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Double Collecting System, A Rare Disease Diagnosis and Management: A Review Article

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1. Abstract

1.1. Introduction: Congenital Anomalies of the Kidney and Urinary Tract (CAKUT) is a group of congenital disorders in the form of kidney malformations and urinary tracts. CAKUT includes renal agenesis, renal hypo / dysplasia, multi cystic kidney dysplasia, duplex renal collecting system (double system), ureter pelvic junction obstruction, mega ureter, posterior urethral valvula (PUV), and vesicoureteral reflux (VUR). The spectrum of congenital renal and urinary tract disorders is very broad, ranging from asymptomatic, mild malformations such as double ureters or minimal ureteral pelvic obstruction to severe, life-threatening pathologies such as bilateral renal agenesis or renal dysplasia. single or as part of a syndrome involving conditions outside the urinary tract. One of CAKUT non syndromic is Double Collecting System (DCS) or Duplication of the Ureter or Duplex Kidney. Aims of the article is to review duplication of the ureter, a rare disease diagnosis and management.

Discussion: Double Collecting System, also known as ureteral duplication, is a condition in which there are two ureters and / or the pielokalicseal system. This disorder is one of the rarest disorders of the urinary tract. This condition can occur completely and incompletely. Incomplete DCS occurs more frequently than complete DCS. This disorder can occur in one or both kidneys. Incomplete duplication is also known as the bifid collectivus system. This condition occurs when one ureteral bud branches before reaching the methanefric blastema. Duplicated ureters then fuse and there is only one uterine orifice. When there are two separate and fused pyelonallicseal systems at the ureter pelvic junction (UPJ), this condition is considered a bifid pelvis. Meanwhile, if there are two separate ureters which then fuse at a point below the UPJ before entering the bladder, this condition is considered a bifid ureter. In contrast to incomplete duplication, complete duplication is a condition where there are two separate ureters that lead to the bladder.

1.3. Conclusion: The Double Collecting System (DCS) is often found incidentally on the investigation of other abnormalities. This disorder is one of the rarest disorders of the urinary tract. Several supporting examinations that can help identify a Double Collecting. Management that have been recommended so far are urethrectomy or hemi nephrectomy pole top kidney.

2. Introduction

Congenital Anomalies of the Kidney and Urinary Tract (CAKUT) is a group of congenital disorders in the form of kidney malformations and urinary tracts. CAKUT includes renal agenesis, renal hypo / dysplasia, multi cystic kidney dysplasia, duplex renal collecting system (double system), ureter pelvic junction obstruction, mega ureter, posterior urethral valvula (PUV), and vesicoureteral reflux (VUR) [1].

Congenital anomalies of the kidneys and urinary tract (CAKUT)

are found in 3-6 out of 1000 newborns, or according to some statistics, CAKUT occurs in 0.5% of all pregnancies. Congenital Anomalies of the Kidney and Urinary Tract (CAKUT) can occur as a result of a complex hereditary pattern, including genetic factors and underlying environmental factors [2].

However, in most cases, genetic defects cannot be identified, so the underlying mechanism is difficult to determine by The process of formation of the kidney and urinary tract depends on the interaction between the ureteric bud and the surrounding metanephric mesoderm. This process starts from the ureteral bud that originates from the Wolffian duct to the metanephric mesoderm, where the ureteric bud affected by the metanephric mesenchyme then differentiates into the ureter, pelvis, and collecting ducts, while the metanefric mesoderm induced by the ureteric bud differentiates into proximal and distal tubules, and glomerulus [1, 3].

The spectrum of congenital renal and urinary tract disorders is very broad, ranging from asymptomatic, mild malformations such as double ureters or minimal ureteral pelvic obstruction to severe, life-threatening pathologies such as bilateral renal agenesis or renal dysplasia. single or as part of a syndrome involving conditions outside the urinary tract4,5. One of CAKUT non syndromic is Double Collecting System (DCS) or Duplication of the Ureter or Duplex Kidney [5-9]. Aims of the article is to review duplication of the ureter, a rare disease diagnosis and management.

3. Discussion

3.1. Double Collecting System

Double Collecting System, also known as ureteral duplication, is a condition in which there are two ureters and / or the pielokalicseal system. This disorder is one of the rarest disorders of the urinary tract. This condition can occur completely and incompletely. Incomplete DCS occurs more frequently than complete DCS. This disorder can occur in one or both kidneys [5, 11, 12].

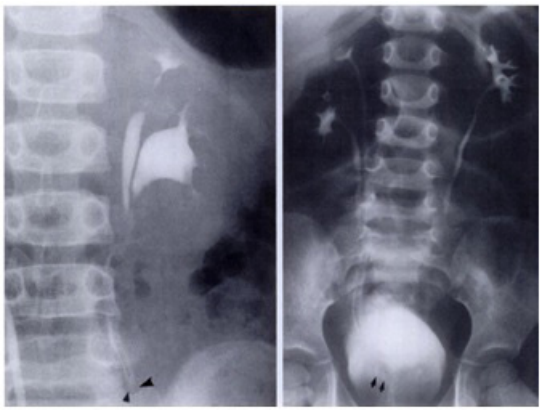

Figure 1: a) Incomplete DCS, excretory urogram showing the ureter which is attached just above the sacrum (arrow); b) Complete DCS, excretory urogram showing less calix image on top pole and more on bottom pole. The ureter from the upper pole enters below and medially from the lower pole ureter and enters separately in the bladder [12].

Incomplete duplication is also known as the bifid collectivus system. This condition occurs when one ureteral bud branches before reaching the methanefric blastema. Duplicated ureters then fuse and there is only one uterine orifice [5, 12]. When there are two separate and fused pyelonallicseal systems at the ureter pelvic junction (UPJ), this condition is considered a bifid pelvis. Meanwhile, if there are two separate ureters which then fuse at a point below the UPJ before entering the bladder, this condition is considered a bifid ureter. In contrast to incomplete duplication, complete duplication is a condition where there are two separate ureters that lead to the bladder.

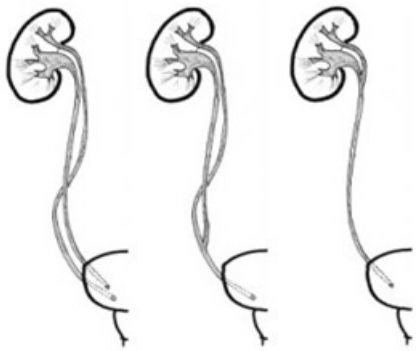

Figure 2: Double Collecting System. a) Duplication of the complete collectivus system, characterized by the presence of two separate ureters leading to the bladder; b) Duplication of the bifid ureter type incomplete collectivus system, characterized by the presence of two ureters that separate and fuse under the UPJ before entering the bladder; and c) Duplication of the bifid pelvic type incomplete collectivus system, characterized by the presence of two ureters separated and fused above the UPJ [5, 11].

This condition can occur due to interference during nephrogenesis where there is excessive branching of the ureteric buds. It can also occur when the embryo has an additional ectopic ureteric bud originating from the Wolffian duct that lies above the normal bud. Worldwide, the Double Collecting System occurs with a prevalence of 1: 150 (0.67%) [11]. incomplete DCS is more common than complete DCS which is estimated to occur in 1 in 500 people [12].

3.2. Clinical Features

Signs and symptoms in the Double Collecting System case include: [11, 12]

a. Recurrent UTI

b. Recurrent fever for no apparent reason

c. Abdominal mass

d. Enuresis

e. Urinary incontinence

Complications of this condition including in incomplete DCS, it often occurs: [12]

a. Ureterourethral reflux

b. UPJ (Ureteropelvic Junction) obstruction

Often it occurs at the bottom of the system, hitting more boys than girls

c. Vesicoureteric reflux (VUR)

Often occurs at the bottom pole of the system, is the most common complication found in complete DCS, and is more common in women.

d. Ectopic ureterocele

Often occurs at the pole top of the system. It is a disorder in which there is a dilatation of the intravesical component which causes the formation of a "cele" in the ureteral orifice, which often results from dilatation of the calix and the ureter above it. This abnormality is easily identified by means of a cystouretrographic examination.

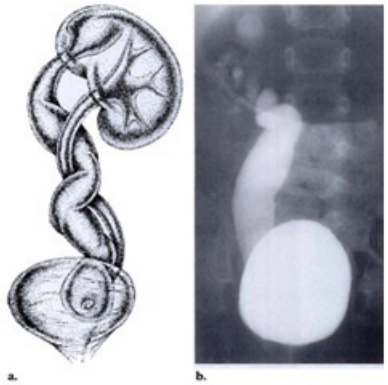

Figure 3: Ectopic ureterocele. a) The image shows the dilatation and twisting of the upper pole ureter which intersects the lower pole ureter and produces an image of extrinsic indentation in the ureter's orifice; b) Voiding systouretrogram shows the contrast material refluxes to the upper pole of the ureter, indicating the large caliber of the ureter which explains why the lower pole ureter appears to be twisted [12].

e. Ectopic ureter insertion

Often occurs at the top pole of the system, often causing enuresis in women

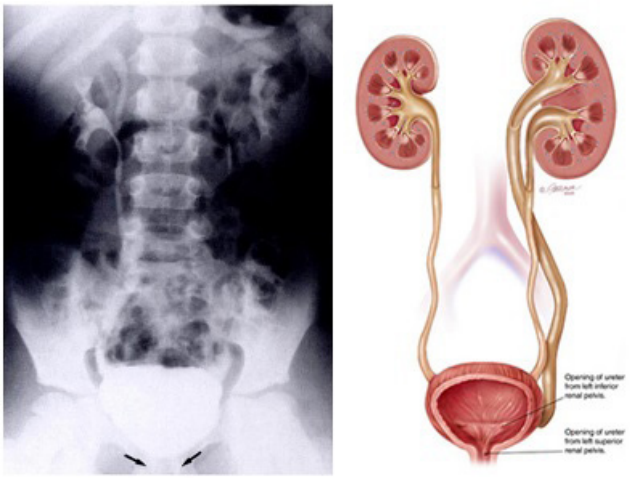

Figure 4: Ectopic ureter insertion. a) Urogram shows faint opacification from the upper pole, which drains through the dilated ureter. Visible contrast material coloring the vagina (arrow) indicates the ureter empties into the vagina. At the time of surgery, both ureters were found to drain into the vagina. b) Schematic image of the ectopic ureter on complete DCS [12].

3.3. Supporting Investigation

The Double Collecting System (DCS) is often found incidentally on the investigation of other abnormalities. Several supporting examinations that can help identify a Double Collecting System include: [4, 11]

a. Ultrasonography (USG) of the abdomen

If not accompanied by other malformations, renal ultrasound examination is often not found abnormal findings. A DCS can be detected on ultrasound if there are two renal pelvis separated by the renal parenchyma. Identification of DCS via ultrasound can be easier in cases of DCS with ureterocele or hydronephrosis, which often occurs in the pole top system of the kidney.

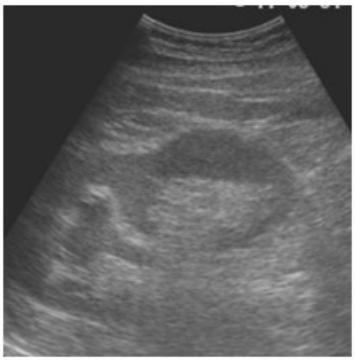

Figure 5: Renal ultrasound in case of DCS with hydronephrosis of the upper pole of the kidney due to ureteral stones [11].

This examination is carried out using a plain photo modality and contrast fluid which is then evaluated for a while to determine the function and anatomy of the kidneys and urinary tract. If the system at the pole top has poor kidney function, the urography examination may not be able to identify the upper portion of the DCS, because identification requires contrast-stained portions or tissue.

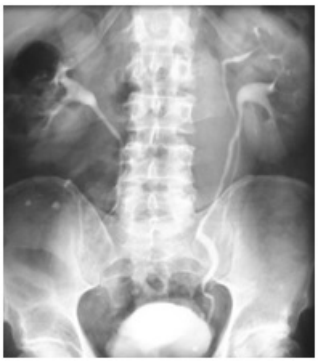

Figure 6: IVU examination in cases of DCS. The results of the examination showed a left DCS without hydro nephrosis [11].

c. System program

Cystouretrogram voids are indicated in cases of recurrent urinary tract infections or dilatation of the pyelonallicseal system. Ureterocele can be detected in the initial filling phase. Reflux of urine into the lower renal system can be detected by 50%.

d. Cystoscopy

Cystoscopy and retrograde pyelonography may be required depending on the complaint.

e. Renal scintigraphy

Renal scintigraphy allows us to distinguish the renal function originating from the upper and lower parts of the double collecting system, making it easier to determine subsequent therapy.

f. CT scan

CT scans of the abdomen with and without contrast can be performed to more clearly identify the anatomy of the kidneys and urinary tract and can more accurately identify the anatomical location based on the slices made.

If it causes symptoms or disorders, management can be carried out in accordance with the symptoms and accompanying malformations. The measures that have been recommended so far are urethrectomy or hemi nephrectomy pole top kidney [4, 11].

4. Conclusion

The Double Collecting System (DCS) is often found incidentally on the investigation of other abnormalities. This disorder is one of the rarest disorders of the urinary tract. Several supporting examinations that can help identify a Double Collecting. Management that have been recommended so far are urethrectomy or hemi nephrectomy pole top kidney.

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