

Congenital broncho-oesophageal fistula

A case report

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Summary

A case of broncho-oesophageal fistula causing bronchiectasis of the left lung is reported. Oesophagorespiratory fistulas without atresia of the oesophagus often have an insidious clinical course and most commonly present in adulthood. This rare congenital anomaly should be considered as a cause of chronic pulmonary sepsis. The clinical, radiographic and therapeutic features of this lesion are discussed.

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Case report

A female infant aged 26 months was referred to the Department of Cardiothoracic Surgery, J. G. Strydom Hospital, Johannesburg, with coughing productive of purulent sputum associated with choking after feeds. She had previously been treated with antibiotics for recurrent chest infections. She was malnourished and anaemic and weighed 6,5 kg with a height of 72 cm. The vital signs were normal and there were no signs of acute distress. The presence of subcutaneous oedema suggested hypoproteinaemia. Diffuse crepitations and rhonchi were audible in the left chest. Haematological examination showed a microcytic anaemia with a haemoglobin concentration of 9 g/dl and a leucocyte count of 12 100/ μ l. The urea and electrolyte levels were normal but the serum albumin level was abnormally low. Sputum examination was negative for *Mycobacterium tuberculosis* and the PPD test result was positive (1+). Chest radiographs showed a diffuse pneumonic change with cavitation of the left lung (Fig. 1). A presumptive diagnosis of bronchiectasis was made and the child was started on antibiotics and physiotherapy.

Rigid bronchoscopy performed under general anaesthesia showed pitting at the base of the left main bronchus above the division of the left upper lobe bronchus. Mucopurulent secretions were present in both major airways. A bronchogram with aqueous dye revealed diffuse saccular bronchiectasis with volume loss of the left lung while the right lung appeared normal (Fig. 2). During the procedure, the oesophagus was found to be outlined, although this was not due to overspill since the dye had been instilled under fluoroscopy (Fig. 2). An oesophagoscopy was then performed and a fistula at mid-oesophageal level was

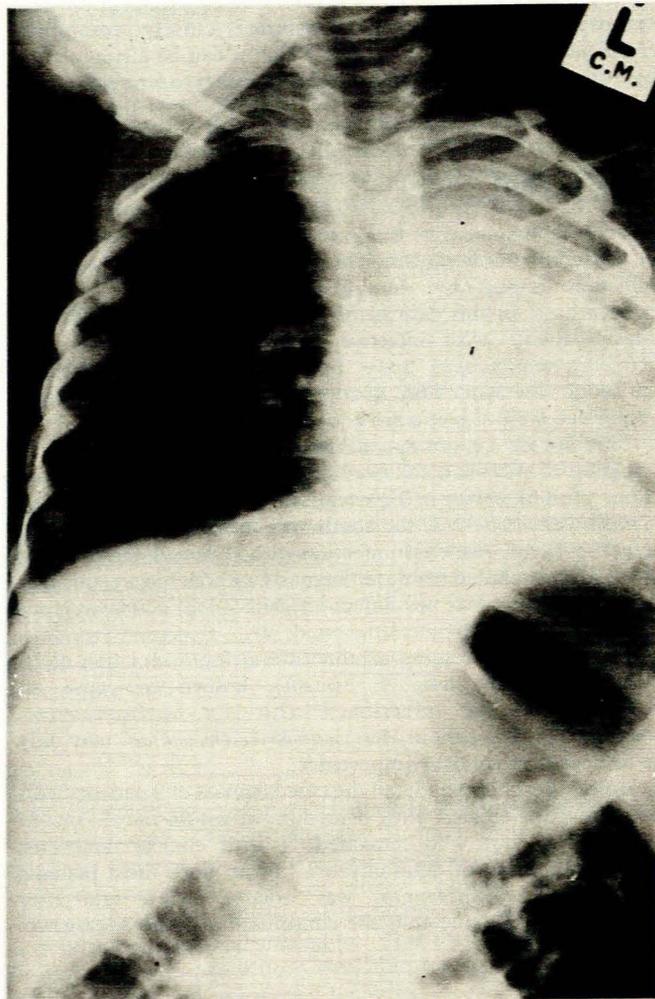


Fig. 1. Chest radiograph showing a diffuse pneumonic change with cavitation of the left lung.

identified quite easily by the leakage of inspired gases into the oesophagus. Barium examination a week later showed a fistulous connection between the mid-oesophagus and the left main bronchus with oesophageal dilatation (Fig. 3).

The lung was mobilized from the chest wall by a right posterolateral thoracotomy and extrapleural dissection. Dissection was carried down to the hilum of the lung and the left main bronchus mobilized. A 5 mm long fistula of 3 mm internal diameter, communicating with the oesophagus, was identified. No inflammatory process was present at this site. A left pneumonectomy was performed and the oesophagus repaired with interrupted 5/0 prolene sutures. The repair was reinforced with a pleural flap. On the 7th postoperative day a Gastrografin swallow showed no leak and good oesophageal function. Fig. 4 is a postoperative chest radiograph. Histological sections of the lung showed destructive saccular bronchiectasis with an acute inflammatory infiltrate. The fistula was lined with squamous epithelium with no evidence of an inflammatory process.

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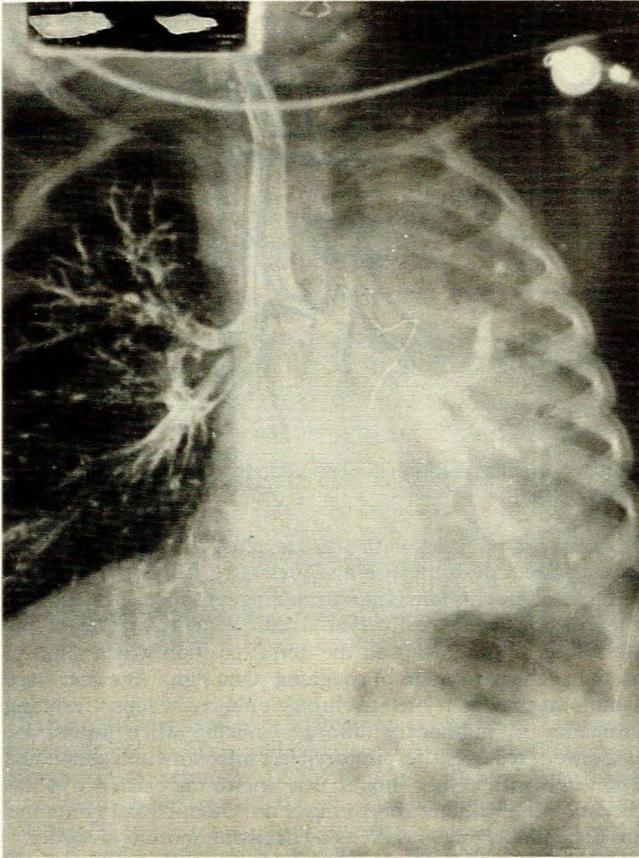


Fig. 2. Bronchogram showing a destroyed left lung and filling of the oesophagus with dye from the endobronchial tree.

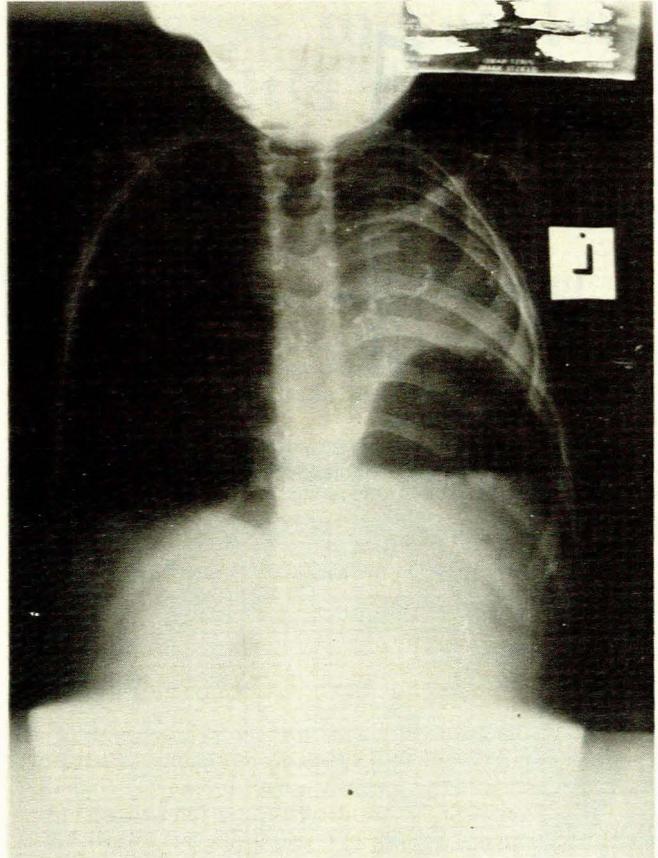


Fig. 4. Chest radiograph taken 5 days postoperatively.

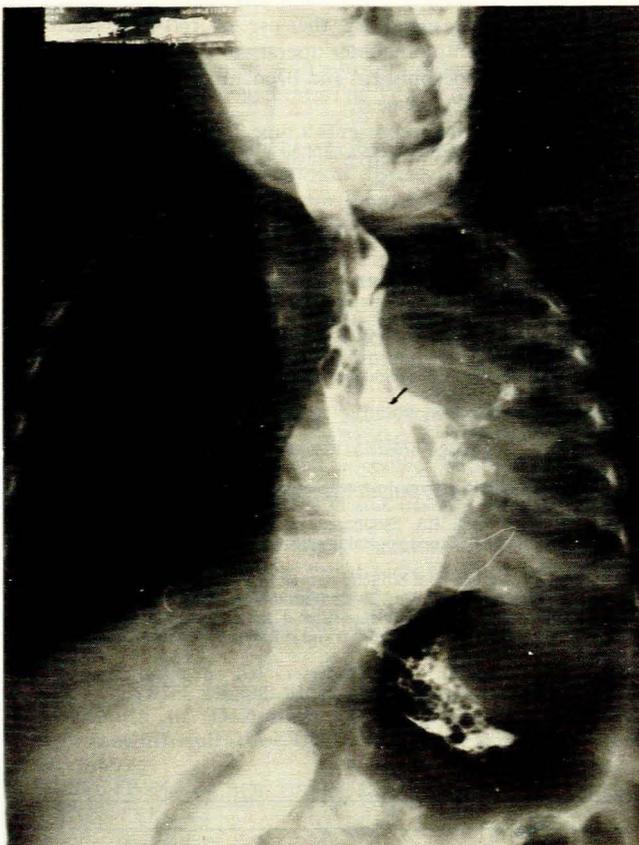
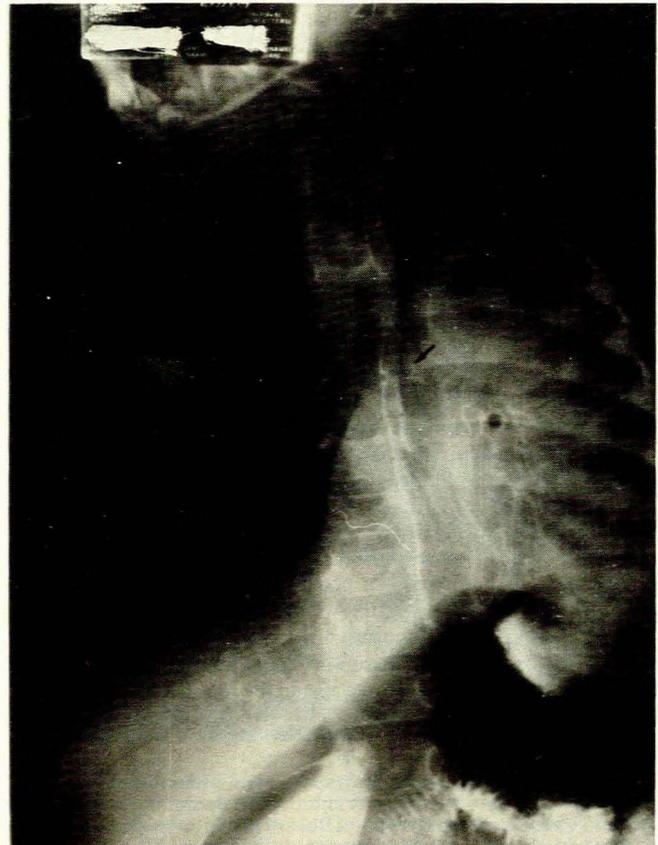


Fig. 3. Barium swallow showing spill-over of dye from the oesophagus into the left main bronchus. The arrows point to the site of the fistula.



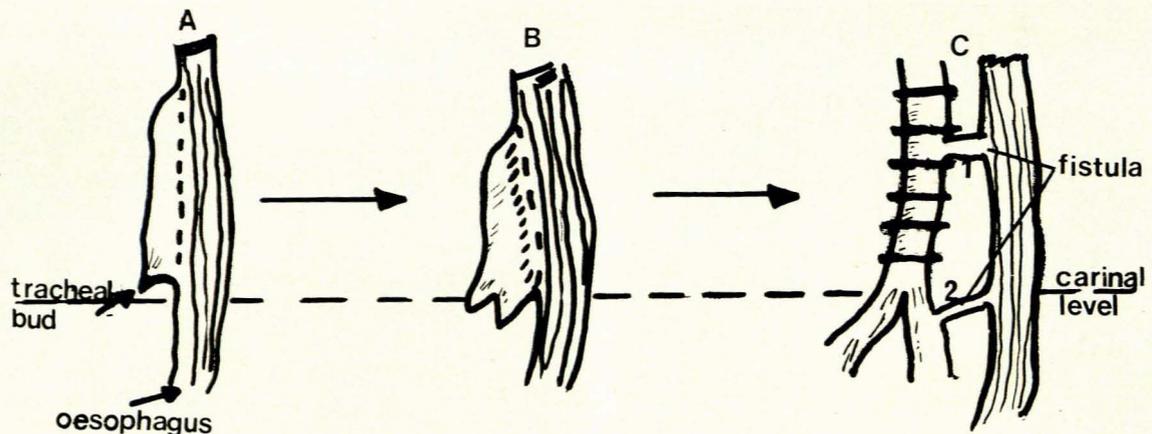


Fig. 5. Embryological development of the trachea from the primitive gut. Failure of separation may result in the formation of a fistula either at point 1 (tracheo-oesophageal fistula) or at point 2 (broncho-oesophageal fistula).

Postoperative dysphagia was relieved by dilatation, and weight gain was rapid. On the 24th postoperative day the patient was discharged and was swallowing well.

Discussion

Broncho-oesophageal fistula is the most infrequent of all abnormal communications between the respiratory and digestive tracts. Patients with this anomaly usually present with it in later life.¹⁻³ In a series of 7 cases described by John *et al.*⁴ only 1 patient presented at the age of 6 years. The late appearance of symptoms may be explained by the presence of a fold of membrane occluding the fistula in the newborn, which breaks down in later life. However, it is more likely that there is interference with the normal mobility of the oesophagus owing to the progressive adherence of the affected lung to the chest wall and mediastinal pleura. The free descent of the oesophagus is thus restricted and the fistula is pulled open, whereas at an earlier stage free movement of the lung, bronchus, fistula and oesophagus closed the fistula by a fold of oesophageal mucosa.⁴

The congenital nature of these fistulas is presumed on the basis of histological verification of a squamous mucosa, muscularis mucosae and no evidence of a neoplastic or inflammatory process.⁵ The embryological formation of these fistulas takes place in the 3rd - 4th week of gestation, when the embryo measures 3 mm. A ventral protrusion of the foregut along its entire thoracic length forms the primitive trachea, which rapidly elongates in a caudal direction to form the main bronchial system. Failure of complete separation at any point will result in an oesophago-pulmonary fistula.^{6,7} The formation of a broncho-oesophageal fistula will depend on the degree of tracheo-oesophageal separation before caudal elongation of the trachea (Fig 5). The more rapid the growth the more distal the communication in the bronchial tree.⁸

Braimbridge *et al.*² described four types of fistula: type 1 is associated with a wide-necked congenital diverticulum of the oesophagus. Inflammation or perforation of this may result in an oesophagobronchial fistula. Type 2 is a short tract communicating directly between the oesophagus and a lobar or segmental bronchus. Type 3 consists of a fistulous tract connecting the oesophagus to a cyst in the lobe of the lung, which

in turn communicates with the bronchus. Finally, type 4 is a fistula communicating with a sequestered lobe.

The classic symptom is of a strangling sensation a few seconds after the ingestion of food or drink. Patients may learn the advantage of swallowing in the dorsal position which tends to minimize the paroxysms of coughing (Ono sign). At a later stage symptoms may be related to chronic pulmonary sepsis resulting ultimately in pulmonary failure.⁵ Choking when liquids are swallowed, recurrent respiratory tract infections and abdominal distension with crying should alert one to the existence of this condition in the infant and young child. Oesophageal symptoms are uncommon unless there is a coexisting motility disorder.

The diagnosis is best confirmed by a typical history and barium swallow findings. Bronchoscopy and oesophagoscopy should be performed together with bronchography to identify the fistulous opening and any damaged pulmonary tissue. The diagnosis may be aided by the use of rod-lens telescopic endoscopes.⁹ Therapy includes operative resection of the fistula with repair of the oesophagus and bronchus and resection of any diseased pulmonary tissue.¹⁰

The case described represents a type 2 fistula and a rare cause of chronic pulmonary sepsis with bronchiectasis in a young child.

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