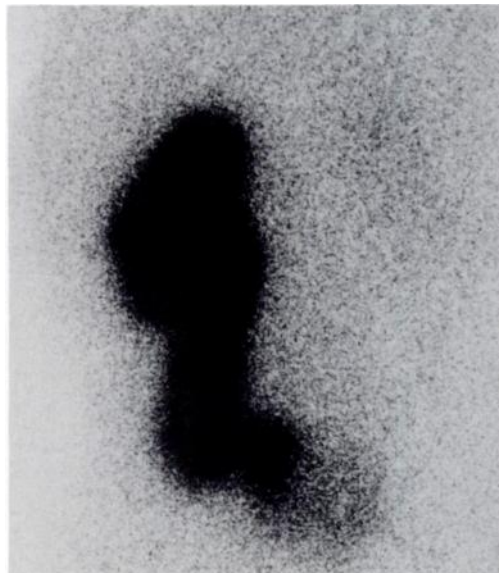
**Persistent vesicoureteric reflux** This boy had posterior urethral valves diagnosed at birth. He had a primary valve ablation in infancy, but massive right vesicoureteric reflux persisted. (A) A voiding cystourethrogram performed at three years of age demonstrates the magnitude of the vesicoureteric reflux. Renal scans indicated no renal function on the right side. A right nephrectomy was performed. (B) A clinical photograph of the resected specimen shows the non-functioning, pyelonephrotic, hydronephrotic right kidney.

**Figure 37**

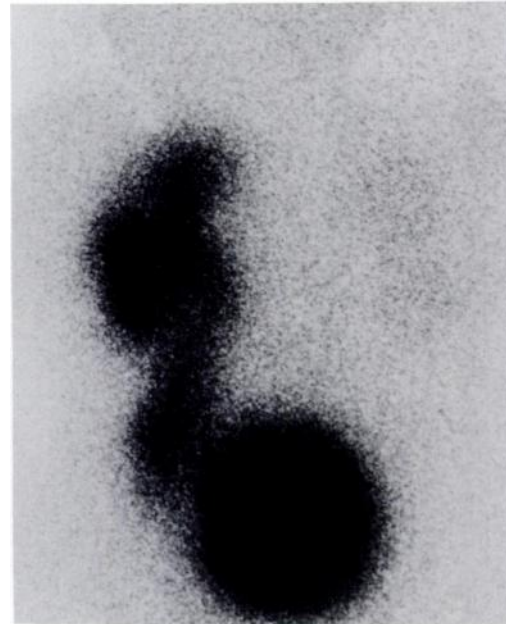
**Valve bladder syndrome** This four year old boy had a resection of posterior urethral valves at 13 months of age and a left nephrectomy at two years. A 99m Tc-DTPA renal scan was used to study the persistently dilated right urinary tract. (A) With the bladder full, the system is obstructed. (B) Immediately after voiding (without reflux), the system is maximally dilated. (C) Later, the dilated system decompresses into the empty bladder. This ureteric obstruction occurring with a full bladder and relieved by bladder emptying is consistent with the "valve bladder syndrome".



**37A**



**37B**



**37C**

### Conclusions

1. We have presented the current concepts of the morphology and embryology of posterior urethral valves.
2. It is a condition with a variety of clinical and radiological manifestations affecting all age groups, including the unborn.
3. Sonography provides us with a tool that can identify this condition prenatally and help us more fully to understand its effects postnatally.
4. Voiding cystourethrography remains the definitive test for demonstrating posterior urethral valves.
5. The management of patients who have posterior urethral valves is a complex and controversial problem.
6. Radiology plays a major role in identifying the complications of posterior urethral valves following treatment.

### References

1. Barry JM, Anderson JM, Hodges CW. The subcapsular C sign: A rare radiographic finding associated with neonatal urinary ascites. *J Urol* 1974; 112:836-839.
2. Chin DH, Filly RA. Ultrasound diagnosis of fetal genitourinary tract anomalies. *Urol Radiol* 1982; 4:115-123.
3. Churchill BM, Krueger RP, Fleisher MH, Hardy BE. Complications of posterior urethral valve surgery and their prevention. *Urol Clin North Am* 1983; 10:519-530.
4. Cobb BG, Wolf JA, Jr., Ansell JS. Congenital stricture of the proximal urethral bulb. *J Urol* 1968; 99:629-631.
5. Emmett JL. Obstruction of the vesical neck of a male infant produced by hypertrophy of the verumontanum: Report of a case. *Proc Staff Meet Mayo Clinic* 1940; 15:364-365.
6. Gilanz V, Miller JH, Reid BS. Ultrasonic characteristics of posterior urethral valves. *Radiology* 1982; 145:143-145.
7. Glassberg KE. Current issues regarding posterior urethral valves. *Urol Clin North Am* 1985; 12:175-185.
8. Glassberg KI, Schneider M, Haller JO, Moel D, Waterhouse K. Observations on persistently dilated ureter after posterior urethral valve ablation. *Urology* 1982; 20:20-28.
9. Glazer GM, Filly RA, Callen PW. The varied sonographic appearance of the urinary tract in the fetus and newborn with urethral obstruction. *Radiology* 1982; 144:563-568.
10. Greenfield SP, Hensle TW, Berdon WE, Wigger HJ. Unilateral vesicoureteric reflux and unilateral nonfunctioning kidney associated with posterior urethral valves - a syndrome? *J Urol* 1983; 130:733-738.
11. Hadlock FP, Deter RL, Carpenter R, Gonzalez ET, Park SK. Sonography of fetal urinary tract anomalies. *AJR* 1981; 137:261-267.
12. Hoover DL, Duckett JW. Posterior urethral valves, unilateral reflux and renal dysplasia: A syndrome. *J Urol* 1982; 128:994-997.
13. Kaplan GW. Posterior urethra in Kelalis PP, King LR, eds. *Clinical Pediatric Urology* 1st ed., W. B. Saunders Company, Philadelphia 1976; 301-327.
14. Kelalis PP. Anterior urethra in Kelalis PP, King LR, eds. *Clinical Pediatric Urology* 1st ed., W. B. Saunders Company, Philadelphia 1976; 328-341.
15. Koff SA, Thrall JH, Keyes JW. Diuretic radionuclide methods of investigating hydronephrosis. *Eur Urol* 1982; 8:82-87.
16. Macpherson RI, Gordon L, Bradford BF. Neonatal urinomas: imaging considerations. *Pediatr Radiol* 1984; 14:396-399.
17. Mitchell ME. Persistent ureteral dilatation following valve resection. *Dialogues Pediatr Urol* 1982; 5:8.
18. Pagan RA, Smith DW, Shepard TH. Urethral obstruction malformation complex: A cause of abdominal muscle deficiency and the "prune belly". *J Pediatr* 1979; 94:900-906.
19. Sanders RC, James AE. Ultrasonic assessment of genitourinary anomalies in utero in. *The principles and practice of ultrasonography in obstetrics and gynecology*, 3rd edition, Appleton-Century-Crofts, Norfolk, Conn. 1985; 195-209.
20. Scott JES. Management of congenital posterior urethral valves. *Brit J Urol* 1985; 57:71-77.
21. Stephens FD. Congenital intrinsic lesions of the posterior urethra in congenital malformations of the urinary tract. 1st ed. Praeger Publishers, New York, 1983:95-125.
22. Tanagho EA. Persistent ureteral dilatation following valve resection. *Dialogues Pediatr Urol* 1982; 5:4-5.
23. Vezina WC, Morin FR, Winsberg F. Megacystis-microcolon-intestinal hypoperistalsis syndrome: Antenatal ultrasound appearance. *AJR* 1979; 133:749-750.
24. Weiss RM. Persistent ureteral dilatation following valve resection. *Dialogues Pediatr Urol* 1982; 5:6-7.
25. Williams DI, Retik AB. Congenital valves and diverticula of the anterior urethra. *Br J Urol* 1969; 41:228-234.
26. Winestock D, Macpherson RI. Neonatal ascites: a case for diagnosis. *JCAR* 1971; 22:272-274.
27. Young HH, Frontz WA, Baldwin JC. Congenital obstruction of the posterior urethra. *J Urol* 1919; 3:289-365.

The authors would like to acknowledge the contributions of the following individuals:

Drs. Robert Lorenzo and Barbara Bradford, our predecessors at the University of South Carolina, who were involved in the initial care of many of these patients; Dr. Nancy Curry, our adult urologist, who provided Figures 17 and 28A; Dr. Alan Decter, pediatric urologist from the Children's Hospital of Winnipeg, Canada, who kindly let us use Figure 7; The Florence Radiologic Associates, for lending us Figures 13 and 27; the editors of "Pediatric Radiology" for permitting us to republish Figures 22, 23, 24 and 25; the Division of Audiovisual Production at the Medical University of South Carolina for their help in the preparation of the many reproductions, particularly Brooks Hart for the artwork and John Sanders for the photography, and Mrs. Suzanne Hall for typing the manuscript.