CASE REPORT

Oesophageal atresia associated with congenital oesophageal stenosis

Mauricio Antonio Escobar Jr,¹ Michael K Pickens,² Randall M Holland,¹ Michael G Caty³

¹Department of Pediatric Surgical Services, Mary Bridge Children's Hospital & Health Center, Tacoma, Washington, USA

²Department of Pediatric Gastroenterology, Mary Bridge Children's Hospital & Health Center, Tacoma, Washington, USA

³Department of Pediatric Surgery, Yale-New Haven Children's Hospital, New Haven, Connecticut, USA

Correspondence to

Dr Mauricio Antonio Escobar,
mauricio.escobar@multicare.org

SUMMARY

Congenital esophageal stenosis (CES) is associated with oesophageal atresia and tracheoesophageal fistula (EA-TEF). Tracheobronchial (cartilaginous) rests are typically found within the area of stenosis. The authors present a case report of EA-TEF associated with CES and present a modification of a novel technique to facilitate the management of CES. The impact of gastro-oesophageal reflux disease is discussed in this unique entity. Finally, the literature is reviewed for this unique entity.

BACKGROUND

Esophageal atresia (EA) has been described as 'the epitome of modern surgery'.¹ In 1936, Thomas Lanman performed the first primary extrapleural anastomosis of this anomaly, but the child lived only 3 h. Leven² and Ladd³ reported the first staged survivors in 1939, and Cameron Haight⁴ performed the first successful primary anastomosis in 1941.

EA with varying types of tracheoesophageal fistula (TEF) affects approximately 1:2400 to 1:3000 neonates.⁴⁻⁵ The majority of infants with EA have an associated congenital malformation. Rarely, EA is associated with a second oesophageal anomaly, such as congenital esophageal stenosis (CES). Spitz⁶ was the first to demonstrate a congenital basis for distal oesophageal stenosis associated with EA by showing the presence of tracheobronchial rests in an oesophagectomy specimen.

CASE PRESENTATION

A 38-week gestational age infant was noted to have thick secretions and difficulty in breathing on the first day of life. An orogastric tube (OG) was unable to be passed into the stomach, and chest x-ray was concerning for EA. Antibiotics were started.

INVESTIGATIONS

A VACTERL workup including echocardiogram, renal ultrasound, spinal films and a voiding cystourethrogram was normal. VACTERL or VATER association is an acronym used to describe a series of characteristics which have been found to occur together. V stands for vertebrae, which are the bones of the spinal column. A stands for imperforate anus or anal atresia, or an anus that does not open to the outside of the body. C is added to the acronym to denote cardiac anomalies. TE stands for tracheoesophageal fistula, which is a persistent connection between the trachea (the windpipe) and the oesophagus (the feeding tube). R stands for

renal or kidney anomalies. L is often added to stand for limb anomalies (radial agenesis).

TREATMENT

The patient underwent right thoracotomy and the TEF was repaired without difficulty, and attention was turned to oesophagoplasty. During the repair of the oesophagus, an obstruction distal to the level of the anastomosis was encountered during the attempted passage of an OG. The diagnosis of CES was suspected intraoperatively. The patient underwent an oesophagram on the seventh postoperative day. The anastomosis was intact, and a mid-oesophageal stenosis was confirmed (figure 1). Three days later, a Stamm gastrostomy was created. The patient was discharged to home from the neonatal intensive care unit on postoperative day 20.

OUTCOME AND FOLLOW-UP

At 2 months of age, the patient developed wheezing with feeds as well as a rough barking cough. Tracheomalacia was suspected. An oesophagram was performed to determine if an anastomotic stricture could be contributing to the respiratory symptoms. The oesophagram revealed a narrowing of the anastomosis. Rigid bronchoscopy and oesophagoscopy were performed. An 85–90% collapse of the trachea with spontaneous breathing confirmed the diagnosis of tracheomalacia. The oesophageal anastomosis was dilated with the 2.5 mm rigid bronchoscope; however, the CES could not be dilated.

Figure 1 Oesophagram demonstrates the esophageal atresia (EA) with distal EA associated with a congenital oesophageal stenosis.

To cite: Escobar MA Jr, Pickens MK, Holland RM, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-009620

Subsequently, the patient had two attempts at balloon dilation of the CES. While radial dilation was certainly an option, we decided to wait based on the currently available literature highlighting the success of balloon dilation with the CES. Additionally, the patient was tolerating feeds, and it was felt to be safe to wait until a balloon dilator of her size was available. By 6 months of age, however, the family wanted to introduce baby food. During oesophagoscopy, a two-French Fogarty (Edwards Lifesciences, Irvine, California, USA) catheter could be passed. Given the tightness of the stenosis, the decision was made to offer an operation.

Following a viral illness, the patient presented for surgery at 9 months of age. Preoperative oesophagram revealed oesophagitis distal to the CES. The patient underwent a segmental oesophagectomy with primary anastomosis via a redo right thoracotomy (figure 1). A Fogarty catheter was placed by oesophagoscopy to guide finding the CES. Pathology revealed cartilaginous rests within the specimen. Oesophagram on the sixth postoperative day revealed a large leak (figure 2). Her course was also complicated by significant reflux mandating maximum medical therapy (H₂ receptor antagonist and proton pump inhibitor). The patient was ultimately managed as an outpatient with nutrition supplied via a gastrojejunostomy tube. Her leak healed by 6 weeks postoperatively and resulted in an anastomotic stricture (figure 3). The patient underwent a Nissen fundoplication to manage her severe reflux 4 months later and subsequently underwent successful dilation of stricture.

The patient did well for almost a year transitioning well to solid foods. At the age of 20 months, the patient once again developed dysphagia. Oesophagram revealed recurrent stricture of the distal anastomosis. The patient has had two successful balloon dilations with almost a complete resolution of the dysphagia.

DISCUSSION

A variety of lethal tracheobronchial abnormalities have been reported in association with EA, including tracheal agenesis, laryngeal atresia, congenital tracheal stenosis and pulmonary agenesis. Other associated tracheobronchial malformations include ectopic bronchi, such as tracheal bronchus and trifurcated trachea, and absent right upper lobe bronchus. The

Figure 2 Postoperative oesophagram demonstrates the anastomotic leak following resection and primary anastomosis of the congenital oesophageal stenosis.

Figure 3 Oesophagram demonstrates resolution of the anastomotic leak complicated by an anastomotic stricture.

observation of a trifurcated trachea in patients with EA supports the current theories of its embryology.

The most commonly accepted theory of the pathogenesis of EA is abnormal embryogenesis of the laryngotracheo-oesophageal groove and septum formation. Abnormal septation allows for persistent communication between the trachea and oesophagus.⁷ Gittes and colleagues⁵ have demonstrated foregut trifurcation in adriamycin-induced EA in Sprague-Dawley rats. Two of the tracheal branches formed lungs and the third formed the TEF and distal 'oesophagus'. It is interesting to note the identification of trifurcated tracheas in infants with EA⁸ and the association of tracheobronchial remnants with EA.⁹

Distal oesophageal stenosis associated with EA is associated with congenital tracheobronchial rests. A de novo CES may be encountered, or one may present more commonly postoperatively after EA repair. Complications arising from tracheobronchial rests are similar to those of postoperative anastomotic stenosis unrelated to tracheobronchial remnants. Gittes' model of EA with TEF formation readily explains the presence of tracheobronchial remnants in the distal oesophageal pouch.⁵ A tracheobronchial rest, an oesophageal membranous diaphragm and segmental hypertrophy of the muscularis and diffuse fibrosis of the submucosa are different aetiologies of CES. Bouginage or balloon dilation is recommended. If symptoms and stricture persist after three dilations, surgical resection of the stenotic segment, with end-to-end anastomosis or oesophageal replacement, is the treatment of choice.^{9–11} Romeo *et al*¹¹ also demonstrated the usefulness of endoscopic ultrasound in diagnosing the aetiology of the CES. Because the tracheobronchial remnants are limited to a short 1–2 cm length, a limited resection rather than a more extensive resection with oesophageal substitution is all that is usually required.¹⁰

If the paediatric surgeon encounters an oesophageal stenosis from tracheobronchial remnants during the initial repair, the authors recommend the following approach. The anastomosis should be completed and a gastrostomy tube should be placed to preserve the native oesophagus. A small nasogastric should be used to stent, or placed proximal to the anastomosis, to ensure adequate drainage of saliva in the immediate postoperative period. The patient should then be allowed to grow, and after the anastomosis has healed, the stenosis may be evaluated with an oesophageal swallow or CT scan. There is no literature

addressing the role of an MRI at this time; however, such a modality would likely require sedation or general anaesthesia in an infant or child. Dilation should be attempted as described above.

The patient should then undergo laparotomy if the CES is distal, or redo thoracotomy if it is proximal, if obstruction is present or persists. Caty *et al*¹² describe a technique for repairing the CES via laparotomy. A Fogarty or Foley (BARD, Covington, Georgia, USA) catheter may be placed through a gastrostomy, and passed proximally above the stenosis and the balloon inflated. The surgeon may then gently retract the oesophagus into the abdomen, and the segment of oesophagus with the tracheobronchial remnants may be resected. The specimen is sent for frozen section and the margins should be clear of any tracheobronchial rests. A primary anastomosis should then be completed in the abdomen. The authors used a modification of this technique by passing a Fogarty catheter antegrade from the oropharynx past the CES and having the anaesthesiologist gently retract. This allowed for ease of discovery of the stenotic area in the reoperative field. A postoperative oesophagram should be performed 5–7 days postoperatively to check for an oesophageal leak. Non-operative management of oesophageal leak was successful in this case and based on outcomes of conservative treatment for anastomotic leakage after surgical repair of EA.¹³ Anastomotic stricture may develop following a major leak, and our patient underwent successful dilation of her stricture. Importantly, late complication of stricture was also successfully managed by dilation.

Gastro-oesophageal reflux occurs in 30–82% of patients after EA and TEF repair; 30% of these patients require an antireflux operation.¹⁴ The bathing of the anastomotic line in stomach acid results in an increased incidence of stricture and leak. Tension on the anastomosis causing shortened intra-abdominal oesophageal length and blunting of the angle of His,¹⁴ intrinsic oesophageal dysmotility, intrinsic innervation disorders,¹⁵ denervation from extensive oesophageal pouch dissection and gastric dysmotility¹⁶ all contribute to the pathogenesis of gastro-oesophageal reflux disease (GERD) in EA.

Long-term consequences of GERD in these patients are manifold. It is important to note that the presence of pathological changes of the oesophagus, such as oesophagitis and gastric metaplasia, do not always correlate with symptoms. Symptoms tend to decrease over time.¹⁴ Consequences of GERD in patients with a history of EA last well into adulthood,¹⁷ including respiratory symptoms (33%), dysphagia (52%), reflux oesophagitis (58%), Barrett's oesophagus (11%) and strictures (42%). In patients with a history of EA, men older than 35 years of age and individuals with episodes of reflux greater than three times per week have an increased risk of severe oesophagitis and Barrett's oesophagus.¹⁷ There have been two reported cases of oesophageal squamous cell carcinoma in patients with EA, including one at the anastomosis.¹⁷ Long-term follow-up is mandatory to screen for these complications of GERD in EA.

Nissen fundoplication has traditionally been considered the best option,¹⁸ but concerns for debilitating dysphagia and incidence of wrap disruption and recurrent GERD (33%)¹⁸ have challenged this notion. Some paediatric surgeons have begun to use partial wraps such as the Thal or Toupet procedure to ameliorate these problems. No clear improved outcome has been demonstrated using any of these procedures.¹⁸ Some surgeons modify their Nissen technique to create a shorter, floppier wrap (1.0–1.5 cm over a larger dilator).¹⁹ In addition, some surgeons have used the Collis-Nissen fundoplication to gain

intra-abdominal oesophageal length.²⁰ In this case, we proceeded with repair of the CES before performing a fundoplication. The patient had the above noted complications, and in future cases consideration will be given to performing the fundoplication before repair of the CES.

In conclusion, the authors presented a rare case of EA-TEF associated with the CES. Management techniques were offered for when the paediatric surgeon encounters the CES at the original operation, and a novel approach was described to facilitate operative repair. Finally, complications arising from the operation and the management of these complications were discussed with emphasis on the sequelae of GERD in this unique patient population.

Learning points

- ▶ Rarely, esophageal atresia (EA) is associated with a second oesophageal anomaly, such as congenital esophageal stenosis (CES).
- ▶ A tracheobronchial rest, an oesophageal membranous diaphragm and segmental hypertrophy of the muscularis and diffuse fibrosis of the submucosa are different aetiologies of CES.
- ▶ Bouginage or balloon dilation is recommended. If after three dilations symptoms and stricture persist, surgical resection of the stenotic segment, with end-to-end anastomosis or oesophageal replacement, is the treatment of choice.
- ▶ Because the tracheobronchial remnants are limited to a short 1–2 cm length, a limited resection rather than a more extensive resection with oesophageal substitution is all that is usually required.
- ▶ Gastro-oesophageal reflux occurs in 30–82% of patients after EA and tracheo-oesophageal fistula repair; 30% of these patients require an antireflux operation. The bathing of the anastomotic line in stomach acid results in an increased incidence of stricture and leak.

Contributors All authors contributed to the: conception and design, acquisition of data or analysis and interpretation of data; drafting the article or revising it critically for important intellectual content; final approval of the version published.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Myers NA. Oesophageal atresia: the epitome of modern surgery. *Ann R Coll Surg Engl* 1974;54:277–87.
- 2 Leven N. Congenital atresia of the esophagus with tracheoesophageal fistula. *J Thorac Cardiovasc Surg* 1941;10:648–57.
- 3 Ladd W. The surgical treatment of esophageal atresia and tracheoesophageal fistulas. *N Engl J Med* 1944;230:625–37.
- 4 Haight C, Towsley H. Congenital atresia of the esophagus with tracheoesophageal fistula. Extrapleural ligation of fistula and end-to-end anastomosis of esophageal segments. *Surg Gynecol Obstet* 1943;76:672–88.
- 5 Crisera CA, Connelly PR, Marmureanu AR, *et al*. Esophageal atresia with tracheoesophageal fistula: suggested mechanism in faulty organogenesis. *J Pediatr Surg* 1999;34:204–8.
- 6 Spitz L. Esophageal atresia: past, present, and future. *J Pediatr Surg* 1996;31:19–25.
- 7 Rosenthal A. Congenital atresia of the esophagus with tracheoesophageal fistula: report of eight cases. *Arch Pathol* 1931;12:756.
- 8 Usui N, Kamata S, Ishikawa S, *et al*. Anomalies of the tracheobronchial tree in patients with esophageal atresia. *J Pediatr Surg* 1996;31:258–62.
- 9 Yeung CK, Spitz L, Brereton RJ, *et al*. Congenital esophageal stenosis due to tracheobronchial remnants: a rare but important association with esophageal atresia. *J Pediatr Surg* 1992;27:852–5.

- 10 Neilson IR, Croitoru DP, Guttman FM, *et al.* Distal congenital esophageal stenosis associated with esophageal atresia. *J Pediatr Surg* 1991;26:478–81; discussion 81–2.
- 11 Romeo E, Foschia F, de Angelis P, *et al.* Endoscopic management of congenital esophageal stenosis. *J Pediatr Surg* 2011;46:838–41.
- 12 Caty MG, Lau ST, Escobar MA. Complications of esophageal surgery In: Caty MG, Glick PL, Levitt MA, Haynes JH.eds *Complications of pediatric surgery*. New York, NY: Informa Healthcare, 2009:221–56.
- 13 Zhao R, Li K, Shen C, *et al.* The outcome of conservative treatment for anastomotic leakage after surgical repair of esophageal atresia. *J Pediatr Surg* 2011;46:2274–8.
- 14 Guo W, Fonkalsrud EW, Swaniker F, *et al.* Relationship of esophageal anastomotic tension to the development of gastroesophageal reflux. *J Pediatr Surg* 1997;32:1337–40.
- 15 Pederiva F, Burgos E, Francica I, *et al.* Intrinsic esophageal innervation in esophageal atresia without fistula. *Pediatr Surg Int* 2008;24:95–100.
- 16 Kawahara H, Kubota A, Hasegawa T, *et al.* Lack of distal esophageal contractions is a key determinant of gastroesophageal reflux disease after repair of esophageal atresia. *J Pediatr Surg* 2007;42:2017–21.
- 17 Lindahl H, Rintala R, Sariola H. Chronic esophagitis and gastric metaplasia are frequent late complications of esophageal atresia. *J Pediatr Surg* 1993;28:1178–80.
- 18 Taylor AC, Breen KJ, Auldist A, *et al.* Gastroesophageal reflux and related pathology in adults who were born with esophageal atresia: a long-term follow-up study. *Clin Gastroenterol Hepatol* 2007;5:702–6.
- 19 Bergmeijer JH, Tibboel D, Hazebroek FW. Nissen fundoplication in the management of gastroesophageal reflux occurring after repair of esophageal atresia. *J Pediatr Surg* 2000;35:573–6.
- 20 Kawahara H, Imura K, Yagi M, *et al.* Collis-Nissen procedure in patients with esophageal atresia: long-term evaluation. *World J Surg* 2002;26:1222–7.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow