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Intestinal malrotation in newborns and infants: comparative possibilities of ultrasound and radiologic methods of investigation

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ABSTRACT

BACKGROUND: Intestinal malrotation is a congenital pathology that results from abnormal fixation and rotation of the midgut during fetal development. This pathological condition is most often diagnosed during the first month of life and is often manifested by the presence of regurgitation, restlessness, vomiting with an admixture of bile, low weight gain. The narrow root of the mesentery facilitates the formation of midgut ingestion, which can lead to ischemia and necrosis and requires urgent surgical intervention. Ultrasound and radiologic examination with contrast are currently used to diagnose malrotation.

AIM: To improve the diagnosis of intestinal malrotation in newborns and infants by carrying out a comparative analysis of the capabilities of radiologic and ultrasound methods of investigation when they are used in combination.

MATERIALS AND METHODS: Data from 112 patients aged from 1 day from birth to 3 months 26 days between 2016 and 2024 with ultrasound signs of malrotation detected by microconvex and linear transducers were analyzed. 50 children were further followed up with this provisional or final diagnosis. Contrast agent passages and irrigographs were performed on an AXIOM Luminos DRF (Siemens), with 1 to 15 radiographs and up to 8 X-ray examination series obtained.

RESULTS: The most common ultrasound sign of malrotation was atypical location of mesenteric vessels, and the most common radiologic sign was left-sided location of the colon and high standing of the cecum. Assessment of the location of the duodenojejunal junction was complicated in most cases. Surgery was performed in 17 children, one of them for suspected intestinal obstruction, and a ring-shaped pancreas was found in 4 patients. The diagnosis of malrotation was made in 40 children out of 50, in 4 cases this was discordant with the radiologic findings. Concomitant abnormal location of internal organs was detected in 12 patients.

CONCLUSION: Screening ultrasound examination of all newborns should be considered as the pathology may be asymptomatic. It is important to include suspicion of malrotation in the diagnosis because of the possible manifestation of the pathology at a later age. It is currently not possible to completely abandon radiologic examination with contrast in the diagnosis of malrotation.

Keywords: intestinal malrotation; newborns; children; ultrasound method; radiologic method.

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Мальротация кишечника у новорожденных и детей раннего возраста: сравнительные возможности ультразвукового и рентгенологического методов исследования

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АННОТАЦИЯ

Актуальность. Мальротация кишечника — врожденная патология, которая возникает в результате аномальной фиксации и вращения средней кишки во время развития плода. Данное патологическое состояние чаще всего диагностируется в течение первого месяца жизни и часто проявляется наличием срыгиваний, беспокойством, рвотой с примесью желчи, низкими весовыми прибавками. Узкий корень брыжейки упрощает образование заворота средней кишки, что может вести к ишемии и некрозу и требует срочного хирургического вмешательства. Для диагностики мальротации в настоящее время используется ультразвуковое исследование и рентгенологическое исследование с контрастом.

Цель исследования. Улучшение диагностики мальротации кишечника у новорожденных и детей раннего возраста путем проведения сравнительного анализа возможностей рентгенологического и ультразвукового методов исследования при их комплексном использовании.

Материалы и методы. Проанализированы данные 112 пациентов возрастом от суток с момента рождения до 3 месяцев 26 дней в период с 2016 по 2024 г. с ультразвуковыми признаками мальротации, выявленными с помощью микроконвексного и линейного датчиков. 50 детей далее наблюдались с данным предварительным или окончательным диагнозом. Пассаж контрастного препарата и ирригографии проводились на аппарате AXIOM Luminos DRF (Siemens) с получением от 1 до 15 рентгенограмм и до восьми серий рентгеноскопии.

Результаты. Наиболее частый ультразвуковой признак мальротации — атипичное расположение мезентериальных сосудов, рентгенологический — левостороннее расположение толстой кишки и высокое стояние слепой кишки. Оценка расположения дуоденоеюнального перехода в большинстве случаев оказалась затруднительной. Операция была произведена 17 детям, одному из них по поводу заподозренной кишечной непроходимости, при этом у четырех пациентов обнаружена кольцевидная поджелудочная железа. Диагноз мальротации был поставлен 40 детям из 50, в четырех случаях это разнилось с результатами рентгенологического исследования. У 12 пациентов выявлено сопутствующее аномальное расположение внутренних органов.

Заключение. Следует рассмотреть возможность скринингового ультразвукового обследования всех новорожденных, так как патология может протекать асимптомно. Важно выносить подозрение на мальротацию в диагноз в связи с возможной манифестацией патологии в более позднем возрасте. Полностью отказаться от рентгенологического исследования с контрастом в диагностике мальротации в настоящее время не представляется возможным.

Ключевые слова: мальротация кишечника; новорожденные; дети; ультразвуковой метод; рентгенологический метод.

Как цитировать

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新生儿和婴儿肠旋转不良：超声波和放射学检查方法的可能性比较

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摘要

论证。肠旋转不良是一种先天性病理，是由于胎儿发育过程中中肠固定和旋转异常的结果。这种病症最常在婴儿出生后的第一个月被诊断出来，通常表现为反胃、烦躁不安、呕吐并伴有胆汁，以及体重增长缓慢。肠系膜根部狭窄易形成中肠扭转，从而导致缺血和坏死，需要紧急手术治疗。目前，超声波和造影剂放射学检查可用于诊断肠旋转不良。

研究目的。通过对放射学和超声波检查方法联合使用时的能力进行比较分析，改进对新生儿和婴儿肠旋转不良的诊断。

材料和方法。我们分析了2016年至2024年期间112名患者的数据，这些患者的年龄从出生后1天到3个月26天不等，这些患者使用微凸和线性传感器检测到旋转不良的超声体征。50名患儿在得到这一临时或最终诊断后接受了进一步随访。使用AXIOM Luminos DRF（Siemens）进行了造影剂通道和灌流成像，获得了1到15张射线照片和多达8个透视光片。

结果。肠系膜血管位置不典型是肠旋转不良最常见的超声波征象，而结肠左侧位置和盲肠高位则是放射学征象。在大多数情况下，难以评估十二指肠空肠过渡的位置困难的。对17名患儿进行了手术，其中1名患儿因疑似肠梗阻而进行了手术，并在4名患儿身上发现了环形胰腺。50名患儿中有40名被诊断为肠套叠，其中4名患儿的诊断结果与放射学结果不一致。有12名患者发现内脏器官排列异常。

结论。应考虑对所有新生儿进行筛查性超声波检查，因为病理可能没有症状。由于这种病变可能在较晚的年龄才显现出来，因此在诊断时必须怀疑是否存在肠旋转不良。目前在诊断畸形时还不能完全放弃造影剂放射检查。

关键词：肠旋转不良；新生儿；儿童；超声波方法；放射学方法。

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BACKGROUND

Intestinal malrotation is a congenital malformation caused by abnormal midgut rotation and fixation during fetal development.

The mean incidence of intestinal malrotation is 1 : 500, and clinical manifestations are less common (1 : 6,000). In 80% of cases, intestinal malrotation manifests in the first month of life [1].

Intestinal malrotation is often associated with other conditions such as Hirschsprung's disease, esophageal atresia, biliary atresia, annular pancreas, meconium ileus, intestinal duplications, mesenteric cysts, Meckel's diverticulum, anal atresia, and various urologic abnormalities [2].

Rapidly growing midgut loops normally begin to protrude into the extraembryonic cavity at 4 weeks of gestation; then, the pre- and postarterial portions of the intestine make three counterclockwise wraps around the superior mesenteric artery (SMA) before and after returning to the abdominal cavity, completing a 270° turn by 10 weeks of gestation [3]. Therefore, the duodenojejunal junction is located at the left of the SMA and the ileocecal angle at the right. After the rotation is complete, the ligament of Treitz fixes the mesentery to the posterior abdominal wall, preventing the midgut from wrapping around the narrow base of its mesentery.

Depending on the age of the malformation, several rotational abnormalities may occur, including lack of rotation, incomplete rotation, reverse rotation, hyperrotation, and internal herniation.

Incomplete rotation may result in duodenal obstruction caused by Ladd's bands between the large intestine and mesentery. Narrowing of the mesenteric attachment enables the wrapping of the midgut around the SMA Midgut volvulus (Ladd's syndrome) is a serious complication that occurs when the intestine wraps the SMA clockwise [4]. Patients may experience regurgitation, vomiting with bile, delayed transitional stool passage, enteral feeding difficulty, low weight gain, and restlessness. If the ischemia progresses, bowel necrosis, peritonitis, and hypovolemic shock can develop, leading to high death or disability risk [2].

Reducing radiation exposure to newborns and infants, who are particularly sensitive to ionizing radiation, remains debatable [5].

Several studies have investigated the possibility of diagnosing malrotation by ultrasound alone [6–8]. However, some studies continue to emphasize the importance of using radiography when ultrasound findings are inconclusive, because even one missed case of such a serious condition is harmful [9, 10].

This study aimed to improve the diagnosis of intestinal malrotation in newborns and infants by comparing the advantages of radiography and ultrasound when used in combination.

MATERIALS AND METHODS

Ultrasound and radiography were performed at the Federal Perinatal Center of the V.A. Almazov National Medical Research Center of the Russian Ministry of Health. Patient information from November 2016 to September 2020 was retrospectively studied, and that from September 2020 to May 2024 was prospectively investigated.

Ultrasound data of 112 patients with suspected intestinal malrotation were evaluated. The study included 50 patients, of whom 45 underwent contrast-enhanced radiography in addition to ultrasound, including both irrigography and contrast passage in 21 patients, irrigography in 23 patients, and contrast passage in 3 patients. In two patients, diagnosis was made based on radiographic findings and clinical signs; in one patient, ultrasound did not show the relative position of the vessels (probably normal or difficult to visualize); and in two patients, the position of the mesenteric vessels appeared normal. In two other patients, ultrasound was adequate to indicate emergent surgery based on clinical signs and plain radiography. Moreover, in one patient, intestinal malrotation was detected during repair surgery of a congenital high-grade bowel obstruction.

Malrotation was suspected in 47 patients (94%) based on ultrasound and radiographic findings and in 2 other patients (4%) based on clinical signs and radiographic findings.

The age of the patients ranged from 1 day to 3 months and 26 days at the time of ultrasound if no further contrast-enhanced radiography was performed or at the time of radiography.

Abdominal radiography included unenhanced plain abdominal radiography using AXIOM Luminos dRF (Siemens), followed by contrast passage with the patient in the vertical position and/or irrigography in the horizontal position. Nonionic iodinated water-soluble contrast media diluted at 1 : 1 in saline for irrigography and infant formula, glucose solution, breast milk, or water for passage were used as contrast media. Eight radiographic series were conducted, with 1–15 radiographs per examination.

Abdominal ultrasound was performed with abdominal or neonatal scan modes using a 10–11 MHz neonatal microconvex transducer. B-mode, color Doppler ultrasound, and pulsed wave Doppler ultrasound (as an adjunct modality) were used. Additionally, intestinal loops were evaluated using a 10–12 MHz linear transducer.

FINDINGS AND DISCUSSION

The topography of the large intestine was assessed during irrigography. Scanning of the upper gastrointestinal tract primarily focused on the location of the duodenojejunal junction in relation to the midline (the shadow of the spine).

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Tables 1 and 2 show the radiographic signs of intestinal malrotation and their incidence in patients.

Furthermore, they reveal that the most common radiographic signs were left-sided large intestine (30%) and high-riding cecum (39%) (Fig. 1).

In eight cases (18%), the left-sided large intestine was combined with an atypical cecum position (high-riding or high- and medial-riding) (Fig. 2).

This sign should be compared with other clinical and diagnostic findings, as statistics show that in 20% of cases, the cecum is normally positioned in children with malrotation, whereas in normal newborns, the cecum position varies [4].

In most cases, duodenojejunal junction position is difficult to assess during contrast study because it is shadowed by the contrast medium in the stomach and the rotation of the newborn's body should be considered, making it impossible to generate reliable conclusions about its atypical location (Fig. 3).

In two patients, ultrasound did not detect malrotation. In one patient, the relative position of the mesenteric vessels appeared typical (Fig. 4), whereas in the other patient, only the presence of fluid and dilated intestinal loops could be determined.

In one patient, the relative position of the mesenteric vessels could not be assessed owing to shadowing by intestinal loops. However, the predominant right position of the small intestine and left hypochondrial position were demonstrated, which did not allow to rule out intestinal malrotation. In one case, ultrasound showed high intestinal obstruction.

A whirlpool sign, also called the whirl sign (Fig. 5), which is wrapping of the mesentery and superior mesenteric vein (SMV) around the SMA, was the main ultrasound finding that demonstrated presence of a midgut volvulus. This sign was partially or completely observed in three of four patients intraoperatively diagnosed with Ladd's syndrome.

In three cases, this condition was successfully treated with the Ladd's procedure. In another case, the abdominal strands were dissected, the mesentery of the small intestine was straightened, and the ascending colon was fixed in the right flank of the abdomen. The colon was not placed in the left flank because of the proper fixation of

Table 1. Frequency of occurrence of radiologic signs of malrotation during irrigography (*n* = 44)

Таблица 1. Частота встречаемости рентгенологических признаков мальротации при проведении ирригографии (*n* = 44)

Radiographic signs of intestinal malrotation	Patients	
	No.	%
Left-sided large intestine	13	30
High riding cecum	17	39
High and medial riding cecum	8	18
Left-sided high riding cecum	3	7
Atypical location of the large intestine	6	14
Location of the cecum in the left iliac region	1	2
Normal	8	18
Total number of abnormal locations of the large intestine	36	82
Total number of children examined	44	100

Table 2. Variants of duodenojejunal junction location during contrast passage in children with suspected malrotation (*n* = 24)

Таблица 2. Варианты расположения дуоденоеюнального перехода при проведении пассажа контрастного препарата у детей с подозрением на мальротацию (*n* = 24)

Location of duodenojejunal junction	Patients	
	No.	%
On the right side of the spine	9	38
At the midline	2	8
On the left side of the spine (normal)	4	17
Lack of clearly visualized junction location	13	54
Total identified signs of atypical location of the duodenojejunal junction	11	46
Total number of children examined	24	100

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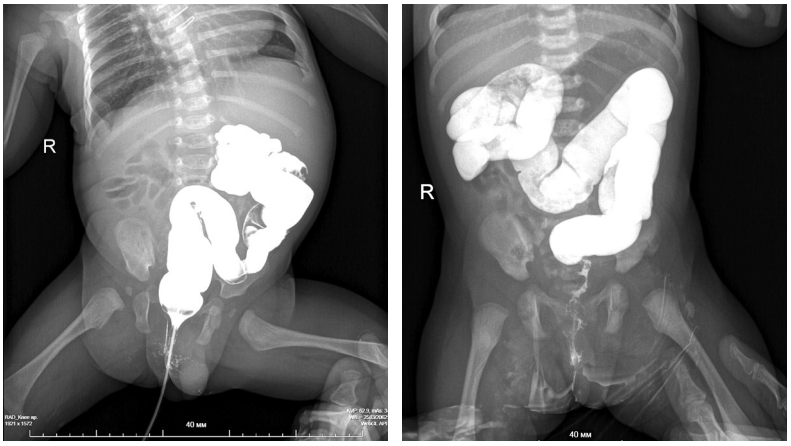

Fig. 1. Left-sided location of the colon (left), high location of the cecum (right) in neonates, irrigography
Рис. 1. Левостороннее расположение толстой кишки (слева), высокое расположение слепой кишки (справа) у новорожденных, ирригография

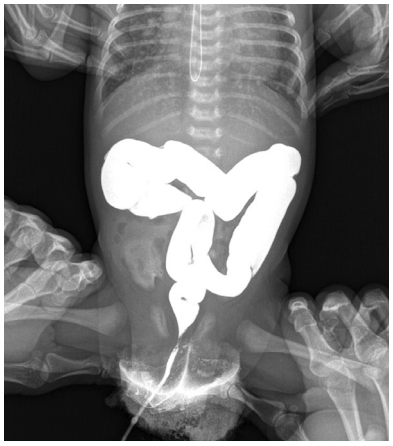

Fig. 2. High and medial position of the cecum in a newborn, irrigography
Рис. 2. Высокое и срединное положение слепой кишки у новорожденного, ирригография

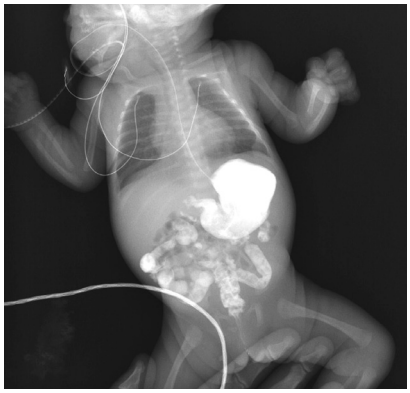

Fig. 3. Duodenojejunal junction probably located to the right of the left vertebral body pedicle, fluoroscopy
Рис. 3. Вероятно расположенный правее левой ножки тела позвонка дуоденоеюнальный переход, рентгеноскопия

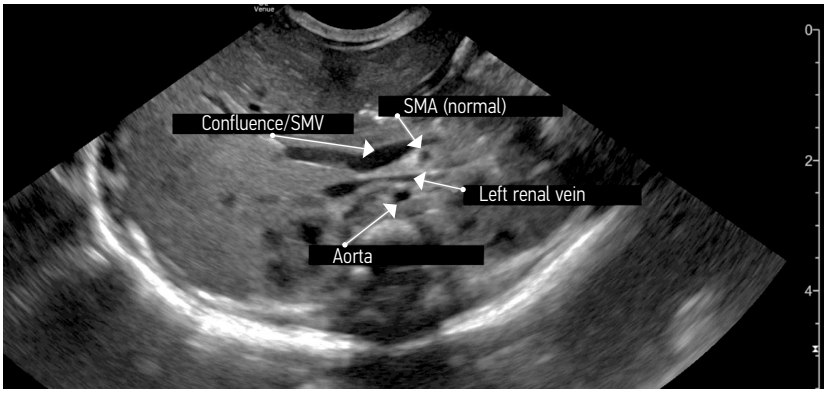

Fig. 4. Normal location of mesenteric vessels in a newborn, B-mode
Рис. 4. Нормальное расположение мезентериальных сосудов у новорожденного, В-режим

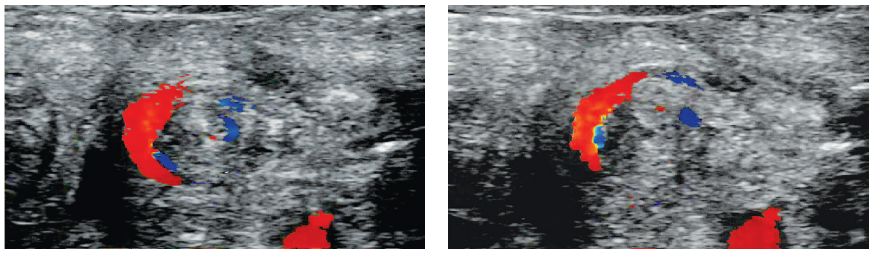

Fig. 5. Whirlpool sign, or whirlpool symptom, detected in a newborn, color Doppler imaging
Рис. 5. Whirlpool sign, или симптом водоворота, выявленный у новорожденного, цветовое допплеровское сканирование

the transverse colon and Treitz ligament. In other cases, the atypical vessel arrangement consisted mainly of a right-sided arrangement of the artery relative to the vein (Fig. 6).

Table 3 presents other ultrasound signs that indicate intestinal malrotation.

In two patients, diagnosis of intestinal malrotation was based only on clinical signs and ultrasound findings. In the first patient, the SMV wrapped incompletely around the SMA, and only regurgitation syndrome was clinically observed. Contrast-enhanced radiography was not performed owing to the serious condition of the patient, who had multiple congenital developmental anomalies, including a heart defect, and underwent two heart surgeries. The patient died at 2 months old. All findings revealed congenital gastrointestinal malformations, and malrotation syndrome was included in the diagnosis as a suspected condition.

The second patient had malrotation with volvulus. Ultrasound showed that the SMV was wrapped around

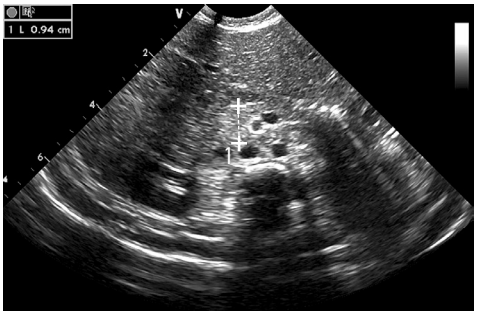

Fig. 6. Atypical location of mesenteric vessels: the artery (vessel with a thick, hyperechogenic wall) is located dorsally, to the right; the vein (vessel with a thin wall) — ventrally and to the left
Рис. 6. Атипичное расположение мезентериальных сосудов: артерия (сосуд с толстой, гиперэхогенной стенкой) расположена дорзальнее, правее; вена (сосуд с тонкой стенкой) — вентральнее и левее

the SMA. Regurgitation, absence of stool from the third day of life, vomiting, and stagnant stomach contents were observed. Ladd's procedure was employed to treat uncomplicated midgut volvulus and duodenal stenosis by peritoneal strands.

One of the surgeries was performed for high intestinal obstruction; ultrasound showed a dilated duodenum, indicating an atresia (Fig. 7). The surgery was performed without prior additional radiography. Examination revealed that the head of the cecum was highly attached to the duodenum by embryonic strands, the small and large intestines had a common mesentery, the intestine collapsed, and the mesentery was twisted by 180°. The embryonic strands were bluntly and sharply divided. When the duodenum was isolated from the strands, a ring-shaped pancreas was noted, which fixed and narrowed the lumen of the intestine.

With adequate experience, the third segment of the duodenum can be visualized using a 10–12 MHz linear transducer. In most cases, specialists do not describe the third segment in the protocol if it was visualized in the normal position and if the mesenteric vessels were not atypically arranged. If the vessels are atypical, the third segment is almost never located in the aorto-mesenteric space, and specialists do not report it. The third segment is described in the protocol if it is visualized in a normal position with an atypical vessel arrangement, as this is not logical.

In 30 patients (60%), ultrasound and radiographic signs were incidental findings of abdominal examination for other disorders, as the patients exhibited no clinical signs of gastrointestinal tract disorders. In another 20 patients, symptoms indicating gastrointestinal disorders included regurgitation, constipation, bloating, poor weight gain, and ineffective feeding.

In 2017–2024, 17 of 50 children (34%) underwent surgeries to repair rotation defects and/or fix the midgut. In four patients (24%), a ring-shaped pancreas was detected during surgery.

Of the 50 patients examined, malrotation was confirmed in 40 (80%) and suspected in 6 (15%) patients. One of the patients showed no radiographic signs of

Table 3. Frequency of ultrasound signs of malrotation in diagnosed or suspected congenital pathology (n = 48)
Таблица 3. Частота встречаемости ультразвуковых признаков мальротации с диагностированной врожденной патологией или с подозрением на нее (n = 48)

Ultrasound signs of malrotation	Patients	
	No.	%
Atypical arrangement of mesenteric vessels (SMA located to the right of SMV)	35	73
Incomplete wrapping (circular motion) of the SMV around the SMA	6	13
Whirlpool sign	1	2
Inverse arrangement of SMA and SMV	2	4
SMV crosses SMA	1	2
Atypical location of the third segment of the duodenum	1	2
The right-sided location of the small intestine and the left hypochondrial location of the large intestine	1	2
No changes	1	2

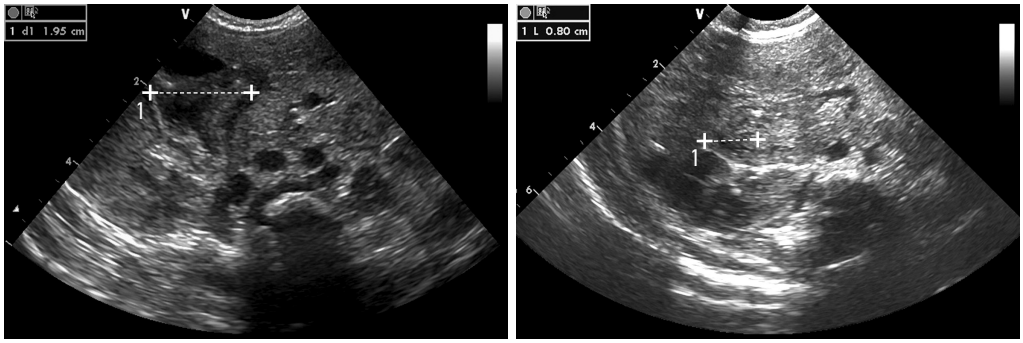

Fig. 7. Example of duodenal dilatation in a newborn on ultrasound, B-mode (left). Second segment of the duodenum in a healthy newborn (right)
Рис. 7. Пример расширения двенадцатиперстной кишки у новорожденного на УЗИ, В-режим (слева). Второй сегмент двенадцатиперстной кишки у здорового новорожденного (справа)

Table 4. Ultrasound data for calculating the sensitivity and specificity of the method
Таблица 4. Частота встречаемости ультразвуковых признаков мальротации с диагностированной врожденной патологией или с подозрением на нее (*n* = 48)

Result	Patients	Healthy subjects
Positive test result	37	75
Negative test result	0	1

malrotation, and three who had such signs were not diagnosed with this condition, as well as the patient whose malrotation was discovered during surgery for intestinal obstruction.

Twelve patients (24%) were found to have concomitant abnormal positioning of internal organs (*situs ambiguus* and *situs inversus*), ten of whom (83%) were diagnosed with malrotation.

At discharge, most patients (78%) had favorable prognoses. However, six patients (12%) were disabled and five patients (10%) died.

Of 112 patients with ultrasound signs of malrotation, 37 (33%) were diagnosed based on clinical signs and radiographic and/or ultrasound findings. The sensitivity of ultrasound was 100%, whereas the specificity was 1.3% (Table 4).

These findings do not reflect the real diagnostic value of ultrasound, as currently, most patients with suspected intestinal malrotation undergo irrigography. However, children may be underexamined, and intestinal malrotation may not be diagnosed if irrigography is not performed accurately (the intestine is partially filled with contrast medium) and if the patient manifests no clinical signs of intestinal obstruction.

CONCLUSION

It is reasonable to consider a screening abdominal ultrasound with assessment of the relative position of the mesenteric vessels as a part of the routine examination of newborns at the age of 1 month as many infants show no clinical signs of intestinal malrotation. A specialist should evaluate these structures and recommend consultation with a pediatric surgeon if required. If an intestinal malformation is diagnosed or suspected, it should be communicated to the child's parents and recorded, as this information may be beneficial in making a differential diagnosis in cases of later manifestation of symptoms.

Based on ultrasound findings, a group of patients should be formed for further radiography; however, the diagnosis can be confirmed only after a comprehensive examination. Diagnostic examinations should be performed accurately (i. e., correct positioning of the patient and complete filling of the large intestine with contrast medium). Therefore, despite the aim to reduce radiation exposure to pediatric patients, which should be achieved primarily by reducing the number of radiographs and fluoroscopy series, it is not currently possible to completely avoid contrast-enhanced radiography in the diagnosis of serious conditions.

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