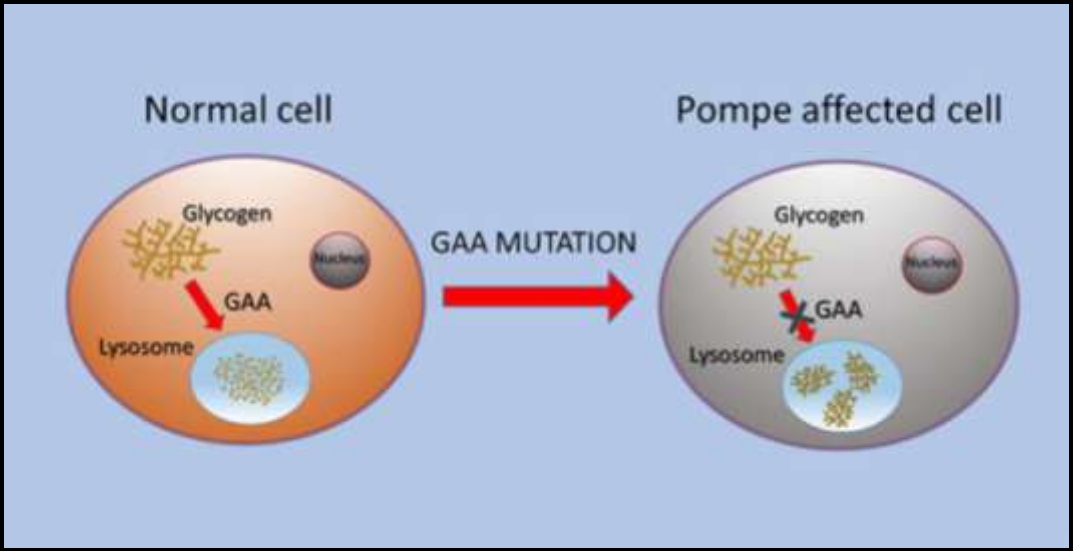
Introduction



- Late onset Pompe's disease (LOPD) caused by acid alpha-glucosidase deficiency → progressive muscle weakness, respiratory compromise.
- Data on obstetric anesthesia management remain sparse

Myopathic Conditions & Anesthesia Risks

- Duchenne Muscular Dystrophy (DMD) linked to MH (malignant hyperthermia) or anesthesia-induced rhabdomyolysis (AIR)
- Congenital myopathies (e.g., central core disease with RYR1 mutation) strongly tied to MH
- **No current evidence** linking LOPD to MH or AIR.

Multidisciplinary Approach

- Essential given potential for respiratory, musculoskeletal complications and need for specialized anesthesia planning.

Case

- ***27-Year-Old G4P2 with LOPD at 36 Weeks Gestation***

- **History & Diagnosis**
  - Diagnosed in mid-20s after progressive proximal muscle weakness in the lower limbs.
  - Family history: brother passed away in his 30s from respiratory complications secondary to severe lung infection s/o hereditary component.

- **ERT (Enzyme Replacement Therapy)**
  - Receiving avalglucosidase alfa (Nexviazyme); monthly infusions with no adverse effects.

- **Clinical Status**
  - Baseline cardiac & pulmonary evaluations unremarkable.
  - At 36 weeks, no significant respiratory or hemodynamic compromise.

- **Delivery Plan**
  - Induction of labor scheduled at 39 weeks+3 days – delivered uneventfully at 39 weeks+4 days
  - Given early epidural to minimize general anesthesia risks.

Conclusion

Impact of ERT

- Improved survival & functional status in LOPD, enabling patients to reach reproductive age.

Pregnancy Challenges

- Potential respiratory compromise & muscular weakness in view of highly variable course of LOPD.
- Distinction from other myopathies (e.g., DMD, central core disease caused by RYR1 mutations) that have known Malignant hyperthermia or anesthesia-induced rhabdomyolysis risk
- No current link between LOPD and MH or AIR.

Neuraxial Anesthesia Benefits

- Literature supports safety in LOPD
- Minimizes airway manipulation & respiratory stress vs. general anesthesia.

Future Directions

- Need for more data on peripartum protocols & long-term outcomes.
- Reinforces a **multidisciplinary approach** (OB, anesthesia, neurology, genetics, critical care)