

# Anesthesiologist's Dilemma in a Patient with Congenital Lobar Emphysema

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## Abstract

Congenital lobar emphysema is a rare congenital condition that can present in the neonatal period as respiratory distress. The clinical and radiological symptoms are often challenging to diagnose. However, the correct diagnosis and surgical resection of the affected lobe can be life saving because of the severity of this anomaly. The anesthetic management of congenital lobar emphysema can be difficult because of the condition's pathophysiology, the potential deleterious effects of positive pressure ventilation during induction and the associated hemodynamic changes.

## INTRODUCTION

Congenital lobar emphysema (CLE) is a rare congenital anomaly that usually manifests in the neonatal period. It is characterized by hyperinflation and distension of the affected pulmonary lobe due to air trapping and subsequent compression of surrounding structures<sup>1</sup>. This can cause mediastinal shift, in decreasing venous return and cardiac output and resulting in hypotension and hypoxia<sup>2</sup>. With neonates, it may present as an acute respiratory distress or a respiratory failure while in older children it may present as recurrent attacks of respiratory infection or progressive respiratory distress. CLE can present as a life-threatening event in neonates, but early diagnosis and immediate surgical intervention can be lifesaving.

## CASE REPORT

A 2 month-old, 4.1-kg female with failure to thrive was seen by her physician with complaints of increased work of breathing. A right-sided tension pneumothorax was seen on the chest radiograph. A 10F chest tube was placed, and the patient was transferred to a pediatric hospital. Upon arrival, a repeated chest radiograph showed a right tension pneumothorax. A chest CT with contrast revealed marked emphysematous hyperexpansion of the right middle lobe with anterior herniation across the midline. There was minimal visualization of the right-lower and right-upper lobes that were compressed by the emphysematous lung. Also noted was mild right-to-left mediastinal shift with subsequent compression of the left lung.

The patient was diagnosed with congenital lobar emphysema and scheduled for a right-middle lobectomy via right thoracotomy. On the preoperative physical exam she was noted to be awake and tachypneic, but not cyanotic. Her vital signs showed a blood pressure of 88/50 mmHg, pulse rate of 110 beats/minute (bpm), respiratory rate of 58/minute and an oxygen saturation of 95% on 1 liter/minute of oxygen via nasal cannula. Upon auscultation, breath sounds were decreased over the right lung fields. There were no cardiac murmurs and no tracheal deviation.

The patient was taken to the operating room and standard ASA (American Society of Anesthesiology) monitors were placed. An inhalational induction was performed using sevoflurane with 100% oxygen while maintaining spontaneous respiration. After obtaining peripheral intravenous access, 0.5 mcg/kg (microgram/kilogram) of fentanyl was administered. The ventilation was manually assisted with peak airway pressure less than 10 cm (centimeters) water pressure when the patient stopped breathing. The airway was easily secured with a 3.5 uncuffed endotracheal tube (ETT). However, following intubation there was neither chest rise nor end tidal carbon dioxide (CO<sub>2</sub>) detected. Also, no breath sounds were heard over either lung field and oxygen percentage saturation decreased to the lower 80s. Any kinking or obstruction of the anesthesia circuit was ruled out. Direct laryngoscopy confirmed the correct placement of ETT but the suction catheter could not be passed through ETT. Possible ETT obstruction was suspected as the patient could not be

ventilated, ETT was removed and manual ventilation was started using the face mask. Fiberoptic endoscope was not used to check the confirmation of ETT placement or obstruction because oxygenation saturation was rapidly decreasing. Still it became increasingly difficult to ventilate the patient even with an oral airway in place. Oxygen saturation remained near 70%, and the heart rate dropped near 80 bpm. It was suspected that the distended emphysematous lobe was not only compressing the normal lung on the right side, but also increasing the mediastinal shift.

The surgeon was asked to emergently open the right chest and relieve the pressure of the emphysematous lobe while the anesthesiologist manually ventilated the patient using a facemask. Immediately after incision of the pleura, part of the emphysematous lobe herniated through the incision, and mask ventilation improved. The patient's oxygen saturation increased to 98%-100% and the hemodynamics stabilized. The patient was reintubated and surgery was completed without further incident. The remainder of the surgery was without incident and the patient was transported back to the neonatal intensive care unit. She was extubated the next day in the intensive care unit and made an uneventful recovery. Finally, she was discharged home a week later in stable condition.

### DISCUSSION

CLE is also known as congenital localized emphysema, congenital lobar over inflation, unilobar obstructive emphysema or congenital hypertrophic lobar emphysema. The prevalence of this condition is 1 in 20,000 to 1 in 30,000 and the incidence is estimated to be 1 in 70,000 to 1 in 90,000<sup>3,4</sup>. Males are affected more than females with a ratio of 3:1<sup>3</sup>.

The cause of CLE is difficult to determine and none is identified in 50% of cases<sup>5</sup>. The most commonly identified cause is congenital bronchial cartilage dysplasia, which is found in 25% of cases<sup>6</sup>. Guidici and colleagues propose polyalveolosis, an increase in number of alveoli within each acinus, as the etiology of CLE<sup>7</sup>. Other causes implicated in the etiology include bronchial obstruction due to redundant mucosal folds or septum, mucous plugging or anomalous vasculature<sup>3</sup>. The upper lobe of the left lung is the most commonly affected (41%) followed by the right-middle lobe (34%) and right-upper lobe (21%). Associated congenital anomalies are found in 10% to 15% of patients, and the most common involves congenital cardiac anomalies such as atrial

septal defect, ventricular septal defect, and pulmonary hypertension<sup>8</sup>.

The pathophysiology is that of a "ball-valve" type of bronchial obstruction due to weakened or absent bronchial cartilage. This allows lung inflation during inhalation but obstructs the bronchus during exhalation, resulting in lobar air trapping and compression of the surrounding normal lung tissue<sup>9</sup>. Hyperinflation of the emphysematous lobe can cause it to herniate across the mediastinum with mediastinal shift (as seen in this case). This results in compression of the contralateral lung. Compression of the normal lung on the ipsilateral or contralateral side increases intra-thoracic pressure and decreases the volume of functional lung tissue causing ventilation/perfusion (V/Q) mismatch<sup>4</sup>. Mediastinal shift may also cause compression of the heart and major vessels resulting in decrease venous return and cardiac output, which further worsens the hypoxia.

Diagnosing CLE may be challenge due to non-specific signs and symptoms, including dyspnea, cyanosis, tachycardia, wheezing, recurrent infections and failure to thrive. CLE may be suspected with asymmetrical chest expansion, hyper-resonance to chest percussion, displaced cardiac sounds, and diminished heart and breath sounds. Only 5% of cases present after the age of 6 months, usually in the form of recurrent respiratory tract infections or as an incidental finding on chest x-ray<sup>5</sup>.

CLE can be present in the antenatal period as an overinflated fluid-filled lobe, and diagnosed using ultrasonography or ultrafast fetal magnetic resonance imaging<sup>10</sup>. Plain chest x-ray is usually the first step in the diagnosis of CLE. A chest radiogram may show marked hyperlucency of the involved lobe, mediastinal shift to the unaffected side, and flattening of the ipsilateral diaphragm. Radiographic appearance of pneumothorax can be similar to CLE and case reports of chest tube placement describe a worsening respiratory distress<sup>8,11,12</sup>. In cases when the diagnosis is not completely clear on plain radiographs, a CT scan of the chest with intravenous contrast often assists in diagnosing CLE. The most commonly reported CT findings associated with CLE include a hyperlucent and hyperexpanded lobe, intact pulmonary vasculature through the involved segment, atelectasis, consolidation, mediastinal shift towards the contralateral side and midline substernal lobar herniation<sup>3</sup>. CLE is commonly confused with tension pneumothorax, which often results in intercostal chest tube insertion. Other differential diagnoses include pneumatocele, diaphragmatic

hernia, congenital cystic adenomatoid malformation, lung abscess and encysted empyema<sup>13,14</sup>.

In CLE patients, surgical excision of the affected lobe is the treatment of choice in infants under 2 months and in older infants with severe respiratory symptoms<sup>14,15</sup>. The critical phase in the anesthetic management of a patient with CLE is the time of induction and intubation when rapid respiratory and hemodynamic collapse can occur. Induction of anesthesia should aim to avoid crying and struggling, because these can increase the amount of air trapped in the emphysematous lobes during patient induced Valsalva maneuvers (occurring during violent inspiratory efforts)<sup>2</sup>. In addition, spontaneous ventilation is preferred prior to thoracotomy, because the risk of hypoxia and hemodynamic disturbances may be minimized during induction and intubation by keeping the patient spontaneously breathing using 100% oxygen<sup>2,16,17</sup>. In this case inhalation induction was done to maintain the spontaneous breathing and to avoiding the crying during intravenous line placement. Crying would increase the airway pressure and increase the size of emphysematous lobes.

Positive pressure should be avoided or kept to a minimum to prevent further inflation of the emphysematous lobe. Nitrous oxide is contraindicated because of its potential for increasing the size of trapped air in the affected lobe<sup>18</sup>. It is imperative that the surgeon be present and ready to open the chest if sudden cardiovascular compromise occurs (as in this case). A thoracotomy will allow the emphysematous lobe to herniate outside of the chest wall, thus relieving the pressure inside the chest. Once the thoracotomy has been performed, muscle relaxants can be administered and positive pressure ventilation safely started. Hypotension is commonly encountered with CLE patients during surgery when a mediastinal shift compresses the heart and great vessels, severely impairing venous return.

Other techniques to diminish complications can be employed during resection of CLE. Selective endobronchial intubation with ventilation of the healthy lung can be used until lobectomy or pneumonectomy of the affected side is completed, after which the endotracheal tube can be repositioned into the trachea<sup>19</sup>. Another option includes infiltration at the incision site using local anesthetic supplemented by intravenous ketamine with patient breathing spontaneously. This has been used in unstable patients to relieve intra-thoracic pressure, which was then followed by general anesthesia with endotracheal intubation

once the patient was more hemodynamically stable<sup>2</sup>. Raghavendran, et al., used a caudally-placed thoracic epidural catheter before thoracotomy in patients with CLE. This provided preemptive analgesia with a reduction of anesthetic requirement and positive pressure ventilation during the critical phase of induction<sup>16</sup>. Finally, postoperative ventilation may be needed for these patients depending on the extent of the resection.

### CONCLUSION

Induction of anesthesia in a patient with congenital lobar emphysema can be challenging for the anesthesiologist because of the risk of overinflating the lung with either infant-induced airway Valsalva effects or positive airway pressure administered by the anesthesiologist. A variety of techniques are available for reducing potential complications, but as this case shows, a surgeon should be scrubbed and ready to do an emergency life-saving thoracotomy.

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