

Malrotation in patients with duodenal atresia: a true association or an expected finding on postoperative upper gastrointestinal barium study?

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Abstract. We retrospectively reviewed the imaging and surgical findings in 17 patients with duodenal atresia to determine (a) the frequency of coexistent malrotation in patients with duodenal atresia and (b) the reliability of the upper gastrointestinal barium study (UGI) in differentiating malrotation from postoperative deformity of the duodenal sweep after repair of duodenal atresia. Postoperatively, 9 (53%) of the 17 patients had UGI findings consistent with malrotation. Of these nine, only two had malrotation coexistent with duodenal atresia, while the other seven had normal midgut rotation demonstrated intraoperatively. The radiographic appearance of malrotation was simulated in two patients in whom the ligament of Treitz had been surgically divided, in three in whom the ligament had not been taken down, and in two in whom the status of the ligament was not specified in the surgical report. Although there is an association between duodenal atresia and malrotation, this cannot be accurately documented on postoperative UGI examination. Malrotation cannot be detected preoperatively because contrast material cannot pass beyond the level of the atresia. Postoperatively, surgical deformity of the duodenal sweep cannot be reliably distinguished from malrotation.

Malrotation of the midgut reportedly occurs in 8–34 % of patients with duodenal atresia [1–4]. However, when there is atresia of the proximal duodenum the radiologist is unable to completely evaluate the duodeno-jejunal flexure during preoperative upper gastrointestinal barium study (UGI) because the contrast material cannot get through. Conversely, postoperative displacement and deformity of the duodenal sweep and proximal jejunum can mimic the appearance of malrotation following repair of duodenal atresia. We retrospectively reviewed the imaging and surgical findings in a group of patients

with duodenal atresia to determine the frequency of coexistent malrotation in patients with duodenal atresia and the reliability of UGI in differentiating malrotation from postoperative deformity of the duodenal sweep after repair of duodenal atresia.

Materials and methods

We used a computerized data base of pediatric surgical patients to retrospectively identify 35 children (23 girls, 12 boys) who underwent operation at our hospital during the neonatal period (median age 1 day) for duodenal atresia over a 17-year period. We reviewed the patients' medical records, including the operative report, and recorded the following information in each case: (a) whether the mesentery of the midgut was normal or abnormal and if malrotation of the midgut was present at operation, (b) whether the ligament of Treitz was mobilized during the repair, and (c) whether any other gastrointestinal anomalies were present. Seventeen (49%) of the 35 patients underwent postoperative UGI in our hospital. An experienced pediatric radiologist (J.M.Z.), who was blind to the operative findings, reviewed each UGI using conventional criteria [5–7] for the diagnosis of malrotation. We then compared the postoperative radiological findings with the surgical diagnoses and procedures.

Results

Thirty-four patients had duodeno-duodenostomy and one had duodeno-jejunostomy. Four (11%) of the 35 patients had abnormal mesenteric fixation with malrotation of the midgut diagnosed during surgical repair of duodenal atresia. Ladd procedure was performed in each of these four patients. Two of them also had post-operative UGI and in both of these patients the course and position of the duodeno-jejunal flexure were abnormal and consistent with the typical appearance seen in cases of malrotation (Fig. 1). However, seven other patients who had normal mesenteric fixation demonstrated intraoperatively also had a postoperative radiographic appearance simulating malrotation (Fig. 2). In two of these seven patients the ligament of Treitz had been surgically divided, in three the ligament had not been di-

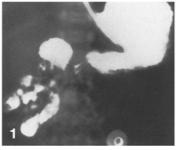




Fig. 1. A newborn boy presented with bilious vomiting and abdominal distension. Abdominal radiographs showed a "double-bubble" appearance with no bowel gas distally, typical of duodenal atresia. At the age of 1 day, upper gastrointestinal barium study (UGI) showed that no barium or gas had passed beyond the proximal duodenum. Duodenal atresia with annular pancreas was found at operation, and duodeno-duodenostomy was performed. Malrotation (without volvulus) was also diagnosed and several Ladd bands were lysed. At postoperative UGI, the duodenum and proximal jejunum were noted to descend completely to the right of the spine, consistent with malrotation

Fig. 2. A girl with Down syndrome and duodenal atresia underwent duodeno-duodenostomy at 2 days of age. Postoperatively, her abdomen remained distended and she continued to vomit. Rectal biopsy was diagnostic of Hirschsprung's disease and she underwent endorectal pull-through. At 7 months of age she again presented with bilious vomiting. At UGI the duodenum and proximal jejunum were dilated and to the right of the midline suggesting malrotation. However, her pediatric surgeon had clearly documented that midgut position and mesentery were normal when he operated on her for duodenal atresia

vided, and in two the status of the ligament was not specified in the surgical report. The sensitivity of post-operative UGI in detecting coexistent malrotation was 1.00. However, the specificity of the postoperative UGI for malrotation was only 0.29 and the overall accuracy was 0.71.

Seven (20%) of the 35 children had additional gastrointestinal anomalies apart from duodenal atresia and malrotation and all underwent postoperative UGI. Five of the seven had coexistent esophageal atresia, and postoperative UGI findings suggested that four of these had malrotation, although this was only confirmed in one child at operation. One child with Hirschsprung's disease also had a radiographic appearance very suggestive of malrotation, but had normal mesenteric fixation demonstrated intraoperatively (Fig. 2). One child with biliary atresia had normal postoperative UGI and no evidence of malrotation at operation.

Discussion

Malrotation and duodenal atresia are among the most frequent causes of bilious vomiting in the neonate [1]. The typical radiographic appearance of a "double bubble" with no gas distal to the duodenum is diagnostic of complete or near-complete duodenal obstruction, most commonly duodenal atresia. Contrast studies of the gastrointestinal tract are usually not required to diagnose duodenal atresia [8–9]. However, malrotation of the

midgut occasionally coexists with duodenal atresia [1–4] (as was the case in 11 % of our patients). It is impossible to detect such malrotation preoperatively at UGI because contrast material cannot pass beyond the level of the atresia [9]. This limitation of UGI is a source of concern in managing these delicate patients because the presence of malrotation indicates more urgent repair than does duodenal atresia alone [1].

Other imaging studies can be useful. Barium enema may show an abnormally positioned or mobile cecum. However, the cecum is normally higher and more medial in the abdomen in the neonate than in older patients [9]. The cecum can also appear to be normally positioned on enema studies, even in some patients with malrotation and midgut volvulus [6, 9]. Inversion of the superior mesenteric vein and artery is a useful sonographic finding and suggests malrotation. However, inversion is present in only about 60 % of cases of surgically proved malrotation [10, 11]. We have also encountered inversion of the mesenteric vessels in neonates with duodenal atresia, without malrotation, presumably because of leftward displacement of the superior mesenteric vein by the dilated duodenum. Coexistent obstruction distal to the duodenal atresia should be suspected when ultrasound reveals dilated, fluid-filled loops of jejunum or ileum [9].

Following repair of the duodenal atresia, the radiographic appearance of malrotation can be simulated at UGI by postoperative deformity and displacement of the duodenum. Although this appearance may result from mobilization or actual surgical division of the ligament of Treitz, it is probably more likely to be secondary to deformity of the sweep related to adhesions and traction on the bowel from the resection of the atretic segment and the anastomosis. A similar appearance of the duodenojejunal loop simulating malrotation can also be seen in patients with distal small bowel obstruction, again probably due to traction on the duodeno-jejunal junction [12]. In some patients, the cecum must also be first mobilized to obtain surgical access to the obstructed duodenum. As a result, the cecum may appear abnormally mobile postoperatively.

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