SUXAMETHONIUM (SUCCINYLCHOLINE)

Suxamethonium (also called Succinylcholine) is a depolarizing neuromuscular blocking agent used to produce short-term paralysis during anesthesia, typically for rapid sequence intubation or surgical procedures.

Mechanism of Action

- Suxamethonium mimics the neurotransmitter acetylcholine and binds to nicotinic receptors at the neuromuscular junction, causing continuous depolarization of the muscle membrane.
- This sustained depolarization prevents further muscle contraction, leading to paralysis.
- It is normally broken down rapidly by the enzyme plasma cholinesterase (pseudocholinesterase), so its effects are short-lived (usually less than 10 minutes).

Why Suxamethonium Should NOT Be Used in Pompe Disease:

Pompe disease causes progressive muscle weakness due to glycogen buildup in skeletal, cardiac, and respiratory muscles.

- The use of suxamethonium in these patients is contraindicated or strongly discouraged for several important reasons:
    - **Risk of Rhabdomyolysis and Hyperkalemia**
        - In patients with underlying muscle disease (like Pompe), muscle membranes are fragile.
        - Suxamethonium can trigger massive potassium release from damaged muscle cells, leading to life-threatening hyperkalemia (elevated potassium levels).
        - This can cause cardiac arrhythmias or cardiac arrest.
    - **Prolonged Paralysis**
        - Some patients with Pompe disease may have altered metabolism or pseudocholinesterase deficiency, causing prolonged paralysis and delayed recovery after administration.
    - **Increased Sensitivity to Muscle Relaxants**
        - Some patients with Pompe disease may be hypersensitive to both depolarizing and non-depolarizing neuromuscular blockers due to reduced muscle mass and impaired respiratory function.
        - This can lead to prolonged apnea and difficulty weaning from ventilatory support.
    - **Respiratory Compromise**
        - Pompe disease frequently affects the diaphragm and other respiratory muscles.

- Even mild residual paralysis or weakness after anesthesia can cause respiratory failure, especially if the patient already relies on ventilatory assistance (BiPAP).

Safer Alternatives

For anesthesia or intubation in patients with Pompe disease:

- Avoid suxamethonium (succinylcholine).
- Use short-acting non-depolarizing agents (e.g., rocuronium or cisatracurium) with careful neuromuscular monitoring.
- Ensure availability of ventilatory support and reversal agents as needed.

References:

1. **Kumbar V, et al.** Anaesthetic management of a patient with Pompe disease. *Indian J Anaesth.* 2016; 60(9): 682–684.
   PubMed: https://pubmed.ncbi.nlm.nih.gov/27695234/
2. **Gurrieri C, et al.** Patients with glycogen storage diseases undergoing anesthesia: a case series. *BMC Anesthesiol.* 2017; 17: 97.
   PubMed: https://pubmed.ncbi.nlm.nih.gov/28619160/
3. **Radeka JZ, et al.** Anesthesia and rare neuromuscular diseases. *Front Anesthesiol.* 2023; 2: 1159436.
   DOI: https://doi.org/10.3389/fanes.2023.1159436
4. **Khan R, Bansal V.** Succinylcholine Chloride. *StatPearls [Internet].* Treasure Island (FL): StatPearls Publishing; Updated 2024.
   NCBI Bookshelf: https://www.ncbi.nlm.nih.gov/books/NBK499984/
5. **ANECTINE® (succinylcholine chloride)** Prescribing Information. U.S. Food and Drug Administration. Revised 2024.
   DailyMed label: https://dailymed.nlm.nih.gov/dailymed/drugInfo.cfm?setid=0b990f84-9515-44bd-a97d-a97bbead065c
6. **Martyn JAJ, Frontera WR.** Succinylcholine-induced hyperkalemia in acquired pathologic states. *Anesthesiology.* 2006; 105(2): 232–239.
   PubMed: https://pubmed.ncbi.nlm.nih.gov/16394702/