

## Case Reports

# Congenital Esophageal Stenosis Due to Tracheobronchial Remnant: 2 Cases Report and Literature Review

HY WONG, KW CHAN, KH LEE

**Abstract** Esophageal stenosis is a rare congenital anomaly. Definitive pre-operative diagnosis is difficult to be made and its therapeutic management is still controversial due to the scarcity of cases. We report the presentation, investigations and management of two Chinese children who suffered from isolated congenital stenosis at lower esophagus causing feeding problems. Both patients remain asymptomatic after radical surgery.

**Key words** Congenital esophageal stenosis; Dysphagia; Tracheobronchial remnant

### Introduction

Congenital esophageal stenosis (CES) is a rare entity found in 1 per 25,000 to 50,000 live births; however its true incidence may be underestimated.<sup>1</sup> Its aetiology remains unknown, but embryologic origin has been suggested. CES is characterised by an intrinsic luminal narrowing of distal segment caused by congenital malformation of esophageal wall architecture. There are three pathological subtypes and the commonest type is tracheobronchial remnant.<sup>1,2</sup> Affected infants are usually asymptomatic during the neonatal period but are present with vomiting when introducing solid food in their early childhood. Due to its rarity and common presentation, it poses diagnostic challenge to both paediatrician and paediatric surgeon. We herein present two isolated cases of CES in this locality.

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### Case 1

A 31-month-old Chinese boy was admitted to the paediatric ward with a diagnosis of pneumonia. A detailed history disclosed that he had repeated vomiting for two years after meal. Contrast swallow was subsequently arranged, and rat tail sign was shown which was suggestive of achalasia (Figure 1a). Endoscopy has been performed and a tight stricture was found at distal esophagus which could only allow a 5.2 mm scope to pass through. Mucosal appearance was normal and balloon dilatation was attempted. His symptoms had been partially relieved after the dilatation. Two more attempts of dilatation had been performed within 3 months but no significant progress was observed; thus radical surgery was decided. Laparoscopic resection of stenosed esophageal segment with end-to-end esophageal anastomosis together with anterior fundoplication was performed when he was 42-month-old. A short course of prokinetic was started during the immediate post-operative period in view of the persistence of high output from nasogastric tube. Feeding was slowly stepped up and he was finally discharged on day 18 after the operation. The histology of the operative specimen revealed tracheobronchial remnant. Contrast study after 2 months of surgery showed smooth passage of contrast. After 5 years of follow-up, patient has been healthy with satisfactory weight gain.

## Case 2

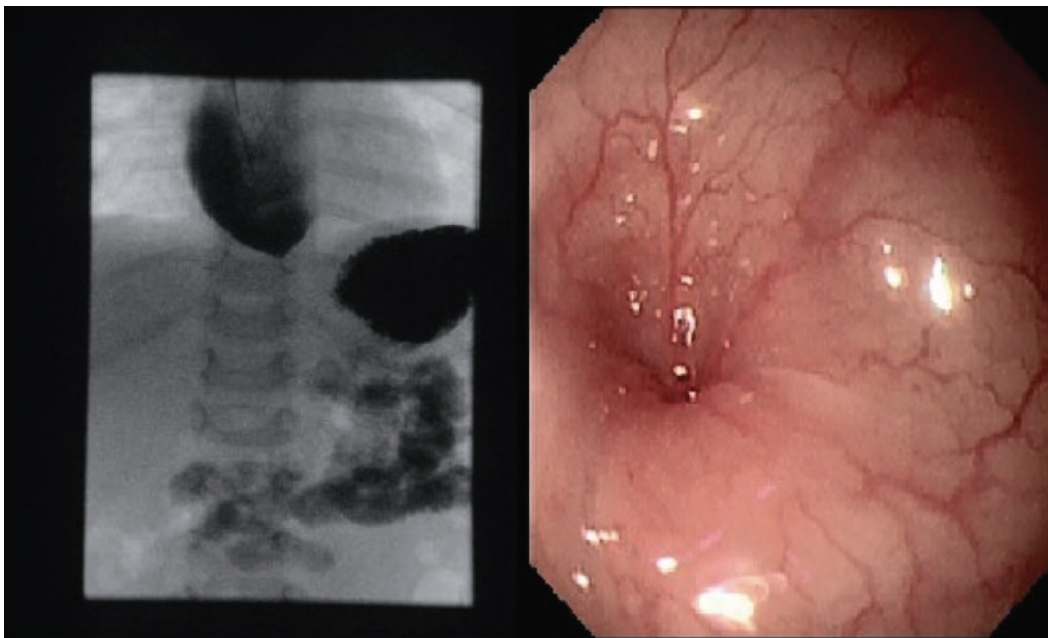
A 14-month-old Chinese girl presented with episodic vomiting after the introduction of solid food, since she was 6 months of age. She was first assessed by paediatrician with the chief complaint of repeated vomiting. Her body weight had dropped from 90th percentile to 25th percentile within 5 months. Nasogastric tube feeding was introduced for nutritional support. An upper gastrointestinal contrast study was performed and showed an abrupt narrowing of distal esophagus with features suggestive of achalasia. Computer tomography showed dilated esophagus without any extrinsic compression and esophageal thickness was within the normal range. In view of suspected achalasia, esophagoscopy was performed which showed a stenosis at distal esophagus with normal mucosa, which further passage with 5.2 mm scope was not feasible (Figure 1b). Balloon dilatation had been attempted but was ineffective. Worsening of symptoms was observed as she occasionally could not swallow her own secretion. In order to minimise the chance of aspiration, a naso-esophageal tube had been inserted for regular suction, after which the condition was much improved. Laparoscopic resection of the stenosed esophagus with end-to-end anastomosis followed by an anterior partial gastric fundoplication was performed when she was 15 months old. Post-operative recovery was

uneventful with oral feeding resumed on day 7 and finally discharged on day 8. The histological examination of the operative specimen (Figure 2) found tracheobronchial remnant. Two months after the surgery, patient could tolerate solid meals without choking or vomiting. However a longer period of follow-up would be essential.

## Discussions

The first case that coined the term congenital esophageal stenosis in the literature was reported by Rossi in 1826. It was a case of distal esophageal membrane which is a subtype of this disease entity. CES is divided into 3 categories according to the histology: (1) tracheobronchial remnant (TBR); (2) fibromuscular hypertrophy with segment muscle thickening and fibrosis and (3) membranous webbing (MW).<sup>2</sup> TBR is the commonest among all the cases and is thought to be a developmental failure in the separation of respiratory system from the foregut. The involved esophageal segment nested cartilage and respiratory epithelium. MW is extremely rare and has been proposed to be due to the incomplete reformation of esophageal lumen upon recanalisation.

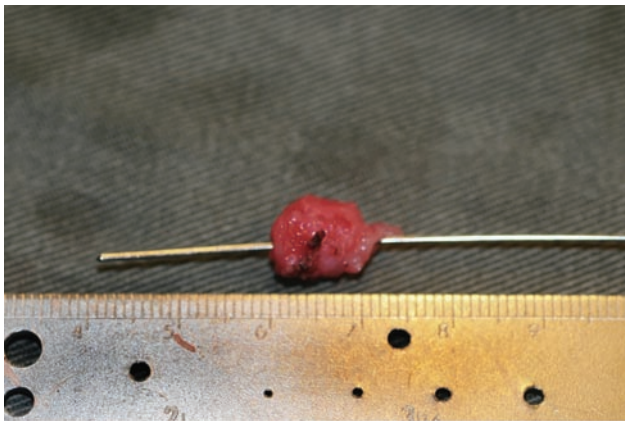
Most of the esophageal stenosis in children is usually acquired due to gastro-esophageal reflux disease, corrosive



**Figure 1** (a) Upper GI contrast study showing abrupt narrowing at distal esophagus which initially misdiagnosed as achalasia. (b) Endoscopic view of intraluminal narrowing.

intake while only 10% are caused by congenital malformation.<sup>1</sup> Diagnosis of CES had been delayed in most patients as the disease is so rare in clinical practice especially in those without associated esophageal atresia.<sup>3</sup> The typical symptoms include vomiting and dysphagia when semisolid food is introduced.<sup>4</sup> However, these functional symptoms do overlap with other more common diseases such as gastro-esophageal reflux disease, achalasia; as a result diagnosis is difficult to be made on clinical ground only. The first diagnostic tool is the upper gastrointestinal contrast study to look for the abnormal configuration and motility of esophagus. If the study result is suspicious, endoscopy should be performed to confirm the diagnosis and to further estimate the level as well as the length of stenosis with the help of fluoroscopy. In order to differentiate the 3 subtypes of CES, some centers will apply endoscopic ultrasonography for better surgical planning.<sup>5</sup>

Up till now, there is no consensus on the management for CES. Both dilatation and surgical resection are commonly used strategies.<sup>1,4,6,7</sup> The effect of dilatation are variable and may be related to the underlying histology.<sup>8</sup> Although data are limited, some authors suggested that dilatation is less effective in TBR form and also with higher perforation rate.<sup>4,6,8</sup> Either laparotomy or thoracotomy were conventionally used for esophageal resection depending on the stenosed level. More experiences in minimal invasive surgical technique have been reported for the treatment of CES.<sup>9</sup> As illustrated in our two cases, laparoscopic esophageal resection with primary anastomosis is a safe and effective method if the stenosed segment is close to the esophagogastric junction; however a longer follow-up period is necessary.



**Figure 2** Operative specimen.

Apart from the method of management, the timing of surgery is another critical issue. As these patients are mostly malnourished, pre-operative nutrition buildup is important. On the other hand, delayed surgery may increase the risk of aspiration and decompensate lung condition. To balance the risk and benefit, we have introduced the idea of regular suction through a separate naso-esophageal tube in addition to a nasogastric tube for feeding. After the insertion of naso-esophageal tube, vomiting and reactive cough has greatly reduced for our patient.<sup>2</sup>

In conclusion, CES is an uncommon yet important diagnosis that may normally be overlooked in daily practice. High clinical suspicion is critical in managing a child with persistent vomiting or dysphagia once solid food started. Early referral is essential as surgical management is effective and with good prognosis.

### Conflict of Interest

No conflict of interest to declare.

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