CHAPTER 22

Donald S. Bae
Peter M. Waters

The Upper Limb

SURGICAL PROCEDURE INDEX

The purpose of this chapter is to discuss the evaluation and treatment of common congenital differences, traumatic and posttraumatic conditions, neuromuscular problems, and growth deformities affecting the upper limb and hand. Each section examines major diagnostic categories and anatomic regions. The reader will find more information regarding upper extremity development (Chapter 33), fractures (Chapter 34), and limb deficiency (Chapter 30) in other chapters of this text.

Treatment of any upper limb or hand condition in a child should address issues of function, growth, aesthetics, and the emotional concerns of the child and family. All are important factors in achieving a successful outcome. The pediatric orthopaedist's goals are to enhance the ability to place the hand in space; to improve deficiencies in grasp, release, or pinch function; to improve skin mobility and sensibility; and to improve the aesthetic appearance of the limb (1). In addition, treatment of physeal abnormalities improves growth-related loss of motion and function and may reduce pain and musculoskeletal deformity (2). Furthermore, extensive time and counseling are important to address the concerns of the child and parents regarding the alteration in self-image that can occur with any hand or upper limb deformity.

CONGENITAL DIFFERENCES

Pathogenesis. *In utero*, the arm bud appears 26 days after fertilization and 24 hours before the appearance of the leg bud. Growth proceeds in a proximal-to-distal manner. Development is guided by the apical ectodermal ridge via fibroblast growth factors, inducing the mesoderm to condense and differentiate (3). The zone of polarizing activity guides radioulnar differentiation and development of the limb, mediated by the sonic hedgehog protein and other growth factors. Similarly, the Wnt signaling center influences dorsal–volar development of the hand. The upper limb anlage is initially continuous and extends to a hand paddle by day 31. The digital rays develop by day 36 with fissuring of the hand paddle, initially in the central rays, followed by the border digits. Mesenchymal differentiation also begins in a proximal-to-distal manner with chondrification, enchondral ossification, joint formation, and muscle and vascular development. Both joint formation and digital separation occur via apoptosis, or programmed cell death. The entire process is complete by 8 weeks after fertilization (4). Other major organ systems develop at the same time as the upper limb, which explains the associated cardiac, craniofacial, musculoskeletal, and renal anomalies that can occur with upper limb differences.

Homeobox, or *HOX*, genes regulate the development of the limb (5). Their genetic expression controls the timing and extent of growth by regulating mesenchymal cells. At present, the understanding of the genetic basis of limb development, and therefore of the occurrence of congenital anomalies, is expanding rapidly (6–12). For example, a mutation at the *HOXD13* site has been identified as a cause of polysyndactyly (13). A further understanding of the role of genetics in limb development may revolutionize the treatment of congenital deficiencies.

Congenital anomalies occur in approximately 6% to 7% of live births, with 1% being multiple anomalies. It has been estimated that between 1 in 531 and 1 in 626 live births involve upper extremity anomalies (14, 15). Only 1% to 2% of these congenital differences are the result of chromosomal abnormalities.

However, 75% of 233 spontaneous abortions studied were noted to have an abnormal karyotype, with 18% having a morphologic defect and normal karyotype (16). At present, only a small percentage of these are known to be caused by defined genetic events. In most cases, the cause of the congenital difference is unknown, but expanding genetic information provides optimism for increased knowledge in the near future.

Classification. There is no perfect classification system for congenital differences of the hand and upper limb. The currently accepted classification system for congenital differences was proposed by Swanson (17) and revised by the Congenital Anomalies Committee of the International Federation of Societies for Surgery of the Hand (18, 19). This classification is based on embryologic or developmental failure and defines deficiencies as terminal or intercalary, with a subclassification into longitudinal and transverse deficiencies. The subcategories are as follows: (a) failure of formation of parts, (b) failure of differentiation of parts, (c) duplication, (d) overgrowth, (e) undergrowth, (f) constriction band syndrome, and (g) generalized skeletal abnormalities. However, there have been reports of inconsistencies in classifying congenital anomalies of the upper limb by this system (20). A more descriptive method has been shown to be valid (19, 21).

This chapter focuses on the major anomalies in each classification group but presents them by anatomic region. Caring for the child with congenital differences involves more than surgical skill. From the time at which the diagnosis is made, these children may potentially be viewed by their parents, family, and society as being impaired; if this premise is left unchallenged, these patients may view themselves the same way (22, 23). It is critical that the treating surgeon helps provide the emotional support and caring that allow the parents and the child to appropriately grieve the loss of a normal hand (24). It is helpful to provide them with in-depth knowledge of the cause and treatment options (25). This process starts with the initial clinical consultation and continues throughout the growth and development of the child into an independent, self-reliant adult (26). Support groups are useful for many of these children and their families.

The children who have normal central nervous systems and cognitive abilities will not be impaired. They will merely develop their skills in a "different" way from their peers. They may need the help of skilled and caring parents, siblings, therapists, teachers, coaches, prosthetists, and surgeons in order to achieve their goals and dreams. Being part of helping these children grow into unique and independent adults is exciting and rewarding for the pediatric orthopaedic surgeon.

ENTIRE LIMB INVOLVEMENT

Neuromuscular

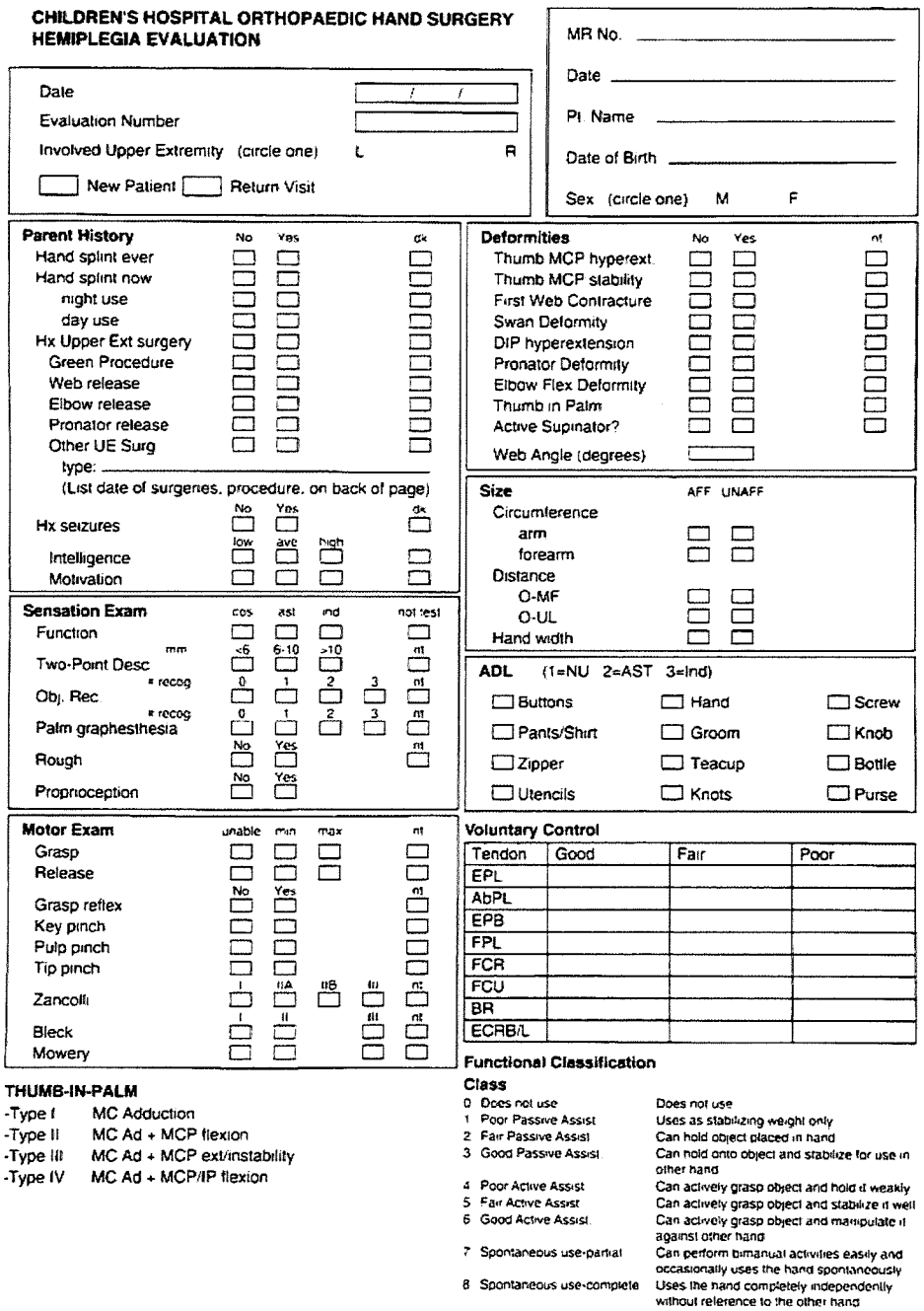
Cerebral Palsy. Cerebral palsy is a nonprogressive disorder of the central nervous system. It occurs in 5 in 1000 live births and may be caused by perinatal anoxia, intraventricular hemorrhage, or congenital cerebral vascular accidents. It occurs most commonly in premature infants weighing <1500g (27, 28). The resultant hemiplegia or quadriplegia can lead to significant upper extremity deformities and functional deficits. In hemiplegia, these individuals predominantly use the affected extremity as an assist for the unaffected extremity. In the quadriparetic patient, both upper limbs will have deformities and deficits. The quality of use of an affected extremity is dependent on many factors, including the presence of contractures, voluntary motor control, discriminatory sensibility, learning disabilities, and visual deficits (29–33). This section focuses on the deformities and deficits relating to elbow flexion, forearm pronation, wrist palmar flexion and ulnar deviation, finger flexion, and thumb-in-palm deformity in these patients.

Upper Limb Contractures. Elbow flexion contractures are often mild in patients with hemiplegia (34–36). Although approximately 50% of patients will have an elbow flexion contracture, most of these contractures are <30 degrees and do not limit function (37, 38). There may be an associated radial head dislocation in a small number of patients, and this should be assessed radiographically before operative intervention (39). Patients with quadriparesis have greater degrees of elbow flexion contracture. However, these contractures rarely affect their ability to use their motorized wheelchairs, computers, or communication boards. In the maximally dependent quadriparetic, contractures may become severe enough to affect hygiene and care. If skin breakdown develops or is imminent, surgery may be indicated.

Nearly three-fourths of patients with hemiplegia develop a forearm pronation contracture (34). The presence of a significant pronation contracture limits the ability to perform bimanual tasks (32, 35, 38). Individuals with contractures >60 degrees will either perform activities with one hand or use the dorsum of the affected hand or forearm to assist the unaffected hand. These individuals may benefit from surgical correction of their pronation deformity in order to improve the assistive function of that extremity. This can often be performed with simultaneous procedures to improve thumb-in-palm, wrist palmar flexion, or digital flexion deformities (40).

Wrist and hand involvement are common in cerebral palsy. Limited motor function occurs with (a) poor release because of wrist and finger flexor spasticity and weak digital extension, (b) inadequate grasp because of wrist palmar flexion spasticity and weak wrist extension, and (c) minimal pinch because of thumb-in-palm deformity. Discriminatory sensibility is deficient in more than 50% of these children (41). Their discriminatory sensibility may improve with hand surgery. Poor voluntary control of the upper extremity limits functional placement of the hand in space (32, 34). In addition, many of these children have visual and cognitive abnormalities that further impair hand use. At best, most patients with spasticity have assistive hand function.

These children generally posture into elbow flexion, forearm pronation, wrist and palmar flexion, thumb-in-palm, and interphalangeal swan-neck deformities. These deformities may result from both neuromuscular spasticity and contractures. Pronation deformity and thumb-in-palm contractures seem to affect function the most (34). The combination of neurologic impairment and disuse affects growth in length and girth of the affected arm and hand (34).



CHILDREN'S HOSPITAL ORTHOPAEDIC HAND SURGERY HEMIPLEGIA EVALUATION

MR No. _____
Date _____
Pt Name _____
Date of Birth _____
Sex (circle one) M F

Date / /
Evaluation Number
Involved Upper Extremity (circle one) L R
☐ New Patient ☐ Return Visit

Parent History — No / Yes / dk
Hand splint ever
Hand splint now
night use
day use
Hx Upper Ext surgery
Green Procedure
Web release
Elbow release
Pronator release
Other UE Surg
type: _____
(List date of surgeries, procedure, on back of page)
Hx seizures — No / Yes / dk
Intelligence — low / ave / high
Motivation

Deformities — No / Yes / nt
Thumb MCP hyperext
Thumb MCP stability
First Web Contracture
Swan Deformity
DIP hyperextension
Pronator Deformity
Elbow Flex Deformity
Thumb in Palm
Active Supinator?
Web Angle (degrees)

Size — AFF / UNAFF
Circumference
arm
forearm
Distance
O-MF
O-UL
Hand width

Sensation Exam — cos / ast / ind / not test
Function
Two-Point Desc (mm) — <6 / 6-10 / >10 / nt
Obj. Rec (# recog) — 0 / 1 / 2 / 3 / nt
Palm graphesthesia (# recog) — 0 / 1 / 2 / 3 / nt
Rough — No / Yes / nt
Proprioception — No / Yes

ADL (1=NU 2=AST 3=Ind)
☐ Buttons ☐ Hand ☐ Screw
☐ Pants/Shirt ☐ Groom ☐ Knob
☐ Zipper ☐ Teacup ☐ Bottle
☐ Utencils ☐ Knots ☐ Purse

Motor Exam — unable / min / max / nt
Grasp
Release
Grasp reflex — No / Yes / nt
Key pinch
Pulp pinch
Tip pinch
Zancolli — I / IIA / IIB / III / nt
Bleck — I / II / III / nt
Mowery

Voluntary Control

Tendon	Good	Fair	Poor
EPL			
AbPL			
EPB			
FPL			
FCR			
FCU			
BR			
ECRB/L			

THUMB-IN-PALM
-Type I MC Adduction
-Type II MC Ad + MCP flexion
-Type III MC Ad + MCP ext/instability
-Type IV MC Ad + MCP/IP flexion

Functional Classification

Class	
0 Does not use	Does not use
1 Poor Passive Assist	Uses as stabilizing weight only
2 Fair Passive Assist	Can hold object placed in hand
3 Good Passive Assist	Can hold onto object and stabilize for use in 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 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 the hand completely independently without reference to the other hand

FIGURE 22-1. Data sheet for prospective analysis of hemiplegic function used at Boston Children's Hospital.

Upper extremity classification systems have been used for assessing function in patients with cerebral palsy (39, 42) (Fig. 22-1) The House classification of function has 9 levels, extending from 0 (does not use) to 8 (complete spontaneous use) (Table 22-1). In this useful scheme, there are four subgroups of patient function: 0 (no use), 1 to 3 (passive assist), 4 to 6 (active assist), and 7 and 8 (spontaneous use). Because spasticity changes with stress, growth, and central nervous system changes, it may be difficult on any one visit to accurately define a patient's level of function. This system is used with the input of the patient, family, and physical therapist in order to best define a patient's overall status. It is used for assessing the outcome of treatments (40).

In addition, the Melbourne Assessment of Unilateral Upper Limb Function and the Pediatric Evaluation of Disability Inventory (PEDI) have been validated for upper-limb function assessment in children with cerebral palsy. The Melbourne Assessment of Unilateral Upper Limb Function has very high internal consistency and high inter- and intraobserver reliability, making it a reliable tool in assessing function and outcome of interventions in patients with cerebral palsy (43, 44). More recently, the Shriner's Hospital Upper Extremity Evaluation (SHUEE) and the Assistive Hand Assessment (AHA) have been validated to characterize upper-limb function and assess outcomes from nonoperative and surgical interventions (45, 46).

TABLE 22-1 House Classification of Upper Extremity and Hand Function for Patients with Cerebral Palsy

Level	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 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 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

Treatment

Nonoperative Care. In general, nonoperative treatment options include observation of the patient's growth and development; the use of therapy, including splints; injections, such as phenol or Botox; and performance of surgical reconstruction.

Physical therapy, starting in infancy, is the standard treatment for children with cerebral palsy. The rationale is that, although the central nervous system deficit is static, the peripheral manifestations of spasticity and muscle imbalance are dynamic and may be progressive with growth. By maintaining range of motion with passive therapy, it is hoped that contractures will be prevented (34, 47). In addition, it is hoped that the affected child is capable of learned motor behavior leading to functional improvement over time, developmentally, and through formal therapy (48, 49). At present, formal therapy is used during the period of infancy. This is most intense in the first year of life and progresses to a home program with less formal supervision. In many states, early intervention programs end at 3 years of age. Monitoring of function and range of motion are performed less regularly thereafter, facilitated in many instances through the school system. During growth spurts that increase spasticity and lessen range of motion, or with specific activities that the patient finds difficult to do, formal therapy is often reinitiated, though the therapeutic benefits of such interventions have not been statistically established (34).

In addition to passive range-of-motion and active-use programs, splints are often used. These may be daytime or nighttime splints. As Manske (47) has observed, it is unclear whether they are cost-effective and alter long-term outcome. However, most caretakers use splints in children with developing contractures. Daytime splints are recommended only if they improve function in patients with dynamic contractures.

Recently, there has been increasing enthusiasm for constraint-induced movement therapy (CIMT). Constraint therapy with casting or immobilization of the unaffected limb has been advocated in order to improve the function of the affected limb in children with hemiplegia and prevent "developmental disuse" of the affected hand and limb (50). A single randomized study has shown this to be effective. It has been shown that patients with hemiplegia do not maximally utilize their motor capabilities in the affected limb in functional tasks (51). Constraint therapy may better enable these patients to maximize their motor function in the affected limb, but there are emotional issues that make this treatment difficult for some families and caregivers. In a small cohort of patients with cerebral palsy (52), it has been shown that functional electrical stimulation (FES) is effective in the short term (up to 3 months) in improving hand function, when applied to the extensor muscles of the wrist and hand. Its long-term effectiveness and applicability to all types, degrees, and ages of patients with cerebral palsy is still unclear.

Injection may provide useful information about the outcome of surgical procedures. At present, botulinum toxin A (Botox, Allergan, Irvine, CA) is the most commonly used pharmacologic agent for neuromuscular injection (40, 53), replacing xylocaine (54, 55), and phenol (56, 57). It is used at an initial dose of 1 to 2 U/kg of body weight and should not exceed 6 U/kg/mo. Injections into the pronator teres, flexor carpi ulnaris (FCU), and adductor pollicis are most often performed. Therapy should be performed aggressively to stretch agonistic musculotendinous units and strengthen antagonists. To date, botulinum toxin A has been most effective in patients with high motivation, good motor learning capacity, and no fixed contracture or limiting spasticity (58). Its role in patients with contractures is limited and less effective, although these patients may show the greatest involvement. Its effectiveness in young children has not yet been studied critically (59–61). There are several ongoing prospective studies of botulinum toxin A injections in the upper extremity and hand, so more definitive information should soon be available on the indications and effectiveness of its use in all age groups and at all levels of involvement. At this stage, in our institution, we use botulinum toxin A injections in the upper limbs in (a) younger patients with marked spasticity or developing contractures and (b) older patients with limitations, for whom surgery is not indicated. Complications involve the formation of antibodies to Botox that limit further effective injections and leading to deterioration of upper limb function for the first 1 to 3 weeks post injection in some patients.

Operative Care. The broad indications for surgery in patients with cerebral palsy include (a) contractures that cause hygiene and care problems not solved by therapy, splints, or casts; (b) muscle imbalance or contractures that cause functional deficits that may be improved by tendon transfers, musculotendinous lengthening, and/or joint stabilization procedures; and (c) aesthetic concerns (29, 32–34, 62). It may be difficult to identify the individual who will have improved function through surgical reconstruction. As Smith so aptly pointed out, careful preoperative assessment is necessary in order to select the appropriate patients and operations (28). Video recordings of activities of daily living and validated multiple-task assessment scales, such as the Jebsen scale, can be helpful in defining functional limitations. Preoperative video assessments using standardized and validated classification systems (e.g., House classification, SHUEE) are reliable and useful for surgical planning (63).

Surgery has been shown to effectively improve the level of function in all forms of cerebral palsy (40, 64, 65). The best candidates are patients with hemiplegia and good voluntary control, sensibility, and motivation. The principle of surgery is to restore muscle imbalance by lengthening or releasing tight, spastic muscles and by augmenting weak, stretched muscles via musculotendinous lengthenings, tendon transfers, and tenodesis procedures. Unstable joints need to be stabilized by capsulodesis or arthrodesis procedures in order to maximize the outcome of tendon reconstruction. Multiple upper extremity rebalancing procedures performed under one anesthesia are preferred. This can also be performed in conjunction with simultaneous lower-extremity procedures. It cannot be stressed enough to the patient and the family that surgery will not achieve a normal hand. Even the best outcome will result in deficiencies of function, aesthetics, and sensibility. However, in properly selected patients, surgery will clearly improve function and result in patient satisfaction (40, 64, 65). This is particularly evident in individuals using the dorsum of the hand or forearm for bimanual tasks or those with considerable thumb-in-palm deformity (64). The goal must be well defined and specific to the peripheral manifestations of the incurable, central nervous system disorder of cerebral palsy.

Mital (36) cited excellent results with surgical release of elbow flexion contractures in patients with hemiplegia. He recommended release of the lacertus fibrosus, Z-lengthening of the biceps tendon, and musculotendinous lengthening of the brachialis. In mild contractures, release of the lacertus fibrosus and musculotendinous lengthening of the brachialis alone may be sufficient.

More extensive elbow contractures are present in severe quadriparetics. The Z-lengthening of the biceps tendon and the release of the brachialis fascia advocated by Mital (36) are not sufficient to obtain adequate release in these patients. In patients with contractures >90 degrees and skin breakdown, extensive release of the muscle origins from the medial and lateral epicondyles, lengthening of biceps and brachialis tendons, and anterior elbow capsule is necessary so as to solve the hygiene and care-related problems that accompany these conditions. The neurovascular bundle becomes the length-limiting factor. Manske (66) has alternatively proposed additional peritendinous adventitial stripping in efforts to ablate the afferent nerve signals causing elbow flexion spasticity.

As cited above, forearm hyperpronation significantly limits hand function (66) in patients with hemiplegia, and is often seen with wrist and finger flexion deformities. The flexor carpi ulnaris (FCU) is usually the major deforming force at the wrist. Transfer of the FCU to the wrist extensors alleviates the deformity and improves the strength of the antagonist. On occasion, the extensor carpi ulnaris (ECU) is the primary deforming force, as noted by more ulnar deviation than palmar flexion at the wrist. In these situations, the ECU is transferred to the extensor carpi radialis brevis (40). Simultaneous musculotendinous lengthenings of the finger flexors are necessary if the extrinsic finger flexors are tight in the neutral wrist position (33). Otherwise, the patient will develop a disabling clenched fist postoperatively. Z-lengthenings, superficialis-to-profundus flexor tendon transfers, and bony pro cedures are reserved for patients with severe contractures and limited function. In the patient with passive but no active digital extension, the FCU, ECU, or pronator teres (PT) can be transferred into the extensor digitorum communis, with or without additional tendon transfers to the wrist extensors. This will improve both wrist and digital extension.

Transfer of Flexor Carpi Ulnaris for Wrist Flexion Deformity. Wrist flexion deformity is a frequent problem in children with cerebral palsy (Figs. 22-2 to 22-4). There are two aspects to the problem. The first aspect, and the one most often discussed in relation to correction of the deformity, is function. The wrist is often held in flexion, pronation, and ulnar deviation, with the inability to dorsiflex the wrist or to release a grasp.

The second aspect is cosmetic. Most authorities on the subject rarely consider this to be a worthwhile goal of surgery. However, for many patients, especially those with hemiparesis who are attending regular schools, this can be an important consideration.

The criteria for obtaining a good result with this operation were briefly mentioned in the follow-up article on the patients who have undergone this procedure (67, 68), which lists eight prerequisites for this procedure. These requirements are mentioned here as factors to be considered, some more strongly than others, rather than as absolute prerequisites.

1. The flexor carpi ulnaris should have good motor power.
2. There should be good passive dorsiflexion of the wrist, extension of the fingers, and supination of the forearm.
3. The patient should be able to extend the fingers actively, with the wrist held in neutral position.
4. The patient should have good voluntary control over placement of the arm.
5. There should be adequate sensory function in the hand.
6. The patient should have reasonable intellect.
7. The patient should be old enough to comply with the postoperative therapy program.
8. No movement disorder, such as athetosis, should be present.

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Transfer of Flexor Carpi Ulnaris for Wrist Flexion Deformity (Figs. 22-2 to 22-4)

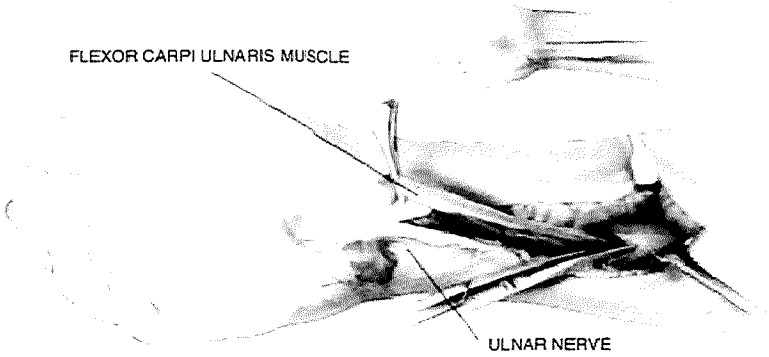

FIGURE 22-2. Transfer of Flexor Carpi Ulnaris for Wrist Flexion Deformity. Although the procedure is usually performed with the patient in the supine position, the prone position facilitates exposure in the patient with an internal rotation contracture of the shoulder coupled with a pronation contracture of the forearm.

The procedure begins by detaching the flexor carpi ulnaris tendon and by freeing up the muscle belly from its extensive origin along the ulna. Although two separate incisions were made originally, it makes more sense to make one incision because most of the dissection is done in the distal aspect of the forearm. The incision starts distally, at the flexor crease of the wrist and directly over the flexor carpi ulnaris tendon, where it inserts into the pisiform bone. The incision extends about midway up the forearm. A right-angled retractor can be used to elevate the skin at the proximal extent of the wound, allowing dissection to extend proximally as far as the junction of the middle and distal one-third of the forearm. The fascia over the tendon and the lateral aspect of the muscle are divided.

Because the ulnar nerve lies directly under the tendon, caution must be exercised in freeing it from the pisiform bone. After the tendon is divided, the muscle fibers along the lateral aspect of the muscle originating from the ulna are identified easily. These fibers must be freed by dissecting them off the periosteum of the ulna. The flexor carpi ulnaris receives its nerve supply from the underlying ulnar nerve. As the dissection proceeds proximally, it is important to identify and protect these branches. This dissection needs to extend proximally at least to the upper one-third of the forearm—far enough to allow the muscle belly to be directed around the medial border of the ulna in a straight line.

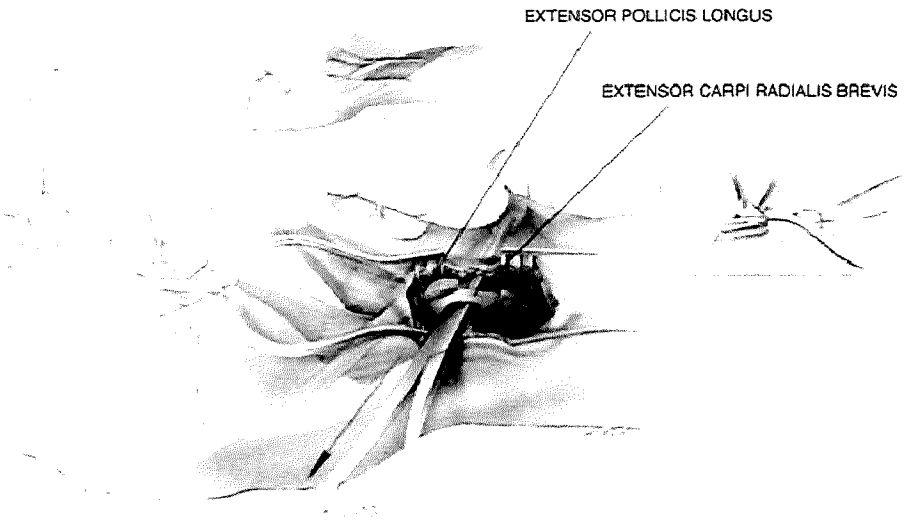

FIGURE 22-3. The second incision is made directly over the extensor carpi radialis and brevis tendons, starting at the extensor crease of the wrist and extending proximally for 3 to 4 cm. After incising the fascia, the two tendons can be identified: the most radial tendon is the extensor carpi radialis longus and the more ulnar one is the brevis. Inserting the transfer into the extensor carpi radialis longus provides a better supination force and is more effective in overcoming ulnar deviation, whereas inserting the transfer into the brevis provides a more central pull. A subcutaneous tunnel is dissected from the proximal extent of the volar incision around the subcutaneous medial border of the ulna. A tendon forceps is used to bring the flexor carpi ulnaris around the medial aspect of the ulna through the subcutaneous tunnel and into the second incision on the dorsal aspect of the wrist. When the surgeon is confident that a sufficient portion of the intermuscular septum has been excised and that the tendon is running along a relatively straight path, the first incision is closed.