ANESTHESIA MEDICATION SAFETY LIST
FOR POMPE DISEASE

Overview:
People diagnosed with Pompe disease are at increased risk for respiratory and cardiac complications during and after anesthesia due to weakness of the diaphragm and other breathing muscles. Careful choice of anesthetic agents, close monitoring, and postoperative respiratory support are critical for safety.

Medications to Avoid:
These medications can cause dangerous side effects or worsen muscle weakness in Pompe disease.

- Succinylcholine (Suxamethonium): ***Avoid***. May trigger rhabdomyolysis, hyperkalemia, or cardiac arrest in patients with muscle disease.
- Inhaled volatile anesthetics (e.g., Halothane, Isoflurane, Sevoflurane, Desflurane): ***Use with caution***. Risk of malignant hyperthermia–like reactions or prolonged muscle weakness.
- High-dose opioids (e.g., Morphine, Fentanyl): May depress respiration.
- Long-acting neuromuscular blocking agents (e.g., Pancuronium, Vecuronium): Can cause prolonged paralysis due to decreased clearance.
- Muscle relaxants without TOF monitoring: ***Avoid use without Train-of-Four response monitoring*** to prevent incomplete recovery of breathing muscles.

Medications That May Be Used With Caution

- Short-acting anesthetic agents such as Propofol or Etomidate: Generally considered safer, but require close monitoring.
- Non-depolarizing muscle relaxants (e.g., Rocuronium, Atracurium): May be used with Train-of-Four (TOF) monitoring and full reversal before extubation.
- Local or regional anesthesia (nerve blocks, epidural, spinal): Often preferred when feasible to avoid systemic respiratory depression.
- Short-acting opioids (e.g., Remifentanil): May be used carefully under close observation.
- Dexmedetomidine (Precedex): Can be useful for sedation but monitor for bradycardia and hypotension.

Monitoring and Support

- Continuous capnography and pulse oximetry during and after surgery.
- Train-of-Four (TOF) monitoring to assess neuromuscular recovery.
- Avoid extubation until full spontaneous breathing is confirmed.

- Postoperative respiratory support (BiPAP or mechanical ventilation) should be available for all Pompe patients.
- Avoid supplemental oxygen alone without ventilatory support if CO_2 retention is suspected.

Postoperative Care

- Monitor for respiratory depression and delayed recovery.
- Resume noninvasive ventilation (BiPAP) as soon as possible.
- Provide pulmonary hygiene (cough assist, suctioning).
- Minimize sedating medications postoperatively.

References

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Glossary of Terms:

Anesthetic agent – A medication used to block pain or cause sleep during surgery or medical procedures.

BiPAP (Bilevel Positive Airway Pressure) – A noninvasive breathing device that provides two air pressures—one for inhaling and one for exhaling—to support weak breathing muscles.

Bradycardia – A slower than normal heart rate, usually below 60 beats per minute in adults. During anesthesia, it can occur as a side effect of certain medications or due to the body's response to surgical procedures. In most cases, it can be corrected by adjusting anesthesia depth, medications, or monitoring heart rhythm closely.

Capnography – Continuous monitoring of the amount of carbon dioxide (CO_2) a person exhales; used during anesthesia or ventilation to check how well someone is breathing.

Dexmedetomidine – A sedative drug used to calm patients or reduce anxiety during procedures. It causes minimal breathing suppression and is sometimes preferred for patients with weak respiratory muscles.

Hyperkalemia – A dangerous condition where potassium levels in the blood become too high, which can cause abnormal heart rhythms or cardiac arrest. It may occur with certain anesthesia drugs such as succinylcholine.

Hypotension – Low blood pressure that may reduce blood flow to vital organs. It can occur during anesthesia or recovery, often due to the effects of anesthetic drugs, muscle relaxants, or dehydration. Careful monitoring and medication adjustments help maintain safe blood pressure levels.

Malignant hyperthermia – A rare but life-threatening reaction to certain anesthesia drugs that causes a rapid rise in body temperature, muscle stiffness, and irregular heartbeat.

Mechanical Ventilation – Use of a machine to move air in and out of the lungs through a breathing tube when a person cannot breathe adequately on their own.

Neuromuscular Blocking Agents – Drugs that temporarily paralyze muscles during surgery, making it easier to place a breathing tube or perform delicate procedures. Patients with Pompe disease are especially sensitive to these medications.

Non-depolarizing Muscle Relaxants – A safer group of neuromuscular blockers that prevent nerve signals from reaching muscles without first activating them. They are preferred for people with Pompe disease.

Noninvasive Ventilation (NIV) – A method of supporting breathing using a mask or nasal interface instead of a breathing tube. Devices like BiPAP and CPAP provide this type of ventilation.

Pulse Oximetry – A simple test that measures oxygen levels in the blood using a small sensor on the finger or ear.

Respiratory Support – Medical methods used to help a person breathe, including oxygen therapy, BiPAP, or mechanical ventilation.

Rhabdomyolysis – The breakdown of muscle tissue that releases proteins and electrolytes into the bloodstream, which can harm the kidneys and heart. It may occur after use of certain anesthesia drugs in muscle disorders.

Suxamethonium (Succinylcholine) – A short-acting depolarizing muscle relaxant used for rapid muscle paralysis. It is **contraindicated** in Pompe disease due to the risk of hyperkalemia and rhabdomyolysis.

TOF (Train-of-Four) monitoring – A method used during anesthesia to check the level of muscle relaxation by delivering small electrical impulses to a nerve.

Ventilatory support – The use of devices such as BiPAP or a ventilator to help move air in and out of the lungs when the breathing muscles are weak or when recovering from anesthesia.