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Current Concept Review

Use of Hyperselective Neurectomy in the Management of the Pediatric Spastic Upper Extremity

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Abstract

In children, upper extremity spasticity is a complex clinical finding with functional deficits and social implications that can be substantial. It can create significant challenges and cause distress in both the patients and their caregivers. Unfortunately, spasticity is incurable, and available treatment options are imperfect. Historically, surgical treatments for this condition were predominantly bone and soft tissue-based procedures. More recently, there is a growing body of evidence to support nerve-based procedures to decrease the degree of spasticity within select muscle groups, while maintaining volitional control. The term "hyperselective neurectomy" (HSN) has been used to describe a procedure where a specific, partial neurectomy is performed on peripheral nerve branches in close proximity to the level of the motor endplates. The result is less dysfunctional spasticity while maintaining selective native innervation to allow for continued volitional function. In this review, we discuss the role of HSN in the treatment of the spastic pediatric upper extremity. Additionally, we describe our groups' early clinical experience with this procedure and how we have implemented it into our established practice of single-event multilevel surgery (SEMLS). HSN techniques may be applicable to lower extremity cerebral palsy surgeons doing similar SEMLS.

Key Concepts

- HSN disrupts the stretch-reflex arc pathophysiology in upper extremity spasticity and is an applicable surgical procedure for upper extremity spasticity in the setting of volitional control of the muscle group of interest.

- Multiple clinical exams amongst a team of physicians promotes better surgical planning to address a patient or caregiver's goals.
- Single-event multilevel surgery decreases operating room time and anesthetic risks to children with significant spasticity.

Introduction

In children, upper extremity spasticity is often associated with a global diagnosis such as cerebral palsy (CP). Spasticity is a complex clinical finding that can come from various central nervous system pathologies. The functional deficits and social implications of a spastic upper extremity can be substantial, creating significant challenges and causing distress in both the patients and their caregivers.[1,2] Appropriate management of patients with spasticity often requires a multidisciplinary team of medical professionals. Even so, the highest level of care is imperfect, and there is clearly room for advancement. Without the ability to "cure" spasticity, treatment must be tailored to the specific patient, their needs, and the needs of their caregivers. In this article, we will review a relatively new, promising surgical technique that is targeted at the underlying pathophysiology of spasticity.

Background Information and Foundational Concepts

Spasticity is a peripheral manifestation of a central neurologic pathology. It is defined as excessive, velocity-dependent muscle contraction resulting from hyperexcitability of the stretch reflex arc and loss of inhibition.[3,4] To understand the management of this problem, we must first recognize the "stretch reflex arc" and how it is altered in spasticity. In a normal neuromuscular system, stretch of muscle spindles (located within the muscle) sends afferent potentials through sensory nerves (group Ia, II) to the dorsal root ganglia of the spine. An action potential is then carried to interneurons within the spinal cord. If activated, these interneurons will excite efferent motor neurons in the anterior horn of the spine causing contraction of the stretched muscle and relaxation of antagonistic muscles (Figure 1). Descending signals from the central nervous system coordinate the activation of this pathway and have an inhibitory effect on the interneurons which prevents contraction from occurring. In spasticity, these

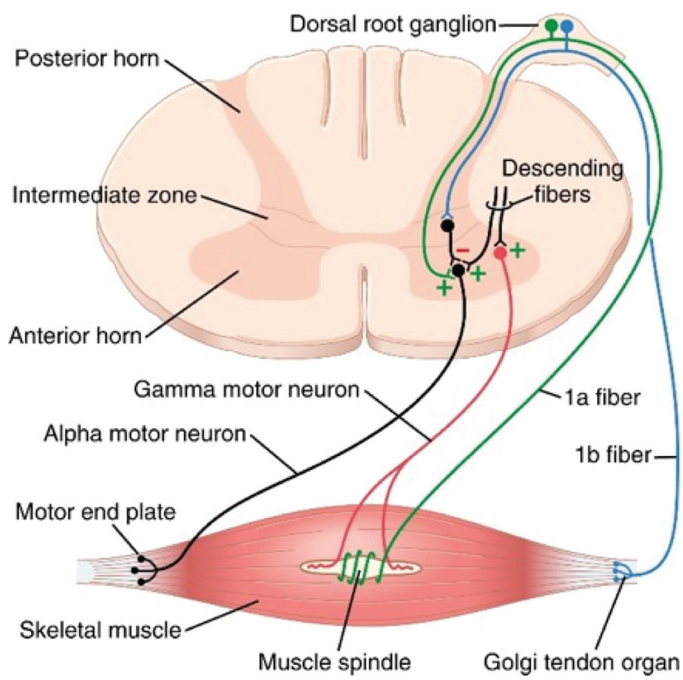

Figure 1. Stretch Reflex Arc: *Representative diagram of the biceps stretch reflex arc. When the muscle spindle is stretched, afferent potentials are carried via the group Ia, II sensory nerves to the dorsal root ganglia of the spine. An action potential is then carried to interneurons within the spinal cord (1 & 2). These interneurons lead to contraction of the stretched muscle (biceps) and relaxation/inhibition of antagonistic muscle groups (triceps). Importantly, when the descending inhibitory signals from the brain are interrupted, this leads to a hyperactive reflex arc which results in spasticity. Source: Figure 55-1, Chapter 55 Spinal Cord Motor Functions; the Cord Reflexes in Pocket Companion to Guyton and Hall Textbook of Medical Physiology published by Elsevier in 2021 (copyright permission obtained-License Number 5387930552763).*

descending inhibitory signals are decreased or absent resulting in hyperexcitability within the spinal cord and aberrant muscle contraction.[5,6]

With this knowledge, we can appreciate why many disease processes of the central nervous system cause some degree of extremity spasticity. In fact, spasticity afflicts up to 42.6% of stroke survivors, 67% of patients with spinal cord injuries, 78% of patients with multiple sclerosis, 50% of patients with traumatic brain injury, and 80% of children with cerebral palsy.[3,7] Importantly, spasticity is only one component of a constellation of findings known as "Upper Motor Neuron Syndrome" (UMN). UMN syndrome encompasses all the clinical manifestations that arise from injury to the primary motor cortex/descending motor efferents from the brain and includes weakness, decreased volitional extremity control, hyper- and/or hypotonicity, clonus, dystonia, and dyskinesia.[6] Pediatric patients with conditions like CP may exhibit some or all these characteristics. It is very important to delineate these findings because spasticity may be amenable to surgical intervention, whereas movement disorders like dystonia can be relative contraindications for limb reconstructive surgery.[4,8]

CP is defined as a fixed, nonprogressive perinatal anoxic brain injury that results in varying amounts of motor, sensory, and cognitive impairment. It is the most common etiology of spasticity in the pediatric population with an incidence of 2 per 1,000 births in the industrialized world.[8,9] In children with CP, 69% demonstrate some degree of upper extremity involvement ranging from minor disability to severe deformity and pain.[8,10] Upper limb dysfunction is typically noted by at least 1 year of age, and treatment can be initiated at the time of diagnosis. This begins with nonoperative interventions such as bracing, serial casting, oral medications, Botox injections, occupational, and physical therapy. Many of these conservative measures can and should be continued throughout the duration of these patients' lives. For some CP patients, surgical intervention may be considered. The goals of upper extremity surgery for cerebral palsy are dependent on the level of motor involvement, cognitive function, and type of movement disorder. Hemiplegic patients and their families are often most interested in both functional and aesthetic improvements. Achieving normal function is not feasible, but improvement in level of function can be achieved in properly selected patients.[11] Traditionally, goals of surgery may include muscle rebalancing by weakening antagonistic and strengthening agonist muscles with musculotendinous releases/lengthenings and tendon transfers, contracture releases, and aligning and stabilizing joints, as indicated. Surgical intervention to directly address the hyperexcitable stretch reflex arc includes dorsal root rhizotomy and complete motor neurectomy.[5] Both procedures risk impairment of the patients' remaining volitional function.

Over the past decade, there is a growing body of evidence to support nerve-based procedures to decrease the degree of spasticity within select muscle groups, while maintaining volitional control. The term "hyperselective neurectomy" (HSN) has been used to describe a procedure where a specific, partial neurectomy is performed on peripheral nerve branches in close proximity to the level of the motor endplates. The end result is less dysfunctional spasticity while maintaining selective native innervation to allow for continued volitional function.[5,12,13] While the results of these surgeries have been encouraging, the majority of the results have been limited to adult patients with spasticity secondary to cerebrovascular accidents. The purpose of this article is to review the use of hyperselective neurectomy in the spastic upper extremity, with a particular focus on its role in treating pediatric patients with spasticity and our institution's recent case-based experience with this technique.

Physical Exam/Indications

On gross examination, the upper extremity of patients with CP and UMN syndrome can appear quite similar. Classically, spasticity predominantly affects the flexor and adductor muscles of the upper extremity. This leads to the characteristic appearance of shoulder adduction/internal rotation; elbow flexion, forearm pronation; wrist flexion, digital flexion, and thumb flexion and adduction, also known as "thumb-in-palm" deformity[3] (Figure 2). At times, surgery is indicated to improve function, aesthetics, and daily care.

However, on closer evaluation, there are subtle but critical differences that must be elucidated prior to pursuing any form of surgical intervention. Thus, wholistic, detailed, stepwise, and repeated exams are essential in this patient population.[3,5,12,14] When examining a patient with CP or upper extremity spasticity, it is very important that it is done in a warm, quiet, and welcoming environment, as emotional distress and ambient conditions can significantly impact exam findings and exacerbate their spasticity.[5] It is also important to examine patients more than once and with more than one provider, if possible, as their exam may vary day-to-day based on a variety of environmental, physical, and emotional factors.

Patient Specific Goals

As surgeons, we strive to use objective clinical findings to guide our interventions. However, we recommend initiating every evaluation by ascertaining the patient's and family's goals in seeing an upper extremity surgeon. In the pediatric population, these goals are often influenced by the parents, especially in pre-school-age children. Adolescents progressing to young adults have more independence in defining their health needs. Understanding their goals can change our suggested interventions as well as help parents and patients better understand the limits of our abilities.

IQ/Sensibility/Functional Capacity

Prior to recommending any treatment, it is critical to understand the capacity of each patient. This begins with a thorough evaluation of their intellectual function. As mentioned, cerebral palsy results from perinatal injury to the cortical brain. This can result in some degree of intellectual impairment. Despite family or caregiver desires, it may be unrealistic to perform functional surgery on a child who does not have the intellectual capacity to use their extremity with intention. In fact, some surgeons suggest that in patients with an intelligence quotient (IQ) of less than 50, surgical goals should focus primarily on improving hygiene and upper extremity position in space.[11]

Figure 2. Spastic Upper Extremity: *Representative image of a patient's spastic upper extremity with the arm classically being held in elbow flexion, wrist flexion, forearm pronation, and digital flexion. The presentation can vary depending on the exact etiology and severity of spastic deformity.*

Additionally, the primary sensory cortex can also be affected by the initial brain injury. If possible, it is important to assess children's sensibility, as it has important implications for their clinical outcome. While impaired sensibility is not a contraindication to functional surgery, studies suggest that patients with less than 10 mm of two-point discrimination, patients able to discriminate three of five objects, and/or those with sensibility discrimination in the palm have better outcomes following surgery.[11] Of note, healthy children usually do not develop normal discriminatory sensibility until 5-7 years old.

Finally, in patients able to participate, it is highly recommended that clinicians obtain repeated, validated assessments of a patient's function. There are several excellent tools that exist. In pediatric patients, we use the House classification serially, preoperatively, and postoperatively (Table 1). There are many choices of validated tools for children and adults including Shriners Hospital for Children Upper Extremity Evaluation (SHUEE), Jebsen-Taylor, Melbourne, and others used by various academic medical centers.[15] When feasible, the use of validated tools by trained therapists serially is recommended. Video recordings and in-person assessments of spontaneous functional analysis and dynamic positional analysis are also often used to determine a patient's ability to perform pinch, grasp, and release activities.[9,16] The HOUSE classification provides insightful information into the patient's baseline function for their hand. Evaluating before and after Botox treatments (which will be discussed later) can also provide information about the degree of spasticity versus contracture in the extremity and potential expected improvements with surgical intervention. Regardless of the assessment tool used, obtaining a thorough, functional evaluation is critical for setting goals and surgical planning, with a higher level of baseline use and recognition of the affected extremity typically correlating with improved outcomes following reconstructive surgery.[9]

Movement Disorder Classification (Spasticity vs. Dyskinesia)

As alluded to previously, one important differentiation to make for patients with cerebral palsy is if their movement disorder is predominantly spastic or dyskinetic. Spasticity is a result of injury to the descending corticospinal tract leading to velocity-dependent hyperexcitability of the stretch reflex arc.[6] This re-producible finding creates a potential target for operative intervention.

Table 1. House Functional Classification of Hand Function in Children with Cerebral Palsy

Grade	Designation	Activity Level
0	Does not use	Does not use
1	Poor passive assist	Uses as stabilizing weight only
2	Fair passive assist	Can hold on to object placed in hand
3	Good passive assist	Can hold on to object and stabilize it for use by the other hand
4	Poor active assist	Can actively grasp object and hold it weakly
5	Fair active assist	Can actively grasp object and stabilize it well
6	Good active assist	Can actively grasp object and then manipulate it against other hand
7	Spontaneous use, partial	Can perform bimanual activities easily and occasionally uses the hand spontaneously
8	Spontaneous use, complete	Uses hand completely independently without reference to the other hand

Source: Arner M, Eliasson AC, Nicklasson S, Sommerstein K, Hägglund G. Hand function in cerebral palsy. Report of 367 children in a population-based longitudinal health care program. J Hand Surg Am. 2008;33(8):1337-1347. doi:10.1016/J.JHSA.2008.02.032.

Dyskinesia, by contrast, results from extrapyramidal lesions and can be classified as predominantly dystonic or choreoathetotic. Patients with dystonia exhibit rigidity and involuntary muscle contractions that are typically exacerbated by voluntary movement. Patients with athetosis demonstrate random fluctuations in tone (predominantly hypotonia) with writhing uncoordinated movements.[5,9,11] As one could imagine, unpredictable patterns of movement and tonicity make targeting surgical treatments quite difficult and are frequently considered relative contraindications for functional surgery in children with cerebral palsy.

Spasticity vs. Contracture

When considering hyperselective neurectomy, it is critical to differentiate between spasticity and contracture in a child with CP. In general, when a purely spastic muscle is relaxed, the extremity in question should demonstrate full, passive range of motion. If spasticity persists without intervention, the constant myostatic shortening can ultimately lead to intramuscular fibrosis and joint contractures. Unlike spasticity, a contracture cannot be overcome even with full muscular relaxation.[3,5] Differentiating these findings can be quite difficult because the degree of spasticity that an extremity exhibits can vary between patients and circumstances.

We recommend using validated tools for spasticity over multiple visits to grade patients' spasticity. In this way, surgeons can develop a firm understanding of the patients' deformity and where surgery should be focused. Validated tools include the modified Ashworth score and the Tardieu scale, both of which provide a 0-4 numeric value for the spasticity exhibited by a muscle group[5,9,11] (Table 2).

One important technique for differentiating between spasticity and muscular contracture is the use of botulinum toxin (Botox) injections. Botox functions by preventing acetylcholine release from motor neurons at the motor endplate. This results in decreased motor function or even flaccid paralysis of the muscles injected, which can last up to 3 months.[12,17] If a patient has full range of motion following injection, we can assume that contracture plays little to no role in their deformity.

An additional benefit of Botox injection is that it allows for a more thorough evaluation of patients' antagonistic musculature. Often, in patients with CP, muscles like the wrist extensors and digital extensors are overpowered by their spastic flexor antagonists, which makes it difficult to assess if patients have any volitional extensor function. Similarly, intrinsic muscle spasticity to the hand may be obscured by more dominated extrinsic finger and

Table 2. Spasticity Grading Scales

Grade	Modified Tardieu Scale	Modified Ashworth Scale
0	No resistance to passive ROM	No increase in muscle tone with passive ROM
1	Slight resistance with no clear catch	Slight increase in tone; catch and release or minimal resistance with ROM
2	Clear catch at precise angle, halting passive ROM, followed by release	Marked increase in muscle tone, catch in middle range, and resistance through remainder of ROM, still easily moved
3	Fatigable clonus (<10 seconds with resistance) occurring at precise angle	Considerable increase in muscle tone, passive movement is difficult
4	Infatiguable clonus (>10 seconds)	Rigid in flexion or extension

Two of the more common grading scales for spasticity: the Modified Tardieu and Ashworth scales. Both graded from 0 to 5 with a higher grade correlating with more spasticity. Using these scales is helpful in preoperative planning and fully evaluating a patient with spasticity. ROM: Range of motion. Source: Van Heest, Ann; Kozin S. Spasticity: Cerebral Palsy and Traumatic Brain injury. In: Green's Operative Hand Surgery.; 2022:1243-1264.

wrist spasticity and contractures. Botox treatment can unmask a patient's potential level of function. Patients who respond to Botox treatments with both absences of contracture and retained antagonist function tend to have a better prognosis following soft tissue reconstruction and hyperselective denervation.[5]

Repeat Examination

While it was mentioned several times above, it cannot be overstated that repeat examination is critical prior to performing surgery on a patient with upper extremity spasticity. In 2009, Carlson et al. found that by performing multiple clinical evaluations and reviewing a videotaped physical examination in children with CP, surgical plans were changed in roughly 72% of cases. Surgical treatment of the spastic upper extremity and hand is not emergent. Taking time to get to know patients, their exam, their goals, and their functional capacity will provide clarity and improve outcomes following treatment.

Traditional Treatment

Nonoperative treatment

All treatment plans should start and potentially continue long-term with nonoperative care in the form of occupational and physical therapy, splinting, serial casting, oral medications (such as baclofen), and serial injections (Botox) if effective in improving function and health state of the patient. This review will not delve into the full details and outcomes of those treatment modalities as our purpose is to review the current state of the role of selective denervation in surgical intervention for patients with cerebral palsy.

Operative Treatment

Following appropriate nonoperative management, some patients are ultimately indicated for surgical intervention to address their spastic upper extremity. Single-event multilevel surgery (SEMLS) is preferred in the upper limb similar to lower limb surgery in CP patients.[18] Historically, this would typically involve some combination of the following: musculotendinous fractional or z-lengthenings as indicated for the agonistic deforming force muscles, tendon transfers to strengthen and improve antagonistic muscle function, and/or contracture releases and joint stabilization procedures.[9] The specific type and number of procedures performed are highly dependent on the patient's goals, clinical evaluation, and baseline functional capacity. One of the major challenges of performing spastic upper extremity surgery is that altering anatomy at one level can impact the function of another (i.e., correcting wrist flexion contracture and providing an active wrist tendon transfer may result in a digital flexion deformity) may result in a digital flnger flexion deformity if there is no active digital extension. For this reason, we and others advocate for SEMLS in order to limit anesthesia exposure in children and address the anticipated effects of surgery on each level.[8]

Hyperselective Neurectomy

More recently, several centers that treat upper extremity spasticity in adults have found hyperselective neurectomy to be an effective addition to this historic algorithm; in particular for joints that have nearly full passive range of motion and are predominantly affected by spasticity. Most often this involves the elbow flexors (biceps, brachialis, and brachioradialis) but also the pronator teres, wrist flexors (flexor carpi radialis and flexor carpi ulnaris), and, even in some cases, hand intrinsic muscles.[12]

Historical Perspective

The concept of selective neurectomy was first introduced by Dr. Adolf Stoffel in 1913 when he described partially sectioning the median nerve in the upper limb to treat spasticity.[19] Ultimately, this technique was abandoned due to unsatisfactory results and high rates of recurrence, partially due to the limitations of anesthesia and antimicrobial medications at that time. The technique was later adapted by Brunelli and Brunelli in 1983, after being almost forgotten for 7 decades. In their technique paper, they advocated for use of large exposures, magnification, and intraoperative nerve stimulation to identify all branches of motor nerves adjacent to the level of the motor endplate. Initially, they advocated for sectioning one-half (50%) of the motor nerve branches to the affected muscle (i.e., musculocutaneous motor branches to the biceps and brachialis) because their concern was not to overly denervate the muscle and lose volitional, important function. However, they and others found a higher-than-expected rate of recurrent spasticity. They attributed this to the "adoption" phenomenon, stating that the intact nerve endings ultimately reinnervated nearby muscle fibers.[5,20] Thus, surgeons who regularly perform these operations now advocate for sectioning between two-thirds and three-quarters of the intact nerve endings.[4,12]

Theorized Mechanism

Hyperselective neurectomy offers a potentially longer-lasting or even permanent solution for upper extremity spasticity while maintaining volitional function of the affected muscle. By sectioning the neurons just proximal to their motor endpoint, one is dividing both the motor efferents as well as the proprioceptive afferents from the intramuscular spindle cells and Golgi tendon organs. Thus, the stretch reflex arc is impaired through sectioning both the afferent and efferent fibers. The close proximity of the severed motor nerve endings allows for re-growth of the motor efferents into the muscle to maintain volitional control and strength, while the proprioceptive dendrites are unable to regrow. This allows for improved spasticity not only in the immediate postoperative period; but with results from several studies suggesting a lasting effect.[13]

Technique for Hyperselective Neurectomy

After multiple clinical examinations have been performed to identify the spastic component of a patient's deformity, a "hyperselective" neurectomy (HSN) as defined by Leclercq et al. can be performed as an isolated surgical intervention or as a part of SEMLS.[5] The ideal patient for HSN has no to minimal contracture, impairing spasticity, and some volitional control that is functional in the affected muscle(s). HSN does not address joint contractures and is contraindicated in patients with dystonia or that lack volitional control of the targeted muscle group.

Presently, between two-thirds to three-quarters of the total number of nerve fascicles are resected at their entry point into the muscle.[5] The goal of the procedure is to decrease the spastic component of the deformity while retaining some active control of the involved muscle.

To perform a HSN, a high-level understanding of peripheral nerve anatomy and the variations therein must be obtained; several cadaver studies by Leclercq et al. provide a blueprint for planned HSN to peripheral nerves for HSN[21,22] (Figure 3). The number of peripheral nerve branches to resect has been a point of much research and anecdotal learning. When selective neurectomy was brought back into popularity by Brunelli, they reported a recurrence rate in over half of their patients that ultimately returned to the operating room[20] for further intervention. The higher-than-desired recurrence rate is now believed to be from not resecting enough of the peripheral nerve fascicular branches (they initially only advocated resecting 50% of the fascicles for the HSN).[5,20] Moreover, a simple neurotomy is not recommended, but an actual resection of at least 5 mm of the nerve root fascicle that has been identified should be performed for at least two-thirds of the motor fascicles identified under loupe magnification. For example, if nine terminal motor branches from the musculocutaneous nerve innervating the biceps are identified, then neurectomy would be performed on six of the terminal branches, leaving three branches in continuity. The neurectomies of the distal motor end branches would involve resection of at least 5 mm of the nerve within the confines of the muscle just before the motor end plates. There are certain muscle groups, however, that are not optimal for HSN given their deeper anatomy and the morbidity associated with identification of all terminal branches. In particular, the flexor digitorum superficialis, flexor digitorum profundus, and flexor pollicis longus innervation are such that these muscles are preferably treated with fractional lengthenings when they are involved in the deformity in the upper limb.[13,23]

In terms of pre-clinical research, a veterinary study of 113 double-muscled Belgian blue calves with spastic paresis were treated surgically by partial tibial neurectomy under caudal epidural anesthesia. Follow-up

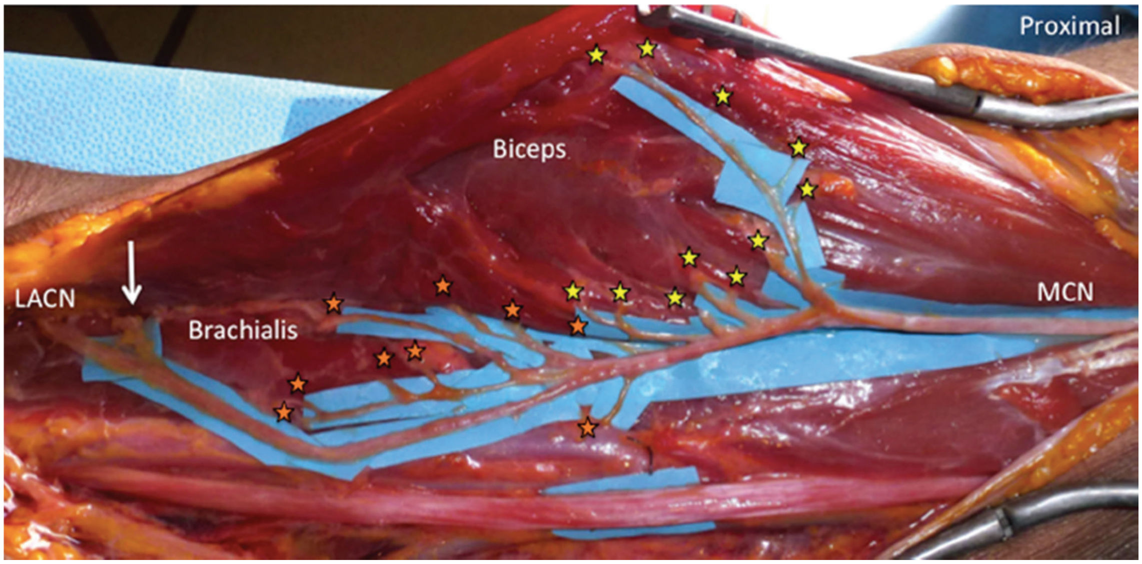

Figure 3. Branching Pattern of the Musculocutaneous Nerve: *Anterior view of the right arm with four trunks from the musculocutaneous nerve towards biceps brachii muscle with 11 terminal branches (yellow stars) dissected within biceps musculature and 9 terminal branches to brachialis muscle (orange stars). Note an accessory cutaneous sensory branch (white arrow) that exits the nerve at 82 % of the arm length (coracoid process-lateral epicondyle distance). MCN musculocutaneous nerve, LACN lateral antebrachial cutaneous nerve (color figure online). Source: Anatomical Study of the Musculocutaneous Nerve Branching Pattern: Application for Selective Neurectomy in the Treatment of Elbow Flexors Spasticity–Adeline Cambon-Binder, Caroline Leclercq.*

over 3 months postoperatively established that good results were obtained in 83.2% of the calves.[24] A large improvement was also found in 4.4% which still had intermittent spastic contractions. Persistent severe hyperflexion foot deformity resulted in early slaughtering in 4.4% of the calves; in 8% there was little or no improvement.

A recent prospective trial by Leclercq et al. reviewed their results of 42 patients undergoing HSN (29 adults and 13 children) where they resected at least two-thirds of the identified fascicular branches to the target muscle and had excellent outcomes. The only two patients undergoing repeat surgery were for muscle groups not targeted by the original HSN procedure, for instance, the brachioradialis innervation that was not addressed in correcting elbow flexor spasticity initially because it was felt that the spasticity was more from the biceps and brachialis.[12] The study specifically evaluated HSN performed for muscles of elbow flexion, forearm pronation and wrist flexion and evaluated for their short-term results (average 6 months) and long-term outcomes (average 31 months). All patients had a significant reduction in spasticity with no evidence of recurrence. While sensory nerve disturbances are the main complication reported from HSN, there were no incidents of the complication in this prospective series; this is believed to be because predominantly motor branches are resected with the use of loupe magnification and intramuscular dissection to limit damage to sensory branches. Interestingly, and important to highlight, there was no reported loss in strength in the muscles undergoing HSN for any of the patients in this study.

Our Experience with Hyperselective Neurectomy in Pediatrics

Our group's experience using HSN in pediatric patients was a product of collaboration amongst three physicians each with different areas of expertise and at different stages of their career. DL, BJL, and RGG have many years of experience treating adult stroke, brachial plexus, and tetraplegia through their time leading our brachial plexus, peripheral and central nerve injury clinic; PW has had a career of experience treating and researching the pediatric upper limb as it relates to cerebral palsy, brachial plexus, and pediatric central and peripheral nerve injuries; and finally, MG, a recently graduated orthopaedic hand fellow, gained first-hand experience from Dr. Caroline Leclercq in the winter of 2022 and her techniques and outcomes treating upper limb spasticity with HSN (through a traveling fellowship funded by the OrthoCarolina Hand Fellowship). With their combined knowledge, these physicians sought to apply hyperselective neurectomy to the standard treatment of the spastic pediatric upper extremity and hand. In addition to care of adult patients with spasticity, we have now performed hyperselective neurectomy in several pediatric patients for the spastic elbow, in combination with other more typical procedures at the forearm, wrist, and hand level as a part of SEMLS.

In general, our group has taken a team-based approach to optimize treatment and expand the surgical options available for patients with complex upper extremity problems. To do so, we have created multiple multi-disciplinary, specialty clinics designed to treat patients wholistically and facilitate innovation and education through thoughtful collaboration between experts and learners. We now have a dedicated clinic for the treatment of the spastic upper limb for both adult and pediatric patients. Having multiple upper extremity surgeons with both adult and pediatric expertise working together allows for input from multiple perspectives when deciding on the appropriate indications and surgical techniques when treating these complex patients. In addition, through simultaneous surgery on different levels of the arm and hand, we have experienced more efficient SEMLS.

Representative Case

MU is a 13-year-old female who recently underwent a single-stage, multilevel surgery for a spastic left upper extremity. She has a past medical history of a hemorrhagic stroke that occurred in the setting of viral myocarditis in 2016 with residual deficits. Her main complaint was her limited hand function due to lack of active wrist and finger extension as well as her thumb-in-palm deformity. In addition, she had elbow flexion spasticity during ambulation and activities of daily living (Video 1). She had reasonable volitional control around the shoulder and elbow, and her parents noted that when she was asleep and relaxed, her elbow and hand were supple. In addition, she was distraught about her appearance and how her marked wrist flexion and forearm pronation deformity was creating independent care and dressing problems for her.

See Video 1. Preoperative video of upper extremity posture: *This is a video of the patient obtained by the patient's parents in a relaxed home environment. Note the posture of the patient's left upper extremity. As she walks across the room, she holds her arm adducted, elbow flexed, forearm pronated, and her wrist and digits flexed with her thumb in her palm.*

Her physical exam of the left upper extremity demonstrated that her elbow rested in a flexed position of 30-40 degrees, wrist in full flexion at nearly 90 degrees, with digits and her thumb flexed into her palm. She had full strength in shoulder internal rotation, external rotation, and forward elevation. In terms of shoulder range of motion, her external rotation was up to 45 degrees with her arm by her side and forward elevation to 180 degrees actively. Her passive elbow range of motion was full but with increased spasticity in the elbow flexors as she was brought through the last 40 degrees of extension. She had full passive and active pronation. There was limited active supination 50 degrees short of neutral with spasticity of her pronators. She had passive wrist extension to neutral but with significantly increased tone in her wrist and digital flexors. Passively, she had full digital extension with her wrist 30 degrees or greater. She had no active wrist extension, supination, digital extension, intrinsic function of the abductor pollicis brevis (APB), 1st dorsal interosseous (DI), or abductor digiti quinti (ADQ). She did, however, have full strength to pronation, elbow flexion, and elbow extension with reasonable volitional control. She also had full strength for flexor digitorum profundus (FDP) to the index, long, and ring as well as flexor digitorum superficialis (FDS) to those same digits but with mild spasticity. Wrist flexion strength was primarily through palmaris longus (PL) and flexor carpi radialis (FCR) with weak flexor carpi ulnaris (FCU) function. There was no extensor carpi ulnaris (ECU), extensor carpi radialis longus (ECRL), or brevis (ECRB) function either.

Her main issues not resolved by non-operative means were: (1) elbow flexion spasticity, (2) forearm pronation spasticity and contracture, (3) marked wrist flexion contracture with no active wrist extension, (4) no active digital extension, (4) concern regarding tightness of her digital flexors and thumb FPL once her wrist deformity was corrected. After several ambulatory clinic visits and consultations independently with all members of the hand and upper limb nerve surgical team with her and her parents, we all agreed that surgical intervention was warranted and had a reasonable likelihood of improving her function and her aesthetic concerns. Our plan was (1) HSN for her elbow flexors (brachialis and biceps) rather than traditional lengthening, (2) pronator teres rerouting tendon transfer rather than just a release in hopes of not only correcting her deformity but gaining some active supination, (3) release of FCU and PL and performing a proximal row carpectomy and wrist fusion as her donors were limited and weak for wrist extension tendon transfer, (4) FCR transfer to EDC and FDS/FPL musculotendinous lengthening to prevent a clenched fist deformity, (5) PL to EPL tendon transfer with rerouting through 1st dorsal compartment to improve thumb palmar abduction and lessen retropulsion.

Intraoperative

To place her hand in a more functional position as well as improve her spasticity, a multilevel, single-stage reconstruction of her left upper extremity was performed by two surgical teams working simultaneously. SEMLS has been shown in the literature to not only decrease operative time and operating room costs but also improve outcomes in patients with spasticity conditions.[18,25] Moreover, the benefit of decreasing anesthetic risk for the child by avoiding multiple surgeries is of great value.[26] One team performed the selective denervation of her biceps and brachialis proximally while positioned in the axillae; meanwhile, the other surgical team performed a left wrist fusion with proximal row carpectomy, bone grafting from carpal bone donors, and plating via a dorsal approach with the arm resting on a fluoroscopic hand table (Figure 4). The radial physis was preserved and the fusion was from the radial epiphysis to the mid-carpus. A FCU tenotomy was performed, and the FCR was harvested as a donor for tendon transfer to the extensor digitorum communis (EDC) for index, long, ring, and small fingers. Palmaris longus transfer to EPL was performed with rerouting of the EPL volar to the first dorsal compartment. With the wrist in 5 degrees of flexion, the FDS to the index, middle, ring, and small fingers as well as FPL were lengthened at the musculotendinous juncture due to tightness in the corrected wrist position. Both surgical teams then met in the middle of the forearm to perform a pronator teres rerouting tendon transfer.

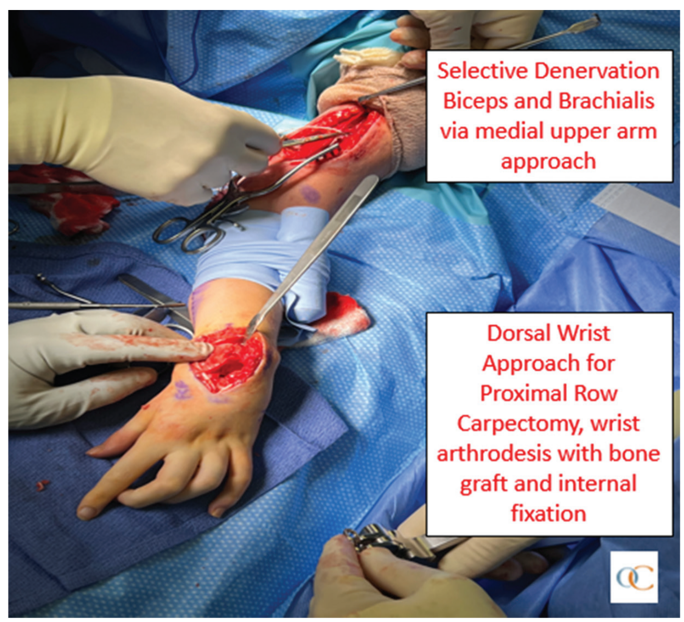

Figure 4. Single-Event Multilevel Surgery: *Intraoperative photo of two teams performing single-event, multi-level surgery on MU. Proximally, one team is performing hyperselective neurectomy of the patient's biceps and brachialis. Distally, the second team is performing a proximal row carpectomy and wrist fusion.*

For the hyperselective neurectomy of the elbow flexors, a 10-15 cm incision was made in the brachial groove along the medial arm. This is done without a tourniquet given the proximal level of the first branch to the biceps, which would be inaccessible with the use of a tourniquet. The brachial sheath was incised. The musculocutaneous nerve was identified and carefully neurolysed several centimeters proximally and distally between the biceps and brachialis muscles. A nerve stimulator was used to confirm the primary biceps branch with stimulation causing isolated biceps muscle contraction; meanwhile, the brachialis branch was identified distally near the transverse vascular pedicle that is often adjacent and a helpful landmark. Nerve stimulation of the primary brachialis branch confirmed its identification as it caused isolated contraction of the brachialis muscle. Distally, the lateral antebrachial cutaneous nerve was identified and stimulated, with no distal muscle contractures as would be expected.

After careful neurolysis of the brachialis branches into the brachialis musculature, a total of six branches from the main trunk were identified. Proximally, the neurolysis of the biceps branch into the biceps musculature also revealed six smaller branches (Figure 5). Complete identification of all branches requires careful intra-muscular dissection to follow the nerve a few centimeters into the muscle where it can be seen dividing into multiple branches. Four of the six branches were divided sharply for each nerve using micro-scissors, performing a neurectomy of about 2-3 mm in length for each identified branch just proximal to its entrance into the muscle. After performing the selective neurectomy, the nerve stimulator was again used to stimulate both primary nerve branches to show visible, retained contraction of biceps and brachialis musculature, respectively. The wound was copiously irrigated and closed in layers with absorbable sutures. After closure of all wounds, she was placed into a long arm cast with the forearm in neutral.

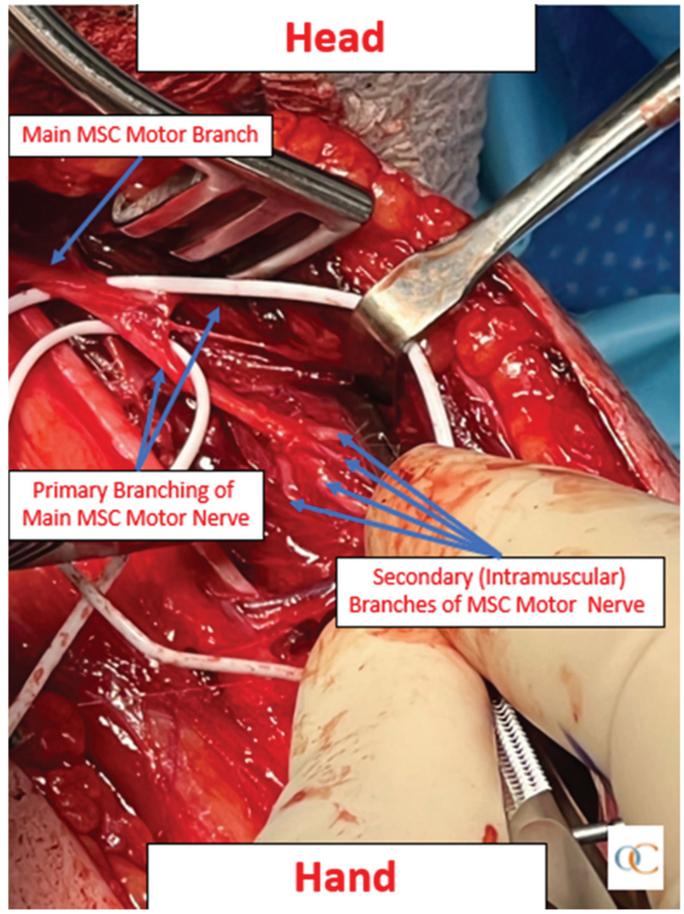

Figure 5. Musculocutaneous (MSC) Nerve Neurolysis: *Intraoperative photo of the MSC after its motor branches were identified. Proximally, we can see the main motor nerve branches. Distally, we can see how the nerve further divides and dives into the muscle belly towards the motor endplate. The secondary (intramuscular) branches of the MSC are where the hyperselective neurectomies were performed. In this image, four secondary branches are visualized entering the biceps muscle belly.*

Short-Term Follow-Up

At her short-term postoperative appointment following wound healing, cast immobilization, and early therapy she had significant improvement in her spastic elbow flexor tone and maintained active elbow flexion to 125 degrees with 4+/5 strength biceps and brachialis strength. Her resting posture was much improved at near full elbow extension (Figure 6). Additionally, the patient was able to ambulate with much less spastic flexion of her elbow compared to preoperatively (Video 2 A & B). Although in a position of pronation at rest, she could actively rotate her forearm to neutral with her wrist held in a stable neutral position. Her digits were held in slight flexion at her PIP and DIP joints. Her thumb was noted to remain slightly in her palm.

Figure 6. Early Postoperative Clinical Photo: *Resting posture of MU's left upper extremity shortly following hyperselective neurectomy. We can see that she holds her arm in significantly less elbow flexion when compared to preoperatively. The forearm is in slightly less pronation in this image as well.*

See Video 2 A & B. Short-Term Follow-up: *(A) Volitional flexion: shortly following her hyperselective neurectomy to the biceps and brachialis, MU has clearly maintained active elbow flexion from 5 to 125 degrees with significantly less spasticity compared to preoperatively. (B) Ambulation: additionally, MU was asked to walk as she had in the video that was obtained preoperatively. Notice that her elbow is held in near full extension with little to no inadvertent elbow flexion.*

Longer-Term Follow-Up (Video 3)

At her longer-term postoperative follow-up, she has improvement in her grip strength with better ability to grasp objects. Her parents noted improved posturing at the elbow with a more natural elbow resting position with marked diminution of her spasticity (Video 3). She improved from modified Ashworth grade 3 preoperatively to 0 postoperatively. Additionally, she improved from a HOUSE 0 preoperatively to 3 postoperatively as she can now actively assist in holding objects weakly such as her water bottle, use her operative hand now for assisted dressing and hygiene which she never did before, and stabilize a book for reading. She was able to flex and extend her digits and thumb from a balanced cascade. On exam, she has passive elbow range of motion from full extension to 150 degrees of flexion. She has no spasticity in her elbow flexors with passive elbow extension and minimal with gait, mostly when she does not wear her leg brace. She has 4+ out of 5 elbow flexion strength. At the forearm, she has active forearm supination beyond neutral now. Her wrist rested in neutral position from her wrist fusion. She has limited digital grasp with a Volkmann angle at 0 degrees, achieving full digital and thumb extension with her neutral wrist position. She does still have a tendency for a thumb-in-palm active positioning when stressed, but this is supple and improved clinically. Radiographs reveal neutral wrist position, hardware in place with bridging trabeculae across her radiocarpal joint (Figure 7).

See Video 3. Longer-Term Follow-Up: *At her longer-term postoperative follow-up, we can see that MU has improved volitional control of her elbow flexion from 0 to 125 degrees. She has no spastic co-contraction noted throughout the arc of motion.*

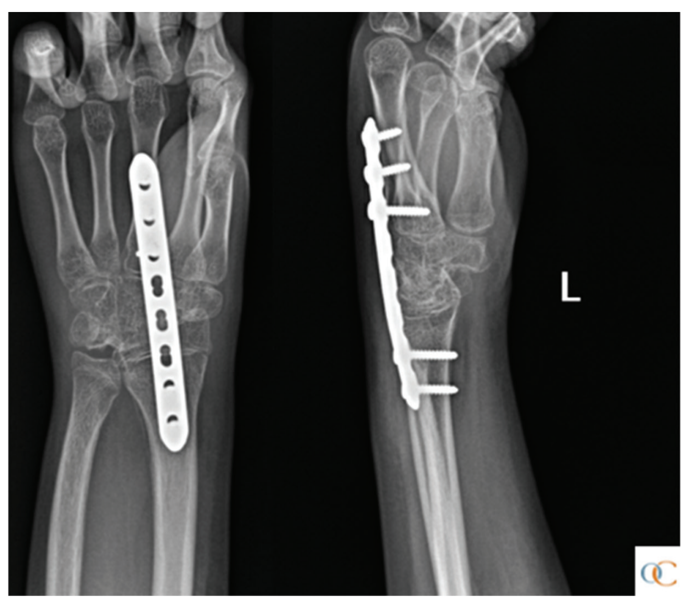

Figure 7. Longer-Term Follow-Up Radiographs: *AP and lateral wrist radiographs obtained at longer-term follow-up visit. Bridging trabeculae from the radial epiphysis to the midcarpal bones indicates a successful wrist fusion.*

Conclusion

Given the recent advances and better understanding of the pathology involved in upper extremity spasticity, nerve-based procedures such as hyperselective neurectomy should now be considered along with more traditional approaches for patients affected by these conditions. When multiple physical examinations of a child reveal true spasticity of a muscle group, volitional control present, and nearly full passive range of motion, HSN should be considered as an alternative solution or adjunct to an entire upper extremity reconstruction. While we have experience in treating the spastic elbow flexors with HSN, we intend to expand these offerings at the forearm and hand level for forearm pronation, wrist flexion, and hand intrinsic spasticity based on the experiences reported by others. In the setting of upper extremity reconstruction, consideration should be given to deploy multiple surgical teams to decrease the anesthetic risk for the child as well as decrease operating room costs and time. There may be applicability for adaption of HSN in SEMLS for lower extremity CP surgery as well.

Additional Links

- POSNA Study Guide: Cerebral Palsy
- Whitaker, A. T. (2022). What's New in Orthopaedic Management in Neuromuscular Disorders: Invited Perspective. *Journal of the Pediatric Orthopaedic Society of North America*, *4*(3).

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