

Peripheral Nerve Blocks in Pediatric Charcot-Marie-Tooth Patients: a Case Series and Expert Positions

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Background

- **Charcot-Marie-Tooth (CMT)** disease is an inherited neuromuscular disorder affecting 1:2,500 people worldwide and is the most common genetic cause of neuropathy^{1,2}
- There is a paucity of short- and long-term outcome data in the CMT population following regional anesthesia (RA), particularly in the pediatric population³
- We aim to share **retrospective data and expert opinions** regarding RA practices at our institution for **pediatric patients with CMT**

Methods

- Following IRB approval, **patients with CMT who had surgery** between 2014 and 2023 were identified.
- Data was abstracted from the electronic medical record including demographics, **surgery type**, and whether **RA was performed**.
- A **survey of all pediatric regional anesthesiologists** at our institution was conducted to better understand variability in practices regarding regional anesthesia for CMT patients.

Results

Race/Ethnicity, n/total N (%)	
White	13/25 (52)
Hispanic/Latino	8/25 (32)
Black	1/25 (4)
Asian	3/25 (12)
Female, n/total N (%)	
	9/25 (36)
Age (yr) at 1st Surgery, median (range)	
	12 (2mo-18yr)
Number of Surgeries Per Patient, median (range)	
	1 (1-10)
Surgery with PNB, n/total N (%)	
	20/61 (33)

Table 1: Demographic Characteristics

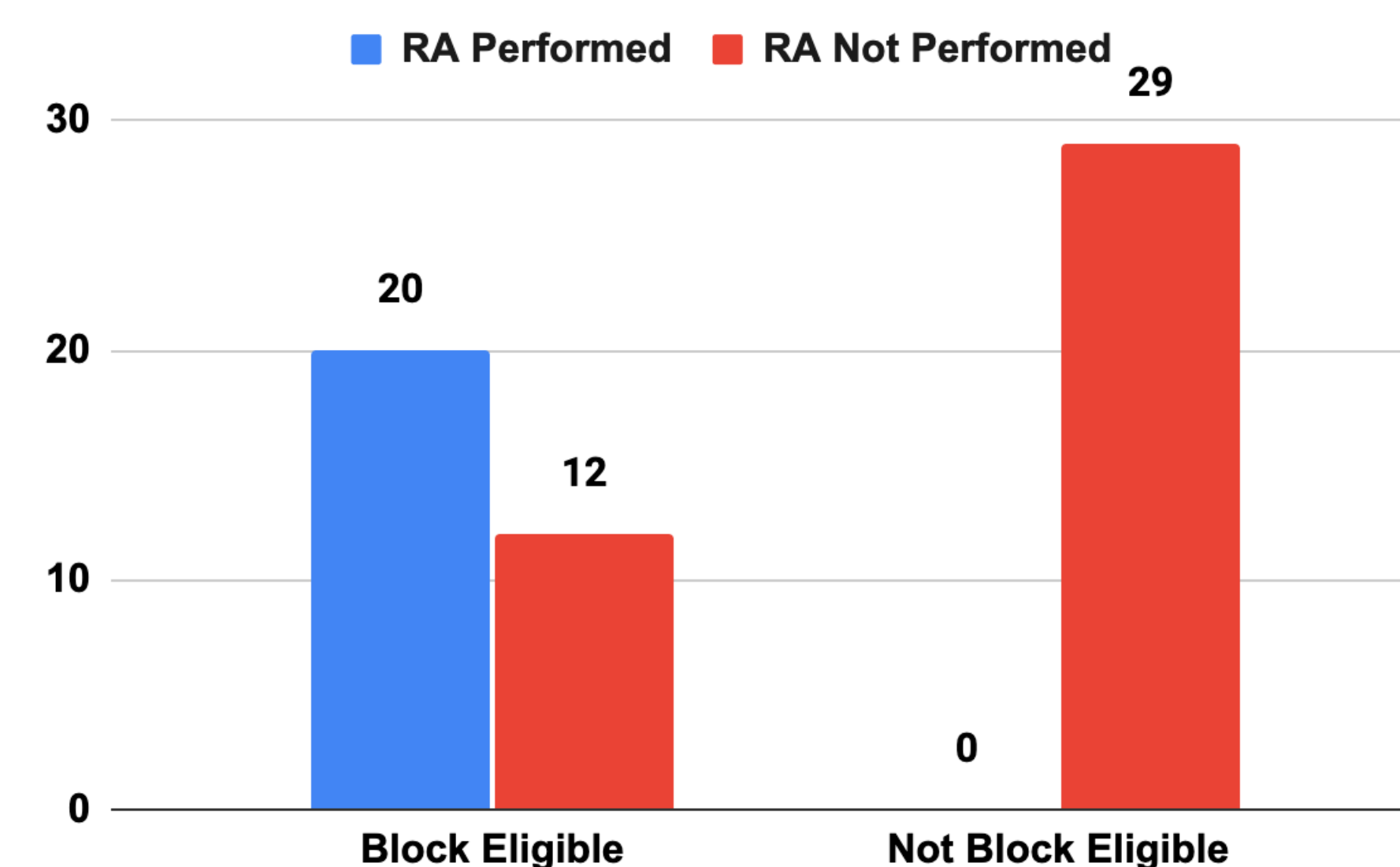


Figure 1: Regional anesthetic use by surgical subtype. Block eligibility determined by whether a block would be offered for a given procedure if the patient did not have CMT.

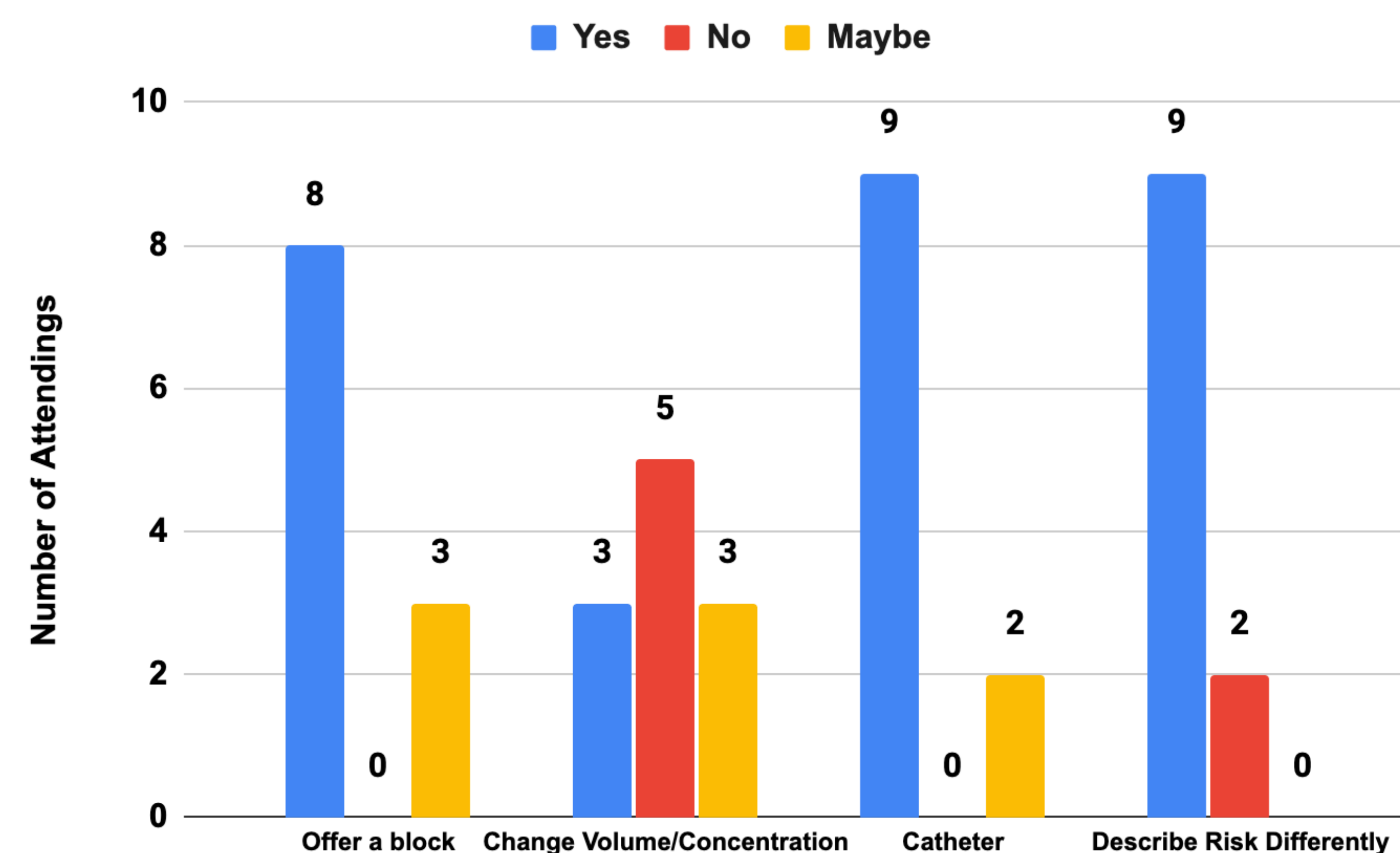


Figure 2: Pediatric regional anesthesiologist expert opinion regarding the following questions: would you offer a block, would you change the volume and/or concentration of the local anesthetic, would you leave a catheter if procedurally indicated, and would you describe the risk differently to the patient/family.

Discussion

- On chart review, **no evidence of nerve injury** in the 20 CMT patients who received blocks.
- **Practice variability remains**, related to historical teaching, lack of large studies, and anecdotal evidence
- **Risks of regional vs general anesthesia** should be weighed.
- Consensus among our institution's RA attendings and pediatric orthopedic surgeons to still **consider RA in CMT patients**.

Conclusion

- **Need for more data** in pediatric and adult populations with CMT.
- **RA in CMT patients considered safe** with low risk of neuropathy by our institutional **expert anesthesia and surgical opinions**.
- We encourage patient- and family-centered **shared decision making** regarding the risks, benefits, and alternatives.

References

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