Developmental defects of the kidneys

Renal dysplasia and agenesis

- A range of serious malformations that require dialysis and transplantation in the first years of life.
- E.g. ``Multicystic dysplastic kidney *where, due to failure of nephron development and failure of branching of the ureteral bud, the collecting ducts have not developed at all. The numerous drainage canals are thus surrounded only by undifferentiated cells.*
- Similar causes can lead to kidney involution and agenesis.

Congenital kidney defects

Congenital polycystic kidney disease

In this disease, multiple cysts are formed, which can be congenital as autosomal recessive disorders, but also autosomal dominant disorders.

Autosomal recessive polycystic kidney disease

It affects 1:5000 newborns, with cysts arising gradually from the collecting and draining ducts. The kidneys thus gain volume and their subsequent failure occurs in childhood. At the same time, cysts are also formed in the liver, which is affected by fibrosis and portal hypertension. The cause is a mutation of the geneu, which codes for the membrane protein ``polyductin*.*

Autosomal dominant polycystic kidney disease

Although this disorder is more common in newborns (1:500–1:1000), it is also less progressive. Cysts here arise from all segments of the nephron and lead to kidney failure only in adulthood. The cause is a disorder in the genes that code for *polycystin 1 and 2* and the result of their mutation is a disorder in the composition of the extracellular matrix and at the same time a change in the resorptive function of the epithelium to a secretory function, which leads to the formation of cysts. Cysts are also formed in other organs (liver, pancreas, spleen). The most serious complication, however, is the formation of an aneurysmt on the blood vessels of the brain, as their subsequent rupture leads to severe subarachnoid hemorrhage.

Cleft ureteral bud

The result of this malformation is the doubling of the ureter, the split can be partial or complete and the metanephros can be divided into 2 parts, each with its own ureter. More often, however, both parts have some lobes in common due to the uneven distribution of drainage canals. Rarely, there may be a situation where one ureter opens into the bladder and the other ectopically opens into the vagina, urethra, or vestibule.

Abnormal kidney disorder

Ren pelvis congenitus (dystopian kidney)

The rise of the kidney between the umbilical arteries did not take place on the one hand - the kidney lies in the pelvis near the common iliac artery.

Horseshoe Kidney

The kidneys have been pressed together during their ascent so that their lower poles fuse. It usually lies at the level of the lower lumbar vertebrae (the inferior mesenteric artery prevents further descent). The frequency of occurrence is 1:600.

Accessory aa. renales

This abnormality is common, the resulting arteries originate from persistent embryonic vessels that formed during the ascent of the kidney.

References

Related Articles

Development of the genitourinary system

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ISBN 80-85866-94-3 .

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