

Anaesthesia recommendations for **Congenital Pulmonary Airway Malformation**

Disease name: Congenital Pulmonary Airway Malformation (CPAM)

ICD 10: Q33.0

Synonyms:

Congenital cystic adenomatoid malformation of lung (CCAM)
Congenital honeycomb lung
Cystic adenomatoid malformation
Cystic lung, congenital
Single lung cyst

Disease summary: Congenital Pulmonary Airway Malformation (CPAM), formerly called Congenital cystic adenomatoid malformation of lung (CCAM), is a congenital lung lesion in children as a result of an embryologic insult in early gestation (7th week of Intrauterine life) causing maldevelopment of the terminal bronchiolar structures. Incidence is 1:25,000–1:35,000 births and it represents up to 25% of congenital lung abnormalities. There is increased cell proliferation but decreased apoptosis, resulting in adenomatoid proliferation of terminal respiratory bronchioles with intercommunicating cysts. The lesion is connected to the airway but a normal intrapulmonary bronchial system is missing. CPAM is classified on the basis of the cyst size (Type I: cysts 2-10 cm; Type II: cysts 0.5-2 cm; Type III: micro-cystic). It may be diagnosed by prenatal ultrasound and can regress or increase in size. It may present ante-natally in exceptional cases (less than 1%) as polyhydramnios or foetal hydrops with foetal demise. At birth it may present as respiratory distress. Small CPAMs may remain asymptomatic and present later in life or are found incidentally. Associated (renal, intestinal, bony, cardiac) anomalies may be present in up to 25% patients (usually in Type II). Except for cardiac anomalies, the other associated malformations worsen the prognosis.

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Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong



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Typical surgery

Open foetal surgery: Not conventionally realised. Prenatal chest tube insertion in case of compressive cysts

Foetoscopic procedures

EXIT procedure: Intervention while on placental support-intubation, ECMO, thoracotomy and resection

Post-natal surgery: Thoracotomy/thoracoscopy and resection, lobectomy, lung sparing surgery

Type of anaesthesia

General anaesthesia with spontaneous ventilation is preferred till lobectomy or resection of the cystic mass due to fear of rupture of cysts with positive pressure ventilation (PPV) which may cause further cardiorespiratory compromise. Muscle relaxation may be instituted after resection / General anaesthesia with muscle relaxation may be advocated if there is no cardiorespiratory compromise. Local anaesthesia in the form of infiltration or regional thoracic epidural or paravertebral block can be given either pre incision or post procedure.

Necessary additional pre-operative testing (beside standard care)

Chest X Ray, CT scan is indicated in all patients to delineate the anatomy. Chest X Ray will reveal marked hyperlucency of involved lobe which may be mistaken for a pneumothorax. There may be mediastinal shift, downward displacement of the diaphragm, compression atelectasis of surrounding lung tissue and herniation of involved lobe across the mediastinum. The left upper lobe is most commonly involved [3]. CT scan may detect abnormally narrowed bronchus with the affected lobe or collapsed lobe. Scintigraphy may reveal absent perfusion of the affected lobe. Occasionally, a Bronchoscopy may be indicated to rule out a foreign body or mucous plug. The association of CPAM with congenital heart disease is rare and accounts for 15-20% of cases. Cardiac evaluation with a 2D echo is warranted in patients presenting with a murmur or failure to thrive to rule out associated congenital heart disease.

Particular preparation for airway management

In neonates, full preparation of appropriate sized airway equipment is necessary. Pre oxygenation is mandatory. There may be difficulty in oxygenating or ventilating the patient in severe respiratory distress. Humidified high frequency nasal cannula or nasal CPAP may help in airway management till tracheal intubation. Case should be discussed with the surgical team beforehand and if the risk of pneumothorax is believed to be significant, then spontaneous breathing and surgical standby is a must for emergent decompression in case of rupture of cysts with positive pressure ventilation (PPV) and tension pneumothorax. In most cases, muscle relaxant with gentle PPV during induction is the method of choice for ventilation and does not result in significant pneumothorax. In thoracoscopic resections, one lung ventilation may be considered and instituted with Fogarty's embolectomy catheter or

Arndt endobronchial blocker. However, care should be taken to avoid right upper lobe collapse in case of right-sided bronchial blockers. Continuous vigilance for rising ETCO₂ concentrations is warranted in thoracoscopic repairs and immediate conversion to two lung ventilation may be needed.

Particular preparation for transfusion or administration of blood products

Blood supply of lesions is from the pulmonary circulation. During resection, there may be massive haemorrhage. Transfusion of 10-15 ml/kg of 5% albumin or 10-15ml/kg of packed RBC's or whole blood has been reported.

Particular preparation for anticoagulation

There is no evidence to support the need of particular anticoagulation.

Particular precautions for positioning, transportation and mobilisation

Ante-natally diagnosed giant CPAM may present as neonatal respiratory distress. In these cases, intubation immediately in the delivery room is indicated. Following this neonate needs to be stabilized in NICU. Arterial line and nasogastric tube should be inserted. The neonate should be transported to OT in a heated isolette along with nasal CPAP or a T-piece resuscitator. Following induction of anaesthesia, lateral position will be required for thoracoscopy/thoracotomy. General anaesthesia, neuromuscular blockade and mechanical ventilation cause a decrease in FRC of both lungs. In infants with unilateral disease, oxygenation is worse with the healthy lung down. Infants have a soft, easily compressible rib cage that cannot fully support the underlying lung. Therefore, the infant's functional residual capacity is closer to residual volume, making airway closure likely to occur in the dependent lung even during tidal breathing. The infant's small size results in a reduced hydrostatic pressure gradient between the nondependent and dependent lungs. Consequently, the favourable increase in perfusion to the dependent, ventilated lung is reduced in infants. This coupled with the high oxygen consumption in infancy, compression of underlying lung with bolsters and diaphragm and surgical retraction leads to desaturation in the lateral position. In order to avoid or minimize the risk of desaturation in infants, single lung ventilation (SLV) is usually avoided. Techniques used for SLV in infants and young children should include the option of providing oxygen to the operative lung or immediate conversion to two lung ventilation.

Interactions of chronic disease and anaesthesia medications

None reported.

Anaesthetic procedure

Induction of general anaesthesia should be smooth. A crying, struggling child may trap increased amounts of air during violent inspiratory efforts. Preoxygenation followed by

inhalational induction with sevoflurane. Muscle relaxation may be given if cysts are small. In case of giant CPAM and risk of rupture with PPV, spontaneous ventilation is best preserved with sevoflurane and IV fentanyl till the cysts are ruptured. Anaesthesia related myocardial depression and/or vasodilation associated with halogenated anaesthetics may cause severe hypotension. This can be prevented by limiting the concentration of inspired sevoflurane and opioid analgesia with fentanyl in doses of 2-4 µg. Alternatively, induction with IV ketamine and oxygen with local infiltration of the surgical site has been considered a safer combination till the intrathoracic pressure is relieved. Positive pressure ventilation and positive end expiratory pressure should be minimized to prevent further inflation of the involved lobe. Surgeon standby during induction is mandatory for decompression in high risk patients and in those with haemodynamic collapse. One lung ventilation is established with either of the following options: Selective mainstem endobronchial intubation with a single-lumen endotracheal tube (ETT), balloon-tipped bronchial blockers in infants [Fogarty catheters (3, 4,5 Fr G), Arndt endobronchial blocker (smallest 5 Fr G)], Marraro DLT for neonates and infants. Univent tube, or a double-lumen tube (DLT) use is restricted to bigger children above 6 years of age and hence is rarely useful in CPAM.

Thoracic epidural catheters or ultrasound guided paravertebral block is recommended for analgesia in thoracotomies. If instituted after induction, these blocks may help limit the inspired concentration of inhalational anaesthetic and opioid dose. Use of Erector Spinae Plane block has not been reported in CPAM but its use can be considered. Short acting opioids like fentanyl/remifentanyl or analgesics like paracetamol as a part of multimodal analgesia.

During maintenance of anaesthesia, hyperinflation of cysts can be prevented by avoiding the use of nitrous-oxide before delivery of affected lobe.

Postoperatively, patient may be extubated if respiratory and haemodynamic parameters are normal. In unstable babies, ventilation is continued postoperatively in NICU until successful weaning can be done. Alternatively, babies may be shifted to NICU on ventilatory support and extubated early there.

Particular or additional monitoring

Pre- and post-ductal oxygen saturation monitoring is a must to detect shunting and desaturation. Invasive monitoring like arterial line is indicated in giant CPAM with risk of haemodynamic compromise. In addition to routine monitoring like temperature, electrocardiogram and capnometry, a precordial stethoscope is recommended for one lung ventilation in infants.

Possible complications

Large CPAMs may cause difficulty in oxygenation and ventilation during positioning, lung retraction, compression and tracheobronchial kinking. Catastrophic cardiovascular deterioration may occur due to rupture of cysts at induction of anaesthesia. Compression atelectasis, decreased compliance, mediastinal shift and finally pneumothorax may occur. In such cases, the surgeon may need to perform emergency thoracostomy which will allow herniation of the overdistended lobe out of the thorax and haemodynamic stabilization.

Because of the limited chest space, respiratory difficulties or surgical exposure, thoracoscopic procedures may need conversion to thoracotomy, especially if there is a preoperative

history of lung infection, if the patient's weight is under 10 kg and if the lung exclusion is insufficient.

Endotracheal tube displacements may occur especially during positioning. Displacement of endobronchial blockers (Fogarty's embolectomy catheter, Arndt Blocker) may occur causing complete inability to ventilate both lungs. In such cases, simple deflation of the cuff will restore ventilation.

Hypoventilation with carbon dioxide retention and acidosis may occur during spontaneous ventilation or during one lung ventilation. This may cause severe pulmonary vasoconstriction and may precipitate acute right heart failure in a child with congenital heart disease or cause shunting of blood at the atrial or ventricular level.

Surgical retraction and chest compression in the lateral position can cause additional haemodynamic effects like decrease in venous return, aortic compression, mechanical arrhythmias due to cardiac retraction and bleeding.

Post-operative care

Postoperatively, the patient may be extubated if respiratory and haemodynamic parameters are normal.

In unstable babies, ventilation is continued in NICU until successful weaning can be done.

Alternatively, babies may be shifted to NICU on ventilatory support and early extubation may be considered.

Those on ECMO may be decannulated in the OR at the end of resection or may remain on ECMO.

Those on HFOV may be continued on HFOV or may be transitioned to conventional ventilation in NICU.

Disease-related acute problems and effect on anaesthesia and recovery

Clinical manifestations such as tachypnea, tachycardia, cyanosis, retractions, wheezing, tympanic chest percussion, asymmetric breath sounds or displaced cardiac tones may easily be mistaken as a Pneumothorax.

Repeated episodes of respiratory distress associated with respiratory infection, agitation, crying on feeding, failure to thrive may be evaluated for a congenital cardiac disease.

Ambulatory anaesthesia

An infant with CPAM may present for CT scan. This can be considered on ambulatory basis only in incidentally diagnosed CPAM on Chest X ray. In all other cases, admission and High Dependency Unit care should be advocated even for minor diagnostic procedures.

Obstetrical anaesthesia

There is no indication of premature delivery and no specific maternal procedures are required.

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Date last modified: July 2019

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Disclosure(s) The author has no financial or other competing interest to disclose. This recommendation was unfunded.

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Disclosures The reviewers have no financial or other competing interest to disclose.
