

Congenital broncho-oesophageal fistula in the adult

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ABSTRACT The case of a 46-year-old man with congenital broncho-oesophageal fistula is presented. The patient had had recurrent chest infections since childhood and a recent history of heartburn and flatulence. A barium-swallow examination showed a small sliding hiatal hernia and an oesophageal diverticulum communicating via a fistula with the apical and posterior segmental bronchi of the left lower lobe. Bronchography showed bronchiectasis in this lobe. At thoracotomy resection of the broncho-oesophageal fistula and left lower lobectomy were performed. The patient's recovery was uneventful. Twenty published cases are reviewed. The classification of congenital broncho-oesophageal fistulae should take account not only of the type of fistula but also of the type and size of the bronchus with which the fistula communicates, the age of the patient at the first appearance of symptoms, and the duration of symptoms.

The common origin of the respiratory tract and the oesophagus from the same embryonic structures sometimes gives rise to anomalous communications such as tracheo-oesophageal fistula and broncho-oesophageal fistula. Most patients with tracheo-oesophageal fistula associated with oesophageal atresia present in infancy.¹ In patients who have congenital fistula without oesophageal atresia symptoms are usually insidious in onset and the patients may present in adult life.¹⁻⁴ The rarity of congenital broncho-oesophageal fistula in adults has been emphasised by various authors and by 1979 there were probably no more than 75 published cases.

The purpose of this communication is to report the clinical details of a recent case of congenital broncho-oesophageal fistula with associated bronchiectasis and hiatal hernia in an adult and to review the present classification of the disease proposed by Braimbridge and Keith.¹

Case report

A 46-year-old sailor, a non-smoker, presented at Llandough Hospital in 1977 with a longstanding history of recurrent chest infections accompanied by cough with expectoration of whitish sputum. Six months before admission he had developed some heartburn and flatulence. This was treated with antacids but his symptoms did not subside completely. He did not give any history of choking or

coughing when he ate. Physical examination showed no abnormal signs. The haemoglobin concentration, white cell count, serum electrolyte and urea concentrations, and results of liver function tests were normal.

A barium-swallow examination showed a small sliding hiatal hernia and an oesophageal diverticulum in the middle third of the oesophagus. The contrast medium spilled into the left lower lobe through a fistulous communication between the oesophageal diverticulum and the apical and posterior basal segmental bronchi (fig 1). Bronchography showed bronchiectasis in the left lower lobe, particularly noticeable in the apical and posterior segments. There was no evidence of sequestration or leakage of the contrast medium into the oesophagus. Bronchoscopy showed an inflamed orifice of the apical segmental bronchus of the left lower lobe. At oesophagoscopy the oesophagus appeared normal and the broncho-oesophageal fistula could not be located. Methylene blue (3 ml) injected into the left-lower-lobe bronchus did not appear in the oesophagus.

At operation (a left thoracotomy) there was a fistulous communication between the mid-portion of the oesophagus and the apical segment of the lower lobe (fig 2). The fistula was about 2 cm long and 0.4 cm in diameter. The fistula was excised and the oesophageal end repaired. Left lower lobectomy was performed. The small hiatal hernia was not repaired. The postoperative period was uneventful and at the time of discharge the patient was symptom free. He was followed up for 10 months before

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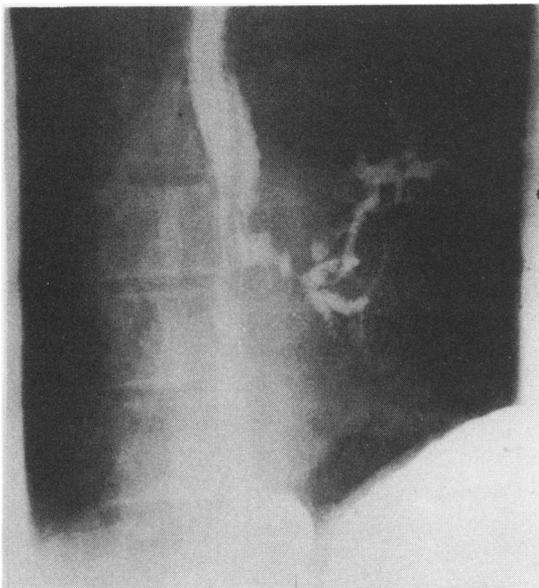


Fig 1 Preoperative barium-swallow radiograph showing a communication between the oesophagus and the apical segment of the left lower lobe.

he returned to his duties in the Merchant Navy.

Careful examination of the lobe after fixation showed that the oesophageal end of the fistula entered the lobe on the mediastinal surface of the posterior basal segment near its upper limit. The fistula was tubular, with external and internal diameters of 0.4 and 0.1 cm, the thick wall being

muscular and only 1 cm long, presumably owing to retraction. Inside the lobe the fistula branched just below the pleura into two intrapulmonary parts. The longer branch passed superiorly and subpleurally for 4.2 cm to end by communicating with a medial subsegmental branch of the apical bronchus. The shorter branch extended inferiorly and subpleurally for 0.8 cm, ending blindly in the posterior basal segment.

Microscopic examination showed that the oesophageal segment of the fistula was lined by stratified squamous epithelium, supported by loose connective tissue. Its outer wall was made up of bundles of smooth muscle fibres, arranged in an irregular but predominantly longitudinal manner. The intrapulmonary or bronchial segment was virtually identical to a bronchus, but with numerous mucous lining cells, no glands, and scanty muscle. Surrounding the fistula there was an area of organising and organised pneumonia with bronchiectasis.

Discussion

In over 75 documented cases of congenital broncho-oesophageal fistula in adults, the patients have mostly presented with symptoms of chronic respiratory infection; cough is usually predominant. Sixty-five per cent of patients have had cough on swallowing liquids (Ono's sign) or food in the sputum, though some have been only mildly affected.¹

The term "congenital broncho-oesophageal fistula in the adult," as it appears in published



Fig 2 Operative photograph showing the fistulous communication between the oesophagus and the apical segment of the left lower lobe. ASLL — apical segment of the left lower lobe; F — broncho-oesophageal fistula; Oes — oesophagus.

Duration of symptoms, age at onset of symptoms, and type of bronchial communication in 21 cases of congenital broncho-oesophageal fistula

Case No	Authors	Year of publication	Age (y)	Age at onset of symptoms (y)	Duration of symptoms (y)	Bronchial communication
1	Braimbridge and Keith ¹	1965	42	8	34	RLL (pinpoint opening)
2	Braimbridge and Keith ¹	1965	19	1	18	Apical segment RLL
3	Braimbridge and Keith ¹	1965	34	2	32	Lateral basal segment LLL
4	Ashley and Evans ¹⁷	1966	45	32	13	Sequestered lung
5	Sacks <i>et al</i> ¹¹	1967	39	30	9	Subsegmental bronchus
6	Harris <i>et al</i> ¹²	1967	63	38	24	Bronchus intermedius
7	Le Roux and Williams ¹³	1968	54	52	2	Bronchus intermedius
8	Le Roux and Williams ¹³	1968	23	Childhood	Many years	Bronchus intermedius
9	Le Roux and Williams ¹³	1968	44	42	2	RLLB
10	Mascarella and Wylie ⁵	1968	18	18	0	Sequestered lung
11	Kinley and Lang ⁴	1969	59	20	39	Posterior basal segment RLL
12	Smith ¹⁵	1970	31	25	6	Postbasal segment RLL
13	Paulin <i>et al</i> ¹⁰	1970	66	65	1	Superior segment RLL
14	Hill ¹⁸	1972	29	13	16	Postbasal segment RLL
15	Vaages ⁴	1973	56	16	40	Postbasal segment RLL
16	Bekoe <i>et al</i> ²	1974	60	?	Many years	RLLB
17	Darakaju <i>et al</i> ¹⁶	1974	20	15	5	Apical segment RUL
18	Darakaju <i>et al</i> ¹⁶	1974	11	Childhood	Many years	Postbasal segment RUL
19	Darakaju <i>et al</i> ¹⁶	1974	10	5	5	Postbasal segment RUL
20	Becker <i>et al</i> ³	1976	62	61	5/12	LMB
21	Osinowo <i>et al</i>	Present report	46	Childhood	Many years	Subsegmental bronchus of apical segment LLL

RLL — right lower lobe; RUL — right upper lobe; RLLB — right lower lobe bronchus; LLL — left lower lobe; LMB — left main bronchus.

reports, gives the impression that there is a very long delay in the onset of symptoms. The observation by Bekoe *et al*² that perhaps there is not a late onset of symptoms but that initially mild complaints have not been thoroughly investigated until complications have appeared, is closer to the observed facts. In 20 cases from reports published since 1965, as well as the present case, the duration of symptoms and the type of bronchus affected were related to the age of the patient at the first appearance of symptoms

(table). The analysis shows that in patients over 40 years of age at the onset of symptoms the duration of symptoms is usually less than three years. The site of the affected bronchus does not appear to influence the duration of symptoms in this group of patients (fig 3). In most of the patients who have had symptoms for less than three years the communication is with a lobar or main bronchus. The single patient with communication with a main bronchus had the shortest duration of symptoms (five months).

All 12 patients with symptoms for more than three years were below 40 years at the first appearance of symptoms. In this group the fistula communicated with a segmental bronchus in seven, a lobar bronchus in two, and a subsegmental bronchus, sequestered lung, and lobar bronchus via a pinhead in one patient each. An 18-year-old patient with sequestered lung was symptom free and the diagnosis was made during investigation of a right basal shadow on routine radiography.⁵

Various attempts have been made to explain why symptoms first appear in the adult. Jackson and Coates⁶ postulate that there is an occlusive membrane that ruptures. Mullard⁷ and Negus⁸ believe that a mucosal fold acts as a valve and loses its occlusive effectiveness after chronic inflammation. Demong *et al* believe that the obliquity of the fistulous tract permits its closure during swallowing. Paulin *et al* hold the view that the first of these

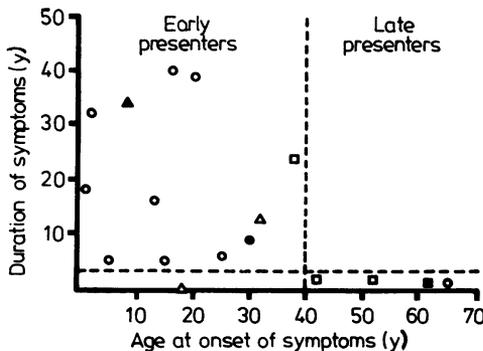


Fig 3 Relation between age at onset of symptoms and duration of symptoms in 21 patients with congenital broncho-oesophageal fistula. ○ Segmental; □ lobar/bronchus intermedius; ▲ lobar (pinpoint opening); ● subsegmental; ■ main bronchus; △ sequestered lung.

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theories is only conjectural, the second is merely plausible, and the third seems to be more in accordance with the facts.¹⁰ A possible theory is that the fistula, which is always invested with a smooth-muscle coat, has the ability to contract, thus narrowing or completely occluding its lumen. This muscle spasm in conjunction with the obliquity of the fistulous tract may be the reason why symptoms do not appear soon after birth.

The diagnosis is usually made by oesophagography when contrast medium spills into the bronchial tree through the fistula.^{1,2} A barium-swallow examination should therefore be carried out in any patient with unexplained chronic cough. Bronchography may sometimes show the fistulous communication with the oesophagus^{3,10} but is more important in assessing the extent of lung damage.¹⁻³ Oesophagoscopy and bronchoscopy are indicated and in some cases the openings of the fistulae have been identified.

There is no controversy about the management of broncho-oesophageal fistulae. The fistula is divided any damaged lung tissue is resected.^{1-5,11-16}

Braimbridge and Keith classified broncho-oesophageal fistulae into four types.¹ Type I is associated with a wide-necked congenital diverticulum of the oesophagus with an inflammatory fistula at the tip. Type II is a simple fistula running directly from the oesophagus to a lobar or segmental bronchus. Type III consists of a fistulous track connecting the oesophagus to a cyst in the lobe, which in turn communicates with the bronchus. In type IV the fistula runs into a sequestered segment; the sequestration connects by one or more tracks with the bronchus.

Since the appearance of this classification most authors have adopted it.^{3,10,16} In delineating types II-IV the type and size of the bronchus with which the fistula communicates are not considered, though this has been shown in our study to be an important determinant of the duration and severity of symptoms. In addition to this classification therefore we must take into account three other factors: (a) *Age of the patient at first appearance of symptoms*—The two groups of patients identified are those not older than 40 years at the first appearance of symptoms (early presenters) and those over 40 at the first appearance of symptoms (late presenters). (b) *Duration of symptoms*—Of the three groups, the first comprises those without symptoms, in whom the fistula is a chance finding, such patients usually having communication with a sequestered lung segment without any bronchial communication; the second group of patients have symptoms for less than three years; and the third group have symptoms for over three years (fig 3). (c) *Type and size of*

bronchial communication—Six types of communications between the fistula and the bronchus are identified: the fistula may communicate with a sub-segmental bronchus, a segmental bronchus, a main bronchus, a lobar bronchus, a pinhole opening into a lobar bronchus, a main bronchus, or sequestered lung tissue.

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