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Distal atresia of the ureter associated with double excretory system

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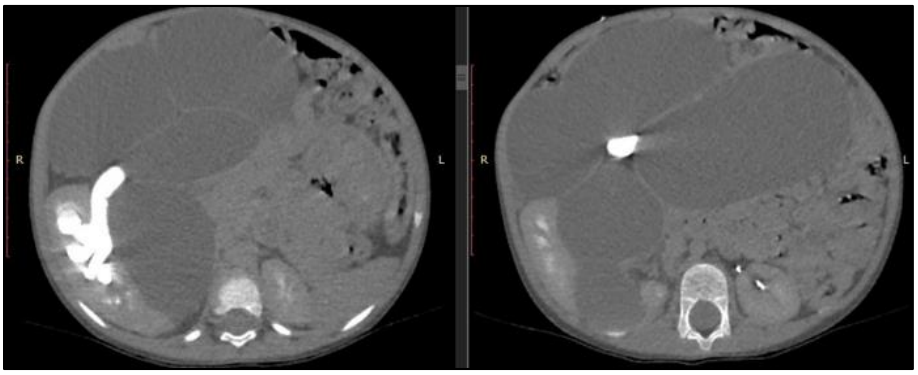
Abstract
Ureteral atresia is rare and usually associated with loss of renal function. Association with other urinary malformation remains exceptional. We report a case of distal ureteral atresia associated with a double excretory system, with preservation of renal function in a 3-year-old boy. Abdominal ultrasound showed voluminous cystic mass. CT-scan demonstrated a malrotated right kidney with double excretory system one of which was atretic. The child had a right partial superior ureter nephrectomy with removal of the atretic ureter. Outcome was good after 6 months follow-up.

Keywords: distal ureteral atresia, double excretory system, renal function, abdominal mass

Introduction
Ureteral atresia is an extremely rare urological malformation. This malformation known as usually associated with a nonfunctional kidney is rarely accompanied by other malformation of the urinary tract [1]. We report a case of distal ureteral atresia associated with a double excretory system and with preservation of renal function in a 3 years-old boy in order to present the specificity of this malformation.

2. Case presentation
A 3-year-old boy was adressed to our department for a big belly. In his antecedents, her mother was not followed-up during pregnancy, no prenatal ultrasound scan was done. The increase in abdominal volume has been observed by her mother since a year and evolve gradually with appearance of episodes of constipation. An episode of unexplored infectious syndrome would have appeared at the age of one year. At the entrance, the child was afebrile (37.2 °C) and the general condition was preserved. Clinical examination showed a big belly with an abdominal perimeter at 65 cm. Percussion revealed diffuse dullness corresponding to a soft abdomino-pelvic mass that was motionless and irregular. There was no spontaneous or palpation pain of the abdomen. Biological examination showed no particular abnormality apart from a mild normocytic anemia (Hb = 10.8g/dl); blood urea (3mmol/l), serum creatinine (32µmol/l) and liver function were normal. Abdominal ultrasound revealed a important tortuous cystic mass. Contrast enhanced CT-scan confirmed the tortuous cystic mass. The right kidney had a double excretory system and the cystic mass corresponded to a monstrously dilated and tortuous ureter (Figure 1), from the upper excretory system, and that ended by a bulky cul de sac behind the bladder and not communicated with the bladder. Opposite, the bladder had a small "diverticulum image" at the lower part of its posterior face (Figure 2). This cul de sac contained declive calcification. Abdomino-pelvic viscera were repressed or compressed. The right kidney was insufficiently rotated. The upper portion of the right kidney supporting the dilated and atretic ureter was thin. Nephrography of the right kidney was preserved but heterogeneous and pale compared to that of the left side; excretion into the lower system was good, whereas in the upper system excretion was delayed and to as much lesser (Figure 3). The ureter of the inferior system was dilated, by compression of the dilated and atretic ureter of the upper excretory system, before arriving in the bladder. Child had a right superior partial ureter nephrectomy with removal of the atretic ureter. No urologic and nephrological abnormality was occured after 6 months of follow-up.



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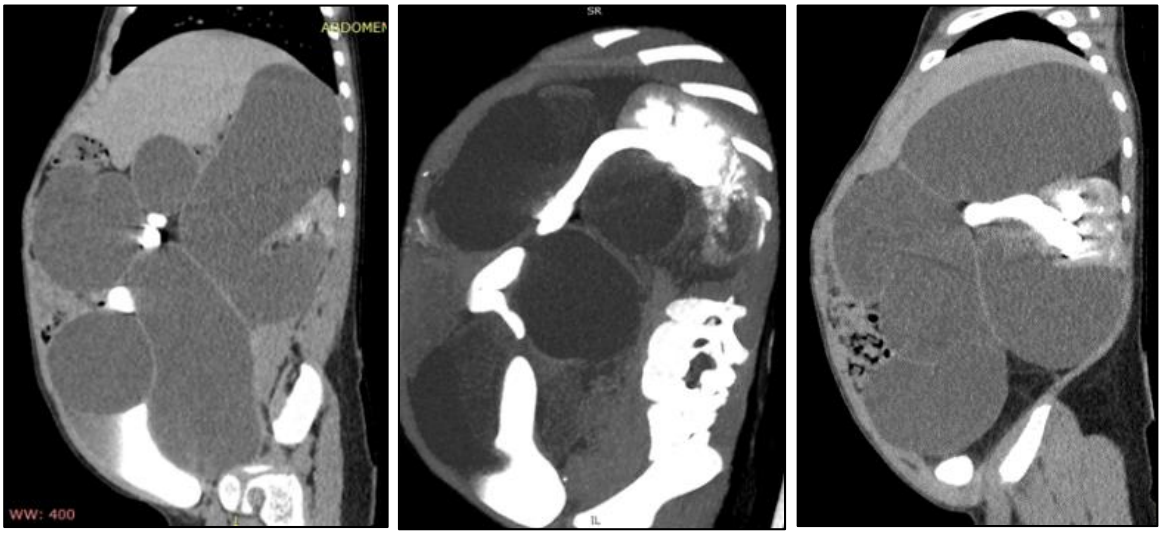
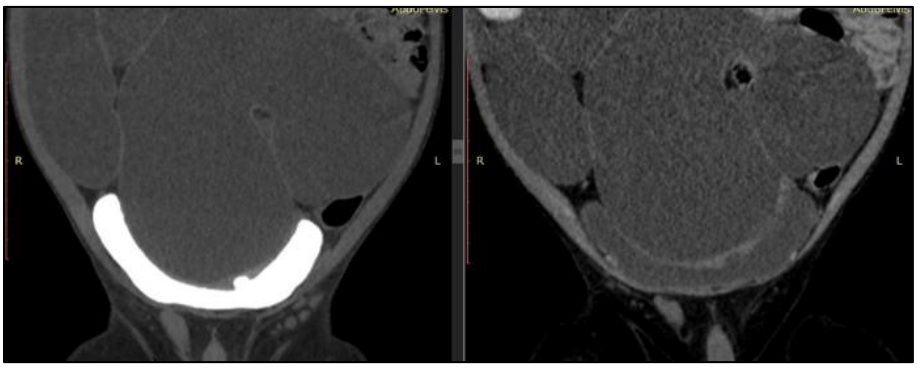
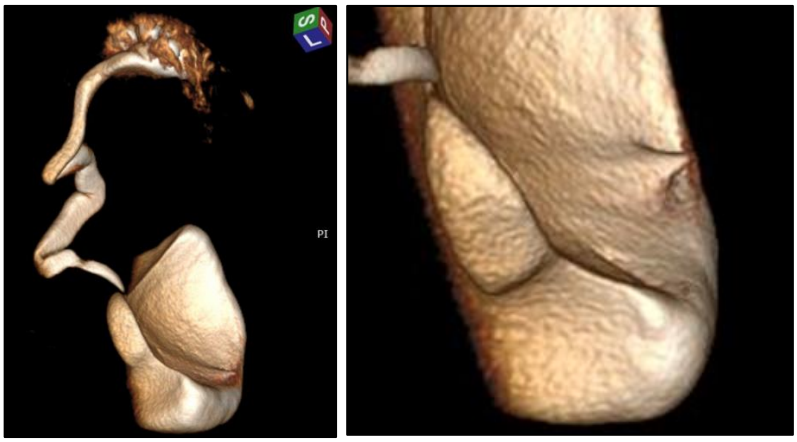


C) **D)** **E)**

Fig 1: CT-scan images of the abdomen, axial (a, b), sagittal reconstruction (c, e) and MIP oblique coronal reconstruction (d) showing a doublé excretory system of the right kidney; the inferior system (arrow) collected by an opacified and dilated pyelo-ureteral system gets into the bladder; the superior system (arrowhead) collected by a monstrously dilated ureter (stars) and terminating with a cul de sac behind the bladder (c). The upper system is barely excreting (dotted arrow) (b, d).



A)



B) **C)**

Fig 2: CT-scan images in coronal reconstruction (a), 3D VR (b, c) showing an ectopic meatus (dotted arrow) at the lower part of the posterior face of the bladder, notching but not communicating with the ureteral cul de sac in opposite. Note the bladder compression and dense image with fluid level (arrows) already visible on the early images before excretion (not shown here) related to calculi.

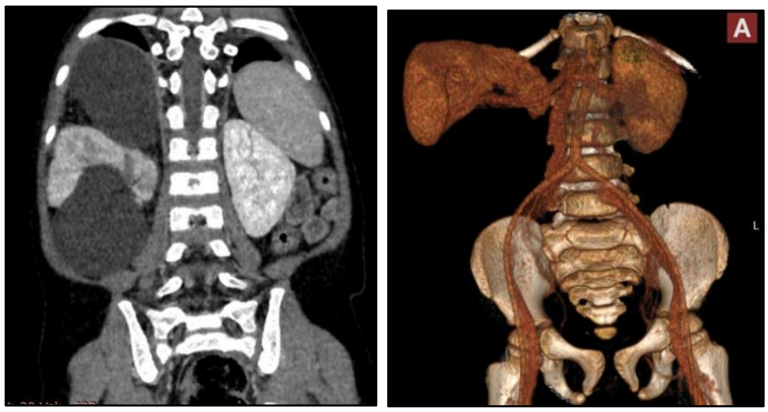


Fig 3: CT-scan images in coronal reconstruction (a) and 3D VR (b) showing a discrete asymmetry of the nephrography, an atrophy of the medial part (superior) of the parenchyma of the right kidney (arrow). Note the insufficient of rotation of the right kidney.

3. Discussion

Atresia of the ureter is one of the rarest malformations. Research in the English and French literature revealed 15 cases reported for the proximal and distal forms; 2 were in females (1 child and 1 adult), 2 unspecified and 11 in males (including one adult); 14 were distal. Distal ureteral atresia is associated with failure of the ureter to communicate with the bladder during embryonic development [2]. Thus, the ureter terminates by a cul de sac, the height of which is a function of the level of ureteral developmental arrest since any ureteral segment can be reached [3]. This lesion is usually unilateral and frequently associated with a non-functional kidney and usually dysplastic [4]. In our case, the involved kidney had only moderate partial atrophy and a slight decrease in nephrography, but with preservation of renal excretion in the lower system. This atrophy is related to the chronicity of urinary stagnation but also to compression of the large ureteral dilation. The bilateral form, never reported, would thus be incompatible with life. However, a unilateral form on single kidney was diagnosed antenatally; the renal function was stabe after cesarean delivery and after ureteroplasty [5]. Ureteral atresia associated with another urological malformation is extremely rare [6, 7]. Association with a double excretory system has never been reported in the English and French literature. The presence of this duplicity would protect at least part of the kidney that excretes into the non-atretic ureter. This involvement is to be differentiated from the "blind ending" ureter, which is an abnormality of ureteral duplication and affects the lower third of the ureter without a hydroureter [9, 10].

Ureteral atresia is pauci-symptomatic and fortuitously revealed by routine antenatal or newborn ultrasound by the discovery of ureteral dilation associated or not with a multi-cystic kidney or later by a gradual increase in abdominal volume that will lead to exploration [1]. This condition is a function of the secretion and excretion ability of the overlying kidney; and determines the earliness or delay of the diagnosis. Ultrasonographic determination of ureteral atresia is difficult because ureteral ectopia or small ureteral diverticle or obstructive mega-ureter may have the same appearance. Intravenous urography may show the ureteral atresia by demonstrating the absence of communication with the bladder and the presence of a cul de sac aspect of the distal ureter if the kidney is functional. The absence of opacification of the excretory cavities makes the analysis difficult and the presence of a large space between the bladder and the rectum on the lateral view could suggest an ureteral atresia; corresponding to the mass effect achieved by the dilated ureter [3]. Noncontrast CT-scan shows the dilation of the excretory cavities which is clearly visible on the multiplanar reconstructions and allows to visualize certain associated

morphological anomalies. Contrast enhanced CT-scan aids in diagnosis especially if the atretic ureter is opacified (in case of fonctional kidney). This technic allowed to demonstrate the double excretory system and the functional aspect of the inferior part; and the atrophic appearance of the upper part supported by the atretic dilated ureter, of the insufficiently rotated kidney. Chronic accumulation of urine in the atretic ureter is the cause of the lengthening and progressive dilation of this obstructed canal realizing an abdominal mass which is a circumstance of discovery in children or the adult [4, 6, 11]. The importance of ureteric dilation is sufficient to slow the digestive transit by compression. No lithiasic formation has been reported to date in these atretic ureters. In our case, spontaneous hyperdense images with fluid level at the bottom of the ureteral cul de sac correspond to calculi. The small "diverticulum image" of the posterior face of the bladder corresponds to a blind ectopic ureteral meatus. The main complications of the distal ureteral atresia are hydronephrosis, urinary tract infection and loss of renal function [1], but also digestive compression; no malignant degeneration has been reported. Although ureteral atresia is known to be associated with a loss of kidney function, 3 cases have been reported with preservation of renal function after treatment [7, 12, 13]. Thus, the therapeutic trend is rather the ureteroplasty than the nephroureterectomy. The preservation of renal function in our case has been largely related to the presence of double excretory system.

4. Conclusion

Distal ureteral atresia is rare. Association with a double excretory system protects or limits the loss of kidney function. CT-scan can be used to make the diagnosis by highlighting the termination of the ureter by a cul de sac behind the bladder and the presence of meatus on the posterior face of the bladder at its opposite, and of course the absence of communication of the ureter lumen with the bladder.

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