KIDNEY CYSTS IN CHILDREN

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https://doi.org/10.5281/zenodo.11412174

Abstract*. Cysts in childhood are a common occurrence. Among them there are quite a large group consists of sporadic cases, as well as random detection of single cysts in the kidney during routine ultrasound; the cause of the appearance of such cysts remains the subject of research. Modern genetics has made a great contribution to the differentiation of various kidney cysts. Polycystic kidney disease is a hereditary nephropathy associated with a mutation of genes that determine the structure of the renal tubules in their embryonic development, manifested by the formation of cysts in the renal parenchyma, an increase in which leads to sclerosis of the kidney tissue and the development of chronic renal failure.*

Keywords*: cysts, ADPKD (autosomal dominant polycystic kidney disease), abdominal pain syndrome, vascular anomalies, the adult type of polycystic disease.*

Relevance. The disease is characterized by fusiform cystic dilatation of the collecting ducts. Dysgenesis of the hepatic collecting system with rapid enlargement of the bile ducts and the formation of hepatic fibrosis is also typical. Children with ARPKD have a history of ischemia during the antenatal period of development; mothers had a severe pregnancy and oligohydramnios. The child has detected > 5 MAR. Due to enlarged kidneys, which is observed in the antenatal period, hypoplasia of the lung tissue occurs at birth manifested by distress syndrome, which can be complicated by pneumothorax. Congenital portal fibrosis leads to liver failure and hepatosplenomegaly. Under our supervision for 6 years there was 4 children with a relatively favorable course of ARPKD. Until relatively recently, ADPKD was called "the adult type of polycystic disease" [Kutyrina I.M., 2000]. This was due to the fact that the main clinical manifestations diseases: hematuria, hypertension, chronic renal failure - appear more often after 30 years of life. Modern ultrasound diagnostic methods can detect the presence of ADPKD in children of the first years of life and even in the antenatal period of their development, which makes the term "adult type ADPKD" questionable. Several stand out genetic variants of ADPKD:

-type 1, which occurs in approximately 80% of patients, is associated with a mutation in the PKD1 gene, located on the short arm of chromosome 16 and a defect in the gene product - polycystin-1;

-type 2, which occurs in approximately 15% of cases of the disease, develops due to a mutation in the PKD2 gene, located on the long arm of chromosome 4 (4q21-q23), gene product - polycystin-2;

-type 3, a rare variant, its frequency is not determined, more often detected in adults, there is no convincing evidence for mutations in the PKD1 genes and PKD2.Polycystin-1, a product of the PKD1 gene, is a large molecular protein. Polycystin-2 is a product of the PKD2 gene, which has a significantly lower molecular weight. Analysis of the primary structure of polycystins showed that they may be a hub part of the calcium channel. Polycystin widely expressed in the body, it is found in epithelial cells not only the kidneys, but also the pancreas, liver and intestines. It is in these Cysts or diverticula are often found in organs in patients with PCD.Polycystin-1 and

polycystin-2 are necessary for normal development, as death of experimental animals was observed in the absence of these proteins in late embryogenesis. They were admitted to the nephrology department of the Moscow Research Institute of Pediatrics and Children's Diseases at the age of over 3 years. Everyone noted more than 5 MAR. Proteinuria occurred in all patients; almost all had hematuria in combination with crystalluria. These children were lagging behind in physical development, complaining of abdominal pain, weakness, and fatigue. Ultrasound revealed large kidneys with multiple small cysts were found in the liver in 2 children, in the spleen - in 1 child. The purpose of this study. ADPKD is based on an incorrect connection during antenatal development of the fetus of straight and convoluted tubules, as a result of which the outflow of primary urine from the proximal tubules is disrupted, as a result cyst formation occurs. Cysts in ADPKD are formed mainly from main cells of the collecting ducts, initially they are connected to the mother cell, but subsequently this connection is broken, and an increase in volume cysts occur by proliferation of cyst-lining cells and secretion fluids in it.

Materials and methods of research. The study of the molecular basis of cyst formation led to the conclusion that the most important links in this process are:

-cellular proliferation, which leads to tubular obstruction, increased intratubular pressure, resulting in stretching tubules;

-excessive accumulation of fluid, which may be associated with disturbances of the sodium pump and the corresponding entry of sodium into the lumen tubules, and not into the blood. Aquaporins take part in this process 1 and 2 expressed on cyst epithelium.

-pathological, hereditarily caused increased stretching BM of the tubules, which contributes to their expansion. Deviations in the structure of the extracellular matrix of the tubules lead to disturbances in the structure and function of the tubular epithelium. Study defective nucleotides of genes involved in extracellular metabolism matrix in experimental PCB, confirms the above position. Children have a disease may be asymptomatic and detected by ultrasound of the kidneys. However, some children during physical activity or during intercurrent illnesses pain appears in the abdomen or lower back. In rare cases, it is possible to palpate sharply enlarged kidneys. Finding hematuria or UTI is fairly typical. With age, there is a tendency to increase Blood pressure, which in children may have the character of latent hypertension, i.e. determined only for ABPM. There is evidence that the age of onset of increased Blood pressure and its severity depend on the presence or absence of hypertension in parents child. A third of patients have liver cysts,10% - in the pancreas, 5% - in the spleen. In our observations of 30 children aged 4 to 16 years with ADPKD, the familial nature of ADPKD occurred in 24 cases, in the rest it was considered as diseases de novo. Ultrasound revealed 5 or more cysts in both kidneys ranging in size from 1 to 3 cm in diameter. There is information in the literature [Andreeva E.F.et al., 2008], that the increase in cyst size that occurs after 7–15 years life of the child, correlates with the nature of abdominal pain syndrome, Hypertension and changes in urine. In our observations, except for cysts in the kidneys, in 10% In two cases, children had cysts in the liver; two children had cysts in the spleen. Sclerosis develops in the kidney, and is still on going discussions - sclerotic changes are secondary to cysts increasing in size and ischemia of the surrounding tissue or is it an independent process that has a different genetic basis. Molecular genetic studies have shown that the ARPKD gene is PKHD1 - located on the short arm of chromosome 6 (6p21-p12). Product gene - fibrocystin. It was experimentally discovered that already in the formed kidney in the cystic

epithelium the product of the SGP-2 gene is detected -clusterin, the functioning of which is characteristic only of immature epithelium [Klingel R. et al., 1992].

Researches and discussion. ADPKD is one of the most common genetically determined diseases in the population. On average, the incidence of ADPKD in the world is 1:1000, in the European population - 1:400. This frequency of the disease is associated with the high penetrance of mutating genes. Due to the fact that ADPKD is often combined with vascular anomalies, experimentally tested the possibility of influencing the occurrence of polycystic kidney disease in the embryonic period of the formation of vascular endothelial growth factor (VEGF). Children have a disease may be asymptomatic and detected by ultrasound of the kidneys. However, some children during physical activity or during intercurrent illnesses pain appears in the abdomen or lower back. In rare cases, it is possible to palpate sharply enlarged kidneys. Already during early anatomical studies of the kidneys in PKD, it was noted that some cysts in the kidneys of patients with ADPKD are surrounded by thickened BM, that suggested the presence of special collagen or its degradation by metalloproteinases [Norman J., Wilson P, 1996]. However, further research showed that, according to its main characteristics, sclerosis of renal tissue with ADPKD is identical to sclerosis in any chronic kidney disease. ARPKD occurs much less frequently in the population than ADPKD. The detection of pathology is reported in 1:6000-1:40,000 newborns [PancrykTomaszenska M., Hoppe V., 2002]. There is a high mortality rate in the perinatal and early neonatal periods, although with modern treatment methods There is also a relatively long life expectancy.

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