Large spectrum of complete urinary collecting system duplication exemplified by cases. Pictorial essay.

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Abstract

Urinary collecting system duplication is a congenital anomaly and can associate various types of urinary tract pathologies. Authors will illustrate by cases some of associated anomalies such as ureterocele, extravesical ectopic ureter, vesicoureteral reflux or reflux nephropathy; one case presented an association between posterior urethral valve and bilateral collecting system duplication.

Keywords: collecting system duplication, ureterocele, vesicoureteral reflux, posterior urethral valve.

Duplications of the urinary collecting system (CS) are congenital reno-urinary anomalies; these entities may have a very large spectrum of variants. In a previous study performed in our hospital duplex CS was found in 7.2% patients with congenital urinary tract anomalies; literature has provided similar data [1-3]. An accurate diagnosis can be established antenatally [4]. The authors will present some of the most important imaging aspects found in duplication of the CS variants, especially aspects assessed by ultrasonography (US). Won’t be neglected findings detected by other imaging techniques such as voiding cystourethrography, renal scintigraphy, magnetic resonance or computed tomography [5].

Duplex CS can be complete, when urinary tracts have different distal ureteral implantation or incomplete when the ureters are getting together before distal implantation [6,7]. In incomplete duplications the pathology is represented by uretero-ureteral reflux. Weigert-Meyer law is observed in cases with complete duplex CS: the ectopic ureter arrives from the upper moiety and the distal end may have an intra or an extravesical implantation or can bulging into the bladder to determine an ureterocele formation. The upper moiety associates pathology due to the abnormal ureteral implantation as obstruction and rarely vesico-ureteral reflux (VUR). The management of patients with CS duplication and ureterocele is controversial [8-13]. The ureterocele sometimes prolapsed into the urethra and can determine bladder outlet obstruction with acute urinary retention [12]. In most cases an endoscopic treatment with ureterocele puncture or a conservative management is the first choice therapy in infancy [13]. An extravesical ectopic ureter leads to urinary incontinence in girls; it is very important to assess in this respect all girls with incontinence [14]. The lower moiety, with a normal or almost normal ureteral implantation presents usually a “normal” urinary pathology, irrespectively VUR or ureteropelvic junction obstruction [15-19].

The authors will present imaging features in pediatric patients diagnosed with some variants of complete CS duplications.

Case 1. Boglarka, girl with antenatal detected urinary tract anomaly. At 2 days of age presented acute urinary retention; physical examination of the urethral meatus detected a herniated purpule cystic “mass” (suggesting
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an ectopic ureterocele prolapsed in the urethra). US detected left duplex CS with ureterocele “in tension” (fig 1). The outcome was favorable after cystoscopic ureterocele decompression (fig 2).

**Case 2. Sara,** girl, 5 days of age, antenatal detected urinary anomaly.US revealed bilateral duplex CS with dilatation of the upper moiety CS (fig 3-5).

**Case 3. Zorica,** girl, 9 years of age. Diagnosis: urinary incontinence. US and computed tomography showed bilateral duplex CS with hypoplasplastic right upper moiety and right ectopic extravesical ureter (with the distal end implanted in the vagina), responsible for urinary incontinence (fig 6-8). In girls with incontinence is necessary to assess a possible ectopic extravesical ureter.

**Case 4. Tudor,** antenatal detected urinary anomaly; first postnatal US was performed at the age of 3 days (figure 9-11). Imaging evaluation was completed with voiding cystourethrography after urinary tract infection treatment (fig 12) and magnetic resonance (fig 13). Diagnosis was right duplex CS with important dilatation of the both CS and an ureterocele (ureterocele eversion was detected during voiding). After ureterostomy there was no dilatation of the CS and ureterocele was flattened (fig 14).
Fig 6. Case 3: Longitudinal scan of the right kidney – narrow upper moiety with thin and irregular parenchyma and mild dilatation of the upper CS. Normal US aspect of the lower moiety.

Fig 7. Case 3: Urinary bladder US – longitudinal scan revealed liquid in the vagina with an anechoic pattern.

Fig 8. Case 3: 3D computed tomographic reconstruction of the renourinary system – bilateral duplication of the CS; hypoplastic aspect of the right upper pole, right upper ureter is dilated and tortuous with an extravescical (vaginal) implantation.

Fig 9. Case 4: Longitudinal scan of the right kidney – both right CS present important dilatation, it can be seen narrowing of the right kidney parenchyma.

Fig 10. Case 4: Transverse scan of the right upper pole revealed renal pelvis and proximal ureter dilatation and detritus/fluid level in the upper CS (white arrow). Urine culture showed urinary tract infection. Cortico-medullary differentiation was present.

Fig 11. Case 4: a) Longitudinal scan of the bladder – there are two enlarged right ureters and ureterocele with a thick wall was detected into the bladder (there was no suspicion of increased pressure in the ureterocele); b) Transverse scan of the bladder performed after voiding: mild vesical residue, ureterocele decreased in size and wall is thicker.
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Fig 12. Case 4: Voiding cystourethrography with an anteroposterior incidence – eversion of the ureterocele during voiding (with a pseudodiverticula appearance), normal urethra and absence of the VUR.

Fig 13. Case 4: Magnetic resonance urography (MIP T2). Right CS duplication with important dilatation; tortuous ureters and ureterocele.

Fig 14. Case 4: a) After ureterostomy there was no urinary tract dilatation; renal parenchyma presented a good corticomedullary differentiation; b) Transverse scan of the urinary bladder showed ureterocele collapse.

Fig 15. Case 5: a) Left kidney US, longitudinal scan: CS duplication with dilatation of the lower pelvicalicealsystem, reduced lower moiety parenchyma and a normal US aspect of the upper moiety; b) Longitudinal scan of the left vesicoureteral junction: ureteral dilatation (diameter of 7 mm) and ureteral wall thickening; c) Color Doppler interrogation of the left vesico-ureteral junction revealed retrograde flow from the bladder into the ureter (VUR).
Fig 16. Case 5: Magnetic resonance urography confirms duplex CS with significant distension of the lower pole, convex shape of the calices and tortuous ureter.

Fig 17. Case 5: 99m technetium dimercaptosuccinic acid (DMSA) scintigraphy: significant decrease in radionuclide uptake in the left lower moiety.

Fig 18. Case 6: a) Left kidney longitudinal scan: dilatation of both CS, more obvious on the lower moiety, poor differentiation of the parenchyma; b) Right kidney, longitudinal scan using a high resolution linear transducer. Dilated duplex CS with parenchyma cystic areas of dysplasia.

Fig 19. Case 6: Longitudinal scan of the symphysis pubis (in neonates is incomplete ossified and permits proximal urethra assessment): dilatation of the proximal urethra with thick wall, there is also bladder wall thickening.

Fig 20. Case 6: Voiding cystourethrography, anteroposterior view: dilatation of the proximal urethra; multiple bladder pseudodiverticula; high degree (Vth degree) VUR in both right CS with intrarenal reflux.
Case 5. Rares, 4 months of age, recurrent urinary tract infections. US (fig 15), magnetic resonance urography (fig 16), renal scintigraphy (fig 17) revealed left CS duplication with lower moiety VUR and reflux nephropathy.

Case 6. Stefan, 1 day of age, antenatal detected posterior urethral valve. US (fig 18, fig 19) and voiding cystourethrography (fig 20) confirmed antenatal diagnosis and revealed bilateral CS duplication, cystic dysplastic parenchyma areas and right high degree VUR.

In conclusion, the authors presented some of the most frequent imaging aspects encountered in daily practice in the large variety of CS duplication; US plays a very important role in the assessment of CS duplication variants.

Conflict of interest: none.

References