Urgent cesarean delivery in a patient with Charcot-Marie-Tooth disease, scoliosis, and preeclampsia

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Background

- Charcot-Marie-Tooth (CMT) disease is a genetically heterogenous group of hereditary polyneuropathies
- Key considerations:
  - Autonomic dysfunction
  - Cardiac conduction abnormalities
  - Respiratory muscle weakness
  - Positioning challenges due to vertebral anatomy
  - Increased sensitivity to neuromuscular blocking agents
- CMT symptoms may significantly impact anesthetic interventions during labor

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Clinical Case

37 y.o., G3P1011, seen as pre-anesthesia consult for CMT

- Mixed obstructive/restrictive lung disease, on home NIV
- Obesity (BMI 35), OSA, orthopnea, remote hx of tracheostomy
- Scoliosis with L S1 radiculopathy

Neuraxial US identified L4-5 epidural space at 10cm in lateral position.
While supine: breathless, distressed, with SpO2 to 90%

Multidisciplinary discussion → GA + post-op ICU admission

Presented at 36 weeks with preeclampsia requiring urgent repeat cesarean section

- ➢ Preoxygenation with high flow nasal cannula and RSI
- ➢ Uneventful case, EBL 600cc
- ➢ Planned ICU admission postoperative for extubation to BiPAP on POD0
- ➢ Required Mg postoperatively, intermittent BiPAP

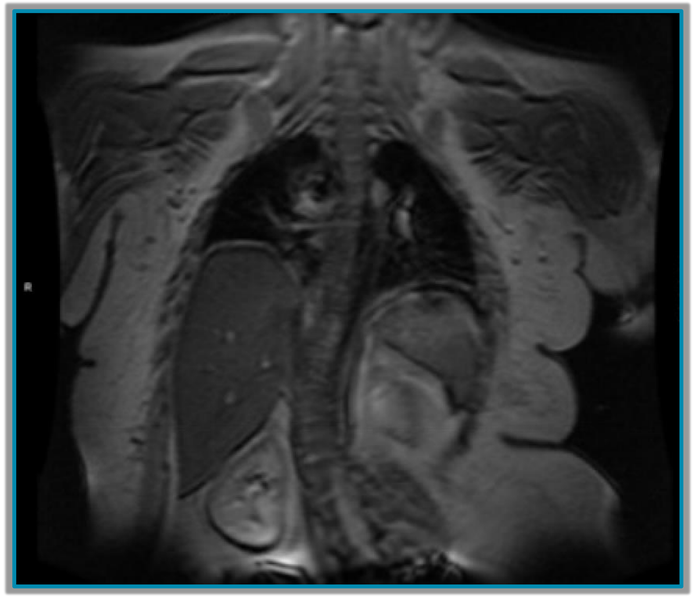

Fig 1. Thoracolumbar MRI demonstrating dextroscoliosis

Teaching Points

- Pregnancy has the potential to exacerbate CMT symptoms & warrants thorough pre-evaluation by obstetric anesthesia
- Both neuraxial and GA have successfully been used in CMT parturients
- If undergoing GA with known respiratory weakness from CMT, consider postoperative needs + disposition
- Multidisciplinary discussion and shared decision-making with the patient can facilitate a safe and positive birth experience