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Pyloric atresia: Comorbidity determines outcome☆

Linda M. Schutzman [a], Meggen Walsh [b], John M. Draus Jr. [c,*]

[a] College of Medicine, University of Kentucky, Lexington, KY 40536, USA
[b] Department of Pathology, University of Kentucky, Lexington, KY 40536, USA
[c] Department of Surgery, Division of Pediatric Surgery, University of Kentucky, 800 Rose Street, MN-102 Chandler Medical Center, Lexington, KY 40536, USA

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ABSTRACT

Pyloric atresia is a congenital form of gastric outlet obstruction. The obstruction is relieved surgically, but survival is determined by the presence of additional anomalies. Herein, we report two cases of pyloric atresia. Our first patient was a neonate with epidermolysis bullosa who died of sepsis. The second was an infant who presented with postprandial emesis and had an uncomplicated postoperative course. We discuss the current understanding and management of this rare malformation.

Pyloric atresias are uncommon gastrointestinal defects that represent less than 1% of all bowel atresias. Traditionally, pyloric atresias have been classified by their anatomic morphology. Type 1 atresias are characterized by the presence of a mucosal web between the stomach and duodenum. Type 2 atresias contain a solid longitudinal segment at the pylorus with an intact mesentery. Type 3 atresias are gap aplasias with a disruption between the blind-ending segments of the stomach and duodenum.

The surgical correction of pyloric atresia results in excellent long-term function with few complications when it is an isolated defect. Unfortunately, more than half of these children are born with concomitant anomalies—most commonly epidermolysis bullosa and other intestinal atresias. Such confounding disorders drastically alter the morbidity and mortality associated with this diagnosis.

1. Case report 1

A 1290 g girl was delivered by cesarean section at 33 weeks gestation. The pregnancy had been complicated by polyhydramnios, premature prolonged rupture of the membranes, and late decelerations. Admission radiographs showed a large gastric bubble and absent intestinal gas which was concerning for pyloric atresia (Fig. 1). The baby was found to have fragile skin, especially at areas of friction, which quickly developed into tense vesicles with scattered areas of erosions, bullae, and crusts (Fig. 2A). Early management included placement of an orogastric tube and initiation of parenteral nutrition. A punch biopsy was diagnostic for junctional epidermolysis bullosa (Fig. 2B).

Laparotomy was performed at 1 month of age. We identified a type II pyloric atresia with a 1 cm segment of solid pylorus and an intact mesentery (Fig. 3). A pyloric resection and gastroduodenostomy was performed. Orogastric decompression was discontinued on postoperative day 17. Breast milk feeds were initiated on postoperative day 23 but were soon stopped because the infant developed malabsorption and suspected septicemia. A septic workup was initiated, and prophylactic antibiotics were given. Enteral nutrition was restarted on multiple occasions and always held because of similar consequences. Parental nutrition was continued for a suspected protein-losing enteropathy. Our patient developed intermittent fevers between 38°C and 41°C, and several courses of prophylactic antibiotics were administered. Septic workups never identified an organism. Stool cultures, *Clostridium difficile* assays, and stool leukocytes remained negative. Eventually, she was intubated for a respiratory distress syndrome, and her general condition continued to deteriorate despite maximum respiratory and nutritional support. On postoperative day 66, the medical staff held a care conference with the girl's family. A do-not-resuscitate order was obtained, and the baby died at 15 weeks of age.

* Corresponding author. Tel.: +1 859 323 5625; fax: +1 859 323 5289.
E-mail addresses: jmdr226@uky.edu, lcomb4@uky.edu (J.M. Draus).

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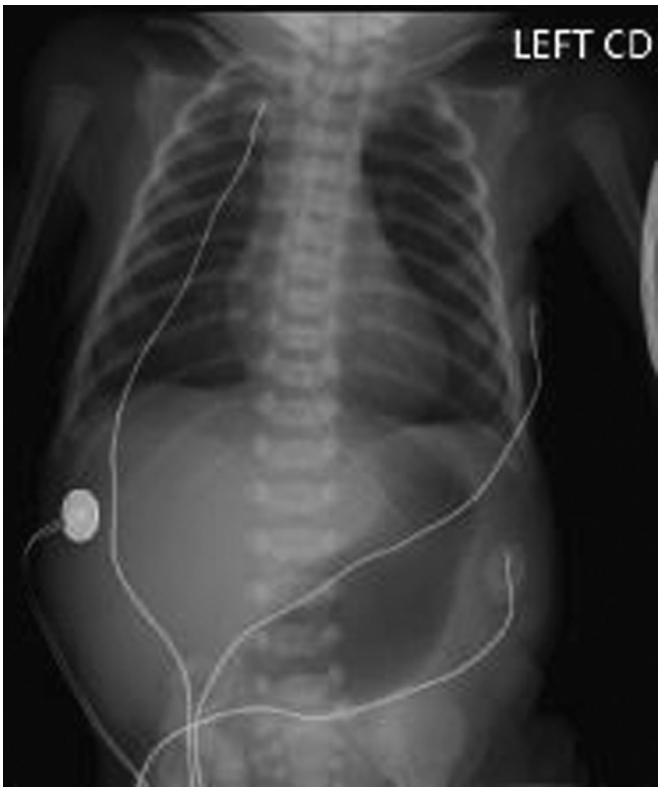

Fig. 1. Abdominal radiograph reveals a large gastric bubble with a paucity of intestinal gas.

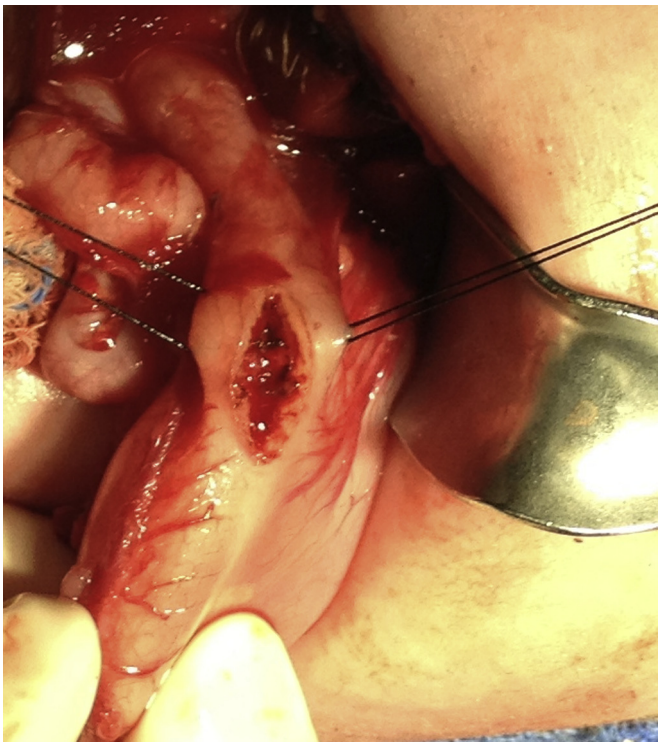

Fig. 3. Intraoperative photograph showing a type II pyloric atresia.

2. Case report 2

A 9-month-old boy presented to the emergency room with a several week history of non-bilious emesis after most feedings. The boy's pediatrician had previously started him on ranitidine for the empiric treatment of gastroesophageal reflux. In the emergency room (ER), an acute abdominal series showed a normal bowel gas pattern with the incidental finding of an opaque foreign body in the left upper quadrant. He tolerated an oral fluid challenge, was discharged home, and instructed to follow-up with his pediatrician.

The next week, he returned to the ER with continued emesis. Our pediatric gastroenterologist performed a flexible endoscopy. A zipper slider and pull tab was removed from the child's stomach (Fig. 4A). The pyloric aperture was found to be quite small, and the neonatal scope could not be advanced through the pylorus (Fig. 4B). Further workup included an abdominal ultrasound to rule out pyloric stenosis and an upper gastrointestinal series which revealed slow passage of contrast out of the stomach (Fig. 5). A pediatric surgical consultation was obtained, and the child was taken to the operating room for exploration. We identified a type 1 pyloric atresia with small perforation within the pyloric web. The web was resected, and a Heineke-Mikulicz pyloroplasty was performed. Oral feedings were resumed on the fourth postoperative day and advanced without complication.

3. Discussion

Pyloric atresias are rare, but well-documented forms of gastric outlet obstruction. The exact etiology of this anomaly remains unknown. An autosomal recessive inheritance pattern is displayed in those with a familial predisposition for pyloric atresia [1]. The

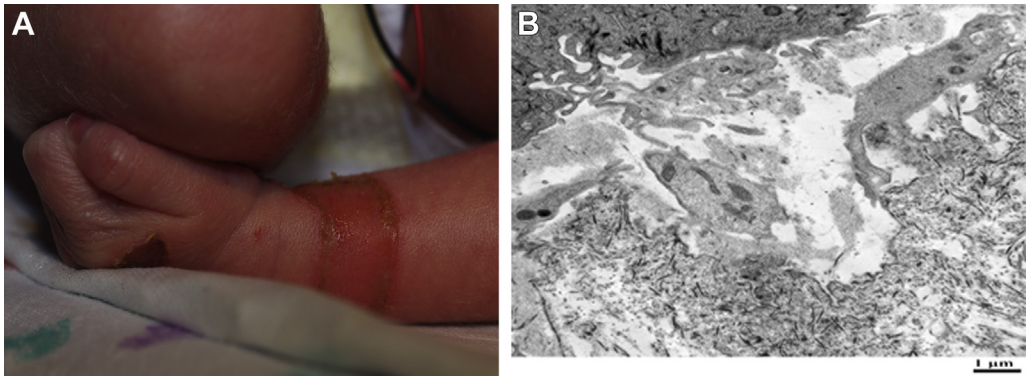

Fig. 2. A) Photograph illustrating the skin lesions of our patient with epidermolysis bullosa. B) Electron micrograph of our patient's skin biopsy. Note the widened dermal/epidermal space. There are areas of lamina densa present; however, the lamina lucida is missing multifocally.