Anesthesia in Patients with LAM

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Patients with LAM may receive varying levels of anesthesia during procedures. Most often, these procedures are related to their underlying LAM, such as transbronchial or video-assisted thoracoscopic surgery (VATS)-guided lung biopsy, chest tube placement for pneumothorax or chylothorax, thoracic duct embolization, drainage of chylous ascites, pleurodesis, and lung transplantation. However, LAM patients may also need other procedures that are not directly related to LAM, such as obstetric indications, joint replacements, etc. The presence of LAM creates a unique set of challenges that need to be carefully considered by anesthesiologists prior to undertaking any procedures.

1. Careful pre-operative planning should involve the patient's primary care doctor or LAM specialist, surgeon and anesthesiologist. Some centers around the country may have preoperative anesthesia clinics. Surgeries should be performed at a center that has the capacity to insert a chest tube if a pneumothorax should occur. At minimum, a phone call and discussion should occur between the patient and their anesthesiologist prior to the day of surgery for even the most minor surgery.
2. A thorough history and physical exam should be performed by the anesthesiologist prior to surgery. Review of all data should include electrocardiogram, pertinent labs, chest imaging to determine disease severity, and pulmonary function tests. Preoperative echocardiography is not typically necessary, unless the patient has a history of pulmonary hypertension, cardiac dysfunction, or a new clinical indication.
3. Patients should be instructed to take all of their medications on the day of surgery. Special attention should be paid to the use of mTOR inhibitors such as sirolimus and everolimus. mTOR inhibitors can delay wound healing and patients should be instructed to stop these drugs for at least 1 week before and after surgeries. Preoperative bronchodilators should be taken if a patient is already on one, but there is little evidence to suggest that they should be used empirically for bronchodilator naïve patients prior to induction of anesthesia.
4. Induction of general anesthesia and intubation may have increased risks. Depending on patient factors and type of surgery, monitored anesthesia care with local anesthesia, nerve blocks, or a primary regional technique including neuraxial technique should all be considered and carefully planned prior to surgery.
5. Standard ASA monitors should be used. Invasive arterial access and central venous access should be considered if hemodynamic instability, high likelihood of blood loss or need for frequent blood sampling is anticipated.
6. Patients should be well-preoxygenated with an FiO_2 of 100% for 5 minutes prior to induction of anesthesia. LAM is not associated with upper airway deviation or obstruction, so direct laryngoscopy and tracheal intubation can be achieved with a standard blade. If there is any concern about the success of rapid tracheal intubation, video laryngoscopy should be used, as these patients often have little reserve and cannot tolerate prolonged periods of hypoventilation.
7. The range of pulmonary dysfunction can vary in LAM. As such, it is paramount that preoperative pulmonary function testing and imaging be reviewed prior to surgery.
8. Intraoperative ventilation strategies should be tailored based on type of pulmonary dysfunction. Generally, low stretch tidal volumes (4-6cc/kg ideal body weight) are recommended. Airway pressures should be kept low (<30mmHg). Use the minimal FiO_2 that keeps SaO_2>92%. Low levels of PEEP are well tolerated and recommended, higher PEEP can increase the risk of barotrauma and development of pneumothorax.

9. Extreme intraoperative vigilance is required for pneumothorax. Consider this diagnosis if unexpected hypotension, tracheal deviation and/or high peak airway pressures occur.
10. Intraoperative fluid management should be restrictive to avoid pulmonary edema.
11. There have been no studies comparing inhaled anesthetics and total intravenous anesthetics for maintenance of anesthesia. Baseline patient characteristics and type of surgery should guide anesthetic choice, and the goal should be to minimize barotrauma and need for prolonged ventilatory support after the procedure.
12. Nitrous oxide should be avoided during general anesthesia in patients with cystic lung disease or closed pneumothorax. Nitrous oxide accumulates faster in closed gas spaces than it can diffuse out, which can rapidly expand a closed space.
13. Postoperative monitoring is essential for LAM patients, as diminished pulmonary reserve can lead to respiratory failure and prolonged ventilatory support. Any muscle relaxation used should be fully reversed and all extubation criteria should be met. Vital signs including pulse oximetry should be monitored in post anesthesia care units. End tidal capnography, when available, is encouraged in the immediate postoperative phase. LAM patients may require supplemental oxygen in the postoperative period, and incentive spirometry should be encouraged. Early mobilization is also critical to ensure rapid recovery.
14. Management of pain is critical. Use opioids judiciously to maintain an optimal balance between pain relief and adequate respiratory function. Strongly consider local or regional anesthetic techniques if possible and use of other adjuncts, such as NSAIDs, acetaminophen, gabapentin, ketamine.
15. Depending on baseline patient characteristics and type of surgery, patients with LAM can go home the same day. However, hospital admission and intensive care monitoring should be considered if surgery is prolonged or complicated.

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