CONGENITAL ESOPHAGEAL STENOSIS: A REPORT ON TWO CASES

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Abstract

Introduction. Congenital esophageal stenosis (CES) is a very rare malformation, occurring in 1:25.000 – 1:50.000 live births.

Purpose. We report two cases of CES treated in our unit.

Materials and Methods. The charts of two patients having CES were retrospectively studied, special attention being paid for clinical aspects, radiological features, surgical treatment and results.

Results. Both of our patients were admitted for repeated episodes of vomiting and dysphagia. The esophagogram revealed the stenotic lesion, without any episode of gastro esophageal reflux. They initially underwent bougienage dilatation, with no significant effect. In one case we performed the stenotic segment removal followed by eso-esophageal anastomosis. In the other one we replaced the long stenotic portion of the esophagus with distal ileum. The histopathology study found fibro muscular thickening. The postoperatory esophagogram showed good results in both cases.

Conclusions. The curative treatment of our cases was represented by surgical removal of the stenotic esophageal segment. In cases with long, distal esophageal stenotic segment, the replacement with distal ileum is a good alternative.

Key words: congenital esophageal stenosis, esophageal substitution with ileum.

Introduction

CES is a consequence of a congenital malformation of the esophageal wall architecture (1). This rare condition was found in 1:25.000 – 1:50.000 live births, the incidence being higher in Japan (3,4). Associated anomalies, including esophageal atresia, cardiac anomalies, intestinal atresia, anorectal malformations, and chromosomal anomalies, were found in 17% - 33% of cases (1,2). More and more CES cases successfully treated with conservative meaning (bougienage or balloon dilatation) were reported, although with a significant consequent morbidity (5,6,7). In failed dilatation cases surgical removal of the lesion is mandatory.

Herein we report our experience with 2 CES patients treated in our hospital.

Materials and Methods

Between 2008 and 2009, two patients (4 months and 2 years of age) having CES were diagnosed and treated in our hospital (one boy and one girl). In both patients the diagnosis was made performing an esophagogram. In this retrospective study we reviewed the clinical picture, the therapeutic particularities and the results.

Results

The first case, a 4 month old female, was admitted in our unit for repeated episodes of vomiting, treated in another medical unit as GER. The esophagogram revealed a tight, stenotic, portion located in the middle of the thoracic esophagus, with no gastro-esophageal reflux (fig.1). A bougienage dilatation was attempted twice, without results. We performed the stenotic segment resection (about 2 cm) and end-to-end esophageal anastomosis (fig.2-4), with good results (fig.5,6). The pathological examination revealed a fibro muscular thickening of the resected esophageal segment.

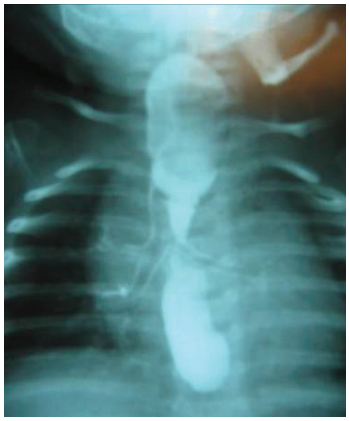
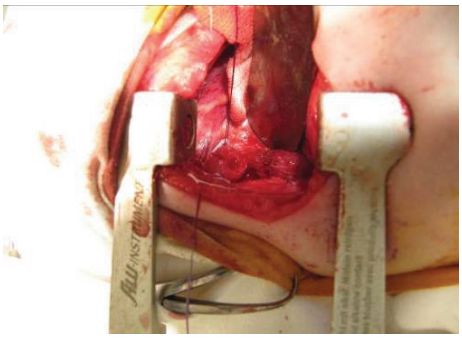


Fig.1: preop. esophagogram.



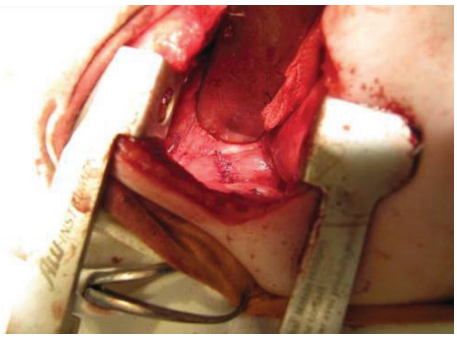


Fig.2,3: intraop findings, eso-esophageal anastomosis.

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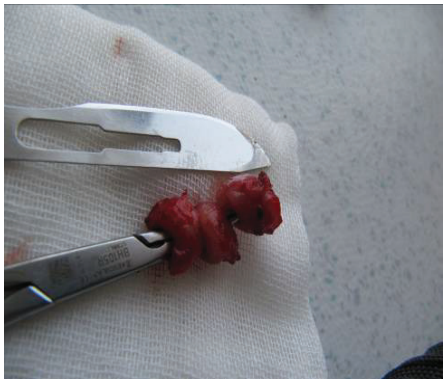


Fig 4: the stenotic segment removed.

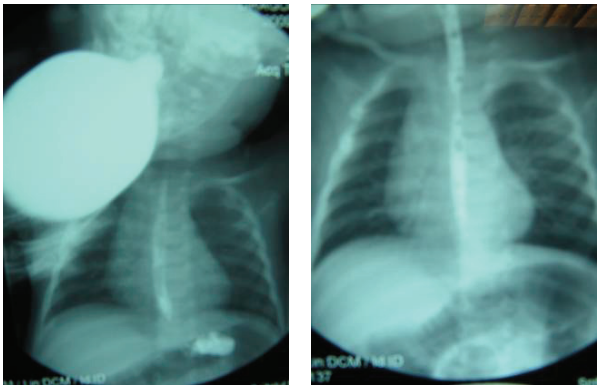


Fig. 5,6: postop esophagogram: no stenosis, no GER.

The second case was a 2 years old boy having Down syndrome associated with ventricular septal defect (previously operated) and cleft palate (previously operated). He was admitted in our department for repeated episodes of vomiting. The esophagogram (fig.7) showed a tight, long (about 4 vertebral bodies) stenotic segment. A bougienage was attempted, without results. In this circumstances we decided to operate him and to remove the stenotic segment (fig.10,11), performing a low esophageal substitution with distal ileum. The blood supply of the graft was given by the ileocolic a., preserving the junction with right colic a. We preserved the last 2 cm of the ileum and we performed an ileum-ileum anastomosis (fig.8,9). The postoperatory esophagogram (3 months postop) showed satisfactory results (fig.12).

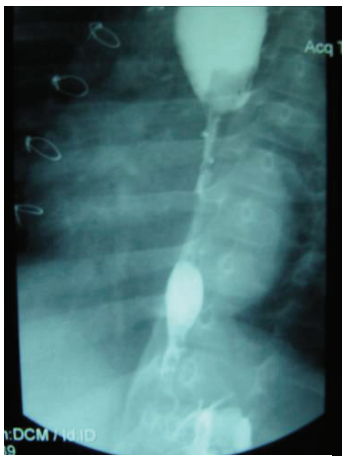


Fig.7: preop. Esophagogram.

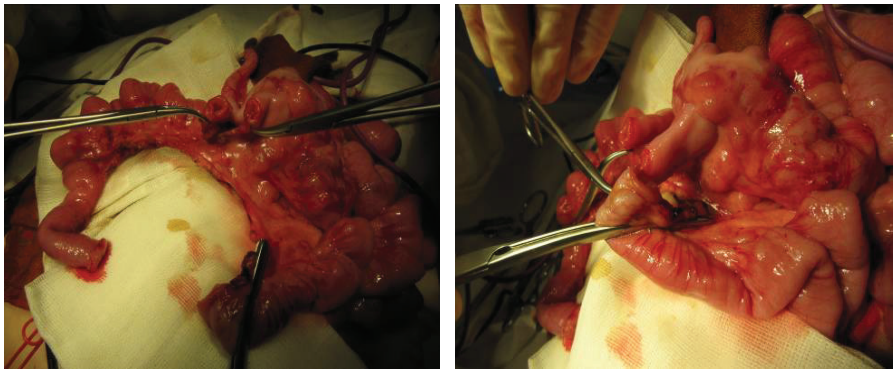
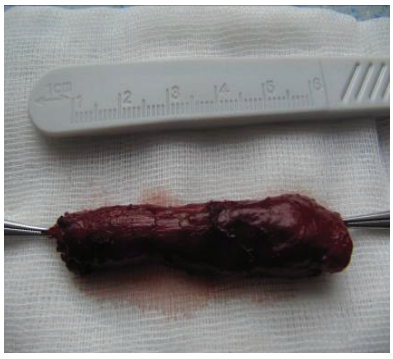


Fig.8,9:the ileal graft (the blood supply given by the ileocolic a.)



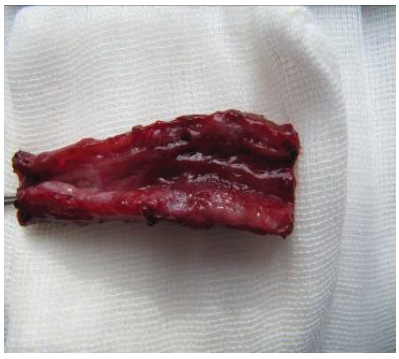


Fig.10,11: The esophageal stenotic segment.

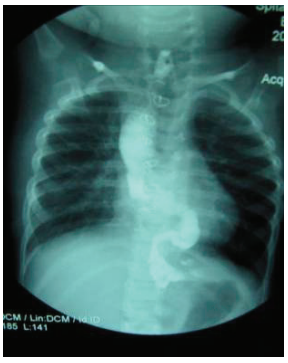


Fig.12: Postop esophagogram.

The pathologic exam revealed a fibro muscular thickening of the esophagus. An important aspect was that the distal thoracic and the abdominal esophagus removed together with the stenotic portion had a normal appearance, with no signs of reflux disease.

Discussion

Despite its rarity, CES must be taken into account in infants having repeated episodes of vomiting. The esophagogram, together with the clinical picture are highly suggestive for the disease. There are 3 histologically types of CES: 1-fibromuscular thickening, 2-tracheobronchial remnants, and 3-membranous webbing. There are 2 possible therapeutic options for this anomaly: dilatation (bougienage or balloon) and surgery. Shintaro and coworkers summarized, on 115 cases founded in 28 English-language studies that about 70% of the cases required surgical treatment (1). When the stenotic segment is relatively short, it is feasible to remove the lesion and to perform an end-to-end anstomosis. In this perspective, a very useful tool is represented by the traction sutures, as are described by Foker (8). When dealing with a long segment, involving the lower half of the thoracic esophagus, we found that esophageal substitution with distal ileum is an effective alternative, the blood supply being offered by the ileocolic a.

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