

# Congenital lobar emphysema: diagnosis and treatment options

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**Abstract:** Although congenital lobar emphysema is a rare lung disease, it can cause severe respiratory distress in the newborn. Lobectomy can be difficult because of the hyperinflated lobe and limited space to carry out surgery. During the past two decades, conservative treatment options have increased for patients with mild and moderate disease.

**Keywords:** congenital lobar emphysema, overinflation, lobectomy

## Introduction

Congenital lobar emphysema (CLE) is a rare developmental lung malformation. During the third week of gestation, the development of the respiratory system begins and aberrations in this developmental stage may cause parenchymal lung malformations.<sup>1–3</sup> Congenital alveolar overdistension, congenital large hyperlucent lobe, and congenital lobar overinflation are other terms synonymous with CLE.<sup>4</sup> In this review, we chose to use CLE owing to the widespread use of this term. CLE is defined as the hyperinflation of one or more pulmonary lobes due to the partial obstruction of the bronchus, which causes pressure symptoms on the adjacent organs.<sup>5</sup> CLE is one of the rarest causes of respiratory distress in newborns.<sup>6</sup> It was first described by Nelson in 1932 and later by Robertson and James in 1951.<sup>7,8</sup>

## Epidemiology

CLE is a rare lung malformation. Its incidence is 1/20,000–30,000 live births.<sup>9,10</sup> The prenatal incidence is unknown because of diagnostic difficulties with ultrasonography (USG) in this period.<sup>2,11</sup> It is more common in males, and the male to female ratio is 3:1. One-third of cases are symptomatic at birth and nearly all of them are diagnosed in the first 6 months of life.<sup>12–14</sup> The most common observation of this disease is left upper lobe involvement (43%), followed by right middle lobe (32%) and right upper lobe (21%) involvement. Lower lobe involvement (2%) is the rarest form.<sup>12–16</sup> In the literature, more than one lobe and bilateral involvement have been described.<sup>17,18</sup>

## Histopathology

No tissue damage is observed histologically such as that found in acquired emphysema. Pathological investigation shows a normal acinus structure and overgrown alveoli.<sup>19</sup> The majority of cases show normal radial alveolar numbers. However, there is no significant maturation with age compared to age-matched controls, suggesting that acinar development in the affected lung tissue has stopped in the

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postpartum period. In a small number of cases, true alveolar hyperplasia with increased radial alveolar counts is seen. This entity is called polyalveolar lobe.<sup>12</sup>

## Etiology

In half of CLE patients, the etiology is unknown. Absence of bronchial cartilage, hypoplasia, or dysplasia is present in one-quarter of cases. Incomplete or defective cartilage structure causes weakening of the bronchial wall, which allows bronchial collapse leading to air trapping in expiration.<sup>20,21</sup> In a patient series, Lincoln et al<sup>22</sup> demonstrated an absence of bronchial cartilage or hypoplasia in 22 of 28 patients. The etiology is undetermined in approximately 50% of patients.<sup>20,23–27</sup> Table 1 shows a summary of the etiologies in CLE patients. Parenchymal diseases are one of the rare causes of CLE. In the 1970s, Hislop and Reid proposed the polyalveolar lobe theory for the etiology of CLE.<sup>28</sup> Histopathological examination showed that the alveolar number in the affected lobes was three to five times higher than in the other, normal parts.<sup>12</sup> It is not known why the air is trapped in this polyalveolar lobe. Tapper et al demonstrated that six of 16 cases reported previously as CLE were diagnosed with polyalveolar lobe.<sup>29</sup>

Bronchial disease is another cause of CLE. Bronchial stenosis, atresia, bronchomalacia, bronchiectasis, and

abnormal bronchi are congenital causes of bronchial diseases, which lead to CLE. Meconium aspiration, hypertrophic mucous membranes, dark mucous plaques, and foreign body aspirations are acquired causes of CLE.<sup>19,24,25,30–34</sup>

Diseases stemming from organs adjacent to the lungs and bronchus may cause CLE. Vascular abnormalities such as pulmonary arterial sling anomalies and abnormal pulmonary venous return anomalies may cause CLE.<sup>5,35–37</sup> Bronchogenic cysts and mediastinal tumors are rare extrinsic causes of CLE.<sup>12,19,38,39</sup>

Genetic transition is unknown. CLE was shown in twin babies, in a mother and her daughter, and in a father and his son. The disease is more common in white people than in black people. Although CLE is sporadic, data suggest that hereditary transition may be responsible for the disease.<sup>40,41</sup>

Data from molecular genetics and embryonic organ culture studies showed that primary branching patterns in lung development are regulated by repetitive signaling of the fibroblast growth factor-10 pathway. Nkx2.1 and thyroid transcription factor-1 play a role in the secondary branching pattern.<sup>3,41–44</sup> Although significant mutations in the genetic material controlling these factors result in major anomalies, small errors in transcription may cause localized deficiencies in bronchial cartilage leading to the development of CLE.<sup>3,42–45</sup>

**Table 1** Etiology of congenital lobar emphysema\*

1.	<b>Idiopathic 50%</b>
2.	<b>Bronchial cartilage absence, hypoplasia or dysplasia 25%</b>
3.	<b>Parenchymal diseases</b> – Polyalveolar lobe – Pulmonary alveolar glycogenesis
4.	<b>Internal bronchial obstruction</b> – Bronchial stenosis – Bronchomalacia – Meconium aspiration – Hypertrophic mucosa membranes – Mold mucous plaques – Foreign body aspiration – Bronchial polyp
5.	<b>External bronchial obstruction</b> – Pulmonary artery sling anomaly – Pulmonary rotation anomaly – Bronchogenic cyst – Lymphadenopathy – Mediastinal mass – Duplication of esophagus

**Note:** \*Adapted from references<sup>5,12,16,20–28,34–39,81,82</sup>

## Clinical presentation

Nearly half of patients are symptomatic at birth, while the other half mostly develop symptoms in the first 6 months of life. The affected lobe is overinflated, and ventilation and perfusion are impaired in the overinflated lobe. With progressive overinflation, compression occurs in the adjacent organs.<sup>19</sup> Thus, ventilation and perfusion are impaired in these parts of the lung parenchyma, which leads to progressive respiratory failure. Retractions, wheezing, cyanosis, and difficulty in feeding can be observed. In infancy, wheezing, chronic cough, and recurrent respiratory tract infections can be seen.<sup>12</sup> In the literature, it was reported that a 15-year-old boy was diagnosed with CLE without respiratory symptoms.<sup>46</sup> Some patients can be mistakenly diagnosed with pneumothorax and pneumonia instead of CLE in later life.<sup>47</sup>

On physical examination, hyperresonance on percussion in the afflicted lobe is likely to be detected because respiratory sounds are diminished in that part of the lung. Wheezing and rhonchus can be heard rarely. The affected area cannot be expanded by respiration despite excessive respiratory efforts.<sup>19</sup>

Congenital cardiac defects are likely to accompany CLE. If a cardiac murmur or unexpected cyanosis is observed, patients should be evaluated for cardiac anomalies. Patent ductus arteriosus, atrial septal defect, ventricular septal defect, total pulmonary venous return anomaly, and tetralogy of Fallot are the most common congenital cardiac defects seen with CLE.<sup>36,48–52</sup> The cartilage structure of the bronchial system occurs between 4 and 6 weeks of the intrauterine period and major changes occur in the development of the heart during this period.<sup>52</sup> Therefore, the effect at this stage may affect both lung development and cardiac development. CLE is accompanied by cardiac anomalies 14–20% of the time. For this reason, CLE patients should be investigated for possible cardiac diseases before surgery.<sup>20,22,25,54,55</sup> Other system anomalies and syndromes may accompany CLE rarely. These are shown in Table 2.

## Diagnosis

In the prenatal period, with the routine use of USG, the incidence of congenital lung diseases is rising worldwide. Prenatal USG shows hyperechogenicity in lung segments without abnormal blood flow. A mediastinal shift and/or polyhydramnios may accompany this situation. When an echogenic lung is determined with USG, CLE, congenital cystic adenomatoid malformation, and pulmonary sequestration should be considered as differential diagnoses. Prenatal magnetic resonance imaging (MRI) findings are consistent with the USG findings, with a solid-appearing lesion with hyperintensity in the T2 sequence. While the absence of concomitant cystic lesions is compatible with CLE, it has been reported rarely in patients with CLE in the literature. Today, MRI is superior to USG in prenatal diagnosis owing to the properties of better tissue contrast, greater field of view, increased anatomical evaluation, and identification of other congenital anomalies.<sup>5,14,56–59</sup>

A posteroanterior chest X-ray is the first choice for an examination procedure in patients with respiratory problems. Figure 1 shows the chest X-ray of a patient's right middle lobe. Overinflation and hyperlucency of lung can be seen on the affected side. If overinflation is excessive, the affected lobe will be herniated to the opposite side of the thoracic cavity. A tracheal and mediastinal shift to the opposite side accompanies the herniation of the lobe. Atelectasis and increased density can be seen in the adjacent lobes as a result of the compression.<sup>12,19</sup>

Lung computed tomography (CT) is the gold standard in the diagnosis of CLE. It is useful for evaluation of the anatomy

**Table 2** Concomitant malformations accompanying congenital lobar emphysema\*

Cardiac malformations 14–20%	Patent ductus arteriosus Atrial septal defect Ventricular septal defect Tetralogy of Fallot Pulmonary stenosis Pulmonary valve atresia Aortic coarctation Pulmonary hypertension Left aortic arch Right descending aorta Left ligamentum arteriosum Double superior vena cava
Renal anomalies	Aplastic kidney Horseshoe kidney
Musculoskeletal anomalies	Pectus excavatum Hiatal hernia Diaphragmatic hernia
Gastrointestinal tract	Omphalocele Pyloric stenosis
Others	Cleft palate Chondroectodermal dysplasia Chondrodystrophy Cystinosis Bronchial atresia Tracheal bronchus
Syndromes	Williams–Beuren syndrome Miller–Dieker syndrome Niemann–Pick disease Fanconi aplastic anemia

**Note:** \*Adapted from references<sup>5,12,16,20–28,32,34–39,48–53,86–92</sup>

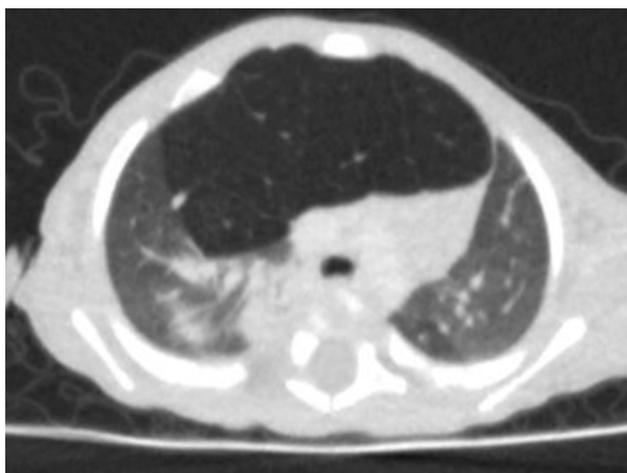
of the emphysematous lobe and herniated lobe. It is also useful in assessing the status of the adjacent lobes and determining whether the contralateral lung tissue is hypoplastic. Contrast-enhanced CT gives information about vascular anomalies and mediastinal masses.<sup>20,60</sup> Figure 2 shows an emphysematous right middle lobe herniated to the opposite side.

Angiography can be performed in cases thought to have CLE as a result of compression of a vascular structure.<sup>61,62</sup>

The use of bronchoscopy in CLE is controversial. Its use in newborn patients with respiratory distress might be dangerous. Thus, in older patients and in patients, who are planned to be treated conservatively, it can be used to remove bronchial plugs, to evaluate anatomical variations, or to distinguish foreign bodies. Figure 3 shows a bronchoscopic image of a bronchomalacia in the right middle lobe in a patient with CLE.<sup>20,63,64</sup>



**Figure 1** Chest X-ray showing overinflation in the right upper and middle zone, and mild shift of the mediastinum to the left.



**Figure 2** Thorax computed tomography showing that the right middle lobe is overinflated and herniated to the left side. Segmental atelectasis is seen in the right lower lobe.

Although radionuclide studies can be very difficult to administer in pediatric patients, they can be used to show that the affected lobe is non-functional and that the compressed lung is functional. Karnak et al used radionuclide imaging in planning patient treatment. However, its use in treatment planning in the literature is very rare owing to difficulties in application.<sup>20</sup>

## Treatment

The first point to be considered in the treatment of CLE is a differential diagnosis. The chest X-ray, which is the first step used in the diagnosis of respiratory distress of



**Figure 3** Bronchoscopic image of bronchomalacia in the right middle lobe.

a patient's image, shows hyperinflation of the affected lobe, which can be confused easily with pneumothorax, pneumatocele, lung hypoplasia, and opposite lung hyperinflation due to atelectasis.<sup>20,65–67</sup> There are patients in the literature who have been diagnosed mistakenly with pneumothorax and have had a chest tube inserted. This situation makes treatment more severe and has a potential for mortality.<sup>9</sup>

The treatment choice for patients with CLE depends on the clinical severity. Approximately half of patients show symptoms in the first month of life. Although there are few asymptomatic patients described in the literature, most of them are diagnosed in the first 6 months of life.<sup>9,20,68</sup> If the symptoms are mild or moderate, conservative treatment is recommended. If the patient has clinical progression or is clinically severe, the traditional treatment option is a lobectomy.<sup>65</sup> Most of the treatment strategies in the literature are case reports or case series. Karnak et al<sup>20</sup> postulated that patients older than 2 months with moderate and mild symptoms should be followed with conservative management. Similarly, Mei-Zahav et al<sup>10</sup> showed that one of eight asymptomatic patients needed a lobectomy in follow-up. In contrast to these findings, Thakral et al<sup>9</sup> presented case series for 21 patients. Unfortunately, four of seven patients needed a lobectomy at follow-up. These findings suggest that asymptomatic CLE patients should be followed closely. In 2018, Fierro et al<sup>69</sup> retrospectively published their experience. In a 3-month-old baby diagnosed with CLE, they occluded the bronchial lumen of the affected lobe with a balloon catheter. They observed the oxygenation, gas exchange, and chest X-ray of the patient. If these parameters did not change with temporary balloon

occlusion of the hyperinflated lobe, they concluded that lobectomy would be likely to be useful in such patients. Therefore, this procedure may assist in the decision to perform a lobectomy.<sup>69</sup>

Induction of anesthesia is very important in the treatment of children with CLE. Excessive crying may cause a valve effect in the lung, which may increase hyperinflation in the affected lobe and increase mediastinal shift. This situation may lead to increased pressure on the vascular structures in the mediastinum. Because of this entity, the circulation deteriorates and cardiac arrest may occur.<sup>70,71</sup> Inhalation induction is a preferred procedure in anesthesia of CLE patients. In the induction period, nitric oxide gas should be avoided because it can increase the mediastinal shift in the lung owing to its rapid spread in the closed cavity.<sup>72</sup> It has been reported that anesthesia induction can be performed safely using oxygen, sevoflurane, or halothane instead of nitric oxide. The critical airway pressure to be applied in these patients is not completely known. It is recommended that high-pressure ventilation should not be performed until thoracotomy and lung tissue is released, and airway pressure should not exceed 20–25 cmH<sub>2</sub>O.<sup>72</sup> If necessary, patients should be ventilated gently. Prabhu and Joseph<sup>73</sup> used mild intermittent positive pressure ventilation following muscle sparing. If possible, the pressure-controlled volume control mechanical ventilator mode may be useful. Goto et al<sup>74</sup> reported the successful use of high-frequency ventilation in CLE patients.

Single-lumen tubes are usually used for tracheal intubation. Double-lumen tubes are not commercially available in this age group. In young infants, lung isolation is technically difficult and is not widely applied. There are cases in the literature of isolated healthy lung intubation, until lobectomy is performed.<sup>75,76</sup>

Although segmentectomy in children with CLE has been published in case reports,<sup>77,78</sup> the traditional treatment of CLE worldwide is still lobectomy. CLE is rarely treated with thoracoscopy, while thoracotomy is still the most common surgical technique for resection of CLE in childhood. In adults, the use of thoracoscopic approaches for the treatment of congenital lung diseases, including CLE, is increasing. In contrast, this practice is controversial in the management of children with CLE because there are several problems in children, such as problems with anesthesia, limited surgical space, more sensitive pulmonary anatomy, complex structure of the disease process and difficulties with surgical dissection, and

decreased pulmonary reserve.<sup>79,80</sup> Kunisaki et al<sup>81</sup> described 51 patients with a diagnosis of CLE, who were treated with lobectomy. Thoracotomy was preferred in 45 of these patients, whereas thoracoscopic approaches was favored in six of these patients. However, the surgical approach for two patients who underwent thoracoscopy returned to thoracotomy because of inadequate surgical space. It should be noted that surgery with video-assisted thoracoscopic surgery in this age group is quite difficult and rarely performed.

Non-anatomical wedge resection has never been reported in the literature. The lobectomy that is performed in CLE patients is different from that carried out in other patients. Depending on the location of the lesion, the patient is approached with a right or left thoracotomy. Single-lumen intubation may expose the affected lobe to positive pressure. This may result in increased hyperinflation and cardiac compression. Therefore, the surgeon should open the thoracic cavity as soon as possible after induction of anesthesia. As soon as the thoracic cavity is opened, the emphysematous lobe will be herniated out of the thorax. The affected lobe cannot be surgically removed during the procedure and this is another difficulty in CLE lobectomy.<sup>82</sup> Therefore, some authors suggest selective intubation to decrease the effect of positive pressure.<sup>83</sup> Although lobectomy is more difficult in children than in adult patients, the rate of complications and mortality is lower.<sup>10,20,65</sup> Bilateral CLE patients are even rarer. In these patients, bilateral lobectomy is unnecessary. In a four-patient case series, only one patient needed bilateral surgery. It was reported that three patients were prescribed one-sided lobectomy, with good survival.<sup>18</sup> It should be kept in mind that external compression is one of the causes of the disease during surgery, and even if preoperative tests have been carried out, the surgeon should check the anatomical structures and make sure that there is no variation in the anatomy of the airway or external pressure to the airway. In a pathological study, an examination of the resection tissue revealed no accessory lobes or vessels. This is perhaps the only good news for the surgeon.<sup>84</sup> In the literature, comparisons of conservative treatment and surgical treatment in long-term follow-up are infrequent, but the results are similar in these publications.<sup>85</sup> Conservative treatment of CLE should be preferred in mild and moderate disease, and lobectomy should be considered in severe disease cases. However, conservative treatment of the disease seems to be increasing as a result of increased antenatal diagnosis and intrauterine regression.

## Disclosure

The authors report no conflicts of interest in this work.

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