Chapter 21 General Principles of Cystourethroscopy

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Abstract Many children require cystourethroscopy to diagnose, characterize, or treat their genitourinary disorder. Several diseases or anomalies are best treated by cystoscopic minimally invasive means, including posterior urethral valves, ureteroceles, vesicoureteral reflux, urethral stricture/fistula/diverticulum, hematuria, urinary stones, tumors, or other rare birth defects. In this chapter, preoperative, operative, and postoperative techniques and tips of cystourethroscopy are discussed, highlighting its versatility.

Keywords Cystoscopy • Cystourethroscopy • Pediatric • Infant • Genitourinary • Anomalies

As a form of minimally invasive surgery, endoscopy of the lower genitourinary tract of the pediatric patient can achieve diagnostic and therapeutic goals for a broad range of pathological entities. Advances in instrumentation have permitted endoscopic treatment of even premature infants and in utero fetal surgery [1]. This chapter focuses on general principles of pediatric cystourethroscopy. The reader is referred to other chapters in this text for more detailed discussions of the management of other clinical entities.

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Fig. 21.1 Cold-knife incision of posterior urethral valves. The "half-moon" knife, seen in the *center* of the image, is cutting through the right valve leaflet. The verumontanum is seen in the right third of the image. The left valve leaflet is out of the image



Indications and Contraindications

Recurrent urinary tract infections (UTIs), urinary incontinence, obstructive uropathy, urosepsis, and radiological anomalies are the usual indications for lower tract endoscopy. Although many diagnoses are made before cystoscopy by using ultrasound, cystourethrography, CT scan, nuclear scan, IVP, and/or MRI, many pediatric cases require further delineation of the anatomy and physiology by endourological techniques. Cystoscopy followed by transurethral incision of posterior ure thral valves for obstructive uropathy [2, 4, 5] is a common indication (Fig. 21.1). Similarly, transurethral incision of ureterocele(s) for outlet obstruction or urosepsis [3] is another clear-cut indication (Fig. 21.2), while prophylactic intervention after prenatal detection is more debated. Cystoscopically guided ureteral or bladder neck injection of bulking agents is frequently employed to treat vesicoureteral reflux (VUR) and urinary incontinence, respectively (see Chaps. 22 and 23). Some surgeons recommend routine cystoscopy before open ureteral reimplantation to assess for the configuration of a prior refluxing ureter, missed ureteral duplication (Fig. 21.3), or cystitis, which would cancel the open surgery. Male urinary incontinence should be evaluated cystoscopically after hypospadias repair or abnormal retrograde urethrogram, assessing for urethral stricture (Fig. 21.4), urethral duplication (Fig. 21.5), or urethrocutaneous fistula. In rare cases, gross hematuria in the pediatric patient may warrant study after a thorough negative medical and radiological evaluation. If clot retention occurs, clot evacuation can be achieved Fig. 21.2 Ureterocele. Figure shows a right moderately sized ureterocele associated with febrile UTIs, right complete ureteral duplication, and a multicystic dysplastic hydronephrotic upper pole moiety. It was transurethrally incised



Fig. 21.3 Complete ureteral duplication. View of the right trigone reveals two ureteral orifices, the lateral, cephalad refluxing orifice (*black arrow*) serving the lower pole, and the medial, caudal orifice (*white arrow*) serving the upper pole duplex kidney



cystoscopically with the instillation of therapeutic agents if indicated. Cystourethroscopy can serve the purpose of ureteral access for retrograde or antegrade upper tract imaging and lithotripsy techniques; however, a trial of medical

Fig. 21.4 Urethral stricture. Urethroscopy revealed a pinpoint lumen in the bulbar urethra (*arrow*) at the site of a prior visual internal urethrotomy. Open primary urethroplasty was required to correct this recurrent urethral stricture



Fig. 21.5 Urethral duplication. When a dorsally foreshortened foreskin was noted, cystoscopic inspection revealed a partial urethral duplication to the symphysis. In this image, with the foreskin retracted, the *black wire* enters the dorsal nonfunctioning urethra, and the metal urethral sound enters the ventral functioning urethral meatus



therapy is warranted because many stones pass in children. Retrograde placement of an occlusion balloon at the ureteropelvic junction can prevent antegrade migration of stone fragments during percutaneous nephrolithotripsy. Retrograde ureteral stenting may be useful at the time of extensive tumor resection or at the time of laparoscopic pyeloplasty. Bladder stones can be endoscopically removed or fragmented via urethra, appendicovesicostomy, or percutaneous cystostomy approaches. At the time of cystoscopy in the child with an open bladder neck due to epispadias (Fig. 21.6) or classic bladder exstrophy, a ballooned catheter can be used for



Fig. 21.6 Female epispadias with bilateral VUR. Exam under anesthesia reveasl subtle case of female epispadias associated with bilateral VUR and urinary incontinence. (a) External genitalia of female epispadias - note the horizontally wide urethral meatus with open urethral plate dorsally. The clitoris is bifid. The refluxing left (b) and right (c) ureteral orifices are seen



Fig. 21.7 Intersex. (a) Urogenital sinus of congenital adrenal hyperplasia (*CAH*). Cystourethroscopic evaluation of the urogenital sinus orifice reveals the bifurcation of the urethra (*white arrow*) and the vagina (*black arrow*). Cystourethroscopic placement of Fogarty balloons into the urethra and vagina aids the surgical reconstruction of the urogenital sinus. (b) Urethroscopic view of entrance into large utricle (*arrow*) on the verumontanum of an intersex patient with mixed gonadal dysgenesis

cystography to measure bladder capacity under anesthesia and assess for vesicoureteral reflux.

Cystourethroscopy with vaginoscopy is indicated in the patient with intersex (Fig. 21.7), urogenital sinus, or cloaca to delineate the surgical anatomy for repair. Tissue diagnosis of genitourinary malignancy (rhabdomyosarcoma, urothelial cancer) can be achieved by cystoscopy with tumor biopsy.

Contraindications include active bleeding disorders, hemodynamic instability, or untreated UTI/urosepsis.

Preoperative Investigation

Cystourethroscopy generally requires general anesthesia. Therefore, a standard preoperative evaluation, considering cardiopulmonary, endocrinological, and hematologic disorders that increase anesthetic risks, is necessary. Children with identified disorders may require preoperative blood chemistries, and children with congenital adrenal hyperplasia require stress steroid dosing. Preoperative radiological investigations often include ultrasound, cystourethrography, CT scan, nuclear scan, IVP, and/or MRI. Sterile urine is required to reduce risk of upper tract UTI prior to invasive instrumentation.

Preoperative Patient Preparation

Once cleared for surgery and meeting NPO restrictions, an oral sedative is given to prevent separation anxiety. At this time, IV antibiotics may be administered in the child with recurrent UTIs, depending on physician preference.

Specific Instrumentation

Most cystoscopic suites are equipped with monitors for fluoroscopic and video camera imaging, which allow multiple viewers, teaching, optical magnification, and video recording. A fiberoptic xenon light source and electrocautery are also required. Cystoscopic irrigant (sterile normal saline or sterile water) should be warmed to body temperature to diminish hypothermia. Several companies manufacture pediatric endoscopic equipment, including Wolf, Storz/Olympus, and ACMI. Given the delicate nature of this equipment, it is crucial to have several scopes available in case of equipment malfunction or unanticipated needs. Rigid pediatric cystoscopes range from 5 Fr to adult sizes, and the pubertal status of males should be noted to help judge the equipment needed. The 5 Fr "all-in-one" cystoscope is a one-piece instrument with united telescope and sheath; the 2.5 Fr to 3 Fr working channel is rather limiting. However the working channel increases in the larger scopes, with greatest caliber in the "all-in-one" cystoscopes. Other scopes consist of two pieces: the interchangeable telescope $(0^\circ, 30^\circ, \text{ and } 70^\circ)$ and the sheath. A range of reusable and disposable equipment (graspers, biopsy forceps, bugbee electrode, wires, catheters, stents, balloons, baskets, laser fibers, and STING needles, to name a few) exist to achieve the indicated therapy but may be impossible if the working channel caliber is <5 Fr. Pediatric cystoscopes with an offset lens allow straight entry into the working channel. 7.5 Fr flexible or semirigid ureteroscopes should be on hand if ureteral access is necessary. Pediatric resectoscopes, ranging from 7.5 FR to adult sizes, require loops, balls, blades, or hooks unique to the FR size of the resectoscope. Resectoscopes can be used cold or hot (with electrocautery); however, most recommend sparing use of electrocautery to minimize thermal damage and stricturing with the highest stricture rates reported with loop resection [4]. Some have used holmium or Nd:YAG laser to cut valves or strictured tissue [4, 5]. It is convenient to have urethral sounds and/or bougies available for urethral dilation if needed.

Endoscopic bladder stone management requires the use of rigid and flexible cystoscopes. If percutaneous access to the bladder is needed, cystoscopically guided suprapubic access sheaths can be quite useful and come in an assortment of sizes, with 13 Fr to 18 Fr the most useful. To achieve stone fragmentation, electrohydraulic, ultrasonic, combined ultrasonic and pneumatic (Swiss lithoclast), or holmium laser lithotripsy can be used. Rigid probes include the electrohydraulic probes (3 Fr or 5 Fr), ultrasonic probes (as small as 5 Fr), and the Swiss lithoclast (3.3 and 3.8 mm). Of the flexible probes, holmium laser fibers are 200, 400, 600, or 1,000 μ m, and Swiss lithoclast has a 0.9 mm flexible pneumatic probe.

Operative Technique

After the induction of anesthesia, the patient is properly padded, positioned, and grounded for electrocautery. In the infant, the supine frog leg position may be adequate; however, an alternative is dorsal lithotomy position with leg suspension via towel rolls and tape at the padded knees. If fluoroscopy is not necessary, position the infant close to the anesthesiologist perpendicular on the bed to increase anesthetic safety (Fig. 21.8). Otherwise, the child will need to be moved down on the foot of the bed so the fluoroscopy arm can pass beneath (Fig. 21.9). The older child should be placed in dorsal lithotomy position with the legs in properly fitted stirrups.

Prior to the surgical preparation, a thorough examination under anesthesia is performed. The external genitalia are closely inspected for anomalies (genital configuration (Fig. 21.10), masses (Fig. 21.11), or ectopic orifices (Fig. 21.12)). After securing properly functioning instrumentation, a lubricated cystoscope is chosen of appropriate size for the child.

Cystourethroscopy of a female is straightforward, and often the greatest challenge is entering the urethral meatus. To minimize bacterial contamination, every effort should focus on endoscopy of the urethra and bladder prior to vaginoscopy. In some challenging cases, the urethral meatus can be identified by gentle outward (not lateral or downward) pull on the labia majora and can be found in a hypospadic position in some. The female urethral meatus should accept a 7.5 Fr to 8 Fr cystoscope in the term infant. Although the female urethra is significantly shorter than the male urethra, it shares the same mucosal vascular striations of the posterior urethra of the male, which should run parallel to the cystoscope. The female urethra is coapted to the bladder neck. On bladder entry, the yellow urine should be evacuated to aid visualization. Once distended with irrigant, the bladder should appear spherical with smooth walls and homogenous epithelium. One slit-like ureteral orifice is



Fig. 21.10 Vaginal agenesis. A thorough examination under anesthesia reveals







Fig. 21.11 Perineal mass. A thorough examination under anesthesia reveals perineal mass which bulges with valsalva. The mass was a right upper pole large ectopic ureterocele. Radiographic contrast was needle injected into the mass, retrograde filling the massively dilated upper pole ureter. Cystoscopic retrograde right lower pole ureterogram revealed an equally massive lower pole grade 5 refluxing ureter. The entire right kidney was nonfunctional and removed laparoscopically



Fig. 21.12 Ectopic ureters. (a) A thorough exam of an incontinent female under anesthesia detected a right ureter exiting on the perineum. The opaque catheter is in the urethra, and the black wire enters the ectopic ureteral orifice. One left and one right orthotopic ureteral orifices were seen within the bladder cystoscopically. Vaginoscopy revealed an ectopic left upper pole ureteral orifice just within the hymenal ring. This child had bilateral complete duplication with bilateral upper pole ureteral ectopia. (b) In a different patient, cystourethroscopic view of an ectopic left upper pole ureter (*arrow*) entering the female urethra. (c) An ectopic right ureter entering the top of the verumontanum serves this solitary functioning kidney in a male patient with recurrent febrile UTIs, Grade 5 left VUR, right trigonal diverticulum, and incompetent bladder neck. Intravenous administration of indigo carmine proved useful in locating the single system orifice (*arrow*), as blue dye consistently swirled from behind the tip of the cystoscope positioned cephalic to the bladder neck.

usually seen on each lateral edge of the trigone, a triangular zone on the floor of the bladder. Pubertal estrogens will stimulate normal squamous metaplasia changes on the trigone. The location, number, and configuration of the ureteral orifices are noted, as abnormal orifices may reflux. The experienced cystoscopist will monitor the quantity of irrigant within the bladder, preventing overdistension and mucosal hemorrhage. If indicated, the same scope can be atraumatically passed thru the hymen into the vagina. To achieve complete visualization, the vaginal introitus must be compressed with gauze sponge to gain distension with irrigant. One midline cervix with os is typically seen with no vaginal mucosal or muscular wall lesions (Fig. 21.13). In general, the female urethra, bladder, and vagina are thoroughly inspected for possible anomalies, which are listed in Table 21.1.

Cystourethroscopy of a male varies from the female procedure mainly by techniques to negotiate the male urethra. In the term, male pediatric patient, the urethra typically can accept a 7.5 Fr or 8 Fr caliber cystourethroscope. Occasionally, the foreskin and the urethral meatus will require dilation in order to admit this. The cystoscope tip is inserted with lubricant. With flow on, the scope is negotiated thru the uniform tubular anterior urethra. At all times, the lumen should be visualized ahead or the scope should be backed until lumen is seen. At the external urethral sphincter, the urethra becomes tighter even with irrigant flow. The mucosal vascular striations begin in this zone, indicating entry into the posterior urethra. At this point, the urethra turns sharply upward. To negotiate this turn, the cystoscopist must lower the penis, so the camera and eyepiece of the scope are below the level of the buttocks. As the scope is advanced, the round raised pink verumontanum is seen on the



Fig. 21.13 Vaginoscopy. With irrigant filling the vagina, the vaginoscopic appearance of a prepubertal cervix and its os is seen (*arrow*)

dorsal midline of the urethral wall. The bladder neck follows the verumontanum and then the bladder is entered. It is cystoscopically identical to the female bladder. The male urethra and bladder are thoroughly inspected for possible anomalies, which are also listed in Table 21.1.

Several other general cystoscopic tips are discussed below:

1. Posterior urethral valves are an obliquely oriented membrane extending from the distal verumontanum and attaching anteriorly to the urethral wall, with a small eccentric aperture. Prior urethral catheterization often alters the form of the valves. In the older child, minivalves can be missed. To improve detection, the bladder should be filled retrograde via the cystoscope. With the irrigant flow shut off, the cystoscope is placed just distal to the external sphincter, and the Credé maneuver is performed. Antegrade flow will further open the valve leaflets. An alternative method is to guide a resectoscope hook blade in the troughs lateral to the verumontanum. Membranous valve leaflets can be identified and cut at 5 o'clock and 7 o'clock with this technique. Several techniques, such as electrocautery, Fogarty balloon, or laser, have been used to ablate/fragment PUV [2, 4, 5], urethral polyps, or urethral strictures. The author's preference is cold-knife incision, followed by temporary catheterization; this technique may result in less tissue damage. In the preterm male infant with obstructive uropathy, cystourethroscopy may be impossible if the urethra is small. To circumvent this problem, some have performed antegrade posterior urethral valve ablation via percutaneous cystotomy access effectively [7]. If this is also ineffective, a Foley catheter, suprapubic catheter, or vesicostomy may be necessary to temporarily divert the obstructed system.

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	Location of			
Sex of child	pathology	Pathology	Cystoscopic findings	Therapy options
Male or female	Anterior urethra	Urethral diverticulum	Dilated cavernous segment of urethra. In males, may have a wide mouth in the penoscrotal region +/- purulent debris or hair. In females, small mouthed with mass effect	Open urethroplasty
Male		Urethral stricture	Mild narrowing to pinpoint narrowing of urethral lumen, short membrane, or long narrowed segment with whitish scarred epithelium	Endoscopic (VIU) via resectoscope or open urethroplasty
Male		Anterior urethral valves	Fenestrated diaphragmatic membrane or mucosal cusp arising from the ventral wall of the bulbar, penoscrotal, or penile urethra	Endoscopic incision or open urethroplasty
Male or female		Urethral duplication	Additional channel with or without communication to the ventral urethra or bladder	Test for communication via contrast or indigo carmine injection
Male		Megalourethra	Marked penile urethral dilation	Open urethroplasty
Male	Posterior urethra	Posterior urethral valves	Valve leaflets at the verumontanum. Bladder neck hypertrophy. Posterior urethral dilation. Bladder trabeculation	Endoscopic transurethral resection of posterior urethral valves (TUR valves) or urinary diversions
Male or female		Ectopic ureteral orifice	Male orifice proximal to the external sphincter on veru or posterior urethra. Female orifice in the bladder neck, urethra, perineum, vagina, or cervix. If functional, can be identified by indigo carmine excretion	If symptomatic, open surgery (ureteral reimplantation, ureteroureteros- tomy, heminephroureterectomy)
Male		Prostatic utricle/vaginal remnant	Frondlike mucosal projections surrounding orifice to utricle/vaginal remnant on the center of the verumontanum	If symptomatic, endoscopic fulguration of orifice or laparo- scopic/open resection of utricle
Male or female	Bladder neck	Ectopic ureteral orifice	Stenotic or gaping orifice at the bladder neck. If functional, can be identified by indigo carmine excretion	If symptomatic, open surgery (ureteral reimplantation, ureteroureteros- tomy, heminephroureterectomy).

Table 21.1 Genitourinary anomalies identified by cystourethroscony

Male or female	Trigone/ ureteral orifices	Ureterocele	Deformed trigone with ballooning bladder mucosa. May extend into bladder neck, urethra, and perineum	Anatomy can be defined by needle retrograde ureteropyelogram. If indicated, transurethral incision of ureterocele for decompression
Male or female		Deformation of the floor	May be feces, megaureter, or ureterocele	Consider fecal disimpaction, retrograde ureterogram, or ureterocele incision
Male or female		Tumors	Papillary or sessile. Often bleeding. Screen in an augmented bladder with gross hematuria, new pain, new incontinence or >4 symptomatic infections/year, abnormal US, or >50 RBC/HPF [6]	Cold cup biopsy for diagnosis. Consider endoscopic resection
Male or female		Ureteral duplication	Distal, medial orifice serves the upper pole moiety. Proximal, lateral orifice serves the lower pole moiety	May require no intervention. Retrograde ureterography may confirm occult or partial duplication. Consider STING for VUR if indicated
Male or female		Diverticula	Hutch diverticula are adjacent to the ureteral orifice (paraureteral). Periureteral diverticula may have the ureteral orifice within the diverticulum	Typically no intervention. May be resected at ureteral reimplantation
Male or female		Patulous ureteral orifice	Ureteral orifice configurations are not predictive of vesicoureteral reflux	If indicated, STING of refluxing ureter
				continued

Table 21.1 (co)	ntinued)			
	Location of			
Sex of child	pathology	Pathology (Cystoscopic findings	Therapy options
Male or female	Bladder	Trabeculation	Irregular small bands of hypertrophied detrusor muscle beneath the bladder mucosa	Evaluate etiology (obstruction, neurogenic cause, functional elimination disorder) and treat
Male or female		Diverticula	May have a small or large mouth to the small or large sac	May require resection if recurrent UTIs unresponsive to medical therapy
Male or female		Calculi	Free-floating stone in the bladder or diverticulum	Open or endoscopic stone ablation techniques
Male or female		Urachal anomalies	Urachal diverticulum may be seen in prune belly syndrome. Urachal sinus may appear infected at the dome	Resect open or laparoscopically
Female	Urogenital sinus		The orifice beneath the clitoris is the urogenital sinus. It biforates at a variable distance into the urethra and vagina	Total urogenital sinus mobilization
Female	Vagina	Duplication/septation	 Hematometrocolpos may cause mass. Septum can be horizontal or vertical and partial or complete. May see two cervices 	Perforate obstructing membranes. Resect septum. May require formal vaginoplasty
		Imperforate hymen	Bulging perineal mass	Incision of hymen. Assess for urinary obstruction
Female		Foreign object	Persistent vaginal discharge prompts vaginoscopy, identifying the object	Removal of foreign object
Female		Cloaca	Single perineal opening with variable configurations leads to the urethra, vagina, and anorectum	Diverting colostomy and possible urinary diversion. Total mobiliza- tion of the cloaca



Fig. 21.14 Bladder diverticulum. Large wide-mouthed trigonal diverticulum viewed from the incompetent deformed bladder neck (Same patient as Fig. 21.12c)

- 2. Bulging masses on the trigone may be the result of a significant fecal impaction elevating the floor of the bladder or, alternatively, a megaureter or large ureterocele. Needle retrograde injection of contrast can delineate ureterocele versus megaureter and can define their extent toward the perineum.
- 3. On the trigone, inspection of the ureteral orifices may reveal a hutch diverticulum. All trigonal diverticula (Fig. 21.14) should be inspected with low volumes of intravesical irrigant to rule out an effacing ureterocele.
- 4. The configuration of the ureteral orifice can be variable even within normal patients but oftentimes takes on a golf-hole configuration when vesicoureteral reflux is present.
- 5. If double J ureteral stenting is planned in the male child, a two-piece scope is crucial. In this case, once wire access is achieved within the ureter, the scope is withdrawn, and the sheath only is back loaded on the wire. Under fluoroscopic guidance, the stent can then be passed over the wire through the cystoscope sheath positioned over the ureteral orifice, preventing wire coiling within the bladder.
- 6. In the intersex patient, close inspection of the verumontanum may show frondular projections around a central orifice, a hallmark sign indicating a prostatic utricle/vaginal remnant (Fig. 21.7). The lengths of the urogenital sinus, vagina, and urethra aid surgical planning. Cystoscopically guided placement of Fogarty balloons into the urethra and vagina of a urogenital sinus can guide surgical repair.

Postoperative Management

Routine postoperative care is indicated, and most cases are performed on an outpatient basis. If purulence was detected, then antibiotics should be administered.

Complications

Possible complications can include bladder or urethral perforation, hemorrhage, pain, urinary retention, ureteral obstruction, infection, and urethral or ureteral trauma with stricture formation or irritative voiding symptoms. Fortunately, these are rare.

Author's Remarks

Cystourethroscopy is an extremely versatile tool for the urologist. It is used to confirm clinical suspicion of disease or to delineate the unusual case. As the technology has advanced, endoscopic tools have permitted minimally invasive therapeutic interventions. In many cases, these procedures negate the need for open reconstructive surgery and have revolutionized the management of these disorders.

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