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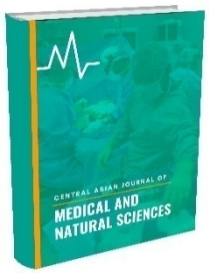

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Ultrasound Diagnosis of Hirschsprung's Disease in Children

1. **Akhmedov Yakub Amandullaevich**
2. **Juraev Kamoliddin Danabaevich**

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[1,2] Samarkand State Medical Institute, Department of Medical Radiology, Republic of Uzbekistan, Samarkand

Abstract: The use of ultrasound diagnostics reduces the number of uninformative, invasive and radiation-related procedures. The objectivity of the results of the ultrasound structure of the intestinal wall depends on many subjective parameters: on the device on which the examination is carried out, on the experience of a specialist, on the degree of bowel preparation, on the contrast solution used, on the severity of the urge to defecate when filling the large intestine.

Keywords: ultrasound diagnostics, Hirschsprung, colon, disease, congenital malformation.

Introduction. Hirschsprung's disease is a congenital malformation of the colon, in which the laying of intramural nerve ganglia is disturbed in embryogenesis. The incidence of this defect is 1 in 5000 newborns.

High and low forms of defect are distinguished, while dividing the sections of the colon according to the degree of damage. The clinic is largely determined by the length of the aganglionic segment: the longer this affected area, the faster the picture of intestinal obstruction grows. With short zones of agangliosis, most authors note the smoothness of clinical symptoms. Constipation does not occur immediately after birth, but at the age of 2-3 years, and even later.

The problem of treating Hirschsprung's disease still remains relevant due to the controversy not only of the operative manual, but also the lack of an algorithm for postoperative rehabilitation. None of the used surgical techniques are complete without complications and are characterized by traumatism, severe postoperative and long recovery period. Due to the development of new medical technologies in recent years, minimally invasive endoscopic operations, which are used in coloproctology, are widely introduced into surgical practice. But, in spite of this, a frequent complaint of patients and their parents after surgical treatment in catamnesis is caloamation and encopresis. Surgeons associate this complication with cicatricial changes in the anorectal area or damage to the sphincters.

Encopresis and kalomazanie - functional disorders, which, although not directly threatening the patient's life, but are undoubtedly socially significant, leading to a limitation of the patient's mental and physical activity, complicating his social adaptation in society, leading to problems in communicating with peers, lag in study, conflict situations in the family and school.

Known methods for diagnosing Hirschsprung's disease include examination of the colon with a focus on clinical, rectal, radiographic, and biopsy of the intestinal mucosa.

X-ray examination and biopsy for acetylcholinesterase of the colon wall remain the only methods confirming Hirschsprung's disease.

The anorectal zone is currently examined only by functional methods. In this case, electrocolography, profilometry of rectal pressure, study of the motor function of the rectum, internal and external anal sphincters, electromyography of these sphincters and determination of the recto-anal reflex are used. However, the results may not be reliable due to technical errors and concerns of young children.

An important task facing coloproctologists and gastroenterologists is the development of informative, accurate, evidence-based methods for the study of patients with Hirschsprung's disease before and after surgical treatment, allowing timely selection of adequate restorative therapy and improving their quality of life.

The main methods for diagnosing Hirschsprung's disease at present are: X-ray contrast irrigography, ultrasound diagnostics of the colon, functional diagnostics and mucosal biopsy to determine the activity of acetylcholinesterase. All survey methods complement each other.

The classic version of irrigography makes it possible to assess the anatomical and topographic position of various parts of the large intestine, their length, diameter, and estimate the puborectal angle. The disadvantages of this method are:

- radiation load on the child's body, which excludes the possibility of frequent repeated examinations (once a year);
- with retrograde forced administration of a contrast agent, the response from the smooth muscle structures of the colon does not always allow one to judge its true anatomical and functional state;
- when examining the rectoanal zone, only the anorectal angle is measured, which is not enough to assess the state of the entire colon;
- irrigography allows to reliably reveal the picture of megarectum, dolichosigma, megacolon, but not answer the question about the etiology of these changes
- in patients with short zones of agangliosis, irrigography can diagnose Hirschsprung's disease in only 75% of cases.

Also, there is a known method for diagnosing Hirschsprung's disease by the method of ultrasound examination of the colon - hydroechocolonography. The use of ultrasound diagnostics reduces the number of uninformative, invasive and radiation-related proceduresIvl. However, the objectivity of the results of the ultrasound structure of the intestinal wall depends on too many subjective parameters: on the device on which the examination is carried out, on the experience of a specialist, on the degree of bowel preparation, on the contrast solution used, on the severity of the urge to defecate when the colon is filled.

Thus, the known diagnostic methods do not make it possible to conclusively accurately assess the nature of colon disorders.

Currently, there are no reliable evidence-based data on the state of the anorectal region in children with Hirschsprung's disease, which determines the urgency of this problem and further attempts to effectively treat it.

The key factor for early diagnosis, timely and adequate treatment of Hirschsprung's disease is the assessment of the anatomical and topographic parameters of the colon, anorectal zone and pelvic floor. For the objectivity of such an assessment, dynamic research is needed.

In recent works, to study the anatomical and functional state of the muscles and organs of the small pelvis, it is proposed to use transperineal ultrasound scanning, which is used along with conventional

research methods. The conduct of such a study in Hirschsprung's disease is not described in the modern literature.

Thus, the goal of our work is to create a non-invasive, informative and objective method for diagnosing Hirschsprung's disease in children, which allows us to give recommendations for adequate effective treatment.

Material and methods. The authors have developed a technology based on precise objective qualitative and quantitative criteria obtained from the results of transabdominal and transperineal echography and characterizing the anatomical and topographic signs of the colon and anorectal zone: the location of the intestine, the fecal mass, the severity of the haustra, the diameter of the wall and muscle, mucous layer colon, the state of the zone of agangliosis and anastomosis, the diameter of the rectum, the state of the mesanterial lymph nodes; the ratio of the length and width of the anal canal, the value of the anorectal angle, the thickness of the external and internal sphincters, the thickness of the puborectal muscle, the blood flow velocity in the branches of the rectal arteries, the presence of changes in the puborectal angle during the functional Valsalva test, reflecting the functional or organic nature of the process.

As a result of evaluating a large statistical material, the authors suggested and unexpectedly found that in Hirschsprung's disease, not only the large intestine is affected, but also the anorectal zone, which makes it possible to increase the information content and accuracy of the method and carry out radical treatment.

The key point in the diagnosis is the assessment of the anatomical and morphological characteristics of the anorectal angle and puborectal loop during the functional Valsalva strain test.

Consecutive carrying out of transabdominal scanning in longitudinal and transverse projections and transperineal scanning in longitudinal and longitudinal-oblique projections allows optimal visualization of the examined organs and increase the information content and objectivity of diagnosis.

In the process of carrying out transabdominal and transperineal studies in children, the authors were guided by the criteria of the norm developed by them on the basis of statistical processing of a large number of patients.

The criteria for the norm of the following indicators in children:

- length of the anal canal - 17.9-20.1 mm,
- width of the anal canal - 13.1-15.2 mm,
- thickness of the internal sphincter - 2.5-2.8 mm,
- thickness of the external sphincter - 2.5-2.8 mm,
- the value of the anorectal angle - 83.9-88.6 *,
- thickness of the puborectal loop - 4.5-4.9 mm,
- blood flow velocity in the vessels of the anorectal zone
- (branches of rectal arteries) - 6.3-7.1 cm / sec,
- the diameter of the rectal ampulla - 30-33 mm,
- thickness of the colon wall - 2-2.5 mm,
- thickness of the muscle layer of the colon wall - 0.5-0.7 mm
- thickness of the mucous layer of the colon wall - 1 mm

The essence of the invention lies in the fact that a method for diagnosing Hirschsprung's disease is characterized by the fact that a pediatric patient with a symptom of persistent constipation after general clinical and special examinations is sequentially carried out ultrasound transabdominal examination of the colon, including the distal part of the rectum, and transperineal examination of the anorectal zone, transabdominal scanning of the colon the intestines are carried out natively without preliminary preparation of the patient in his position on the back in two perpendicular projections - longitudinal and transverse, gaustra are taken as reference points for visualization, while their severity is determined, the thickness of the intestinal wall, its muscle and mucous layers is measured, the state of the aganglionic zone is assessed when it is detected in the descending part of the colon, its length is measured, the diameter of the rectal ampoule, transperineal studies of the anarectal zone are carried out in the patient's position on the left side with legs bent at the joints and, the sensor is installed in the anal fossa, cmwhile scanning it from the bosom to the sacrum in the process of scanning in two projections - longitudinal and longitudinal-oblique, focusing on the sacrum, symphysis, anal canal for visualization, the state of the distal ampulla of the rectum, the aganglionic zone when it is detected in this section is assessed in detail echographic anatomy of the anal canal: length and width, determine the presence and safety of the internal hypoechoic and external hyperechoic anal sphincters, measure their thickness, use pulsed dopplerometry to assess the blood flow velocity in the branches of the rectal arteries, measure the value of the anorectal angle, at the border of the transition of the rectal ampulla into the anal canal the sensor is placed in a longitudinal-oblique projection, the branch of the puborectal loop is determined, its thickness is measured, the echogenicity and structure are assessed at rest of the patient, then a functional Valsalva test with straining is performed, while determining the thickness of the puborectal loop and the presence of changes in the anorectal angle, the obtained indicators are compared with the criteria of the norm, and in the presence of changes from the colon to the aganglionic zone: the thickness of its wall is more than 2.5 mm, the thickness of its muscle layer is more than 0.7 mm, and the thickness of the mucous layer is more than 1 mm, there is no muscle layer in the aganglionic zone itself , lack of expression of haustra, expansion of the diameter of the rectal ampulla more than 30 mm, lack of differentiation and increased echogenicity in the aganglionic zone detected in the descending colon or distal rectal ampulla, as well as in the presence of changes from the anorectal zone: a decrease in the thickness of the outer and internal sphincters of the anal canal less than 2.5 mm, no fixation of blood flow velocity values in the branches of rectal arteries, an increase in the thickness of the puborectal loop more than 4.9 mm and an increase in its echogenicity, an increase in the anorectal angle of more than 100 degrees at rest and in the absence of its change during the Valsalva test, the presence of hypertrophy of the muscle layer of the proximal colon, dysfunction of the puborectal loop and muscles of the pelvic floor, hypoplasia of the sphincters is judged, and Hirschsprung's disease is diagnosed.

The proposed method has passed clinical trials at the Izmailovo Children's City Clinical Hospital in Moscow in the Department of Coloproctology and Gastroenterology.

A total of 50 children aged from 1 month to 17 years old with complaints of persistent chronic constipation were examined.

All those admitted to the hospital with complaints about the absence of independent acts of defecation underwent a comprehensive examination: general clinical and special studies, including radiography, sigmoidoscopy with biopsy and ultrasound.

All children underwent a study on the proposed method, according to the results of which 50 children with suspected Hirschsprung's disease were diagnosed with Hirschsprung's disease, taking into account X-ray data and biopsy results.

As a result of transabdominal examination, all 50 children revealed: hypertrophy of the muscular layer of the proximal intestine to the zone of agangliosis, the very zone of agangliosis, lack of expression (smoothness) of haustra, expansion of the rectal ampulla more than 30 mm.

Transperineal scanning revealed in all children: hypoplasia of the external and internal sphincters, an increase in the anorectal angle and thickening of the puborectal loop and its increased echogenicity.

The functional test of Valsalva, which was carried out in children after seven years of age, was negative in 15 children.

All children underwent radical surgery.

In the postoperative period, there were no complications in the form of kalomazaniya and encopresis.

Discussion and results. For the first time, the authors established that in Hirschsprung's disease (before surgery), there is a thickening of the walls of the colon to the zone of agangliosis, the diameter was 3-5 mm, while the muscle layer is 2-3 mm, the mucous layer is 2-3 mm. The gaustra of the large intestine are smoothed. With changes in the wall of the colon, there is an increase in the mesanterial lymph nodes. The area of agangliosis is represented by the intestinal wall of increased echogenicity, without a clear differentiation of anatomical layers.

With transperineal scanning, the anatomotopographic characteristics change. From the side of the rectum, an expansion of the rectal ampulla is revealed, a decrease in the external and internal sphincters, a thickening of the puborectal loop and an increase in its echogenicity, as well as an increase in the anorectal angle, are noted, while the length and width of the anal canal are not changed.

It has been established that, normally, during the Valsalva maneuver with straining, the anorectal angle decreases to 5-10 degrees. In children with Hirschsprung's disease, this test is negative: there is no contraction of the puborectal loop and, as a consequence, a change in the anorectal angle.

Example 1. Boy N. 1 month. Was admitted with complaints of stool retention up to 7 days. Defecation acts only after cleansing enemas. He was admitted for examination at the Department of Coloproctology. Irrigorgafia revealed distal agangliosis. Manual examination revealed a narrowing of the anal canal at the height of the finger. Conducted ultrasound transabdominal and transperineal research according to the proposed method.

Transabdominal examination was performed with the boy supine to visualize the large intestine in transverse and longitudinal projections. Then, sequential parallel studies of the rectum and perineal zone were performed with a detailed assessment of the anatomy anal canal, the state of the rectal ampulla. In this case, the child was on his left side with legs bent at the knee and hip joints. The sensor is installed directly into the anal fossa. Scanning was carried out in two perpendicular projections: longitudinal and longitudinal-oblique. At the same time, the location of the large intestine, the fullness of the lumen, the severity of the haustra, the thickness of the wall, muscle and mucous layers of the colon were determined, the area of agangliosis was assessed, its length was measured, the state of the mesanterial lymph nodes, the blood flow of the rectal arteries, the diameter of the rectal ampulla.

When examining the anal canal, its length and width were measured. The presence and safety of anal sphincters: internal and external, was determined, their thickness was measured. The anorectal angle was measured. At the border of the transition of the rectal ampulla into the anal canal, the sensor was placed in a longitudinal oblique projection. The branch of the puborectal loop has been identified. Its thickness was measured, its echogenicity and structure were assessed.

A functional test was not performed on this child due to his young age.

Transabdominal examination revealed: the descending section of the transverse colon is lateralized to the right, heterogeneous contents in the lumen, the haustra are smoothed, the intestinal wall is thickened to 4 mm, the muscular layer is up to 2 mm, the mucous membrane is 1.5 mm, mesanterial nodes were not visualized. The ampulla of the rectum was enlarged to 45 mm.

Transperineal scanning revealed: the length and width of the anal canal is not changed. The thickness of the external and internal sphincter is reduced to 1.3 mm. The anorectal angle was 110, the puborectal loop was increased to 6 mm, increased echogenicity. The blood flow was not determined. The area of agangliosis was 20 mm, which passed into the anal canal.

As a result, the presence of hypertrophy of the muscle layer of the proximal colon, dysfunction of the puborectal loop and pelvic floor muscles, hypoplasia of the sphincters, and Hirschsprung's disease (low form of agangliosis) were diagnosed.

The child was operated on laparoscopically according to the Soave method. The postoperative period was uneventful. The child was discharged on the 10th day in a satisfactory condition. No complications were found in the catamnaz.

Example 2. Boy G. 3 years old. Was admitted for surgical treatment at the Department of Coloproctology. Operated for large bowel obstruction in 1 year - a colostomy was applied (in the ascending section of the transverse colon) at the place of residence. Irrigography and bowel biopsy diagnosed Hirschsprung.

In laboratory analyzes, inflammatory changes were not revealed: scatology and analysis of feces for dysbiosis are normal.

A transabdominal examination revealed: the topography of the intestine was not changed, the descending sections were emptied, the haustra were smoothed, there was a thickening of the intestinal wall to the zone of agangliosis by 5 mm: the muscle layer was up to 2 mm, the mucous membrane was up to 3 mm. Mesanterial nodes were identified multiple up to 10 mm, without changes in echo structure. The area of agangliosis was 30 mm.

With transperineal access, the length and width of the anal canal are not changed. The thickness of the external and internal sphincter was reduced to 1.5 mm. The anorectal angle was 111 degrees, the thickness of the puborectal loop was increased to 6.5 mm, with increased echogenicity. The blood flow of the rectal arteries was not recorded.

Clinical diagnosis: Hirschsprung's disease, colostomy carrier.

The child was operated on laparoscopically using the Soave method. The postoperative period was uneventful. The child was discharged on the 10th day in a satisfactory condition. Complications in the late postoperative period were not observed.

Thus, the method made it possible to make an accurate diagnosis, to identify all the diagnostic signs of the disease due to high accuracy and information content, and to carry out radical treatment.

The absence of postoperative complications confirms the effectiveness of the proposed method for diagnosing Hirschsprung's disease.

The test results proved the high accuracy, information content and effectiveness of the method, which made it possible to perform a radical operation and exclude postoperative complications, which makes it possible to recommend it for wide use in medical institutions on devices of both expert and middle class as a diagnostic method for diagnosing children with Hirschsprung's disease without the use of invasive research methods.

Conclusions. A highly effective pathogenetically substantiated method for diagnosing Hirschsprung's disease before and after treatment in children is proposed, based on precise objective criteria obtained from the results of transabdominal and transperineal echography.

The method is informative, it has high accuracy and objectivity, it allows on the basis ofprecise quantitative and qualitative criteria to assess the condition of the colon throughout its entire length and the anorectal zone, identify and verify anatomical abnormalities and prescribe adequate therapy.

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