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ЖУРНАЛ ГЕПАТО-ГАСТРОЭНТЕРОЛОГИЧЕСКИХ ИССЛЕДОВАНИЙ

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БОЛЕЗНЬ И СИНДРОМ КАРОЛИ: ОБЗОР ЛИТЕРАТУРЫ И СОБСТВЕННОЕ НАБЛЮДЕНИЕ СИНДРОМА КАРОЛИ У РЕБЁНКА, ЗАВЕРШИВШЕЕСЯ УСПЕШНОЙ ТРАНСПЛАНТАЦИЕЙ ПЕЧЕНИ

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АННОТАЦИЯ

Цель представления: Представлен обзор отечественной и зарубежной литературы по болезни и синдрому Кароли. Обзор включает историческую справку и современный взгляд на этиологию и патогенез болезни, её место среди фиброкистозных болезней печени. Демонстрация клинического случая синдрома Кароли у ребёнка, наблюдавшегося авторами с рождения. Заболевание манифестировало врождённой кистозной трансформацией почки. Печёночная патология была выявлена в возрасте 5 лет, с последующим быстрым прогрессированием поражения печени с развитием тяжёлых осложнений: цирроза печени, портальной гипертензии, отёчного синдрома, печёночной энцефалопатии. В возрасте 11 лет пациенту успешно проведена ортотопическая трансплантация печени.

Основные положения. Болезнь Кароли – редкая генетическая патология, характеризуется широким клиническим и морфологическим полиморфизмом от тяжёлых вариантов с фиброзом в неонатальном период до случаев с манифестацией в позднем возрасте. Изучены генетические причины патологии, в частности, мутации, связанные с аутосомно-рецессивной и аутосомно-доминантной формами поликистозной болезни почек. Большое количество описанных мутаций и преобладание среди пациентов сложных гетерозигот объясняет разнообразие клинических форм патологии. Взаимоотношение генотипа и фенотипических вариантов патологии не определены. При болезни Кароли помимо кистозных изменений в печени и почках, может наблюдаться поражение других органов и систем, в частности церебральных сосудов. Описаны случаи сочетания с различными генетическими заболеваниями.

Заключение. Представленный случай демонстрирует позднее выявление печеночного поражения, быстрое его прогрессирование. Несмотря на наличие тяжёлых осложнений, трансплантация печени имела успех и в настоящее время пациент находится в стабильном компенсированном по всем функциям состоянии.

Ключевые слова: болезнь Кароли, синдром Кароли, поликистоз почек, фиброкистоз печени, трансплантация печени.

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DISEASE AND CAROLI SYNDROME: LITERATURE REVIEW AND OWN CASE STUDY OF CAROLI SYNDROME IN A CHILD RESULTING IN SUCCESSFUL LIVER TRANSPLANTATION

ANNOTATION

The purpose of the presentation: A review of domestic and foreign literature on Karoli's disease and syndrome is presented. The review includes a historical background and a modern look at the etiology and pathogenesis of the disease, its place among fibrocystic liver diseases. Demonstration of a clinical case of Karoli syndrome in a child observed by the authors from birth. The disease was manifested by congenital cystic transformation of the kidney. Hepatic pathology was detected at the age of 5 years, followed by rapid progression of liver damage with the development of severe complications: cirrhosis of the liver, portal hypertension, edematous syndrome, hepatic encephalopathy. At the age of 11, the patient successfully underwent orthotopic liver transplantation.

The main provisions. Karoli's disease is a rare genetic pathology, characterized by a wide clinical and morphological polymorphism from severe variants with fibrosis in the neonatal period to cases with manifestation at a late age. The genetic causes of pathology have been studied, in particular, mutations associated with autosomal recessive and autosomal dominant forms of polycystic kidney disease. The large number of described mutations and the predominance of complex heterozygotes among patients explains the diversity of clinical forms of pathology. The relationship between genotype and phenotypic variants of pathology has not been determined. In Karoli's disease, in addition to cystic changes in the liver and kidneys, there may be damage to other organs and systems, in particular cerebral vessels. Cases of combination with various genetic diseases are described.

Conclusion. The presented case demonstrates the late detection of liver damage, its rapid progression. Despite the presence of severe complications, liver transplantation was successful and the patient is currently in a stable condition compensated for all functions.

Key words: Caroli disease, Caroli syndrome, polycystic kidney disease, fibrocystic liver disease, liver transplantation.

Conflict of interest. Authors declare no conflict of interest.

KAROLI'S DISEASE AND SYNDROME: LITERATURE REVIEW AND OWN OBSERVATION

The last decade has seen an increase in the number of publications of clinical cases and studies of Karoli's disease, which is associated with the development of genetic and molecular technologies, as well as with the success of transplantology [1-4]. Jacques Caroli, a French gastroenterologist, his colleague C. Couinaud and co-authors published articles in 1958 in which they described in detail a patient with cystic liver and kidney damage. Morphological changes were defined by the authors as "non-obstructive saccular (fusiform) multifocal segmental dilation of the intrahepatic bile ducts" [5, 6]. More than half a century before that, in 1906, British doctors H. Vachell and W. Stevens published an article entitled "The case of an intrahepatic stone", in which, no less detailed, they described a similar patient. The work of British authors remained without attention, and only a few decades later, the newly discovered pathology entered the medical annals under the name of Karolyi's disease. Karolyi's disease is rare, 1 case per 1 million. human, a genetic disease characterized by non-obstructive segmental cystic (sac-like or fusiform) extensions of the intrahepatic bile ducts. The largest number of patients was identified in the Asian population, more than 30% of the reports were made by Japanese authors.

In most patients, a mutation is identified in the PKHD1 gene (6p21.1-p12) encoding the synthesis of the fibrocystis-na/polyductin protein complex, which is secreted in the kidneys, bile ducts, pancreas and lungs. This mutation corresponds to autosomal recessive polycystic kidney disease (ARPK). Fibrocystin is one of the numerous proteins involved in the formation of primary cilia, which allows us to consider this pathology as ciliopathy [7-11]. More than 100 mutations have been described in the PKHD1 gene, in most cases these are complex heterozygotes. Numerous studies show clinical and morphological heterogeneity of patients, however, to date there are no reviews that draw parallels between the genotype and phenotype of patients.

The morphological basis of the disease is laid at the embryonic level in the form of a defect of the flow plate. It is possible to detect malformations in the fetus in the third trimester of pregnancy with ultrasound examination. The morphological changes detected in patients are diverse, including segmental absence of the muscle layer and collagen fibers, periductal fibrosis and eosinophilic infiltration, secondary changes may include cholangiolithiasis, cholangitis and liver abscesses. Cystic changes in the intrahepatic ducts are often combined with the expansion of extrahepatic and common ducts.

There are two forms of the disease [12]. The first (simple, true or isolated) is actually Karolyi's disease, described primarily in adults, characterized by a bag–like expansion of the main intrahepatic ducts with more frequent involvement of a separate lobe or segment of the liver. It is characterized by a manifest course with the development of

cholelithiasis, cholangitis and abscesses, which is manifested by fever and pain syndrome. Clinical manifestation is usually preceded by a long period of latent existence of cysts, at least until puberty. Cystic changes of the kidneys and pancreas are not uncommon, on the contrary, cirrhosis and portal hypertension do not develop often [13, 14].

The second form (complex, complex) is referred to as Karoli syndrome and is described in young children. Morphological changes are diffuse in nature with the expansion of small intrahepatic ducts, early development of fibrosis, in some cases existing already in utero, and portal hypertension. Stones in the intrahepatic ducts and cholangitis can form at an older age [15]. Karolyi syndrome is considered as a potentially precancerous condition. The probability of developing cholangiocarcinoma is 2.5–17.5%. There are isolated descriptions of the diagnosis of tumors (in particular hepatoblastoma) under the age of 3 years [16, 17].

The relationship between the syndrome and Karolyi's disease has not been studied enough. With Karoli syndrome, hereditary burden is relatively more often detected. Due to differences in the age of manifestation, patients with Karolyi syndrome and disease are more often analyzed in different reviews. Patients whose clinical picture was dominated by nephrological changes, as a rule, are considered as patients with polycystic kidney disease, and those with predominant hepatic pathology fall into the group of Karoli's disease and syndrome. The involvement of the same genes in Karoli's disease and syndrome on the one hand, and in polycystic kidney disease on the other, allows us to consider melon pathologies as phenotypic variants of autosomal recessive (ARPK) and autosomal dominant polycystic (ADPK) kidney diseases [18].

APK is diagnosed in utero or at birth, characterized by a more severe course, hematuria, arterial hypertension and the development of renal failure. Hepatic pathology is diagnosed in patients with this form, as a rule, later. In ARP, Potter syndrome with characteristic facial dysmorphic disorder, bone abnormalities and pulmonary pathology is described, congenital liver fibrosis and cystic changes in the liver are often mentioned.

The autosomal dominant type of inheritance is detected much less frequently in Karolyi's disease [13, 19, 20]. At APK, mutations are detected in PKD1 (85%) or PK D2 genes encoding the synthesis of a transmembrane receptor like polycystin-1 protein and polycystin-2 membrane protein homologous to Na+ and Ca2+ channels.

APK is diagnosed mainly in adults. Intrahepatic duct cysts are more often not clinically manifested, but may have manifestations similar to those described in Karoli's disease. Valvular heart defects and brain aneurysms have been described as concomitant pathology in ADPK. Vascular aneurysms also occur in ARPK, although less frequently [21]. There is a description of a combination of Karoli syndrome and Galena vein malformation in a 2-month-old boy [22].

Liver fibrosis (in particular congenital) is secondary to flow changes, while the ratio of cystic and fibrotic changes can vary in a wide range. In this regard, patients with so-called idiopathic liver fibrosis deserve attention. In a study by Watson ML et al. more than half of these patients had dilated renal tubules [23].

Numerous cases of combination of Karolyi syndrome and disease with other congenital diseases, in particular, with Lawrence-Moon-Bidl syndrome and with trisomy on chromosome 21 have been described [24]. Cystic and fibrous changes in the liver are described in Meckel–Gruber syndromes (ciliopathy described in the Finnish population), Joubert (cerebellar malformation), Jeune, short rib - polydactyly, Ivemark, vaginal atresia, tuberous sclerosis and juvenile nephronophthysis [25]. It can be assumed that in some patients, the clinical and morphological picture, similar or close to that of Karoli syndrome / disease, may be caused by other mutations (not associated with ARPK and ADPK).

The clinical picture (a set of symptoms and the sequence of their development), the severity of manifestations, as well as the age of manifestation of the disease vary widely, but in most cases its cyclical progressive character is noted, sooner or later leading to the development of characteristic complications. There are no differences in the frequency of symptoms between patients with the disease and Karoli syndrome. Pain syndrome associated with cholelithiasis can be considered as the earliest clinical manifestation of the disease.

No less characteristic is the development of recurrent cholangitis, manifested by episodes of fever with chills, pain syndrome and jaundice with itching of the skin. There are a large number of descriptions of cases of detection of the disease for the first time during examination for portal hypertension, gastrointestinal bleeding and splenomegaly.

The manifestation of developing cirrhosis of the liver and recurrent infection of the ducts can be protein-energy deficiency, anorexia and asthenia. Some patients had diarrhea, vomiting, hepatomegaly and splenomegaly. Isolated splenomegaly may reflect the latent development of portal hypertension. In some cases, liver pathology can only be detected by biopsy examination. So, K. Janowski et al. a 9-year-old patient with splenomegaly and portal hypertension was described, in whom fibrosis and duct malformation were determined during morphological examination [26].

Ultrasound examination of the hepatobiliary system is considered as the first stage of diagnosis, for detailed diagnosis, preference should be given to magnetic resonance cholangiopancreatography with contrast (MRCPG) [18]. Taking into account the geno- and phenotypic diversity of cystic changes in the liver and kidneys, it is advisable to conduct genetic studies on ARPK and ADPK in all cases of detection of hepatobiliary and renal cysts, as well as idiopathic liver fibrosis.

Drug treatment for Karoli's disease is aimed at complications of the disease: treatment of portal hypertension and hepatic insufficiency, antibacterial therapy for cholangitis, as well as for liver failure [18]. There are mixed reports about the efficacy of ursodeoxycholic acid in relation to the treatment and prevention of cholelithiasis, especially in relation to slowing the progression of fibrosis, however, in most descriptions of clinical cases and reviews, patients received this drug.

Surgical intervention should be performed before the development of complications such as portal hypertension. In Karoli's disease with an isolated lesion, resection of the affected lobe or segment is possible [27]. In the case of Karoli syndrome, the only way to treat patients is liver transplantation. One of the first reports of successful orthotopic transplantation belongs to M. Sens et al, 1976. [15, 28-32].

In recent years, active studies of Karolyi's disease have been conducted on animal models, which has revealed a number of subtle mechanisms of pathogenesis and cytokines that induce duct damage and fibrosis. In particular, the role of β-catenin, interleukin 1ß-dependent chemokine (CXC-motif) and ligand 10 (CXCL10) attracting tissue macrophages is shown.The use of inhibitors of these chemokines reduced the formation of cysts and the progression of fibrosis in the experiment [33, 34].

We present a clinical case of Karoli syndrome in a child who was under our supervision from birth to the age of 15. The boy D., born in 2006, was born from 14 pregnancies (10 medical and 2 spontaneous abortions), which proceeded with the threat of termination at 18 weeks, 3 deliveries by caesarean section at 36 weeks due to premature placental abruption. Anthropometric indicators at birth corresponded to gestational age (weight 2700 g, length 48 cm). He was born in severe asphyxia (2/6 points on the Apgar scale). He was in the Department of Pathology of newborns with a diagnosis of congenital pneumonia, cerebral ischemia. At the age of 1 month, a nephrectomy was performed due to multicystic left kidney. The postoperative period was complicated by enterocolitis, perforation of the small intestine with the development of purulent peritonitis, abscess of the abdominal cavity. Family history of liver or kidney diseases is not burdened. Up to 5 years old, he developed normally, underwent regular examination, rarely had respiratory infections, was vaccinated according to the national calendar. No pathology was detected in general and biochemical blood and urine tests. Ultrasound showed gradually increasing vicar hyperplasia of the right kidney, in which small single cysts were detected.

At the age of 5, ultrasound revealed cystic changes in the liver for the first time. In July 2011, against the background of high fever, there was an increase in the abdomen and the appearance of edema, an increase in the liver and spleen, ictericity of the sclera was noted. The blood test revealed anemia, thrombocytopenia and hypoalbuminemia. The child was hospitalized in the department of hematology due to suspected lymphoproliferative disease. Against the background of intensive therapy, which included transfusion of erythrocytes and albumin, antibacterial therapy, a significant positive dynamics was obtained.

The liver and spleen had a dense consistency, were determined 2 cm and 11 cm below the costal arch

In October 2011, he was hospitalized with a clinic of gastrointestinal and nasal bleeding, symptoms of liver failure-news with encephalopathy. There was a decrease in physical activity, daytime drowsiness and a violation of night sleep, speech disorders, tremors, refusal to eat. On examination, an increase in the circumference of the abdomen (71 cm) with symptoms of ascites, depletion of subcutaneous fat (weight 20 kg with a height of 120 cm), widespread edema, dilation of the veins of the lower abdominal wall, shortness of breath (Fig. 1, 2).

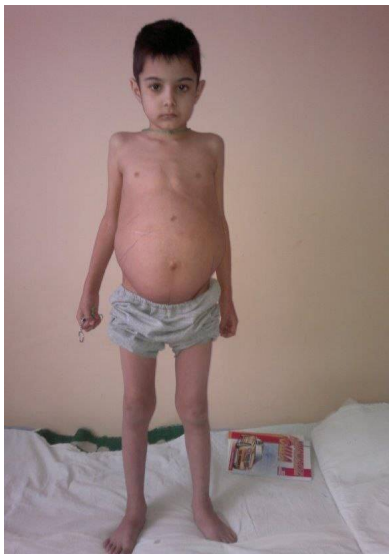

Fig 1.

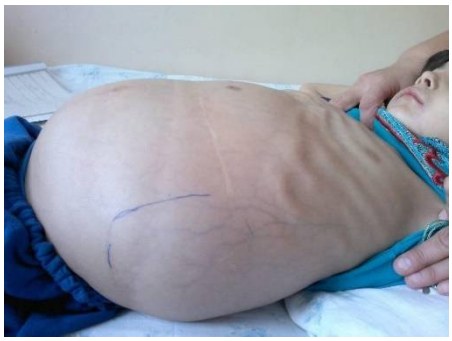

Fig 2.

Fig. 1. A patient aged 5 years (October 2011). Hospitalization in case of deterioration of the condition.
Fig. 2. A patient aged 5 years (October 2011). Ascites. Splenomegaly.

The blood test showed normochromic hyporegenerative anemia with a minimum hemoglobin level of 52 g/l, a tendency to leukopenia and thrombocytopenia up to 144x1012/l, an increase in ESR up to 73 mm/h, hypoalbuminemia up to 22 g/l, an increase in the globulin fraction up to 41 g/l, a maximum level of total bilirubin up to 29.2 mmol/l mainly due to indirect. The levels of transaminases, creatinine and urea did not increase, the level of alkaline phosphatase did not exceed the norm. There were signs of hypocoagulation in the coagulogram. In urine tests, there is no constant leukocyturia (up to 60 in the field of view), bacteriuria, proteinuria up to 0.099 g / l, erythrocyturia up to 5-7 in the field of view, the maximum specific gravity is 1017. GFR according to the Schwartz formula is 67.1 ml/min.

Endoscopically revealed the expansion of the veins of the esophagus of the I st . During ultrasound examination of the abdominal organs, the liver structure was described as uniformly compacted with multiple cystic formations, oblique vertical size of the right lobe 146 mm, enlargement and deformation of the gallbladder with thickening and layering of its wall, expansion of the oral and splenic veins to 14 and 11 mm, respectively. The spleen 175 x 77mm had a compacted heterogeneous structure. The right kidney is 137x60 mm with a compacted parenchyma up to 15 mm. An MRI of the abdominal organs visualized the expansion of the intrahepatic bile ducts up to 16 mm. Single cysts were detected in a single vicar-hypertrophied right kidney.

The combination of clinical laboratory and instrumental data made it possible to formulate the diagnosis: Karoli syndrome: Cystic dysplasia of the intrahepatic bile ducts, cirrhosis of the liver, portal hypertension, splenomegaly with hypersplenism, anasarca, hepatic insufficiency with hepatic encephalopathy of the first degree. Cystic dysplasia of the single vicar-hypertrophied right kidney. Secondary (hepatic) coagulopathy with hemorrhagic syndrome. Anemia of the III degree. Protein-energy deficiency of the II degree.

Treatment included transfusions of erythrocyte mass and freshly frozen plasma, vikasol, dicinone, diuretics, antibacterial therapy, lactulose, ornithine, ursodeoxycholic acid, propranolol. Positive dynamics was noted in the form of improvement of neurological status, reduction of edematous and relief of hemorrhagic syndromes.

During the next 2 years, the patient's condition was relatively stable against the background of planned therapy. There was a positive dynamics of nutritional status, complete relief of edematous syndrome and symptoms of encephalopathy. At that time, the child did not actively complain, was well adapted socially, continued to attend school.

At the age of 8 years (2014), the boy was examined in the surgical department of the Russian Children's Clinical Hospital and consulted in the Department of Liver and Kidney Transplantation of the Russian National Research Center. Academician B.V. Petrov, registered under the program of preparation for transplantation. However, the patient did not show up for subsequent consultations. During hospitalizations in 2015 and in May 2016, the condition remained relatively stable, signs of cytopenia were noted, mainly due to anemia.

Deterioration of the condition at the age of 10 years (September 2016): persistent edematous syndrome in the form of an anasarca, the symptoms of encephalopathy persisted and sharply increased against the background of often recurrent cholangitis. Pronounced ascites and splenomegaly, led to a sharp increase in the abdomen. There was shortness of breath with the participation of auxiliary muscles in breathing, the child took a forced position, sitting with support on his hands. At the age of 11, with a height of 134 cm , he weighed 39,5 kg .

Anemia and thrombocytopenia were detected in the laboratory, leukocytosis with severe intoxication was absent, acceleration of ESR, hypoalbuminemia up to 18 g/l, hyperglobulinemia up to 59 g/l, hyperbilirubinemia up to 179 mmol/l, including direct bilirubin - up to 90 mmol/l, LDH increase up to 1556 units, cholesterol reduction up to

1.1 mmol/l, while the level of alkaline phosphatase remained not high. Instrumental examination revealed the expansion of the veins of the esophagus of the III st., the presence of multiple portal lymph nodes enlarged to 3 cm.

At each emergency hospitalization, the patient received multiple transfusions of erythrocytes, plasma and albumin, diuretics, hemostatic and antibacteri

al therapy. Against this background, the child's condition has improved significantly. Planned therapy included diuretics, lactulose, propranolol, ursodeoxycholic acid, ornithine, decontaminating antibiotics, iron and erythropoietin preparations, fat-soluble vitamins, famotidine.

Preparation of the patient for transplantation was carried out with considerable difficulties, which was associated with family social problems. In 2017, the child had chickenpox, despite the abundance of rashes, the course of the disease was of moderate severity and did not cause complications (Fig. 3).

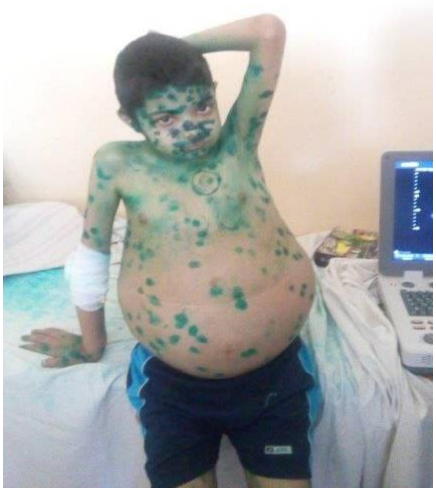

Fig. 3. 2017. Hospitalization for chickenpox.

In November 2017, V.I. Shumakov National Research Center for Transplantology and Artificial Organs performed hepatectomy with preservation of the inferior vena cava, splenectomy and orthotopic liver transplantation from a mortal donor. The early postoperative period was complicated by renal, respiratory, cardiovascular insufficiency, perforation of the small intestine with the development of intra-abdominal bleeding. Immunosuppressive therapy included tacrolimus and mycophenolic acid.

For the period 2017-2022, the patient's condition is stable, there are no complaints, there is a significant improvement in the quality of life: he tolerates physical activity well, copes with the training program (Fig. 4, 5). At 15 years of age, with a height of 158 cm, he weighs 54.5 kg. Indicators of general and biochemical blood tests, acid-base state and coagulogram are normal. The graft function is satisfactory.

The peculiarity of the presented case is that the relatively late clinically manifested hepatic pathology had a rapid progression with the formation of severe complications after 2 years. Periods of decompensation of the condition were associated with exacerbation of cholangitis, and the therapy, up to a certain time, led to a significant clinical improvement in the condition. The presented case demonstrates the reserve capabilities of the child's body and the potential for recovery after liver transplantation, despite the far-reaching complications

The photos are provided by the patient and his mother with consent for publication.

Fig 4,5.

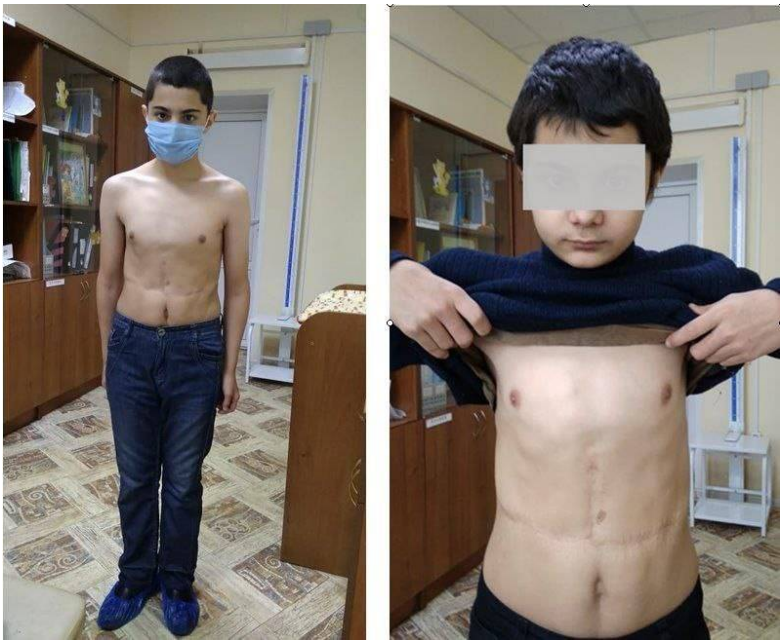

Fig. 4, 5. Condition after transplantation (March 2017).

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