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Congenital esophageal stenosis in 3 children: A case series☆

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ABSTRACT

Congenital esophageal stenosis (CES) is rare condition found in 1 per 25,000 to 50,000 live births. It is characterized by intrinsic narrowing of the esophagus secondary to congenital malformation of the esophageal wall architecture. Diagnosis is often difficult to definitively establish as the symptoms are often initially attributed to esophageal strictures secondary to reflux, or occur within the context of a tracheo-esophageal fistula (TEF) in the newborn. Endoscopic dilation and surgical repair are the mainstays of treatment. We report a series of three cases seen recently at our institution, the University of Alberta/Stollery Children's Hospital.

1. Introduction

Congenital esophageal stenosis (CES) is a rare condition found in 1 per 25,000 to 50,000 live births [1]. It is characterized by intrinsic narrowing of the esophagus secondary to congenital malformation of the esophageal wall architecture [1]. Diagnosis is often difficult to definitively establish as the presenting symptoms are often attributed to esophageal strictures secondary to reflux, or are discovered during the repair of a tracheo-esophageal fistula (TEF) or in the post-operative period on contrast esophogram. Endoscopic and surgical management are the mainstays of treatment. We report a series of three cases recently seen at our institution, the University of Alberta/Stollery Children's Hospital.

2. Case report

2.1. Patient A

8-month-old female initially presented to a rural community hospital with intermittent vomiting with both solids and fluids since birth, initially attributed to colic and reflux. The vomiting became progressively worse with significant weight loss over a few weeks. There was no other significant medical history and no abnormalities on physical exam. An upper GI series revealed esophageal stenosis (see Fig. 1). The patient was referred to a Pediatric Gastroenterology service, who performed upper endoscopy and balloon dilatation. Post-procedure the patient became restless, tachypnic and febrile. A chest radiograph revealed soft tissue emphysema in the neck and mediastinum. Further investigation with a barium swallow revealed a small amount of extravasation from the lateral esophagus into the mediastinum, without extension into either pleural cavity. She was started on broad-spectrum antibiotics and kept NPO, with the plan to manage conservatively. Later that evening she clinically deteriorated and a CT chest showed worsening pneumomediastinum with a small effusion. In consultation with the pediatric surgical service, the patient was taken to the operating theater for exploration and definitive management. An esophageal perforation was identified, located at a long mid-esophageal stricture (see Fig. 2). Given the extent of mediastinitis, the patient underwent a transhiatal esophagectomy, gastric tubularization with cervical gastroesophageal anastomosis and feeding J-tube insertion. She did well postoperatively and started J-tube feeds on post-operative day four. Surgical pathology did not specify the subtype of the stricture, nor did they see any signs of GERD

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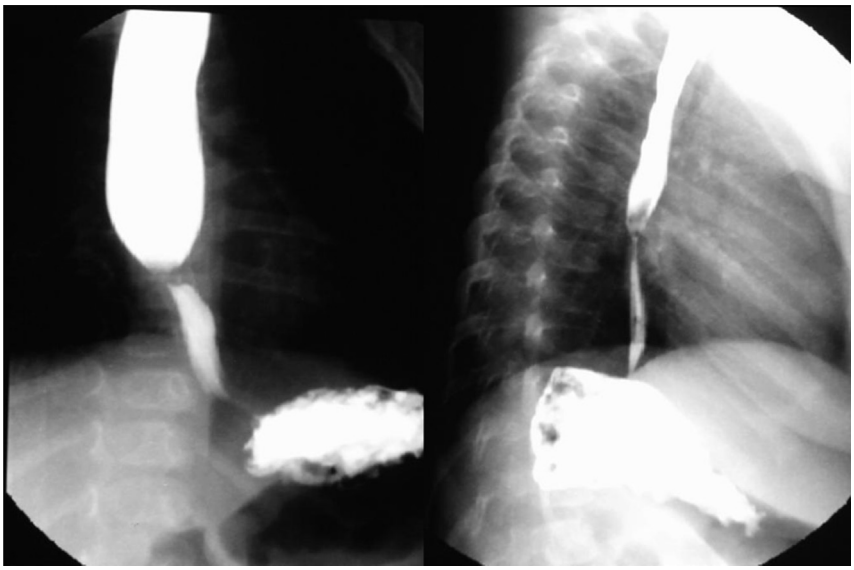

Fig. 1. Patient A - Initial barium swallow.

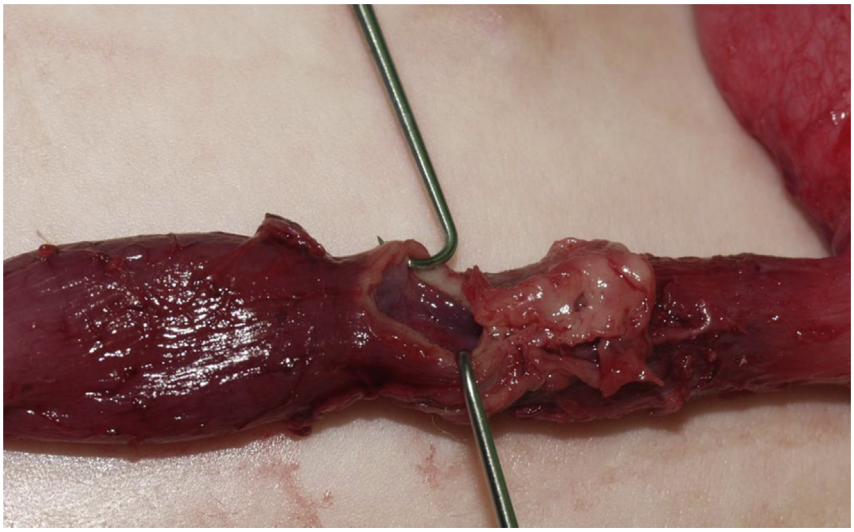

Fig. 2. Patient A - Perforation at mid-esophageal stricture.

within the stomach or esophagus. Of note, the initial endoscopy reported the presence of a web during the procedure. The patient required five endoscopic dilatations in the 6 months following the operation. After this point she moved away and was lost to follow-up.

2.2. Patient B

1-year-old male with a history of tracheomalacia, laryngomalacia, GERD, vomiting since birth and previous difficulties with NG tube insertions. He was investigated for his chronic emesis/regurgitation with an esophagogram, which revealed a high-grade distal esophageal stricture. Further work-up via gastroscopy revealed an additional proximal esophageal stricture. The patient went on to have a G-tube placed endoscopically and the first of several esophageal balloon dilatations. Approximately one month from the time of diagnosis, the decision was made to surgically manage the stricture, as there were ongoing concerns about feeding, feeding tube care, and geographic difficulties. He underwent resection of the stricture and GE junction with an esophagogastric anastomosis, partial anterior fundoplication and insertion of a balloon gastrostomy. Postoperatively, he continued to have feeding difficulties, failure to gain weight, severe reflux symptoms and issues of esophageal dysmotility. Repeat endoscopy revealed a patent anastomosis and changes consistent with reflux. Approximately 1.5 years after the initial operation, a transhiatal esophagectomy with gastric tubularization and pull-up, with pyloroplasty was performed. In addition, bilateral truncal vagotomy and feeding jejunostomy were performed. After the second surgical repair, there was mild narrowing at the new anastomosis, which required minimal dilatation. The patient was eating well and gaining weight and the feeding tube was removed. To date, the patient has not required any dilations within the past year. Surgical pathology confirmed fibromuscular hyperplasia.