

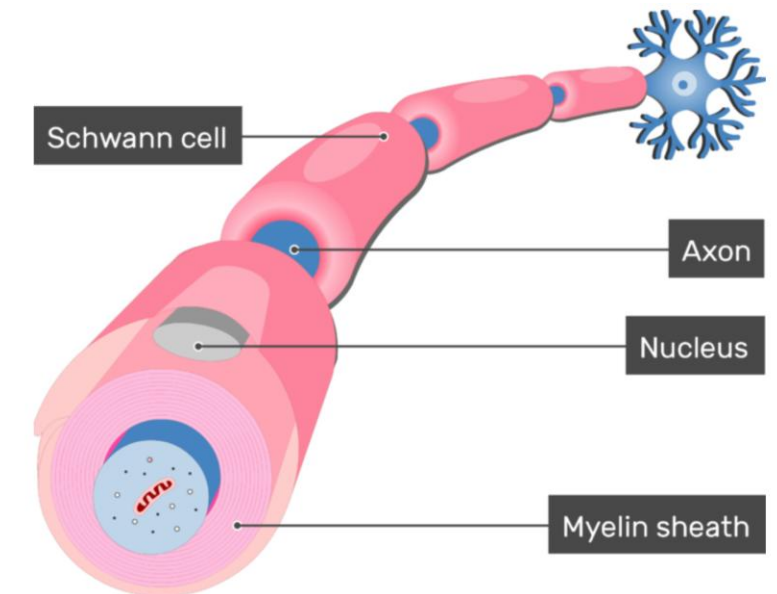
# No Pressure: Labor Management of Patient with Hereditary Neuropathy with Pressure Palsy

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## What is Hereditary Neuropathy with Pressure Palsy (HNPP)?

- Autosomal dominant condition with estimated incidence 7-16 per 10,000<sup>1</sup>
- One copy of PMP22 gene instead of two → Schwann cells malform peripheral nerve myelin<sup>1</sup>
- Increased risk of developing **motor and/or sensory neuropathies**, particularly with pressure and trauma, that can last hours to months<sup>1</sup>
- **Little information to guide anesthetic care**, including in peripartum period<sup>1</sup>
- Case reports describe neuraxial use; risk of causing exacerbation of condition **unknown**<sup>2</sup>



<https://cmtrf.org/what-is-cmt-disease/types-of-cmt/what-is-hnpp/>

1. Laudanski K, Elmadhoun O, Mathew A, et al. Anesthetic Considerations for Patients with Hereditary Neuropathy with Liability to Pressure Palsies: A Narrative Review. Healthcare (Basel). 2024;12(8):858. Published 2024 Apr 19. doi:10.3390/healthcare12080858
2. Lepski GR, Alderson JD. Epidural analgesia in labour for a patient with hereditary neuropathy with liability to pressure palsy. Int J Obstet Anesth. 2001;10(3):198-201. doi:10.1054/ijoa.2000.0828

# Case Overview



Patient: 33-year-old G1P0 with history of GERD, type 1 diabetes, and HNPP induced at 36w1d in the setting of new onset fetal growth restriction and challenging glycemic control

## Pre-delivery: Consult with OB Anesthesia Team

- Patient's history and current symptoms
- Reviewed current literature on HNPP management during labor; counseled patient about uncertain risk with neuraxial anesthesia
- Strategized on plans to minimize exacerbation including patient bringing her mattress topper, padding pressure points, and minimizing use of BP cuffs and SCDs when able

## Labor and Delivery Course

- On admission: Team reviewed precautions
- Patient initially developed sensory neuropathy from wrist IV → resolved after anesthesia team replaced with IV in forearm
- Patient requested labor epidural → uneventful placement without complication or aggravation of HNPP
- Outcome: vaginal delivery and discharged on post-delivery day 2

# Learning Points



- Although relatively rare, HNPP can complicate a parturient's course and requires communication and collaboration of care team
- **Strategies to minimize exacerbation include:**
  - Redundant padding to minimize compression or pressure injuries
  - Emphasize frequent position adjustments
  - Extend time between BP cuff inflations or reduce maximal cuff pressure; consider arterial line placement
  - Avoid use of SCDs to avoid nerve injury
- Neuraxial anesthesia is not contraindicated in these patients, but they should be counseled that **risk of HNPP flare is unknown**