

# A rare case of H-type tracheoesophageal fistula associated with congenital oesophageal stenosis

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## Summary

H-type tracheoesophageal fistula (TOF) occurring in conjunction with congenital oesophageal stenosis (CES) is an infrequently reported condition. We present such a case in an infant with respiratory difficulty since birth. This report displays the challenges in diagnosis and the subsequent surgical management of this complex anomaly. A staged surgical approach, prioritising surgical ligation of the TOF followed by balloon dilatation of the CES proved to be effective with a favourable outcome.

**Keywords:** H-type tracheoesophageal fistula, congenital oesophageal stenosis, oesophageal atresia, VACTERL, paediatric surgery case reports

## Case presentation

A female infant was born at 40 weeks gestational age via normal vaginal delivery with a birth weight of 2860 g. The patient was discharged home with the mother after a brief observation period. On day 1 of life, she presented to the hospital with respiratory distress. A rapid deterioration led to intubation and ventilation in the neonatal intensive care unit (NICU). The chest X-ray was consistent with a multi-lobar pneumonia, and the orogastric tube tip was noted in the stomach.

After an initial recovery, the infant continued to have difficulty with feeds, with severe respiratory distress and a spluttering cough while drinking. On day 20 of life, a computed tomography (CT) chest was performed which indicated an ongoing multi-lobar pneumonic process.

There was a delay in obtaining a contrast oesophagogram due to the patient being dependant on high flow oxygen support. Once performed at 2 months of age, it indicated an oesophageal narrowing at the level of T6/T7 and contrast bronchograms without evidence of proximal aspiration. A formal prone oesophagogram was not done due to the patient experiencing severe oxygen desaturation when positioned prone. Subsequent bronchoscopy confirmed the presence of a proximal TOF just distal to subglottic area with subtle tracheomalacia.

A right-sided neck exploration and fistula ligation was performed 2 weeks later. Flexible oesophagoscopy was done on-table prior to neck exploration revealing a distal CES without features of reflux oesophagitis. Balloon dilatation of the oesophageal stenosis was delayed until 3 weeks after fistula ligation. Bedside laryngoscopy after fistula ligation revealed a right-sided vocal cord palsy which was managed

conservatively and improved symptomatically. Repeat contrast swallow confirmed successful ligation of the fistula with improved patency of distal oesophageal stenosis.

Oral feeds were slowly introduced under the guidance of the speech therapist in hospital. On discharge the child was tolerating bolus milk feeds. She underwent one further oesophageal dilatation with rigid dilators 8 weeks after the first dilatation. At follow-up, the parents had introduced baby cereal followed by soft purees and the child was gaining weight without any further respiratory morbidity.

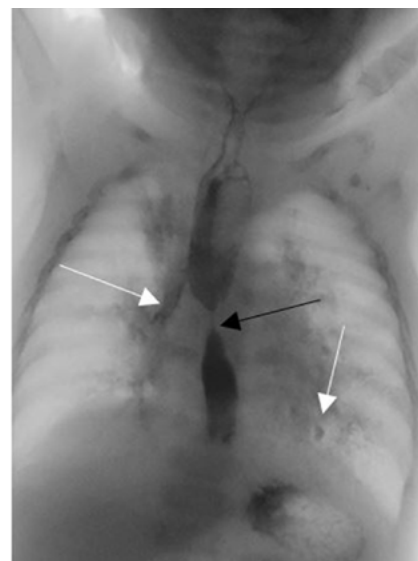


Figure 1: Contrast oesophagogram showing oesophageal stenosis (black arrow) and contrast bronchograms (white arrows)

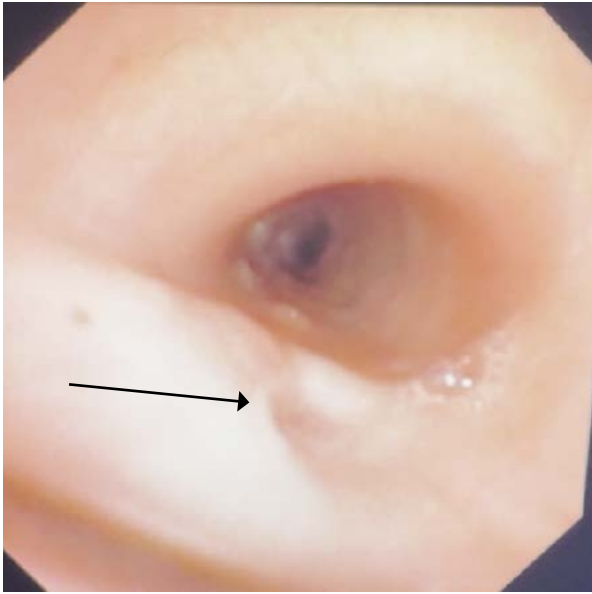


Figure 2: Bronchoscopy showing opening of H-type tracheoesophageal fistula (black arrow)

## Discussion

Gross' succinct classification of oesophageal atresia (OA) with or without TOF into 5 types collates the majority of congenital oesophageal pathology seen in practice. However, the exhaustive "Atlas of Esophageal Atresia" by Kluth, documenting over 90 variations, reflects the true spectrum of this anomaly.<sup>1</sup> H-type fistula (Kluth VII, Gross Type E) makes up about 4% of all OA cases.<sup>2</sup> Congenital oesophageal stenosis or CES (Kluth VIII) accounts for approximately 1% of OA cases and has traditionally been classified into three distinct histopathological types: tracheobronchial remnants (TBR), fibromuscular thickening or membranous diaphragm.<sup>2</sup> CES can infrequently occur in combination with a proximal OA, typically Gross C, and may be missed at initial diagnosis.<sup>3</sup> However, the combination of H-type TOF associated with CES (Kluth Type VII<sub>4</sub>) is exceptionally rare with only 16 published cases to date.<sup>1,2,4,5</sup> This combination of pathologies introduces several complexities in diagnosis and management.

The diagnosis of H-type fistula is often delayed due to non-specific symptoms and subtle radiological signs. It is predominantly diagnosed when a high index of suspicion leads to a tube-injected prone contrast oesophagogram, delineating a communication between oesophagus and trachea. This study has a sensitivity range between 50–73%.<sup>6</sup> It may also be detected or confirmed on flexible or rigid bronchoscopy.<sup>2</sup> CES is usually diagnosed on contrast swallow. The level of stenosis frequently occurs at the distal oesophagus near the oesophageal gastric junction (OGJ) but can occur more proximally.<sup>3</sup> It may also be diagnosed by oesophagoscopy or, uncommonly, with cross-sectional imaging with CT or Magnetic Resonance Imaging (MRI) with oral contrast.<sup>3,7</sup>

Differentiating between the three histopathological types of CES is often not possible without resection. Endoscopic ultrasound has shown promise in detecting TBR; however, its use is limited by availability.<sup>3</sup> Radiological appearance is not helpful in determining the type of CES, unless TBR are seen on CT scan which is rare.<sup>3,7</sup> The combination of CES with H-type fistula in previous case reports has been diagnosed

either on contrast oesophagogram or as an incidental finding during surgery.<sup>2,4,5</sup> In this case, we suspected the diagnosis on contrast oesophagogram.

The initial study showed an obvious distal stenosis and the presence of contrast bronchograms raised the suspicion of a proximal fistula. Prone contrast study was abandoned due to aspiration and acute decompensation during the study, highlighting some of the difficulties with obtaining a quality study in these patients.

Due to the uncertainty of the diagnosis, we performed flexible bronchoscopy to visually confirm the H-type TOF with corrective surgery planned two weeks later, to allow for full recovery from aspiration pneumonia and an improved nutritional state. Previous case reports have suggested doing both bronchoscopy and oesophagoscopy at the time of surgical correction to avoid multiple anaesthetics, allow for cannulation of the fistula tract to assist with intraoperative visualisation, and to document any preoperative vocal cord palsy.<sup>2,5,8-10</sup>

Preoperative identification of associated anomalies remains important, with special attention to cardiac defects that may increase the risk of anaesthesia. Both H-type fistula and CES in isolation are associated with VACTERL anomalies and therefore it can be extrapolated that the two in combination should prompt similar screening.<sup>7,8</sup>

CES as an isolated entity, can be managed endoscopically with balloon or bougienage dilatations or with surgical resection and anastomosis via thoracotomy, laparotomy or minimally invasive technique.<sup>2,4,7</sup> Traditional understanding is that stenosis due to TBR is not amenable to dilatation, however, due to limitations in determining the type of stenosis together with reports of successful management of CES with dilatations alone, this is being challenged.<sup>3,7</sup> Neither balloon nor bougienage dilatation technique has been shown to be superior, however most authors have described balloon dilatation as their preferred initial approach.<sup>3,7</sup> H-type fistula is usually managed with surgical ligation.

More than 90% of fistulas occur above the level of T2 and are amenable to a cervical approach through a right- or left sided neck incision.<sup>8,10</sup> Fistulas below this level are best approached through thoracotomy or thoracoscopy, while endoscopic approaches have proven to have a high recurrence rate and are best avoided.<sup>6,8,10</sup> Preoperative cannulation of the fistula via endoscopy can assist in identifying the tract and may allow the anaesthetist to apply gentle traction, bringing the fistula into the surgical field.<sup>6,9</sup>

We opted for fistula ligation through a right-sided neck incision as the priority intervention, followed by balloon dilatation of the CES. A gastrostomy was not done at the time of fistula ligation, as the patient was tolerating feeds via a nasogastric tube passing adequately through the stenosis.

Identifying the fistula during neck dissection was a challenge and there was iatrogenic injury to the proximal oesophagus that was primarily repaired with a drain left in situ. Retrospectively, this dissection may have been easier if approached from the left, due to the anatomical position of the proximal oesophagus. Additionally, preoperative cannulation of the fistula via bronchoscopy may have aided with identification of the fistula. Fortunately, a prone contrast study done subsequently confirmed successful ligation of the fistula with no leak. The distal stenosis responded appropriately to two subsequent dilatations, at 3 weeks and

11 weeks post fistula ligation, and oral feeds are progressing adequately on follow-up at 4 months post fistula ligation.

The most common complication of H-type fistula ligation is vocal cord palsy and is present postoperatively in 22% of cases. It is likely due to recurrent laryngeal nerve injury.<sup>6,8,10</sup> We opted for conservative management of this complication as most published cases improve without intervention.<sup>10</sup> This patient had a normal swallow on subsequent contrast studies and vocal changes have improved. The reported risk of oesophageal perforation during dilatation for CES is between 3.8% and 44%.<sup>7</sup> TBR are thought to have a higher risk of perforation than the other variants.<sup>3</sup> If dilatation is opted for, balloon dilatation under fluoroscopic guidance rather than bougienage is most likely to be the safest, although neither technique has proven superiority.<sup>7</sup> Oesophageal leak, stricture, fistula recurrence, tracheomalacia and gastrooesophageal reflux are other potential complications.<sup>2,7,8,10</sup>

This case report describes a patient with a rare combination of an H-type TOF associated with a distal CES and highlights the diagnostic difficulties and the complexity of the decision-making required to correct the anomalies surgically. A staged surgical approach with prioritisation of fistula ligation via a neck incision followed by balloon dilatation of the stenosis is effective. Identifying the H-type fistula in these cases may prove difficult during neck dissection and preoperative bronchoscopy with fistula cannulation and potentially a left-sided neck approach for more proximal fistulas may help prevent iatrogenic injury to the oesophagus or recurrent laryngeal nerve.

#### **Conflict of interest**

The authors declare no conflict of interest.

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#### **Ethical approval**

Informed written consent was obtained from all patients being included in the study.

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