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THE CONTEMPORARY APPROACHES TO SURGICALLY TREATING HIRSCHSPRUNG'S DISEASE IN CHILDREN

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Abstract: This study presents recent literature on the current surgical treatment of Hirschsprung's disease in children. Despite the numerous literature data on Hirschsprung's disease, the problems of diagnosis and treatment remain relevant. Analysis of the literature data showed that these problems can be solved by developing an examination program, taking into account all the existing complications and determining the appropriate treatment tactics.

Keywords:Hirschsprung'sdisease,treatment, children.

Relevance. Hirschsprung's disease (HD) is associated with impaired localization and migration of ganglion cells in the intestinal tube during intrauterine development. Due to a failure to relax the distal ganglionic region of the intestine and to participate in normal peristalsis, such patients suffer from congenital constipation [1,2,4,6,11]. Typically, in patients with Hirschsprung's disease, meconium excretion is delayed at 24-48 hoursof life. In most cases, thefaeces appearafteranorectal stimulation. If the condition is left untreated, chronic constipation, dysbiosis, enterocolitis,transmural bacterial translocation and sepsis develop. For these reasons, a high degree of cautionwith regard to BG is particularly important in the condition of congenital constipation [9].

It should be noted that, to date, the incidence of the disease in Uzbekistan has not been identified. TheincidenceofmalformationshasbeenreportedbyauthorsfromCIScountriesataratioof1:2,000 to 1:30,000 to the number of newborns [3,5]. According to Rosstat, in children under14 years ofage, the incidence of first-time detected congenital malformations (CHD) in 2015 was 277,900 (a ratio of 1,154.8 per 100,000 children) [2]. Theincidence of HD in the structureof CHD has not been established. According to the European association of CHD registries (2011- 2015), the incidence of HD is 404 per 10,000 newborns [9].

Confirmation of the diagnosis of Hirschsprung's disease is an indication for surgical treatment. Advances in neonatal care, anaesthesia and resuscitation over the last decade, have allowedpaediatric surgeons to remove total aganglionosis and prevent a mild variant of Hirschsprung's disease in one step. The diagnosis is often made in the neonatal period. Manycentres use a one-stage correction with excellent results. The main obstacles to transanal endorectal proctoplasty are malformations of severe vital importance, severe enterocolitis, marked dilatation of the proximal bowel and somatic disorders. In this intervention, the aganglionic bowel is lowered transanally and resected 10 cm above the transition zone. A coloanal anastomosis is inserted between the ganglionic anus (14,18). In cases of extended ganglionic zone, laparoscopic mobilisation of the colon and its transanal endorectal proctoplasty may be used [7,10]. In the presence of enterocolitis, water- electrolytic equilibrium is first stabilised and intensive treatment, including purging enemas, is carried out [10,13]. In complicated forms of Hirschsprung's disease (intestinal obstruction, necrotizing enterocolitis with toxicdilatation ofthecolon, perforation of theterminal small intestine, other parts of the cecum or large intestine), biopsies are taken at different levels of the colon and a stoma with rectal biopsy is taken. Once the diagnosis of Hirschsprung's disease has been made, and the

aganglionic region of the intestine has been identified, the type of surgical treatment is determined [22]. The intestinal type of agangliosis is a serious omen. An upper small bowel stoma will inevitably cause short bowel syndrome and a dubious prospect after radical surgery [8]. Carefully considered technique, adequate hemostasis, good blood supply to prevent ischaemia, avoidance of intestinal entanglement and distension should prevent complications during transanal endorectal descending.

Another serious complication is the formation of scar stenosis after radical lowering. The rate of stenosis varies from 0% to 35% [11, 16]. Ischaemia and anastomosis failure, also circular anastomosisaredangerousfactorsofstenosis,inaddition,asmentionedabove,stenosisisahighrisk of developing postoperative enterocolitis. The vast majority of stenoses are treated by dilatation, conservatively and only a few require deeper surgical correction. An oblique anastomosis on theanus can reduce the formation of stenosis. Peritonitis, as perianal phlegmon or pararectal fistula, requires removal of the stoma until inflammation is prevented and the coloanal anastomosis is suitable. Such complications are extremely rare, as the anastomosis is protected by the walls of the anal canal. However, in the presence of ischaemia or secondary infectious haematoma in the anastomosisarea,afocus ofinflammationcanoccur. In complicatedforms ofHirschsprung'sdisease (intestinal obstruction, necrotizing enterocolitis with toxic dilatation of the colon, perforation of the terminal small intestine, other cecum or large intestine sections) biopsies are taken at different levels of the colon, a stoma is drawn and a rectal biopsy is taken. Once the diagnosis of Hirschsprung's disease has been made, and the aganglionic region of the intestine has been identified, the type of surgical treatment is determined [22]. The intestinal type of agangliosis is a serious omen. An upper small bowel stoma will inevitably cause short bowel syndrome and a dubious prospect after radical surgery [8]. Carefully considered technique, adequate hemostasis, good blood supply to prevent ischaemia, avoidance of intestinal entanglement and distension should prevent complications during transanal endorectal descending.

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aganglionic bowel or persistent constipation, irrigoscopy and biopsy may be required. The method of treatment depends on the findings and may include active dilatation, Botox, myectomy and recurrent prolapse (13,16,20).
Encopresis is usually found in children over 4 years of age. The vast majority of examinations show the absence of encopresis after transanal proctoplastic surgery. Some authors have pointed out the presence of encopresis after bowel obstruction due to abnormal faeces or liquid stools after extended aganglionic zone surgery in children at this time. 44% of children have to follow a diet to prevent constipation or encopresis [7,14,17]. Other authors point to changes in the consistency of the faces and aspects of constipation over the years[16,19].Enuresis occurs in5-26%of cases and is attributed to iatrogenic damage to pelvic nerves or neuropathies [13]. The use of laparoscopy or transanal lowering is aimed at reducing iatrogenic damage. Defecation disorders, constipation, encopresis and enuresis have a major impact on the lives of patients with Hirschsprung's disease.The shortness of the colon after resection of the extensive aganglionic zone leads to a worsening of the quality of life, but this will change within a year, after the patient learns to control the defecation process and helps himself through different methods (enema, diet, etc.). The surgeon's task is to develop an all-round operative approach to these complications, reducing them through post- operative and long-term follow-up care.
Surgical correction should be carried out in hospitals with experience in treating such patients. Operative tactics include open laparoscopic surgery such as Swenson[9,21],Boley-Soave[12,19]or Duhamel [11,15]. For the first time, in the early 1950s, O. Swenson and F. Rebein, independently described a surgical approach for Hirschsprung's disease. While O. Swenson presented an extracorporeal anastomosis [18], F. Rebein first described an intracorporeal anastomosis, an inferior anterior abdominal resection [8]. Subsequently, B. Duhamel published data on post-rectal descending bowel with side-to-side anastomosis [15]. To date, this procedure is an elective surgery for total Hirschsprung's disease and can be performed laparoscopically [11]. F.Soave [12] and S.Boley [18] independently described similar operations for Hirschsprung's disease. They consist of submucosal dissection of the aganglionic segment. The muscularis retains the muscle box of varying lengths and the healthy bowel is lowered through it, resulting in an anastomosis.
The authors often use video-assisted endorectal surgery for transanal lowering. These include laparoscopic seromuscular biopsy to locate the extended transit zone, laparoscopic mobilisation of the distal colon and intermediate submucosal circular dissection, followed bybowel lowering and an anastomosis of the healthy bowel. The laparoscopic technique, preferred by the authors, was described in 1999 [7]. It is performed without changing the position of the patient's body in both laparoscopic and intermediate interventions from the lower extremities to the thorax of the patient in the operative field, and enables to do it without additional antiseptic measures. In order to decompress the bladder and create a close-up view during the manipulation of the pelvic organs, catheterisation is performed. First the transit area is determined macroscopically by visual assessment, the dilated proximal segment and its passage point to the retracted distal segment. A seromuscular biopsy is performed from the border of the designated healthy proximal zone to the altered distal colon. Biopsies are sent for express biopsy. At this time a distal rectal dissection is performed by mobilising down the mesorectum. Care must be taken when dissecting the intestine to avoid damaging the laterally passing urinary duct and the vas deferens. Rectal dissection is performed circularly, up to the level of the levator complex. After confirmation by pathomorphologists of the transit zone, electroligation of the mesenteric blood vessels of the aganglionic segment to the proximal region of the healthy bowel is performed. In cases of transition of the aganglionic zone into the rectosigmoid portion,

mobilisation of the left colonic half andsplenic angle is performed. After mobilisation of the abdomen, the patient's lower extremities are lifted upwards and an intermediate operation is performed. Using circular sutures connected to the skin of the perianal area, the edges of the anus are turned out. A circular incision is made, traction sutures are tightened and the submucosal dissection continues proximally until the muscle cuff is turned out through the anus. A longitudinal incision is then made at the back of the cuff and the cuff is returned to the abdomen. The intestine can now descend through the anus to the level of the "healthy" biopsy. At the same point, the intestine is cut off. The proximal healthy bowel is sutured to the small part of the submucosa, lower directly to the dentate line.

Laparoscopic endorectal descending of the colon can also be performed by birch-port penetration, leaving an unpleasant looking scar (15). The functional results of this type of intervention can be compared with conventional three-port laparoscopy. After surgery, patients drink water until transit is restored and the volume of the peritoneum is reduced. The child is then transferred to an age- appropriate diet. Postoperative antibiotic treatment may last for 24 hours at the surgeon's recommendation. After the patient starts eating, regular defecation will begin and the child can be discharged home. Within two weeks, the child is regularly examined for anastomosis retraction,which may require folding.

Patients requiring enterostomy before radical surgery are at risk of common peristomal complications such as bowel prolapse and retrostomy, peristomal infection and inflammation. Some patients suffer from postoperative complications at the end of treatment, abscess formation, constipation from damage to the internal sphincter during surgery or encopresis. Stable distal aganglionosis may require revision if the segment is longer than 2-3 cm. If this segment is short, relaxation can be achieved by botulinum toxin injection or sphincterotomy.

Patients who have been cured of Hirschsprung's disease in the long postoperative period especially have positive results after reaching adulthood. However, enterocolitis persists in 10% of patients, regardless of the surgical technique. Some patients suffer from constipation and this requires additional treatment. Occasionally, the sphincter complex can be damaged after repeated operations. This in turn leads to encopresis (21). This can be the cause of poor postoperative results when compared to the total variant of Hirschsprung's disease. The technical features of bowel depressions lead to a reoperation of the residual aganglionic zone [9].

Conclusions

1. In spite of modern development of coloproctology, accurate data on distribution, classification of HD in children of various ages are insufficient and the questions of its diagnostics and treatment are still actual.
2. Great progress has been made in the diagnosis and treatment of Hirschsprung's disease. However, the desired functional results have not yet been achieved. The reason for this is the time required for surgical correction of the defect as well as the choice of a surgical method suitable for each patient. A study of the literature shows that while questions about the pathogenesis of the disease are not controversial, new methods of examination and modern surgical treatment remain difficult.
3. The team of most specialists supports the dynamics of HD, modern and effective surgical treatments, however, strict ad er enceto standards and the organization of the collection of severe and difficult patients and treatment measures in large colorectal centres is essential.

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