Bronchopulmonary foregut malformation is a group of rare congenital anomalies affecting the respiratory and upper gastrointestinal (GI) tract. Congenital bronchoesophageal fistula (BEF), if not associated with esophageal atresia, is a rare anomaly.

In 1965 Braimbridge and Keith classified congenital BEF. They suggested four subtypes based on anatomic description of the fistulous tract.1

Type 1: An esophageal diverticulum is associated with a fistula.

Type 2: A short tract directly runs from the esophagus to the bronchus or the trachea.

Type 3: A fistula communicates between the esophagus and a cyst in the lung lobe.

Type 4: A fistula runs into a sequestered lung segment.

Type 2 is the most prevalent and comprises almost 90% of all cases.1 Most of reported cases are type 2. The largest literature review by Risher et al. identified 100 cases of BEF, of which only three were classified as type 4.2

Our patient underwent rigid bronchoscopy that showed absence of right main bronchus. An upper GI contrast study showed leakage of contrast from the lower esophagus into the right lung confirming a right BEF (Figure 4).

At right posterolateral thoracotomy, a heavily consolidated hypoplastic right lung with a single right pulmonary artery and vein was discovered. The right main bronchus was originated from the lower third of the esophagus (Figure 5). The right lung was removed and the fistula was ligated.

We describe an unusual form of bronchopulmonary foregut malformation (type 4 of BEF) in which the right main bronchus originated from the lower third of the esophagus to supply a sequestered lobe.

In 1874, Kiebs, for the first time, described the occurrence of an abnormal communication between accessory lung tissue and the GI tract.3 In 1946, Pryce coined the term "sequestration" to describe a "disconnected bronchopulmonary mass or cyst with an anomalous systemic arterial supply".4 Although the term pulmonary sequestration has been refined over time, Pryce’s original description endures.4

In 1968 Gerle et al. asserted that "all sequestrations have a common embryogenesis" and introduced a new term "bronchopulmonary foregut malformation" (BPFM).3 This has come to include a seemingly disparate group of congenital anomalies affecting the cardiorespiratory and upper GI systems. As more cases came to light, it became clear that a sequestered lobe might have a pulmonary vascular supply, as was in our case.3–5

The embryogenesis of BPFMs has been the source of speculation since first described. Epinger and Schauenslein, in 1902, postulated

---

**Figure 4.** Upper gastrointestinal contrast study demonstrating a right bronchoesophageal fistula.

**Figure 5.** The right main bronchus originating from the lower third of the esophagus.

---

**Photoclinic Diagnosis:** Right Pulmonary Hypoplasia with Bronchoesophageal Fistula
that an additional tracheobronchial bud developed from the primitive foregut distal to the normal tracheobronchial bud, with its own blood supply, migrated caudally, giving rise to a sequestered lobe. 3–5

Gerle et al. emphasize two additional points. 3 Firstly, the timing of the accessory budding is believed to be the principal determinant of the final pathology. And secondly, the absence of a patent foregut connection in most cases of sequestered lobes can be explained by its involution as it outgrows its blood supply. This concept has wide, though not universal, acceptance. There are at least six other widely published models. 6 Clements and Warner’s wheel theory proposes that an insult to the tip of the developing bronchial bud is responsible for the array of morphologic abnormalities. 7 The nature of this “insult” is yet to be determined; trauma, ischemia, infection, and adhesions are postulated. Probably more important to the eventual outcome, as proposed in other models, is the timing and severity of the insult. 5

BPFMs are rare. In 1976, after an extensive review of the published literature, Heithoff et al. described 10 cases of a sequestered lobe involving the entire right lung and maintaining a connection to the GI tract. 6 In 1992 De León-Cantú et al. described an infant with right pulmonary hypoplasia and BEF. 8

Another case was presented in 2006 and our case presented here, changes the number of cases recorded in the medical literature to 13. 5

References