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## CONGENITAL ANOMALIES OF THE KIDNEYS AND URETERS IN CHILDREN.

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**Abstract:** From this article we describe the most severe kidney lesions in children, which lead to end stage renal failure and subsequently require transplantation, develop against the background of congenital malformations of the kidneys and urinary tract. Timely diagnosis and correction of these conditions are necessary to reduce the incidence of adverse outcomes and disability in children.

**Keywords:** congenital anomalies, kidneys, urinary tract, newborn, cystatin C, various defects, renal damage, horseshoe-shaped kidney, hydronephrosis.

Despite the progress made in the diagnosis of congenital malformations of the kidneys and urinary tract, there are still controversies in assessing the degree of implementation of antenatal signs of violations of the structure of the urinary system, as well as the clinical significance of markers of renal damage in assessing the state of the urinary system after the birth of a child.

Kidney formation can be accompanied by various defects. Sometimes the location of the kidneys (dystopia), their orientation (incomplete rotation of the kidney), they can grow together (horseshoe-shaped kidney), one kidney may be completely absent (renal agenesis), the presence of additional renal arteries is sometimes disturbed. When both kidneys are missing, the child does not survive (Potter's syndrome). Kidney tissue may also develop abnormally (aplasia, hypoplasia, or dysplasia of the kidney). For example, sometimes the kidney contains many cysts - polycystic kidney disease is diagnosed.

Possible anomalies of the ureters include: the presence of additional ureters, their abnormal location, narrowing (narrowing or fusion of the ureter occurs in 0.6% of children) or expansion. Urine from the bladder can back up into the abnormal ureters, making it more likely to develop an infection in the kidneys (pyelonephritis). Narrowing of the ureter makes it difficult for urine to pass from the kidney to the bladder, causing the kidney to enlarge (hydronephrosis) and damage it.

Polycystic kidney disease may not manifest itself for a long time, however, cysts compress the renal parenchyma, and its blood circulation is disturbed. Polycystic disease begins to manifest after 30 years: there is polyuria, hypostenuria, hypertension. On palpation, enlarged, dense, tuberous kidneys are found, and simultaneous cystic damage to the liver is possible. A contrast x-ray examination (pyelography) reveals a typical picture of elongated pelvises and cups in the form of "spider legs".

Renal agenesis is the congenital absence of one or both kidneys (arenia). Such newborns have folded skin, a puffy face, low-lying auricles, a wide and flat nose, and protruding frontal tubercles. It is combined with congenital malformations of other organs. Children are not viable. Renal agenesis is rare.

Renal hypoplasia is a congenital decrease in the mass and volume of the kidneys. It can be one- or two-sided.

Accessory renal arteries are more common on the left, they go around and compress the ureter, which leads to hydronephrosis and renal hypertension.

Frequent malformations of the urinary tract - doubling of the pelvis and ureters; agenesia, atresia, stenosis of the ureters, ectopia of their mouths.

Ureteral stenosis can be unilateral or bilateral. Bilateral atresia is a fatal defect, accompanied by hydronephrosis or kidney dysplasia. The ureter at the same time comes to an end blindly and is sharply expanded above an atresia. It may look like a fibrous band.

The valves of the ureter are most often located in the upper third and are folds of the mucosa with muscle fibers. They are detected in childhood, since infection of the kidney (pyelonephritis), hydronephrosis easily occurs, and stones are formed.

A diverticulum of the ureter is a protrusion of its wall. It can also be single or double sided. Complicated by the formation of stones, inflammation, rupture of the wall. A tumor may develop in the diverticulum.

Dilatation of the ureter - the expansion of the ureter with atrophy of the wall. The normal diameter of the ureter in adults is 1 cm, in newborns it is 0.6 cm. Dilation is most often the result of obstruction and is located above the site of narrowing, but primary dilation without obstruction is also possible.

Hydroureter - dilation and dropsy of the ureter due to obstruction. May be combined with hydronephrosis or alone.

Megaloureter - expansion and lengthening of the ureter. It can be primary and secondary. Occurs with vesicoureteral reflux or obstruction at various levels, as a result of metabolic, toxic, inflammatory and postoperative disorders.

Increased anthropogenic impact on the environment, deterioration of women's reproductive health, an increase in the number of infections, including intrauterine and perinatal infections, pregnant women taking various drugs that affect the development of the fetus (for example, aminoglycosides, cyclosporine A, prostaglandins, dexamethasone, furosemide, cyclophosphamide, etc. ) leads to an increase in the frequency of congenital malformations. The availability of ultrasound (ultrasound) and its introduction as a screening method for examining pregnant women has increased the frequency of antenatal detected fetal anomalies. In the population, congenital anomalies in the development of various organs and systems in newborns are recorded at a frequency of 50-60 per 1000 births. Among the variety of congenital malformations, anomalies of the kidneys and urinary tract are leading and account for up to 20-50% of all detected fetal malformations. Usually they are detected on a screening ultrasound performed on decreed terms (18-20 weeks of pregnancy). Every year in Russia, about 1,000 children under 18 are recognized as disabled due to diseases of the genitourinary system. A significant contribution to the formation of disability is made by congenital anomalies of the kidneys and urinary tract, which, in particular, are the main cause of renal failure in pediatrics.

The problem of the correct choice of diagnostic and therapeutic tactics for managing newborns with malformations of the kidneys and urinary tract (English congenital Anomalies of the Kidney and Urinary Tract, CAKUT) has remained

relevant for many decades for a number of reasons. First, the frequency of occurrence and detection of these diseases is high and continues to increase. Secondly, until now, the causes and risk factors for their development are not completely clear. Thirdly, the factors that determine the choice between conservative and surgical management of children with malformations of the kidneys and urinary tract, as a rule, are assessed already in the presence of complications. Abnormally developed kidneys have their own characteristics of blood circulation, innervation, changes in urine outflow, they are characterized by a decrease in local immunity, which can lead to the addition of a secondary infection, which also affects the work of abnormally formed kidneys. Newborns with malformations of the kidneys and urinary tract also need to create new organizational models for the provision of highly qualified medical care, including, among other things, the possibility of their transportation to surgical departments of the appropriate level. With megaloureter, the ureter is significantly elongated, expanded (up to 2 cm in diameter or more), it is tortuous, and its wall is thickened. All this is revealed by contrast radiography. Dysplasia of the ureter is a congenital disorder of the structure of the muscular wall of the ureter, in which there is a sharp expansion of the ureter.

Ureter dysplasia has an unfavorable prognosis. Surgical treatment is ineffective. Hypoplasia of the ureter - congenital underdevelopment of the ureter - complete or incomplete. Ureterocele (intravesical cyst of the ureter) - a protrusion in the bladder wall of the ureter. More common in women. It can be one- or two-sided. Most often manifested by vesicourethral reflux and pyuria (leukocytes in the urine). The most common complications are hydronephrosis, pyelonephritis, and stone formation.

Kidney dysplasia is hypoplasia with the simultaneous presence of embryonic tissues in the kidneys. With bilateral severe hypoplasia and kidney dysplasia, children are not viable.

Milder developmental anomalies - fusion of the kidneys (horseshoe-shaped kidney) and kidney dystopia are not clinically manifested and are detected only with instrumental research methods or with the development of complications.

I know that dystopia - congenital displacement of the kidney (usually down), can be combined with an abnormal ureteral discharge, the presence of additional renal arteries. Kidney dystopia also leads to hydronephrosis, stones are easily formed in it, and renal hypertension develops. Malformations occur when the fetus is exposed to teratogenic factors during the formation of one or another section of the urinary tract (4-8 weeks of embryogenesis. Many malformations are hereditary or familial. In addition, they are often found in chromosomal syndromes. The reason for the obliteration of the ureter is the absence of sewerage (the appearance of a canal) of the ureteral cord. This can cause various teratogenic factors that affect the fetus during the formation of the ureters. There is a family predisposition to ureteral obstruction. A polygenic mode of inheritance is assumed. In addition, there is a gender-linked anomaly: boys are twice as likely to be affected. Ureteric strictures may coexist with strictures in the gastrointestinal tract, suggesting that their cause is identical. Often this anomaly is combined with dysplasia of the corresponding kidney. Kidney dystopia is detected by X-ray examination. It is distinguished from

the prolapse of the kidney by a short ureter (when prolapsed, the ureter is long and curved).

Clinical outcomes in patients with antenatal hydronephrosis depend on the degree of dilatation of the pelvicalyceal system.

The severity of hydronephrosis is assessed by measuring the anterior-posterior size of the renal pelvis (the maximum diameter of the renal pelvis in the transverse plane). The size of the renal pelvis during normal development also increases with the growth of the fetus, so the literature suggests its threshold values depending on the gestational age. So, in the second trimester of pregnancy, the initial degree of expansion is considered to be the size of the pelvis from 4.0 to 7.0 mm, moderate - from 7.0 to 10.0 and pronounced - more than 10.0 mm. In the III trimester of pregnancy, a size from 7.0 to 9.0 mm is considered only an initial degree, from 9.0 to 15.0 mm - moderate, and more than 15.0 mm - a pronounced degree of expansion of the pelvis.

However, the method of measuring the anterior-posterior size of the renal pelvis has limitations, since it does not take into account the increase in calyces and thinning of the renal parenchyma. There is currently no consensus as to what size of the renal pelvis will require intensive follow-up or treatment. However, a number of authors point out that an anteroposterior size of less than 4.0-5.0 mm at follow-up requires only conservative tactics, while an increase in the anterior-posterior size of the pelvis > 15.0 mm is strongly associated with an increased risk of developing urinary tract infections and the need for surgical treatment. To clarify the nature of the pathology and subsequent prognosis, it is also important to assess the presence of a megaureter and / or an enlarged bladder. The presence of ureterohydronephrosis without an enlarged bladder suggests possible obstruction of the vesicoureteral junction. Conversely, ureterohydronephrosis (particularly bilateral) with an enlarged bladder indicates an obstruction at the level of the urethra.

The subsequent management of the patient is fundamentally influenced by the period of formation of the pathology, as well as whether the hydronephrosis is bilateral and whether the fetus has oligohydramnios. The latter suggests the presence of a serious developmental anomaly, which with a high probability may require urgent intervention after birth, since in the third trimester most of the amniotic fluid consists of fetal urine.

Thanks to new technologies for ultrasound, it became possible not only to state the presence of an anomaly of development, but to describe the changes in more detail, which can be a key point in determining the tactics of managing such pregnancies. At the moment, one of the promising methods for a detailed assessment of the features of congenital malformations of the kidneys and urinary system of the fetus is three-dimensional echography. This technology allows you to automatically determine the contours and calculate the volume of structures filled with water, and the volume of the parenchyma (kidney), makes it possible to calculate their absolute dimensions, average volume and diameter, which can be a promising marker of the state of the urinary system .

For intrauterine assessment of kidney function, the definition of renal blood flow has been proposed. In obstruction of the urinary tract, developing atrophy of the parenchyma is manifested by a violation of vascularization, which is visually assessed by power Doppler mapping. The parenchyma of such kidneys has a depleted blood flow, a vascular pattern that cannot be traced to the capsule, expanded elements of the collecting system that push the segmental and interlobar branches apart. Pulsed wave Doppler allows assessing renal hemodynamics, and using three-dimensional Doppler angiography, one can clearly assess the structure of the kidney and the distribution of blood vessels in it, the state of blood flow.

Various markers of amniotic fluid, fetal urine, and plasma are used to assess the condition of the fetal kidneys. The main metabolites that attract attention in the urine of the fetus are osmolarity, sodium, potassium, calcium, glucose,  $\beta$ 2-microglobulin. An unfavorable prognosis at a gestation period of less than 20 weeks was noted with an increase in sodium more than 100 meq/l, chlorine - more than 90 meq/l, osmolarity - more than 210 mosm/l.

Postnatal assessment of the degree of dilatation of the renal collector system

After birth, all children with antenatally detected hydronephrosis at the 1st week of life are shown ultrasound of the kidneys and urinary tract. The proposed SFU scale for assessing hydronephrotic changes allows for a comprehensive assessment of the severity of kidney damage, taking into account the expansion of the pelvis, deformation of the calyces, the degree of thinning of the parenchyma, and to select the appropriate therapeutic tactics.

In accordance with the proposed scale, the following degrees are distinguished:

- HN 0 norm, absence or slight visualization of the collecting system of the kidney;
  - HN I the pelvis is visualized, but its size is less than 5-7 mm;
- HN II several groups of enlarged calyces are visualized with unchanged fornic structure and expansion of the pelvis from 5 to 10 mm;
- HN III all groups of enlarged cups are visualized with smoothness of the contours of the formic apparatus and expansion of the pelvis by more than 10 mm, but without disturbing the structure and thickness of the parenchyma;
  - HN IV visualized expansion of all cups, thinning of the parenchyma;
  - HN V almost complete absence of parenchyma.

Dynamic observation is indicated for patients with I-II degree of dilatation, as well as with III-IV degree, provided that more than 40% of kidney function is preserved and there are no complications. Dilatation of the III-V degree requires additional examinations and the decision on surgical treatment suggested using the hydronephrotic index (HI) as an alternative method for assessing the degree of expansion of the renal collector system - the ratio of the area of the renal parenchyma to the area of the entire kidney, multiplied by 100% showed the presence of a correlation between the GI and the SFU scale and its greater sensitivity in determining the degree of hydronephrosis. The GI can also be used for long-term monitoring of children with hydronephrosis, including pre- and postoperative follow-up. As a more sensitive measure of renal parenchymal thickness, GI allows informed clinical decisions to be made by evaluating changes that, as noted by these researchers, are not identified by the current SFU system.

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