Extralobar pulmonary sequestration receiving its arterial supply from the innominate artery

Abstract Pulmonary sequestration is a rare bronchopulmonary foregut malformation, for which the arterial blood supply is usually derived from the descending thoracic or abdominal aorta. A 5-week-old infant is described with an extralobar pulmonary sequestration supplied by a large artery originating from the innominate artery, an arrangement only described once previously. Helical CT scan now makes arteriography unnecessary in the evaluation of these malformations.

Introduction
Pulmonary sequestration is characterized by a segment of nonfunctioning lung parenchyma that does not communicate with the normal tracheobronchial tree and receives its blood supply from anomalous systemic arteries. Two forms exist: intralobar sequestration, which is located within the visceral pleura of the normal lung tissue, and extralobar sequestration, which is invested with its own visceral pleura [1]. Extralobar sequestrations have also been referred to as accessory lobes, accessory lung, pulmonary aberrations, supernumerary lung, Nebenlunge, and Rokitansky lobes [2].

Case report
A 5-week-old male presented with failure to thrive and cough associated with feeding. He was a full-term infant, with a birth weight of 3.78 kg. On admission his temperature was 36.7 °C, respiratory rate was 38/min, pulse was 138/min, blood pressure 115/82 mm Hg, weight 3.43 kg (5th percentile), length 53.5 cm (10th percentile), and head circumference 37 cm (10th percentile). He was visibly malnourished. Cardiac examination revealed a visible precordial heave and a grade II/VI low-pitched systolic apical murmur radiating to the axilla. Neurological examination was significant for extremit hypotonia. All other physical findings were normal. Frontal and lateral radiographs of the chest revealed a large, non-specific mass in the right lower hemithorax. No bowel, air bronchograms, or other distinguishing features were seen. A helical chest
CT scan with off-axis reconstruction showed a large mass in the right lower lung field that was supplied by a very large vessel (approximately the size of the descending aorta) arising from the innominate artery (Fig. 1a,b). The mass was drained by a large vein emptying into the left atrium (Fig. 1a,b), and it contained a large bronchus with an anomalous connection to the distal esophagus (Fig. 2a,b). (Technique: Helical CT with 3-mm collimation and a pitch of 1:1 was performed from the lung apices to the upper abdomen; 10 cc of Omnipaque 300 [2 cc/lb] was injected at a rate of 0.3 cc/s. Data were reconstructed at overlapping 2-mm intervals.) Arteriography confirmed the vascular findings seen on the CT scan. A contrast swallow and upper gastrointestinal contrast study confirmed the esophageal bronchus and also demonstrated malrotation of the intestine. An echocardiogram revealed normal intracardiac anatomy, a mildly dilated left ventricle with normal systolic function, and a mildly increased right ventricular systolic pressure.

The radiologic findings were confirmed at thoracotomy. There was an additional large vein which drained towards the esophageal hiatus and which, in retrospect, was visible on the CT scan. The mass replaced the right lower lobe. The upper and middle lobes were normal. The sequestered pulmonary tissue was lobulated, was invested by its own visceral pleura, and contained a bronchus arising from the lower esophagus. One branch of this bronchus passed through the mass and entered the normal middle lobe, but the ventilation of the entire middle lobe was through its normal bronchus.

The abnormal lung tissue and the anomalous bronchus were resected. The diagnosis of extrapulmonary sequestration was confirmed histologically. The infant’s postoperative course was uneventful.

**Discussion**

Extralobar sequestrations most commonly receive their blood supply from the descending thoracic or abdominal