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Management and the main complications of gastroschisis: A systematic review

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ABSTRACT

Objective: This systematic review article aims to compile and analyze the evidence on complications and strategies for the management of gastroschisis, providing a comprehensive and up-to-date view to guide future research and clinical practice. Methodology: The systematic review used the PVO

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strategy to investigate the main surgical treatments for gastroschisis and its complications. Searches were carried out in the PubMed Central (PMC) and Virtual Health Library (VHL) databases with specific descriptors, resulting in 134 articles. After applying the inclusion and exclusion criteria, 14 articles were selected for analysis. Discussion: Immediate postnatal care is crucial to avoid complications such as fluid loss, hypothermia, and infections. Infants with gastroschisis should be treated in neonatal intensive care units. Initial measures include protection of the herniated viscera, regulation of body temperature, and gastric decompression. Results: Surgically, timely reduction of herniated viscera is essential to avoid severe abdominal syndromes. The choice between primary or delayed correction depends on the patient's conditions. An integrated and vigilant approach is vital to improve neonatal outcomes and reduce the morbidity and mortality associated with gastroschisis.

Keywords: Gastroschisis, Management and complications.

INTRODUCTION

Gastroschisis is a congenital defect of the abdominal wall, usually to the right of the midline. It occurs when the abdominal organs cannot properly return to the abdominal cavity. Risk factors include young maternal age, smoking, and infection. Although there are several theories about the etiology of gastroschisis, such as vascular lesions, abnormalities in the folding of the ventral wall, and failures in the insertion of the yolk sac, none of them has been definitively proven. The ventral defect associated with gastroschisis is less than 4 cm in diameter and has no membrane covering the bowel. Gastroschisis involves a visceral hernia visible on ultrasound examination, with the abdominal contents floating in the amniotic fluid with no covering membrane. The cause is still unknown in most newborns, but there is a strong association with young maternal age. The global incidence is about one case per 1,953 births. (FERREIRA. et al; 2022 and DIYAOLU et al; 2021)

Generally, gastroschisis is located to the right of the umbilical cord and contains primarily the midgut, as well as the stomach and possibly the gonads and distal colon [1,8]. Because the intestine is exposed to the uterine environment, it can become thick, tangled, edematous, and covered by a fibrinous shell. These changes can also occur after birth, when the intestine is exposed to the external environment. Newborns with gastroschisis are more likely to be premature, have respiratory complications, and have low weight for gestational age. (DIYAOLU et al; 2021)

Risk factors include low maternal socioeconomic status, early maternal age, and low preconception body mass index. Exposure of the intestine to amniotic fluid leads to pathological changes, such as thickening and stiffness of the bowel loops. In addition, the disproportion between abdominal growth and abdominal cavity volume can make postnatal bowel replacement difficult. Intestinal dysmotility is common in gastroschisis, but its exact cause is not yet understood. A "two-stroke" model suggests that mesenteric ischemia also plays a role. Prognosis has improved with advances in maternal-fetal medicine, neonatal intensive care, and pediatric surgery. (DURMAZ et al; 2022)

Complex gastroschisis, with intestinal complications, has a poor prognosis, whereas simple gastroschisis does not have such complications. The fetal development of gastroschisis is dynamic, occurring until birth. Despite being identified on prenatal ultrasound, there is no consensus on the optimal time for delivery and the treatment strategy, requiring systematic review of the evidence. On the other hand, in neonates with complex gastroschisis (which includes at least one of the following complications: intestinal atresia, perforation, necrotic segments, or volvulus), mortality and morbidity rates are significantly higher compared with simple gastroschisis. (FERREIRA. et al; 2022) (DIYAOLU et al; 2021) (RADUMA. et al; 2021)

Complicated gastroschisis is associated with gastrointestinal conditions such as atresia, perforation, stenosis, volvulus, or necrosis. These cases have higher morbidity and mortality

compared with uncomplicated gastroschisis. Studies show that neonates with complicated gastroschisis spend more days on mechanical ventilation, have prolonged periods of adynamic ileus, longer hospitalizations, and take longer to tolerate full enteral feeding. There is also an increase in gastrointestinal, respiratory and infectious complications.

On the other hand, simple or uncomplicated gastroschisis is not associated with these specific pathologies. The bowel dilation seen on ultrasound may be predictive of complicated gastroschisis. (DIYAOLU et al; 2021)

The prognosis of gastroschisis has improved due to advances and increased collaboration between the disciplines of maternal-fetal medicine, neonatal intensive care, and pediatric surgery. However, in neonates with complex gastroschisis (which includes at least one of the following complications: intestinal atresia, perforation, necrotic segments, or volvulus), mortality and morbidity rates are significantly higher compared with simple gastroschisis (without these complications). In addition, there are significant differences in clinical behavior, postoperative complications, and length of hospital stay between infants with simple and complex gastroschisis. The exact cause of complex gastroschisis is not yet completely understood, but it may be related to ongoing inflammation due to exposure to amniotic fluid and the formation of a shell around the exposed intestines. Other mechanisms may also be involved, but they are not yet clear. (DURMAZ et al; 2022)

Despite advances in maternal-fetal medicine, neonatal intensive care, and pediatric surgery, the optimal management of gastroschisis, especially in terms of the timing of delivery and treatment strategies, remains a topic of debate. This systematic review article aims to compile and analyze the available evidence on complications and strategies for the management of gastroschisis, in order to provide a comprehensive and up-to-date view that can guide future research and clinical practice.

METHODOLOGY

This is a systematic review that seeks to understand the main aspects of gastroschisis, aiming to demonstrate the main methods used in the treatment of the pathology, and the complications that appear concomitantly with the condition, aiming to ensure a greater clinical elucidation of this condition. For the development of this research, a guiding question was elaborated through the PVO (population, variable and objective) strategy: "What are the main surgical treatments for gastroschisis, as well as its complications? ``

The searches were carried out through searches in the PubMed Central (PMC) and the Virtual Health Library (VHL) databases. Four descriptors were used in combination with the Boolean term "AND": Gastroschisis, Digestive System Surgical Procedures, Postoperative Period, Short Bowel Syndrome. The search strategy used in the PMC database was: Gastroschisis and Digestive System

Surgical Procedures AND Gastroschisis and Postoperative Period AND Gastroschisis. In the VHL database, the following combinations were searched: Short Bowel Syndrome AND Gastroschisis and Postoperative Period AND Gastroschisis. From this search, 134 articles were found, which were subsequently submitted to the selection criteria. The inclusion criteria were: articles in English, Portuguese and Spanish; published in the period from 2019 to 2024 and that addressed the themes proposed for this research, in addition, review, observational and experimental studies, made available in full. The exclusion criteria were: duplicate articles, available in the form of abstracts, that did not directly address the proposal studied and that did not meet the other inclusion criteria.

After associating the descriptors used in the searched databases, a total of 134 articles were found. After applying the inclusion and exclusion criteria, 14 articles were selected from the PubMed database, and a total of X studies were used to compose the collection.

DISCUSSION

The optimal time for delivery in cases of gastroschisis is widely debated. Opting for a late delivery may prolong the exposure of the herniated intestine to toxic amniotic fluid and increase the risk of intrauterine complications, such as closure of gastroschisis. These risks must be balanced against the complications of preterm birth. Limited evidence suggests that elective late preterm birth (planned between 35-37 weeks' gestation) is associated with fewer infectious complications and a faster recovery of enteral nutrition, compared with expectant management (term delivery). On the other hand, non-elective preterm birth is linked to a longer time to normal bowel function. Term preterm delivery (just after 37 weeks) has shown better results compared to expected full-term delivery. The decision about the timing of delivery should consider gestational age, ultrasound results, and fetal tests. Currently, most centers prefer planned delivery at 37 weeks. (BIELICKI et al; 2021)

The main purpose of immediate postnatal care is to prevent fluid loss (through evaporation) and hypothermia, as well as to prevent infections (BIELICKI et al; 2021) All patients with gastroschisis should be treated in a neonatal intensive care unit under the supervision of neonatal intensivists, respiratory therapists, and pediatric surgeons. Initial management of gastroschisis includes intestinal protection with a translucent pouch, temperature regulation and homeostasis, and reduction of evaporation losses. An initial examination of the intestines is required to rule out obvious findings such as atresia or volvulus. An orogastric tube is inserted for proximal decompression. Intravenous access is obtained for volume replacement, with a peripherally inserted central catheter being placed for eventual initiation of total parenteral nutrition (DIYAOLU et al; 2021)

The herniated viscera are covered with warm gauze soaked in saline, and the lower half of the newborn is placed in a plastic bag. This is especially important if the patient needs to be transported to another facility. If respiratory support is required, continuous positive airway pressure (CPAP) or high-flow O2 (to prevent the bowel from filling with air) should be avoided (BIELICKI et al; 2021)

Overall, there is conflicting evidence about the importance of the place of birth for congenital surgical anomalies. There is limited evidence to suggest that birth outside of an operating theatre may be associated with poorer neonatal outcomes, particularly for infants with congenital diaphragmatic hernia, which has a high mortality rate in the first few days after birth. Similarly, the need to transfer a defect such as gastroschisis for surgical repair can lead to a delay in the establishment of enteral feeding and a prolonged hospital stay (MALDONADO. et al; 2023).

There may also be a benefit to early exposure to breast milk, the preferred choice for feeding newborns, especially after gastrointestinal surgery. Breast milk is known to provide anti-inflammatory properties and nutrients that are uniquely adapted to the situation of neonates and are easily digestible. Due to exposure to amniotic fluid, infants with gastroschisis may experience decreased intestinal motility. This dysmotility can manifest as symptoms of food "intolerance" and can delay the advancement of enteral nutrition. Infants with gastroschisis who receive diets with human milk have been shown to have a shorter dwell time, achieving complete enteral feeding more quickly than those fed with formula (TUCKER. et al; 2020).

Early enteral feeding provides immunoprotective benefits to the gut by improving enzyme activity, nutrient delivery, gastrointestinal hormone/peptide function, surface immunity, gut microflora, and intestinal villus regeneration. These factors can lead to reduced risk of complications, especially necrotizing enterocolitis (NEC) and cholestatic injury, while increasing nutritional benefits. Studies show that the appropriate time of initiation and the rate of advancement of enteral feeding (EF) may be associated with better outcomes, lower complication rates, early interruption of parenteral nutrition (PN), shorter length of hospital stay (LOS) and, therefore, reduced cost of care in children with gastroschisis (RADUMA. et al; 2021).

The overriding goal in the surgical management of gastroschisis is to achieve a timely reduction of the herniated viscera, thereby avoiding damage to the viscera and preventing abdominal compartment syndrome (BIELICKI et al; 2021). After childbirth, the exposed intestine is susceptible to dehydration, mechanical trauma, pressure necrosis, or infections. To avoid these complications, two postnatal techniques have become standard in the treatment of gastroschisis, and can be addressed through primary or delayed correction (DIYAOLU et al; 2021), (DURMAZ et al; 2022).

In primary correction, the intestine is repositioned, ideally in its anatomical location, followed by the placement of the remaining umbilical cord over the defect and application of an occlusive dressing. Primary reduction with subsequent sutureless closure can be performed at the bedside,

often using a silo bag with warm saline, whereas primary reduction and suture closure are typically performed in the operating room. Regarding suture closure, after reduction, the edges of the fascia are closed with absorbable sutures. It is crucial to avoid increasing intra-abdominal pressure (IAP) (>20 mmHg) after fascia closure. In cases of mild increase in IAP, temporary use of muscle relaxants and sedation may be used to reduce fascial tension. If tension and/or IAP prevent primary closure of the fascia, simple closure of the skin over the defect or closure of the abdominal wall using prosthetic material can be performed (DIYAOLU et al; 2021), (BIELICKI et al; 2021)

Sutureless closure can be performed at the bedside, avoiding the need for general anesthesia, by applying a waterproof and non-adherent dressing. In addition, the umbilical stump can be placed over the defect to cover the viscera (BIELICKI et al; 2021). Alternatively, when the surgeon is unable to reduce the abdominal contents, a delayed correction may be performed, in which the intestines are placed in a silastic silo and gradually reduced into the abdominal cavity. Once the reduction is completed, the sutureless method can then be applied, placing a cushion and dressing over the abdominal defect. The dressings are left in place and replaced every 4-5 days until epithelialization occurs, after which petroleum ointment and a light gauze dressing are used. Delayed correction of gastroschisis is often used when the bowel is thickened or dilated and the reduction of the bowel causes significant respiratory impairment and/or abdominal compartment syndrome (DIYAOLU et al; 2021)

Fetal surgery for a relatively benign malformation, such as gastroschisis, is certainly very controversial. Although morbidity in the neonatal period can be significant, the overall survival rate is over 90% and with good long-term outcomes. This should be weighed against the risk of intrauterine death or prematurity, as well as possible surgical complications in intrauterine gastroschisis repair. Interestingly, the condition of complex gastroschisis carries a much higher morbidity and mortality than the general population of children affected by gastroschisis and is much more impressive when compared to the condition of simple gastroschisis. Therefore, complex gastroschisis should be the primary target of fetal intervention, since patients with simple gastroschisis do not have significant impairment in gastrointestinal function or health-related quality of life compared to healthy controls in adolescence and adulthood (DURMAZ et al; 2022).

Gastroschisis can be classified into complicated and uncomplicated. Complicated gastroschisis is associated with a gastrointestinal condition such as atresia, perforation, stenosis, volvulus, or necrosis, whereas simple gastroschisis is not linked to these pathologies. In addition, complicated gastroschisis is related to an increased morbidity and mortality compared to uncomplicated gastroschisis (DIYAOLU et al; 2021). Neonates with complicated gastroschisis have more days on mechanical ventilation, a prolonged period of adynamic ileus, longer hospitalizations, higher incidence of sepsis and catheter-associated infection, cholestasis, short bowel syndrome,

longer days to achieve complete diet, longer parenteral nutrition time, and prolonged dependence on parenteral nutrition can trigger liver disease associated with parenteral nutrition, with possible progression to severe liver failure (DIYAOLU et al; 2021) (BIGIO. et al; 2021).

Concomitant complications in gastroschisis are frequent. Up to 30% of patients initially present with complex gastroschisis, revealing intestinal atresia or intestinal stenosis. A subset of patients also suffer from secondary diseases such as transient or persistent intestinal dysmotility, necrotizing enterocolitis (NEC; 3.8–8.2%), volvulus (0.5–3.0%), intestinal necrosis (4.5%), and evanescent gastroschisis (<1.0%). These conditions can lead to long-term complications such as enteral feeding intolerance, short bowel syndrome (3.2–11.4%), bowel failure, and adhesive bowel obstruction (20.4–27.0%) (HAGHSHENAS. et al; 2021).

Surgical site infection (SSI) after surgery for an abdominal birth defect is a frequent and significant clinical problem that has been poorly studied in infants (less than three years of age) who have undergone surgery due to abdominal birth defects. There are different forms of SSI, such as wound infection, wound dehiscence, anastomotic leakage, postoperative peritonitis, and fistula development. These complications can lead to longer hospital stays, increased medical costs, impact on quality of life, and increased mortality rates (LD, GD, et al., 2021).

Gastroschisis, omphalocele, and small bowel atresia are usually at higher risk of wound infections compared with other types of defects. This can be partly explained by neonatal age and the relatively high proportion of babies born prematurely with these birth defects. More than half of patients with gastroschisis are born prematurely (LD, GD, et al., 2021).

Based on the data collected by the study (1), 154 studies were included, representing 11,786 patients. The overall pooled percentage of wound infections after surgery for abdominal birth defect was 6% (95% CI: 0.05–0.07), ranging from 1% (95% CI: 0.00–0.05) for common bile duct cyst surgery to 10% (95% CI: 0.06–0.15) after gastroschisis surgery. Wound dehiscence occurred in 4% (95%CI:0.03–0.07) of infants, ranging from 1% (95%CI:0.00–0.03) after surgery for duodenal obstruction to 6% (95%CI:0.04–0.08) after gastroschisis surgery (LD, GD, et al., 2021).

Abdominal compartment syndrome is a feared complication of gastroschisis reduction and closure, defined by persistent IAP above 20 mmHg (measured indirectly by bladder pressure), along with loss of function of one or more organs (e.g., anuria). The actual incidence of compartment syndrome is unknown and appears to be quite rare, but delayed closure is thought to reduce the risk of compartment syndrome (DIYAOLU et al; 2021)(BIELICKI et al; 2021)

About 10–15% of patients with gastroschisis have concomitant intestinal atresia, with the small intestine being more commonly affected than the colon (80% versus 20% of cases). Generally, there are three different treatment options: if there are no excessive signs of excessive intestinal inflammation/scaling, the atresia can be resected and the primary anastomosis is performed before

the abdominal wall is reduced and closed. If there are excessive signs of inflammation, a stoma is created or a silo is made, leaving the atresia untouched, and resection with anastomosis or stoma creation is performed when the abdominal wall is closed (i.e., after about 7 to 10 days). Whether primary resection and anastomosis and subsequent reduction (primary or staged) should be performed depends on the same basic factors as for all intestinal anastomoses, i.e., stable patient, adequate perfusion, absence of tension at the anastomotic site, absence of distal obstruction, absence of mesenteric torsion, and no incompatibility of intestinal lumens (BIELICKI et al; 2021)

Among the various congenital anomalies, patients with gastroschisis are at higher risk of incisional hernia after abdominal surgery, with an incidence of approximately 10%. In the subgroup analysis, complex gastroschisis and SILO closure were identified as risk factors for incisional hernia. In contrast, studies show that simple gastroschisis has an odds ratio of 0.18 for developing an incisional hernia compared to complex gastroschisis (EEFTINCK SCHATTENKERK. et al; 2020).

Ventral hernias are a well-known complication after gastroschisis repair. However, few studies have compared the incidence of ventral hernia based on whether the patient received immediate or silo closure. In this report, we observed that patients who underwent immediate closure had a higher incidence of ventral hernias than those who had silos placed for a short period. The hernia rate increased with longer silo duration and was equivalent to the immediate group when the silo was present for more than 10 days. The reason for this observed difference is unclear, but may be related to greater abdominal wall tension and fascial stress in patients with immediate closure compared to those who have a short-term silo placed with staggered closure whose abdominal dominance has increased. Silo placement may also facilitate fluid management with reduced intestinal edema and less strain at the time of definitive fascial closure (HAWKINS. et al, 2023).

As Sosnowska (2021) assures us, neonatal survival was 91.29% with a rate of 8.71% of postnatal deaths; fourteen deaths occurred in neonates with simple gastroschisis and sixteen deaths in neonates with complex gastroschisis (FERREIRA. et al; 2022). Despite the outstanding results of the treatment, it is essential to maintain continuous investigations to progressively improve the results and the complete recovery process after surgical intervention. For this, time and the support of a multidisciplinary medical team are required. The complete rehabilitation of an infant depends not only on the severity and management of gastroschisis, but also on the patient's clinical condition and other associated congenital malformations (SOSNOWSKA-SIENKIEWICZ. et al:2021).

Among the risk factors related to mortality in newborns with gastroschisis are low birth weight, prematurity, complex gastroschisis, sepsis, lack of prenatal diagnosis, and poor hospital care conditions. Properly identifying these risk factors for gastroschisis mortality can play a crucial role in the formulation of strategies by public health authorities and hospital managers to improve survival rates of neonates with this congenital anomaly (MUNIZ. et al; 2023).

CONCLUSION

Immediate postnatal care for patients with gastroschisis is essential to avoid complications such as fluid loss, hypothermia, and infections. It is critical that these infants are treated in neonatal intensive care units, under the supervision of neonatal intensivists, respiratory therapists, and pediatric surgeons. Initial measures include protection of herniated viscera with the use of a translucent bag, careful regulation of body temperature, and insertion of an orogastric tube for decompression. Additionally, it is crucial to perform a detailed initial evaluation of the intestines to rule out complicating conditions such as intestinal atresia or volvulus.

The early introduction of breast milk feeding has demonstrated significant benefits in the recovery of these patients, providing adapted nutrients and anti-inflammatory properties that favor gastrointestinal function. This nutritional care also contributes to reducing complications such as necrotizing enterocolitis and facilitates the transition to complete enteral feeding more quickly.

From a surgical point of view, it is essential to perform timely reduction of herniated viscera to avoid severe abdominal syndromes such as abdominal compartment syndrome. The choice between primary or late repair depends on the specific conditions of each patient, always aiming to minimize abdominal tension and prevent postoperative complications, such as incisional hernias. Therefore, an integrated and vigilant approach, focusing on the early identification of risk factors and the implementation of effective management strategies, is crucial to improve neonatal outcomes and reduce the morbidity and mortality associated with gastroschisis.

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