

Anaesthetic Management of Adult Unilateral Pulmonary Hypoplasia with Post-tuberculosis Sequelae and Cervical Vertebral Anomalies: A Case Report

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ABSTRACT

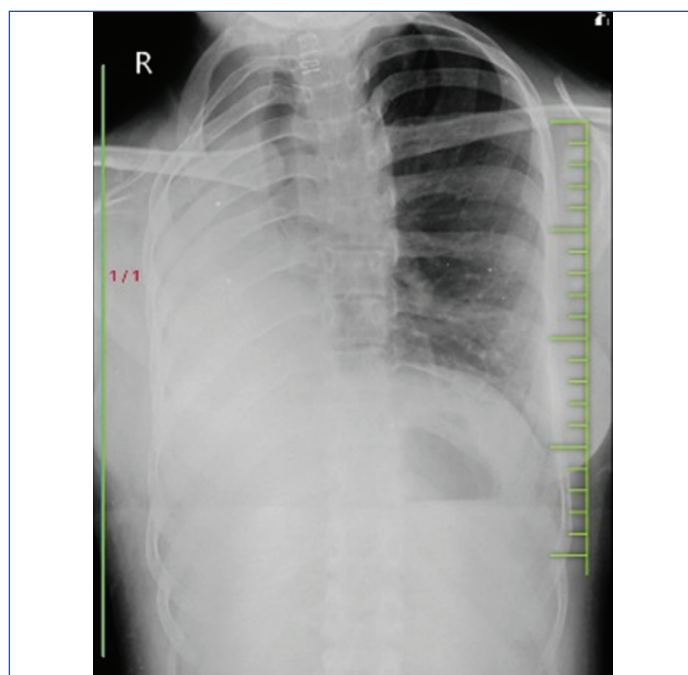
Pulmonary hypoplasia is a rare congenital anomaly associated with underdeveloped lungs with incomplete lung volume. It can be unilateral or bilateral and often occurs in people with other anomalies. Contributing factors include oligohydramnios, congenital diaphragmatic hernia, and genetic syndromes. In addition, sequelae from Pulmonary Tuberculosis (PTB) may further exacerbate the cases of pulmonary hypoplasia, causing fibrosis and lung damage. The present case report, presents an account of a 20-year-old female with congenital pulmonary hypoplasia with exertional dyspnoea and upper limb paresthesia. On imaging, she had hypoplastic right lung and cervical rib anomalies with vertebral segmentation defects. A multidisciplinary team assessment of her condition was done, and she underwent general anaesthesia for surgical intervention. Preoperative pulmonary function tests were Forced Vital Capacity (FVC) and Forced Expiratory Volume in 1 second (FEV1) at 60% and 58% of predicted values, and thus, bronchodilator therapy with nebulised albuterol was initiated. Rapid sequence induction was done using propofol, followed by rocuronium. Maintenance was done using sevoflurane, oxygen, and nitrous oxide. She was then transferred to the Surgical Intensive Care Unit (SICU) for close monitoring. A multimodal analgesia strategy with fentanyl and ketorolac was initiated, along with careful fluid management to avoid overload. Tailored pulmonary rehabilitation and optimised nutritional support were given for her recovery. The present case underscores the difficulties of managing pulmonary hypoplasia in adulthood and the benefits one could reap from a multidisciplinary approach.

Keywords: Airway management, Cervical rib anomaly, Exertional breathlessness, Mechanical ventilation, Pulmonary tuberculosis, Vertebral segmentation defect

CASE REPORT

A 20-year-old, American Society of Anaesthesiologists (ASA)-III female weighing 60 kg with a known case of pulmonary hypoplasia for the past four years came with complaints of severe breathlessness for a day. She had been experiencing breathlessness on exertion which was modified Medical Research Council (mMRC) dyspnoea scale grade 3 for the past three months and a tingling sensation in both upper limbs for one month. She was posted for cervical fusion at C-7. The patient had a history of PTB four years back, for which she had Anti-Koch's Treatment (AKT) and the treatment lasted for six months. At present, there was no history of weight loss, anorexia, or substance abuse. Her chest X-ray revealed that the right hemi thorax was completely opacified with a shift of the trachea and mediastinum to the right, with an impression that there was a hypoplastic right lung as seen in [Table/Fig-1]. The patient was oriented to time, place, and person. Her temperature was 100.8°F, pulse was 88 beats/minute, respiration was 18 breaths/minute, blood pressure was 110/80 mmHg, and oxygen saturation was 96%. Blood investigations are seen in [Table/Fig-2].

An Electrocardiogram (ECG) showed evidence of sinus tachycardia. A Computed Tomography (CT) scan of the cervical spine revealed multiple anomalies, including fusion of several cervical vertebrae indicating a segmentation anomaly, the presence of cervical ribs on both sides (incomplete on the right and fused with the first rib on the left), and platybasia with a skull base angle of 143 degrees [Table/Fig-3]. A bifid spinous process of one of the lower cervical vertebrae and complete atlantooccipital assimilation were also noted. The lung assessment on the CT scan confirmed a mediastinal shift to the right-side, along with a fibro calcific nodule in the apicoposterior segment of the left upper lobe and mosaic attenuation in the



[Table/Fig-1]: Chest X-ray showing opacified right hemi thorax.

visualised left lung field. After thorough counselling of the patient and her relatives, consent for surgery was obtained. The procedure was planned to be performed under general anaesthesia.

Anaesthetic Management

The patient required careful planning and a multidisciplinary approach because of her complex medical history and anatomical

Name of test	Patient value	Reference value
Haemoglobin	12.2	11-15 g/dL
Packed cell volume	24.9	32-46 %
Red blood cell	3.79	3.8-5.8 million/mm ³
Mean corpuscular volume	89	76-96 fl
Mean cell haemoglobin	29.0	27-32 pg
Mean corpuscular haemoglobin concentration	32.5	31-35 g/dL
Platelet count	2.60	1.5-4 lakhs/mm ³
Red cell distribution width	18.2	11.5-15 %
Total leukocyte count	5700	4000-11000/mm ³
Differential leukocyte count		
Neutrophils	64	40-75 %
Lymphocytes	30	20-45 %
Eosinophils	03	1-6 %
Monocytes	03	2-10 %
Basophils	00	0-1 %

[Table/Fig-2]: Laboratory investigation of the patient.



[Table/Fig-3]: CT-cervical spine revealed multiple anomalies, including fusion of several cervical vertebrae indicating a segmentation anomaly.

anomalies. Thus, a thorough pre-operative assessment was crucial, and it started with a comprehensive review of her medical history. Pulmonary function tests were done to evaluate her respiratory status appropriately. The FVC was recorded at 60% of predicted values, and the FEV1 at 58% of predicted, showing a restrictive lung disease pattern. There were minimal breath sounds on the right-side, consistent with the known hypoplastic lung. To optimise the patient's respiratory function before surgery, bronchodilator therapy was initiated with albuterol 2.5 mg every four hours to alleviate bronchospasm and to improve her lung function for the best possible baseline pulmonary status before anaesthesia. On the day of the surgery, standard monitoring was done for electrocardiography, non-invasive blood pressure and pulse oximetry. Premedication was done with midazolam at a dose of 2 mg to reduce the patient's anxiety. Glycopyrrolate was administered at 0.2 mg to minimise the secretions, preventing any potential aspiration during induction. Fentanyl at a dose of 50 mcg was administered for analgesia and to achieve haemodynamic stability during induction. Rapid sequence induction was done to reduce the risk of aspiration. Propofol was administered at 100 mg, followed by rocuronium at 60 mg (1 mg/

kg) for quick intubation with a good neuromuscular blockade. Due to the anomalies of the cervical ribs, a video laryngoscope and fibre-optic bronchoscope were kept on standby. She was intubated with a 7.5 mm cuffed endotracheal tube following direct laryngoscopy. The position of the tube was confirmed by 5-point auscultation and capnography. The patient was put on volume control mode with the following settings- TV- 300 mL, RR-18/min, FiO₂-50%, I:E-1:2. Lung isolation was not used. Using ultrasound guidance, an arterial line was established in the left radial artery with a 20-G catheter.

Anaesthesia was maintained with sevoflurane, oxygen and nitrous oxide. Following this, the surgery was initiated. Fentanyl boluses were administered for analgesia and were adjusted according to her reaction to pain or surgical stimulation. Constant changes to tidal volumes and respiratory rates were made to match ventilation strategies to her reduced lung volume. Regular Arterial Blood Gas (ABG) assessments were done to track the earliest signs of respiratory acidosis or hypoxemia. The duration of surgery and anaesthesia were six hours, respectively. Following the surgery, the patient was extubated. She was transferred to the SICU for close follow-up.

In the SICU, a multimodal approach for analgesia was used with the target of reducing postoperative pain with opioids in combination with Nonsteroidal Anti-Inflammatory Drugs (NSAIDs), to minimise the adverse effects of respiratory depression. Fentanyl was titrated to a maintained dose of 25 mcg/h for analgesia along with ketorolac 15 mg every six hours for additional pain relief. Fluid management was carefully managed with Lactated Ringer's solution to ensure hydration without risking fluid overload. A precise schedule of follow-up with careful daily assessment by the surgical team for complications like infection, respiratory distress, or fluid overload was done. The pulmonology team assessed lung function and adjusted respiratory support as required.

Further measures to enhance recovery included a customised pulmonary rehabilitation program. The program consisted of chest physiotherapy techniques to facilitate the mobilisation of secretions and lung expansion. She was encouraged to use incentive spirometry regularly with set goals for maintaining optimal lung function and preventing atelectasis. Nutrition was the other critical element in her postoperative care. Given that she had a history of tuberculosis and would be undergoing surgery, a dietitian was consulted to prepare a high-protein, high-calorie nutrition plan, which was supposed to support healing and provide much-needed energy during the recovery period. Her nutritional status was constantly monitored, and dietary modifications were undertaken as required to ensure an excellent outcome at discharge. The patient was found normal and healthy, with no complications on her follow-up visit at three months.

DISCUSSION

Pulmonary hypoplasia, a rare developmental anomaly characterised by incomplete development of lung tissue, presents itself as a challenge in the management of the patient clinically, especially when complicated with other congenital abnormalities and sequelae of pulmonary diseases such as Tuberculosis (TB) [1]. Pulmonary hypoplasia develops due to impaired growth of the lungs, resulting in a decreased volume of the lungs and a risk for respiratory impairment. Usually, these anomalies develop alongside other congenital anomalies, as was the case of this patient, who suffered from cervical rib anomalies and defects of vertebral segmentation. Such anatomical abnormalities worsen the symptoms concerning the respiratory system and create problems with the management strategy [1,2]. The patient had a history of PTB that was treated with AKT. Post-tuberculosis sequelae in the form of fibrosis and residual lung damage must have played a role in her symptomatology even after completion of AKT. This is yet another scenario that addresses the issue of long-term complications TB can cause in pulmonary

function and the need for extended follow-up in such individuals [3,4]. Anaesthetic management of such a patient with unilateral pulmonary hypoplasia complicated by post-tuberculosis sequelae and multiple skeletal abnormalities is exceptionally challenging because of severely compromised pulmonary reserve, changed respiratory mechanics, and possible airway challenges. Restrictive lung pattern on pulmonary function tests combined with minimal breath sounds on the hypoplastic side warranted scrupulous preoperative optimisation with bronchodilators to secure the optimal possible baseline status [1,4].

The level of lung hypoplasia, as well as other skeletal abnormalities, was found with the aid of thoracic imaging, chest X-rays, and follow-up CT scans. The findings of mediastinal shift, fibrocalcific nodules, and mosaic attenuation on imaging contributed to the definition of structural lung abnormalities that have compromised lung function and ventilation-perfusion dynamics [5]. Expected airway challenge due to cervical skeletal abnormalities necessitated readiness with sophisticated airway equipment, such as a video laryngoscope and fibre-optic bronchoscope. During operation, the main anaesthetic challenge was to ensure proper oxygenation and ventilation without causing barotrauma to the solitary functional lung, accomplished by individualising tidal volumes and respiratory frequencies based on serial arterial blood gas analysis [6,7]. Management involved coordinating pulmonologists, thoracic surgeons, anaesthesiologists, and radiologists for appropriate diagnosis and perioperative planning in a delicate manner and, subsequently, long-term management. Proper and thorough preoperative evaluation of the patients with optimisation of their respiratory status was fundamental to ensuring that the surgical procedure was performed without complications. The lung disease pattern was restrictive in pulmonary function tests, but optimal perioperative management was feasible using bronchodilators and close monitoring during anaesthesia. Haemodynamic stability was maintained by judicious use of opioids and titrated anaesthetic depth. Postoperatively, the emphasis shifted to opioid-induced respiratory depression, which was minimised using a multimodal analgesic regimen, instituted aggressive pulmonary rehabilitation measures including incentive spirometry and chest physiotherapy, besides optimal nutrition for enhancing recovery [4,8]. Postoperative care, including pain management, pulmonary rehabilitation, and nutrition support, ensured the patient's optimal recovery. She was monitored continuously for complications such as respiratory distress or infection [9].

In the case reported by Subramanian S and Bapat M, anaesthetic care of a 1-year 10-month-old patient with unilateral lung hypoplasia for ophthalmic surgery demonstrated the significance of adequate preoperative imaging to recognise airway compression and direct ventilation planning. The point to note is that detailed intraoperative monitoring, diligent control of ventilation settings, and immediate response to desaturation or hypotension can make the perioperative period uneventful in such patients [10].

In the case reported by Selvaraj S et al., effective anaesthesia management of an adult patient with congenital right lung agenesis

and undergoing neurosurgery in an MRI suite highlighted the need for individualised ventilation strategies to safeguard the solitary functional lung. Lung-protective ventilation with reduced tidal volumes, optimised airway pressures, and judicious titration of Fraction of Inspired Oxygen (FiO₂) reduce the risk of barotrauma and hypoxemia. The main message is that, with careful preoperative planning, intraoperative caution, and a multidisciplinary team, even high-risk patients with severe structural lung abnormalities can be treated with complex procedures safely in complicated settings like faraway places [11].

CONCLUSION(S)

The present case report highlights the difficulties involved in managing pulmonary hypoplasia in an adult in the context of other congenital anomalies and the long-term effects of tuberculosis. Such exertional breathlessness and respiratory symptomatology of the patient could be more directly correlated with structural abnormalities in the lungs, as well as the sequelae of previous TB infection. The betterment of her respiratory function and the quality of her life was thus managed through this multidisciplinary approach, including detailed diagnostic imaging, surgical intervention, and careful perioperative management. The present case underscores the need for accurate diagnosis, comprehensive assessment, and personalised treatment strategy to maximise outcomes in patients with complex congenital anomalies and chronic pulmonary conditions. Long-term follow-up and monitoring are essential for guiding subsequent management, establishing an early warning system for potential complications, and maintaining improvement in respiratory functions.

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