Duodenal Atresia With an Anomalous Common Bile Duct Masquerading as a Midgut Volvulus

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In a patient with duodenal atresia, a "double bubble" is classically present on plain radiographs. When bowel gas exists distal to the duodenum, duodenal atresia often is excluded from the differential diagnosis. The authors present a case in which contrast can be seen in the small bowel and biliary system on upper gastrointestinal series in a patient with duodenal atresia and an anomalous common bile duct. One always must consider duodenal atresia with an anomalous biliary system as a possible cause of bilious vomiting with a high grade proximal bowel obstruction in a neonate. *J Pediatr Surg 36:956-957. Copyright © 2001 by W.B. Saunders Company.*

INDEX WORDS: Duodenal atresia, small bowel obstruction, malrotation, duplicated common bile duct.

THE HALLMARK radiographic sign of duodenal atresia is the "double bubble" with gaseous distension of the stomach and proximal duodenum and total absence of intestinal gas distally.[1] If small bowel gas is observed distal to the double bubble, the differential diagnosis includes duodenal stenosis, duodenal web, and intestinal malrotation with midgut volvulus. Duodenal atresia also may present with small bowel gas distal to the double bubble when an anomalous biliary system exists. Thirteen documented cases of duodenal atresia with biliary communication between the proximal and distal duodenal segments exist.[2-9] We offer the 14th case of duodenal atresia with an associated biliary duct anomaly as confirmed by upper gastrointestinal series.

CASE REPORT

A 1,532-g boy born at 31 weeks' gestation presented with bilious drainage from the orogastric tube shortly after birth. An abdominal x-ray showed no dilated segments of bowel and a nonspecific gas pattern. An upper gastrointestinal series was obtained to rule out malrotation. The stomach and duodenal bulb were dilated moderately with contrast narrowing at the duodenal bulb. A small amount of contrast also was seen entering the jejunum and opacifying the common bile duct (Fig 1). A preoperative diagnosis of intestinal malrotation was made. At laparotomy, duodenal atresia was found, and a duodenoduodenostomy was performed. Bile was observed in the distal duodenum. The neonate began feeding on postoperative day 8 and was discharged on postoperative day 38 tolerating 205 mL/kg/d of full-strength formula.

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DISCUSSION

Duodenal atresia is assumed to result from failure of recanalization of the duodenum.[10] During fetal development the hepaticopancreatic duct ends blindly in the duodenum. Vacuoles coalesce forming 2 duodenal lumens, each communicating with 2 separate openings in the hepaticopancreatic duct.[11] Boyden et al[12] postulates that delayed recanalization of the bile duct takes precedence over the vacuolization of the duodenal portions creating a "traffic jam," thereby suggesting bile duct entry into the duodenum restricts the recanalization of the duodenum.[12] Eighty-three percent of the time, the obstructed duodenal segment is found at the preampul-

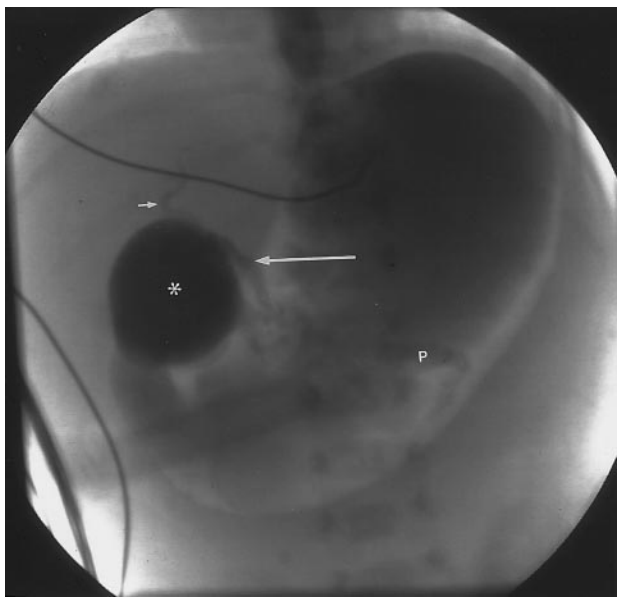

Fig 1. Upper gastrointestinal series shows a dilated proximal duodenum (*), common hepatic duct (short arrow), distal common bile duct (long arrow), and proximal jejunum (P).

lary level. The biliary anomalies described in this case and those previously described by Karpa[3] and Katz[4] can be explained in that the atresia occurred between the 2 orifices of the bile ducts.

Duodenal atresia usually presents as a complete proximal bowel obstruction. In the presented case of a neonate with bilious nasogastric tube aspirate and distal small bowel gas on plain abdominal radiograph, intestinal malrotation must be excluded as the etiology of the bilious emesis. Results of an upper gastrointestinal series showed a dilated proximal duodenum and contrast in the proximal jejunum. However, the presence of distal bowel gas and contrast in the jejunum does not exclude complete duodenal obstruction. Although uncommon, gas bypassing a duodenal atresia via the biliary system can be sufficient enough to give a misleading impression of partial obstruction when, in fact, complete intestinal obstruction exists.[2] This is an important distinction, because a clinical diagnosis of malrotation with a midgut volvulus requires emergent operative intervention.

One must consider duodenal atresia with an anomalous biliary ductal system in the differential diagnosis of bilious emesis with a high grade proximal bowel obstruction. A contrast study is not required if there is a total absence of bowel gas beyond a double bubble. However, in the presence of distal bowel gas, intestinal malrotation must be the leading diagnosis. When performing the duodenoduodenostomy, the enterotomy should be made at the closest point to the atresia to avoid injury to an anomalous ampulla.[13] The papilla is located by observing bile flow, and extreme care must be taken not to injure the anomalous dual ampulla biliary system.

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