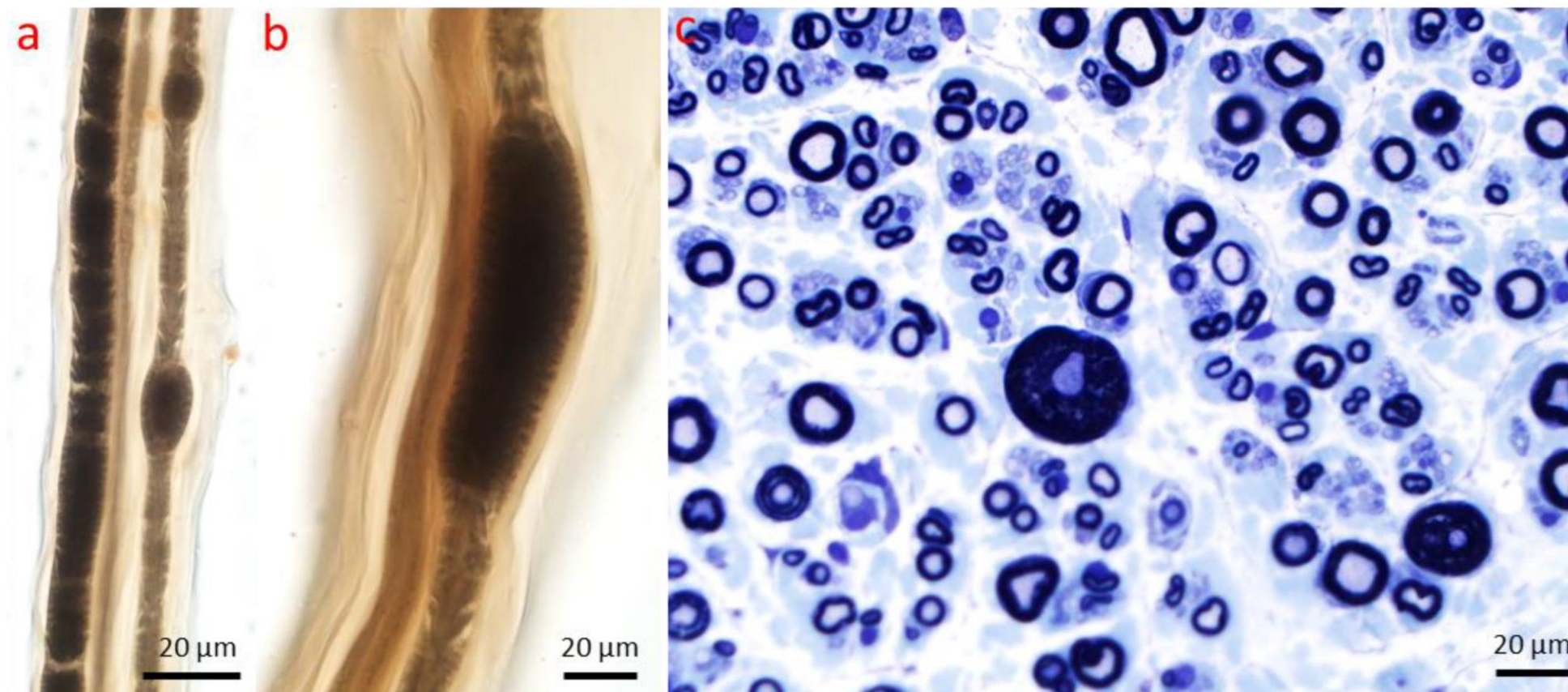


# Peripartum Anesthetic Considerations and Management in a Patient with Hereditary Neuropathy with Liability to Pressure Palsies (HNPP)

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## BACKGROUND

- Deletion of peripheral myelin protein-22
- AD with 70-100% penetrance
- Development of myelin sheath swelling (tomacula)
- Prevalence of 7-16 per 100,000
- Onset at 20-40 years old
- Classically presents with painless recurrent, transient, focal mononeuropathies after *minor* pressure, stretch, or sheer injury
- Areas of nerve entrapment particularly vulnerable
- Recovery in 50% of cases within days or weeks



**Figure 1.** Myelinopathy. Tomacula. (a,b) Teased nerve fiber preparation shows focal thickenings of the myelin membrane (magnification: (a) 60× oil immersion; (b) 100× oil immersion). (c) Sural nerve biopsy displaying fibers with abnormally thick myelin sheath relative to axon diameter (magnification: 100× oil immersion). Schenone et al. 2025.

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## Patient

- 34 year old G1P0000 with HNPP
- Right ulnar motor neuropathy diagnosed at 11 years old

## Pregnancy

- Diffuse numbness, tingling, weakness with right foot drop
- GDM, hypothyroid, symptomatic hypokalemia, RUE DVT

## Pre-Op

- OB pre-anesthesia visit at 32 and 34 weeks
- Presented at 36 weeks and 2 days for urgent CS due to severe IUGR and NRFHT

## Intraop

- Lumbar spinal anesthesia
- Standard ASA monitors with a **non-invasive continuous blood pressure device**
- Meticulous pressure point padding

## Post-Op

- PONV and hypoglycemia with resolution after PO intake
- No new or exacerbated neuropathies





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## TEACHING POINTS

- Multidisciplinary and individualized approach
- GA and NA both safe\*
- Shared decision-making
- Caution with NMB
- Meticulous attention to positioning
- NIBP monitoring with Acumen cuff
- Thorough pre/post-anesthesia neurologic evaluation

## REFERENCES

1. Laudansky, K et al. Anesthetic Considerations for Patients with Hereditary Neuropathy with Liability to Pressure Palsies: A Narrative Review. Healthcare 2024; 12(8):858.
2. Attarian, S et al. Hereditary Neuropathy with Liability to Pressure Palsies. Journal of Neurology 2020; 267:2198-2206.

