CHAPTER 22

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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

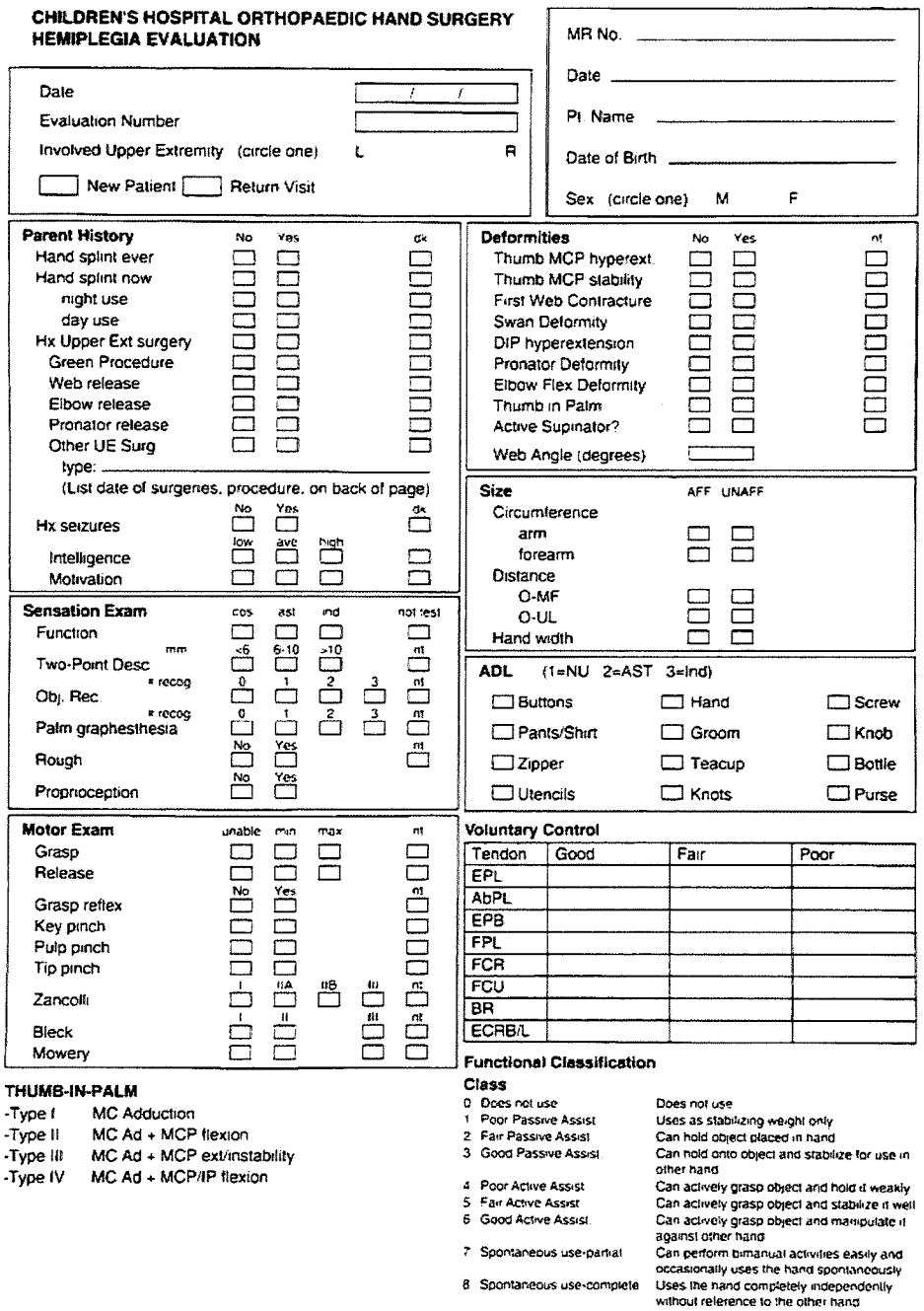
Cerebral Palsy. Cerebral palsy is a nonprogressive disorder of the central nervous system. It occurs in 5 in 1000 live births and may be caused by perinatal anoxia, intraventricular hemorrhage, or congenital cerebral vascular accidents. It occurs most commonly in premature infants weighing <1500g (27, 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)

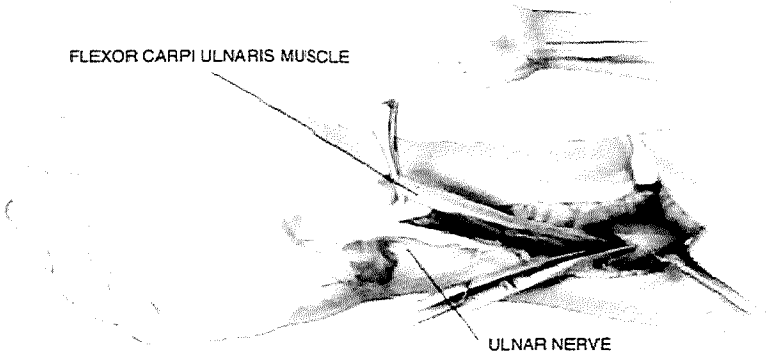

FIGURE 22-2. Transfer of Flexor Carpi Ulnaris for Wrist Flexion Deformity. Although the procedure is usually performed with the patient in the supine position, the prone position facilitates exposure in the patient with an internal rotation contracture of the shoulder coupled with a pronation contracture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

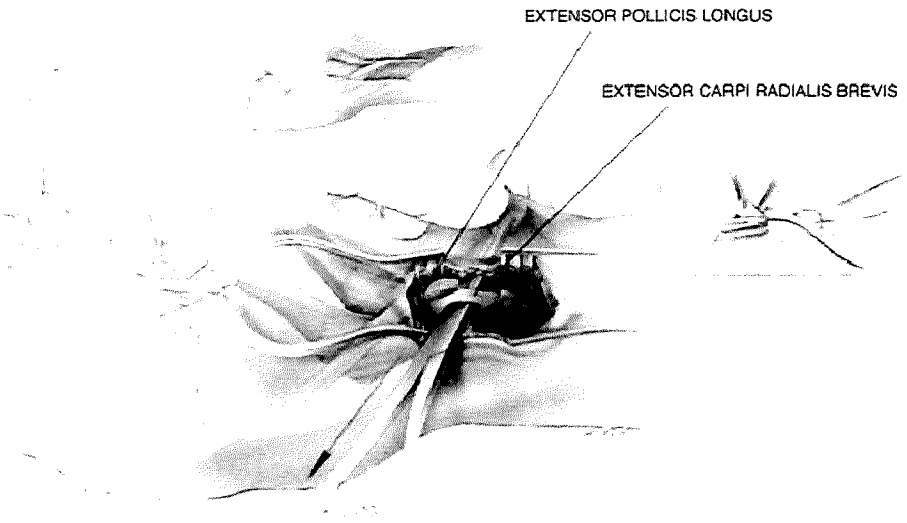

FIGURE 22-3. The second incision is made directly over the extensor carpi radialis and brevis tendons, starting at the extensor crease of the wrist and extending proximally for 3 to 4 cm. After incising the fascia, the two tendons can be identified: the most radial tendon is the extensor carpi radialis 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.

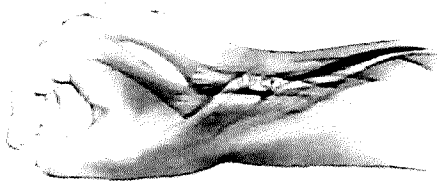

FIGURE 22-4. The flexor carpi ulnaris is then sutured into the desired tendon. During this procedure, the wrist is held at about 45 degrees of extension and the forearm is held in maximal supination. After the tendon anastomosis is complete, the wrist should flex passively at least 15 degrees past neutral with the fingers simultaneously going into extension.

The second wound is closed, and the patient is placed in a long arm cast with the wrist in slightly less than maximal dorsiflexion and the forearm in full supination. Because the underlying pathology is spasticity, the thumb should be incorporated in the cast in a position of abduction, with the metacarpal joints flexed about 15 degrees and the interphalangeal joints in neutral position.

Hoffer et al. (69) studied patients with spasticity by using dynamic electromyography and noted that the flexor carpi ulnaris cocontracted with the finger extensors. Because releasing is often more of a problem than grasping, they suggested transferring the flexor carpi ulnaris into the extensor digitorum communis to improve both release of grasp and wrist extension. In a subsequent report, Hoffer et al. (70) demonstrated the effectiveness of this in carefully selected patients and described the indications. In addition to failure in achieving the desired functional goals, the most common complication of this procedure is a wrist extension contracture. Hoffer and colleagues claimed that transferring the flexor carpi ulnaris into the extensor digitorum communis obviates this problem (70).

Thumb-in-palm deformity will limit dynamic pinch and grasp function, and make hygiene difficult to maintain in severe contractures. Static contractures in the web space are corrected with web-space Z-plasties and adductor releases. Hoffer et al. (71) have shown by dynamic electromyography that release of the transverse adductor alone may lead to better pinch postoperatively in selected patients. At times, the static contractures include the flexor pollicis longus and brevis, and these muscles need to be appropriately lengthened or released. Dynamic rebalancing is performed with tendon transfers to the weak abductors and extensors of the thumb. The potential donor muscles used are numerous, and include the palmaris longus, flexor carpi radialis, and brachioradialis, among others. The recipient tendons include the extensor pollices brevis and longus and the abductor pollicis longus. The treatment for each patient should be individualized in order to correct his or her deformity and imbalance. Finally, the metacarpophalangeal (MCP) joint should be stable postoperatively. In most patients, this is achieved by muscle rebalancing. On occasion, a capsulodesis or an arthrodesis procedure should be performed. Selected patients with thumb-in-palm deformity respond very favorably to surgical intervention (64).

Correction of Thumb-in-Palm Deformity in Cerebral Palsy. Children with cerebral palsy frequently have difficulty with hand function, and often the most noticeable associated deformity is of the thumb (Figs. 22-5 to 22-8). Several authors have discussed in detail the indications for correction of such deformities (38, 72, 73). Most authors have stressed the importance of the preoperative evaluation of outcome, with assessment of voluntary control, sensation, cognition, and the ability to cooperate with a postoperative program, these being the most important factors to consider in the assessment of outcome.

House et al. (38) have classified the deformities by an assessment of the thumb's function rather than by its static position.

Type I: Metacarpal adduction contracture—this is the most common deformity and is usually associated with a contracture of the first thumb web space. It is caused by spasticity and contracture of the adductor pollicis and first dorsal interosseous muscles.

Type II: Metacarpal adduction contracture and metacarpophalangeal flexion deformity—in this deformity, the interphalangeal joint remains mobile and the metacarpophalangeal joint is fixed in flexion by contracture of the flexor pollicis longus.

Type III: Metacarpal adduction contracture combined with a metacarpophalangeal hyperextension deformity or instability—this deformity is caused by spasticity of the extensor pollicis longus in the absence of spasticity in the flexor pollicis longus.

Type IV: Metacarpal adduction contracture combined with metacarpophalangeal and interphalangeal flexion deformities—this is usually caused by spasticity of the flexor pollicis longus and the intrinsic muscles of the thumb, but it may be caused by isolated spasticity of the flexor pollicis longus.

The steps that are taken to correct the deformities are considered in three categories: release of the skin and muscle contractures, augmentation of the weak muscles, and stabilization of joints.

Finally, some patients with cerebral palsy have disabling swan-neck deformities of the interphalangeal joints. If the fingers extend at the proximal interphalangeal (PIP) joint beyond 40 degrees and lock, the position can limit grasp and cause pain. Multiple operations have been advised, including flexor digitorum superficialis tenodesis (33), intrinsic muscle slide (33), lateral band rerouting (74), spiral oblique ligament reconstruction, and resection of the motor branch of the ulnar nerve. The lateral band rerouting procedure provides both intrinsic and extrinsic rebalancing and is effective in correcting the problem (75).

In summary, patients with cerebral palsy who have functionally limiting dynamic spasticity and fixed contractures of the wrist and hand may benefit from surgical reconstruction. Over a 25-year experience, House reported that, for 718 procedures in 134 patients with cerebral palsy, the functional improvement was 2.6 functional levels on the House scale of 0 to 9 (35). Patients with fair and good voluntary control had significantly better improvement in functional use scores than those with

Text continued on page 906

Correction of Thumb-in-Palm Deformity in Cerebral Palsy (Figs. 22-5 to 22-8)

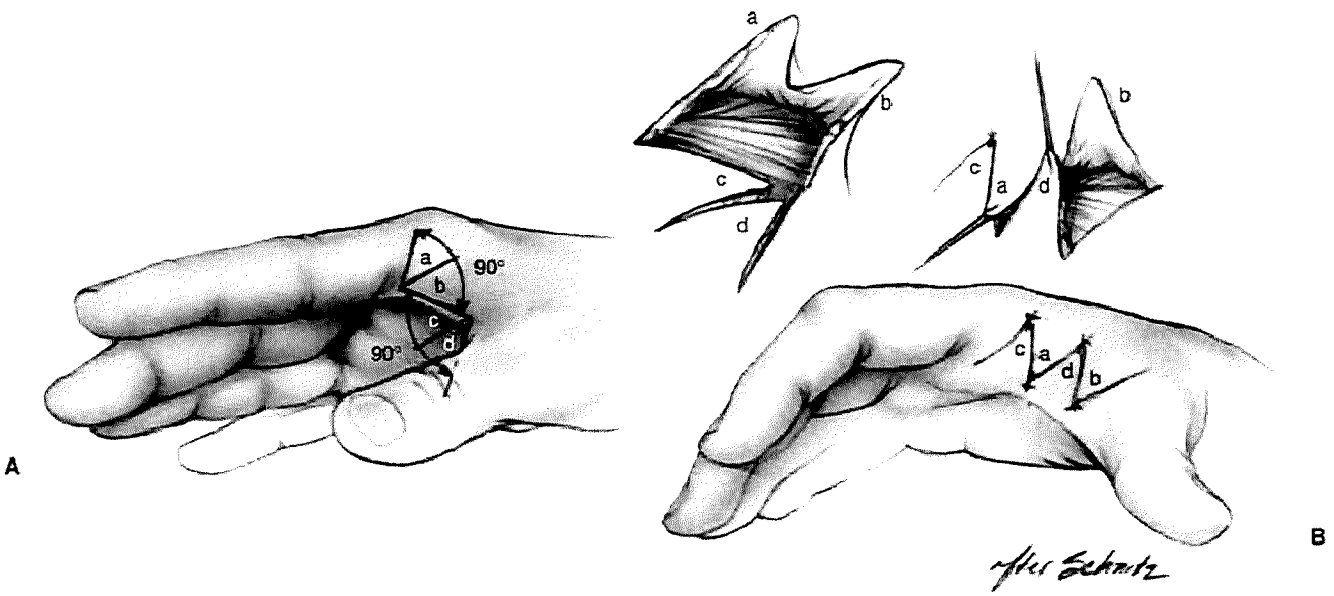

FIGURE 22-5. Correction of Thumb-in-Palm Deformity in Cerebral Palsy. The release of the contracted first thumb web space is achieved by Z-plasty incision, through which the tight dorsal fascia and the muscles causing the contracture in the first place, the adductor pollicis longus and the first dorsal interosseous muscles, can be divided. The incision is a four-flap Z-plasty. This Z-plasty has been described using angles of 120 and 60 degrees or, as illustrated here, using angles of 90 and 45 degrees. Each limb of the incision should be of equal length. The first limb of the incision is made along the line of the maximal contracture. **A:** At each end of this incision and at 90 degrees to it, another incision is made. This limb should be equal in length to half the length of the longitudinal limb. Finally, a third limb is added to each end of the incision, which bisects the right angle made by the first two limbs. This should be equal in length to the second limb. **B:** The incision is closed by transposing the flaps.

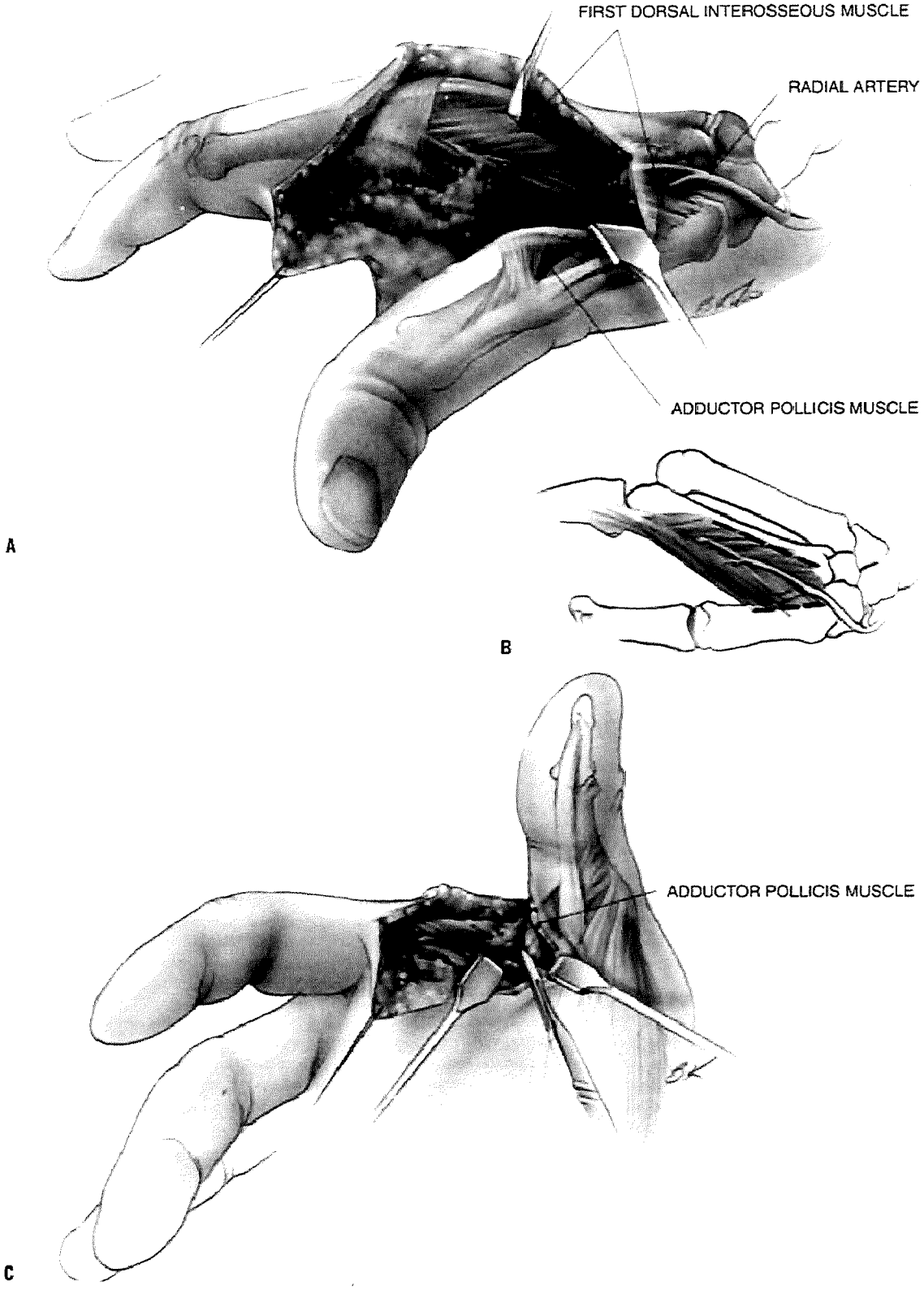

FIGURE 22-6. **A:** After the flaps of the incision are developed and retracted and the dorsal fascia is divided, the tight adductor pollicis and the first dorsal interosseous muscle are identified easily. The origin of the first dorsal interosseous muscle that arises from two heads, one on the first metacarpal and one on the second metacarpal, is released first. Care must be taken as the radial artery passes between these two heads to form the deep palmar arch. The portion inserting on the first metacarpal is released first. **B:** It usually is necessary to release at least a portion of the head originating on the second metacarpal because the two heads join together close to their origin. After this, the adductor pollicis muscle is released by partially dividing it in its intramuscular portion. This muscle can be found running obliquely beneath the first dorsal interosseous muscle. **C:** Its division is accomplished more easily, however, from the palmar aspect of the wound. If this does not provide sufficient abduction, it is necessary to release it from its origin on the third metacarpal, as subsequently described.

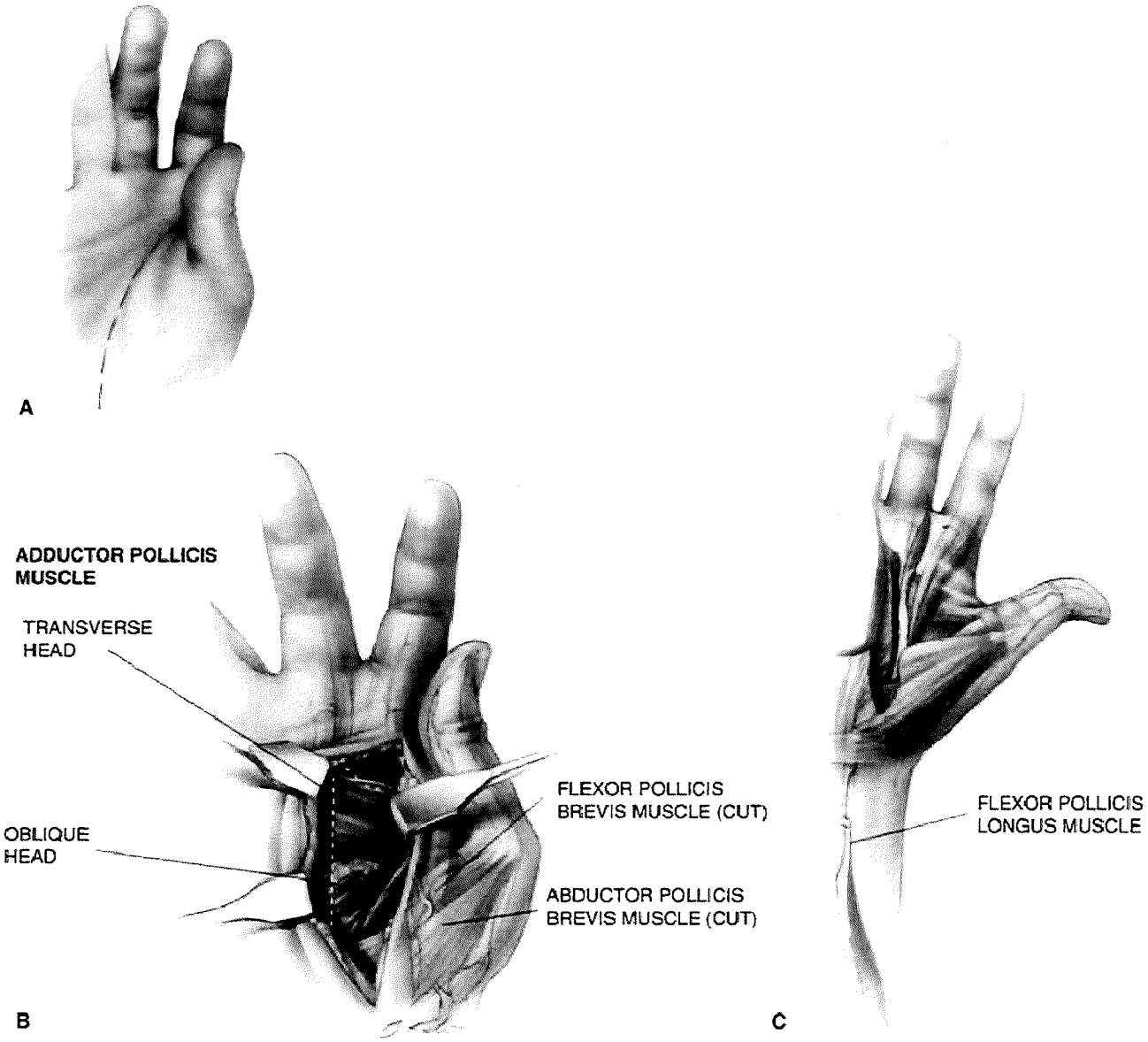

FIGURE 22-7. In the more severe type II deformities, it is usually necessary to release the origin of the adductor pollicis muscle from the third metacarpal and the origin of the flexor pollicis brevis from the flexor retinaculum. If necessary, a portion of the abductor pollicis brevis can also be released (Ulthoff HK. *The Embryology of the Human Locomotor System.* Berlin, Germany: Springer-Verlag, 1990.). A palmar incision following the crease of the thenar eminence is used. **A:** The proximal portion of this incision lies over the third metacarpal. **B:** After the skin and the fascia are divided, the flexor tendons of the middle finger are retracted in the ulnar direction, whereas the neurovascular bundle and the superficial palmar arch, along with the flexor tendons of the index finger, are retracted in the radial direction. This exposes (distally to proximally) the transverse head of the adductor pollicis, the oblique head of the adductor pollicis, the flexor pollicis brevis, and the abductor pollicis brevis, overlying the opponens pollicis muscle. **C:** The adductor pollicis muscle is stripped off the third metacarpal, whereas the origin of the flexor pollicis brevis is detached from the flexor retinaculum (transverse carpal ligament).

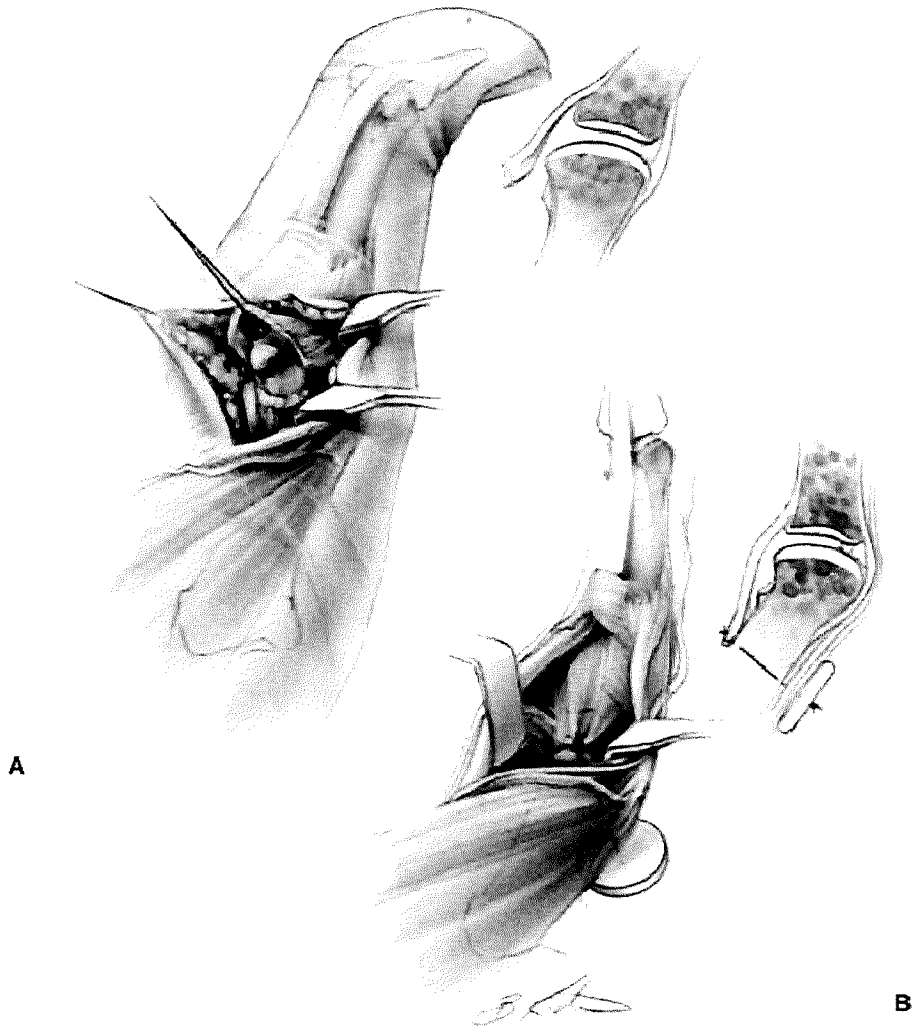

FIGURE 22-8. In type III deformities, it may be necessary to stabilize the metacarpophalangeal joint of the thumb. This can be done by arthrodesis. In the growing child, the stabilization can be accomplished by denuding the cartilage from the joint surface and by fixing the joint with an intramedullary pin, thereby sparing the growth of the physis.

Another method that preserves more of the function of the thumb, however, is described by Filler and colleagues (Tabin C. The initiation of the limb bud: growth factors, Hox genes, and retinoids. *Cell* 1995;80:671–674.). Through a V-shaped incision over the volar aspect of the metacarpophalangeal joint, as described for release of trigger thumb, the sheath of the flexor pollicis longus is partially excised to expose the tendon. As with release of trigger thumb, it is important to identify and retract the neurovascular bundles carefully, particularly the radial digital nerve that lies just beneath the skin and crosses the operative site. This exposes the volar plate or capsule. Its proximal insertion **(A)** is incised and freed. Both sides are then incised just outside of the sesamoid bones so that only the distal attachment remains. The joint is flexed 30 to 35 degrees and transfixed with a small Kirschner wire. The capsule is advanced proximally until it is taut. At this new point of insertion, a small groove is cut into the cortical bone, and a small drill hole is made from this groove to the dorsal surface of the metacarpal. A pull-out wire or a strong absorbable suture is passed through this hole and tied over a button on the dorsum of the thumb **(B)** to secure the insertion of the volar plate into this groove.

In type IV deformity, it is necessary to lengthen the flexor pollicis longus. AZ lengthening is easily accomplished proximal to the wrist (see Fig. 22-7C).

Numerous muscles can be used to augment the functions of the abductor pollicis longus, the extensor pollicis brevis, or the extensor pollicis longus. The palmaris longus, the brachioradialis, and the flexor carpi radialis muscles are commonly used.

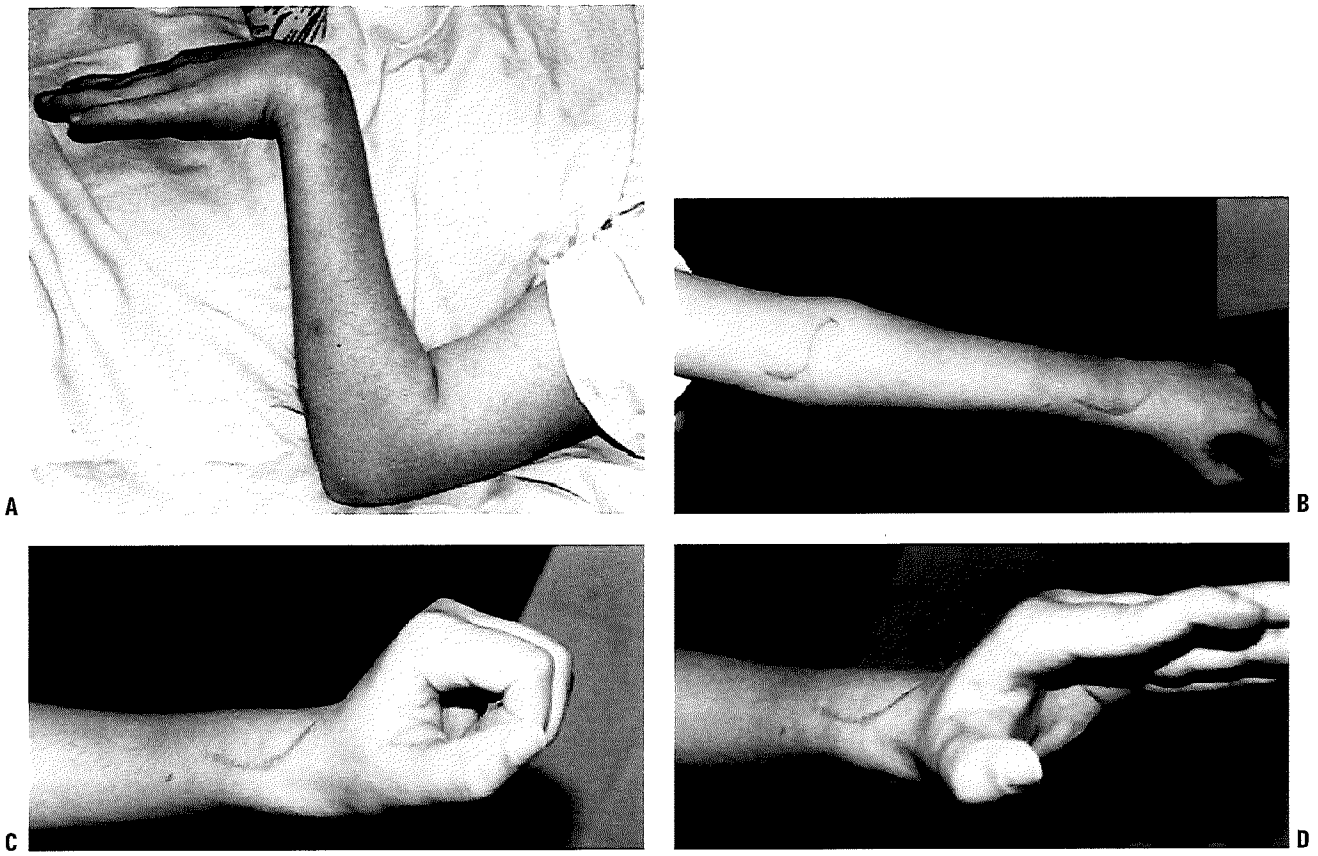

FIGURE 22-9. Clinical photographs of hemiplegic tendon transfers for improving hand and upper limb function. **A:** Preoperative view of dynamic elbow flexion, forearm pronation, wrist flexion, and ulnar deviation with poor assist function. **B:** Postoperative active elbow extension with maintenance of active elbow flexion. **C:** Postoperative active wrist extension with the thumb out of the palm for active pinch. **D:** Postoperative active grasp function with the thumb abducted and extended actively. (Courtesy of Ann Van Heest, MD.)

poor voluntary control. Often, the more severely involved patients (House levels 0 to 2) respond best to musculotendinous lengthenings, tenodesis, and joint stabilization procedures. More functional patients (House levels 3 to 6) improve with dynamic tendon transfers and releases. Both groups of patients tolerate multiple simultaneous procedures (Fig. 22-9). However, surgery will not create a normal hand. The goals of surgery need to be realistic and attainable. In properly selected patients, surgery will improve assistive function and cosmesis. For many of these children, especially adolescents, and their families, the functional and cosmetic improvements are quite marked and satisfying.

Complications. Deformity may recur, or function may fail to improve. Proper preoperative selection so as to assess functional deficits and the patient's level of cooperation may lessen the risk of these problems postoperatively (29). Athetosis and movement disorder patients can get the opposite deformity. Hematoma formation, wound breakdown, and infection can occur after extensive elbow releases (30). The institutionalized patients with quadriparesis may be most at risk. If wound dehiscence occurs and the joint is exposed, coverage with a rotation flap is the treatment of choice.

Brachial Plexus Birth Palsy. Brachial plexus birth palsy is rare, with an incidence between 0.1% and 0.4% of live births (76–78). Fortunately, most infants with minor birth palsies recover fully. These are the infants who initiate recovery of all muscle groups in the first 1 to 2 months of life. However, permanent impairment does occur in infants who do not initiate antigravity motor recovery before 5 to 6 months of life (79–83). Most infants have involvement of the upper trunk (C5-C6, causing Erb palsy and, often, additional involvement of C7). Less often, the entire plexus (C5-T1) is affected. In rare instances, the lower trunk (C7-T1, causing Klumpke palsy) is most affected.

Perinatal risk factors include infants who are large for gestational age; prolonged labor; previous births with brachial plexopathy; difficult delivery, including extraction techniques; and fetal distress. Shoulder dystocia is the mechanical factor that leads to an upper trunk lesion in the difficult vertex delivery. Difficult arm extraction in a breech delivery can result in an avulsion injury of the upper trunk (31). The degree of impairment is related to the level and magnitude of injury to the plexus. Neural injury is defined by the type (stretch, rupture, avulsion) and severity (Sunderland grades I to V). Prognosis

by natural history has been best defined by the spontaneous rate of recovery of muscle strength in the first 3 to 6 months of infancy. Gilbert and Tassin (83) described the recovery of antigravity biceps function in infancy as a predictor of the outcome of spontaneous recovery. This finding was confirmed in a similar study by Waters (82). These studies demonstrated that infants who did not recover biceps function by 3 months of life were not normal after 2 years of age. Gilbert et al. recommend microsurgical reconstruction of the plexus in the first 3 to 6 months of life for infants who fail to recover biceps function (83–85)(61). Michelow et al. (86) noted that return of biceps function alone had a 12% error rate in predicting outcome, as defined by long-term antigravity muscle strength. By using elbow flexion, elbow extension, wrist extension, finger extension, and thumb extension (the Toronto scale), their error rate for predicting outcome decreased to 5%. In this system, each muscle group is scored as 0 (no motion), 1 (motion present but limited), or 2 (normal motion), for a maximum score of 12. A score of <3.5 predicted a poor long-term outcome without microsurgery. In all studies, the presence of Horner syndrome, total plexus involvement, and failure of return of function by 3 to 6 months of life portend a poor long-term outcome.

Clinical examination of an infant for motor-sensory function can be challenging. It is important to distinguish true paralysis from the pseudoparalysis that comes with a neonatal clavicle fracture, humeral fracture, or septic shoulder. There can be clinical overlap because fractures also occur in infants with shoulder dystocia and infantile brachial plexopathy. Plain radiographs will identify the infant with clavicle and humeral fractures. In the neonate, these fractures heal within 10 to 21 days. If restriction in range of motion persists after 1 month, there is most likely a concomitant brachial plexopathy. In the rare infant with a septic shoulder, there will be evidence of systemic illness (altered vital signs, change in appetite, toxicity), marked irritability with glenohumeral range of motion, and abnormal white blood cell count (87). If there is doubt, ultrasonography will reveal the effusion, and arthrocentesis will be confirmatory.

The pupils should be assessed for Horner syndrome. Motor examination is limited to observation of spontaneous activity and stimulated movement by primitive reflexes in the infant. The Moro startle reflex and the asymmetric tonic neck reflex can elicit upper trunk movement in infants in the first 6 months of life. Classification of nerve injury in the ambulatory child has included physical assessment according to the Mallet system. The modified Mallet system classifies upper-trunk function by grading hand-to-mouth, hand-to-neck, and hand-on-spine activity, global abduction, and global external rotation from 0 (no function) to V (normal function). Grades II, III, and IV are illustrated (Fig. 22-10). The Hospital for Sick Children Active Movement Scale is also utilized to define the degree of motor recovery, grades I through IV being gravity-assisted and grades V through VII being against gravity. These classification systems have been shown to be reliable in intra- and interobserver analysis (88).

Radiography can demonstrate an associated fracture of the clavicle or proximal humerus. Radiographic assessment of the severity of brachial plexus injury may be attempted using

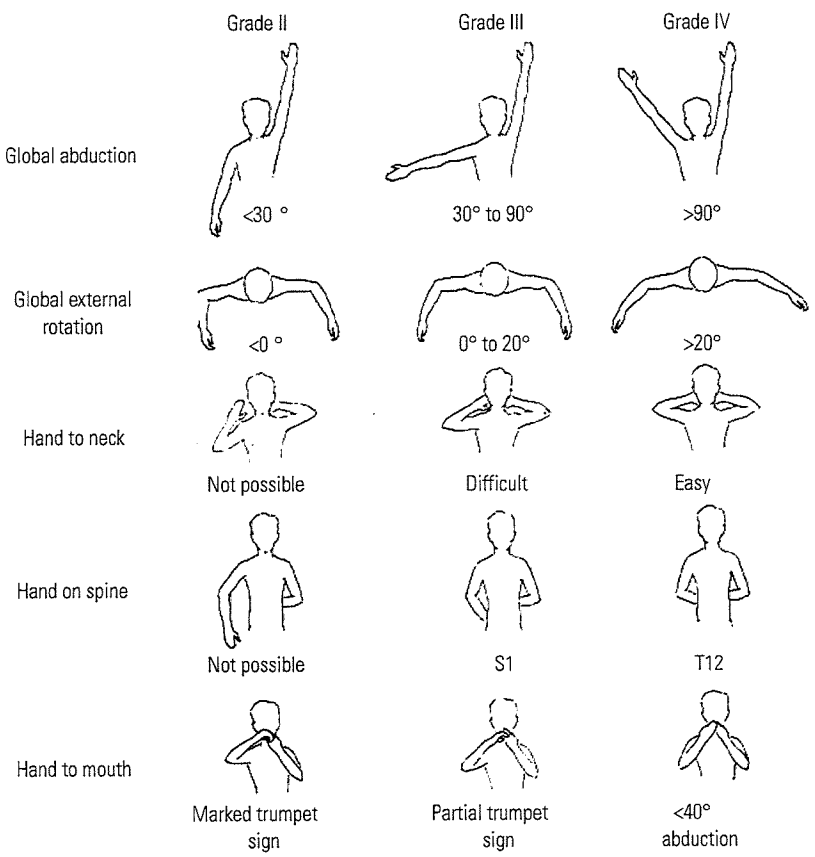

FIGURE 22-10. Mallet classification for function about the shoulder in patients with brachial plexus birth palsy. Grade 0 is no function; grade V is normal function; and grades II through IV are depicted for hand-to-mouth, hand-to-neck, external rotation, and hand-to-sacrum activity.

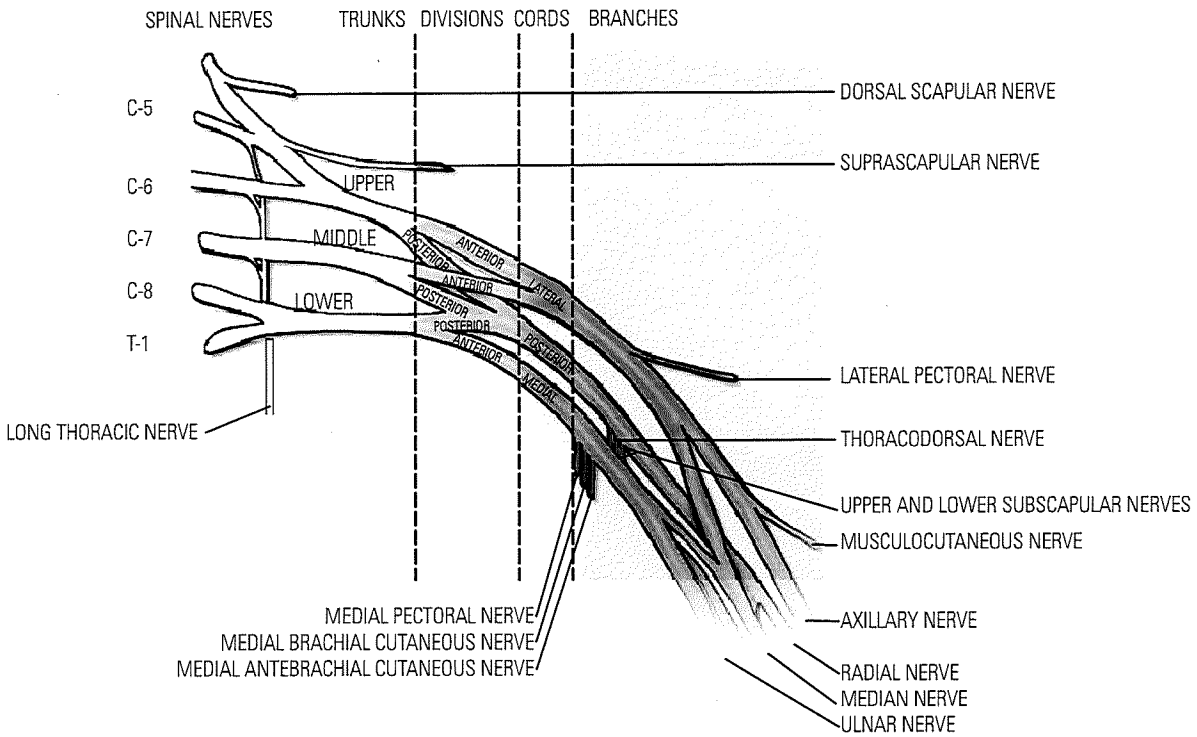

FIGURE 22-11. Brachialplexus anatomy.

myelography, combined computed tomography (CT) scan and myelography, and magnetic resonance imaging (MRI). Kawai et al. (89) compared the results of all three techniques with operative findings. MRI and combined myelography and CT scan were more reliable than myelography alone. The presence of large diverticulae and meningoceles was indicative of root avulsion. Small diverticulae were diagnostic only 60% of the time. Electrodiagnostic studies, with electromyography and nerve conduction studies, are diagnostic of avulsion if there is no reinnervation after 3 months of age. However, the presence of reinnervation does not indicate the long-term quality of muscle recovery.

Pathoanatomy. Understanding the normal anatomy of the brachial plexus is critical to assessing and caring for an infant or a child with brachial plexus palsy (Fig. 22-11). The brachial plexus supplies every muscle of the upper extremity except the trapezius. It is made up of spinal cord nerve root contributions from C5 to T1. Prefixed cords (22% of the specimens) receive a contribution from C4. Postfixed cords are rare (1%) and receive a contribution from T2. The C5 and C6 roots join to form the upper trunk. The C7 root alone becomes the middle trunk. The C8 and T1 roots become the lower trunk. Each trunk has an anterior and a posterior division. The anterior divisions of the upper and middle parts of the trunk form the lateral cord. The posterior divisions of all three parts of the trunk form the posterior cord. The anterior division of the lower trunk continues as the medial cord. The terminal branches of the cords form the major nerves of the upper extremity. The upper and lower subscapular and thoracodorsal nerves branch off from the posterior cord before it bifurcates into the radial and axillary nerves. The medial cord branches are the medial pectoral, medial brachial cutaneous, and medial antebrachial cutaneous nerves, terminating in the medial contribution to the median nerve and the ulnar nerve. The lateral cord supplies the lateral pectoral nerve and the lateral branch of the median nerve, and terminates as the musculocutaneous nerve. In infantile brachial plexopathy, any of these nerves can be affected. However, the most severe injuries are avulsions of the nerve roots. The most common injuries are postganglionic ruptures of the upper trunk.

Treatment

Nonsurgical. As mentioned above, all infants with brachial plexus birth palsies should be monitored for spontaneous recovery during the first 3 to 6 months of life. During this time, it is important to maintain glenohumeral range of motion, especially passive external rotation (90). This will lessen the risk of progressive glenohumeral dysplasia and dislocation (80–82, 90–93).

Many infants will initiate recovery in the first 6 to 8 weeks of life, and progress to a normal result. Infants who do not demonstrate recovery until after 3 to 6 months of life may be candidates for microsurgery or reconstructive surgery.

Microsurgery. The optimal timing for microsurgical intervention is still debated. The range used clinically is from 1 month to after 6 months of life (31, 76, 82–84). The indications include absence of biceps recovery, Toronto score <3.5, and total plexopathy with Horner syndrome. At present, most centers throughout the world agree that an infant with a flail extremity and Horner syndrome should have microsurgical reconstruction between by 3 months of life. A child with complete absence of upper trunk function (shoulder abduction, elbow flexion)

TABLE 22-2 Computed Tomography/Magnetic Resonance Imaging (CT/MRI) Classification of Glenohumeral Deformity in Chronic Brachial Plexus Birth Palsies

Type	CT scan/MRI Findings
I	Normal glenohumeral joint
II	Minimal glenoid hypoplasia (>5 degrees increased retroversion)
III	Posterior subluxation of the humeral head
IV	Development of a false glenoid
V	Posterior flattening of the humeral head
VI	Infantile dislocation
VII	Proximal humeral growth arrest

Findings are additive, with increasing severity from type I to type V.
From Waters PM, Smith GR, Jaramillo D. Glenohumeral deformity secondary to brachial plexus birth palsy. *J Bone Joint Surg Am* 1998;80:668–677, with permission.

should have surgery between 3 and 6 months of life. However, the controversy regarding the best timing for microsurgery, whether it should be done at 3, 4, 5, 6, or even 9 months, is still unresolved. This creates difficulties for parents who are trying to do what is best for their infants. There is an ongoing prospective study sponsored by the Pediatric Orthopaedic Society of North America (POSNA) that hopes to resolve this issue.

Microsurgery involves resection of the neuroma and bypass nerve grafting or nerve transfer procedures. On the basis of the information published in peer-reviewed journals, there is no role for neurolysis alone in a patient at any age, especially in the infant older than 6 months of age (94). The recommended surgical technique involves exploration of the brachial plexus and reconstruction of avulsion and nonconducting rupture injuries. If the proximal trunk or nerve roots are intact, sural nerve grafting across the neuroma is preferred. In the presence of an avulsion, intercostal and spinal accessory nerve transfers or distal neurontizations may be performed (95). The surgery will not restore normal function, but there is improvement when compared to natural history outcome alone (82, 83).

Shoulder Surgery. Children with chronic upper trunk plexopathy may develop external rotation and abduction weakness and internal rotation contractures about the shoulder. This muscle imbalance will progressively alter glenohumeral joint morphology (80, 80) (Table 22-2). Function, especially with the arm in above-horizontal activities, will be impaired (79, 81). These children clearly benefit from surgical intervention (81, 86). In the situation of glenohumeral dislocation in an infant, open reduction and capsulorrhaphy or arthroscopic release and reduction are indicated (96) (Fig. 22-12). Such children have limited external rotation that affects function.

In young children with nearly normal glenohumeral joints (normal or mild increase in glenoid retroversion, grades I and II) or slight posterior subluxation (mild, grade III), anterior musculotendinous lengthening of the pectoralis major and/or subscapularis muscles and posterior latissimus dorsi and teres major transfer to the rotator cuff (92, 97) will improve function (82). In addition, dynamic rebalancing of the muscle forces about the shoulder at a young age has the potential advantages of restoring more normal anatomy and prevent ing progressive glenohumeral joint deformity (98). However, glenohumeral joint remodeling appears to have limited utility with extra-articular musculotendinous rebalancing procedures alone. The benefit of arthroscopic release and reduction, as opposed to open reduction and stabilization, is still unclear in terms of long-term functional outcomes, but both techniques have been shown to be effective in reducing the humeral head and inducing early glenohumeral remodeling, as verified by postoperative MRI (99–102). In the older child with more established and progressive deformity of the glenohumeral joint (more severe posterior glenoid flattening, advanced grade III), development of a false glenoid (grade IV) (Fig. 22-13), or humeral head dislocation and deformity (grade V), the deterioration of the joint is usually too advanced to tolerate a soft-tissue procedure. In these situations, humeral derotation osteotomy will improve shoulder function, but will not affect glenohumeral joint morphology (92, 103).

On rare occasions, there are patients who need both osteotomy and tendon transfer. These patients are in the middle range of deformity (grade III). To date, it has been difficult to identify this small subset of patients preoperatively. Therefore, the transfer alone is performed initially, and only if the result is suboptimal more than 1 year later is the secondary-stage osteotomy performed. The role of glenoid osteotomy, its risks, and its benefits are still being defined as it relates to grade III and mild grade IV patients.

Elbow and Forearm Reconstruction. Elbow flexion and forearm supination deformities can occur with a permanent Klumpke (C8-T1) or a mixed brachial plexus lesion. Contractures, bony deformity, and joint instability are the result of muscle imbalance in a growing child. In the rare case of a patient with residual C8- T1 neuropathy with recovery of C5-C6 function, the elbow and forearm deformities are secondary to an intact biceps muscle in the presence of weak or absent triceps, pronator teres, and pronator quadratus muscles. Progressively, the biceps creates an elbow flexion and supination deformity from unopposed muscular activity. Soft-tissue contractures develop, followed by rotation deformities of the radius and ulna (104). Radial head dislocation may occur (105). The wrist and hand are often in extreme dorsiflexion because of unopposed wrist dorsiflexors. In the position of forearm supination, gravity further exacerbates the dorsiflexion deformity. The patient is left without use of the hand, and performs bimanual activities using the volar and ulnar forearm as an assist. Often, shoulder abduction and internal rotation are required in order to improve assistive function. Activities that require simultaneous elbow flexion and forearm pronation, such as dressing, eating, and writing (106), are significantly limited. In addition, the forearm and hand posture is a major cosmetic concern to both the patient and the family (107).

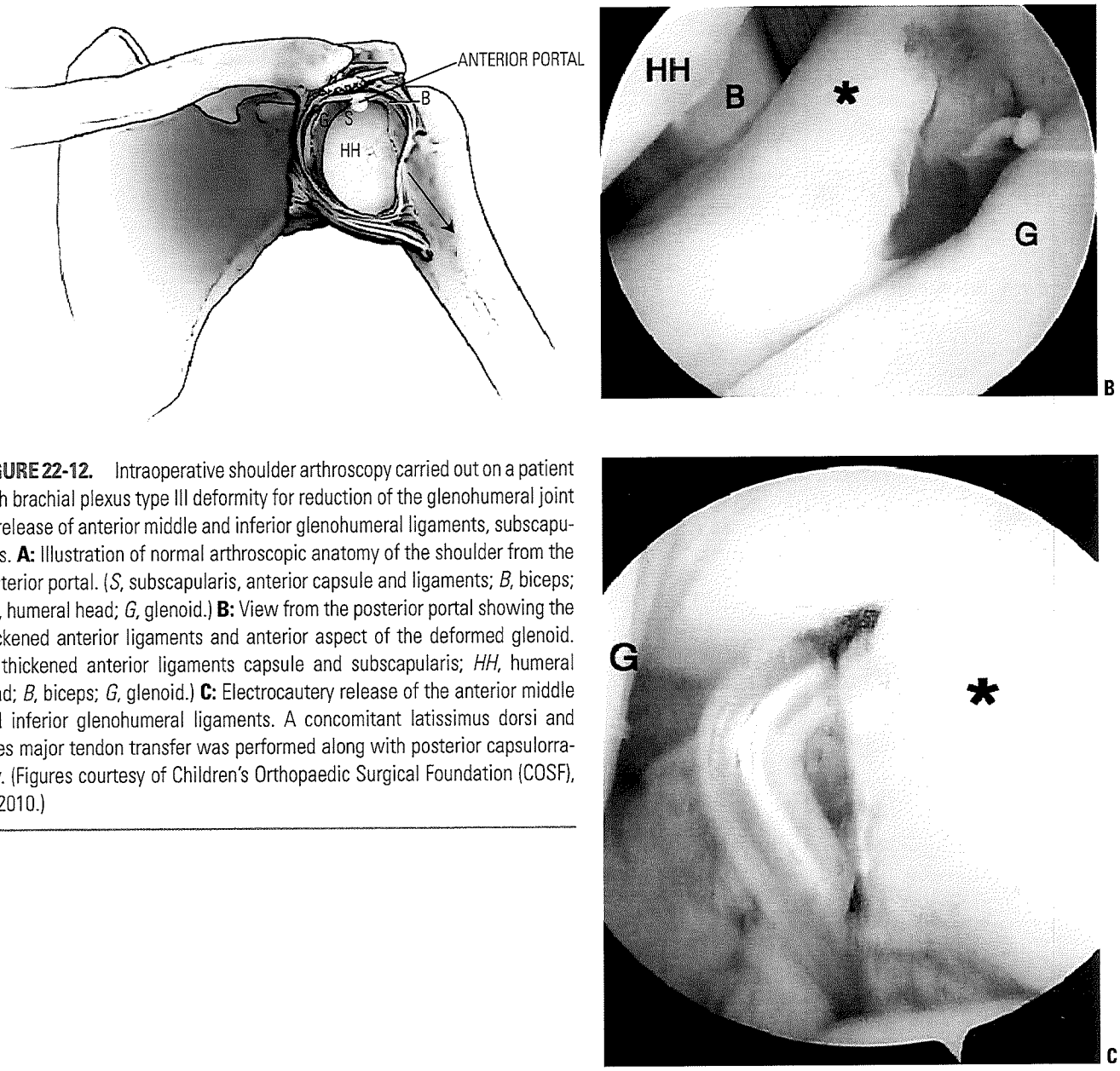

FIGURE 22-12. Intraoperative shoulder arthroscopy carried out on a patient with brachial plexus type III deformity for reduction of the glenohumeral joint by release of anterior middle and inferior glenohumeral ligaments, subscapularis. **A:** Illustration of normal arthroscopic anatomy of the shoulder from the posterior portal. (*S*, subscapularis, anterior capsule and ligaments; *B*, biceps; *HH*, humeral head; *G*, glenoid.) **B:** View from the posterior portal showing the thickened anterior ligaments and anterior aspect of the deformed glenoid. (*, thickened anterior ligaments capsule and subscapularis; *HH*, humeral head; *B*, biceps; *G*, glenoid.) **C:** Electrocautery release of the anterior middle and inferior glenohumeral ligaments. A concomitant latissimus dorsi and teres major tendon transfer was performed along with posterior capsulorrhaphy. (Figures courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

The biceps tendon can be treated by Z-lengthening and rerouting around the radius to convert it from a supinator to a pronator. This will improve elbow extension and forearm pronation. Surgically, the biceps tendon is identified as it inserts into the radial tuberosity. By dissecting lateral to the tendon, the brachial artery and the median nerve are protected. A long Z-plasty of the tendon is performed from the musculotendinous junction to the insertion site. The distal attachment of the tendon is rerouted posteriorly around the radial neck, from medial to lateral. Care must be taken to stay adjacent to the radial neck so as to avoid injury or compression of the radial nerve. The distal tendon is reattached to its proximal counterpart in a lengthened position. This converts the biceps into a forearm pronator (30, 106, 108).

In the presence of a fixed supination contracture, if the rerouting procedure alone is carried out, it will fail because of recurrence of the deformity. Zancolli (106) suggested performing simultaneous interosseous membrane release. However, active pronation was maintained in only 50% of patients who underwent this procedure. Bony correction of the forearm deformity can be performed more predictably. Manske et al. (30) proposed staged procedures of tendon rerouting and forearm osteoclasis. Waters and Simmons (107) described simultaneous tendon rerouting and osteotomy, using internal fixation to avoid multiple operations and loss of alignment. In both

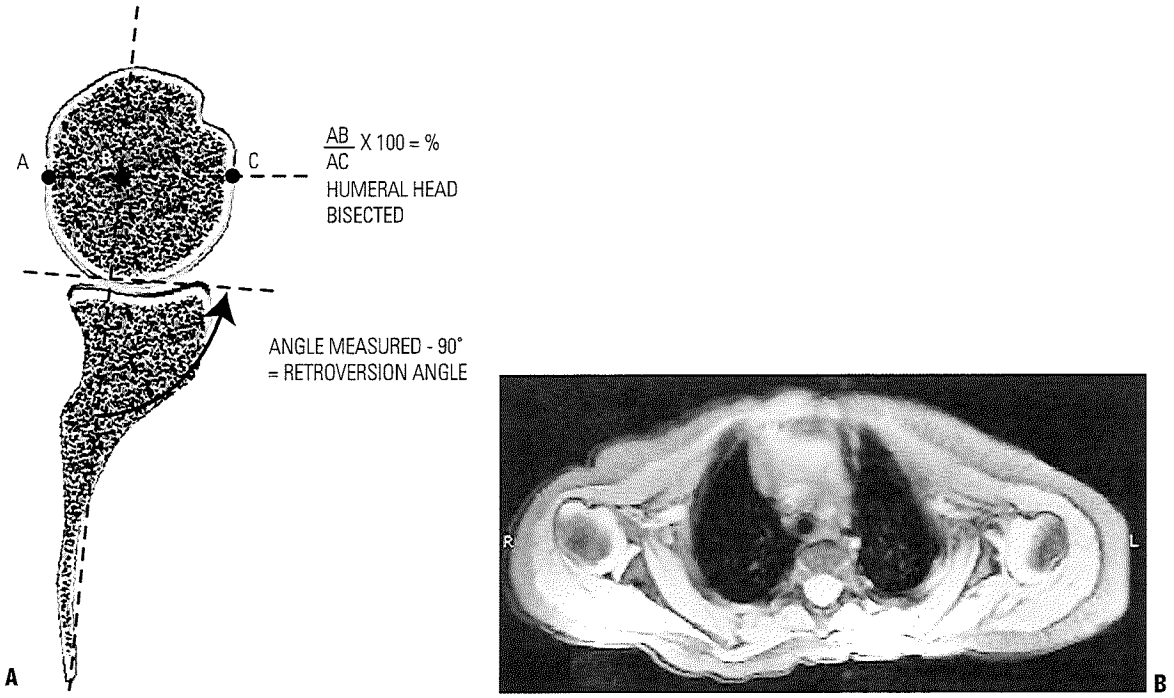

FIGURE 22-13. **A:** Schematic showing the method of measuring the glenoscapular angle (glenoid vision) and the percentage of posterior subluxation of the humeral head. To measure the glenoscapular angle, a line is drawn parallel to the scapula and a second line is drawn tangential to the joint. The second line connects the anterior and posterior margins of the glenoid. The cartilaginous margins are used on magnetic resonance images. The osseous margins are used on computed tomographic scans. The intersecting line connects the center point of the first line (approximately the middle of the glenoid fossa) and the medial aspect of the scapula. The angle in the posterior medial quadrant is measured with a goniometer (*arrow*), and 90 degrees is then subtracted from this measurement to determine the glenoid version. The percentage of posterior subluxation is measured by defining the percentage of the humeral head that is anterior to the same scapular line. The greatest circumference of the head is measured as the distance from the scapular line to the anterior portion of the head. This ratio [the distance to the anterior aspect of the humeral head (*AB*) divided by the circumference of the humeral head (*AC*), multiplied by 100] is the percentage of subluxation. **B:** Magnetic resonance imaging of a type IV deformity with posterior humeral head subluxation and the development of a false glenoid. The glenoid is markedly retroverted. The contralateral glenohumeral joint is normal for age. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

techniques, the forearm is positioned in approximately 20 to 30 degrees of pronation. Others have described corrective osteotomies of the radius and ulna with plate fixation with good reported correction (109).

These patients clearly have significant improvement in their functional capabilities. Bimanual tasks, such as lifting, carrying, and transferring, are easier. The affected extremity becomes a better assistive extremity to the unaffected side. The wrist and hand now have greater assisted palmar flexion and resolution of their dorsiflexion deformity. In addition, the patients are usually pleased with the aesthetic results.

Arthrogryposis. Arthrogryposis multiplex congenita is a syndrome of unknown cause that presents at birth characterized by congenital joint contractures and muscle weakness (110). The incidence is approximately 1 in 3000 live births (111). The clinical syndrome is variable and includes classic arthrogryposis (amyoplasia), distal arthrogryposis, and syndromic involvement (112). The classification of arthrogryposis makes the distinction between myopathic and neurogenic types; however, muscle biopsies and electromyography have not been shown to be helpful in determining the mode of therapy for these children (113). Intelligence is usually average or above average. Sensibility is normal. Upper extremity involvement is frequent, with 72% of the 114 patients in the Gibson and Urs study being affected (114). The wrist was most commonly involved, followed by the hand, elbow, and shoulder. In the classic presentation, the elbow is usually contracted in extension at birth. The shoulder is internally rotated with the forearm pronated. Often, there is wrist palmar flexion and ulnar deviation, and the fingers have flexion deformities. The thumb is usually adducted and flexed in the palm (114–116). These children often have incomplete syndactylies of all web spaces. The first web-space contracture is usually the most functionally significant. There is usually marked intrinsic muscle weakness. There may be camptodactyly or symphalangism of the

PIP joints. All of this will limit hand function in these children (117).

Involvement is generally bilateral. The absence of both passive and active elbow flexion is a significant functional liability in these children. The goal of orthopaedic management of the arthrogrypotic elbow is to improve self-feeding and independent hygiene skills by achieving both passive and active elbow flexion. The goal of treatment of the hand and wrist is to improve pinch, grasp, and release functions.

Treatment

Nonoperative Care. Initial care is with physical therapy to improve passive range of motion. Repetitive, gentle, passive manipulation of the involved joints may progressively lessen the contracture. This process is tedious and requires meticulous, gentle care by both the therapist and the family. Corrective splints and serial casts have been used with varying success (118). Caution is necessary because of the risk of fractures or dislocations that can occur as a complication of aggressive treatment of resistant contractures. At the elbow, the goal of therapy is to achieve at least 90 degrees of passive elbow flexion by 2 years of age. Most patients can achieve the desired passive elbow flexion through therapy (117). However, if this is not obtained, operative posterior elbow release and triceps V-Y lengthening are recommended (119–124). When passive elbow flexion is obtained, therapy should then emphasize the use of adaptive trunk sway, head tilt, and table-assisted passive elbow flexion to improve feeding and hygiene tasks. Finally, if subsequent active elbow flexion does not develop, active elbow flexion tendon transfer can be considered beyond approximately 5 years of age (119). Most of these children will have deficient biceps and brachialis musculature, and will fail to develop active elbow flexion.

Initial treatment of the wrist and hand should involve passive range of motion and nighttime splinting. The goal of therapy in the hand is to improve motion of the joints and digital strength. Fortunately, in many children, the condition improves with growth and therapy during the first several years of life. Serial casting or splinting is often used for the wrist deformity, but this procedure is associated with a high rate of recurrence. If passive motion cannot be improved, surgical releases and tendon transfers may be necessary (125).

Operative Care

Posterior Capsulotomy and Triceps Lengthening. As mentioned in the preceding text, children who fail to achieve a functional arc of flexion at the elbow with manipulative therapy, splints, and casts are candidates for operative elbow posterior capsulotomy and triceps lengthening. Surgery can be performed when the child is 2 years of age. If it is delayed well beyond this age, progressive bony deformity can occur. The goal of operative intervention is to achieve at least 90 degrees of passive elbow flexion. Initially, the dominant extremity should have surgery. The presence of passive elbow flexion will improve independence in feeding, hygiene, play, and school activities (107, 119).

Surgical exposure is by a standard posterior approach to the elbow. The triceps tendon is incised in an inverted V. The angle of the V should be acute enough to allow for appropriate triceps lengthening. The ulnar nerve is protected during the medial incision of the tendon. The distal flap of the triceps is elevated from the elbow capsule, but often the triceps and the capsule are confluent distally. The triceps lengthening alone usually does not improve passive elbow flexion. A transverse incision in the elbow capsule is then made. Full passive elbow flexion is gained. The triceps tendon is lengthened in a V-Y manner at 90 degrees of flexion.

Tendon Transfers for Elbow Flexion. Children with arthrogryposis who have passive elbow flexion of >90 degrees and no active elbow flexion are candidates for tendon transfer. The transfers to be considered include (a) triceps (119, 120), (b) pectoralis major using the sternocostal origin, (c) pectoralis major using the entire musculature on a neurovascular pedicle (121), (d) latissimus dorsi (117, 122), (e) lateral and proximal reinsertion of the flexor–pronator origin, (f) sternocleidomastoid with a free tendon graft, and (g) pectoralis minor with a free tendon graft. Each of these transfers has been described in limited series in the arthrogrypotic elbow. Until recently, no objective criteria had been proposed for comparing the results of these various transfers (107, 119). The muscle considered for transfer must be expendable and of sufficient strength to function actively against gravity after transfer. Each transfer has its inherent negative attributes: triceps transfer may weaken assistive ambulation in patients with lower extremity involvement, and may result in a flexion contracture; pectoralis major transfer may create asymmetric breast appearance in women; Steindler flexorplasty may worsen wrist and finger flexion contractures. Information gained to date indicates that the triceps transfer is most effective in improving strength, active range of motion, and function (107, 119).

The triceps muscle is strong in most children with arthrogryposis. With transfer, it is usually successful in providing active elbow flexion in a functional arc. However, the triceps is important for crutch ambulation, rising from a sitting position, and wheelchair transfers in patients with lower extremity involvement and should be used cautiously for tendon transfer in these children. This operation involves the transfer of the antagonist to elbow flexion and leaves the patient without an active elbow extensor postoperatively. This can lead to progressive elbow flexion deformity with growth (117, 122). By transferring only the long head of the triceps (separately innervated), active flexion can be achieved while preserving elbow extension. This lessens the risks of the above complications noted using complete triceps transfer.

There are two options for transfer of the pectoralis major muscle for elbow flexion. The first choice is transfer of the sternocostal origin, as originally described by Clark (126). This transfer can be problematic because the partial transfer may be too weak to provide antigravity strength for feeding and facial

hygiene. In addition, the pectoralis major muscle crosses the shoulder and may lose strength in trying to move both the shoulder and the elbow.

The second choice is transfer of the entire pectoralis major muscle on its neurovascular pedicles, as advocated by Carroll and Kleinman (121) and Doyle et al. (127). This operation has had favorable results in limited series of arthrogrypotic elbows. It involves transferring the insertion of the pectoralis major to the acromion. The origins of the clavicular and sternocostal heads, with attached anterior rectus abdominis fascia, are inserted distally into the proximal radius. The medial and lateral pectoral nerves and the lateral thoracic vessels are preserved. This transfer has the mechanical advantage of a linear contraction for elbow flexion and does not involve the loss of any strength in stabilizing or moving the shoulder. The proximal advancement of the insertion to the acromion or clavicle improves the lever arm and the mechanical advantage of the transfer. In addition, the pectoralis minor can be transferred with the pectoralis major for further strength of transfer. However, it may create an asymmetric appearance of the breasts in women, and this has been raised as an argument against transfer (107).

In patients with significant lower extremity involvement, with weak triceps or pectorals, or with failed pectoralis major or triceps transfers, a bipolar latissimus dorsi transfer, as described by Zancolli and Mitre (122), may be the optimal choice. Preoperative assessment of the strength of the latissimus is important before transfer, but at times this is difficult to assess. An experienced pediatric physical therapist with extensive muscle evaluation experience may be helpful. Biopsy of the muscle has been tried, but is not predictive of outcome with transfer.

In summary, physical therapy should be initiated in infancy to obtain and maintain passive range of motion of the elbow. This will frequently result in passive elbow flexion of >90 degrees. If by 24 months of age nearly full passive elbow flexion has not been achieved, elbow capsulotomy and triceps lengthening are recommended. After 4 years of age, tendon transfer for elbow flexion in the dominant arm can be considered with consideration given to intelligence, ipsilateral and contralateral upper limb function, lower extremity involvement, and available motors for transfer. All transfers have had same success, but partial or complete triceps-to-biceps transfer gives the most predictably good results (107, 119).

Wrist and Hand Reconstruction. The wrist palmar flexion contracture is addressed with FCU lengthening or transfer to the wrist extensors. Unfortunately, in many of these children, the transfer is more of a tenodesis procedure than a dynamic transfer. In addition, there is often bony deformity, even in the very young. Smith (28) had recommended a proximal row carpectomy to correct the wrist flexion contracture. However, there are frequently carpal coalitions present that preclude that procedure. A dorsal, carpal, closing-wedge osteotomy can correct the deformity in the presence of a carpal coalition. This is an excellent procedure to correct the bone and wrist joint deformity that does not respond to therapy (116). Simultaneous FCU transfer can be performed to rebalance the wrist. An alternative to carpal osteotomy is a dorsal closing osteotomy of the radius and ulna dorsal osteotomies (112). However, these create an "S" deformity to the distal forearm, and physeal remodeling with growth tends to lead to recurrent wrist flexion deformity.

The thumb-in-palm contracture is addressed with a Z-plasty syndactyly release. Alternatively, rotation flaps may be utilized to increase the first web space and provide new tissue to the volar aspect of the thumb (Ezaki and Oishi) Care must be taken not to overrelease the adductor, because it may be providing the bulk of the pinch strength. Dynamic transfers for thumb abduction and extension are predominantly tenodesis procedures because of the limited strength of the donor muscles. Many of these children will have permanent limited motion and strength in their hands. Fortunately, their high level of intelligence allows them to be very adaptive in their functioning.

SHOULDER REGION

Sprengel Deformity. Children with Sprengel deformity (128) often present with a decreased neck line, limited motion about the shoulder, or both. This is secondary to the embryonic failure of one, or sometimes both, scapulae to descend in utero. The abnormal elevation is in conjunction with hyperplasia and abnormal alignment of the scapula. Most often, the scapula is small and shaped like an equilateral triangle rather than having a long medial border. The scapula in Sprengel deformity usually has abnormal anterior bending of the superior pole into the convexity of the upper thoracic region. There is often limited forward flexion and abduction of the shoulder because of lack of normal scapulothoracic motion and malpositioning of the glenoid. In up to 50% of cases, there may be an associated omovertebral bar, which consists of a fibrous, cartilaginous, or bony connection between the superior medial angle of the scapula and the cervical spine (129). Frequently, there is associated abnormal regional anatomy including scoliosis, spina bifida, clavicular abnormalities, rib anomalies, and Klippel-Feil syndrome, among others (130, 131). Systemic abnormalities include renal and pulmonary disorders. In 10% to 30% of the cases, the condition is bilateral.

A classification by Cavendish (130) grades the severity of deformity in a rudimentary way: grade I is mild, with level glenohumeral joints and no deformity visible when the patient is dressed; grade 2 has level glenohumeral joints, with a lump in the neck region with the patient dressed; grade 3 is a moderate deformity with 2 to 5 cm of shoulder elevation; and grade 4 is severe, with elevation of the scapula to the vicinity of the occiput. The more severe the deformity, the more likely there are to be limitations

of motion and function and associated regional anatomic anomalies. Surgery is indicated in children with severe aesthetic and functional limitations. Surgery does not correct the scapular hypoplasia but is indicated for improving shoulder motion by restoring more normal positioning of the scapula and the glenoid. This often consists of excising any omovertebral connections and surgically derotating and caudally relocating the scapula. Most of the procedures that are described include extraperiosteal resection of the superior pole of the scapula (130, 132). Subperiosteal resection is associated with a high rate of recurrence (133, 134). In addition to functional indications for surgery, most patients and families welcome the improvement in the appearance of the neck line.

In the mild deformities, extraperiosteal excision of the superior pole of the scapula and any omovertebral connections alone may be satisfactory treatment. In the moderate and severe deformities, the scapula is also derotated and moved more distally in order to bring the glenoid into a more vertical orientation. The purpose of surgery is to improve the neck contour along with shoulder motion and function. Indications for functional improvement have been cited for preoperative abduction <110 to 120 degrees (129, 135). Surgery is recommended most often in patients between 3 and 8 years of age (136–138). Surgery after 8 years of age is associated with the highest risk of nerve impairment; clavicular osteotomy or morsellization is recommended in the older child in order to lessen the risk of brachial plexus impingement with scapular descent.

Surgical procedures for scapular descent have included the Woodward procedure (139), the Green procedure (140, 141), and a vertical scapular osteotomy (142). The Green procedure involves extraperiosteal detachment of the scapular insertion of the paraspinal muscles, and reattachment after the scapula has been moved distally with traction cables. Wilkinson and Campbell described a vertical scapular osteotomy in conjunction with a clavicular osteotomy in the older child for improving anterior release and scapular relocation and for lessening the risk of neurologic injury. The Woodward procedure moves the scapula distally by detachment and reattachment of the parascapular muscles at their origins on the spinal process. The modified Woodward procedure includes resection of the superior pole of the scapula in conjunction with surgical scapular descent and realignment. The results have been reported to have improved abduction in the range of 40 to 50 degrees (129, 143) and achieved a satisfactory aesthetic result. Hypertrophic scar formation, however, has been cited often as a complication.

Woodward Repair of Sprengel Deformity. Congenital high scapula, commonly known as *Sprengel deformity*, is not a common condition. It can be seen in all degrees of severity. As a result of the school-screening programs, we have seen numerous children with minor degrees of scapular elevation and smaller scapulae on one side. Minor degrees of high-riding scapulae need no treatment and are usually not associated with other developmental abnormalities around the shoulder. At the other end of the spectrum is the child diagnosed at birth or shortly thereafter. The physician seeing the infant or the small child should realize that the deformity usually becomes worse with growth. This can be difficult to judge when the child is between 4 and 8 years of age. However, 4 to 8 years is the ideal time for optimal correction. The condition develops during the 9th to 12th weeks of gestation; therefore, other organs, as well as those structures around the shoulder girdle, may be affected. An understanding of the pathologic anatomy is important for the correction of the deformity and for the avoidance of complications. The scapula is shorter in its vertical height than the opposite normal scapula and is more concave anteriorly to fit the convex shape of the superior aspect of the thoracic cage. In addition, the supraspinous portion of the scapula is usually tilted forward and its superior medial portion may be larger. The clavicle may also be higher and shorter, lacking its usual anterior convexity (136). In about one-third of the cases, an omovertebral bone connects the superior medial angle of the scapula to the posterior elements of the fourth and fifth cervical vertebrae. This may actually be bone, cartilage, or fibrous tissue. Finally, the muscles of the shoulder girdle are usually affected, and hypoplasia of the trapezius and rhomboids is the most common problem.

Two operations for the correction of Sprengel deformity have stood the test of time and are the most commonly used. The Green procedure (140, 144–146) detaches the muscles from the scapula, whereas the Woodward procedure (139, 147) detaches the origins of the trapezius and rhomboids from the spinous processes (Figs. 22-14 to 22-19). We have had experience with both procedures and find the Woodward procedure to be easier (but not easy) and to produce the same results with a shorter period of hospitalization and lower morbidity. Surgery is expected to improve the cosmetic appearance and the function of the shoulder (129, 148).

One of the most important complications is radial nerve palsy resulting from compression of the brachial plexus between the clavicle and the first rib when the scapula is pulled down. This is more common in children aged 7 years or older and is relatively uncommon in children aged 3 to 4 years. Some authorities have advocated division or morcellation of the clavicle to prevent this complication (136, 149). This is an effective measure, but it is important to determine which patients require it because the incidence of radial nerve palsy is low (139, 140, 143, 150), especially in young children. This additional procedure may be reserved for affected children older than 8 years and for those younger children who have an unusually severe deformity. When nerve palsy is noted after the Woodward procedure, division of the clavicle can be done.

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Woodward Repair of Sprengel Deformity (Figs. 22-14 to 22-19)

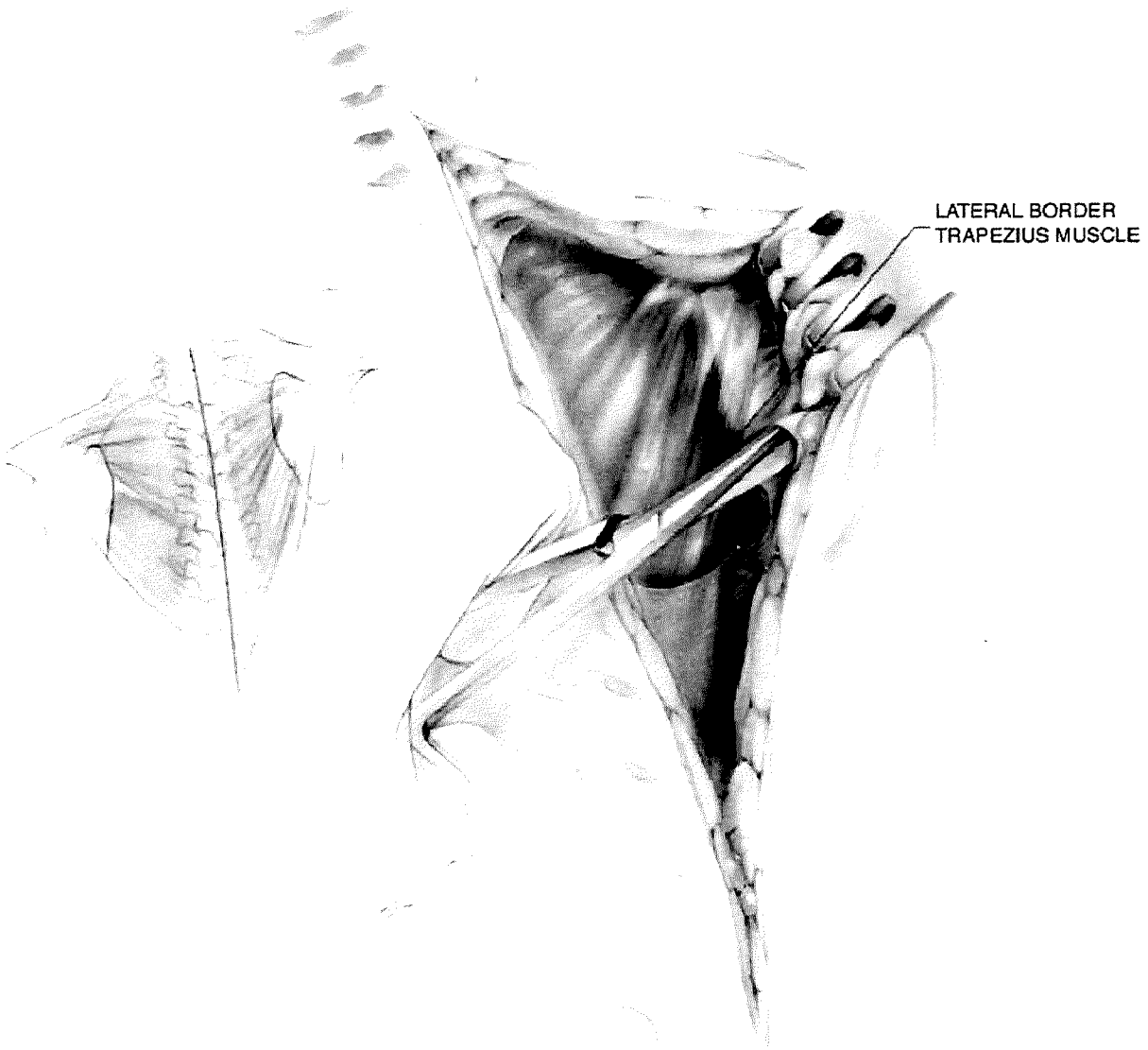

FIGURE 22-14. Woodward Repair of Sprengel Deformity. The patient is positioned prone. The arm and the shoulder on the affected side should be draped free. It may be helpful if the entire posterior thorax is in the sterile field so that the level of the opposite scapula can be observed. It is also helpful if the head is positioned as if looking straight ahead. The incision should be in the midline and should extend from the level of the upper cervical spine (C3, C4) to the lower thoracic spine (T9-T10). The incision is deepened through the subcutaneous tissue and is undermined on the affected side. This dissection should be carried far enough laterally to identify the lateral border of the trapezius muscle in the inferior aspect of the wound and the lateral border of the scapula in the midportion and far enough to allow exposure of the medial half of the supraspinous portion of the spine of the scapula in the superior portion.

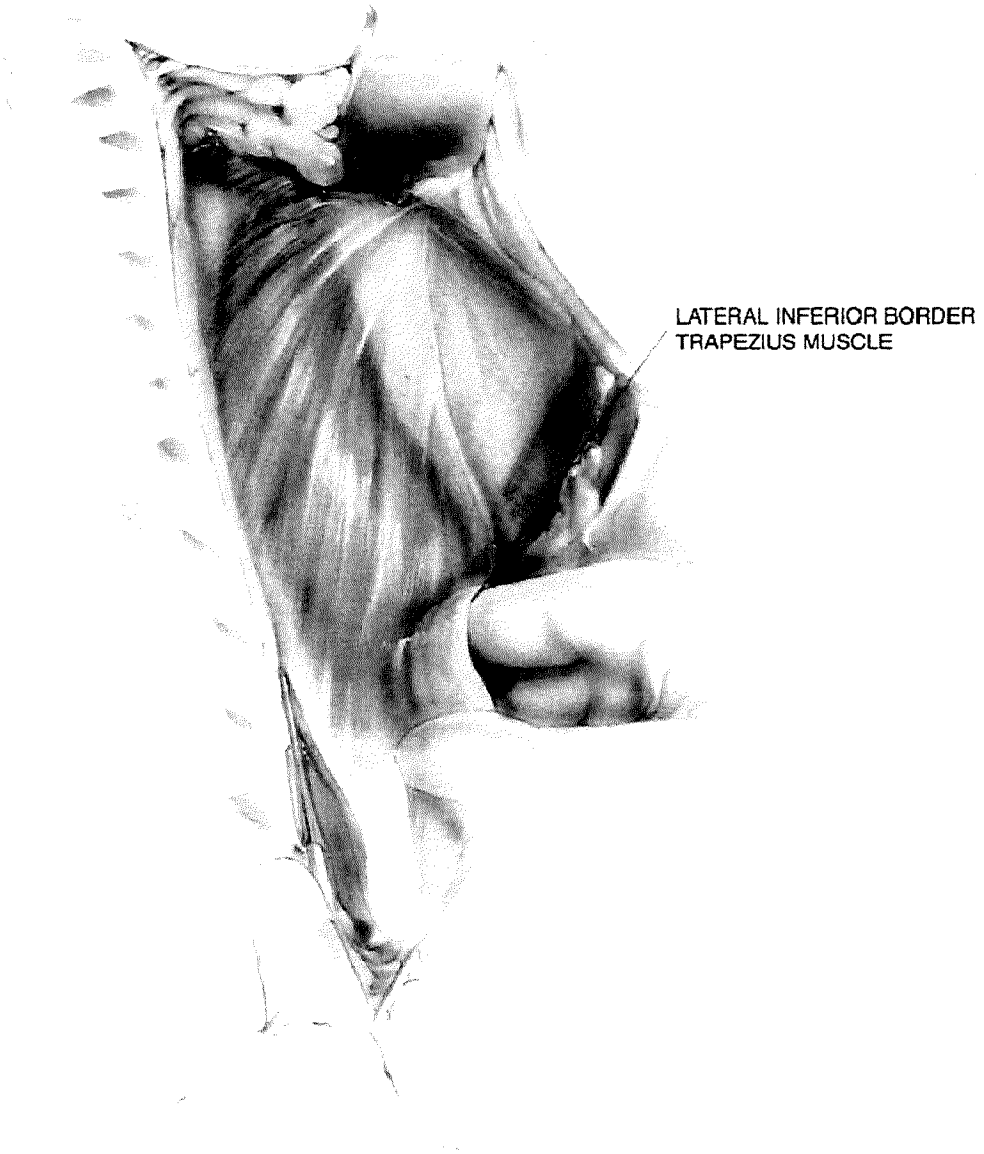

FIGURE 22-15. Although Woodward described detaching the trapezius and rhomboid muscles by directly detaching their origins from the midline, this is virtually impossible because they blend inseparably with all the other muscles with origins at the midline. First, the lateral border of the trapezius muscle in the inferior aspect of the wound must be identified and, by blunt finger dissection, separated from the well-defined thoracolumbar fascia and the latissimus dorsi muscle, which cover the serratus and erector spinae muscles. The maneuver eases identification of the origin of the trapezius, which can be detached without cutting into the deeper muscle layers. This detachment of the trapezius is begun distally and extends to the level of the fourth cervical vertebra, where it can be cut transversely to complete its release. After the trapezius muscle is detached and reflected laterally, the attachments of the rhomboid muscle to the scapula are identified.

Blunt finger dissection can be used to separate them from the underlying deep fascia, aiding in detaching them from their origins like the trapezius muscle. Although this dissection is straightforward in an adult cadaver, it is much more difficult in a 4-year-old child with hypoplastic muscles and abnormal fibrous bands. Nevertheless, this step is the key to the exposure of the surgical area and the further steps in the procedure.

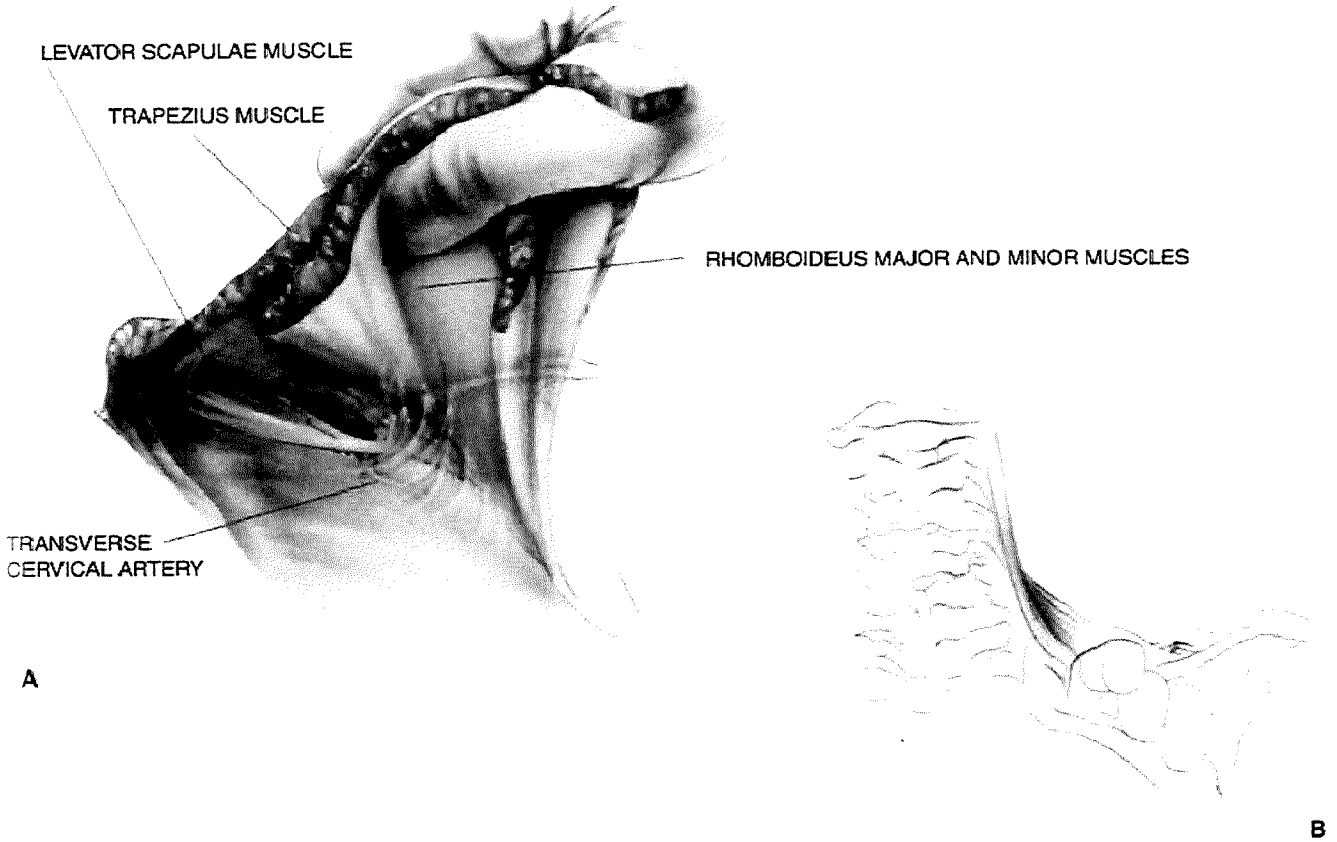

FIGURE 22-16. With the trapezius muscle retracted laterally **(A)**, the levator scapula muscle can be identified as the structure originating from the superior medial corner of the scapula and running toward the cervical spine. Although it lies in the same plane as the rhomboid muscles, it is difficult to identify as a distinct structure. In about one-third of cases, an omovertebral bone (not illustrated here), consisting of actual bone, cartilage, or dense fibrous tissue, originates from this corner of the scapula, usually lying beneath the levator scapulae muscle. If present, it is rarely connected to the cervical spine by bone and can usually be detached by sharp dissection after the bone has been exposed by extraperiosteal dissection. It is essential to release all structures in this region because they will prevent downward displacement of the scapula. Fibrous bands, as well as the levator scapulae muscle, are most easily isolated and divided at the superior medial border of the scapula. Notice the transverse cervical artery running deep to the levator scapulae muscle. Care should be taken to avoid cutting the artery **(B)** by inserting a finger behind the muscle before dividing it.

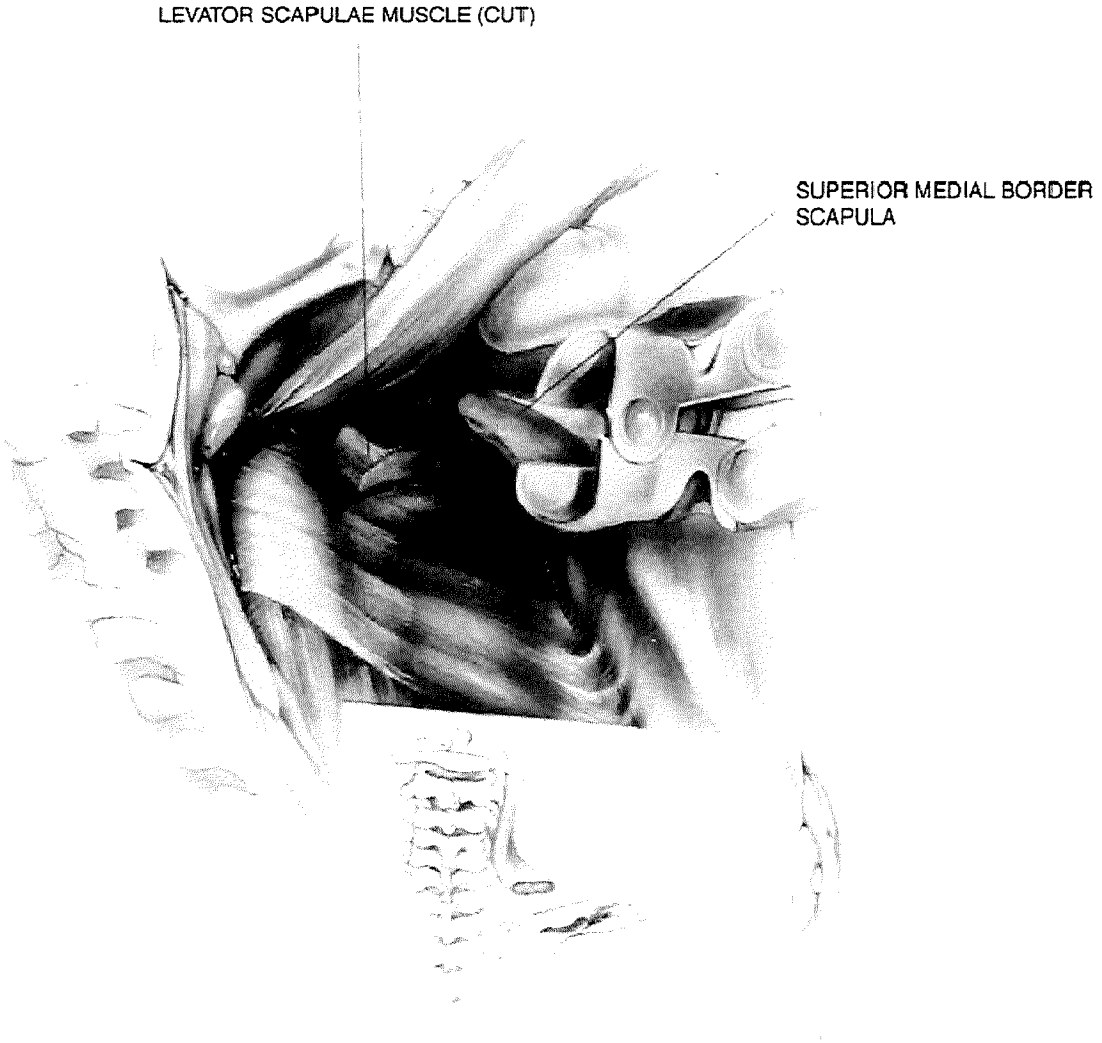

FIGURE 22-17. With the division of the structures originating from the superior medial border of the scapula, it becomes easier to appreciate the contribution that the large anterior-curving, medial supraspinous portion of the scapula makes to the deformity. This portion of the scapula should be exposed extraperiosteally and excised with large bone-cutting forceps. The surgeon should proceed no farther laterally than the scapular notch to avoid causing injury to the suprascapular artery or nerve. With this completed, the scapula can be everted. This usually reveals multiple fibrous adhesions between the scapula and the chest wall. This is especially true in cases with associated anomalies of the chest wall (e.g., missing ribs). These adhesions should be divided. The scapula can be pushed downward and observed for any other tight structures. In severe cases, it may be necessary to divide a portion of the serratus muscle insertion into the scapula.

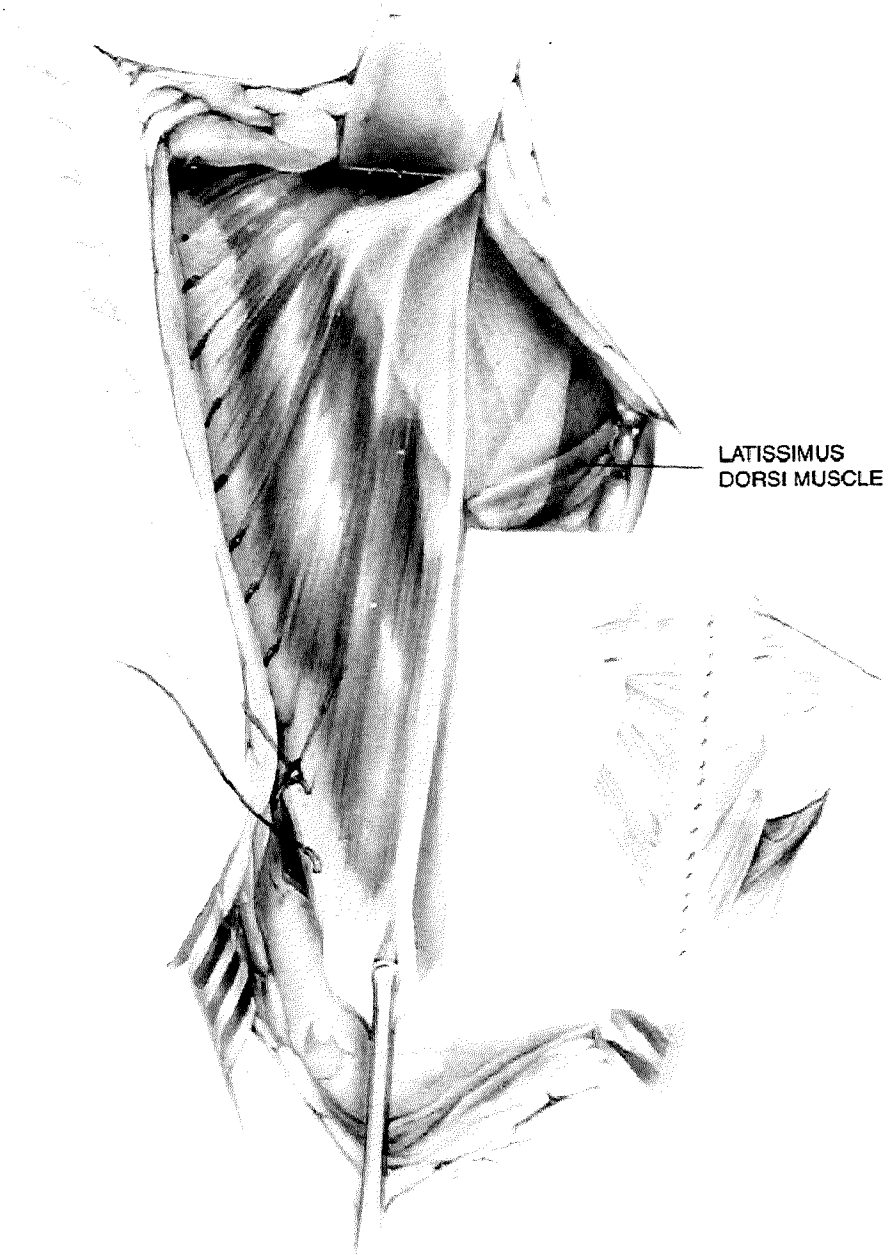

FIGURE 22-18. The latissimus dorsi muscle is elevated to allow the scapula to be displaced beneath it. The rhomboid and trapezius muscles are pulled downward, displacing the scapula to the desired level. The affected scapula is smaller than normal; therefore, displacing it so that its inferior border is level with the inferior border of the opposite normal scapula results in overdisplacement. Rather, it should be displaced so that the spines of the two scapulae lie on the same level. The suprascapularis and subscapularis muscles can be repaired by suturing them together over the resected area of the superior medial border of the scapula. If the serratus muscles were detached, they can be resutured to the scapula in a more cephalad location. The rhomboid and trapezius muscles are reattached to their midline origin in a new, more caudal location. Because the most distal origin of the trapezius muscle (extending to T12) was left intact, there is a redundant segment of muscle and fascia distally, which can be excised. If desired, the tip of the scapula can be sutured to an underlying rib by an absorbable suture as a temporary means of fixation. We find this process useful in maintaining proper rotation of the scapula. Finally, the latissimus dorsi muscle is reattached to the tip of the scapula and the wound is closed.

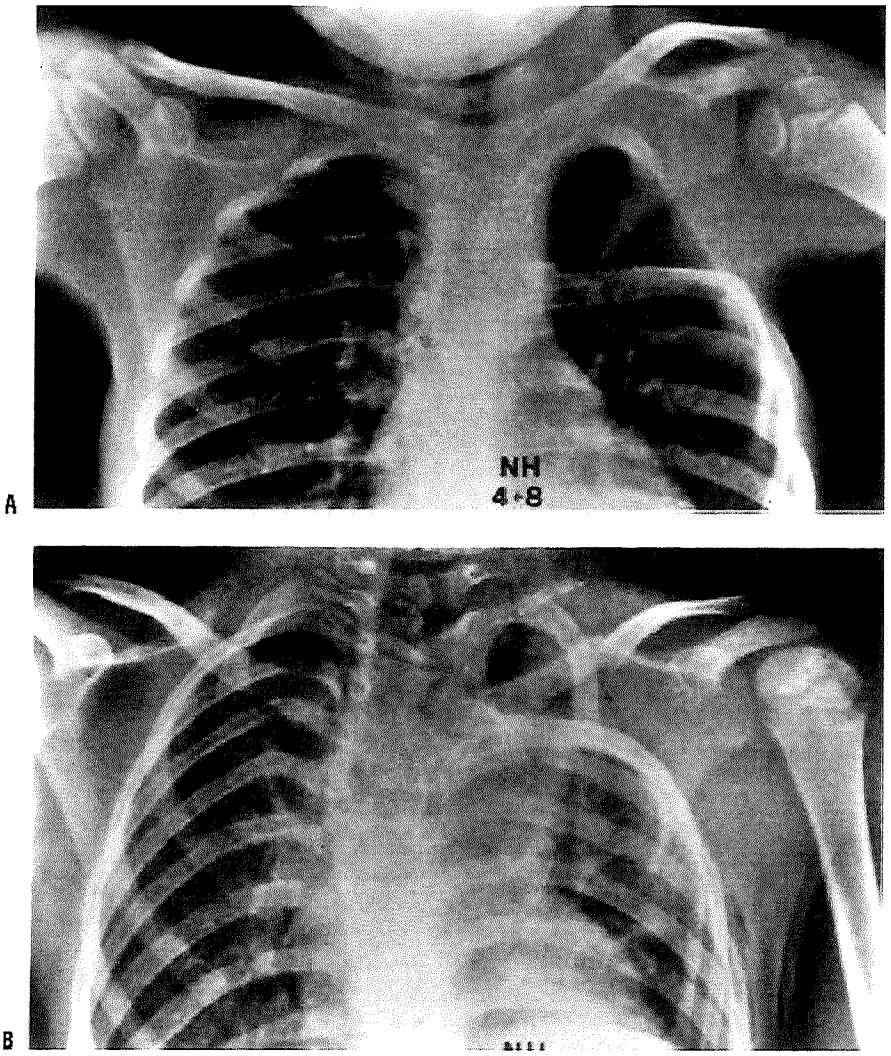

FIGURE 22-19. NH is a 4-year, 8-month-old girl who presented with a high left shoulder and restricted motion, which the parents had observed. They had consulted an orthopaedic surgeon 2 years previously and were told that treatment would not be advisable as she would just be trading a slightly high shoulder for a very large scar. However, the parents were convinced that the deformity was becoming worse. **A:** A preoperative radiograph shows many of the skeletal anomalies seen in association with the congenital elevation of the scapula. The most obvious is that the scapula is high, but it is also smaller than the opposite scapula. There is a defect in the chest wall, with missing and deformed ribs and mild scoliosis with a vertebral anomaly. **B:** Postoperatively, the spine of the left scapula is on the same level as in the normal scapula. Its smaller size is obvious and demonstrates that the affected scapula should not be brought so far inferior that the inferior angle is on the same level as the normal scapula.

Repair of Congenital Pseudarthrosis of the Clavicle. Congenital pseudarthrosis of the clavicle is an unusual condition of unknown etiology. Although the name often causes congenital pseudarthrosis of the clavicle to be confused with congenital pseudarthrosis of the tibia, there is no similarity in the etiology or the natural history of these conditions. The pathology of congenital pseudarthrosis of the clavicle, unlike that of the congenital pseudarthrosis of the tibia, is of two bone ends covered with cartilage and often encapsulated with synovial tissue and fluid. It is clear, however, that resection of the pseudarthrosis, bone graft, and internal fixation are necessary to obtain union (Figs. 22-20 to 22-24) (151–154).

Patients may present at any age with a painless lump in the clavicle. Although usually mild in terms of cosmetic deformity and typically asymptomatic in younger children, the deformity worsens with age. Dissatisfaction with the cosmetic appearance usually develops by adolescence, and discomfort, especially with throwing activities, may develop. For these reasons, surgical repair is usually recommended when the condition is noted at a young age. The ideal time to repair the pseudarthrosis is between 3 and 4 years of age to take advantage of remodeling of bone with

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Repair of Congenital Pseudarthrosis of the Clavicle (Figs. 22-20 to 22-24)

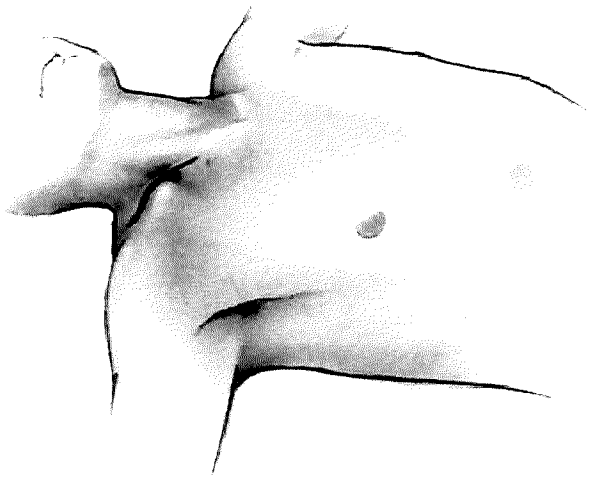

FIGURE 22-20. Repair of Congenital Pseudarthrosis of the Clavicle. The patient is placed supine on the operating table with a sandbag under the upper thoracic spine to allow the head and shoulder to fall posteriorly and to improve exposure of the clavicle. The arm, the shoulder, and the clavicle are draped free. The anterior iliac crest is also prepared for the bone graft, which will be necessary in the repair. The skin incision is placed along the cephalad edge of the clavicle. Its length depends on the child's size, but because the skin in this region is so mobile, it does not have to be excessively long.

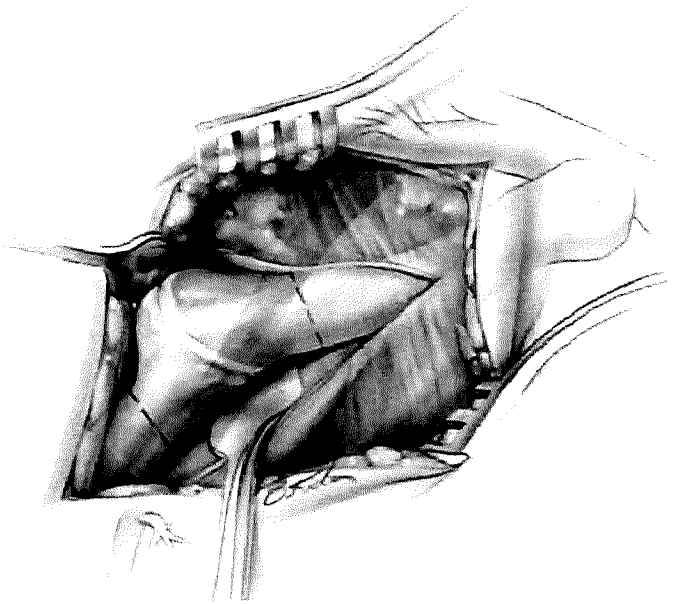

FIGURE 22-21. After dividing the skin, the periosteal surface of the clavicle is exposed below the platysma muscle. The normal clavicle and the maximum possible extent of the bulbous ends of the pseudarthrosis should be exposed subperiosteally. In older children, the bulbous ends may be large, in which case it is impossible to remain subperiosteal. The surgeon must be careful during the dissection because of the proximity of the subclavian artery and vein and the apex of the pleural cavity. The pseudarthrosis is then excised with a rongeur. If the surgeon wishes to preserve the entire pseudarthrosis for histology, a Gigli saw or bone biter can be used, provided that the circumferential dissection is sufficient.

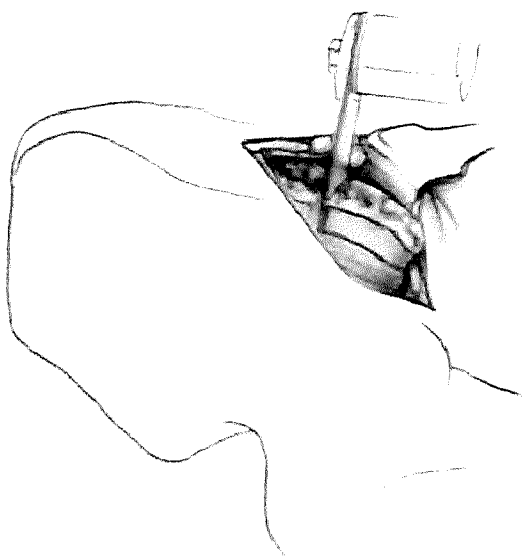

FIGURE 22-22. After resection of the pseudarthrosis, a bone graft may be necessary both to secure osteosynthesis and to maintain the length of the clavicle. A full-thickness (tricortical) piece of bone can be harvested from the anterior iliac crest just behind the anterosuperior iliac spine. The portion of bone just beneath the apophysis is thicker than the thin plates of bone that make up most of the iliac wing and provides a better fit with the two ends of the resected pseudarthrosis. A larger piece of bone than is judged necessary should be removed to allow it to be fashioned to the appropriate size and contour.

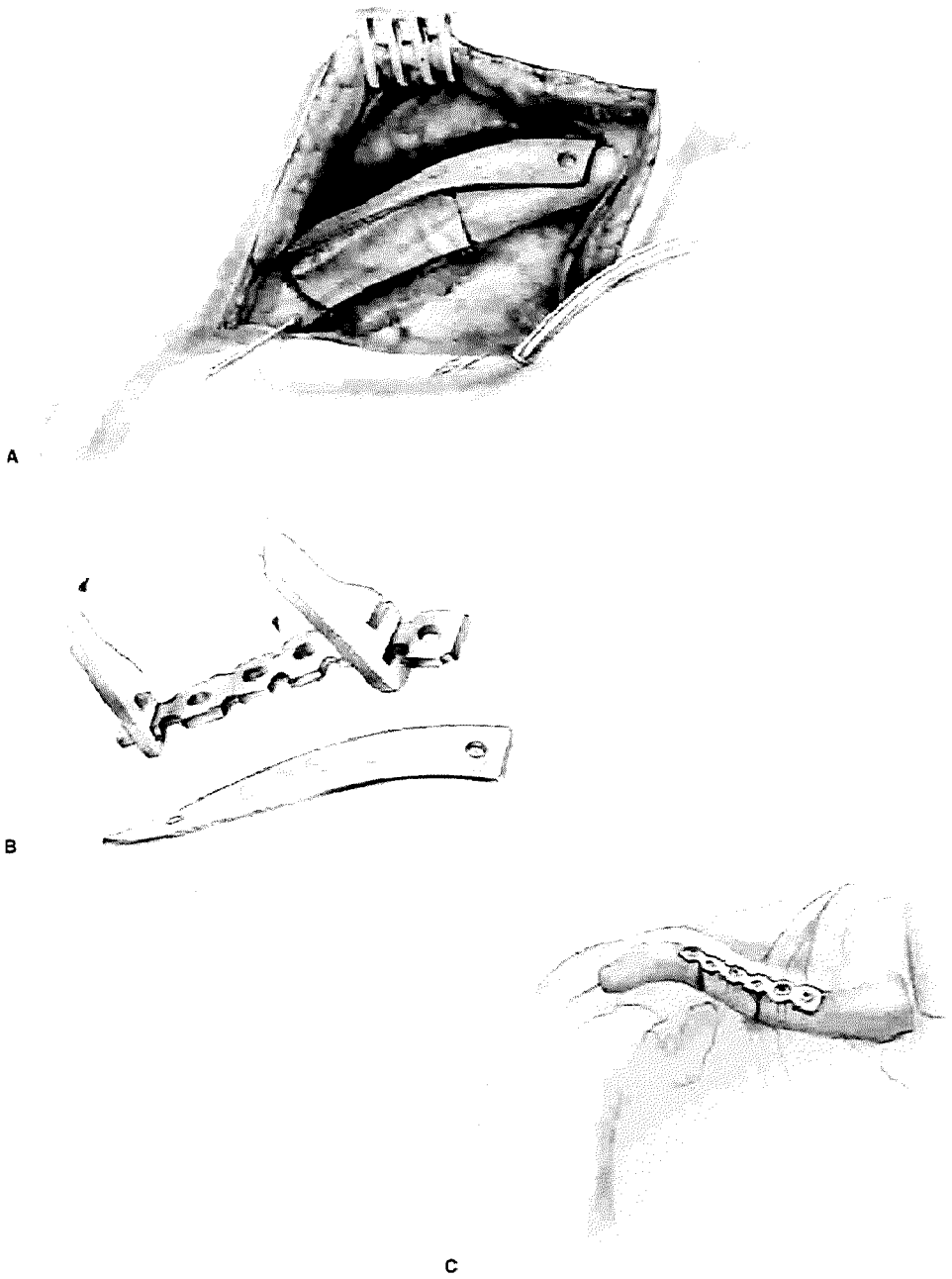

FIGURE 22-23. Various forms of fixation have been used, all more or less with success. Grogan and colleagues (6) recommend only a suture and no graft. If a graft is used, it is possible to use a Kirschner wire, which is drilled out laterally from the osteotomy site and then through the graft and the proximal fragment. If a Kirschner wire is used, it is necessary to leave the wire outside the skin, bending it at 90 degrees to prevent its migration. It is best to remove this wire within 3 to 4 weeks to prevent infection. Because of the clavicle's complex shape, it is impossible to keep a pin or wire of any strength within the clavicle's medullary canal. A thin Kirschner wire that can be passed through the medullary canal of the bone provides little fixation and, unless left outside the skin and bent, risks migration. The use of a pin or wire also risks migration. To avoid these problems, the surgeon can use the small reconstruction plates.

The plates come in two sizes—2.7 mm and 3.5 mm. Their flexibility makes it possible to contour them exactly to the shape of the clavicle and the graft. Because immobilization is required in an active child, regardless of the method of fixation, the plates are sufficiently strong. **A,B:** With the graft held temporarily in place between the two resected ends of the clavicle by a small Kirschner wire, the appropriately sized reconstruction plate is contoured using the template provided. **C:** When the proper shape has been achieved, the plate is attached by screws to both ends of the clavicle and the graft. Each end of the clavicle should be fixed with a minimum of two screws, and at least one screw should hold the graft. The wound is irrigated, a small drain is placed adjacent to the clavicle and brought out through the skin lateral to the incision, and the wound is carefully closed in layers. In young children, a Velpeau dressing is applied and reinforced with a roll of plaster if deemed necessary. In older children, who may be more cooperative with the postoperative immobilization, a commercial sling with a strap that passes around the waist to hold the arm next to the trunk is sufficient.

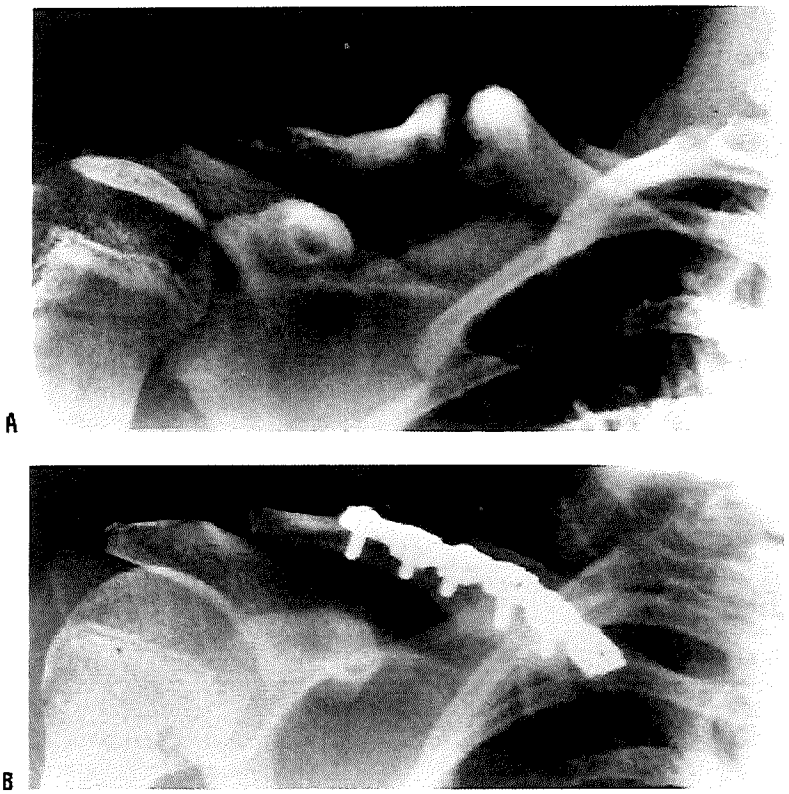

FIGURE 22-24. BC is a 10-year-old boy who noticed a lump on his collar bone and experienced discomfort with throwing activities since an injury 3 years earlier. At the time of the injury he was told that he had fractured his clavicle; at the time of these radiographs he was referred for a persistent nonunion. **A:** Radiographs demonstrate a typical congenital pseudarthrosis of the right clavicle. **B:** Results of excision, grafting, and plating are shown. Healing was prompt, and he returned to all activities without discomfort in 6 months.

growth. Repair, however, can be accomplished with improved cosmesis and with elimination of discomfort at any age.

Several methods of repair have been recommended. Fixation with a Kirschner wire is difficult because of the shape of the clavicle and the potential migration of the wire. However, successful outcome with this technique has been reported (155). This method is less than ideal because the small flexible wire needed to traverse this convoluted shape often breaks. A series of successful cases has been reported using only a suture to secure the bone ends (156). We believe that more rigid fixation is necessary and have used the technique described here with a small reconstruction plate.

ELBOW AND FOREARM REGION

Congenital Dislocations

Congenital Radial Head Dislocations. Congenital dislocation of the radial head is a rare condition that may not be diagnosed until school age. It is usually an isolated condition, but it may be present in association with other congenital malformations and syndromes, including arthrogryposis and Cornelia de Lange, Larsen, and nail-patella syndromes (123, 157–159). It may be associated with radioulnar synostosis (160, 161) or other musculoskeletal anomalies, such as congenital hip dislocation, clubfeet, brachydactyly, clinodactyly, tibial fibular synostosis, congenital below-elbow amputation, and radial or ulnar clubhand. Dislocations associated with Madelung deformity or familial osteochondromatosis (161) may be acquired, and will be considered elsewhere in this chapter.

Congenital radial head dislocation may be bilateral or unilateral (162). It is defined by the direction of subluxation or dislocation. Most congenital dislocations are posterior or posterolateral. It is important to distinguish the congenital dislocation from the posttraumatic dislocation. Because the condition frequently presents late, this distinction can be confusing (162, 157). This is especially true for unilateral anterior dislocations in otherwise healthy children (163–166). Radiographic criteria have been established to distinguish this lesion from a chronic, traumatic dislocation. These include a small, dome-shaped radial head; a hypoplastic capitellum; ulnar bowing with volar convexity in the anterior dislocation and dorsal convexity in the posterior dislocation; and a longitudinal axis of the radius that does not bisect the capitellum. The presence of these characteristics in the absence of any history of trauma to the affected elbow has been seen as evidence of a congenital radial head dislocation (107, 157, 166–171). In addition, bilateral involvement, the presence of other musculoskeletal or systemic malformations, and a positive family history make a congenital cause more likely.

Clinical and Radiographic Features. Children with radial head dislocations often present after infancy. The most common reasons for presentation are (a) limited elbow extension; (b) posterolateral elbow mass/prominence; and (c) pain with activities, especially athletics (107, 172). Often the diagnosis is made after innocuous trauma based on incidental findings noted on radiographs. The elbow extension loss is frequently <30 degrees and rarely of functional significance. This loss of motion is usually not noted early in life. The mass may be

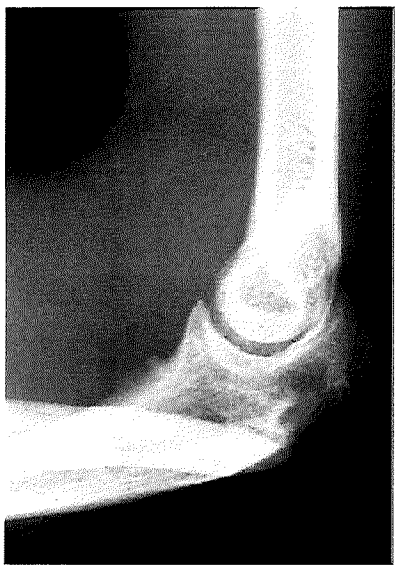

FIGURE 22-25. Lateral radiograph of congenital posterolateral dislocation of the radial head. There is evidence of tapering of the radial head and neck posteriorly, bowing of the ulna posteriorly, and a small dome-shaped radial head. These patients often have limited elbow extension and develop intra-articular pain at the abnormal radiocapitellar articulation in adolescence. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

noted in infancy. Radiocapitellar incongruity can be a cause of pain and disability later in life (162, 172). Unfortunately, many children present late with pain resulting from radiocapitellar articular changes. There is often chronic discomfort with school and sports activities. On occasion, these children may present with an acute loss of motion attributable to a loose osteochondral fragment. Some individuals remain asymptomatic, and the aesthetics of the deformity is their major concern.

On physical examination, the elbow may have cubitus valgus. A flexion contracture of up to 30 degrees often occurs with a posterior subluxation/dislocation. Hyperextension and/or loss of flexion may occur with an anterior dislocation. The radial head is palpable in its dislocated position. A congenital dislocation is not reducible by forceful manipulation, and should not be misinterpreted as a nursemaid's elbow or a Monteggia lesion. There is usually limited forearm rotation, with supination being affected more than pronation. Clicking and crepitus may be present when there is advanced intra-articular pathology (107).

Radiographs reveal the subluxation/dislocation (Fig. 22-25). The longitudinal axis of the radius does not bisect the capitellum, regardless of the angle of the radiograph. The radius and ulna are of different lengths. The ulna is bowed, with volar convexity in an anterior dislocation and dorsal convexity in the more common posterior dislocation. The capitellum is hypoplastic. The radial head will be convex or dome-shaped, with a long, narrow radial neck.

Natural History. The presence of a congenital dislocated radial head is not an indication for operative intervention. Many patients with this disorder have no functional limitation and no pain. Their mild limitation of motion may not restrict them in any significant way. The degree of cubitus valgus is usually mild and does not seem to put them at risk for ulnar neuropathy. Therefore, in most cases, a definitive diagnosis followed by observation is most appropriate. If the patient develops pain, functional or progressive limitation of motion, or restriction of elbow-related activities, then surgery may be considered ((107).

Treatment

Operative Care. Ideally, the care of a congenital dislocated radial head would involve open reduction and restoration of normal anatomy. This has led many surgeons to consider open reduction of a congenital dislocation if the child presents in infancy (123, 160, 166, 173). The logic is that if the radial head can be reduced early in infancy, the deformity of the capitellum and the forearm may not occur or remodel with growth. This may prevent the long-term complications of pain, loss of motion, and osteochondral loose bodies. However, there have been only a small number of published cases of open reduction of congenital radial head dislocations (160, 166, 173, 174). Techniques have included ulnar osteotomy and lengthening, radial shortening and osteotomy, annular ligamentous reconstruction, and the use of limb-lengthening devices to reduce the radial head (170, 173, 175, 176). Sachar and Mih's report of open reduction through an anconeus approach, followed by annular ligament reconstruction, is the most promising series to date. They described seven cases of open reduction of a congenitally dislocated radial head with good success (173). Their operative findings included an abnormality of the annular ligament that was surgically correctable. The indications for this procedure, and the age limit, are still being defined in this relatively rare condition. It is reasonable in specialized centers to consider open reduction of the congenitally dislocated radial head in the infant younger than 1 to 2 years of age, provided the family is well informed of the limited nature of the information regarding this procedure. Hopefully, clinical surgical research in this area will define the indications and techniques for open reduction and annular ligament reconstruction in congenital radial head dislocations.

Most children with congenital radial head dislocation present later than infancy. Therefore, the most common procedures for this problem are excision of loose bodies and excision of the radial head. The indications for excision of a loose osteochondral fragment are the presence of pain, clicking or locking, and loss of motion. Usually, degenerative changes are too advanced for repair of the osteochondral fragment. There is some controversy regarding the indications and the timing for excision of the radial head. In the skeletally immature patient, the concern is the potential development of postoperative complications (see "Complications," below). These concerns have not been supported in the published literature on excision of the congenitally dislocated radial head. Most of these children do not present until adolescence with pain or progressive restriction of motion. In our series, the youngest patient with excision of a symptomatic congenital radial head without complication was 8 years of age (162). However, the presence of an

asymptomatic dislocated radial head alone, without painful, progressive restricted range of motion, is not an indication for radial head excision. Indications for radial head excision must include progressive pain, progressive loss of motion, and progressive restriction of activities (174), regardless of age (107).

Complications. Throughout the 20th century, standard textbooks and journal articles denounced the concept of radial head excision in the skeletally immature individual. Postoperative complications of progressive cubitus valgus and potential associated ulnar neuropathy, proximal migration of the radius with recurrent radiocapitellar impingement, radioulnar synostosis, and reformation of the radial head have been cited (166, 167, 177–179). However, most of these problems occurred after radial head excisions to treat trauma. The admonishment never to excise a radial head in a skeletally immature individual still holds true in the posttraumatic situation. These complications are rare after excision for congenital radial head dislocations (160).

"Reformation of the radial head" by bony overgrowth of the proximal neck is the most common problem with excision of a congenital dislocation (178, 180, 181). If it leads to recurrent radiocapitellar impingement, limitation of motion, and/or pain, then repeat bony excision should be performed. Wrist pain does occur in the long term, though most published reports suggest this wrist pain is mild and nonrestrictive (162). Fortunately, iatrogenic radial nerve injury is rare.

Congenital Humeroulnar Dislocations. Dislocation of the ulnotrochlear joint is exceedingly rare. Mead and Martin described a family with aplasia of the trochlea and humeroulnar dislocations (182). Ulnotrochlear dislocations have also been seen in hyperelasticity syndromes. These situations are rarer than the unusual posttraumatic persistent or recurrent dislocation.

A congenital dislocation will result in limited range of elbow motion that can affect function. The dislocation is usually palpable on examination. There may be axial malalignment, such as cubitus valgus. If severe, the valgus deformity can result in ulnar neuropathy. In recurrent dislocations secondary to hyperelasticity or associated with syndromes such as Rubinstein-Taybi syndrome (183), the elbow instability is palpable and even audible on examination. On occasion, the recurrent instability can lead to osteochondral injury that will cause pain, clicking, or even locking on examination.

Elbow dislocation can also be seen with ulnar dysplasia and ulnar dimelia (184–187). The dysplastic ulnotrochlear joint in ulnar dysplasia can lead to elbow problems that limit motion and function. Ulnar dimelia, or mirror hand, is exceedingly rare. The forearm and elbow in this condition consist of two ulnae and no radius. This means that there are two olecranon processes articulating with the distal humerus. There are usually two poorly defined trochleae and no capitellum present. The olecranon processes may face one another. There is significant limitation of elbow and forearm rotation (107, 188, 189).

If the child presents before ossification of the secondary centers, it may be difficult to define the dislocation anatomically by plain radiography. MRI will be diagnostic, but will require sedation or general anesthesia in infants. Ultrasonography may be diagnostic in skilled hands (107).

Natural History. Children with congenital dislocations will have limited elbow and forearm range of motion and strength, and this will affect function. They must compensate with shoulder, wrist, or trunk range of motion to perform recreational activities and activities of daily living. If left unreduced, chronic arthritic pain could develop. However, this is not well documented.

In children with recurrent instability, pain may develop secondary to osteochondral injury. This can lead to osteochondral loose bodies and arthrosis-like pain.

Treatment

Operative Care. The isolated, congenital elbow dislocation has been rarely treated with open reduction (107, 183). These cases and operations are rare enough that generalized comment is difficult. The more abnormal the anatomy, the less likely that operative intervention will be successful.

In recurrent instability, ligamentous reconstruction, transposition of the biceps tendon insertion to the coronoid process, and an anterior bone-block procedure have all been advocated (183, 184). The choice or combination of procedures depends on the pathologic anatomy and the degree of instability.

It is the rare congenital elbow dislocation associated with ulnar dimelia and ulnar dysplasia that may warrant surgical reconstruction. Although ulnar dysplasia will be described in more detail in the section dealing with the wrist, it is worthwhile to discuss elbow reconstruction in this section. In type II ulnar dysplasia, there is partial absence of the ulna distally (184, 186). The proximal ulna articulates with the humerus but is usually unstable. With growth, the radius migrates proximally, leading to progressive loss of elbow flexion and extension. A supination deformity of the forearm may develop that limits forearm rotation (186). In these circumstances, creation of a single-bone forearm may improve cosmesis, stabilize the forearm, and improve elbow motion (184, 188). As described by Bayne (182), with this procedure, the ulnar anlage is completely excised and the adjacent ulnar artery and nerve are protected. Radial osteotomy is then performed proximally. The radius is placed distal to the ulna in an end-to-end manner. Intramedullary fixation is performed to connect the proximal ulna to the distal radius. If there is significant bowing of the radius distal to the osteotomy site, a second osteotomy is performed with passage of the intramedullary wire. If it is difficult to attain end-to-end fixation, then side-to-side fusion is acceptable. Resection of the dislocated proximal radius can be performed simultaneously or up to 6 months later. If there is any question of neurovascular compromise, it is advisable to delay the proximal radius excision (184). At the time of proximal radius excision, the posterior interosseus radial nerve should be exposed and protected.

Wood recommends that reconstruction of the complex elbow deformity associated with ulnar dimelia should begin at the elbow with excision of the lateral olecranon process (188). Reconstruction of ligamentous structures may be necessary

after excision in order to provide elbow stability. Excision of the lateral olecranon will reportedly provide improved passive elbow flexion and extension, but limitation in active elbow flexion may continue because of deficiencies in the biceps and the brachialis musculature. Tendon transfers for active elbow flexion have reportedly had limited success (188). This condition (and reconstruction) is so rare that in-depth analysis of treatment options is not possible.

Congenital Synostoses. These entities are classified as failure of differentiation of parts with skeletal involvement. In this section, congenital radioulnar and elbow synostoses will be discussed.

Congenital Radioulnar Synostosis. Congenital synostosis of the proximal radius and ulna is a rare malformation of the upper limb. It is caused by a failure of the radius and ulna to separate, usually proximally.

During the embryonic period of fetal development, the humerus, radius, and ulna are conjoined. Longitudinal segmentation begins distally and proceeds proximally. For a time, the proximal ends are united and share a common perichondrium. Genetic or teratogenic factors that are as yet unknown may disrupt proximal radioulnar joint development, leading to a bony synostosis. This represents a type I deformity. If rudimentary joint development occurs before developmental arrest, a rudimentary radial head will develop with a less severe degree of coalition. This is a type II deformity (190).

During this period of intrauterine development, the forearm is anatomically in a position of pronation (191). Failure of formation of the proximal radioulnar joint at this stage of differentiation will leave the forearm in its fetal position of pronation. With rare exceptions (192), the forearm is fixed in pronation with congenital radioulnar synostosis (191).

Congenital radioulnar synostosis is usually an isolated event. There is a 3:2 ratio of male to female. Positive family histories have been reported (157, 193, 194). It is a bilateral occurrence 80% of the time (195). The condition is also seen in disorders such as acropolysyndactyly (Carpenter syndrome), acrocephalosyndactyly (Apert syndrome), arthrogryposis, acrofacial dysostoses of Najjar and mandibulofacial dystosis, and Klinefelter syndrome and its variants (196, 197).

Although radioulnar synostosis is usually an isolated event, there may be associated anomalies of the musculoskeletal, cardiovascular, thoracic, gastrointestinal, renal, and central nervous systems. Cardiac anomalies include tetralogy of Fallot and ventricular septal defects. Thoracic anomalies include hypoplasia of the first and second ribs and the pectoral musculature. Renal anomalies involve anatomic malformations that can be screened by ultrasonography. In the central nervous system, associated problems include microcephaly, hydrocephalus, encephalocele, mental retardation, delay in attaining developmental milestones, and hemiplegia. Musculoskeletal problems include clubfeet, dislocated hips, polydactyly, syndactyly, and Madelung deformity (107, 160, 195, 196, 198).

Clinical and Radiographic Features. These children present for evaluation when they have a functional deficit. Generally, the degree of fixed forearm pronation determines the disability and the age of presentation. The presence of bilateral synostosis in marked pronation significantly limits function and leads to an earlier presentation. Most children will present for evaluation by school age. Radioulnar synostosis is often first noted by a teacher or a daycare worker when comparing the affected child with peers performing the same tasks (107).

Functional complaints are variable and include (a) difficulty in holding or using small objects such as spoons or pencils, (b) inability to dress owing to poor manipulation of belt buckles or buttons, (c) backhanded positioning when holding objects such as bottles or toys, and (d) difficulty competing in sports requiring upper extremity dexterity. Feeding and accepting objects with an open palm in forearm supination are often difficult (107, 195).

On physical examination, the elbow often has loss of its normal carrying angle and has a flexion deformity. The flexion contracture is usually minimal. Shortening of the forearm is more apparent in unilateral cases. Rotational hypermobility of the wrist compensates for the lack of forearm rotation (192, 194). Despite this ligamentous laxity, patients do not appear to develop symptoms of carpal instability.

Almost all patients present in fixed pronation. In the series by Simmons et al. (195), approximately 40% of patients presented with pronation of <30 degrees, 20% had pronation fixed between 30 and 60 degrees, and 40% had more than 60 degrees of pronation. Pronation of >60 degrees is most limiting.

Radiographs of patients with congenital radioulnar synostosis show anatomic variations from minor radial head deformities in patients with limited forearm rotation to full synostosis and absence of the radial head in patients with no rotation (160) (Fig. 22-26). The more extensive synostoses are usually fixed in more pronounced pronation. Plain radiographic classifications have distinguished partial and complete synostoses. In the partial synostosis there is often a rudimentary radial head present, but it is posteriorly or posterolaterally subluxated. In the complete synostosis the radial head is absent, and the proximal radius and ulna are a single bony mass. There is always an increased anterior bow of the radius. On occasion, the synostosis can extend into the middiaphysis of the forearm.

Occasionally, a patient will present with limited forearm rotation and normal radiographs. MRI of the proximal radius and ulna may reveal a cartilaginous synostosis that has yet to ossify or a fibrous tether that limits motion (107).

Natural History. In the absence of functional limitation, children with radioulnar synostosis should be observed. Children can often compensate for lack of forearm rotation if they have (a) synostosis in neutral-to-mild pronation (<60 degrees), (b) significantly compensatory radiocarpal and intercarpal wrist rotation, and (c) unilateral disease (107). These children present because they, their parents, and/or their teachers notice them performing home, school, or recreational tasks differently from their peers. However, when questioned

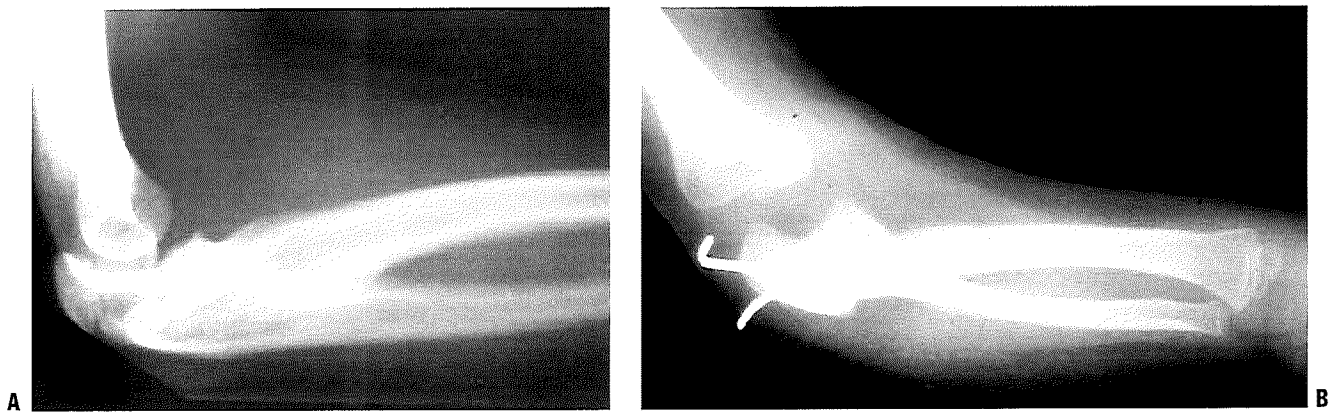

FIGURE 22-26. A: Preoperative radiograph of a congenital radioulnar synostosis. There is complete fusion of the proximal radius and ulna, and posterior dislocation of the radial head. The entire ulna is mildly hypoplastic. **B:** Postoperative radiograph of a derotation corrective osteotomy for this patient. A longitudinal wire is passed down the medullary canal of the ulna across the synostosis site. This Kirschner wire starts from the proximal ulnar apophysis. The osteotomy cut is performed through the synostosis. The transfixing wire is obliquely placed to secure the corrective derotation to a position of 0 to 20 degrees of pronation. (Figures courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

extensively, it becomes apparent that they are without pain or functional disability (194) These children and their families are best served by counseling regarding the diagnosis and functional issues of their problem, and reassurance that operative intervention would be unlikely to improve their condition.

Treatment

Operative Care. The ideal treatment would be to restore normal forearm rotation. Many surgical attempts to do so have been tried. Reported procedures have included division of the bony bridge (191); resection of the synostotic proximal radius to save the bicipital tuberosity, with (199–201) and without (202) muscle interposition; division of the interosseous membrane; and muscle, fat, fascia, or silastic interposition after synostosis excision (193, 203). All had limited success at restoring motion. Artificial joint replacement, with a metallic swivel in the intramedullary canal of the radius between the supinator and pronator teres, also failed (201). Tagima et al. (203) reported improved forearm rotation with synostosis takedown, radial osteotomy, and interposition of either a silastic or a free fascial lateral arm flap. Intraoperatively, synostosis takedown procedures can dramatically improve motion, but there is a high incidence of loss of motion in 6 to 12 months after surgery. At present, the functional gain does not seem to warrant this surgical intervention.

The alternative to synostosis excision is derotation osteotomy. The goal is to place the hyperpronated hand in a more functional position. The dominant extremity is given priority in bilateral cases. It is easiest to perform the osteotomy through the synostosis distal to the coronoid process. Before the procedure, an intramedullary ulnar Kirschner wire is placed to maintain control of the osteotomy. After completion of the osteotomy, the forearm can be rotated into the desired position of correction and can be held in this position by either percutaneous pins or external fixation (204). Generally, patients undergoing derotation osteotomy have a fixed preoperative position of 60 to 100 degrees of pronation. The final corrected position is often 0 to 20 degrees of pronation (107). Ogino and Hikino advocated measuring the preoperative compensatory wrist supination to define the desired operative osteotomy correction (192). Once this position is achieved, a second percutaneous Kirschner wire transfixes the osteotomy site obliquely, from the proximal ulna to the distal radius, across the derotated synostosis (Fig. 22-26). Because there is a high risk of compartment syndrome postoperatively (195, 205), it is important to avoid internal fixation that would require a second operation for removal if neurovascular compromise occurs. Resection of bone at the synostosis site (192), or dorsal and volar fasciotomies through the operative incision, lessen the risk of compartment syndrome postoperatively (107) and should be performed routinely. Others have advocated single or double osteotomies of the radius and/or ulna distal to the synostosis site (206–209).

Patients undergoing derotation osteotomies have been noted to show significant improvement in function and aesthetics. Bimanual tasks are easier. Single-handed tasks, such as holding a fork, no longer require backhanding in extreme hyperpronation. Activities of daily living, such as dressing and feeding, are performed more independently and with less adaptive shoulder and trunk motion.

Complications. The most significant complication is postoperative compartment syndrome. It has been reported in one-third of patients undergoing derotation osteotomy. This is attributable to changes in the vascularity and volume of the forearm compartments with derotation osteotomies in the range of 60 to 90 degrees. Compartment syndrome is more common in osteotomies with >85 degrees of rotational correction. Prophylactic forearm fasciotomy, or resection of a segment of synostotic bone, reduces the incidence of this

complication. If compartment syndrome is developing, the compressive dressings should be removed promptly, and the limb should be placed horizontally at the level of the heart. Compartment pressure measurements are routinely performed in the presence of tense compartments in a child with the clinical appearance of compartment syndrome. In pediatric patients, an increasing analgesia requirement and a high level of anxiety are the most diagnostic clinical signs of compartment syndrome (210). Removal of the oblique transfixing Kirschner wire is performed if removal of dressings and proper elevation fail to improve the situation. Removal of the oblique Kirschner wire allows the forearm to rotate to its preoperative position, lessens the tension on the interosseous vessels, and may reduce the pressure of the forearm compartments. Finally, if these maneuvers do not resolve the problem, emergent skin and fascia decompression is mandatory (107, 195).

With removal of the Kirschner wire in compartment syndrome, there is a risk of loss of operative correction. The longitudinal ulnar Kirschner wire helps maintain control of the osteotomy site and allows for controlled, repeat derotation 5 to 10 days later. Although more rigid internal fixation may seem more desirable, it unnecessarily complicates the procedure, especially if compartment syndrome develops. The use of external fixation will achieve the same goals.

Elbow Synostosis. Elbow synostosis is very rare. It occurs in isolation or in association with syndromic conditions. Humeroradial synostosis is more common than ulnotrochlear synostosis (211–213). The elbow flexion may range from 60 to 90 degrees. Often, there is also limited or no forearm motion. On examination, there will be no elbow motion.

Elbow synostosis is often associated with other upper-limb malformations, such as ulnar dysplasia (213). It has been described in siblings with humeroradial synostosis, indicating a potential genetic inheritance pattern. It frequently occurs with phocomelia variants (211). The limitation of elbow motion limits function, particularly if the affected extremity is dominant. The placement of a functional hand in space is limited by the lack of flexion-extension at the elbow. Compensatory trunk, head, and shoulder motion is difficult to adapt. Associated hand anomalies can further limit function (107).

Treatment. Attempts at synostosis excision and restoration of elbow motion have had minimal success. Techniques have included excision with muscle, fat, silastic interposition, or distraction arthroplasties. Although intraoperatively the motion can be improved, recurrence of the synostosis usually develops postoperatively. The use of continuous passive motion devices or distraction elbow hinge devices has not improved results (107). If the ankylosis leads to dysfunctional positioning of the hand in space, such as in the presence of an ulnar dysplasia, corrective osteotomy is indicated. Most often, this is a derotation osteotomy at the level of the synostosis (211). Correction of a marked flexion deformity acutely increases the risk of neurovascular compromise. There is no role for total elbow arthroplasty in the child because of the possibility of early mechanical failure (107).

Musculoskeletal

Osteochondromatosis. Deformity of the forearm is common in multiple hereditary osteochondromatosis, with between 30% and 60% of patients affected in various series (214–216). The most frequent problem seems to be distal ulnar osteochondroma, which selectively slows the growth of the ulna in the presence of continued radial growth. The resultant relative shortening of the ulna can lead to progressive bowing of the radius and/or possible radial head dislocation. At the wrist there is increased radial angulation of the distal epiphysis, with ulnar deviation of the hand and ulnar translocation of the carpus (216–219). These deformities can lead to progressive loss of forearm rotation. If radial head dislocation occurs, loss of elbow motion can occur and pain may develop. This section focuses on the treatment of ulnar shortening, progressive radial bowing, and radial head subluxation. The principles outlined here for osteochondromatosis have also been used in congenital syndromes with forearm growth discrepancies, such as Conradi and Morquio syndromes (107).

Natural History. There are very limited natural history data on patients with deformity secondary to osteochondromatosis of the upper extremity. There is ample information on the indications for, and the results of, surgical excision of osteochondromas and forearm reconstruction for these patients with deformities (220–223). The Shriners group in St. Louis (224) attempted to obtain natural history data by surveying their patients by telephone. Their data suggest that adults with forearm, wrist, and hand deformities from osteochondromatosis do reasonably well with activities of daily living and occupational tasks. Unfortunately, their data were limited because they could not reach many of their patients, and no patients were examined.

Treatment Indications. The presence of an osteochondroma alone is not an indication for surgical excision. Excision of an osteochondroma may improve growth when one bone is affected, but will not predictably improve growth or prevent recurrence with both bone involvement growth. However, if the osteochondroma is a source of pain, limitation of motion, or neurovascular or muscular impingement, then excision is indicated. In addition, children with forearm osteochondromatosis may present with progressive deformity, loss of pronation and supination, and wrist or elbow pain related to joint subluxation. The limitations of forearm rotation may be caused by impingement of osteochondromas distally or proximally. When the loss of motion is secondary to impingement alone, rotation will improve with osteochondroma excision (216, 217). In the presence of progressive forearm deformity, loss of rotation may also be related to bony malalignment, proximal radial head subluxation, or distal radioulnar joint dislocation. In these situations, rotation and radiocapitellar alignment can be improved by corrective radial osteotomy and ulnar lengthening (221). In the presence of radial head dislocation, reconstruction is very difficult. Attempts at reduction of the radial head by osteotomy or distraction lengthening techniques have had limited long-term success. Radial head excision has been advocated after skeletal maturity (222, 223). The creation of a single-bone

forearm may be the necessary salvage procedure in this complex deformity, and this can be performed at a very young age with successful results (216, 220, 223, 225, 226).

Operative Management. Operative intervention is indicated in the presence of either progressive deformity that limits motion or radiocapitellar joint instability. The indications, specifically in terms of deformity, are ulnar shortening by more than 1.5 cm, increasing distal radial articular angle >30 degrees, ulnar carpal translocation >60%, progressive radial bowing, and radial head subluxation (221). The key to managing radiocapitellar instability is to treat it before frank dislocation occurs. Once the radial head is dislocated, obtaining and maintaining reduction is difficult.

Most patients with forearm deformities secondary to osteochondromatosis can be treated with a single-stage operative correction. The ulnar shortening is addressed by simultaneous excision of the osteochondroma and Z-lengthening of the ulna. After the Z-osteotomy, distraction lengthening is carried out intraoperatively with an external fixator. When the desired lengthening is achieved, a plate is applied to maintain the length until bony healing is complete (107) (Fig. 22-27). This lengthening technique is, in essence, a rebalancing of the forearm skeleton. It realigns the proximal and distal radioulnar joints. In one series, lengthenings of 1 to 2.3 cm, leading to a neutral ulnar variance at the wrist, were obtained in a single stage (221). In most patients, forearm rotation was improved by an average of 40 degrees (107, 221). Results indicate improved range of motion and function with minimal risk of complications.

There are rare situations in osteochondromatosis in which correction cannot be obtained in a single procedure (176, 227, 228, 229). The option then is to perform serial lengthenings or gradual distraction osteoclasis. Up to 13 cm of length has been obtained by distraction techniques (228, 227). However, the rate of complications with distraction osteoclasis in the forearm has been cited as between 60% and 100%. Therefore, forearm lengthenings by distraction techniques should be performed cautiously by those skilled in the technique. The techniques available for distraction lengthening include unilateral external fixation frames (230–232), classic Ilizarov technique (231), and hybrid fixation using transverse Ilizarov wires fixed at 90 degrees to half pins (229, 233). Most surgeons performing distraction lengthening now use a hybrid technique so as to lessen the risk of neurovascular and muscle entrapment complications (228, 233, 227, 229). The fixator is preassembled as part of preoperative planning, with a half ring proximally and a full ring distally. In situations requiring angular correction, appropriate hinges need to be applied in order to obtain correction. Because each case is unique, the specifics of application are difficult to address in a review such as this. However, certain principles need to be adhered to. The pins need to be placed in the safe zone so as to lessen the risk of complications. Passive digital flexion and extension need to be at the full range intraoperatively after pin placement to ensure

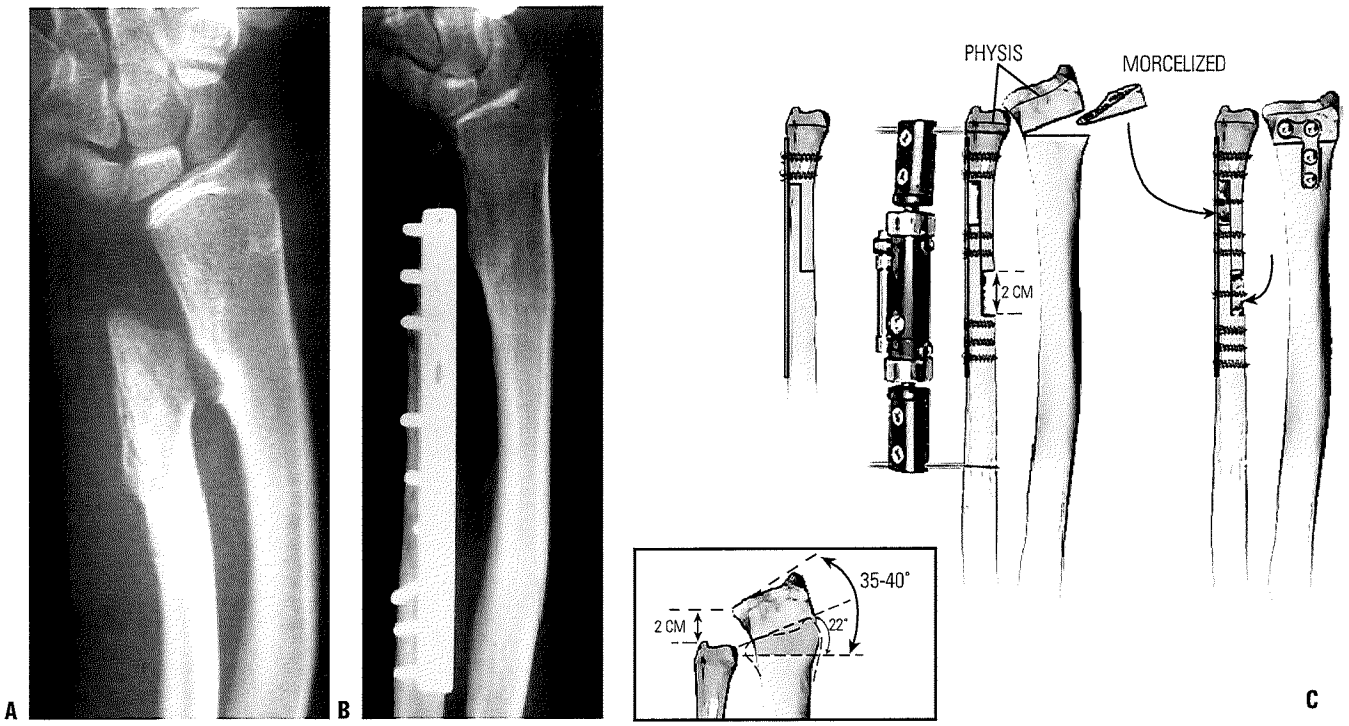

FIGURE 22-27. **A:** Preoperative radiograph of a patient with osteochondromatosis, ulnar shortening, and mild radial deformity, with recent progressive loss of forearm rotation. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.) **B:** Postoperative radiograph of single-stage lengthening of the ulna and radial osteotomy. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.) **C:** Illustration of the lengthening technique.

postoperative maintenance of motion. The preferred site for corticotomy is the proximal ulna metaphysis to enhance regeneration of bone (229). Lengthening begins 3 to 5 days after surgery and progresses at a rate of 1 mm/d, usually with an advance of 0.25 mm four times per day. Maintenance of passive and active range of motion of the shoulder, elbow, and digits is critical. Clearly, the loss of hand function is not worth the advantage of increased forearm length. Prevention and treatment of expected pin-track infection require meticulous pin care and judicious use of oral antibiotics. After the desired lengthening is achieved, the external fixator is left in place until there is sufficient regenerate bone to allow removal without the risk of fracture. In general, the fixator is left in place for at least twice the time necessary to obtain lengthening (107).

In the presence of radial head dislocation, the distraction technique has been used in an attempt to reduce the radial head before correcting the forearm deformity (229, 233, 176). A separate ring and an olive wire are placed in the proximal radius. Progressive distal migration of the radial head has been used for radiocapitellar reduction. Once the radial head is reduced, the forearm correction is performed as described in the preceding text. However, recurrent subluxation, stiffness of the joint, and pain have occurred after radial head reduction (107). This procedure should be used cautiously in the young.

The creation of a radioulnar synostosis is indicated for either painful radial head dislocation or radius and ulna instability that is not salvageable by other means. In these circumstances, it can result in a stable, pain-free extremity (222). Radial head excision is performed to decompress the radiocapitellar joint and improve the range of motion of the elbow. Correction of the deformities of the radius and ulna is performed at the same time as the radioulnar synostosis with internal fixation and bone grafting. Some increased length can be achieved with the single-stage procedure. The distal ulna and proximal radial resected bone are utilized as bone graft. Neutral rotation to 20 degrees of pronation is desired. Although this procedure is rarely indicated, these patients have excellent long-term results (222).

Pseudarthrosis. Congenital pseudarthrosis of the forearm is rare and clearly associated with neurofibromatosis. Wood (234) summarized the cases of forearm pseudarthrosis in the medical literature, and noted that, according to the published papers, 5% of patients with neurofibromatosis have pseudarthrosis of the upper or lower limb, whereas more than 50% of patients with congenital pseudarthrosis of the forearm have definitive neurofibromatosis, multiple café-au-lait spots, or a positive family history of neurofibromatosis. Congenital pseudarthrosis is most often seen in the tibia, but it has been described in all the long bones.

The survey by Wood (234) found 46 cases of forearm pseudarthrosis. The ulna alone was involved in 20 cases, the radius alone in 15 cases, and both ulna and radius were involved in 11 cases. Twenty-three of these patients had either neurofibromatosis (18 patients) or a positive family history of neurofibromatosis (5 patients). Reports of this disorder range from a single case to up to 6 patients (235).

As in tibial pseudarthrosis, all reports describing treatment options for this problem outline the difficulty of obtaining union with conventional cast immobilization or corticocancellous autografting or allografting, with and without internal fixation techniques. The role of distraction lengthening techniques for congenital pseudarthrosis of the forearm is unclear. There are several reports of the use of vascularized fibular grafts (235–237) to heal the pseudarthrosis. These reports indicate a high rate of union when vascularized fibular transfer is carried out. This is the preferred treatment for this disorder at present (Fig. 22-28). In the forearm, the fibula is internally fixed to the proximal ulna. Distally, the fibula is secured either to the soft tissues of the distal radioulnar joint and the triangular fibrocartilage complex (TFCC) or to the residual distal ulna, provided it is not too dysplastic and dysvascular. The vascular anastomosis is end to side in relation to the ulnar artery. At the donor site of a skeletally immature patient, the distal fibula is fixed to the tibia so as to prevent valgus ankle instability after harvesting a vascularized fibular graft (234, 238). The proximal fibular epiphysis can be transferred in the young patient to allow for growth (239). In this situation, the separate vascular supplies

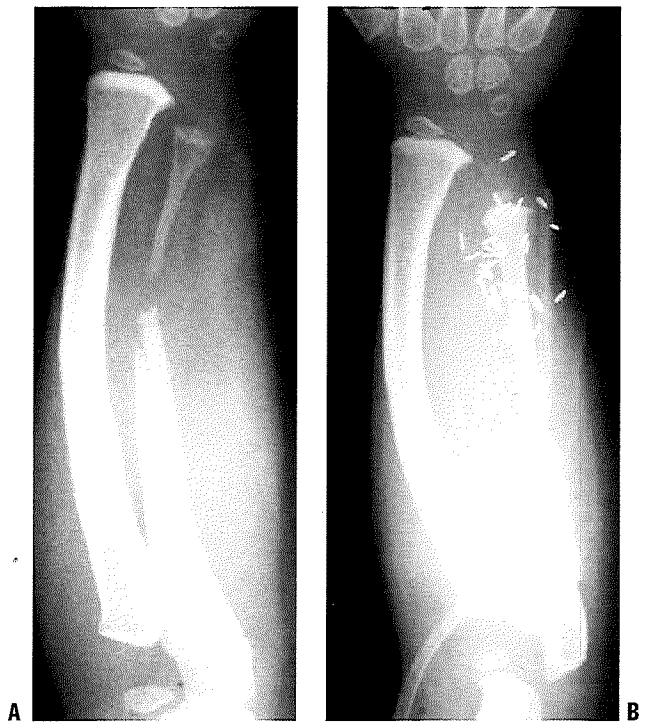

FIGURE 22-28. A: Preoperative radiograph of congenital pseudarthrosis of the ulna. Note the hypoplasia and tapering of the distal ulna. **B:** Postoperative radiograph of the vascularized fibular transfer, with proximal epiphyseal and physeal transfer, to establish distal ulnar growth. If the patient is very young, the microvascular transfer must include revascularization of both the diaphysis and the epiphysis of the fibula so as to obtain physeal growth. The most distal metallic clip indicates the top of the fibular epiphysis in the reconstructed distal radioulnar joint. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

to the fibular diaphysis and epiphysis need to be preserved and maintained with the transfer and vascular anastomosis. The soft-tissue support of the lateral knee must be reconstructed.

Creation of a single-bone forearm has been performed successfully as a salvage procedure (172). In the presence of an associated radial head dislocation, this may be the only successful option (107).

WRIST REGION

Congenital

Radial Dysplasia. Radial dysplasia is classified as a failure of longitudinal formation. It occurs in 1 in 30,000 to 1 in 100,000 live births (240, 241). The underdevelopment, or aplasia, of the radius is universally associated with hypoplasia or absence of the thumb and of the radial aspect of the carpus (242). The radial or preaxial deficiency has been classified by Bayne and Klug (241) into four types, ranging from a present but defective distal radial epiphysis (type I), to complete absence of the radius (type IV). James et al. modified this classification system to include type N, which has normal radial- and carpal-length thumb hypoplasia, and type O, which has normal radial length but carpal abnormalities (243). The severity of the radial deficiency determines the extent of the associated deficiencies of the thumb, digits, ulna, and elbow (Fig. 22-29). Therefore, the wide spectrum of anatomic deficiency includes mild radial deviation of the wrist and minimal thumb hypoplasia; complete absence of the thumb and radius; camptodactyly of the index, long, and ring fingers; foreshortening of the ulna; and a stiff elbow.

Pathogenesis. The pathogenesis of longitudinal deficiency of the radius is unknown. It has been postulated that abnormalities of the apical ectodermal ridge during upper limb development are the cause (7). Factors such as intrauterine compression, an inflammatory process, vascular insult, maternal drug exposure (thalidomide, insulin), and irradiation have all been raised as possible causes (1). There is no known genetic cause except when radial deficiency is associated with other congenital abnormalities. The pattern of inheritance is autosomal dominant or autosomal recessive, depending on the syndrome. There are many associated congenital syndromes, including cardiac, craniofacial, hematopoietic, musculoskeletal, gastrointestinal, and renal organ syndromes. There are associated chromosomal abnormalities, including trisomies 13, 18, and 21. The occurrence is most often sporadic.

Associated Anomalies. Although radial longitudinal deficiency can occur in isolation, it is commonly associated with other congenital malformations. Forty percent of patients with unilateral radial dysplasia and 27% of patients with bilateral radial dysplasia have associated malformations (244). It is imperative that these problems be assessed by clinical, radiographic, and laboratory evaluations as appropriate. These organ system malformations may present in a nonsyndromic pattern. Congenital cardiac, genitourinary, respiratory, skeletal, and neurologic problems occur in children with radial dysplasia. Similarly, many syndromes have been described in association with longitudinal deficiency of the radius. The most common are Holt-Oram syndrome, Fanconi anemia, thrombocytopenia with absent radius, and the "VACTERLs" syndromes of abnormal vertebrae, anus, cardiovascular tree, trachea, renal system, and limb buds. (The classic "VATER" syndrome comprises vertebral anomalies, anal atresia, tracheoesophageal fistula, and renal anomalies.) (245–247). Holt-Oram syndrome is an autosomal dominant disease characterized by upper limb malformations and major cardiac malformations (248). The genetic mutations for Holt-Oram syndrome have been identified as involving the TBX5 gene in over 70% of patients (249). Fanconi anemia also has an autosomal dominant inheritance pattern. In infancy, there are usually characteristic facial features (microphthalmos, strabismus, hearing deficits) (250). Pancytopenia often does not present until later in childhood. Fanconi anemia can be identified by a mitomycin C test, but it is now advocated that all infants with radial-sided defects be assessed early for Fanconi anemia with the diepoxybutane (DEB) test since a delay in diagnosis can have potentially life-threatening consequences (251). Thrombocytopenia with absent radius is also from autosomal recessive inheritance. While no causative genetic mutations have been identified, patients demonstrated deletions on the long arm of chromosome 1 (252); both the c-mpl gene and the HOX genes have been implicated (253, 254). The thrombocytopenia is present at birth. The platelet count usually improves with growth, and hand surgery should be delayed until it is safe (248, 253).

Clinical Features. The clinical presentation of radial dysplasia depends on the severity of the malformation. Bayne et al. (256) have tried to clarify the spectrum of clinical deformity by classifying radial dysplasia from type I to type IV. Type IV deficiency is the most common. Type I deformity involves a defective distal radial physis. This leads to a minor foreshortening of the radius and a prominent distal ulna. Although there is mild radial deviation of the wrist throughout life, problems with radioulnar incongruity, such as triangular fibrocartilage tears, ulnocarpal impaction syndrome, and distal radioulnar joint pain or loss of motion, usually do not occur. The major clinical issue is the associated thumb hypoplasia with opposition weakness. Type II deficiency involves limited proximal and distal radial physeal growth (the so-called radius in miniature). As a consequence, the wrist is more radially deviated, and the ulna bows. The thumb hypoplasia is usually more significant, with more deficiency of the radial carpus. Type III deficiency is the absence of the distal two-thirds of the radius. The wrist is more severely deviated, and the hand has limited mechanical support. The ulna is thickened and bowed. The associated thumb and finger abnormalities of hypoplasia and camptodactyly are more common and severe. Type IV deficiency involves complete absence of the radius. The ulnar bowing is marked. The thumb is usually absent. The index, long, and even ring fingers are often involved. The elbow may have limited range of motion. There is marked limitation of hand, wrist, and forearm function.

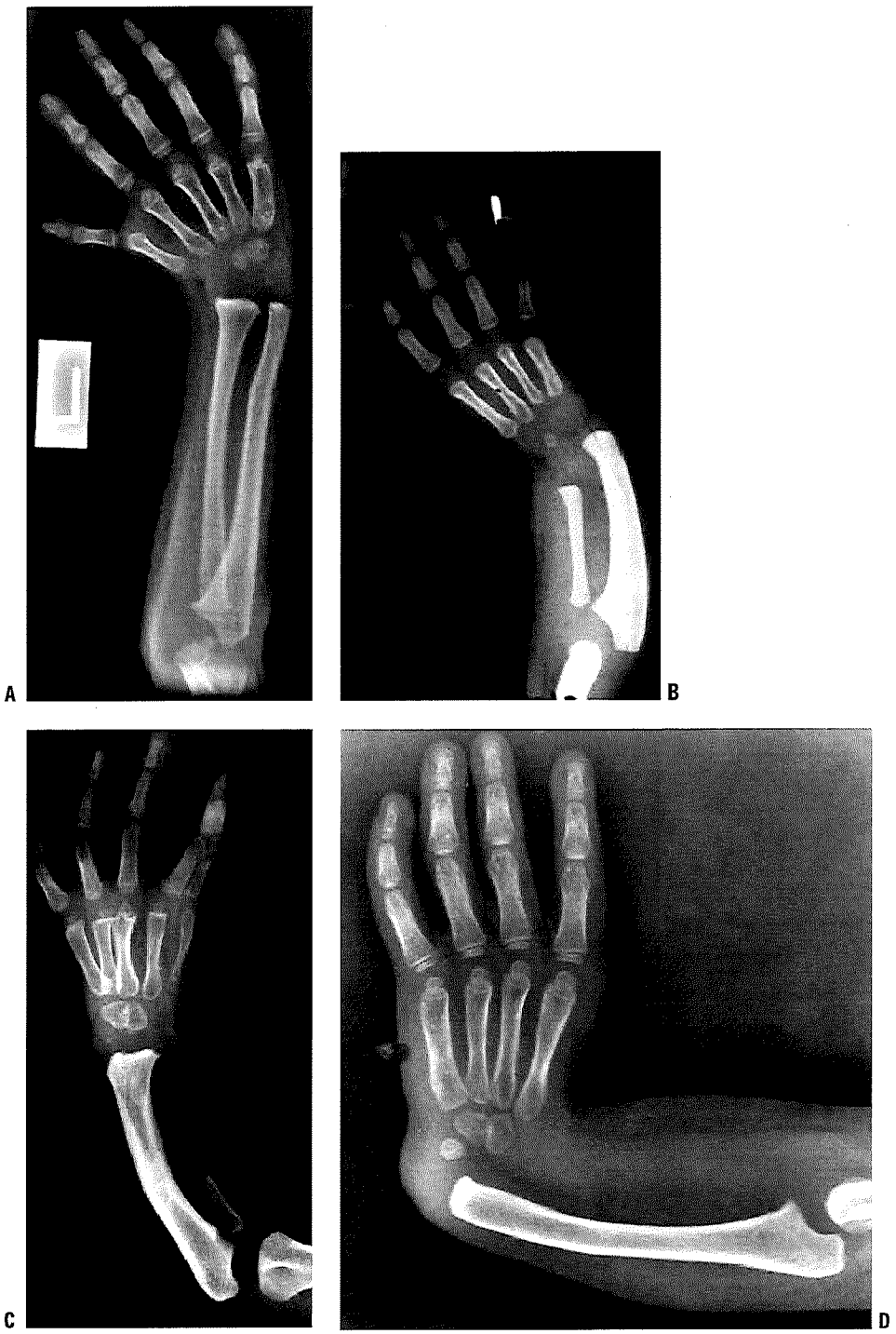

FIGURE 22-29. Classification of radial dysplasia types I through IV as represented by radiographs. **A:** In type I, the ulnar variance is positive as a result of the foreshortened distal radius. **B:** In type II, both the proximal and distal radial physes have deficient growth, with more radial shortening and ulnar bowing. **C:** In type III, the radius is partially absent. **D:** In type IV, the radius is completely absent.

Pathoanatomy. Radial dysplasia involves skeletal malformations and soft-tissue deficiencies on the preaxial or radial side of the hand, wrist, and forearm. The severity of the soft-tissue loss parallels the skeletal deficiency. The preaxial muscles arise from the lateral epicondyle and are normally innervated by the radial nerve. Therefore, the radial wrist extensors and brachioradialis are often absent or deficient. The pronator–flexor muscle mass is affected when its skeletal insertion sites are absent or hypoplastic. These structures may consist of only fibrous tissue (radial anlage) that maintains or worsens the deformity of the wrist and

hand with growth. Similarly, the neurovascular structures will be affected. The posterior interosseous and sensory branches of the radial nerve will be absent in a severe deformity. The radial artery is usually absent. The ulnar nerve and artery are usually present and unaffected. The blood supply to the hand comes through the ulnar artery, and at times the interosseous vessels or a persistent median artery. The median nerve is usually present and serves as a neural supply to the hand along with the ulnar nerve. However, the more severe the deformity of the hand and wrist, the more limited the neural and vascular supply will be to the hand.

Natural History. Generally, children with longitudinal deficiency of the radius have an unaffected central nervous system. As with any congenital upper limb malformation, children's creative minds allow them to perform all activities of life. However, they may need to use adaptive mechanisms. These generally include a spherical grip and lateral pinch to compensate for the absence of opposition (257). Fifty to sixty-two percent of patients with radial dysplasia have unilateral involvement. Even with severe unilateral radial dysplasia, these children will adapt their skills by increasing their use of the contralateral, normal hand and upper limb. Lamb (258) noted no functional impairment in patients with unilateral involvement. Individuals with bilateral involvement have more difficulty. Activities of daily living, such as hygiene, eating, and dressing, are affected. Adaptive techniques and alteration of clothes are often necessary. However, as Bora et al. (259, 260) reported, despite these adaptive modifications, patients without surgical correction were more limited than surgically treated patients. Finally, the issue of the aesthetic and psychological impact of radial dysplasia is difficult to quantify. The social setting is constantly changing, and peer perception plays a major role in an involved individual's self-perception. Parental and family perception and coping will surely have a profound effect on a developing child's self-image. There is limited objective psychological information at present.

Treatment. Treatment should address the following problems with radial dysplasia: (a) unstable wrist with lack of support for the hand, (b) digital weakness secondary to radially deviated wrist, (c) intrinsic digital weakness and deformity, (d) thumb hypoplasia or aplasia that results in lack of opposition, and (e) foreshortened ulna. All of these deformities affect function in the patient with radial dysplasia. In addition, there are significant aesthetic deformities in these children that can be improved by intervention.

Nonsurgical Intervention. The options for nonsurgical intervention in these children are corrective casting, bracing, and physical therapy. In infancy, the first goal is to achieve passive correction of the radial deviation deformity. In mild cases, this involves merely a home exercise program of wrist ulnar deviation, extension, and distraction stretching. In more severe cases, care involves corrective casting or splinting to gradually stretch the contracted soft tissues, and then maintain the correction. The splints are used in conjunction with a passive range-of-motion program. If attempts to correct the static radial deviation contracture are not successful within 6 to 12 weeks of vigorous therapy and skilled bracing or casting, the use of distraction external fixation to obtain soft-tissue and musculoskeletal alignment should be considered (261).

Once passive motion has been achieved, it is necessary to maintain the correction. Again, in mild forms of the condition, this can be done nonsurgically. A nighttime corrective splinting program during infancy and times of rapid growth is useful. This treatment is adequate for most type I and II malformations. In severe cases, the lack of a stable wrist out of the splint impairs hand and limb function. These children are candidates for operative correction at 6 to 12 months of age.

Surgical Intervention. There are two indications for surgery: persistent wrist radial deviation contracture (Fig. 22-30) and functionally limiting thumb deficiency. The surgical options for the wrist contracture and the lack of support for the hand have ranged from bone graft procedures to the ulna, centralization, radialization, ulnarization, and wrist fusion (262, 263). Surgical options for thumb aplasia are pollicization and microvascular toe-to-thumb transfer. Thumb hypoplasia can be surgically corrected with first web-space deepening, MCP joint stabilization, and opponensplasty tendon transfer. Less clear indications exist for the surgical treatment of digital camptodactyly, ulnar foreshortening, and radial hypoplasia in type II deformities.

Potential contraindications for surgery include (a) lack of elbow flexion, such that the wrist deviation enables the patient to perform hand-to-mouth and hand-to-neck activities;

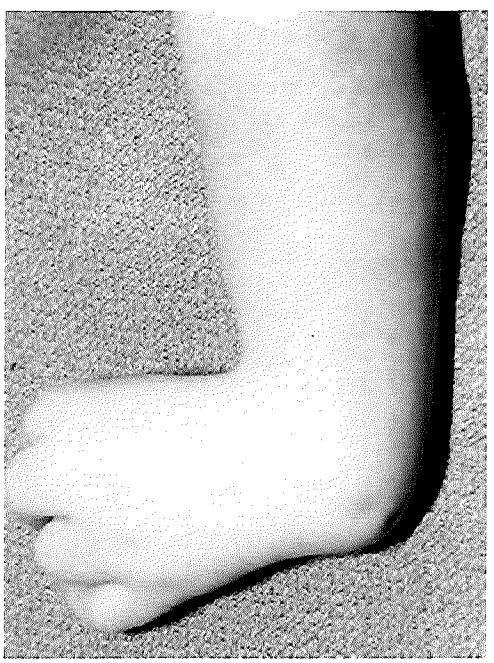

FIGURE 22-30. A patient with complete absence of the radius and thumb aplasia. Note the foreshortening of the forearm, 90-degree radial deviation at the wrist, and dimpling of the skin over the distal ulna, with the proximal and radially subluxated carpus and hand. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

(b) severe index finger deformity and weakness that will result in failed pollicization; and (c) severe medical problems that pose a risk to the patient's well-being and preclude surgical treatment.

The earliest forms of surgical correction for radial dysplasia involved improving the radial deviation deformity and lack of wrist support for the hand by grafting bone to the ulna. Albee (264), Starr (265), Entin (266), and Riordan (186) described the use of nonvascularized bone grafts from the proximal fibula to the ulna in Y-configuration to support the carpus and hand. These procedures resulted in significant short-term improvement. However, the transplant usually failed to grow, leading to recurrent deformity. Vascularized bone grafting has recently been advocated by Vilkki (267), in rare circumstances. Centralization of the carpus over the third metacarpal has been a standard treatment (258–260). Soft-tissue release of the radial contracture, contouring of the ulna to match the carpus, ulnar capsular reefing, and tendon transfers are typically performed. Stabilization is performed with pin fixation until healing is achieved. The problem with centralization is a high incidence of recurrence. Lamb (258) advocated modifying the technique by notching the carpus to inset the distal ulna. This lessened recurrence, but also decreased wrist motion and increased early ulnar physeal closure postoperatively. Function is clearly impaired when there is <30 degrees of wrist motion postoperatively. Buck-Gramcko (268) introduced radialization during the thalidomide crisis (269). Centralization procedure is modified by aligning the ulna with the second metacarpal. Tendon transfers from the radial aspect of the wrist (extensor carpi radialis and flexor carpi radialis, if present) to the dorsal, ulnar wrist are performed in order to rebalance the wrist and hand. The quality of the radial muscles clearly affects the success of the radialization procedure. With both centralization and radialization, correction is performed at the wrist. If there is a concomitant ulnar bow of >30 degrees, ulnar osteotomy should also be performed. This usually involves a multiple-level open osteotomy and intramedullary fixation.

In the rare situations in which passive correction of the wrist is not possible by splinting, casting, or therapy, distraction and deformity correction with an external fixator is performed. (270–273) As described by Kessler (261), this can be performed in infancy (Fig. 22-31). Often, after 3 to 6 weeks of external fixation an open centralization or radialization procedure is

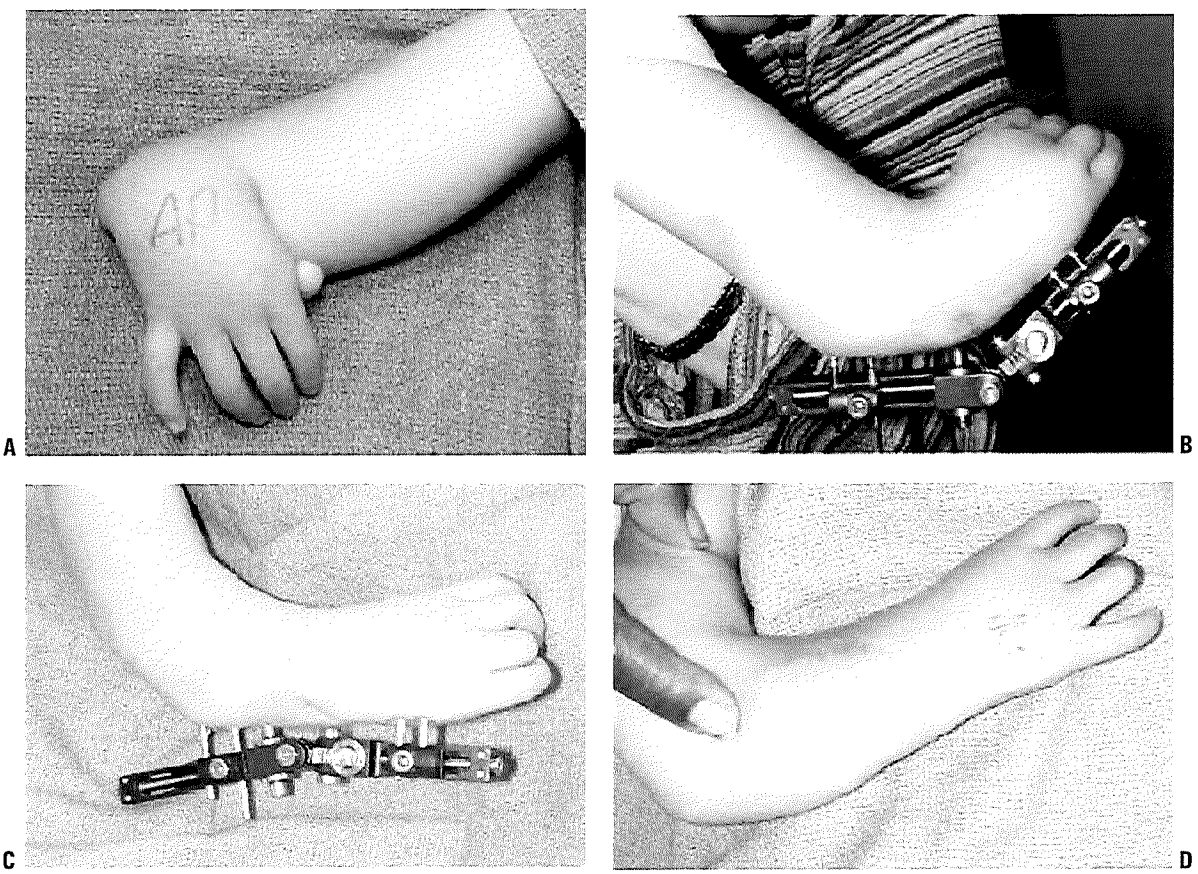

FIGURE 22-31. Clinical photographs of a patient undergoing progressive distraction lengthening to stretch the soft tissues, bring the hand out to length, and reduce the wrist over the ulna. This patient has a markedly foreshortened forearm. Preoperatively, there was volar, radial, and proximal subluxation of the carpus and hand that was not correctable with exercises and splinting. **A–D:** Preoperative **(A)**, early postoperative after fixator application **(B)**, after correction just before fixator removal **(C)**, and final result after centralization **(D)**. (Case and illustrations courtesy of Dr Allan Peljovich.)