Urethral Imaging

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Martin Kathrins, Dana A. Weiss, Kavita Gupta, Douglas A. Canning, and Richard D. Bellah

M. Kathrins, MD
Division of Urology, Department of Urology, Children's Hospital of Philadelphia, Philadelphia, PA, USA

Division of Urology, Department of Surgery, University of Pennsylvania Health System, Philadelphia, PA, USA

D.A. Weiss, MD
Division of Urology, Department of Surgery, The Children's Hospital of Philadelphia, 34th Street and Civic Center Blvd, 3rd Floor – Wood Center, Philadelphia, PA 19104-4399, USA
e-mail: weissd1@email.chop.edu

K. Gupta, BA, MD
Division of Urology, Department of Surgery, The Children's Hospital of Philadelphia, 34th Street and Civic Center Blvd, 3rd Floor – Wood Center, Philadelphia, PA 19104-4399, USA
e-mail: kgupta77@gwmail.gwu.edu

D.A. Canning, MD (✉)
Division of Urology, Department of Surgery, The Children's Hospital of Philadelphia, 34th Street and Civic Center Blvd, 3rd Floor – Wood Center, Philadelphia, PA 19104-4399, USA

Division of Urology, Department of Surgery, Perelman School of Medicine, University of Pennsylvania, Philadelphia, PA, USA
e-mail: canning@email.chop.edu

R.D. Bellah, MD
Perelman School of Medicine, University of Pennsylvania, Philadelphia, PA, USA

Department of Radiology, The Children's Hospital of Philadelphia, 34th Street and Civic Center Blvd, Philadelphia, PA 19104, USA
e-mail: bellah@email.chop.edu

Imaging of the pediatric and adolescent urethra is largely the purview of dynamic fluoroscopy. However, recent advances have led to the use of ultrasound and MR imaging in an attempt to reduce the traditionally invasive nature of urethral imaging.

Normal Anatomy (Fig. 15.1)

The male urethra is composed of an anterior segment and a posterior segment. The anterior urethra is distal to the urogenital diaphragm and includes the bulbar and pendulous (penile) urethra. The posterior urethra is composed of the membranous (sphincteric) and prostatic urethra. The one constant landmark in fluoroscopic imaging of the urethra is the verumontanum, which is seen as an indentation in the membranous urethra. On oblique urethrogram, narrowing of the urethra may be seen as it traverses the urogenital diaphragm. The most distal aspect of the penile urethra is the fossa navicularis (the histologic convergence of urothelium and distal squamous epithelium) which is visualized as a slightly narrowed segment just proximal to the meatus.

The female urethra extends from bladder neck to introitus. The striated sphincter complex encompasses the distal two-thirds of the urethra. Dynamic contraction of the complex leads to urethral narrowing and has been mistaken for meatal stenosis in the past. Reflux into the vagina is frequently seen after voiding on urethrograms.

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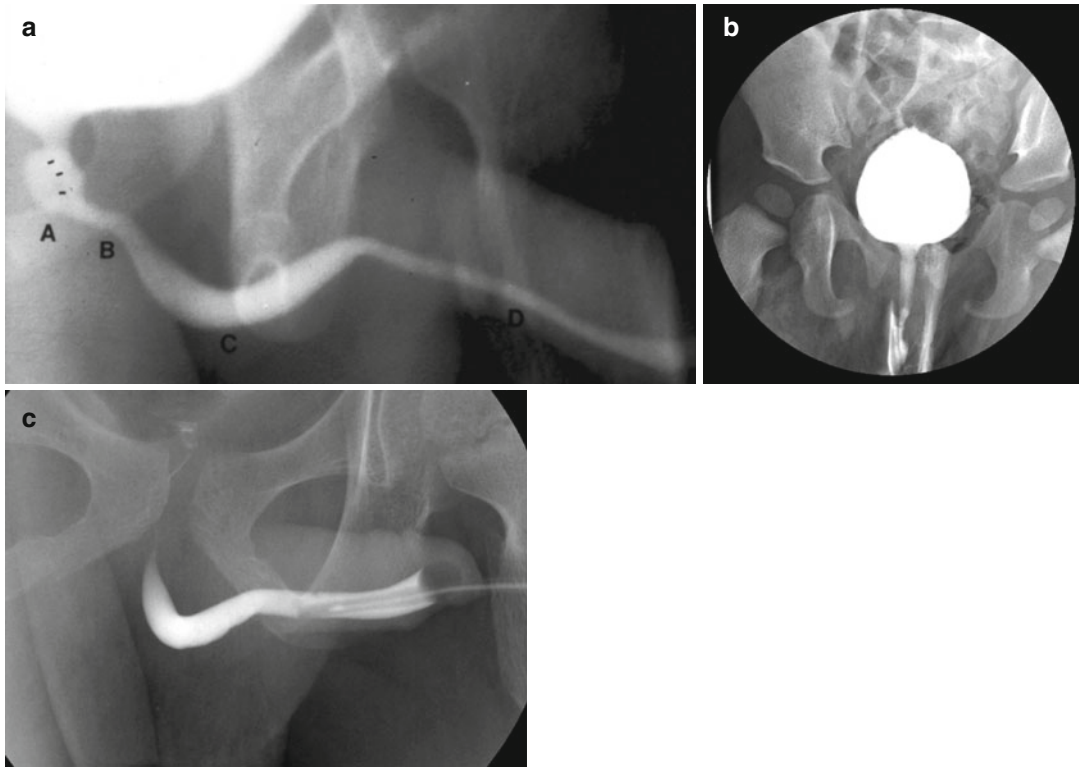

Fig. 15.1 (**a**) Normal male VCUG. *A* prostatic urethra; *B* membranous urethra; *C* pendulous urethra; *D* penile urethra. (**b**) Normal female urethra on VCUG. (**c**) Normal male RUG

Voiding Cystourethrogram

The voiding cystourethrogram (VCUG) is the most effective method to dynamically image the urethra. Clinicians also use the VCUG to evaluate the upper tracts in cases where vesicoureteral reflux exists. A "cyclic" VCUG where the bladder empties multiple times is helpful to identify vesicoureteral reflux (VUR) in duplicated systems. An anterior-posterior scout fluoroscopic image is first performed to evaluate for radiopaque stones and bony abnormalities, such as pelvic diastasis. It is our practice to use periprocedural antibiotics with gram-negative coverage and to obtain urinalysis and urine culture in select children prior to imaging.

An appropriately sized catheter is inserted into the bladder through the urethra or an indwelling suprapubic tube, without inflation of the catheter balloon. The bladder is drained and a urine culture obtained.

In males, the patient is placed supine with the penis laterally displaced for oblique images, while females remain supine for AP images. Iodinated contrast of an age and weight appropriate volume is instilled to opacify the bladder. Filling the bladder beyond predicted age and weight capacity may overestimate vesicoureteral reflux. The child then voids while monitoring with intermittent fluoroscopy. The voiding component evaluates the bladder neck for funneling and radiographic bladder emptying several minutes post-void. The appearance of the sphincter is monitored as the bladder neck begins to open at the initiation of voiding.

The VCUG provides essential imaging of the posterior urethra. Anterior-posterior images are obtained to evaluate the bladder and lateral/oblique images for the bladder neck and the urethra. Close communication between dedicated consistent radiology and urology staff is important

for the best outcomes. Interobserver reliability may be poor, specifically when evaluating obstruction at the bladder neck and in the prostatic urethra in retrospective series [1].

Retrograde Urethrogram

The retrograde urethrogram—without voiding phase—is the preferred study to evaluate the anterior urethra, particularly in the male. For this study, a catheter is inserted into the fossa navicularis and the balloon partially inflated and placed on gentle traction to occlude the distal urethra. With the child in the lateral decubitus position, contrast is gently injected with slow steady pressure under fluoroscopy to overcome resistance of the external sphincter opacifying the posterior urethra as much as possible. In attempting to diagnose a suspected stricture to facilitate surgical planning, we try to opacify flanking aspects of the normal urethra by rotating the child or the fluoroscopy unit. To perform an adequate retrograde urethrogram in the shorter female urethra, a "double-balloon" catheter is sometimes used in adults to occlude both the meatus and bladder neck; this is not commonly used in children.

Newer Modalities

Magnetic resonance imaging (MRI) is used more commonly to image the upper urinary tract in children. Despite the use of triple-phased contrast-enhanced imaging (MR urogram) for functional imaging of the upper urinary tracts, a regular role for urethral imaging is yet to be identified.

Voiding enhanced urosonography is a newer imaging modality that reduces radiation exposure. This technique is under active investigation but has not yet been widely used [2]. Researchers have used it most often as an alternative to VCUG in children with suspected VUR. Nonlinear imaging techniques are used to differentiate anatomic structures from contrast-enhanced bubbles (galactose-palmitic acid) in this technique [3]. Transperineal imaging is used for males with a full bladder and normal micturition. Alternatively, the transpubic approach may be used to diagnose males with inability to control micturition and in females to better examine the bladder neck. Isolated reports describe the use of transpubic ultrasound to identify posterior urethral valves in two patients, and a dilated prostatic utricle and anterior urethral diverticulum were also diagnosed in two patients [4]. In a prospective comparison of VCUG and contrast-enhanced ultrasonography in patients with suspected urethral pathology, ultrasonography accurately diagnosed posterior urethral valves, anterior valves, urethral stenosis, and detrusor sphincter dyssynergia in all. Two patients with syringocele were not identified with ultrasound but were diagnosed with VCUG [5].

While ultrasonography is useful for studying stricture length and depth in adults with anterior urethral strictures, we use it less in children. One study examined differences in echogenicity, luminal narrowing, and changes in periurethral tissue in non-contrast-enhanced ultrasound to diagnose anterior urethral stricture. The authors estimated the degree of urethral distention to determine stricture length. According to the authors, the use of perioperative ultrasound led to a change in planned surgical approach in 58 % of patients [6]. However, VCUG remains the study of choice for diagnosis of posterior urethral pathology.

Imaging of Acquired and Congenital Defects

Adequate imaging of the urethra is important for proper diagnosis of both acquired and congenital urological abnormalities. A wide range of congenital anomalies affects the urethra and requires urgent and repeated imaging. These include posterior urethral valves (PUV), prune belly syndrome, megalourethra, anterior urethral diverticula, anterior urethral valves, and urethral duplication. Acquired conditions may include trauma, strictures, diverticula, infections, urethrorrhagia, and sources of obstruction such as polyps or stones.

Congenital Urethral Anomalies

Congenital Urethral Polyps

Congenital urethral polyps arise from the verumontanum. They may result in bladder outlet obstruction in boys. Boys may complain of discomfort and straining to void. The diagnosis is usually made by VCUG, but polyps may be seen on ultrasound as well. The diagnosis is confirmed and the polyp is managed with cystoscopy and transurethral resection [7].

Posterior Urethral Valves (Fig. 15.2)

PUV are the most common congenital cause of bladder outlet obstruction in children. A defect in the developing Wolffian duct folds is thought to be causative, and the valves become obstructive at variable times after the 8th week of development [8]. PUV are often diagnosed before birth—with ultrasound, with as many as 45 % of cases identified in utero [9]. Findings include a distended bladder often with bilateral hydronephrosis and a variable finding of oligohydramnios, depending on the severity of the obstruction and degree of renal injury. Postnatal renal bladder ultrasound (RBUS) and VCUG confirm the diagnosis. The sensitivity of RBUS in detection of valves is 95 % [10]. Findings of RBUS include a dilated, thick walled bladder often with diverticula/cellules, dilated upper tracts, and a dilated posterior

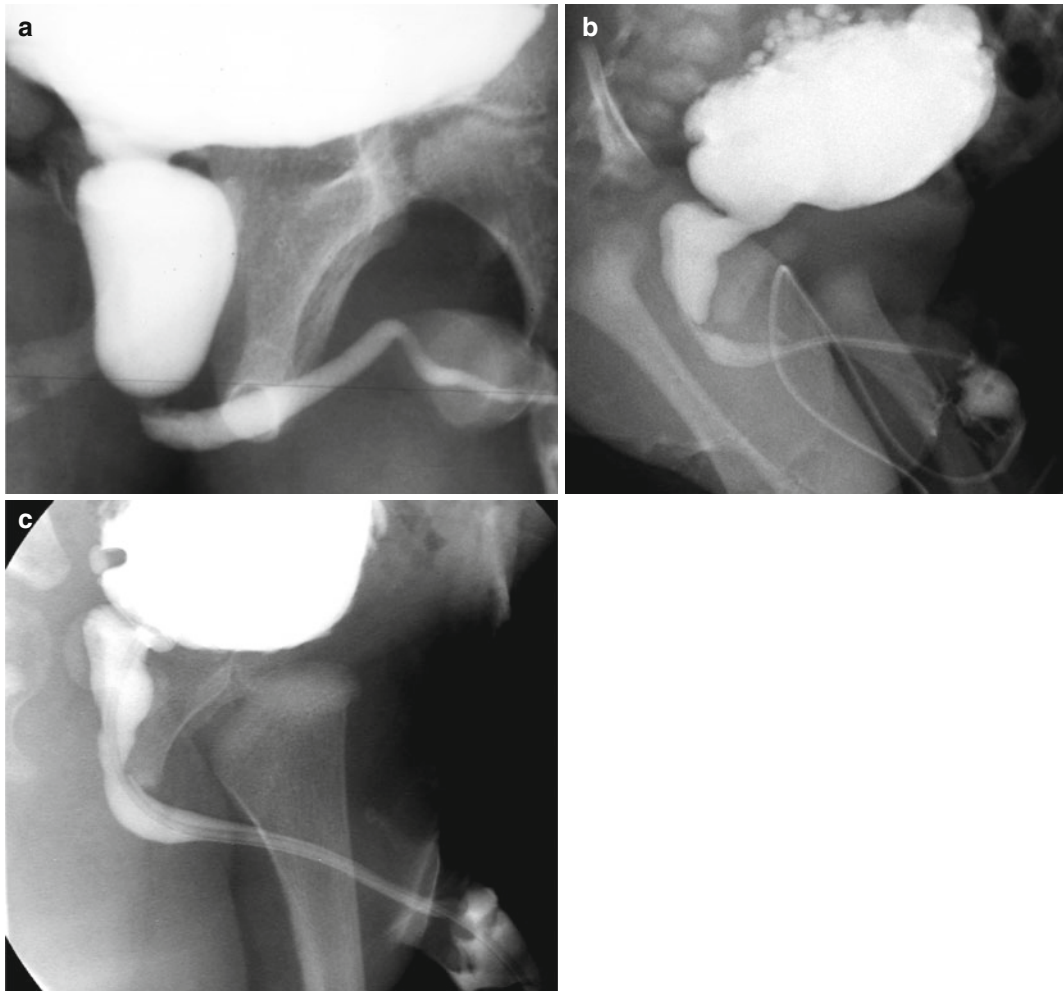

Fig. 15.2 Posterior urethral valves. (**a**) PUV showing dilated and elongated urethra. (**b**) PUV showing dilated and elongated urethra and multiple bladder diverticula. (**c**) PUV after valve ablation showing smooth transition across posterior urethra

urethra, the "key-hole" appearance. Urethral catheterization is the initial treatment prompted either by retention and/or ultrasound imaging.

VCUG/RUG is performed via urethral catheter or suprapubic tube, if present. The voiding phase is critical for differentiation from other forms of bladder outlet obstruction. While removal of the urethral catheter during the voiding phase is not always necessary, as it does not obscure the identification of the valves [11], our practice does remove the catheter unless contraindicated. Pathognomonic findings include the appearance of the valve which may be seen as an indentation on lateral imaging along with a dilated and elongated posterior urethra. Incomplete bladder emptying is also present. VUR is present in the majority of cases. After valve ablation, a follow-up imaging study is recommended in 4–6 weeks to assess decompression. If the degree of dilation of ureters and kidneys does not improved despite a decompressed bladder, a repeat VCUG could be performed to rule out residual obstruction.

Prune Belly Syndrome

Prune belly (Fig. 15.3) or triad syndrome (PBS) describes a triad of laxity of abdominal musculature, bilateral undescended testicles, and GU tract abnormalities, which include hydronephrosis, renal dysplasia, and urethral dilation. Variations of PBS may occur along a spectrum of these three clinical characteristics—so-called pseudo-prune belly syndrome [12]. Urethral dilation in PBS may arise from one of three etiologies. The first, urethral obstruction, occurs early in gestation and is believed to occur in 20 % of cases, from urethral atresia, urethral valves, or urethral diverticulum. Alternatively, the urethral dilation may be related to a functional abnormality of bladder emptying without obstruction. In the absence of an obstructive lesion, the dilation may result from prostatic hypoplasia. VCUG in PBS shows tapering of the dilated posterior urethra to the membranous urethra. A prostatic utricle is often present [13, 14].

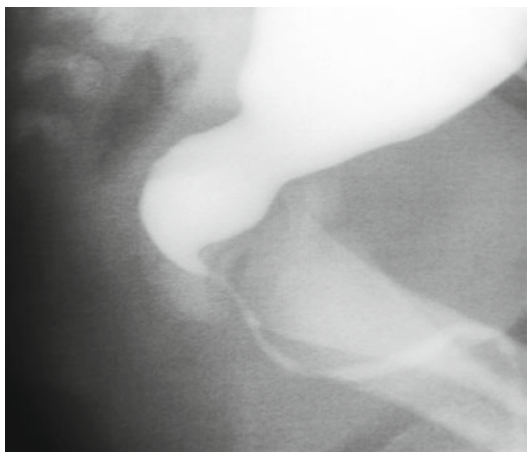

Fig. 15.3 Prune belly with wide open bladder neck, dilated but short posterior urethra

Congenital Anterior Urethral Obstruction

At least 260 reports have described cases of congenital anterior urethral obstruction, either congenital anterior urethral diverticula or anterior valves. Astute clinicians will suspect diverticula in boys with ventral penile swelling, post-void dribbling, dysuria, and recurrent urinary tract infection. When the diverticulum fills during voiding, it may progress to obstruction. Two forms of diverticula have been identified—saccular (Fig. 15.4) and globular. Diverticula are differentiated radiographically from congenital

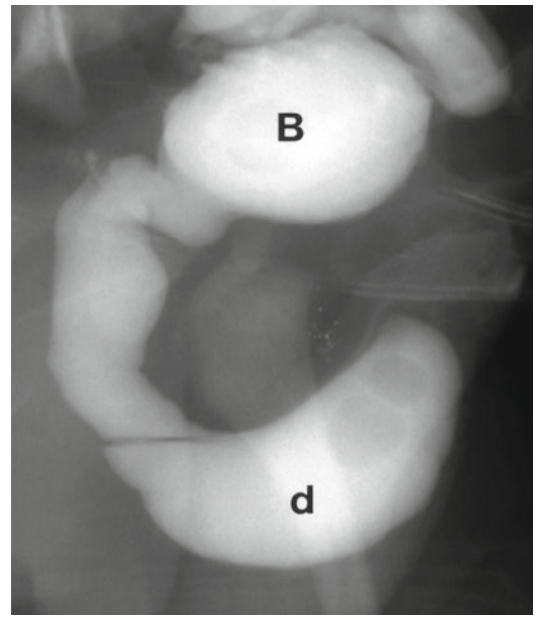

Fig. 15.4 Image from a VCUG of a boy with a saccular anterior urethral diverticulum. The bladder (*B*) is nearly empty, and contrast is seen in the posterior urethra and then filling the diverticulum (*D*)

anterior valves by the presence of an acute angle between the proximal parts of the dilated urethra, which is not present with valves [15]. Iatrogenic anterior urethral diverticula may also develop after hypospadias repair due to distal obstruction and after repair of anorectal malformation if the rectourethral fistula is not trimmed close to the urethra. Diagnosis is made by VCUG/RUG.

Anterior urethral valves are a rare, obstructing condition of unclear embryologic origin. The valves originate close to the penoscrotal junction or bulbar urethra. They arise from the ventral portion of the urethra and obstruct urine flow during voiding. Many believe that valves are part of the spectrum of congenital urethral diverticula. Indeed, valves may develop into diverticula secondary to outflow obstruction. However, pathologic findings indicate that congenital valves are always bordered by the corpus spongiosum and true congenital urethral diverticula develop outside the corpus spongiosum [16]. The gold-standard diagnostic study remains VCUG/RUG; however, contrast-enhanced ultrasonography has been used to make this diagnosis.

Urethral Duplication (Fig. 15.5)

Urethral duplication more commonly occurs in the sagittal axis rather than the coronal according the aggregate of case reports. The duplication develops from misalignment of the cloacal membrane, the genital tubercle, and the urogenital sinus. Dorsal duplication may occur with a single normal meatus, dorsal penile curvature, and a second epispadiac meatus. The dorsal urethra may communicate with the more normal, ventral urethra. Ventral duplicated urethra, which is less common is also known as "Y-type duplication," splits at the prostatic urethra. The duplicated ventral urethra may track to the rectum or perineum [17]. Urethral duplication is most easily diagnosed with VCUG/RUG. In some cases, additional retrograde urethrogram of a small, accessory urethra is necessary to fully image the aberrant duplicated urethra.

Cowper's Syringocele (Fig. 15.6)

Cowper's syringocele is a condition resulting from a rare, cystic dilation of Cowper's duct. The Cowper's glands are located in the deep perineal pouch within the urogenital diaphragm and are involved in secretion of pre-ejaculate fluid. Cowper's syringocele may be open to the urethra or closed. Either may present with pain. Closed syringocele may also present with obstructive voiding symptoms due to passive expansion and mass effect. Open syringocele may present with post-void dribbling, urethral discharge, and recurrent urinary tract infection. It may also obstruct

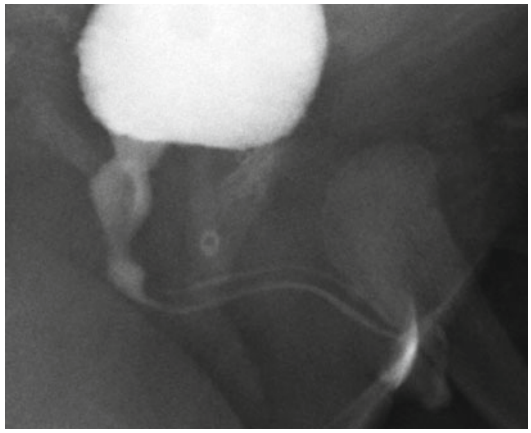

Fig. 15.5 Urethral duplication. The ventral urethra is of normal caliber while the dorsal one is narrow but does emanate from the bladder and terminate on the glans penis

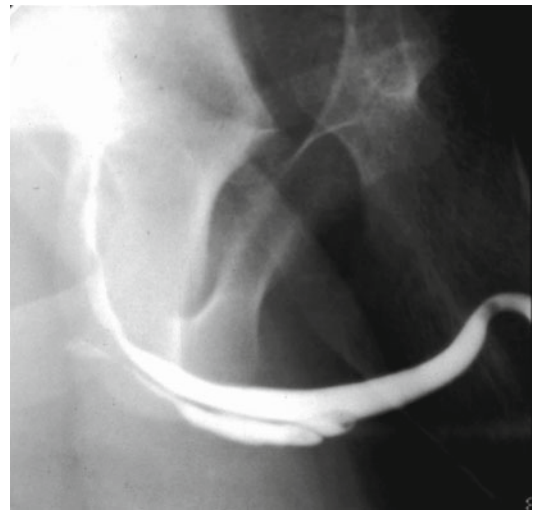

Fig. 15.6 Cowper's syringoceles. They are oblong contrast-filled spaces running parallel to the urethra starting from the bulbous urethra

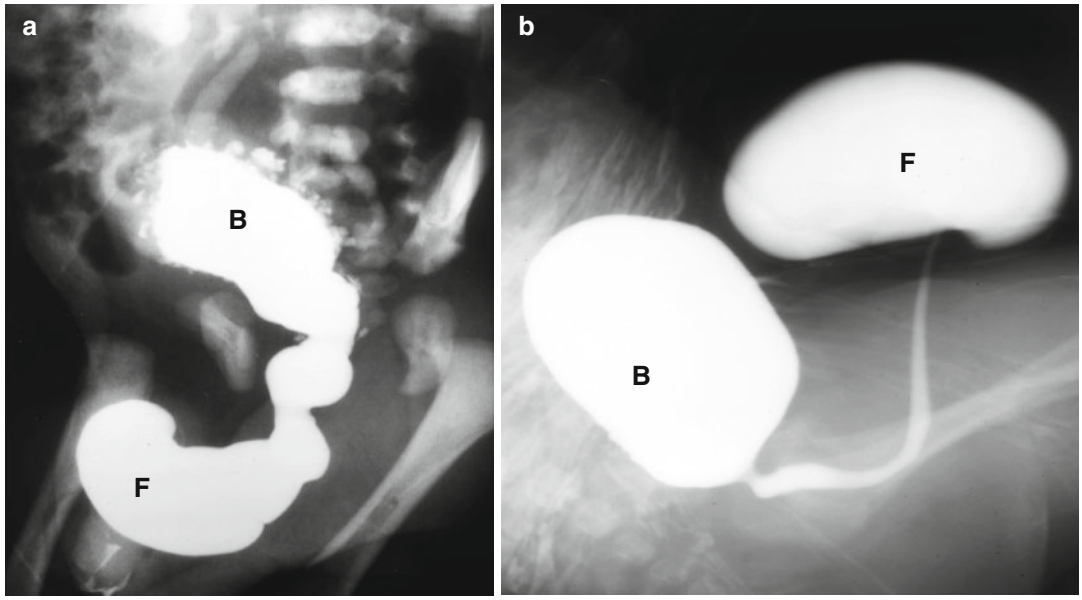

Fig. 15.7 Scaphoid (**a**) and fusiform (**b**) megalourethrae are seen demonstrating the billowing out of the urethra during voiding. The entire urethra is affected in the fusiform type while only a segment of the urethra is affected in the scaphoid type. *B* bladder; *F* megalourethra

urethral flow in some cases. Either transrectal or transperineal ultrasound may reveal a cystic structure (closed) or, in one case report, an open syringocele [18]. Retrograde urethrogram or VCUG will aid diagnosis. Cystoscopy with careful attention to the proximal bulbar urethra is diagnostic.

Congenital Megalourethra (Fig. 15.7)

Megalourethra is a congenital, non-obstructive dilation of the anterior urethra. It presents with an enlarged, deformed phallus with distention during voiding. It is more commonly seen in patients with PBS than in other syndromes. Two forms are recognized: fusiform and scaphoid. Fusiform megalourethra is secondary to a deficiency of the corpus cavernosum. On VCUG, the entire urethra dilates with voiding. Scaphoid megalourethra is caused by a deficiency of the corpus spongiosum. Only the ventral urethra dilates with voiding in these cases. Catheterization of these patients may be difficult and may require urethroscopy.

Urethral Fistula

Urethral-enteric fistulae may develop secondary to congenital anorectal malformations. Eighty percent of boys with anorectal malformations have uro-enteric fistula, most often associated with imperforate anus. An initial VCUG/RUG as a screening test is diagnostic [19]. Alternatively, MR imaging in the initial evaluation may provide a noninvasive alternative [20]. However, diagnosis of the fistula is often made in the operating room during colostomy creation or distal colostogram performed prior to definitive pull-through procedure. Congenital urethral cutaneous fistula is extremely rare. They may be associated with penile curvature or hypospadias. We have seen at least two of these associated with anorectal anomalies. One case report cites a congenital urethro-perineal cutaneous fistula in a 12-year-old male [21]. The appearance of the VCUG/RUG appears similar to urethral duplication; however, the dorsal channel is invariably functional. Clinically, boys with congenital fistulae may present with recurrent UTI, pneumaturia, or perineal dribbling.

Urethral Trauma (Fig. 15.8)

Urethral trauma is far more common in males than in females but can be devastating in both.

Male urethral trauma has been divided into anterior and posterior disruption [22] and has been subclassified within these groups based on the injury extent seen on RUG. In pediatrics, the most common source of trauma is blunt pelvic trauma causing posterior urethral injury from pelvic fracture (still a rare event occurring in only 0.47–4.2 % of pelvic fractures and pelvic fractures have an incidence of only 2.4–4.6 %). Because the developing prostate is more prone to rupture than in adults and due to the relative intra-abdominal location of the bladder in children, blunt pelvic trauma poses an increased risk to the bladder neck and penopubic ligaments [23, 24]. Anterior urethral disruptions due to straddle injuries have an incidence of 0.6–10 % [25]. Radiologic grading of male urethral injuries varies by the location of injury.

In boys, the initial imaging in the trauma setting remains a RUG; however, caution is warranted because imaging that indicates a complete disruption does not rule out a partial tear [26] (Fig. 15.8). Opacification of a periurethral urinoma proximal to urethral defect may be mistaken for the lumen of the posterior urethra after a complete disruption injury. The use of several oblique angles during urethrography may avoid this misinterpretation. In girls, due to the limitations of a RUG in the shorter urethra, a cystoscopy and vaginoscopy is superior to any radiographic imaging and is indicated if clinical suspicion is present.

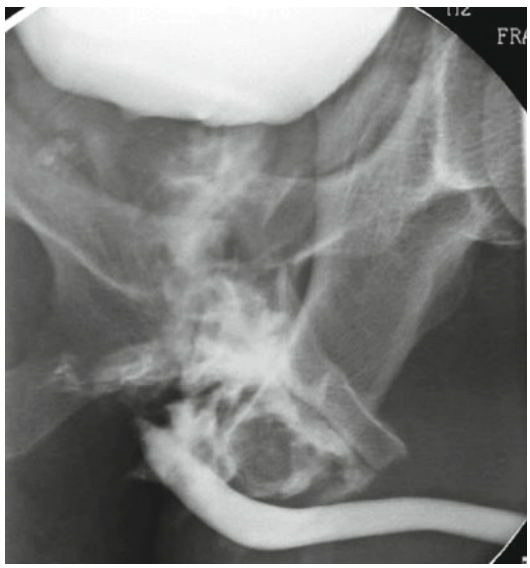

Fig. 15.8 RUG showing complete disruption of posterior urethra

Urethral Strictures (Fig. 15.9)

Urethral strictures may be due to infection (especially C. trachomatis in adolescent males and gonococcal urethritis, which can be seen at any age), iatrogenic injury such as instrumentation, or trauma [27]. The gold standard for imaging strictures is a RUG/VCUG. However, in children, the combination of RUG/VCUG is not as accurate in assessing the length of a defect. This is due to an inability to opacify the posterior urethra due to an inability to relax the sphincter [8]. A static cystogram performed via an indwelling suprapubic tube that shows opacification of the posterior urethra may indicate the presence of an incompetent bladder neck. Overestimation or underestimation of the defect may lead to a suboptimal surgical approach between the options of transpubic or perineal [9].

Sonourethrography, with sterile lubricating jelly instilled retrograde has become a common adjunct to RUG in order to guide treatment planning and help estimate length of stricture. While MR urethrography has been evaluated in adults for stricture disease, it has still not been widely accepted. In adults, comparisons have indicated equal or better accuracy compared with RUG, with improved anatomic detail of associated soft tissue injury provided [28, 29].

Postoperative strictures from hypospadias repairs are often evaluated functionally rather than by a specific appearance, so there are fewer data on the optimal imaging in this setting. At our institution, children are evaluated with noninvasive urine flow when they present with complaint of difficulty voiding. If there is diminished flow, we consider RUG and more commonly surgical exploration and reconstruction.

While rarer in young boys than adults, lichen sclerosis (LS) may contribute to stricture formation. LS causes genital scarring with possible voiding and sexual consequences [30]. The incidence is confused by a historical failure to identify and diagnose this condition. LS can lead to

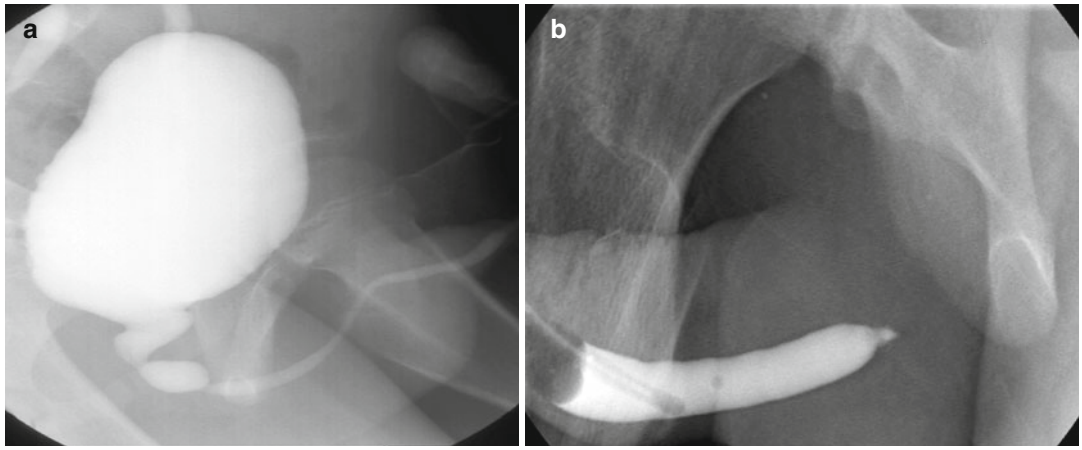

Fig. 15.9 (**a**) VCUG of bulbar urethral stricture. (**b**) RUG of urethral stricture

true stricture formation in 2–40 % of cases. LS may also develop after failed hypospadias repair with distal urethral obstruction and can progress to cause severe pan-urethral stricture disease. Notable findings include development of squamous metaplasia involving the ducts of periurethral glands that stiffen and open. RUG/VCUG is used to evaluate the urethra and may even demonstrate these open periurethral glands [31].

The myriad of pediatric urethral conditions may be effectively diagnosed with a RUG and VCUG. Newer technologies have at times been found to be equally effective but have yet to achieve the proliferation and practiced results of the traditional fluoroscopic urethrography. Facility with these techniques and their interpretation is essential for any pediatric urologist.

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