

Hereditary Neuropathy with Liability to Pressure Palsies (HNPP) in Obstetric Care

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Genetic Basis: Rare autosomal dominant demyelinating disease caused by a deletion on Chromosome 17p12 affecting the **PMP22** gene.

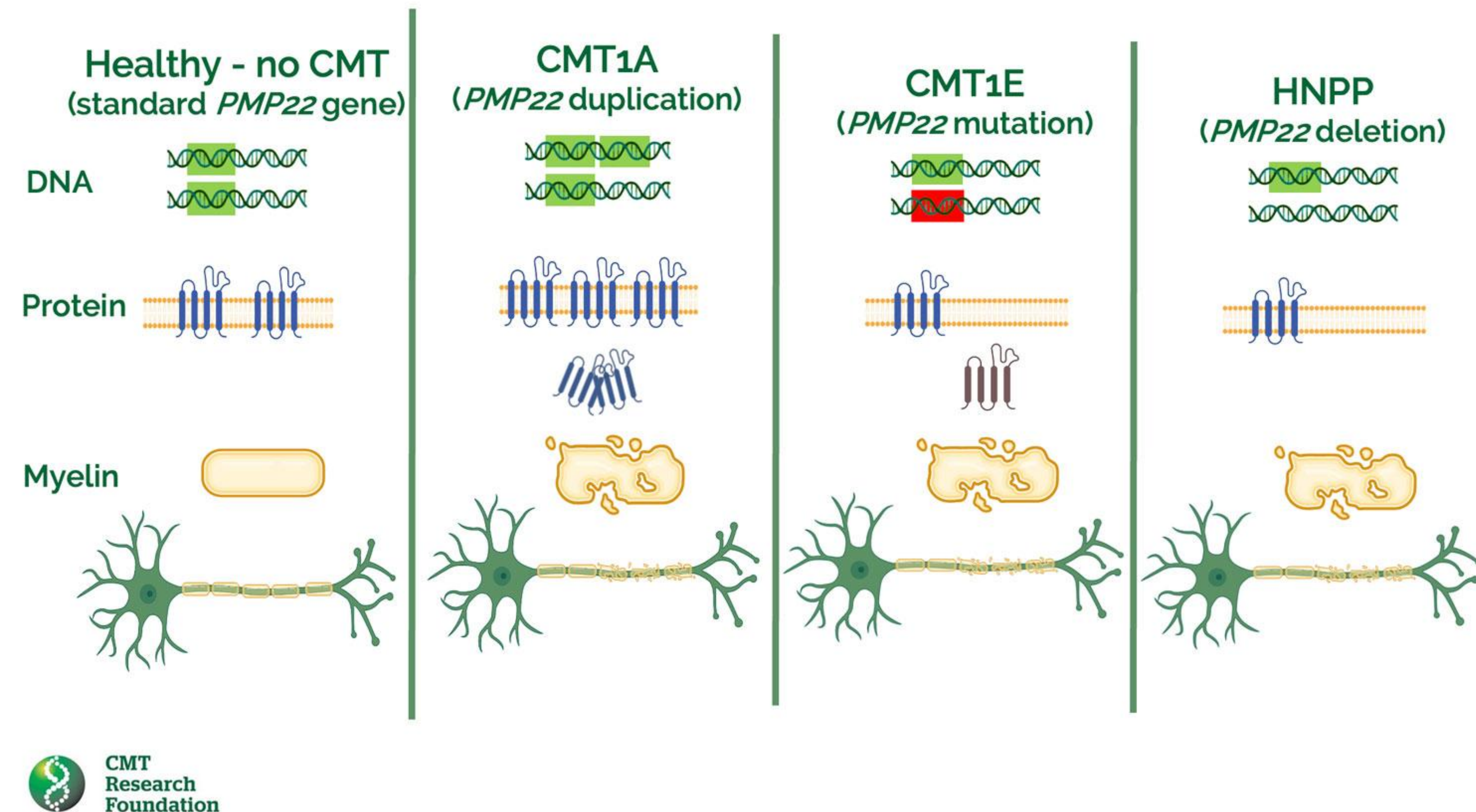
Pathophysiology: Myelin becomes exquisitely sensitive to pressure injury.

Clinical Presentation: Recurrent, transient focal neuropathies; 75% of patients report associated pain.

Prognosis: Resolution of symptoms can take days, weeks, or months.

Obstetric Implications

- Prolonged immobility during labor or Cesarean Delivery (CD) poses a risk for permanent disability.
- Management requires specialized anesthetic adaptations to prevent focal nerve compression.



The Case

G2P0 with known history of HNPP presented at 40 weeks gestation in early labor

- **Labor analgesia strategy:**

- Epidural Placement: L4-5 level.
- Ultra-low dose PCEA (Bupivacaine 0.625% + Fentanyl 2mcg/ml) to maintain patient mobility.
- Protective Measures: Bed was padded; patient asked to self reposition every 15–20 minutes.

- **Surgical anesthesia strategy:**

- After 14 hours, cesarean delivery indicated for arrest of dilation with category 2 fetal heart rate tracing.
- Anesthesia plan: epidural chloroprocaine 3% (20ml) and fentanyl (100mcg) followed by PF morphine 2mg post delivery
- Intraoperative precautions: OR bed padded; legs manually moved every 15 minutes by staff. BP Cuff site switched throughout case.
- Outcome: Lower extremity strength regained within 30 minutes of arrival to PACU; no neuropathies reported

Clinical Management Strategies for HNPP

Anesthetic Selection

- Neuraxial vs. General: Neuraxial is preferred to avoid oropharyngeal neuropathies associated with endotracheal tubes.

Pharmacology:

- Use ultra-low dose epidurals for labor to maintain sensation/mobility.
- Use Chloroprocaine for surgery to minimize the duration of post-operative immobility.
- Avoid succinylcholine

Vigilance & Positioning

- Frequent Repositioning may help prevent neuropathies from continuous focal pressure.
- Padding: Ensure all surfaces and bony prominences are well padded.

Monitoring Considerations

- Pressure Cuffs: While some literature suggests invasive monitoring to avoid cuff-related injury, standard cuffs may be used if the patient maintains sensation in the extremities to report discomfort.

Balancing Act: *Recognize that higher-dose analgesia reduces the patient's ability to detect an evolving neuropathy.*