Journal of Neonatal Surgery
ISSN(Online): 2226-0439
Vol. 14, Issue 21s (2025)
https://www.jneonatalsurg.com

OPEN ACCESS

Emergency Rescue Lobectomy in A Two-Month-Old with Congenital Pulmonary Airway Malformation

Dr Sonal Khatavkar[1], Dr Brinda Badam*[2]

[1]Professor, Dr D Y Patil Vidyapeeth, Dr D Y Patil medical college hospital and research centre

[2*]Resident, Dr D Y Patil Vidyapeeth, Dr D Y Patil medical college hospital and research centre

Email ID: drbrindabadm@gmail.com

***Corresponding Author**

Dr Brinda Badam,

Resident, Dr D Y Patil Vidyapeeth, Dr D Y Patil medical college hospital and research centre

Email ID: drbrindabadm@gmail.com

Cite this paper as: Dr Sonal Khatavkar, Dr Brinda Badam, (2025). Emergency Rescue Lobectomy in A Two-Month-Old with Congenital Pulmonary Airway Malformation. *Journal of Neonatal Surgery*, 14 (21s), 855-859.

ABSTRACT

Congenital Pulmonary Airway Malformation (CPAM) is a rare developmental anomaly of the lung that can cause severe respiratory distress in infants. Type I CPAM, characterized by large cystic lesions, may result in life-threatening complications such as pneumothorax and mediastinal shift. We report the case of a two-month-old male infant presenting with respiratory distress due to Type I CPAM involving the right upper and middle lobes. Imaging revealed large cystic lesions with a right-sided pneumothorax and mediastinal shift. During admission the infant developed bradycardia and desaturation, requiring cardiopulmonary resuscitation and intubation, after stabilizing further evaluation and investigation patient was posted for emergency thoracotomy and lobectomy , which were successfully performed. Postoperatively, the infant was managed in the Paediatric Intensive Care Unit with ventilatory support. Early recognition and urgent surgical intervention are critical in symptomatic CPAM cases. Multidisciplinary management is essential for optimizing outcomes in critically ill infants.

Keyword: *Congenital Pulmonary Airway Malformation (CPAM), Neonatal Respiratory Distress, Emergency Lobectomy, Paediatric Thoracic Surgery, Mediastinal Shift.*.

1. INTRODUCTION

Congenital Pulmonary Airway Malformation (CPAM), previously known as Congenital Cystic Adenomatoid Malformation (CCAM), is a rare developmental anomaly of the lung arising from abnormal airway branching during fetal life. It is characterized by cystic or solid mass lesions that can disrupt normal lung function [1]. CPAM has an estimated incidence of approximately 1 in 25,000 to 1 in 35,000 live births, with Type I being the most common form, typically presenting with large, single, or multiple cysts lined by respiratory epithelium. Clinical presentation of CPAM varies significantly. While some cases remain asymptomatic and are detected incidentally, others present early with symptoms such as respiratory distress, pneumothorax, recurrent infections, or mediastinal shift. Symptomatic neonates and infants often require urgent intervention to relieve respiratory compromise [2]. Large cystic lesions, particularly those causing compression of adjacent lung tissue or mediastinal structures, can rapidly deteriorate and lead to life-threatening complications. Early diagnosis is crucial and often involves a combination of prenatal imaging, chest radiography, and computed tomography (CT) postnatally. Management strategies depend on the size, location, and clinical impact of the lesion. Elective surgical resection is recommended for symptomatic cases or to prevent complications in asymptomatic patients [3]. Emergency surgery is indicated when significant respiratory distress, tension pneumothorax, or hemodynamic instability occurs.

We report a rare case of a two-month-old infant who presented with severe respiratory distress due to Type I CPAM complicated by right-sided pneumothorax and mediastinal shift, requiring emergency thoracotomy and lobectomy. This case highlights the importance of timely surgical intervention and multidisciplinary management in critically ill neonates

CASE PRESENTATION

A two-month-old male infant, weighing 3.1 kilograms, presented to the emergency department with complaints of progressive respiratory distress, poor feeding, and increasing tachypnoea over several days. The infant was born at term via an uneventful vaginal delivery, with no significant prenatal findings noted. However, postnatal respiratory symptoms gradually worsened, prompting hospital admission. On examination, the infant was tachypnoeic with subcostal retractions and diminished breath sounds on the right side. Pulse oximetry revealed an SpO_2 of 96% on supplemental oxygen via nasal cannula. His pulse rate was 120 beats per minute, with intermittent episodes of bradycardia recorded during monitoring. Other systemic examinations were within normal limits. Given the clinical signs, an urgent chest X-ray followed by a high-resolution CT scan of the thorax was performed. Imaging revealed multiple large cystic lesions measuring 6.3 × 6.5 × 5.3 cm occupying the right upper and middle lobes. Hyperlucent lung fields, right-sided pneumothorax, and significant mediastinal shift to the left were evident. The findings were consistent with a diagnosis of Type I Congenital Pulmonary Airway Malformation (CPAM).

The patient was started on broad-spectrum intravenous antibiotics (piperacillin-tazobactam and meropenem) due to the risk of secondary infection. Inotropic support with an adrenaline infusion was initiated to manage hemodynamic instability. Antiepileptic prophylaxis with levetiracetam was started in view of his bradycardic episodes. Analgesia was maintained with fentanyl infusion. During admission, the patient experienced acute desaturation followed by bradycardia. Immediate cardiopulmonary resuscitation (CPR) was initiated. The infant was successfully intubated with a 3.5 mm uncuffed endotracheal tube fixed at 9 cm from the lips. After successful resuscitation and stabilization. In view of worsening respiratory compromise and radiological evidence of tension pneumothorax with mediastinal shift, the surgical team decided to proceed with an emergency thoracotomy and lobectomy.

The infant was stabilized preoperatively on medications including piperacillin-tazobactam, meropenem, levetiracetam, fentanyl, and an adrenaline infusion. Anaesthesia was maintained using sevoflurane in a 50:50 mixture of oxygen and air, along with fentanyl, propofol, Controlled ventilation was provided via a Jackson-Rees circuit. Emergency right thoracotomy and lobectomy were performed between 2:30 PM and 4:10 PM. The estimated blood loss was 20 ml, with an equivalent urine output. Intraoperative arterial blood gas analysis showed a pH of 7.38, pCO_2 of 56 mmHg, pO_2 of 188 mmHg, HCO_3^- of 30.1 mmol/L, lactate of 0.7 mmol/L, and hemoglobin of 9.0 g/dL. Electrolyte imbalances were noted, with a potassium level of 2.6 mmol/L and calcium at 1.08 mmol/L. The infant remained intubated and was transferred to the Pediatric Intensive Care Unit (PICU) for continued management. Adrenaline infusion was maintained, and supportive care included gradual weaning of ventilatory and inotropic support. Postoperative imaging demonstrated re-expansion of the remaining lung and resolution of the mediastinal shift.

Surgical and Anaesthetic Management

Anaesthetic management was initiated with total intravenous induction using fentanyl and propofol ,Sevoflurane in a 50:50 mixture of oxygen and air, combined with regional anaesthesia with a 19 gauge caudal epidural catheter fixed at 15 cm a dose of 0.7 – 1ml of 0.2% Inj Ropivicaine was given to allow the surgeons proceed with the incision , while maintaining the patient on spontaneous ventilation. This approach was chosen to prevent worsening of the existing pneumothorax due to the potential risk of tension pneumothorax from intermittent positive pressure ventilation (IPPV). Following the thoracotomy incision, once the pneumothorax was relieved and the chest was open, muscle relaxation was achieved using atracurium to facilitate controlled ventilation. Anaesthesia was then maintained with a combination of Sevoflurane in a 50:50 mixture of oxygen and air, along with continued infusions of fentanyl and propofol. Controlled ventilation was provided via a Jackson-Rees circuit. Continuous intraoperative monitoring included electrocardiography (ECG), pulse oximetry (SpO_2), end-tidal carbon dioxide ($ETCO_2$), and core temperature. The surgical team proceeded with a right thoracotomy, successfully performing a lobectomy of the right upper and middle lobes. Intraoperatively, hemodynamic fluctuations were observed and managed with fluid boluses and maintenance fluids, including Ringer's Lactate with 5% dextrose. The estimated intraoperative blood loss was 20 ml, with an equivalent urine output of 20 ml.

An intraoperative arterial blood gas (ABG) analysis performed at 4:00 PM showed: pH 7.38, pCO_2 56 mmHg, pO_2 188 mmHg, bicarbonate (HCO_3^-) 30.1 mmol/L, lactate 0.7 mmol/L, hemoglobin (Hb) 9.0 g/dL, sodium 148 mmol/L, potassium 2.6 mmol/L, and calcium 1.08 mmol/L. Notably, the hypokalemia and hypocalcemia were corrected during the procedure. The surgical incision was closed without complication. Given the patient's critical intraoperative course and preoperative instability, the decision was made to continue mechanical ventilation postoperatively. The infant remained intubated and was transferred to the Paediatric Intensive Care Unit (PICU) for ongoing ventilatory and inotropic support.

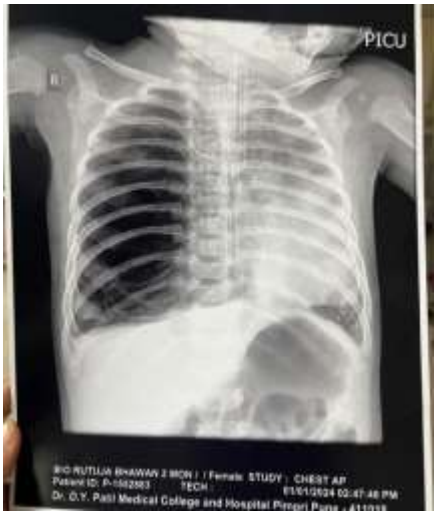

FIGURE 1: Chest X ray (AP view) demonstrating a right-sided pneumothorax and mediastinal shift .

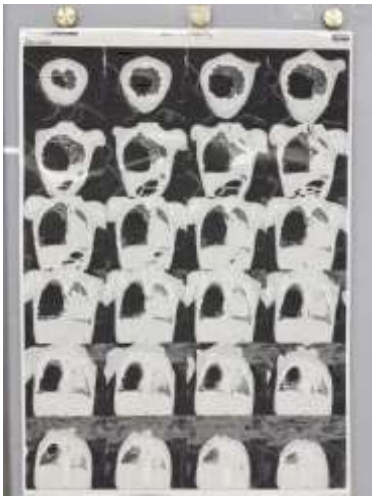

FIGURE 2: Coronal CT scan showing a large heterogenous cystic lesions occupying the right lung with right-sided pneumothorax and mediastinal shift.

FIGURE 3: Intraoperative view showing a large lesion in the thoracic cavity prior to lobectomy.

FIGURE 4: Intraoperative image of dissection of the affected lung lobe.

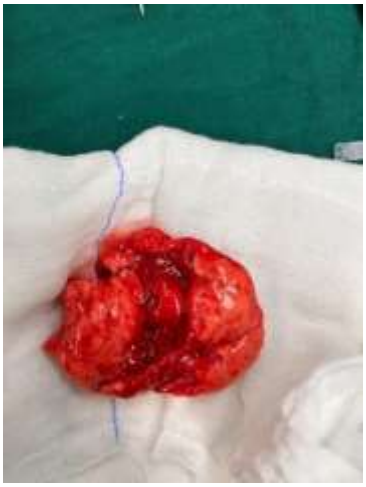

FIGURE 5 Gross specimen of the resected lung lobe.

FIGURE 6. Surgical field after right lobectomy, exposing the bronchial stump and surrounding structures.

Postoperative Course

In the PICU, the infant was maintained on mechanical ventilation with careful oxygen supplementation. The adrenaline infusion was continued to support cardiovascular stability. Analgesia and sedation were provided with a combination of fentanyl and midazolam infusions. Electrolyte imbalances were closely monitored and corrected as needed. A chest drain was placed intraoperatively and monitored for air leak and drainage [4]. Serial chest radiographs showed re-expansion of the remaining lung and resolution of the mediastinal shift. Gradual weaning from ventilatory and inotropic support was initiated over the following days, depending on the patient's respiratory and hemodynamic status[5] and was extubated on postoperative day 4 . The patient remained under close observation by a multidisciplinary team comprising paediatric surgeons, intensivists, anaesthesiologists, and neonatologists, with plans for gradual extubating once respiratory parameters stabilized.

2. DISCUSSION

Congenital Pulmonary Airway Malformation (CPAM) represents a rare developmental anomaly of the lower respiratory tract, characterized by abnormal cystic proliferation within the lung parenchyma. Type I CPAM, the most common variant, typically presents with large cysts and carries a relatively favourable prognosis when surgically managed [6]. In neonates and young infants, symptomatic CPAM may lead to life-threatening respiratory distress, infection, and compression of adjacent thoracic structures. In the present case, the two-month-old infant developed acute respiratory compromise secondary to CPAM-associated pneumothorax and mediastinal shift, necessitating emergency surgical intervention. Emergency thoracotomy and lobectomy in infants pose significant aesthetic and surgical challenges [7]. Preoperatively, our patient exhibited instability with desaturation episodes and bradycardia, indicating critical cardiorespiratory compromise requiring immediate cardiopulmonary resuscitation and intubation. Such events highlight the precarious balance of oxygenation and ventilation in infants with large thoracic cystic lesions, where positive pressure ventilation may worsen pulmonary dynamics and exacerbate mediastinal shift [8]. Intraoperatively, careful hemodynamic monitoring and management were crucial.

A pneumothorax is a medical condition characterized by the presence of air between the parietal and visceral pleura. Even a small, uncomplicated pneumothorax can quickly enlarge and become more severe during general anaesthesia, as positive pressure mechanical ventilation may exacerbate the condition and potentially result in a tension pneumothorax.[9] Therefore our anaesthesia plan was initiated with a combination of total intravenous induction using fentanyl and propofol, combined with regional anaesthesia with a caudal epidural catheter to allow the surgeons proceed with the incision , while maintaining the patient on spontaneous ventilation, so as to prevent worsening of the existing pneumothorax due to the potential risk of tension pneumothorax from intermittent positive pressure ventilation (IPPV). Following the thoracotomy incision, once the pneumothorax was relieved and the chest was open, our plan was converted to controlled ventilation via a Jackson-Rees circuit to minimize airway pressures [10]. Fluid management was optimized with isotonic solutions and glucose supplementation, crucial for maintaining perfusion without precipitating pulmonary edema. Arterial blood gas analysis indicated adequate gas exchange and tissue perfusion despite minor hypercapnia, reflecting the delicate ventilatory management required.

Postoperative care focused on continued respiratory and cardiovascular support. The decision to maintain intubation and

adrenaline infusion in the PICU ensured a controlled transition to spontaneous ventilation while minimizing the risk of respiratory decompensation [11]. Early postoperative imaging confirmed successful re-expansion of the remaining lung, an important indicator of surgical success and recovery trajectory. This case underscores the importance of early diagnosis, multidisciplinary planning, and prompt surgical intervention in symptomatic CPAM. Preoperative stabilization, careful anaesthetic induction, rapid response to intraoperative events, and meticulous postoperative care are essential elements that influence outcomes. Although elective surgical resection is often preferred in asymptomatic patients, emergency lobectomy remains lifesaving in critically ill infants [12]. The favourable outcome in this case illustrates that even in unstable patients, with vigilant perioperative management, emergency surgical correction can achieve good results. Future advancements in prenatal diagnosis, neonatal intensive care, and minimally invasive surgical techniques may further improve the prognosis for infants with CPAM requiring early intervention [13].

3. CONCLUSION

This case highlights the critical role of early recognition and prompt intervention in managing congenital pulmonary airway malformation (CPAM) in neonates. Type I CPAM, while generally presenting with favourable outcomes, can lead to severe respiratory distress and life-threatening complications, particularly when complicated by pneumothorax and mediastinal shift, as demonstrated in this infant. The timely decision to perform an emergency thoracotomy and lobectomy in this case was essential to restoring respiratory function and stabilizing the patient. The case also underscores the complexity of managing such patients in the perioperative period, particularly in terms of anaesthetic considerations, ventilation strategies, and fluid management. The risk of intraoperative desaturation and bradycardia required swift action and highlighted the need for a skilled multidisciplinary team, including paediatric anaesthesiologists, surgeons, and intensivists, to ensure optimal care. Ultimately, with careful preoperative stabilization, effective intraoperative monitoring, and diligent postoperative care, patients with CPAM can recover successfully, even in critical conditions. This case serves as a reminder of the importance of thorough planning and vigilant monitoring in achieving favourable outcomes in paediatric thoracic surgery, especially for rare and complex conditions like CPAM.

REFERENCES

[1] M. L. Stocker, "Congenital cystic lesions of the lung: Congenital pulmonary airway malformation (CPAM)," Seminars in Pediatric Surgery, vol. 16, no. 4, pp. 231-237, Nov. 2007.

[2] J. E. Darragh, L. H. Friedberg, and M. M. Sussman, "Pneumothorax in a neonate with congenital pulmonary airway malformation," Pediatric Pulmonology, vol. 41, no. 7, pp. 631-634, Jul. 2006.

[3] R. S. Ladd, J. P. Aprahamian, and S. J. Macgregor, "Congenital pulmonary airway malformation: Diagnosis and management," The Journal of Thoracic and Cardiovascular Surgery, vol. 152, no. 5, pp. 1184-1190, Nov. 2016.

[4] M. A. Chaudhary and R. T. Deterding, "Congenital lung malformations: Diagnosis and management," The Journal of Pediatrics, vol. 150, no. 4, pp. 352-358, Apr. 2007.

[5] T. M. Papageorgiou, G. M. N. Parikh, and M. A. V. Bhagat, "Emergency management of a newborn with congenital pulmonary airway malformation," Pediatric Surgery International, vol. 32, no. 2, pp. 217-219, Feb. 2016.

[6] J. D. L. Weitzman and S. M. Meyer, Pediatric Anesthesia and the Management of Respiratory Compromise, 2nd ed. New York, NY: McGraw-Hill, 2014.

[7] S. H. Lee, W. R. Eason, and M. C. Beauchamp, "Anesthetic considerations for pediatric thoracic surgery," Anesthesia & Analgesia, vol. 120, no. 6, pp. 1269-1275, Jun. 2015.

[8] R. S. Gupta, K. S. Taylor, and J. R. Mohr, "Critical care management of pediatric patients after lobectomy," Pediatric Critical Care Medicine, vol. 16, no. 3, pp. 237-245, Mar. 2015.

[9] Bacon AK, Paix AD, Williamson JA, Webb RK, Chapman MJ. Crisis management during anaesthesia: pneumothorax. Qual Saf Health Care. 2005;14:e18.

[10] M. W. S. Clark, "Thoracic surgery in neonates: Case study review," Journal of Pediatric Surgery, vol. 45, no. 6, pp. 1341-1345, Jun. 2010.

[11] T. T. Chan, D. P. Shen, and P. M. Hull, "Preoperative stabilization of neonates with congenital lung malformations," Journal of Neonatal Surgery, vol. 2, no. 2, pp. 55-61, Dec. 2013.

[12] A. G. Shapiro, M. L. Hoffman, and J. K. Brezinski, "Management of congenital pulmonary airway malformation in neonates and infants," Neonatal Intensive Care, vol. 21, no. 3, pp. 123-129, Mar. 2018.

[13] S. B. Patel, E. T. Johnson, and N. L. Gregory, "Surgical resection of CPAM and its complications in neonates: A retrospective study," Pediatric Surgery International, vol. 34, no. 6, pp. 745-750, Jun. 2018.

..