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CLINICAL CASE OF LEDD'S SYNDROME IN ADOLESCENCE

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Abstract. Ledd's syndrome is a common cause of intestinal obstruction in newborns, and in 90% of cases — in children under 1 year. At the same time, diagnostics and surgical treatment in this age period are not difficult. The authors analyzed 15-year experience of treating 51 children of older age groups with Ledd's syndrome and showed that its course in adolescence often has a nonspecific clinical picture, as well as an asymptomatic course, which can cause such conditions as hypotrophy, malabsorption syndrome, constipation, gastroesophageal and duodenogastric reflux and gastroduodenitis. The article presents the experience of treating a child with Ledd's syndrome in adolescence, demonstrating the complexity of diagnosis and treatment of older children.

Keywords: *Ledd's syndrome, Ledd's syndrome in adolescents and adults, diagnostics, surgical treatment*

КЛИНИЧЕСКИЙ СЛУЧАЙ СИНДРОМА ЛЕДДА В ПОДРОСТКОВОМ ВОЗРАСТЕ

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Резюме. Синдром Ледда — нередкая причина кишечной непроходимости у новорожденных, и в 90% случаев — у детей до 1 года. При этом диагностика и хирургическое лечение в этом возрастном периоде не представляет сложностей. Авторами проанализирован 15-летний опыт лечения 51 ребенка старших возрастных грууп с синдромом Ледда и показано, что его течение в подростковом периоде часто имеет неспецифическую клиническую картину, а также бессимптомное течение, что может явиться причиной таких состояний, как гипотрофия, синдром мальабсорбции, запоры, желудочно-пищеводный и дуодено-гастральный рефлюкс и гастродуоденит. В статье представлен опыт лечения ребенка с синдромом Ледда в подростковом периоде, демонстрирующий сложность диагностики и лечения детей старшей возрастной группы.

Ключевые слова*: синдром Ледда, синдром Ледда у подростков и взрослых, диагностика, хирургическое лечение*

INTRODUCTION

Ledd's syndrome (LS) is an uncommon cause of intestinal obstruction in neonates, with a incidence of 0.8:1000 [1, 2]. About 90% of cases of Ledd's syndrome manifest prior to 1 year of age. K. Fang et al. indicate that only 10% of cases of Ledd's syndrome are detected after the first year of life [3, 4]. The incidence of Ledd's syndrome in adult patients ranges from 0.0001 to 0.2%, and about 15% of such people do not have any clinical manifestations for a long period of life [5, 6].

Diagnosis and surgical treatment of Ledd's syndrome as a cause of intestinal obstruction in neonates is not difficult. However, the prolonged asymptomatic course of LS in children and adolescents lead to late diagnosis, and further formation of acute and chronic pathological conditions that threaten the child's life. In this regard, children with LS are often repeatedly hospitalized in various departments (more often — in gastroenterological ones), receive symptomatic treatment and are discharged in a state of compensation without receiving etiotropic treatment. As an intraoperative finding in a number of cases, Ledd's syndrome causes significant difficulties in adopting the correct surgical tactics during surgery [7].

CLINICAL CASE

The clinical case presented in this article demonstrates the complexity of diagnosis and treatment of adolescents with LS.

Patient E., 14 years old, male, came to the clinic with complaints of abdominal pain, irregular stools, nausea and vomiting. It is known from the anamnesis that the child has been having nausea and vomiting periodically since birth. Recently, abdominal pain, constant vomiting, weight loss has been noted. The child has repeatedly received inpatient treatment at the place of residence, as well as in the department of gastroenterology, but with no positive results. He was hospitalized in the surgical department of the clinic. Objective examination showed hypotrophy of the I degree. General condition is satisfactory, in consciousness. The skin is dry, pale pink, without rashes. Subcutaneous fatty tissue is moderately developed, skin turgor and elasticity are preserved. There are no visible swellings on the face. Breathing through the nose, free. The chest participates in the act of breathing, symmetrical. Vesicular breathing is heard above the lungs. Tones are clear, pulse is rhythmic. The mucous membrane of the oral cavity is clean, pale pink, the tongue is moist, without rashes. The abdomen is oval, symmetrical, not bloated, participates in the act of breathing. During the palpation the abdomen is soft, painful around the umbilicus. There is no dulling in flanks. Stools are irregular.

The perianal area is featureless. Tone of anal sphincter is not changed. The ampulla of the rectum is filled with fecal mass of hard consistency. There were no significant changes in clinical and biochemical blood and urine analysis parameters. According to the results of ultrasonography (USG), no signs of gallbladder deformation were revealed. According to esophagogastroduodenofibroscopy (EFGDS) there is insufficiency of esophageal cardia of the I degree. Reflux esophagitis of the I degree. Diffuse gastroduodenitis with hyperaemia of the mucosa. Enlargement of the duodenum. Multislice computed tomography-angiography was also performed, which revealed signs of narrowing of the aortomesenteric angle — compression of the left renal vein between the aorta and the superior mesenteric artery cannot be excluded. Partial small bowel obstruction — an abnormality of bowel rotation and fixation cannot be excluded. The angle of superior mesenteric artery branching is 9°.

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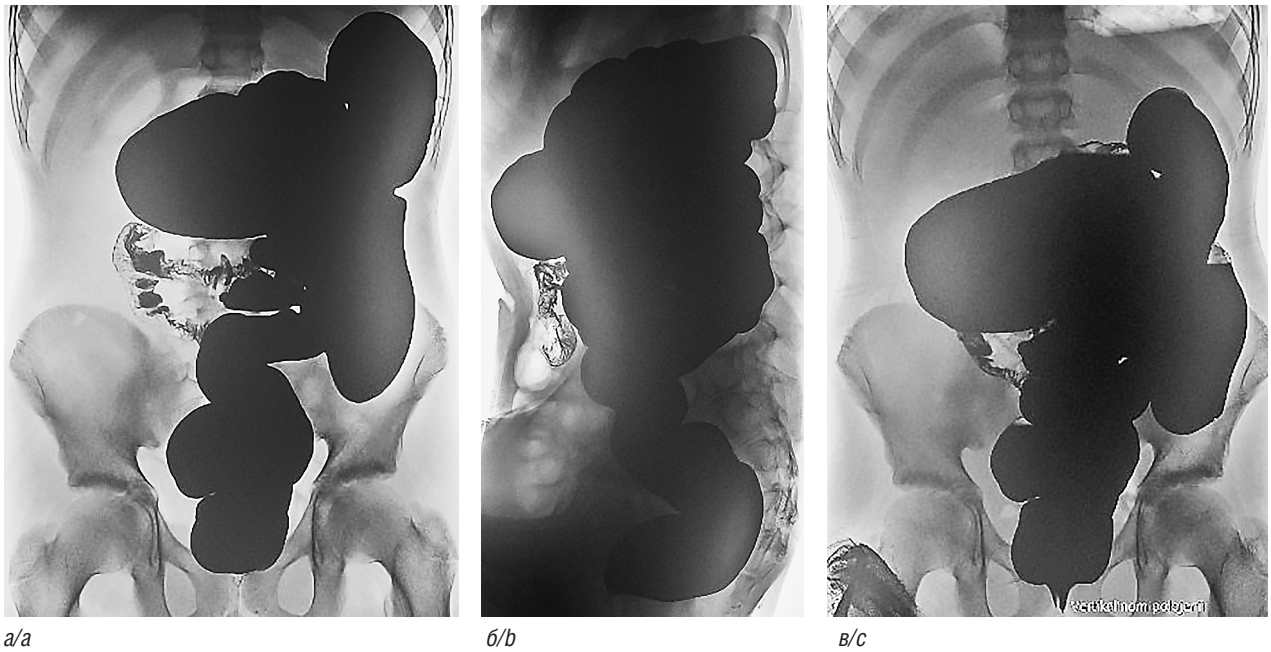
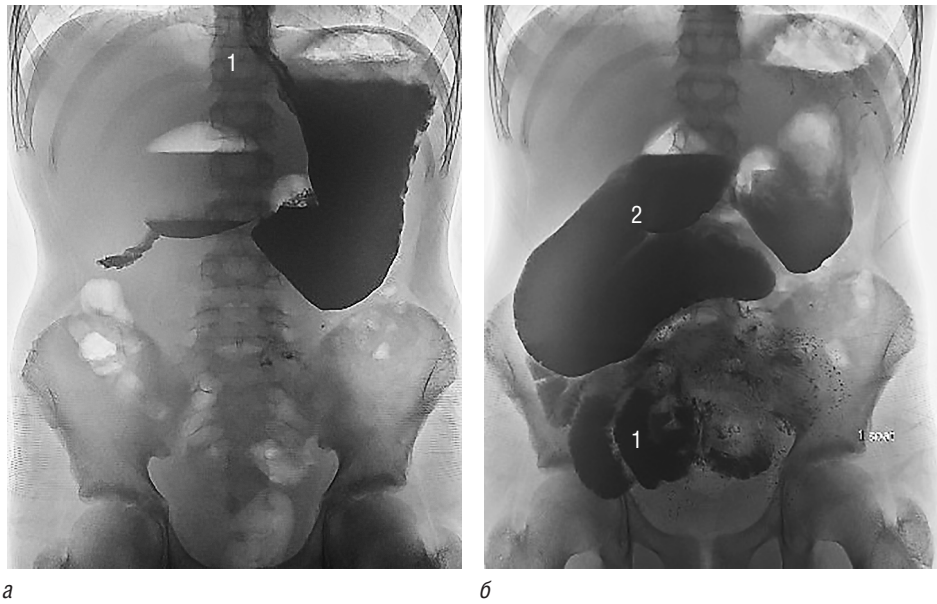


а/a *б/b* *в/c*

Fig. 1. Irrigography. High location (*a*) of the dome of the cecum, median (*b*) and left-sided (*c*) location of all parts of the large intestine

Рис. 1. Ирригография. Высокое расположение (*а*) купола слепой кишки, срединное (*б*) и левостороннее (*в*) расположение всех отделов толстой кишки



а *б*

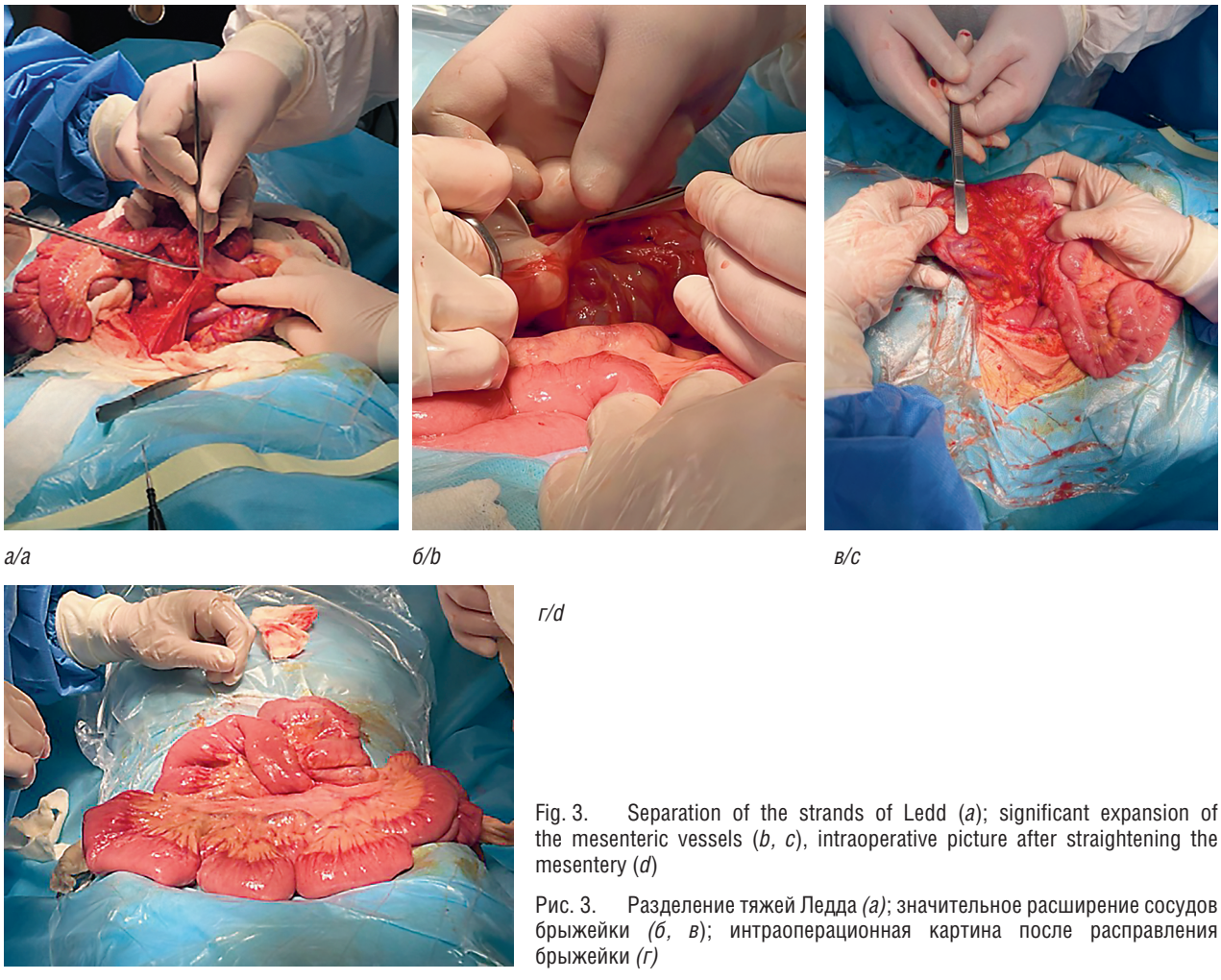
Fig. 2. X-ray contrast study of the upper gastrointestinal tract with barium sulfate (*a*: 1 —gastroesophageal reflux; *b*: 1 — symptom of "spring"; 2 — megaduodenum)

Рис. 2. Рентгеноконтрастное исследование верхних отделов желудочно-кишечного тракта с сульфатом бария (*а*: 1 — желудочно-пищеводный рефлюкс; *б*: 1 — симптом «пружины»; 2 — мегадуоденум)

Irrigography revealed a high location of the cecal dome, predominantly on the left side and medial location of all parts of the colon (Fig. 1).

X-ray contrast study of the upper gastrointestinal tract revealed gastresophageal reflux (Fig. 2, *a*), megaduodenum (Fig. 2, *b*), "spring" symptom (Fig. 2, *b*), reflecting the spiral course of the initial parts of the small intestine, predominantly right-sided location of the small intestine (Fig. 2).

A laparotomy was performed, which revealed Ledd's tracts, small intestine fasciculation, and significantly dilated mesenteric vessels (Fig. 3, *b*). The congenital fetal adhesions were



а/a *б/b* *в/c* *г/d*

Fig. 3. Separation of the strands of Ledd (*a*); significant expansion of the mesenteric vessels (*b*, *c*), intraoperative picture after straightening the mesentery (*d*)

Рис. 3. Разделение тяжей Ледда *(а)*; значительное расширение сосудов брыжейки *(б, в)*; интраоперационная картина после расправления брыжейки *(г)*

separated (Fig. 3, *a*), detorsion and spreading of the pericardium were performed. Intestinal passage was restored on the 3rd day. The child was discharged home on the 7th day.

Over a fifteen-year period we have accumulated experience in treating 51 children with Ledd's syndrome. Obviously, the course of LS in adolescence often has a nonspecific clinical picture, as well as asymptomatic course, which, due to its rare occurrence in children and adolescents, is the cause of pathological conditions of gastroenterological profile (such as hypotrophy, malabsorption syndrome, constipation, gastresophageal and duodenogastric reflux, gastroduodenitis, bulbitis). Children with a gastroenterological complaints, episodes of vomiting with light intervals should undergo a comprehensive examination, including X-ray contrast examination, fibrogastroduodenoscopy, expert ultrasound scanning, and, if a characteristic clinical picture is detected, laparotomy with separation of Ledd's ligaments, detorsion of the ductus and mesenteric spreading should be performed.

ADDITIONAL INFORMATION

Author contribution. Thereby, all authors made a substantial contribution to the conception of the study, acquisition, analysis, interpretation of data for the work, drafting and revising the article, final approval of the version to be published and agree to be accountable for all aspects of the study.

Competing interests. The authors declare that they have no competing interests.

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Consent for publication. Written consent was obtained from legal representatives of the patient for publication of relevant medical information within the manuscript.

ДОПОЛНИТЕЛЬНАЯ ИНФОРМАЦИЯ

Вклад авторов. Все авторы внесли существенный вклад в разработку концепции, проведение исследования и подготовку статьи, прочли и одобрили финальную версию перед публикацией.

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Конфликт интересов. Авторы декларируют отсутствие явных и потенциальных конфликтов интересов, связанных с публикацией настоящей статьи.

Источник финансирования. Авторы заявляют об отсутствии внешнего финансирования при проведении исследования.

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