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:

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Glossary of Terms:

Acetylcholine – A chemical messenger (neurotransmitter) that helps nerves communicate with muscles, triggering muscle contraction.

Anesthesia – A medically controlled loss of sensation or awareness used during surgery or procedures, achieved with medications that prevent pain and relax muscles.

Apnea – A temporary pause in breathing, often caused by weak muscles or certain anesthesia medications.

BiPAP (Bilevel Positive Airway Pressure) – A machine that provides two levels of air pressure through a mask to help people with weak breathing muscles inhale and exhale.

Cardiac arrest – When the heart suddenly stops beating effectively, causing loss of blood flow to the body; a medical emergency.

Cholinesterase (Plasma or Pseudocholinesterase) – An enzyme in the blood that breaks down certain medications like suxamethonium; deficiency can cause drugs to last longer than expected.

Depolarizing muscle relaxant – A medicine that briefly activates and then blocks muscle signals, causing temporary paralysis (suxamethonium works this way).

Diaphragm – The main breathing muscle under the lungs that helps draw air in; it can be weakened in Pompe disease.

Hyperkalemia – A high level of potassium in the blood that can cause dangerous heart rhythms.

Intubation – The placement of a breathing tube through the mouth into the airway to help a patient breathe during surgery or emergencies.

Neuromuscular blocker – A medication that temporarily paralyzes muscles by blocking communication between nerves and muscles; used in anesthesia.

Paralysis – The inability to move a muscle or group of muscles, sometimes caused intentionally during surgery using neuromuscular blockers.

Plasma cholinesterase deficiency – A condition where the enzyme that breaks down suxamethonium works too slowly, leading to prolonged paralysis after anesthesia.

Rhabdomyolysis – The breakdown of muscle tissue that releases muscle proteins and electrolytes into the bloodstream, which can harm the heart and kidneys.

Rocuronium / Cisatracurium – Non-depolarizing muscle relaxants used as safer alternatives to suxamethonium in people with neuromuscular conditions.

Suxamethonium (Succinylcholine) – A fast-acting muscle relaxant used during anesthesia; it is generally avoided in people with muscle diseases like Pompe because of dangerous side effects.

Ventilatory support – The use of machines such as BiPAP or mechanical ventilators to assist breathing when muscles are too weak to move air effectively.