Restoration of Prehensile Function for Motor Paralysis in Hopkins Syndrome: Case Report

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Hopkins syndrome is a rare cause of poliomyelitis-like paralysis affecting 1 or more extremities after an acute attack of asthma. The exact etiology of Hopkins syndrome is not known. A 4-year-old girl developed acute asthma followed by complete flaccid paralysis of the left upper extremity. She underwent staged reconstruction using the double free muscle transfer technique. Rigorous postoperative physiotherapy was carried out to achieve a good functional outcome. At recent follow-up, 27 months after the first procedure, the patient was able to effectively use the reconstructed hand for most daily activities. She had good control and could perform 2-handed activities. The selection of a suitable operative treatment and suitable donor nerves is critical, and there are no clear guidelines in the literature. The double free muscle transfer can be effectively employed in similar cases to restore grasping function. *(J Hand Surg Am. 2014;39(2):312−316. Copyright © 2014 by the American Society for Surgery of the Hand. All rights reserved.)*

Key words Brachial plexus palsy, double free muscle transfer, Hopkins syndrome, poliomyelitis-like paralysis, postasthmatic amyotrophy.

Hopkins[1] reported a syndrome in 1974 characterized by sensory-sparing poliomyelitis-like motor paralysis of 1 or more extremities after an acute attack of bronchial asthma.[1−5] The etiology of Hopkins syndrome is unknown. The pathology likely occurs at the level of the anterior horn cell or ventral motor root.[2,4] An immunologic cause, induced by a viral infection or activated in an immunosuppressed state, or an allergic mechanism may trigger an acute asthmatic attack or cause direct anterior horn cell damage.[5]

All reported patients had a mean age of 5 to 6 years (range, 13 mo to 13 y) and had a history of an acute attack of bronchial asthma. The resulting paralysis was not related to the severity of the attack.[1−5] The mean duration of onset of paralysis is 6 days (range, 1−18 d) after the start of the asthma attack. The weakness evolves rapidly in hours to days and results in total or near-total paralysis. The weakness is usually localized to 1 extremity, although some patients can have patchy weakness. A few patients may have minimal spontaneous recovery. The residual weakness is severe and disabling.

We restored prehensile hand function in a patient with Hopkins syndrome by double free muscle transfer (DFMT). We discuss our surgical technique and technical considerations in relation to Hopkins syndrome.

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CASE REPORT

A 4-year-old girl presented with complete loss of movement of the left upper extremity. She had a history of bronchial asthma since age 1 year. She had had a normal birth and developmental and immunization history. Three months before presentation, she had a severe attack of wheezing and respiratory distress. She was treated with intravenous medications and was discharged after 4 days. Three days

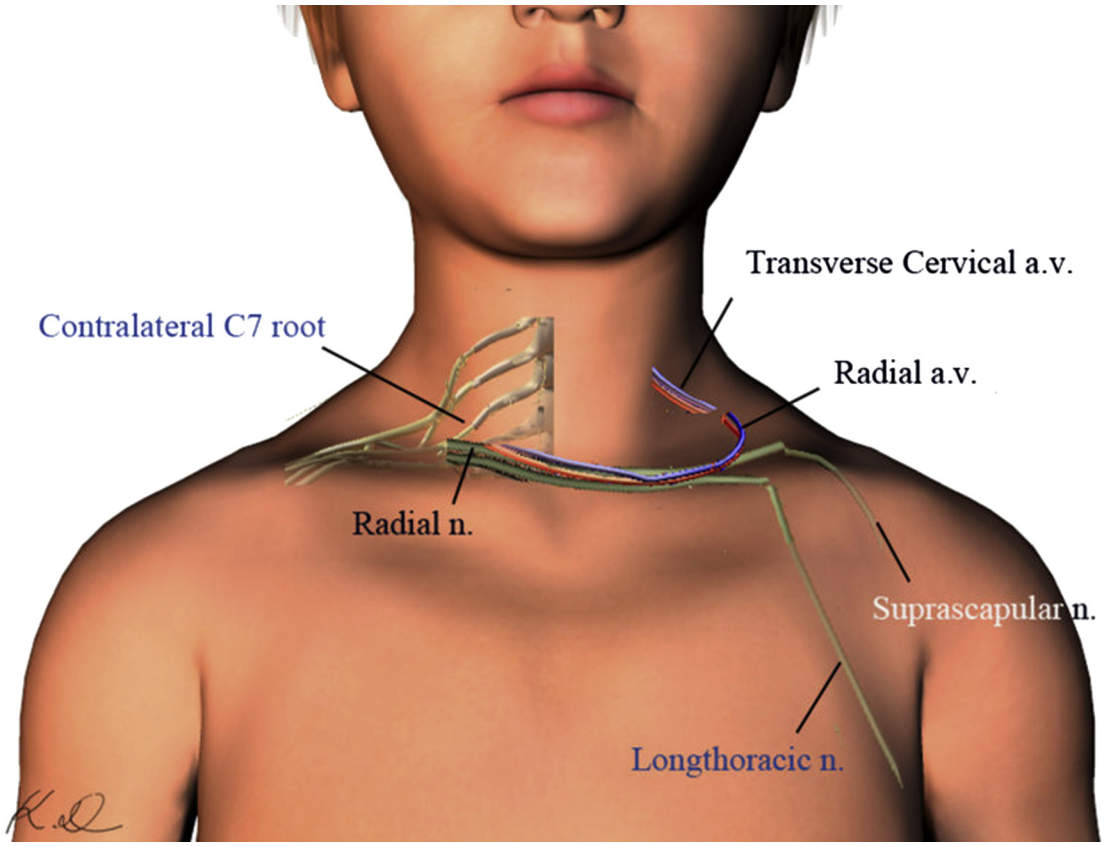

FIGURE 1: Stage 1 of the DFMT–contralateral C7 transfer for restoration of shoulder function. A vascularized radial nerve (n.) graft is used to connect the C7 root to the suprascapular and long thoracic nerve. The radial artery and vein (radial a.v.) were anastomosed to the transverse cervical artery and vein (transverse cervical a.v.).

later, her mother noted complete loss of movement of the child's left upper limb. The biceps, triceps, and brachioradialis tendon reflexes were absent. Sensory examination was normal. She was conscious and alert, and had no neck stiffness or pain. Blood tests revealed a rise in eosinophil count (9%) and immunoglobulin E levels (1,522 IU/mL). Cerebrospinal fluid examination showed mild pleocytosis and raised protein levels. Viral studies were negative. Magnetic resonance imaging displayed increased T2 signal intensity of the anterior horn cell region from C3 to C7 disk space. Hopkins syndrome was diagnosed, and she was treated with steroids and gamma globulins, but there was no improvement in the paralysis.

Clinical examination showed total paralysis of the left upper extremity. We counselled the parents and discussed possible treatment options. We decided to observe the child for a few months and assess any spontaneous recovery. After 6 months, there was no sign of motor recovery. Hence, we decided to perform DFMT to restore upper limb function.

We explored the left brachial plexus (stage 1) and confirmed the diagnosis of a total preganglionic injury. In the absence of suitable ipsilateral donors, we decided to use the contralateral C7 root for shoulder reinnervation. The radial nerve was harvested from the left forearm along with the radial artery and vena comitantes. It was used as a free vascularized nerve graft between the contralateral C7 root and the suprascapular nerve and long thoracic nerve. The vascular pedicle was anastomosed with right transverse cervical vessels (Fig. 1).

Four months after stage 1, we performed a gracilis free functioning muscle transfer (FFMT) to restore finger flexion (Fig. 2). Simultaneously, the third and fourth intercostal nerves were transferred to the motor branch of the triceps. At 2.5 months later, we performed another gracilis FFMT to restore elbow flexion and finger extension (Fig. 3).

Postoperative management

After each muscle transfer, the upper limb was immobilized with the shoulder in 30° abduction and flexion and 60° internal rotation. The elbow was in 100° flexion, the wrist in neutral, and the fingers in full flexion or extension for 8 weeks. Early passive mobilization of elbow, wrist, and fingers was started 1 week after FFMT. Electromyographic biofeedback