Duodenal atresia with apple-peel jejenoilial deformity: Case report and review of the literature

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Abstract
Duodenal atresia (DA) with small bowel atresia is an extremely rare condition. The widely accepted theory beyond DA development differs from that for small bowel atresia. We report here a case of a baby having both conditions.

1. Case report
A 2-day-old male neonate (1.9 kg) born at 33 weeks' gestation by cesarean section was referred from another hospital at the age of 30 h because of feeding intolerance, a greenish gastric aspirate, a "double-bubble" sign on an abdominal radiograph, and failure to pass meconium. An antenatal ultrasound scan was normal apart from the presence of polyhydramnios.

On examination, the infant appeared comfortable on room air. Upper abdominal distention was appreciated. The abdomen was otherwise soft and unremarkable. The anus was patent, and the external genitalia were normal. Apart from left microphthalmia, there were no anomalies and no features of Down's syndrome detected. An abdominal radiograph revealed a "double-bubble" sign with a gasless remainder of the abdomen (Fig. 1). Echocardiography was normal.

After adequate hydration with a working diagnosis of DA, an exploratory laparotomy was performed. There was a hugely distended duodenum with atresia at the third part and an apple-peel configuration of the rest of the small intestine, which was twisted around and supplied by a single feeding vessel with the absence of the superior mesenteric artery and dorsal mesentery. The proximal jejunum was in the pelvis and widely separated from the duodenum, while the cecum was intimately adherent to the duodenum by Ladd's bands (Fig. 2). Malrotation was corrected, Ladd's bands were released, and small bowel patency was confirmed by injecting saline at the proximal jejunum that reached the rectum. A side-to-end duodenojejunostomy after spatulation of the jejunal end was done without a transanastomotic tube with single layer, interrupted 5/0 Vicryl sutures; no drains were inserted. The postoperative course was uneventful apart from expected delayed gastric emptying. Feeding was started on postoperative day 11, and the patient was discharged home on milk formula. At follow up at the age of 16 months, he had gained weight and he had no gastrointestinal issues.

2. Discussion
The incidence of intestinal atresia varies according to the site and type. The incidence is 0.8–1.0 per 10,000 total births for DA and
0.7–0.8 per 10,000 per life for jejunoileal atresia [9–12]. Apple-peel atresia is the rarest type of small bowel atresia, occurring in 7%–11% of all patients with small intestinal atresia [13–16]. A familial occurrence of combined DA and apple-peel atresia has been reported, which supports the possibility of a genetic cause for such an association [17,18].

The pathogenesis of DA is believed to involve failure of recanalization at the end of the second trimester following a temporary solid stage. However, the fact that the rat duodenum does not undergo a solid core stage makes the belief of a solid core stage of the human duodenum rather questionable. Atresia of the jejunum or ileum is believed to be due to an intrauterine vascular accident. These long-held theories of failure of recanalization and vascular events have been taken for granted and have not been adequately investigated [1–4].

The number of studies that have explored the vascular hypothesis and the role of vascular impairment in duodenal, jejunal, or ileal atresia is limited. Ligation of the mesenteric vasculature in utero supplying the intestine has been shown to produce atresia in animal models, such as dogs and chickens. Further support for the vascular hypothesis comes from epidemiology studies in which maternal exposure to vasoconstrictors has also been associated with an increased risk of small intestinal atresia. However, the vascular hypothesis is limited in that it does not address how a substantial ischemic event could lead to necrosis of a segment of the intestine despite its extensive vascular anastomoses [3,4].

### Table 1
Outcomes of reported cases.

<table>
<thead>
<tr>
<th>Author (year)</th>
<th>Sex</th>
<th>GA</th>
<th>Age at OR</th>
<th>Weight at OR</th>
<th>Presentation</th>
<th>Associated anomalies</th>
<th>Site of DA</th>
<th>Procedure</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arbell et al. (2006)</td>
<td>Boy</td>
<td>32</td>
<td>2 days</td>
<td>1.5 kg</td>
<td>Double-bubble sign</td>
<td>Choledochal cyst</td>
<td>1st part</td>
<td>End-to-end anastomosis, Appendectomy, Excision of choledochal cyst, &amp; Cholecystoduodenostomy.</td>
<td>Died 2nd day postoperatively</td>
</tr>
<tr>
<td>Ahmad et al. (2009)</td>
<td>Boy</td>
<td>34</td>
<td>8 days</td>
<td>1.2 kg</td>
<td>Polyhydramnios Bilious gastric aspirate Double-bubble sign</td>
<td>–</td>
<td>2nd part</td>
<td>End-to-end anastomosis</td>
<td>Died at age of 41 days due to sepsis.</td>
</tr>
<tr>
<td>Patil et al. (2011)</td>
<td>Girl</td>
<td>33</td>
<td>7 days</td>
<td>1.6 kg</td>
<td>Polyhydramnios Bilious vomiting Jaundice Double-bubble sign</td>
<td>–</td>
<td>2nd part</td>
<td>Duodenojejunalostomy, Release of Ladd’s bands &amp; inversion Appendectomy</td>
<td>Died at age of 41 days due to sepsis.</td>
</tr>
</tbody>
</table>

DA, duodenal atresia; GA, gestational age in weeks; OR, operation; VSD, ventricular septal defect.
Gupta et al. performed an exploratory case-control study that considered the vascular hypothesis by exploring the risks of small intestinal atresia associated with 32 single nucleotide polymorphisms (SNPs) of genes involved in the vascular processes of homocysteine metabolism, coagulation, cell-cell interactions, inflammatory response, and blood pressure regulation. The study's preliminary findings have identified a number of genes that merit further exploration for their association with small bowel atresia [4].

A few reports have described an atresia at the end of the duodenum that extends to the mid-small intestine [6,18–20]. Our patient had atresia at the third part of the duodenum and the typical apple-peel configuration (type IIIb small intestinal atresia) with an absent dorsal mesentery, an absent superior mesenteric artery, malrotation, and Ladd's bands. There was no family history of atresia; however, he had a left microphthalmia and ocular anomalies have been reported to be associated with such a combination of atresia [21].

In our patient, as well as in the four reported cases, the correction of atresia was done by duodeno-jejunoanastomosis without performing a reduction duodenoplasty. Since the introduction of duodenoplasty for the management of duodenal atresia in the early 1980s, there has been no consensus on performing such a procedure in all patients to overcome the problems of megaduodenum. Moreover, biliary injuries after performing tapering duodenoplasty have been reported. Also, in many infants, a dilated proximal duodenum resolves after relief of obstruction without performing a reduction duodenoplasty. However, if megaduodenum persisted and the patient became symptomatic, a future plan was to perform a reduction duodenoplasty either by simple plication, tapering, or serial transverse enteroplasty (STEP) according to the function of the bowel [22,23].

The outcome of intestinal atresia varies according to the site and type of atresia. In general, the outcomes of isolated duodenal atresia and isolated jejunal atresia are excellent, while the outcome of apple-peel atresia is poor [13,15,16]. The outcomes of the reported similar four cases are as follows: two mortalities, one at 2 days and one at 41 days postoperatively. One other patient suffered from failure to thrive, and the fourth patient survived on enteral feeding. Our present patient survived on regular feeding (Table 1).

The plausible explanation is that one mechanism is responsible for creating both anomalies rather than two different mechanisms. It appears likely that under certain circumstances, both duodenal atresia and jejunoileal atresia are caused by a common intrauterine event.

**Conflict of interest statement**

I certify that I have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or topics discussed in this manuscript.

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**References**