

ANAESTHETIC MANAGEMENT OF A CHILD WITH ONE LUNG APLASIA UNDERGOING PERINEAL ANORECTOPLASTY

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PRESENTATION OF CASE

An eighteen month old female child, known case of aplasia of right lung, dextroversion and malrotated kidney was posted for stage 2 operation (perineal anorectoplasty). No history of fever, cough and no other complaints. Accepting feeds well.

DIFFERENTIAL DIAGNOSIS

Total lung collapse, atelectasis, consolidation, haemothorax, pneumothorax.

CLINICAL DIAGNOSIS

We are reporting an extremely rare case where an eighteen month old baby with aplasia of one lung, dextroversion and malrotation of right kidney underwent perineal anorectoplasty, ie stage 2 (pull through) operation of the three stage surgical procedure, the first stage being colostomy and the final stage, closure of colostomy. Patient is a case of anovestibular fistula with aplasia of right lung, dextroversion and malrotated right kidney. There is no history of fever, cough or other complaints. Child is accepting feeds well.

Stage 1 operation (colostomy) was done eight months ago. Child was born through normal vaginal delivery with birth weight of 2.75 kilograms and immunized as per schedule. There is history of delayed mile stones. Not a consanguineous marriage. On examination child is conscious, afebrile and malnourished, ill built, looks pale and weighs 6.5 kgs.

Vesicular breath sounds heard on left side, apex beat heard on right side of chest. Per abdomen soft, no organomegaly.

PATHOLOGICAL DISCUSSION

A thorough search of google did not yield any similar case of a patient with aplastic lung undergoing reconstructive surgery.

Pulmonary agenesis, aplasia and hypoplasia are few congenital abnormalities of the lung which are rare. Pulmonary agenesis is the complete absence of the lung parenchyma, its vasculature, and its bronchus.¹ Pulmonary aplasia, the most common variant, consists of a carina and the main-stem bronchial stump with absence of the distal lung.² Presentation is with usual respiratory symptoms like noisy breathing, fast breathing, repeated respiratory tract infections and respiratory distress.² prognosis depends on two factors. Firstly, the severity of associated congenital anomalies and secondly involvement of the normal lung in any disease process. If patient survives the first five years without major infection, an almost normal life span can be expected.³

Pulmonary agenesis is exceedingly rare, and the vast majority of paediatric surgeons' cumulative experience will barely surpass the single digits. The incidence is estimated at around 1 in 15,000 pregnancies. It was first described by De Pozze in 1673 as an incidental finding during an autopsy of an adult woman.⁴ Patients frequently have other anomalies, including congenital heart disease. The long-term survival into adulthood is estimated at around 50% for agenesis of the left lung, and 30% in patients with the right lung affected.⁵ Lung isolation is being used more frequently in both adult and paediatric age groups due to increasing incidence of thoracoscopy and video-assisted thoracoscopic surgery in these patients. Lung isolation and one-lung ventilation (OLV) are indicated in various surgical and nonsurgical procedures. Video assisted thoracoscopic surgery (VATS) is a less invasive approach for thoracoscopic surgery.⁶ The management of hypoxia resulting with OLV is a stepwise drill of increasing inhaled oxygen, adding positive end-expiratory pressure (PEEP) to ventilated lung, and continuous positive airway pressure or high-frequency jet ventilation (HFJV) to non-ventilated side.

OLV causes more airway damage secondary to onsite manipulation. It also leads to significant physiological derangements such as ventilation-perfusion (V/Q) mismatching and early development of hypoxia. Main problem is obligatory shunt through the non-ventilated lung. Main compensatory mechanism is hypoxic pulmonary vasoconstriction (HPV). Development of hypoxemia (arterial oxygen saturation <90%) caused by OLV can be explained by following factors-

- Reduction in oxygen stores of the body, due to the disease process and collapse of one lung, the functional residual capacity (FRC), and hence, the oxygen stores of the body get significantly reduced in a situation of OLV.⁵

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- Poor oxygenation as effects of anaesthesia and the lateral decubitus position
- Compromised ventilation due to compression of ventilated, dependent lung by the weight of mediastinum and by abdominal contents after diaphragmatic paralysis further adds to the gravity of atelectasis of the ventilated lung. Increased closure of small airways with old age, reduced elastic recoil, and the lateral position leads further to more atelectasis.⁵
- Dissociation of oxygen from haemoglobin due to non-ventilation of one out of two lungs causes a reduction in arterial oxygen partial pressures, increase in CO₂ levels, and respiratory acidosis. It leads to rapid dissociation of oxygen from haemoglobin (Bohr effect), shown by the steep slope of the oxygen-dissociation curve
- Mismatched V/Q relationship caused by reduced FRC and atelectasis of both lungs, surgical retraction, and/or for children and adults with unilateral lung disease, V/Q matching is optimal when the patient is placed in the lateral decubitus position with the "healthy" lung in a dependent position. This holds true for both spontaneous and controlled ventilation. Due to gravitational forces, this position results in increased perfusion to the "healthy," dependent lung and decreased perfusion to the diseased, non-dependent lung.

Steps to Be Taken Sequentially to Evaluate and Treat Hypoxemia during One-Lung Ventilation

- Place patient on 100% FiO₂
- Evaluate position of tube or blocker by auscultation for breath sounds or through fiberoptic bronchoscopy
- Apply CPAP (5 cm of H₂O) or HFO or HFJV to the non-ventilated lung. Apply low levels of PEEP (10 cm of H₂O) to the dependent lung to improve the FRC
- Ensure there is no kinking or obstruction of the tube from secretions
- Intermittent lung recruitment manoeuvres can be used on operated side of lung
- Optimization of Hb levels and cardiac output

Lastly, in case of intractable hypoxia, the surgeon should be informed and asked for reinflation of operative lung or clamping of the pulmonary artery of the non-ventilated lung.

Investigations- Complete haemogram-
Haemoglobin 9.1 gm/dl
Total leucocyte count -17600 cells/cumm
Platelets -8.3 lakhs
PCV- 31%
Blood group- O positive
Electrolytes: sodium-144 meq/l
Potassium-4.4 meq/l
Renal function tests: blood urea-22 mg/dl
Serum creatinine-0.7 mg/dl
Chest x-ray showed right lung consolidation

CT scan of chest showed

1. Right upper lobe and middle lobe aplasia with mediastinal shift towards right side.
2. Fused 7th, 8th and 10th and 11th ribs on right side.
3. Bilateral cervical ribs.
4. Left severe hydronephrosis with thinning of cortex.
5. Malrotated right kidney.
2D ECHO shows dextroversion, no shunt or obstructive lesion.

Discussion of management

Child was taken up for surgery after explaining the consequences of surgery and anaesthesia to the parents and taking high risk consent from them.

Premedication was given with injection glycopyrrolate 0.05 mg iv and injection fentanyl 10 mcg iv, preoxygenated for 5 minutes with 100% oxygen; induced with injection thiopentone sodium 40 mg and intubated under suxamethonium 25 mg with 3.5 mm oral endotracheal tube (cuffed). Anaesthesia was maintained with sevoflurane and non-depolarising muscle relaxant vecuronium. Intraoperative vitals were stable

Surgery lasted for one and half hours. Neuromuscular blockade reversed with inj. neostigmine 1 mg and inj. glycopyrrolate 0.05 mg iv. Child was extubated after full recovery. Rectal suppository paracetamol 90 mg was inserted for postoperative analgesia. Patient was shifted to PICU and kept under observation for 24 hrs, and shifted to paediatric surgery ward. Child was discharged after 5 days.

Final Diagnosis

Anaesthetic management of a rare case of unilateral lung aplasia posted for perineal anorectoplasty.

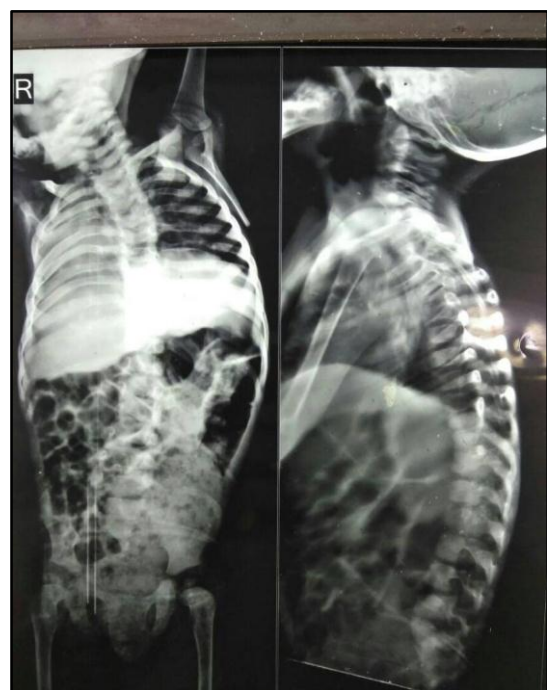


Figure 1. Lung Aplasia 2



Figure 2. Lung Aplasia

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