Congenital anomalies of urinary system in children presented by voiding cystourethrography

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Learning objectives

This poster aims

• To imply the significance of voiding cystourethrography in diagnosis of congenital anomalies of urinary system in children
• To depict most frequent congenital urinary tract anomalies presented by VCUG, followed by description of pathologic findings.
• To provide an insight to the anatomical anomalies of urinary system that may be associated and/or lead to vesicoureteral reflux.
Background

Voiding cystourethrogramy (VCUG, synonym being micturating cystourethrogram or MCUG) refers to a fluoroscopic technique complemented by radiographic imaging performed as an examination of lower urinary tract, by introducing contrast media into the bladder via a catheter.

Congenital anomalies of urinary system affect 10% of general population, and present approximately 30% of all congenital anomalies. Therefore clinical indications for voiding cystourethrogramy include congenital anomalies of urinary tract (American College of Radiology and Society for Paediatric Radiology guidelines). The advantages of VCUG are precise presentation of anatomical features and abnormalities, especially of bladder and urethra (lower urinary tract), followed by high sensitivity in detection of vesicoureteral reflux (VUR). If vesicoureteral reflux exists, a number of anatomical anomalies in upper urinary tract may also be confirmed by this exam. Hindrances of the technique are patient discomfort during examination and absorbed radiation dose. Based on the images acquired by the examination, radiologist can determine the presence and grade of VUR, and describe anomalies associated with and/or leading to it.
Findings and procedure details

PROCEDURE DETAILS

VCUG examination is performed combining fluoroscopic and radiographic imaging that starts with abdominal radiography prior to the aseptic catheterization of the bladder, followed by application of nonionic iodinated contrast media via a catheter. Fluoroscopic screening and radiographic images are obtained within ALARA principle (As Low As Reasonably Achievable) during contrast application, when the bladder is filled (frontal view with full bladder, left and right oblique), during voiding (oblique in male patients) and following micturition.

The volume of contrast material depends on estimated bladder capacity, which can be calculated as follows: for children less than 1 year old V (mL) = Weigh (kg) X 7, for children older than 1 year V (mL) = [Age (years) + 2] X 30

Preparation for the exam includes prophylactic use of antibiotics.

FINDINGS

Congenital anomalies of urinary tract most frequently presented by VCUG in children are abnormalities of the urethra (posterior urethral valves in boys, and less frequently hypospadia), bladder (neurogenic bladder, bladder diverticulum), ureters and kidneys (ureterocoele, congenital megaureter, duplex collecting system, and even renal malposition) and may be associated or lead to VUR.

Hypospadias

Hypospadias is an abnormality of anterior urethral and penile development referring to ectopic location of urethral opening (meatus urethrae), which is, instead of opening at the tip of the glans, positioned on ventral penile surface, anywhere along the line from the penoscrotal region to the glans penis. It occurs in male children with a frequency of 1:300. After catheterization through an ectopic urethral meatus and filling the bladder, during voiding we can assess urethra and confirm hypospadias, whereas in accordance with clinical examination the exact type is determined. Illustration represents proximal penile hypospadias, VCUG finding being a normal posterior urethra and bulbar part of anterior urethra, while there is a lack of opacification of pendulous part of anterior urethra, depicted as a short channel ending in a voiding jet [Fig. 1].

Posterior urethral valves
Posterior urethral valves (PUV) are the most common obstructive anomaly in male children, occurring with an incidence of 1:5000-8000. Voiding cystourethrography is the only procedure that confirms PUV, showing a filling defect followed by reduced caliber of urethra between disproportionately dilated posterior urethra and a narrow anterior urethra, associated with secondary changes- bladder neck hypertrophy, and trabeculation or sacculation of the bladder [Fig. 2-3], with or without vesicoureteral reflux (depending on bladder pressure).

In this case [Fig. 3], a small pouch connected to the prostatic urethra by a narrow channel, the prostatic utricle, is also opacified by contrast media, due to obstruction.

**Megaureter**

Any ureter with a diameter over 8 mm is considered abnormal, and may be described as megaureter. Primary or secondary, it usually belongs to one of the three groups: refluxing, obstructive or non-refluxing non-obstructive megaureter. Refluxing megaureter is always seen in voiding cystourethograms [Fig.3].

**Duplicated ureter**

Duplicated ureter is the most common congenital abnormality of ureter, arising with duplicated collecting system, presented as two pyelocaliceal systems draining a single kidney, continuing into a bifid (ureter fissus, ureter bifidus, partial duplication) or double ureter (with a separate ureteric orifice each). Incidence of this anomaly is 1:150, more frequently unilaterally, with female predominance (3:1). Possible complications are obstruction, ureterocele, or vesicoureteral reflux -in which case it can be confirmed by VCUG [Fig.4].

**Ureterocele**

Ureterocele denotes a congenital dilatation of the distal part of the ureter, protruding into the bladder when filled with urine. It is more common in girls, incidence being 1:5000-12000, and it may be associated with ureteral duplication. There are two types: simple (intravesical) and ectopic. Herein we present a case of simple ureterocele, with its VCUG features, an oval lucency near the trigone on the left [Fig.5].

**Bladder diverticulum**

Bladder diverticulum is a herniation of the bladder mucosa through a defect in a muscle layer. It can be acquired or congenital. Primary congenital bladder diverticulum located at vesicoureteric junction, also called Hutch diverticulum, occurs almost always in boys, in 1.7% of cases, very often associated with VUR, because of altered ureteral orifice. Voiding cystoureterography findings present opacified outpouching arising from
the bladder at the point of vesicoureteric junction, often followed by vesicoureteral reflux [Fig.6].

**Neurogenic bladder**

Neurogenic bladder is a bladder dysfunction caused by neurologic disorders, acquired or congenital (including spinal cord or CNS abnormalities). Congenital neurogenic bladder is typically associated with sacral abnormalities at birth (detrusor sphincter dyssynergia), leading to cystogram appearance of a Christmas tree bladder in VCUG, referring to abnormal elongated shape of the bladder with a trabeculated outline (or sacculations) due to bladder wall hypertrophy [Fig.7].

**Crossed renal ectopia**

Crossed renal ectopia stands for an anomaly where the kidneys are located on the same side of the midline, ectopic kidney usually lower than orthotopic, with separate ureters, and orthotopic ureteral orifices. It occurs with an incidence 1:3000, more often in males. Crossed ectopia with fusion is present in over 90% crossed renal ectopias, and may be diagnosed by VCUG when associated with VUR. The abnormal positioned left kidney (to the right of the midline) attached to the left ureter indicates crossed ectopia, while distortion of the pyelocaliceal system in ectopic kidney suggests fusion anomaly. Presence of contrast media in the parenchyma adjacent to the calices is due to calicotubular reflux (arrows). Calicotubular backflow is usually found in very young children and does not affect grading of reflux (grade IV in this case) [Fig.8].
**Fig. 1:** short penile urethra with urethral meatus (arrowhead) approximately 1 cm from bulbar urethra, without opacification of orthotopic channel in a penile shaft distally.

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**Fig. 2:** Posterior urethral valve. Oblique voiding cystourethrogram shows an elongated dilated posterior urethra (arrowheads) with filling defects (arrow) and a marked decrease of urethral caliber at the level of the defects, followed by transition to a normal-calibre anterior urethra

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Fig. 3: Posterior urethral valves. The secondary changes crucial to the diagnosis - trabeculated bladder (arrowheads), and bladder neck hypertrophy associated with the right-sided vesicoureteral reflux, grade V, demonstrating dilated and tortuous ureter-megaureter (arrow). Opacified prostatic utricle (circle).

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Fig. 4: Unilateral ureteral duplication. Anteroposterior voiding cystourethrogram of a patient with grade IV reflux presenting duplicated pyelocaliceal system on the left

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Fig. 5: Ureterocele - oval lucency, resembling filling defect, near the trigon on the left of the bladder (circle).

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Fig. 6: Bladder diverticulum (Hutch diverticulum)- opacified bladder outpouching at the site of vesicoureteric junction (arrow), associated with high grade VUR.

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Fig. 7: Neurogenic bladder and spina bifida- cystogram appearance resembling christmass tree or pine cone

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Fig. 8: Crossed renal ectopia (arrowheads), with malrotation of orthotopic right kidney, and bilateral VUR, grade IV, complicated by intrarenal reflux (arrows)

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Conclusion

Although VCUG is used for a long time, it still represents the gold standard in diagnostics of VUR, but it is also extremely useful in diagnosis of congenital anomalies of urinary tract, especially of bladder and urethra.

Apart from lower urinary tract anomalies, in presence of VUR even the abnormalities of upper urinary tract are disclosed.

Therefore it can be concluded that voiding cystourethrography is a significant diagnostic procedure, still playing a major role in diagnosis of congenital urinary tract anomalies in children.
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