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Abstract

This article analyzes the incidence and treatment approaches for congenital gastrointestinal tract (GIT) malformations in children.

This study utilizes comparative analysis of our own data and a literature review of congenital GIT pathologies.

The study found that the most common anatomical locations for this condition are the small intestine and anus.

It is concluded that a comprehensive approach to the diagnosis and treatment of GIT malformations is essential for a favorable prognosis and quality of life in children.

Title : CONGENITAL GASTROINTESTINAL MALFORMATIONS IN CHILDREN

Keywords : congenital malformations, gastrointestinal tract, surgical treatment, children

Introduction

Intestinal malformations are the most common defects of the digestive tract. They occur with varying frequency for each disease entity, with an average incidence of 1 in 500-5,000 newborns. Their high relevance in clinical pediatrics is due to the need for early surgical intervention, which in some cases may only be palliative (for example, in Hirschsprung's disease).

This article presents current data from Russian and international literature on the main issues in surgical correction of congenital gastrointestinal malformations in children. The effectiveness of various surgical treatment methods and their potential for practical application are discussed. New concepts for providing surgical care to newborns at various stages of treatment are presented.

RELEVANCE OF THE PROBLEM

Every year, the number of neonatal infants developing various surgical pathologies increases worldwide. Among newborns with surgical diseases, the majority are children with congenital malformations. A significantly smaller group also includes children with surgical infections: enterocolitis, peritonitis, and purulent processes in soft tissues and bones.

Methods

Rational tactics and a preoperative preparation program based on objective criteria for assessing the severity of the child's condition are essential in neonatal surgery. For most congenital malformations, preoperative preparation begins in the maternity hospital. Its duration is determined by the condition of the infant with the surgical pathology and ranges from several hours to 2-3 days. Extending the preoperative preparation period reduces the risk of renal dysfunction in newborns with surgical pathology and prevents the development of acute renal failure [7].

Maintaining an adequate temperature regime during transportation, using multicomponent endotracheal anesthesia, cardiotonic drugs, synchronized prolonged artificial ventilation, prolonged pain relief, appropriate antibiotic therapy with continuous monitoring of the microecological status, and early detection and correction of concomitant pathology are essential for optimizing treatment outcomes for newborns with surgical pathologies. Care regimens for newborns with malformations should comply with treatment protocols for each nosological entity [10]. The survival rate of patients with esophageal atresia (EA) has increased significantly over the past decade, driven by improvements in surgical techniques, the introduction of minimally invasive surgical methods, and changes in preoperative and postoperative management of children. In patients with esophageal atresia, concomitant malformations, which occur with a frequency of 35–50%, have a significant impact on prognosis. Correction is particularly challenging in the presence of the VACTERL association [25]. The first successful operation for AP without fistula was performed by J. Donovan in 1935, and the first successful operation for AP with tracheoesophageal fistula (TEF) was performed by W. Ladd and N. Leven in 1939. The operation consisted of creating a gastrostomy, ligating the tracheoesophageal fistula, and creating a cervical esophagostomy. N. Leven subsequently performed esophagoplasty using a section of small intestine, and W. Ladd created an artificial esophagus using a flap from a skin tube [1].

Serious complications may occur in the early postoperative period, including leakage of the esophago-esophageal anastomosis with the development of mediastinitis and sepsis, recurrence of the pathological communication between the trachea and esophagus, and stenosis of the esophago-esophageal anastomosis. Less commonly, clinical manifestations of severe dysphagia and respiratory distress due to secondary tracheomalacia, requiring long-term ventilation therapy, are observed. In addition to segment tension, anastomotic technique, and the quality of suture material, gastroesophageal reflux undoubtedly plays a role in the development of esophago-esophageal anastomosis leakage. In cases of "major" esophagoesophageal anastomosis leakage, characterized by the presence of a significant amount of drainage, the development of pneumomediastinum, respiratory distress, and significant leakage of contrast medium beyond the esophagus during X-ray control, preference is given to rethoracotomy and reanastomosis. In rare cases, with severe mediastinitis and a large esophageal wall defect, it is preferable to perform an esophagostomy and gastrostomy [4]. Nevertheless, cases of conservative treatment of significant esophageal-esophageal anastomotic defects with favorable outcomes have been described. "Minor" leaks often have no clinical manifestations and are detected radiographically. In most cases, this complication is managed conservatively: adequate drainage, gastrostomy placement, transition to parenteral nutrition, and appropriate antibacterial therapy. With adequate drainage, 95% of esophageal-esophageal anastomotic leaks resolve spontaneously, and, moreover, without stenosis [29]. Some authors perform prophylactic bougienage of the esophago-esophago-anastomosis area 3-4 weeks after the anastomosis is created, while other researchers recommend dilating the anastomotic area if the patient presents with certain symptoms (dysphagia, aspiration, pneumonia, foreign bodies). Bougienage is typically performed using a thread or string. Balloon dilation is quite popular; if it is ineffective, esophagoplasty is performed using various methods [21].

Later complications following surgical correction of esophageal atresia include the development of secondary gastroesophageal reflux, the genesis of which is associated with both tension of the lower segment and a change in the angle of His, as well as with impaired esophageal motility in the postoperative period, which, according to D.N. Levin et al. (2011), is detected in 26–70% of cases. Based on the results of pH-metry and fluoroscopy of the esophagus and stomach, clinical presentation, and medical history, the advisability of antireflux surgery is determined on an individual basis. In the postoperative period, endoscopic monitoring is necessary to detect Barrett's esophagus in the future [23].

In order to reduce the adverse factors associated with pain in the postoperative period following thoracotomy and chest wall deformities, minimally invasive procedures have been used in recent years to create an esophageal anastomosis. In March 1999, T. Lobe and S. Rothenberg performed the first successful thoracoscopic reconstruction of a fistula-free form of esophageal atresia in a two-month-old child. Currently, the widespread adoption of the thoracoscopic technique for creating an esophageal anastomosis in newborns is noted [21].

A significant advantage of thoracoscopy is its excellent visualization of mediastinal structures, allowing for careful mobilization of esophageal segments without damaging the small branches of the vagus nerve, which is important for the function of the distal esophagus in the postoperative period.

The overwhelming majority of pediatric surgeons note a clear trend toward increased survival in newborns with esophageal atresia, reaching 70–90%. High mortality (up to 65–80%) is associated only with cases of esophageal atresia combined with severe congenital cardiovascular defects, prematurity, and multiple congenital malformations [32]. According to L. Spitz (2007), the favorable outcome of esophageal atresia in newborns weighing more than 1500 g and in the absence of significant cardiac problems reaches 100%, but can decrease to 80% in

the presence of one of the risk factors and to 30–50% in the presence of two or more factors [26].

Congenital intestinal obstruction is a common malformation, occurring in 1:1500–1:2000 newborns. Prenatal diagnosis is possible from the 18th to 20th week of gestation, especially in cases of severe obstruction. The most reliable signs of visualization of the defect are identified by the 24th to 30th week. Antenatal complications such as meconium peritonitis (pseudocysts, hyperechoic ascites) are detected in 20% of cases [5]. The ratio of duodenal and small intestinal (jejunal and ileal) obstruction is approximately the same, but associated malformations are more common with high obstruction - on average in 33.7-67% of cases, while almost every fourth infant with duodenal atresia has Down syndrome [14].

In recent years, significant changes have occurred in the surgical treatment of intestinal obstruction in newborns. For severe (duodenal) obstruction, the Kimura diamond-shaped anastomosis is preferred. For small bowel obstruction, "straight" end-to-end anastomoses are most often used, while T-shaped anastomoses are used less frequently. In cases of diameter discrepancy, some authors recommend performing de Lorimier-Harrison plication anastomoses. Staged treatment with enterostomy is resorted to only in cases of perforation of the afferent colon leading to fecal (meconium) peritonitis. Interintestinal anastomoses in newborns are performed using a single-row continuous suture made of thin biodegradable material [6].

Various minimally invasive approaches are used as an alternative to laparotomy, including subumbilical mini-laparotomy and various selective approaches. In recent years, video-assisted surgery (laparoscopy) has been increasingly used in the treatment of congenital intestinal obstruction—duodenal obstruction and congenital volvulus. In the postoperative period, some patients continue to experience short bowel syndrome and malabsorption, which is associated with congenital shortening of the intestinal tube due to gastrointestinal tract

malformations, as well as the extent of resection of the dilated afferent loop, especially in cases complicated by volvulus. To maximize intestinal tube length preservation in cases of multiple atresias, multiple interintestinal anastomoses are performed between the atretic segments with a proximal T-shaped anastomosis to decompress the intestine.

In cases of extensive necrosis secondary to congenital volvulus, the technique of delayed relaparotomy remains relevant for assessing the viability of intestinal loops after the volvulus has been corrected and a parsimonious resection has been performed. In addition, various methods of "lengthening" the intestine are used. Due to improved early diagnosis of intestinal obstruction in newborns, severe complications such as perforation of the afferent loop, volvulus, and so on are less common. This fact is also associated with improved survival rates for gastrointestinal atresia.

Mortality is mainly related to the severity of concomitant pathology (congenital heart defects, immaturity, respiratory distress syndrome, intraventricular hemorrhage, etc.). In general, according to various authors, survival rates for congenital intestinal obstruction range from 75.4 to 96% [11, 22].

Anorectal malformations are a fairly common congenital pathology of the neonatal period, occurring with a frequency of 1:4000–1:5000 newborns [5]. According to a number of authors, combined malformations are observed in 38–44% of cases, most often of the genitourinary, cardiovascular, and musculoskeletal systems. In addition, anorectal atresia is part of the VACTERL association [25]. Surgical treatment of this defect depends on the height of atresia, the presence of fistulas, concomitant pathology, and the general condition of the infant (its maturity, acute pathology of the neonatal period, etc.) [13].

The surgical approach used for anorectal malformations remains traditional: if the atresia is low and the newborn's condition allows for radical surgery, reconstruction is performed in the neonatal period. Good functional results are

observed with such early correction of the defect. The presence of wide fistulas allows for a delay of 1–2 months without a preventive colostomy. In all other cases —high rectal atresia, cloaca, or ureteral anastomoses—the first step is definitely a preventive colostomy [2].

The main stage of surgical treatment is performed primarily during the first six months of the patient's life. The surgical technique and the placement level of the iatrogenic stoma are debated in the literature. A split or loop colostomy is recommended (many authors rule out the possibility of intestinal contents refluxing into the diverting loop during a loop fistula), as well as dissection of the right or left colon or the proximal sigmoid colon. The surgeon's decision on this issue depends on the height of the atresia and the proposed type of reconstructive surgery. When choosing a proctoplasty method, the leading role is played by the ability to preserve the holding function (continence) and independent regulation of the act of defecation [18]. The method proposed by Alberto Pena at the end of the 20th century (PSARP – posterior sagittal anorectoplasty) fundamentally changed the understanding of proctoplasty techniques. Many authors attribute improved functional outcomes in the treatment of anorectal atresia to the posterior sagittal anorectoplasty method, which allows for delicate mobilization of the rectum in cases of rectourethral and rectovaginal fistulas [20].

In recent years, endoscopic surgery has made significant inroads into pediatric proctology. Laparoscopic mobilization of the rectum with the release of high fistulas allows for precise bowel reduction through the center of the perineum and the thickness of the levator vertebrae, maintaining the principle of minimal invasiveness at both the abdominal and perineal stages. Along with endosurgical methods of surgical interventions for anorectal malformations, the successful methods of combined abdominoperineal plastics according to Romualdi-Rehbein, the original operation of A.I. Lenyushkin - excision of a rectovestibular fistula with a formed anus, the use of parasacral access, methods of anterior plastics of the levator complex, etc. have not lost their significance [19].

In anorectal anomalies, combined regional osteoneural defects account for 40–50%, which causes secondary neurogenic dysfunctions and leads to urinary incontinence and various defecation disorders (Shaul D.B. et al., 1997; Yoshida A. et al., 2010; Goossens W.J. et al., 2011). For the early diagnosis of Currarino syndrome and other anomalies of the sacrococcygeal region, MRI is included in the diagnostic algorithm. The higher the location of the atretic section of the rectum, the more pronounced the dysmorphism of the anorectal zone [30].

According to S. De Vos et al. (2011), after primary plastic surgery for low forms of rectal atresia, normal functional results were achieved in 93% of cases, while after staged treatment of high and intermediate forms, the results were 41% normal, 17% good, 38% satisfactory, and 4% unsatisfactory. Functional disorders of the rectal obturator apparatus after radical correction of anorectal defects, according to various authors, are observed in 30–60% of cases and are accompanied by impaired social adaptation of children in society. PSARP for intermediate and high forms provides good results in 77% of patients [15, 24].

Conclusions

In recent years, endorectal colon pulldown has been widely used in the surgical treatment of congenital colon aganglionosis in newborns and infants, alongside classical abdominoperineal proctoplasty techniques. In cases of extensive resections, video-assisted colon mobilization is used [17].

Thus, a review of domestic and international literature has demonstrated a variety of surgical approaches for newborns with congenital gastrointestinal defects. In recent years, a significant number of scientific papers have appeared on the treatment of intestinal malformations, highlighting the existing challenges in the treatment and rehabilitation of this critical group of patients. The solution to these problems lies in further scientific research and the improvement of treatment and rehabilitation measures for children with congenital gastrointestinal anomalies.

Intestinal malformations are the most common defects of the digestive tract. They occur with varying frequency for each disease entity, with an average incidence of 1 in 500-5,000 newborns. Their high relevance in clinical pediatrics is due to the need for early surgical intervention, which in some cases may only be palliative (for example, in Hirschsprung's disease).

Anal atresia is currently highly prevalent. This defect is a major cause of childhood disability, as it is virtually impossible to cure. Furthermore, intestinal malformations can coexist with other gastrointestinal defects, significantly complicating treatment and worsening the child's prognosis.

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