Surgery of the Upper Extremity in Cerebral Palsy

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Functional activities of the upper extremity are limited in most individuals with a diagnosis of cerebral palsy (CP). However, surgical interventions are applied in fewer than 20% of pediatric patients with an upper extremity affected by CP, in marked contrast to the lower extremity in which surgery is more frequently indicated. Apart from improving function, surgical procedures may decrease pain, prevent or fix upper limb deformity, and have a positive impact on the patient’s caregiving, self-esteem, and appearance. Several conservative treatment methods are also available (eg, therapy, casting, electrical stimulation, oral spasmolytic medications, and parenteral neuromuscular blocking agents). These methods are primarily used to preserve joint range of motion (ROM), to delay tendon and muscle contractures, and to prevent upper extremity osseous deformities. In patients without competently functional antagonist muscles, passive stretch for a minimum of 6 out of 24 hours is required to maintain muscle length and to avoid development of a fixed contracture. Occupational therapy and splinting alone do not accomplish long-term reduction of the involuntary spasm. Moreover, pharmacologic agents designed to decrease spasticity have not been validated as a definitive means of providing lasting improvement. In selected cases, surgery following conservative treatment has been reported to give satisfactory results.

This article covers the surgical interventions used for the reconstruction of the upper limb in patients with CP. The optimal surgical approach for each deformity type is described. In addition, the various evaluation techniques of the upper extremity, the general principles of an operative treatment plan, and the appropriate postoperative care of these patients is presented.

EVALUATION

History and Physical Examination

A team approach to the management of the patient with CP is important; medical history and physical examination is the basis for the successful assessment of each individual. Input from the patient and his or her caregiver, and any involved health care worker should be provided. Functional classification systems (eg, House scale) as well as standardized testing regimens, such as the Melbourne Assessment of Unilateral Upper Limb Function (Melbourne), are helpful evaluation tools. The Melbourne scale allows objective measurement of the upper extremity function in patients with CP. Both active and passive ROM of the upper extremity joints should be assessed in a reliable and reproducible manner. In the authors’ institution, the Upper Extremity Rating Scale (UERS) is used for this purpose. The UERS provides a composite score of the active and
passive ROM of the shoulder, elbow, forearm, and wrist.\textsuperscript{15} Degree of spasticity, and absence or presence and magnitude of involuntary movement disorders should also be evaluated and recorded. For patients with mild to moderate involvement, the use of the Melbourne scale and the Jebsen-Taylor Hand Function Test is suggested. A global instrument such as the WeeFIM (Uniform Data System for Medical Rehabilitation, University at Buffalo Foundation Activities, Inc, New York) is also available to measure self-care and functional skills. If possible, the patient is observed during ambulation, standing, and sitting to evaluate certain posture and motion patterns. Several factors including fatigue level, anxiety, and even time of the day may affect the clinical findings; therefore, serial evaluations are necessary. Preoperatively, sensibility testing including evaluation of proprioception, stereognosis, and 2-point discrimination should also be performed.

**Imaging Studies and Ancillary Testing**

Evaluation of patients with CP is complex. Several examinations are necessary as part of the surgical planning process. Plain radiographs, computed tomography imaging studies, and magnetic resonance imaging studies are helpful in assessing preoperative joint congruency. An individualized and detailed functional evaluation is also a critical component for developing the appropriate surgical plan. Dynamic electromyography (EMG) provides a qualitative and quantitative assessment of voluntary motor control and the type of motor activity of muscles being considered for transfer.\textsuperscript{14–16} A videotaped evaluation of the upper extremity in children with CP provides an objective assessment of a patient’s motor performance and functional capacity. Carlson and colleagues\textsuperscript{17} reported changes to the initial preoperative plan following the study of videotaped evaluations, especially for procedures addressing the wrist, digit, and thumb. Motor blockade produced by injections of botulinum toxin A (BTX-A) or a topical anesthetic agent (eg, bupivacaine) into the muscles identified for surgery may serve as a diagnostic tool to select the proper operative interventions.\textsuperscript{18}

**General Principles of an Operative Treatment Plan**

Diagnostic evaluation is helpful in identifying the suitable candidates for complex reconstructive procedures of the upper extremity. Selection of the appropriate intervention is required to achieve both specific and global outcomes individualized for each patient. However, it must be recognized that although some improvement in function and appearance may be a realistic goal for properly selected patients, normality can rarely, if ever, be achieved. The type of joint deformity, the underlying neuromuscular disorder, the preoperative sensibility and functional capacity of the limb, the patient’s intellectual status and goals, and the surgeon’s preferences are the factors used to devise a treatment plan.\textsuperscript{19}

Global surgical goals for best outcomes include (1) improved function, (2) facilitation of care, (3) pain reduction, and (4) enhancement of self-esteem. Specific surgical interventions may be performed at one or more levels (ie, shoulder, elbow, wrist, hand) to release overactive muscle groups, stabilize joints, and augment selective motor control of weak muscles by tendon transfer to achieve each patient’s specific goals. Surgical interventions may be reduced to a checklist of options (Table 1). Proactive planning with multiple team members, including the development of an immediate postoperative regimen (eg, period and type of immobilization) and rehabilitation program, is the key to the success of the operative procedure.

**Operative Procedures**

**Shoulder**

Adduction and internal rotation shoulder deformity is common in patients with CP; the deformity is due to unbalanced spasticity of the internal rotators of the arm at the glenohumeral joint (pectoralis major, latissimus dorsi, subscapularis, and teres major).\textsuperscript{20} A fixed contracture of the muscles (mentioned earlier) and the joint capsule may contribute to the deformity posture. Surgical interventions may need to address (1) the muscle/tendon and/or capsule contractures, and (2) the subluxation or dislocation of the humeral head in one or more planes of the shoulder joint. Inferior subluxation is the most common form in the hemiplegic shoulder.\textsuperscript{21} Moreover, dysplasia of the glenoid or humeral head and/or arthritis of the glenohumeral joint should be encountered in the treatment plan. In dynamic deformities, the glenohumeral articulation is typically stable with congruous articular surface contact. Treatment options include muscle lengthening, tendon transfer, humeral osteotomy, and shoulder joint fusion.

Although surgical treatment is rare, the pectoralis major and subscapularis muscles should be lengthened to correct shoulder adduction and internal rotation deformity; capsular release may also be performed. Transfer of the latissimus dorsi and teres major may augment active external rotation of the arm (Fig. 1). In severe cases, release of the latissimus dorsi and teres major muscles in conjunction with the procedures described above
may be required. If tendon/muscle release and/or transfer fail, a proximal or distal osteotomy of the humerus may be used to improve rotation of the arm. Osteotomy is also indicated for patients with dysplastic/subluxed or arthritic shoulder joints. Refractory arthritic pain may be addressed by shoulder fusion in individuals with CP; however, there is no experience with this procedure in the authors’ institution.

**Elbow**

Flexion contracture of the elbow may interfere with the use of the limb by limiting reach activities of the hand. The associated abnormal attitude of the extremity, appearing during gait, is also a cosmetic disability and may impair self-image unless addressed. Increased muscle tone of the biceps brachii, brachialis, and brachioradialis muscles results in dynamic motor imbalance around the elbow joint. Secondary fixed contractures of the capsule and the adjacent flexor-pronator muscle/tendon units may contribute to elbow flexion deformity.\textsuperscript{22} Subluxation/dislocation of the radial head due to associated hyperpronation of the forearm, accompanied with secondary dysplasia of the head may be an additional feature of elbow flexion deformity in children with CP. Pletcher and colleagues\textsuperscript{23} identified a radial head dislocation in approximately one-fourth of the children, with a combination of a flexion deformity of the elbow and a pronation contracture of the forearm. Most dislocations were posterior. The investigators proposed that preoperative elbow radiographs are required for all children with this combined deformity type.

Anterior elbow release is indicated for spastic elbow flexion deformity in patients with a diagnosis of CP. Operative treatment is seldom indicated unless extension loss exceeds 30°. For elbow deformities between 30° and 60°, soft tissue procedures (including excision of the lacertus fibrosus, Z-lengthening of the biceps, and fractional lengthening of the brachialis aponeurosis) are usually sufficient, and reliably decrease the degree of deformity (Fig. 2). For deformities exceeding 60°, a flexor-pronator origin slide accompanied with anterior elbow capsulotomy may also be required.

Anterior elbow release can improve active extension of the elbow, as well as both the functional use and aesthetic appearance of the involved upper extremity. Mital\textsuperscript{22} reported the only large series of patients in whom elbow flexion deformity was addressed by weakening of both the brachialis (fractional lengthening) and the biceps muscles (release of lacertus

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**Abbreviations:** APL, abductor pollicis longus; BR, brachioradialis; ECRB, extensor carpi radialis brevis; ECRL, extensor carpi radialis longus; ECU, extensor carpi ulnaris; EDC, extensor digitorum communis; EPB, extensor pollicis brevis; EPL, extensor pollicis longus; FCU, flexor carpi ulnaris; FDP, flexor digitorum profundus; FDS, flexor digitorum superficialis; FPL, flexor pollicis longus; LD, latissimus dorsi; MCP, metacarpophalangeal; PIP, proximal interphalangeal; PT, pronator teres.
aponeurosis, tendon Z-lengthening). Following this procedure the flexion posture angle was decreased, and the patient’s ability to flex the elbow or supinate the forearm was retained. However, the risk of increasing pronation deformity following a biceps-lengthening procedure should be noted; rerouting or lengthening of the pronator teres (PT) muscle may prevent increased pronation deformity after anterior elbow release.

Elbow fusion is considered as the last option for operative treatment of the elbow flexion deformity. It is reserved for patients who experience intractable elbow pain. However, this procedure is seldom used and has never been performed at the authors’ institution.

Fig. 1. For the modified Sever-L’Episcopo procedure, Z-lengthening of the pectoralis major is performed (A), the subscapularis is lengthened on the “flat” (B), and the latissimus dorsi and teres major are transferred to the posterolateral humerus using 1 incision (C) or 2 incisions (not shown). (Reproduced from Koman LA, editor. Wake Forest University School of Medicine orthopedic manual 2001. Winston-Salem (NC): Orthopaedic Press; 2001. © Wake Forest University Orthopaedic Press; with permission.)
Forearm

Pronation of the forearm is part of the typical pattern of the upper limb in patients with CP. The deformity may be fixed, dynamic, or a combination of both; hypertonicity of the PT and pronator quadratus muscles is the primary cause. Inability to supinate the forearm in the absence of shoulder compensation interferes with hand function and compromises activities such as turning a doorknob or using a key. Multiple nonsurgical measures (muscle stretching, plaster casts, splints, braces, and BTX-A) have been advocated. Following extensive conservative treatment, operative procedures in properly selected patients are appropriate to improve forearm position and function. Allowing active supination to neutral should be considered a successful outcome.

Operations can be classified into 2 groups. In group I, procedures primarily designed to improve functions such as wrist dorsiflexion, elbow motion, and hand grasp and release are included. The release of the pronation contracture is incidental to these procedures. The flexor-pronator release, primarily advocated to decrease flexion deformity of the wrist and fingers, has been reported to improve supination of the forearm. However, the risk of supination deformity following excessive flexor-pronator release is increased. Greer described a method to correct dynamic flexion deformity of the wrist; the flexor carpi ulnaris (FCU) tendon is transferred to augment either the extensor carpi radialis brevis (ECRB) or the extensor carpi radialis longus (ECRL) muscles. By transferring the FCU tendon from ulnar to radial, a supination moment is created and pronation deformity is potentially decreased.

In group II, procedures are designed exclusively for pronation deformity of the forearm. PT release tenotomy, fractional lengthening, Z-lengthening, and rerouting are described. In addition, rerouting of the brachioradialis has been reported to improve supination. In none of the techniques mentioned here is the pronator quadratus released at the same time; thus, the risk of loss of active pronation is minimized. Dynamic EMG as well as clinical examination of the forearm helps decide the treatment plan. In the presence of passive supination, PT release, Z-lengthening, or fractional lengthening is appropriate for patients with a continuously firing PT by EMG. However, for patients with a phase-dependent PT muscle activity, PT rerouting is the preferred surgical option at the authors’ institution (Fig. 3). If soft tissue procedures fail, radial osteotomy may be performed to improve forearm position. In severe cases, inferior radioulnar joint fusion may also be an option.

Wrist

Flexion deformity of the wrist impairs grasp-and-release function of the hand. Common causes include hypertonic wrist flexors and/or weak wrist extensors with subsequent capsular contracture. Wrist joint deformities may be dynamic or static in nature. Management options include soft tissue and/or osseous procedures, depending on the severity and nature of the deformity.

Passively correctible deformities may benefit from transfer of a wrist flexor to the weak radial wrist extensors. Preoperative dynamic EMG and clinical testing of the muscles involved in active motion of the wrist determine the appropriate surgical intervention. Functional sufficiency of the flexor carpi radialis muscle is crucial for a successful FCU tendon transfer. Dynamic flexion deformities of the wrist can be managed by transferring the FCU to either the ECRB or the ECRL or the extensor digitorum communis (EDC) tendon (Fig. 4). Preferably, in patients with continuous FCU activity throughout the wrist motion arc, the FCU is transferred to the EDC; with this procedure, wrist and finger

Fig. 2. Z-lengthening of the biceps tendon and fractional lengthening of the musculotendinous portion of the brachialis with or without flexor pronator release is appropriate for elbow flexion contractures. (Reproduced from Koman LA, editor. Wake Forest University School of Medicine orthopedic manual 2001. Winston-Salem (NC): Orthopaedic Press; 2001. © Wake Forest University Orthopaedic Press; with permission.)

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extension is improved, finger flexion is permitted, and wrist extension deformity by FCU overpull is avoided.\textsuperscript{14} If the combination of EMG and clinical testing demonstrates a phase-dependent FCU activity, improved voluntary wrist extension is achieved by transferring the FCU to the ECRB; when ulnar deviation of the wrist is an issue, the FCU to ECRL transfer is preferred. The latter procedure may be enhanced by transferring the extensor carpi ulnaris (ECU) tendon radially to the fourth metacarpal (Fig. 5).

Static wrist deformities are commonly associated with more than 45° of palmar flexion and may not respond to soft tissue release procedures in isolation. Static wrist deformities may require flexor tendon release combined with procedures such as wrist fusion (with or without carpal incisional resection)\textsuperscript{34} and/or proximal row carpectomy (PRC).\textsuperscript{35} Tendon transfers combined with PRC and flexor tendon releases are reported to allow active extension of the wrist.\textsuperscript{36} Wrist fusion maintains the wrist in a fixed neutral position and is indicated for patients that require pain relief and an improved cosmetic result.\textsuperscript{37} Before considering a wrist stabilization procedure, finger flexion and extension capability should be evaluated in the desired corrected position. Wrist flexion is crucial for effective release in many patients, and wrist extension is often needed to achieve effective grasp. Therefore, procedures that limit motion should be reserved for patients with effective grasp-and-release in a position of the wrist close to neutral.\textsuperscript{5,38}

**Thumb and Finger Deformities**

**Finger flexion deformity**

Finger flexion deformity typically is synchronous to other upper limb deformity types. Flexor digitorum

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*Fig. 3.* The PT may be detached from the radius and rerouted to improve supination of the forearm. (*Reproduced from Koman LA, editor. Wake Forest University School of Medicine orthopedic manual 2001. Winston-Salem (NC): Orthopaedic Press; 2001. © Wake Forest University Orthopaedic Press; with permission.*)
superficialis (FDS) and/or flexor digitorum profundus (FDP) spasticity is present; the digital extensors may also be weak. Lengthening of the shortened flexors is the appropriate treatment method for this deformity; power reinforcement of the finger extensors may also be performed. Preferably, the FCU or the FDS tendon of the ring finger is transferred to the EDC. One or two FDS tendons may be used; most frequently, the long and ring finger tendons are selected.

Depending on the severity of the deformity, flexor lengthening may be achieved by various interventions. Intramuscular BTX-A injections followed by serial muscle stretching is a nonoperative option. In mild to moderate cases, fractional lengthening of the individual FDS tendons combined with BTX-A injections into both the lengthened FDS and the nonlengthened FDP musculotendinous units is preferred in the authors’ institution; stretching, casting, and therapy should be applied after the initial procedure. More involved deformities may require an additional fractional or Z-lengthening of the FDP tendons. The FDS to FDP transfer may also be performed.33

In severe cases, a flexor-pronator origin release (slide) is recommended. Caution should be exercised when performing this procedure; the risk of overlengthening of the flexor-pronator mass is increased. This risk may be minimized by suturing the muscle mass in the optimal position to prevent overlengthening by injecting BTX-A or by a combination of both modalities.

**Swan-neck deformity**

The abnormal posture of the digits may adversely affect grasp function of the hand because of the inability to flex the fingers from the hyperextended position at the proximal interphalangeal (PIP) joint. Although effective treatment is difficult, the deformity can be managed by central slip tenotomy,39 PIP tenodesis,40 and lateral band translocation procedures.41 Intrinsic origin release may also be beneficial.

**Thumb-in-palm deformity**

Thumb-in-palm deformity limits the grasp-and-release function of the hand in patients with CP. The deformity consists of static (due to contracture) and/or dynamic (due to spasticity) shortening of the adductor pollicis, first dorsal interosseous, and flexor pollicis longus (FPL) muscles; the adductors and flexors of the thumb may be

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**Fig. 4.** Transfer of the FCU to the ECRB (A) or transfer of FCU to the EDC (B) improves wrist extension. The tendon may be transferred subcutaneously to provide a supination moment or through the interosseous membrane to provide more dorsiflexion without supination. (Reproduced from Koman LA, editor. Wake Forest University School of Medicine orthopedic manual 2001. Winston-Salem (NC): Orthopaedic Press; 2001. © Wake Forest University Orthopaedic Press; with permission.)
affected in various combinations. In addition, muscles involved in active extension and abduction of the thumb may be weak; extensor pollicis brevis (EPB), extensor pollicis longus (EPL), and abductor pollicis longus (APL) are overstretched due to prolonged abnormal thumb and wrist position. The wide spectrum of contributing factors is completed by instability of the metacarpophalangeal (MCP) joint and the fixed contracture of the skin and soft tissue covering the first web space.

Surgical treatment of this complex deformity usually includes various combinations of muscle releases, tendon transfers, and joint stabilization procedures. Preoperative voluntary muscle control quality has been reported to be one of the most important factors in predicting the success of the operation. Limited sensory capability is a relative contraindication for the use of complex surgical procedures. Therefore, a careful assessment of the patient’s thumb function must serve as the basis for all treatment decisions.\(^9\)

Thumb adduction deformity can be managed with careful release of the adductor pollicis and/or first dorsal interosseous muscles. Z-lengthening of the adductor pollicis is another choice (Fig. 6). Adductor pollicis release is obtained by sectioning its origin from the third metacarpal through a palmar incision, along the thenar eminence crease.\(^{42}\) In addition, a 4-flap Z-plasty rearranges the skin and soft tissues of the first web space (Fig. 7).

Surgery is indicated to hold the thumb out of the palm during grasp and to permit lateral pinch.

**Fig. 5.** Transfer of the ECU from its insertion on the fifth metacarpal to the fourth metacarpal decreases ulnar deviation and assist extension of the wrist. The extensor retinaculum should be released to allow dorsal translation of the ECU tendon. (Reproduced from Koman LA, editor. Wake Forest University School of Medicine orthopedic manual 2001. Winston-Salem (NC): Orthopaedic Press; 2001. © Wake Forest University Orthopaedic Press; with permission.)

**Fig. 6.** The adductor pollicis may be released from its origin on the third metacarpal through a volar incision (A); alternatively, the adductor tendon may be Z-lengthened or transferred proximally to the first metacarpal to increase the first web space, (B) the first dorsal interosseous muscles may be released from the first metacarpal. (Reproduced from Koman LA, editor. Wake Forest University School of Medicine orthopedic manual 2001. Winston-Salem (NC): Orthopaedic Press; 2001. © Wake Forest University Orthopaedic Press; with permission.)
Adduction deformity is not fully corrected unless the APL is reinforced. This reinforcement can be accomplished by transfer of the brachioradialis to the APL. Rerouting of the EPL tendon radial to Lister’s tubercle has also been described to augment thumb abduction.

Flexion deformity of the thumb may be an additional feature of the thumb-in-palm posture. Therefore, procedures designed to reinforce the thumb extensors should be included in the therapeutic protocol. The EPL tendon may be sectioned, rerouted through the first dorsal compartment, plicated, and reattached to the EPB at the MCP joint. The brachioradialis or FDS tendon of the ring finger may be used to augment the thumb extensor. Before improving extensor power, fractional or Z-lengthening procedures are used to address FPL fixed contractures (Fig. 8). The contracted and spastic FPL is usually weak; either the brachioradialis or another tendon may be transferred to improve its overall strength.

Pure MCP joint hyperextension instability may be improved by volar plate capsulodesis; however, if this procedure fails, MCP fusion can be used to control thumb hyperextension. Arthrodesis of the MCP joint has also proved to be a successful method for global instability of the thumb. In children, damage to the epiphyseal line can be minimized by the use of transitory thin fixation pins. If tendon transfers fail to overcome an MCP flexion deformity, MCP fusion may be performed to improve the position of the thumb.

**POSTOPERATIVE CARE**

Appropriate postoperative care is necessary to achieve the optimal surgical result. Preferably, input from therapists regarding the postoperative therapy and splinting regimen should be included in the preoperative planning. The initial 3 to 4 weeks of postoperative care are crucial for...
a successful outcome; active and passive ROM of the contiguous joints (the operative site is protected by pins and rigid splinting or casting during this period) should be maintained or improved, and tendon transfers should be protected. Intra-muscular injections of BTX-A may be used to protect tendon repairs associated with lengthening and/or transfer procedures; with this intervention, the postoperative pain is decreased, earlier active motion is permitted, and overlengthening following fractional procedures is prevented. Following the initial healing phase, active and passive ROM protocols are implemented, and static splinting is used as indicated. At 6 to 10 weeks following surgery, a strengthening exercise program is initiated; the inclusion of a home program is an important component of the rehabilitation process.

SUMMARY

Although only a small percentage of children and adults with CP are candidates for upper extremity surgery, the judicious use of the surgical procedures discussed in this article can result in positive outcomes and improve the health-related quality of life of patients. However, proper selection of patients is crucial to ensure reasonable postoperative results, and postoperative care must be coordinated with appropriately trained hand therapists for optimal results.

REFERENCES