Simultaneous Opposition Tendon Transfer with Median Nerve Decompression for Severe Bilateral Carpal Tunnel Syndrome in Adolescents with Hunter Syndrome: A Case Report

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Summary: Although carpal tunnel syndrome (CTS) is exceedingly rare in children, its prevalence in those with Hunter syndrome, mucopolysaccharidosis type II, is high. With the advent of hematopoietic stem cell transplantation and enzyme replacement therapy, the survival of patients with Hunter syndrome has dramatically improved. In addition, with the advent of hematopoietic stem cell transplantation and enzyme replacement therapy, the survival of patients with Hunter syndrome has dramatically improved. In patients with Hunter syndrome, the severity of CTS progresses with age.1,3 However, the diagnosis and subsequent treatment of CTS are often delayed due to the presence of atypical symptoms and cognitive impairment.1,3 In addition, with the advent of hematopoietic stem cell transplantation and enzyme replacement therapy, the survival of patients with Hunter syndrome has dramatically improved.5 Thus, the number of delayed and advanced presentations of CTS in older adolescents with Hunter syndrome have been increasing.1,5 So far, no studies have reported the simultaneous opposition tendon transfer with OCTR for severe CTS in adolescents with Hunter syndrome. Here, we present a mid-term follow-up of a 16-year-old patient with Hunter syndrome associated with severe bilateral CTS successfully treated by the simultaneous opposition tendon transfer with an OCTR to improve the thumb function. Intraoperatively, the median nerve was constricted and flattened with congestion by the transverse carpal ligament. External and internal neurolysis of the scarred median nerve were performed and found epineural fibrosis and tethered epineurium. An intraneural lipoma of the left median nerve was especially resected with epineurectomy. During neurolysis and tendon transfer, the soft tissue was very viscous, a characteristic of mucopolysaccharidoses. Transferring the tension of the palmaris longus tendon to the abductor pollicis brevis for the thumb palmar abduction should be stronger than routine adult patients because the soft tissue such as the tendon excursion is stickier and more contracted in patients with Hunter syndrome. Postoperatively, a thumb spica splint was applied for 3 weeks, and then active motion exercises were cautiously started to prevent joint contracture. Early recognition and surgical intervention for CTS are essential in patients with Hunter syndrome. (Plast Reconstr Surg Glob Open 2020;8:e3251; doi: 10.1097/GOX.0000000000003251; Published online 30 November 2020.)

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CASE REPORT

The patient was diagnosed with mucopolysaccharidosis type II at 2 years of age and started on enzyme replacement therapy with human iduronate-2-sulfatase, Elaprase, at 6 years of age. At 16 years of age, he was referred to our hand surgery clinic by his pediatrician due to suspected bilateral CTS with impaired thumb function, particularly pinching something small, from severe abductor pollicis brevis (APB) muscle atrophy (Fig. 1). He had a gargoyle-like face and obstructive sleep apnea without mental retardation. A nerve conduction study showed that both the compound muscle action potential (CMAP) of the APB muscle and the sensory nerve action potential (SNAP) were non-recordable. First, simultaneous Camitz opposition tendon transfer with OCTR for the right hand was performed under tourniquet control, and the second for the left hand at 12 months after the first surgery. Intraoperatively, the median nerve was constricted and flattened with congestion by the transverse carpal ligament. The transverse carpal ligament was released. An external neurolysis of the scarred median nerve, which was adherent to surrounding tissues, was performed circumferentially. As epineural fibrosis and tethered epineurium were found, an internal neurolysis with epineurotomy was performed to release the constriction. As an intraneural lipoma of the median nerve was found in the left hand, an intraneural lipoma excision and internal neurolysis with epineurotomy were performed under a microscope (Fig. 2). Pathological findings of the internal lipoma were fibrolipoma-like fibromatous hamartoma without depositing glycosaminoglycans. Next, transferring the palmaris longus tendon to the APB was performed to improve the thumb function. During neurolysis and tendon transfer, the soft tissue was very viscous, a characteristic of mucopolysaccharidoses.

Fig. 1. Preoperative appearance of impaired palmar abduction of both thumbs.

Fig. 2. Intraoperative findings. The left median nerve with an intraneural lipoma (A) after excision of the intraneural lipoma and neurolysis (B).
Hence, it was necessary during the tendon transfer to keep maximum tension with the wrist 30 degrees flexed and the thumb in maximum palmar abduction. This is a stronger tension than that used for the routine Camitz procedure in adults with CTS. Postoperatively, a thumb spica splint was applied for 3 weeks. Following this, active progressive motion exercises were initiated, as Hunter syndrome patients are prone to development of joint contractures. Weight bearing and power grip were restricted until 12 weeks postoperatively. The strength of the patient’s grip, tip pinch, and key pinch in both hands had increased 4 years postoperatively. Sensory function assessed with Semmes–Weinstein monofilament test and 2-point discrimination test had also improved. The patient-reported outcome scores from the Quick Disability of the Arm, Shoulder, and Hand improved from 45.4 preoperatively to 9.1 postoperatively and those of the hand from 54.5 to 12.5. Although the CMAP of the right median nerve remained non-recordable at 4 years postoperatively, the distal motor latency of the left median nerve became recordable (6.0 ms) 3 years postoperatively. The patient has been continuing weekly infusions of Elaprase with improved function in both hands (Fig. 3).

**DISCUSSION**

CTS starts in very early childhood in patients with Hunter syndrome, as early as 26 months; however, its diagnosis may be difficult and delayed because of atypical clinical symptoms as well as developmental delays, difficulty with communication, and an inability to express symptoms. Moreover, its severity progresses with age. The CMAP and SNAP amplitudes of the median nerves decreased with age. Because of long-standing nerve compression, thenar muscle atrophy would persist in patients with Hunter syndrome, as was found in the present case. In severe CTS with thenar muscle atrophy, pinch strength and activities of daily living improved to a greater extent with simultaneous opposition transfer when compared with the OCTR alone.

In this case, unilateral lipofibromatous hamartoma of the median nerve was accompanied with CTS. Numerous cases of lipofibromatous hamartoma have been reported to involve the median nerve. However, lipofibromatous
hamartoma, without deposition of glycosaminoglycans, associated with mucopolysaccharidosis has not yet been reported. Although OCTR is the mainstay management, no clear guideline has been established for the treatment of lipofibromatous hamartoma that involves a wide range of options from close observation to complete nerve resection. Agarwal and Haase recommended epineurotomy and intraneural dissection with intraneural lipoma resection if patients have worsening nerve function such as in this case.8

CONCLUSIONS
Simultaneous opposition tendon transfer with median nerve decompression was more effective in improving fine motor function of the thumb, even in longer surviving adolescents with Hunter syndrome with severe CTS due to enzyme replacement therapy. The tension of the transferred palmaris longus tendon to the APB intraoperatively should be stronger because the soft tissue such as the tendon excursion is stickier and more contracted in patients with Hunter syndrome. Lipofibromatous hamartoma in the median nerve for patients with Hunter syndrome was successfully treated with interfascicular excision of the intraneurial lipoma with epineurotomy.

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