Dysphagia in a 30-year-old woman: too old for a congenital abnormality?

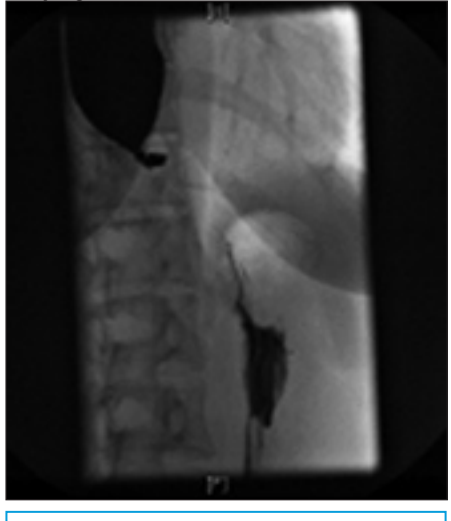
Introduction

Congenital abnormalities are usually thought to be the domain of paediatricians. However, they can present in adult life. This case highlights this in a 30-year-old woman with swallowing difficulties.

Discussion

Oesophageal atresia occurs in approximately 1 in 4425 live births. The classical picture is where the oesophagus forms a blind passage through which no nutrition can be delivered. Less commonly a tracheoesophageal fistula can be found, where there is an abnormal opening between the trachea and oesophagus. Both of these can occur together. They are normally diagnosed soon after birth, in babies producing excess salivation with coughing or choking.

Figure 1. Barium swallow demonstrating the oesophageal stricture.



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More rarely oesophageal stenosis is found, seen in 1 in every 25 000 live births. This is also known as oesophageal stricture, and may go undiagnosed until adulthood. The stenotic segment is anything from 2 to 20 cm in length and found in the middle or lower oesophagus. Dilatation proximal to the stenosis is common.

Figure 2. Barium swallow. Note the dilatation of the distal oesophagus.

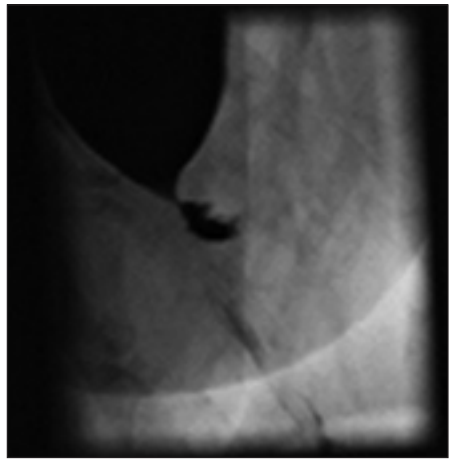
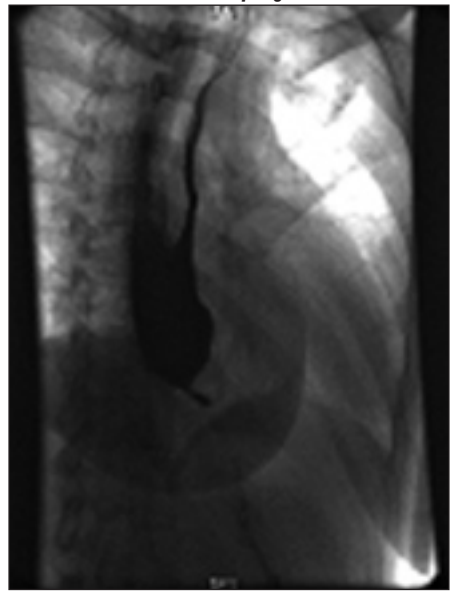


Figure 3. Barium swallow demonstrating both dilatation of the distal oesophagus and stricture.



Case Report

A 30-year-old woman presented to the acute medical take with nausea and vomiting of 48 hours' duration. Clinical examination showed no evidence of bowel obstruction. X-rays showed no abnormalities and blood tests were unremarkable. A gastroenterology opinion was sought. A closer probing of the history revealed that the vomiting consisted of regurgitation of completely undigested food contents shortly after eating. As an infant she had been diagnosed with a 'narrow' oesophagus, but had been told by paediatricians that she would 'grow out of it'. She was of short stature, and on closer questioning revealed that she had always found eating difficult and had a poor appetite. The possibility of partial oesophageal atresia was raised.

A gastroscopy was performed which revealed almost complete obstruction of the oesophagus at 35 cm. On barium swallow a thin string-like 4 cm passage into the stomach was evident, enough to allow the passage of fluids (*Figures 1–3*). A computed tomography scan was performed to further delineate the anatomy and to exclude a mass lesion associated with the stricture. Owing to the severity of the stenosis, it was felt that dilatation would only provide temporary relief and could be potentially dangerous, with a high risk of perforation. A surgical review was requested and it was decided that a Merendino jejunal transposition may be required to allow effective swallowing. The patient was consented for this elective procedure, and she was fed on a liquid diet of build-up products for 3 weeks. Surgery went ahead as planned. However, during the operation it was decided that the changes were not as severe as previously felt and that a simple myotomy would provide adequate relief, with the option to proceed to more aggressive surgery at a later date if needed. It was probable that a degree of the narrowing seen on imaging was caused by inflammation from eating solid foods, and that this had improved on the liquid diet. The postoperative period was complicated by an anastomotic leak. However, the patient recovered quickly from this and was discharged home shortly afterwards. In the 3 months post surgery she continued to improve, and she is now able to eat a normal diet without difficulty.

mon. In these individuals the oesophagus is fully intact and connected to the stomach (Feldman et al, 2006).

When resected many stenotic walls contain tracheobronchial remnants, which are sequestered respiratory tissue, suggesting the origin may be incomplete separation of lung bud from the primitive foregut (Zhao et al, 2004). Fibromuscular hypertrophy can be found with associated damage to the myenteric plexus. Sometimes the muscle layer is not involved at all and only a membranous diaphragm is found (Minkes, 2008). Although often symptomatic in infancy, these tend to present in childhood when more solid food is ingested. With time the stenosis gradually narrows, leading to a deterioration in symptoms and a sensation of food 'catching' just above the diaphragm.

Management can be challenging. The main options are endoscopic dilatation, oesophageal myotomy, distal oesophagectomy or Merendino jejunal transposition. Owing to the uncommon nature of the condition there is a limited evidence base for all of these approaches, especially in cases detected later in life. Oesophageal stricturing involving fibromuscular stenosis and membranous webs can respond to dilatation. Those caused by tracheobronchial remnants rarely improve with bougienage, and require resection. The risks of dilatation are that the connection between the stomach and oesophagus can be fibrous tissue only. Stretching this carries a high risk of mucosal tears and perforation (Takamizawa et al, 2002). Furthermore, the benefits are likely to last no more than 6–12 months, after which time surgery may be more difficult. Therefore endoscopic ultrasound is important in assessing the strictures.

A surgical myotomy is considered the treatment of choice in infancy, providing good symptomatic results. The evidence of benefit in later life is less clear, however, and this is not suitable if the stricture is too long or severe. In these cases a more radical approach is required. Unfortunately a distal oesophagectomy does not always produce satisfactory results. Furthermore, it is prone to failing 15–20 years down the line – in a patient of 30 years of age this is clearly not ideal. Currently jejunal trans-substitutions are rare. The replacement is unique because it retains peristaltic activity, so it must be positioned isoperistaltically. Peristalsis is not synchronous with swallowing and may slow food transit. It tends to result in a low incidence of leaks and is less likely to fail at a later stage. However, it is a technically difficult procedure as it requires preservation of a vascular pedicle, so there is a risk of infarction and graft failure.

Conclusions

In this patient a long-term solution was necessary given her very young age, with a failure in middle life being undesirable, so a Merendino transposition was felt to be the best option. It was only during the procedure that it became clear that a less invasive approach was likely to provide an acceptable result. It is not uncommon that inflammation can subside when a patient is put on to a liquid diet, as occurred in this case. A surgical transposition can still be performed at a later stage if required, and she will be under long-term follow up to assess the need for any further treatment. **BJHM**

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