Review Articles

Congenital Urethral Anomalies in Boys. Part II

Abdolmohammad Kajbafzadeh*

Department of Pediatric Urology, Children's Hospital Medical Center, Tehran University of Medical Sciences, Tehran, Iran

ABSTRACT

Introduction: In the second part of this article, congenital urethral anomalies other than posterior urethral valve were reviewed.

Materials and Methods: The anomalies considered in the current review were anterior urethral valve, lacuna magna, syringocele, Cobb's collar, duplication of urethra, megalourethra, and prostatic urethral polyps. The literature was extensively reviewed concerning the presentations, diagnosis, different types of treatment modalities, morbidity, mortality, and new concepts for the above disorders.

Result: Anterior urethral valves or diverticula are the most prevalent congenital anomalies of anterior urethra. The lacuna magna is the largest depression in the dorsal aspect of the fossa navicularis. It is demonstrable on a well-performed voiding cystourethrography of the distal urethra. The dilated Cowper's gland duct is the other missed diagnosed anomaly of the urethra in boys. The congenital narrowing of the bulbar urethra with a variable clinical presentation and obstruction grade and different types of anterior urethral obstruction are the most common presentation of these anomalies. However, other symptoms or signs including, hematuria, bloody spotting on underwear, discomfort or sever pain in the vicinity of the glance, interrupted voiding, infection, bulging of anterior urethra, enuresis, and postvoiding dribbling are the only nonspecific manifestations of these disorders.

Conclusion: All of these disorders are demonstrable on a well-performed voiding cystourethrography of the distal urethra. The urologist must be aware about these uncommon congenital anomalies and the anterior urethra should be carefully evaluated for such anomalies. Diagnosis of these entities is elusive unless the physician is looking for them. Nonspecific symptoms mentioned here besides radiographic findings can be a valuable clue for diagnosis.

KEY WORDS: urethral anomalies, valves, obstruction, duplication, syringocele, megalourethra, polyp, Lacuna Magna, diverticula, boy

Anterior Urethral Obstruction

Anterior urethral valve (AUV) is a rare cause of congenital urethral obstruction in boys. AUV is associated mainly with a proximal diverticulum (Figure 1). It may be found anywhere distal to the membranous urethra, usually proximal to the penoscrotal junction. Depending on the age of the patient and the severity of obstruction, clinical presentation highly varies from a stream that dribbles and voids poorly to hydroureretonephrosis and end-stage renal disease.

The most common cause of congenital urethral

*Corresponding author: No 36, 2nd Floor, 7th St, Aadat-abad Ave, Tehran 19987, Iran.
Tel: +98 21 2208 9946, Fax: +98 21 2206 9451
E-mail: kajbafzd@sina.tums.ac.ir
obstruction is posterior urethral valve (PUV), and AUV is reported to be 7 times less common; however, it can be equally as devastating.\(^1\) The embryological pathophysiology of AUV remains uncertain. Various proposed etiologies exist, including an abortive attempt at urethral duplication, failure of alignment between the proximal and distal urethra, incomplete formation of the ventral corpus spongiosum, congenital cystic dilation of the periurethral gland, and a ruptured Cowper's duct cyst.\(^3\) AUV may locate in every portion of the anterior urethra with almost equal incidence.\(^4\) It may even be found in fossa navicularis.\(^5\) It is likely that a pathophysiologic spectrum exists from valve to diverticulum formation associated with the degree of urethral dilation.\(^1\) Intrauterine urethral obstruction can result in severe bladder dysfunction after birth, which does not necessarily resolve after valve resection. If severe neonatal obstruction exists, urinary diversion by vesicostomy, and antibiotics, electrolyte management, and assessment of renal function improvement are recommended.\(^6,7\) The abnormal bladder function can occasionally be so severe as to end with augmentation cystoplasty.

An anterior urethral diverticulum should be considered in every case of anterior urethral obstruction. Urethral obstruction usually can be detected by antenatal scanning. When anterior urethral obstruction is suspected, an antegrade cystourethrogram is the investigation method of choice as any attempt at urethral catheterization may disrupt the pathology, and the exact diagnosis can be missed. Urodynamic studies through a suprapubic catheter before and after valve ablation can be a helpful baseline investigation.\(^6\) If secondary pathological changes such as bladder dysfunction and upper tract dilation do not improve after valve resection, other treatment options should be considered. The rapid improvement in bladder function and upper tract dilatation in these cases must be good prognostic factors. We recommend that urodynamic studies be performed in all patients with urethral obstruction before and after treatment to help predict the future prognosis.

The association of AUV with PUV is extremely rare in the literature. In 1982, Graham and colleagues\(^2\) have reported an association of an anterior urethral diverticulum with PUV in a newborn. Patients may present with poor urinary stream, recurrent urinary tract infections, or kidney failure. Depending on the severity of anatomic obstruction, patients may present early at birth or later in childhood if obstruction is minimal.\(^1\)

An AUV in almost all cases is actually a congenital urethral diverticulum.\(^9\) During voiding, the diverticulum expands, ballooning ventrally and distally beneath the thinned corpus spongiosum. The flaplike dorsal margin of the diverticulum then extends into the urethral lumen, occluding urinary flow (an obstructing valve). Anterior urethral valves have been described in every portion of the anterior urethra with almost equal incidence. They may be small, minimally obstructive, and of limited clinical concern. Often, though, they are severely obstructing and result in all the findings seen with PUVs.

Diagnosis of AUVs is confirmed on voiding cystourethrography. At times, difficulty with catheterization may be encountered because the catheter preferentially slips into the diverticulum.

**Fig. 1.** Typical radiologic appearance of anterior urethral valves
However, this occurs less often than one might expect, because the proximal wall of the diverticulum is often not hollowed out nearly as extensively as the distal wall. Because the diverticula almost always are placed ventrally in the midline, a dorsally oriented coudé-tipped catheter usually can be negotiated with less difficulty into the more proximal urethra. To establish the diagnosis, the entire penile urethra must be included in the voiding phase of the cystourethrography, or more distally located lesions will be missed.

The etiology of these anomalies is not entirely clear, but they seem to represent incomplete fusion of a segment of the urethral plate. Another possible cause might be a focally incomplete development of the corpus spongiosum with ballooning of the urethral mucosa due to inadequate support. Small, nonobstructing diverticula often appear stable for many years and do not show continuous enlargement and progressively worsening obstruction.

Initial management of a congenital AUV is the same as that for the more commonly seen PUV. Initial imaging studies assess the extent of hydronephrosis, the thickness and quality of the renal parenchyma (echogenicity on renal ultrasonography and uptake on renal scan), and the presence or absence of vesicoureteral reflux. Infants presenting with urosepsis and/or severe renal insufficiency require a period of transurethral or suprapubic (by percutaneous route) catheter drainage for stabilization, antibiotics, electrolyte management, and assessment of renal functional improvement. As with infants with PUVs, a temporizing tubeless diversion (vesicostomy, loop ureterostomy) may be chosen on an individual basis.6

Management of the urethral anomaly may be endoscopic or open. A hooked, single-wire, electrocautery knife can be used to engage the distal margin of the diverticulum and incise it in the midline. When performing this procedure, the surgeon must be very careful not to place the tip of the wire too close to the floor of the diverticulum. At this location, the wall of the urethra can be very thin, and thermal injury may result in development of a urethrocutaneous fistula. Even after satisfactory destruction of the leaflet, postoperative urethrogramy is often disappointing, because the appearance of the diverticulum may be unchanged. One must carefully assess the quality of flow (flow rate if the child is old enough) and the extent of filling of the urethra distal to the anomaly, to evaluate the results of the procedure.

Some surgeons have advocated open resection and reconstruction of the diverticulum.9 This technique allows one to completely excise the distal lip and provide a more homogeneous caliber to the urethra. In most cases, a patch graft urethroplasty is the preferred procedure. If the diverticulum is on the penile shaft, a sleeve dissection of the penile shaft skin from the corona to the penoscrotal angle allows the urethroplasty to be completed without overlapping suture lines.

Some AUVs may not be associated with a urethral diverticulum. De Castro and colleagues10 have described 3 children with anterior urethral membranes without the associated diverticulum.

**Lacuna Magna or Sinus of Guerin**

Formation of the distal glanular urethra may occur by a combination of 2 separate processes: proximal fusion of the urethral folds and distal ingrowth of ectodermal cells. It is generally thought that the stratified squamous lining of the fossa navicularis results from an ingrowth of surface ectoderm as far proximally as the valve of Guerin. The lacuna magna (also known as the sinus of Guerin), which can produce symptoms of hematuria and dysuria in some boys, may form as a result of dorsal extension of this ectodermal ingrowth. It has been suggested that the entire penile urethra might differentiate from fusion of the urethral plate via the mechanism of epithelial-mesenchymal interactions.11,12 The lacuna magna is the largest depression in the fossa navicularis. Guerin13 has described the valve of Guerin as a septum between the lacuna magna and the urethral lumen, which may act as a valve (Figure 2). The anomaly has been described in the urologic literature since 1980, at which time Somner and Stephens14 first drew attention to the fact that it may present with specific symptoms and/or dysuria, hematuria, blood in urethra, and spotting of underclothes.14-18 Although it may be present in 30% of boys as a small pit or sinus,17 only a few become symptomatic owing to the valvelike effect of the septum. If the inferior wall of the sinus balloons during micturition, it may elongate and trap urine in the lacuna magna. Chronic distention and irritation of the diverticulum probably are
responsible for the dysuria and hematuria.\(^ {14-16}\)

Micturating cystourethrography must be performed meticulously if this lesion is not to be missed, and the entire distal urethra must be included on the radiographic films (Figure 2). Contrast medium on towels or clothing may obscure or simulate the valve of Guerin. Similarly, retrograde studies, particularly those that employ the Zipser clamp, may be less accurate, because the lacuna is compressed or a catheter may be placed beyond its orifice.\(^ {19}\)

Another fallacy is to misinterpret the diverticulum as contrast medium within the prepuce or on the skin. One must remember to keep the end of the urethra always in sight.

**Syringocele**

The dilated Cowper's gland duct is referred to as a syringoceles (in Greek, syrinx means "tube" and cele means "swelling"). These anomalies seem to be more common than previously reported. Syringoceles are cystic dilations of Cowper's gland duct within the bulbous urethra.\(^ {20}\) They are usually small, inconsequential lesions. But rarely, they can be of sufficient size to cause varying degrees of outlet obstruction.

Lesions of Cowper's gland duct have various appearances. A system to classify each of these appearances is offered to diagnose these lesions more precisely. The urethrographic and endoscopic characteristics of dilated Cowper's gland ducts are classified in 4 groups:

1. Simple syringoceles, a minimally dilated duct
2. Perforate syringoceles, a bulbous duct that drains into the urethra via a patent ostium and appears as a diverticulum
3. Imperforate syringoceles, a bulbous duct that resembles a submucosal cyst and appears as a radiolucent mass
4. Ruptured syringoceles, the fragile membrane that remains in the urethra after a dilated duct rupture\(^ {20}\)

Cystic dilatation of Cowper's gland ducts (Cowper's syringoceles) is uncommon in children and is frequently asymptomatic; however, it may cause urinary infection, hematuria, dysuria, and obstructive voiding symptoms. Only a quarter of children with obstructive syringoceles need surgical intervention. The true clinical significance of Cowper's syringoceles lies in its potential to cause urethral obstruction. Careful clinical, radiologic, endoscopic, and urodynamic evaluation is necessary to avoid unnecessary surgery.\(^ {21}\)

Management, where necessary, is usually by endoscopic unroofing. After unroofing, a diverticulum-like defect may result on the posterolateral wall of the bulbous urethra. However, these defects are rarely obstructing and do not need further management.

Marsupialization of the syringoceles can cure urinary infection and hematuria, but voiding symptoms may persist.

**Cobb's Collar**

The Cobb's collar or Moormann's ring is a congenital narrowing of the bulbar urethra with variable clinical presentation and obstruction grade. Tubular or cystic dilatation of Cowper's gland duct has been called a syringoceles, while congenital urethral narrowing is known as Cobb's collar.

It is important to distinguish congenital urethral obstruction, Cobb's collar, from type-III PUV as described by Young.\(^ {22}\) Congenital urethral obstruction, Cobb's collar, should be differentiated from a congenital obstructive posterior urethral membrane. Congenital obstructive posterior urethral membrane may be a variation of a type-I urethral valve, supporting the theory of unified morphology in boys with congenital obstruction of the posterior urethra. In some congenital obstructive posterior urethral membrane cases, the membrane may prolapse as far as the bulbar urethra. However, the leaflet is supported by attachments to the verumontanum in congenital obstructive posterior urethral membrane, but not in Cobb's collar.

Meticulous cystourethroscopy is indispensable for detecting a clinically significant bulbar narrowing. Transurethral incision of the lesion is useful as a primary treatment in the majority of
cases, even with concurrent vesicoureteral reflux and unstable bladder. A cold knife is preferable to electrocautery for incising this fine anterior urethral lesion.\(^{(23)}\)

It appears that syringoceles are often associated with a Cobb’s collar, in keeping with the possible origin of both structures from the region of the urogenital membrane. Narrowing in the bulbar urethra may, however, be an incidental finding in many of the cases.\(^{(24)}\)

The more proximal lesion is a membranous obstruction that is able to prolapse as far as the bulbar urethra, but has paramedian folds that attach along the posterior wall of the urethra to the verumontanum and is due to persistence of an embryologic attachment between the distal verumontanum and the anterior wall of the posterior urethra. The more-distal narrowing is not always obstructive and is primarily a bulbar urethral membrane; it is independent of the verumontanum and external sphincter and may represent a persistence of part of the urogenital membrane. There are 2 distinct types of congenital obstruction of the proximal urethra, with an association to the verumontanum being the distinguishing feature.\(^{(25)}\) Cobb’s collar or Moormann’s ring is a stricture of the bulbar urethra that is largely unrecognized but has considerable relevance urologically as the site of congenital or postinstrumental strictures.\(^{(26)}\)

**Duplication of Urethra**

Accessory urethra or duplication of the urethra is a rare anomaly. The vast majority occurs in the sagittal plane; however, collateral or side-by-side duplications in the absence of bladder duplication also exist.

Urethral duplications are divided into sagittal and collateral types.\(^{(27,28)}\) Those not associated with bladder duplications occur almost exclusively in males. They are often accompanied by other anomalies such as anorectal anomalies or duplication of the penis.\(^{(28,29)}\) Urethral duplication in females is almost always associated with bladder duplication (Figure 3).

The etiology of urethral duplication is not clear.\(^{(27)}\) It is probably due to misalignment of some sort between the termination of the cloacal membrane and its relationship with the formation of the genital tubercle and urogenital sinus.\(^{(28)}\) The following classification has been accepted in the literature:

- **Type I**, incomplete duplication;
- **Type II**, complete duplication, IIa. two meatuses, noncommunicating urethras arising independently from the bladder; IIb, a second channel arising from the first and exiting independently, complete duplication joining at one meatus;
- **Type III**, duplication as a component of caudal duplication.

This classification may apply to either dorsal or ventral duplications. The perineal or rectal type associated with a stenotic, normally located penile urethra, is placed in the IIa category. The Y-type fistula usually takes its origin from the prostatic urethra and is commonly associated with stenosis of the anterior portion of the normally situated urethra.\(^{(28)}\) Children with complete and incomplete forms of accessory urethras usually present with 2 urinary streams. Infection in the partially stenotic orifice is sometimes the presenting symptom.\(^{(28)}\)

A proper clinical examination (under anesthesia, if needed), a micturating cystourethrogram, an ascending urethrography, and a urethrocystoscopy will give a complete picture of the altered anatomy. Based on the findings with reference to adequacy of channels, abnormality of location, and symptoms, individually tailored treatment is advised. Most often, the ventral urethra is functionally better and can be utilized by plastic procedures, as necessary. The dorsal urethra becomes obliterated either spontaneously or by ablative intervention.
Many asymptomatic children can be left untreated. Cosmetic correction of the division of the septum and creating an apparently single terminal orifice may be necessary when the orifices are close to each other at the tip of the glans. In some cases an end-to-side urethrourethrostomy may be done.\(^{30}\)

**Megalourethra**

Congenital megalourethra is a rare disorder characterized by the congenital absence of the corpus spongiosum and/or corpus cavernosum, leading to dilatation of the urethra.

Megalourethra was described originally by Nesbitt\(^{31}\) as a rare congenital abnormality characterized by dilatation of the penile urethra. This defect has been classified as a scaphoid or fusiform deformity. In the scaphoid variety, the corpus spongiosum is thought to be the only abnormal segment, whereas the fusiform variety is also associated with defects of the corpora cavernosa. However, this distinction is arbitrary and is not based on any recognized embryologic difference between the two varieties.\(^{29}\)

Megalourethra is especially common in association with the prune-belly syndrome and may represent a defect in development of the mesoderm, one of the proposed causes for the prune-belly anomaly.\(^{32}\)

**Prostatic Urethral Polyps**

Congenital posterior urethral polyps are rare, benign lesions that can cause a variety of symptoms in young boys; the diagnosis is usually made by cystourethrography and ultrasonography where the polyp appears as a soft tissue mass arising at the base of the urinary bladder.

Hunter is credited with the first documented case of urethral polyp, and Thompson reported the first case in a human.\(^{33}\)

Urethral polyps are rare and anterior urethral polyps are even rarer. They are usually of congenital origin. Urethral polyps in children occur exclusively in boys; the average age is 5.2 years. Mostly, they arise in the posterior urethra, usually proximal to the membranous urethra. But, anterior urethral polyps are still very rare.\(^{34}\) They are usually single and rarely multiple.

Urethral polyps have been speculated to represent a developmental error in the invagination process of the submucous glandular material of the inner zone of the prostate.\(^{35}\) They are benign lesions (not to be confused with the polypoid masses of a sarcoma botryoides), and transurethral excision of the polyps is curative.\(^{36}\)

Presentation of urethral polyps is varied consisting of dysuria,\(^{37,38}\) retention,\(^{37}\) secondary enuresis,\(^{39}\) and hematuria.\(^{33}\) They may present with obstructive symptoms.\(^{40}\) Occasionally, they present in adults with hematuria.\(^{33}\) Williams\(^{41}\) has reported associated vesicoureteral reflux.

Diagnosis of anterior urethral polyps is by voiding cystourethrography and cystourethroscopy.\(^{41}\) However, the penile and bulbar urethra should be palpated in patients presenting with retention or dysuria. Indurated area or lump in the absence of radio-opaque stone on radiography is suggestive of stricture or rarely a polyp.

The main symptom is bladder outlet obstruction, either intermittent or acute. Color Doppler ultrasonography may be diagnostic by revealing the polyp's vascular stalk arising from the posterior urethra.\(^{42}\) Transurethral resection of the polyp is the treatment of choice.\(^{36}\) However, when this is complicated by the displacement of the polyp into the bladder, transvesical removal could be an acceptable alternative.\(^{36-43}\)

Histologically, these lesions are benign and there have been no reported recurrences when they have been removed completely. The histologic examination reveals a fibroepithelial core with transitional epithelium with squamous metaplasia at times.

**References**


