A Proposed Classification for the Spectrum of Vanishing Gastroschisis

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Abstract

Infants born with gastroschisis in association with intestinal atresia are well described. We are the proposing the classification of vanishing gastroschisis. In this series of six cases, at one end of the spectrum is an infant having gastroschisis with a much narrower defect on the right side of umbilicus. The ischemic bowel loops were connected to bowel inside the abdomen by a fibrous band compressing the exposed bowel mesentery. On the other end of spectrum, an infant having extensive bowel atresia and complete closure of abdominal wall defect (gastroschisis) detected on antenatal ultrasound. These cases should raise awareness of this devastating complication in prenatal management of gastroschisis.

Keywords

- gastroschisis
- intestinal atresia
- abdominal wall defect
- vanishing gut

Introduction

Gastroschisis occurs in approximately half in 1,000 to 4,000 births. Gastroschisis is a ventral abdominal wall defect characterized by extrusion of the midgut from the coelom with variable degrees of malrotation and the absence of a membranous covering. This defect is nearly always located to the right of the umbilical ring. In general, survival is very good, though initial hospital stay is often plagued with numerous morbidities related to the state of the exposed bowel at the time of birth and coexisting abnormalities. One of the most serious comorbidities is likely to be intestinal atresia of varying significance. Intestinal atresia occurs in approximately 10 to 20% of children with gastroschisis.[1] This is a significant complication with great impact on morbidity and mortality for these patients.

Materials and Methods

This study include the six cases from two institutes (West Virginia University Hospital, Morgantown, West Virginia, United States and Cardinal Glennon Children's Medical Center, St. Louis, Missouri, United States) over 8-year period, January 1996 through December 2003 and 40 infants were admitted to the WVU Children's Hospital neonatal intensive care unit (NICU) with the primary diagnosis of gastroschisis. Three infants presented with different stages of vanishing gastroschisis (VG). From April 2008 to December 2009, three newborns were admitted with primary diagnosis of gastroschisis and turn out to be at different stages of VG at the Cardinal Glennon Children's Medical Center.

Detailed Descriptions of the Cases Are As Follows

Case 1

A 20-year-old gravida 2, para 1 mother was followed in high-risk obstetrics clinic with the diagnosis of gastroschisis. Labor was induced at 36 weeks after fetal ultrasound showed progressive dilation of fetal bowel loops. A 2,500-g male infant was delivered by cesarean section. In the delivery room, the exposed bowel was poorly perfused. After initial stabilization, the infant was taken to the operating room for an exploratory laparotomy. In the operating room, the surgeon noted that the bowel was dark maroon or black,

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edematous, and matted together. The gastroschisis defect was noted to very small, less than 4 mm diameter, and located to the right of the umbilicus. It was severely constricting the mesentery. The exposed bowel was connected to the rest of intra-abdominal bowel by a fibrous atrophied intestinal segment where it came through the defect. Operatively, the defect was extended superiorly and inferiorly in the midline. Exploration of the abdominal contents was performed, and the patient was found to have viable bowel from pylorus to the level of the ligament of Treitz and from mid-transverse colon distally. The rest of his bowel was found to be severely ischemic. There was a tight band of tissue around the base of the mesentery above the level of the abdominal wall. The bowel was strictured at this point. This band was divided and it was then realized that it was a scarred piece of bowel wrapping around the base of the mesentery. The divided ends were ligated. The mesenteric vessels were exposed and found to have organized clot in many of the vessels. A silo bag was fashioned around the bowel, and the infant was returned to the NICU. After extensive discussions, the parents opted for comfort care measures only. Comfort care which was decided after discussion with parents includes pain control, no further intervention as well do not resuscitate the child, and no more life supporting medications. The infant died after 5 days.

Case 2

An 18-year-old gravida 1, para 1 mother was followed in high-risk obstetrics clinic with the diagnosis of gastroschisis. Labor was induced at 37 $^{2/7}$ weeks after fetal ultrasound showed progressive dilation of fetal bowel loops and right hydronephrosis. A 3-kg female infant was delivered by vaginal delivery. Her abdominal wall defect was less than 1 cm with the exteriorized bowel loops disconnected proximally and distally (►**Fig. 1**).

On arrival in the NICU, patient was reassessed and taken to the operating room. On exploratory laparotomy, dilated proximal small intestine was hugely dilated and 30 cm in length and bowel lengthening procedure (serial transverse enteroplasty [STEP]) was performed, proximal and distal stomas were created. The exteriorized part of bowel back into peritoneal cavity. The re-exploration was done after 6 weeks and found that it was normal looking bowel with lumen and decision was taken to perform ileocolic anastomosis after checking patency and creating mucus fistula and keeping the original proximal stoma. Recently, she underwent bowel transplant at the age of 2 years.

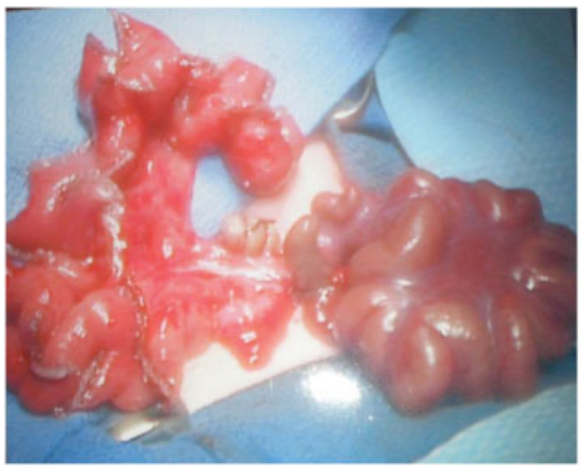

Fig. 1 Type I: vanishing gut with lumen.

Case 3

A 23-year-old gravida 1, para 0 mother was followed in high-risk obstetrics clinic with the diagnosis of gastroschisis. Near term, a follow-up ultrasound showed a decrease in size of the extra-abdominal bowel loops. Labor was induced, and a 3,050-g male infant was delivered. In the delivery room, the gastroschisis was noted to be a small nubbin of tissue; it was atrophied exposed bowel loops protruding through less than 1 cm abdominal wall defect at the right edge of the umbilical cord. No vascular supply was noted. The tissue was removed and microscopic examination showed the remnants of intestinal mucosa. The infant was taken to the operating room the next day. Exploratory laparotomy was performed and found that bowel loop was terminated abruptly at the duodenojejunal junction. There was no evidence of midgut. The large bowel was shortened, of small caliber with a narrow lumen. The abdomen was closed, and the patient was returned to the NICU. He died several days later.

Case 4

A 25-year-old gravida 1, para 0 mother received prenatal care from the second trimester and with the diagnosis of gastroschisis. Mother was presented to the hospital in labor at 33 $^{1/7}$ weeks and female child was born by vaginal delivery with birth weight of 2,122 g and her abdominal wall defect was barely seen with the exteriorized bowel loops disconnected proximally and distally.

On arrival in the NICU, patient was assessed and found the exteriorized bowel loops were atrophic and fibrotic at the multiple places (►**Fig. 2**). Following that patient was taken to operating room and on exploratory laparotomy, dilated proximal small intestine was dilated and approximately 20 to 22 cm in length and bowel lengthening procedure (STEP) was performed and length was doubled to ~40 cm The jejunocolic anastomosis was performed at the level of mid-transverse colon. Following that patient was transferred to transplant center for further management.

Case 5

An 18-year-old gravida 1, para 0 mother was followed in high-risk obstetrics clinic with the diagnosis of gastroschisis. Over a 6-week period, repeat fetal ultrasound examinations showed a disappearance of the extra-abdominal bowel loops. A 3,500-g female infant was born at term. Her physical examination, including abdominal wall, was normal. However, she did not pass any meconium. An orogastric tube was placed, and an X-ray of the abdomen revealed dilated loops of small bowel seen in the upper abdomen. No colonic air was seen. A Gastrografin enema was performed which revealed a small left colon that ended abruptly at the middle portion of the transverse colon. She was taken to the operating room for exploration. The proximal jejunum measured 13 cm from the ligament of Treitz was extremely dilated and came to an

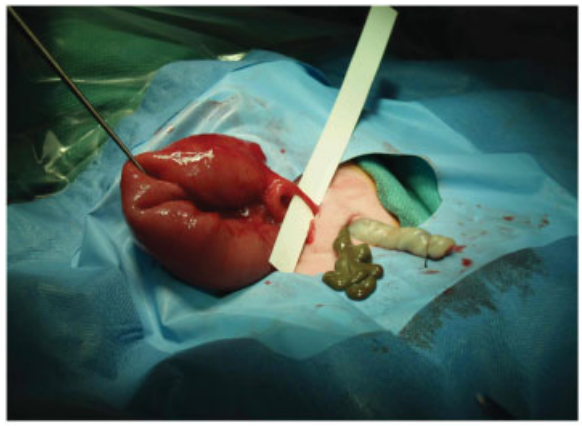

Fig. 2 Type II: vanishing gut without lumen or nubbin of tissue.

abrupt end. There was a fibrous band leading from the end of the proximal jejunum to the umbilicus. Similarly, the left colon was identified which was a microcolon and ended abruptly at the mid-transverse colon. The proximal jejunum and the mid-transverse colon were not connected but were adherent by fibrous tissue. There was no evidence for malrotation. The C-loop of the duodenum was identified, and the ligament of Treitz was in the normal position. The abdomen was closed, and the infant was returned to the NICU. The parents opted for comfort care measures only.

Case 6

A 25-year-old gravida 4, para 3 mother received prenatal care. Antenatal ultrasound done at 20 weeks of gestation showed no abnormality. During pregnancy, she was having poor weight gain with hyperemesis and presented to the hospital with preterm labor. Fetal ultrasound has done a week prior delivery with concern of abdominal distension and repeat ultrasound after 1 week showed double bubble and delivered the male child weighing 2,230 g at 35 $^{1/7}$ weeks through vaginal delivery.

On arrival in the NICU, patient was assessed and 70 mL of bilious fluid was drained after placing an orogastric tube. X-ray abdomen showed finding suggestive of intestinal obstruction. Hence, the patient was taken to the operating room.

On exploratory laparotomy, dilated proximal small intestine was hugely dilated (7 to 8 cm) and microcolon present from mid-transverse colon and whole midgut was absent. On the operating table, a decision was taken to perform jejunocolic anastomosis as there was not enough small bowel to perform either Bianchi or STEP procedure. Following stabilization, patient was transferred after 15th postoperative day to transplant center for further management with regards to small bowel transplant.

Discussion

The term "vanishing gut" has been given to infants with gastroschisis accompanied by extensive midgut atresia and partial or complete closure of the abdominal wall defect detected on antenatal ultrasound. The development of gastroschisis has been studied experimentally as well as clinically.[2–6]

The vanishing gut in gastroschisis appears to be due to ischemic changes of the bowel secondary to compression of the bowel loops and mesentery in a narrowed abdominal wall defect. In our series, cases 1 and 2 (type I) was on the one end of the spectrum showing the compromised exposed bowel loops connected to the rest of the intra-abdominal bowel by a fibrous and atrophied segment of bowel through a tight narrow defect. A scarred, fibrotic piece of bowel was wrapped around the mesentery. The mesenteric vessels were exposed and found to have organized clot in many of the vessels. Cases 3 and 4 (type II) was a small nubbin remnant of atrophied exposed bowel loops. It was protruded through a tiny defect in the abdominal wall to the right edge of the umbilical cord. Cases 5 and 6 (type III) was a complete closure of the abdominal wall defect detected on prenatal ultrasound. On exploratory laparotomy, there was extensive atresia with the proximal jejunum and the mid-transverse colon adherent by fibrous tissue but not in continuity. The details of normal abdominal wall embryology and the pathogenesis of gastroschisis are not completely clear.[7–11] Several mechanisms could explain the findings in our series. First, the anterior abdominal wall defect seen in early gestation may have closed spontaneously around the exposed bowel and its mesentery from progressive narrowing of the defect leading to complete closure. A second possibility is that the fetal bowel loop may have become incarcerated within the defect and become atrophied and causing compression over the mesentery to the remaining exposed bowel loops.[11–14] Third, it could be due to volvulus of the midgut with subsequent infarction and resorption.[3,12,14] The endpoint of all the hypotheses is that the vanishing gut and atresia associated with prenatally closed gastroschisis are the result of an intrauterine vascular accident.

We are proposing the classification of the spectrum of the VG in three types for easy understanding of different types of VG.

1. Type I: vanishing gut with lumen (►**Fig. 1**).
2. Type II: vanishing gut without lumen or nubbin of tissue (►**Fig. 2**).
3. Type III: antenatal evidence of gastroschisis and at birth total absence of midgut.

Above classification may be helpful in antenatal care of the patients with gastroschisis and developing the VG of varying degree, particularly looking for increasing bowel dilatation with bowel wall thickening as well as the size of abdominal wall defect on antenatal ultrasound. Early intervention can prevent the progression of gastroschisis to VG.

From surgery point, VG can be managed differently on the basis of classification.

In type I, the exteriorized bowel that is disconnected from rest bowel proximally as well as distally should be placed back into peritoneal cavity and at the same time, attempt should be made for bowel lengthening of proximal dilated bowel using STEP or Bianchi procedure at the surgeon preference and diverted. Patient should be re-explored after 6 to 12 weeks for the evaluation of exteriorized bowel, which was placed back into peritoneal cavity at the initial surgery and managed accordingly.

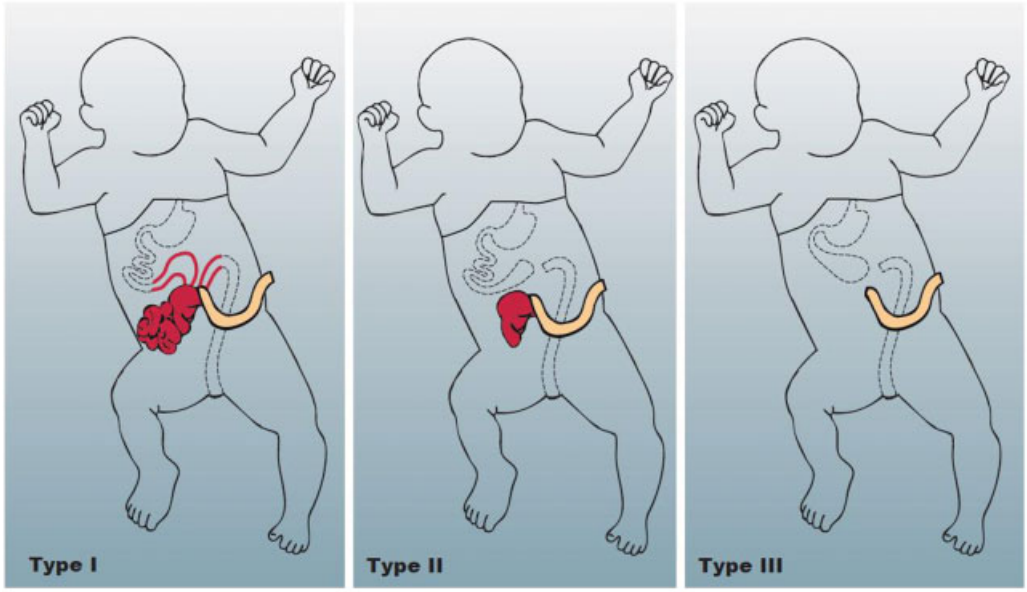

Fig. 3 Proposed classification of vanishing gastroschisis (dotted line showing the intra-abdominal portion of bowel loops, and solid lines and shaded area showing the exposed portion of bowel loops).

In types II and III, there is total absence of midgut with dilated proximal bowel and microcolon beyond the mid-transverse colon. In type II VG, the dilated proximal bowel is larger than type III. During initial exploratory laparotomy, attempt should be made for bowel lengthening of proximal dilated bowel using STEP or Bianchi procedure at the surgeon preference and diverted or connected on the basis of newborn condition and on the choice of surgeon.

Pictorial description in ►**Fig. 3** has shown all the three types on single slide to help understand the types of VG.

The proposed classification will make perinatologist aware of spectrum of VG and also open the venue for future research in the early detection of VG by the following means:

1. During antenatal ultrasound, by comparing the blood flow in the mesenteric blood vessels passing through the abdominal wall defect with previous studies.
2. By comparing the measurement of bowel loop dilatation and bowel wall edema with previous studies.
3. By assessing the size of abdominal wall defect. Nowadays with advancement of ultrasound quality, these are possible measurement going to help in early detection of VG and will help in possible elective delivery as preterm.

From surgery point, it will help surgeons to prepare to make the strategy for surgery as well as for future management in term of bowel lengthening procedure and possible bowel transplantation and parents will also prepared for long-term challenges of this problem.

Acknowledgments

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Conflict of Interest

None

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