

Factors Influencing the Survival of Patients with Gastroschisis in A Lower-Middle Income Country: A Retrospective Observational Study

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Keywords

Gastroschisis
Abdominal wall defects
Silo Treatment

Abbreviations

LMIC - Low and middle
income countries

Abstract

Background: Survival rate of gastroschisis has improved worldwide due to advancements in antenatal diagnosis, perinatal care and neonatal intensive care. This is in stark contrast with the persistently poor outcomes observed in low and middle-income countries. Aim of this study is to identify factors affecting survival in a resource constraint center, where intensive neonatal care and parenteral nutrition could not be provided.

Methods: This single-center retrospective observational study was done at our institute from September 2022 to 2023. Data were collected from hospital records. All patients admitted with gastroschisis during the study period were included and grouped into A (survivor) and B (non-survivor). Factors influencing the outcome were compared between the two groups. All patients were systematically categorized based on antenatal diagnosis, age at presentation, maturity, birth- weight and mode of repair. Statistical analysis was done using SPSS version 22.

Results: Among a cohort of 48 patients, only 8 (16%) survived. Twenty patients presented within 24 hours of birth of whom 5 survived. Age at presentation did not significantly impact the outcomes ($P=0.25$). Seven patients were full term and no statistically significant difference in terms of maturity was observed between the two groups ($P=0.05$). A pre-formed silo procedure done in 30 patients yielded a survival rate of 13.3% with four individuals demonstrating positive outcomes. Twelve patients underwent traditional silo procedures and only one of them survived. Among the 6 patients who underwent primary repair, 3 died and 3 survived.

Conclusion: Without adequate intensive care and nutritional support, no technique of closure can improve the survival rate of patients with gastroschisis. Prompt antenatal diagnosis and enhanced perinatal care are critically important to improve the outcome of gastroschisis in resource-constrained settings.

INTRODUCTION

Gastroschisis is a congenital defect of anterior abdominal wall on the right side of umbilical cord causing protrusion of abdominal contents through the defect without any covering of amniotic membrane.⁽¹⁾ Incidence of gastroschisis is 1 in 4000 live birth.^(2,3) The mortality rate ranges between 33 and 100% in the low and middle-income countries (LMIC). In contrast, survival rate is above 95% in high income countries with the advanced antenatal diagnosis, improved perinatal care and intensive neonatal care.^(4,5)

Neonates with gastroschisis are at the risk of losing water and body heat from the exposed bowel, compromised intestinal circulation, and mechanical irritation to the gut.⁽⁵⁾ Factors known to influence the survival of gastroschisis are antenatal diagnosis, time of delivery, gestational age, birth weight, time lapse in reaching the tertiary care center, associated anomalies and the surgical technique.^(6,7) In high income countries, uncomplicated gastroschisis has very low morbidity with a mortality rate nearing zero. But in developing countries like Bangladesh, mortality rate is still high.⁽⁶⁾ In LMIC accessing specialist care is delayed due to absence of prenatal recognition and poor neonatal referral and transport system.⁽⁹⁾ When there is delay in initial management, the exteriorized gut becomes matted and edematous, which makes its replacement into abdominal cavity difficult.⁽¹⁰⁾

Depending on the accommodation capacity of abdomen and the condition of the gut, either primary fascial closure or staged closure (using traditional hand sewn silo or spring loaded preformed silo) is performed to reduce the exteriorized gut and close the defect.⁽¹¹⁾ In comparison to delayed closure, primary repair of gastroschisis is associated with improved neonatal outcomes, including early initiation of enteral feeds, early discontinuation of parenteral nutrition, shorter hospital stay and lower risk of surgical wound infection.⁽¹²⁾ When delayed closure is resorted to, several studies have shown the benefits of pre-

formed silo in low resource center as it is thought to avoid abdominal compartment syndrome thereby circumvents the need of ventilatory support.⁽⁹⁾ However, staged-closure demands longer period of parenteral nutrition as enteral feeding cannot be started until fascial closure is achieved.⁽¹³⁾

Aim of this study is to identify the factors contributing to the survival of neonates with gastroschisis, where neonatal intensive care and parenteral nutrition could not be provided.

MATERIALS AND METHODS

This single-center retrospective observational study was done at Bangladesh Shishu Hospital and Institute from September 2022 to 2023. All new born with gastroschisis admitted to this hospital were included. Neonates with gastroschisis, who were discharged against medical advice, were excluded from the study. Clinical data retrieved from hospital records included age, sex, antenatal and postnatal history, gestational age, birth weight, mode of delivery, clinical findings, time from delivery to hospital admission, and associated anomalies. Management was initiated with resuscitation and stabilization of the neonate by maintaining a warm environment, keeping the baby dry, and preventing heat loss. Immediately after admission, herniated viscera were kept enclosed in a sterile saline-bag. Nasogastric decompression and bladder catheterization were done. Intravenous access was established for fluid resuscitation and broad-spectrum prophylactic antibiotics were started. The choice of surgical procedure (primary closure *vs.* silo application) was decided according to the general condition of the neonate, the size of defect, nature of herniated organs and presence of viscera-abdominal disproportion (VAD). Post-operatively, patients were kept on nothing per oral and nasogastric suction. In case of silo repair, it was squeezed every day and final repair was performed when complete repositioning was possible.

Clinical data were carefully logged in the Excel sheet. All patients were grouped into A (survivor) and B (non-survivor). Factors influencing the outcome were compared between the two groups. The Statistical analysis was done by using a statistical package for social sciences (*SPSS*). Continuous variables were compared with student t-test and discrete variables were analyzed with chi-square test where applicable. A $p < 0.05$ was considered as significant.

RESULTS

A total of 50 neonates with gastroschisis were admitted during the study period. Two neonates were excluded as they were discharged against medical advice and the remaining 48 were included in this study. Among these 31 were male and 17 were female. Only 8 (16%) of them survived. Prenatal diagnosis was made in only 3 patients, among them two (66%) patients survived.

Table 1: Factors affecting the survival of gastroschisis

Factors	Group A n=8	Group B n=40	P
Antenatal diagnosis (n=3)	2	1	-
Age at presentation			
< 24 hr (n=20)	5	15	0.25
> 24 hr (n=28)	3	25	
Maturity			
Term (n=25)	7	18	0.05
Preterm (n=23)	1	22	
Birth Weight in Kg (Mean \pm SD)	2.32 (\pm 0.28)	2.13 (\pm 0.35)	0.56
Mode of repair			
Preformed silo (n=30)	4	26	0.06
Traditional silo (n=15)	1	11	
Primary repair (n=6)	3	3	

A total of 20 patients (42%) were admitted to the hospital within 24 hours of birth; among them 5 (25%) survived. The remaining 28 patients (58%) reached hospital after more than 24 hours of birth; among them 3 survived (10%). There were 23 preterm (48%) and 25 (52%) term neonates (Table 1). Among the preterm neonates, only one survived (4%). However, no significant difference ($P = 0.05$) was seen in terms of maturity in the survivor and non-survivor groups. Mean birth weight was 2.32 ± 0.28 kg in the Group A and 2.13 ± 0.35 kg in the Group B. The small intestines were herniated 100%, the colon in 83%, and the stomach in 60% of the patients. Pre-formed silo application was performed in 30 patients, of which 4 survived. Traditional silo was performed on 12 patients, of which 1 survived. Among the 6 patients who underwent primary repair, 3 survived (Table 1).

Death occurred within 48 hr of hospital admission in 24 patients (60%) and the causes of death in these patients were late presentation (>24 hours of birth), hypovolemia, hypothermia, and sepsis. Among the primary closure group, 2 patients expired within the first 72 hours due to abdominal compartment syndrome, multi-organ failure, and sepsis. In our patients, the diagnosis of abdominal compartment syndrome was made based on clinical signs and symptoms such as abdominal distension, paralytic ileus and oliguria. Duration of hospital-stay in gastroschisis survivors ranged from 1 to 4 weeks.

DISCUSSION

The incidence of gastroschisis has risen throughout the world in the last 3 decades.⁽¹⁴⁻¹⁶⁾ A survey was carried out among pediatric surgeons by Wright and colleagues in 2012. They reported 2 cases on an average per institution per year in low-income countries, while middle-income and high-income countries reported 12 and 15 cases per institution per year, respectively.⁽¹⁷⁾ A study was performed in our institution in 2017 found 75 cases within 3 years.⁽⁶⁾ In the present study, we

found 48 cases within one year. In the absence of formal registry for congenital anomalies in Bangladesh, it is difficult to calculate the exact incidence.



Fig 1. *Traditional silo*



Fig 2. *Preformed silo*

Birth weight is an important factor affecting the outcomes. In a study, no newborn with a birth weight less than 1500 gm survived.⁽¹⁾ We also observed that survival was more in term neonates with higher birth weight, though the difference

was not statistically significant. In developed countries, most of the gastroschisis are diagnosed antenatally and these neonates are delivered in a specialist center where surgery can be performed. In contrast to the high income countries, these defects are diagnosed postnatally at primary health care centers in Bangladesh and then referred to tertiary referral institutes. In our study, 20 mothers had antenatal ultrasound scan at least once during pregnancy. But only 3 had the diagnosis; they all had their delivery at a private hospital from where the neonates were shifted to our hospital within 24 hours of birth and 2 of them survived. We have observed that antenatal sonographers often focus on basic obstetrics parameters and fetal viability rather than detection of congenital anomalies. Similar observation was made by Gurjar.⁽¹⁾ In their series of 58 cases, no one had an antenatal diagnosis. In our study, most of the neonates were delivered at home, primary health-care clinics or secondary health-care centers, and these babies were transported, usually over long distances, without adequate initial resuscitation. Most of these patients arrived at our hospital without a functional intravenous line or any protective covering of the exposed gut. A study by Stevens *et al*/showed that poor resuscitation is a more significant predictor of mortality than postnatal transfer time. They highlighted the importance of adequate initial care.⁽¹⁹⁾ Likewise, the survival rate was less in our study in those who presented after 24 hours of birth. Among the 48 cases, 28 reached our hospital after more than 24 hours of birth and 25 (89%) of them expired.

The intestinal morbidity observed in patients with gastroschisis may be due to the damage of the bowel wall that occurs by exposure to amniotic fluid as well as by constriction of the intestine and its blood supply at the abdominal defect.⁽²⁰⁻²²⁾ Based on this hypothesis, elective preterm delivery by caesarian sections is the standard management in some parts of the world.^(23,24) On the contrary, some authors mentioned that there is no advantage of routine preterm delivery.⁽¹⁸⁾

Baud *et al* found induction at 37 weeks was associated with a lower rate of sepsis, bowel damage, and neonatal death compared with pregnancies managed expectantly beyond 37 weeks.⁽²⁵⁾ In our study term neonates survived more than preterm neonates, though the difference was statistically not significant. It supports the hypothesis that there is no survival advantage of routine preterm elective delivery in gastroschisis.

The abdominal wall closure method is thought to be an influencing factor in the outcome of gastroschisis. Fraga *et al* found that primary closure has benefits over delayed closure.⁽¹⁸⁾ In our study, we did not find any significant difference in the outcomes following different approaches of reduction. In fact, primary reduction is possible only when the intestine is in good condition (thin wall, no peel) and abdominal cavity is adequate. Patients who have the intestine in worse condition (wall edema, thick peel, shortened mesentery) generally undergo silo reduction. Exposed viscera in almost all of our cases (42 out of 48) were swollen and edematous. These patients were in severe hypovolemia and hypothermia. Thus, silo (preformed or traditional) was performed in most patients though primary repair is the treatment of choice. We used traditional silo only when it was difficult to accommodate the contents within the preformed silo bag or when the preformed silo was unavailable. The survival rate did not differ significantly and hence we could not conclude that one closure method was better than the other. However, several studies have shown that the use of preformed silos is associated with lower requirements for mechanical ventilation, reduced time to feeding, lower infection rates and lower risk of abdominal compartment syndrome.^(13,26,27)

In our study, 60% of our patients died within 48 hours after hospital admission. Most of them suffer from hypothermia, sepsis, and acidosis. Our institution, like other developing countries, has significant difficulty in caring for these patients in an appropriate ICU setting due to a lack of

availability of NICU beds. Non-availability of total parenteral nutrition was also a challenge in our country in managing these babies. On the contrary, parenteral nutrition, and neonatal intensive care support were invariably given in developed countries where the survival rate of gastroschisis was more than 95%.⁽⁹⁾

This present study is limited by the relatively small sample size from a single center and by the retrospective nature of the research. The survivor group was too small to reflect any statistically meaningful conclusion. Another limitation is long-term outcome could not be evaluated due to poor follow-up and resource limitations

To conclude, inadequate antenatal diagnosis and delay in initial resuscitation are the important factors that contribute to poorer outcome of gastroschisis. Condition of the bowel, adequate nutritional and intensive care support rather than the operative technique appears to determine favorable outcome.

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