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CLINLCAL CASE OF MULTICYSTIC DYSPLASIA OF THE LEFT KIDNEY IN A NEWBORN CHILD – SCIENTIFIC AND APPLIED REALITIES

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Summary

The clinical picture of congenital anomalies of the kidneys and urinary tract can vary from isolated renal anomalies to syndromic phenotypes. The modern components of management of congenital anomalies of the kidneys and urinary tract, described in the article on the example of a clinical case of congenital polycystic dysplasia of the left kidney, will help to deepen the scientific and applied competencies of doctors who are part of a multidisciplinary team of medical process of this congenital developmental anomaly, as well as parents or persons responsible for the child.

Keywords: *Congenital Polycystic Dysplasia of the Left Kidney; Congenital Anomalies of the Kidneys and Urinary System; Newborns.*

When working with a patient, each physician must carefully consider, systematize and justify a plan to achieve the goal, to be implemented according to the patient's needs, followed by an evaluation of the results of the diagnostic search, treatment management and prognosis. All components of the medical process in which the patient is the primary participant should not only be documented in detail, but also communicated in an accessible form to family members/caregivers, as these individuals, after appropriate training, will become full members of the multidisciplinary treatment and follow-up team, learning and performing the skills necessary to improve the patient's health outcomes. In addition, health care providers should place information materials on the main clinical manifestations of diseases in places accessible to patients, as well as on their websites and official pages of health care facilities.

Despite modern technologies and possibilities of medical-genetic counseling and medical care for women of reproductive age, congenital malformations still occupy a leading position in the structure of perinatal morbidity and mortality.

Thus, in 2022-2023 in the Lviv region 78 congenital malformations were registered out of 15489 live births. Among the 84 children who died before the age of one year, 19 % of children had severe isolated or combined malformations, including multicystic kidney dysplasia (Latin: dysplasia renum multicystica, English: multicystic dysplastic kidney), which is defined as a large group of genetically determined diseases, a type of kidney development disorder, the visible manifestation of which is multiple cysts of different sizes and the absence of a normal pelvic system (Fig. 1).

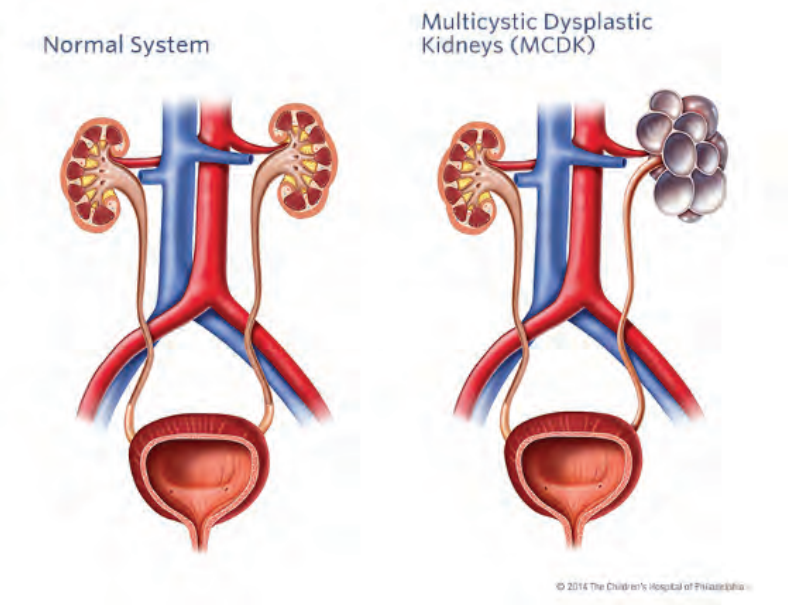

Fig. 1. Comparative view of the normal structure of the urinary system (left) and multicystic dysplasia (right)

In general, cystic kidney disease is a common cause of end-stage renal disease in children, with an overall incidence of about 4 cases per 10,000 births [1]. Multicystic kidney dysplasia is a sporadic genetic or inherited disorder. The unilateral form is more common in 1/4300 live births; if the defect affects two kidneys, the life prognosis is poor. [2]. It is inherited as an autosomal dominant trait, i.e. the patient's children have a 50 % risk of inheriting the disease. The frequency of occurrence is 1:400-1:1000. The incidence in Finland is 7-8/1000000. The cause of the disease is a mutation in the PKD1 or PKD2 genes [3]. The group of congenital anomalies of the kidney and urinary tract (CACUT) is a diverse group of structural malformations that occur as a result of abnormalities in embryonic development of the kidney and urinary tract at any stage. It is the leading cause of acute kidney injury in childhood (~40 %) with a frequency of 3-6 affected children per 1000 births [4].

The expected question of the child's parents will be an explanation of the causes of multicystic kidney dysplasia, which can be answered as follows: at about 5 weeks of pregnancy, the germ of the tissue that forms the ureter connects with the tissue that forms the kidney. If this process is abnormal, a kidney is formed that consists of numerous cysts and is called a multicystic kidney. Due to the lack of communication with the ureter, the diseased kidney is unable to excrete urine and function. At the same time, it should be explained to the family members/caregivers that multicystic kidney disease is a genetic and/or inherited developmental disorder characterized by the formation of cysts in the kidneys that can increase in size and impair kidney function [1].

Pathology of renal development is most often diagnosed prenatally, using ultrasound, which can visualize an increase in kidneys compared to their normal size, multiple echonegative hollow formations, ranging in size from point to 2-7 mm in diameter. The cysts occupy almost the entire renal parenchyma. There is a focal or diffuse increase in echo density in the remaining parenchyma. Cysts are found in the liver in one in five children [1] However, in the early stages of fetal development it is not always possible to make a differential diagnosis with other renal and upper urinary tract anomalies leading to dilatation of the urinary tract cavity system, especially in the case of bilateral localization of the defect by ultrasound. Renal anomalies are found in 10-20 % of prenatal ultrasound examinations [1]. These anomalies are most commonly described as bilateral enlarged and/or hyperchogenic kidneys and can be divided into three main groups. The first includes hereditary hepatorenal fibrocystic disease (HFD), a complex of monogenic disorders characterized by fibrocystic kidney abnormalities and portobiliary dysgenesis, a disorder associated with mutations in genes encoding proteins that function in the primary cilium or centrosome, and thus HFD is considered a renal subset of the larger group of ciliopathy disorders [1]. The second group includes congenital anomalies of the kidney and urinary tract (CAKUT) – a spectrum of disorders that can be manifested by renal hypoplasia, uretero-vesical junction obstruction, primary megaureter, vesicoureteral reflux, uretero-vesical junction obstruction or posterior urethral valves, with or without concomitant cystic kidney disease [1]. The latter group includes patients with chromosomal or syndromic disorders. The clinical presentation in all groups can vary from mild to severe with anhydramnios and pulmonary hypoplasia causing significant perinatal mortality [1]. In terms of sonographic imaging, these different disorders often mimic or phenocopy each other.

In order to increase the efficiency of defect detection up to 95 %, the use of MRI for early differential diagnosis of multicystic kidney dysplasia with upper urinary tract defects accompanied by marked expansion of the cavernosal system can be a clarifying imaging method in doubtful cases [1]. In addition, MRI in the prenatal period allows not only to diagnose multicystic kidney, but also to verify the parenchyma of the affected kidney, which gives reason to consider prenatal MRI as an effective method of diagnosing multicystic kidney, which will determine the tactics of pregnancy management and delivery.

The sonographer should inform the child's parents that in most cases the cyst will involute, and the remaining renal tissue is sometimes too small to be detected by standard imaging techniques. The second kidney is usually normal in structure but larger in size as a result of compensatory hypertrophy. Multicystic kidney dysplasia may present with kidney pain, blood in the urine, arterial hypertension, and other kidney problems. Treatment of a child with congenital polycystic kidney disease is usually aimed at relieving symptoms and maintaining kidney function [1].

Thus, the interpretation of the obtained fetal kidney ultrasound data is provided to the parents of the «fetal child» by the ultrasound physician and later by the physician who manages the pregnancy at the outpatient stage – this confirms the multidisciplinary work of clinical case management already at the outpatient stage of medical care. It is important to focus the attention of parents with prenatally detected renal dysplasia on dynamic monitoring of the child, which allows to avoid nephrectomy in 80 % of cases [1]. In 50 % of children with multicystic kidney disease, the organ dries up over time and gradually stops growing, resulting in replacement of the parenchyma by connective tissue and gradual shrinkage of the kidney. This is the most favorable course of the disease in humans. In the case of a PKD2 gene mutation, the disease has a milder course, the need for dialysis in such patients occurs on average at the age of 69, while in the case of a PKD1 gene mutation, the average age of initiation of dialysis is 53. In 85 % of patients with polycystic kidney disease, the disease is caused by a mutation in the PKD1 gene [1].

The most common questions that members of the multidisciplinary medical team may hear from parents of a child with prenatally diagnosed multicystic kidney dysplasia include What is the best way to have my baby? Will I be able to breastfeed my baby? What can our baby expect after birth? Will my baby be transferred to another hospital? What does the future hold for our baby?

To help doctors involved in the management of a newborn with congenital polycystic kidney disease, to structure the sequence of conversations to comprehensively inform parents of a child with this kidney disease, to develop a memo for parents, the presentation of a clinical case of congenital anomaly of the kidney and urinary tract will help.

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Objective: to describe the features of clinical management of a case of congenital anomaly of the kidney and urinary tract to deepen the applied knowledge of doctors who are part of a multidisciplinary team for the management of congenital multicystic dysplasia of the left kidney in a newborn child

Clinical case

Medical history: The child was born with a weight of 3450 g, length of 54 cm. Apgar score was 8/8 b. After birth he was admitted to the neonatal unit of the perinatal center with the diagnosis of cystic kidney dysplasia, unilateral. Congenital megaureter. Open oval window. Isolated dilatation of the left lateral ventricle at the level of the posterior and temporal horn.

The child was born in the second pregnancy, first delivery at 38 weeks. Delivery by cesarean section. Features of this pregnancy: isthmic cervical insufficiency, corrected by pessary. Maternal hereditary predisposition. Maternal congenital renal disease (left-sided multicystic kidney disease).

On the 4th day the child was discharged home with recommendations for further examination and treatment of the congenital defect of the left kidney. On the 16th day of life the parents and the child went to the reception of the Clinical Center of Pediatrics for further examination and treatment. The child was fed with an adapted formula due to antibiotic therapy for pneumonia in the mother.

The newborn boy at the age of 16 days was admitted to the post-intensive care of newborns and premature infants center of the Supercluster Clinical Center of Pediatrics for inpatient treatment and further examination on the referral of a family doctor.

Diagnosis: Other clarified congenital kidney defects: giant multicystic kidney disease of the left kidney with compression of the abdominal organs. Megaureter on the left. Another candidiasis of the newborn. Open oval window.

Objective examination data: T – 36.6 °C, body weight on admission – 3630 g, BP – 60/min, HR – 160 beats/min, SpO2-98 %, OH – 35 cm, WC – 41 cm.

On admission to the center for post-intensive care of newborns and premature infants of the Supercluster Clinical Center of Pediatric Medicine, the general condition of the child was moderate due to a congenital kidney disease. During the examination, the child is restless. The skin is subchlorotic and dry. Visible mucous membranes are pale pink, white plaque in the oral cavity. The head height is 3.0 x 3.0 cm, at the level of the skull bones. Breathing is independent, uncomplicated, respiratory rate – 54 per minute. To ensure sufficient blood oxygenation – 92-95 % requires oxygen therapy through nasal cannulas, flow rate – 1 liter/min.

Auscultation, vesicular breath was heard over the lungs throughout the lungs, on both sides. The heart sounds were rhythmic, sound. Heart rate was 146 beats per minute. Hemodynamics, according to the assessment of mean arterial pressure (44 mm Hg), the state of peripheral perfusion, determined by checking the «white spot» symptom – 2.5 seconds, filling, symmetry, tension, rhythmic pulse on the radial arteries, external temperature of the distal parts of the body – stable, pulsation on the femoral arteries is good, symmetrical.

The abdomen is enlarged in volume, pronouncedly distended, with clearly contoured lines of the boundaries of enlargement, a venous network is visualized on the anterior abdominal wall, soft to palpation (Fig. 2).

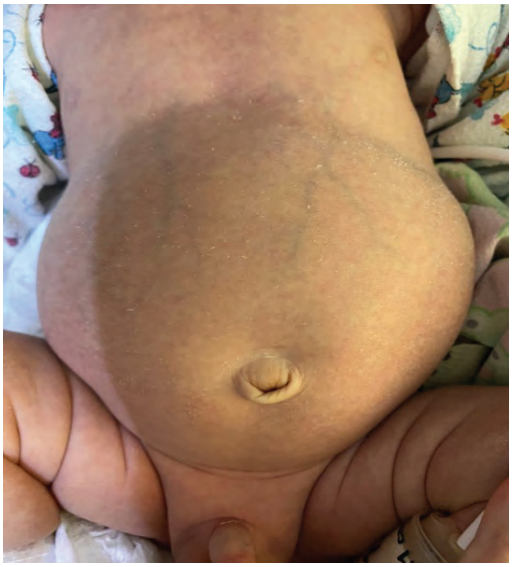

Fig. 2. Appearance of the abdomen of a child with congenital multicystic kidney dysplasia: enlarged in volume, markedly distended, with clearly contoured lines of the enlargement boundaries, a venous network on the anterior abdominal wall is visualized.

Peristalsis is heard on the right. The umbilical ring is epithelialized. Liver: rounded edge, elastic consistency, protrudes 2 cm from under the right edge of the rib arch. The genitals are of the male type, the testicles are lowered into the scrotum. Urination is active and sufficient. The stools are independent, without pathological impurities.

During the stay in the center for post-intensive care of newborns and premature infants of the Supercluster Clinical Center for Pediatric Medicine, the child underwent the following tests, examinations, and consultations with narrow specialists:

Laboratory data:

Table 1

Hemogram of a child during a stay in the center for post-intensive care of newborns and premature infants of the Supercluster Clinical Center for Pediatric Medicine

Blood cells	Normal values (0-1 months)	Child's age 16 days	Child's age 21 days
Red blood cells 10 12/l	3,9-6,2	4,73	4,42
Hemoglobin (Hb) g/l	160-230	160	154
White blood cells 10 9/l	10,0-28,0	11,39	9,97
Platelets 10 9/l	150-400	314	326
Segmented neutrophils, %.	30-55	27	17
Neutrophils with stick nuclei, %.	1-5	1	7
Eosinophils, %.	1-5	0	5
Lymphocytes, %.	45-65	60	57
Monocytes, %.	До 8	6	20

Mild neutropenia in terms of segmented neutrophils and monocytosis in the hemogram in the dynamics is explained by the presence of candidal infection that occurred from the first day of hospitalization and pathological microbial colonization of the mucous membranes of the nose, eyes, pharynx, and umbilicus.

Sowing from the umbilicus on the 16th day of life: St. aureus.

Nasal culture 16 days of life: St. aureus, Haemophilus influenzae.

Sowing from the eye on the 16th day of life: Candida albicans.

Sowing from the pharynx on day 16 of life: Candida albicans, St.aureus, Haemophilus influenzae.

Urine culture on day 16 of life: no pathogenic flora was detected.

Blood culture on day 16 of life: blood is sterile

According to the results of the general urinalysis, no signs of urinary tract infection were detected.

Table 2.

Indicators of general urinalysis of a child with congenital multicystic kidney dysplasia

Date	color	transparency	protein	epithelium	white blood cells	red blood cells	mucus
16 days	straw yellow	transparent	not detected	1-2 in the field of view	3-5 in the field of view	0-1 in the field of view	+
1 days	straw yellow	transparent	not detected	2-3 in the field of view	4-7 in the field of view		+
22 days	straw yellow	transparent	not detected	0-1 in the field of view	1-3 in the field of view	0-1 in the field of view	not detected

Biochemical blood test on the 17th day of the child's life: urea – 2.57 mmol/l, normal; creatinine – 63.8 mmol/l, elevated (normal 0.035-0.06 mmol/l), signaling the development of renal failure in the child.

The glucose level was 6.23 mmol\l, bilirubin (total) – 166.0 μmol\l – elevated, AST – 26.5 U\l, ALT – 16.0 U\l, total protein – 52.7 g\l, CRP – (–) negative.

Biochemical blood test from the 22nd day of life: urea – 1.91 mmol\l, creatinine – 75.3 mmol\l, in the dynamics the indicator increased; glucose – 4.76 mmol\l, bilirubin (total) – 107.8 μmol\l, decreased in the dynamics; AsAT – 27.2 U\l, (normal) AlAT – 14.7 U\l (normal), total protein – 60.3 g\l, CRP – (–) negative.

Radiography of the chest and abdominal organs on day 17 of life (Fig. 3).

On the first day of hospitalization, the child underwent an ultrasound of the internal organs and urinary system **(Fig. 4).**

Right kidney: 5.7 x 2.2 cm in size. The parenchyma is homogeneous, 1 cm. Cortico-medullary differentiation is preserved. The pelvic system is not dilated. Left kidney: multiple anechogenic masses 2.1-7.4 cm in size in the projection. Cortico-medullary differentiation is not visualized. Ureter: tortuous on the left, dilated from 1.6 to 2.2 cm. Bladder: with clear, even contours, homogeneous contents. Conclusion: The sonographic picture may correspond to a multicystic left kidney, megaureter on the left.

Neurosonography at the age of 17 days: The sonographic picture may be consistent with right choroid plexus cysts.

Audiogram performed at 22 days of age: click-induced otoacoustic emission is present on both sides.

Consultation with an ophthalmologist at the age of 18 days: OD\OS pupils are sufficiently dilated. Optical media are clear. Fundus is age-appropriate.

Neurologist consultation at 22 days of age: no pathological changes were found in the neurological status at the time of the examination.

Consultation of a geneticist at the age of 21 days: the band has a VSD of the urinary system: multicystic left kidney.

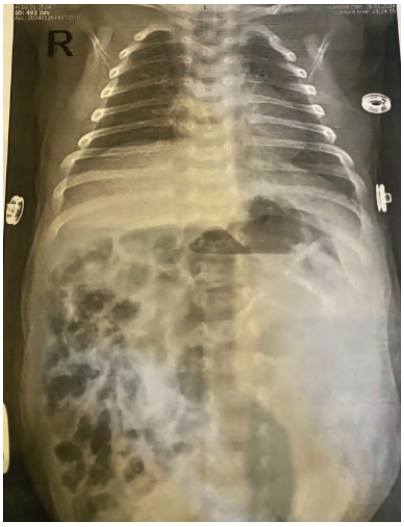

Fig. 3. X-ray of the chest and abdominal organs on day 17 of life: no focal or infiltrative shadows were detected. Pulmonary pattern is enhanced in the lower medial regions. The diaphragm is contoured. The sinuses are free. The abdominal cavity is enlarged in size, the intestinal loops are unevenly pneumatized, the right half of the abdominal cavity is sharply displaced. No free air is found in the abdominal cavity.

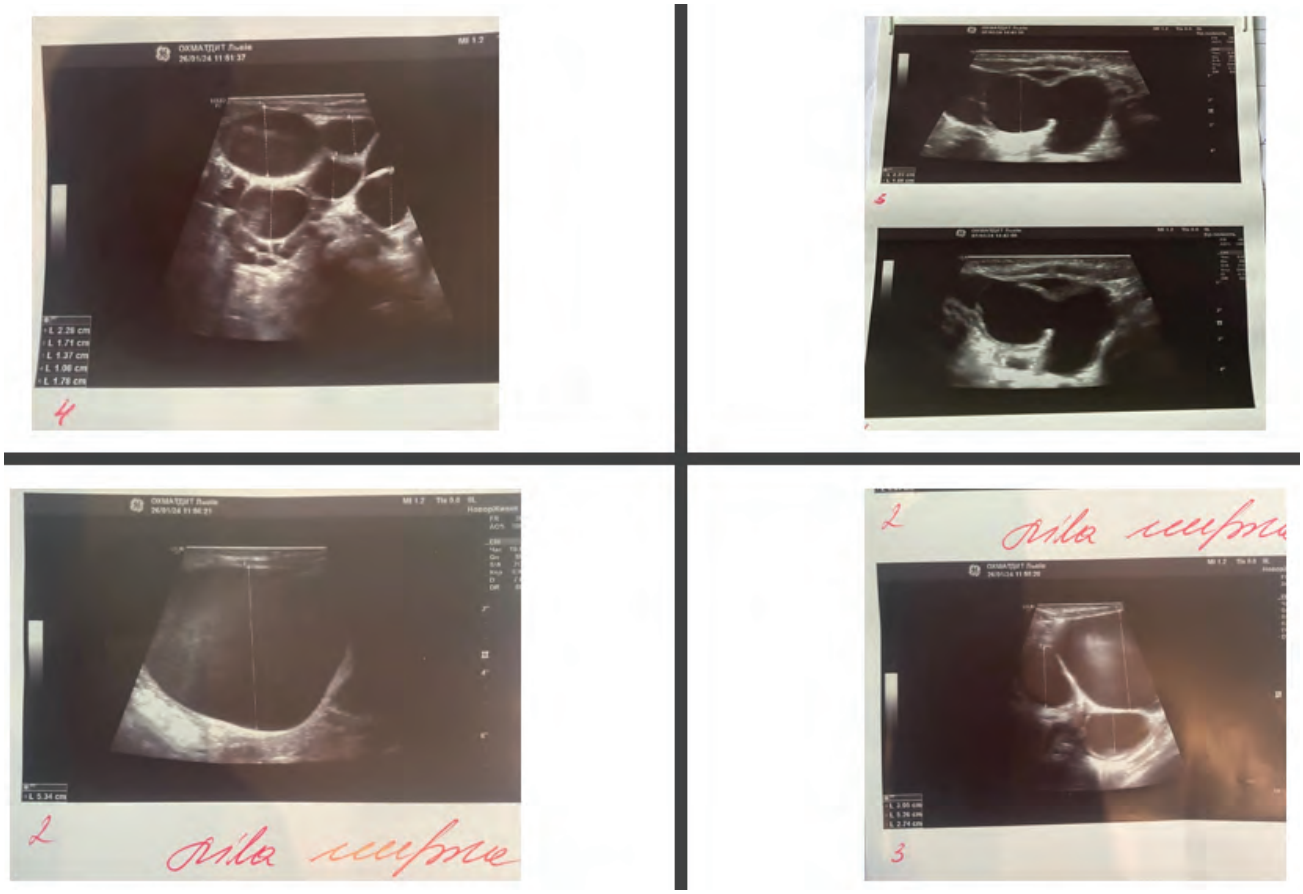

Fig. 4. Ultrasound of internal organs and urinary system on the 16th day of life.

The following conclusions can be drawn from the results of genetic sequencing:

Autosomal dominant polycystic kidney disease, hepatorenal fibrocystic disease, can manifest in the perinatal period, childhood or young adulthood and is caused mainly by mutations in two genes, PKD1 (78 % of cases) and PKD2 (15 % of cases), with the remaining 5-10 % of cases due to rare mutations at other loci []. In comparison, autosomal dominant polycystic dysplasia is less common, with an incidence of 1 in 26,500 live births []. The disease is mainly caused by mutations in the PKHD1 gene. In its most severe prenatal manifestation, autosomal dominant polycystic kidney disease is characterized by bilaterally enlarged echogenic kidneys and oligohydramnios, with a perinatal mortality rate of 21 % due to lung hypoplasia []. Surviving patients usually reach adulthood due to medical

advances in pediatric renal replacement therapy and kidney transplantation []. The rarer forms of hepatorenal cystic kidney dysplasia, NPHP, JBTS, MKS, and BBS, which are characterized not only by cystic kidney lesions but also by cystic kidney disease, may present with a variety of extrarenal features, including polydactyly, occipital encephalocele, liver fibrosis, obesity, and retinal degeneration.

The clinical presentation of congenital anomalies of the kidneys and urinary tract can vary from isolated renal anomalies to syndromic phenotypes. Familial clustering is observed. More than 50 genes are involved in CAKUT. The most common mutations involve 12 genes, such as BMP7, CDC5l, CHD1l, EYA1, GATA3, HNF1B, PAX2, RET, ROBO2, SALL1, SIX2, and SIX5. However, single gene mutations may occur in 10 % to 20 % of CAKUT cases. []. In addition, copy number variants (CNVs), i.e. duplications or deletions of genomic segments, underlie 5-10 % of cases of congenital anomalies of the kidney and urinary system.

Consultation with a nephrologist at the age of 20 days: multicystic left kidney was diagnosed in a child with a complicated history (maternal multicystic left kidney, secondary chronic pyelonephritis). At present the function of the right kidney is preserved. Recommended: treatment by a urologist, monitoring of urine analysis, kidney function.

Consultation by urologist at the age of 16 days: conclusion: congenital malformation of the left kidney: multicystic kidney disease?

Consultation by urologist at the age of 20 days: the child was diagnosed with a severe congenital malformation of the urinary system – giant multicystic left kidney with compression of the abdominal organs.

Recommendations:

1. Additional examination: cystography.
2. Conditionally urgent left-sided nephrectomy.

Echo-CG at the age of 16 days: the heart chambers are not dilated. No septal defects were found. Open oval foramen –3-4 mm. The arterial duct is at the stage of closure. The course of the main vessels is correct. The function and structure of the valves are good. Myocardial contractility is good. The ejection fraction is 68 %. There is pulsatile blood flow in the abdominal aorta. There is no evidence of aortic coarctation. There is no fluid in the pericardium and pleural cavities. Conclusion: There is no evidence of congenital heart disease or coarctation of the aorta. The open oval window is hemodynamically insignificant.

Based on the results of subjective and objective symptoms of a child with congenital multicystic kidney dysplasia with a tendency to develop acute renal failure, the results of laboratory and instrumental studies, consultations with narrow specialists, the responsible doctor summarized and summarized all the data and developed further treatment and follow-up tactics for the child, namely: the child should be under the supervision of a pediatric urologist. In most cases, the disease regresses, leaving the patient with one kidney. If the cysts increase in volume, surgical treatment (nephrectomy) is required.

Current guidelines for the management of congenital renal and urinary tract anomalies state that: any antenatal suspicion/diagnosis of congenital renal dysplasia is confirmed by neonatal ultrasound (US), avoiding the routine use of further imaging unless other renal/urinary tract anomalies are detected, including MRI. Nephrectomy for large cysts can now be performed using a laparoscopic technique. The main indication for surgery to remove the organ damaged by cysts in patients with multicystic renal dysplasia is the syndrome of abdominal compression by a bulky mass accompanied by pain and the presence of reflux into the affected kidney with signs of chronic pyelonephritis.

Possible complications of congenital cystic kidney dysplasia may include: compression of vital organs located in the abdominal cavity. suppuration of cysts, their subsequent inflammation and rupture, possible peritonitis, and high blood pressure, which cannot be treated with medication.

According to Han J. H., Lee Y. S., Kim M. J. et al. [15] A multicystic kidney regresses in 60 % of cases and does not require surgical intervention.

The congenital single kidney without additional anomalies may undergo compensatory enlargement, which should be evaluated by ultrasound. Regular urinalysis, but not blood tests or genetic analysis, should be performed at diagnosis in infants and children with compensatory enlargement of the single kidney. Extrarenal malformations, especially genital tract malformations in females, should be sought. Excessive protein and salt intake should be avoided and exercise should not be restricted. In children with dysplastic kidney or single kidney, lifelong follow-up is recommended, which should be adapted to the risk stratification, namely: low risk: congenital single kidney with compensatory enlargement, medium risk: congenital single kidney without compensatory enlargement and/or additional congenital anomaly of the kidneys and urinary tract, and high risk: decreased glomerular filtration rate and/or proteinuria and/or hypertension.

Children at low risk should have regular ultrasound, urinalysis, and blood pressure measurements; children at moderate risk should also have serum creatinine measured; children at high risk should have the replacement therapy schedule adjusted according to renal function and clinical data.

Prognosis: In the first year of life, the child may become constipated if pressure is applied to the abdomen, or faint if the vena cava is compressed. The follow-up plan for children with congenital renal anomalies is determined by risk stratification as follows: low risk: kidney length > 50th percentile in the first 2 years of life and ≥ 95th percentile thereafter, and absence of ipsilateral congenital renal and urinary tract anomalies; intermediate risk: CAKUT without compensatory renal enlargement and/or with ipsilateral renal and urinary tract enlargement: reduced congenital dysplastic kidney (i.e., mean glomerular filtration rate for age – 1 SD in children younger than 2 years, < 90 ml/min/1.73 m2 in children older than 2 years, < 90 ml/min/1.73 m2 in children younger than 2 years and/or proteinuria and/or hypertension [1].

Low-risk children can be seen by general pediatricians (if possible, based on the local health care system), while intermediate-risk children requiring specialized care should be seen by a pediatric nephrologist, and high-risk children should be seen in pediatric nephrology units. In all risk classes, women with SAKUT should undergo abdominal

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ultrasound after telarche and before menarche to assess the condition of the genital apparatus. In addition, follow-up should continue into adolescence in all risk classes. Finally, the transition of patients from pediatric to adult care should be carefully planned, as this is a critical period in terms of maintaining renal function. [1].

Conclusions

1. The management of newborns with polycystic kidney disease should be carried out by a multidisciplinary team of competent professionals who provide treatment, monitoring, prevention and comprehensive education of parents/caregivers about this congenital kidney disease, starting from the prenatal counseling stage.

2. The main indication for surgery to remove an organ damaged by cysts in patients with multicystic renal dysplasia is the syndrome of compression of the abdominal cavity by a mass, which occurs with pain, the presence of reflux into the affected kidney with signs of chronic pyelonephritis, suppuration of cysts, their subsequent inflammation and rupture, peritonitis, increased blood pressure, which cannot be treated conservatively.

3. Newborns, infants with congenital multicystic kidney dysplasia should be under constant, dynamic observation of a pediatric nephrologist until adulthood, who, based on the results of stratification of the risk of complications, can refine the observation of children with congenital kidney dysplasia. Children with low risk can be followed by pediatricians and general practitioners on an outpatient basis, while children with medium risk should be followed by a pediatric nephrologist, and children with high risk should be followed in pediatric nephrology units.

Prospects for further research: to supplement the existing data on the experience of clinical course and management of congenital polycystic kidney dysplasia with a series of cases to deepen the existing scientific and practical data on this congenital developmental problem.

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КЛІНІЧНИЙ ВИПАДОК МУЛЬТИКІСТОЗНОЇ ДИСПЛАЗІЇ ЛІВОЇ НИРКИ У НОВОНАРОДЖЕНОЇ ДИТИНИ – НАУКОВО-ПРИКЛАДНІ РЕАЛІЇ

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Резюме.

Клінічна картина вроджених аномалій нирок і сечовидільної системи може варіювати від ізольованих ниркових аномалій до синдромальних фенотипів. Сучасні складові менеджменту вроджених аномалій нирок і сечовидільних шляхів, описані у статті на прикладі клінічного випадку вродженої полікістозної дисплазії лівої нирки, допоможуть поглибити науково-прикладні компетентності лікарів, які входять у мультидисциплінарну команду медичного процесу цієї вродженої аномалії розвитку, а також батьків або осіб, відповідальних за дитину.

Ключові слова: вроджена полікістозна дисплазія лівої нирки; вроджені аномалії нирок і сечовидільної системи; новонароджені.

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