

Management of Congenital Lobar Emphysema: The Current Challenges

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Abstract

Congenital lobar emphysema (CLE) is a rare but serious developmental lung malformation. It has a wide range of clinical presentation, but commonly with life-threatening acute respiratory distress at neonatal period. Asymptomatic cases may present in late childhood or early adulthood. This malformation may associate with other congenital malformations practically of cardiac origin. Misdiagnosis on initial chest X-rays with pneumothorax is common with a probably fatal insertion of intercostal tube. Early diagnosis and treatment of CLE improves outcome and reduces mortality. A controversy remains regarding diagnostic methods, and the choice of appropriate conservative or surgical treatment. This review targets the literature for the current established and controversial management issues about CLE in practice.

Abbreviations: Congenital cystic adenomatoid malformation (CCAM), Congenital heart disease (CHD), Congenital lobar emphysema (CLE), Computed tomography (CT), Magnetic resonance imaging (MRI), Patent ductus arteriosus (PDA), Pressure regulated volume controlled (PRVC), Video-assisted thoracoscopic surgery (VATS)

Keywords: Lung Malformations, Congenital Lobar Emphysema, Thoracic Surgery, Pediatric Surgery

1. INTRODUCTION

Development of the respiratory system begins at the 3rd week of gestation. The developmental aberrations may result in structural abnormalities called bronchopulmonary foregut malformations that may be presented at neonatal period as parenchymal abnormalities of the lung [1]. Congenital lobar emphysema (CLE) is one of these rare and serious developmental lung malformations, occurring in 1:20,000 to 1:30,000 deliveries [2], and characterized by partial obstruction of the bronchus resulting in hyperinflation of lung lobe.

It is usually unilateral with a male preponderance. The most commonly involved lobes are the left upper lobe (40-50%) and the right middle lobe (30-40%) [3, 4]. However, involvement of pulmonary segments [5], and bilateral lung involvement were reported [6].

CLE commonly presented at the neonatal period, but rare cases were reported in late childhood or early adulthood [7, 8]. There is no uniform etiology of CLE. Up to 50% of cases have no cause, while congenital defect of

cartilage presents in 25% and other causes of bronchial obstruction underlying the remaining 25% of cases [9]. There is an additional evidence for inherited factors in the etiology of congenital lobar emphysema. Roberts et al [10] described two cases of CLE involving the right upper and middle lobes in a father and son secondary to relative deficiency of the bronchial cartilage.

CLE may associate with other congenital cardiac malformations, such as patent ductus arteriosus, atrial septal defect, ventricular septal defect, total anomalous pulmonary venous return, and Tetralogy of Fallot [11]. In addition, CLE may associate with double superior vena cava, horseshoe kidney [12], and polysplenia [13].

Controversy remains concerning the diagnosis and treatment of congenital lobar emphysema, with a wide variation in practice regarding its management [14, 15]. Therefore, this review of literature highlights the established and controversial concerns about the management of CLE in the current practice of thoracic pediatric surgery.

2. CLINICAL PRESENTATION

Clinical presentation of CLE varies from no symptoms to severe respiratory distress [3]. It may present as life threatening respiratory distress commonly within the first 6 months of life due to compression atelectasis, mediastinal shift, hypoxia, and associated hypotension [16].

Unusual presentations of CLE reported in literature. Rusakow and Khare [17] described radiographically occult CLE presented as unexplained neonatal tachypnea. Arnaud et al [18] presented a case of a 33-year-old lady with progressive exertional dyspnea.

3. DIAGNOSTIC METHODS

Ultrasound and fetal magnetic resonance imaging (MRI) help prenatal diagnosis of CLE and associated complications such as polyhydramnios and fetal hydrops [19]. Ultrasound may demonstrate gradual increase or decrease of the lesion echogenicity [20]. However, increased echogenicity of the lungs that could be too subtle to be appreciated in utero, affects prenatal ultrasound diagnosis of CLE [21]. Fetal MRI should differentiate CLE from microcystic congenital cystic adenomatoid malformation (CCAM) and bronchopulmonary sequestration because of the former's homogeneity and intact lung structure with stretched hilar vessels [22]. It is important to continue postnatal investigations of prenatal ultrasound abnormalities even when they appear to have resolved [23].

Early neonatal diagnosis is crucial and in many cases is complicated due to the variety of its clinical presentation that varies from mild tachypnea to severe respiratory distress [19]. Diagnosis is often confused with pneumonia and pneumothorax [24]. A case of CLE with pneumothorax due to a large bulla in an adult has been reported [25]. It is a significant risk during an aesthetic induction if undiagnosed preoperatively [26].

The differential diagnosis of CLE includes pneumothorax, cysts, and diaphragmatic hernia that can be ruled out by linear bronchovascular and alveolar markings on chest x-ray. Other conditions need differential diagnosis include pulmonary sequestration, CCAM and upper airway obstruction that can be ruled out by ultrasonography, color doppler imaging and magnetic resonance imaging [27].

The initial diagnosis carried by chest X-ray (Figure 1) and confirmed by computed tomography scan (Figure 2) and histopathological studies (Figure 3) [28]. The

typical chest X ray features include presence of bronchovascular markings with transillumination, and secondary compression of the over expanded lobe (tracheal or a mediastinal shift to the opposite side, atelectasis of the adjacent lobes or herniation of the expanded lobe across the midlines). A region of increased density rather than a hyperlucency may lead to a misleading diagnosis of pneumonia [29].

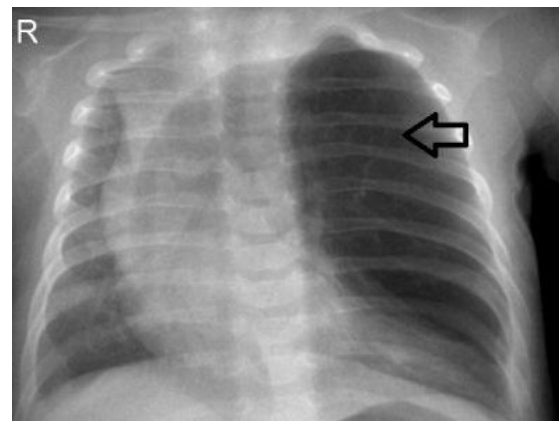


Fig1. Chest X-ray showing overexpanded hyperlucent left upper lobe (Arrow) with mediastinal shift. R: Right side.



Fig2. Chest computed tomography scan (lung window) showing congenital lobar emphysema of right middle-lobe (Arrow) compressing adjacent tissues with mediastinal shift. R: Right side.

Chest computed tomography (CT) has a definite role in early diagnosis with a special importance to avoid inadvertent chest tube insertion in such cystic lucent lung lesion on chest X-ray, and hence avoid associated complications [30]. Chest imaging with CT or MRI can save life of a neonate when there is a high suspicion of CLE. The non-improvement of the distress and non-expansion of the lung in neonate presented with respiratory distress increases the suspicion of congenital lobar emphysema, and indicates chest CT or MRI [31].

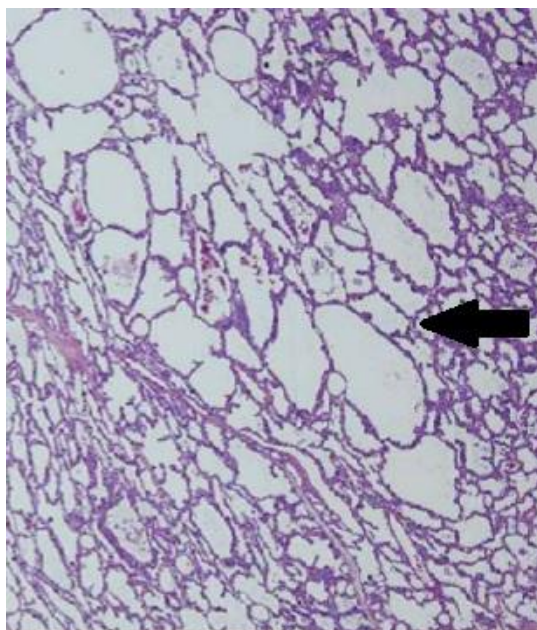


Fig3. Histopathological specimen of left upper lobe showing over distension of alveoli and thin interalveolar septa (Arrow).

Nuclear imaging by ventilation perfusion scanning (Figure 4) is useful to represent the reduced perfusion of affected lobe due to vessel compression and increased perfusion of normal lobes due to shunting [32]. It is helpful in older patients with fewer symptoms to demonstrate the extent of surgical resection for segmental lobar involvement [33].

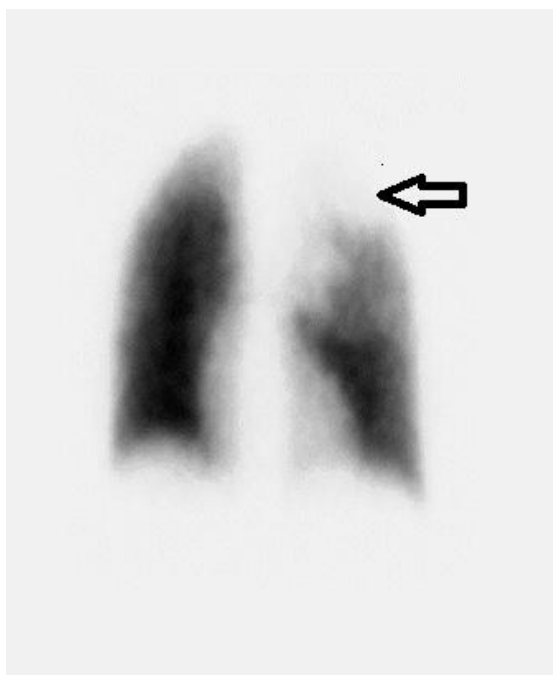


Fig4. Pulmonary ventilation perfusion scan showing reduced perfusion in left upper lobe (Arrow).

Bronchoscopy is useful in evaluating airway patency and dynamic changes in the airway, and in excluding obstruction by aspirated material or

from external compression, but its routine use is often unnecessary. Flexible bronchoscopy is superior to rigid bronchoscopy in demonstrating dynamic changes in the airway [34, 35].

Multislice CT with reconstructions of the bronchial tree and virtual bronchoscopy are important imaging tools for differential diagnosis of CLE and bronchial foreign bodies, as both may present with pulmonary hyperinsufflation findings and nonspecific clinical features [36]. Presence of other congenital cardiac malformations may require further echocardiographic evaluation and/or cardiac catheterization [37].

3.1. Anesthetic Challenges

Anesthetic management of a child with CLE is challenging. Precautions during induction of anesthesia and endotracheal intubation avoid further gas in the emphysematous lobes to prevent an increase of intra thoracic pressure, which further decreases the respiratory reserve. During induction, it is critical to avoid crying and struggling, and maintaining the airway pressure of 20–25 cm of H₂O through gentle manual ventilation. In addition, it is important to reduce the time between anesthesia induction and thoracotomy [38, 39].

A debate surrounds optimal technique of ventilation. Advocates to avoid positive pressure ventilation is based on subsequent further inflation, thus, lung isolation or selective main stem bronchus intubation is desirable. The useful techniques of ventilation for resection of CLE include pressure regulated volume controlled (PRVC), high frequency ventilation, and pressure controlled ventilation keeping airway pressure below 20 cm H₂O to control the delivered tidal volumes [40–42].

For a single stage excision of bilateral CLE, Iodice et al [43] suggested inhalational induction, maintained spontaneous ventilation with sevoflurane in 100% oxygen, and only instituted positive pressure ventilation after femoral and arterial line access. This allowed minimize the duration of positive pressure ventilation prior to surgical incision and reduce the potential for hyperinflation of the emphysematous lobes. This method may facilitate safe induction of anesthesia and establishment of invasive monitoring, avoid the potential for cardio respiratory compromise.

3.2. Treatment

Although surgery is the traditional routine management of congenital lobar emphysema, the contemporary management directs toward

conservative non-operative approach with follow-up in asymptomatic or mild symptomatic patients [44]. Long-term evaluation of surgically and conservatively treated asymptomatic children showed no difference in lung growth which confirmed that asymptomatic or mild symptomatic patients do not benefit from surgical treatment [45]. The conservative management involves close follow-up of the children, alarming the family about the disease features [46].

Severe respiratory distress is the main indication for surgical excision. Evidence of mediastinal shift with subsequent compression of the unaffected lung lobes indicates surgical excision. Some surgeons recommended surgery in all infants younger than 2 months or older than 2 months with severe respiratory symptoms [19, 35].

The surgical options for treatment of CLE include lobectomy through thoracotomy or video-assisted lobectomy [47]. Segmental resections can be performed safely while conserving healthy lung tissue. In one study, median hospital stay was longer for the lobectomy at 7 days when compared to the segmentectomy group at 2 days. There was not a difference in complication rate (21% vs. 19%, $p=1$) or in median number of chest tube days (2 vs. 3 days, $p=0.07$) for segmentectomy versus lobectomy patients [48].

Management of concomitant congenital heart disease (CHD) and CLE depends on severity of cardiac lesion and ease of management. In cases with high pulmonary artery pressure, the management is palliative or corrective surgery for CHD with lobectomy for CLE. Clinical follow-up of CHD is sufficient treatment after lobectomy for small lesion without high pulmonary artery pressure. When CLE is a result of compression associated with patent ductus arteriosus (PDA), ligation of PDA before lobectomy may be considered [37].

Video-assisted thoracoscopic surgery (VATS) is a serious alternative to traditional thoracotomy for children with CLE as it results in reduced length of postoperative period, lesser amount of complications and good cosmetic and functional results [49]. Granato et al [50] reported first case of endoscopic parenchymal sparing resection in CLE carried out by 3-portal VATS using single-lung ventilation leading to precise determination the limit to resect due to the obvious and clear-cut distinction between functioning and non-functioning parenchyma of the lobe.

Surgically treated patients remain asymptomatic with normal growth and development. They may have abnormal postoperative pulmonary function tests [51], or have some compensatory growth of the remaining lung tissue which rule out functional impairment owing to loss of lung tissue or residual disease [52].

The contemporary reported unfavorable postoperative outcomes after surgical excision of CLE include death in up to 13% of cases, pneumothorax, delayed weight gain, permanent oxygen dependency, pneumonia, atelectasis, post-operative bleeding, and wound infection. The number of affected lobes and base deficit at the time of admission are associated with mortality [53, 54].

4. CONCLUSIONS

Congenital lobar emphysema is a rare and serious developmental malformation still with multiple controversial issues about its etiology, presentation, natural history, diagnostic workup, and treatment options. High index of suspicion and careful differential diagnosis help to save life of patients with CLE. Combination of clinical features, radiological characteristics, and histological abnormalities provides the appropriate diagnosis. Approach by multidisciplinary team of pediatrician, surgeon, radiologist, and anesthetist is important to obtain better outcome of such surgically correctable cause of neonatal respiratory distress. Surgical techniques to preserve more of healthy lung parenchyma and segmental resection need further evaluation.

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