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On-line Case Report

Successful management of short gut due to vanishing gastroschisis – case report and review of the literature

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ABSTRACT

Vanishing gastroschisis is a rare in utero complication of gastroschisis. It is associated with a high mortality. We present a case report of an infant with vanishing gastroschisis that was managed with a combination of reconstructive bowel surgery and hepatosparing parenteral nutrition. The technique is described and a review of the literature is provided.

Keywords: Vanishing gastroschisis – Bowel lengthening – Parenteral nutrition

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Gastroschisis is the evisceration of the fetal intestine into the amniotic cavity through a defect in the para-umbilical anterior abdominal wall. The incidence of gastroschisis has been quoted as 1 in 2000.[1] In recent years, however, there has been an increasing incidence particularly amongst younger mothers.[2] The aetiology remains uncertain. One theory suggests that gastroschisis is possibly due to an early vascular interruption in the omphalomesenteric artery.[3] Associations have also been made with the use of recreational drugs.[4] The overall survival rate is greater than 90%.[5]

Vanishing gastroschisis is a rare complication, with only few cases reported in the literature. The abdominal wall defect closes spontaneously, leading to sloughing off of the extra-abdominal bowel *in utero*. This can lead to catastrophic loss of bowel. The case mortality is high, with 11 reported deaths and two fetal deaths *in utero* (FDIU) and five alive at the time of publication. The majority of these were not managed by active surgical techniques.

The authors present a case that had active surgical management from day 1 of life with a successful outcome. At 24 months of age, this patient is now established on full enteral feeds, with no total parenteral nutrition (TPN) or hepatotoxicity and managed as an out patient.

Case history

A 20-year-old woman had a raised α-fetoprotein level on the triple test at 16 weeks' gestation. An early detailed scan showed extra-abdominal bowel and a presumed diagnosis of gastroschisis. At 18 weeks of gestation, an isolated gastroschisis was confirmed and at 33 weeks of gestation, no extra-abdominal bowel loops could be visualised and the intra-abdominal bowel loops were dilated. The maximal dilatation of the intra-abdominal bowel was 40 mm. No loops of extra-abdominal bowel were visualised at any further point. At 36 weeks of gestation, labour was induced. A live male infant was delivered by normal vaginal delivery weighing 2530 g.

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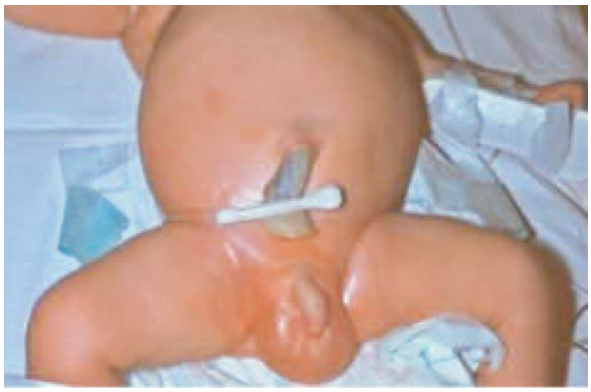

Figure 1 Immediate post delivery photograph showing complete closure of abdominal wall.

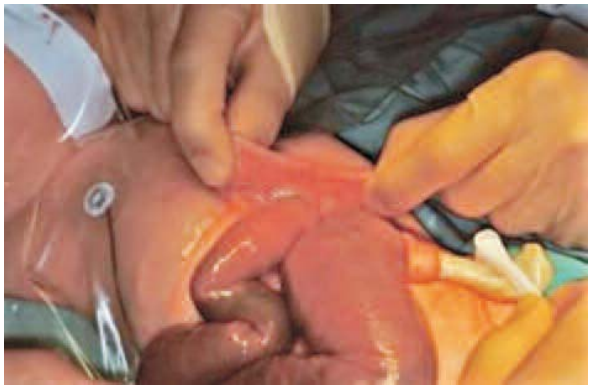

Figure 2 Intra-operative photograph showing dilated proximal jejunum and collapsed distal colon.

On delivery, there was abdominal distension with no defect in the anterior abdominal wall (Fig. 1).

On day 1 of life, a laparotomy was performed which revealed a single, dilated, blind-ending, jejunal loop that measured 30 cm in length (normal, 300 ± 45 cm)[6] distal to the duodenal flexure. There was no ileocaecal valve, ascending or transverse colon. There was an unconnected microcolon (Fig. 2). Due to the serious bowel loss, the patient underwent formation of bowel tube stomas (Fig. 3) for controlled bowel expansion and colonic recycling of jejunal contents.[7] A central line was inserted for administration of liver-sparing TPN. This involved a regimen of 'no fat' TPN for the first few weeks of life with gradual introduction of fat in the TPN to a maximum of 1.0 g/kg/day, 3.75 g/kg/day of protein and 14 g/kg/day of glucose.[7] The total calories derived from this TPN regimen were 81.8 kcal/kg/day. The colonic recycling of jejunal contents was achieved by performing a tube jejunostomy and a tube colostomy. After enteral feeding, the jejunal tube was temporarily clamped to induce partial obstruction of the small bowel, causing dilatation.

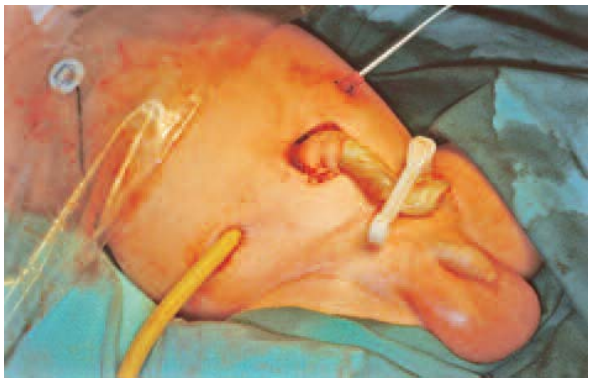

Figure 3 Intra-operative photograph showing tube stomas performed.

This was done in a graded fashion with the length of time of proximal tube clamping increased as the bowel expanded. This was in preparation for a definite bowel lengthening operation at a later date. After 6 months of bowel expansion, a longitudinal intestinal lengthening and tapering (LILT) procedure was undertaken as described by Bianchi in 1980.[8] Total length of bowel after surgery was 75 cm. Enteral feeding was established on day 5 postoperatively and continued alongside TPN. TPN was then gradually reduced to allow TPN 'free' days. By 15 months of age, he was on full enteral feeds. At the age of 2 years, he is gaining weight appropriately with a healthy liver.

Discussion

Vanishing gastroschisis has been attributed to the spontaneous closure of the abdominal wall defect *in utero* causing strangulation and subsequent necrosis of the extra-abdominal bowel loops.[9] These babies are born with large portions of their mid- and hind-gut absent, usually with a blind-ending loop of proximal small bowel and a microcolon with or without a fibrous band connecting them.

Eighteen cases have been identified in the literature. Two of these died *in utero*.[10,11] Six cases died in the first week of life. Two of these had a palliative duodenostomy[12,13] and four cases had no surgical intervention at all.[9,14,15] Five cases died of TPN-related hepatotoxicity. Three of these had a bowel length of 25 cm each.[16–18] The fourth child had 65 cm of bowel[19] and the fifth child had 30 cm of bowel.[20] Five cases were alive at the time of their publication. One child had TPN hepatotoxicity at 8 months and had undergone a Bianchi bowel lengthening procedure.[21] Two children had a bowel lengthening using the serial transverse enteroplasty technique (STEP). One of these two was well at 40 months of age but was still

Table 1 Summary of the outcomes of 18 case reports of vanishing gastroschisis

Reference	Post natal findings	Procedure	Outcome
Pinette (1994)[10]	Agenesis of small intestine		FDIU
Tawil (2001)[11]	56 cm of bowel		FDIU
Johnson (1991)[12]	Agenesis of small bowel	Palliative duodenostomy	Died aged 4 days
Bromley (1995)[13]	Dilated duodenum only	Palliative duodenostomy	Died aged 7 days
Morris-Stiff (1996)[14]	10 cm of jejunum	No surgery	Died aged 2 days
Kimble (1999)[9]	Blind ending duodenum	No surgery	Died aged 7 days
Anveden-Hertzberg (1996)[16]	25 cm duodenojejunal length	End jejunostomy	Died of TPN-related hepatic failure at 10 months
Bhatia (1996)[17]	25 cm of dilated small bowel		Died of TPN-related hepatic failure at 18 months
Celayir (1999)[18]	25 cm of dilated small bowel	Anastomosis	Died of TPN-related sepsis at 4 months
Fowler (1999)[19]	65 cm of dilated small bowel	Anastomosis	Died of TPN-related hepatic failure at 12 months
Basaran (2002)[20]	30 cm of jejunum	Jejunocolostomy	Died of TPN-related hepatic failure at 2 months
Barsoom (2000)[21]	Duodenum and proximal jejunum	Bianchi bowel lengthening at 5 months	At 8 months alive but with TPN hepatotoxicity
Sandy (2006)[22]	30 cm small bowel	Anastomosis/STEP at 30 months.	40 months old and on TPN and oral feeds
Ogunyemi (2000)[23]	15 cm of jejunum	Anastomosis and liver and intestinal transplant	53 months old and well on enteral feeding
Winter (2005)[24]	17 cm of dilated small bowel	Bianchi bowel lengthening/ liver and intestinal transplant	32 months old and on full enteral feeds
Vogler (2008)[15]	3 patients: 2 less than 10 cm and one 23.5 cm	Two with less than 10 cm died without intervention. The child with 23.5 cm had STEP	Child with STEP alive with TPN cholestasis

dependent on TPN.[22] The other child underwent the STEP at 6 weeks of age and presently is on a weaning regimen of TPN but has TPN cholestasis for which he is receiving supplemental fish oils.[15] The other two children were on full enteral feeds at 53 months and 32 months of age.[23,24] Both had a liver and intestinal transplant, one of which had a Bianchi bowel lengthening prior to transplant.[24] A summary of the 18 cases is presented in Table 1.

It can be seen from the few cases in the literature that patients with vanishing gastroschisis have a high mortality and morbidity especially due to TPN-related hepatotoxicity. Only two cases are on full enteral feeds and both have had transplant.

The authors believe that this is the first published case of vanishing gastroschisis that was successfully managed without TPN-related hepatotoxicity and without liver and intestinal transplant and presently on full enteral feeds. An early, active, surgical intervention was employed with clearly defined enteral and parenteral feeding regimens in an attempt to restore sufficient bowel length to allow full enteral feeding whilst preserving the child's liver. The initial fat-free TPN followed by a regimen of fat-reduced TPN (a maximum of 1.0 g/kg/day) was successful in avoiding the hepatotoxicity that is common in short-gut patients. Bowel expansion allowed greater absorption of nutrients, thus decreasing the amount of calories needed from TPN and allowing for weight gain whilst preserving liver function. It was observed that the child had a static weight gain in the first weeks of life as the bowel expansion had not yet reached its optimum size. However, by 6 weeks of life, a steady gain in weight was observed.

The initial stasis in weight in the first few weeks did not have any long-term effects as the child, now aged 2 years, is developing normally without any neurological deficits. The tube stomas were vital in achieving a controlled expansion. Clamping of the proximal stoma allowed graded expansion of the bowel. Recycling into the distal colonic stoma not only enhanced distal colonic adaptation but also promoted the colon to increase in diameter to avoid size discrepancy between proximal and distal bowel in the final anastomosis.

Bowel lengthening procedures should be performed with the hope of achieving full enteral autonomy. The advantage of attempted bowel reconstruction and lengthening is that, even in the event of failure, it may enable the child to reach an age where transplantation becomes an option.[25] This case is the first reported success of vanishing gastroschisis managed by the combination of bowel expansion, lengthening and hepatosparing TPN. The authors advocate the use of these procedures in these

rare cases. Centres taking such cases should have specialised intestinal failure units with multidisciplinary teams experienced in the management of short gut. As both the medical and surgical management of these patients are complex, it is recommended that these cases be centralised to specialist centres.

Conclusions

Vanishing gastroschisis is associated with high morbidity and mortality. Early surgical interventions and hepato-sparing parenteral nutrition may lead to a successful outcome in these patients.

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