Bilateral congenital lobar emphysema: staged management☆,☆☆

Lindsey Perea a,*, Thane Blinman b, Joseph Piccione c, Pablo Laje b

a Department of Surgery, Philadelphia College of Osteopathic Medicine, 4170 City Avenue, Philadelphia, PA, 19131, USA
b Division of Pediatric General, Thoracic and Fetal Surgery, Department of Surgery, Children's Hospital of Philadelphia, 3401 Civic Center Blvd, Philadelphia, PA, 19104, USA
c Division of Pulmonary Medicine, Department of Pediatrics, Children's Hospital of Philadelphia, PA, 19104, USA

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A B S T R A C T

Background: Only a few isolated cases in the literature exist to guide management of bilateral congenital lobar emphysema (CLE). Here, we review our experience in infants with bilateral CLE.

Methods: A case series of all infants presenting with bilateral CLE from 2014 to 2015 in a single institution.

Results: Four patients underwent intervention, with all having right middle lobe (RML) and left upper lobe (LUL) affected. Preoperative planning with computed tomography angiography (CTA) chest allowed a tailored approach based on specific radiologic features. All patients also underwent bronchoscopy to evaluate the anatomy and to assess for alternative causes of airway compression. Three patients underwent unilateral lobectomies, two RML and one LUL. All are growing normally and on room air more than one year later. The last patient underwent a staged procedure beginning with left upper lobectomy followed by right middle lobectomy two weeks later after exhibiting rebound hyperexpansion of the remaining diseased lobe. Thoracoscopy was precluded by mass effect in all patients. No patients underwent emergent lobectomies. One patient had pulmonary interstitial glycogenosis (PIG) in the setting of CLE, first reported case of bilateral CLE with PIG.

Conclusions: This study supports a staged, image-guided, physiology-based operative approach to bilateral CLE. Excision of both diseased lobes does not appear to be mandatory, at least in the short-term follow up, and comport with a “the least intervention that is the most effective” philosophy. CTA is critical for planning, but the role of V/Q scan is not defined. Thoracoscopy appears to have no role.

Level of evidence: Treatment Study; Level IV.

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Congenital lobar emphysema (CLE), or congenital hyperlucent lung, is a rare condition in which over-inflation of one or more lobes occurs secondary to obstruction, either intrinsically or extrinsically. It is thought to result secondary to multiple disruptions in bronchopulmonary development [1]. As air enters the lung, a ball-valve mechanism occurs, trapping air from exiting the lung, further exacerbating the condition [1]. Infants become symptomatic early on in life with about one third symptomatically at birth [2]. Fifty percent of children are symptomatic by one month of life and almost all by six months of age [2].

Single lobe, congenital lobar emphysema occurs with an incidence of 1 in 70–90,000, and a prevalence of 1 in 20–30,000 [2]. The most commonly affected lobe is the left upper lobe (40–50% of cases), followed by the right middle lobe (25–35% of cases) [2]. Oftentimes diagnosis can be made from plain radiographs demonstrating overinflation and mediastinal shift with compression of unaffected lobes/lung. At times, CLE can be diagnosed prenatally with ultrasound. CLE can be distinguished from other congenital lung lesions such as congenital cystic adenomatoid malformations and bronchopulmonary sequestrations, because of increased echogenicity and reflectivity [3].

Only a few isolated cases in the literature exist to guide management of bilateral CLE [4]. Our institution seems to have acquired a cohort of patients with bilateral CLE. This study looks to report our experience of patients from 2014 to 2015 in what we believe is the largest series of patients found to have bilateral congenital lobar emphysema. Additionally, we look to report on our management and outcomes of these patients.

1. Methods

A case series including all infants presenting with bilateral CLE from 2014 to 2015 in a single institution.

2. Results

Table 1 summarizes the pertinent findings regarding each patient described in the following cases.
2.1. Case one

A 39 1/7-week female presented to our facility as a transfer from an outside hospital at day of life (DOL) 82. She required transfer to the neonatal intensive care unit (NICU) because of tachypnea on her second day of life. At the referring facility, bronchoscopy demonstrated narrow right middle lobe and left main bronchi. Right upper lobe atelectasis was also noted. Plain radiographs demonstrated both right middle and left upper lobe air trapping. Computed tomography angiography (CTA) of the chest noted small aberrant bronchi (Image 1). Because of continued respiratory distress the patient underwent tracheostomy. Her course included inhaled nitric oxide, as well, that was started before transfer to our facility.

The patient underwent repeat bronchoscopy with our pulmonary team and was found to have non-pulsatile compression of the left mainstem bronchus at the takeoff of the left upper lobe, which failed to improve despite increasing positive end expiratory pressure (PEEP). The right middle lobe bronchus orifice was severely compressed as well. In order to assess the contribution of these lobes to the patient’s alveolar ventilation and their impact on ventilation–perfusion relationships, selective bronchial occlusion was performed under fiberoptic flexible bronchoscopic guidance using a balloon catheter. During occlusion of the right middle lobe, the adjacent lobes re-expanded and the PEEP and peak inspiratory pressure (PIP) requirements decreased dramatically.

Given the improvement in right upper and lower lobe re-expansion following balloon catheter occlusion of the right middle lobe, the infant underwent a right middle lobectomy via thoracotomy. Prior to discharge her tracheostomy was decannulated and she was breathing room air. Pathology revealed an alveolated lung with congenital lobar emphysema features along with chronic lung disease. She was discharged to home almost three months after surgery.

2.2. Case two

A 38 2/7-week male presented to our facility as a transfer from an outside hospital at day of life ten for further management and pulmonary evaluation. At the referring facility, the infant was noted to have intermittent desaturations, tachypnea and increased work of breathing. Chest radiograph at the outside hospital demonstrated right upper lobe atelectasis. CTA chest was obtained demonstrating relative overinflation of both the right middle lobe and left upper lobe without ability to visualize their respective bronchi (Image 2). Of note the right middle lobe extended into the mediastinum anterior to the thymus. These findings were concerning for either bronchial atresia or congenital lobar emphysema.

The patient underwent flexible bronchoscopy at 18 days of life demonstrating bronchomalacia of both the right middle lobe and left upper lobe atelectasis. CTA chest was obtained demonstrating relative overinflation of both the right middle lobe and left upper lobe without ability to visualize their respective bronchi (Image 2). Of note the right middle lobe extended into the mediastinum anterior to the thymus. These findings were concerning for either bronchial atresia or congenital lobar emphysema.

The patient underwent flexible bronchoscopy at 18 days of life demonstrating bronchomalacia of both the right middle lobe and left upper lobe bronchi and apparent non-pulsatile compression. The patient was taken for right middle lobectomy via thoracotomy given the bronchoscopic findings coupled with the herniation of the right middle lobe across midline. Post operatively the patient had continued collapse of the right lung and underwent repeat bronchoscopy to evaluate for mucus impaction and attempt to assist lung re-expansion.
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