Early Recurrent Carpal Tunnel Syndrome in Patients with Mucopolysaccharidoses

Keon Min Park, MD
Chau Tai, MD
Paymon Rahgozar, MD

Summary: Early-onset carpal tunnel syndrome (CTS) is a well-known manifestation of mucopolysaccharidoses (MPS) due to excessive deposition of glycosaminoglycans in soft tissues. Standard treatment has been carpal tunnel release surgery, with the conventional technique of dividing the transverse carpal ligament. With advancement of treatments for MPS, these patients now have a longer life expectancy and are presenting with recurrent CTS. Management of recurrent CTS in these patients is not well studied. Here, we report 2 cases of recurrent CTS in MPS patients after a carpal tunnel release operation. We describe the findings on repeat operations and propose a unique technique for treating CTS in MPS patients to minimize recurrence during the initial CTS surgery. Our method involves resection of a portion of the transverse carpal ligament and use of a hypothenar fat pad flap over the median nerve. (Plast Reconstr Surg Glob Open 2021;9:e3505; doi: 10.1097/GOX.0000000000003505; Published online 18 March 2021.)

Mucopolysaccharidoses (MPS) manifest with various symptoms, including skeletal dysplasia, cardiac abnormalities, and central and peripheral neuropathy, including carpal tunnel syndrome (CTS). Up to 58% of pediatric CTS cases may be due to MPS, and many hospitals regularly screen for CTS from an early age. Furthermore, up to 14% of patients experience recurrence after the initial operation, with the timing varying from 1 to 11 years.

In our study, we present 2 patients who had undergone carpal tunnel release earlier in life and presented with recurrence of CTS after 4 and 8 years. We suggest an operative strategy to reduce the risk of future recurrence.

CASE REPORTS

Patient A

A 13-year-old girl with MPS type I presented with bilateral hand numbness for 3 months. Previously, she had bilateral carpal tunnel release surgery at age 9. During the initial surgery, the transverse carpal ligament (TCL) was found to be 4-mm thick and was sharply bisected. Postoperatively, the patient had unrestricted function for 4 years, but CTS symptoms re-developed. EMG showed severe median neuropathy bilaterally at the level of the wrist. We proceeded with a repeat but modified bilateral carpal tunnel release. Again, 5-mm-thick TCL was found in each upper extremity and was fused back together (Fig. 1A). The TCL was divided, and in order to prevent recurrence of TCL fusion, 3 mm of the cut edges on both sides was excised with precautions to protect the recurrent branch of the median nerve (Fig. 1B). Then, we elevated a previously described hypothenar fat pad flap to create a vascularized barrier between the 2 cut ends of the TCL, preventing their fusion and to provide additional protection to minimize scarring over the median nerve. The skin over the flap was raised and tethering bands were released, allowing the vascularized fat pad to cover the median nerve (Fig. 1C).

At 9-month follow-up, the patient endorsed resolution of carpal tunnel symptoms and increased grip strength with good thenar muscle group bulk. (See figure, Supplemental Digital Content 1, which displays patient A at 9-month follow-up with good thenar muscle group bulk. http://links.lww.com/PRSGO/B609.) EMG showed that bilateral median nerve conduction had markedly improved with her right median nerve study within normal limits. (See appendix, Supplemental Digital Content 2, which displays preoperative and postoperative EGM reports of patient A. http://links.lww.com/PRSGO/B610.)
Patient B

A 24-year-old man with MPS type VI with a history of bilateral carpal tunnel release surgery at the age of 16 presented with bilateral hand numbness. After the initial surgery, he had temporary resolution of CTS symptoms. However, bilateral hand numbness began to recur over several years, and EMG of his upper extremities confirmed severe median neuropathy bilaterally. The patient underwent the modified carpal tunnel release surgery described above. Bilateral TCLs were found to be thick at 5 mm.

At 10-month follow-up, the patient denied CTS symptoms and had mildly improved grip strength. His repeat EMG showed similar severe bilateral median nerve neuropathies.

**DISCUSSION**

CTS is a well-known finding in MPS patients, but recurrent CTS after surgical release had not been a frequent concern until advances in management of MPS that have extended patients’ life expectancy. A recent study reported pediatric patients with metabolic disease who are experiencing recurrent CTS. The mean age of initial diagnosis of CTS in MPS patients is 3–11 years. The current literature supports early surgical management for MPS patients and does not recommend conservative management because delayed surgeries have correlated with a poor outcome. However, surgeries expose this patient population to the risks of general anesthesia, which are compounded by difficult airways and other medical comorbidities. Due to their young age and common developmental delays, IV sedation or local/regional anesthesia is often not an option, and emergent intubation can be highly dangerous in these patients.

At our medical center, only 1 anesthesiologist can manage these patients intraoperatively given their difficult airway and unstable C-spine, as well as many comorbidities. Both patients were seen by a cardiologist, a pulmonologist, and an endocrinologist before surgery due to their complex medical conditions. Intraoperative neuromonitoring was utilized to verify neurologic integrity during the surgery. To minimize the time under anesthesia, we used a 2-team approach with 1 attending surgeon operating on each extremity simultaneously. Given the difficulties in managing these patients intraoperatively, it is crucial to effectively manage CTS during the initial treatment in this specific patient population.

In our study, we present 2 cases of recurrent CTS in MPS patients. To our knowledge, we are the first group to suggest a unique combination of surgical techniques in treating this particular group of patients to prevent multiple surgeries. We recommend excising a portion of TCL rather than performing a simple release, and to place a hypothenar fat pad flap over the median nerve during the initial surgery. Resection of TCL provides further decompression and a larger space for median nerve in case of further glycosaminoglycans deposition in the TCL, which is thought to be the etiology of CTS in MPS patients. It may also decrease the likelihood of repeat scarring, thus recurrence of CTS. The hypothenar fat pad flap has demonstrated effectiveness in preventing CTS recurrence because the flap prevents adherence and scarring to the median nerve. Thus, the combination of resection of TCL and hypothenar flap during the initial surgery can significantly lower the risk of recurrent CTS.

Although EMG study of our second patient did not show improvement, likely because of the chronicity of his neuropathy leading to irreversible damage, the patient reported relief of symptoms. Multiple EMG studies over several years before his second surgery showed severe CTS, but the surgery was delayed due to patient preference. Both patients had flexion contracture release on multiple digits due to progression of chronic trigger fingers, and postoperative hand therapy and serial splinting were recommended. However, for simple CTS surgery, we recommend 1–2 weeks of wrist splint to protect the surgical site. Limitations of our study included short length of follow-up and small number of patients. Additional studies are needed to show long-term effectiveness for a larger patient group.

**CONCLUSIONS**

Our method of TCL resection with hypothenar fat pad flap has shown promise in effectiveness over short-term follow-up in recurrent CTS. We recommend using our method as the initial treatment to minimize recurrent symptoms in high-risk MPS patients and the need for a repeat operation and potential risks of anesthesia.

**Keon Min Park, MD**
Division of Plastic and Reconstructive Surgery
Department of Surgery
University of California San Francisco
San Francisco, CA 94143
E-mail: keon.park@ucsf.edu
ACKNOWLEDGMENTS

No IRB was obtained given the nature of case reports. All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975.

REFERENCES

1. Galimberti C, Madeo A, Di Rocco M, et al. Mucopolysaccharidoses: early diagnostic signs in infants and children. Ital J Pediatr. 2018;44(Suppl 2).
2. Van Meir N, De Smet L. Carpal tunnel syndrome in children. J Pediatr Orthop B. 2005;14:42–45.
3. Patel P, Antoniou G, Clark D, et al. Screening for carpal tunnel syndrome in patients with mucopolysaccharidosis. J Child Neurol. 2020;35:410–417.
4. Dabaj I, Gitiaux C, Avila-Smirnow D, et al. Diagnosis and management of carpal tunnel syndrome in children with mucopolysaccharidosis: a 10 year experience. Diagnostics (Basel). 2019;10.
5. Jadhav T, Kornberg AJ, Peters H, et al. Carpal tunnel syndrome in pediatric mucopolysaccharidoses. Clinical Neurophysiology. 2014;125:e4–e5.
6. Strickland JW, Idler RS, Lourie GM, et al. The hypothenar fat pad flap for management of recalcitrant carpal tunnel syndrome. J Hand Surg Am. 1996;21:840–848.
7. Karthik K, Nanda R, Stothard J. Recurrent carpal tunnel syndrome—analysis of the impact of patient personality in altering functional outcome following a vascularised hypothenar fat pad flap surgery. J Hand Microsurg. 2012;4:1–6.
8. Velicki K, Goldfarb CA, Roberts S, et al. Outcomes of pediatric and adolescent carpal tunnel release. J Hand Surg Am. 2020.
9. Moretto A, Bosatra MG, Marchesini L, et al. Anesthesiological risks in mucopolysaccharidoses. Ital J Pediatr. 2018;44(Suppl 2).