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Pneumonectomy in Congenital Lobar Emphysema: A Case Report

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Abstract

Background Congenital lobar emphysema is a rare congenital disease which caused by anomaly on bronchial cartilage development. Usually, it is limited to a single lobe. It could be treated conservatively or surgically depend on the clinical condition.

Objective In this case report we would like to discuss the management strategy for congenital lobar emphysema that affected the whole lung in 17 days old baby.

Methods The patient was admitted to NICU due to respiratory distress. The patient had a difficulty to wean from ventilator. Radiologic examination revealed the whole left hemithorax is filled by multiple bullae which push mediastinal organ contralaterally. Intraoperatively, we found the whole left lung is emphysematous and there is PDA which is not detected preoperatively. Surgeon decided to do pneumonectomy and PDA ligation. After operation, patient recovered on NICU slowly and discharged from hospital 34 days post operation.

Result our case of congenital lobar emphysema which affect one side of lung is a rare case. Our decision to do pneumonectomy was challenging post operatively.

Conclusion Multidisciplinary team should be involved in treatment of congenital lobar emphysema. Team should be prepared for prolonged intensive care if decision to do lung resection is chosen.

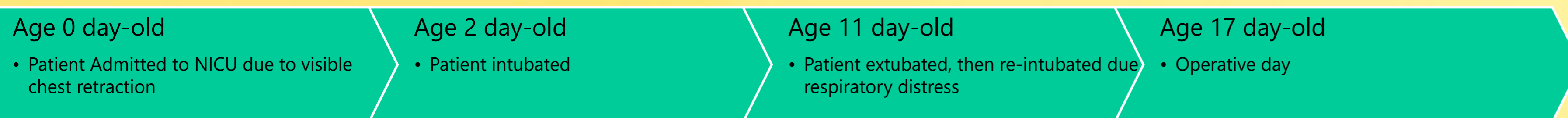
Keywords: Congenital lobar emphysema, pneumonectomy, neonates

Case Presentation

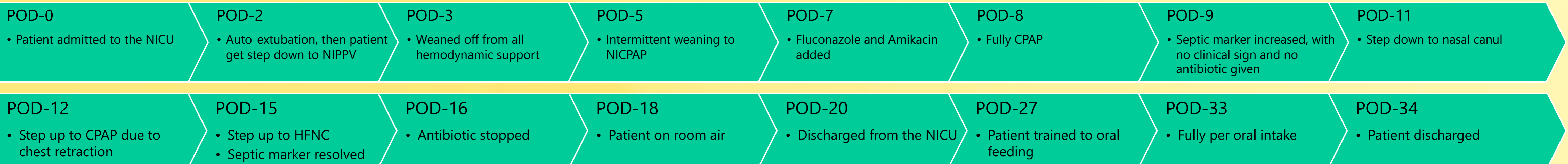
History

- A 12-day-old baby was consulted to our cardiothoracic surgery due to respiratory distress and difficulty with ventilator weaning.
- The baby was born by SC operation due to impending eclampsia at 37 weeks gestation.
- On the second-day patient gets intubated due to respiratory distress. The patient had a history of reintubation due to desaturation and respiratory distress. Since then the patient had difficulty with ventilator weaning..
- On physical examination baby weight 2450 grams, intubated on a ventilator
- The patient had increased septic marker procalcitonin by 2,41 no other laboratory results were remarkable
- **Working Diagnosis** → Congenital Lobar Emphysematous
- **MDT Discussion** → left thoracotomy and upper left lobectomy with the possibility of pneumonectomy

Preoperative Timeline



Postoperative Timeline



Pre-operative Radiological Findings

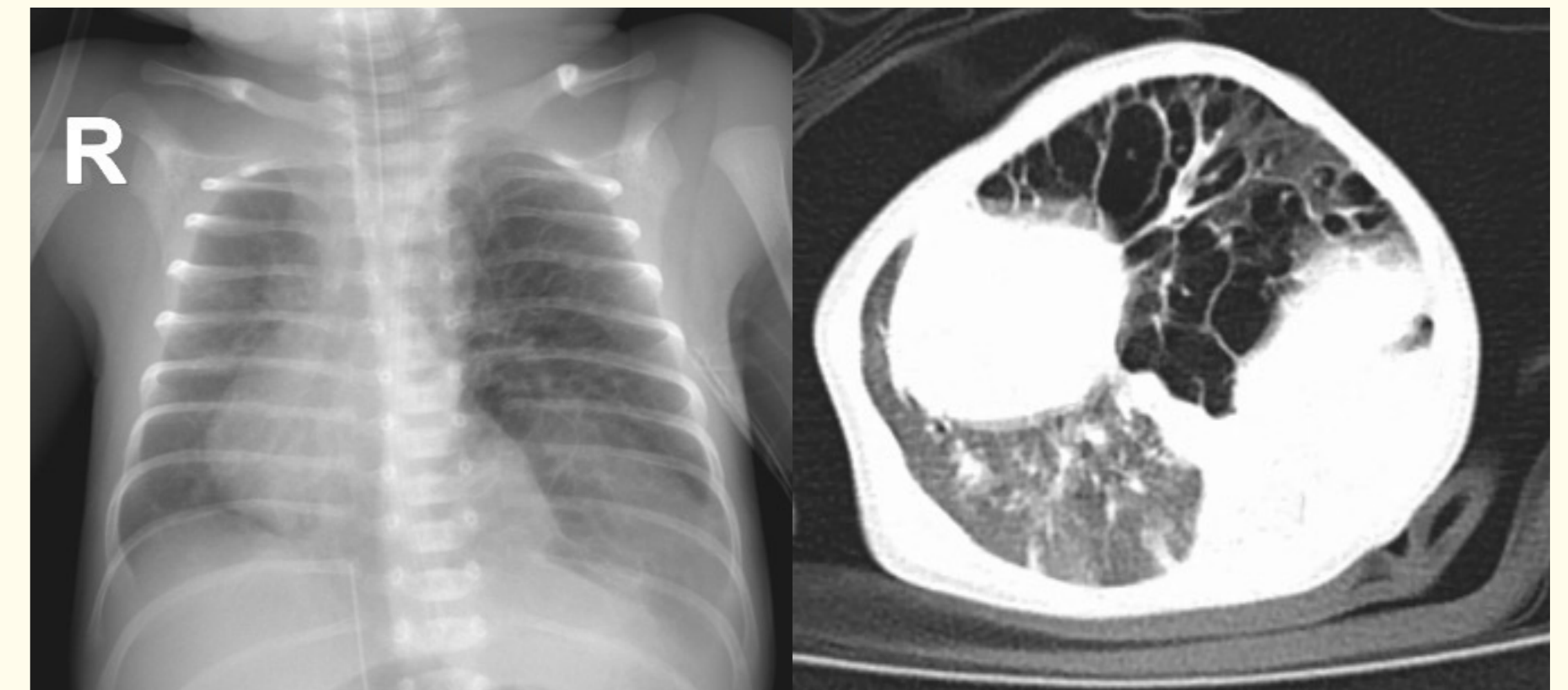


Figure 1 Chest Xray shows hyperaeration on left upper hemithorax which push the mediastinal organ to the right hemithorax (left). CT scan shows multiple bullae on left hemithorax (right).

Post-operative Radiological Findings

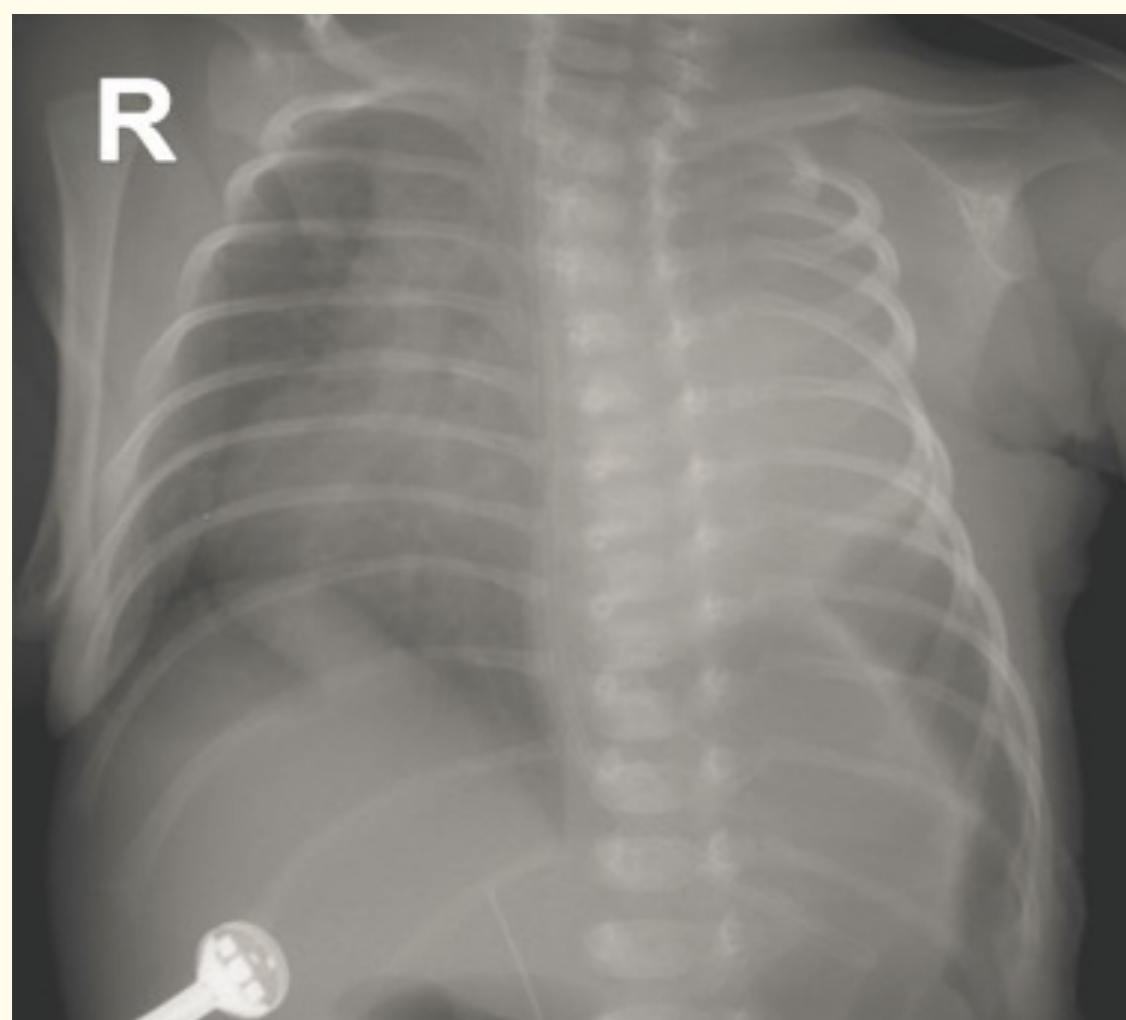


Figure 2 Post operative CXR shows right lung has fully expanded

Operative Techniques

- Patient under general anesthesia and put on right lateral decubitus position.
- Thoracic cavity opened by right posterolateral thoracotomy on 5th intercostal space.
- It was difficult to expose the hilum due to the upper lobe overinflation so the upper lobe was pulled out from thoracic cavity.
- From the inspection of the left lower lobe, the lobe was emphysematous too
- We decided to do pneumonectomy rather than lobectomy We decided to do pneumonectomy rather than lobectomy
- The left pulmonary artery, upper left pulmonary vein, lower left pulmonary vein, and bronchus were identified.
- The left pulmonary artery, upper left pulmonary vein, and lower left pulmonary vein is ligated and then cut.
- The left main bronchus is clamped and cut. While the proximal punctum is sutured.
- After bleeding control, we did a bubble test, which shows no sign of air leak.
- A drain then was placed into the intrathoracic space
- The thoracic cavity is closed, and the chest muscle is reconstructed layer by layer. Skin is sutured
- The surgery is done



Figure 3 Intraoperative finding shows emphysematous left upper lobe. Take attention to the left lower lobe which is still inside thoracic cavity is also emphysematous.

Discussion

CLE is a rare disease with an incidence of ¹ out of 20000-30000 life birth found the most on the left upper lobe (57%), right middle lobe (30%), then right upper lobe (27%).¹ On this report, 25% cases have two lobes involved.² It rarely affects the lower lobe but it can affect both sides of the lungs or whole side of the lung.

Treatment of CLE could be surgically or conservatively. Surgical treatment is usually chosen for patients with a severe cases. In our case, the patient had severe respiratory distress and get intubated. The weaning process was difficult so we decide to operate on the patient.³

On preoperative evaluation, we couldn't sure whether the affected was one lobe or a whole lung. From preoperative plain radiography and CT scan evaluation, we suspect only the left upper lobe was emphysematous and the left lower lobe was atelectasis.

About 12-20% of CLE case is concomitant with congenital cardiac disease.⁴ Cardiac evaluation to detect congenital cardiac disease must be done on all patients with CLE. We had done echocardiography but we missed PDA on our echocardiographic evaluation. This might be caused by the anatomical disturbance which causes the mediastinal organ to shift to the right and air trapped on the affected lung to disturb the echocardiography imaging. Kylat RI recommends evaluation by CT scan or MRI to evaluate cardiac anomaly on CLE patient.⁴

We found that exposing the hilum intraoperatively was difficult in this case since the left upper lobe was overinflated. To overcome this problem we pull the overinflated lobe out. We could expose the hilum better and inspect another lobe this was.

Pneumonectomy is rarely done for this disease since it's rarely found to affect the whole lung. Tempe et al (2010) described their experience in treating congenital lobe emphysema which affects the whole lung and then underwent pneumonectomy on one patient. In their case, the patient recovers faster post-operatively. The patient was only intubated for six hours and discharged from the hospital after eight days post-operative.⁵

In our case, the patient recovers slowly after the operation. It might be caused by an infection that is difficult to handle and laboratory result which shows bad renal function so we can't aggressively administer antibiotic which also affects renal function. But after the surgery, this case was resolved so we could administer antibiotics more aggressively.

Conclusion

- CLE is a rare disease that usually affects only one lobe. Patient with this disease which affects the whole lung is even rarer.
- CLE could be managed surgically or conservatively depending on the clinical condition of the patient.
- All patients with CLE should undergo a cardiac evaluation to detect congenital cardiac anomalies.
- CLE patients whose been undergone pneumonectomy could recover fast if there is no complication which causes patients difficult to wean from respiratory support.

Citation

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