Anesthesia in an infant with congenital lobar emphysema (CLE)

Author(s): Jesus Apuya, MD and Shailesh Shah, MD
Affiliation(s): Arkansas Children’s Hospital

ABSTRACT BODY: A 2-month old, 4.1-kg baby girl was scheduled for right thoracotomy and lobectomy for congenital lobar emphysema. On preoperative physical exam, she was noted to be awake and tachypneic but not cyanotic. Her vital signs showed a blood pressure of 88/50, pulse rate of 110, respiratory rate of 58 with an oxygen saturation of 95% on 1 liter per minute of oxygen via nasal cannula. There was air entry in both lung fields but was decreased on the right. There were no cardiac murmurs heard and no tracheal deviation noted. She had a right chest tube placed in the emergency room. Her laboratory results were unremarkable. The chest X-RAY showed marked hyperinflation of the right lung extending across the midline with marked left mediastinal shift possibly secondary to suspected congenital lobar emphysema. There was no 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 which were compressed by the emphysematous lung. There was right-to-left mediastinal shift with compression of the left lung. The trachea and left bronchial tree were patent. The patient was brought to the operating room and ASA standard monitors were placed. The chest tube was connected to suction. Her peripheral intravenous access was infiltrated. A mask induction was started using sevoflurane and 100% oxygen while keeping the patient spontaneously breathing. After a peripheral intravenous access was obtained, fentanyl (1 mcg/kg) was administered in preparation for intubation. Peak airway pressure was kept at a minimum. At this point, the patient’s oxygen saturation was 100% with a stable heart rate and blood pressure. The trachea was intubated easily using 3.5 endotracheal tube. However, there were no breath sounds heard over either lung field or gastric region. There was neither chest rise nor end-tidal carbon dioxide noted. The oxygen saturation started to decrease to the low 70’s. The endotracheal tube was removed and mask ventilation resumed. At this time it was noted that it was increasingly difficult to ventilate the patient even with an oral airway in place. No breath sounds were audible and the oxygen saturation remained in the 80’s. However, the systolic blood pressure dropped to the low 60’s and the surgeon was asked to emergently open the right chest. Immediately after part of the emphysematous lobe herniated out of the chest, mask ventilation became easier and oxygen saturation increased to 98-100%. The systolic blood pressure stabilized. The patient was reintubated and surgery started. The remainder of the case was uneventful.
Discussion:
Congenital lobar emphysema (CLE) is a rare anomaly of the lung that can occur in children. It is characterized by hyperinflation of the affected pulmonary lobe with air trapping leading to compression of the surrounding structures. Mediastinal shift and herniation of the emphysematous lobe across the anterior mediastinum (as shown in this case) may occur. In roughly 50% of cases of CLE the etiology is unknown (1). Regardless of the cause, the end result is an overdistended lobe that produces compression atelectasis of the normal lung, mediastinal shift, and impaired venous return with subsequent hypoxemia and hypotension. The upper lobe of the left lung is the most commonly affected followed by the right middle and upper lobes. It can manifest during the first several days of life up to several months of age. Signs and symptoms are non-specific and include dyspnea, cyanosis, tachycardia, wheezing, recurrent infections and failure to thrive. Unless suspected or a chest XRAY is done, a diagnoses of CLE can be easily missed until the patient is severely affected. A chest XRAY 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 very similar to CLE and case reports of chest tube
placement describe a worsening respiratory distress (2, 3). A plain film is also useful in excluding other causes of respiratory distress such as a mediastinal mass or an enlarged heart. In our patient, a CT scan was done based on the result of the chest X-RAY. Congenital heart disease is present in 14% of cases and therefore should always be considered. Induction of anesthesia should be smooth, avoiding crying and struggling, both of which can increase the amount of air getting trapped in the emphysematous lobes during violent inspiratory efforts (4). 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. The surgeon should be present and ready to open the chest if sudden cardiovascular compromise occurs. A thoracotomy will allow the emphysematous lobe to herniate outside of the chest wall thus relieving the pressure inside the chest. Several case reports have been published regarding how the airway is secured in these patients (5, 6). A right mainstem endobronchial intubation can be done if CLE is present in the left lung. However, in our patient the affected lung was the right middle lobe. Endobronchial intubation of the left lung is not easy to achieve. Gupta and others described a successful first attempt left-sided endobronchial intubation in an infant with CLE (6). The head is turned to the right side and the endotracheal tube is rotated 180º. The authors were not able to try this technique because the patient deteriorated rapidly immediately after intubation.

Summary: Although CLE is a rare disease in children, anesthesiologists should be aware of its anesthetic implications to avoid potential life-threatening consequences.

References: