Posterior urethral valve

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Background: **Posterior urethral valve (PUV) is a significant cause of morbidity, mortality and ongoing renal damage in children. It accounts for end-stage renal disease in a proportion of children. This article aims at highlighting the current trend in the management of boys with posterior urethral valve.**

Data sources: **PubMed/Medline and bibliographic search for posterior urethral valve was done. Relevant literatures on presentation, pathology, evaluation, management and outcomes of PUV were reviewed.**

Results: **PUV which is increasingly diagnosed prenatally presents a spectrum of severity. The varied severity and degree of obstruction caused by this abnormality depend on the configuration of the obstructive membrane within the urethra. The decision to intervene prenatally is dependent on gestational age, amniotic volume, and renal function of fetal urine aspiration. Identification of the patients who may benefit from early intervention remains inconclusive. Endoscopic ablation of the valve is the gold standard of treatment but use of Mohan's valvotome and other modalities are invaluable in developing countries where endoscopic facilities are limited. Proximal urinary diversion may result in poor bladder compliance and should be reserved for patients with persisting or increasing upper urinary tract dilatation, increasing serum creatinine or inappropriate instruments. The behavior of the bladder and its subsequent management after valve ablation may influence the long-term renal outcome in PUV patients.**

Conclusions: **The care of children with PUV continues to improve as a result of earlier diagnosis by ultrasound, developments in surgical technique and meticulous attention to neonatal care. The ultimate goal of management should be to maximize renal function, maintain normal bladder function, minimize morbidity and prevent iatrogenic problems.**

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Review article

Introduction

Posterior urethral valve (PUV) is a common cause of lower urinary tract obstruction in male infants and the most common congenital cause of bilateral renal obstruction. PUV continues to be a significant cause of morbidity, mortality and ongoing renal damage in infants and children.[1] The incidence of PUV is estimated to be 1 in 5000 to 8000 male births, but it may be more common for some fetal demise.[2] The incidence of this condition in the African population is unknown. High bladder outlet obstruction throughout gestation leads to severely compromised renal function secondary to renal dysplasia in many children with PUV.[3] Treatment of PUV remains a clinical challenge, requiring active management from infancy to adulthood to avoid progressive renal dysfunction and deterioration of the upper and lower urinary tracts.

Historical background

Morgagni[4] was the first to describe PUV in 1717. However the most frequently referenced, earliest description of PUV is credited to Langenbeck in 1802, who commented on valve-like folds in autopsy specimens. Langenbeck did not infer any clinical significance to these findings.[2] Thirty years elapsed before the subject was again referred to by Velpeau (1832), who described several anatomical specimens in which there were posterior urethral valve-like folds that might be of clinical importance. It was not until 70 years before the first comprehensive discussion of valves was

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present in 1870 by Tolmatschew.[5] He was also the first to recognize this as a pathological entity and propose a theory about its embryology. Finally, Young et al[6] described PUV as a clinical and pathological condition in 12 patients while giving an accurate description of the clinical presentation and the anatomy of valves in 1919. This classic anatomical description of the valves was the subject of a recent review.[7]

Etiology and embryology

The exact etiology of PUV is unknown, but it appears to be a multifactor gene mediated embryopathy. Familial inheritance is not common, but has been reported.[8]

During embryogenesis, the most caudal end of the Wolffian duct is absorbed into the primitive cloaca at the site of the future verumontanum in the posterior urethra. In healthy males, the remnants of this process are the posterior urethral folds, called plicae colliculi. Histological studies suggest that PUV is formed at approximately 4 weeks' gestation, as the Wolffian duct fuses with the developing cloaca. When the insertion of the mesonephric ducts into the cloaca is anomalous or too anterior, normal migration of the ducts is impeded, and the ducts fuse anteriorly resulting in the formation of abnormal ridges or folds, which are believed to be the origins of 95% of PUVs;[9] this type is referred to as type I PUV. Although Young[6] has described type II PUV, most pediatric urologists believe that these are not obstructing valves but simply hypertrophy of the plicae colliculi. Type III valves represent the other 5% and consist of a ring-type membrane distal to the verumontanum with a perforation present centrally. The cause of these valves is an incomplete dissolution of the urogenital membrane.[10]

Classification

Young in 1919 described three different types of valves based on the orientation of the valves and their relationship to the verumontanum (Fig. 1):

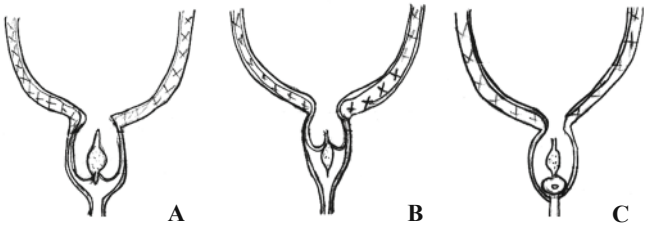

Fig. 1. Young's classification of posterior urethral valves based on the orientation of the valves and their relationship to the verumontanum.[6] **A:** Type I; **B:** Type II; **C:** Type III.

Type I: Two membranous structures in the posterior urethra originating from the caudal end of the verumontanum rising along the lateral margin of the urethra on each side meeting at 12 o'clock.

Type II: Membranes arising from verumontanum and attached cranially to the bladder neck.

Type III: Circular diaphragm in the region of the caudal end of the verumontanum with a central defect.

They further subdivided the classification of types I and III into a and b. Type III lesions were thought to consist of a membrane with a hole above (type IIIa) or below (type IIIb) the verumontanum, but without attachment to the verumontanum. Most patients were diagnosed by digital palpation of the lesion through the bladder neck, the passage of urethral sounds and/or autopsy, and not all lesions were visualized. This has been challenged recently by Dewan et al.[7] They studied the pristine uninstrumented posterior urethra of babies with PUV after suprapubic drainage and believed that the three types of valves probably represent a single diaphragm like structure with a central defect, which can assume different appearances due to either an antenatal rupture or postnatal instrumentation. This protocol was subsequently followed up in several babies and the concept of congenital obstructing posterior urethral membrane (COPUM) has been proposed.[11] This concept proposes that, instead of a true valve, a persistent oblique membrane is ruptured by initial catheter placement and, secondary to the rupture, forms a valve like configuration.[12]

Pathophysiology

PUV presents a spectrum of severity. The varied degree of obstruction caused by this abnormality depends on the configuration of the obstructive membrane within the urethra. The morbidity of PUV is not only limited to transient urethral obstruction but, the congenital obstruction of the urinary tract at a critical time in organogenesis may have a profound and lifelong effect on the function of the kidney, ureter, and bladder.[13]

Primary or secondary pathological manifestations of PUV

The macroscopic appearance of an obstructive membrane is the primary pathology causing a mechanical obstruction in the urethral conduit leading to sequential secondary changes. The severity of the changes depends on the degree and timing of the primary obstruction.[1]

Pathophysiology of the fetus

After 20 weeks, the kidneys provide over 90% of

amniotic fluid. An adequate amount of amniotic fluid is of vital importance for lung growth and skeletal development.[14] Severe oligohydramnios or anhydramnios secondary to decreased fetal urine output produces an abnormally small uterine cavity. This compresses the fetus and interferes with normal growth and expansion of the fetal thorax, resulting in pulmonary hypoplasia and soft tissue deformities.[1,15] The typical phenotype appearance of the fetus includes low-set ears, wide-set eyes, micrognathia, limb contractures, and talipes (Potter sequence).[16] An appropriate volume of amniotic fluid (produced by the kidneys) is necessary for complete and proper branching of the bronchial tree and alveoli.[13] Bladder distension and urinary ascites expand the fetal abdomen and compromise the development of the abdominal wall muscle, resulting in the prune belly appearance. Spontaneous upper urinary tract decompression can lead to urinary ascites, perirenal urinoma and peritoneal calcification.[1] High grade oligohydramnios, azotemia and severe pulmonary hypoplasia can lead to fetal death.

Urethra

The posterior urethra is dilated and elongated, the verumontanum is distorted, and the ejaculatory duct is dilated with reflux of urine into the vas deferens.[17]

Bladder

Obstruction in the posterior urethra causes high intravesical pressures, progressive muscle thickening (hypertrophy and hyperplasia), traberculation, sacculation and, in severe cases, diverticular formation. A large diverticulum affords some degree of protection to the upper renal tract because of the pop-off mechanism which is associated with good prognosis. There are increased collagen and connective tissue elements in the severely traberculated bladder. This bladder leads to the following changes:[1] high intraluminal pressure; increased residual urine; high pressure voiding; and bladder dysfunction even after the ablation of the valves. On urodynamic studies, three patterns of bladder dysfunction have been identified: myogenic failure with overflow incontinence, hyperreflexic bladder, and small capacity bladder with poor compliance.

Ureter

Ureteric dilatation may occur in PUV because of vesico-ureteric reflux (VUR). It is present in 50% of patients with PUV, vesico-ureteric junction obstruction, inefficient ureteric drainage due to high vesical pressure or dysplastic ureter. Significant proportion of vesicoureteric reflux will be resolved following the relief of obstruction.[18] VUR and renal dysplasia are attributable to abnormal location of the ureteric bud arising from the mesonephric duct. VUR may also be secondary to the high intravesical pressure.

Kidney

The pathology of the kidney includes hydronephrosis and progressive renal damage.

Hydronephrosis is due to 1) vesicoureteric reflux; 2) obstruction: high pressures within the bladder, which are transmitted to the ureter and kidney directly by hydrostatic pressure; 3) abnormal ureteric bud resulting in dysplastic kidney and a dilated collecting system.

Renal damage is due to the following two factors:

1. Renal dysplasia: Renal dysplasia is probably due to damage in early fetal life or abnormal embryogenesis. The renal dysplasia is microcystic and occurs primarily in peripheral cortical zone. The diagnostic histological findings include disorganized renal parenchyma, presence of embryonic tubules, cartilages, and cysts and presence of mesenchymal connective tissue. These changes are not reversible and account for long-term renal failure.

2. Obstructive uropathy: Renal injury due to obstruction results in both glomerular and tubular dysfunction with diminished glomerular filtration rates, decreased renal function, some degree of fibrosis and scarring of renal parenchyma, tubular damage which results in failure of concentration and acidification of urine. This leads to high urine output, increased sodium loss and electrolyte imbalance. These changes are potentially reversible.[11]

Presentation

Prenatal presentation

Most PUVs are diagnosed after detection of hydronephrosis by routine prenatal ultrasound.[19] Typical prenatal findings include bilateral hydroureteronephrosis, distended bladder and a dilated prostatic urethra (keyhole sign).[3,20] Discrete focal cysts in renal parenchyma are diagnostic of renal dysplasia.

Postnatal presentation

Neonates

Neonates with PUV who are not diagnosed before births may present with delayed voiding or poor urinary stream, abdominal mass, failure to thrive, poor feeding, lethargy, urosepsis or urinary ascites. In addition, respiratory distress at birth due to pulmonary hypoplasia may be the initial sign of urethral obstruction.

Infants

In infants, poor urinary stream and recurrent urinary tract infection are common.[21] Delayed post natal presentation is often with specific urological symptoms

like voiding disturbance, urine retention and urinary tract infection (UTI). Non-specific symptoms secondary to azotemia or sepsis are not uncommon. Rare presentations include urinary ascites or urinoma. This may occur as a result of spontaneous bladder perforation or fornicial urinary leak.

Older children

Older boys may present with recurrent UTI, diurnal enuresis in boys older than 5 years, secondary diurnal enuresis, voiding pain or dysfunction, and decreased force of urinary stream. PUVs are sometimes discovered during evaluation of abdominal mass or renal failure. Hydronephrosis and proteinuria found on evaluation of unrelated conditions may be the first sign of PUV.

Physical examination

Examination findings in the newborn include poor breathing movements, small chest cavity, palpable walnut-sized bladder (hypertrophy detrusor muscle), abdominal distension (ascites), potter facies and limb deformities (skin dimpling) and indentation of the knees and elbows due to compression within the uterus.

Evaluation

Antenatal

The antenatal diagnosis and assessment of PUV is based on maternal ultrasound and fetal urinary biochemical assesssment for renal function. The prenatal sonographic appearance of the fetus with urethral obstruction varies. The cardinal signs of the fetus include dilatation of the fetal urinary bladder and proximal urethra, and the key hole sign[20] with thickening of the bladder wall. Cohen et al[20] have also documented the presence of an echogenic line in the dilated posterior urethra confirmed postnatally to be a valve. Other findings include discrete renal cysts diagnostic of renal dysplasia. The diagnosis of PUV should be suspected in a male fetus with bilateral hydronephrosis, distended bladder with or without upper tract anomaly, and oligohydramnios around 18-20 weeks gestation.[22] Fetal urinary electrolytes and β2-microglobulin provide the most accurate results of evaluating fetal renal function. Normal fetal urine is hypotonic with sodium less than 100 mEq/L, chloride less than 90 mEq/L, osmolality less than 210 mEq/L, and β2-microglobulin less than 4 mg/L. Elevated fetal urine electrolytes and β2-microglobulin levels are indications of irreversible renal dysfunction.

Postnatal

Renal and bladder ultrasonography

Postnatally, ultrasound may show a thickened bladder wall and classically a dilated and elongated posterior urethra. Hydronephrosis varies in degree and may be unilateral or bilateral. Renal scan may demonstrate perirenal collection of urine, echogenic kidneys, subcortical cyst, and corticomedullary junction. Echogenic kidney and subcortical cyst are unfavorable signs.[9,19] Because newborns commonly have oliguria during the first few days of life, repeat ultrasound after the first week of life may be necessary if previous findings are normal in a child with previously diagnosed antenatal hydronephrosis before making a final determination that hydronephrosis has resolved. Ultrasound through the perineal approach may confirm the dilated posterior urethra with valve leaflet visualized. The diameter of the posterior urethra before and after voiding is larger than in normal children.[17,23]

Voiding cystourethrography

The key to the workup of any child with antenatal hydronephrosis is voiding cystourethrography (VCUG) performed during voiding and under fluoroscopy, with imaging of the posterior urethra. The diagnosis of PUV is based on: (1) thickened trabeculated bladder, dilatation and elongation of the posterior urethra (Fig. 2); (2) circumferential filling defect at the level of the pelvic floor; (3) prominence of the bladder neck and vesicoureteric reflux in many infants.

Renal scintigraphy

Functional imaging of the upper tract is usually deferred in neonates to about 4 weeks of age to allow some maturation of the developing kidneys. Radionuclide studies are useful in situations when either kidney shows thin or abnormal parenchyma and in infants in whom a postoperative ultrasound scan shows no improvement to help to distinguish between persistent obstruction and cystic dysplasia.[1,9] A dynamic study using tracer that is taken up by the kidney and excreted into the urine,

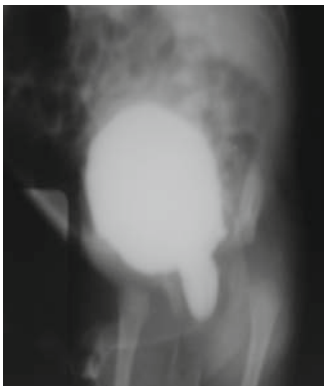

Fig. 2. Voiding cystourethrogram showing distended bladder, dilated and elongated posterior urethra.

either diethylenetriaminepentaacetic acid (DTPA) or mercaptoacetyl triglycine (MAG3) is used. The latter has high protein binding and thus remains mainly in the intravascular space compared with DTPA which is a pure glomeruli filtrates. The renal extraction of MAG3 is virtually double that of DTPA, making MAG3 the isotope of choice in the neonate or infant with a relatively large extracellular space and a low glomerular filtration rate. The parameters that can be estimated include differential renal function (DRF) and drainage. The drainage function is evaluated when dilatation is present. Up-sloping renogram pattern with diuretic (furosemide) is used to determine obstruction.[24]

A static scan using dimercaptosuccinic acid (DMSA) can estimate DRF and focal parenchymal defects. DMSA is extracted by proximal tubules and fixed in these cells. The static scan should not be performed in the neonatal period because of renal immaturity. DMSA is characterized in the newborn period by high background activity, excess excretion of isotope in urine, and relatively low fixation in the renal tubules. The scan may be done at the first 3-6 months for detection of renal scars.[24,25]

Laboratory investigation

Serum biochemistry

Serum biochemistry is performed to evaluate routine electrolytes, urea and creatinine which may be deranged. The serum values in the first 48 hours may be misleading as they may represent maternal values. Serum electrolyte is necessary to determine the level of renal derangement, to monitor responses to drainage, and to guide management. Serum creatinine 0.8 mg/dL is of prognostic importance.

Arterial blood gas (ABG)

ABG is determined to exclude metabolic acidosis which may be present. Urine sample should be sent for microscopic study, culture, and sensitivity test before commencing antibiotics especially in setting where most patients present with infected urine. Colony counts of more than 105 in samples collected through transurethral cauterization denote infection.

Urodynamic studies

Urodynamic evaluation provides information about bladder storage and emptying. Normal bladder should store urine at a low pressure, less than 30 cm H_2O and then completely empty at appropriate pressures. Patients with PUV have a spectrum of urodynamic abnormalities.[1] Urodynamic studies are helpful in directing the type of bladder therapy especially after valve ablation. Cystometry measures intravesical pressure and volume as the bladder fills at a measured rate. Pressure and volume data can be plotted against each other to produce a cystometrogram, which can detect dynamic abnormalities such as hyper-reflex contraction, hypocontractility, myogenic failure and detrusor-sphincter dyssynergy. Bladder compliance and post void residual volume can also be measured. A poorly compliant bladder is likely to cause persistent hydronephrosis.[9,26]

Management

Antenatal

The antenatal management of a patient with antenatally diagnosed PUV is based on serial examinations of a baby, with full parental consent. The decision made to intervene is dependent on gestational age, decreasing amniotic volume, and deteriorating renal function of fetal urine aspiration (Fig. 3). No intervention is considered in babies with the poorest prognosis.[27] Because of the morbidity related to antenatal fetal intervention, it is hard to justify intervention in a baby with good prognosis. If PUV is diagnosed late (third trimester) in the pregnancy without decreasing amniotic fluid, the best action is for nature to take its course as this period is towards the end of renal structural development. With improvement of prenatal ultrasonography, earlier intervention with vesicoamniotic shunting would improve postnatal renal function. However, identification of the patients who may benefit from early intervention remains elusive.[1] If a fetus is demonstrated to have PUV early in pregnancy (second trimester) and there are decreased volume of amniotic fluid and deteriorated renal function from urine aspiration, intervention would be appropriate.[27]

Three procedures are recommended such as vesicoamniotic shunt, vesicostomy, and fetal endoscopic valve ablation.

Selection criteria for intervention are as follows: Na^+ <100 mEq/L; Cl^- <90 mEq/L; osmolarity <210 mOsm/L; β2-Microglobulin <4 mg/L; total protein <40 mg/dL; Ca^{2+} <8 mg/dL.

Three bladder taps should be performed in the intervals of 48-72 hours. Documented sequential fall in values indicates salvagebility of the kidneys.

Vesico-amniotic shunt

The vesico-amniotic shunt has been a popular technique for in utero relief of obstructive uropathy. Inadequate decompression and patency of the shunt appeared for 1-2 weeks. Shunt migration is also common. The overall outcome is not satisfactory at present.[27]

Fetal vesicostomy

Decompression is adequate but it is more invasive with

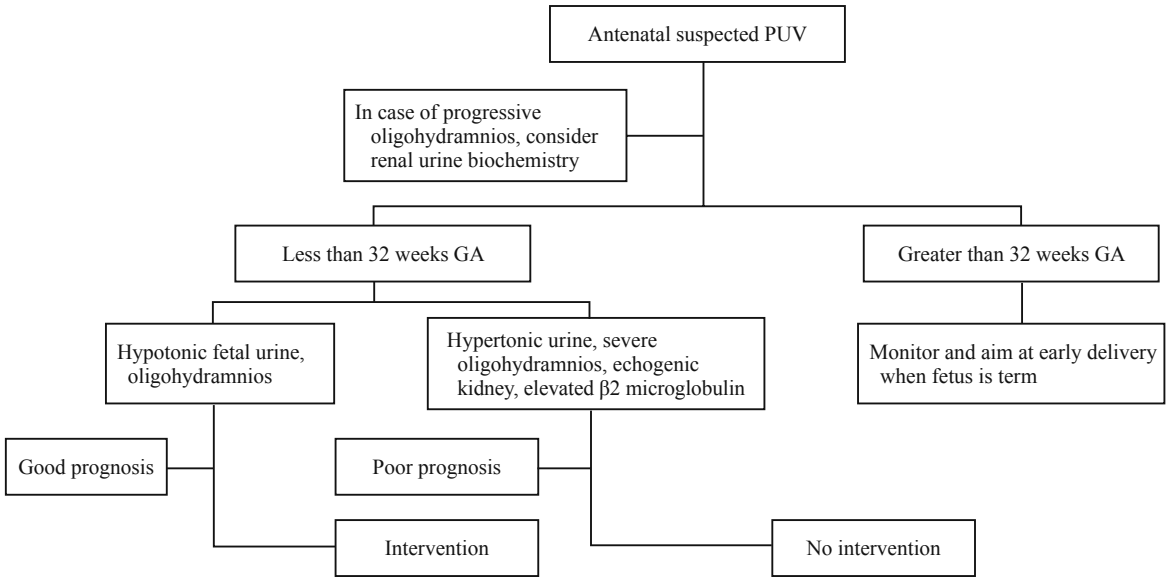

Fig. 3. Algorithm for the management of antenatally detected PUV. PUV: posterior urethral valve; GA: gestational age.

chances of fetal loss. It is usually done at 20-24 weeks gestation. Elective caesarian section is done at 10-14 weeks after open fetal surgery since most of these pregnancies will go into preterm labor around this time. Only a few cases were subjected to the procedure so far with higher maternal risk and premature labor.[27]

Fetal endoscopic valve ablation

Decompression with this procedure is less than with vesicostomy and is associated with preterm labor and fetal loss.

Postnatal treatment

Initial clinical examination, ultrasound and serum chemistry are followed by a voiding cystourethrography and a period of catheterization. The temporary vesical drainage is achieved by passing a feeding tube (size 5 or 6 Fr gauge) transurethrally under strict aseptic condition. Balloon catheters are not suitable for vesical drainage in PUV obstruction because they may accentuate spasm.[1] The relief of bladder obstruction by catheterization, either urethral or suprapubic, is often followed by a period of post obstructive diuresis and a fall in serum creatinine. Adequate medical stabilization is the priority at this stage. Urine is sent for routine microscopic examination and culture. Blood is also sent for calculation of complete blood count, culture and sensitivity test. In severe cases of PUV associated with pulmonary hypoplasia, catheter drainage is the only management until pulmonary function is improved. Such patients may need mechanical ventilatory support. Many of the neonates with PUV may have some of the following problems: dehydration and electrolyte imbalance, uremia, metabolic acidosis, infection, nutritional problems, and respiratory problems.

The patient is resuscitated with intravenous fluids, correction of electrolytes, and acid base deficits. Infection is controlled with broad spectrum antibiotics. Once the general condition is stable and diagnostic work up is complete, the definitive treatment is planned. From a practical point of view, management of neonates with posterior urethral valves begins with placement of a urethral catheter. The upper urinary tract should be assessed by renal sonography before placing the catheter and repeated after several days of bladder decompression. Improvement in renal function often corresponds with diminution in hydroureteronephrosis, whether the child has vesicoureteral reflux or not. When this occurs, primary valve ablation should be considered.[28] An algorithm for the management of newborns with PUV is shown in Fig. 4.

Ablation of valves

With miniaturized instrumentation at present, even the small urethra will accommodate a working cystoscopic sheath through which a 3 Fr coated ureteral wire can be inserted and used for valve ablation. Valve ablation as a primary procedure can be achieved by the following methods:

a) Endoscopic ablation: Transurethral ablation under direct vision is the treatment of choice for PUV. Pediatric cystoscope sized 6 or 8 is passed through the urethra. Bugbee electrode is used to fulgurate the valves. The obstructing membrane is incised at 5, 7 and 12 o'clock position. A 3 Fr ureteric catheter with a metal stylet that can be used to coagulate is a satisfactory alternative.

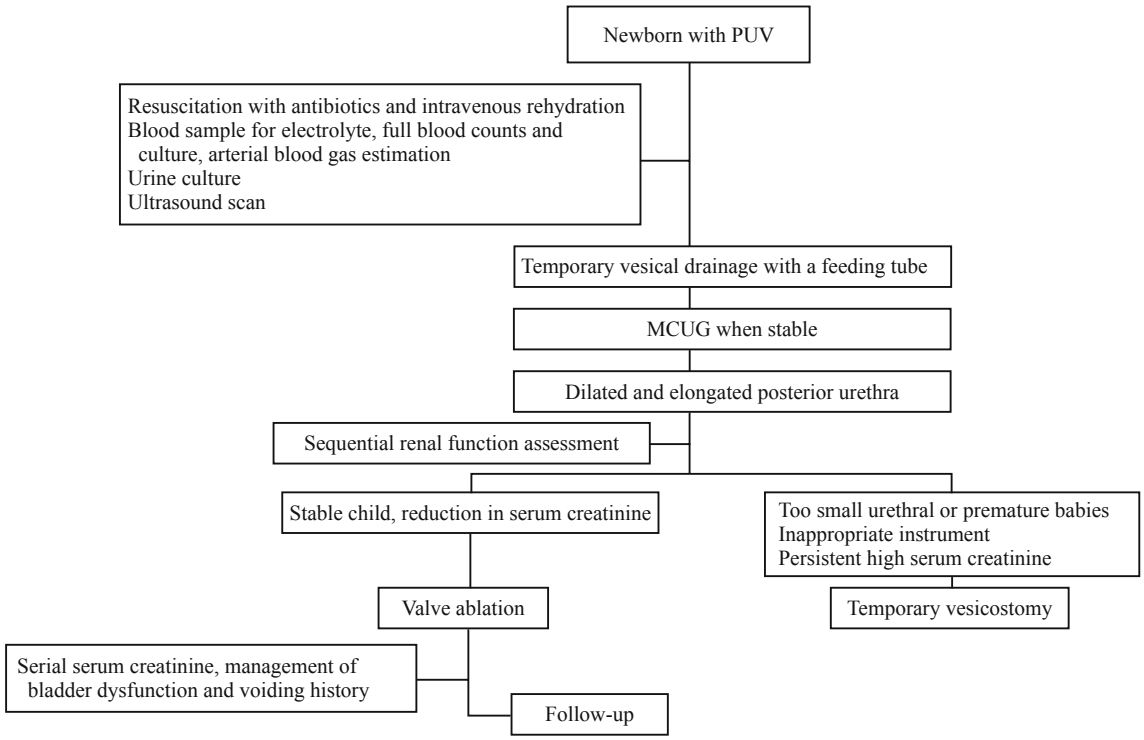

Fig. 4. An algorithm for the management of newborns with PUV. PUV: posterior urethral valve; MCUG: micturating cystourethrogram.

b) Whitaker-Sherwood hook: This is a size 6-7 Fr instrument, which has a crochet hook at the end. It is lubricated and passed up to the urethra, pointing to the 12 o'clock position, with the bladder full. The valve is engaged and diathermy applied to ablate it.

c) Laser ablation: neodymium-YAG laser has also been used successfully to fulgurate the valves.[29]

d) Mohan's valvotome: This is a 2 mm diameter instrument for newborns and a bigger 3 mm diameter instrument for older patients. With the patient in a supine position under general anesthesia, a well lubricated 6-Fr or 8-Fr feeding tube is inserted through the urethra and the urinary bladder filled up with normal saline. The feeding tube is then withdrawn and suprapubic pressure applied to demonstrate the initial preoperative urinary stream. A well-lubricated Mohan's valvotome is introduced through the urethra until urine/saline begins to come out of its end. The valvotome is gently withdrawn while applying sustained suprapubic pressure so as to display PUV. When the hook of the valvotome has engaged the valves, the valvotome is withdrawn from the urethra thereby ablating the valves. The procedure is carried out at 5, 7 and 12 o'clock positions. The bladder is again filled up with normal saline and suprapubic pressure applied to demonstrate the postoperative urinary stream indicating that the obstruction has been adequately relieved. This can be confirmed with cystoscopy when available. Mohan's valvotome is invaluable in the developing world where pediatric cystourethroscopes are not readily available.[21,30-33]

e) Fogarty or Foley's balloon catheter: The baby is anesthetized and a size 6 Fr catheter is introduced transurethrally. The bladder is filled with contrast material or saline until the posterior urethra is filled. A size 4 Fr Fogarty balloon catheter or an appropriate size Foley's catheter is placed into the bladder and balloon inflated with 0.75 ml of saline. With gentle withdrawal, the operator places the balloon at the level of the valves. Sharp withdrawal of the catheter ruptures the membrane without injury to the urethra.[34] This procedure should be preferably done under fluoroscopic or ultrasonographic guide to ensure that the balloon is only inflated proximal to the valve to avoid urethral injury.

Urinary diversion

An upper urinary tract diversion should be reserved for the infant in whom a small urethra limits the passage of available instrumentation. When there is improvement in upper urinary tract dilatation without significant improvement in serum creatinine, renal dysplasia is likely present and there remains little justification for upper urinary tract diversion over primary valve ablation. Proximal urinary diversion results in the absence of urine going to the bladder and thereby prevents bladder cycling, which may cause poor bladder compliance.[3] In case of persisting or increasing upper urinary tract dilatation, increased serum creatinine or

urinary tract infections, upper urinary tract diversion should be contemplated.

Option for urinary diversion

Vesicostomy: Vesicostomy is the diversion of choice for neonates with PUVs. The dome of the bladder should be brought to the skin midway between the umbilicus and pubis, such that the posterior bladder wall will not prolapsed into the stoma. The vesicostomy should be calibrated to 24 Fr. This allows bladder cycling and maintains bladder compliance. It is said to improve and stabilize VUR in 90% of patients.[35] However, formation of a small stoma results in stomal stenosis and inadequate emptying of the bladder. A large stoma induces bladder prolapse. The use of vesicostomy has decreased because most patients can be safely drained or undergo primary valve ablation.

Cutaneous ureterostomy: Dilatation of the ureter leads to failure of coaptation, this in turn leads to failure of ureteric peristalsis and stasis. Hence in the grossly dilated ureter, vesicostomy is not able to drain the upper tract; ureterostomy may be needed to establish a drainage. Ureterostomy for short duration has the advantage of not interfering with bladder function. Jaureguizar et al[36] did not find any significant change in bladder function between children who underwent initial pyeloureterostomy and those who received initial valve ablation. They concluded that temporary pyeloureterostomy did not affect bladder function adversely for a long duration.

Bilateral cutaneous ureterostomy can also be performed for urinary drainage. Techniques for cutaneous ureterostomy include end stomal ureterostomy, loop ureterostomy, Y-ureterostomy (in which the ureter is divided and one end is brought to the skin and the other is re-anastomosed by uretero-ureterostomy), and ring ureterostomy.[10] Potential complications of cutaneous ureterostomies include ureteral devascularization, inadequate drainage, and stomal stenosis.

Sober-en-T diversion: This is a temporary high diversion for posterior urethral valves, in which a cutaneous ureterostomy is performed, and the distal ureter is sutured to the upper ureter just distal to the renal pelvis.[37] The advantage of this approach is that the upper tract is diverted, but some urine drains to the bladder for long-term cycling.

Pyelostomy: In selected cases, cutaneous pyelostomy may be necessary if the child has urosepsis secondary to pyonephrosis.

Nephrostomy: Tube insertion for temporary diversion during percutaneous nephrostomy may be done to identify those patients who may benefit from cutaneous ureterostomy/pyelostomy.[38]

Secondary bladder management

Bladder management after ablation of the valves is a key to improve outcome in patients with PUV. The patients should be evaluated by voiding history, urinalysis, urine microscopy, culture, sensitivity test, serum creatinine and bicarbonate levels after ablation. VCUG should be repeated after ablation for 3 months and cystoscopy can be done if VCUG suggests incomplete ablation of the valves. When residual valves are ruled out, post voiding residual urine should be assessed with ultrasound after 1-week voiding after ablation of the valves, thus ensuring the bladder neck and urethral spasm has subsided. Post voiding residual urine volume of greater than 10% of expected bladder capacity for age is considered significant. Children with a significant post voiding residual urine volume are given Terazosin, a selective α1 adrenergic blocker dose ranging from 0.25 to 2 mg (0.02 to 0.4 mg/kgOD)[39] or an anticholenergic agent. Bladder training with a regular voiding program is imperative.

Indications for bladder augmentation include low bladder storage volume and high bladder pressure despite anticholinergic medication and clean intermittent catheterization. The ileum is most commonly used; however the use of the large bowel, stomach, and ureter is dependent on clinical conditions and surgeon preference.

Continent appendicovesicostomy, also called the Mitrofanoff's technique, is an option. The procedure involves placement of a nonrefluxing tubular conduit for catheterization between the bladder and skin to provide an alternative channel for catheterization. In children with PUVs, institution of intermittent catheterization through a sensate urethra can be difficult. In addition, some patients may have a dilated proximal urethra which may not be easily catheterized. The stoma often can be hidden in the umbilicus to provide acceptable cosmesis. The appendix, ureter, and tubularized bowel can be used for formation of this channel. However, since many of these bladders dilate with time, augmentation should be done only after sufficient time is given for the natural resolution of hypertrophy.

Complications

Newborns

Pulmonary hypoplasia secondary to intrauterine renal dysfunction and oligohydramnios is the primary cause of patient death. Other complications of PUV are generally secondary to chronic bladder changes, leading to elevated detrusor pressures. This, in turn, leads to progressive renal damage, infection, incontinence and renal insufficiency. Historically, in patients with

adequate pulmonary function, approximately 25% died of renal insufficiency in the first year of life, 25% died later in childhood, and 50% survived to adulthood with varying degrees of renal function. With the recent advent of better techniques in the treatment of pediatric renal insufficiency, most of these children can be expected to survive. The goal of treatment is to preserve the maximal obtainable renal function for each patient. This entails aggressive treatment of infections and bladder dysfunction.[13]

Vesicoureteral reflux

Vesicoureteral reflux is commonly associated with PUV and is present in one third of patients. When associated with PUV, reflux is generally secondary to elevated intravesical pressures. Persistent VUR following valve ablation is best treated conservatively with antibiotics. Persistent reflux from poor bladder emptying may be treated with anticholinergics or alpha blockers. Indications for reconstruction in persistent VUR include failed medication, recurrent UTI with renal damage, and severe reflux, which interfere with bladder emptying.[17]

Urethral stricture

Urethral stricture constitutes a significant complication of transurethral approach of valves ablation.[21,40] It occurs in up to 50% of cases and is due to urethral trauma of instrumentation.[1] The availability of miniature endoscope results in a decreased incidence of urethral stricture.[40] Urethral stricture is also due to thermal burn from the diathermy for fulguration, i.e., electro-coagulation of urethral tissue or heat generated when electric current passes through glycine or water during fulguration.

Urinary tract infections

Recurrent UTIs are common in patients with PUVs. Elevated intravesical pressures predispose patients to infection, possibly by altering urothelial blood flow. Additionally, patients with PUV may have elevated post voiding residual urine volumes, leading to stasis of urine. Dilated upper urinary tracts, with or without vesicoureteral reflux, further elevate UTI risk. UTI management is directed at lowering bladder pressures (anticholinergic medication), lowering post voiding residual urine volume (via alpha blocker[39] or/and clean intermittent catheterization), and timely administering prophylactic antibiotics.

Urinary incontinence

The same factors for vesicoureteral reflux and UTI also lead to urinary incontinence. Correct management of bladder function depends on adequate bladder evaluation with urodynamic studies. Lowering bladder pressure, improving bladder compliance, and minimizing post voiding residual urine volume contribute to the attainment of urinary continence.

Long-term prognosis

The prognosis of boys with PUV depends on the status of the kidneys and the bladder at the time of diagnosis and the methods of bladder management as the child grows.[9] A vast majority of the patients who developed end-stage renal diseases have voiding dysfunction.[41] Researchers have noted different variables that may have predictive value or be responsible for long-term renal failure in patients with PUV.[42,43] Prenatal detection of posterior urethra obstruction at or before 24 weeks of gestation could predict a poor outcome[22] (Table). The bladder dysfunction that some of these patients have will be decisive in the development of their renal failure.[44,45] Poor compliance is associated with the worst prognosis[46] but other factors implicated in the worsened prognosis in the boys include age at diagnosis, renal dysplasia, renal function before and after valve ablation, VUR, UTI, proteinuria, hypertension, and initial treatment.[42,43]

The level of serum creatinine at 1 year old is shown to have a greater predictive value than the level at the time of diagnosis.[42] Because the levels of serum creatinine after relief of obstruction could reflect more faithfully a kidney mass than those obtained before, and indeed, serum creatinine levels at 1 year old correlate strongly with the final outcome of patients. A serum creatinine level of less than 8 mg/L at 1 year old is associated with normal renal function.[47,48] Recent studies[42,43] have shown that there is a clear prognostic relationship between the serum creatinine levels at 4-5 days after bladder catheterization and renal function.

The glomerular filtration rate at 1 year old is also strongly correlated with final renal function. Patients

Table. Prognostic factors of posterior urethral valves

Good	Poor
	Prenatal diagnosis on or before 24 wk gestation
Presentation after 1 y	Respiratory distress at birth
Nadir serum creatinine less than 0.8 mg/dL at 1 y	Presentation before 1 y
Nadir serum creatinine less than 0.8 mg/dL after 4-5 d of initial drainage	Bilateral vesicoureteric reflux
Identification of corticomedullary junction	Persistent serum creatinine higher than 1.0 mg/dL after initial therapy
Presence of pop-off mechanism	Persistent incontinence after 5 y
	Echogenic kidney on ultrasonogram
	Subcapsular renal cyst
	Proteinuria
	Hypertension

with glomerular filtration rate of ≥80 mL/min per 1.73 m^2 at 1 year old have a normal renal function.[49]

Identification of at least one kidney with good corticomedullary differentiation on ultrasonographic diagnosis of a boy with PUV is associated with a good prognosis.[3,50,51] The presence of hyperechoic kidneys from the beginning as shown by poor corticomedullary differentiation on ultrasonogram is closely correlated with the development of chronic renal failure.[49]

The presence of pressure pop-off mechanism is another good prognostic factor. This includes a massive reflux into a non-functional kidney termed VURD syndrome (valves, unilateral reflux and dysplasia), urinary ascites, or large bladder diverticulum. This mechanism prevents deleterious effects of high vesical pressure on the opposite kidney. Bilateral high-grade reflux is significantly correlated with poor prognosis.[41,48]

The appearance of proteinuria during infancy is related to a poor prognosis.[52]

Pathogenically, this result could be related to decreased kidney functional reserves, which cannot respond to the increased demand for child's growth. Thus, functioning nephrons cause hyperfiltration to maintain a normal renal function. Excess filtration produces proteinuria, focal segmentary glomerulosclerosis, and finally kidney failure. In a significant proportion of children, renal impairment leads to anorexia, nutritional and growth failure, and by the second decade of life, loss of libido and end stage renal failure may occur.[53]

Other prognostic factors are listed in the Table.

Long-term management

Bladder dysfunction

Since the long-term outcome of patients with PUV depends on the behavior of the bladder and its subsequent management after valve ablation, close follow-up is imperative after valve ablation. A VCUG should be done to document that bladder outlet obstruction is relieved. Serial renal sonograms and examination of serum creatinine and electrolytes are necessary. Renal scintigraphy may be necessary if there is evidence of renal dysfunction. Serial VCUG is important if VUR is present until reflux is minimal or absent.[3] Urodynamic evaluation is required in the presence of persistent hydroureteronephrosis to confirm that resting and filling bladder pressures are in the safe range of less than 30 cm H_2O. This will also guide further management of dysfunction bladder.

Renal dysplasia

Renal dysplasia is irreversible, but treatment of other issues including management of urinary infection and bladder dysfunction can decrease or delay ongoing renal deterioration. It has been suggested that removal of the dysplastic kidney and dilated ureter may improve voiding efficiency and decrease the possibility of potential infection.[54] Recent evidence however suggests that retaining this dysplastic unit does not affect infection or function and the unit may be left in place.[55]

Renal transplantation

Despite optimal management, about one third of boys with PUV develop end-stage renal disease. Impaired renal function can be stabilized during childhood, but insufficient renal reserve developed in adolescence makes dialysis or renal transplantation necessary.[41]

Fertility

Few long-term studies revealed the reproductive status of men who had PUV during childhood. It has been postulated that prostate function might be affected because of elevated urethral pressure during embryonic development and the ongoing voiding dysfunction.[3]

Conclusion

Management of posterior urethral valves is still a clinical challenge in pediatric urology. There is still much to know about obstructive bladder physiology. The long-term outcome depends on the degree of renal damage, upper tract changes and bladder dysfunction. The ultimate goal of management is to maximize renal function, maintain normal bladder function, minimize morbidity and prevent iatrogenic problems.

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References

1 Imaji R, Moon D, Dewan PA. Congenital posterior urethral obstruction. In: Prem P, eds. Newborn Surgery, 2nd ed. London: Arnord, 2003: 856-866.
2 Krishnan A, de Souza A, Konijeti R, Baskin LS. The anatomy and embryology of posterior urethral valves. J Urol 2006;175:1214-1220.
3 Elder JS. Management of antenatally detected hydroneprosis. In: Prem P, eds. Newborn Surgery, 2nd ed. London: Arnord, 2003: 793-808.
4 Morgagni JB. Seats and Causes of Diseases investigated by Anatomy. In: Millar A, Cadell T, eds. Five books, containing a

Great Variety of Dissections with Remarks to which are added very accurate and copious indexes of the principal things and Names Therein contained. London: Johnson and payne, 1769: 540-556.
5 Tolmatschew N. A case of Semilunaren flaps of the urethra and vergrosserter vesicula prostatice. Archiv Path Anat 1870;11:348.
6 Young HH, Frontz WA, Baldwin JC. Congenital obstruction of the posterior urethra. J Urol, 3: 289-365, 1919. J Urol 2002;167:265-268.
7 Dewan PA, Zappala SM, Ransley PG, Duffy PG. Endoscopic reappraisal of the morphology of congenital obstruction of the posterior urethra. Br J Urol 1992;70:439-444.
8 Borzi PA, Beasley SW, Fowler R. Posterior urethral valves in non twin siblings. Br J Urol 1992;70:201.
9 Elder JS, Shapio E. Posterior urethral valves. In: Ashcraft KW, Holcomb GW, Murphy JP, eds. Pediatric Surgery, 4th ed. Philadelphia: Elsevier Saunders, 2005: 781-792.
10 Murphy JP, Gatti JM. Abnormalities of the Urethra, Penis, and Scrotum. In: Grosfeld JL, O'Neill JA, Fonkalsrud EW, Coran AG, eds. Pediatric Surgery, 6th ed. Philadelphia: Mosby Elsevier, 2006: 1899-1910.
11 Dewan PA. Congenital obstructing posterior urethral membrane (COPUM): further evidence for common morphological diagnosis. Pediatr Surg Int 1993;8:45-50.
12 Dewan PA, Goh DG. Variable expression of the congenital obstructive posterior urethral membrane. Urology 1995;45:507-509.
13 Bomalaski MD, Windle ML, Koo HP, Rauch D, Cendron M. posterior urethral valves. http://www.emedicine.com/urology/puv (accessed November 7, 2008).
14 Vanderheyden T, Kumar S, Fisk NM. Fetal renal impairment. Semin Neonatol 2003;8:279-289.
15 Cuckow PM, Nyirady P, Winyard PJ. Normal and abnormal development of the urogenital tract. Prenat Diagn 2001;21:908-916.
16 Christianson C, Huff D, McPherson E. Limb deformations in oligohydramnios sequence: effects of gestational age and duration of oligohydramnios. Am J Med Genet 1999;86:430-433.
17 Kulshrestha R. Posterior urethral valves. In: Common problems in pediatric surgery, 2nd ed. New Delhi: CBS, 2006: 322-328.
18 Close CE, Carr MC, Burns MW, Mitchell ME. Lower urinary tract changes after early valves ablation in neonates and infants: is early diversion warranted? J Urol 1997;175:984-988.
19 Hodges SJ, Patel B, McLorie G, Atala A. Posterior urethral valves. ScientificWorldJournal 2009;9:1119-1126.
20 Cohen HL, Zinn HL, Patel A, Zinn DL, Haller JO. Prenatal sonographic diagnosis of posterior urethral valves: identification of valves and thickening of the posterior wall. J Clin Ultrasound 1998;26:366-370.
21 Sudarsanan B, Nasir AA, Puzhankara R, Kedari PM, Unnithan GR, Damisetti KR. Posterior urethral valves: a single center experience over 7 years. Pediatr Surg Int 2009;25:283-287.
22 Hutton KA, Thomas DF, Arthur RJ, Irving HC, Smith SE. Prenatally detected posterior urethral valves: is gestational age at detection a predictor of outcome. J Urol 1994;152:698-701.
23 Chowdhury DN, Kabiraj SP, Kundu AK. Posterior urethral valves: transperineal ultrasonography: a diagnostic tool, pictorial essay. Indian J Urol 2002;18:140-142.
24 Gordon I, Riccabona M. Investigating the newborn kidney: update on imaging techniques. Semin Neonatol 2003;8:269-278.
25 Gordon I. Imaging of the renal tract in neonate. In: Puri P, eds. Newborn Surgery, 2nd ed. London: Arnord, 2003: 787-792.
26 Kim YH, Horowitz M, Combs AJ, Nitti VW, Borer J, Glassberg KI. Management of posterior urethral valves on the basis of urodynamic findings. J Urol 1997;158:1011-1016.
27 Chowdhary SK, Wilcox DT, Ransley PG. Posterior urethral valves: antenatal diagnosis and management. J Indian Assoc Paediatr Surg 2003;8:163-168.
28 Joseph DB. Editorial: posterior urethral valve and the 11th commandment. J Urol 2000;164:149-150.
29 Bhatnagar V, Agarwala S, Lal R, Mitra DK. Fulguration of posterior urethral valves using the Nd:YAG laser. Pediatr Surg Int 2000;16:69-71.
30 Abraham MK. Mohan's valvotome: a new instrument. J Urol 1990;144:1196-1198.
31 Shittu OB, Asinobi AO. Long-term outcome of posterior urethral valves ablation using the Mohan's urethral valvotome. W Afr J Med 2004;23:35-37.
32 Mukhopadhyay B, Sen S, D'Cruz AJ, Abraham MK, Ghosh SI, Mitra SK. Posterior urethral valves—a multi center review. J Indian Assoc Pediatr Surg 2003;8:140-143.
33 Ikuerowo SO, Omisanjo OA, Balogun BO, Akinola RA, Alagbe-Briggs OT, Esho JO. Mohan's valvotome for the ablation of posterior urethral valves. J Pediatr Urol 2009;5:279-282.
34 Kyi A, Maung M, Saing H. Ablation of posterior urethral valves in the newborn using Fogarty balloon catheter: a simple method for developing countries. J Pediatr Surg 2001;36:1713-1716.
35 Krahn CG, Johnson HW. Cutaneous vesicostomy in the young child: indications and results. Urology 1993;41:558-563.
36 Jaureguizar E, Lopez Pereira P, Martinez Urrutia MJ, Espinosa L, Lobato R. Does neonatal pyeloureterostomy worsen bladder function in children with posterior urethral valves? J Urol 2000;164(3 Pt 2):1031-1033.
37 Liard A, Seguier-Lipszyc E, Mitrofanoff P. Temporary high diversion for posterior urethral valves. J Urol 2000;164:145-148.
38 Ghali AM, El Malki T, Sheir KZ, Ashmallah A, Mohsen T. Posterior urethral valves with persistent high serum creatinine: the value of percutaneous nephrostomy. J Urol 2000;164:1340-1344.
39 Abraham MK, Nasir AR, Sudarsanan B, Puzhankara R, Kedari PM, Unnithan GR, et al. Role of alpha adrenergic blocker in the management of posterior urethral valves. Pediatr Surg Int 2009;25:1113-1115.
40 Lal R, Bhatnagar V, Mitra DK. Urethral strictures after fulguration of posterior urethral valves. J Pediatr Surg 1998;33:518-519.
41 Parkhouse HF, Barratt TM, Dillon MJ, Duffy PG, Fay J, Ransley PG, et al. Long-term outcome of boys with posterior urethral valves. Br J Urol 1988;62:59-62.
42 Denes ED, Barthold JS, Gonzalez R. Early prognostic value of serum creatinine levels in children with posterior urethral valves. J Urol 1997;157:1441-1443.
43 Onuora VC, Mirza K, Koko AH, Al Turki M, Meabed AH, Al Jawini N. Prognostic factors in Saudi children with posterior urethral valves. Pediatr Nephrol 2000;14:221-223.
44 Nguyen HT, Peters CA. Long-term complications of posterior urethral valves. Br J Urol 1999;83(Suppl 3):23-28.
45 Lopez Pereira P, Jaureguizar E, Martinez Urrutia MJ, Meseguer C, Navarro M. Does treatment of bladder dysfunction prior to

renal transplant improve outcome in patients with posterior urethral valves? Pediatr Transplant 2000;4:118-122.
46 Lopez Pereira P, Martinez Urrutia MJ, Espinosa L, Lobato R, Navarro M, Jaureguizar E. Bladder dysfunction as prognostic factor in patients with posterior urethral valves. BJU Int 2002;90:308-311.
47 Drozdz D, Drozdz M, Gretz N, Mohring K, Mehls O, Scharer K. Progression to end-stage renal disease in children with posterior urethral valves. Pediatr Nephrol 1998;12:630-636.
48 Lal R, Bhatnagar V, Mitra DK. Long-term prognosis of renal function in boys treated for posterior urethral valves. Eur J Pediatr Surg 1999;9:307-311.
49 Lopez Pereira P, Espinosa L, Martinez Urrutina MJ, Lobato R, Navarro M, Jaureguizar E. Posterior urethral valves: prognostic factors. BJU Int 2003;91:687-690.
50 Duel BP, Mogbo K, Barthold JS, Gonzalez R. Prognostic value of initial renal ultrasound in patients with posterior urethral valves. J Urol 1998;160:1198-1200.
51 Hulbert WC, Rosenberg HK, Cartwright PC, Duckett JW, Snyder H. The predictive value of ultrasonography in evaluation of infants with posterior urethral valves. J Urol 1992;148:122-124.
52 Dinneen MD, Duffy PG. Posterior urethral valves. Br J Urol 1996;78:275-281.
53 Duffy PG, Chowdhary SK. Posterior urethral valve: the current perspective. J Indian Assoc Pediatr Surg 2003;8:169-173.
54 Hoover DL, Duckett JW Jr. Posterior urethral valves, unilateral reflux and renal dysplasia: a syndrome. J Urol 1982;128:994-997.
55 Kim YH, Horowitz M, Combs AJ, Nitti VW, Glassberg KI. The management of unilateral poorly functioning kidneys in patients with posterior urethral valves. J Urol 1997;158:1001-1003.

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