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Communicating bronchopulmonary foregut malformation associated with esophageal atresia and tracheo-esophageal fistula

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Abstract. We report on two neonates who had esophageal atresia with a distal tracheo-esophageal fistula (type III) and a second fistula between the distal esophagus and an intralobular sequestrated lung segment. In both patients the bronchopulmonary foregut malformation became evident only when the entire esophagus was in continuity following primary repair and a contrast swallow was performed to evaluate the anastomotic site.

Esophageal atresia (EA) with a tracheo-esophageal fistula (TEF) is a well-recognized fetal developmental abnormality. It can occur in association with other abnormalities of the VATER/VACTERL syndrome [1]. However, an added bronchopulmonary foregut malformation (BPFM) with a fistulous communication between the lower esophagus and a sequestrated lung segment is rare. Only 9 cases of a type III EA–TEF associated with a communicating BPFM have been reported [2–8] and we report 2 further cases. The importance of a careful postoperative radiological evaluation of the entire esophagus following repair of EA is stressed as the distal esophagus is not available for examination before or during routine surgery.

Case reports

Case 1

This full-term male baby was delivered by Cesarian section following a pregnancy which was complicated by maternal hypertension and polyhydramnios. No antenatal ultrasound examination had been performed. At delivery the baby was small for gestational age, mildly distressed and drooling. A nasogastric tube could not be passed and radiographs showed a type III EA–TEF, dextroversion of the heart and 13 pairs of ribs. The lungs were clear (Fig. 1). A standard ligation and division of the TEF and primary end-to-end repair of the esophagus was performed via a right posterior extrapleural thoracotomy. On the tenth postoperative day a routine contrast swallow demonstrated an intact anastomotic site and a fistula from the distal esophagus to the posterior right lower lobe (Fig. 2).

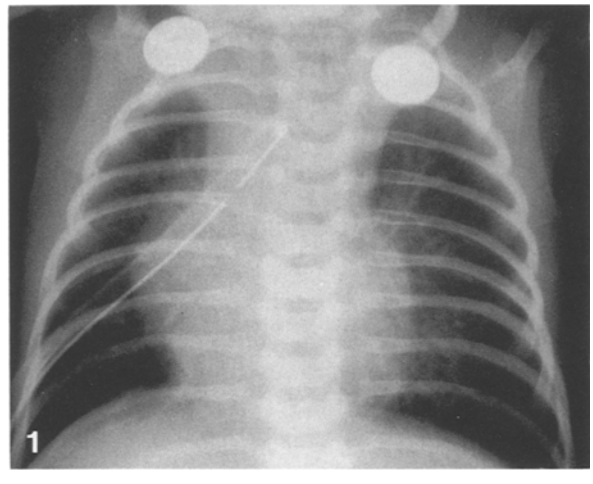
A second thoracotomy identified this tract, which originated 4 cm below the esophageal anastomosis, communicating with a small intralobar sequestion. There was a systemic arterial supply from the adjacent thoracic aorta. Ligation and complete resection were performed.

Histological examination confirmed an intralobar sequestration with aerated alveoli and features of acute bronchopneumonia. Contrast material was present in the small bronchial tree.

Tracheomalacia and gastro-esophageal reflux complicated the baby's further progress and an aortopexy and a surgical antireflux procedure were subsequently performed. The child has done well since then.

Case 2

This full-term male baby presented at birth with drooling and no anal opening. Radiographs showed a type III EA–TEF, high anorectal atresia and a sacral deformity. The lungs were clear. Cardiac evaluation revealed an atrial septal defect. A standard ligation and division of the TEF and an end-to-end anastomosis of the esophagus were performed via a right posterior extrapleural thoracotomy;



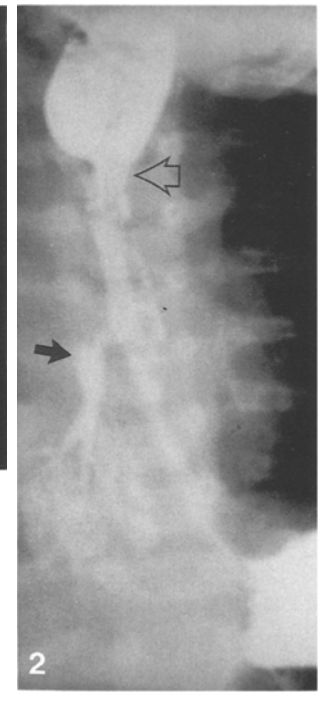


Fig. 1. Case 1. Chest radiograph following primary repair of esophageal atresia. The lungs are clear

Fig. 2. Case 1. Postoperative contrast swallow showing the distal fistula communicating with the right basal sequestration *(filled arrow)* and the intact proximal anastomotic site *(open arrow)*

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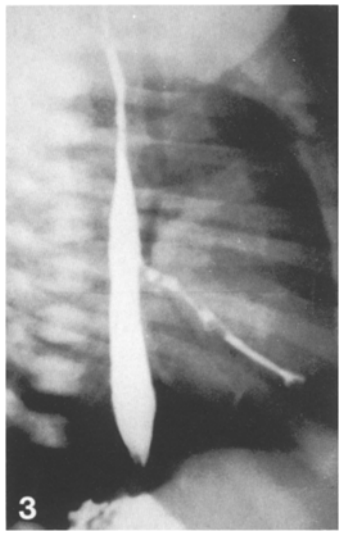

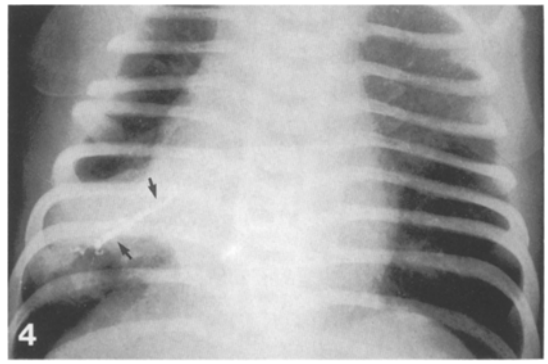

Fig. 3. Case 2. Postoperative contrast swallow showing the distal fistula

Fig. 4. Case 2. Chest radiograph following postoperative contrast swallow showing contrast remaining in the distal fistula *(arrows)* which communicated with the consolidated right mid and lower lung

and a defunctioning sigmoid colostomy was exteriorized. Postoperatively consolidation developed in the right mid and lower lung and on the tenth postoperative day a routine contrast swallow demonstrated an intact anastomotic site and a fistula from the distal esophagus to the opacified right lung (Figs. 3, 4).

A second thoracotomy confirmed this distal fistula which communicated with an intralobar sequestration in the right mid and lower lung, which had an inferior pedicle of vessels with a systemic arterial supply. No fissure existed between the middle and lower lobes. A complete resection was performed.

Histological examination demonstrated a marked inflammatory response, large tortuous lymphatics and arteries, and areas of atelectasis and emphysema. The larger bronchioles showed both mucosal atrophy and hypertrophy with deficiency of cartilage plates.

Further progress was complicated by gastro-esophageal reflux and tracheomalacia which required an antireflux procedure and aortopexy. The anorectal atresia and atrial septal defect will be treated later.

Discussion

The primitive foregut develops a ventral outpouching at about 26 days of embryonic life. A tracheo-esophageal septum then separates the primitive trachea from the primitive esophagus [9]. Insult to the fetus at this critical period affecting the intimate relationship of the trachea and esophagus results in the anomalies of EA–TEF [4, 10]. The ventral outpouching rapidly divides into left and right lung buds which grow into mesodermal tissue having a systemic vascular supply from ventral and dorsal aortae. The lung buds take with them the sixth branchial arch vasculature – the future pulmonary arteries. Pulmonary development will attain differentiated lobar bronchi by 5 weeks [9]. Anomalies of this process lead to BPFM, which includes bronchogenic cysts, pulmonary sequestrations (comprising abnormal parenchyma, abnormal bronchial connections and an abnormal vascular supply) and sequestrations communicating with the foregut [4, 10, 11]. Because the lung buds develop after tracheal separation from the primitive foregut, complex embryological theories have been advanced to explain communicating BPFM. One accepted theory is that supernumerary, accessory lung buds arise from the distal foregut [12]. No single hypothesis has yet adequately and fully explained this large and complex grouping of disorders [4, 12].

Surgical repair of EA–TEF in the newborn is now a routine and highly successful procedure in a modern pediatric surgical unit [13]. The thoracotomy involves a posterior extrapleural approach on the side opposite the aortic arch. Following ligation of the TEF an end-to-end anastomosis of the esophagus is performed. The distal esophagus cannot be visualized during this procedure. Postoperatively, when the entire esophagus is in continuity, swallowed saliva can enter the more distal fistula and pass into the sequestration. This explains the delay in clinical signs and symptoms. EA–TEF and communicating BPFM is so rarely encountered that radiographic lung opacification may be attributed to aspiration, pneumonia, mucus plugs and anastomotic leaks. The first likelihood of diagnosis follows a routine postoperative contrast swallow which is performed primarily to examine the anastomotic site. This will then reveal the more distal fistula from the esophagus to a sequestrated lobe.

We have described 2 further cases of the association of EA–TEF (type III) with a communicating BPFM – an association that has only rarely been reported. The responsibility for this diagnosis rests with the radiologist performing the postoperative contrast swallow in the routine evaluation of the entire esophagus as well as the anastomotic site.

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