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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.

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).

Case 1			Case 2 – Left Hand			Case 2 – Right Hand		
Outcome Measure	Pre-Operative	Post-Operative 11 Months	Outcome Measure	Pre-Operative	Post-Operative 19 Months	Outcome Measure	Pre-Operative	Post-Operative 9 Months
Gross Grasp	4 pounds	2 pounds	Gross Grasp	10 pounds	7.5 pounds	Gross Grasp	5 pounds	6.5 pounds
Lateral Pinch	Unable	1 pound	Lateral Pinch	Unable	1 pound	Lateral Pinch	Unable	1 pound
Palmar Pinch	Unable	1 pound	Palmar Pinch	Unable	1 pound	Palmar Pinch	Unable	0 pounds
Tip Pinch	Unable	0 pounds	Tip Pinch	Unable	1 pound	Tip Pinch	Unable	0 pounds
9 Hole Peg Test	40.53 seconds	30.50 seconds	9 Hole Peg Test	127 seconds	50.37 seconds	9 Hole Peg Test	68.9 seconds	46.84 seconds
Functional Dexterity Test	95 seconds	82 seconds	1/2 Functional Dexterity Test	72 seconds	38.68 seconds	Functional Dexterity Test	96 seconds	88.88 seconds
Palmar Abduction	None	30 degrees	1/2 Functional Dexterity Test with penalty	192 seconds	58.68 seconds	Functional Dexterity Test with penalty	336 seconds	238 seconds
Radial Abduction	30 degrees	40 degrees	Palmar Abduction	None	40 degrees	Palmar Abduction	None	40 degrees
Opposition	None	All but Small Finger	Radial Abduction	15 degrees	35 degrees	Radial Abduction	15 degrees	35 degrees
			Opposition	None	All digits	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 CMT-Peds 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 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.

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, scissoring grasp between her index and middle fingers, and bilateral palm to palm grasp. Functional impairments included difficulty opening door knobs, grasping paper and playing cards, manipulating clothing fasteners, opening snack/drink containers, and using the computer mouse.

4. Case report 2

An 11 year old girl with clinical features and electrodiagnostic studies indicative of a length-dependent axonal sensorimotor polyneuropathy, with the clinical diagnosis of CMT type 2, presented with bilateral symmetric limited thumb mobility, decreased active digit extension, hand weakness, wasting of the hand and forearm, and impaired dexterity of both hands. No mutation was identified in testing PMP22 gene sequencing and deletion/duplication assay, sequencing of Cx32, MPZ, EGR2, NF-L, PRX, GDAP1, LITAF/SIMPLE, MFN2, SH3T2C, GARS, HSPB1, LMNA, RAB7 and DNM2 genes, and genome-wide 550 K SNP microarray. She had bilateral limitations as follows: no palmar abduction, 15° of radial abduction, and was unable to actively oppose her digits. She relied on compensatory grasp patterns including: gross raking grasp, modified lateral pinch, scissor grasp between her index and middle fingers, and a palm to palm grasp. She complained of difficulty managing clothing fasteners, opening containers, stabilizing objects, and handwriting fatigue. She reported limited left hand use during daily tasks.

5. Surgical intervention

Long finger Flexor Digitorum Superficialis (FDS) Opposition tendon transfers of the non-dominant, left hands were performed at ages 6 and 11 respectively. Case 2 underwent dominant, right long finger FDS tendon transfer at age 12. FDS strength testing was completed to select the optimal tendon for transfer. The tendon was cut at the base of the finger (Fig. 1c) and pulled into a separate volar forearm incision (Fig. 1d). The flexor carpi ulnaris (FCU) was used to create a FCU loop. One-half of the tendon was cut 2 cm proximal to the pisiform leaving its distal attachment. The proximal cut portion of the tendon was sutured to the distal attachment to form a loop. The FDS tendon was passed through the FCU loop (Fig. 1e) and advanced through a subcutaneous tunnel to the radial side of the thumb. The FDS tendon was then woven through the abductor tendon (Fig. 1f). Tension was adjusted until there was substantial thumb opposition with the wrist into extension. The patients were casted for 3 weeks in a long arm thumb spica cast with the elbow at 90°, the wrist in slight flexion, and the thumb in opposition.

6. Post operative therapeutic protocol

During immobilization (weeks 1–3), active range of motion, tendon glides, and DIP blocking exercises were performed on digits II through V to prevent adhesions at the donor site. Following cast removal, and initiation of the mobilization phase (weeks 3–6), a dorsal forearm based splint was fabricated with the wrist slightly flexed and thumb in opposition. The splint was worn continuously, except for bathing and exercise. Active movement, place and hold exercises, and tendon glides were completed to facilitate opposition and to prevent adhesions at the donor site. Functional retraining was initiated at week 6 with the splint worn only for resistive activities during the day and while sleeping at night. Functional grasp, manipulation, and use of the hand for ADL's, was then added to the current exercise regimen. Strengthening exercises were started at week 8 with light resistive activities progressing in resistance over the next 4 weeks, and day time splint wear was discontinued. At 12 weeks, the postoperative splint was discontinued completely, and therapy focused on maximizing hand function for ADL's.

7. Results

Case 1 was re-evaluated 11 months post-operatively and demonstrated increased palmar abduction (30°), radial abduction (40°), and opposition (able to oppose her index, middle, and ring fingers) enabling her to execute lateral, pincer, and palmar grasp. A 2 lb decrease in gross grasp strength was noted bilaterally. She attains proper positioning and generates force for lateral (1 lb), pincer (0 lb), and palmar (1 lb) grasp with her left hand. Dexterity improved by 1.03 s on the 9 Hole Peg test and 13 s on the FDT (Table 1). She is using her left hand in the dominant manipulative role and her right as the stabilizer for some bimanual tasks. New skills developed include; executing and maintaining a precise and secure grasp on the Zip lock bag pull tab and cover of her yogurt container to obtain/access food as well as on buttons/zippers during clothing management. She can grasp small snack foods more efficiently for self feeding, and pick up playing cards easier. The patient/parent reported 8/10 satisfaction with the surgery, and are requesting surgery on the dominant hand.

Case 2 was reevaluated at 19 months (left hand) and 9 months (right hand) postoperatively (Table 1). She demonstrates increased active palmar abduction (40°), radial abduction (35°), and opposition (Fig. 2a–b) (able to oppose index, middle, and ring fingers with pad to pad contact, and tip of her small finger), for both hands enabling execution of lateral, pincer (Fig. 2c–d), and palmar grasp. She demonstrates a 2.5 lb decrease in left gross grasp strength and a 1.5 lb increase in right gross grasp. She attains the standard position and generates force for lateral (1 lb) pincer (1 lb), and palmar (1 lb) grasp with her left hand. With her right hand she generates less than 1 lb of force for palmar and pincer grasp, while lateral pinch is 1 lb. Dexterity testing improved on the 9 Hole Peg Test by 77 s (left hand) and 22 s (right hand). However, her immediate preoperative trial (left hand) was unreliable as she was highly distractible and unmotivated. She showed a 42 s improvement compared to her previous 9 Hole Peg Test (6 months prior). On a modified FDT (1/2 board completed) she improved left hand raw time by 33.32 s and penalty assessed time by 133.32 s. Right hand completion of FDT (full board) improved by 8 s raw time and 98 s penalty assessed time (Table 1). She reports incorporating her left hand into bimanual tasks and using it interchangeably with the right. New skills developed since the surgery include; picking up and moving the computer mouse, turning pages in a book, using her left hand to assist with self-care tasks, improved grasp on utensils, and picking up small items off a table. She reports a 7/10 level of satisfaction following surgery.

8. Discussion

There is some concern regarding the viability of donor muscles to sustain function over time in CMT, as it's a progressive disease and patients often present with weakness/atrophy at evaluation. This shouldn't prevent patient referrals to a hand surgeon for evaluation of tendon transfer. Previous literature supports surgical management (tendon transfer, arthrodesies, and soft tissue releases) of adults with CMT with long standing (average onset of symptoms 10 years prior to surgery) [6] impairments and deformities [4–6]. The sustainability of functional gains following tendon transfer is described as “remaining functional over time” [4] and “achieved remarkable and sustained improvement” at follow up 18 years postoperatively [5]. However, surgical management in pediatric CMT hasn't been formally addressed previously. Here we evaluate the short term functional change seen in two children with CMT to assess the initial impact of the surgical intervention. A longer follow up will need to be completed to assess the sustainability of the effects we have observed.

We found that FDS opposition tendon transfer improves active thumb mobility, supports the acquisition of new grasp patterns, and facilitates increased hand function. Increased precision grasp strength was minimal; however, these new movement patterns may require longer follow up to assess the full impact of tendon transfer on strength. Improvements in active range of motion, strength, and grasp patterns were similar for both patients. Timed testing improved, with the non-dominant (surgical hand) surpassing the dominant hand on the 9 Hole Peg Test and FDT (Graph 1). However, the degree of change varied between patients. Case 2 had sufficient intrinsic digit function to complement the newly acquired thumb mobility, whereas Case 1 had poor intrinsic function, resulting in a lesser degree of improvement.

The FDT may be a more sensitive measure, in the authors' opinion, in CMT as it relies heavily on the combination of intrinsic digit function and thumb mobility and captures qualitative data via a penalty assessed time. As stated within the FDT guidelines, a penalty time of 10 s was added for a dropped peg and 5 s for compensatory movements used (supination past neutral, touching of the peg to the board, chest, opposite hand, etc.). Improvement in raw time suggests better fine motor dexterity; however the ability to capture a qualitative change in movement pattern via penalty assessed time may offer a better indicator of functional performance as daily fine motor tasks require accurate, coordinated, and controlled movement and manipulation of objects within the hand.

Most importantly, both patients acquired new functional skills. This is consistent with adult literature describing improved function including: turning doorknobs, picking up a can of soda, and picking up keys from the table without having to scoop them off [5].

Small sample size, variability in retrospective data, limited postoperative follow up, and non-standardized post surgical therapeutic interventions are limiting factors in our study. We were unable to establish pre-operative trends for all outcome measures selected for this study. Our postoperative data points are limited in frequency and duration which further limits examination of the emergence of fine motor skill and strength over time, as well as the ability to evaluate long term sustainability of hand function following tendon transfer.

9. Conclusion

We describe two pediatric patients with CMT that benefited from Flexor Digitorum Superficialis Opposition tendon transfer, and showed continued improvements over time without any observed/reported negative effects. It's been suggested that tendon transfers followed by hand therapy can improve function, cost effectively reduce disability, and

improve quality of life for patients with severe CMT or other slowly progressive polyneuropathies [5]. This case series, along with support from the adult literature, suggests that surgical management for children with CMT can immediately improve hand function and lead to sustained performance over the lifespan. Further research in a larger patient population is needed. Future considerations include: the role of EMG in selection of donor muscles, the impact of surgery on the dominant vs. non-dominant hand, the rate of motor learning and skill acquisition, development of appropriate therapeutic interventions, and further evaluation of the sensitivity of outcome measures including the exploration of kinematics.

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Fig. 1.
Intraoperative photographs.

Table 1

Newly aquired thumb mobility allows for improved grasp and hand dexterity.