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POSTOPERATIVE COMPLICATIONS IN HIRSCHSPRUNG'S DISEASE IN CHILDREN

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Abstract: Hirschsprung's disease (HD) is a congenital condition characterized by the absence of ganglion cells in one segment of the colon, leading to functional obstruction and severe constipation. Surgical intervention, usually involving resection of the aganglionic segment and anastomosis of the healthy bowel, is the mainstay of treatment for HD. Although surgery can significantly improve bowel function and quality of life, postoperative complications remain a concern. At the site of surgical reconnection, the anastomosis may leak, which requires additional intervention. Intestinal obstruction can be caused by an adhesion or stricture, which requires careful observation and treatment.

Key words: Hirschsprung's disease, postoperative complications, enterocolitis, intestinal obstruction, anastomotic leakage, pediatric surgery.

Hirschsprung disease (HD) is a motor disorder of the colon caused by the failure of neural crest cells (precursors of enteric ganglion cells) to migrate completely during intestinal development. The resulting aganglionic segment of the colon fails to relax, causing a functional obstruction. In the majority of patients, the disorder affects a short segment of the distal colon and rectum, with a transition zone in the rectosigmoid. In other patients, the aganglionosis involves longer segments of the colon; in rare cases, the entire colon and parts of the small bowel may also be involved. Hirsprung's disease is a congenital loss of ganglion cells in the intestine, and the lack of these cells causes intestinal obstruction.

These babies may present at birth with delayed bowel movements or bowel movement abnormalities associated with bowel movements and vomiting.What is the diagnosis?Diagnosis is made by clinical history and examination, followed by tests such as abdominal x-ray, barium/Gastrografin enema. Confirmation is done by biopsy (rectal biopsy or laparotomy and biopsy). Girschprung's disease (GD) is a developmental disorder of the enteric nervous system (ENS) due to defects in neural crest migration, proliferation,

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differentiation, and survival, leading to agangliosis of the intestinal wall. The neural crest (cristaneuralis) is a very important embryonic structure that forms the basis of a diverse spectrum of cell populations, including those that make up the enteric nervous system. Since the most important etiopathogenetic factor in the development of Girshprung's disease is the disease of the nerve sheath, this condition belongs to the category of neurocristopathy.

As a result, the complete absence of ganglia in the affected part of the intestine leads to functional obstruction, which is clinically manifested after birth. Although this disease was described by the Danish physician Harald Girschprung in 1886 and named after him, it was actually the Dutch anatomist and botanist Frederik Rüysch who first mentioned the condition in 1691 in his report on the disease of a 5-year-old girl as a "gross dilatation of the intestine". frequency is from 1:4400 among live births up to 1:7000. In terms of gender, the short form of Girschprung's disease is mainly in favor of the male sex, ranging from 3:1 to 4:1. The gender shift is 1:2 to 2:1 between males and females in both subtotal and total forms of the disease. A high index of suspicion for Girschprung's disease should be endorsed in children with congenital malformations. The most noticeable early symptom is the delay of the primary stool (meconium), which leads to further clinical-instrumental and histological examinations and confirmation of the diagnosis. The inevitable therapeutic approach is the resection of the aganglionic part of the intestine, but 30-50% of patients have fecal retention symptoms after the operation. If the symptoms of the disease persist despite treatment, the weight of the remaining content of the intestine can worsen the patient's condition and lead to a terrible complication, enterocolitis associated with Girshprung. Enterocolitis is the most common cause of death caused by this pathology.

Enterocolitis associated with Girschprung's disease is characterized by severe diarrhea, hyperthermia, and abdominal rest. According to the data presented in the foreign literature, enterocolitis associated with Girshprung is observed in 6-60% before surgery and in 25-37% after surgery. Mortality from this pathology can reach from 1% to 10%. Surgical treatment of Girsprung's disease has improved significantly in recent decades. Advances in modern approaches, such as one-stage transanal drainage, have resulted in smoother postoperative scarring, less pain, shorter hospital stays, as well as greater safety and efficacy. Nevertheless, the majority of patients suffer not only from somatic aspects of persistent symptoms, but also from some conditions that persist after treatment, such as incontinence, postoperative constipation, and this affects their social and emotional well-being. Another important aspect of the disease is its origin. As mentioned above, GK is included in neurocristopathies, therefore, the examination of a patient with the diagnosis of GK should be aimed at looking for other disorders associated with the defective development of the neural crest, for example, medullary carcinoma of the thyroid gland, neurofibromatosis, or multiple endocrine neoplasia at an older age. A general X-ray for diagnosis is not always informative. Irrigography with a water-soluble contrast agent is desirable.

Hyperosmolar solutions should not be used because they can cause fluid and electrolyte imbalance. Except in infancy, when occult perforation is less likely, barium is preferred in the diagnosis of GC because it provides a clearer view. Contrast enemas are not recommended during active enterocolitis. Classic radiographic findings in Hirschsprung's disease represent a narrow spastic distal bowel segment with dilatation in the proximal segment. The point of change of caliber or the transition zone is the main radiographic sign in the diagnosis of Girshprung's disease with the help of irrigographic examination. The

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transition point is most often located in the rectosigmovid area, but it can be located anywhere in the colon and ileum. Anorectal manometry (ARM) is used to determine the absence of a relaxation reflex after bolus distension in the rectal cavity. This is the most common method performed in children and requires little training. It is a less invasive method compared to rectal biopsy, and, unlike irrigography, does not require radiation.

Manometric assessment of anorectal function includes measurement of anal canal length and resting pressure, rectoanal inhibitory reflex (RAIR), rectal sensitivity, and finally, the ability to simulate contractions and defecation. The rectoanal brake reflex is the reflex relaxation of the internal anal sphincter in response to rectal distension. Intrinsic innervation of this reflex gut is present in normal individuals and absent in individuals with GC. However, the reported diagnostic accuracy of ARM has significantly lower specificity and positive prognostic value in some studies. In addition, the diagnosis of Girschprung's disease in infants is controversial in the literature. de Lorijn F. et al compared the diagnostic accuracy of anorectal monometry, contrast enema, and rectal aspiration biopsy (RAB) in infants diagnosed with GC in a systematic review. They reported that RAB was the most accurate test with the highest mean sensitivity of 93% and mean specificity of 98%. The diagnostic accuracy of ARM was slightly lower: average sensitivity and specificity were 91% and 94%, respectively. In 1948, F.R. Whitehouse and I.W. Kernohan published the first detailed series of clinical cases that clearly demonstrated that distal rectal aganglionosis and adjacent rectal length variability were primary phenotypic features of Girschprung's disease. Absence of ganglion cells (aganglinosis) in both submucosal tangles and muscle tangles of the affected segment of the intestine is a characteristic microscopic feature of the disease.

Postoperative complications in Hirschsprung's disease can include several issues that may arise following surgical intervention:This is one of the most common complications, characterized by inflammation of the intestines. Symptoms can include diarrhea, abdominal distension, and fever, requiring prompt medical attention. This occurs at the site where the healthy bowel is connected to the remaining bowel. Signs may include fever, abdominal pain, or a change in stoma output if present. Adhesions or strictures can lead to obstructions after surgery, presenting as vomiting, abdominal pain, or failure to pass stool.Some children may experience difficulties with bowel control postoperatively, which can persist and require additional management. Some children may have a slower return to normal bowel function, necessitating prolonged hospitalization or additional interventions.

Children may encounter feeding difficulties or malabsorption issues due to bowel surgery, requiring dietary adjustments and monitoring.The psychological adjustment to surgery and ongoing bowel issues can affect a child's mental health and social interactions. Early recognition and management of these complications are crucial to improve outcomes and enhance the quality of life for affected children. Regular follow-up with a pediatric gastroenterologist may be necessary for long-term care.

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