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**Case Report**

# Esophageal lung associated with VACTERL anomaly: a case report

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## ABSTRACT

Esophageal lung is an extremely rare type of bronchopulmonary foregut malformation in which the main stem bronchus is anomalously connected to the esophagus instead of the trachea and is supplied by branches of pulmonary artery. Less than 25 cases have been reported so far in literature. We hereby report first case of esophageal lung associated with VACTREL anomaly. An 8-month-old female child with weight of 4.7 kg presented with recurrent lower respiratory infections. She was diagnosed to have esophageal lung on basis of clinical examination, chest X-ray f/b computed tomogram of chest and esophagobronchoscopy. There was a bifid vertebra at D9 level with scoliosis, an ectopic left kidney and dextrocardia with patent foramen ovale. Thus, this case was classified as having VACTREL association. Patient was successfully operated for excision of the hypoplastic lung via thoracotomy. She recovered well and gaining weight at 3 months f/u without any respiratory infection since surgery. High index of suspicion is necessary for the diagnosis of esophageal lung when common causes of recurrent respiratory infections are ruled out.

**Keywords:** Esophageal lung, Surgical management, VACTERL anomalies

## INTRODUCTION

Esophageal lung is an extremely rare type of bronchopulmonary foregut malformation in which the main stem bronchus is anomalously connected to the esophagus instead of the trachea and is supplied by branches of pulmonary artery. Less than 25 cases have been reported so far in literature.[1-4] Majority of the cases have been associated with esophageal atresia.[1,2]

Most cases are treated with detachment of the esophageal bronchus, with repair of the esophagus and resection of the hypoplastic lung. We present a case of isolated unilateral esophageal lung that was associated with a bifid thoracic vertebra, an ectopic right kidney and a patent foramen ovale (VACTERL group of anomalies). The patient was successfully treated by resection of hypoplastic lung and repair of esophagus.

## CASE REPORT

An 8-month-old female child with weight of 4.7 kg (less than 5$^{th}$ percentile for age), presented with recurrent lower respiratory infections in form of fever, failure to thrive, productive cough and increased respiratory rate requiring admission to pediatrics ward twice in 3 months. There was no history of cyanosis or other signs of congenital heart disease. Antenatal history revealed no significant fetal anomaly. Baby was exclusively breastfed till 6 months age and complimentary feeds were started as per schedule. She had achieved milestones on time till date. Clinical examination revealed scoliosis in thoracic region with convexity to left. There were crepitations on left side of chest and respiratory sounds were significantly reduced on right side. Apex beat was found on right side in 4$^{th}$ Intercostal space in the midclavicular line and there was no murmur.

Blood workup showed microcytic anemia with hypoproteinemia. Chest x-ray revealed opacity on the right side of chest, right sided deviation of trachea with dextrocardia. There was a bifid vertebra at D9 level on the left side with 2 ribs arising from it along with prominent Broncho vascular markings. Ultrasound abdomen showed right ectopic kidney in iliac fossa. Echocardiography confirmed dextrocardia, normal four chambered heart with small patent foramen ovale of 2mm with insignificant left to right flow.

High Resolution Computer Tomography (HRCT) showed non-visualization of right main bronchus with an abrupt cut off just distal to carina. There was a complete collapse of right lung with patent bronchus seen within the parenchyma. The bronchus could be traced to be communicating with the lower esophagus at vertebral level D7-D8. This tissue was supplied by branches from the right pulmonary artery. Shift of trachea and mediastinum to right gave a false primary impression of dextrocardia.

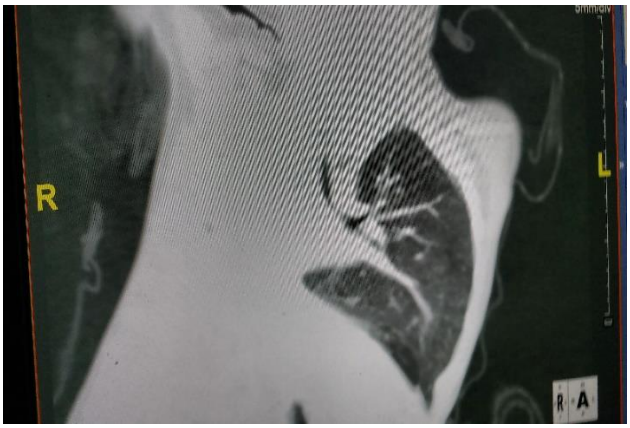


**Figure 1: Non visualisation of right lung.**

Pre - operative esophagoscopy was done to confirm the diagnosis. A fistulous opening was identified at 16cm from the angle of mouth. On bronchoscopy, the trachea was continuous with left main bronchus. Main bronchial division was absent.

At thoracotomy, there was a hypoplastic right lung with a few enlarged lymph nodes at the hilum. The right main stem bronchus was communicating with the lower esophagus. No communication with the trachea could be identified. The esophageal end of the bronchus was ligated with non-absorbable suture and the lung was resected.

The patient was monitored in ICU for 2 days and received intravenous antibiotics for 5 days. Feeds were resumed on 3rd post op day and the intercostal drain was removed on 5th post op day. Histopathology report revealed chronic inflammatory infiltrate in a background of collapsed alveoli. The bronchus showed presence of cartilage.

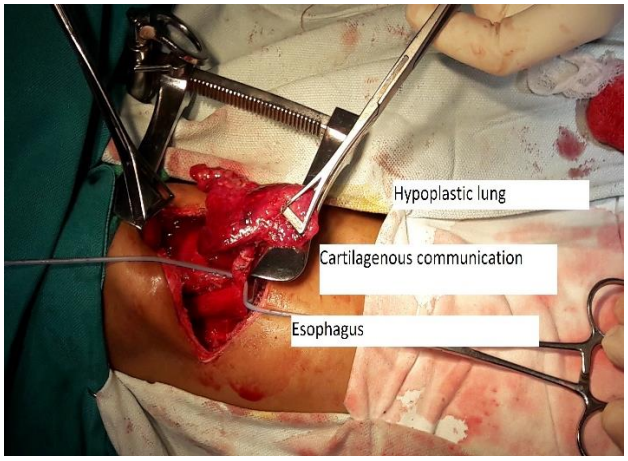


**Figure 2: Intraoperative appearance of the hypoplastic lung and its communication with the esophagus.**

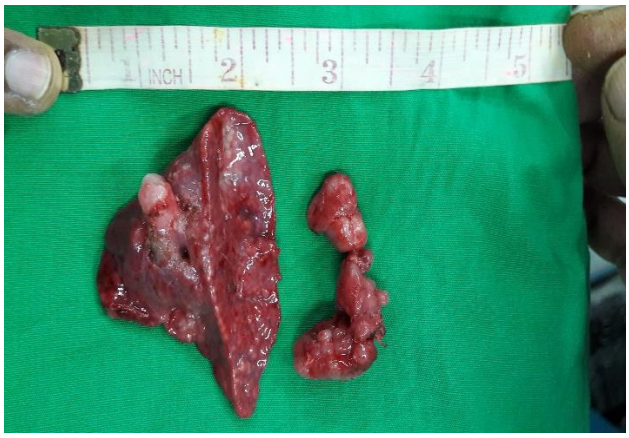


**Figure 3: Hypoplastic lung and necrotic lymph nodes.**

At 2 months of follow-up, patient has no respiratory distress and thriving well. There are no recurrent respiratory tract infections and patient has gained weight.

## DISCUSSION

Bronchopulmonary foregut malformations (BPFM) are rare congenital malformations, which usually present in early neonatal age. The spectrum ranges from Congenital Pulmonary adenomatoid malformation (CPAM), intralobar and extralobar pulmonary sequestration (EPS) with associated foregut diverticulae, duplication cysts, tracheoesophageal fistulae (TEF), and bronchoesophageal fistulae.[3] Esophageal lung is a rare type of communicating BPFM where the entire lung or part of the lung communicates with the esophagus by a bronchus and does not communicate with the native trachea-bronchial tree. It is differentiated from pulmonary sequestration by its arterial supply, extent of pulmonary involvement, and esophageal origin of the involved main stem bronchus.[5]

Esophageal lung is a very rare is very rare and till date, less than 25 cases have been reported in English literature. The left lower lobe and the entire right lung are the most common locations of pulmonary involvement.[1] The communication with the esophagus was found most commonly in the distal portion of the esophagus or gastro-esophageal junction. The majority of the patients with unilateral disease (60%) present in the first eight months of life, and the incidence in females was nearly twice that in males. Almost 50 percent cases have associated Esophageal Atresia/ Tracheo-esophageal fistula.[4,7] The arterial supply comes from either the pulmonary artery or an independent branch from the aorta, or both. A few cases with systemic blood supply have also been observed.[5] The venous drainage is to the pulmonary vein, portal vein or azygous system. Chronic cough, recurrent pneumonias and respiratory distress are the most common presentations.[2] Bronchiectasis, gastrointestinal hemorrhage, dysphagia and hemoptysis may also be the presenting symptoms rarely.[3]

Esophageal lung has been classified by Srikanth et al into 4 major classes:[6]

- Group I: (16%) - Anomaly is associated with esophageal atresia and tracheoesophageal fistula:
    a) IA - Entire lung arises from esophagus or stomach
    b) IB - A portion of one lung or lobe arises from esophagus
- Group II: (33%) - One lung originates from the lower esophagus. Ipsilteral mainstem bronchus is absent from trachea. Trachea extends to form contralateral mainstem bronchus
- Group III: (46%) - An isolated anatomic lung lobe or segment communicates with the esophagus or stomach and is supplied by pulmonary artery, aorta or both
- Group IV: (5%) - A portion of the normal bronchial system communicates with the esophagus. The portion of the lung served by the communicating bronchus receives systemic blood supply.

Accordingly, the index case falls into group II where the entire right lobe of lung was arising from the esophagus, was not associated with a trachea-esophageal fistula and supplied by branches of pulmonary artery.

Diagnosis requires a high index of suspicion with recurrent lower respiratory infections and respiratory distress in early infancy, not responding to treatment. As seen in the index case, the severely hypoplastic lung may give a false impression of dextrocardia on chest x-ray. Confirmation of diagnosis is generally made using HRCT of Chest with pulmonary vasculature. Other complementary modalities of diagnosis are fibreoptic upper gastrointestinal endoscopy, bronchoscopy and 2D Echocardiography. In this context, due to presence of air specs on HRCT films in the lung tissue, rigid bronchoscopy was done in the index case to rule out communication of the lung tissue to the native bronchial tree.

Associated anomalies in BPFM include pulmonary hypoplasia, agenesis, malformations of heart, kidneys, ribs, vertebral column and digestive tract.[3,6] In our patient, there was a bifid vertebra at D9 level with scoliosis, an ectopic left kidney and dextrocardia with patent foramen ovale. Thus, the patient can be characterized as a case of VACTERL group of anomalies. However, there was no associated neurodeficit in the patient and the creatinine was within normal range. The cardiac anomaly was clinically silent. After extensive literature search, this is only the second case report of association of esophageal lung with VACTERL group of anomalies.[8] The associated anomalies are a major cause of morbidity and mortality in patients with unilateral disease. Hence these needs to be assessed pre-operatively and managed individually.

## CONCLUSION

Esophageal lung needs to be suspected in patients with recurrent lower respiratory infections when other causes are ruled out. The condition should be suspected even more, when associated with trachea-esophageal fistula. Patients maybe falsely diagnosed, as dextrocardia and HRCT of chest should be done in doubtful cases. Treatment with thoracotomy and excision of lung relieves the patient off the symptoms. Associated anomalies should be addressed promptly as they are a cause for major morbidity and mortality.

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