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Case report

Flexor digitorum superficialis opposition tendon transfer improves hand function in children with Charcot-Marie-Tooth disease: Case series

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Abstract

Charcot-Marie-Tooth disease limits hand function. Tendon transfer has not been reported in pediatric CMT. We report two severely affected children following long finger flexor digitorum superficialis opposition tendon transfer. Improvement was noted in palmar abduction, (30°/40°), opposition, (thumb to all digits), and acquisition of pincer, palmar, and lateral pinch with measureable force (1 lb). Dexterity testing improved on the 9 Hole Peg Test (1.03 s/77 s, 22 s) and Functional Dexterity Test (13 s/33 s, 88 s). Functional improvements were observed in self feeding, clothing management, and play. These cases support flexor digitorum superficialis opposition tendon transfer surgery to improve hand function in children with CMT.

Keywords: Charcot Marie Tooth; Hand function; Tendon transfer; Children; Hand surgery

1. Introduction

Charcot-Marie-Tooth disease (CMT) is a genetically based progressive peripheral neuropathy often beginning in childhood. The main features are length-dependent impairment of motor (weakness, muscle atrophy) and sensory (reduced sensation) function resulting from either a primary axonal or demyelinating neuropathy [1]. These impairments result in functional limitations in self-care, work, and leisure tasks requiring hand strength, stability, and coordination.

Reduced thumb mobility and intrinsic finger function limit prehension in pediatric patients with CMT, resulting in reliance on compensatory grasp patterns: gross raking to retrieve small items (coins, keys, utensils), palm to palm for larger items (bottled water, glass), scissoring between fingers (pencil, food), or modified lateral pinch to tear open snack packaging/containers.

Tendon transfers can improve motor function for patients with diminished hand function due to nerve injury [2,3] and there are reports of hand surgery in adults with CMT [4–6], however literature supporting surgical management in pediatric CMT wasn't identified. We evaluated the effectiveness of Flexor Digitorum Superficialis (FDS) Opposition tendon transfer to restore thumb mobility and enhance hand function in children with CMT.

2. Materials and methods

2.1. Patient selection

We reviewed the cases of two patients that received Long Finger Flexor Digitorum Superficialis Opposition Tendon Transfer to improve hand function.

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2.2. Patient classification

Level of impairment was classified based on the patients' upper extremity sensory, motor, and functional limitations as measured by the Overall Neuropathy Limitations Scale (ONLS), Charcot-Marie-Tooth Neuropathy Score (CMTNS), and the Total Neuropathy Score (TNS).

healthy controls [11,12] and adults with CMT [13]. In order to obtain accurate and reliable data, a modified FDT (1/2 board – 8 pegs vs. 16) was completed for Case 2 – Left hand to prevent frustration and maintain compliance, as this was an extremely challenging task. A penalty assessed time [11] is also documented for Case 2 to offer additional data capturing quality/efficiency of movement.

Case 1

Outcome Measure	Pre-Operative	Post-Operative 11 Months
Gross Grasp	4 pounds	2 pounds
Lateral Pinch	Unable	1 pound
Palmar Pinch	Unable	1 pound
Tip Pinch	Unable	0 pounds
9 Hole Peg Test	40.53 seconds	39.50 seconds
Functional Dexterity Test	95 seconds	82 seconds
Palmar Abduction	None	30 degrees
Radial Abduction	30 degrees	40 degrees
Opposition	None	All but Small Finger

Case 2 – Left Hand

Outcome Measure	Pre-Operative	Post-Operative 19 Months
Gross Grasp	10 pounds	7.5 pounds
Lateral Pinch	Unable	1 pounds
Palmar Pinch	Unable	1 pounds
Tip Pinch	Unable	1 pound
9 Hole Peg Test	127 seconds	50.37 seconds
½ Functional Dexterity Test	72 seconds	38.68 seconds
½ Functional Dexterity Test with penalty	192 seconds	58.68 seconds
Palmar Abduction	None	40 degrees
Radial Abduction	15 degrees	35 degrees
Opposition	None	All digits

Case 2 – Right Hand

Outcome Measure	Pre-Operative	Post-Operative 9 Months
Gross Grasp	5 pounds	6.5 pounds
Lateral Pinch	Unable	1 pound
Palmar Pinch	Unable	0 pounds
Tip Pinch	Unable	0 pounds
9 Hole Peg Test	68.9 seconds	46.84 seconds
Functional Dexterity Test	96 seconds	88.88 seconds
Functional Dexterity Test with penalty	336 seconds	238 seconds
Palmar Abduction	None	40 degrees
Radial Abduction	15 degrees	35 degrees
Opposition	None	All digits

2.3. Assessments

Upper extremity measures from the CMT Pediatric Scale (CMTPedS) [7], Goniometry, clinical observations, and patient reported measures were completed. The CMTPedS is a multidimensional, age-appropriate, quantitative scale designed to measure disease severity in children with CMT. The CMTPeds upper extremity measures include hand strength and dexterity assessments as described below. Patient's were evaluated pre and post-operatively.

Hand Strength evaluation included Gross, Lateral, Palmar, and Tip prehension force measured using a Jamar Dynamometer/pinchometer, which reliably measure muscle strength [8–10]. Range of Motion for thumb abduction (radial/palmar) was completed using a standard finger goniometer and opposition measured by the ability to oppose the digits.

Hand Dexterity was assessed using the Functional Dexterity Test (FDT), a measure of hand dexterity that provides information regarding use of the hand for daily tasks (buttoning, tying shoes laces, screwing a nut and bolt) requiring manipulation, accuracy, and palmar prehension. The FDT demonstrates excellent validity and reliability in

The Nine-Hole Peg Test was assessed to examine dexterity and hand/eye coordination as a measure of hand function, and has demonstrated reliability and validity in healthy children and adults with CMT [14–16].

Activities of Daily Living–Self Feeding, clothing management, and leisure activities were evaluated by observation/interview. Level of independence and compensatory techniques observed/reported were recorded.

Patient Satisfaction was measured using a 10 point Likert Scale (zero being highly dissatisfied and 10 being highly satisfied).

3. Case report 1

A 6 year old girl with CMT Type 2A, MFN2 gene sequencing showed a heterozygous pathogenic missense mutation c.1090C>T p.Arg364Trp, presented with limited thumb mobility, clawing, hand weakness, wasting of the hand (Fig. 1a and b), and impaired dexterity. She was unable to palmarly abduct or oppose her thumb to any digits. She had decreased radial abduction (30°) of the thumb and clawing of all digits resulting in the inability to execute pincer grasp patterns. She relied on modified lateral pinch,