

Gastroschisis: five cases after primary closure at Sanglah General Hospital, Bali, Indonesia



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ABSTRACT

Background: Gastroschisis is a congenital anterior abdominal wall defect characterized by intra-abdominal organs evisceration without covering the membrane. Several risk factors affect the outcome of a gastroschisis patient. Considering these risk factors might also increase the chance of better prognosis and survival. We report five neonates with gastroschisis after primary closure in Sanglah General Hospital, Denpasar, Bali, in 2017. This case series aims to describe the clinical characteristics and outcome of different babies with gastroschisis managed by primary closure surgery.

Case Presentation: Four of our cases were detected during pregnancy with fetal gastroschisis at the age of 32, 19, 23, 25 weeks of gestation, respectively. We reserve cesarean delivery for the patient who did antenatal care in our hospital, but one of our patients was born by spontaneous labor due to lack of antenatal

care. Four cases had the primary closure of less than 2 hours and one of them after 10 hours. Good outcome was found in three cases that started enteral feeding on the 7th day after post closure. Other cases started oral feeding on the 3rd day and 12th day—both of them shown poor outcomes. Three cases survive, but two others died because of sepsis.

Conclusion: We found poor outcomes in both cases, which neonatal sepsis as a significant cause. One of our cases with poor outcome was referred from another hospital without antenatal care and had a long duration of entering the operating room. Diagnosing these babies in their early prenatal period and transferring them to an experienced medical centre containing multidisciplinary working facilities will contribute to both the mother and the baby's health.

Keywords: Gastroschisis, Primary Closure, Outcome.

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INTRODUCTION

Gastroschisis is a congenital anterior abdominal wall defect characterized by the evisceration of intra-abdominal organs without covering membrane through a defect in the form of cleft on the right side of the umbilical cord.¹ There is no reported epidemiological data in Indonesia about the congenital abdominal defect, but the incidence reported worldwide is in 2000 pregnancies and growing in prevalence worldwide.¹ Neonatal mortality rate from 2009 to 2012 was 26.5 per 1,000 live births, from 7 to 12 per 1,000 in developed countries and up to 50 or more per 1,000 in underdeveloped countries.^{2,3}

Nowadays, gastroschisis is almost always diagnosed prenatally during the first and second-trimester ultrasound examinations.⁴ Neonatal survival and the quality of life of children born with gastroschisis are often expressed as excellent. However, this might result from different treatment strategies or cause by the fact of extra-intestinal

congenital abnormalities.⁴

Surgical management of the infant with gastroschisis remains controversial. While the abdominal defect's primary closure is the preferred surgical approach, each pediatric surgeon must subjectively assess the degree of abdominal wall tension anticipated before deciding the repair's nature.⁴ If primary closure cannot be obtained, the alternative management strategy is a staged silo repair. Because of the increased risk of sepsis and hypovolemic shock, primary closure is considered in all cases where reduction does not cause hemodynamic or respiratory compromise. If primary closure is attempted without sufficient space in the abdominal cavity, potential complications secondary to abdominal compartment syndrome may occur.^{2,4} We present the experience of the Department of Child Health, obstetrics and gynecology, and Pediatric Surgery in managing some cases of gastroschisis with numerous outcomes. This case series aims to describe the clinical presentation and outcome

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Table 1. Baseline characteristic of case studies

Parameters	Case 1	Case 2	Case 3	Case 4	Case 5
Perioperative					
a. Gestational age when fetal gastroschisis was found	32 weeks	19 weeks	23 weeks	25 weeks	(-)
b. Gestational age when the baby was born	37 weeks	36 weeks	37 weeks	37 weeks	37 weeks
c. Mother's age	20 years old	26 years old	23 years old	24 years old	22 years old
d. Delivery method	Cesarean section	Cesarean section	Cesarean section	Cesarean section	Spontaneous labor
e. Duration from delivery until the procedure was done (hour)	30 minutes	35 minutes	60 minutes	120 minutes	10 hours
f. Exposed organ	Intestine	Intestine	Intestine	Intestine	Intestine
g. Sepsis risk factor	None	Greenish of amniotic fluid, multifetal pregnancies, premature	-	Greenish of amniotic fluid	-
h. Birth weight	2,750 grams	2,000 grams	2,800 grams	2,240 grams	2,500 grams
Durante Operation					
a. Operative duration	45 minutes	25 minutes	45 minutes	1 hour, 30 minutes	50 minutes
b. Comorbid	-	-	Distended Colon	-	Band
Postoperative					
a. Duration of mechanical ventilation	Only during operation	1 day	3 days	3 days	4 days
b. TPN duration postoperative	3 days	7 days	7 days	7 days	12 days
c. Fullfeed duration	Cannot reach full feed	12 days	10 days	5 days	Cannot reach full feed
d. Sepsis Marker	Leukocytes 0,34 K/uL (neutrophils 0,10 K/uL (28,35%), lymphocytes 0,11 K/uL (32,01%); platelets 3,86 K / mL; IT ratio 1,60; procalcitonin >100 ng/dL).		Neutrophils 11,52 K/uL (79,82%), platelets 34,83 K / mL; IT ratio 0,63; procalcitonin 182,4 ng/dL		Leukocytes 9,82 K/uL (neutrophils 3,9 K/uL (40,63%), platelets 23,67 K / mL; IT ratio 0,44; procalcitonin 4,33 ng/dL).
e. Complication	Clinically late-onset sepsis	Clinically early-onset sepsis	Late-onset sepsis and pneumonia	No complication	Late-onset sepsis
f. Blood culture	No growth	No growth	Klebsiella pneumonia	No growth	<i>Staphylococcus marcescens</i> , <i>Acinetobacter baumannii</i> , and <i>Candida albican</i>
g. Antibiotic	Ceftriaxone, metronidazole	Ceftriaxone, metronidazole, ampicillin, and amikacin	Ceftriaxone, metronidazole, meropenem, vancomycin	Ceftriaxone, metronidazole	Ceftriaxone, metronidazole, ampicillin amikacin, meropenem
h. Duration of admission	11 days	21 days	30 days	15 days	36 days
Outcome	Death	alive	alive	alive	Death

of different babies with gastroschisis managed by primary closure surgery.

CASE DESCRIPTION

We report five neonates with gastroschisis after primary closure in Sanglah General Hospital,

Denpasar, Bali, in 2017. All cases were born by mothers aged between 20 and 26 years old, with gestational age between 36-37 weeks. Only one of our cases didn't do prenatal care and known gastroschisis after birth. The others did prenatal care and known gastroschisis when gestational age

was 19-32 weeks (Table 1). Only one of our cases was born by spontaneous labor and only two of our cases have a sepsis risk factor. The birth weight of our cases was 2,000 grams until 2,750 grams. Primary closure was done immediately after delivery in 30 minutes until 2 hours. Only one of our cases did surgery 10 hours after delivery because the patient was born in another regency (Table 1).

Duration of surgery was 25 minutes until 1 hour 30 minutes with distended colon and band were found in some cases as comorbid. All cases were intubated for 1 until 4 days. Only one of our patients was extubated immediately after surgery. Total parenteral nutrition was given to all patients with a duration varying depending on patient readiness from 3 until 12 days (Table 1). Only two of our patients cannot reach the full feed. Almost all of our cases have sepsis as a complication, and two died due to sepsis as a leading cause. The duration of admission was 11 days until 36 days (Table 1).

DISCUSSION

Prenatal detection is essential to determine post-birth management.¹ With ultrasound examination in the first trimester, these congenital abnormalities can be detected as early as possible. Most gastroschises were diagnosed in the early second trimester (median 18 weeks estimated gestational age) by ultrasonography.^{1,2} We might found cauliflower reflection, which is gastrointestinal loops floating in the amniotic fluid. If any abnormalities were detected, the frequency of fetal evaluation was increased. Patients were hospitalized for fetal surveillance when indicated. Parents were counseled for scheduled cesarean section at 36 to 38 weeks of gestation. Earlier delivery was performed for indications such as worsening fetal status, progressive bowel dilatation and/or, loss of bowel peristalsis, or maternal indications. Multispecialty counseling was provided for all the couples with gastroschisis fetuses. If gastroschisis is detected early, team preparation and child surgery will be better and provide better infant outcomes.⁵ Our results confirm that correct prenatal surveillance and diagnosis are essential in reducing the mortality and morbidity of patients with gastroschisis. In our country, prenatal screening is still far from ideal. People tend to visit our facilities based on complaints. Moreover, our government limits the number of visits that are covered during the prenatal period. Such as our patients that were diagnosed with these abnormalities late. One of our patients underwent spontaneous labor and found with gastroschisis during labor.

The incidence does not vary concerning race or gender. However, it is indicated that being a

mother at a very young age raises gastroschisis risk.⁶ Another aspect regarding the disease is that for a mother to be a smoker.⁷ We didn't find a history of smoking in our cases, but most of them were in their early third decade of life. These findings support the higher possibilities of gastroschisis babies were born from a younger mother. Our case revealed multifetal pregnancies are sharing similar genetic properties but different outcomes. One of the babies had gastroschisis as described, but the other was normal. By this means only, we can't exclude the possibilities of genetically linked to this abnormality.

The goal of delivering the newborn with gastroschisis is to optimize their outcome by minimizing trauma to the exposed gastrointestinal content. The best mode of delivery for these infants remains controversial. From a theoretical standpoint, one might assume delivery by cesarean section would be more advantageous than vaginal delivery for several reasons.⁸ The first reason is a cesarean delivery is thought to produce less compromise to the mesenteric circulation because there may be less compression and twisting of the bowel during uterine contractions and passage through the birth canal.⁸ Another reason is that the risk of infection to the exposed bowel is decreased by cesarean delivery with intact membranes. The last theoretical disadvantage to vaginal delivery is if a large defect is present with possible liver involvement, there may be an increased risk for avulsion injury.⁸ Otherwise, a previous study have concluded that there is no difference in neonatal outcome between cesarean and vaginal delivery, and some institutions prefer spontaneous vaginal delivery as the obstetric policy.⁹ A descriptive study was done in Germany, which involved all pediatric surgery institutions that treat newborns with gastroschisis, the respective national associations of Germany, Austria, and Switzerland were contacted.¹⁰ All pediatric surgeries with associated neonatology were selected. There were 89 hospitals that treat newborns with gastroschisis (Germany 76, Austria 6, Switzerland 7). Overall, 96% of all obstetricians plan an elective delivery, 89% perform planned cesarean section, 2% either cesarean section or vaginal delivery, and 3% vaginal delivery, concerning the timing of scheduled delivery with cesarean section, 87% plan for a late preterm or term delivery with similar frequencies (38%/39%). Only in Germany, 7% plan for a moderately preterm cesarean section.¹⁰ In our centre, we reserve cesarean delivery, but in our cases, one of our patients was born by spontaneous labor due to lack of antenatal care. Not all patients who were born by cesarean had a good outcome.

It is dependent on many factors like postnatal care. In our cases, cesarean delivery was planned at 37th weeks of gestation. This was supported by Baud D and colleagues, who found that induction of labor at 37 weeks was associated with reduced risk of sepsis, bowel damage, and neonatal death compared with expectant management beyond 37 weeks gestation.¹¹

The goal of surgery is to reduce the intestines and other eviscerated organs, followed by abdominal wall closure with acceptable intra-abdominal pressure. The debate over the different strategies is ongoing. A literature review yields only grade-C evidence (small randomized trials with equivocal results) that there is no significant survival difference between immediate and delayed closure.¹² Although surveys have shown geographic variation in closure preferences, primary closure is generally favored over the staged repair.^{12,13}

The transfer of the baby to a centre in time providing antenatal, obstetric, neonatal, and pediatric surgery care coordinately remain a critical issue in the management of the disease. Operating room (OR) was scheduled for the babies 1.5 hours after the planned cesarean section. Babies were received by the neonatologist and examined by the pediatric surgeon immediately after delivery. Initial management of the babies included (a) rapid physical assessment, (b) placement of bowel in the midline to prevent venous engorgement and distension, (c) placement of a nasogastric tube to low intermittent suction, (d) intravenous fluid bolus with 20 mL/kg of normal saline, (e) endotracheal intubation, (f) muscle relaxant, and (g) intravenous administration of ampicillin and gentamicin. Babies were then transported to the OR. A study by Vegunta RK et al revealed the median time to the OR was 112.5 minutes.⁵ Delay of this management could result in a worse outcome for the patient.⁵ In our case, one of our issues, which the baby underwent the procedure after 10 hours, ended up dead because of sepsis. Those undetected prenatally and referred after delivery without preparation for gastroschisis might have experienced significant fluid and heat loss from the exposed bowel before being stabilized in the neonatal intensive care unit.

Neonates with gastroschisis may require mechanical ventilation due to increasing intraabdominal pressure that can cause diaphragmatic elevation leads to a reduction in static and dynamic pulmonary compliance.^{1,2} Therefore, with a volume-cycled ventilation peak, inspiratory pressures increase.^{1,2} The increase in intra-abdominal pressure also reduces total lung capacity, functional residual capacity, and residual volume. These lead to ventilation-perfusion

abnormalities and hypoventilation, producing hypoxia and hypercarbia, respectively.⁸

Early, trophic enteral feeding has a number of theoretical benefits for the neonate recovering from abdominal surgery. Trophic feeding promotes enterocyte maturation, enzymatic activity, and villous renewal. Furthermore, as long as the feeds are of a small, fixed volume, the risks associated with an intestine's enteral nourishment with incomplete functional recovery are likely outweighed by the potential benefits. Decisions regarding readiness for enteral nutrition initiation must be based on the infant's clinical status (abdominal distension, evidence of gastrointestinal function). A study by Thompson JP et al revealed significant positive correlations were found between the time taken to initiate enteral feeding after abdominal closure and each of the primary outcomes (length of NICU stay, duration of parenteral nutrition, and time is taken to regain birth weight).¹⁴

Regarding the onset of feeds, Singh SJ et al review of 181 neonates with gastroschisis, found that those fed within 10 days of operation had a significantly lower incidence of sepsis than those who were fed after 10 days.¹⁵ This decreased incidence of sepsis could be secondary to a shorter duration of the central venous line due to parenteral nutrition in the group that had their feed commenced within 10 days of operation.¹⁵ In our case, a good outcome was found in three patients were started enteral feeding less than 10 days after post-closure, but one of them had a poor outcome due to late-onset sepsis.

There is delayed bowel function in neonates with gastroschisis, which is thought to be related to premature dilatation of the bowel or increased intestinal thickening and matting.⁵ Simultaneously, this immotility persists, it is necessary to maintain bowel decompression by continuous nasogastric suction and the maintenance of nutrition by total intravenous parenteral nutrition. Abdominal distention appeared in almost all of our cases, and hypoperistalsis or adynamic ileus was suggested to be a significant cause.⁸ Adynamic ileus is frequently seen in the postoperative period and may persist for several weeks.⁸ The initial gastrointestinal drainage is characteristically green because of the backup of biliary and pancreatic secretions in the immediate postoperative period.⁸ A large-bore naso/orogastric tube placed to intermittent suction is needed to prevent gastrointestinal distention caused by hypoperistalsis and we also performed nasogastric suction until bowel function returned. We avoided increase pressure of the abdominal, and we start feeding slowly, monitored daily. In our cases, not all of our patients may reach fullfeed. One could not tolerate feeding and developed abdominal

distention and need a long duration of central line usage for TPN.

A study was done by Singh SJ et al revealed that the duration of central line usage for TPN is another critical factor for sepsis.¹⁵ The longer duration of central line usage, the higher the incidence of sepsis was recorded. In our cases revealed sepsis as the main complications that were observed.¹⁵ Although the blood culture did not always show the causing pathogens, clinically, SIRS was apparent and septic markers were increased. This might happen due to abnormalities of the gastrointestinal tract and translocation of bacteria. We believe that these lead to sepsis and causing multiorgan failure and death.^{16,17} The prognosis of gastroschisis is considered acceptable because having associated anomaly is uncommon among the babies with this abnormality.¹⁷ Still, several factors can affect our patients' outcome, such as older age at closure, necrotizing enterocolitis, more prolonged mechanical ventilation, and central-line associated bloodstream infection (CLABSI) were independently associated with longer TPN duration and hospital stay, with CLABSI being the strongest predictor. In our cases, the poor outcome was found in two patients with neonatal sepsis as a major cause of death.

Primary closure may cause abdominal hypertension and abdominal compartment syndrome, leading to ischemia and necrosis of the bowel, renal failure, respiratory distress, and sepsis. Abdominal hypertension is defined as a prolonged or repeated increase in intraperitoneal pressure above 12 mmHg.¹⁹ When the bowels are reduced into the non-sufficiently developed abdominal cavity, the rise of intraperitoneal pressure leads to compression of the blood vessels causing mesenteric ischemia and bowel necrosis.¹⁹ This kind of ischemia occurs very fast because the bowels in gastroschisis have got, in most cases, primarily lower perfusion than the bowels of a healthy infant. Increased intra-abdominal pressure reduces the blood flow in prominent veins leading to diminished heart preload, causing lower cardiac output and global hypoperfusion. This influence renal blood flow making renal hypoperfusion, anuria, and renal failure.¹⁹ In our setting, we used a clinical sign to predict the increase of peritoneal pressure. Abdominal distension, urine production, and breathlessness are some predictors that we can observe in our setting. In our cases, urine production was within normal limit that suggest no abdominal compartment syndrome were found.

There are mainly two reasons for prenatal monitoring of gastroschisis: first, to plan delivery concerning improve the outcome, and, second,

to predict outcome and counsel parents. an interdisciplinary team consisting of obstetrician, neonatologist, and pediatric surgeon are needed in management patient with gastroschisis.

CONCLUSION

We found poor outcomes in both cases, which neonatal sepsis as a significant cause. One of our cases who has poor outcomes was referred from another hospital without antenatal care and had long duration of entering the operating room. Early enteral feeding is suggested in patient's post-primary closure, because can shorter the length of NICU stays, duration of parenteral nutrition, and time is taken to regain birth weight. In our opinion, that terminating these pregnancies is not a necessity. Therefore, diagnosing these babies in their early prenatal periods and transferring them to an experienced medical centre containing multidisciplinary working facilities will contribute to the health of both mother and the baby.

CONFLICT OF INTEREST

There is no competing interest regarding manuscript.

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AUTHOR CONTRIBUTIONS

All authors contribute to the case study from the conceptual framework until reporting the outcome of patients through publication.

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