Anesthetic Management of a Cesarean Delivery in a Parturient With Glycogen Storage Disease and Multisystem Involvement

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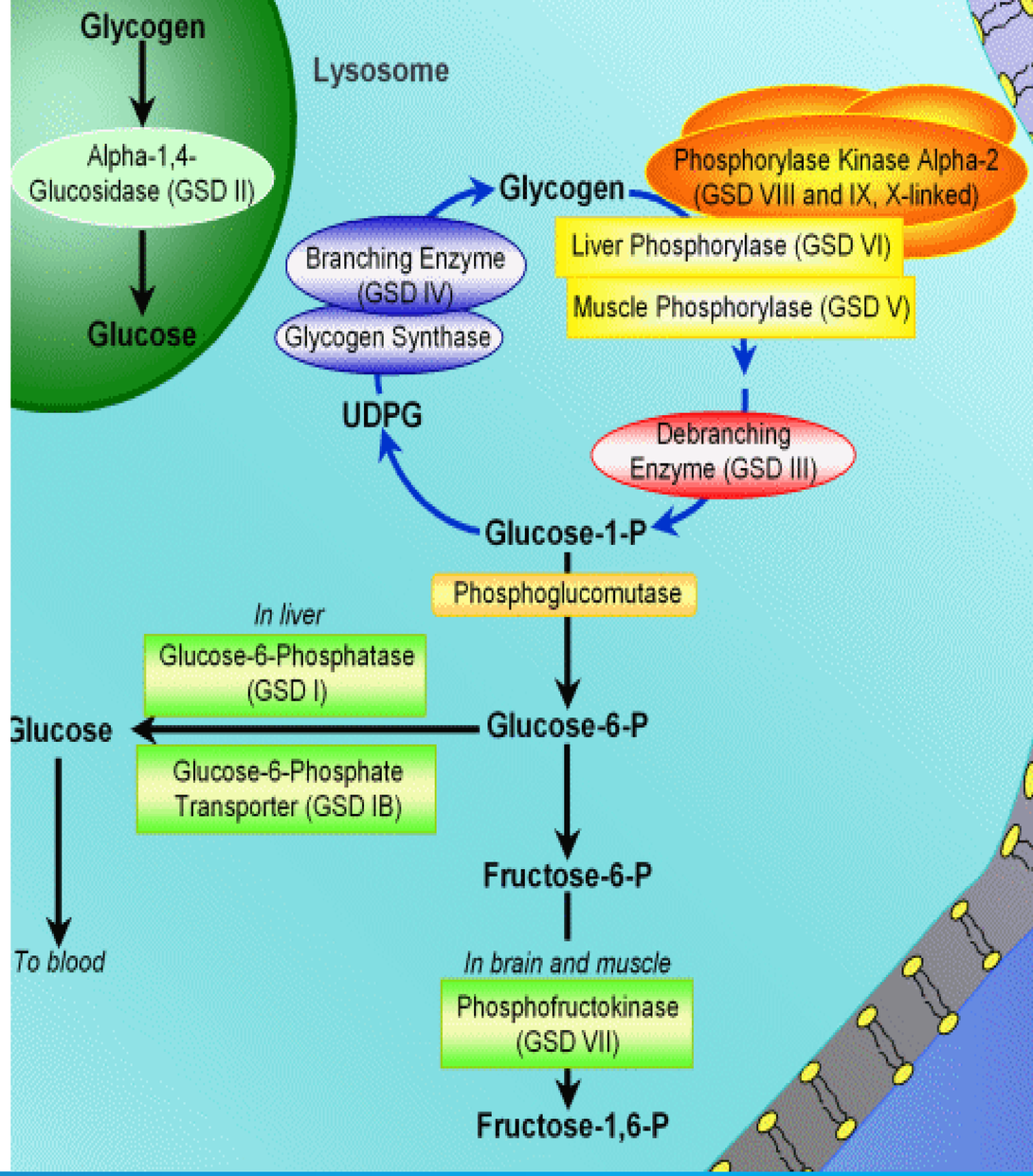
May 3rd, 2026

Mount Sinai

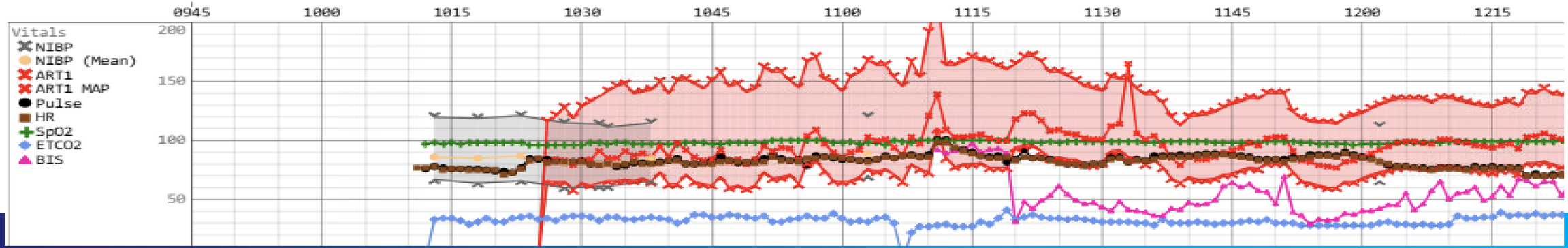
Background

- Glycogen storage disease type IIIa (GSD IIIa) is an autosomal recessive disorder caused by deficiency of the glycogen debranching enzyme, resulting in accumulation of abnormal glycogen in both liver and muscle tissues (including cardiac and skeletal muscle)
- The condition results from mutations in the AGL gene, which encodes the glycogen debranching enzyme
  - **Incompletely degraded glycogen ("limit dextrin") accumulates**
  - This protein has two independent catalytic activities (transferase and glucosidase) necessary for complete glycogen degradation.
- In the liver, without full glycogen breakdown, this process leads to fasting hypoglycemia and hepatomegaly
- In the skeletal muscle, glycogen accumulation can lead to exercise intolerance and muscle weakness
- In cardiac muscle, the accumulation can lead to hypertrophy
- Cardiac involvement occurs in approximately 58% of patients, though most cases manifest as asymptomatic left ventricular hypertrophy rather than clinically significant cardiomyopathy.
- Clinical manifestations typically begin in infancy with hepatomegaly, hypoglycemia, hyperlipidemia, and growth retardation.



The Case

- 26 y/o G1P0, GSD IIIa, presenting for scheduled primary cesarean section
- Comorbidities: HOCM (LVOT gradient ~13 mmHg, LVEF 60%, mild pHTN, mild MR — on carvedilol), cirrhosis with portal hypertension and esophageal varices (EGD + banding at 28w1d), progressive thrombocytopenia (60k, down from 80–100k, with epistaxis — started on TXA, iron; DDAVP planned pre-delivery), recurrent hypoglycemia, chest pain, dyspnea, LE edema
- First presented to MFM at 12w6d; counseled on risks and elected to proceed
- Multidisciplinary team: MFM, Nutrition, Hematology, Genetics, Cardiology, GI, ICU, Obstetric Anesthesiology (first consulted at 22 weeks)
- Decision made to deliver in main OR with cardiac anesthesiology, TEE, and ECMO available
- Admitted at 35w4d; cesarean proceeded at 36w6d
- Monitors: standard ASA + pre-induction radial arterial line + RIJ 14fr introducer
- Induction: etomidate, fentanyl, rocuronium; intubation without complication
- Intraoperative TEE: massive LVH, minimal LVOT obstruction, trace AI, severe MR Hemodynamics managed with phenylephrine and judicious volume Extubated at end of case; transferred to CCU; discharged POD5



Teaching Points

- Parturients with GSD IIIa can have multisystem involvement from the disease.
- Patients with HOCM and bleeding histories present challenges to the anesthesiologist during the peripartum period.
- Multidisciplinary involvement and delivery at a tertiary/quaternary care center with access to advanced interventions is critical to caring for complex obstetric patients.
- While the patient in this case had a relatively favorable outcome, careful management remains crucial, as a deterioration in cardiac function or sudden obstetric complications can pose significant risks.
- With careful planning, early consultation with specialists, and prompt intervention in response to hemodynamic changes, it is possible to safely navigate labor and delivery in a patient with glycogen storage disease IIIa with multi-organ involvement.