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Vanishing gastroschisis: Good outcome after a 10-year follow-up

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ABSTRACT

Gastroschisis (GS) is a full-thickness abdominal wall defect with prolapse of the bowel and sporadically other viscera. The abdominal wall itself may close around the viscera determining entry/exit level intestinal atresia and ischemia or variable midgut infarction, a phenomenon described as vanishing GS (VGS). We report on an infant harboring VGS born to a 35-year-old Gravida 2, Para 0, affected with seizures controlled by anti-convulsive therapy. The infant is well after ten years of follow-up. Guidelines and directives have not been officially issued at international level yet. We suggest that close antenatal monitoring may prevent severe bowel loss in some cases pondering it as the best approach. To the best of our knowledge, after multiple surgical interventions, most patients with closed GS should have a favorable outcome.

1. Introduction

The full-thickness abdominal wall defect, usually right-sided of the umbiculus, with prolapse of the bowel and sporadically other viscera is known as gastroschisis (GS). The abdominal wall itself may close around the viscera determining entry or exit level intestinal atresia and ischemia or variable midgut infarction, a phenomenon described as closed GS or vanishing GS (VGS) [1–6]. Surgically, the resulting associated eventration of an extensive portion of the small and large bowel in the amniotic cavity may be alarming to the Maternal-Fetal Medicine Unit and pediatricians. The gut dislocation is typically accompanied by an abnormal intestinal rotation and/or fixation. Moreover, it may be particularly worrisome according to the defect size and the time the amniotic fluid permeates the intestine [7]. One of the most atypical situations is the combination of GS and intestinal atresia, which may be related to a torsion of the supplying branches of the superior mesenteric artery to the affected bowel. In the scientific literature, the term closing or closed or vanishing gastroschisis has been used for a phenomenon when a gastroschisis closes spontaneously in utero, causing an intestinal atresia, possibly with the persistence of a small extra-abdominal bowel portions [8–12]. Here, we report on an infant harboring VGS born to a 35-year-old Gravida 2, Para 0, affected with seizures controlled by anti-convulsive therapy.

2. Case report

In the fetus of a 35-year-old gravida 2, para 0, affected by seizures, but attack free under anti-convulsive therapy using a Na- and Ca-channel blocker (“Lamotrigine”), a gastroschisis was supposed with a routine ultrasound at the 22nd week of gestation. In the 26th week, the abdominal wall defect was confirmed showing several intestinal loops identified outside of the abdominal cavity (Fig. 1a). At the 28th week, the abdominal wall defect was 28 mm extended, and there was a prolapse of the intestinal loops almost identical to the preliminary investigation, but with beginning intraabdominal dilated bowel loops (Fig. 1b). A sonographic examination was carried out in the 32nd week of gestation and showed similar findings (Fig. 1c), but bizarrely a sonographic analysis in the 34th week revealed a different result with the abdominal wall defect no longer present, and the prolapsed bowel convolute was hardly recognizable. Interestingly, the intra-abdominal bowel loops impressed contrast with marked dilatation suggesting an intestinal atresia (Fig. 1d). A Caesarean section was performed because of the changes in ultrasound findings. The postnatal investigation showed a preterm male baby (body length 44 cm, body weight 2170 g) with a large abdomen. On the right side of the abdomen, in the paraumbilical area, there was a 3-cm long mummified-looking, discolored mesenteric cord with a strand-like bowel portion of 4.5 cm hanging from the abdomen. In this loop, there was the inferior ileum and cecum

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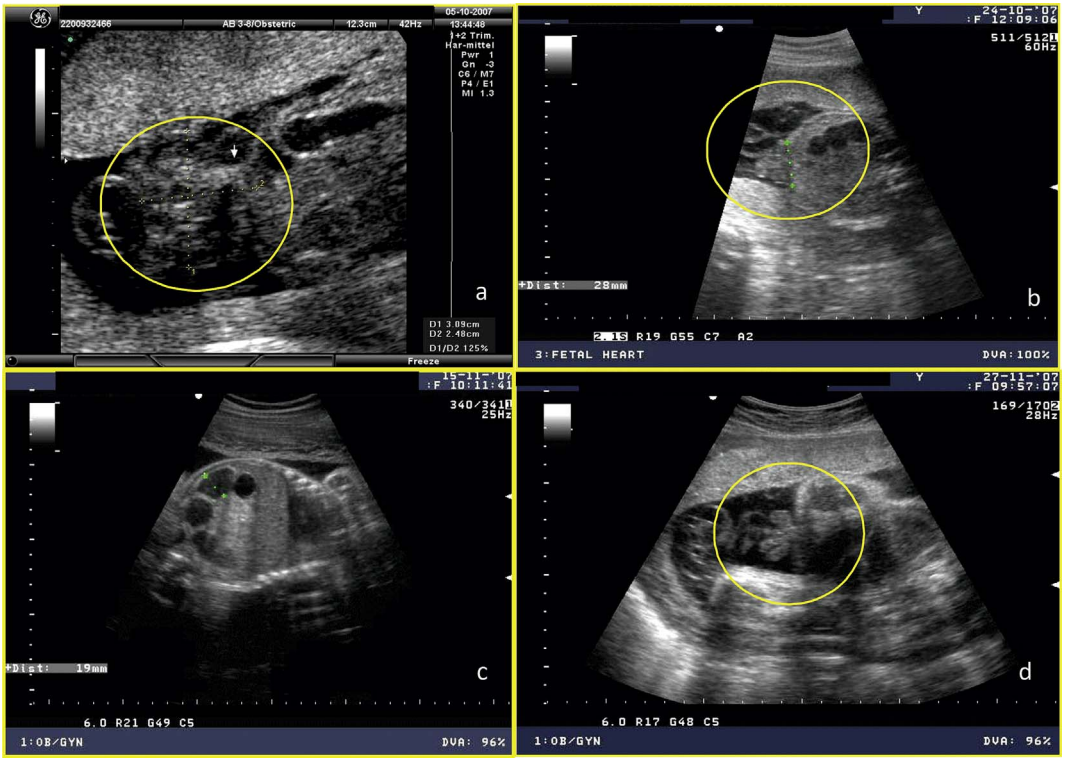

Fig. 1. a. Ultrasound findings at 26 weeks of pregnancy with 28 x 20 mm intestinal convolute in front of the anterior abdominal wall with expanded intraabdominal intestinal loops according to Doppler sonography with good blood flow to the prolapsed intestine. b. Ultrasound findings performed in the 29th week of pregnancy with detectable intestinal convolute in front of the anterior abdominal wall (abdominal wall defect 28 mm in size), which widens the intraabdominal intestinal loops. c. Ultrasound findings at 32 weeks of pregnancy with detection of enlarged bowel loops in the abdomen. d. Ultrasound findings at 34 weeks of pregnancy (immediately prepartal): Intestinal convolution is located in front of the anterior abdominal wall, but the abdominal wall defect can not be shown with certainty.

as blindly closed bowel loops (Fig. 2a–b). Radiographically, there was distension of the rostrally located small intestinal loops compatible with an atresia of the small intestine. Subsequently, an abdominal ultrasound revealed no additional information. During the laparotomy, an atresia of the ileum of type 3A was diagnosed. The diameter of the lumen of the blindly ending ileum was about 5 cm, which the blindly closed aboral intestinal portion (ascending colon) was not superior to 3 mm in size. Th terminal ileum and the cecum were identified with their ileocolic vascular bundles (Fig. 2c–d). This residue had no apparent blood supply. After resection of the remnant the atretic ileum was opened and decompressed, a resection of the distal ileal segment was performed. There was an apparent bluish discoloration, and the surgical team expected that the bowel segment was not going to recover. A resection was completed, and two terminal stomata were built. The histopathologic examination of extra-abdominal located bowel remnants showed necrosis of the terminal ileum over a long distance with superimposed fibrous membranes, although, the cecum was intact and had a mostly unremarkable mucosa (Fig. 2e–f).

The general postoperative course was without considerable problems, but the last massive dilated sling of the ileum showed no signs of recovery, i.e., the lumen remained ident far, a peristalsis did not materialize. The remaining part of the small intestine could only be emptied by ileum flushing enema. On the 9th postoperative day it was decided, therefore, the non-functioning ileal segment, after all, almost 10 cm long to resect and to create a new ileostomy. The course of this engagement was unnoticeable, and the peristalsis could be recorded within a few days. About four weeks later, the stomata were closed, and the bowel continuity was re-established using an end-to-end ileo-ascendostomy. The measured length of the small intestine was about 76 cm. After the transient, about seven-month-long partial short bowel syndrome symptoms, the child received parenteral nutrition. Subsequently, the child developed slowly but increasingly adequately for the age. The boy is now 10-years-old and his length and his weight are about in the 30th percentile of Austrian children. We reviewed the clinical approach and surgical management of VGS using methods used in previous systematic reviews [6,13,14].

3. Discussion

The simultaneous occurrence of gastroschisis with an intestinal atresia is a long-known combination of malformations for whom various causes have been postulated [15]. The genesis of gastroschisis is still not settled, and it may rely on the fact that there are very different conjectures and explanations for the occurrence of intestinal atresia in the setting of gastroschisis. Although a direct connection between the presence of gastroschisis and intestinal atresia has been initially supposed, there are also numerous statements, which did not involve a direct link between the two malformations [15]. According to the literature, up to 25% of children harboring gastroschisis are expected to show also intestinal atresia [7,16]. Some other sources quote that it is an infrequent event occurring in 4.5–6% of cases with any form of abdominal wall defect [4,17,18]. From an embryologic standpoint, the gastroschisis occurs at 4–8 weeks of gestation by an abnormal involution of the right umbilical vein [19]. If Duhamel initially attributed gastroschisis to a localized teratogenic interference with the differentiation of somatopleuric mesenchyme at a time when splanchnic mesoderm and organogenesis could no longer be adversely affected, Bremer credited gastroschisis to somite trauma and consequent failure in the proliferation or distribution of muscle cell primordia [20,21]. The impression that mesenchyme of the body stalk is oddly resistant to resorption is without proper scientific support, while, conversely, the mesenchyme, particularly that of growing somatopleure, is vulnerable in this specific junctional area wherein the umbilical vein involutes [19]. There is also the possibility of an insult of the right omphalo-