Urology

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Congenital Anomalies of The Upper Urinary Tract

Congenital anomalies of the upper urinary tract comprise a group of abnormalities, ranging from complete absence to aberrant location, orientation, and shape of the kidney as well as aberrations of the collecting system and blood supply. These diverse entities are among the most common malformations in newborns.

These anomalies are usually asymptomatic. They may be discovered incidentally or because of complications such as infection or stone formation.

Renal Anomalies:

1. Renal Agenesis (Anomalies of Number): Congenital absence of the kidney. When bilateral, it is incompatible with life and causes early neonatal death. Most commonly it is unilateral and the other kidney shows compensatory hypertrophy.
2. Renal Ectopia (Anomalies of Position): The kidney is found at an abnormal position. It may be found lower down in the pelvis (Pelvic kidney) or it may even cross to the contralateral side (crossed ectopia) and sometimes fuses with the other kidney (crossed fused ectopia). As a rule, the ureter of the ectopic kidney drains to the bladder at its normal position. Ectopic kidney may present a diagnostic problem and may be missed with other conditions such as acute appendicitis.
3. Horseshoe Kidney: A pair of ectopic kidneys fused together at lower poles in front of the vertebral column.

A horseshoe kidney is vulnerable for diseases, possibly due to angulation of the ureter causing urinary stasis and hence increased incidence of infection and stone formation. It can be diagnosed radiologically.

Division of the central isthmus may be needed for operations on the abdominal aorta or vertebral column.

4. Malrotated Kidney (Anomalies of Rotation): The kidney lies in its normal position but not in its normal plane. This may also be liable for stasis.

b. Multicystic Kidney: One kidney is replaced by numerous cysts. It is more common than PKD. Management is by excision of the affected kidney (Nephrectomy)

c. Simple renal cyst: Usually asymptomatic, discovered incidentally. It can present with pain due to superadded infection or bleeding. Renal cysts are diagnosed by ultrasound and CT Scan. They rarely need treatment, usually in form of percutaneous aspiration or surgical deroofing.

   Simple renal cyst should be differentiated from hydatid disease of the kidney which is common in sheep-bearing areas.

6. Aberrant Renal Vessels (Anomalies of Vasculature): Renal arteries are functional end arteries therefore cut of an artery will result in ischemia and infarction to the segment supplied by that artery. Multiple renal arteries are more common on the left side. Aberrant renal vessels may present on the ureteropelvic junction causing hydronephrosis.

Congenital Anomalies of The renal Pelvis and Ureters:

Duplication:

Duplication of the collecting system and ureters is the most common urologic anomaly. Duplication of the renal pelvis (bifid pelvis) is common and each part drains certain calyces.

Ureteric duplication is less common. It may be partial, with a common opening in the bladder or it may be complete with two separate openings each draining its own calyces. As a rule, the ureter draining the upper pelvis opens distally and medially.

A duplex renal system is more likely to get infection, pain and calculus formation. However, most are asymptomatic and discovered incidentally. In cases of complete duplication, ureteric openings may be abnormal. The ureter draining the lower moiety may be refluxing and that draining the upper moiety may end in a ureterocele and / or be obstructing. In females, an ectopic ureter may open in the urethra distal to the sphincter or open in the vagina. This usually presents with continuous incontinence. The history is highly suggestive and a urography is

5. Anomalies of Volume and Structure (Parenchymal Anomalies):

    a. Polycystic Kidney Disease: A hereditary condition in which BOTH kidneys are replaced by a large number of cysts that displace the normal renal tissue and result in impairment of normal renal function. The condition may be associated with other organ cystic anomalies such as the liver and pancreas and may be associated with intracranial vascular aneurysm (Berry aneurysm).

    Polycystic kidney disease is classified into two types:

    1. Adult type: Autosomal dominant inheritance that manifest usually in the third or fourth decade of life. It is slightly more common in females and presents with the following features:

        a. An irregular bilateral flank mass
        b. Loin pain
        c. Hematuria
        d. Hypertension
        e. Recurrent urinary infection
        f. Renal impairment (Uremia)

        The condition is usually diagnosed by imaging which reveal multiple variable size cysts occupying the whole kidneys causing stretching of the collecting system.

        Management of the condition includes management of the complications and of the renal impairment. Dietary restriction may help to delay renal deterioration. Surgery may be needed for large painful cysts or for stones. The definitive treatment of the disease is by renal transplantation.

    2. Infantile type: Autosomal recessive presents early in life with progressive deterioration of renal function. It may be associated with obstructed labour and stillbirth. It is usually fatal in few days.

diagnostic. A girl or a woman who voids normally but dribbles urine continuously is probably having an ectopic orifice.

Treatment: Asymptomatic duplication needs no treatment. An atrophic moiety of the kidney is best treated by excision (Partial nephrectomy or heminephrectomy).

An ectopic or a refluxing ureter needs reimplantation into the bladder.

Congenital Megaureter:

It is an uncommon condition in which dilatation occurs in the lower ureter. The condition may be bilateral and may increase the likelihood of infection. Spontaneous improvement usually occurs. However, it may require reimplantation.

Post-caval Ureter (Retrocaval Ureter):

In this condition the right ureter passes behind the inferior vena cava instead of passing nearby it. If this causes obstructive symptoms, the ureter can be divided and re-anastomosed.

Ureterocele:

This is a cystic dilatation of the terminal (intramural) part of the ureter, possible due to atresia of the ureteric orifice. It is usually asymptomatic and diagnosed by urography or cystoscope. The condition is more common in females and may obstruct the urine outflow when it prolapses through the bladder neck.

Treatment should be avoided unless it causes infection or stone formation. In complicated cases endoscopic incision is effective. An obstructing ureterocele that causes renal damage may necessate nephrectomy.