Navigating the Mitochondrial Maze: Emergent Intrapartum Cesarean Delivery in A Patient with Leigh Syndrome

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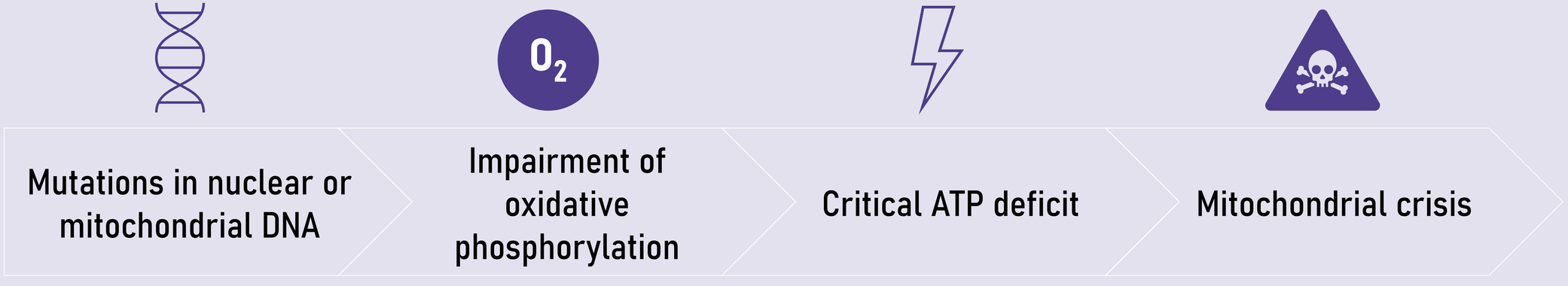
Mitochondrial Disease



Leigh Syndrome

- Incidence is 1 per 40,000; hallmarks are necrotizing lesions in the CNS with **rapid cognitive and motor decline**
- Usually **fatal in early childhood**, adult-onset cases are rare and poorly characterized
- No guidelines for obstetric management in this specific population exist

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Case Timeline

Prenatal

- Dx age 22
- Prolonged interruptions in treatment
- Cognitive decline, progressive lower extremity weakness
- Hx uncomplicated spinal anesthetic with **ropivacaine**

Antepartum

- OB anesthesiology consult
  - Collaboration with pharmacy for **ropivacaine epidural infusion**
  - Contingency planning

Intrapartum

- Exam: motor weakness requiring wheelchair use; MP3, small distances
- Analgesic plan (**prior to labor induction**): arterial line, dural puncture epidural
- 15h later, emergent cesarean delivery for NRFHTs
  - Conversion to **epidural anesthesia** with **2% lidocaine** + bicarbonate + epinephrine

Postpartum

- Uterine atony management with oxytocin, **methylergonovine**, **carboprost**
- Discharged POD5

Discussion and Teaching Points

- There is significant **genetic** and **phenotypic heterogeneity** between mitochondrial diseases
- Common goal of **preventing mitochondrial crisis**

- Increased O2 requirements during labor can **exacerbate ATP deficits**
- Early epidural labor analgesia can mitigate imbalances in O2 supply and demand
- Amide local anesthetics inhibit mitochondrial transport: **bupivacaine > ropivacaine > lidocaine**
  - Bupivacaine implicated in development of ventricular tachyarrhythmias
  - No safety data exists for **chloroprocaine**
- **General anesthesia** can be well-tolerated but most agents **inhibit oxidative phosphorylation**

References

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