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# Pathogenic implications of remnant vitelline structures in gastroschisis

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Yolk sac;
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**Abstract**

**Purpose:** The pathogenesis of gastroschisis is unknown. It may be helpful in understanding its pathogenesis to know the structural relationships among umbilical components including umbilical vessels, urachus, and vitelline structures, and thus, the authors investigated the remnants of vitelline structures in a series of cases of gastroschisis.

**Methods:** Medical records of 41 cases with gastroschisis treated in our institute from 1979 to 2009 were retrospectively reviewed.

**Results:** Paraumbilical bands, possible remnants of vitelline structures, were observed in 4 cases (9.8%). All 4 bands were attached to the skin edge of the abdominal defect without incorporation into the umbilical cord. The band ended at the mesentery in 3 cases and at the antimesenteric site of the ileum in the remaining case. Histologic findings showed fibrous tissues in all cases. One was possibly associated with the development of colonic atresia. Another was noticed after silo reduction when herniated bowels became strangulated by the band. The other 2 cases were uncomplicated.

**Conclusions:** Our findings may support the recently proposed hypothesis that the developmental failure of the yolk sac and related vitelline structures to merge with or to be incorporated into the umbilical stalk might be associated with the pathogenesis of the abdominal wall defect in gastroschisis. Paraumbilical bands derived from vitelline structures may possibly cause intestinal ischemia prenatally or postnatally.


Gastroschisis is an abdominal wall defect that involves paraumbilical visceral herniation usually to the right of the umbilicus. The abdominal wall defect does not involve the umbilicus, unlike other ventral body wall defects including omphalocele. The pathogenesis of gastroschisis is unclear, though several hypotheses have been proposed [1-4]. Recently, new hypotheses have been proposed in which the yolk stalk fails to merge with or to be incorporated into the body stalk [5-6]. We investigated whether there is evidence of a lack of merging between the yolk sac and body stalk in cases with gastroschisis. Such evidence would support the previous reported hypothesis [5-6].

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## 1. Methods

Between 1979 and 2009, 41 newborns with gastroschisis (20 boys and 21 girls) were treated at the University of Tsukuba Hospital (Ibaraki, Japan). The mean gestational age at birth was 36 (range, 26-39) weeks. The mean birth weight was 2102 (range, 880-3080) g. Associated anomalies included colonic atresia (1 case), duodenal stenosis (2 cases), intraabdominal testis (2 cases), a defect in the ileocecal mesentery (1 case), liver cysts (1 case), gastric rupture (1 case), and arthrogryposis multiplex congenita (1 case). Urinary bladder evisceration was observed in 8 cases. The clinical records were retrospectively reviewed, and the remnants of vitelline structures in gastroschisis were studied.

## 2. Results

### 2.1. Meckel's diverticulum

A Meckel's diverticulum was found in 2 cases (4.9%). One was located 10 cm proximal to the ileocecal valve. The location of the other was not described. Both diverticula were found on the herniated loops of intestines.

### 2.2. Paraumbilical bands in gastroschisis

We encountered 4 cases (9.8%) with paraumbilical bands, which attached to the edge of the abdominal wall defect (Table 1). In case 1, the band was attached to the edge of the defect near the umbilicus and ended at the antimesenteric side of the ileum; this band was suspected to be a remnant of vitelline structures (Figs. 1A and 2). In case 2, the band was attached to the lower margin of the defect and ended in the mesentery (Figs. 1B and 2). In case 3, the band was attached to the right margin of the defect opposite the umbilicus and extended with bifurcation to the mesenteries of the colon and ileum (Figs. 1C and 2). In case 4, the band was also connected between the right margin of the defect and the ileal mesentery (Figs. 1D and 2). In all 4 cases, the attachments of the paraumbilical bands to the defect were firm with a wide base, which suggested a congenital rather than inflammatory or adhesive etiology. The locations of the paraumbilical attachments are shown in Fig. 2. The pathologic characterization of these bands was fibrous tissue in all cases.

### 2.3. Clinical courses in infants with paraumbilical bands

In case 1, the band was excised at the primary closure. No associated anomaly or complication was found. The postoperative course was uneventful. In case 2, the band was noticed and removed at the silo reduction surgery. No complication associated with the band occurred. The defect was closed uneventfully at the age of 11 days. In case 3, the patient had arthrogryposis multiplex congenita. At surgery, the band was bifurcated and ended at the mesenteries of the colon and the ileum, respectively. Between the mesenteric attachments of the band, colonic atresia with complete luminal discontinuity was observed (Fig. 1C), and the proximal blind end 3 cm from the ileocolic valve was perforated. The defect was closed and a colonic stoma was created in the right upper abdomen. The stoma was closed 5 months after the initial surgery. In case 4, the band was not noticed at the time of the initial silo reduction using a wound protector and retractor. Soon after the surgery, the bowel within the silo showed severe ischemic changes. When the silo was removed at the bedside, the paraumbilical band was found to be causing strangulation of the herniated bowel. Resection of the band led to an immediate restoration of intestinal blood flow with normalization of the color of the bowel. The clinical course after the closure of the defect at the age of 12 days was uneventful.

### 2.4. Pathologic findings of the umbilical cord in gastroschisis

In all cases, the umbilical cord was located to the left of the defect. In 11 cases, the umbilical cord was removed at an initial surgery, whereas it was preserved in the remaining

**Table 1** Clinical data of cases with paraumbilical bands in gastroschisis

| Case | Sex | Gestation (wk) | Body weight (g) | Mesenteric or ileal attachment of the band | Associated anomalies | Complications |
|---|---|---|---|---|---|---|
| 1 | Female | 38 | 2606 | Antimesenteric site of ileum | None | None |
| 2 | Female | 36 | 1794 | Mesentery of ileum | None | None |
| 3 | Female | 36 | 1850 | Mesenteries of ileum and colon (bifurcated) | Colonic atresia, arthrogryposis multiplex congenita | Perforation of the proximal pouch of colonic atresia |
| 4 | Female | 37 | 3024 | Mesentery of ileum | None | Bowel strangulation by the band |