

## REVIEW ARTICLE

# Role of Surgery in the Outcome Difference of Gastroschisis between High-income and Low-middle-income Countries: A Review

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### Abstract

*Gastroschisis is a major congenital abdominal wall defect where the abdominal viscera comes out through a gap right to the umbilicus. The management of gastroschisis requires prompt and aggressive medical and surgical intervention immediately after birth. However, there is a notable disparity in mortality rates between developed countries and LMICs. Despite various surgical technique adjustments, LMICs still suffer from a high mortality rate, with certain centers reporting a 100% mortality rate. This article seeks to examine the literature and evaluate the impact of surgical techniques on the outcome of gastroschisis.*

**Keywords:** *Gastroschisis, primary fascial closure, preformed silo, staged closure.*

### Introduction

Gastroschisis is a congenital anterior abdominal wall defect with an incidence rate of 5 cases per 10,000 live births. Unfortunately, the incidence is rising worldwide.<sup>1</sup> In this condition, there is a gap right to the umbilical cord through which the intestine emerges from the abdomen.<sup>2</sup> Rarely, gaps left to the umbilical cord have also been reported.<sup>3,4</sup> It is differentiated from the omphalocele by the absence of the covering membrane and the gap lateral to the umbilicus.<sup>2</sup> Gastroschisis contributes to maximum mortality among neonatal surgical emergencies in LMICs. Even in some centers, all babies with gastroschisis die. Interestingly, mortality from gastroschisis in HICs is rare.<sup>5,6</sup> Even in some reported studies, it is almost nil, where the primary outcome measures are the length of hospital stay (LOS) and the duration of total parental nutrition (TPN). This unusual disparity in outcome depends not on the surgical procedure used but rather the intensive perioperative care. Still, there are

endeavors to modify surgical procedures to reduce morbidity in HICs and mortality in LMICs.<sup>1,5,7-10</sup> The uncovered intestine creates two different types of problems. In utero, it is exposed to the amniotic fluid and undergoes chemical changes. These changes alter its normal functions. After birth, the exposed intestine evaporates fluid, loses heat, becomes swollen, and invites infection. In 10-15% of babies, gastroschisis is associated with co-existing intestinal pathologies like atresia, necrosis, and perforation. Therefore, it requires aggressive medical and surgical management immediately after birth.<sup>2,10-12</sup>

The goals of management are- 1. Correction and prevention of hypothermia, hypovolemia, and sepsis 2. Reposition of the intestine and closure of the abdominal wall gap, 3. Establishment of normal enteral feeding.<sup>13,14</sup>

The successful outcome depends on the timely utilization of maximum resources to achieve all three goals. Unfulfillment of any of these goals

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compromises the survival chances of these babies. This article aims to review the role of surgery in the outcome of gastroschisis.

### **Types of surgical procedures in gastroschisis**

The surgical procedures used in gastroschisis can be broadly classified into two groups-

**Primary closure:** Here, the exteriorized gut is reduced into the abdomen, and the abdominal gap is closed immediately after birth. The first primary fascial closure of gastroschisis was reported in 1913.<sup>15</sup> Since then, it has been the first approach for gastroschisis in most centers. It offers an early reduction of the intestine into the abdomen and closure of the defect. It reduces the fluid and heat loss and chances of contamination of the exposed viscera. However, it increases the intra-abdominal pressure due to the visceroperitoneal disproportion in these babies. The reduced intestine produces abdominal compartment syndrome more or less in every patient, the severity of which depends on the degree of the visceroperitoneal disproportion. Therefore, these babies develop respiratory and renal compromise after surgery and require a prolonged period of mechanical ventilation. However, the visceroperitoneal disproportion does not allow primary fascial closure in all patients.<sup>8,13-15</sup> Some centers monitor intravesical and/or intra-gastric pressure to determine the possibility of primary fascial closure.<sup>16,17</sup>

**Modifications of primary closure technique:** Several surgical approaches have been described to reduce postoperative intraabdominal pressure and the need for mechanical ventilation. Several authors with successful outcomes have reported umbilical cord flap coverage of the gap. It maintains lower intra-abdominal pressure and reduces the need for mechanical ventilation. The flap is autogenic, so there is less chance of infection, and no extra cost.<sup>18-20</sup> Resection of various lengths of the gut has been reported to facilitate successful primary closure with lower intra-abdominal pressure. Okoro PE et al. achieved >67% survival rate in African resource-constrained centers with primary closure after extended right hemicolectomy.<sup>21</sup> Negash et al<sup>22</sup> described a successful outcome of primary closure aided by ileocecal resection and ileostomy in two patients.

**Staged or delayed closure:** Here, the exteriorized gut is gradually reduced into the abdomen over time,

and then the abdominal wall is closed. After its introduction in 1969, it has become a routine procedure in many centers. Initially, a silastic sheet was used to cover the exposed viscera, sutured with the skin and fascial margin of the gap. The staged closure became more popular with the invention of a preformed silo. Combined with suture-less closure, this self-retaining device is easy to use and can be placed at the bedside without anesthesia, eliminating the need for anesthesia completely. These bags are transparent and allow inspection of the gut for possible necrosis and perforation.<sup>8,15,23,24</sup>

### **Modifications of staged closure technique:**

Unfortunately, the preformed silo is expensive and unavailable in low- and middle-income countries (LMICs). However, several improvised silo techniques are currently in use in these countries. Sterile urobag is commonly used as an alternative to the preformed silo. Successful outcomes have also been reported using surgical gloves, female condoms, and saline bags. Recently, wound protectors are gaining popularity as a preformed silo. It is self-retaining, available, and cost-effective. It can be placed at the bedside without anesthesia.<sup>25-28</sup>

### **Role of surgery in outcome**

A significant advancement in treating gastroschisis occurred in 1970 with the introduction and widespread use of total parental nutrition (TPN).<sup>29</sup> This, combined with advancements in peri-operative intensive care, greatly increased the survival rate of neonates with gastroschisis. Mortality rates have significantly decreased, and gastroschisis is no longer considered fatal in medical centers with access to these resources.<sup>7-9</sup> Mortality is no longer an outcome measure in developed centers. Instead, the length of hospital stays, duration of TPN and mechanical ventilation contributing to the treatment cost is a matter of concern.<sup>1</sup>

Therefore, in developed centers, the modifications and choice of surgical procedures are directed at reducing the duration of mechanical ventilation, hospital stay, and TPN. Allotey et al. compared the benefits of staged closure with preformed silo against the traditional primary closure. The mean duration of mechanical ventilation was similar between the two groups. However, the primary closure group required higher mean airway pressure and inspired oxygen. The duration of TPN and hospital stay were also similar. They had no mortality in any group.

They concluded that staged closure is associated with less barotrauma and reduced intra-abdominal pressure.<sup>8</sup>

Schlatter et al<sup>29</sup> changed their surgical approach to gastroschisis in the past decade. Previously, they preferred emergency primary closure, which transitioned to staged closure in the past decade. In their series, patients in the staged closure group required fewer days on ventilators and had fewer complications. However, there was no difference in mortality. The overall survival was 98%.<sup>29</sup>

Similarly, Kidd et al<sup>30</sup> started staged closure routinely after 1993 in their practice. They reported longer duration of ventilation and hospital stay in staged closure. However, complications related to high intra-abdominal pressure were less. There was no difference in mortality.<sup>30</sup>

Various studies from developed countries, including multicentre cohorts, randomized controlled trials, and meta-analyses, have concluded that the type of surgery does not impact the survival of neonates with gastroschisis. However, it does affect the overall cost of treatment. A longer duration of mechanical ventilation, hospital stay, and TPN (total parenteral nutrition) are associated with higher treatment costs. As a result, gastroschisis is considered the most expensive non-cardiac congenital anomaly. Each patient requires more than 100,000 USD.<sup>9,31-36</sup>

This is where the survival rate varies between developed centers and LMICs. Despite several modifications in surgical technique, Inadequate infrastructure, unavailability of intensive care facilities, mechanical ventilation, and TPN are primarily responsible for the unacceptably higher mortality in LMICs. The type of surgery has no role in this. According to Ford et al<sup>10</sup>, gastroschisis can serve as a valuable indicator of the capacity of healthcare institutions to provide neonatal surgical care, as it demands considerable resource allocation.

### Conclusion

Gastroschisis is a major birth defect that demands costly intensive care before and after surgery. This care involves mechanical ventilation and total parental nutrition. Surgery is the primary treatment, but without proper pre- and post-operative care, survival rates are low regardless of the surgical approach used.

### References

1. Zalles-Vidal C, Peñarrieta-Daher A, Bracho-Blanchet E, Ibarra-Rios D, Dávila-Perez R, Villegas-Silva R, et al. A Gastroschisis bundle: effects of a quality improvement protocol on morbidity and mortality. *J Pediatr Surg* 2018;**53**:2117-22.
2. Bence CM, Wagner AJ. Abdominal wall defects. *Transl Pediatr* 2021;**10**:1461-69.
3. Mandelia A, Agarwala S, Sharma N, Solanki S, Panda S. Left-sided Gastroschisis: A Rare Congenital Anomaly. *J Clin Diagn Res* 2013;**7**:2300-02.
4. Danso P, Nimako B, Amoah M, Yifeyeh AC, Sagoe R, Davor A, et al. Rare Left-Sided Gastrochisis with Bifid Umbilical Cord. *Open Access Library Journal* 2021;**8**:1-8.
5. Wright N, Abantanga F, Amoah M, Appeadu-Mensah W, Bokhary Z, Bvulani B, et al. Developing and implementing an interventional bundle to reduce mortality from gastroschisis in low-resource settings. *Wellcome Open Res* 2019;**4**:46. DOI: 10.12688/wellcomeopenres.15113.1.
6. Hasan MS, Islam N, Mitul AR. Neonatal surgical morbidity and mortality at a single tertiary center in a low- and middle-income country: A retrospective study of clinical outcomes. *Front Surg* 2022;**9**:817528. DOI: 10.3389/fsurg.2022.817528.
7. Räsänen L, Lilja HE. Outcome and management in neonates with gastroschisis in the third millennium- A single-centre observational study. *Eur J Pediatr* 2022;**181**:2291-98.
8. Allotey J, Davenport M, Njere I, Charlesworth P, Greenough A, Ade-Ajayi N, Patel S. Benefit of preformed silos in the management of gastroschisis. *Pediatr Surg Int* 2007;**23**:1065-69. DOI: 10.1007/s00383-007-2004-9.
9. Kunz SN, Tieder JS, Whitlock K, Jackson JC, Avansino JR. Primary fascial closure versus staged closure with silo in patients with gastroschisis: A meta-analysis. *J Pediatr Surg* 2013;**48**:845-57.
10. Ford K, Poenaru D, Moulot O, Tavener K, Bradley S, Bankole R, et al. Gastroschisis: Bellwether for neonatal surgery capacity in low resource settings? *J Pediatr Surg* 2016;**51**:1262-67.
11. Bradnock TJ, Marven S, Owen A, Johnson P, Kurinczuk JJ, Spark P, et al. Gastroschisis: one year outcomes from national cohort study. *BMJ* 2011;**343**:d6749. DOI: 10.1136/bmj.d6749. PMID: 22089731; PMCID: PMC3216470.

12. Carnaghan H, Baud D, Lapidus-Krol E, Ryan G, Shah PS, Pierro A, et al. Effect of gestational age at birth on neonatal outcomes in gastroschisis. *J Pediatr Surg* 2016;51:734-38.
13. Bielicki IN, Somme S, Frongia G, Holland-Cunz SG, Vuille-Dit-Bille RN. Abdominal wall defects- Current Treatments. *Children (Basel)* 2021;8:170. DOI: 10.3390/children8020170.
14. Rentea RM, Gupta V. Gastroschisis. In: Stat Pearls. Treasure Island (FL): Stat Pearls Publishing; 2023 Jan. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK557894>.
15. Denmark SM, Georgeson KE. Primary closure of gastroschisis: Facilitation with postoperative muscle paralysis. *Arch Surg* 1983;118:66-68.
16. Santos Schmidt AF, Goncalves A, Bustorff-Silva JM, Oliveira-Filho AG, Miranda ML, Oliveira ER, et al. Monitoring intravesical pressure during gastroschisis closure. Does it help to decide between delayed primary or staged closure? *J Matern Fetal Neonatal Med* 2012;25:1438-41.
17. Thompson RJ, Jaffray B. Gastric tonometry after gastroschisis repair. *Archives of Disease in Childhood* 2002;87:339-40.
18. Arafa MA, Elshimy KM, Shehata MA, Elbatarny A, Almetaher HA, Seleim HM. High abdominal perfusion pressure using umbilical cord flap in the management of gastroschisis. *Front Pediatr* 2021;9:706213. DOI: 10.3389/fped.2021.706213.
19. Werbeck R, Koltai J. Umbilical cord as temporary coverage in gastroschisis. *Eur J Pediatr Surg* 2011;21:292-95.
20. Ferdous KMN, Hasan MS, Kabir KA, Zahid MK, Islam MK, Hasanuzzaman M. (). Use of umbilical cord flap for closure of gastroschisis. *Journal of Shaheed Suhrawardy Medical College*, 2018;10:47-50.
21. Okoro PE, Ngaikedi C. Outcome of management of gastroschisis: comparison of improvised surgical silo and extended right hemicolectomy. *Ann Pediatr Surg* 2020;16. DOI: <https://doi.org/10.1186/s43159-019-0012-x>.
22. Negash S, Temesgen F. Primary closure of gastroschisis aided by ileostomy: A new management approach for low resource settings. *J Pediatr Surg Case Rep* 2022; 76:102135.
23. Grabski DF, Hu Y, Vavolizza RD. Sutureless closure: a versatile treatment for the diverse presentations of gastroschisis. *J Perinatol* 2019;39:666-72.
24. Diyaolu M, Wood LS, Bruzoni M. Sutureless closure for the management of gastroschisis. *Transl Gastroenterol Hepatol* 2021;6:31. DOI: 10.21037/tgh-20-185.
25. Elhosny A, Banieghbal B. Simplified preformed silo bag crafted from standard equipment in African Hospitals. *Afr J Paediatr Surg* 2021;18:123-26.
26. Akbar Shirzad M, Tareq Rahimi M, Jurat R, Rahman H. Reduction of gastroschisis using a surgical glove in the absence of standard silos. *J Pediatr Surg Case Rep* 2020. DOI: <https://doi.org/10.1016/j.epsc.2020.101598>.
27. Oyinloye AO, Abubakar AM, Wabada S, Oyebanji LO. Challenges and outcome of management of gastroschisis at a tertiary institution in North-Eastern Nigeria. *Frontiers in Surgery* 2020;7:493403. DOI: <https://doi.org/10.3389/fsurg.2020.00008>.
28. Gomes Ferreira C, Lacreuse I, Geslin D, Schmitt F, Schneider A, Podevin G, et al. Staged gastroschisis closure using Alexis wound retractor: First experiences. *Pediatr Surg Int* 2014;30:305-11.
29. Schlatter M, Norris K, Uitvlugt N, DeCou J, Connors R. Improved outcomes in the treatment of gastroschisis using a preformed silo and delayed repair approach. *J Pediatr Surg* 2003;38:459-64.
30. Ross AR, Eaton S, Zani A, Ade-Ajayi N, Pierro A, Hall NJ. The role of preformed silos in the management of infants with gastroschisis: a systematic review and meta-analysis. *Pediatr Surg Int* 2015;31:473-83.
31. Poola A, Aguayo P, Fraser J, Hendrickson R, Weaver K, Gonzalez K, et al. Primary closure versus bedside silo and delayed closure for gastroschisis: A truncated prospective randomized trial. *European Journal of Pediatric Surgery* 2018;29:203-08.
32. Murthy K, Evans JR, Bhatia AM, Rothstein DH, Wadhawan R, Zaniletti I, et al. The association of type of surgical closure on length of stay among infants with gastroschisis borne <34 weeks' gestation. *J Pediatr Surg* 2014;49:1220-25.
33. Hawkins RB, Raymond SL, St Peter SD, Downard CD, Qureshi FG, Renaud E. Immediate versus silo closure for gastroschisis: Results of a large multicenter study. *J Pediatr Surg* 2020;55:1280-85.
34. Sydorak RM, Nijagal A, Sbragia L, Hirose S, Tsao K, Phibbs RH, et al. Gastroschisis: small hole, big cost. *J Pediatr Surg* 2002;37:1669-72.
35. Wong M, Oron AP, Faino A, Stanford S, Stevens J, Crowell CS. Variation in hospital costs for gastroschisis closure techniques. *Am J Surg* 2020;219:764-68.
36. Hook-Dufresne, DM, Yu X, Bandla V, Imseis E, Moore-Olufemi SD. The economic burden of gastroschisis: costs of a birth defect. *Journal of Surgical Research* 2015;195:16-20.