

Disorders of the Upper Extremity

Introduction,	379	Infections,	476
Principles of Dressing and Splinting,	382	Traumatic Disorders,	479
Principles of Acute Care,	398	Tumors of the Upper Limb,	487
Principles of Reconstruction,	404	Microsurgery,	503
Congenital Anomalies,	410		
Juvenile Arthritis and Other Noninfectious Inflammatory Conditions,	467		

Introduction

Very few congenital anomalies of the upper limb can be restored to normal function and appearance. The goals of treatment are therefore to maximize functional potential, preserve sensation, and minimize scarring. The surgeon should offer support, first to the family and later to the child, to help them cope with the impact of knowing that even with today's medical technology, the limb cannot be restored to normal. Surgical intervention should be undertaken only when the intended procedure is to be done *for* the child, not merely *to* the child, and when the goals are clearly understood by the child, if possible, and the family.

Some surgical indications for common congenital conditions are straightforward, and abundant literature exists detailing the techniques. Surgery for other, less common conditions requires careful, sometimes creative preoperative planning *and* the ability to change those plans intraoperatively in an acceptable fashion if aberrant anatomy precludes a beneficial outcome.

In general, the goal of surgical treatment of congenital upper limb anomalies is to position the hands so that bimanual activity within the field of vision is possible. Preservation of joint motion where possible is desirable to allow the greatest reach of the limbs, especially with regard to reaching the face and the perineum. Because hand function requires intact sensation, scars should be carefully placed and peripheral nerves protected through the limb. Position, motion, and strength for grasp and release are the goals of surgery on the nondominant hand, which will stabilize objects for manipulation by the dominant hand. Position, motion, and strength for pinch and fine motor function are the goals of surgery on the dominant hand.

EMBRYOLOGY

The upper limb bud appears along Wolf's crest at approximately 23 days of gestation. Mesodermal proliferation within

the limb bud depends on a critical vascular structure that grows with the limb and supplies nutrients and oxygenation. The limb bud is innervated from its inception by nerves derived from the neuroectoderm. The leading edge of the limb bud ectoderm is a thickened ridge called the *apical ectodermal ridge*, or AER. It functions as an advancing mobile command center and contains the "architectural blueprint" for the limb structure. The AER is programmed to allow sequential transcription of crucial segments of DNA. The time-linked sequence of genetic transcription directed by the AER controls the proximodistal differentiation of limb structures. A number of *fibroblast growth factors* are involved in maintaining outward growth. Genetically encoded limb anomalies are "blueprint" variations and are built into the limb construction process.

Differentiation of the underlying mesodermal substrate occurs through the interplay of concentration gradients of growth factors and cellular mediators. On the caudal edge of the limb bud a *zone of polarizing activity* (ZPA) has been identified as crucial to the cranio-caudal orientation of the limb. The Hox gene family plays a role in this radioulnar (or tibiofibular) orientation of the limb. The WNT7 group of genes plays a role in dorsoventral patterning. The increasing number of clinically recognized conditions that can be mapped to specific gene mutations is contributing to a better understanding of the control of the three axes of limb development.

Vascular ingrowth supplying the advancing *progress zone* of undifferentiated mesodermal cells is critical to the development of limbs of normal length and size. A disruption of this vascular support limits the amount of mesodermal substrate and results in the spectrum of transverse limb deficiencies known as *symbrachydactyly*.

The final act of the AER is programmed self-destruction via a gene that codes for an endonuclease. This gene triggers a process known as *apoptosis*, or programmed cell death. Dissolution of interdigital webbing occurs at the end of the

embryonic period, at about 56 days of gestation. The limbs continue to grow in size during the fetal period.

A requirement for normal prenatal development is a protected uterine environment. Congenital limb abnormalities may also be due to teratologic, deforming, or disruptive influences on development. Teratogenic agents may affect development at the genetic transcription stage or may interfere with posttranscription growth factors. Physical forces may cause deformations of the growing embryo or fetus or disruption of normal development; an example of the latter is amnion disruption sequence (amniotic band syndrome).

Associated anomalies may occur in a child with an upper limb abnormality due to the timing of development of other organ systems. Recognized associations include anomalies of the heart, thoracic contents, spine, and kidneys, all of which develop concomitantly with the upper limb. Known hereditary associations also include hematopoietic and gastrointestinal conditions and lower limb anomalies.

HISTORY AND EXAMINATION

Pertinent History. The history taking begins with asking the parents to describe the child's problem. This description may focus a more specific inquiry. The expanded description of the presenting problem should include any functional limitations in age-appropriate activities and—elicited by gentle inquiry—the emotional status of the family and child in dealing with a deformity. The offer of occupational therapy to assist with activities of daily living and referral to support networks of families with similar children or to psychological counseling can be made at this juncture.

Questions about the pregnancy should elicit information about previous pregnancies and prior miscarriages (a clue to possible genetic conditions); illnesses and exposures to disease, chemicals, radiation, or drugs during the pregnancy; difficulties with the pregnancy, such as leakage of amniotic fluid or premature labor; and any antenatal testing such as amniocentesis or chorionic villous sampling that might have been done, and the reason for the testing. Information elicited about the delivery includes the infant's gestational age, birth weight, Apgar scores, condition at delivery, and anything abnormal about the placenta or umbilical cord. (A single umbilical artery is abnormal and is one of the abnormalities in the VATER association.)

Information to be obtained about the newborn period includes any extended hospital stay, diagnostic tests or treatment required, bleeding problems or the need for blood transfusions, nutritional status, and attainment of appropriate developmental milestones.

The pertinent family history for a child with a limb abnormality includes questions about similar anomalies in the families of both parents.

Physical Examination. The physical examination is focused by the presenting complaint but includes inspection of the entire child. The examiner should pay special attention to overall muscle tone while handling the child. A floppy baby may be morphologically normal but developmentally delayed. An appreciation of the appropriate reflexes according to the age of the child helps the examiner detect central nervous system (CNS) dysfunction. Craniofacial features such as premature closure of sutures, abnormal head

shape, facial asymmetry, crumpled or abnormal ears, or micrognathia suggest syndromes associated with limb abnormalities. As the observer becomes more familiar with the dysmorphic features, other facial features characteristic of syndromes will be recognized.

The neck and chest should be palpated for symmetry and integrity of the clavicle and ribs, the presence and bulk of the pectoralis and latissimus musculature, and symmetry of the breast tissue. The lower limbs are checked for stability of the hips, length, symmetry, and deformity.

The spinal examination includes a check of alignment and range of motion of the neck, and palpation of the dorsal spinal elements. Any cutaneous manifestation of spinal dysraphism such as a hairy patch or dimple should be investigated.

Examination of the limbs includes observation and documentation of all pertinent findings. The limbs can be inspected while the examiner evaluates the active and passive range of motion of all joints. Any asymmetry in size should be noted. Motor examination includes observation of active range of motion, palpation of muscle bulk, and assessment of any musculotendinous contractures or spasticity. Any anomalies should be noted in anatomic descriptive terms and by classified or named conditions if the diagnosis is clear. Pejorative terms such as “lobster claw” or “clubhand” should be avoided in favor of simpler descriptive terms such as cleft, deviated, bowed, or angled.

Assessment of sensation may be inferred from the texture, temperature, presence of sweat and papillary ridges, and integration of the limb or digit into function. Discrete sensory testing is impossible in the young child. Asymmetry in the size of limbs or parts of limbs may be a clue to asymmetric neural innervation of the limb, vascular or neural malformation, or a possible underlying tumor.

In the case of anomalous formation of limb structures, the examination should focus on both the appearance and the function. Observation of the child as he or she uses the limb in developmentally appropriate activities is important. A description of the functional limitations, such as inability to touch the mouth with the hands or to bring the feet into a plantigrade position, should be included in the record to provide a clearer picture of the abnormality.

The examination of the child may need to be repeated to obtain a clear picture of the problem and the potential treatment. Establishing a rapport with the child may be important in formulating a plan for treatment, and may take more than one visit. Having several small washable, nonlatex toys available for the child to play with is often helpful for observing active range of motion of the upper limbs and fine motor dexterity. White coats tend to provoke conditioned negative behavior, and examining the child will be more difficult.

Examining a young child is very different from examining an older child or adult. Tricks of the trade include using toys, decoys, diversions, and patience.

TIMING OF SURGICAL PROCEDURES

The rational timing of hand procedures in children depends on the parent, the child, the procedure, the surgeon, the hand itself, and the anesthesiologist. All must be ready for the procedure in order to achieve the best result afterward.

A logical approach makes decision making easier for the surgeon and parent and leads to a better result for the patient.

The Parent. Especially in children with a congenital deformity, the attitude of the parents must be one of cooperation. Parents need to work through a bereavement process, mourning the loss of the “perfect baby” they had anticipated prior to birth. The time required for family members to experience this is variable. The grief process may lead to unrealistic expectations of the surgical procedure and the surgeon. The parent and the surgeon must be patient and allow grieving to take its course. This is one reason why all but the most trivial reconstructive surgical procedures on the child’s hand are best deferred until after the first 6 months of life. Bradbury lists three stages of the grieving process;¹ these are discussed below.

DENIAL. In the denial phase the parent tends to minimize the impact of the deficit: “There’s nothing my child can’t do!”

ANGER. This is an important phase for the surgeon to recognize in the parent, since the anger may be directed toward the surgeon or may focus on the obstetrician’s failure to make a prenatal diagnosis. In a diagnosis such as Erb’s palsy, the parent may be unable to get past the assignment of blame. Before reconstructive surgery, the parent must understand that the only real solution is not “what if” but “what now.” Prior to this the parent is not capable of understanding informed consent.

DISTRESS. In the distress phase parents experience feelings of guilt, anxieties about future pregnancies, and loss of control. It may be helpful to consider with the parents the time of fetal development when the deformity occurred. Genetic counseling for the parents may be useful.

At times parents may seek to abdicate their responsibility for decisions until the child is an adult and can decide for him- or herself. This is inappropriate and should be strongly discouraged.

The Child’s Maturity. There are two important considerations relating to the child’s maturity level: cooperation and self-awareness.

Children’s capacity to participate in a helpful way in their care varies with age. We have found Bradbury’s “cooperation milestones” to be clinically useful (Table 14–1). They help the surgeon make reasonable decisions regarding the timing of treatment.

Another consideration is the child’s self-awareness, or when the child with a deformity learns “I’m different.” Although the child may notice a difference in the deformed hand by age 3 or earlier, self-consciousness rarely develops before about age 5, when the child begins to interact with people outside of the family. There are two important times when the deformity poses great emotional stress for the child. The first is between 5 and 7 years and the second is in early adolescence. Adolescent boys seem to be particularly prone to acute self-consciousness, perhaps due to a lack of ability compared with girls to talk about their deformity with their friends.

The Surgeon. It is important for the surgeon treating these children to have appropriate interest, training, and experience. Dealing with the child, the family, and the tediousness

TABLE 14–1 Cooperation Milestones in the Child

Age	Milestones
Infants (<6 mo old)	Accepts strange adults Accepts splinting with encouragement Less accepting of passive stretching Best comforted by parents
Babies (6–18 mo)	Anxious with strangers Easily distracted and reassured Tolerate splinting less well than younger babies Passive stretching more effective
Toddlers (18 mo–4 yr)	Very resistant to control and restrictions Short attention span
Children (4–7 yr)	Want to please adults Understand the rules Flexible in thinking Conjure up strange ideas about surgery—fear of waking up
Children (7–12 yr)	Logical in their thinking Compliant Can make meaningful decisions Still dependent on parents
Adolescents	Fragile body image Very aware of their appearance Socially dependent on peers Fearful of death (anesthesia) Distrustful of adults

From Bradbury E: Counseling People with Disfigurement. Baltimore, Paul H. Brookes Publishing, 1996.

of the operations and dressings demands a special commitment of time and patience to achieve optimal results. The first surgeon who operates on the child’s hand has the best chance for achieving a good-quality result. Even small gains in function may be important to the use of the hand. The hand has many emotional and physical differences from the lower extremity. Although the hand does not bear weight, it needs stability and power, and at the same time precision, flexible movement, and high-quality sensation. A child with a poor foot or leg can often be improved by amputation and fitting with a prosthesis. By contrast, a severely impaired hand is usually better than the best upper limb prosthesis. The hand also has a unique importance cosmetically and is never covered by clothing. Because of these considerations, pediatric hand surgery is usually best done by orthopaedic or plastic surgeons with a special interest in children’s hand deformities. Recently, special training in pediatric hand surgery has become available at some centers. The pediatric orthopaedist should keep in mind that the first surgeon who treats the child’s hand deformity has by far the best chance to optimize the result. Unfortunately, there has been a history of “one-upmanship” regarding who could do the biggest operation on the smallest hand. Sadly, this tragedy has often been facilitated by unknowing parents, who are usually anxious to get the treatment of their deformed child behind them. Poor results have occurred that could have been avoided with a rational treatment approach at a later time. The performance of hand operations is never an emergency (with the rare exception of a congenital band causing vascular compromise), and as a rule, within reason, later tends to be better. As Upton said so well, “The stakes are high, patience is a virtue.”³³

The Hand. Two unique characteristics of the child’s hand direct the appropriate timing of surgical intervention: the

TABLE 14-2 Growth of the Hand

Fetal age 8 wk	5 mm
Birth (40 wk)	60 mm
3 yr	120 mm
Full adult size	200 mm

growth of the hand and the unusual hypertrophic scar created in infants' skin at the site of hand incisions.

The hand increases in size tenfold between the time it is fully formed, at 8 weeks of gestation, and birth. After birth the hand doubles in size nearly twice before reaching adult size. It is helpful for the surgeon to realize that the first doubling occurs early in childhood, at about 2 to 3 years.² Waiting to begin surgical correction until age 2 to 3 years rewards the surgeon with a great increase in size and the opportunity for precision execution of a surgical plan (Table 14-2).

Fetal skin has the remarkable characteristic that it can heal without scar, a property that only bone maintains into adult life. Soon after birth, however, the infant skin makes an unexpectedly robust, hypertrophic scar at the site of surgical incisions. This is in contrast to the more favorable nature of deep scarring around their tendons and joints. This superficial hypertrophy of the skin incision scar is particularly a problem for the hand surgeon, because the hand's great mobility demands absence of scar in those areas of the skin that change length with hand motion. Areas that are especially important are the palmar surface of the digits and the webs (Fig. 14-1). Inaccurate positioning of incisions, especially in these areas, is a common cause of "web creep" after syndactyly. The poor results in children from "tiny Z-plasties" or any small flap are usually related to hypertrophic scar formation in the skin of the young child.

The Anesthesiologist. Finally, because of the elective nature of the surgery and the nonfatal aspect of these conditions, the unexpected anesthetic catastrophe is especially shocking when it occurs during reconstruction of the child's hand. An anesthesiologist trained in the care of the child and whose practice is mostly pediatrics is essential for hand reconstruction, because the surgical procedure itself carries almost no risk of death. The operating room in which the procedure is done should be equipped with appropriate instruments for children. Most anesthesiologists agree that non-life- or limb-threatening conditions requiring operations under general anesthesia are best delayed until after 6 months of age, and even later in children born prematurely, whose lungs may be less mature than would be expected by chronological age. Essentially all hand reconstruction falls into this category.

Suggested Surgical Milestones. We have developed certain guidelines for scheduling surgery on the hand in children. These are "soft" guidelines, and it must be remembered that timing also varies with the experience of the surgeon and anesthesiologist and the emotional readiness of the parent. In addition, children with hand anomalies often have serious visceral and systemic problems that take precedence over treatment of the hand anomalies (e.g., cardiac, renal, hematologic, spinal, and tracheoesophageal problems). Once these problems are attended to, accurate execution of

the hand surgical procedure must be balanced against giving the child the best opportunity to begin using the surgically altered hand. Finally, to some degree, pressure from parents may speed up or delay the time of surgery. Acknowledging all of this, Table 14-3 shows a few milestones we have found useful on the hand surgical service at the Texas Scottish Rite Hospital for Children.

The following example illustrates the importance of a plan for reasonable timing based on the principles discussed above. It is critical for the parent to understand the goal of treatment, the methods used to achieve the goal, and the limitations imposed by the age and size of the child. Such early explanation of a long-term plan makes it easier for the parents to accept what may be a prolonged course of treatment that requires the cooperation of and input from the entire family.

Radial Clubhand

- *Birth to 6 months*—Children at this age often seem to tolerate splints better than stretching exercises.
- *6 months to 12 months*—Passive stretching is added or may replace poorly tolerated splints in the slightly older baby. We have not been impressed with the results (in our hands) of external fixation and distraction.
- *About 1 year*—If passive alignment has not been achieved, we now consider a forearm-muscle-origin slide (and, in the TAR syndrome child, release of the often present anomalous brachioradialis-type muscle which spans the elbow). No bridges are burned, and the slide may occasionally avoid formal centralization, which may damage the distal ulnar epiphysis, the major source of longitudinal growth in these already short forearms. This procedure is an adjunct to therapy. Splinting and stretching are resumed after operation.
- *2 to 3 years*—Now that the first size doubling has occurred, we usually find that when more complex operations such as free joint transfer or centralization are indicated, they are best deferred until this time.

REFERENCES

Introduction

1. Bradbury E: Counseling People with Disfigurement. Baltimore, Paul H. Brookes Publishing, 1996.
2. Feingold M: Clinical evaluation of the child with skeletal dysplasia. *Orthop Clin North Am* 1976;7:291.
3. Mathes K, Manske PR, Upton J: Symposium: Changing concepts in the management of congenital hand anomalies. *Contemp Orthop* 1993; 27:481.

Principles of Dressing and Splinting

DRESSINGS

Dressings applied after an operation are of special importance in hand surgery, where uncomplicated wound healing usually makes the difference between a good result and a disaster. This is particularly important in operating on the hands of children. The combination of a child's wiggly hand and the small flaps and skin grafts that cannot tolerate motion is a recipe for disaster during the postoperative period. The small child seems to do his or her best, even if

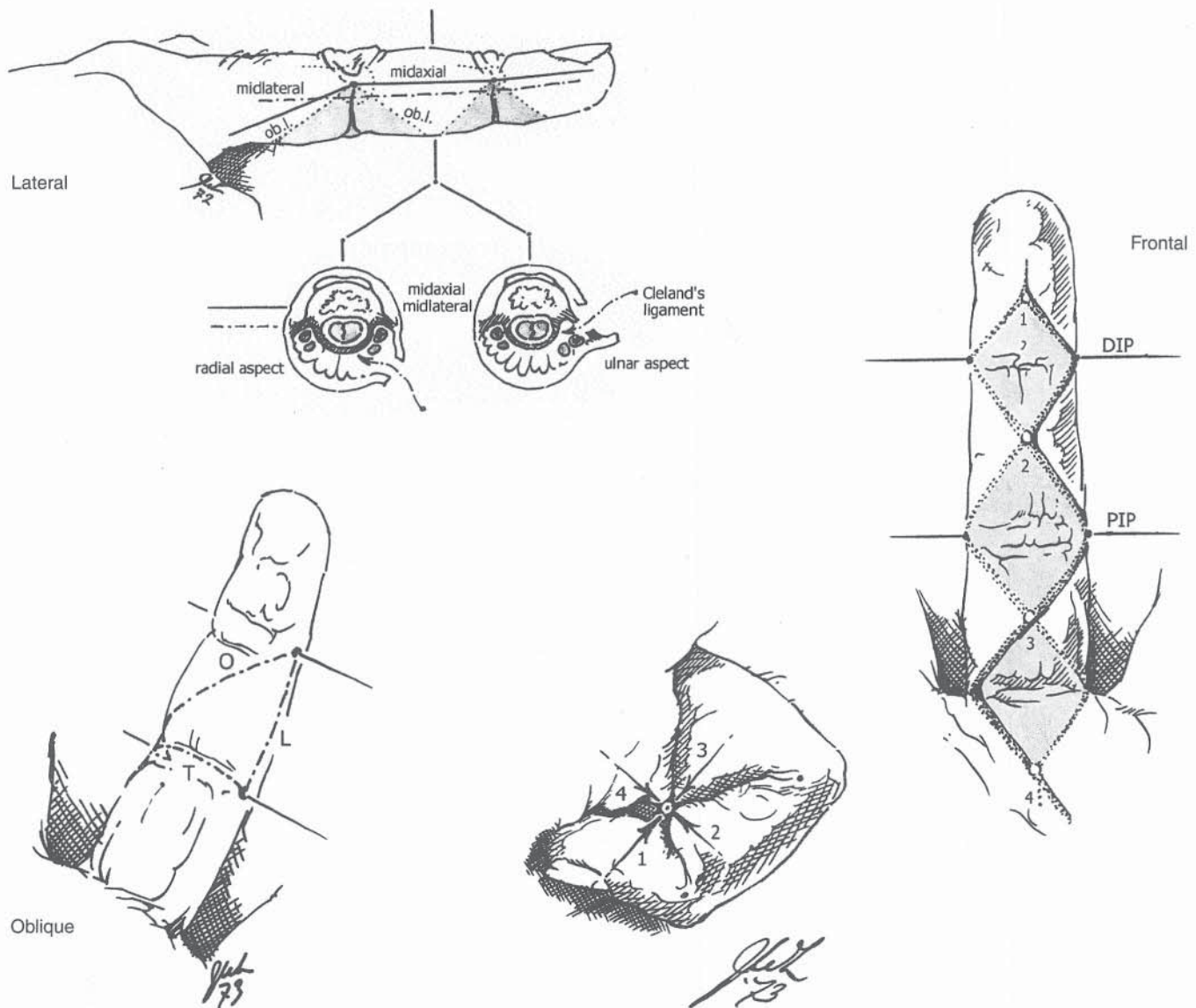


FIGURE 14-1 Precision design of digital skin incisions is even more critical in the child's hand than in the adult's, owing to hypertrophic scarring in the hands of small children. The figure illustrates the fundamental principles for the correct design of these incisions. Note that the incisions are based on the axis of rotation of the interphalangeal joints, as shown in the lateral view illustration. The oblique view shows the three basic interaxial lines from which all correct incisions are derived. The frontal view shows the two digital diamonds containing skin that changes length with finger motion. Any longitudinal scar located in these diamonds will hypertrophy and contract. (From Littler JW, Cramer LM, Smith JW (eds): Symposium on Reconstructive Hand Surgery, vol 9, pp 90-91. St. Louis, CV Mosby Co, 1974.)

unconsciously, to thwart a successful operation. Therefore, the objective of the hand dressing is primarily to protect the child from him- or herself. Preventing problems is by far the favored strategy in hand surgery. The only reasonable means of preventing problems is with a carefully applied, rigid dressing, previously made of plaster but now usually made of fiberglass.

It is most depressing for a surgeon when, after hours of well-planned and careful operating, the child pulls the dressing off in the recovery room, usually taking the skin grafts along with it. The child and surgeon don't get another chance to repeat the operation as well a second time. In a small child, this dressing should not be changed without indication.

What follows is a method we have evolved over the years

at the Texas Scottish Rite Hospital for Children for treating younger children. It is based on avoiding complications painful to the child, parent, and surgeon. Especially in children under age 8 or 9 years or who are not likely to understand the importance of leaving the dressing on, the method has worked well for us.

More than the usual care in the application of plaster or fiberglass dressings in these little hands is mandatory, and failure to do so may lead to the complications that the dressing is planned to prevent. In addition, the anesthesiologist must keep the child completely still during application of the entire dressing because movement by the child during dressing application almost always prevents a well-fitted dressing without pressure points.

TABLE 14–3 Surgical Timing Guidelines Followed by the Hand Service of the Texas Scottish Rite Hospital

Conditions that should be corrected in infancy (under local anesthesia):

- Floating polydactyly: The floating digit is usually ulnar (postaxial) and dangling from a narrow skin and vascular tether. Ligation is well accepted by the mother, who has recently seen the umbilical cord desiccate, just as the polydactylous digit will. If ligation-amputation is not done, as the child gets older the family or patient may develop a curious morbid attachment to the useless part, which may complicate the best plans for reconstruction.
- Strangulating amniotic bands: Bands causing obvious vascular compromise that need to be released to maintain viability (or, more often, amputation of a nonviable part) are rare. Z-plasty correction of the clefts formed by the band in these small hands should wait until the child is larger.

Conditions that should be corrected after age 6 months but before 1 year:

- Border syndactyly: Especially the thumb-index web and the ring-small finger web, which cause serious loss of function of the thumb and tethering of the ring or index finger.
- Complex osseous syndactylies: Especially those with transversely positioned bones, which progressively deform the hand as it grows. Some cases of central polydactyly may be optimally treated now.
- Apert's polysyndactyly: Because of the prodigious number of operative procedures required in these children, we like to start early and often do them bilaterally with two teams of surgeons.
- Macroductyly needing amputation: Although rare, such massive macroductyly may be present at an early age that amputation is the only reasonable course of action.
- Broad-based polydactyly: Cases that could not safely be treated before age 6 months with ligation under local anesthesia.
- Thumb duplications: Here the skill of the surgeon may alter the timing of reconstruction of a duplicated thumb. Preaxial thumb polydactyly is unique in that reconstruction is almost never a simple amputation, and careful consideration of ligament reconstruction is critical for long-term joint stability and alignment.

Conditions in which surgery can wait until the second year of life:

- Simple or cutaneous syndactyly involving the long and ring fingers or the long and index fingers: By 12 to 18 months the hand is much larger than an infant's hand, and precision construction of flaps and skin grafts leads to a more predictable result. Even greater delay may be appropriate in the child with an understanding parent.
- Pollicization: We prefer the age of 18 to 24 months in order to allow technical precision in the execution of this multistep and important procedure.
- Angular deformities requiring osteotomy and fixation: The larger size of the bone allows more accurate and effective use of K-wires.
- Thumb-index web reconstructions: Thumb-index web reconstruction affords the most important opportunity to improve hand function. We do these operations earlier if bony connections are present in the web and do them later when only cutaneous restraint is present.
- Hypoplastic thumbs, hands with ulnar dysplasia, and complex polysyndactyly frequently are treated at this time.

Because of the importance of avoiding complications, there is little if any place for a soft dressing on a child's hand. We follow the admonition of Richard Eaton—"Things heal faster under plaster!" Furthermore, short-arm casts are rarely used in young children. The smaller the child, the larger the dressing required. In children less than 8 or 9 years old, we rarely use anything but a long-arm cast, since putting a short-arm cast on small pudgy arms is much like putting a cast on a carrot. It won't stay where it is applied. This is one area where "more is less" rather than "less is more."

In operations on the fingers or palm of a small child, we also use a safety cast or mitten casts. These casts cover the fingers (which have been loosely wrapped with gauze and cotton padding) entirely in a loose-fitting shell to prevent the child from getting to the fingers and to keep unwanted food, toys, and other items out. The surgeon must be certain of lack of vascular compromise prior to final "finger cage" application, since continuous direct observation is sacrificed for protected wound healing. We have found this trade-off to be overwhelmingly a rewarding one.

Of course, when the surgeon is concerned for any reason (if the child has persistent or unusual hand pain, an unexplained fever, or the odor is foul), the dressing should be removed and the wound inspected. In the small or uncooperative child, this may best be done under general anesthesia so that another

dressing can be carefully applied. Changing the dressing before primary healing in 3 to 4 weeks is rarely necessary when each step of the dressing application receives from the surgeon the same meticulous care he or she gives the operation prior to wound closure. Leaving the responsibility of dressing application to the most junior or inexperienced member of the operating team can destroy the best efforts of the operating surgeon. Conversely, changing the cast for no reason prior to healing may severely complicate such operations as tendon repairs and skin grafts. Unless applied under general anesthesia, a subsequent new dressing on a small uncooperative child rarely fits as well as the one applied initially.

The Texas Scottish Rite Hospital hand dressing application protocol has several steps, which are illustrated in Figures 14–2 through 14–10:

1. Wound closure
2. Primary dressing
3. Gentle compression components
4. Padding and skin protection
5. Rigid outer dressing

Wound Closure. We prefer to use fine absorbable sutures (6-0 plain gut) dyed blue with the marking pencil for easy visualization (Fig. 14–2). This obviates later suture removal, saving time and anxiety on the part of patient, parent, sur-

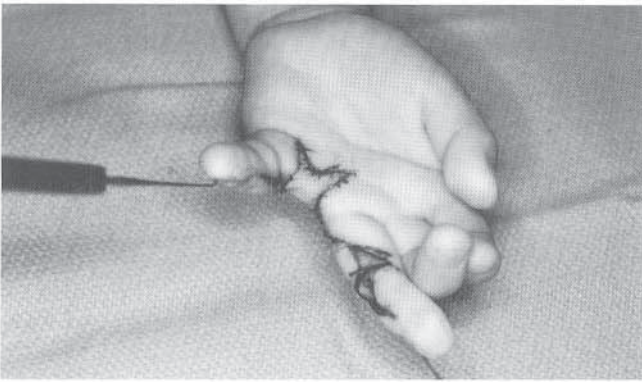
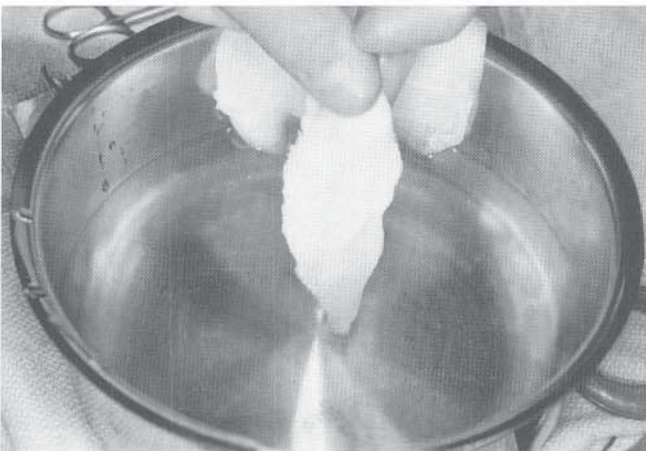


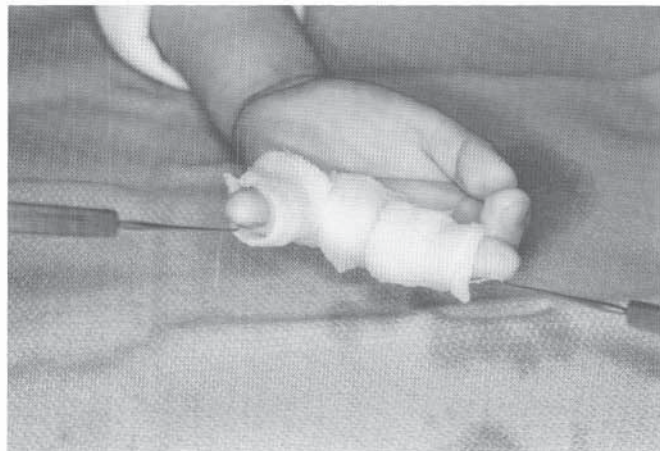
FIGURE 14-2 The use of fine absorbable suture to close the skin obviates later suture removal, which is stressful to the child, parent, and surgeon. Dying the suture blue with the marking pencil makes the suture readily visible.



FIGURE 14-3 The primary dressing material is a gauze treated with petroleum jelly and an antibiotic and applied in a single layer directly to the incision. This kind of dressing prevents adherence and is easily removed at the postoperative visit. Only a single layer is used, to prevent maceration and allow fluids to pass out of the wound into the next layer of the dressing.



A



B

FIGURE 14-4 A and B, A tailored gauze strip is soaked thoroughly in sterile saline solution and applied over the primary dressing. This dressing layer closely coapt to the contours of the hand. More important, the moisture in the dressing allows movement of blood out of the wound and into the gauze by capillary action.

geon, and nurse. These fine absorbable sutures are weak and must be protected with the rigid dressing described below.

Primary Dressing. The primary dressing goes directly on the wound. We prefer a single layer of fine mesh gauze treated with Vaseline and antibiotic (Fig. 14-3). Use of the Vaseline-treated gauze minimizes sticking of the dressing to the wound, and if a single layer is used there will be no maceration of skin, since the blood from the wound can easily pass into the dressing. This important drainage of blood out into the dressing is facilitated by application of a saline-soaked dressing sponge of appropriate size (Fig. 14-4). The well-moistened gauze also contours well to the complex shapes of the hand and digits.

This primary dressing is altered when a skin graft is present by applying the Xeroform in a single sheet combined with a dry gauze just covering the wound. This dressing is then secured with paper tape (Steri-Strips) and, once secured, is moistened with saline. We call this the “Steri-Strip stent” and have found it most useful for securing the skin graft, as is required, for example, in syndactyly reconstruction (Fig. 14-5).

Gentle Compression Components. This layer holds the primary dressing in place and is carefully applied with just enough compression to accomplish this aim but not so much as to restrict circulation. Applying this layer requires some practice and is critical to success. Illustrating the technique of just the correct amount of compression is difficult.

The first layer of the compression components is one of fluffed gauze formed from single dressing gauze opened up and formed into cones. These cones are gently placed between the fingers to prevent maceration but are not forced into the webs so to restrict circulation (Fig. 14-6A). The fluffs are secured by the appropriate size roller gauze (e.g., Conform, Kling) in 2- or 3-inch widths (Fig. 14-6B). Although gentle compression is required to make this first compression layer fit the hand well, all tightness around the



FIGURE 14-5 The “Steri-strip stent” dressing is used for all skin grafts. It is made from a carefully tailored, dry gauze piece held in place by $\frac{1}{4}$ -inch or $\frac{1}{2}$ -inch paper tape. Benzoin or other adhesive liquid may be applied to secure the tapes especially well. When well secured, these stents are also soaked with sterile saline.

forearm must be avoided. After proper application of this layer, the hand should be securely in the proper position of slight wrist extension and thumb abduction. (The exception is following flexor tendon reconstruction, in which case some wrist flexion is required to protect the tendon repair.)

Padding and Skin Protection. Now a layer of cotton cast padding (Webril) is applied to protect the skin and bony prominence and make removing the rigid shell easier (Fig. 14-7). Plaster or fiberglass will stick to gauze dressing and are more difficult and time-consuming to remove later. Orthopaedic felt applied to the upper portions helps protect the skin from the very abrasive fiberglass (Fig. 14-8).

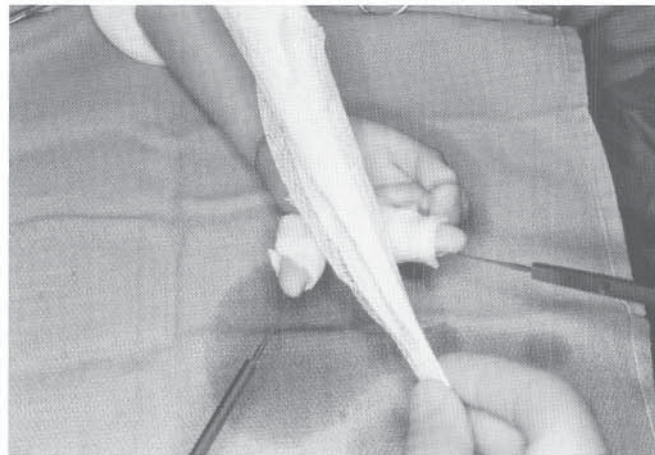
The tourniquet must now be deflated, prior to the next stage, to verify unobstructed and unequivocal digital circulation. If circulation is in doubt, it must be restored, and this may occasionally require removal of the dressing at this point. The hand should be kept elevated now, since during



FIGURE 14-7 Now the fluffed layer is secured with roller gauze (2-inch or 3-inch Kling). Care is taken to secure the deeper layer with just enough compression to hold it in place but not so much that it will occlude circulation into or out of the hand. Special care is taken to avoid any tightness around the forearm or digits. However, some gentle compression is necessary over the hand and palm to compress the fluffed gauze layer enough to provide a total contact fit and security to prevent motion during wound healing.

the next few minutes, during post-tourniquet hyperemia, the arm volume is equilibrating prior to the application of the rigid outer shell.

Rigid Outer Shell. This important layer has two functions. The first is to hold the primary dressing in the position applied, and the second as to function as armor plating to protect the wound against molestation. Plaster-of-Paris casts have worked well in the past and have the advantage that they can be accurately molded. However, fiberglass is lighter and more durable, and simplifies cleanup. Recently a new softer, incompletely setting fiberglass (Softcast) we have found has the advantage that it can be removed weeks later by simply unwrapping it. Softcast obviates use of a noisy cast saw, which is especially frightening to children and parents when used around the hand (Fig. 14-9).



A



B

FIGURE 14-6 A and B, The gentle compression component of the dressing. The fluffed gauze formed into a ghost is gently and carefully positioned between the fingers and in the web of the abducted thumb. This provides the base layer for a total contact dressing to secure the position desired by the surgeon. Between the fingers the surgeon must be especially careful not to pull the fluffed gauze so tight that it occludes the circulation.



FIGURE 14-8 Now a layer of cotton cast padding (Webril) is smoothly applied to the skin and prevent adherence of the fiberglass or plaster to the gauze. This layer greatly facilitates removal of the rigid layer at the patient's postoperative visit. The tourniquet is now deflated and circulation is verified unequivocally. If doubt exists, the dressing must be adjusted to allow unimpeded flow of blood.

Next, in small children, the digits are first well padded and covered with cast padding completely enclosed by a fiberglass mitten. This prevents food, toys, and those around the child from molesting the wound. Some active digital movement by the child under the fiberglass is possible (Fig. 14-10). A 2-inch sling of stockinette can be fashioned and put around the chest or neck of the child as indicated (Fig. 14-11).

SPLINTING

Introduction. Proper splinting of the hand and upper extremity is an acquired skill. It requires knowledge of anat-



FIGURE 14-9 Prior to application of the rigid dressing of fiberglass, a band of orthopaedic cotton felt is usually applied at the top of the roller cotton cast padding to protect the child's sensitive skin from the rough and irritating plaster or fiberglass. The fiberglass is applied as smoothly as possible, with the surgeon carefully avoiding stretching it too tight, until all but the digits are covered. At first a short-arm dressing is applied; when it is secure, in younger children it is always extended up to a long-arm rigid dressing. The long-arm portion should be carefully extended as far proximally as possible; there is little use for a "short" long-arm cast in children. The elbow position should be flexed at least 90 degrees to prevent the child from extracting the arm either partially or completely before the postoperative visit.

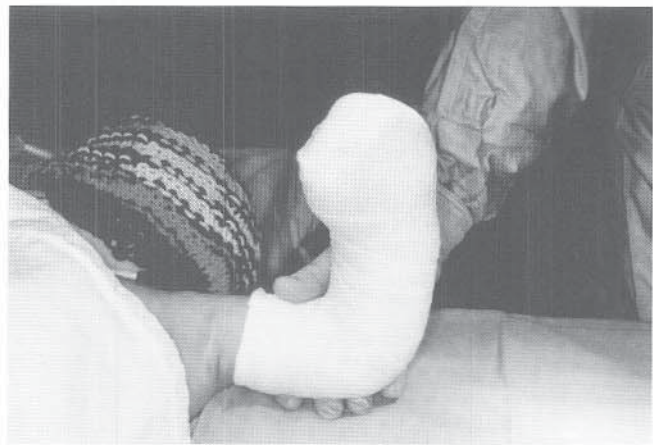


FIGURE 14-10 After the long-arm fiberglass dressing has been applied the digits are carefully and copiously padded and the "safety cast" fiberglass mitten outer shell is applied. This keeps food and toys out of the wound and protects it from unwanted molestation by those around the child, while allowing some limited motion of the digits under the rigid dressing.

omy, an understanding of the condition being treated, attention to detail, and considerable practice. For splints used in the postoperative period, an operative note should be available to the therapist, and it is preferable that the surgeon discuss in detail his objectives and concerns directly with the person who will fabricate the splint. There are several manuals of splinting that detail fabrication of standard splints, and the reader is referred to them in the bibliography.¹⁻⁴ Splinting in pediatrics requires the ability to work quickly as well as the development of an effective method for holding the desired hand position as the splint material cools.

Materials. The first thermoplastic hand splinting material to be introduced was Prenyl (Johnson & Johnson Medical), in 1964. It was soon followed by Aquaplast, Polyform, and Orthoplast (polymeric hydrocarbon).⁴ Since that time a number of other products have become available, some having properties that make them particularly useful to the

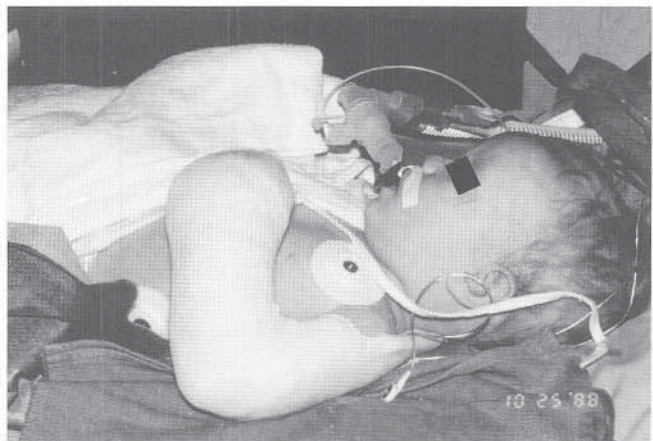


FIGURE 14-11 In some children a length of 2-inch stockinette is formed into a sling or restraint and placed around the neck or chest as needed. This kind of restraint is most useful in the very chubby child who is very active.

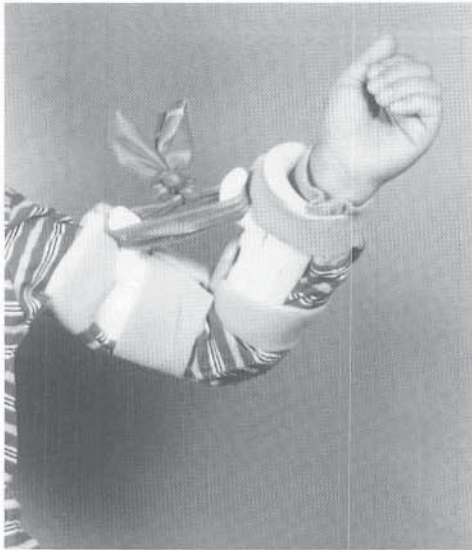


FIGURE 14-12 A dynamic elbow flexion splint fabricated out of Ezeform with a theraband dynamic component. This splint is more comfortable if a sock is placed on the arm or if the splint is worn over a long-sleeved T-shirt.

therapist caring for the hands and upper limbs of children. The therapist should take the time to practice carefully with any new material, learning the often subtle and unique characteristics of the particular polymer. These characteristics include drapability, softness, setup time, and remoldability. The best splints are ones that fit, accomplish their intended purpose, and are worn faithfully. Achieving these goals is more the result of the therapist's skill and his or her understanding of the chosen material than of which polymer is actually used. Complete understanding of the material's characteristics, attention to detail in design and fit, care in application, and close follow-up adjustments are the hallmarks of effective splinting.

EzeForm is a strong and durable material that does not develop fingerprints. Molding is relatively easy, and because the material has no plastic coating, pieces of it can be joined without the use of a solvent. It is strong and rigid and does not age or stretch. However, this can be a disadvantage if draping or precise contouring is needed. The working time is adequate to easily allow positioning of the splint. The

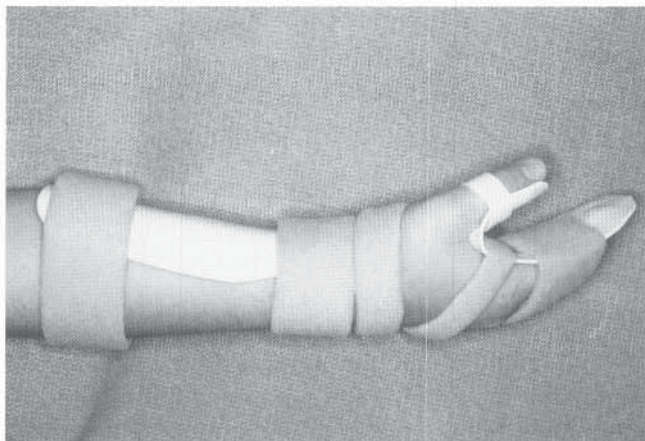


FIGURE 14-13 A resting hand splint fabricated out of Ezeform Light.



FIGURE 14-14 A dorsal hand base C-bar fabricated out of Polyflex II.

standard $\frac{1}{8}$ -inch thickness is particularly useful for fabricating splints for patients with upper limb spasticity.³ The strength and bonding qualities make this a desirable material for a dynamic elbow flexion splint (Fig. 14-12). The $\frac{1}{16}$ -inch EzeForm Light, a thinner version of EzeForm, heats up quickly and hardens sooner, a function of its thinness. The rapid setup characteristic of the material allows less time for adjustments while fitting, and some therapists may find it difficult to use. On the other hand, short setup times may be advantageous in the younger, less cooperative patient who wiggles and squirms. Splints fabricated from the thinner

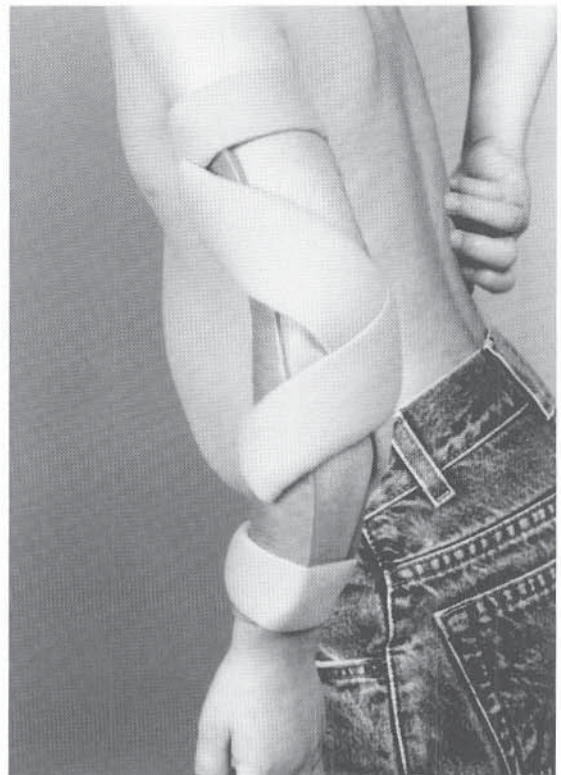


FIGURE 14-15 A static elbow splint fabricated out of Aquaplast. Straps are placed on the splint forming an X across the elbow. This is important to make sure the desired position of the splint is maintained.

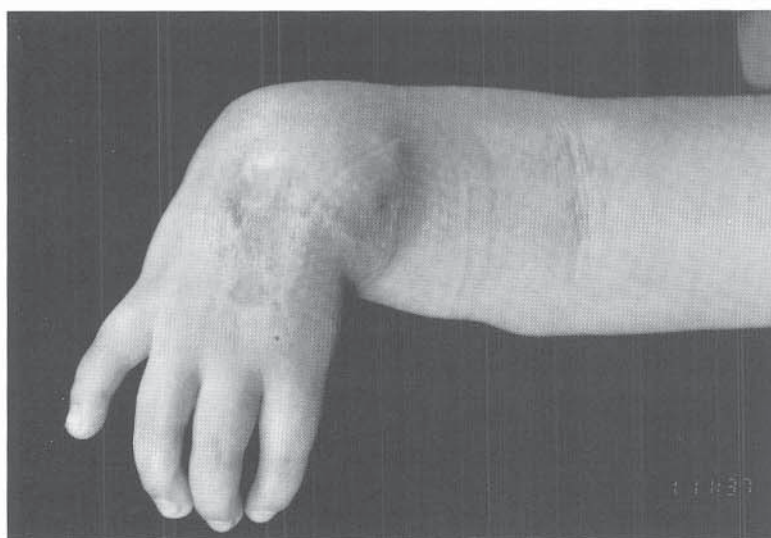
material are lighter, a characteristic that is beneficial in the infant (Fig. 14-13).

Poly Flex II is a thin ($\frac{3}{32}$ -inch) material that has many of the same advantages as EzeForm except that fingerprints may remain on the material, and it can be overstretched. The material is drapable and contours well in the thumb-index web space (Fig. 14-14). Because it has a plastic coating, a solvent must be used to join pieces of it together. The working time is adequate to achieve the correct position of the splint. It is very useful for blocking splints to protect flexor tendon repairs. The lightweight form of the material is useful in infants but becomes malleable especially quickly in the splint water, a feature that makes it easy to oversoften.³

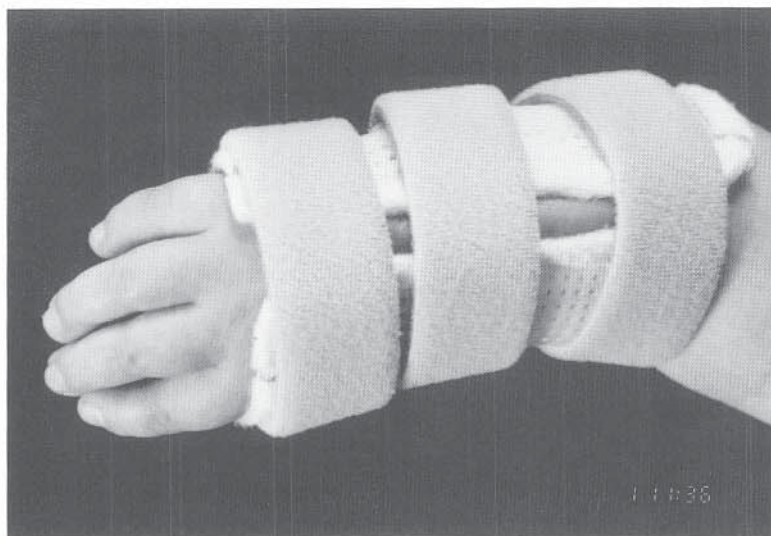
Aquaplast (Noneepanoe, polycaprolactone) is a polymer with moderate resistance that is available in $\frac{3}{16}$ -inch, $\frac{1}{8}$ -inch, and $\frac{1}{16}$ -inch thicknesses and in coated and uncoated versions. It is quite useful in the pediatric patient. The drapability characteristic makes it especially conformable to the precise and variable contour of the upper limb because it clings to

the skin as it hardens. It remains transparent as it hardens so that the extremity can be observed during splinting. This polymer also has a memory characteristic, useful when small alterations in splint shape are needed. It is useful in serial splinting of elbow contractures and when repeated adjustments in fitting are needed, since the material can soften without losing its general shape (Fig. 14-15). Aquaplast comes in several colors, a feature that may improve compliance with splint wearing in children.³

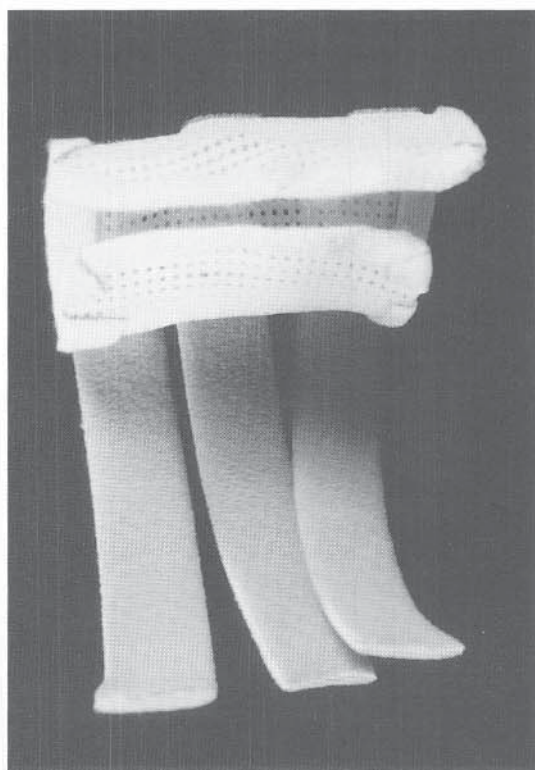
Orfit (polycaprolactone) is a polymer that lacks the plastic coating. This characteristic makes it particularly sticky, and splint fabrication is somewhat more difficult. Fabrication may be eased by adding a small amount of soap to the water in the splint pan. Both perforated and nonperforated forms are available. The particularly good drapability character of the plastic makes it especially useful when precision fit splints are required, such as those used to immobilize the fingers after fracture or tendon injury, or for circumferential splinting (Fig. 14-16).



A



C



B

FIGURE 14-16 A circumferential splint fabricated out of Orfit. A, Child with radial dysplasia. B, Orfit circumferential splint and strap placement. C, Splint placement on the child.

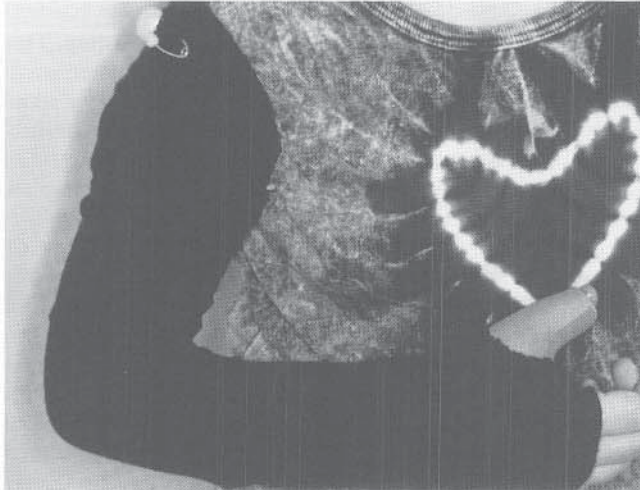


FIGURE 14-17 A sock placed over the splint and secured with a safety pin at the shoulder helps to prevent the child from removing the splint.

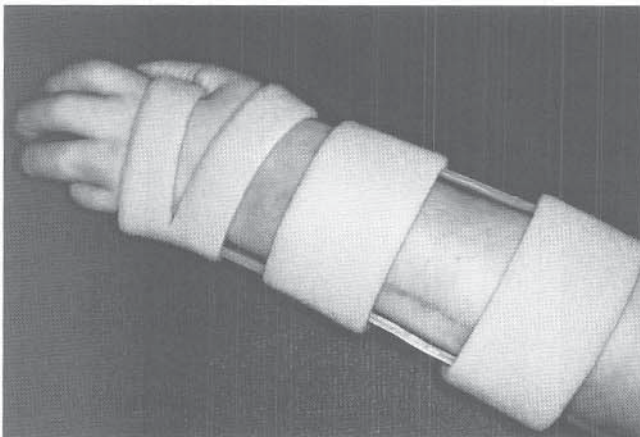
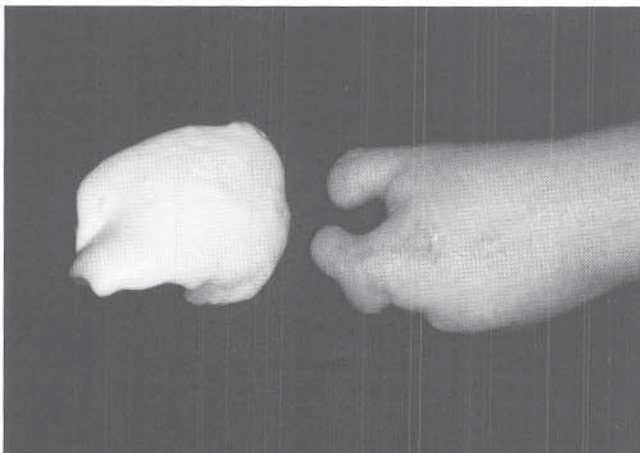
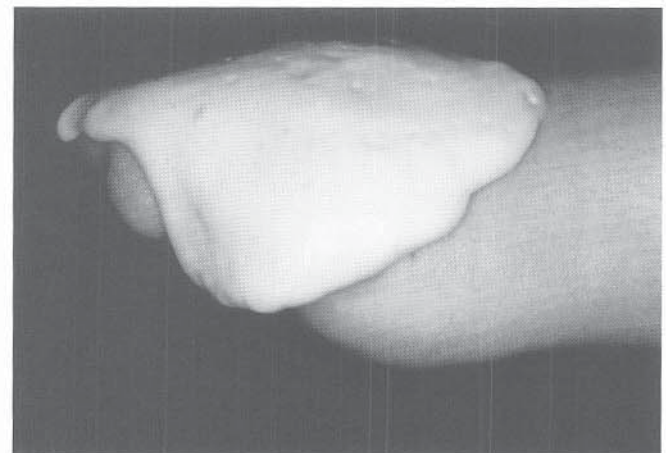


FIGURE 14-18 Splint strap across the wrist. One tail goes around the wrist; the other goes over the dorsal aspect of the hand to keep the hand from migrating out of the splint.



A



B

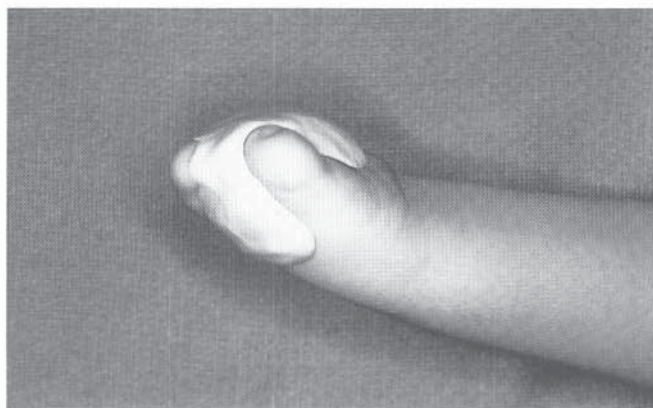
FIGURE 14-19 A, Rolyan silicone elastomer conformer. B, Conformer on the hand.

Securing the Splint. When continual immobilization is required, the splint may be secured by using Coban or adhesive tape strips over the straps at the palm, wrist, and elbow levels to secure the correct splint position. These products need to be applied in a figure-eight fashion, avoiding circumferential pressure. In normal positioning of splints, Velcro straps are used. If the child removes the splint unsupervised, additional measures may be needed to ensure consistent wearing patterns. These measures may include placing a sock over the top of the splint, securing it with a safety pin at the shoulder (Fig. 14-17), or a lace-up method.¹ The wrist strap may be split most of its length, allowing one tail to go around the wrist while the other is placed across the hand and through the web to secure the hand in the splint (Fig. 14-18).

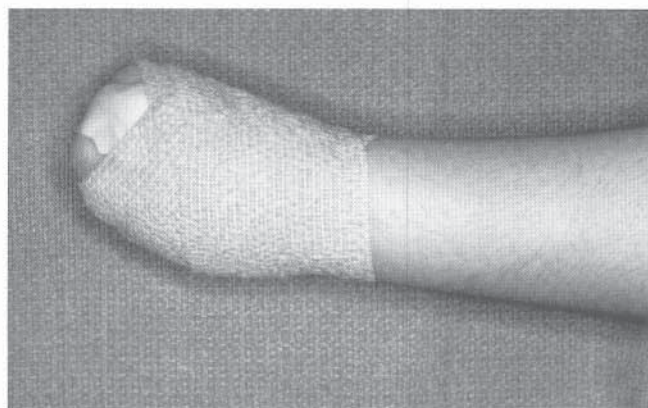
Materials for Fabricating Total Contact Molding Inserts.

Inserts may be used during splint application for compression of scar tissue by the application of firm pressure to the maturing scar. There are many products available, and the choice depends on the condition of the healing tissue. One readily available material is Rolyan Elastomer (tetra-N-alkyl silicate), which is available in a liquid form, allowing application to large areas of scar (Fig. 14-19). Rolyan Elastomer and Otoform (hydroxyl-polydimethylsiloxane with fillers, auxiliaries, not vulcanized) are in a putty form that can be spread thin or formed to fit areas requiring bulk, such as the web areas (Fig. 14-20). These types of scar conformers provide complete contact and must be modified frequently as the scar improves. Because the combination of base and catalyst affects the mixing and setup times, practice prior to use is essential. Silicone or gel sheets are best used when a smooth, flat surface of compression is needed. These sheets are available in a variety of weights (thicknesses) and densities to achieve the desired result as appropriate for the healing tissue. The selection is based on the condition and general sensitivity of the tissue being treated. Scar pads that are not used under a splint require some method of attachment to ensure uniform compression.

The silicone sheet must be cut to cover the entire area of the scar being treated (Fig. 14-21). Cutting the edges near flexion creases in a curved fashion keeps the material from folding over onto itself (Fig. 14-22). Using a compres-



A



B

FIGURE 14-20 A, Otoform K scar conformer in place on the hand. B, Otoform K scar conformer attached with Coban wrap. Coban needs to be wrapped past the wrist joint to secure the conformer onto the hand.

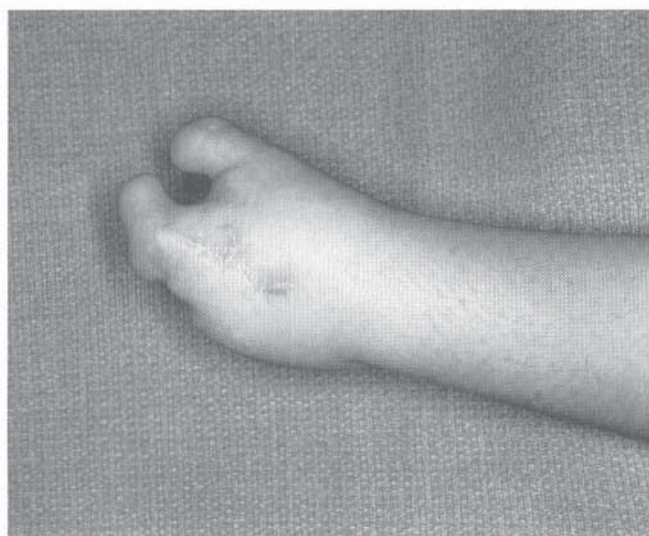


FIGURE 14-21 Dorsal scar extending over the central portion of the hand; the silicone sheet is cut to cover the entire area of scar.

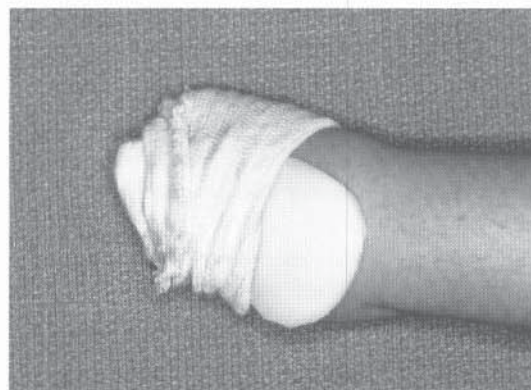


FIGURE 14-23 Maintaining the position of the scar pad while attaching the compressive garment. In this illustration, Tubigrip is used to hold the scar conformer in place.

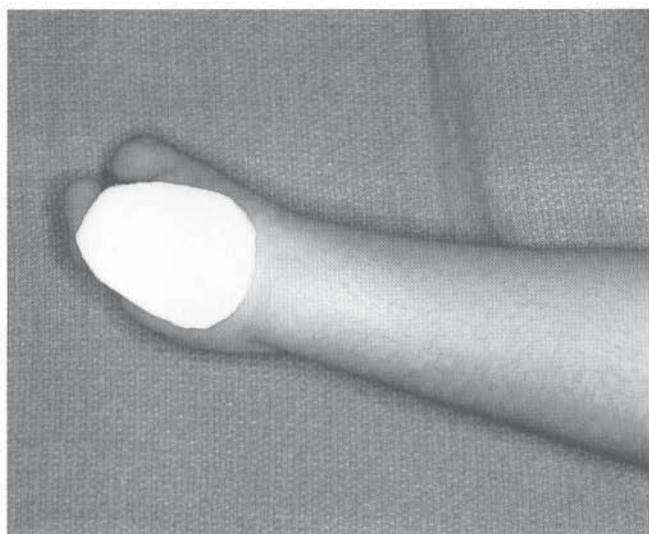


FIGURE 14-22 Placement of Rolyan silicone elastomer sheet.



FIGURE 14-24 Tubigrip extending past the wrist joint to maintain the correct position of the scar pad.

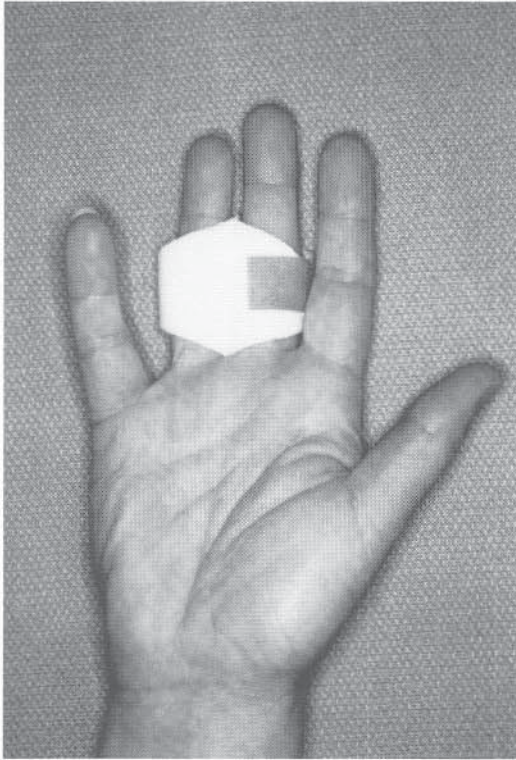


FIGURE 14-25 Buddy strap placed on two adjacent fingers.

sive-type wrap to secure the sheet in place ensures that uniform pressure is applied to the scar (Fig. 14-23). The wrap needs to be secured proximal to the scar pad to ensure that the correct position is maintained (Fig. 14-24).

Coban, Tubigrip, and Medigrip are some effective materi-

als for holding scar conformers in the desired position. The therapist needs to determine the best material to use for each individual patient to ensure the best possible outcome. The attachment of a scar conformer to the finger is a challenge to therapists who treat children, as the fingers are small and it is difficult to keep wraps in place. Thus, the conformer may have to be attached with the use of a splint.

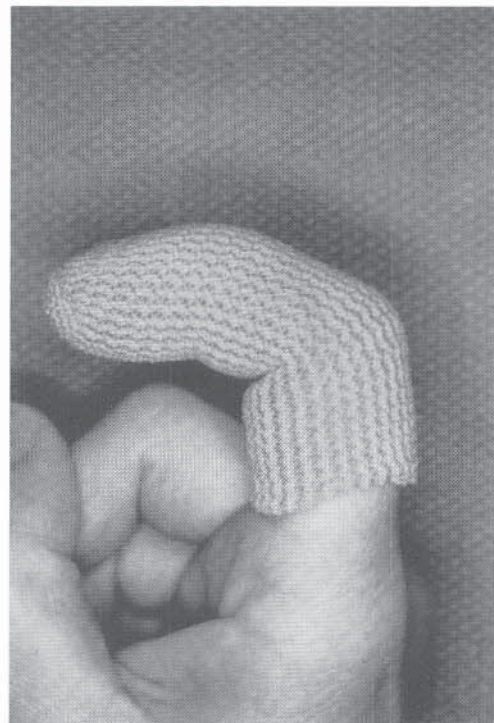
Mechanical Principles. The mechanical principles of splinting are basic principles that need to be understood for effective splinting. The angle of pull needs to be 90 degrees for slings applied to the fingers and for dynamic traction applied to the wrist. Pressure is addressed by using a well-contoured splint that covers the largest surface area possible without affecting uninvolved joints. It is also imperative to remember precautions, including vascular insufficiency, thin tissue, insensate hand, and edema.²

FINGERS. Splinting the fingers of children can be challenging. An injured finger can be effectively splinted by attaching it to an adjacent finger with a buddy strap (Fig. 14-25). The buddy strap also encourages range of motion because the injured finger is attached to a stronger finger. There are a variety of types, although the softer ones tend to be better tolerated by children. Tape or Coban can also be used for this purpose. Care must be taken to ensure that application of the strap or tape does not affect circulation.

Silipos is a unique material very effective for scar management and available in a variety of sizes. This is a stretchy tube lined with a gel pad that is impregnated with mineral oil. The finger tubes come with a closed tip that provide a smooth contour to fingertip amputations. These tubes provide compression and allow full range of motion (Fig. 14-26). They are available in larger sizes for application to the upper extremity.



A



B

FIGURE 14-26 A, Silipos sleeve with finger in full extension. B, Sleeve with finger in flexion.

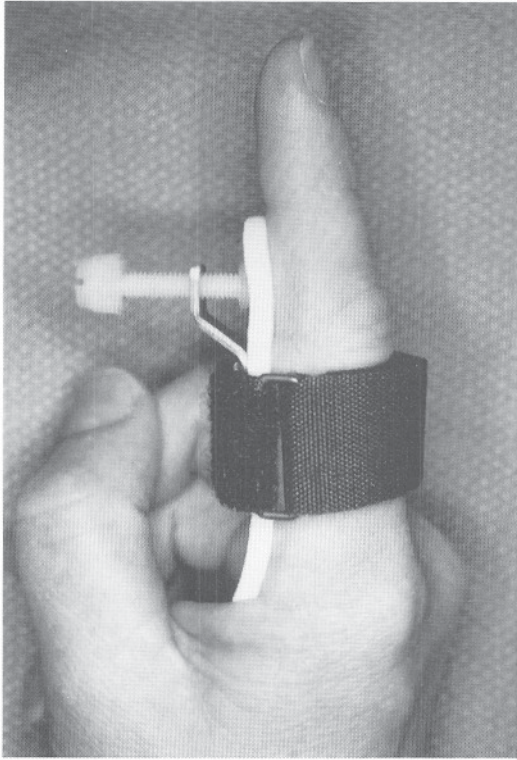


FIGURE 14-27 Adjustable tension spring splint with PIP joint in extension.

An adjustable tension spring is used for *proximal interphalangeal* (PIP) extension (Fig. 14-27). This splint is secured with the tension screw loose, then gradually tightened to increase three-point pressure. The tension spring is better tolerated by children but may not be rigid enough for a larger teenager.

The LMB Finger-Hugger is used for PIP and *distal interphalangeal* (DIP) extension (Fig. 14-28). It comes with two inserts that slip into a pocket. One is more flexible to encourage extension, but allows some motion. The second is a static aluminum insert allowing minimal movement. The dorsal material provides gentle compression, but is comfortable to wear as it better distributes the force across the dorsal aspect of the finger.

Ben's Splint (Fig. 14-29) is a static extension splint used to maintain extension of the PIP and DIP joints. It is constructed of Orfit. The splint can be kept in place with paper tape rather than straps, which may become uncomfortable as they separate the fingers too much. Placing the splint on the dorsal aspect of the finger and careful placement of the tape may allow the pad to be free as a friction surface in hand use. If the dorsal placement allows more motion than the surgeon desires, the splint may need to be fabricated on the volar aspect of the finger.

The LMB extension finger spring is used for PIP extension. It is low-profile and has a light spring force, which allows longer wearing times and greater tolerance (Fig. 14-30). The LMB flexion splint (Fig. 14-30B) may be used in swan neck deformities and extension tightness. Oval-8 ring splints may be used for swan neck deformities, boutonniere deformities, and mallet finger injuries. They are low-profile, which makes them ideal if multiple splints are required on adjacent fingers (Fig. 14-31).



FIGURE 14-28 The LMB Finger-Hugger provides extension of the PIP and DIP joints and has two inserts to vary the amount of tension used to hold the finger in extension.

THUMBS. A Joe Cool splint (Fig. 14-32) is used for increased tone in the thumb adductor muscle. It applies pressure into the index-thumb web space, placing the thumb into a more functional position. Care must be taken to avoid hyperextension of the metacarpophalangeal (MCP) joint during wear.

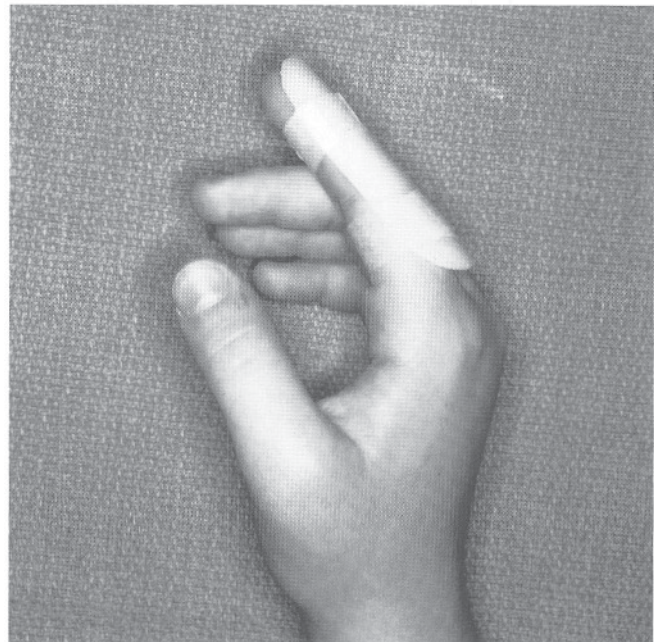
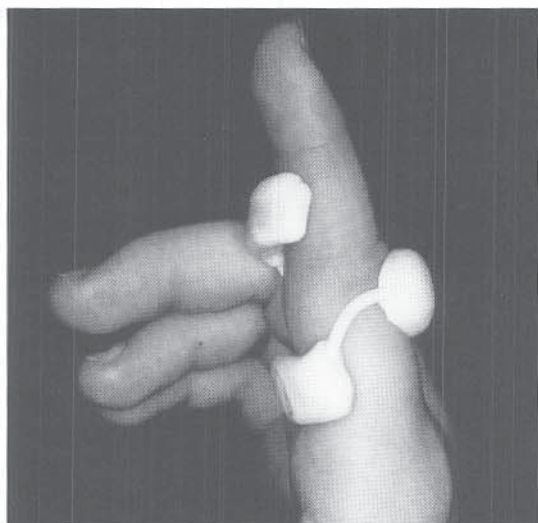
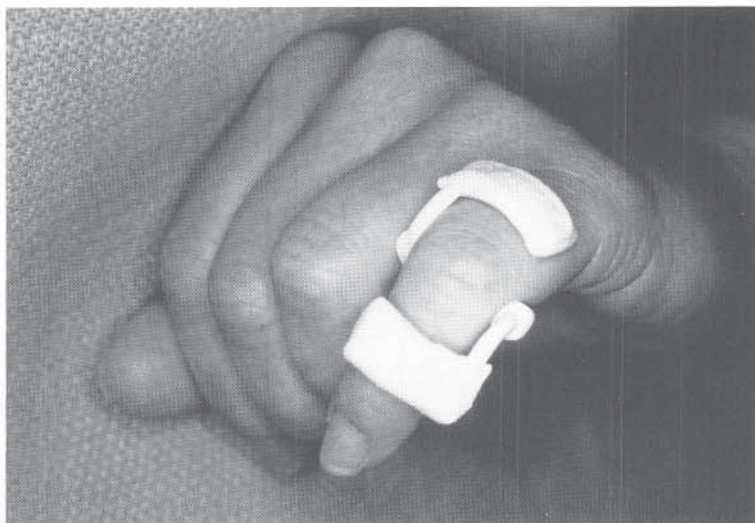


FIGURE 14-29 Ben's Splint, a static extension splint. Here it is held in place with paper tape.



A



B



C

FIGURE 14-30 A, The LMB extension splint. B, The LMB flexion splint. C, This splint allows full finger flexion and blocks hyperextension.

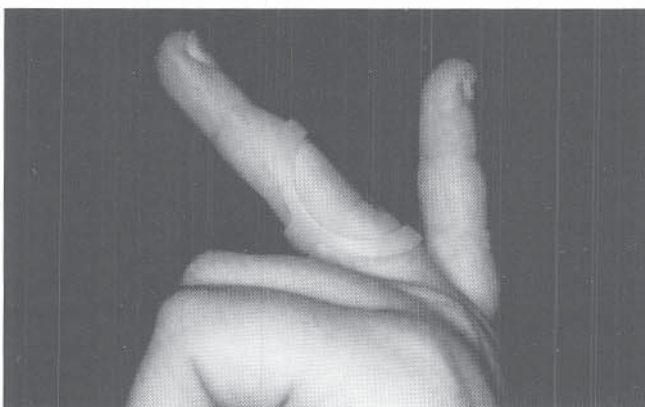


FIGURE 14-31 Oval-8 ring splint. Here this splint is being used for swan neck deformity. It allows full finger flexion. It is very low profile. Also, it is very effective when multiple fingers need to splinted, as the fingers are not overly abducted.

The Benik glove (Fig. 14-33) may be used for moderate tone in the thumb or postoperatively when rigid splinting is not required. Semiflexible splints (Fig. 14-34) may be used for many different diagnoses and may be fabricated or purchased. Support of the thumb postoperatively or following trauma can decrease pain and increase function by providing support to soft tissue during periods of stress to the thumb in the performance of activities of daily living.

WRIST. The most common wrist splints used in pediatrics are the wrist cock-up splint, the extension blocking splint, and the long basic opponens splints. The wrist cock-up splint (Fig. 14-35) is used to block motion at the wrist. Care needs to be taken to ensure that the wrist is not in an ulnar deviated position, especially in children with abnormal tone.

The extension blocking splint (Fig. 14-36) is used for flexor tendon injury or other types of injury to the flexor side of the forearm. Because the splint is placed on the dorsal side of the hand, a drapable material is most desirable.

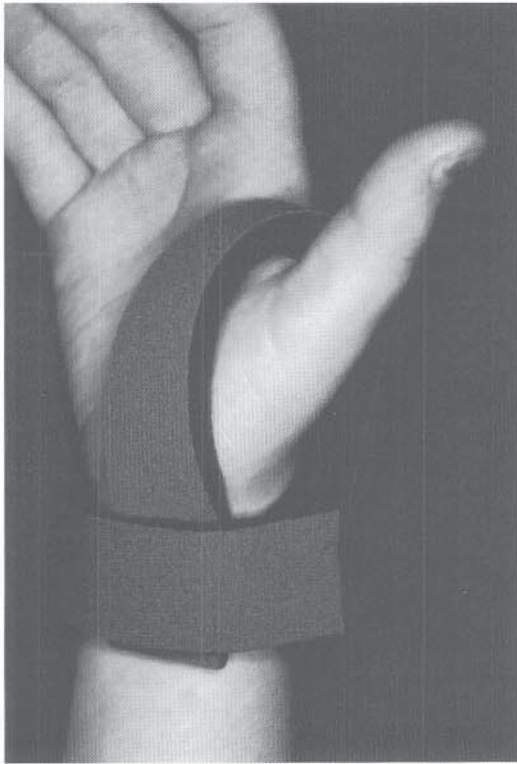


FIGURE 14-32 Joe Cool splint.



FIGURE 14-34 Semiflexible thumb support.



FIGURE 14-33 Benik glove.

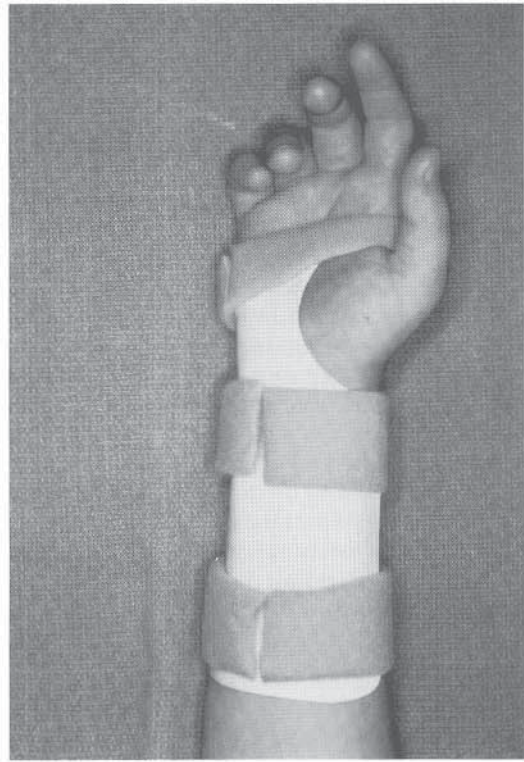


FIGURE 14-35 Thermoplastic wrist cock-up splint.



FIGURE 14-36 Dorsal extension blocking splint, used after tendon flexor repair.



FIGURE 14-37 Long basic opponens splint with a C-bar.

The long basic opponens splint (Fig. 14-37) is used after surgery on the thumb or after trauma. The thumb can sometimes be supported using a hand-based opponens splint.

Two commonly used prefabricated wrist splints are the elastic wrist cock-up splint (Fig. 14-38) and the Benik wrist cock-up splint (Fig. 14-39). The Benik splints are fabricated of neoprene and are available in a variety of bright colors, making them appealing to children.

FOREARM. The Tone and Positioning (TAP) splint is a pronation/supination splint that is effective for lack of range caused by weakness or tone. The forearm strap can be changed if there is a deficit in both directions, rather than two separate splints being required. When pronation is needed for functional tasks such as using the keyboard, the forearm wrap is started over the ulnar border of the forearm (Fig. 14-40).

When a child is carrying objects, such as a lunch tray at school, the hand needs to be in supination. The strap then need only be rewrapped over the radial border of the forearm (Fig. 14-41). Since the neoprene is stretchable, the child can move against the splint. When the child relaxes, the splint returns the forearm to the desired position.

ELBOW. Splinting the elbow is a challenge in pediatrics, as most elbow splints restrict motion. When support is needed for soft tissue, an elbow sleeve (Fig. 14-42) is well tolerated by children. However, when gains need to be made in flexion, a dynamic elbow splint (see Fig. 14-12) may result in better compliance than a heavier hinged splint. Elbow extension is best addressed with a static extension splint worn at night (see Fig. 14-15).

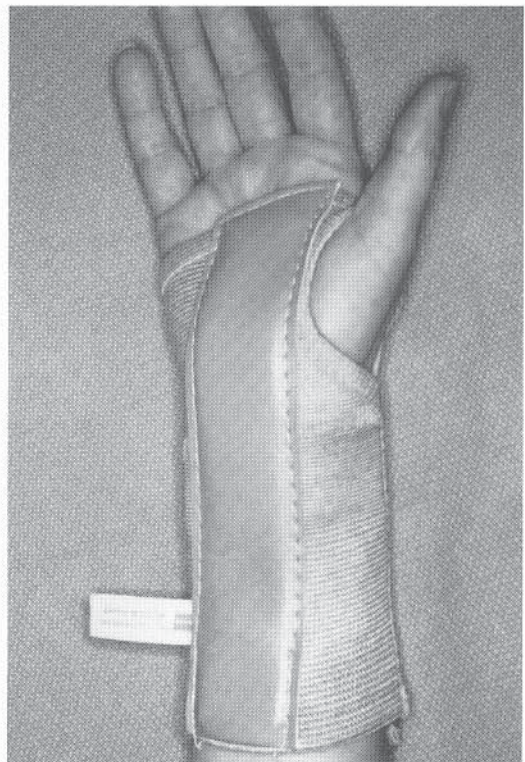
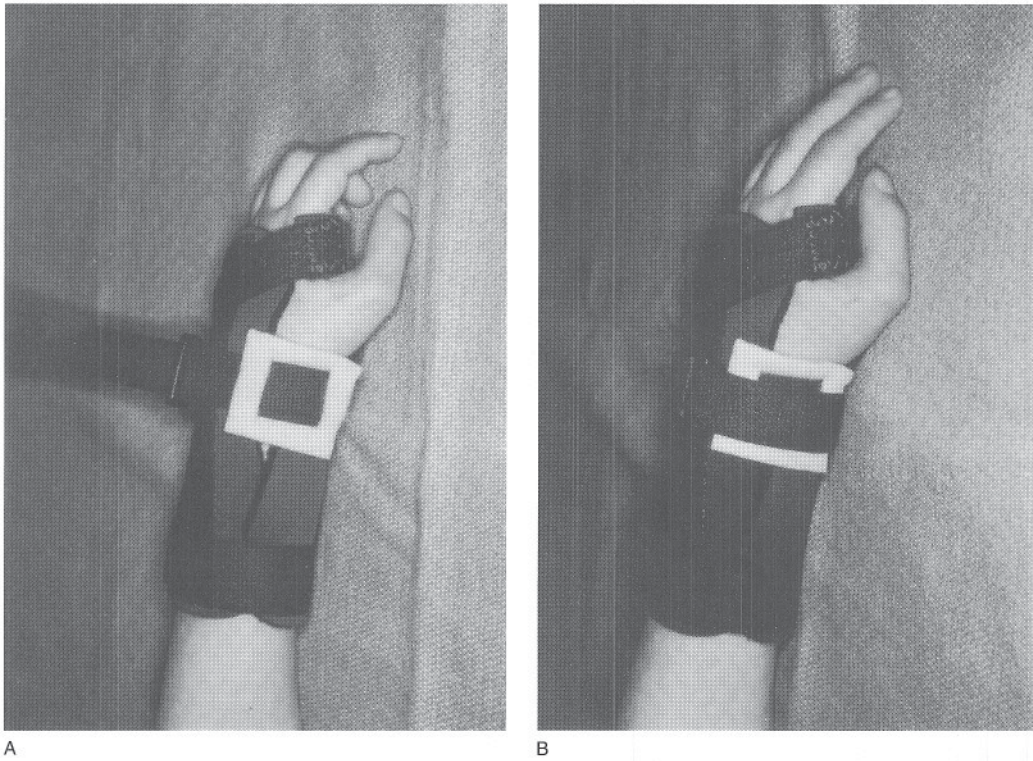


FIGURE 14-38 Elastic wrist support.



A

B

FIGURE 14-39 Benik wrist cock-up splint. A, A piece of strapping material is slit into two and slid over the strap to pad the wrist, protecting the superficial branch of the radial nerve so that the strap does not rub against it. B, Strap securely fastened over the padding.



FIGURE 14-40 The Tone and Positioning (TAP) splint pulling the forearm into pronation. The strap is attached in the palm and is placed over the ulnar aspect of the wrist. Then it is rotated up the forearm, and the Velcro closure is secured above the elbow joint.

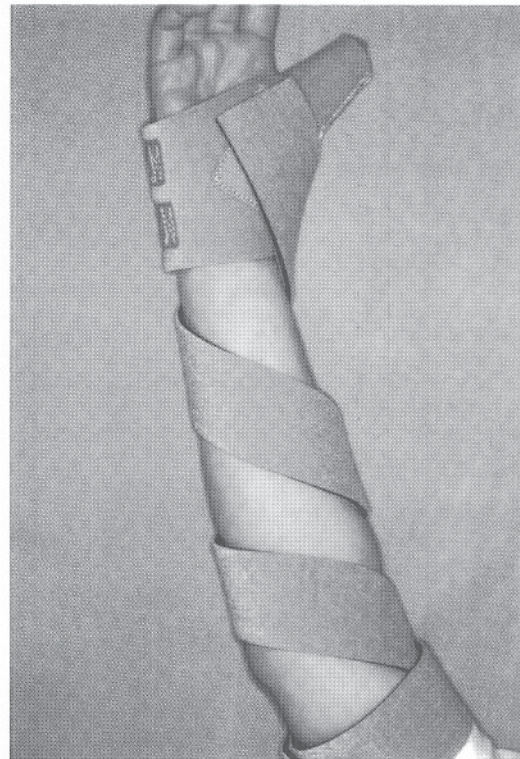


FIGURE 14-41 The Tone and Positioning (TAP) splint pulling the forearm into supination. The strap is attached in the palm and is placed over the radial aspect of the wrist. Then it is rotated up the forearm, and the Velcro closure is secured above the elbow joint.

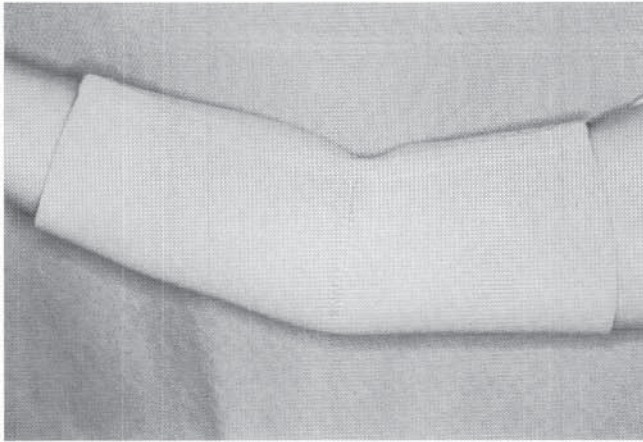


FIGURE 14-42 Neoprene sleeve extending over the central portion of the hand.

Splinting in pediatrics varies from the “normal” methods used in adults. Children are often casted longer postoperatively, as they are unable to cooperate in the acute period of early mobilization programs. These programs are not needed with most children, as they heal quickly without the types of adhesions adults may need to battle.

Use of Distraction Techniques. Children respond well to distraction. A little planning goes a long way in pediatrics. Having toys children can manipulate with one hand is helpful during the splinting process. Pushbutton musical toys and battery-powered toys that spin, light up, or make noise provide the desired distraction. The surgeon should remember that the process of cast removal may be a scary experience for the child and the reaction to a splint may be less than desired.

REFERENCES

Principles of Dressing and Splinting

1. Callahan A, Cannon NM, Mullins PT, et al: *Manual on Management of Specific Hand Problems*, ed. Malick M and Kasch M. Pittsburg, American Rehabilitation Educational Network, 1984.
2. Cannon N, Foltz RW, Koepfer JM, et al: *Manual of Hand Splinting*. New York, Churchill Livingstone, 1985.
3. Fess EE, Philips CA, Strickland JW, et al: *Hand Splinting*, 2nd ed. St. Louis, CV Mosby Co, 1987.
4. Hogan L, Uditsky T: *Pediatric Splinting: Selection, Fabrication, and Clinical Application of Upper Extremity Splints*. San Antonio, Therapy Skill Builders, 1998.

Principles of Acute Care

GENERAL PRINCIPLES

It has been said that the first surgeon who sees the injured hand most affects the final result. This is especially true in the child. Accurate early diagnosis is more difficult in the child than in the adult, since patient cooperation may be minimal or absent. The chance of missing significant injury is reduced when the surgeon and parent realize that a complete and accurate diagnosis may often require seeing the child more than just in the emergency room. It is perfectly acceptable and in fact often crucial to insist that a second

follow-up examination be done a day or so later in the clinic or doctor’s office, when less blood and less hysteria make possible a more complete and reliable examination. A second examination is particularly useful in cases of possible nerve injury, for a sensory examination can be done more reliably in the quiet of the doctor’s office than in a hectic emergency room. Occasionally even this is not sufficient, and when the surgeon cannot rule out a nerve or tendon injury suspected because of the location of the wound, the wound may need to be explored under general anesthesia.

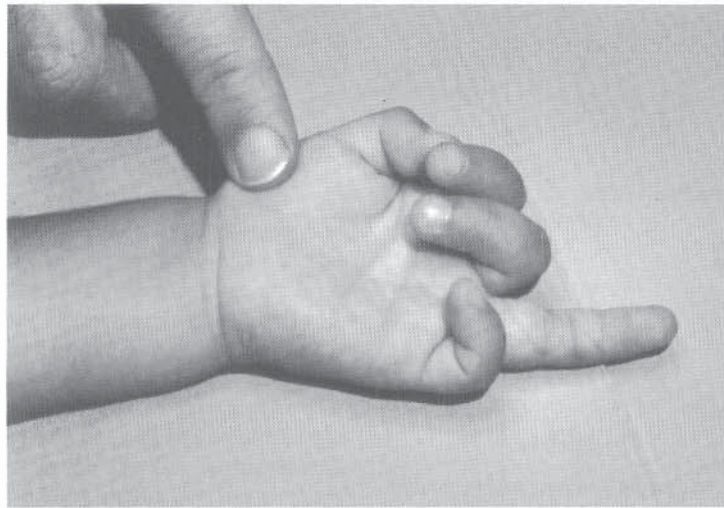
It is important to realize that no hand injury is a life-threatening emergency, and treatment should always be delayed until the anesthetic risk is minimized by a period of fasting and until a well-rested operating team with appropriate light, instruments, and training is available. Simple closure of the skin and delay is the treatment of choice until all of these elements are in place. This is especially important in children with hand injuries, since children eat constantly and frequently are difficult to anesthetize. The stakes are high and patience is a virtue.

As in other areas, the diagnosis is based on the history of injury, observation, physical examination results, and radiographs. Only very rarely are more sophisticated examinations necessary. Careful and thoughtful observation of the injured hand by the surgeon is by far the most important factor in evaluating an uncooperative child. This observation must be made in the context of what has been called “topographical anticipation.” The surgeon’s knowledge of the topographical anatomy of the injured site is used to anticipate the physical findings associated with likely injuries. By first asking the question, “What structures are at risk?” the surgeon is much more likely to notice the critical finding associated with the injury. In most cases, this assessment is best done by critically observing the rest position and use of the hand by the child. More information is gained by simply observing the child’s hand use from across the room than by wrestling the child from the mother’s arms. The lacerated flexor tendon is the prototypical example of the usefulness of topographical anticipation.

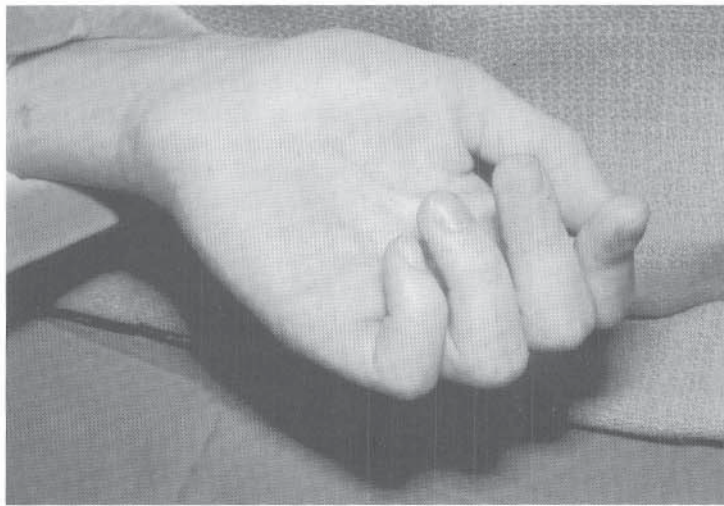
TENDON INJURY

Alteration of the hand’s resting position in a child with a tendon laceration may be obvious or subtle (Fig. 14-43). Furthermore, when wound exploration reveals a tendon sheath laceration, it is very likely that the tendon itself has sustained injury. This is especially true if the finger was flexed at injury. By necessity, the wound is inspected with the finger in extension. The site of tendon laceration then moves up the finger and is hidden from view (Fig. 14-44). Subsequently, during the first 5 to 7 days following injury, the partially injured tendon is especially vulnerable to failure. If a partial injury is suspected, the hand is splinted in a posture to protect the tendon from rupture. Some surgeons prefer to explore such cases in the operating room under general anesthesia so that accurate assessment of the extent of the partial laceration and appropriate repair can be accomplished.

Surgical exploration and the technique of tendon repair are, except for the size of the structures involved, identical to exploration and repair in adults. This topic has been covered thoroughly in other hand surgical texts and will not



A



B

FIGURE 14-43 A, In the small child, the resting stance of the digit may be the best diagnostic sign of tendon injury. The only possible cause of this appearance is disruption of the profundus and sublimus in the child's ring finger. B, Sometimes the diagnosis is more subtle. The normal finger posture of the hand at rest displays a cascade of finger flexion. All fingers are gently flexed at all joints, with the flexion increasing from index finger to small finger. In this child, only the profundus has been lacerated. However, it has been lacerated in both the index finger *and* long finger, and whereas the index finger is hyperextended, the DIP joint of the long finger is completely straight. Careful observation is fundamental to a good hand examination.

be discussed here. Magnification, appropriate instrument size, and careful technique usually can resolve any differences in the technical aspects of tendon repair. The more critical challenges in children are the immediate postoperative care to prevent rupture, and, later, rehabilitation to develop gliding of the tendon repair.

Postoperative immobilization should be complete and uninterrupted in the small child. In very young children and babies, the best treatment regimen usually consists of an extra week or two of immobilization to try to prevent rupture. Excessive immobilization of more than 4 weeks for flexor tendons or 6 weeks for extensor tendons is not warranted and even in small children can lead to stiffness. On the other hand, changing the casts early just to "have a look" may reward the surgeon with rupture and should not

be done unless infection or swelling leading to compromise is suspected.

The type of rehabilitation in very young children is limited but becomes more like that in adults, as the cooperation level of the child improves with age as the child starts elementary school. In adolescence, compliance may sometimes be poor, but this is less of a problem if the teenager can be made to feel responsible and involved in his or her own treatment program.

When a primary repair or tendon grafting fails, we rarely repeat the attempt in a young, uncooperative child. If tendon reconstruction is unsuccessful in the preschool child, repeated surgery should wait until the child's hand is larger and the child is better able to understand and cooperate with the treatment program. This is not to say that good

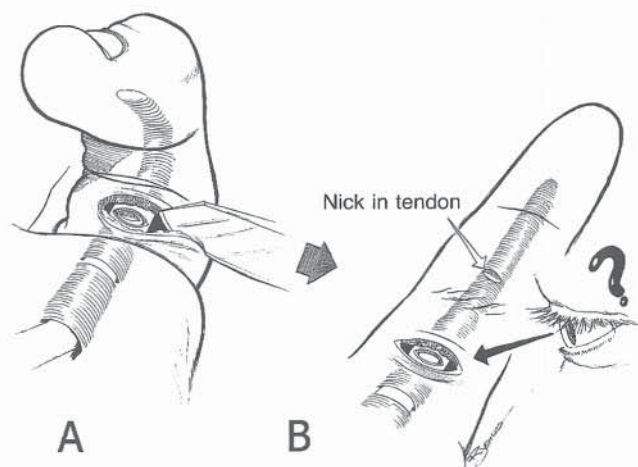


FIGURE 14-44 A and B, Beware of the partial laceration occurring with the finger in flexion or grasping a sharp object. When the finger is extended for examination of the wound, the tendon appears intact, and only a laceration in the tendon sheath is seen. The partial laceration of the tendon is hidden within the tunnel of the flexor sheath. The tendon may rupture in 4 to 5 days if it is not protected. (From Carter P: *Common Hand Injuries and Infections*, p 147. Philadelphia, WB Saunders Co, 1983.)

results cannot be obtained after tendon reconstruction in the preschool child. In fact, a review of our results suggests that the quality and frequency of an acceptable result are similar to that in adults, and therefore one early attempt at repair is justified.

NERVE INJURY

The diagnosis of a nerve injury is usually subtle and also depends on topographical anticipation. The surgeon is well served to expect the worst and not just hope for the best. We have seen a toddler with a minor wound over the posteromedial aspect of the elbow present weeks later with the first recognized sign of nerve injury being an anesthetic fingertip that was bitten off by the child. In addition to being very superficial in the hand, wrist, and elbow, the peripheral nerves are usually accompanied by an artery. When there is a good history of arterial bleeding after a wound, one should carefully examine the patient for a nerve injury. Children with suspected nerve injuries, more than almost any patient, deserve a follow-up assessment in the relative quiet of the clinic or office setting. At this time bleeding is absent, the child is not in pain, and even the parent is often more helpful. Much grief will be avoided if the parent is made aware of this important clinical tool, the "follow-up in the office" examination, since the diagnosis is easily missed the day of injury.

Absence of a sweat pattern of the anesthetic finger is a useful finding, and recognition of this finding is more often possible during the "second examination." Absence of a sweat pattern is best identified by the dry or slick quality of the affected finger when the surgeon gently strokes the injured digit and uses a normal digit for comparison. Loss of intrinsic function may also be a subtle finding (especially in the median nerve) and is best noticed by comparing the resting position of the normal hand and thumb with the injured one. Again, topographical anticipation allows the subtle findings of nerve injury to be noticed. Finally,

little is lost by a competent hand surgeon's taking the child to the operating room and exploring the wound when the diagnosis is sufficiently suspected.

SKIN INJURY

Fingertip injuries in children are very common. As a general rule, the less done for these injuries, the better. Even some exposed bone can heal with just dressing changes if there is no exposed tendon. More extensive wounds often require full- or split-thickness grafts and local rotation flaps. Pedicle grafting in children is rarely indicated in the acute treatment of all but the most catastrophic hand wounds.

Burns are treated as in adults, with the surgeon remembering that the initial assessment frequently underestimates the severity of children's burns. Child abuse may manifest as burns; especially suspicious are cigarette burns and bilateral hand burns.

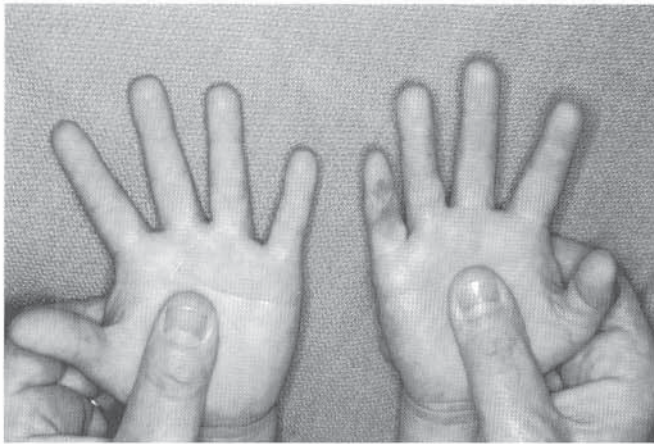
Characteristically, children tend to burn their hands more often in the palms than adults, because they lack understanding of what is too hot to hold. Severe flexion contractures are frequent following these injuries. Fortunately, the palmar skin is thicker, and the deeper and important structures such as the nerves, tendons, and tendon sheath are usually unharmed. Even when the finger seems hopelessly contracted following these palmar burns, reconstruction can frequently yield a marked improvement in function. A careful release of the anterior cicatrix, which protects the tendon sheath, tendons, and nerves, is followed by precision full-thickness skin grafting. First, a transverse incision accurately joining the axis of rotation of the joints is made. This incision must carefully avoid the tendon sheath and nerves. With gentle dissection, the contracted cicatrix is removed, and as the finger extends, the linear incision is converted into diamond-shaped defects. The resulting defects are covered with carefully tailored, full-thickness skin grafts whose margins conform to the rules of hand surgery incision (Fig. 14-45). Frequently this approach can restore near complete movement of the finger and may greatly improve the function of the hand.

BONE INJURY

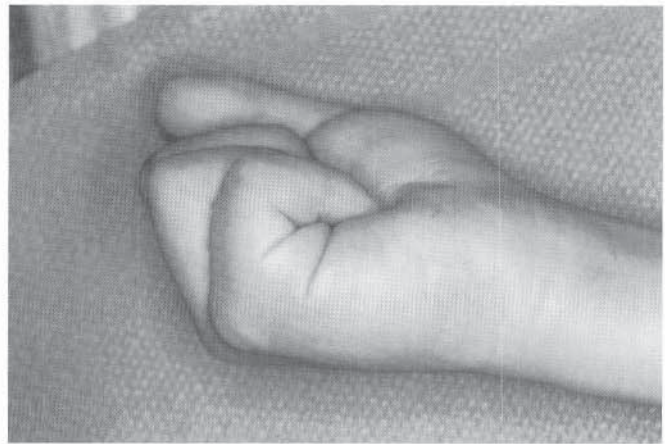
The great advantage of radiographs in diagnosing adult fractures is limited in children by the cartilaginous nature of much of the young child's bone. The value of simultaneous comparison views of the injured side and the normal side cannot be overestimated, especially when the structures are placed side by side on the same radiographic plate. This simple and inexpensive detail can usually provide more information than expensive CT or MRI studies.

A true lateral radiograph of the injured digit is important and may reveal a subtle but often significant injury. The technician should use the fingernail as a topographical landmark to obtain a true lateral view (Fig. 14-46).

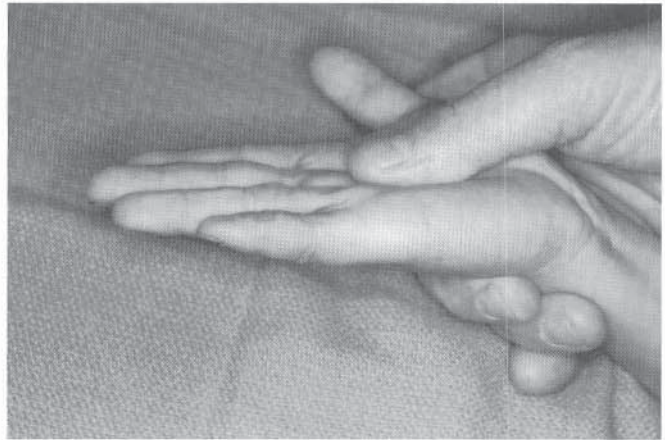
In general, most fractures in the child's hand are treated nonoperatively, since children's hands have a remarkable ability to recover useful movement by remodeling fractures that would clearly need open reduction in an adult. Minimal displacements and malalignments heal better with closed treatment. Simply protecting the hand from use for 3 or 4



A



B



C

FIGURE 14-45 A to C, Burn scar showing release and diamond-shaped skin grafting. To release the contracted finger, a transverse incision was made in the longitudinal scar and a diamond-shaped full-thickness graft was inserted into the defect so created. Preoperatively the finger contracture was so severe that the proximal and distal edges of the diamond-shaped defects had coapted. The surgeon must remember that the edges of a skin graft act like an incision after healing and must be constructed to follow the rules of incisions (see Fig. 14-1).

weeks is usually adequate. Reduction of rotational malalignment is critical prior to immobilization.

Serious rotational malalignment, markedly displaced intra-articular fractures, and some displaced epiphyseal fractures may require open reduction and internal fixation, and these procedures are conducted following the same guidelines as are used in other long bones.

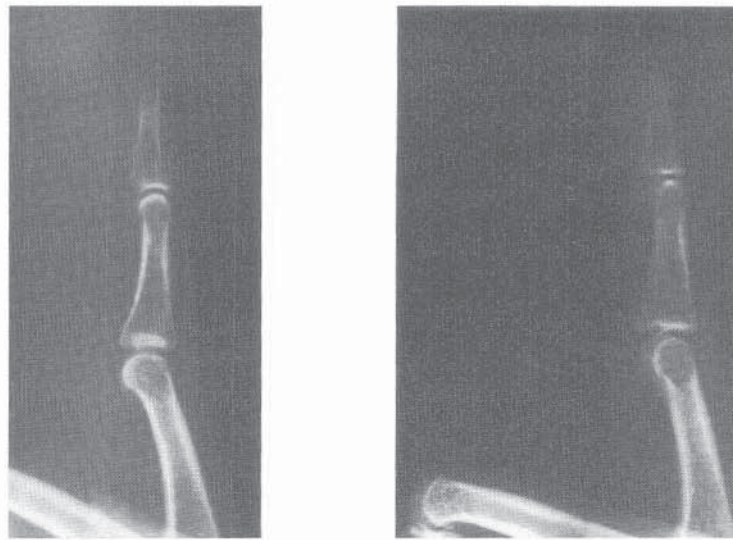
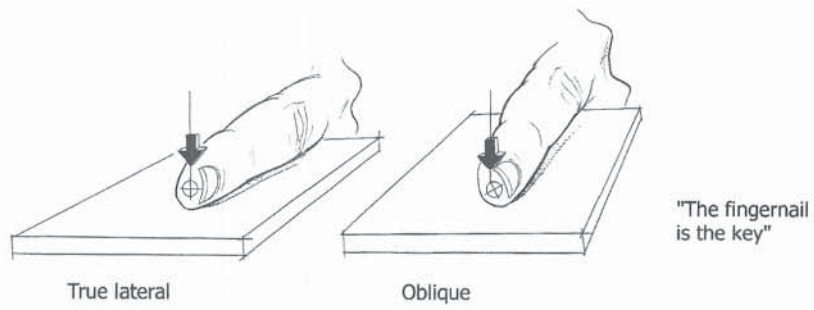
When the fracture is unstable, we prefer to stabilize fractures in the hand with smooth K-wires augmented with protective plaster or fiberglass dressing with the joints of the hand and wrist in the resting position. It is especially critical in children to leave the wires outside the skin and covered by the rigid plaster dressing. They are easily removed later in the clinic, when bone union is adequate. Open reduction of hand fractures with more sophisticated internal fixation devices in order to allow early motion is almost never justified in the child who cannot be trusted to protect the fracture adequately during healing.

Somewhat later, once healing has begun and the fracture site is nontender or callus is seen on the x-ray, the wise surgeon will often delay operative treatment in order to avoid excessive cicatrix, loss of tendon gliding, or avascular fragments near joints. The wisdom of this approach is especially evident for subacute injuries. Open reduction of displaced fractures is especially difficult in the small bones of the hand when substantial fracture healing has taken place. Vigorous dissection around these small bones can occasion-

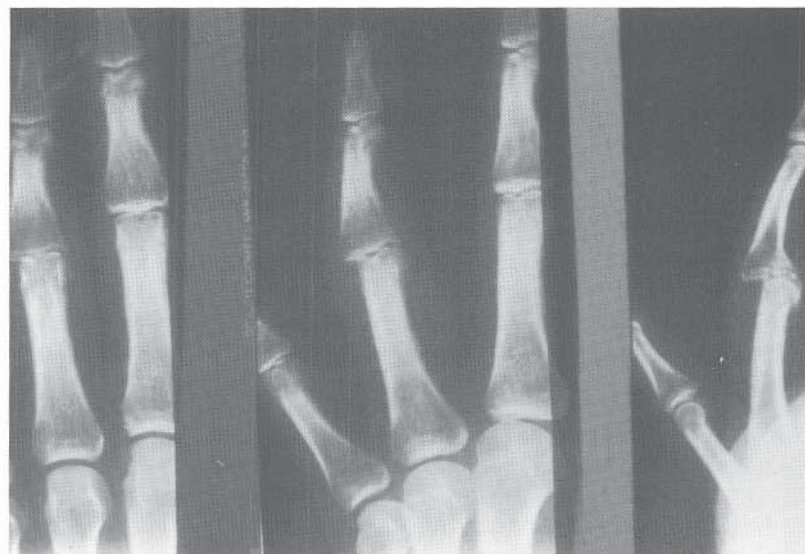
ally result in devascularization and should be avoided. Subacute treatment of fractures of the small bones in the hand can lead to some of the most disastrous complications in hand surgery. The care of these complications may go on for months, only to yield a final result that is most unsatisfactory to the surgeon, patient, and parent (Fig. 14-47). It is often best to let fracture remodeling complete itself and then to address the finger from a clinical rather than just a radiographic aspect. Later, after remodeling has taken place, an osteotomy can be done more safely in an area of the bone that is more likely to heal quickly. Even then, if the finger looks straight, has a functional range of motion, and is painless, aggressive treatment of the radiographic finding is rarely justified. The surgeon must remember that precise control of the small fragments of phalangeal osteotomies in young hands is usually not possible and that small malalignments are difficult to improve even by the most experienced surgeon.

VASCULAR INJURY

Vascular trauma in the upper limb results from direct vascular injury leading to a nonviable distal extremity or a compartment syndrome caused by pressure in an unyielding fascial compartment. Complete loss of distal circulation should be addressed by a vascular surgeon and will not be discussed here. The repair of smaller distal vessels in a



A True lateral Oblique



B

FIGURE 14-46 A, Subtle fractures of the digits can be missed when a true lateral radiograph is not obtained. A true lateral view of the bone is obtained when the true lateral of the fingernail is used as a topographical landmark. (From Carter P: *Common Hand Injuries and Infections*, p 76. Philadelphia, WB Saunders Co, 1983.) B, The resulting radiograph shows a fracture dislocation not seen on the anterior or oblique views.



FIGURE 14-47 Subacute fracture dislocations of the small joints of the hand require considerable clinical judgment on the part of the surgeon. This fracture, although not completely healed, was nontender 3 weeks after injury. Open reduction and the attendant stripping of bone required for repositioning this small articular fragment resulted in complete AVN of the fragment. Subsequently the joint had to be arthrodesed to stop the pain and correct an unstable malaligned finger.

nonviable hand or digits is discussed at the end of this chapter under Microsurgery.

Forearm compartment syndrome has been the dread of the pediatric orthopaedic surgeon since Volkman first described the condition in 1881.³ The clinical setting for compartment syndrome is rarely that of massive open trauma, since in these cases the fascial covering is disrupted enough to prevent a secondary muscle ischemic death due to swelling. Instead, the condition follows closed injury; is all too often unexpected; and, despite aggressive early fasciotomy, may not be preventable.¹

The classic clinical signs of pain, pallor, pulselessness, and paresthesia are important but not reliable. Loss of the pulse may never occur. The surgeon should strongly suspect a compartment syndrome when the child's complaints of pain seem out of the ordinary in a setting where swelling of the forearm is expected. Although a supracondylar fracture of the humerus is the classic association, both-bone forearm fractures, blunt trauma, and even extravasation of blood or fluids may be the inciting cause. It is most useful clinically to compare the size and induration or firmness of the affected extremity with the normal one. Pain on passive digital extension that is referred to the proximal forearm is especially suggestive. Eaton has quantified this by measuring the angle of wrist extension or flexion at which pain is referred (R. G. Eaton, personal communication) (see Fig. 14-54B). By holding the digits in full extension at the MCP and IP joints, this can be accurately assessed and recorded.

Although compartment pressure measurements have been popular in the literature, in our experience they are time-consuming and occasionally falsely negative. If the surgeon is concerned enough to measure the pressure, a fasciotomy rarely complicates the treatment and may save the function of the extremity.

The incision used for fasciotomy should not only allow access to the entire compartment but should also anticipate the possible need for tendon transfer in later reconstruction

(Fig. 14-48). Undermining of skin flaps should be kept to a minimum in the swollen extremity, but a complete release of forearm fascia is required from above the lacertus fibrosis to (and if necessary including) the carpal tunnel.

Rare cases of newborns with compartment syndrome have been reported.² The cause is controversial, but we have occasionally seen late muscle contracture quite similar to what is seen in more typical cases later in life, a fact that

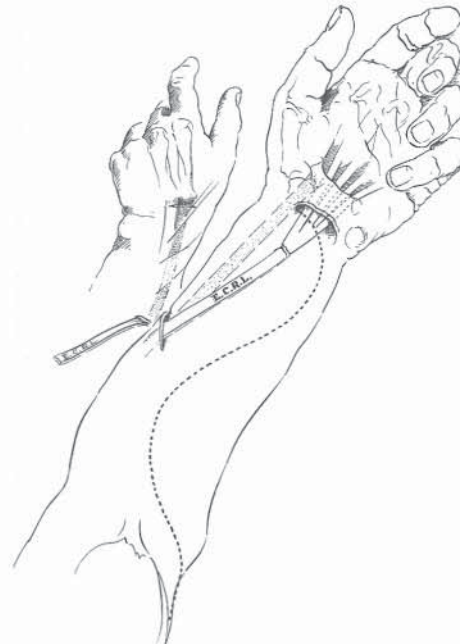


FIGURE 14-48 The proper incision for a forearm fasciotomy should take into account the future reconstruction needs if the forearm muscle cannot be saved. (From Converse JG: *Reconstructive plastic surgery*. In Littler JW (ed): *The Hand and Upper Extremity*, 2nd ed, vol 6, p 3136. Philadelphia, WB Saunders Co, 1977.)

suggests that early fasciotomy of these infants would be appropriate.

Compartment syndrome in the hand is less common but occurs most often in conditions with serious soft tissue trauma associated with intact skin. Recognition depends on suspicion on the part of the orthopaedic surgeon and evidence of the intrinsic plus position (flexion of the MCP joints in conjunction with extension of the IP joints). Release of the fascia of the interosseous muscles is best accomplished with three small longitudinal incisions placed over the dorsum of the hand.

Reconstruction of established Volkmann's contracture is discussed in the following section under Principles of Reconstruction—Established Volkmann's Contracture of the Forearm.

REFERENCES

Principles of Acute Care

1. Converse J: Reconstructive plastic surgery. In Littler JW (ed): *The Hand and Upper Extremity*, vol 6, p 3103. Philadelphia, WB Saunders Co, 1977.
2. Tsujino A, Hooper G: Neonatal compression ischaemia of the forearm. *J Hand Surg* 1997;22-B:612.
3. Volkmann RV: Die ischaemischen Muskellahmungen und Kontrakturen. *Zentralbl Chir* 1881;8:801.

Principles of Reconstruction

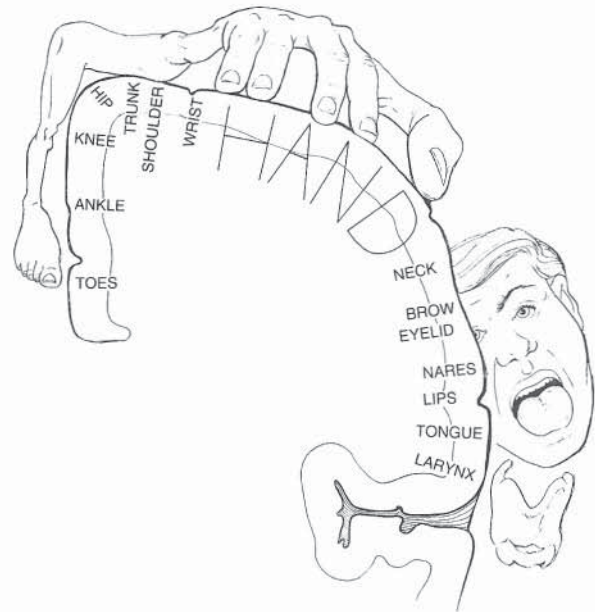
ARCHITECTURE AND HAND PHYSIOLOGY

Successful reconstruction of the child's hand relies on the well-established principles of hand surgery developed for adults, with some special considerations for children. Effective reconstruction is based on understanding the architecture and physiology of the hand as an organ of motion, strength, and sensibility. A short discussion of mechanics and function of this unique element of the musculoskeletal system is appropriate, since any surgeon's reconstructive plan must, as much as possible, restore these elements: *sensibility, motion, and strength*.

NERVE FUNCTION: SENSIBILITY AND POWER

The primary consideration in any hand reconstructive procedure is the ability of the hand to feel. Wilder Penfield's homunculus illustrates the great importance given to the hand by the sensory cortex of the brain (Fig. 14-49). Although operations designed to improve the appearance of the crippled upper limb are an important part of pediatric upper limb reconstruction, the child's ability to feel with and protect the hand is essential to improving hand use. The primary place of sensibility in all treatment for children with hand deformity or injury cannot be overemphasized. It is the fundamental reason for failure of long-term use of upper limb prostheses (Fig. 14-50).

Fortunately for the pediatric hand surgeon, the results of all peripheral nerve reconstructions are considerably better in children than in adults. The better results are due more to the central reintegration of the defective signal from the periphery and less to faster or better growth of the axons or the shorter distances of axonal growth needed in a child's



"Homunculus"
W. Penfield, M.D.

FIGURE 14-49 Wilder Penfield's homunculus, indicating the amount of the sensory portion of cerebral cortex allocated to the hand. The consequence of a lacerated median nerve is obvious when one considers the cerebral cortex watershed of this small structure. (From Carter P: *Common Hand Injuries and Infections*, p 31. Philadelphia, WB Saunders Co, 1983.)

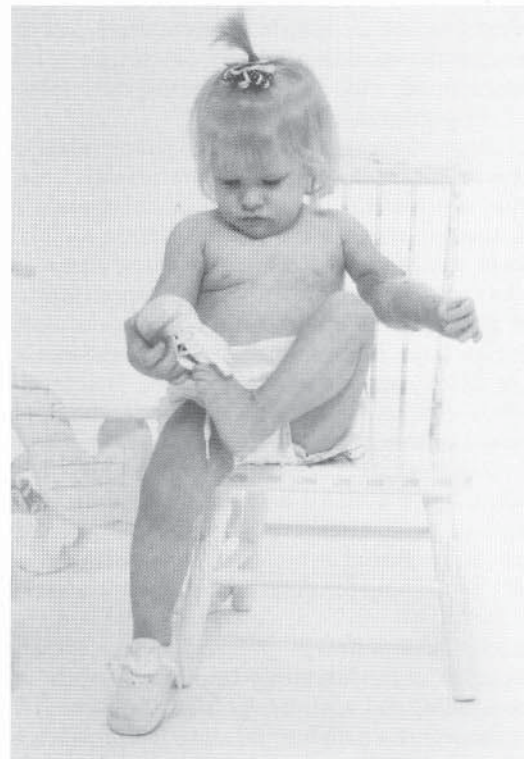


FIGURE 14-50 An illustration of the importance of sensibility. This child avoids using the well-made and handlike-appearing prosthesis because it covers a sensate, below-elbow stump that she would otherwise be using to help put on her shoe. The upper limb prosthesis can only be expected to function as a tool rather than as a hand.

shorter limb. However, the nerve injury must be recognized, repaired, and held in contact long enough for the child to realize these advantages. In addition, the aftermath of nerve injury is not always more favorable in children than in adults. Particularly in the very young and rapidly growing child, nerve injuries can affect the length of growing bones as well as the shape of joints. The shortened extremity in the polio patient or the shortened, denervated digit illustrates the first, and the deformed shoulder joint of an incompletely recovered Erb's palsy patient illustrates the second.

ARCHITECTURE OF THE HAND

Reconstruction of the complex function of the hand can be facilitated by dividing its mechanics into two components—a fixed portion surrounded by a collection of mobile units.

The *fixed component* is made up of the index and long metacarpals and the distal row of carpal bones. These six rigid bones form two arches at right angles to one another. The transverse arch formed by the distal carpal row is a Roman or semicircular arch. At a right angle to this arch, in the sagittal plane, the metacarpals and carpus form another arch whose shape is a common cycloid. The fixed unit of the hand is the foundation of all hand motion and power. For practical purposes, there is no movement between these bony elements. The stability and alignment of this combined

“foundation” are controlled in space by the strong flexor and extensor muscles of the wrist (Fig. 14–51).

The *mobile components* include three groups of movable parts. The first part is the highly mobile thumb ray radiating off the radial volar side of the fixed unit. Its complex conical motion and great strength are the result of a collection of the three joints empowered by the intrinsic and extrinsic muscles of the thumb (Fig. 14–52). The second mobile component consists of the two minimally mobile ulnar metacarpals, which, in conjunction with the thumb, allow flattening and cupping of the palm. The third and highly mobile component is made up of the 12 bones of the four fingers. The interaxial lengths of these bones make it possible for the sensate fingertip to move in a motion commonly found in nature—the equiangular spiral (Fig. 14–53). This spiral motion of the fingers accounts for the adaptability of the hand to any size object and is the consequence of the fact that the distance between the axes of the joints of the bones of the finger follows precisely the Fibonacci sequence.¹

PLANNING RECONSTRUCTION

The reconstruction of any complex hand injury or anomaly must first address the restoration of stability of the fixed unit. Here fusions and osteotomies are used to restore the two arches and their stability. When adequate power in the wrist motors is absent and transfers are not available,

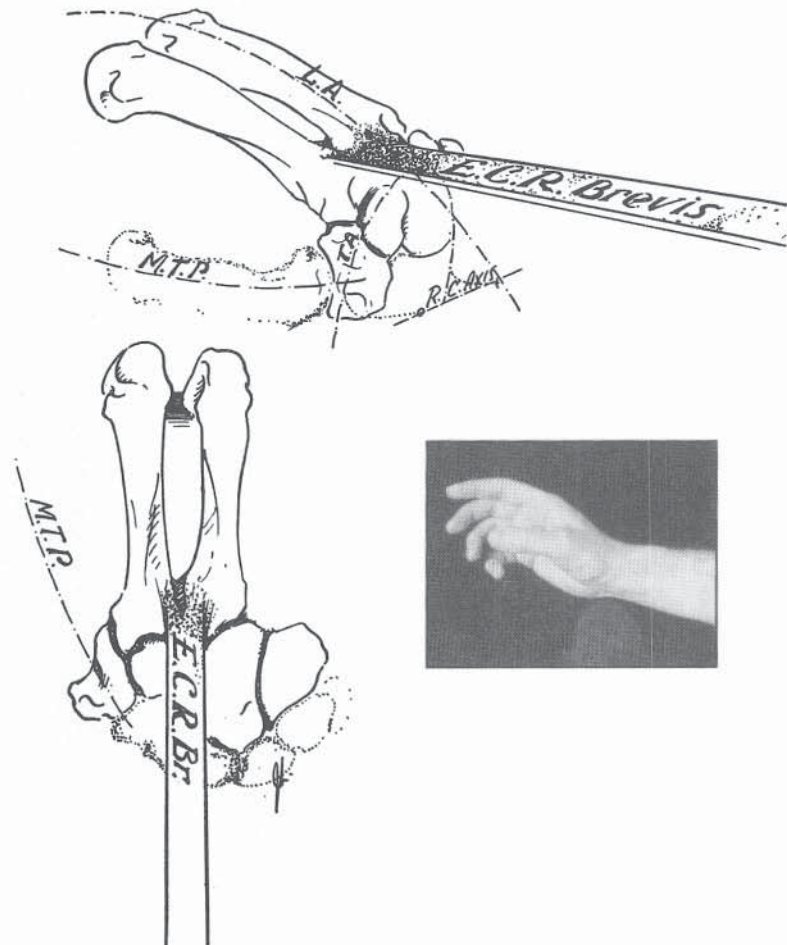


FIGURE 14–51 The fixed unit is the foundation of the hand. Its position and stability are controlled by the extrinsic motors of the wrist. (From Littler JW: Physiology and dynamic function of the hand. *Surg Clin North Am* 1960; 40[2]:261.)

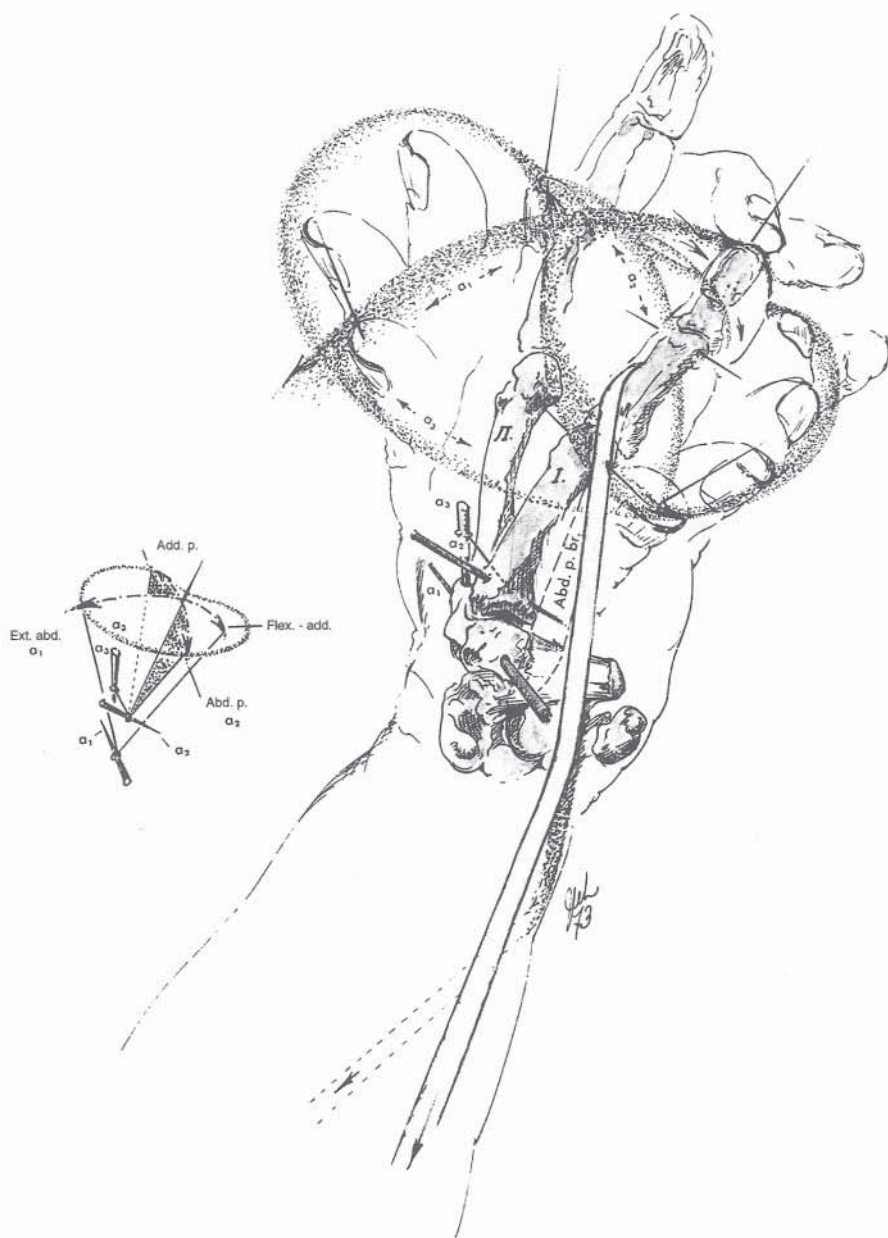


FIGURE 14-52 The thumb ray's mobility and great strength account for almost half of the function of the hand. Stability of the thumb is provided both by the extrinsic muscles of the thumb in the forearm and by the cone of intrinsic muscles of the thenar region. These muscles receive innervation from all three of the great nerves of the upper limb—the median, ulnar, and radial. The motor units of this three-bone cantilever act on the multiplanar first CMC joint and two distal ginglymus joints to make up the radial mobile unit of the hand. (From Carter P: *Reconstruction of the Child's Hand*. Philadelphia, Lea & Febiger, 1991.)

a wrist fusion is required to allow the fingers and digits to experience the motion provided by the muscles originating above the wrist. In general, these operations to restore the fixed unit or other elements of the skeleton require prolonged immobilization. As soon as skin coverage is obtained, bone reconstruction takes priority. Bone reconstruction must not be combined with operations that require early movement, such as tenolysis, tendon repair, or tendon transfer. It is a simple but often ignored surgical principle that one cannot hold something still and move it at the same time.

Reconstruction of the mobile portion of the hand requires wrist stability, flexible digital joints, gliding tendons, and the combined power of extrinsic and intrinsic muscles. Without the balanced function of these four components, the long cantilever of the wrist and multiarticulated finger falls into the familiar claw or swan neck deformity. Although

children maintain joint mobility and tendon gliding better than adults, the surgeon must be certain that the patient has developed passive motion of joints before adding active muscle power. If the surgeon cannot move the child's joints easily with his own hands, there is no possibility that the child's diminutive muscle which the surgeon transfers to the area can move them.

Another important principle in reconstructing the mobile unit is the synergy of wrist and finger motion. Active finger extension cannot be restored without wrist flexion power that is at least equal to the power of the extrinsic finger extensor muscles. Conversely, finger flexion requires the active, strong, simultaneous contraction of wrist extensor. The inability of a patient with a radial nerve palsy to make a fist is convincing evidence of this critical synergism. When the synergistic wrist motor is not available, wrist arthrodesis is required.

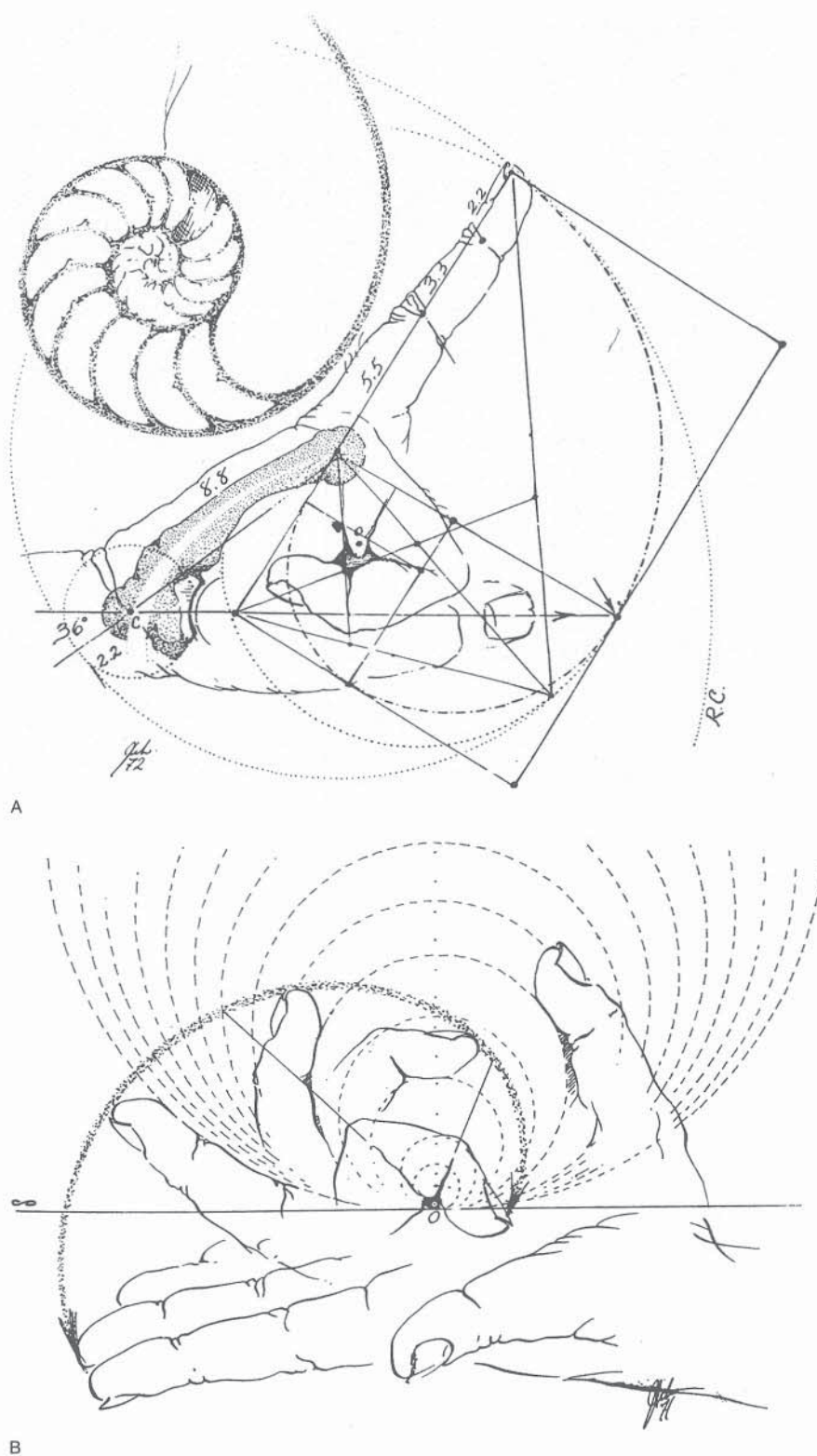


FIGURE 14-53 The adaptability of the mobile finger unit is the result of the course traversed by the sensate tip of the finger. This path is that of an equiangular spiral—one of the most common shapes in nature. **A**, It is the consequence of an arrangement of the distances between the joint axes in a sequence described by the Italian mathematician, Fibonacci: 0, 1, 1, 2, 3, 5. **B**, Since the equiangular spiral is a circle with a continuously increasing radius, it provides an infinite variability in the size (radius) of objects that the digit can surround. (From Carter P: *Common Hand Injuries and Infections*, p 17. Philadelphia, WB Saunders Co, 1983.)

EVALUATION OF THE CHILD FOR RECONSTRUCTION

The decision regarding which children can be helped by surgical reconstruction, at what age it should be carried out, and which operative procedure would be appropriate is the critical challenge for the pediatric hand surgeon. Although

in some cases this decision is straightforward, in others it can be very complicated. The guidelines from adult surgery are important, but there are some additional factors to consider before embarking on reconstruction in children. These factors include the child's variable and often limited ability to cooperate in the postoperative period, the anesthetic risk in early life, the size of the structures, absence of critical

parts, the need for growth, and occasionally the reduced life span of some patients with particular congenital syndromes. The timing of reconstruction in children was discussed in detail earlier in this chapter, and the reader is encouraged to review this material.

Surgical decisions for those treating children can sometimes be difficult to make. The child's disabilities and deformities may seem overwhelming to the surgeon and parent. Furthermore, making a selection from a long list of possible treatments is one of the most important and sophisticated tasks in hand surgery. The goal is to get the most from the least insult to the limb. This is important, because the healing capacities of the tissues are compromised by repeated operations. We have found the following guidelines useful for decision making in complex reconstruction problems. We try to address each of these questions in order.

1. *What is the patient's problem?* This question must be answered *after* the diagnosis has been made. Sometimes it can be difficult to get the child or parent to verbalize how their problem really affects their daily lives. The surgeon must observe and listen carefully to both parent and child to discover the child's exact needs.

2. *Goals of treatment, short and long term.* Treatment of the patient's short-term goal requires the surgeon to remain focused on the patient's problem, as noted above, and to discover those losses that are critical to the patient's daily life. Reduced to its basic elements, the hand needs the power of pinch and grasp with durable, sensate coverage.

The long-term goal of treatment is to make the patient as independent as possible. Care must be taken to avoid making the patient dependent on the medical team any more than is necessary.

3. *Reasonable methods to achieve these goals.* The key here is reasonableness. The surgeon must carefully consider the likely responses to the planned operation by the child, the parent, and the tissues of the child's hand. In addition, the surgeon must realistically evaluate what the surgeon himself or herself *can actually deliver* in the operating room and postoperative period.

4. *Reasonable time schedule.* Both the surgeon and the family find comfort when a realistic time schedule is agreed on at the beginning. This schedule may need revision later, but the child's exit from the medical environment should not be delayed more than necessary. Although follow-up to the treatment is important, the final goal of all hand surgery is the maximum *independent* life for the patient. When a

treatment plan is open-ended, the child and parent tend to delay taking responsibility for this ultimate goal.

5. *Outcome evaluation method.* All too often, failure to take preoperative photographs, to measure joint angles and grip strength, and to obtain functional tests prior to treatment renders an honest, accurate assessment of the result at the end of treatment impossible. This is as important as establishing the type of treatment.

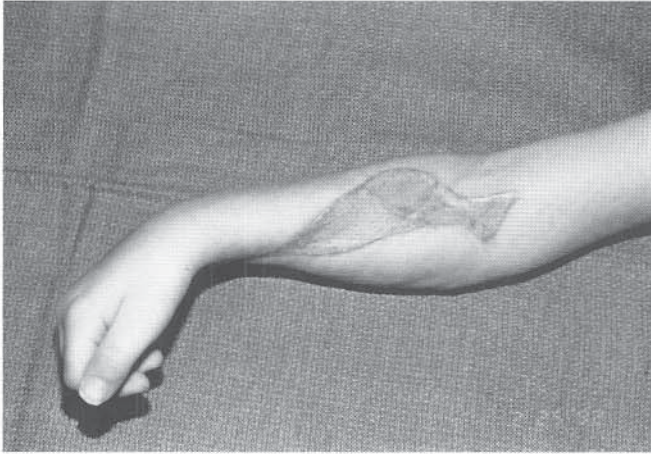
By applying these guidelines to those complex problems that initially seem overwhelming, a more rational approach can be developed by the surgeon in conjunction with the patient and family. As a rule, simple is better. Littler has said it well: "There is no place in surgery for the superfluous" (J. W. Littler, personal communication).

ESTABLISHED VOLKMANN'S CONTRACTURE OF THE FOREARM

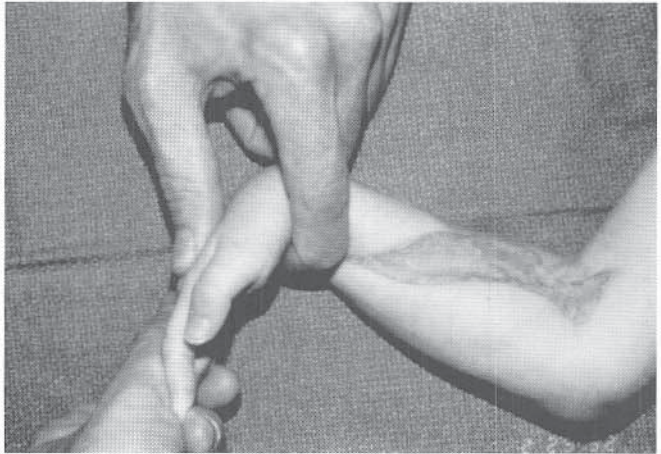
The child with an established Volkmann's contracture of the forearm provides an example where the decision-making guidelines listed above can be helpful. A child with a completely developed Volkmann's contracture presents with a useless extremity fixed in flexion and a complete median and ulnar nerve palsy. The finger contracture is fixed, sensibility of the digits is absent, the intrinsic muscles of the hand are paralyzed. The problem at first seems overwhelming, and future use of the extremity seems hopeless. However, with an understanding of the pathophysiology, the result of reconstruction can be spectacular, and almost complete restoration of function can be achieved (Fig. 14-54).

To achieve this good result, the surgeon must first recognize that the condition is really two separate but related problems—contracture of the forearm digital flexor muscle mass and a compression neuropathy of the median and ulnar nerves as they pass through this fibrotic muscle infarct in the proximal forearm² (Fig. 14-55). Correction is achieved in two separate operations. First, the infarct ischemic muscle mass is completely removed from the forearm, which simultaneously relieves the pressure on these nerves. Usually the nerves themselves are only compressed and not infarcted, so nerve grafting is not required. The wrist flexor muscles are usually not affected because they are in the superficial layer of the forearm and are carefully protected during the dissection. With the extrinsic contracture restricting digital motion now completely relieved, the forearm skin is closed. When sensibility and intrinsic function have

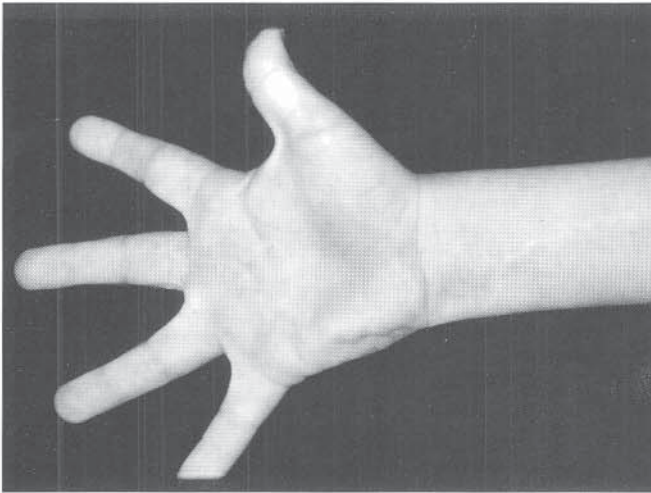
FIGURE 14-54 A, Despite early, aggressive fasciotomy and subsequent skin grafting, Volkmann's contracture developed in this 4-year-old child after a supracondylar fracture of the humerus. His hand was dry and completely without sensibility, and all of the intrinsic muscles were paralyzed—the classic picture of a complete median and ulnar nerve palsy. B, Eaton's angle. The same young patient has the typical forearm muscle infarct due to the digital flexor muscle necrosis. Contracture of these muscles causes a tenodesis effect at the wrist. Eaton's angle is the maximum passive extension of the wrist allowed when the digital joints are held in full extension. Here the angle is almost 90 degrees. The contracture is assessed by stabilizing the digits in neutral position. C and D, The restoration of active flexion and extension required two surgical procedures. The first included both a resection of the entire infarcted digital flexor muscle and neurolysis of the median and ulnar nerves compressed by this infarct. After 6 months, sensibility and intrinsic function returned. Subsequently, at a second operation, a free microsurgical neurovascular transfer of the gracilis muscle was done, attaching the motor nerve of the gracilis muscle to the remaining anterior interosseous nerve stump in the forearm. E and F, The gracilis muscle now provides the patient with actively contracting extrinsic digital flexion and a grip strength of 15 pounds (35 N) at age 10. He has returned to vigorous active function.



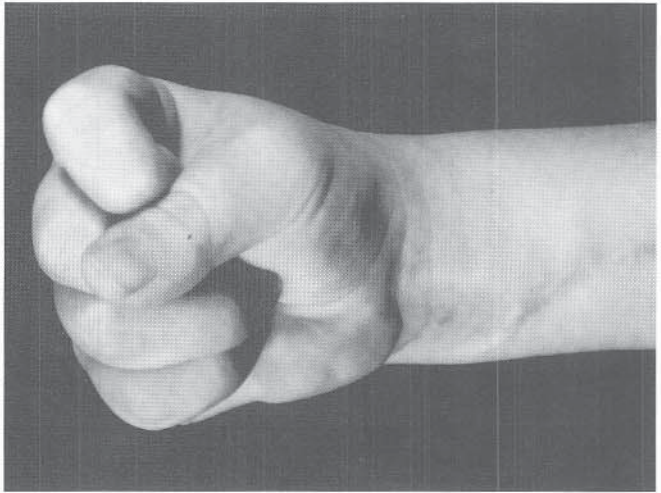
A



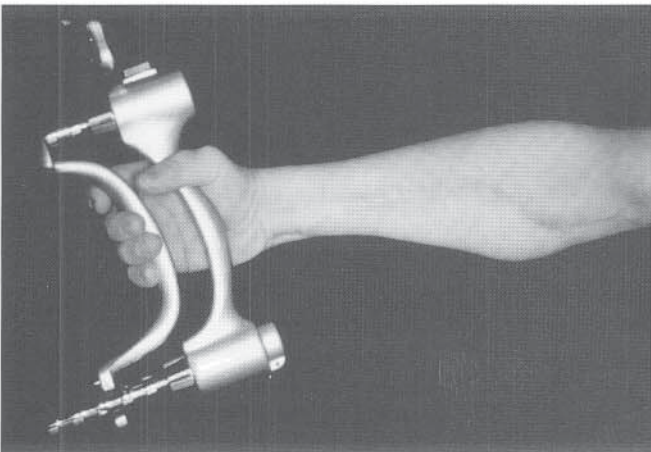
B



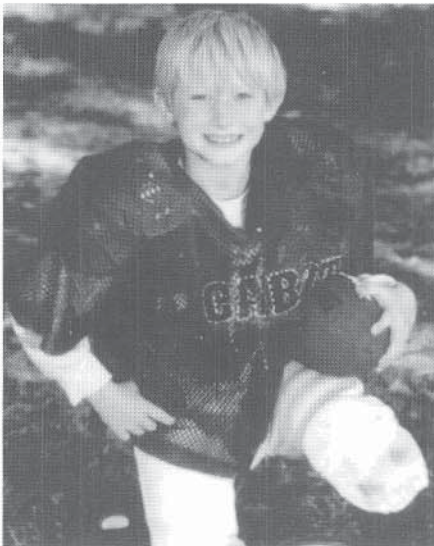
C



D



E



F

FIGURE 14-54 See legend on opposite page

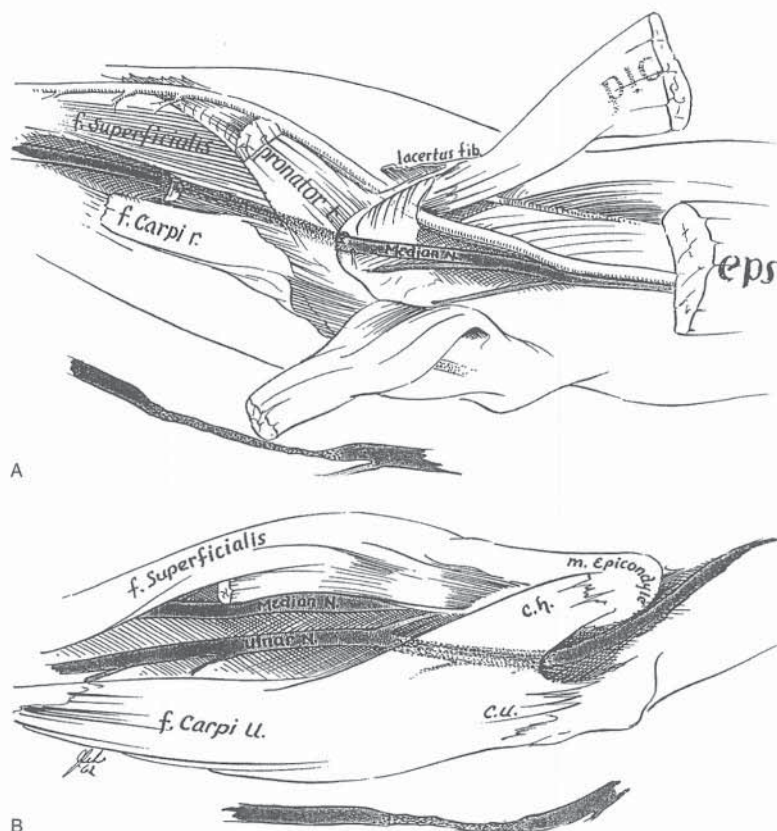


FIGURE 14-55 A, The compression neuropathy in Volkmann's contracture illustrated in Figure 14-54 is caused by the infarct of the sublimis, profundus, and pronator compressing the median nerve. B, Compression of the ulnar nerve in cases like the one shown in Figure 14-44 occurs as the nerve passes under the two heads of the flexor carpi ulnaris. (From Converse JG: *Reconstructive plastic surgery*. In Littler JW (ed): *The Hand and Upper Extremity*, 2nd ed, vol 6, p 3135. Philadelphia, WB Saunders Co, 1977.)

returned, which may take several months, the second procedure is indicated. Extrinsic finger flexor muscle function may now be restored by a transfer of the intact extensor carpi radialis longus (see Fig. 14-48) or a free neurotized gracilis muscle transfer.

REFERENCES

Principles of Reconstruction

1. Littler JW: On the adaptability of man's hand. *Hand* 1973;5:187.
2. Littler JW: "Principles of Reconstructive Surgery of the Hand." In Converse JM (ed): *Reconstructive Plastic Surgery*, 2nd ed, p 3103. Philadelphia, WB Saunders Co, 1977.

Congenital Anomalies

CLASSIFICATION

The classification of congenital limb anomalies is made difficult by the spectrum of presentations for developmentally related conditions, the myriad terms derived from Greek and Latin for the same condition, and the overlap in clinical appearance of conditions with differing etiologies. In particular, etiologic information may erode a neat anatomic classification.

A good classification system should allow a reproducible "fit" by any user, should group the spectrum of presentations of the same anomaly consistently into the same category, and should be specific and useful. It should allow retrieval of information and facilitate communication between geneticists, surgeons, pediatricians, and embryolo-

gists. Moreover, a good classification system should be flexible enough to accommodate new information about developmental and genetic etiologies for the conditions classified.

The classification system adopted by the International Federation of Societies for Surgery of the Hand remains a useful framework for classifying limb abnormalities.¹ It is based on a mixture of anatomic sites and presumed developmental errors. A single anomaly or group of anomalies may fit into more than one category or may not be easily classified into any group. In its simplest form, the classification system adopted by the IFSSH in 1976 is given in Table 14-4. This system can be expanded to include every anomaly. With better understanding of developmental pathways and elimi-

TABLE 14-4 **Classification of Congenital Anomalies**

- Failure of formation
 - Transverse arrest
 - Longitudinal arrest
- Failure of differentiation (separation) of parts
 - Soft tissue involvement
 - Skeletal involvement
- Duplication
- Overgrowth
- Undergrowth
- Constriction ring syndrome
- Generalized abnormalities and syndromes

Adapted from Swanson AB: A classification for congenital limb malformations. *J Hand Surg* 1976;1-A:8.

nation of overlap in terminology, this system may be flexible enough to withstand the test of time.

REFERENCE

Classification

- Swanson AB: A classification for congenital limb malformations. *J Hand Surg* 1976;1-A:8.

DIAGNOSING ASSOCIATED ANOMALIES

It is very important to recognize coexisting anomalies associated with upper limb abnormalities in order to plan treatment, counsel the family, and work with other providers of health care. The conditions may affect treatment decisions, anesthesia techniques, the timing of surgery, and the coordination of multiple disciplines in the treatment of the child.

Anomalies may be characterized as syndromes (patterns whose causation is understood), associations (anomalies that occur in combination but whose underlying causation is not known), and sequences (anomalies whose coexistence results from a cascade of events during development). Anomalies may be associated on the basis of genetic coding, timing of development of other organ systems, or recognized patterns of disruption. Known syndromes of heritable anomalies are now finding explanation in localization of the genetic error and an understanding of how the incorrect protein affects morphogenesis. Multiple anomalies may be associated as a result of a generalized insult to the developing embryo, with the particular association reflecting the timing of the insult. The classic diagnosis in this category is the VATER association.

It has long been known that radial dysplasia is one of the findings in a number of conditions, including the potentially life-threatening Holt-Oram syndrome, Fanconi's anemia, and thrombocytopenia-absent radius syndrome. This list was at one point limited enough to be included in a text such as this. The rate at which new information is discovered and new reports published makes this impractical. General information can be found in books such as *Smith's Recognizable Patterns of Malformation*¹ and *Clinical Syndromes*.³ An extremely useful source of information that is updated continuously is the Online Mendelian Inheritance in Man (OMIM) web site, maintained by the National Institutes of Health (<http://www.ncbi.nlm.nih.gov/Omim/>). Links to PubMed, a reliable source of medical literature maintained by the NIH, and additional informative web sites are provided. A web search by diagnosis will usually produce information posted and maintained by families and support groups for children with a particular diagnosis. These sites are kept current by dedicated volunteers; most information, however, is not peer-reviewed.

In general, anomalies that affect upper and lower limbs simultaneously are often the result of genetic alterations either with known heredity or as new mutations.

Mutations in the regions that code for the fibroblast growth factors (FGF) have been shown to be responsible for a number of limb malformations in association with craniofacial abnormalities. Different base pair substitutions cause different alterations in the three-dimensional configuration of the protein. This change in shape prevents normal binding of the factor with the receptor site. Some of the

TABLE 14-5 **Malformations Associated with Fibroblast Growth Factor or Receptor Mutations**

FGFR	Syndrome	Comments
FGFR 1	Pfeiffer's syndrome	Overlap in syndromes and protein mutation
FGFR 2	Noack's syndrome Carpenter's syndrome	
FGFR 2	Apert's syndrome Crouzon's syndrome Jackson-Weiss syndrome	Different mutations of the same protein producing phenotypic differences
FGFR 3	Achondroplasia Hypochondroplasia Thanatophoric dwarfism	Autosomal dominant mutations, chromosome 4p16.3

resulting syndromes are listed in Table 14-5. Another example are the different phenotypes of cleft hands and feet that map to different chromosomes. Three different mutations have been localized.

Those abnormalities that are associated with radial-sided dysplasia, including hypoplastic thumbs, may be associated with other conditions, including cardiac, renal, vertebral, and hematopoietic disorders. These kinds of known associations must be investigated further as part of the treatment plan. Consultation with other pediatric disciplines is important in planning reconstructive care for the child.¹⁻³

REFERENCES

Diagnosing Associated Anomalies

- Jones KL (ed): *Smith's Recognizable Patterns of Human Malformation*, 4th ed. Philadelphia, WB Saunders Co, 1988.
- Temtamy SA, McKusick VA: An overview of hand malformations. In Bergsma D (ed): *The Genetics of Hand Malformations: The National Foundation—March of Dimes. Birth Defects: Original Article Series*, vol 14, no 3, p 1. New York, Alan R Liss, 1978.
- Weidemann HR, Kunze J, Grosse FR: *Clinical Syndromes*, 3rd ed. London, Mosby-Wolfe, 1997.

ULNAR DYSPLASIA

History. Goller is said to have first described the deformity in 1693.⁹ However, it was Priestly who in 1856 presented a case of a newborn with an absent ulna and a hand with only a thumb and index finger. He recognized a longitudinal deficiency state different from those deformities with transverse amputations.⁶

Etiology. The cause of ulnar deficiency is unknown. From studies in human embryos, the deficiency likely occurs during weeks 4 and 5 of fetal development, in the earliest stages of upper limb formation. Ogino and Kato induced longitudinal deficiency states in rats using Myleran.⁷ Ulnar deficiency occurred 1 week earlier in embryologic development than radial deficiency.

Ulnar dysplasia is seen as a part of several syndromes whose genetic locus has been identified, and these are discussed elsewhere (see Chapter 32, Limb Deficiencies). However, genetic details for the isolated spontaneous and most common form of ulnar clubhand are unknown. A listing of these syndromes can also be found in Chapter 32 under Ulnar Deficiencies.

Clinical Presentation. Although the terms *ulnar* and *radial clubhand* suggest that these two conditions are similar, patients with these conditions differ in almost every respect.

1. Visceral anomalies of the heart, hematopoietic and gastrointestinal system are common in radial dysplasia but, although reported, are rare in ulnar dysplasia.
2. Other musculoskeletal anomalies are usually absent in radial deficiency but are often present in ulnar deficiency. These anomalies vary widely and include proximal femoral focal deficiency, fibular and tibial ray deficiency, phocomelia, scoliosis, clubfeet, absent patellae, congenital dislocation of the hip, coxa vara, and spina bifida.¹¹
3. The wrist is usually unstable and a significant clinical problem in radial dysplasia. The wrist of a patient with ulnar clubhand usually needs little treatment for the deformity, which is usually mild and rarely unstable.
4. The elbow in radial deficiency, if present, is stable. In patients with ulnar deficiency, the elbow may be stable, unstable, or fused.
5. In radial deficiency, either the hand is normal or only the radial components are affected. In ulnar deficiency states, the hand deficiency varies widely and is often severe. The distribution of the deficit in the hands of

patients with ulnar deficiency may occur on either the radial or the ulnar border of the hand, or both.

6. Proximal limb abnormalities of the shoulder and humerus are rare in radial clubhand but may be very severe in ulnar deficiency. Some of these patients blend in with the phocomelic population, suggesting that the developmental error in ulnar dysplasia and phocomelia may be related.
7. Total absence of the radius is the most common presentation of radial deficiency. In ulnar deficiency, partial absence is by far more common.
8. The prevalence of radial deficiency is greater than the prevalence of ulnar deficiency but may not be as high as previously thought. At Texas Scottish Rite Hospital for Children, 102 radial clubhands and 66 ulnar clubhands have been treated. The ratio in the literature varies between 1:2.3 in a German study³ to 1:10 radial deficiencies.⁸ The reported incidence of ulnar deficiency is 1 in 100,000 live births.¹ As an indication of the rarity of ulnar dysplasia, approximately 160 cases have been reported in the world's literature.⁹

Classification. To assist in surgical decision making for patients with this rare deformity, and after consideration of the efforts of many previous authors, we have settled on

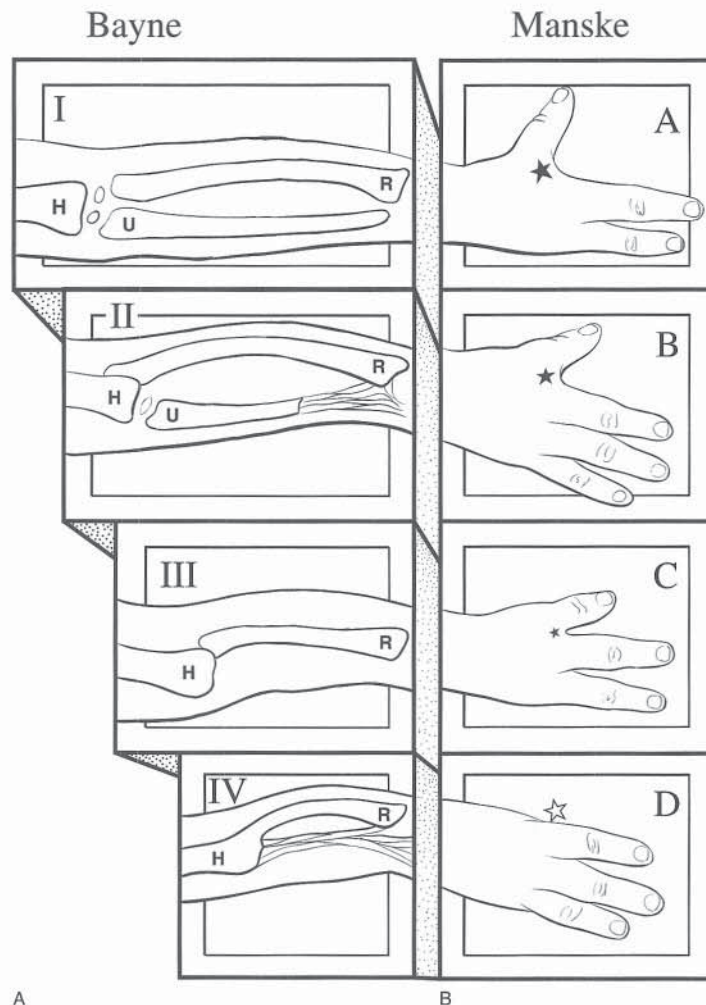


FIGURE 14-56 Of many classifications for ulnar hemimelia, the two most frequently used are those of Bayne and Manske. Both are clinically useful. Bayne's focuses on the forearm and wrist, and Manske's focuses on the hand. There is no necessary connection between the two classification systems; any variety of hand anomaly can be associated with any variety of forearm involvement. Less improvement in function for the child comes from surgical procedures in the forearm/wrist area than from surgical procedures on the hand. The treatment of patients with ulnar dysplasia should focus on the hand rather than the forearm. Substantial gains in function come from operations creating or improving opposition and the thumb-web space.

two clinically useful classification systems: Bayne's classification for forearm and elbow deformities and Manske's classification for hand deformities.

Bayne's classification is widely used and focuses on the elbow and forearm (Fig. 14-56A). The stability of the wrist is usually good in forearm variants unless the ulnar anlage is present.

Manske's classification of the hand defect associated with ulnar dysplasia is more important from a functional standpoint (Fig. 14-56B). Manske's classification is particularly useful clinically because the substantial functional gains are derived more often from surgical operations on the hands of these patients and less often from operations on their forearms or elbows. We believe the surgeon's main focus should be the hand rather than the forearm.⁴

Pathology. In some of the ulnar deficiency states, a curious fibrocartilaginous mass thought to represent the anlage of the absent portion of the ulna may be present. It is seen in Bayne's types II and IV (Fig. 14-56A) and originates proximally in either the distal ulna or the humerus. In the proximal portion the ulnar anlage is formed of hyaline cartilage. Distally the mass continues as fibrocartilage and may insert into the distal radius, the carpal mass, or both. The structure has been described by Riordan as similar to a fiberglass fishing rod that allows bending but no increase in length.^{5(p416)}

Controversy exists about the significance of this fibrocartilage anlage and its effect on progressive bowing of the radius, deviation of the wrist, and dislocation of the radial head at the elbow. Some authors have suggested that the anlage is unimportant and requires no surgical treatment since follow-up in nonoperatively treated patients has not shown convincing evidence of progressive deformity.² Others have suggested that early resection of the anlage at 6 months of life is indicated in healthy babies.^{5(p420)}

Imaging. No unusual imaging techniques are required. As in cases of radial clubhand, clinical measurement of deformity should augment the radiographic measurements of bowing and radial deviations, which are vulnerable to inconsistency in positioning for the radiographic study.

Treatment

GENERAL PRINCIPLES OF MANAGEMENT. Surgeons have been appropriately cautious in operating on these patients since 1926, when Southwood wrote, "From a functional viewpoint the deformed limb is much more useful than its anatomical condition would lead one to expect."¹⁰

Today, reasonable treatments of ulnar clubhand are founded on the fact that the most important functional gains for these patients usually come from operations on their hands, not on their wrists or forearms. Crucial gains are especially derived from improving the thumb-index web space.

Careful clinical assessment of the ulnar deviation of the wrist and passive correction should be recorded initially and at subsequent follow-up examinations because wrist deformity, elbow radial head dislocation, and radial bow may occasionally be progressive. Relying on the radiograph alone is not adequate. Progression verified by careful clinical measurements of the ulnar deviation deformity is the strongest indication for anlage resection in Bayne's types II and

IV. Improving the aesthetic appearance of the malaligned hand and forearm unit is a reasonable goal of this treatment.

NONOPERATIVE TREATMENT. Early splinting and stretching of the ulnar-deviated wrist is reasonable when the wrist is deviated more than 30 degrees. Infants less than 6 months old usually tolerate a splint better than stretching. After 6 months, if the wrist still shows 30 degrees or more of fixed ulnar deviation, surgical correction should be considered.

OPERATIVE TREATMENT

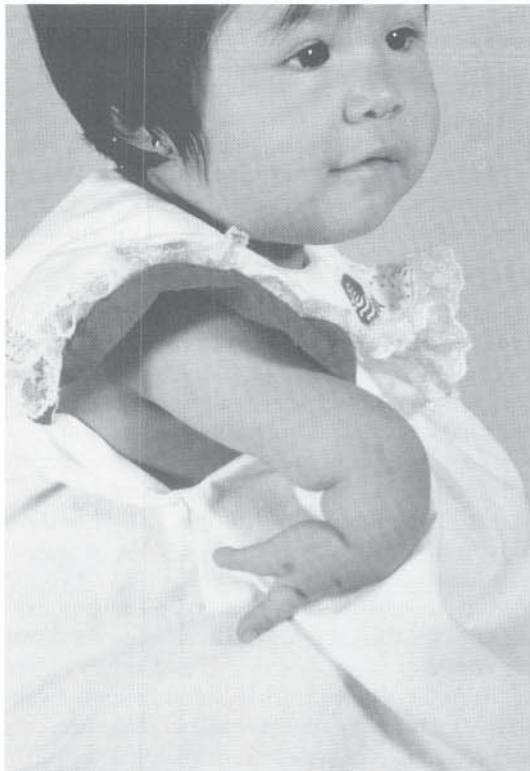
Wrist. When there is evidence of deformity greater than 30 degrees of ulnar deviation or evidence of progression, resection of the anlage is appropriate in Bayne's type II or IV forearm. Most of these children are usually otherwise healthy (unlike their radial dysplastic counterparts). Early treatment at 6 months of age is appropriate, since as the child grows, the forearm will double almost twice in length, and the first doubling occurs in the first 3 years of life. Early resection affords the greatest possibility of reducing the tether of growth by the anlage.

The procedure is carried out with a lazy S-incision over the ulnar aspect of the forearm and wrist. Because the flexor carpi ulnaris is absent, the ulnar nerve and artery, when present, may lie immediately under the incision in the subcutaneous tissue. Once the neurovascular structures are identified and retracted, the anlage is dissected. It is a firm fibrous structure originating from the proximal residual ulna, in type II, or from the humerus, in type IV (Fig. 14-56A). It is critical that dissection distally carefully and completely exposes the attachment of the fibrous anlage to the carpus and, when present, to the radius. The structure is completely resected from the carpus and the distal radius. It should be easy to passively deviate the wrist at least to neutral after resection. Complete proximal excision of the anlage is less important. Osteotomy of the radius is appropriate if excessive bowing is present. Postoperatively a reasonable period of follow-up stretching and splinting is instituted, usually for about 6 months.

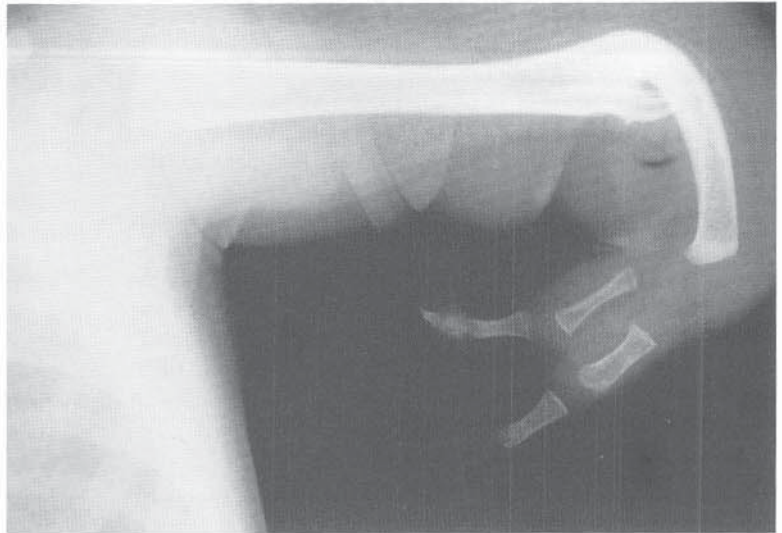
Hand. Although forearm and wrist surgery is best done during the first year of life, hand operations should be done later. Syndactyly and first web reconstructions are important procedures to improve the use of the hands in these children. Better results come from more precisely done operations on slightly larger hands, and we prefer to delay hand surgery in these children until the second year of life.

Rotational osteotomies of the metacarpals are indicated in those hands with digits that are all aligned in the same plane. These flat hands make prehension with the pulp of the digits impossible, and osteotomies to rotate the metacarpals or phalanges into opposition can improve prehension. There is a tendency for the rotation achieved at operation to slowly return to the preoperative state, and concomitant realignment of muscle power with tendon transfers may help prevent derotation.

Forearm. The creation of a one-bone forearm should be reserved for older children with type II dysplasia and is indicated only when the instability of the forearm is truly disabling. This is rarely the case. The price paid in loss of forearm rotation by a child with a severely disabled hand is rarely worth the additional stability or questionable cosmetic improvement afforded by the one-bone forearm. In our experi-



A



B

FIGURE 14-57 A and B, Bayne's type IV forearm may occasionally be associated with marked rotation of the forearm and hyperextension of the elbow fusion. This leads to the hand resting behind the trunk in such a way that the arm at first appears to be on backward. This unusual position of the hand in relation to the trunk and contralateral hand prevents the child from using the hand in front of herself in her field of vision. Repositioning of the hand is especially critical because most of these hands are severely affected enough to require bimanual use by the patient.

Operative intervention, although potentially functionally useful, carries considerable risk owing to the rotational stress on tethered vascular structures.

ence and others', any function gained from increased stability is greatly offset by the loss of forearm rotation needed for positioning the hand for use.

Elbow. In some type IV cases, osteotomy of the elbow synostosis may be very useful. This is especially true when the elbow deformity positions the hand behind the child and away from the opposite uninvolved hand (Fig. 14-57). Although the benefit of this surgery is potentially great, there is a serious risk of vascular compromise owing to tethering of the vessels at the osteotomy. Because of the potential for catastrophic complications and loss of the limb, this is not an operation for the occasional pediatric upper limb surgeon.

Radial head resection in types III and II has often been considered when the head is dislocated. However, we try to avoid radial head resection, since pain is usually not a problem for these children. Radial head excision increases elbow instability and usually should be avoided.

REFERENCES

Ulnar Dysplasia

1. Birch-Jensen A: Congenital Deformities of the Upper Extremities, p 15 (E Aagesen, trans). Odense, Denmark, 1949.
2. Broudy AS, Smith RJ: Deformities of the hand and wrist with ulnar deficiency. *J Hand Surg* 1979;4-A:304.
3. Buck-Gramcko D: Current Practice in Hand Surgery. St. Louis, CV Mosby Co, 1997.
4. Cole RJ, Manske PR: Classification of ulnar deficiency according to the thumb and first web. *J Hand Surg* 1997;22-A:479.
5. Flatt AE: The Care of Congenital Hand Anomalies, 2nd ed. St. Louis, Quality Medical Publishing, 1994.
6. Kelikian H: Congenital Deformities of the Hand and Forearm, p 866. Philadelphia, WB Saunders Co, 1974.

7. Ogino T, Kato H: Clinical and experimental studies on ulnar ray deficiency. *Handchir Mikrochir Plast Chir* 1988;20:330.
8. O'Rahilly R: Morphological patterns in limb deficiencies and duplications. *Am J Anat* 1951;89:135.
9. Peimer C (ed): Surgery of the Hand and Upper Extremity, p 2095. New York, McGraw-Hill, 1996.
10. Southwood A: Partial absence of the ulna and associated structures. *J Anat* 1926;61:346.
11. Tonkin M: Ulnar longitudinal deficiency. In Green TR (ed): Operative Hand Surgery, 4th ed, p 358. Churchill Livingstone, 1998.

SYNSTOSIS OF THE RADIUS AND ULNA

History. The first reported case is said to be by Sandifort, in 1793, approximately a century before Roentgen. Morrison described the classic clinical findings in 1892, 3 years before the discovery of x-rays.⁷

Synostoses represent a failure of differentiation of parts. Although the precise cause of synostosis of the forearm is unknown, the time of occurrence is almost certainly during the earliest portion of embryonic limb development. There is no known teratogen trigger, but in a study by Jaffer and colleagues, two of 15 infants with fetal alcohol syndrome had the anomaly.⁵ At about 5 weeks post conception, the elbow forms from the three cartilaginous condensations representing the humerus, radius, and ulna. For a short period these cartilage analogues share a common perichondrium.⁶ Subsequently a cavitation process ensues, forming the three distinct bones. If this process fails, enchondral ossification results in the bony synostosis. Because the forearm bones separate at a time when the fetal forearm is in pronation, essentially all forearm synostosis is fixed in this position.

The condition occasionally affects other members of the family, usually in an autosomal dominant inheritance pattern.¹²

Because the event that causes radioulnar synostosis occurs so early in fetal development, when all organ systems are forming, it may be seen in conjunction with other syndromes. These include Apert's syndrome (acrocephalosyndactyly), Carpenter's syndrome (acropolysyndactyly), arthrogryposis, mandibulofacial dysostosis, Klinefelter's syndrome, and Poland's anomaly.^{2,13} Approximately one-third of these patients have other anomalies, according to Upton,¹³ but no common pattern is seen.^{2,13} Cardiovascular (tetralogy of Fallot and ventricular septal defects), thoracic (absence of first rib or pectoral muscles), genitourinary, gastrointestinal, CNS (microcephaly, hydrocephalus, encephalocele, mental retardation, delayed milestone attainment, hemiplegia), and other musculoskeletal anomalies are seen in association with radioulnar synostosis. Waters and Simmons, on the other hand, report that although radioulnar synostosis is present in the above syndromes, it is an isolated anomaly in one-third of cases.^{11(p2052)}

Clinical Presentation. Boys are slightly more often affected with radioulnar synostosis than girls (3:2 ratio). The lesion is bilateral in 80 percent of cases.¹¹ The diagnosis is often delayed, and the patients frequently lack functional complaints when the position of fixed pronation is moderate and carpal compensatory hypermobility is adequate. Affected children sought medical attention at an average age of 6 years in Cleary and Omer's study³ and 2.5 years in the study by Simmons and Waters.¹¹

Cleary and Omer followed 23 patients who had been treated nonoperatively.³ On careful functional testing they could identify no advantage of early operation. Their final recommendation was that operative correction was rarely indicated and that when surgical treatment is selected it should be based more on limitations in function than on physical or radiographic findings.

In sharp contrast, Simmons and Waters felt that most patients did have functional limitations related to the synostosis. When the fixed pronation was great, patients complained of difficulty using a spoon or pencil, buckling belts, fastening buttons, and grasping small objects. Some patients also reported difficulty in sporting activities.¹¹

Interestingly, in Ogino and Hikino's Japanese study, the patients' complaints of dysfunction most commonly were in the nondominant arm.⁸ Perhaps the eating modes of Asians are critical in those patients because the nondominant hand must hold the rice bowl in supination. The degree of pronation averaged about 60 degrees in either nondominant or dominant forearms, but it was as little as 30 degrees in the dominant forearm and 10 degrees in the nondominant forearm.⁸ Like other authors, Ogino and Hikino noted the remarkable compensation by wrist hypermobility for lack of forearm motion. They felt that the amount of wrist mobility should be considered alongside the position of fixed pronation when electing surgical repositioning.

It is important that a careful clinical measurement of the patient's exact forearm fixed position be made as accurately as possible, since intraoperative decisions are based in part on this assessment. All degrees of fixed pronation are seen, but the most common are less than 30 degrees of pronation

(40 percent) and more than 60 degrees of pronation (40 percent). By comparing the angle of pronation to the plane of the palm, one may assess the compensatory rotation through the carpus, which is usually increased in these patients and enhances use of the hand, especially when the shoulder is normal.

Imaging. Radiographs usually show proximal radioulnar coalescence, but extensive synostosis extending distally into the forearm is occasionally seen.¹³ There are several classification systems based on radiographic appearance. Tachdjian has noted three types:^{12(p207)}

- I. True congenital radioulnar synostosis, or headless type. Here the radial head is absent and a bony fusion of the radius to the ulna is present. The distal ends are fused and the radius is bowed, thicker than the ulna and not attached to it distally.
- II. Dislocated radial head type. The malformed radial head is posteriorly dislocated and the proximal end of the radius is fused with the ulna just below.
- III. No bony synostosis is present, but a thick fibrous interosseous ligament forms that attaches to each bone just distal to their proximal ends and prevents rotation. This is the rarest type in Tachdjian's classification.

In Omer's classification, four radiographic types represent a continuum from fibrous to complex bony synostosis.³ Omer did not feel this classification was helpful in assessing function or making decisions about treatment:

- I. Fibrous synostosis, with no bony change but a stiff and smaller forearm (6/35, or 17 percent).
- II. Osseous synostosis, radial head present and reduced (3/35, or 19 percent).
- III. Osseous synostosis, radial head present and posteriorly dislocated (20/35, or 57 percent).
- IV. Osseous synostosis, radial head present and anteriorly dislocated (6/35, or 17 percent).

Sachar, Akelman, and Ehrlich have reported progressive dislocation of the radial head in some patients.¹⁰

Treatment. Most patients and parents envision restoration of forearm motion and often are not satisfied with adjustment of the position of the fixed forearm. This has led a few surgeons to attempt correction of the synostosis by inserting various materials, both inert and biologic. Poor follow-up of these patients has precluded the acceptance of any of these techniques by hand and pediatric surgeons other than the authors of the methods.

Even a simple positional change of the forearm is associated with a high complication rate. Surgical intervention has been recommended by most surgeons only when significant amounts of pronation (usually more than 60 degrees) are associated with functional limitations and complaints. Recommendations for the optimal postoperative forearm position vary. Green recommended 20 degrees of supination.⁴ Simmons¹¹ and colleagues recommended 20 degrees of pronation in unilateral cases and, in bilateral cases, 20 degrees of pronation for the dominant arm and neutral position for the nondominant arm. In contradistinction to these North American studies, Ogino and Hikino have noted the need for supination in Asian populations who use chopsticks rather than a knife and fork. They consider the degree of wrist compensatory mobility critical, and try to obtain full

supination of the palm (disregarding the true position of the forearm bones) in the nondominant hand in their Japanese patients. This may require as little as 20 degrees of true forearm supination. For Ogino, 70 degrees of palmar supination in the dominant arm in bilateral cases is adequate. He favors an osteotomy through the fusion mass but emphasizes shortening the arm by resection of 0.5 cm of bone at the osteotomy site.

Although correction by Ilizarov's method has been reported,¹ we have no experience with it.

Results. The reported results of treatment and the complications have varied. Simmons reported excellent results in 70 percent, good results in 12 percent, fair results in 12 percent, and poor results in 6 percent.¹¹ Cleary and Omer noted that poor results were related more to surgical complications or a sedentary lifestyle rather than final forearm position.³ Green and Mital reported excellent results in 50 percent, good results in 33 percent, fair results in 6 percent, and poor results in 6 percent.⁴ Finally in a Japanese population, Ogino and Hikino reported complete relief of symptoms in all patients, all of whom were symptomatic preoperatively.⁸

The complication rates for these series were very high. Ogino and Hikino reported two radial nerve palsies in 13 operations; both patients recovered at 4 and 8 weeks postoperatively. In Simmons's series of 22 cases, there were eight (36 percent "significant complications": one wound infection, three patients with loss of correction (60, 50, and 20 degrees), and four cases of vascular compromise (no permanent disability, but a second operation was often required for remanipulation after the vascular emergency mandated abandoning the original attempt at correction). There was one posterior interosseous nerve palsy, which cleared by 6 weeks postoperatively. Green, out of 13 cases, had two that required repeat osteotomy and one patient with severe Volkmann's contracture.⁴

Author's Preferred Treatment. At the Texas Scottish Rite Hospital for Children, the diagnosis of radioulnar synostosis was made in 131 patients; of these patients, 13 have undergone osteotomy of the forearm. We tend to side with Cleary and Omer and frequently advise against any surgical treatment. Although many patients definitely have functional limitations, most envision full forearm rotation, not just an adjustment of the fixed position. The risk of surgical intervention in these cases barely justifies the benefit of surgical change of the fixed position. When indicated, we have favored proximal osteotomy, and we have recently added the resection of a small, 0.5-cm wafer as described by Ogino and Hikino. Ogino has recently suggested a simpler procedure of distal radial osteotomy in these patients (T. Ogino, personal communication).

REFERENCES

Synostosis of the Radius and Ulna

1. Bolano LE: Congenital proximal radioulnar synostosis: treatment with the Ilizarov method. *J Hand Surg* 1994;19-A:977.
2. Bora F: *The Pediatric Upper Extremity Diagnosis and Management*. Philadelphia, WB Saunders Co, 1986.
3. Cleary JE, Omer GE Jr: Congenital proximal radio-ulnar synostosis: natural history and functional assessment. *J Bone Joint Surg* 1985; 67-A:539.

4. Green WT, Mital MA: Congenital radio-ulnar synostosis: surgical treatment. *J Bone Joint Surg* 1979;61-A:738.
5. Jaffer Z, Nelson M, Beighton P: Bone fusion in the foetal alcohol syndrome. *J Bone Joint Surg* 1981;63-B:569.
6. Lewis W: The development of the arm in man. *Am J Anat* 1901;1:169.
7. Morrison J: Radio ulnar synostosis. *BMJ* 1892;2:1337.
8. Ogino T, Hikino K: Congenital radio-ulnar synostosis: compensatory rotation around the wrist and rotation osteotomy [published erratum appears in *J Hand Surg* 1987;12-B:402]. *J Hand Surg* 1987;12-B:173.
9. Peimer C: *Surgery of the Upper Extremity*. New York, McGraw-Hill, 1996.
10. Sachar K, Akelman E, Ehrlich M: Radioulnar synostosis. *Hand Clin* 1994;10:399.
11. Simmons BP, Waters PM: Congenital abnormalities of the elbow region. In Peimer C (ed): *Surgery of the Hand and Upper Extremity*, p 2052. New York, McGraw-Hill, 1996.
12. Tachdjian M: Congenital radioulnar synostosis. In Tachdjian M (ed): *Pediatric Orthopedics*, vol 1, p 180. Philadelphia, WB Saunders Co, 1990.
13. Upton J: Congenital anomalies of the hand and forearm. In McCarthy JG (ed): *Plastic Surgery*, vol 8, p 5309. Philadelphia, WB Saunders Co, 1990.

MADELUNG'S DEFORMITY

History. In 1878, Otto Wilhelm Madelung presented a comprehensive description of this wrist and forearm deformity.¹ As has been pointed out by many authors, including Madelung himself, he was not the first to describe the condition. Kelikian's exhaustive review of the history of Madelung's deformity lists at least six authors, including Dupuytren, whose descriptions predated Madelung's.⁴

An anterior-ulnar bowing of the radius and a dorsally prominent ulna may be seen in conjunction with trauma, infection, multiple enchondromatosis, and in families with Leri-Weil mesomelic dwarfism. Although some favor reserving the eponym for cases unrelated to these entities, it is common practice to use the term for any anterior-ulnar distal radial bow with a prominent distal ulna.

Etiology. The etiology of true Madelung's deformity is unknown. In spite of its frequent classification as congenital, there is no proof for a defect that is present at birth or even in early childhood. The deformity is usually recognized in late adolescence. Relatively deficient growth in that portion of the growth plate on the anterior-ulnar corner of the radius would seem important in the deformity. However, no convincing explanation exists for why this should occur in adolescence. Although the growth rate increases in this age group, it pales in comparison with growth of a child in the first 3 years of life—a time when Madelung's deformity is not seen. Support for a systemic cause acting locally is provided by the fact that the condition is twice as likely to be bilateral as unilateral, and four times more common in girls than in boys.⁴

The Madelung's deformity associated with Leri-Weil dwarfism has recently been shown to be associated with a specific genetic defect in the homeobox gene known as *Shox*.^{1a}

Pathology. In 1989, Vickers and Nielsen described a thick fibrous structure that begins on the ulnovolar metaphyseal region of the radius and attaches to the lunare and triangular fibrocartilage in Madelung's deformity.¹³ Evidence of this ligament is seen on radiographs, where its fossalike origin causes a small spur to appear on the radius and a flamelike radiolucency distal to the spur (Fig. 14–58). We have found

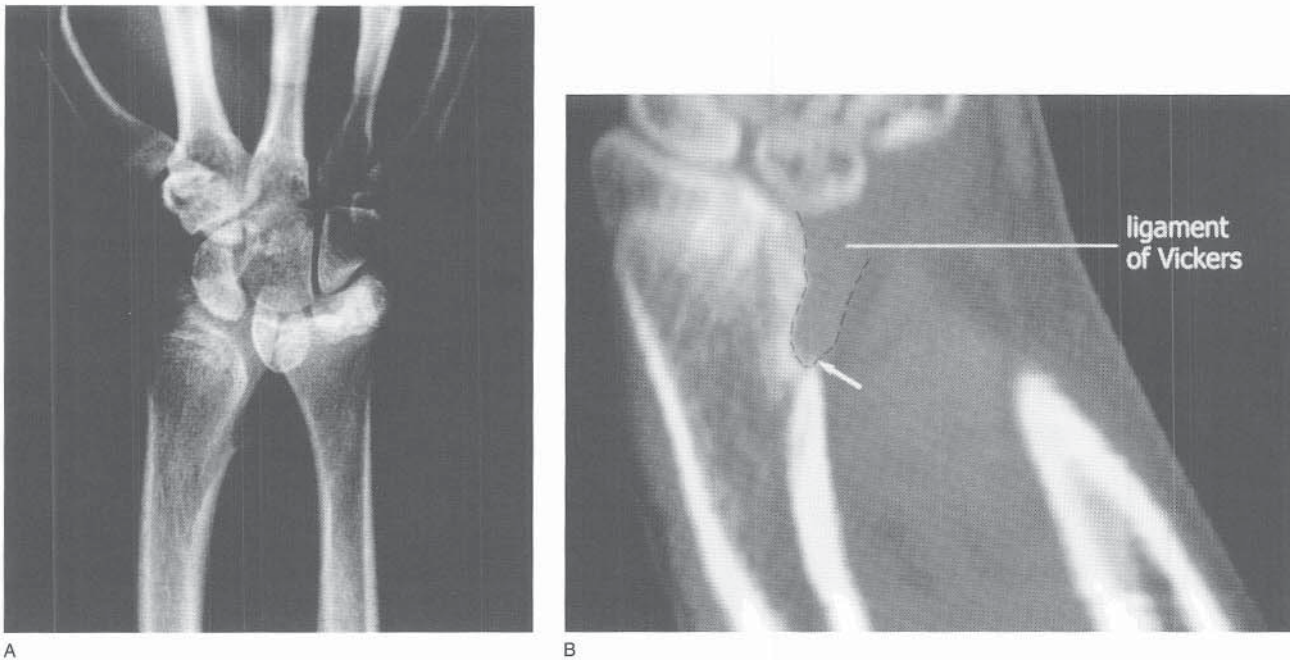


FIGURE 14–58 The ligament described by Vickers in Madelung’s deformity. **A**, Radiographic evidence of the ligament. Its fossalike origin causes a small spur on the radius and a flamelike radiolucency distal to the spur. **B**, MR image demonstrating Vickers’s ligament and the bone spur (*arrow*).

this structure at operation in almost every case of true Madelung’s deformity. Histologically, the ligament has elements of fibrous tissue and fibrocartilage with some areas of hyaline cartilage. Grossly, the ligament is a large fibrous band about 5 to 7 mm thick. It is found under the pronator quadratus, originating well proximal to most of the physis in a fossalike area on the ulnar side of the anterior corner of the radius. From here it flows out onto the anterior surface of the lunate, forming an insertion like that of the radiolunate ligament in the normal wrist. Portions of the ligament insert into the anterior radioulnar ligament portion of the triangular fibrocartilage. It may in fact be a stretched-out coalescence of these normal structures formed as a consequence of delayed growth of the radial physis beneath it. It seems unlikely that this ligament could be the primary cause of Madelung’s deformity and present since birth, because the enormous growth of the child during the first 3 years is never associated with the deformity. The importance of releasing this ligament when reconstructing these wrists is discussed under Treatment, below.

Clinical Presentation. The clinical description by early surgeons was careful and elegant. Here is Madelung in his

original text describing the deformity: “The deformity is most noticeable looking at the subluxation from the ulnar side. The forearm is apparently normally formed. The distal end of the ulna is distinct under the normal, though rather tense skin and the styloid process and articular surface can be recognized with the eye and encircled by the finger. The hand viewed on its own is normal but has dropped forwards. The diameter of the wrist is almost twice normal. The hand, viewed from the radial side, is less obviously displaced forwards. The extensor tendons, which pass over the radius towards the dorsum of the hand, bridge and obscure the step that was so noticeable on the ulnar side.”¹² Although most patients present in mid- to late adolescence, the smooth and symmetric nature of the bony changes suggests that the sequence of events leading to the deformity began in the preadolescent years.

The slowly progressive nature of this process is insidious and often is not noticed until a traumatic event draws attention to the prominent ulna (Fig. 14–59). The patient may present to the orthopaedist complaining of “dislocation” of the ulna. The compensatory remodeling by bony and ligamentous components of the radioulnar joint during the

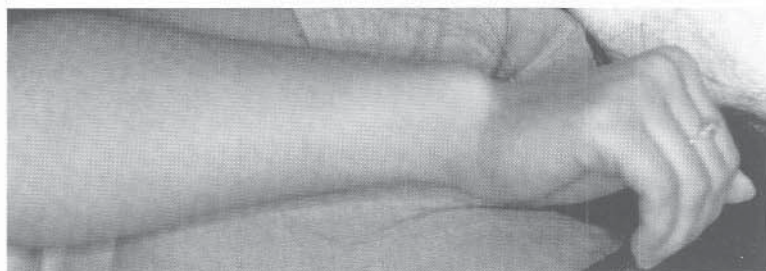


FIGURE 14–59 Madelung’s deformity is most noticeable when the examiner observes the subluxation from the ulnar side.

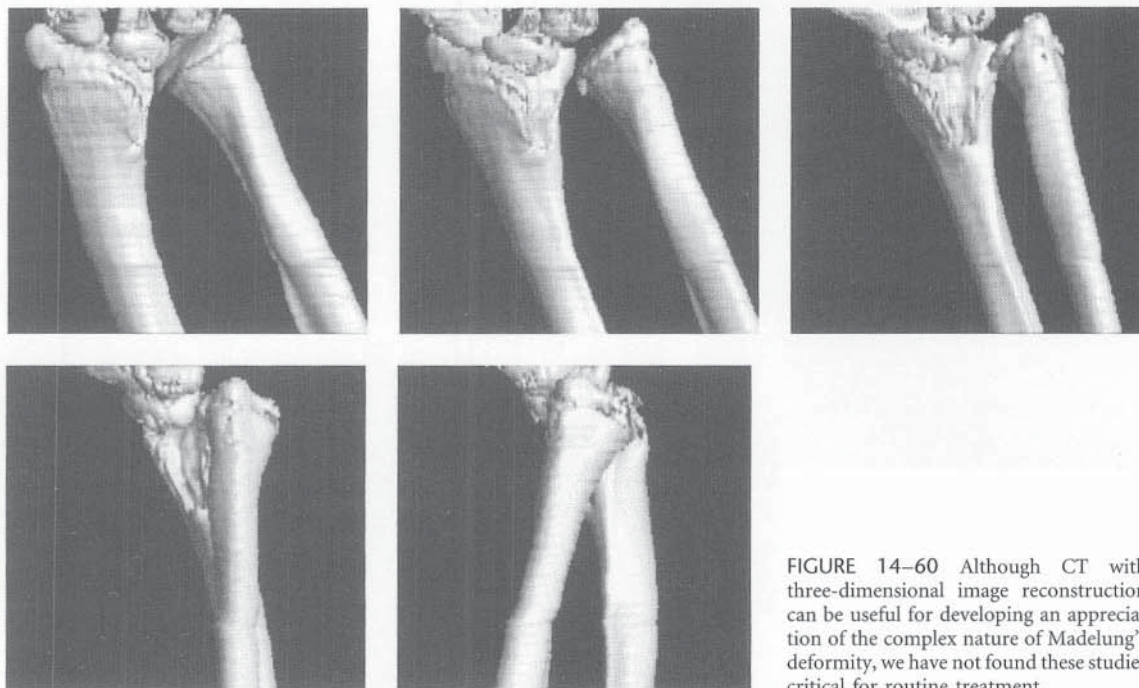


FIGURE 14-60 Although CT with three-dimensional image reconstruction can be useful for developing an appreciation of the complex nature of Madelung's deformity, we have not found these studies critical for routine treatment.

slow development of the deformity complicates reconstruction in late cases. This remodeling often facilitates hand pronation and supination in the presence of significant limitation of forearm motion. The presenting complaint is usually pain over the bony prominence of the ulna that is worse with activity. The appearance of the wrist is usually an important reason for seeking treatment, but often the patient does not immediately volunteer this information.

Imaging. The deformity is very much a three-dimensional one, a fact that cannot be completely appreciated on the standard radiograph. Although CT with three-dimensional image reconstruction can be useful for teaching an appreciation of the complex nature of the deformity, we have not found these studies critical in the routine treatment of Madelung's deformity (Fig. 14-60).

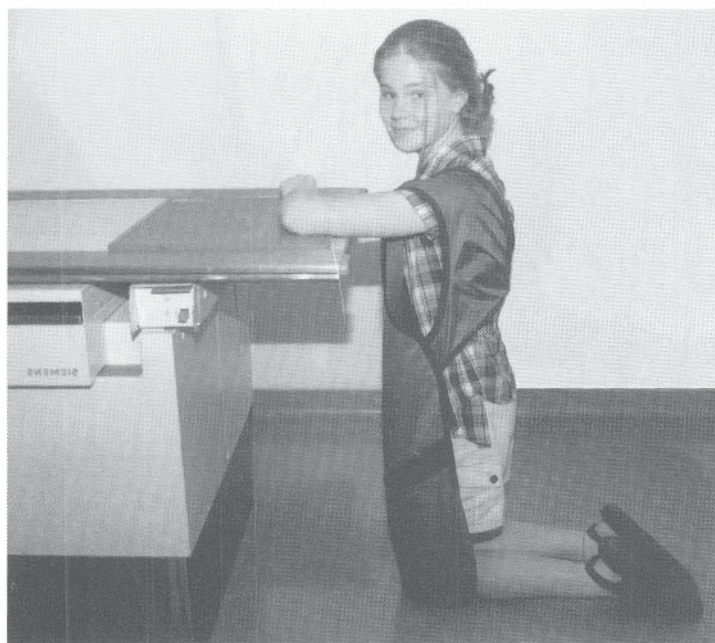
The regular x-ray study is the essential clinical tool, but a more detailed description of the technique and findings needs emphasis. When the standard PA radiograph is obtained, the forearm is in a variable amount of pronation, and the relative shortening of the radius with respect to the ulna may vary. A more clinically useful and reproducible technique is the bilateral, true PA neutral radiograph positioned on the same x-ray plate. The comparison right and left lateral views are then exposed on a second plate (Fig. 14-61). These simple views are easily reproduced by radiology department technicians on subsequent follow-up visits. This not only helps assess the deformity but, in addition, prevents overlooking a milder deformity in the asymptomatic but frequently involved opposite wrist. This technique is also useful in planning the osteotomy as well as explaining it to the patient and parents preoperatively. When the case is truly unilateral, this technique facilitates the postoperative evaluation of correction. Finally, putting the right and left arms on the same film reduces the number of films stored in the packet and keeps exposures made on the same day

together. The bilateral PA film is most useful in the operating room for reference.

The radiographic findings represent a continuum and are less severe in early cases. On the PA radiograph, a dramatically increasing radial tilt develops. As this tilt becomes severe, the lunate follows the ulnar-volar corner of the radius to eventually lie interposed in the expanding space between the radius and ulna as the growth of the ulna carries it away from the lunate facet of the radius. Rather than being in its normal position, resting distally and contained and supported by the radius and ulna, the lunate appears to be wedged between the two forearm bones. The radial epiphysis becomes teardrop in shape and the growth plate is at such an angle that on the ulnar side it may disappear from view on the PA radiograph. There is a marked ulnar plus variance (long ulna). On the lateral view, the radius tilts volarward until the ulna is shown dislocated from its normal articulation with the radius. The ulna finally arrives dorsal to and resting on top of the ulnar side of the proximal carpal row.

Occasionally a reverse Madelung's deformity is seen. In these cases the growth disturbance in the ulnar portion of the distal radial epiphysis is posterior rather than anterior and the typical radial bow is reversed, leading to anterior dislocation of the ulna and a dorsal displacement of the hand. In other unusual cases, the growth disturbance is more central and the bowing is less significant than the wedge-shaped or so-called chevron carpus.¹³

Treatment. There is controversy over the optimal treatment of these young teenagers. Some have suggested that the condition is often self-limiting and not sufficiently symptomatic to warrant surgical treatment.¹ However, most surgeons now accept that many of these patients do have significant and prolonged pain in adolescence and on into adulthood. Certainly the deformity is often considered a significant cosmetic problem for both men and women.



A



B

FIGURE 14-61 A, Radiographic positioning of the wrist and arm in Madelung's deformity. The true PA neutral radiograph of both wrists is positioned on the same x-ray plate. These simple views are easily reproduced by the radiology department technicians at subsequent visits. B, The resultant radiograph with both right and left wrists on the same plate. The comparison right and left lateral views are also combined on a single plate.

Previously, treatment usually included a Darrach or some other modification of ulnar resection arthroplasty and a tendency to ignore the fact that it is the radius that is deformed. Thus, earlier surgeons rarely performed an osteotomy of the radius until the deformity was severe.¹¹ Occasionally some have used the Suave-Kapangi (Lowenstein) technique of distal radioulnar joint arthrodesis combined with simultaneous distal ulnar osteotomy, creating a pseudarthrosis.¹⁴ Only the most enthusiastic proponents of the Ilizarov technique have published their results in the literature.¹⁰

Recently, Vickers has stimulated interest in Langenskiöld's epiphysiolysis of the abnormal ulnovolar corner of the distal radial epiphyseal plate using fat interposition.^{5,13} Linscheid described using Silastic instead of fat as interposition material.⁷

Although we have performed epiphysiolysis in a few patients, we have been unable to correct the deformity adequately in most cases with this technique alone. Our limited success with epiphysiolysis is explained by the following reasons. Peripheral lysis of any epiphyseal bars is less effective than lysis of lesions in the center of the epiphysis. Cases of Madelung's deformity are the result of a focal and almost always intensely peripheral lesion of the distal radial epiphysis. Second, most of our patients have presented in late adolescence, when they are finally significantly symptomatic and with the fully developed deformity. These patients are usually girls, who are at the end or close to the end of normal

growth by age 13. Perhaps, especially in those with Leri-Weil mesomelic dwarfism, we are not forceful enough in insisting that the parents allow us to follow the younger siblings of our patients. Vickers, a strong advocate of epiphysiolysis, is rigorous about following the young siblings of his Leri-Weil patients carefully, and a significant proportion of his cases in young children have been discovered this way (D. Vickers, personal communication, 1999).

Finally, the radial epiphyseal plate appears open on the radiograph for some time after it no longer contributes to the growth of the radius.³ Lysis of the epiphyseal bar in these cases could not be expected to allow correction of significant deformity. Therefore, our usual treatment recommendation for patients who are seen with significant deformity and symptoms has been release of the ligament described by Vickers, combined with a dome osteotomy of the metaphysis of the radius (Fig. 14-62). This requires an anterior approach to the radius, since the ligament cannot be appreciated unless the anterior surface of the radius beneath the pronator quadratus is exposed (Fig. 14-62A). The usual dorsal exposure fails to release this ligamentous tether, which connects the proximal fragment of the osteotomized radius to the proximal carpal row and triangular fibrocartilage ligamentous complex. Containment of the proximal carpal row by the radius can more easily occur after proximal release of the ligament because the release allows the distal radial fragment to rotate under the carpus. This rotation of the orientation of the lunate facet occurs in three dimensions

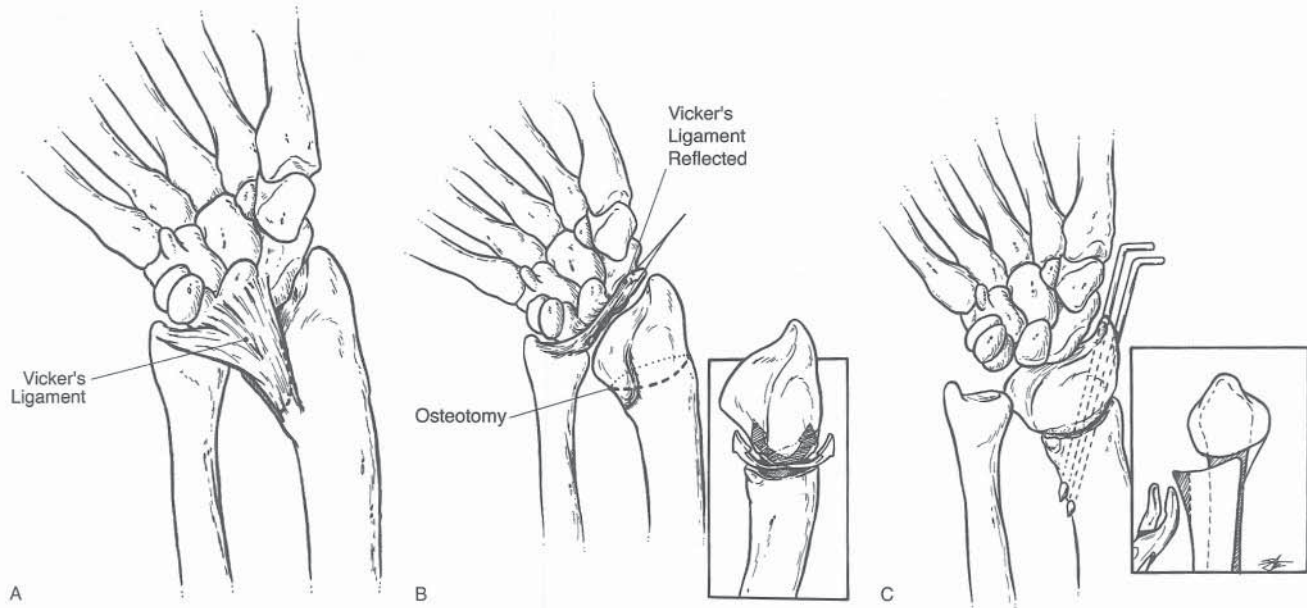


FIGURE 14-62 Our usual treatment recommendation for patients with significant symptomatic Madelung's deformity is release of the ligament described by Vickers combined with a dome osteotomy of the metaphysis of the radius through an anterior approach. **A**, The anterior aspect of the wrist and the ligament described by Vickers. The *dotted line* shows the initial incision in the ligament and where the distal dissection is to begin. **B**, Vicker's ligament detached from the fossa in the radius and reflected distally. The ligament has been dissected until the radiocarpal joint is opened and the lunate and scaphoid are seen. The *dotted line* on the radius is the site of the dome osteotomy. **C**, The osteotomy and repositioned intercalated distal fragment of the radius. The dome shape of the osteotomy allows correction in three planes. This simultaneously improves the containment of the carpus, corrects the intense obliquity of the radiocarpal joint, and resolves the apparent ulnar positive relationship seen on the PA radiograph. *Insert*: Lateral view after repositioning the intercalated distal radial fragment and the beak of anterior bone that results. This beak may be trimmed and used for local bone grafting after fixation. Percutaneous pin fixation with two small Steinmann pins rather than plate fixation has been adequate in our patients when supplemented for 6 weeks with a long-arm cast. This eliminates the need for a radius plate and its later removal.

from an anterior ulnar stance to a more dorsal radial position. The dome nature of the osteotomy seems to facilitate this more three-dimensional movement of the intercalated osteotomized distal radial fragment and better realignment of the wrist and hand on the forearm (Figs. 14-62B and C). Failing to release the ligament described by Vickers may be a cause for incomplete and unsatisfactory resolution of symptoms. Vickers has noted that release of the ligament alone can relieve the symptoms of pain prior to correction of the deformity of the distal radius following epiphysiolysis (D. Vickers, personal communication, 1999). Pin fixation using a small Steinmann pin rather than plate fixation has been adequate in our patients as long as it is supplemented for 6 weeks with immobilization in a long-arm cast. This eliminates the need for a radius plate and its later potential need for removal.

After the corrective osteotomy, the distal radioulnar joint is incongruous, but the ligamentous laxity in these patients seems to work to their advantage in this regard. Usually forearm rotation is incomplete but painless after the osteotomy has healed, but wrist ligament laxity allows the hand to supinate and pronate through a functional range. Patients are pleased that the clinical deformity of the ulnar prominence is well corrected by our combination of dome osteotomy and release of Vicker's ligament (Fig. 14-63). Perhaps the pain relief they report is explained more by the release of the ligamentous tether than by the correction of the malaligned hand-forearm relationship. Only occasionally has ulnar shortening been required, when there was a marked ulnar positive variance. Ulnar shortening is best done later, after the radius has healed and early forearm rotation exercises have restored function.

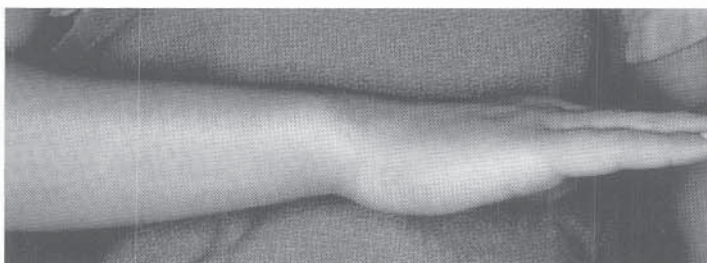


FIGURE 14-63 Postoperative appearance of the patient with Madelung's deformity whose preoperative wrist was shown in Figure 14-59. The clinical deformity of the ulnar prominence is well corrected by the combination of dome osteotomy and release of the ligament described by Vickers. Only occasionally has ulnar shortening been required, when there was a marked ulnar positive variance. Resection of the distal ulna may rarely be necessary, when the forearm rotation is severely limited.

Rarely, resection of the distal ulna and ligament reconstruction may be necessary when the forearm rotation is severely limited.

Results. The results of reconstructive surgery for Madelung's deformity suggest that the cosmetic deformity can usually be minimized and the pain relieved or improved. Moderately long-term results have been reported.^{9,11} Operations that do not correct the appearance of the wrist are rarely associated with a satisfied patient.

Complications. Neurologic complications are rarely seen. Carpal tunnel syndrome was reported by Luchetti and colleagues.⁸ Ulnar nerve compression with reverse Madelung's deformity was reported by Fagg.² Keinbock's avascular necrosis of the lunate has been associated with Madelung's deformity.^{6(p 335)} This is in contradistinction to the association of Keinbock's AVN of the lunate with a short rather than a long ulna.

Surgical complications related particularly to operative procedures on patients with Madelung's deformity include recurrence or incomplete correction of the deformity and distal radioulnar loss of motion or pain. Recurrence is more common in younger patients and those treated with epiphysiolytic procedures rather than osteotomy. Incomplete or incorrect correction of the alignment of the hand on the forearm may be related to lack of release of the anterior ligament described by Vickers.

REFERENCES

Madelung's Deformity

1. Dobyns JH: Madelung's deformity. In Green DP (ed): *Operative Hand Surgery*, p 515. New York, Churchill Livingstone, 1993.
2. Fagg PS: Reverse Madelung's deformity with nerve compression. *J Hand Surg* 1988;13-B:23.
3. Greulich P: *Radiographic Atlas of Skeletal Development of the Hand and Wrist*. Stanford, CA, Stanford University Press, 1959.
4. Kelikian H: *Congenital Deformities of the Hand and Forearm*, p 753. Philadelphia, WB Saunders Co, 1974.
5. Langenskiöld A: The possibilities of eliminating premature partial closure of an epiphyseal plate caused by trauma or disease. *Acta Orthop Scand* 1967;38:267.
6. Lichtman D: *The Wrist and Its Disorders*, p 335. Philadelphia, WB Saunders Co, 1988.
7. Linscheid RL: Madelung's deformity. *American Society for Surgery of the Hand Correspondence Newsletter* 1979.
8. Luchetti R, Mingione A, Monteleone M, et al: Carpal tunnel syndrome in Madelung's deformity. *J Hand Surg* 1988;13-B:19.
9. Nielsen J: Madelung's deformity: a follow up study of 26 cases and a review of literature. *Acta Orthop Scand* 1977;48:379.
10. Paley D: Ilizarov technique: application in congenital deformities of the upper limb. In *Congenital Anomalies of the Upper Limb*. Lahaina, Maui, Hawaii, 1991.
11. Ranawat CS, De Fiore J, Straub LR: Madelung's deformity: an end-result study of surgical treatment. *J Bone Joint Surg* 1975;57-A:772.
12. Rang M: *Anthology of Orthopaedics*, p 36. Edinburgh, E & S Livingstone, 1966.
13. Vickers D, Nielsen G: Madelung deformity: surgical prophylaxis (physiolytic) during the late growth period by resection of the dyschondrosteosis lesion. *J Hand Surg* 1992;17-B:401.
14. White GM, Weiland AJ: Madelung's deformity: treatment by osteotomy of the radius and Lauenstein procedure. *J Hand Surg* 1987;12-A:202.

ADDED REFERENCE

Madelung's Deformity

- 1a. Blaschke RJ, Rappold GA: SHOX: growth, Leri-Weill and Turner syndromes. *Trends Endocrinol Metab* 2000;11(6):227.

POLYDACTYLY

History. Polydactyly, the most common deformity of the human hand, has been recorded in literature since Biblical times, beginning in the eleventh century B.C. In one tribe the inheritance pattern was so complete that absence of the deformity was considered a sign of illegitimacy.¹⁵ In England, polydactyly was once considered a sign of royalty. Queen Anne Boleyn had a sixth finger on her left hand, which "was to her an occasion of additional grace by the skillful manner in which she concealed it from observation."²⁵ Even today, as the child with an extra floppy digit grows older, the patient and family often develop a morbid attachment to and refuse deletion of the useless part. In our society, for both functional and social reasons, a pentadactylous hand is a reasonable goal of surgery. Many times this represents no more than a simple ligature of the floating, useless extra digit. At other times, particularly in central polydactyly, a five-digit hand must be abandoned to give the child the most functioning mobile hand possible.

Etiology. A genetic cause of polydactyly is well established in some cases and absent in others. In ulnar-sided polydactyly in blacks, the inheritance pattern is strongly that of a dominant gene with variable penetrance. Central polydactyly in whites often shows a dominant inheritance pattern. The condition in Caucasians is associated with other visceral abnormalities and syndromes so often that Flatt has said, "An extra digit on the hand of a newborn should signal the need for a complete and thorough physical workup since it may indicate concealed malformations. . . ."²⁵ Although this is true in Caucasians with polydactyly, Upton points out that an associated abnormality is rarely present in the much more common postaxial (ulnar or little fingers) polydactyly of blacks.¹⁵ Syndromal associations, when they do occur, may involve virtually every organ system, producing chromosomal, orofacial, skin, and eye abnormalities, bone dysplasias, and mental retardation (Ellis-van Creveld syndrome, Laurence-Moon-Biedl syndromes, trisomy 13). Autosomal recessive genetic associations are seen as well (Blackfan-Diamond anemia, Bloom's syndrome, Carpenter's syndrome).³

Triphalangeal thumbs, often seen in association and reasonably grouped with radial polydactyly, have been related to thalidomide ingestion by the mother between the 45th and 50th day of gestation.¹⁷

More recent embryologic research on the homeobox (Hox) genes responsible for development of each digital ray has shown these genes to be abnormal in cases of synpolydactyly.⁹

Pathology. Polydactyly is an abnormality in the longitudinal segmentation of the limb bud that manifests not as an additional amount of the substrate but as an increase in the folding of the apical ectodermal ridge—a "ruffle" in the hand plate. The tissues present vary enormously and may contain only a small soft tissue packet attached by a narrow stalk or a completely formed appendage with normal bones, joints, tendons, and neurovascular structures. A classification system based on this spectrum was presented by Stelling and Turek in the 1960s.^{12,14}

- Type I: No skeletal attachment
- Type II: Skeletal attachment to an enlarged or bifid phalanx or metacarpal
- Type III: Complete duplication including a normal metacarpal (rarest)

Radial polydactyly is relatively common and deserves special consideration since the treatment of thumb duplication is so critical and reconstruction is more complex than reconstruction of the typical ulnar polydactyly. Therefore, for useful treatment reasons, hand surgeons have further subclassified these patients.

Clinical Presentation

INCIDENCE. The true incidence of polydactyly is difficult to establish, since extra digits are often amputated in newborns by obstetricians and pediatricians. The condition is considerably more common in blacks than whites (blacks, 1 in 300, whites, 1 in 3,000). In whites the duplication is more common on the radial or preaxial border (thumb), while in blacks, duplications of the ulnar or postaxial border (small finger) are much more common. In Flatt's Iowa series, polydactyly accounted for almost 15 percent of his cases of congenital anomalies.⁵

Clinical Appearance and Classification Systems

RADIAL POLYDACTYLY. In thumb duplications, neither of the two thumbs will be equal in size to a normal thumb, even when they are grossly different from each other. Pointing this out to the parent before surgery is important so the parent will not mistakenly believe that the corrective surgical procedure has made the thumb smaller or grow more slowly than normal. In addition, the surgical repair of radial polydactyly is usually more involved than the surgical repair of ulnar duplications. On the radial side, thumb reconstruction

TABLE 14-6 Ezaki's Classification of Polydactyly

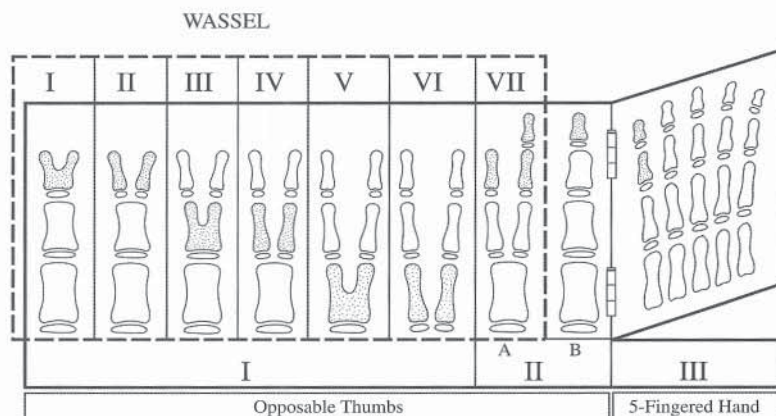
Type I radial polydactyly: Includes the first six types of Wassel
Type II radial polydactyly: Includes two varieties of opposable thumbs
A. A duplicated thumb with a triphalangeal member (Wassel's type VII)
B. A nonduplicated triphalangeal thumb
Type III radial polydactyly: A nonopposable duplication of the index finger—the true five-fingered hand

describes the procedure well, since ligamentous reconstruction, angular correction, and tendon reconstruction are usually required to make the best of this most important digit. Although the thumbs may be symmetric or asymmetric, the tendons usually bifurcate from a normal proximal tendon. The tendon insertions of both flexor and extensor are frequently eccentrically placed and must be repositioned and aligned by the reconstruction. Simple ablation, common in ulnar polydactyly, can destroy the possibility for quality hand function in thumb duplication.

Wassel's classification (based on his analysis of Flatt's cases) is frequently used with the radial polydactylies (Fig. 14-64).¹⁶ The relative frequency of each type is as follows:

- Type I = 2%
- Type II = 15%
- Type III = 6%
- Type IV = 43%
- Type V = 10%
- Type VI = 4%
- Type VII = 20%¹⁶

The distinction between types I through VI is straightforward and is based simply on the level of the duplication being either at or between joints. Type VII is a more complex



McKUSICK & TEMTAMY

FIGURE 14-64 Radial polydactyly is relatively common and deserves special consideration because the treatment of thumb duplication is critical and reconstruction is more complex than reconstruction of the typical ulnar polydactyly. Therefore, for useful treatment reasons, hand surgeons have further subclassified these patients. The figure shows the relationship of two commonly used classification systems. By studying Flatt's⁵ patients, Wassel¹⁶ developed seven types and reported their frequency: type I—2%; type II—15%; type III—6%; type IV—43%; type V—10%; type VI—4%; type VII—20%. The geneticists Temtamy and McKusick divided the preaxial polydactylies into three groups. This classification is particularly useful clinically because of the correlation of the triphalangeal thumbs with and without duplication and the true five-fingered hand. Temtamy and McKusick's type I radial polydactyly includes the first six types of Wassel. Type II radial polydactyly includes two varieties of opposable thumbs: IIA—duplicated thumbs with a triphalangeal member (Wassel's type VII), and IIB—nonduplicated triphalangeal thumbs. Type III radial polydactyly includes nonopposable duplication of the index finger, also known as a five-fingered hand.^{3,13}

category that requires the existence of at least one triphalangeal thumb. Distinguishing between a duplicated index finger and a triphalangeal thumb is sometimes difficult. An interesting bit of detective work has led one group to note that the fingerprint pattern between the normal thumb and the normal index finger is markedly different. The index fingerprint is characterized by a distinguishing pattern called the radial loop, which is not seen in the normal thumb. Triphalangeal thumbs have a high incidence of radial loops.¹¹

Because of this confusion, Ezaki has favored a modification of the geneticists' classification (Temtamy and McKusick¹⁴) in which the preaxial polydactylies are divided into three groups (Table 14-6).² This classification is particularly useful clinically because of its relevance to important treatment decisions (Fig. 14-64).

Associated Conditions. Triphalangeal thumbs are associated not only with thumb duplications but also with duplications of the great toe and with the cardiac pathology of Holt-Oram syndrome (atrial and ventricular septal defects, anomalous coronary arteries, great vessel anomalies, and patent ductus arteriosus). In Fanconi's pancytopenia, according to Wood, a triphalangeal thumb is always present,¹⁷ although we have seen the occasional case in which it is not.

CENTRAL POLYDACTYLY. Central polydactyly is much less common than a border polydactyly and is frequently seen in association with syndactyly (synpolydactyly). Central polydactyly is frequently bilateral and has an autosomal dominant inheritance pattern. Duplications of the three central rays are more common ulnarly, with index duplications being the rarest. The true incidence of central polydactyly is complicated by the fact that thumb duplications may be associated with triphalangeal thumbs. This can be occasionally confused with index duplication.

Central polydactyly, syndactyly, and cleft hand not infrequently occur together. Polydactylous elements may be buried in the hand of the patient with both central polydactyly and cleft hand (Fig. 14-65 and 14-66D). In patients with

a cleft hand on one side, a central polydactyly is often present on the other.⁸ Foot anomalies are present in many of these patients.

The classification system most commonly used for central polydactyly is a modification of the previously outlined one by Stelling and Turek:

Type I central polydactyly: No skeletal attachment of a soft tissue mass.

Type II central polydactyly: Duplication of a common metacarpal or phalanx.

Type II cases are further subdivided into A and B by the presence or absence of syndactyly.

Type IIA: No syndactyly (Fig. 14-66A).

Type IIB: Syndactyly to adjacent digits that often presents as an extra digit hidden within a syndactyly (Fig. 14-66B).

Type III central polydactyly: A complete duplication, including the metacarpal. This anomaly is rare (Fig. 14-66C).

ULNAR POLYDACTYLY. Duplication of the little finger is such a common anomaly that a system classifying it into two types has been used. Type A has a skeletal attachment (Fig. 14-67A). Type B is a floating digit attached by only a soft tissue stalk (Fig. 14-67B). Type B is strongly inherited in black patients in an autosomal dominant pattern.

MIRROR HAND. This fascinating and exceedingly rare condition is also known as ulnar dimelia, or mirror hand. Less than 80 cases were present in the literature when reviewed by Kelikian in 1974. Most reports are of isolated cases, and Adrian Flatt, one of the foremost and prolific writers on congenital hand anomalies, said he had never operated on a patient with mirror hand.⁵ In these patients a duplicated ulnar component entirely replaces the radial components from the elbow distally. Although there is no radius or thumb, two ulnas and a plethora of fingers are present. Mirror hands are rarely truly symmetric, and seven rather than eight fingers may be present (Fig. 14-68). The function of these very unusual upper limbs is often very limited,

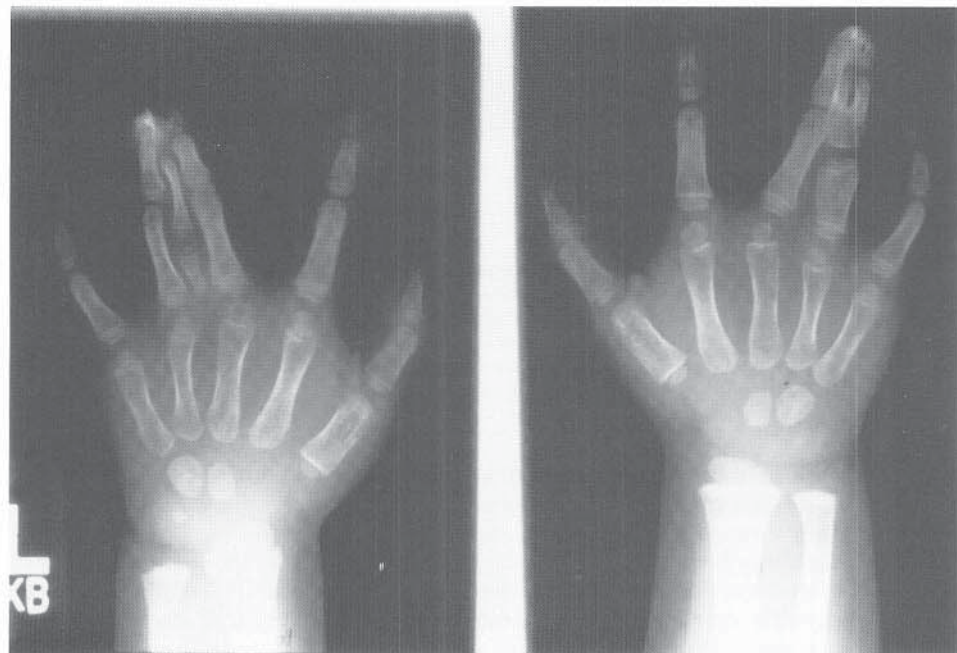


FIGURE 14-65 Central polydactyly, syndactyly, and cleft hands are related. Polydactylous elements may be buried in the hand of the patient with both central polydactyly and cleft hand.

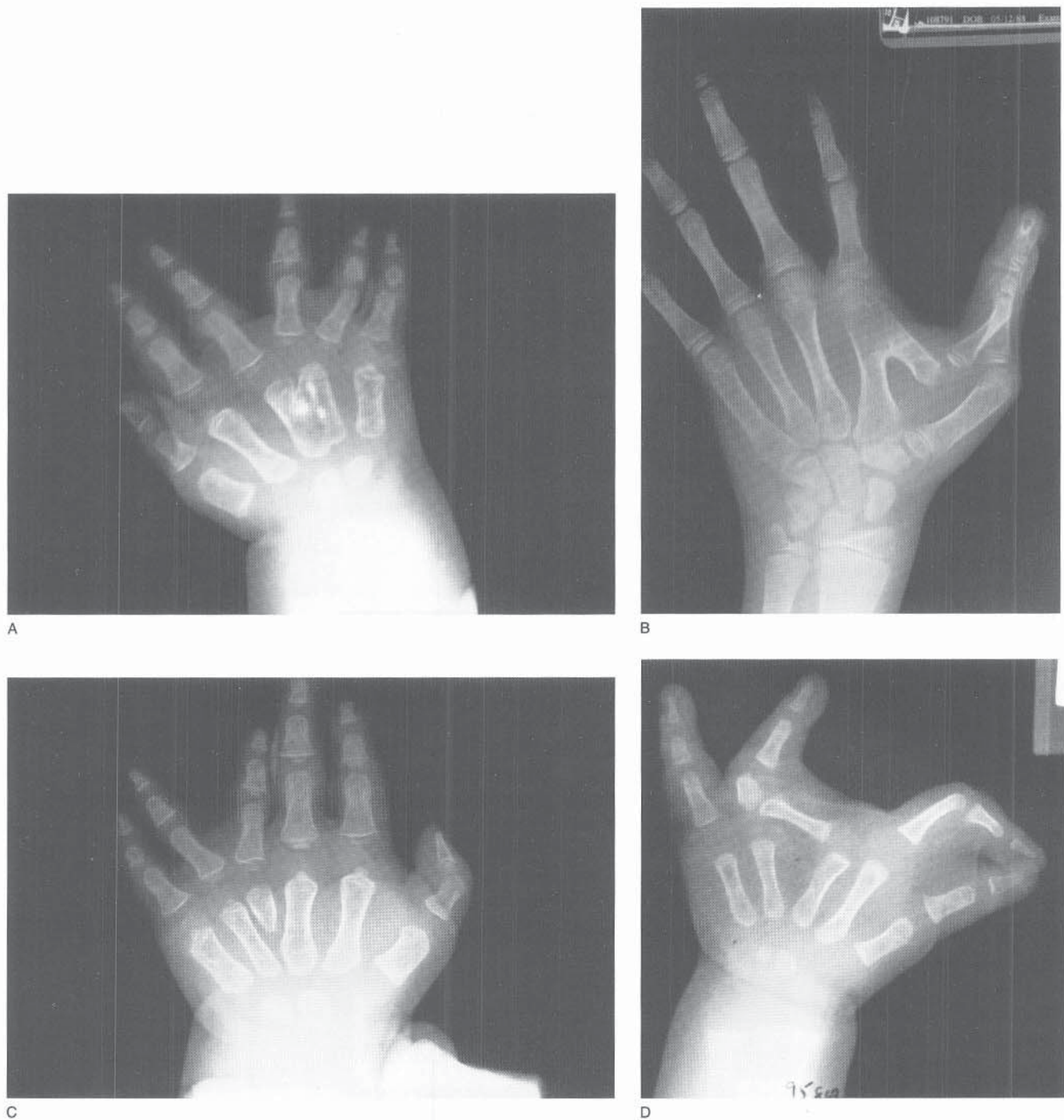


FIGURE 14-66 There are several types of central polydactyly. In type I central polydactyly there is no skeletal attachment of a soft tissue mass. (There is no illustration of this unusual type.) **A**, Type IIA central polydactyly, or duplication on a common metacarpal or phalanx without syndactyly. **B**, Type IIB central polydactyly, or duplication on a common metacarpal or phalanx with syndactyly to adjacent digits, which often manifests as an extra digit hidden within a syndactyly. **C**, Type III central polydactyly. A complete duplication, including the metacarpal, is a rare anomaly. **D**, Relationship of central polydactyly and cleft hand.

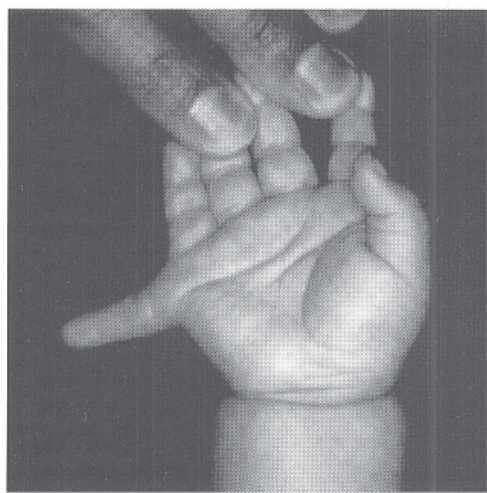
especially because of the associated abnormalities of the forearm and elbow. The condition may be seen with fibular dimelia in a family through an autosomal dominant inheritance. Mirror hand may be associated with tibial hemimelia. There are no known associated visceral anomalies.

The wrist and elbow are enlarged and the forearm may be short. The whole upper limb is frequently shorter because

of other associated proximal anomalies of the scapula, clavicle, humerus, and shoulder joint.

At the elbow the distal humerus is always markedly abnormal. The biceps and brachialis may fail to cross the elbow and instead insert into the distal humerus, resulting in a very stiff elbow with little function. A deformed pair of trochleae is present, and the capitellum is absent. The two

FIGURE 14-67 Ulnar polydactyly is classified into two types. Type A is broad-based and has a skeletal attachment (A). Type B is a floating digit attached by only a soft tissue stalk (B). Type B is strongly inherited in black patients in an autosomal dominant pattern.



A



B

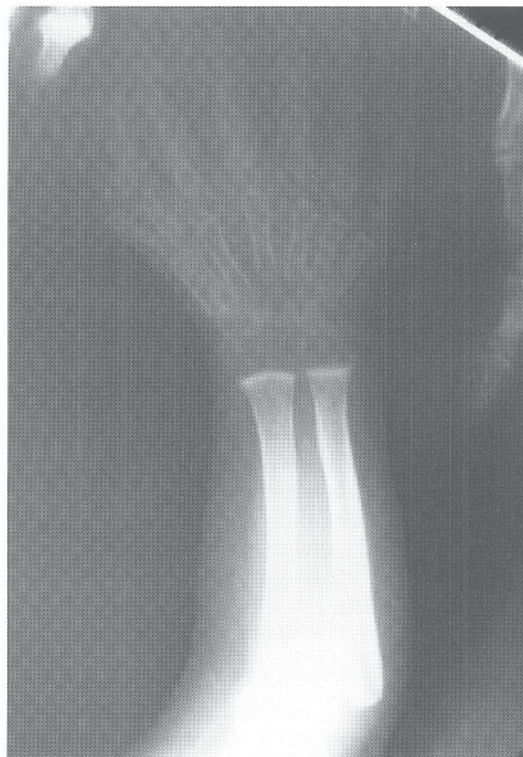
olecranon fossae of the ulnae virtually face one another at approximately 120 degrees. The distal ulnae are also abnormally enlarged compared to the ulna of a normal forearm. Elbow and forearm motion is severely limited because of these anomalies. The forearm muscles contain two sets of flexor and nonexistent or hypoplastic extensor muscles and tendons.

The carpal bones are also duplicated, with pairs of pisiforms, hamates, and triquetra. The lunates and capitates may either be paired or fused. There are no scaphoid, trapezium, trapezoid, or thumb elements. The wrist tends to lack extension and to be fixed in excessive flexion, resting in either radial or ulnar deviation.

In the hand and forearm the arterial pattern is one of



A



B

FIGURE 14-68 Mirror hand or ulnar dimelia, a fascinating and exceedingly rare condition. A and B, In these patients a duplicated ulnar component entirely replaces the radial components from the elbow distally. Although there is no radius or thumb, two ulnae and a plethora of fingers are present. Mirror hands are rarely truly symmetric, and seven rather than eight fingers may be present.

two ulnar arteries feeding a common superficial arch, with a pinwheel of common digital arteries feeding the finger cluster.

Although in copious supply, the fingers are often stiff, with poor intrinsic musculature, and may share bifurcated extrinsic flexor tendons. The ulnar or, more accurately, postaxial set of digits tends to be the most functional.

Embryologists have created the condition in chick embryos by transplanting the zone of proliferating activity (ZPA) to the preaxial (radial) side.¹⁰

Imaging. The PA radiograph is a critical tool in the evaluation and treatment of polydactyly. In the wiggly infant, not only can it be difficult to obtain, it may also be deceptive owing to lack of complete ossification of the bones. Nevertheless, prior to surgery a radiograph is essential in anything but the most simple type I case.

More sophisticated tests such as arteriography, MRI, and CT are not necessary.

Treatment. Treatment of the various forms of polydactyly varies from simple to complicated as the deformities become more complex. The discussion here focuses on principles of treatment and is presented in more detail only for the more common patterns of the anomaly. The reader is referred to hand surgical texts for more complex treatments of the rarer forms of duplication.^{2,5,7,15}

RADIAL POLYDACTYLY. The thumb is by far the most critical single element of the hand's anatomy, and in treating radial polydactyly, care must be taken not to simply delete the extra thumb. Only rarely is the treatment of preaxial polydactyly as simple as treatment of a type B postaxial polydactyly, but when a floating thumb with a narrow stalk is present, simple ligation can be done in infancy. The vast majority of thumb duplications have a broader-based attachment, and reconstruction should be deferred until some time between ages 6 and 18 months. In these cases, unless careful reconstruction of ligaments and tendons with appropriate fitting of joints is done, instability, unsightliness, and limited function of the remaining thumb will result. Even when one of the duplicated thumbs is larger and more appropriate for retention, the parent should be shown preoperatively that the duplicated thumb to be saved is still smaller than the normal contralateral thumb. This averts inappropriate parental concern after the operation that the treatment damaged the growth of the remaining thumb. Generally, the ulnar thumb is preserved when possible to save the normal ulnar collateral ligament, which must resist the power of pinch. When this is not possible, ligament reconstructions around the epiphyseal plate are limited, and late instability of the joint due to failure of the reconstructed ligament may occur. This may require later arthrodesis or more formal ligament reconstruction at the end of growth.

The McKusick and Temtamy type I or Wassel types I through VI (see Fig. 14–64) will be considered first.

Wassel Type I and II Thumbs. In these patients the distal phalanx is bifid, but a common joint is shared. When one thumb is much smaller, it can be simply abated. When the duplication has resulted in two markedly smaller thumbs of equal size, excision of one entire duplicated thumb leaves a small thumb with an unnatural appearance. The Bilhaut-Cloquet procedure or modifications of it have been used to effectively

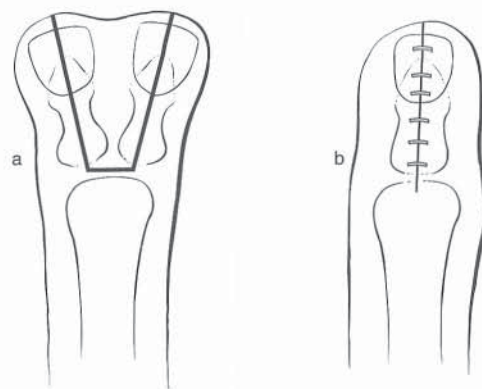


FIGURE 14–69 The Bilhaut-Cloquet procedure. When duplication has resulted in two smaller but equal-sized thumbs, excision of one entire duplicated thumb leaves a small thumb with an unnatural appearance. The Bilhaut-Cloquet procedure or modifications of it have been used effectively to resect the central portion and join the two.

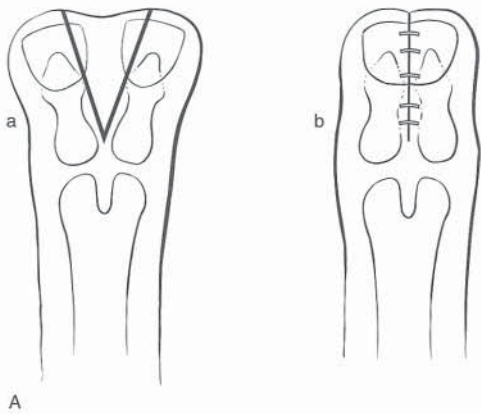
resect the central portion and join the two (Fig. 14–69).¹ The nail closure is critical. It may be possible to join the two distal phalanges, and the epiphysis can continue to grow. The tip of the thumb often remains broad and somewhat stubby but functional. Removal of the nail plate and careful, accurate closure of the sterile and germinal matrix is essential to provide the best nail later. Even then a groove or ridge is essentially always present. Soft tissue closure should preserve the outer margins of the pulp and its digital nerve supply. Sometimes the bony duplication is best left alone and reconstruction is confined to soft tissue and the nail (Fig. 14–70).

Wassel Type III Thumbs. When one thumb is smaller it is best deleted, but when the condition represents a truly bifid thumb, the Bilhaut-Cloquet operation may be possible. However, IP joint motion is usually limited.

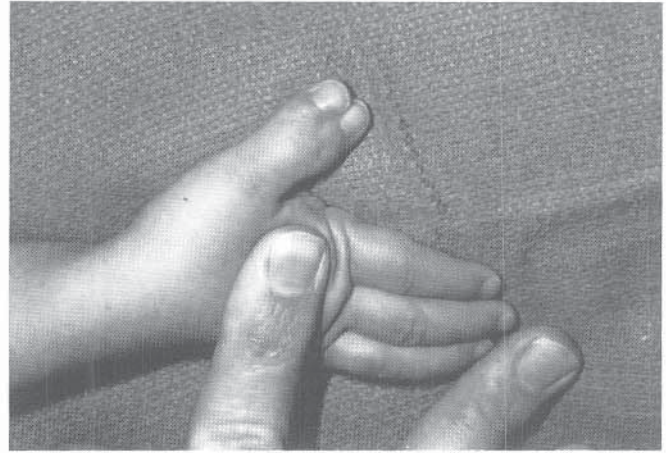
Wassel Type IV Thumbs. This is the most common variety, accounting for almost half (43 percent) of Flatt's original series, which was reviewed and classified by Wassel (Fig. 14–71). Here reconstruction of ligaments and tendons is especially critical. Unless the condyle of the metacarpal is reduced, reconstruction of the radial collateral ligament is compromised, and an unsightly bump rather than a more natural taper is left behind (Fig. 14–72).

For the Wassel type IV thumb, the incision should allow careful exposure of the extensor and flexor surfaces so that the tendon bifurcations and digital neurovascular structures can be well seen. Exposure usually entails a racquet-shaped incision of some type with extension proximally and distally (Fig. 14–73). The abductor pollicis brevis and some of its fibrous insertion should be identified and preserved. The extensor tendon to the ulnarmost thumb is detached from the abductor aponeurosis and as far distally as possible (Fig. 14–74). It is then reflected off the dorsal capsule of the MP joint of the two thumbs. The insertions of the flexor and extensor tendon into the remaining thumb's distal phalanx should be inspected for eccentric insertions and realigned. Failure to do so can result in a later zig-zag deformity of the thumb as the child grows (see Fig. 14–72).

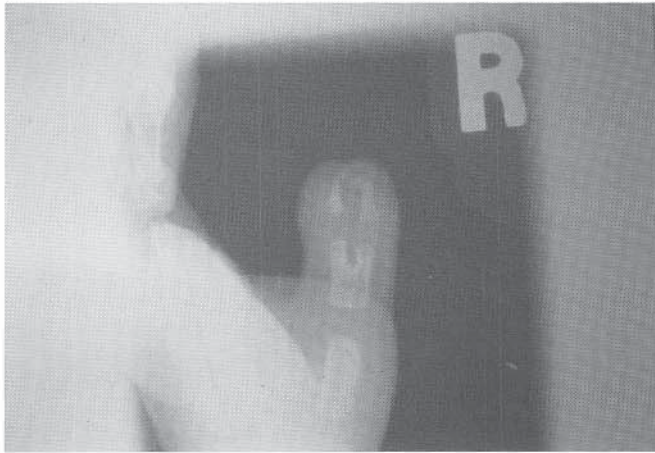
The next goal is to detach the radial collateral ligament as far distally as possible so as to preserve the ligament for reconstruction. This is conveniently done through a



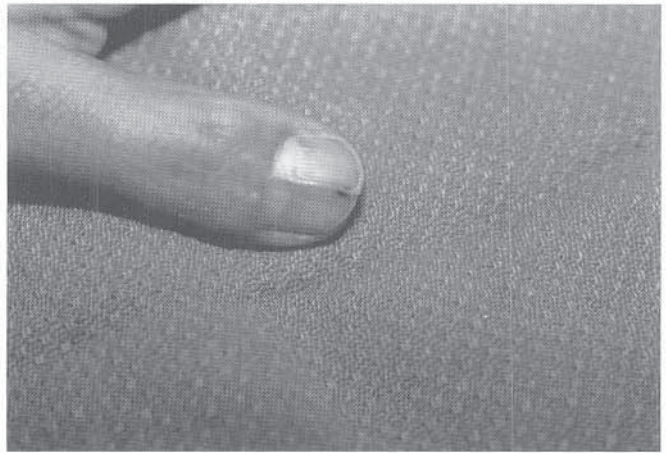
A



B



C



D

FIGURE 14-70 A, Sometimes the bony duplication is best left alone and reconstruction is confined to soft tissue and nail. B to D, A clinical example with 20-year follow-up.

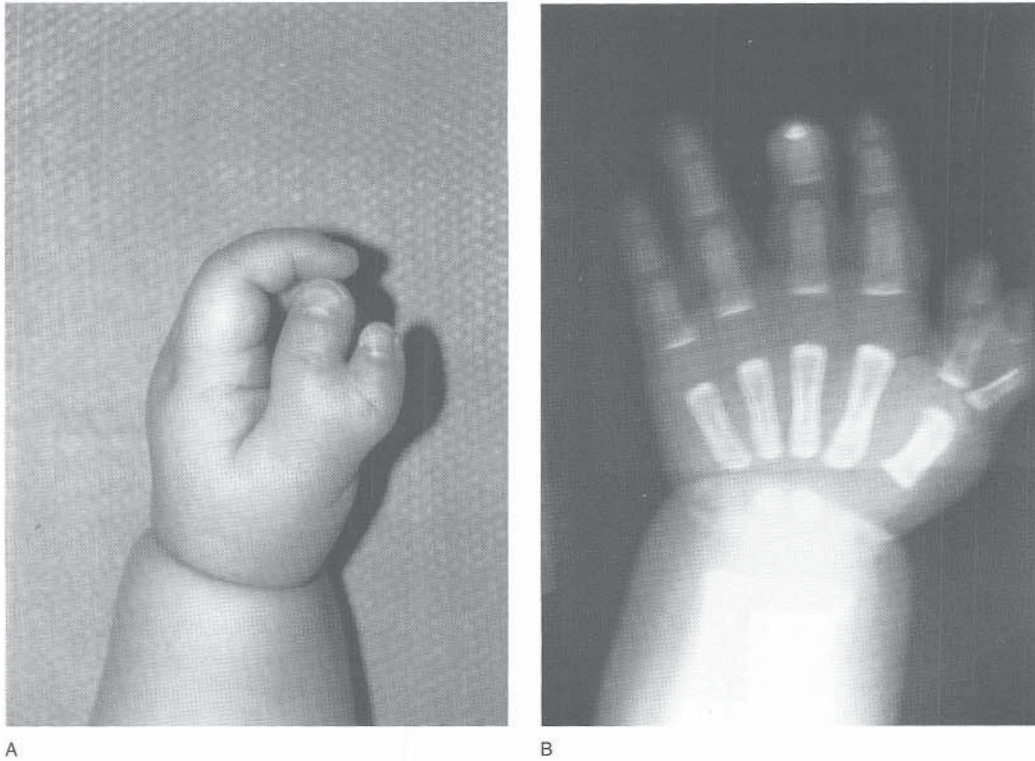


FIGURE 14-71 A and B, Wassel type IV is by far the most common of the duplicated thumbs. To help the surgeon avoid the problems of instability and get the best possible appearance and function from the thumb, the details of correct technique are illustrated in Figures 14-73 through 14-78.

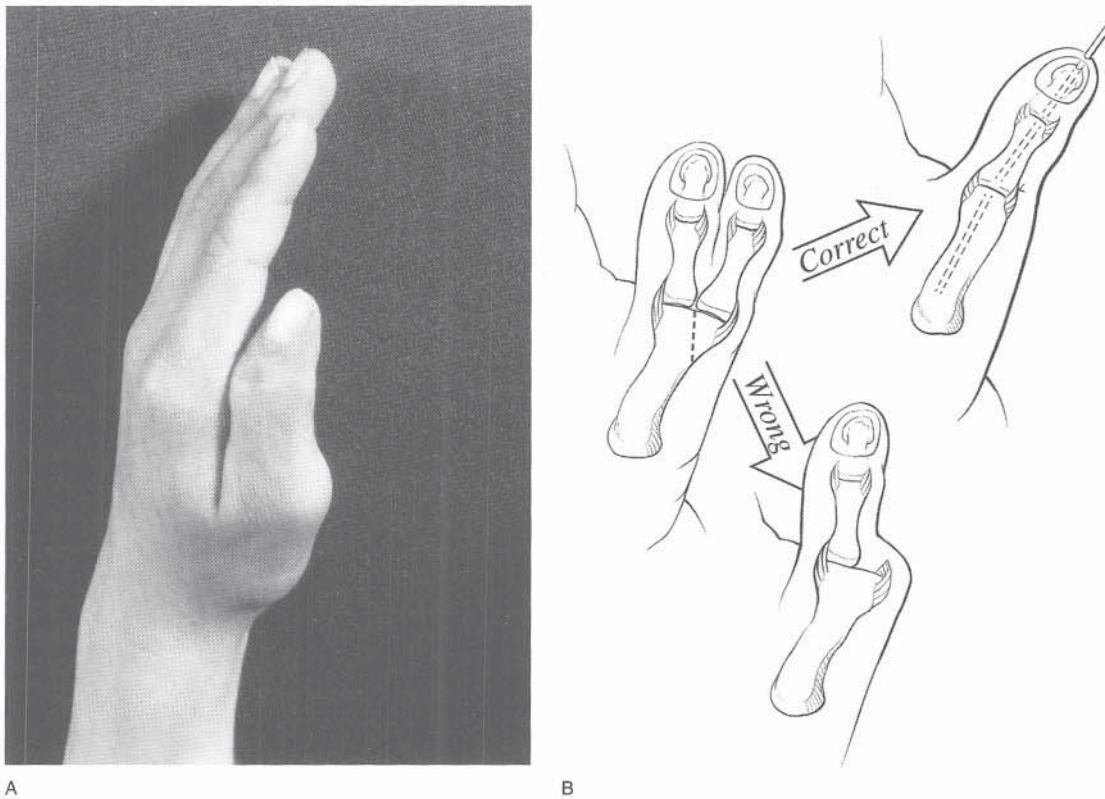
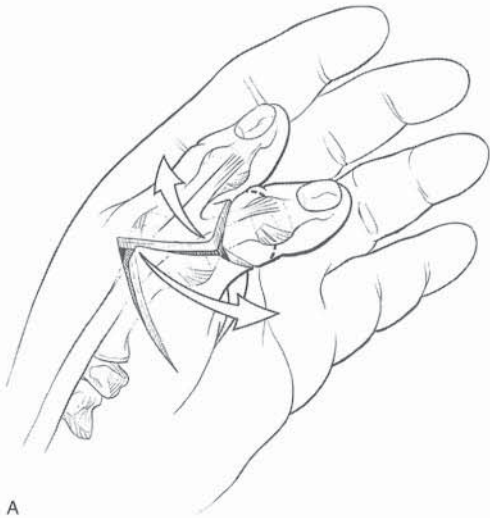
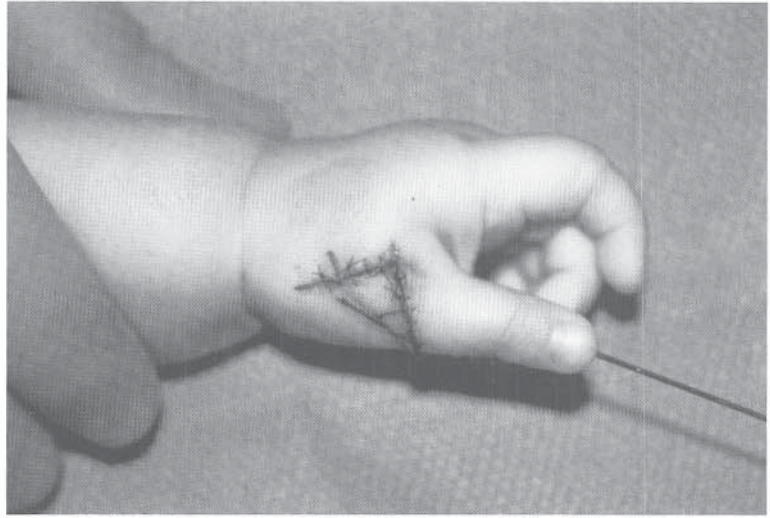


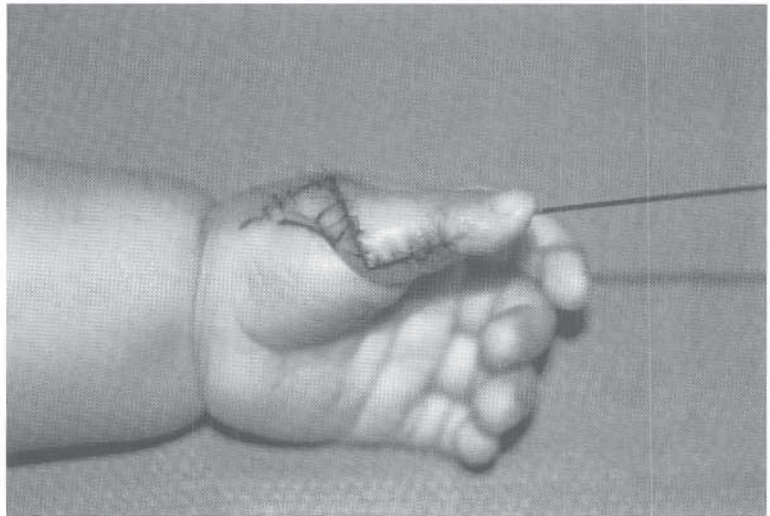
FIGURE 14-72 A, Unless the condyle of the metacarpal is reduced, an unsightly bump rather than a more natural taper is left behind. B, More important, reconstruction of the radial collateral ligament is compromised and instability of the joint is the result.



A



B



C

FIGURE 14–73 The incision should allow careful exposure of the extensor and flexor surfaces so that the tendon bifurcations and digital neurovascular structures can be well seen. Exposure usually entails a racquet-shaped incision of some type with extension proximally and distally. **A**, The resulting closure after reconstruction and removal of the duplicated thumb follows the hand surgery principles for incision design and avoids the problem of hypertrophy of the surgical incisional scars. **B** and **C**, Closure of this incision in the patient whose hand is shown in Figure 14–72.

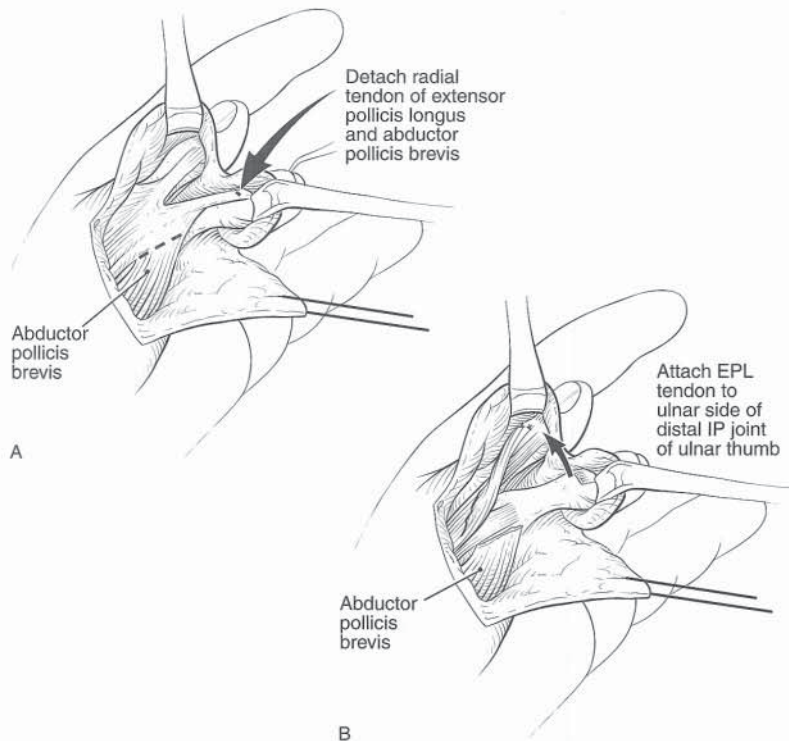


FIGURE 14-74 The abductor pollicis brevis and its tendinous insertion should be identified and preserved. **A**, The extensor tendon to the ulnar thumb is detached as far distally as possible and from the abductor pollicis brevis portion of the extensor hood. It is then reflected off the dorsal capsule of the MP joint of the two thumbs. **B**, When the thumb to be preserved has an eccentric extensor tendon insertion, the radial thumb's tendon can be oversewn radially to balance the extensor force.

longitudinal incision in the dorsal capsule of the MP joint. Sharp dissection of the radial joint tissues is first carried distally, protecting the origin of the ligament off the broad metacarpal head proximally. Next, working proximally with sharp dissection, the operator carefully makes a cuff of radial periosteum confluent with the collateral ligament (Fig. 14-75).

The radial thumb is then shelled out of the wound. This leaves the ulnar thumb, the extensor and flexor mechanism of the dorsal capsule, and the important ulnar collateral of the MP joint untouched in the remaining ulnar thumb. The metacarpal head usually has two facets, one for the excised thumb and one for the remaining thumb. Using a no. 15 blade as an osteotome, the operator removes the radial facet

with a triangular portion of the radial shaft (Fig. 14-76). In older children with more ossification of the metacarpal, a rongeur or bone biter may be required. Only when marked angulation of the articular facet is present is a supracondylar osteotomy of the metacarpal needed (Fig. 14-77). During the osteotomy, care must be taken to protect not only the origin but also the entire ulnar collateral ligament.

A substantial K-wire (0.035 or 0.045 inch) is placed in retrograde fashion from the articular face of the remaining proximal phalanx out the tip of the thumb. The thumb

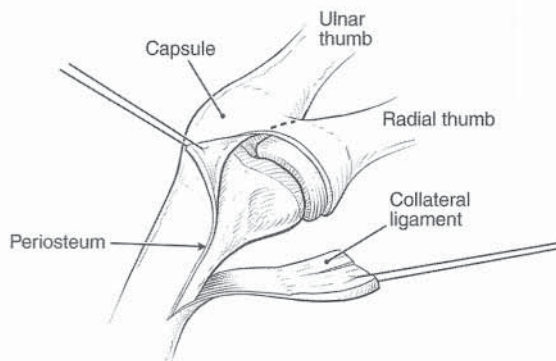


FIGURE 14-75 Sharp dissection of the radial joint tissues is begun distal to the MP joint of the thumb to be sacrificed (the ulnar thumb, in most cases). The collateral ligament is carefully detached as far distally as possible. Working distally to proximally, the operator preserves the origin of the ligament off the broad metacarpal head. Now sharp dissection of a cuff of radial periosteum confluent with the collateral ligament is carefully done.

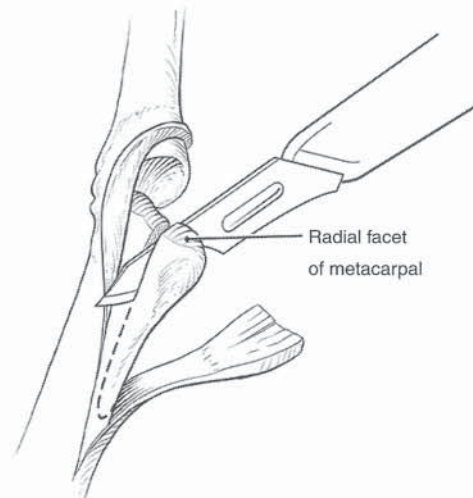


FIGURE 14-76 The metacarpal head can usually be noted to have two facets, one for the excised thumb and one for the remaining thumb. Using a no. 15 blade as an osteotome, the surgeon removes the mostly cartilaginous radial facet along with a triangular distal portion of the radial shaft of the metacarpal.

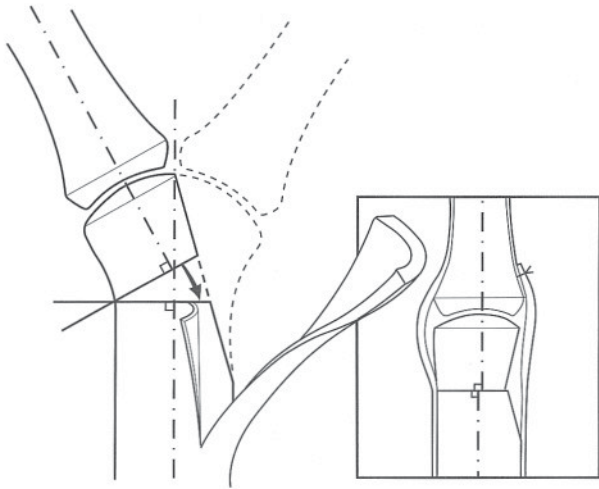


FIGURE 14-77 Supracondylar osteotomy of the metacarpal is rarely required, and only when there is marked angulation of the remaining articular facet. It is useful to remember in these small bone osteotomies that when the osteotomy of each side of the wedge is made at a right angle to the long axis of the metacarpal shaft proximally and the long axis of the distal thumb distally, a straight thumb results from wedge removal and closure. Special care must be taken to protect the origin and entire ulnar collateral ligament. The bone fragments are stabilized with a substantial K-wire positioned longitudinally, and the reconstruction is protected with a safety cast. See section on dressings and Figures 14-2 through 14-11.

is relocated in slight radial deviation to protect the radial collateral ligament reconstruction. Radial deviation is not done when an angular correction osteotomy of the metacarpal has been required. The K-wire is then inserted across the MP joint into the metacarpal to protect the ligament reconstruction during healing. The ligament is repaired with one or two substantial (3-0) nonabsorbable sutures. The K-wire is left outside the skin for easy removal later. The abductor pollicis brevis tendon is attached to the proximal phalanx over the radial collateral ligament repair and an extensor hood is reconstructed (Fig. 14-78).

The hand and thumb are completely immobilized in a long-arm "safety cast" for 4 to 6 weeks to protect the K-wire and reconstruction during healing (see Fig. 14-10). After healing, the cast and K-wire are removed. Postoperative rehabilitation is impractical in these young children, and

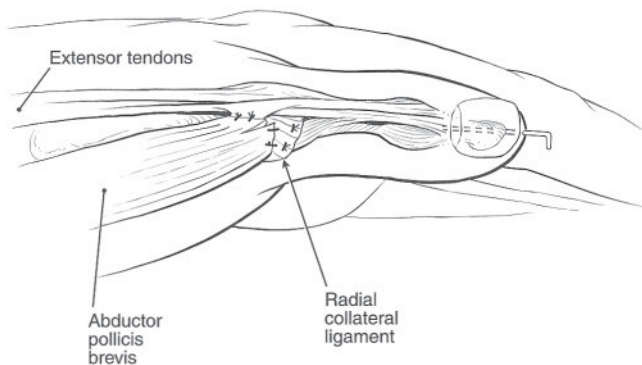


FIGURE 14-78 The extensor hood is reconstructed by attaching a portion of the abductor pollicis brevis tendon to the proximal phalanx over the radial collateral ligament repair and extensor tendon.

they are allowed to use the thumb freely after the cast and pin are removed, 4 to 6 weeks after the operation.

Wassel Type V and VI Duplications. These rare and complex duplications require reattachment of the abductor tendons and reconstruction of the basal joint of the thumb. The reader is referred to hand surgery texts for a full discussion of the treatment.

TRIPHALANGEAL THUMBS. It is important to remember that triphalangeal thumbs may be associated with many serious cardiac anomalies and hematopoietic disorders. Clearance of these disorders prior to treatment is essential. In Wassel's classification of Flatt's cases, this is a type VII thumb and represented 20 percent of the thumb duplications in Flatt's series. Although many untreated adults claim normal function in the long and frequently angled triphalangeal thumb, careful observation shows reduced fine motor skills. When the triphalangeal thumb occurs without duplication, angulatory deformities are common. We prefer to shorten these thumbs by resecting one of the joints which moves the least. Angulatory deformity can also be corrected.

When the triphalangeal thumb occurs as part of thumb duplication, often the ulnar thumb may be more normal from the supracondylar area of the metacarpal distally and the radial triphalangeal component may be attached to a more normal basal joint (Fig. 14-79). In these cases, the best elements of each thumb are used in the reconstruction.

CENTRAL POLYDACTYLY. The treatment of central polydactyly may be one of the most challenging problems in all of congenital hand reconstruction surgery. At times these duplicated digits are so severely abnormal that, if all other digits are normal, a ray resection of both duplicated digits may be the best choice. It is often difficult for the parents to give up a five-fingered hand for a more mobile one.



FIGURE 14-79 When the triphalangeal thumb occurs as part of a thumb duplication, the ulnar thumb may be more normal from the supracondylar area of the metacarpal distally and the radial triphalangeal component may be attached to a more normal basal joint. In these cases, the best elements of both thumbs are used. The distal portion of the radial thumb is deleted, while the basal joint and proximal metacarpal are saved and attached to the supracondylar area of the ulnar thumb, since it is more normal distally.

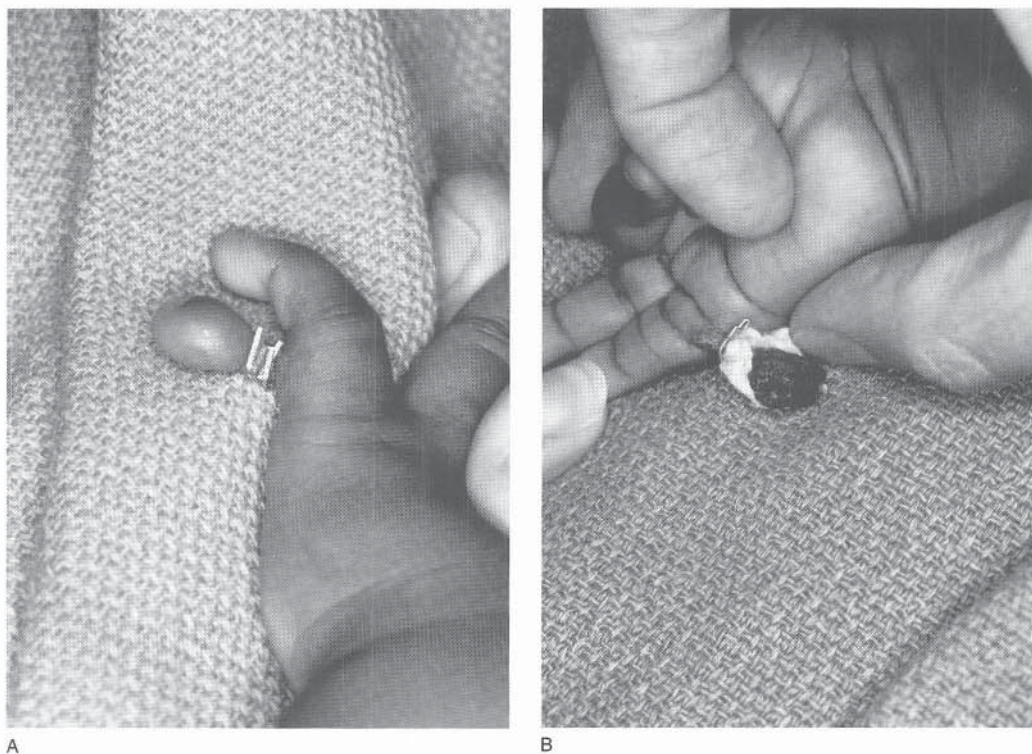


FIGURE 14-80 **A**, After injection of the base with local anesthetic, a type B ulnar polydactyly (see Fig. 14-67B) can be treated with a small ligaclip to autoamputate the extra digit as an outpatient procedure. **B**, The digit is then dressed and the child is seen weekly until it is desiccated and drops off.

Nevertheless, they must understand that a stiff central digit acts as a “functional parasite” on the remaining normal hand. Graham and Ress wisely recommend saving one digit only when that digit remaining is 75 percent of normal size and has tendon and joint development adequate to allow a movable segment.⁶

Transverse polydactylous elements in the central portion of the hand have the potential to cause increasing deformity, and early excision is warranted (see Figs. 14-66B and D).

ULNAR POLYDACTYLY. Extra digits without skeletal attachment (type B) are often treated by ligation of the stalk in the nursery (see Fig. 14-67B). Ligation should only be used in digits held by a narrow stalk. Flatt cites a case of known death from exsanguination from ligation.⁴ A broad-based attachment should await safe general anesthesia, good light, good instruments, and a more formal excision (see Fig. 14-67A). However, delay in removing the simple type B digit should be avoided, because families occasionally develop a curious attachment to these floppy, useless appendages. A small ligaclip placed after injection of the base with local anesthetic is very effective (Fig. 14-80A). The digit is wrapped and the child is seen weekly until the digit is desiccated and autoamputates (Fig. 14-80B). The mother usually grasps the concept of autoamputation since she has recently experienced it with the child’s umbilical cord.

MIRROR HAND. The details of treatment for this extremely rare and fascinating disorder are beyond the scope of this general pediatric text, and the reader is referred to the hand surgical literature. The pediatric orthopaedist is strongly encouraged to refer patients with mirror hand to a hand

surgeon with a special interest in pediatric hand surgery. In addition to the hand, the elbow and wrist require treatment, and the results are unpredictable.

Complications

RADIAL POLYDACTYLY. The complications from radial polydactyly reconstruction center on unstable joints and deformity (Fig. 14-81). Careful reconstruction of tendon insertions and joint collateral ligaments reduces the incidence of complications. However, occasionally joint reconstruction by fusion or ligament reconstruction may later be necessary. Nail deformities after the two distal duplications are joined are frequent and sometimes a problem for the patient.

CENTRAL POLYDACTYLY. Complications are especially severe when a stiff digit is left behind after reconstruction. The central digit, which does not move, is poorly vascularized, and a digit that is numb is not a functional asset. It is rarely a cosmetic one. When other digits are nearly normal, deletion and accepting less than a pentadactylous hand is appropriate.

ULNAR POLYDACTYLY. Upton has said that a wart noted over the ulnar border of the hand of a black teenager is often a cicatrix resulting from a ligation of postoperative polydactyly in the nursery. Cicatrix and keloid scars are often a nuisance but rarely are serious complications.

REFERENCES

Polydactyly

1. Bilhaut M: Guérison d’une pousse bifide par un nouveau procédé opératoire. *Congres Franc Chir* 1890;4:576.

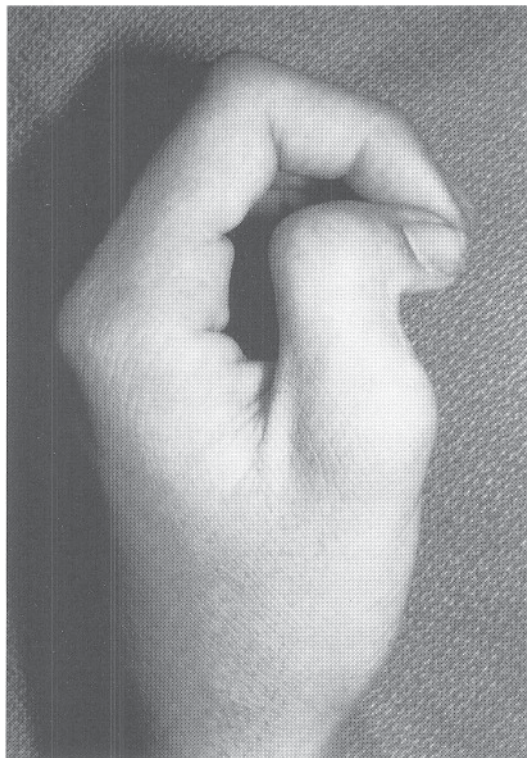


FIGURE 14–81 Careful reconstruction of tendon insertions and joint collateral ligaments at the IP joint in bifid thumb treatment reduces the incidence of complications. In some cases, later joint reconstruction by fusion or ligament reconstruction may be necessary.

2. Ezaki M: Five-fingered hand. In Green TR (ed): *Operative Hand Surgery*, 4th ed, p 449. New York, Churchill Livingstone, 1999.
3. Ezaki M: Radial polydactyly. *Hand Clin* 1990;6:577.
4. Flatt AE: *The Care of Congenital Hand Anomalies*, 1st ed, p 292. St. Louis, CV Mosby Co, 1977.
5. Flatt AE: *The Care of Congenital Hand Anomalies*, 2nd ed, p 292. St. Louis, Quality Medical Publishing, 1994.
6. Graham T, Ress A: Finger polydactyly. *Hand Clin* 1998;14:49.
7. Light TR: Congenital anomalies: syndactyly, polydactyly, and cleft hand. In Peimer C (ed): *Surgery of the Hand and Upper Extremity*, p 2111. New York, McGraw-Hill Health Professions Division, 1996.
8. Light TR: Congenital hand deformities/Development of the hand. In Green DP (ed): *Operative Hand Surgery*, 4th ed, vol 1, p 432. New York, Churchill Livingstone, 1999.
9. Muragaki Y, Mundlos S, Upton J, et al: Altered growth and branching patterns in synpolydactyly caused by mutations in HOXD13. *Science* 1996;272:548.
10. Saunders JW Jr, Cairns JM, Gasselting MT: The role of the apical ridge of ectoderm in the differentiation of the morphological structure and inductive specificity of limb parts in the chick. *J Morphol* 1957;101:57.
11. Shiono H, Ogino T: Triphalangeal thumb and dermatoglyphics. *J Hand Surg* 1984;9-B:151.
12. Stelling F: The upper extremity. In Ferguson AB (ed): *Orthopedic Surgery in Infancy and Childhood*, p 304. Baltimore, Williams and Wilkins Co, 1963.
13. Temtamy SA, McKusick VA: Polydactyly. In Bergsma D (ed): *The Genetics of Hand Malformations: The National Foundation—March of Dimes. Birth Defects: Original Article Series*, vol 14, no 3, p 364. New York, Alan R Liss, 1978.
14. Tureck SL: *Orthopaedics Principles and Their Application*. Philadelphia, JB Lippincott, 1967.
15. Upton J: Congenital anomalies of the hand and forearm. In McCarthy JG (ed): *Plastic Surgery*, vol 8, pt 2, p 5213. Philadelphia, WB Saunders Co, 1990.
16. Wassel HD: The results of surgery for polydactyly of the thumb. *Clin Orthop* 1969;64:175.

17. Wood V: Congenital hand deformities. In Green DP (ed): *Operative Hand Surgery*, 4th ed, vol 1, p 445. New York, Churchill Livingstone, 1999.

SYNDACTYLY

Syndactyly shares the rank of most common congenital anomaly with polydactyly, and because it is so common, surgeons may fail to recognize the importance of careful treatment. The terms simple or complex are commonly used to describe types of syndactyly (referring to the presence or absence of bony connection between the digits). That the phrase simple syndactyly has been associated with this condition is unfortunate, since the treatment of even the cutaneous form of the condition is far from simple. Light lists 14 steps to the operation!⁵ More important, a complication of the initial treatment of syndactyly precludes a good result and may lead to frequent revision throughout childhood. With appropriate modern treatment, carefully done with precision and attention to detail, the results from correction are usually good and the need for later revision is rare.

History. Descriptions of the treatment of syndactyly began to appear in the surgical literature in the nineteenth century and are now so various as to bewilder the surgeon trying to prepare for his or her first case. Upton lists more than 40 different techniques to treat the “simple” form of the condition.⁹ However, significant milestones in the history of the treatment have led to principles, which are well discussed by Littler and Hentz.⁶ The standard of care for the condition today reflects the contributions of many earlier surgeons. Especially critical was the work of Zeller, who in 1810 devised the dorsal triangular flap; Lennander, who in 1891 used free skin grafts; and Pieri, who in 1920 used the Z-shaped incisions.

Etiology. Upper limb development between weeks 5 and 7 of fetal life is described elsewhere in this text. All hands are initially webbed until cessation of the elaboration of the *apical ectodermal ridge maintenance factor* (AERMF) by the cells at the periphery of the web. At a preprogrammed moment, these cells cease the production of AERMF, and the process of web recession by *apoptosis*, or programmed cell death, begins. The process begins distally and continues proximally to a variable depth.^{1,5} The cause of failure of the process of recession of webbing in patients with syndactyly is unknown. A family history of the condition has been reported in 15 to 40 percent of cases.⁹ When a genetic inheritance pattern is identified in a patient with syndactyly without other associated conditions, it is of an autosomal dominant type with variable penetrance.⁴

Pathology. Several tissues may be involved in syndactyly. The skin is usually normal unless a constriction band causing chronic edema causes it to be edematous and thicker. The subcutaneous tissues are normal in quality, but some surgeons have felt the copious fat compromises wound closure and suggest partial defatting of the finger.⁴ Others feel that the amount of fat is normal in a child’s chubby finger and, if removed, may compromise a normal profile in adult life.¹

The fascial ligaments of Cleland and Grayson are frequently coalesced and thickened. Palmar fascia may be more extensive and thickened, particularly in the first web, causing a significant reduction in the potential thumb-index span.

Joints may be partially or completely stiffened. The radiograph may or may not demonstrate a cleft, falsely suggesting the presence of a joint. Because of this, careful clinical examination for movement in all joints is important in these children. One must not assume that a joint or even the potential for reconstructing a joint is present solely from the presence of a cleft on the radiograph. This is a common misconception of parents when they see the child's radiograph. Ligaments of joints are usually normal in the less complicated forms of syndactyly. However, in more complicated cases, such as central polysyndactyly, a shared ligament of a multifaceted proximal bone may complicate reconstruction. The syndactyly is best left unseparated in some of these cases.

In complex syndactyly, the bone tissue of adjacent digits is by definition fused. Fusion may vary from only the tip of the tuft to complete coalescence. In the case of distal phalangeal coalescence, a synonychia or fusion of adjacent fingernails is present—a useful clinical sign. This association reflects the common ectodermal origin of these two elements, the nail and the tuft portion of distal phalanx. In some cases the radiograph may reveal whole or partial skeletons of unexpected digits enclosed by the skin syndactyly (Fig. 14–82).

Tendons and their sheaths are usually completely duplicated and normal in a cutaneous syndactyly but may be grossly abnormal in complex cases. This is especially common in the acrocephalosyndactylies such as Apert's syndrome.

Nerves and vessels are usually duplicated in cutaneous syndactyly, providing a complete set for each digit after separation. They may be abnormal to a varying extent in

complex cases. Abnormality may manifest only as a more distal bifurcation of the nerve and vessel, but in severe cases a single vessel or nerve may entirely supply both digits. The latter is most commonly seen in the acrocephalosyndactylies and is very unusual in the simple cutaneous form of syndactyly.

Clinical Presentation

Terminology

Cutaneous (simple)—Cases without bone fusion.

Complex or osseous—Cases with evidence of any bone fusion.

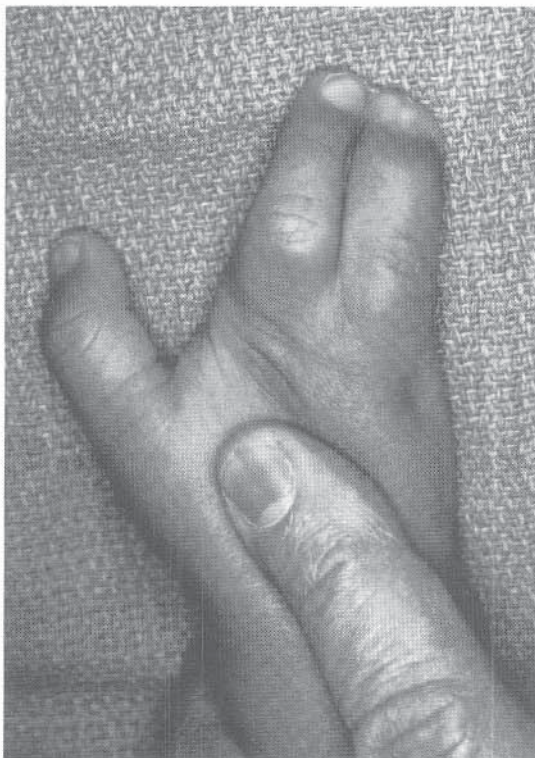
Complicated—Cases of syndactyly that are complicated by polydactyly, severe coalitions, transverse elements, and so on. The term *complicated* was introduced by Dobyns to separate these more difficult cases from those with a small bony fusion at only the distal phalanx tuft.

Incomplete—In these cases, the web began to recede but stopped short of the normal level.

Complete—The entire web is absent and the tips are coalesced.

Acrosyndactyly—This term should be reserved for cases resulting from congenital band syndrome (Streeter's syndrome), for the condition results from a traumatic fetal injury and does not represent a failure of formation, as other syndactylies do. Because it is different in etiology and treatment, acrosyndactyly is discussed later in this chapter under Congenital Band Syndrome.

Incidence. The incidence of syndactyly is about 1 in 2,000 live births. The condition is 10 times more common in white children than in black children. Boys are affected twice as



A



B

FIGURE 14–82 A and B, The web may hide an extra digit within the syndactyly.

often as girls. Bilateral cases occur at the same rate as unilateral involvement.³

A family history of the condition is not unusual in syndactyly. Familial cases not associated with other syndromes account for 15 to 40 percent of cases and tend to be more complex forms of syndactyly. The inheritance pattern follows an autosomal dominant pattern but with variable penetrance.^{3,9}

The distribution of the frequency of web involvement is shown in Figure 14–83. By far the most common is long-ring syndactyly. The rarest is the thumb-index web, perhaps because the first web separates earlier in fetal life than the other three webs.⁶

Associated Syndromes. Syndactyly is often an isolated anomaly and may be a part of many syndromes. These are listed in Table 14–7.

Imaging. Much of the hand and the entire carpal skeleton are not ossified in the infant, making radiographic information always incomplete and occasionally deceptive at this age. However, a PA radiograph of both hands obtained prior to operation is useful. The radiograph often needs to be obtained again immediately before the planned surgical reconstruction since it may often have been properly delayed from the initial visit to the surgeon. Even in toddlers, false negative studies may occur in regard to bony coalitions, delta phalanges, duplications, and symphalangism. On the other hand, false positive studies are a problem in evaluating joint spaces. It is important not to assume that a joint or even the potential for reconstructing a joint is present solely from the presence of a cleft on the radiograph. This is a common misinterpretation by parents if they see their child's x-ray.

Arteriography is not indicated in simple syndactyly. It may be rarely indicated in the most complicated forms of complicated syndactyly, such as the severe coalitions seen in Apert's syndrome. In general, however, the risk-benefit ratio of more sophisticated imaging in these small children who need general anesthesia for studies such as MRI, arteriography, and CT makes their use limited in even the most complex syndactyly.

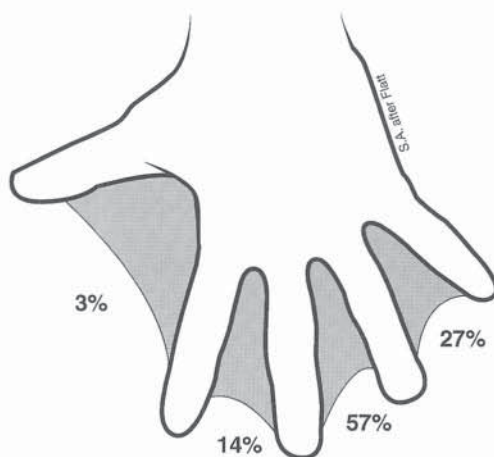


FIGURE 14–83 The incidence of syndactyly of the various webs when only true syndactyly not associated with other conditions is considered.

TABLE 14–7 Conditions Associated with Syndactyly

Chromosomal

Trisomy 13
Trisomy 14
Trisomy 21
Partial trisomy 10q
Triploidy syndrome
5p–
13q– ring, D+

Craniofacial Syndromes

Aglossia-adaactylia syndrome
Apert's syndrome
Carpenter's syndrome
Cohen's syndrome
Cryptophthalmos syndrome
Ellis–van Creveld syndrome
Familial static ophthalmoplegia syndrome
Glossopalatine ankylosis syndrome
Greig's cephalopolysyndactyly syndrome
Hanhart's syndrome
Hypertelorism and syndactyly syndrome
Möbius' syndrome
Noack's syndrome
Oculodentodigital syndrome
Oculomandibulofacial syndrome
Orofacial-digital
Otopalatal-digital
Pfeiffer's syndrome (acrocephalosyndactyly type V)
Pierre Robin syndrome
Saethre-Chatzen syndrome

Other Syndromes

Acropectoral-vertebral dysplasia, F form (F syndrome)
Aarskog syndrome
Bloom syndrome
Brachydactyly A2
Brachydactyly B
Chondrodysplasia punctata (Conradi's syndrome)
Cornelia de Lange's syndrome
EEC syndrome (ectrodactyl-ectodermal dysplasia-clefting syndrome)
Escobar syndrome
Fraser's syndrome (cryptophthalmos)
Goltz's focal dermal hypoplasia syndrome
Holt-Oram syndrome
Incontinentia pigmenti syndrome
Jarcho-Levin syndrome
Laurence-Moon-Biedl syndrome
Lacrimeoauriculodentodigital syndrome
Lenz's microphthalmos syndrome
Lenz-Majewski hyperostosis syndrome
McKusick-Kaufman syndrome
Meckel's syndrome
Miller syndrome
Neu-Laxova syndrome
Pallister-Hall syndrome
Poland's syndrome
Popliteal pterygium syndrome
Prader-Willi syndrome
Roberts-SC phocomelia syndrome
Rothmund-Thomson syndrome
Sclerosteosis syndrome
Scott craniodigital mental retardation syndrome
Short rib-polydactyly (Saldino-Noonan) syndrome
Smith-Lemli-Opitz syndrome
Spondylothoracic dysplasia syndrome
Summit syndrome
Thrombocytopenia-absent radius syndrome
Waardenburg's syndrome

From Ezaki M, et al: Syndactyly/congenital hand deformities. In Green DP (ed): *Operative Hand Surgery*, 4th ed, vol 1, p 415. New York, Churchill Livingstone, 1999.

Treatment

TIMING. A consensus is finally developing among pediatric hand surgeons that early surgery is indicated only when border digits such as the thumb or small finger are involved, and in cases of complex complicated syndactyly (especially those with transverse elements). Such cases are best addressed surgically in the second half of the first year of life.

However, in cases of cutaneous or simple distal tuft osseous syndactyly of the long/ring and long/index webs, the operation is best deferred until the size of the child's hand is more favorable for the precision surgery required for an optimum result. These cases account for more than two-thirds of all cases of syndactyly (see Fig. 14–83). The surgeon should remember that normal growth of the hand provides for a doubling in size about twice during the approximately 15 years of growth, and that the first doubling occurs during the first 3 years of life.² Therefore, judiciously waiting until the child is about 18 to 24 months old rewards the surgeon with a significant increase in size of the operative site. In addition, the relatively hypertrophic scar produced by the young child's skin does not tolerate mistakes in the layout of incisions. These incisions must follow the standard diamond-shaped patterns described by Littler (see Fig. 14–1).

CUTANEOUS (SIMPLE) SYNDACTYLY. The term *simple* may suggest that this operation requires little skill and can be done by a novice. That is not the case. Expert execution of a syndactyly reconstruction is not simple. The surgeon who treats a cutaneous syndactyly must realize that complications of this operation are difficult to treat and are usually the result of poor incisional design, tight closure, and flap or graft necrosis.

Critical elements for obtaining a quality result were succinctly identified by Littler and Hentz almost 30 years ago:⁶ "The goal of surgery is separation of independent digits without deformity. Inherent is the establishment of a definite commissure and the avoidance of deforming scars. Examination of the [normal] hand demonstrates that the cleft skin has the unmistakable quality of dorsal skin. To use a volar flap here ignores this observation. The interdigital volar cleft edge is a definitive transition between dorsal and palmar skin. However, to end the dorsal skin flap at this point could leave a restricting scar at a crucial position. Therefore, the dorsal flap must be cut sufficiently long to be inset just palmar to the new cleft margin. . . . That a skin deficit exists in every syndactyly is demonstrated by envisioning that two circles are to be created from an oval whose long axis equals twice the diameter of each circle."

Figures 14–84 and 14–85 illustrate the technical maneuvers of reconstruction and the absolute deficit of skin in cases of syndactyly, which mandate a skin graft. In our own experience, we have found that when possible, a dorsal flap of adequate length, interdigitating skin incisions, and full-thickness skin grafts are critical elements in the treatment of cutaneous syndactyly. We find arguments for avoiding skin grafting to be specious.

Attention to detail in incisions, dissection, skin graft harvest, suture, and careful dressing application is tedious but rewards the surgeon and patient with a digit that is close to normal. Anything less compromises the result. Less interested surgeons help their patients and themselves by refer-



FIGURE 14–84 A to C, The principles of creating the dorsal flap, reciprocal zig-zag incisions, and free skin grafts are illustrated, along with the harvest site in the glabrous region of the lateral portion of the inguinal fold. (From Converse JG: Plastic and Reconstructive Surgery, 2nd ed, vol 6, p 3318. Philadelphia, WB Saunders Co, 1977.)

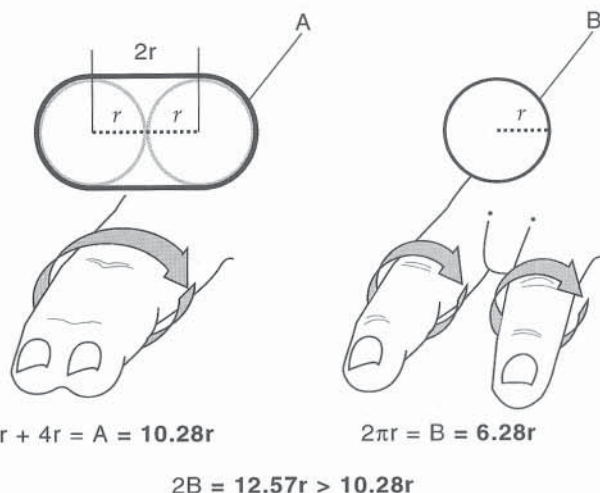


FIGURE 14–85 There is an absolute deficit of skin in cases of syndactyly which dooms the attempt to reconstruct a proper, scar-free, mobile web without skin graft.

ring these children to a surgeon who does these cases routinely. For the pediatric orthopaedist who is interested in performing syndactyly reconstruction and achieving an optimal result, the details of a cutaneous syndactyly release are illustrated here:

Technique for Cutaneous (Simple) Syndactyly

1. *Preparation.* All syndactyly reconstruction must be carried out under general anesthesia. A tourniquet must be used to ensure a completely bloodless field during the operation. Although the pneumatic tourniquet provides a numerical figure for monitoring pressure, in small arms it may fail to control bleeding well. We have found that the Martin elastic bandage used to exsanguinate the arm can be wrapped around the upper arm three times and tucked in, giving predictable and safe hemostasis. Ruby and his associates have shown that each wrap of the elastic bandage is equivalent to about 80 mm Hg pressure; hence, three wraps is about 250 mm Hg (L. Ruby, personal communication).

The skin graft donor site should be prepared and draped at the same time. Our site is usually the outer groin, in the natural crease of the skin formed when the hip is flexed. Care must be taken to use skin from the lateral portion of the groin, as full-thickness skin carries the pubic hair-forming elements to the hand when harvested. When more skin is required in multiple syndactylies, we prefer the area just below the umbilicus. Here large amounts of glabrous skin are available and the long transverse closure line is acceptable.

There is controversy over the use of prophylactic antibiotics in regard to type, duration, dose, and even whether they should be used at all. The skin grafts are especially vulnerable to *Staphylococcus* or *Streptococcus* infection, and in our community, where summer heat as well as the casual hygiene of some of our patients is a problem, we have settled on one intravenous dose of a cephalosporin given just prior to inflation of the tourniquet.

2. *Design of the incisions.* It has been said, with some justification, that after the incisions have been drawn in a syndactyly, the case is over. The importance of thoughtful planning of these incisions cannot be overemphasized. Healing guarantees that there is no place for the superfluous in surgery, and simplicity is critical when planning these incisions. Flaps should be as generous as possible and out-

lined so that the incisions, when closed, will provide a web space covered with dorsal skin and free of transverse scars. Interdigitation of flaps usually can be done only partially. We do not attempt to completely cover one finger with flaps alone and prefer instead to use a skin graft on both sides of the web. The goal is to have incisions resulting from the combination of flap closure and graft insertion that conform to the previously mentioned rules of digital incisions (see Fig. 14-1).

The surgeon begins by flexing the MP joints to identify the metacarpal heads. (Fig. 14-86). The proximal extent of the dorsal flap should start at about the MP joint line. The distal extent of the dorsal flap usually ends just short of the PIP extension wrinkle on the dorsum of the unseparated digit. The dorsal flap must be quite long, since it must swing obliquely downward, fold slightly on itself, and insert into the palm proximal to the web (Fig. 14-87).

After the dorsal flap is correctly laid out, the incision is carried distal from the flap in a zig-zag manner. The remaining dorsal flaps created by continuing the incision are designed to resurface the sides of the fingers by interdigitating with their palmar counterparts (Fig. 14-88). This group of flaps has two critical design elements. First, the flaps must be kept large and simple, since the infant's healing skin tends to produce a robust scar that will consolidate any small flaps into a single thickened line. Second, the flaps must be designed so that after closure, the resulting incision lines on the separated fingers satisfy the rules for digital incisions detailed in Figure 14-1. Any incision lines oriented in such a way that they will change length or be under tension with digital movement will result in an incisional scar that hypertrophies. Similarly, in syndactyly reconstruction, incision lines from flaps as well as the edge of skin grafts react in this same way. Scarring from poorly designed incisions is the major cause of "web creep" after syndactyly. The often quoted failure of the skin graft to grow with the child is much less significant than poorly planned incisions or loss of skin grafts. Therefore, the palmar incision must

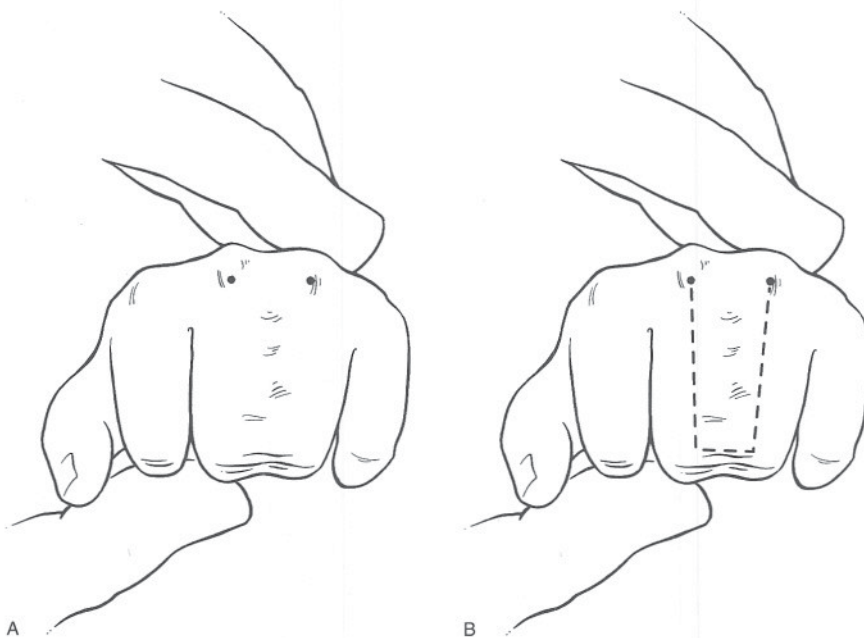


FIGURE 14-86 A and B, To design the incisions for a syndactyly reconstruction, the surgeon first locates the metacarpal heads by flexing the MP joints, then draws the dorsal flap.

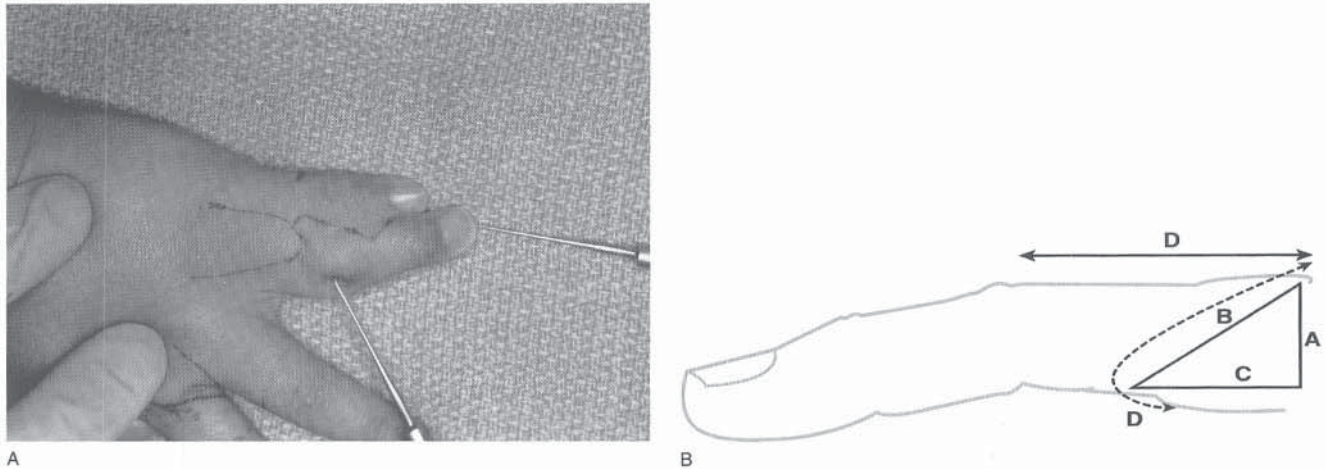


FIGURE 14-87 A, The dorsal incision with a long, proximally based flap for the web. Side-based flaps of generous size are designed to interdigitate with the mirror flaps palmarly. On the unseparated syndactyly the distal extent of the dorsal flap must extend to a point just short of the PIP skin wrinkle and is well distal to the final web level. B, This ensures adequate skin to create the normal slant of the web (the hypotenuse of the triangle). In addition, this allows the flap tip to fold back on itself and rotate slightly into the palm. This prevents the potential complication of leaving the incision at the leading edge of the web.

allow the dorsal and palmar finger flaps to interdigitate correctly. When this is not possible, the full-thickness graft must be designed and inserted so as to prevent hypertrophy.

The palmar incision should end proximally just proximal to the intended web so that the flap from the dorsum will insert correctly. The flap should fold back on itself and avoid a transverse scar at the distal or leading edge of the web (Fig. 14-89).

3. *Separation of the digits.* We usually begin with incision and elevation of the dorsal flap distally, carrying the dissection down to the extensor tendon peritenon. The flap is then gently elevated and the base of this flap dissected as little as possible, since it will rotated down and into the web. A 4-0 silk retention suture helps avoid unnecessary

handling of the flaps with forceps. Proceeding distally, the other dorsal flaps are dissected away from the thicker than normal Cleland's ligament.

The palmar dissection is done from proximal to distal on the digit. The use of loupe magnification allows dissection of the neurovascular bundle to be accomplished more safely. Care and patient, gentle dissection are especially important during this step because damage to the vessels may compromise separation (Fig. 14-90). The nerve is superficial to the artery at the level of the proximal phalanx and can be found coursing through the subcutaneous fat. Gentle spreading with scissors in line with the nerve and artery is the safest way to find these critical structures. Once the vascular supply to each digit has been identified at this level in the finger,

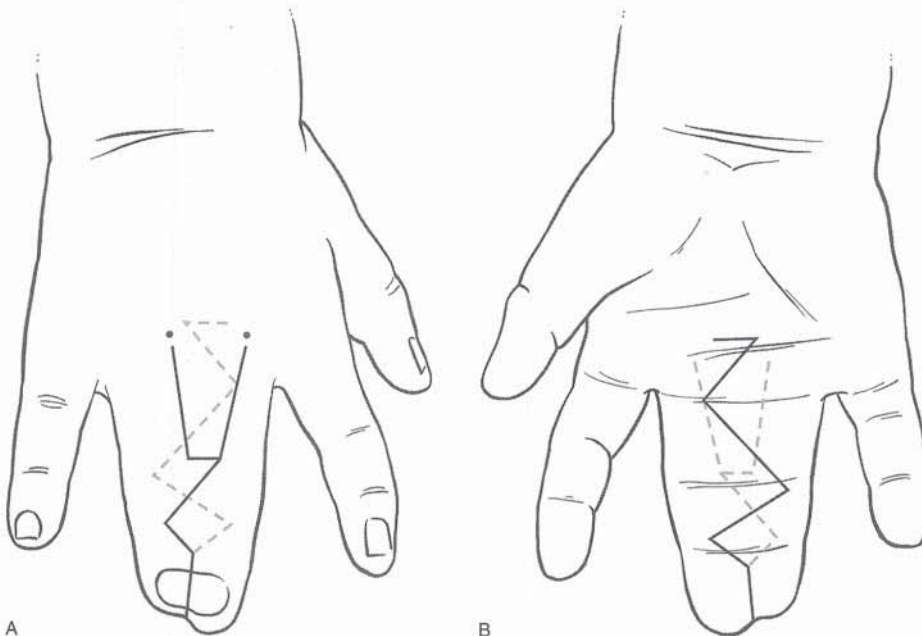


FIGURE 14-88 A, The palmar incision is designed to mirror the distal flaps and allow the dorsal flap creating the web to insert at a point well proximal to the final distal edge of the web. B, The design of the web and the palmar incision.

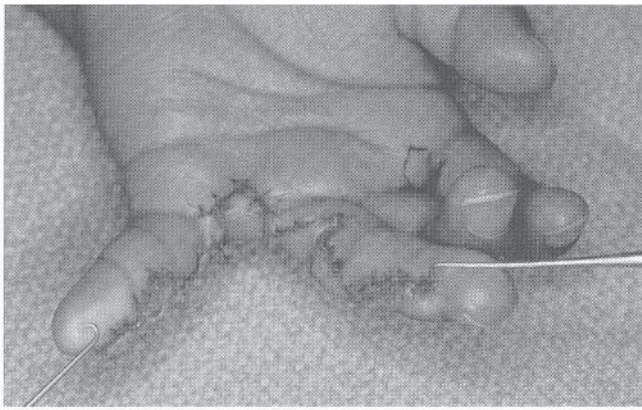


FIGURE 14-89 The truncated dorsal flap should fit well proximal to the distal edge of the reconstructed web.

the bifurcation of the common digital vessel can be found with careful proximal dissection. Now the dissection can be carried distally directly along the anterior surface of the neurovascular bundles, using care to leave them in the floor of the wound. After the bundles are separated out to the distal pulp, a simple skin hook in each finger tip allows gentle tension to be used to facilitate release of a thickened Cleland's fascia. After the coalesced Cleland's fascia is cleaned of its subcutaneous fat and well identified, we usually separate the fascia sharply with a knife, teasing the two digits apart but staying out of the joints and ligaments, and mindful of the neurovascular structures (Fig. 14-91).

4. *Flap closure.* The flaps are rotated into position and held at first by the silk retention suture while being meticulously closed with fine (6-0) absorbable suture (Fig. 14-92). If a suture that does not have to be removed is used, the child, parent, and surgeon have a more pleasant experience in the aftercare period. We use plain ophthalmic gut, dyed blue with a skin pencil. There should be very little tension

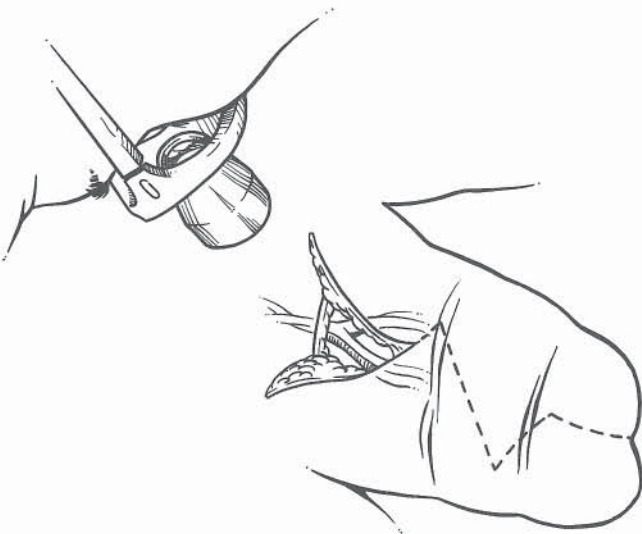


FIGURE 14-90 The palmar dissection is also done proximally to distally. The use of loupe magnification allows dissection of the neurovascular bundle to be carried out more safely. Care and patient, gentle dissection are especially important at this step, since damage to the vessels at this point may compromise separation.

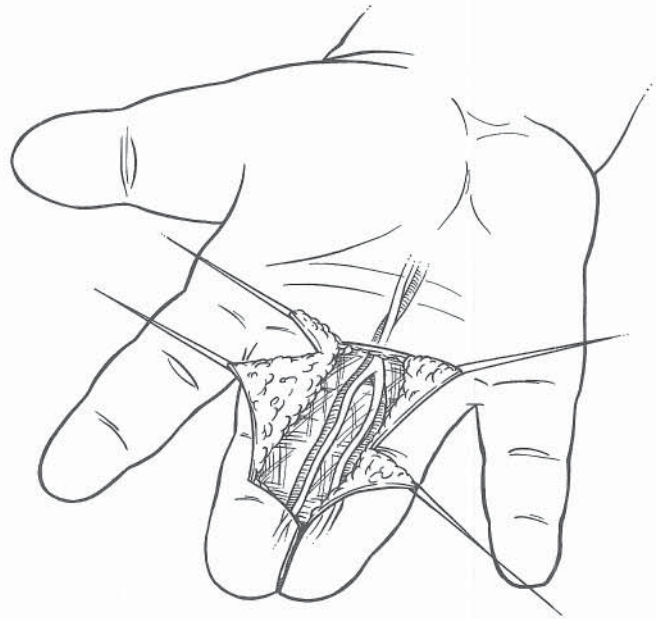


FIGURE 14-91 Once the vascular supply to each digit has been identified at this level in the finger, the bifurcation of the common digital vessel can be found with proximal dissection. Now the dissection can be carried distally directly along the anterior surface of the neurovascular bundles with care to leave them in the floor of the wound.

on the wound edges. Tension is prevented by the adequate use of skin grafts.

5. *Skin graft patterns.* The defects requiring grafting are now carefully measured by making a precise paper pattern of the defect. Making a small dot at the end of the pattern to identify the proximal or distal end makes later orientation of the graft easier (Fig. 14-93). The pattern is labeled as to location. After it has been used in outlining the skin graft donor site incision, the pattern can be used by the scrub nurse to make a precision stent dressing slightly larger than the pattern. The dressing is cut from a single layer of nonadherent gauze (Xeroform) and a dry 4 × 4-inch gauze (see Fig. 14-98A).

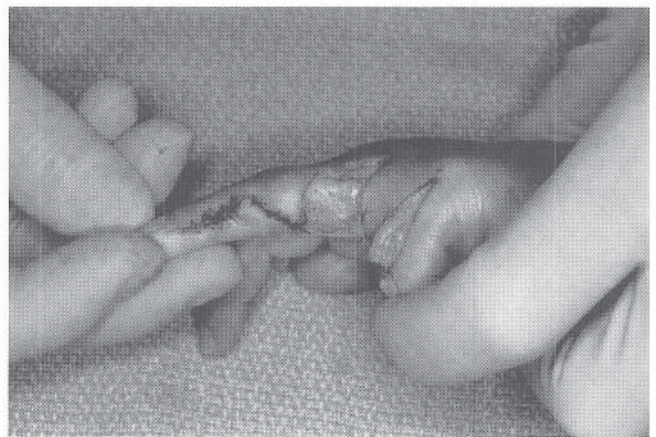


FIGURE 14-92 The flaps are rotated into position and held at first with a silk retention suture while being meticulously closed with fine (6-0) absorbable suture. We use plain ophthalmic gut, dyed blue with a skin pencil. There should be very little tension on the wound edges. Tension is avoided with the adequate use of skin grafts.

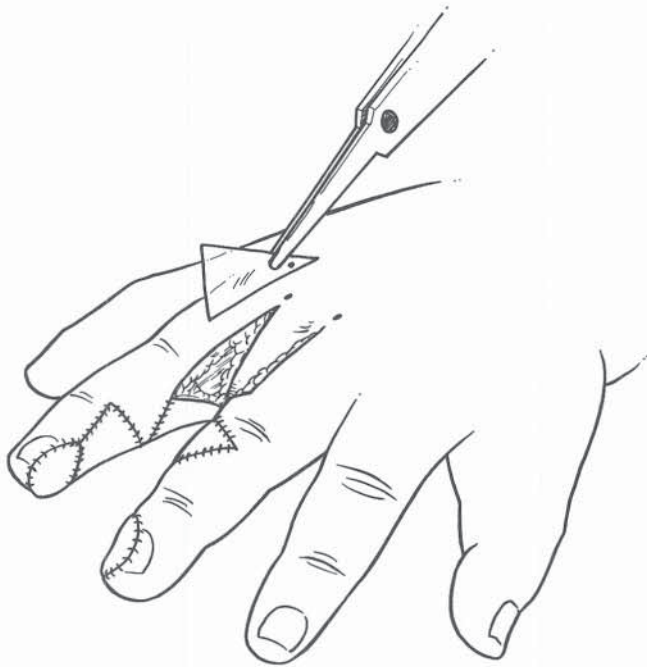


FIGURE 14-93 The defects requiring grafting are carefully measured by making a precise paper pattern of the defect. Making a small dot at the end of the pattern to identify the proximal or distal end makes later orientation of the graft easier. The pattern is labeled as to location. After it has been used in outlining the skin graft donor incision, the pattern can be used by the scrub nurse to make a precision stent dressing slightly larger than the pattern. The dressing is cut from a single layer of nonadherent gauze (Xeroform) and a dry 4 × 4-inch gauze (see also Fig. 14-98A).

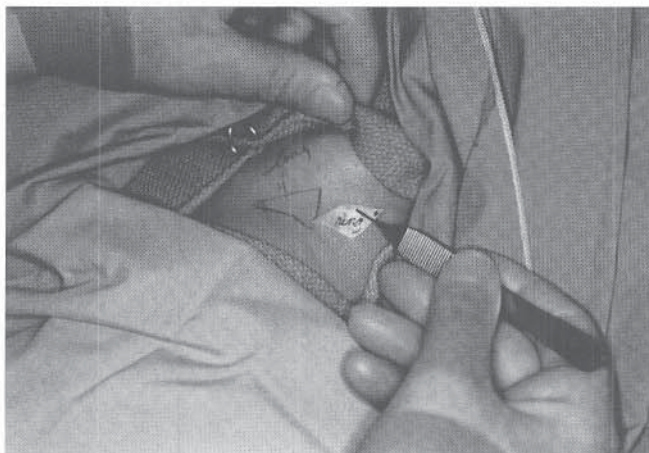
6. *Release of the tourniquet.* Now the tourniquet is released and circulation of the tips verified. If the circulation is not brisk and adequate, the sutures must be released from the flaps until it is. Once circulation has been verified, the wounds are covered with a moist dressing and the arm is elevated while the grafts are harvested.

7. *Graft harvest and donor site closure.* Precise, labeled outlines of all grafts are carefully drawn on the donor site, with the grafts arranged so that an ellipsoid defect is created that can be easily closed (Fig. 14-94). The grafts are defatted carefully as they are being removed, without attention to the exact shape of the donor site defect, which is tailored later.

Using a fresh no. 15 blade, and rolling the skin back with a gauze sponge rather than with forceps, the surgeon cuts a full-thickness graft of uniform thickness, free of fat, and exactly the correct size and shape. This provides for proper skin tension when the graft is sutured into place. We have found the care taken in graft harvest to be especially critical for good take of the graft and a quality result.

The donor site defect in the groin is converted into an ellipse and closed with skin tapes and a subcuticular, absorbable monofilament suture of adequate strength to tolerate unrestricted motion (3-0 or 4-0). A waterproof dressing is applied to the donor site and left in place for a week.

8. *Attaching grafts to the digital skin defects.* By the time the grafts have been harvested and the donor sites closed, hemostasis is usually adequate. The tourniquet is then reinflated to facilitate accurate suturing of the grafts into place. The dry field greatly facilitates precision attachment of these grafts. We use fine (6-0) absorbable suture, first anchoring the corners and then completing the attachment with either an interrupted or a fine running suture. To prevent infolding of the graft edge, the suture must be as close to the edge as possible (Fig. 14-95).



A



B

FIGURE 14-94 A, Graft harvest and donor site closure. Precise, labeled outlines of all grafts are carefully drawn on the donor site, with the grafts arranged so that an ellipsoid defect is created that can easily be closed. B, The grafts are defatted carefully as they are being removed, without attention to the exact shape of the donor site defect, which is tailored later. Using a fresh no. 15 blade, and rolling the skin back with a gauze sponge rather than with forceps, the surgeon cuts a full-thickness graft of exactly the correct size and shape. The graft should be of uniform thickness, free of subcutaneous fat, and of the proper size. This will provide the proper skin tension when sutured in place. Care in harvesting the graft is critical to good take of the graft and a quality result. The donor site defect is converted into an ellipse and closed with subcuticular, absorbable monofilament suture of adequate strength to tolerate unrestricted motion (3-0 or 4-0). A waterproof dressing is applied to the donor site and left in place for a week.

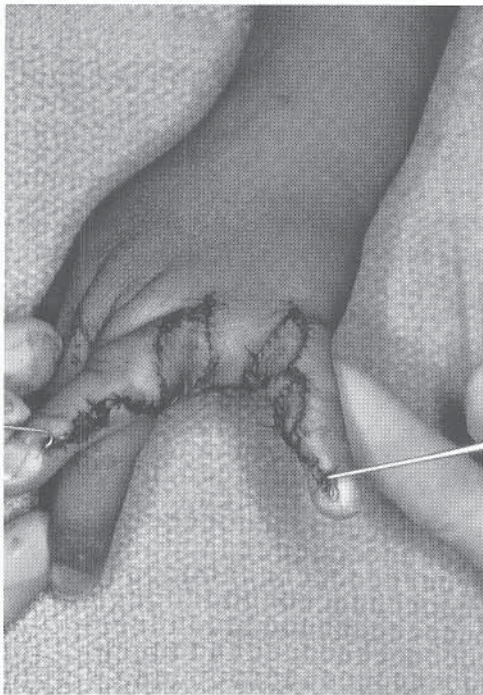


FIGURE 14-95 The tourniquet is deflated and the arm is elevated while the grafts are being harvested. By the time the grafts are ready for insertion and the donor sites closed, hand hemostasis is usually adequate, and the tourniquet is reinflated while the grafts are carefully sutured into place. A dry field greatly facilitates the precision attachment of these grafts. We use fine (6-0) absorbable suture, first anchoring the corners of the graft and then completing the attachment with either an interrupted or fine running suture. To prevent infolding of the graft edge, the suture must be as close to the edge as possible.

9. *Tip reconstruction.* If the syndactyly is incomplete at the tip, the flaps and grafts described above suffice to reconstruct the digits. However, in a complete osseous syndactyly, especially one in which the distal phalanx is fused at the tip, reconstruction of a padded surface of the distal phalanx is more difficult and important. Failure to do so by putting a full-thickness graft on bare bone or allowing the wound to heal by secondary cicatrix usually will result in a nail bed deformity that is difficult to care for, unsightly, and painful. We have found two methods that offer predictably good results in these cases. We tend to favor a free composite graft when there is a more severe shortage of skin available at the tip and use Buck-Gramcko flaps only when an unusually abundant amount of skin is available.

a. *The composite graft: The Hentz “pulp-plasty.”* A composite graft of skin and fat can be useful to resurface the raw bone surface associated with a complex syndactyly fused at the tip (Fig. 14-96). Hentz first used these composite grafts on the tips of syndactyly releases, and their successful use was subsequently reported by others.⁸ Small amounts of these composite tissues (skin and fat) can be used, but the size of the graft must not exceed the revascularization time for the vessels to reestablish circulation. The vessel ingrowth in the composite graft, unlike thinner skin grafts, occurs from the periphery or “shoreline” of the defect. The surgical closure technique is critical for success and requires careful edge approximation, since this is the only site at which neovascularization occurs.

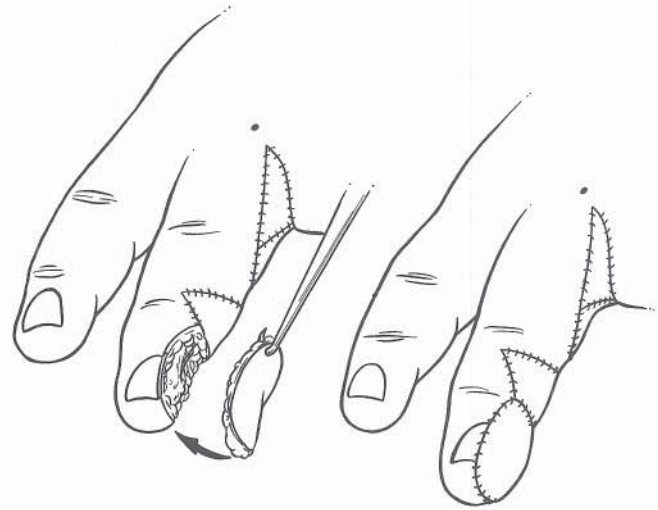


FIGURE 14-96 Composite grafts of skin and fat can be useful for resurfacing the raw bone surface associated with a complex syndactyly fused at the tip.

Either hypothenar skin or the inner surface of the great toe makes an acceptable donor area for these small grafts.

b. *Buck-Gramcko “stiletto flaps” reconstruction.* We use this method when more skin is available. The flaps are so long and attenuated that healing is similar to the healing of a composite graft harvested locally. Again, precision closure is important for optimum healing (Fig. 14-97).

10. *Dressing application.* This step is too often accorded inadequate respect by the neophyte hand surgeon. The goal of syndactyly surgery is to obtain complete and primary healing of all wounds and skin grafts. Anything short of this compromises the result and usually results in reoperation. The dressing protects the surgeon’s careful work from the patient and those around the patient. The dressing must also secure the grafts so as to prevent motion and seroma formation without congested fingers developing. The critical nature of the closing pressure in a digit is evident when one considers the simple application of a Band-aid circumferen-

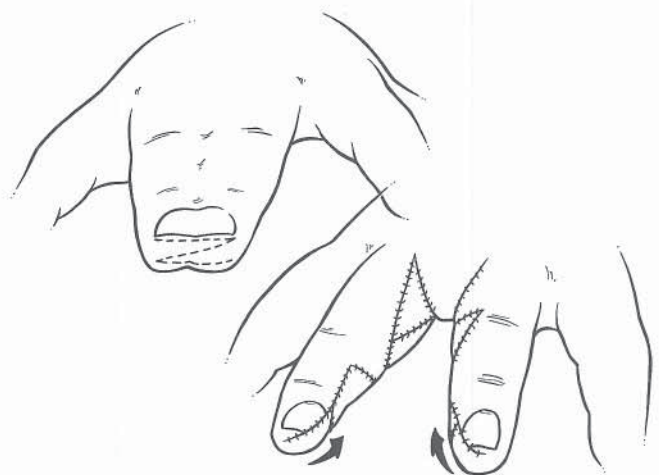


FIGURE 14-97 The Buck-Gramcko technique of flap reconstruction is preferred when there is more and better-quality skin available. The flaps are so long and attenuated that healing is similar to that of a composite graft harvested locally. Precision closure is required for optimum healing.

tially around any finger. Once the grafts are secure, the entire dressing must be made safe from the infant, who simply does not have the capacity to protect it on his or her own. In these small limbs, lack of a precision dressing will reward the surgeon with the cast separating prematurely from the patient, usually along with the grafts. We have found over the years that making a concession regarding continuous observation of the digits in the postoperative period has rewarded us with a more secure dressing and a much higher take of the skin grafts. Since healing is, in the end, the major determinant of the success of the endeavor, the risk has been worth the benefit. We call the dressing "the safety cast." Care in application of this dressing is critical.

The primary dressing of the grafted areas is a layered dressing consisting of a base of nonadherent gauze (Xeroform) followed by a pledget of regular gauze. These layers are carefully tailored to match the skin graft area using the patterns previously used to size the skin grafts (Fig. 14-98A). They are then applied as a stent dressing secured with skin tapes directly over the graft. Circumferential tape must be avoided.

The remaining flap incisions are also covered with a single layer of nonadherent gauze followed by a pledget of a carefully tailored 4 × 4-inch gauze/square soaked in saline. This is followed by fluffed gauze held in place with a roller gauze (Kling, Conform) and then cast padding (Webril) (Figs. 14-98B to D). The roller gauze should apply just enough compression to hold the primary dressing in place without causing congestion or constriction of the digits; application is a skill which requires some practice. The tourniquet is then released and circulation at the digital tips is validated. If it is not brisk, the dressing must be reapplied until unquestioned circulation is present.

The entire dressing is then supported with a long-arm mitten dressing or "safety cast" that covers the hand entirely and extends to the axilla. The elbow must be bent to 90 degrees prior to application of the dressing and cast. There is no place in children for the use of a "short" long-arm cast (Fig. 14-98E).

11. *Aftercare.* The wound is left undisturbed under the cast for 3 weeks unless fever, increasing pain, or unusual odor is noticed. If there is concern on the part of the surgeon regarding any of these reasons, the cast is removed. Over the years, however, we have found that nothing is gained and much can be lost by an unnecessary dressing change. Accurately fitting dressings that were applied in the op-

erating room under general anesthesia can never be duplicated in the awake small child. At 3 weeks the dressing is removed and the parent reassured that the dry crusts in the area (Fig. 14-99) will respond well to a few days of repeated soap-and-water washings followed by a hand lotion massage carried out by the parent. We have found more recently that the newer fiberglass casting materials that do not completely set up (e.g., Softcast, 3M) are useful, since this type of fiberglass tape can be unwound 3 weeks later, allowing removal of the cast without the use of a cast saw. This reduces the anxiety of the postoperative clinic experience for both the parent and the child.

When this regimen is followed, we expect and usually achieve complete primary healing of the operative sites. In the unusual event that this does not occur, we do not immediately regraft an area of graft slough (although some have recommended it). Any benefit of immediate surgery is usually outweighed by the need for mobilization of the hand and child.

SYNDACTYLY OF THE FIRST (THUMB-INDEX) WEB. Of all the operations done to treat congenital hand problems, we strongly agree with Upton that none rewards the surgeon and patient with more functional gain than those for the treatment of the absent or insufficient web between the thumb and fingers.⁹ Although a complete syndactyly affects the first web in less than 10 percent of cases of isolated syndactyly, a deficiency of this web is common in many other hand anomalies. To obtain the maximum first web span, in addition to correction of the skin deficit, careful and complete release of interosseous fascia and palmar fascia may be necessary. Rarely, a CMC joint release may also be required, and when it is, temporary K-wire fixation traversing the midportion of the first and second metacarpals is useful to maintain correction until soft tissue healing is complete. Although pedicle flap coverage is often used for reconstruction of a thumb adduction contracture in adults, there is little use for it in first web reconstruction in children. We acknowledge that a true tetrahedral space cannot be duplicated in the completely adducted thumb using only local flaps and grafts. However, in most cases a useful first web can be predictably and adequately reconstructed without the nursing difficulties associated with managing a young child with an attached pedicle flap. Later, in the rare case that it proves necessary, pedicle skin can be obtained when the child can better understand the importance of his or her own contribution

FIGURE 14-98 A, After it has been used in outlining the skin graft donor incision, the pattern can be used by the scrub nurse to make a precision stent dressing slightly larger than the pattern. The dressing is cut from a single layer of nonadherent gauze (Xeroform) and a dry 4 × 4-inch gauze. B to E, The remaining flap incisions are also covered with a single layer of nonadherent gauze followed by a pledget of a carefully tailored 4 × 4-inch gauze soaked in saline. This is followed by fluffed gauze held in place with a roller gauze (Kling) and/or cast padding (Webril). The roller gauze should apply just enough compression to hold the primary dressing in place without causing congestion or constriction of the digits; its application is a skill that requires some practice. The tourniquet is then released, and circulation at the digital tips is validated. If it is not brisk, the dressing must be reapplied until unquestioned circulation is present. F, The "safety cast." Once circulation has been determined to be adequate, the entire dressing is supported with a long-arm mitten dressing or "safety cast" secured high in the axilla and protected with orthopaedic felt to prevent molestation of the surgical site. Inability to directly observe the fingers is a small risk which is offset by the important benefit of undisturbed healing. Unless fever, increasing pain, or severe odor is present, we leave the rigid dressing in place for 3 weeks. Therefore, the surgeon must apply this dressing carefully. We use a fiberglass dressing for its light weight and durability, and we prefer the "soft cast" variety because it can be later removed without a saw.

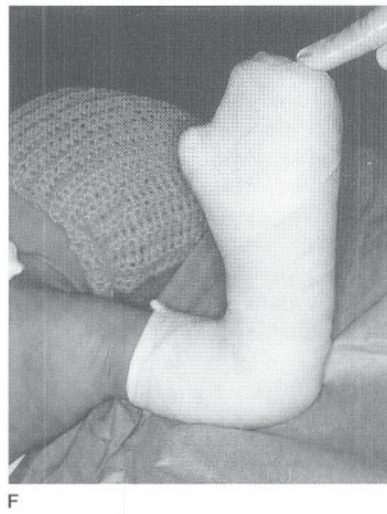
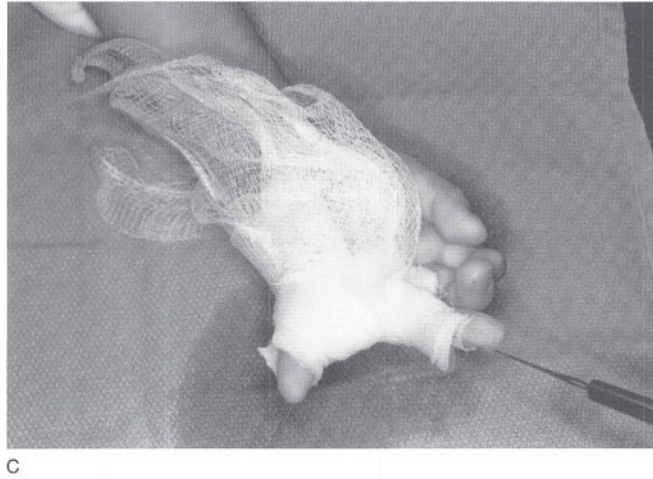
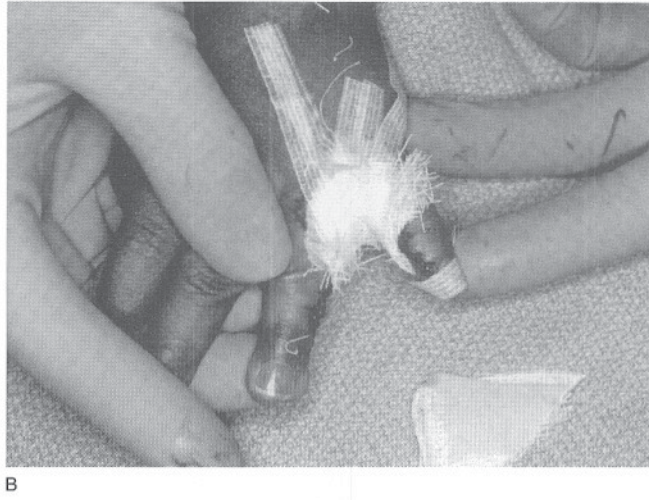
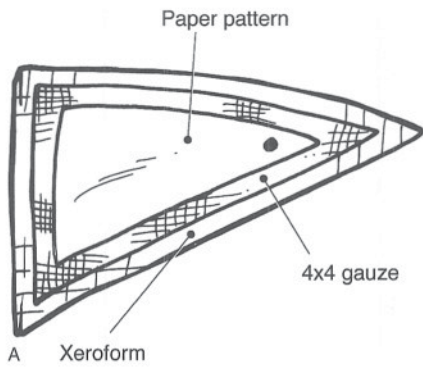


FIGURE 14-98 See legend on opposite page



FIGURE 14–99 When the cast is removed, parent and patient are reassured and instructed that the small sutures and dried skin will quickly wash off with a soap-and-water washing routine followed by a massage using petroleum jelly several times a day. With this routine, the skin rapidly returns to normal texture and softness. The scars are reddish for several months.

to a good result. Our selection of the type of treatment varies with the severity and type of web deficiency. The mainstays of our treatment are local flaps, grafts, and variations on the Z-plasty, used alone and in combination.

The milder forms of incomplete webbing of the thumb-index web are often well treated with the four-flap Z-plasty (Fig. 14–100A). The Z-plasty is ideally suited for application here, since it rearranges the excessively distal skin of the partial syndactyly by 90 degrees to increase the span between the metacarpal heads of the thumb and index finger. The four-flap variant improves on the simple Z-plasty in this location because it forms a more smoothly rounded web. As smaller amounts of palmar skin that have no pigment are brought onto the dorsum, a more natural color match appearance is obtained. It is important that the initial three limbs be constructed to be (1) of equal length and (2) oriented at 90 degrees to each other. Then the two flaps created are simply bisected to create 45-degree-angle flaps (Figs. 14–100B and C).

When the syndactyly is cutaneous but more complete, the dorsal flap augmented by skin grafts, just as used in reconstruction of the finger webs, can be very effective (Fig. 4–101).

In the child with clasped thumb or similar variants of the arthrogryptic hand, the skin web of the index finger and thumb has an unusual anterior orientation unlike that seen in the usual first web syndactyly. In the arthrogryptic child, we have often found that combining a transverse

incision to release the thumb web anterior to the MP joint with an index finger rotational flap to fill the defect on the thumb is unexpectedly effective for these special thumbs (Fig. 14–102). Depending on the amount of anterior webbing on the index finger, this may or may not require back grafting the index finger donor site. This method can also be used for the moderately severe types of the more typical deficient first web/adduction contracture but usually requires a back graft in these cases.

More severe loss of first web skin is better treated with a larger rotation flap from the dorsum of the hand, followed by skin grafting of the donor defect.

Complications. The majority of complications from syndactyly treatment come from loss of grafts and improper design of flaps. This can usually be averted with careful planning or referral to a hand surgeon prior to treatment. There are some complex syndactyly digits that are better left unseparated when the resulting digit would be unstable or damage to a growth plate would result in progressive angulation of a digit. Occasionally, vascular anomalies may prevent separation.

“Web creep” has been blamed unjustly for failure of a graft to grow in the child’s finger. Web creep is most commonly due to poorly designed incisions, loss of grafts, operating on too young a child, or infection.

The treatment of complications of inadequate treatment is beyond the scope of this book. Such cases should be referred to a hand surgeon with a special interest in pediatric hand surgery.

REFERENCES

Syndactyly

1. Ezaki M: Syndactyly. In Green DP (ed): *Operative Hand Surgery*, 4th ed, p 414. New York, Churchill Livingstone, 1999.
2. Feingold M: Clinical evaluation of the child with skeletal dysplasia. *Orthop Clin North Am* 1976;7:291.
3. Flatt AE: *The Care of Congenital Hand Deformities*; 1st ed, p 170. St. Louis, CV Mosby Co, 1977.
4. Flatt AE: *The Care of Congenital Hand Anomalies*, 170-2nd ed, p 228. St. Louis, Quality Medical Publishing, 1994.
5. Light TR: Congenital anomalies: syndactyly, polydactyly, and cleft hand. In Peimer CE (ed): *Surgery of the Hand and Upper Extremity*, p 2112. New York, McGraw-Hill Health Professions Division, 1996.
6. Littler JW, Hentz R: Hand and upper extremity. *Reconstr Plast Surg* 1977;6:3313.
7. Peimer CA (ed): *Surgery of the Hand and Upper Extremity*, p 2095. New York, McGraw-Hill Health Professions Division, 1996.
8. Sommerkamp TG, Ezaki M, Carter PR, et al: The pulp plasty: a composite graft for complete syndactyly fingertip separations. *J Hand Surg* 1992; 17-A:15.
9. Upton J: Congenital anomalies of the hand and forearm. In McCarthy JG (ed): *Plastic Surgery*, vol 8, p 5279. Philadelphia, WB Saunders Co, 1990.

THUMB DYSPLASIA

History. A thumb abnormality in a child often leads to the diagnosis of other conditions. For purposes of discussion, thumb abnormalities can be grouped into tight thumbs, the “duplicated” thumb, small or hypoplastic thumb, angled thumbs, and miscellaneous types. Each of these categories starts with the presenting physical finding and generates a differential diagnosis to guide treatment. Detailed discus-

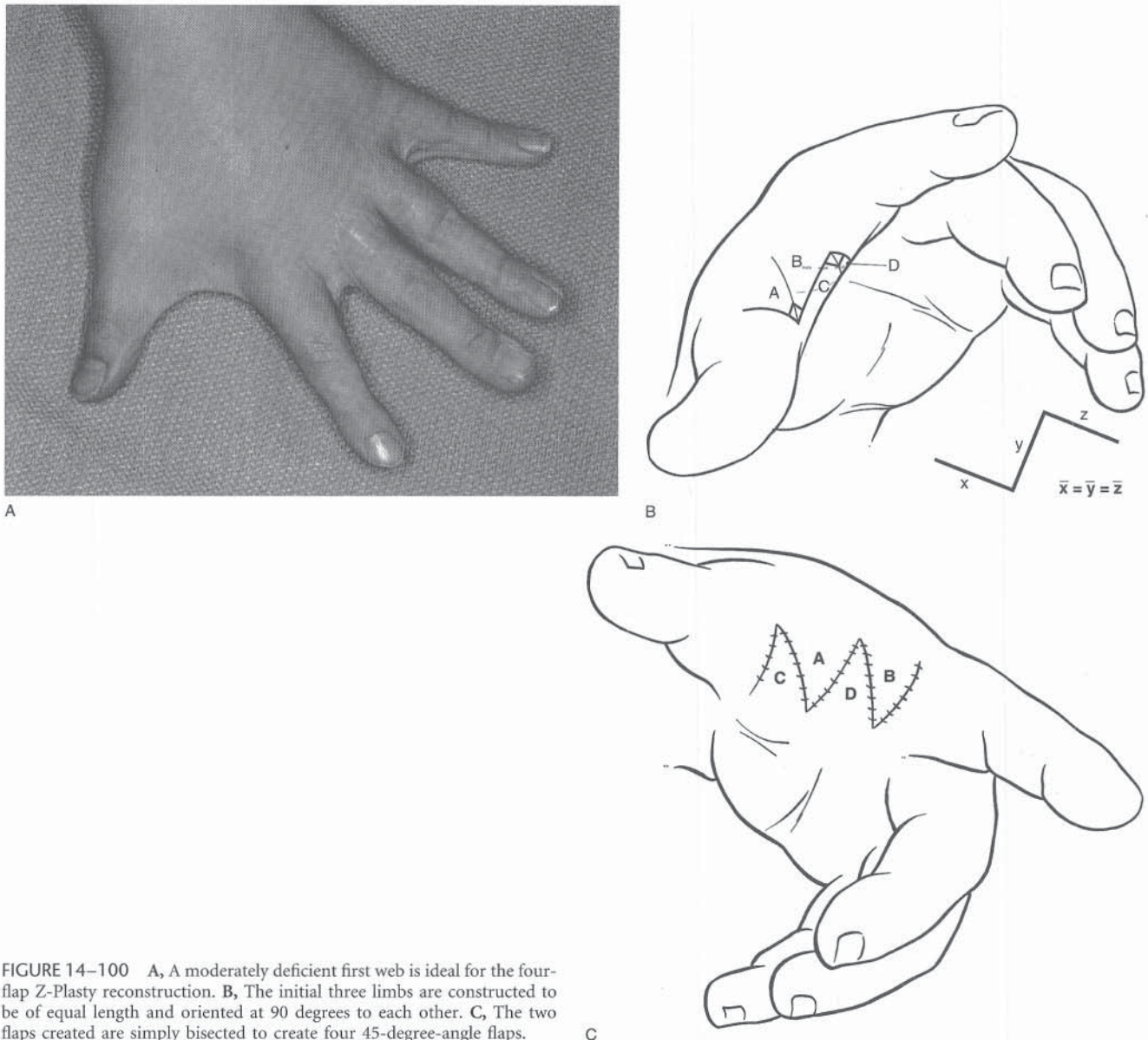


FIGURE 14-100 A, A moderately deficient first web is ideal for the four-flap Z-Plasty reconstruction. B, The initial three limbs are constructed to be of equal length and oriented at 90 degrees to each other. C, The two flaps created are simply bisected to create four 45-degree-angle flaps.

sions of classification, etiology, and treatment can be found in the references to this section.¹⁻⁷¹

Tight Thumbs. Tight thumbs include the trigger thumb, the spastic thumb, and the clasped thumb.

TRIGGER THUMB

Clinical Presentation. The IP joint of the thumb is held in fixed flexion. On occasion, the thumb will have been noted to trigger, but most are discovered after the thumb has settled into the locked position. The thumb is morphologically normal. A palpable nodule at the base of the MCP joint, known as Notta's node, can be palpated moving within the tendon as the distal joint is gently manipulated.

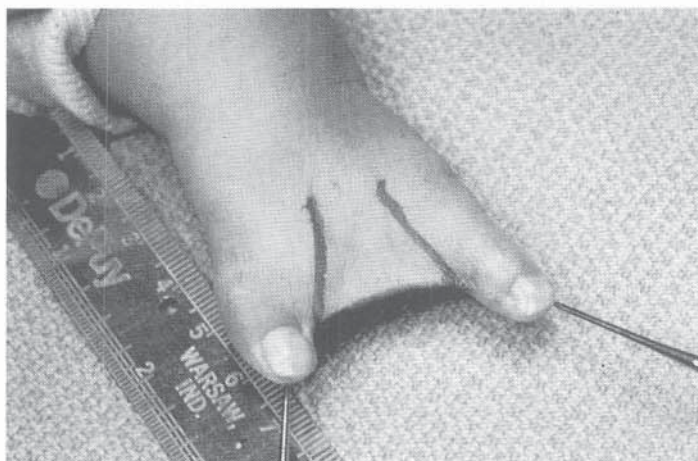
Etiology. Trigger thumb is not a congenital deformity. Trigger thumbs are developmental and often occur within the first 2 years of life. It has been reported that up to 25 percent of cases are bilateral, but may not present simultaneously.

Pathology. A thickening within the tendon forms a nodule at the region of the first annular pulley of the flexor sheath mechanism. With time the nodule enlarges to the point of preventing free excursion of the tendon within the sheath.

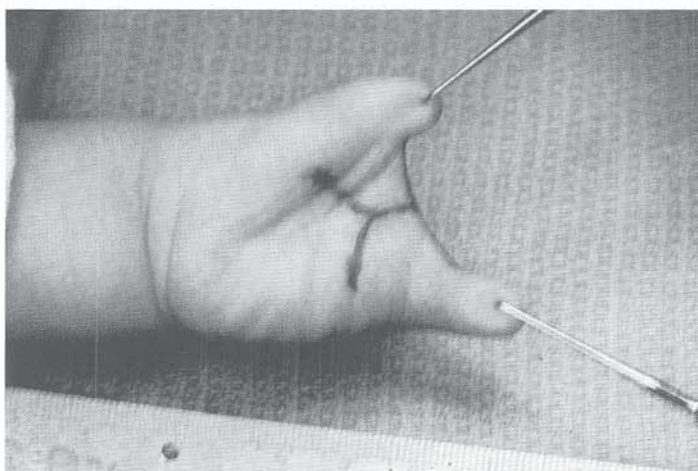
Imaging. Radiographs are not usually necessary but should be obtained if trauma is suspected.

Treatment. Although there are reports of nonoperative treatment with long-term splinting, this condition is a mechanical problem and the most expeditious treatment is trigger thumb release. This should be done under a general anesthetic and tourniquet control. Complete release of the offending proximal pulley will result in full motion and no recurrence. No more of the pulley mechanism should be released than is necessary.

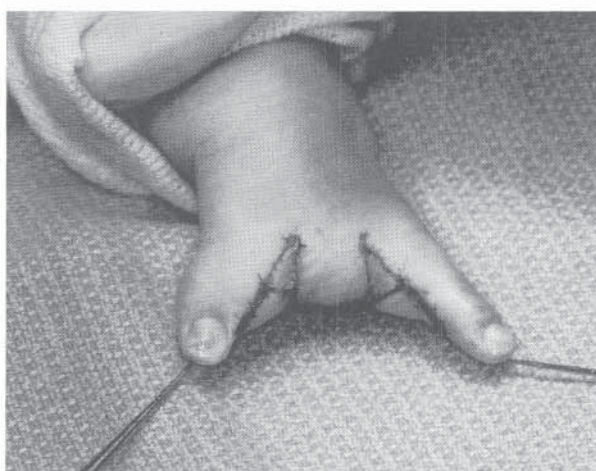
Complications. Complications of treatment include infection, damage to digital neurovascular structures, and excessive release of the pulley system with resultant bowstringing.



A



B



C

FIGURE 14-101 A to C, In more severe skin webs of the thumb, such as in this child with ulnar hemimelia, a dorsal flap, augmented with skin grafts much like those for a digital syndactyly, can be very effective. This procedure was followed by a shortening and rotational osteotomy of the first metacarpal and advancement of the abductor indicis to provide prehension.

THE SPASTIC THUMB

Clinical Presentation. The thumb-in-palm position is a normal one for a baby until about 3 months of age. After that time the digits begin staying out of the fist as the child begins to develop more advanced motor patterns, especially grasping for objects. Persistence of the thumb-in-palm posture becomes worrisome in the presence of other motor delay or asymmetric use of the upper limbs. The thumb is morphologically normal with no limitation in passive motion but with increase toned and failure to spontaneously bring the thumb out of the palm. The diagnosis of cerebral palsy should not be assigned until one is certain. Consultation with a neurologist may be helpful in identifying an etiology.

Etiology. Spasticity is the end pathway for the upper motor neuron dysfunction that may result from antenatal infarct, cerebral or meningeal infections, or some forms of syndromic spasticity.

Imaging. Imaging studies are usually not necessary unless bone procedures are contemplated.

Treatment. Initially, manipulation and perhaps splinting may help bring the thumb out of the palm. Any splint applied

must be carefully fitted to prevent hyperextension and instability of the MCP joint. Many children will improve greatly during the first several years of life, and surgical treatment should be delayed until the specific goals can be well defined and understood by the child's family. Surgery addresses the peripheral manifestations of CNS dysfunction and cannot produce a normally functioning limb.

Surgical treatment depends on the type and degree of deformity, the severity of the spasticity, and the age of the patient. Realistic goals can be set only after evaluation of all of the above. Surgical procedures include lengthening and release of spastic and contracted muscles, tendon transfers to effect better balance of the thumb, and stabilization of joints with capsulodeses or arthrodeses.

Results. The results of both surgical and nonsurgical treatment can be encouraging if the goals are carefully selected and normal function is not the goal. The long-term results may show deterioration because of the continual muscle imbalance, but in general, the position of the thumb can be improved and the results of surgery prolonged with careful attention to maintaining the correction with splinting until the completion of growth.

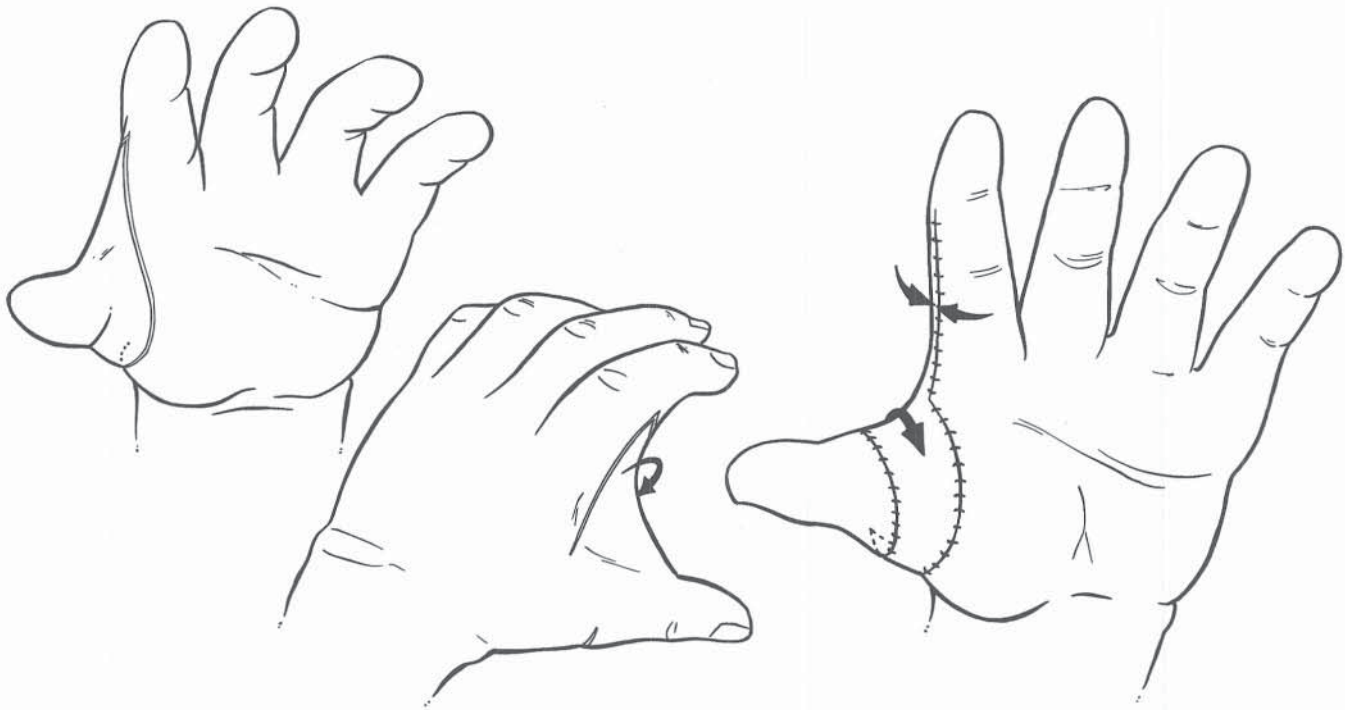


FIGURE 14-102 The rotation flap off of the index finger is very useful in cases of clapsed thumb or arthrogryposis. The unique anterior webbing in these patients often allows the donor area to be closed primarily without a skin graft.

CLASPED THUMB

Etiology. Clapsed thumb is the term given to a thumb that has morphological deficiencies in both intrinsic and extrinsic function as well as inadequate soft tissue and skin coverage. The condition is often associated with the forms of the distal arthrogryposis, and other malformations of the limbs may be present. Syndromic forms of arthrogryposis such as Freeman-Sheldon (“whistling face”) syndrome or Pena-Shokar syndrome are associated with a clapsed thumb as part of the spectrum of malformation. Careful examination of the child for other anomalies and genetic screening are indicated in a child with a clapsed thumb.

Clinical Presentation. The clapsed thumb cannot be passively extended at the CMC, MCP, or IP joint. There is a paucity of skin on the palmar aspect of the thumb, and the thumb-index web space is severely contracted. In some of the syndromic forms, the wrist has an extension contracture. In the mildest form the extensor brevis appears to be the only extrinsic tendon that is deficient. In the more severe forms, the other fingers are also involved and are held in a flexed and ulnarly deviated position, the so-called windblown hand deformity. There is a spectrum of presentation, and this type of thumb is often found in syndromic conditions.

Imaging. Imaging studies will demonstrate the abnormal posture of the thumb but will show no deficiencies in the bony anatomy. Later, joint irregularities from the lack of passive range of motion will become apparent.

Treatment. Treatment consists of passive stretching and splinting until there is no further change in position. Surgical treatment must address all of the components of the deformity and includes a comprehensive release of the intrinsic and extrinsic contracture, augmentation of the extrinsic ex-

tensors with tendon transfer, and soft tissue coverage with a either rotational flap and full-thickness skin graft or a free tissue transfer to cover the thumb-index web space. Failure to maintain adequate thumb extension can be addressed with a thumb MCP arthrodesis. Individual finger deformity can be addressed separately.

Results. The results of nonoperative treatment are always disappointing, owing to the extensive deficiencies in soft tissue elements. Surgical treatment is limited because of the extent of the involvement of soft tissue and the degree of contracture. Positional change and correction can be maintained with arthrodesis. Limitation in range of motion of the digit is to be expected, and hand function is compromised, especially when the other digits are involved.

THE “DUPLICATED” THUMB

Etiology. The so-called duplicated thumb is more appropriately termed a radial or preaxial polydactyly and is due to an error in segmentation. This abnormality can be due to one of two general mechanisms, an extra segmenting signal from the degrading apical ectodermal ridge in the final phase of differentiation, or an inborn patterning that results from a genetically programmed abnormality. In the first case the error occurs distally to proximally; in the second case the abnormal formation occurs proximally to distally.

Clinical Presentation. Genetic and dysmorphological classification separates thumb polydactyly into several broad categories. The first is the nonhereditary, unilateral, nonsyndromic thumb polydactyly that is due to an abnormality in the distal to proximal influence of the segmenting effect of the apical ectodermal ridge. This presents as either a symmetric or asymmetric “extra” thumb. Radiographically, the tissues are normal to the level of the bony bifurcation. The distal pre-

sentation depends on the symmetry, the tendon insertion, the joint involvement, and the condition of the thumbnail. The sum of the composite tissues of the two smaller thumbs is equivalent to the mass of a normal thumb. It must be pointed out to parents that on the abnormal side, the two smaller thumbs together make up about the same amount of tissue as the opposite normal thumb. This type of thumb is described by the Wassel I through VI classification.

The second type of thumb has an extra phalanx. This may be seen on radiographs or suspected from a widened space for a phalanx that is delayed in its ossification. This type of thumb is classified as a Wassel VII and further subclassified according to the complexity of the metacarpal and phalangeal anatomy. The clinical findings include bilaterality and a positive family history, both of which indicate a genetic pattern. This is the most frequent type of polydactyly in patients of Chinese descent.

The third type of thumb polydactyly is really a "duplication" of the index finger. The hand appears to have five fingers, the most radial lacking opposition. This type of radial polydactyly is occasionally associated with other malformations, including cardiac and hematopoietic ones.

The fourth type of thumb polydactyly is associated with multiple polydactyly and syndactyly and may also be associated with craniofacial syndromes.

Imaging. Plain radiographs establish the level of bony abnormality and are important in planning surgical reconstruction. Other imaging studies are not needed.

Treatment. The polydactylous thumbs are treated electively when the child is sufficiently large enough to ensure safe anesthesia, to fit a tourniquet, and to make surgical manipulation of the digits easier. Because the size of the child's hand nearly doubles in the first year after birth, waiting for a year is a reasonable surgical plan.

The surgical treatment is one of reconstruction of all of the components of the digit in one surgical procedure. Joints should be aligned with an osteotomy or judicious cartilage shaving, the capsule and ligament must be reconstructed and secured, the tendons must be balanced and centralized, and the soft tissue envelope must be repaired to leave a sensate, well-covered thumb. Usually the part of the thumb to be deleted is quite clear. In the case of nearly symmetric thumb components, priority should be given to saving the ulnar thumb because of the integrity of the ulnar collateral ligament of the MCP joint. Each of the musculotendinous units inserting onto or affecting the motion of the thumb must then be addressed to balance the adductor with the abductor, the short abductor with the long and short extensors, and the long flexors and extensors. Centralization of the flexor and extensor tendons at their insertions must be done carefully to avoid damage to the physis of the distal phalanx.

In the more complicated segmentation anomalies of the thumb, priority should be given to the bony stability of the base of the thumb and to the quality of the soft tissue envelope at the thumb tip. Reconstruction of some types of thumbs may require combining components of both thumbs. Decisions about the best reconstructive option may have to be made intraoperatively.

Results. The results of reconstruction of the radial polydactylies depend on the pattern of thumb anomaly present, the

success of realigning the joints in the thumb, and the forces of pull of the tendons. Most reconstructed thumbs are functional and are used constantly. Most complaints after reconstruction relate to the appearance of the thumb, in particular the size and scarring of the nail, lack of flexion of the IP joint, and residual angulation. The best results from surgical reconstruction are consistently with the types of duplications that do not involve either the IP or MCP joints. Residual instability of the IP joint is common and difficult to control by soft tissue reinforcement. Procedures that attempt to recombine the outside halves of nail and distal phalanx often result in unsightly thumb tips and dissatisfaction with the appearance of the thumb and thumbnail. Some of the postoperative results are to be expected as the best reconstruction possible given the quality and amount of available tissue. Other late complaints are related to failure to achieve skeletal and soft tissue balance to maintain alignment. Parents should be prepared for the potential for residual angulation or instability, and for the persistence of size discrepancy between the two thumbs.

THE SMALL OR HYPOPLASTIC THUMB

Etiology. Thumb hypoplasia is part of a dysplasia that involves the developing radial half of the forelimb. The radius may or may not be hypoplastic; however, the carpus almost invariably is underdeveloped. Each child with thumb hypoplasia must be evaluated for other anomalies known to be associated with radial dysplasia. These include but are not limited to Holt-Oram syndrome, the VACTERL association (vertebral, anal, cardiac, tracheoesophageal fistula, renal, radial, and leg abnormalities), Fanconi's anemia, thrombocytopenia-absent radius syndrome, and other blood dyscrasias. The child with TAR syndrome will invariably have abnormal thumbs but will have no radii.

Pathology. Diminished size or absent intrinsic and extrinsic musculature, small skeletal elements, and instability of the joints of the thumb contribute to the functional deficit.

Clinical Presentation. Depending on the severity of involvement, the child with a hypoplastic thumb may not be recognized until fine motor skills are required and the thumb is noted to be too small. Blauth classified the types of thumb hypoplasia,⁴ and the definitions have been modified by Manske and others. The mildest, type 1, involves smaller than normal size of the thumb and diminution of the bulk of the intrinsic musculature. Type 2 adds a diminished thumb-index web space, instability of the MCP ulnar collateral ligament, and inability to bring the thumb into palmar abduction. Type 3A adds to this further diminution of the size of the thumb metacarpal, although with a stable CMC joint. Type 3B has an unstable CMC joint. Type 4 is the "floating" thumb, or *pouce flottant*, held to the hand by a small pedicle and not stabilized to the skeleton of the hand by any bony connection. Type 5 is the completely absent thumb. With greater involvement of the thumb, one can expect abnormalities in the radial fingers as well.

Imaging. Plain radiographs help to assess the quality of the basal joint of the thumb and to examine the other digits.

Treatment. Reconstruction of the hypoplastic thumb addresses each of the missing components. In types 1, 2, and 3A thumbs, opponens tendon transfer with either a ring

superficialis or an adductor digiti quinti, plus stabilization of the ulnar collateral ligament of the MCP joint and opening of the thumb-index web space with a four-flap Z-plasty, will augment function.

Reconstruction of a severely hypoplastic type 3B, 4, or 5 thumb is rarely attempted in Western societies. In societies that demand five fingers on the hand, microsurgical reconstruction with free toe and soft tissue transfers have found a place. The alternative to retaining a nonfunctional digit is pollicization of the index finger. This procedure moves the index finger to the thumb position by shortening, rotating, and repositioning the hyperextensible MCP joint of the index finger. The function of the hand is improved in both grasp and pinch, and the cosmetic result is very acceptable.

Results. Results of reconstruction of the hypoplastic thumb depend directly on the quality of the residual thumb and radial side of the hand. If the tissues are supple, tendons are present, and the skeleton is stable, reconstruction will augment function and appearance. If the radial dysplasia is more severe and the joints and soft tissues are rigid, the results of reconstruction will be poor in terms of pinch and grip strength and flexibility. Function may, however, be improved by a change in position of the index finger.

ANGLED THUMBS

Etiology. The angled thumb may be found in some of the forms of aberrant segmentation, particularly in the triphalangeal types. It is also a characteristic thumb finding in Apert's syndrome, Rubenstein-Taybi syndrome, and some familial forms of polydactyly. The developmental etiology is unknown, but in the case of Apert's syndrome, the genetic defect is known to be in a fibroblast growth factor receptor site.

Pathology. Either the proximal phalanx or the additional triphalangeal bone is shaped abnormally such that the articular surfaces of the phalanx are not parallel. The shape may be trapezoidal or triangular, the so-called delta phalanx. The growth aberration may be a bracketed epiphysis in one plane only, or in some cases the growth plate may be abnormal in both radioulnar and dorsopalmar planes. In the case of the triphalangeal component, occasionally the additional ossification center may be within a single cartilage block lacking differential motion at the IP joint.

Imaging. Plain radiographs usually suffice to define the abnormality in the growth plate.

Treatment. It is important to determine whether the abnormal bone is a single block of cartilage with two ossification centers or truly two separate bones. If the offending bone segment moves as a unit, the treatment is to remove a wedge to bring the distal segment into alignment with the long axis of the thumb. If the abnormal bone is separate and trapezoidal in shape, a wedge osteotomy can be done to correct the malalignment after the bone is large enough to minimize damage to the growth plate. In the case of the delta phalanx with a true bracketed epiphysis, excision of the nonhorizontal portion of the growth plate with fat interposition may lead to longitudinal growth and effectively convert the delta to a trapezoidal shaped bone. The potential for longitudinal growth of the angled thumb is limited; therefore, the surgical intervention should be timed to max-

imize the length of the thumb along with correcting the deformity.

Complications. Iatrogenic injury to the intrinsically abnormal growth plate may cause further shortening of the thumb. Injury to the collateral ligament has the potential to cause instability of the joint.

MISCELLANEOUS THUMB ANOMALIES

Diastrophic Dwarfism. A characteristic finding in diastrophic dwarfism is the widely abducted "hitchhiker's thumb." The thumb is small but not hypoplastic in the spectrum of radial dysplasia. The extrinsic abductors appear to function, but the intrinsic function is severely deficient. The soft tissue envelope on the radial side is also deficient. The deformity is difficult to correct cosmetically, and functional results are usually poor.

REFERENCES

Thumb Dysplasia

1. Bayne LG: The hypoplastic thumb. In Dobyns JH, Wood VE, Bayne LG (eds): *Congenital Hand Deformities*, p 385. New York, Churchill Livingstone, 1993.
2. Bilhaut M: Guérison d'un pouce bifide par un nouveau procédé opératoire. *Congr Fr Chir* 1889;4:576.
3. Blair WF, Buckwalter JA: Congenital malposition of flexor pollicis longus: an anatomy note. *J Hand Surg* 1983;8-A:93.
4. Blauth W: Der hypoplastische Daumen. *Arch Orthop Unfallchir* 1967; 62:225.
5. Broadbent TR, Woolk RM: Flexion-adduction deformity of the thumb: congenital clasped thumb. *Plast Reconstr Surg* 1964;34:612.
6. Brunelli GA, Brunelli GR: Reconstruction of traumatic absence of the thumb in the adult by pollicization. *Hand Clin* 1992;8:41.
7. Buck-Gramcko D: Pollicization of the index finger: method and results in aplasia and hypoplasia of the thumb. *J Bone Joint Surg* 1971;53-A:1605.
8. Burton RI, Littler JW: Thumb reconstruction. In Everts CM (ed): *Surgery of the Musculoskeletal System*, vol 2, p 501. New York, Churchill Livingstone, 1983.
9. Call WH, Strickland JW: Functional hand reconstruction in the whistling-face syndrome. *J Hand Surg* 1981;6-A:148.
10. Carroll RE, Louis DS: Anomalies associated with radial dysplasia. *J Pediatr* 1974;84:409.
11. Chestnut R, James HE, Jones KL: The VATER association and spinal dysraphia. *Pediatr Neurosurg* 1992;18:144.
12. Dinham JM, Meggitt BF: Trigger thumbs in children: a review of the natural history and indications for treatment in 105 patients. *J Bone Joint Surg* 1974;56-B:153.
13. Dobyns J: Duplicate thumbs. In Green DP (ed): *Operative Hand Surgery*. New York, Churchill Livingstone, 1993.
14. Dobyns J: Trigger digits. In Dobyns JH, Wood VE, Bayne LG (eds): *Congenital Hand Deformities*, p 374. New York, Churchill Livingstone, 1993.
15. Erhardt R: *Developmental Hand Dysfunction: Theory, Assessment, Treatment*. Tucson, AZ, Therapy Skill Builders, 1989.
16. Flatt A: *The Care of Congenital Hand Anomalies*, 2nd ed, p 67. St. Louis, Quality Medical Publishing, 1994.
17. Ger E, Kupcha P, Ger D: The management of trigger thumb in children. *J Hand Surg* 1991;16-A:944.
18. Gilbert A: Toe transfers for congenital hand defects. *J Hand Surg* 1982;7-A:118.
19. Goldberg MJ: *Syndactyly and polydactyly in the dysmorphic child: an orthopaedic perspective*. New York, Raven Press, 1987.
20. Harris M: Reconstruction of the child's hand. In Carter P (ed): *Milestones in Pediatric Anesthesia Risk*, p 3. Philadelphia, Lea & Febiger, 1991.
21. Hentz VR: Congenital anomalies of the thumb. In McCarthy JG, May JW, Littler JW (eds): *Plastic Surgery*, p 5106. Philadelphia, WB Saunders Co, 1990.

22. Hentz VR, Littler JW: Abduction-pronation and recession of second (index) metacarpal in thumb agenesis. *J Hand Surg* 1977;2-A:113.
23. Jaeger M, Refior HJ: The congenital triangular deformity of the tubular bones of hand and foot. *Clin Orthop* 1971;81:139.
24. Jennings JF, Peimer CA, Sherwin FS: Reduction osteotomy for triphalangeal thumb: an 11-year review. *J Hand Surg* 1992;17-A:8.
25. Johnson CF: Abnormal thumbs and physical diagnosis. *Clin Pediatr (Phila)* 1970;9:131.
26. Kitayama Y, Tsukada S: Patterns of arterial distribution in the duplicated thumb. *Plast Reconstr Surg* 1983;72:535.
27. Kleinman WB: Management of thumb hypoplasia. *Hand Clin* 1990;6:617.
28. Lewis O: *Functional Morphology of the Evolving Hand and Foot*. Oxford, Clarendon Press, 1989.
29. Light TR, Ogden JA: The longitudinal epiphyseal bracket: implications for surgical correction. *J Pediatr Orthop* 1981;1:299.
30. Linscheid RL: Presidential address: The hand and evolution. *J Hand Surg* 1993;18-A:181.
31. Linscheid RL, An KM, Gross RM: Quantitative analysis of the intrinsic muscles of the hand. *Clin Anat* 1991;4:265.
32. Lister G: Pollex abductus in hypoplasia and duplication of the thumb. *J Hand Surg* 1991;16-A:626.
33. Littler JW, Cooley S: Opposition of the thumb and its restriction by abductor digiti quinti transfer. *J Bone Joint Surg* 1963;45-A:1389.
34. Manske PR, McCarroll HR Jr: Abductor digiti minimi opponensplasty in congenital radial dysplasia. *J Hand Surg* 1978;3-A:552.
35. Manske PR, McCarroll HR Jr: Index finger pollicization for a congenitally absent or nonfunctioning thumb. *J Hand Surg* 1985;10-A:606.
36. Manske PR, McCarroll HR Jr: Reconstruction of the congenitally deficient thumb. *Hand Clin* 1992;8:177.
37. Marks TW, Bayne LG: Polydactyly of the thumb: abnormal anatomy and treatment. *J Hand Surg* 1978;3-A:107.
38. Marzke MW: Evolutionary development of the human thumb. *Hand Clin* 1992;8:1.
39. McCarroll HR Jr: Congenital flexion deformities of the thumb. *Hand Clin* 1985;1:567.
40. McCarroll HR Jr, Manske PR: The windblown hand: correction of the complex clasped thumb deformity. *Hand Clin* 1992;8:147.
41. Miura T: Duplicated thumb. *Plast Reconstr Surg* 1982;69:470.
42. Miura T: Triphalangeal thumb. *Plast Reconstr Surg* 1976;58:587.
43. Moy OJ, Peimer CA, Sherwin FS: Reconstruction of traumatic or congenital amputation of the thumb by distraction-lengthening. *Hand Clin* 1992;8:57.
44. Mubarak SJ, O'Brien TJ, Davids JR: Metatarsal epiphyseal bracket: treatment by central physiylisis. *J Pediatr Orthop* 1993;13:5.
45. Ogino T, Minami A, Fukuda K: Abductor digiti minimi opponensplasty in hypoplastic thumb. *J Hand Surg* 1986;11-B:372.
46. Penfield W, Boldrey E: Somatic motor and sensory representation in the cerebral cortex of man as studied by electrical stimulation. *Brain* 1937;60:389.
47. Penfield W, Rasmussen T: *The Cerebral Cortex of Man: A Clinical Study of Localization of Function*. New York, Macmillan, 1950.
48. Poznanski A: *The Hand in Radiologic Diagnosis*. Philadelphia, WB Saunders Co, 1984.
49. Poznanski AK, Garn SM, Holt JF: The thumb in the congenital malformation syndromes. *Radiology* 1971;100:115.
50. Romer AS, Parson TS: The nervous system. In Romer AS (ed): *The Vertebrate Body*. Philadelphia, WB Saunders Co, 1977.
51. Romer AS, Parson TS: Supporting tissues: the skeleton. In Romer AS (ed): *The Vertebrate Body*, p 203. Philadelphia, WB Saunders Co, 1977.
52. Skov O, Bach A, Hammer A: Trigger thumbs in children: a follow-up study of 37 children below 15 years of age. *J Hand Surg* 1990;15-B:466.
53. Smith RJ: Osteotomy for "delta-phalanx" deformity. *Clin Orthop* 1977;123:91.
54. Strauch B: Dorsal thumb flap for release of adduction contracture of the first web space. *Bull Hosp Jt Dis* 1975;36:34.
55. Strauch B, Spinner M: Congenital anomaly of the thumb: absent intrinsic and flexor pollicis longus. *J Bone Joint Surg* 1976;58-A:115.
56. Swanson AB: A classification for congenital limb malformations. *J Hand Surg* 1976;1-A:8.
57. Tada K, Yonenobu K, Tsuyuguchi Y, et al: Duplication of the thumb: a retrospective review of two hundred and thirty-seven cases. *J Bone Joint Surg* 1983;65-A:584.
58. Temtamy SA, McKusick VA: Polydactyly. In Bergsma D (ed): *The Genetics of Hand Malformations: The National Foundation—March of Dimes*. Birth Defects: Original Article Series, vol 14, no 3, p 364. New York, Alan R Liss, 1978.
59. Theander G, Carstam N: [Longitudinally bracketed diaphysis]. *Ann Radiol (Paris)* 1974;17:355.
60. Tsuyuguchi Y, Tada K, Kawaii H: Splint therapy for trigger finger in children. *Arch Phys Med Rehabil* 1983;64:75.
61. Upton J: Congenital anomalies of the hand and forearm. In McCarthy JG (ed): *Plastic Surgery*, vol 8, p 5213. Philadelphia, WB Saunders Co, 1990.
62. van Genechten F: Familial trigger thumb in children. *Hand* 1982;14:56.
63. Vickers D: Physiylisis for treatment of delta phalanx of the thumb. Presented at the Third International Workshop for Congenital Hand Differences, Salt Lake City, UT, 1994.
64. Wassel HD: The results of surgery for polydactyly of the thumb: a review. *Clin Orthop* 1969;64:175.
65. Weckesser E: Congenital flexion-adduction deformity of the thumb (congenital "clasped thumb"). *J Bone Joint Surg* 1955;37-A:977.
66. Weckesser EC, Reed JR, Heiple KG: Congenital clasped thumb (congenital flexion-adduction deformity of the thumb): a syndrome, not a specific entity. *J Bone Joint Surg* 1968;50-A:1417.
67. Wood VE: Polydactyly and the triphalangeal thumb. *J Hand Surg* 1978;3-A:436.
68. Wood VE, Biondi J: Treatment of the windblown hand. *J Hand Surg* 1990;15-A:431.
69. Woolf RM, Broadbent TR: The four-flap Z-plasty. *Plast Reconstr Surg* 1972;49:48.
70. Zaleske DJ: Development of the upper limb. *Hand Clin* 1985;1:383.
71. Zaleske DJ, Tsukurov O, Boehmer A, et al: Determining the locus of a human complex bilateral polysyndactyly with the polymerase chain. Presented at the annual meeting of the Pediatric Orthopedic Society of North America, Memphis, TN, 1994.

MACRODACTYLY

History. In the typical busy children's orthopaedic hospital, enlarged extremities are an unusual but not rare occurrence. Wood reports that Klein described the first authenticated case of true macrodactyly in 1824.²⁵ In the 175 years that have followed, 300 cases of macrodactyly of the hand and 60 cases of macrodactyly of the toes have been reported.²⁵ During this time it has become obvious that the causes of large extremities are many and varied. Even today misdiagnosis and confusion persist, partly because of the multitude of synonyms for the conditions and the relative rarity of many of the diagnoses. In this section the discussion will focus in some detail on true macrodactyly as it affects the hand. However, in order to put the condition in the proper context of other causes for enlarged parts, the differential diagnosis is briefly considered.

Enlarged digits, hands, and upper limbs are also seen in fibrous dysplasia, lymphedema, arteriovenous fistula and malformations (lymphangioma, hemangioma, Klippel-Trénaunay-Weber syndrome), multiple enchondromatosis, and the associated vascular variant—Mafucci's syndrome. In addition, Proteus syndrome, with its varied presentation and high association with enlarged fingers, is often misdiagnosed. The "protean" presentations of the syndrome may make it impossible to recognize initially. Its effects are seen in all three germ layers and are characterized by macrodactyly with or without hemihypertrophy (in 54 of 61 cases, 89 percent), thickening of the skin, lipomas and subcutaneous tumors, verrucous epidermal nevi, and macrocephaly. The reader is referred to the review article by Stricker on Proteus syndrome.¹⁹

Classification. Temtamy and McKusick used the word *pseudomacrodactyly* to include vascular malformations, enchondromatoses, fibrous dysplasia, congenital band syn-

drome, lymphedema, and lipomatosis.²⁰ They reserved the term *true macrodactyly* for those conditions in which the bone was also involved.²⁰

For our purposes, use of the term macrodactyly will be limited to the way it is most often used by the hand surgeon (who often has a less complete understanding of the many varied causes of macrodactyly than the pediatric orthopaedist). The discussion considers four types of macrodactyly whose treatment involves primarily the hand. The classification used by Upton is clinically convenient and is used here.²⁴ Type I and its treatment will be examined in some detail because of its frequency and unique characteristics. Other hand surgery references are included in the rare event that one of the more unusual causes presents to the reader.

Type I. Macrodactyly with nerve-oriented lipofibromatosis

- a. Static subtype
- b. Progressive subtype

Type II. Macrodactyly with neurofibromatosis (von Recklinghausen's disease)

Type III. Macrodactyly with hyperostosis

Type IV. Macrodactyly with hemihypertrophy

In type I, sometimes referred to as true macrodactyly, the enlarged portion of the hand is in a distribution of a major peripheral nerve, most often the median nerve. The area of enlargement is always associated with abnormally large nerves that are both grossly and microscopically infiltrated by large amounts of fat (Fig. 14–103). Importantly, no additional abnormalities of organs or other areas of the skeleton occur in this type. The many synonyms for type I macrodactyly include macrodystrophia lipomatosa, dactylo-megaly, gigantomegaly, local gigantism, clubfinger, megalodactyly, digital gigantism, fibrofatty hamartoma of nerves, and nerve-oriented lipofibromatosis. Tsuyuguchi has proposed practical criteria for the clinical diagnosis of this type.²³ The condition is present at birth or by the age of 3 years, and the enlarged digit is longer and wider than normal.

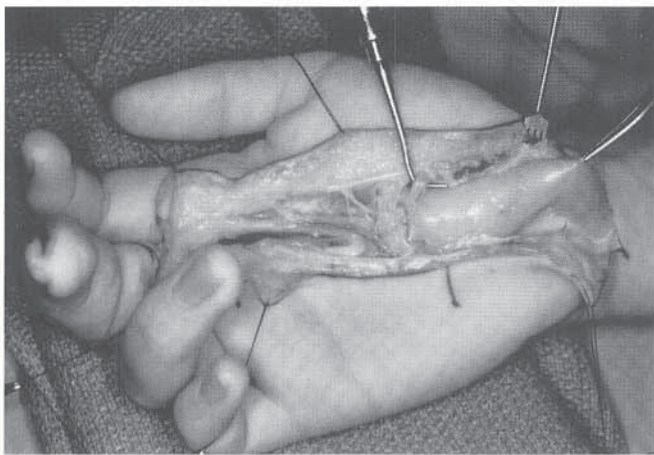
In type II macrodactyly, the digital enlargement is associated with the typical stigmata of neurofibromatosis: six or more café-au-lait spots, pedunculated cutaneous tumors,

and nodular peripheral nerves secondary to the multiple neurofibromas. The myriad of other skeletal problems associated with von Recklinghausen's disease, such as pseudarthrosis of the tibia and scoliosis, may be present in this type. Later in life, cartilaginous masses develop around the IP joints of the digits.

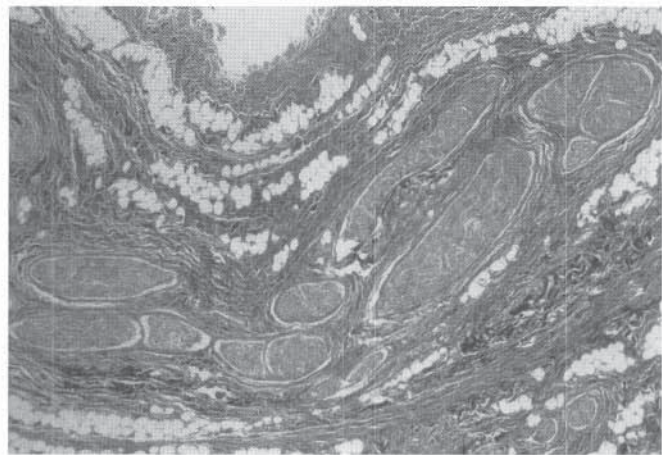
Type III macrodactyly is a very rare type of macrodactyly (17 reported cases) without abnormal nerves but associated with unusual hyperostosis or hypertrophic cartilaginous masses (Fig. 14–104). These masses result in enlarged metacarpal heads and phalanges without enchondroma. It is distinctly different from but has been confused with hereditary multiple osteocartilaginous exostoses. None of the stigmata of von Recklinghausen's disease are present.

Type IV macrodactyly is associated with a true hemihypertrophy of the ipsilateral upper and lower extremity. Present in the hand are multiple, atavistic, enlarged intrinsic and extrinsic muscles with abnormal insertions. These bizarre muscles cause a characteristic and distinct appearance of the hand, recognized by ulnar drift and flexion contracture of the MCP joints as well as by enlarged thenar and hypothenar eminences (Fig. 14–105). Hemihypertrophy is known to be associated with renal, adrenal, and brain tumors.^{3,8}

Etiology. The etiology of macrodactyly is unknown, but there has been abundant speculation. Early investigators postulated something inherently wrong with the part from its earliest embryologic formation, the so-called defective germ plasm.¹⁸ Often the word hamartoma is used in association with this condition since the enlargement involves multiple tissues but with one (the nerves) predominating.¹⁰ Steadman defines hamartoma as a focal malformation that resembles a neoplasm which results from faulty development. Hamartomas are composed of an abnormal mixture of tissue elements or an abnormal proportion of a single element naturally present at the site. Upton had noted that the derivation of hamartoma is from the Greek *hamartia*, to sin.²⁴ Because of the grotesque appearance of the enlarged part, a profane stigma frequently follows many of these children into adult life, and all too often they are the brunt



A



B

FIGURE 14–103 Type I (true) macrodactyly. **A**, The enlarged portion of the hand is in a distribution of a major peripheral nerve, most often the median nerve. **B**, The area of enlargement is always associated with abnormally large nerves that are grossly and microscopically infiltrated by large amounts of fat.

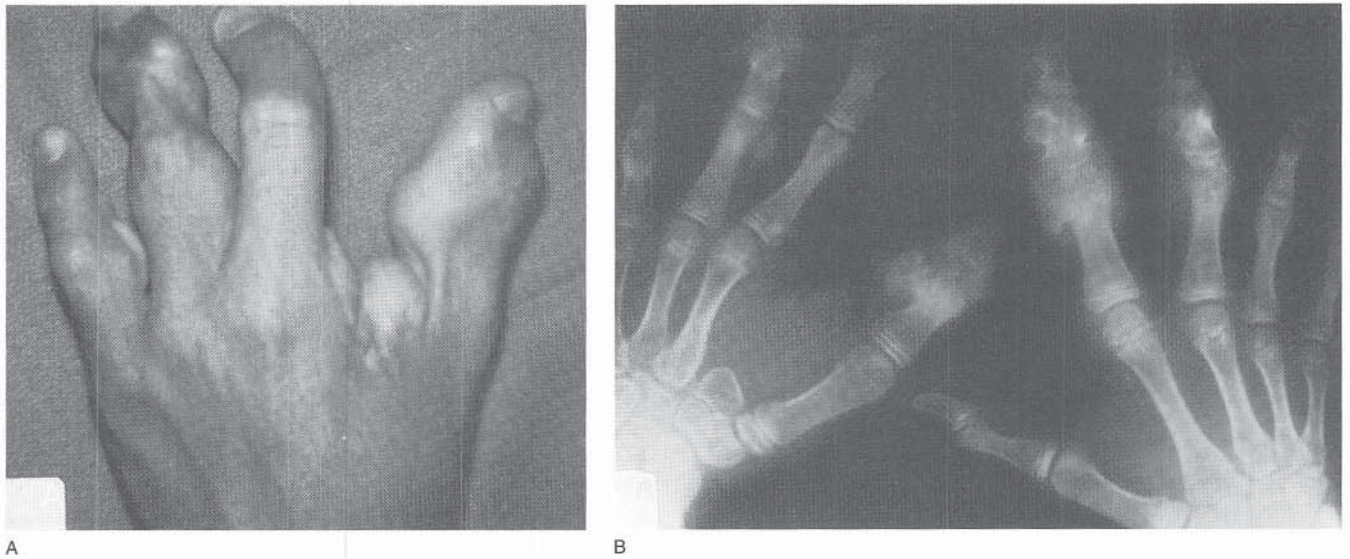


FIGURE 14-104 Type III macrodactyly, a very rare type of macrodactyly without abnormal nerves but associated with unusual hyperostosis or hypertrophic cartilaginous masses. **A**, The masses result in enlarged metacarpal heads and phalanges without enchondroma. **B**, Type III macrodactyly is distinctly different from but has been confused with hereditary multiple osteochondromatous exostoses. None of the signs of von Recklinghausen's disease are present. (From Dell PC: Macroductyly. *Hand Clin* 1985;1:517.)

of jokes and psychological injustice. We agree with Upton and feel that the term hamartomata is so nonspecific as to be more confusing than helpful. Considering its true derivation, the use of the word to describe these cases is, in fact, cruel.

Because of the usual association of macrodactyly with abnormalities of nerves, some have felt that all types were a localized variant or *forme fruste* of neurofibromatosis.³ Conversely, since many of the cases fail to exhibit the hallmark signs of von Recklinghausen's disease, other authors have felt that the condition is simply an impairment of peripheral nerves.¹⁵

Because of the important connection between normal growth and several hormones, investigators have looked for and found evidence of association with other conditions related to hormone abnormalities.¹¹ However, since the enlarged tissues in macrodactyly demonstrate such a local rather than generalized distribution, an aberrant end-organ or receptor of hormonal influence has been postulated. In fact, some evidence exists for an abnormal but localized growth hormone receptor.¹⁴

Still others have insisted that the process must involve a defect involving growth control mediated by the enlarged nerve. This was suggested by Japanese surgeons after the clinical observation that incision or resection of an involved nerve in some cases altered progressive growth.²²

Flatt has summed up the confusion well: "The unknown etiology of this peculiar state has prompted much speculation. . . . The best that can be said is that since these en-

largements occur in the sensory supply area of a major peripheral nerve, an abnormal neural control of growth is the most respectable current explanation."⁸

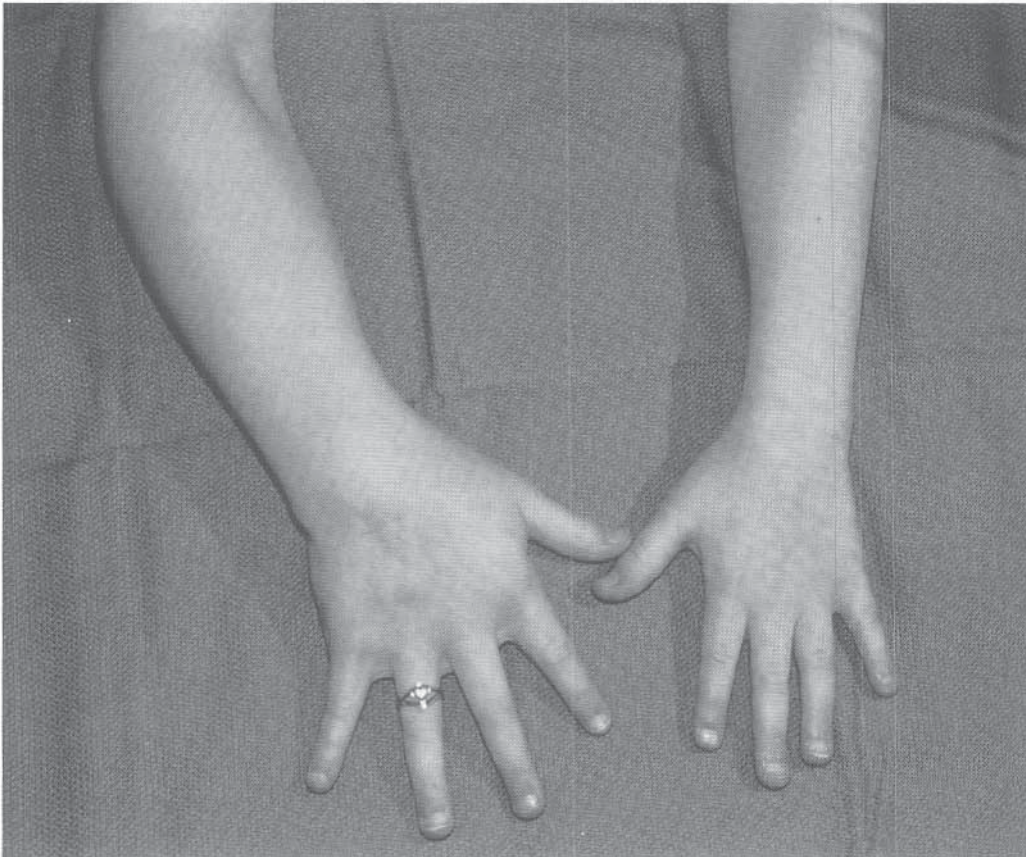
Pathology. The four types of macrodactyly have a characteristic histologic appearance, but it is not clear whether the condition is true hyperplasia or an enlargement of the cell type.²⁴

In type I macrodactyly, the major finding is the presence of excessive fat infiltrating the involved nerves and muscles. The nerves are distorted by significant endoneural and perineural fibrosis but the epineurium may or may not be fibrotic. The axons, while greatly distorted, remain normal in size and caliber.

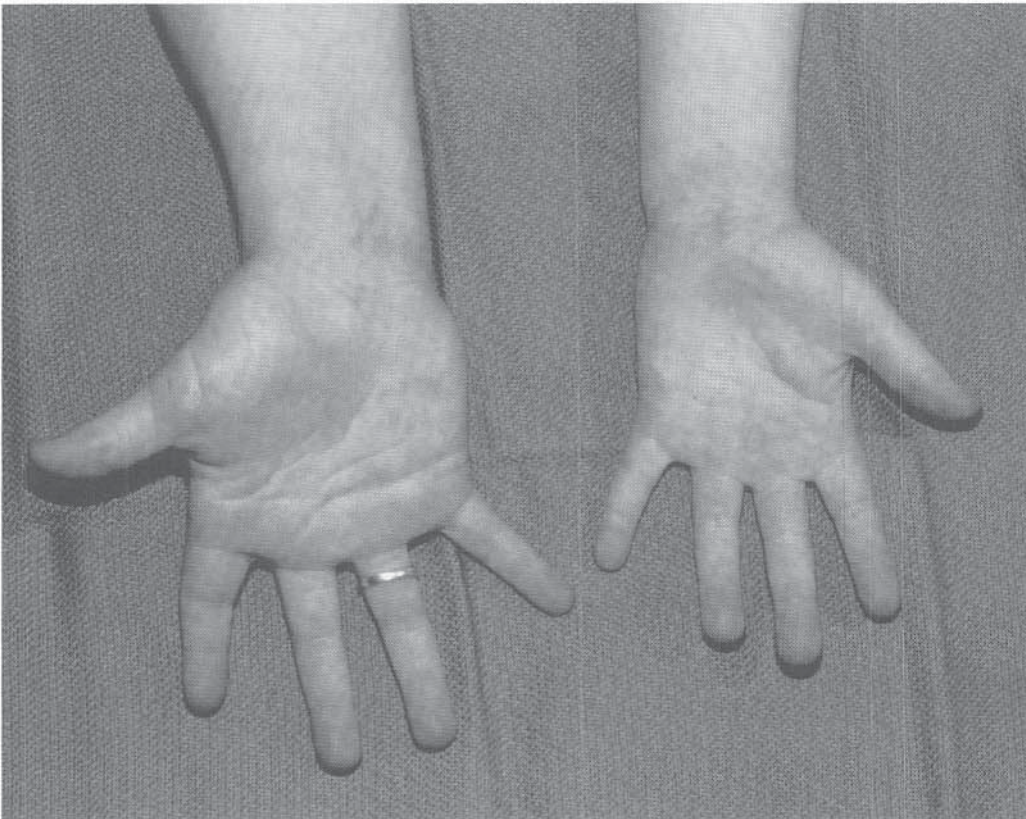
The bone contains large numbers of osteoblasts and osteoclasts and the periosteum is markedly thickened. There is an increase in enchondral and intramembranous bone formation. In the immature skeleton, bone remodeling is normal but occurs at an increased rate at the periosteal level.¹⁴ Pathologists describe this lesion using a variety of names: interfibrillary lipoma, fatty infiltration, lipomatous hamartoma, or lipofibroma of the median nerve. However, they are identical histologically.^{3,7}

Type II macrodactyly, the neurofibromatosis type, is associated with all of the usual signs of von Recklinghausen's disease. Histologic examination shows massive hypertrophy and distortion of nerve but with features distinctly different from those of type I macrodactyly. There is less excessive fat, and the predominant feature is proliferation of all of

FIGURE 14-105 Type IV macrodactyly is associated with a true hemihypertrophy of the ipsilateral upper and lower extremity. **A**, Present in the hand are multiple atavistic, enlarged intrinsic and extrinsic muscles with abnormal insertions. **B**, These bizarre muscles cause a characteristic and distinct appearance of the hand recognized by ulnar drift and flexion contracture of the MP joints as well as enlarged thenar and hypothenar eminences.



A



B

FIGURE 14-105 See legend on opposite page

the fibrous elements of the nerve (epineurium, endoneurium, and perineurium).

In type III, the very rare hyperostosis type, typical hypertrophic cartilaginous masses are present with somewhat larger nerves but no proliferation of fat or neural supporting cells. The enlarged metacarpal heads and phalanges do not contain enchondromas, and the condition is not to be confused with hereditary osteocartilaginous exostoses.

In the hemihypertrophy type IV macrodactyly, multiple, atavistic, enlarged intrinsic and extrinsic muscles with abnormal insertions are present. The muscles are not neoplastic, but the condition, like all hemihypertrophy, may be associated with renal adrenal and brain tumors.^{4,9}

Clinical Presentation. The condition is rare. Flatt reported 26 cases in 2,758 patients with hand deformity.⁸ The upper limb is more often involved than the lower limb. The condition may affect a single digit, multiple digits, the whole limb, or half of the entire body.⁸ Multiple digits are three times more likely to be involved than a solitary digit.⁴ The predilection of type I for the median nerve makes digital involvement much more common on the radial side, and the index and long fingers are by far the most frequently involved, the border digits least. Flatt's case distribution within the hand is shown in Figure 14-106. This distribution has been confirmed in both Dell's⁶ and Wood's²⁶ extensive reviews of the literature. There is equal sex distribution except in the lipofibromatosis form, where males outnumber females by about 3:2. Ninety percent of cases are unilateral, although the lipofibromatosis form is much less likely to be bilateral. While the type I lipofibromatosis nerve involvement is not usually associated with other conditions, it is seen occasionally with syndactyly, polydactyly, cryptorchidism, and nevi.⁸

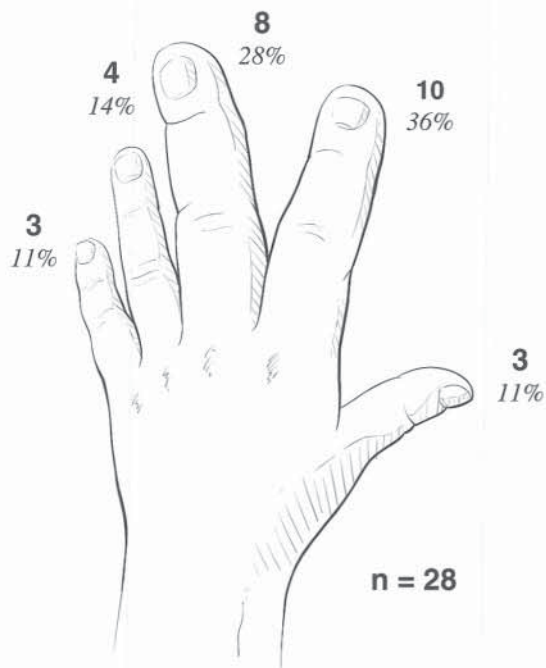


FIGURE 14-106 The predilection of type I macrodactyly for the median nerve makes digital involvement much more common on the radial side. The index and long fingers are by far the most frequently involved, the border digits least. Flatt's case distribution in the hand is shown. This distribution has been confirmed in both Dell's⁶ and Wood's²⁶ reviews of the literature.

In type I macrodactyly, with nerve-oriented lipofibromatosis, typically the enlargement is greatest distally and tapers proximally. The nerve enlargement, however, extends well proximal to the macrodactyly, often beginning in the distal forearm and gradually increasing to the tip of the involved digit. Nerve enlargement may occur without gigantism. In a series of lipofibromatosis hamartomas of the median nerves, six of 18 had no associated macrodactyly.¹

Clinodactyly of the enlarged digit is common, as is deviation into extension. The curvature is most severe when one side of the digit is involved and the other is not. The deformity always deviates toward the more normal side of the digit.

In type I macrodactyly, two different clinical presentations are seen. The child may be born with a large digit that continues to enlarge proportionately with the child's growth. This is the so-called static subtype. In the second or progressive subtype, the digits may be normal or nearly normal at birth, but by age 3 the digit begins to enlarge in a nonproportional and progressive manner. Digital overgrowth may be symmetric or asymmetric. When only half of the digit is involved, clinodactyly is the result. Overgrowth on the anterior surface usually exceeds that on the dorsal surface of the hand and digit, and because of this, most of these digits become hyperextended.

The natural history of the condition is one of progressive growth until epiphyseal closure. Joints become progressively stiff with age for two reasons, one physiologic and one psychological. The enlarged tissues prevent flexion, and the patient tends to hide the grotesque digit and avoid using it.

The symptoms of carpal tunnel syndrome—hand pain, finger numbness, and tenderness of the enlarged nerve at the carpal tunnel—occur commonly. However, measurable sensibility loss is rare.

In grotesquely enlarged extremities, symptoms of vascular insufficiency—cold intolerance and discoloration—often occur. The vessels, although larger, are normal. The clinical appearance of the various types is distinctive.

Type I or nerve territory-oriented macrodactyly (Keliian) is the most common type, is unilateral, is not inherited, and is not associated with other malformations. There are no signs of neurofibromatosis. Affected males outnumber females by a ratio of 3:2. The median nerve distribution and second web space are most commonly affected. When the involved fingers were large, 10 percent were associated with syndactyly.^{24(p 362)} Multiple digits are involved three times as often as a single digit, and in at least 90 percent of cases the condition is unilateral. Carpal tunnel syndrome is common in older patients. Barsky described two types, the progressive and static forms of the condition. Early in life it may be difficult to diagnose the progressive type or to differentiate it from the static form. Because of this, the children should be followed carefully during childhood. Documentation of hand and digit size with PA hand radiographs is essential.

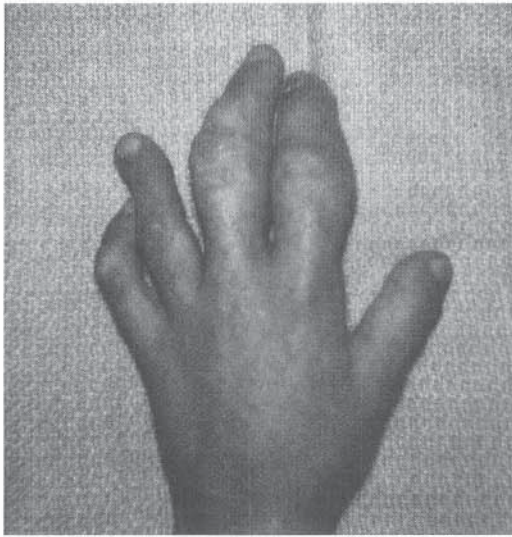
Frequently the static type, especially when not grotesque, tends to present in adolescence for treatment. Reasonably good function in spite of the unsightliness of the deformity can be present if the condition is limited to one or two digits.

In the progressive type, many affected babies are essentially normal at birth. However, by about 2 years of age, slow but unremitting growth begins and does not stop until the epiphyseal plates close in adolescence.

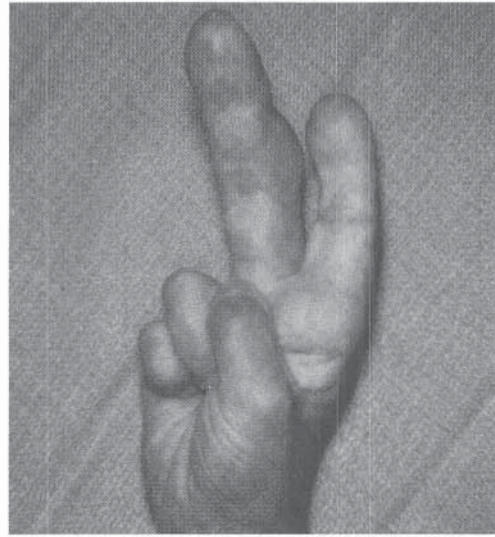
In type II macrodactyly, the gigantism is by definition associated with neurofibromatosis (von Recklinghausen's)—a condition with variable clinical expression, including the hallmark six or more café-au-lait spots, pedunculated cutaneous tumors, and multiple tumors of the peripheral nerves that become distinctively nodular. The condition is not particularly uncommon, having an incidence of 1 in 3,000. Although strongly inherited as an autosomal dominant condition, it is one of the most common spontaneous mutations resulting in new cases of a subsequently inherited disease. Skeletal involvement in other areas is seen in 30 percent of cases and includes scoliosis, bowing, and pseudarthrosis of the tibia. In addition to macrodactyly,

the many other associated problems of neurofibromatosis include seizures, mental retardation, astrocytoma, glioma, and pheochromocytoma. The onset of puberty aggravates the disorder, increasing the number and size of the cutaneous lesions.

There are distinctly different characteristics of digital gigantism in von Recklinghausen's disease. Involvement is often bilateral. The enlargement of digits is often associated with osteochondral masses around the epiphyses of the phalanges and metacarpals (Fig. 14–107). These masses, which are near joints, may obstruct motion. Particularly troublesome are cartilaginous masses in the volar plate, which prevent flexion of the digit. Although the nerves are enlarged



A



B



C

FIGURE 14–107 Type II macrodactyly. There are distinctly different characteristics of digital gigantism in von Recklinghausen's disease. A and B, Involvement is often bilateral. The enlargement of digits is often associated with osteochondral masses around the epiphyses of phalanges and metacarpal. These masses, which are near joints, may obstruct motion. Particularly troublesome are cartilaginous masses in the volar plate, which prevent flexion of the digit. C, Although the nerves are enlarged in the macrodactyly associated with neurofibromatosis, there is no fatty infiltration so characteristic of the lipofibromatous form of macrodactyly. (From Dell PC: *Macrodactyly*. *Hand Clin* 1985;1:516.)

in the macrodactyly associated with von Recklinghausen's neurofibromatosis, the fatty infiltration characteristic of the lipofibromatous form of macrodactyly is lacking. Type III macrodactyly, in which gigantism is associated with hyperostosis but not with neurofibromatosis, is very rare. Upton cites only 17 cases in literature.²⁴ This medical curiosity manifests with enlargement of the digits with localized overgrowth of skeletal structures. It may be bilateral but is usually asymmetric. The typical osteochondral growths, like those in neurofibromatosis, are adjacent to the epiphysis and palmar plates. Enlarged metacarpal heads and phalanges cause progressive loss of joint motion. These patients may have associated hyperostosis of the capitellum, producing subsequent radial head dislocation. The growths are to be distinguished from osteochondral exostosis, a condition with which they are often confused.

Type III macrodactyly is not inherited, there are no cutaneous spots or other manifestations of von Recklinghausen's disease, and there is no nerve abnormality. The lack of abnormal nerves is the important feature distinguishing this type from types I and II. However, most cases trace the sensory distribution of the median nerve in spite of its apparent normality.

Hypertrophic postoperative scars have tended to occur commonly in hand incisions after operation, and for this reason, careful placement of incisions in the midaxial line is especially important in type III macrodactyly.

In type IV macrodactyly, the enlargement is usually less grotesque and is seen invariably with enlargement of the entire upper extremities, or hemihypertrophy. Often, however, the parent may seek consultation after noticing only one enlarged extremity. Both forearm and arm enlargement

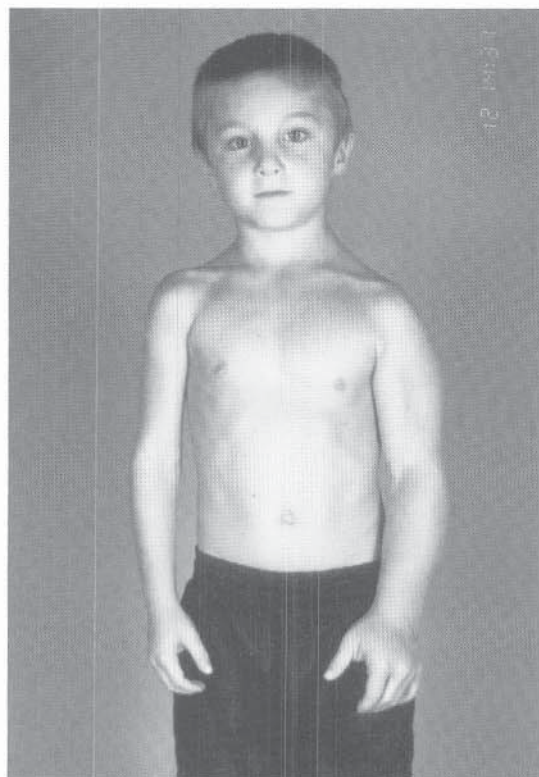
is usually present to some degree. The condition is rare and present at birth. It is not inherited and is always unilateral. There is no evidence of vascular malformation.

The hand in type IV macrodactyly has a unique and characteristic appearance. There is massive hypertrophy of the thenar and hypothenar musculature, adduction contracture of the thumb, and ulnar deviation of the fingers (Fig. 14–108). Although the joints are usually supple at birth, the natural history is one of progressive stiffness with flexion and ulnar deviation of joints by adolescence. Symptoms of vascular insufficiency are not seen, and the patient's symptoms relate to the progressive flexion contracture, ulnar deviation of the fingers, and the adduction contracture of the thumb.

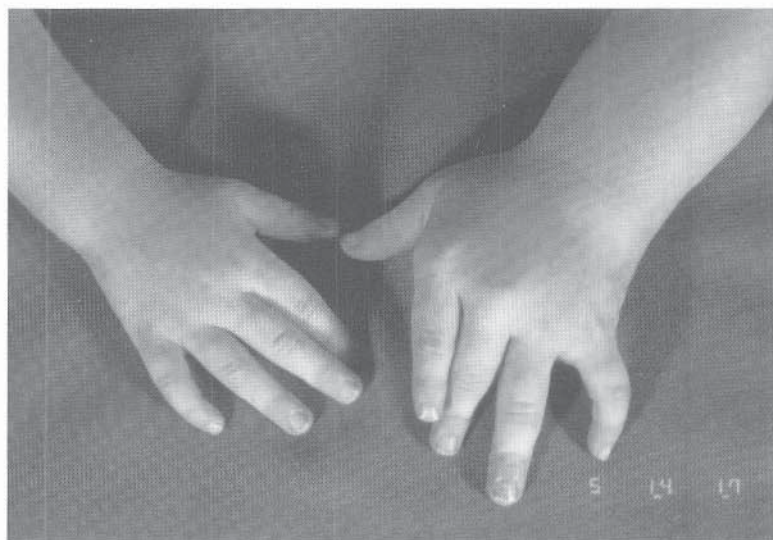
Imaging. Comparison radiographs should be obtained periodically to document growth. In milder cases only the phalanges are involved. In more severe cases the metacarpal is also enlarged. Periarticular growths are seen in the von Recklinghausen and hyperostotic macrodactyly types (types II and III). These growths may be confused with osteochondral exostosis (in which there is no true macrodactyly).

Arteriography differentiates the enlargement associated with vascular malformations from true macrodactyly. Vascular studies of true macrodactyly show enlargement of the digital artery on the side of the enlarged nerve.

Treatment. The reader will note that the same line drawing illustrations of ingenious operations devised by Barsky, Millesi, and Tsuge keep cropping up in hand surgery textbooks discussing the treatment of macrodactyly, but that the authors' presentations rarely include postoperative photographs. This should make the reasonable surgeon skeptical



A



B

FIGURE 14–108 A, The hand in type IV macrodactyly associated with hemihypertrophy has a unique and characteristic appearance. The thumb has massive hypertrophy of the thenar musculature and also enlarged hypothenar musculature. B, There is an adduction contracture of the thumb with MP flexion and ulnar deviation of the fingers.

about the quality of the postoperative result of an ingenious operation conceived on a sketch pad.^{8,17,24,25} As Dell points out, none of the illustrations in current textbooks adequately portray the deformities as they actually exist, and case reports as well as larger series usually do not show long-term postoperative results. The end result is never normal.⁶

It is an understatement to say that the treatment of these cases is among the most challenging and frustrating in all of pediatric hand surgery. Several previous authors have expressed this frustration. Upton noted, “There is no predictable way to inhibit local growth . . . the surgery tends to be repetitive and ablative.”²⁴ Flatt wisely emphasized the importance of explaining this carefully to the parents at the outset of treatment: “The parents of these unfortunate children must be honestly told of the potential number of operations necessary and of the limitation of function produced by the surgery. They must accept that at the end of it all the finger may look better but will still seem abnormal to strangers seeing it for the first time.”⁸

Unfortunately, the inadequacies of surgery are all the more distressing for the surgeon and family because of the social problems the deformity inflicts on these children. The psychological effect on a small child and family can be devastating; I know of one case of suicide in an otherwise attractive young woman, allegedly related to massive macrodactyly of her thumb.

On the other hand, when the parents understand the palliative nature of the treatment, repetitive, aggressive surgery may result in reasonably good appearance and function in many less severe cases. We agree with Upton that “the child should be followed carefully during infancy and early childhood and a disproportionately large digit(s) should *not* be allowed to go untreated through school ages.”²⁴ Although an early, aggressive approach is imperfect, it is far better than making the growing child endure the taunting of classmates while awaiting the completion of growth. As Flatt said, “Hopefully, by operating early, excessive growth can be minimized by epiphyseal arrest and some of the important secondary problems such as psychological disturbances, joint stiffness, and curvature of the finger may be avoided.”⁸

The goals of operative treatment of macrodactyly are:

1. To control or reduce size
2. To maintain useful sensibility
3. To maintain motion, especially at the MCP joint of the finger and the CMC joint of the thumb.

The methods used to achieve these goals can involve essentially all of hand surgery methodology:

1. Resection of skin and subcutaneous tissue, occasionally with full-thickness grafting
2. Nerve stripping or resection
3. Epiphysiodesis
4. Angulation osteotomy
5. Recession osteotomy
6. Narrowing osteotomy
7. Arthrodesis
8. Amputation
9. Multiple and combination operations

In younger children, an epiphyseal arrest should be done when the digit reaches adult length. This may be quite early in life; a comparison with the finger of the parent of the same sex is helpful. All involved epiphyses should be arrested. Some authors prefer to excise rather than drill the

phalangeal epiphysis, but in the small child this can make the joint very unstable. The resection of the joint during arthrodesis can effectively include the epiphysis.

The width of the bone must also be addressed, and this usually involves longitudinal resection. This surgery is always followed by tendon and joint fibrosis with a resultant stiff finger.

The soft tissue and nerve lesion is harder to treat. There have been many approaches to the enlarged nerve. Tsuge and Ikuta in type I and II (nerve-induced macrodactyly) recommended stripping of the nerves while attempting to preserve the main nerve trunk to avoid major sensory deficits. They reported a 6-year follow-up of 12 patients in whom growth stopped after the nerve-stripping operation.^{21,22} Kelikian in 1974 recommended excision of the tortuous segment of the nerve with direct repair of the two ends (Fig. 14–109).¹² McCarroll in 1950, reporting on gigantism induced in type II cases (von Recklinghausen’s), recommended removal of the nerve at a very early age in an effort to decrease the trophic influence of the nerve on growth.¹³

When the patient presents in adolescence with a grotesquely enlarged digit, the treatment is difficult and must be individualized. In older children with periarticular growths of the fingers, the PIP or DIP may need to be sacrificed with arthrodesis, and attention focused on maintaining as much MCP motion as possible. In general, in the older child or young adult with the severe form of macrodactyly, the operations are extensive and the quality of the result is more limited. Shortening is often the most practical treatment, ablating distal segment and nail with debulking of the remainder of the digit. Shortening but retaining part of the nail or distal phalanx is associated with a dog ear either dorsally or palmarly. Barsky resected the DIP joint and retained palmar sensation and a palmar dog ear (Fig. 14–110).² Tsuge resected palmar skin distally, the anterior half of the distal phalanx, and the dorsal half of the middle phalanx. The nail is moved dorsally with the phalanx, and a dog ear remains dorsally (Fig. 14–111).^{21,22}

When the enlargement is asymmetric, a resection of one

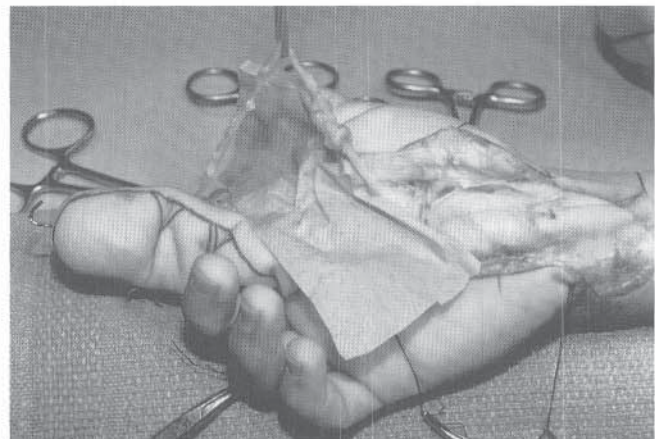


FIGURE 14–109 Kelikian in 1974 recommended excision of the tortuous segment of the nerve with direct repair of the two ends. As shown in this case, the digital nerve is not only increased in circumference but exceedingly redundant.

Barsky

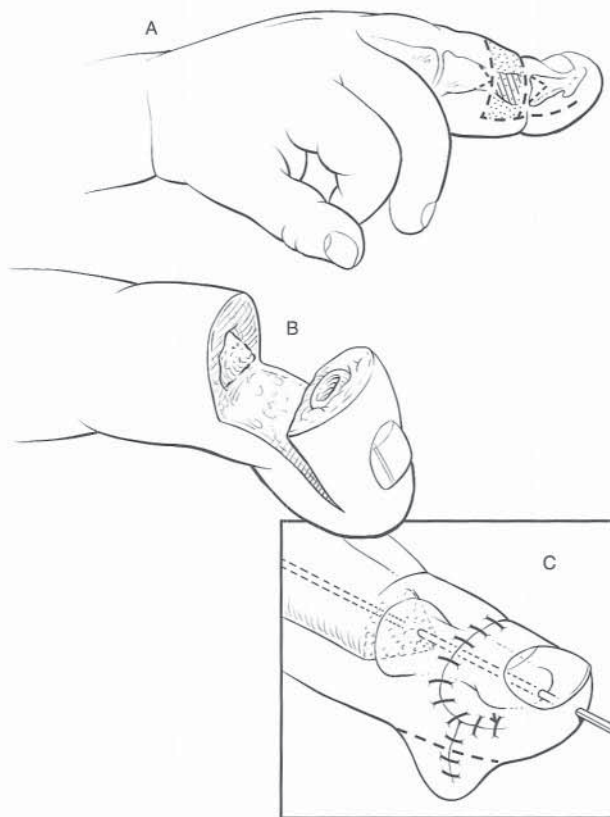


FIGURE 14-110 Shortening but retaining part of the nail or distal phalanx is associated with a dog ear either dorsally or palmarly. Barsky resected the DIP joint and retained palmar sensation and a palmar dog ear.

side of the digit with removal of the entire digital nerve can be effective. Skin slough and slow healing can be a problem (Fig. 14-112).

Surgical reduction of the width of the finger is challenging and often is unsatisfying. The improved appearance usually is associated with stiffness and nail deformity (Fig. 14-113).

Correction of deviated digits may require closing wedge osteotomies with or without epiphyseal plate closure, resection, or IP arthrodesis.

The thumb is a special case, since significant shortening can be gained by resecting the MCP joint and surrounding bone while preserving the CMC joint and IP joint. Soft tissue debulking is more difficult and in severe cases can be only palliative (Fig. 14-114).

The place of amputation in macrodactyly should not be forgotten. Although parents are usually reluctant to accept amputation unless the size is truly gargantuan, it is appropriate to gently include the subject of amputation in initial treatment discussions with the family. After a series of failed attempts to improve the appearance with lesser procedures, it may be more readily accepted by parent and child. Amputation is worthwhile when the digit is so insensate that it is a liability rather than an asset to the patient. Sometimes even at a very early age, amputation is a reasonable choice when the enlargement

is severe (Fig. 14-115). Unfortunately, after amputation the process in surrounding and less involved digits may increase, negating the favorable functional and cosmetic effects of the initial resection.⁶ For the osteochondral masses seen in neurofibromatosis and the rare hyperostotic gigantism, excision early to try to preserve joint motion is the goal. Later, arthrodesis of the digit in an optimal position reduces size and positions the digit in the optimal position for use.

Hemihypertrophy and macrodactyly (type IV) is a rare and special case when considering treatment. Early, many of these children are still relatively supple and may tolerate and respond to stretching and splinting. We have tried exploration and resection of the multiple enlarged and abnormal intrinsic muscles in these bizarre palms, with limited success. If necessary, when the child is older, a three-part treatment for the fully developed deformity may be necessary to improve hand opening, finger alignment, and the cosmetic appearance of thenar and hypothenar enlargement. These operations can be combined or staged as required. The first surgery is closing wedge metacarpal osteotomies to

Tsuge

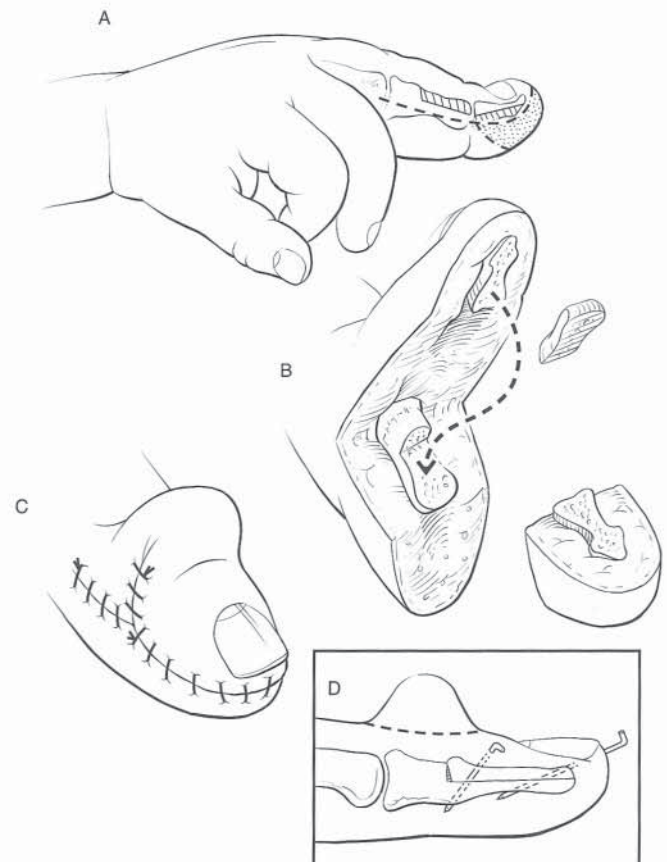


FIGURE 14-111 Shortening but retaining part of the nail or distal phalanx is associated with a dog ear either dorsally or palmarly. Tsuge resected the distal palmar skin, the anterior half of the distal phalanx, and the dorsal half of the middle phalanx. The nail is moved dorsally with the phalanx, and a dog ear remains dorsally.

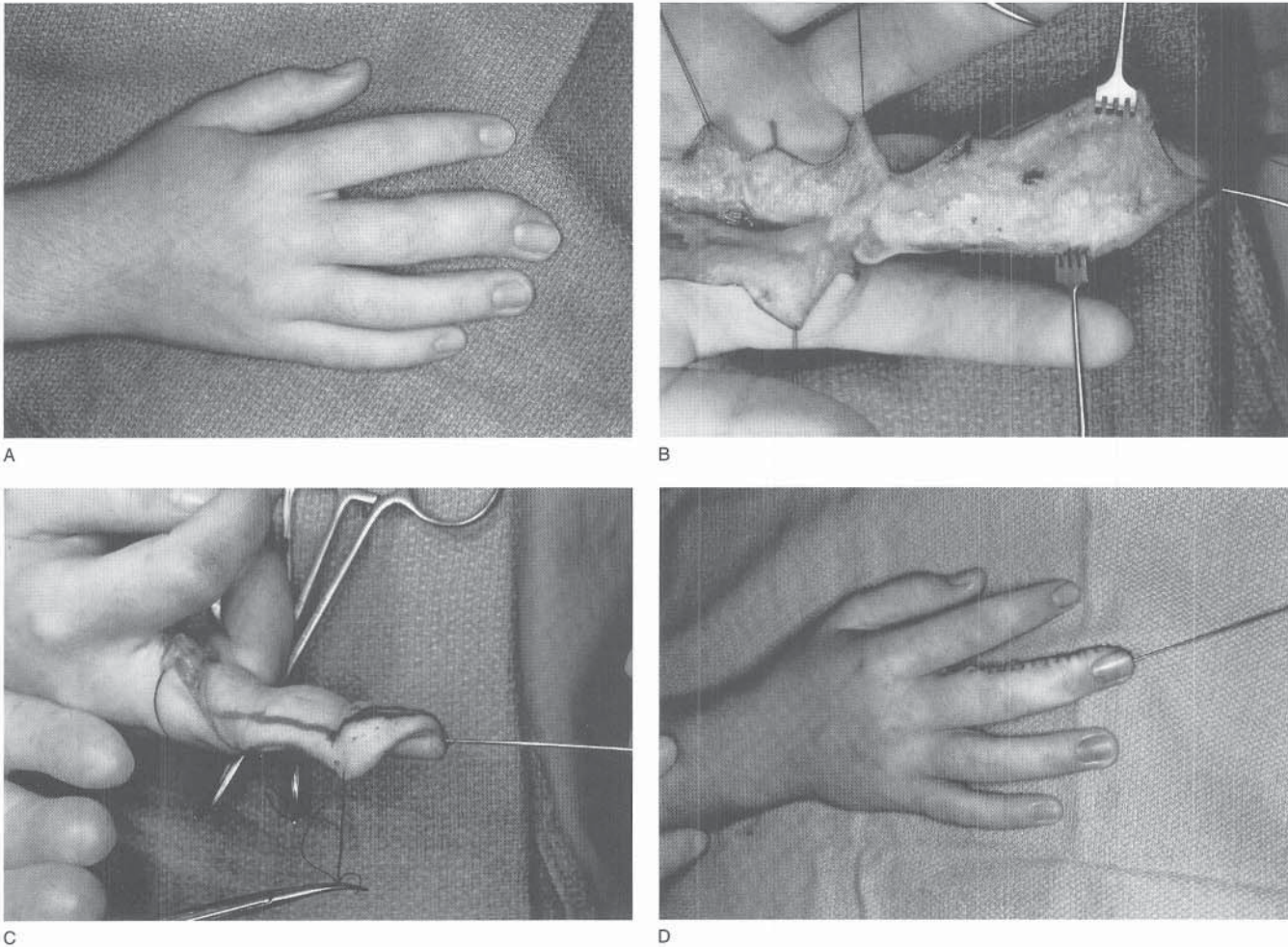


FIGURE 14-112 A to D, When the enlargement is asymmetric, resection of one side of the digit with resection of the entire digital nerve can be effective. Skin slough and slow healing can be a problem.

correct ulnar drift, the second surgery is release of deforming forces on the abnormal intrinsic muscles, and the third surgery is debulking of the thenar and hypothenar muscle masses.

Complications. The most distressing complication is the result in severe cases: The digit is still large, covered in scar,

and still attracts the morbid attention of the public. It may be stiff, insensate, and useless. After aggressive debulking, nail bed deformities, stiffness of joints, persistent grotesque enlargement, loss of sensibility, and skin and flap necrosis all too often are the rewards for this monumental surgical effort, the deceptively neat and clever line drawings of opera-

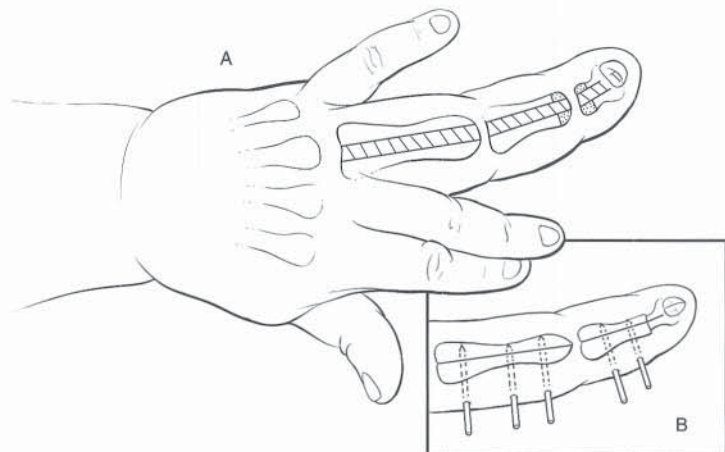


FIGURE 14-113 Reduction in the width of the finger is challenging and often unsatisfying. The improved appearance usually is associated with stiffness and nail deformity.

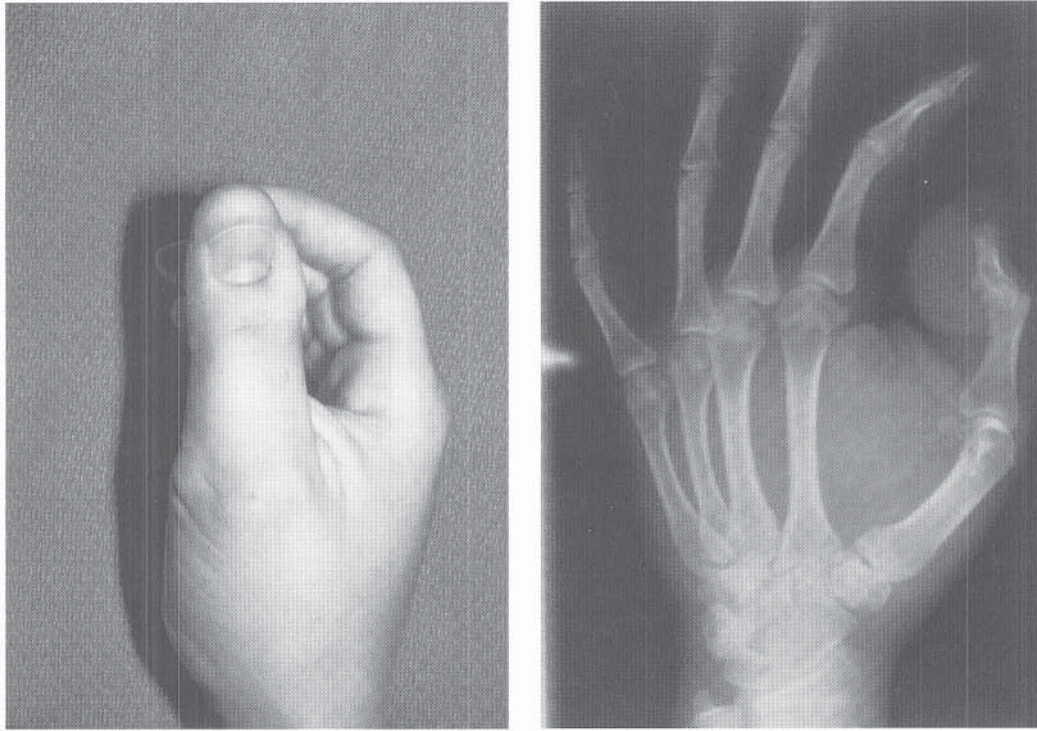


FIGURE 14-114 The thumb is a special case, since significant shortening can be gained by resecting the MP joint area and preserving the CMC joint and IP joint. A and B, Soft tissue debulking is more difficult and in severe cases can only be palliative.

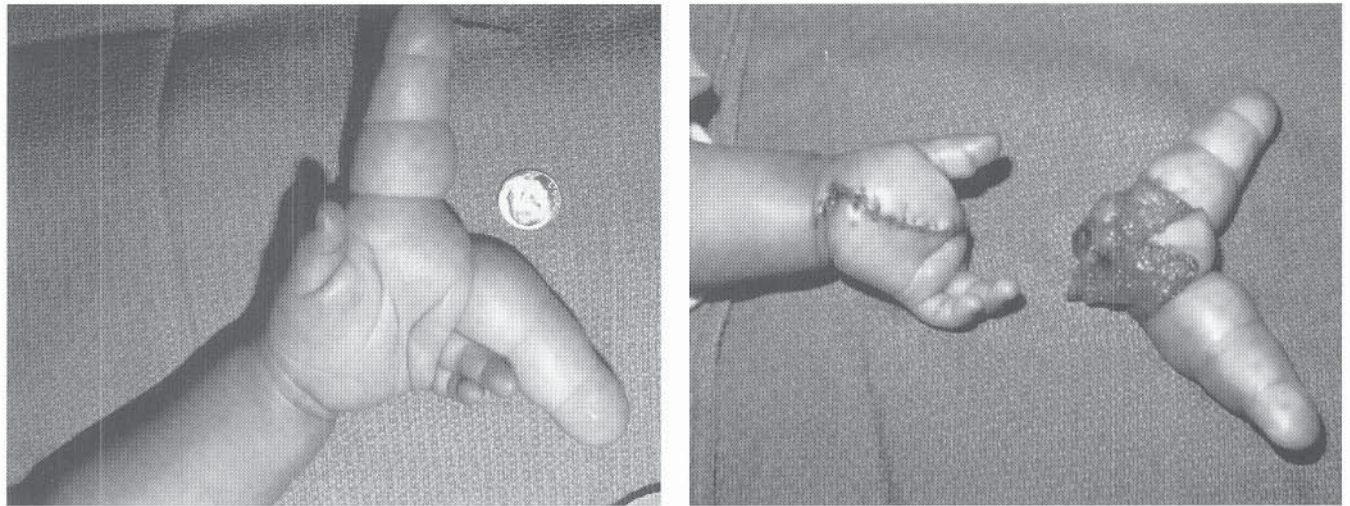


FIGURE 14-115 A and B, The place of amputation in macrodactyly should not be forgotten. Although the parent is usually reluctant to accept amputation unless the size is truly gargantuan, it is appropriate to gently include amputation in initial treatment discussions with the family. Even at a very early age, amputation may be a reasonable choice if the enlargement is severe. Unfortunately, after amputation the process in surrounding, less involved digits may increase, negating the favorable functional and cosmetic effects of the initial resection.

tions shown repeatedly in textbooks of hand surgery notwithstanding.

REFERENCES

Macroductyly

1. Amadio PC, Reiman HM, Dobyns JH: Lipofibromatous hamartoma of nerve. *J Hand Surg* 1988;13-A:67.
2. Barsky AJ: *Congenital Anomalies of the Hand Their Surgical Treatment*. Springfield, IL: Charles C Thomas, 1958.
3. Ben-Bassat MC, Kaplan I, Laron Z: Congenital macroductyly: a case report with a three-year follow-up. *J Bone Joint Surg* 1966;48-B:359.
4. Bjorklund S: Hemihypertrophy and Wilms's tumor. *Acta Paediatr* 1955;44:287.
5. Brooks L: The changes of the bone in von Recklinghausen's Neurofibromatosis. *Surg Gynecol Obstet* 1964;38:587.
6. Dell P: Macroductyly. *Hand Clin* 1985;1:511.
7. Edgerton MT, Tuerk DB, in Littler JW, Cramer LM, Smith JW (eds): *Symposium on Reconstructive Hand Surgery*, vol 9, p 157. St. Louis, CV Mosby Co, 1974.
8. Flatt AE: *The Care of Congenital Hand Anomalies*, 2nd ed. St. Louis, Quality Medical Publishing, 1994.
9. Fraumeni JF Jr, Miller RW: Adrenocortical neoplasms with hemihypertrophy, brain tumors, and other disorders. *J Pediatr* 1967;70:129.
10. Hueston JT, Millroy P: Macroductyly associated with hamartoma of major peripheral nerves. *Aust NZ J Surg* 1968;37:394.
11. Inglis K: Local gigantism (a manifestation of neurofibromatosis). *Am J Pathol* 1950;26:1059.
12. Kelikian H: *Congenital Deformities of the Hand and Forearm*. Philadelphia, WB Saunders Co, 1974.
13. McCarroll H: Clinical manifestations of congenital neurofibromatosis. *J Bone Joint Surg* 1950;32-A:601.
14. Minkowitz S, Minkowitz F: A morphological study of macroductyly: a case report. *J Pathol Bacteriol* 1965;90:323.
15. Moore B: Peripheral nerve changes associated with congenital deformities. *J Bone Joint Surg* 1944;26-A:282.
16. Ochi M, Yasunaga H, Ikuta Y, et al: Study on pathogenesis of macroductyly-somatomedin-c receptor on the growth plate. *J Jpn Soc Surg Hand* 1986;4:567.
17. Ogino T: Macroductyly. In Buck-Gramcko D (ed): *Congenital Malformations of the Hand and Forearm*, p 183. New York, Churchill Livingstone, 1998.
18. Streeter GL: Focal deficiencies in fetal tissues and their relation to intrauterine application. *Contrib Embryol* 1930;22:1.
19. Stricker S: Musculoskeletal manifestations of Proteus syndrome: report of two cases with literature review. *J Pediatr Orthop* 1992;12:667.
20. Temtamy SA, McKusick VA: Macroductyly. In Bergsma D (ed): *The Genetics of Hand Malformations: The National Foundation—March of Dimes. Birth Defects: Original Article Series*, vol 14, no 3, p 507. New York, Alan R Liss, 1978.
21. Tsuge K: Treatment of macroductyly. *Plast Reconstr Surg* 1967;39:590.
22. Tsuge K, Ikuta Y: Macroductyly and fibro-fatty proliferation of the median nerve. *Hiroshima J Med Sci* 1973;22:83.
23. Tsuyuguchi Y, Yonenobu S, Kawai H: Macroductyly: Treatment and Prognosis. *Rinshouseikeigeka* 1983;18:255.
24. Upton J: Congenital anomalies of the hand and forearm. In McCarthy JG (ed): *Plastic Surgery*, vol 8, p 5309. Philadelphia, WB Saunders Co, 1990.
25. Wood VE: Macroductyly. In Green DP (ed): *Green's Operative Hand Surgery*, vol 1, p 533. New York, Churchill Livingstone, 1999.
26. Wood VE: Macroductyly. *J Iowa Med Soc* 1969;59:922.

CONGENITAL BAND SYNDROME

History and Etiology. The syndrome of congenital banding has puzzled authors for centuries. Hippocrates wrote about children with amputations and constrictions related to amniotic bands. During the nineteenth century, it was postulated that there was an external or mechanical cause for the syndrome.¹ In the 1930s, Streeter popularized the concept of an internal defect within the embryo and discounted

the amniotic theory.⁵ Torpin reopened the strangulation argument,⁶ and the preponderance of the current literature favors the amniotic band theory.⁷ Even today the facts do not completely fit either argument, and there are thoughtful proponents of both arguments. Although Upton feels the association of clubfeet and cleft lip with this disorder cannot be explained by the mechanical theory,⁷ Ezaki² and others suggest that cleft lip can result from swallowing an amniotic strand. The clubfoot association is unexplained. Perhaps the perspective of Adrian Flatt is useful here: "A review of the literature is entertaining but unenlightening. Hippocrates first suggested that amniotic bands might press on a limb and produce deformities or amputations. Since then the etiological battle has swayed back and forth between intrinsic and extrinsic causes, with iatrogenic lesions of chickens and rats being used as evidence."³

There is general agreement that cases are not genetically predetermined and occur at random. Today the condition is considered by most to be a disruption sequence due to external mechanical causes.

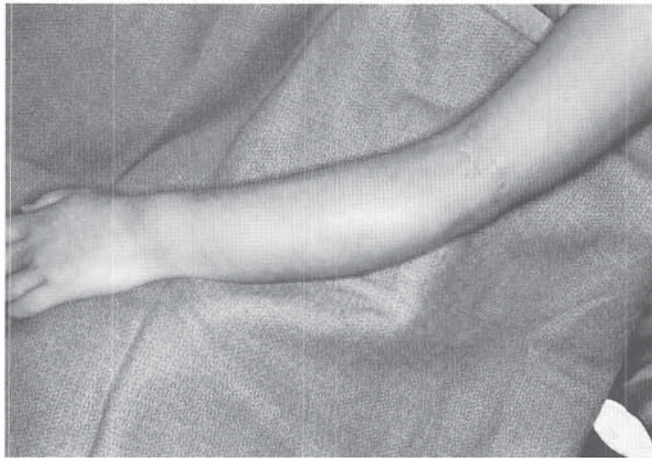
Pathology. Pieces of amnion and other material can be found in the depth of some clefts encircling and strangulating the digits. After the initial trauma, the defect heals and the resultant cleft may be superficial, involving only the skin and part of the subcutaneous tissue, or it may involve veins, nerves, and arteries. Fusions of bone and skin may develop. Neurologic deficits distal to these deep clefts may be severe (Fig. 14–116). Occasionally even the bone can be involved, and abnormalities of the tibia may occur in association with a band (Fig. 14–117).

Clinical Presentation. The prenatal history may include oligohydramnios, premature uterine contractions, and leakage of fluid. However, the mother often reports an uncomplicated pregnancy. Although prenatal diagnosis by ultrasonography is occasionally possible, for most parents the affected child comes as a surprise. Newer techniques, such as three-dimensional ultrasonography, may improve the prenatal diagnosis.

Although the condition of extremity banding of newborn children is well known to the pediatric and orthopaedic communities, confusion is caused by the existence of multiple names for the same condition, including Streeter's syndrome or dysplasia, annular bands, grooves or rings, constriction ring syndrome, congenital band syndrome, amniotic band syndrome, and amniotic disruption sequence.

Band syndrome is also confused with symbrachydactyly. In symbrachydactyly, a subclavian disruption sequence is thought to occur at about week 5 or 6. This vascular catastrophe affects the subectodermal mesenchyme while sparing the normal ectoderm (the source of the fingernail and distal phalangeal tuft). The child with symbrachydactyly has abnormal nerves, vessels, bones, and tendons proximally, but nails and a distal tuft are always present to some degree. The intrauterine event in band syndrome occurs later, after all of the elements of the limb have completely formed. In the amputated digits of band syndrome, the fingernails are absent but the remaining proximal tissues, which were forming normally prior to the traumatic amputation, tend to be normal (Fig. 14–118).

Acrosyndactyly may be seen in band syndrome and may result in very complex deformities. The distinguishing fea-



A



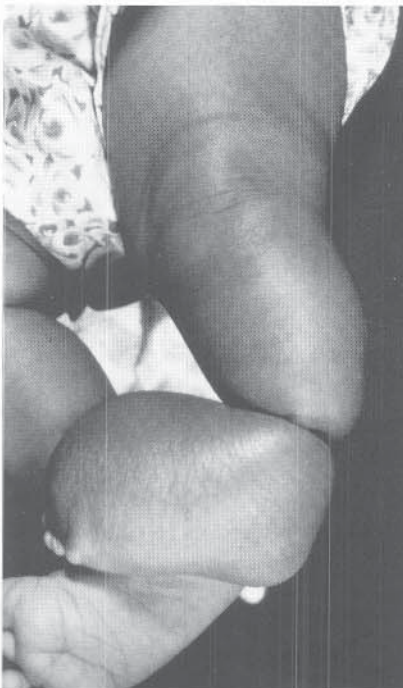
B

FIGURE 14-116 Congenital band syndrome. **A**, This child had a deep band just above the elbow with a complete median and ulnar nerve palsy. A Z-plasty improved the cosmetic defect, but paralysis persisted. **B**, This view shows the hand with a high median and ulnar palsy. Note the severe thenar atrophy and the supinated position of the paralyzed thumb.

ture of acrosyndactyly caused by banding is the presence of clefts or fistulas (Fig. 14-119). These clefts are the result of distal fusion of the healing fetal skin after the intrauterine injury. The clefts or sinus tracts tend to be distal to the normal web level. This can complicate to a variable degree the design of the dorsal flap to reconstruct a normal web more proximally.

Classification. Patterson has developed a useful classification for the extent of banding (Figs. 14-119 and 14-120):⁴

1. Simple constriction ring
2. Constriction ring with deformity of the distal part
3. Constriction with fusion of distal parts—acrosyndactyly
4. Complete intrauterine amputation



A



B



C

FIGURE 14-117 Congenital band syndrome of the lower extremity. **A**, This deep cleft on the distal tibia was associated with marked bowing of the tibia. **B** and **C**, After band reconstruction, the tibia and soft tissues improved.

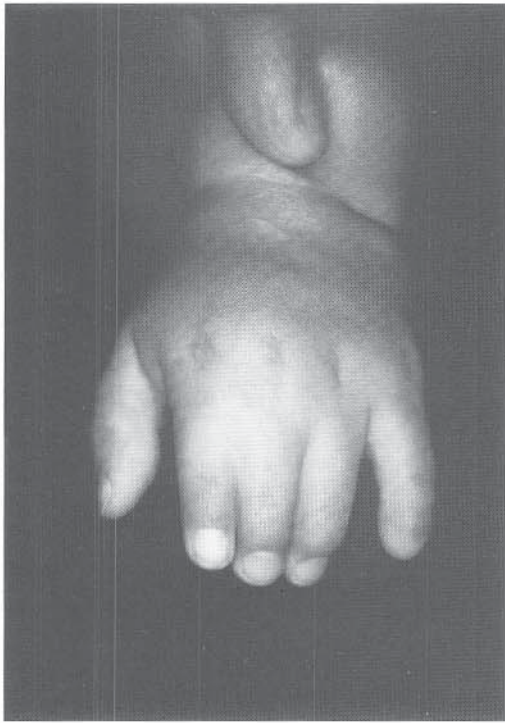


FIGURE 14-118 Digits amputated from congenital band syndrome are distinctly different from the digits in symbrachydactyly. Note the presence of the nails on the shortened fingers. The nail is absent on the short finger in Figure 14-120.

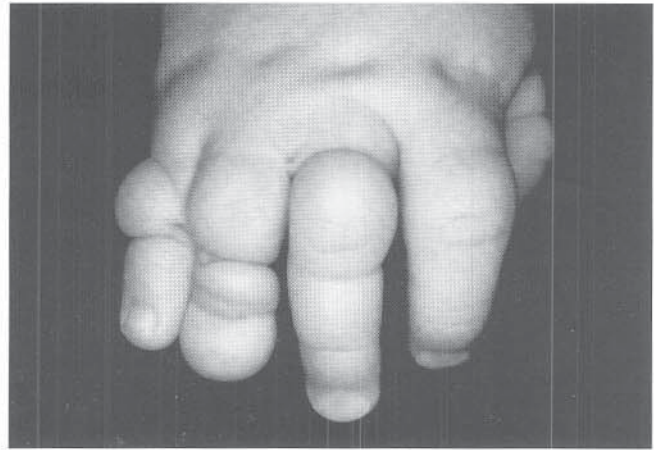
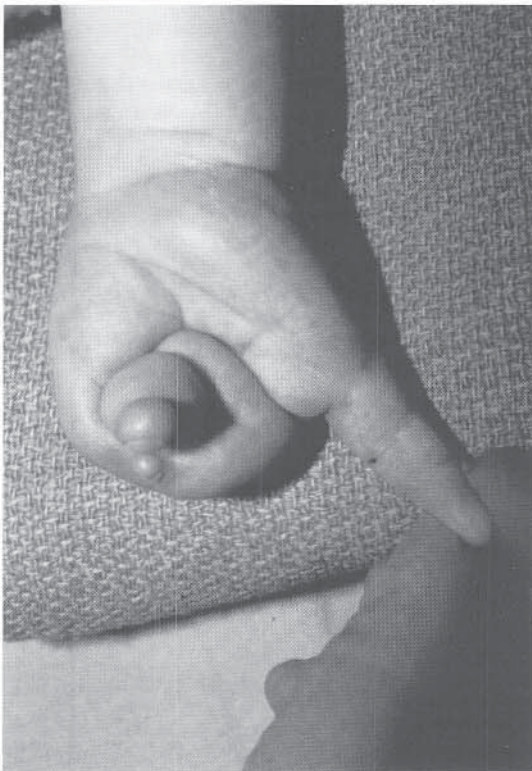


FIGURE 14-120 Three of Patterson's types of congenital banding are seen in this patient. The amputation of the tip of the ring finger is a type IV band (amputation). A type II band with deformity distal to the band is present in the small finger, in the midportion of the ring finger, and proximally in the long finger. A simple type I band with no deformity is present in the midportion of the long finger. For type III banding (acrosyndactyly), see Figure 14-119.

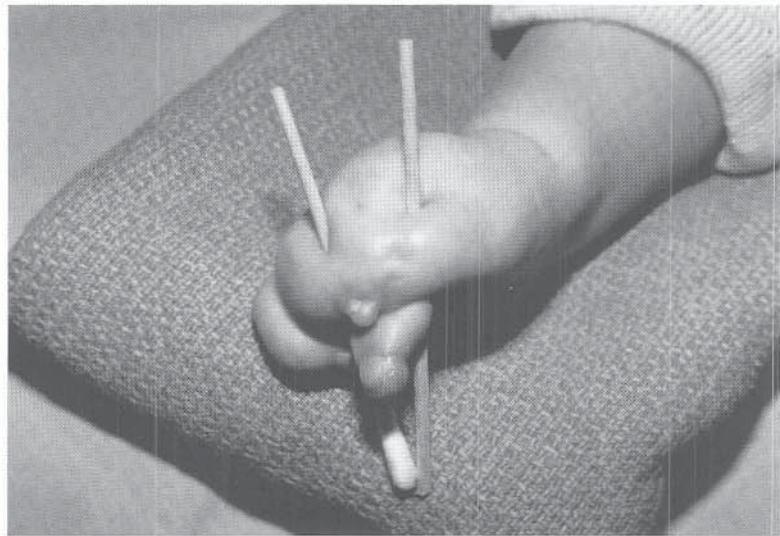
Imaging. No imaging techniques are required in cases of band syndrome other than radiography, which is useful to discover bone abnormalities.

Treatment

ACUTE TREATMENT. Since in most cases the amputation has occurred before delivery, it is rare that a newborn has an impending amputation that can be saved by the surgeon.



A



B

FIGURE 14-119 Acrosyndactyly may be seen in congenital band syndrome (Patterson's type III) and may result in very complex deformities. **A**, The distinguishing feature of acrosyndactyly caused by banding is the presence of clefts or fistulas. **B**, Note the applicator sticks passed through the sinus tracts left over from the base of the former web.

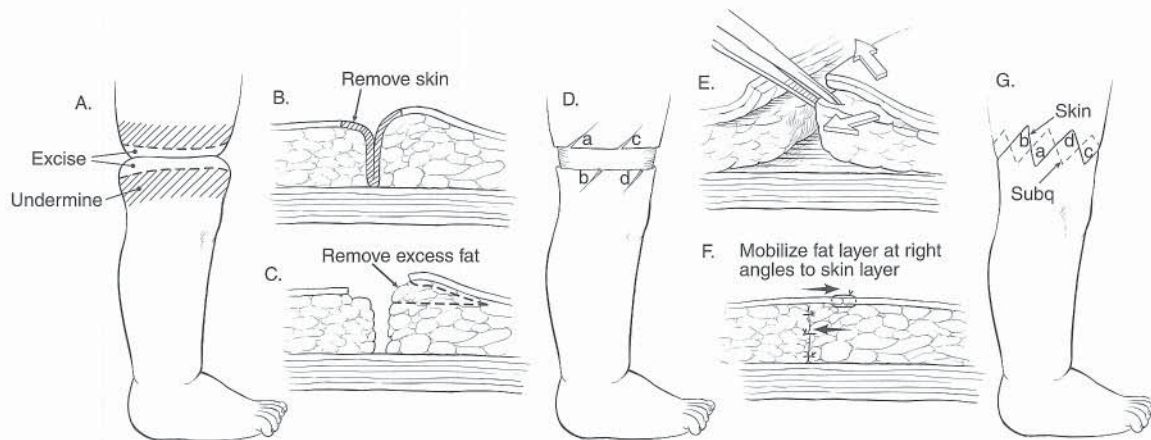


FIGURE 14-121 We like Upton's two-layer closure on the more proximal bands of the limb, but rarely is there enough soft tissue on the digit to make it reasonable. The Z-plasty in the skin is opposite to the one in the deeper tissues.

More often the newborn presents with a dried necrotic part that cannot be saved. This tissue may be surgically removed or allowed to slough. When the digit is severely compromised but has potential for salvage, we try to keep the operation on an infant's small digit as simple as possible. A straight longitudinal incision over the dorsum and through the band will release the constriction and assist in extracting amnion or tissue from the groove. It is best to delay Z-plasty and formal reconstruction until the digit's size and skin healing are more favorable. Digits that are puffy but show no progressive circulatory compromise are also best left alone until the hand is larger.

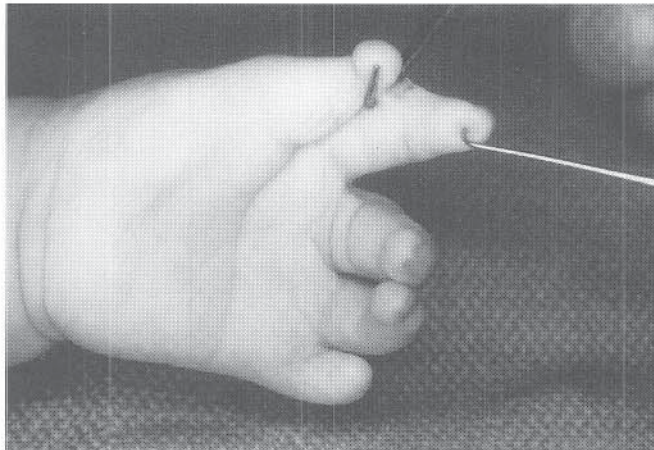
RECONSTRUCTION. When the band is not associated with loss of nerve function, delay of cosmetic reconstruction is important. When small Z-plasties heal with the typical prolific scar of the infant, the result is usually poor. Because the hand doubles in size during the first 2 to 3 years of life, the parents should be encouraged to be patient in their requests for a more normal appearance, and surgery should be postponed.

In more proximal banding of the arm, forearm, leg, or thigh it is also best to wait until the chubby infant fat rolls

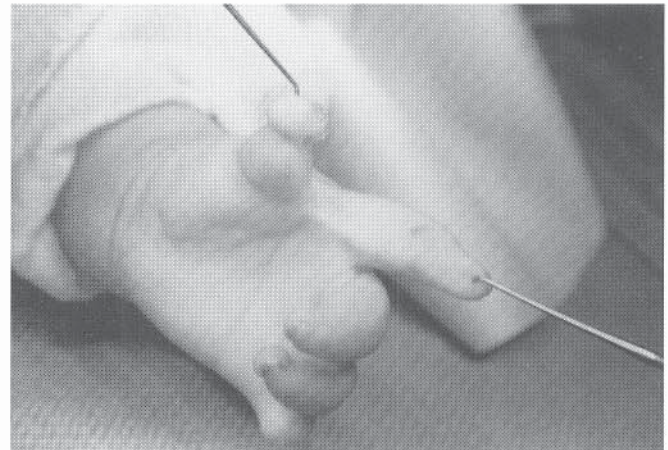
have subsided to assess the real need for the surgery. When the procedure is done, the ring of abnormal skin must be taken out completely. The circumferential line is then converted into generous size Z-plasties for better healing and cosmetic appearance. We agree with the two-layer closure recommended by Upton and Tan in areas where there is enough subcutaneous layer present (Fig. 14-121).⁷ On the digits, this is usually not practical, and here we use a single-layer closure.

Controversy has historically existed over whether to stage the reconstruction into two separate operations involving half the circumference of the band so as to not stress venous return. The surgeon should use judgment, but often the entire circumference of the band can be safely excised at one operative setting.

If a band of the forearm or arm is associated with a severe proximal neurologic defect, it may be appropriate to move up the timing of surgical treatment. The return of neurologic function distal to bands affecting major nerves in the upper limb has not been good, and early exploration and appropriate treatment of the nerve lesion is reasonably considered after 6 months of age.

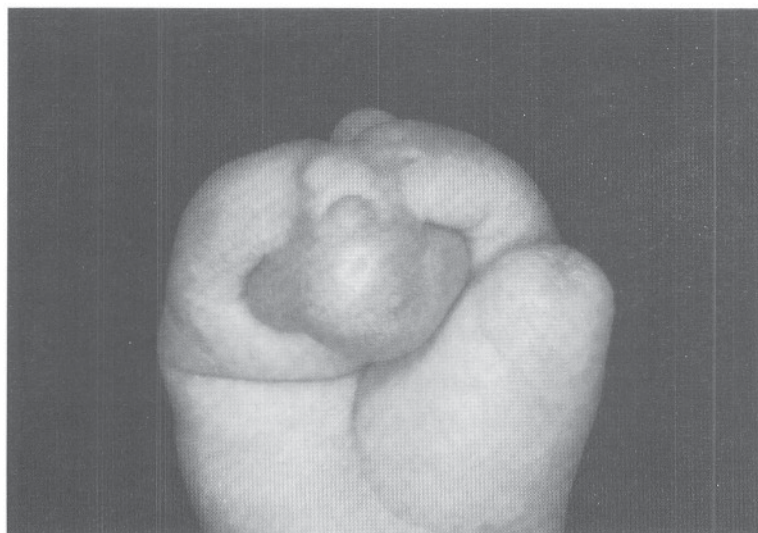


A



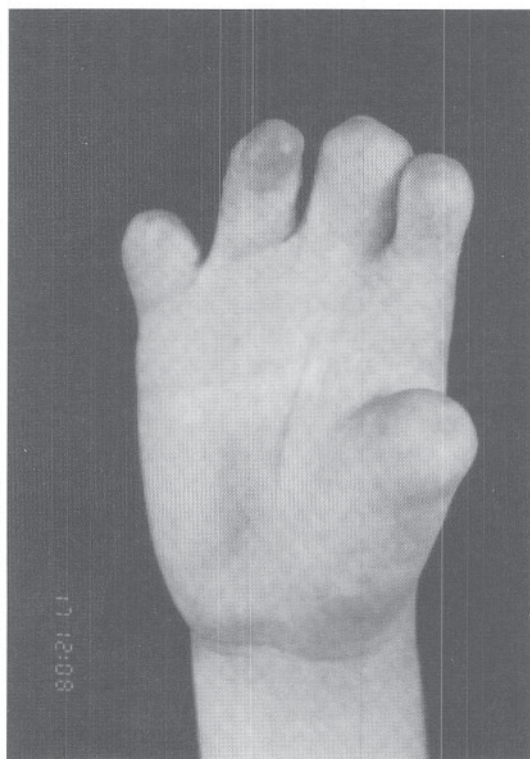
B

FIGURE 14-122 A and B, Simple release of the tip of the index finger and thumb in this 4-month-old child does not require grafting and greatly improves use of the digits. Later (at about 2 to 4 years) a more formal reconstruction of the thumb index web may be required.



A

FIGURE 14-123 A, Sometimes the initial appearance of the hand seems hopeless, but early separation of the fingertips can allow better decisions to be made later about salvage of these parts. B, The surgeon may be surprised by the functional value of parts that were considered worthless. An admonition traditionally attributed to Sterling Bunnell seems particularly appropriate here: “When you don’t have anything at all, a little bit means a lot!”

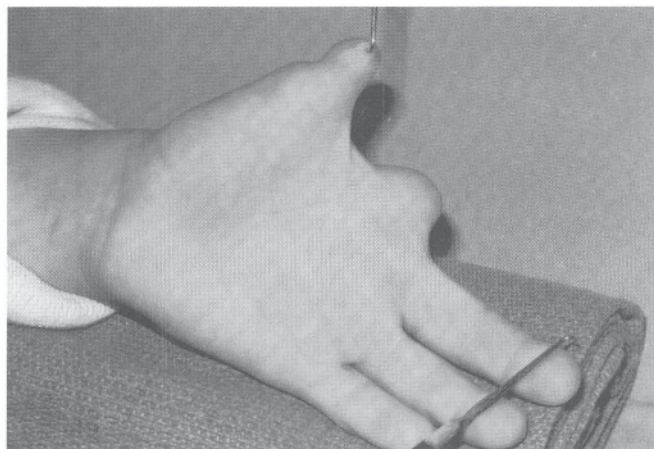


B

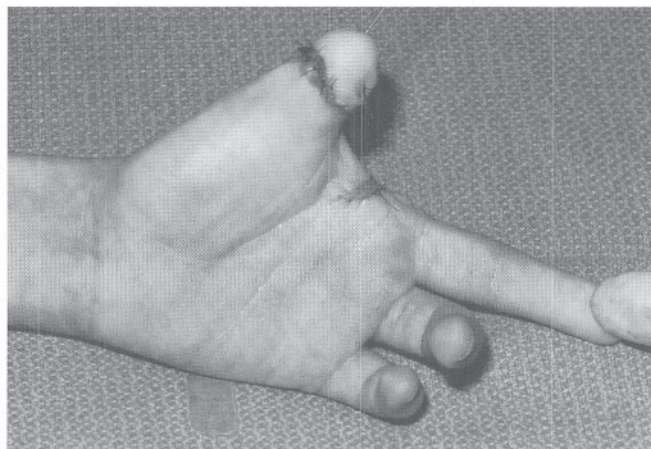
Early but minimal surgery is warranted in the hands of band acrosyndactyly which are fused by a small skin bridge. Simple release with only an appropriate dressing and no skin grafts or flaps is adequate to free the digital growth and allow use (Fig. 14-122). Later, better decisions about salvage of these parts are possible, since the surgeon may be surprised at the value of parts that were considered worthless (Fig. 14-123). On the other hand, a malformed digit is best deleted if it cannot be covered with durable, padded, sensate skin.

Maximizing pinch and apposition by deepening the thumb web is an important consideration, and the Z-plasty or flap reconstructions outlined in the section on syndactyly should be reviewed when this is needed. The island pedicle transfer of tissue is reasonable in some cases of band syndrome when the band amputations are arranged in a fortuitous manner. Length of nerve and vessel pedicles is usually the limiting factor (Fig. 14-124).

Occasionally microsurgical toe transfer may be considered in the appropriate case, usually when a good thumb



A



B

FIGURE 14-124 A, Occasionally a short thumb and a useless index finger can be combined into an “on-top-plasty” done much like a pollicization. B, The proximal resection of the index metacarpal makes for a deeper web space, and the island pedicle tissue transferred to lengthen the thumb is sensate and vascularized.

lacks any member to oppose. Children with amputations due to band syndrome usually have more normal vessels, nerves, and tendons than those with symbrachydactyly or other failures of formation and because of this tend to make better candidates for this more complex surgical solution. A discussion of this treatment is found at the end of the chapter under Microsurgery.

Results. The cosmetic results of more proximal limb banding not associated with neurologic deficit can be quite acceptable (Fig. 14–125). Results in the digits in general depend on the severity of deformity distally. With less severe banding, results are usually good and salvage is remarkably good. Even in more severe cases the surgeon may be surprised that what appears hopeless initially can result, with patience and careful surgery, in a very useful hand (see Fig. 14–123). Occasionally, when a padded, sensate digit with mobility and strength cannot be achieved, amputation of useless, unsightly parts may prove to be the best approach.

Complications. The complications from treatment of banding are usually related to inappropriate timing or execution of the surgical treatment. Any surgery on infants, unless limited to release of distal tethering, has a poor track record. This is because the required Z-plasties are small and often coalesce into a single ring of keloid-like scarring. Simple excision of the ring without Z-plasty is ineffective and unwarranted since it results in a recurrence of the band. The use of many small Z-plasties, even in the larger band or older child, should not be done because more generous flaps that are carefully designed and fitted will deliver a better cosmetic result.

REFERENCES

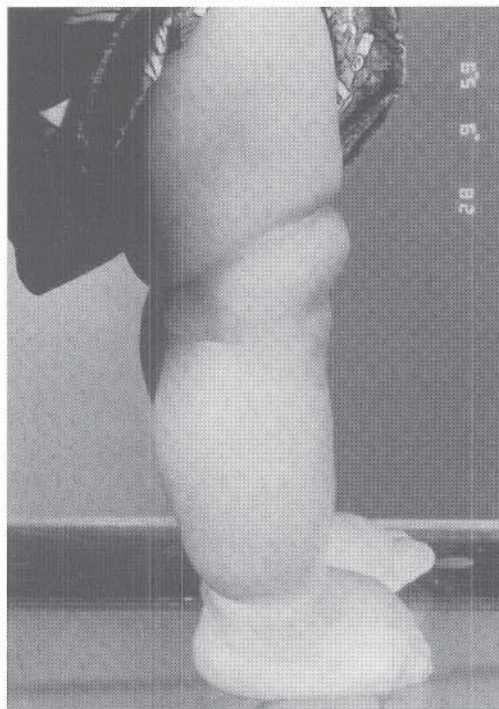
Congenital Band Syndrome

1. Converse JM: Reconstructive plastic surgery. In Littler JW (ed): *The Surgical Management of Congenital Hand Anomalies*, vol 6, p 3306. Philadelphia, WB Saunders Co, 1977.
2. Ezaki M: Amnion disruption sequence. In Green DP (ed): *Green's operative hand surgery*, 4th ed, p 429. New York, Churchill Livingstone, 1999.
3. Flatt A: Constriction ring syndrome. In *The Care of Congenital Hand Anomalies*, 2nd ed, p 277. St. Louis, Quality Medical Publishing, 1994.
4. Patterson TJS: Congenital ring-constrictions. *Br J Plast Surg* 1961;14:1.
5. Streeter GL: Focal deficiencies in fetal tissues and their relation to intrauterine application. *Contrib Embryol* 1930;22:1.
6. Torpin R: Malformations Caused by Amnion Rupture During Gestation. Springfield, Charles C Thomas, 1968.
7. Upton J, Tan C: Correction of constriction rings. *J Hand Surg* 1991; 16-A:947.

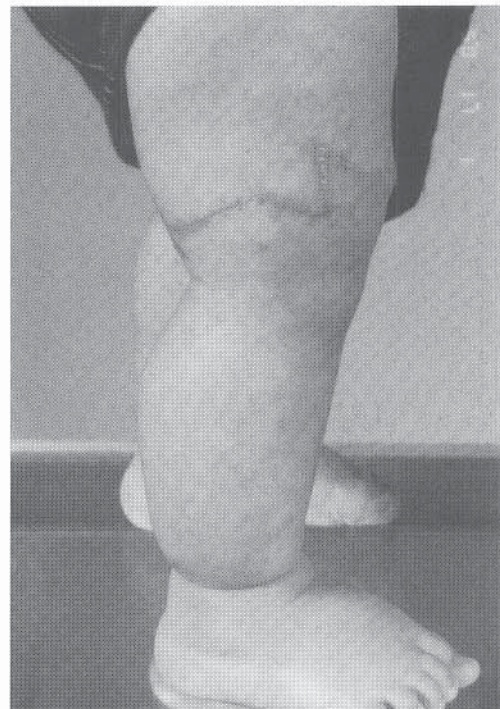
APERT'S SYNDROME

Apert's syndrome is one of a group of relatively rare deformities known as the acrocephalosyndactyly syndromes. In 1906, Eugene Apert described a group of children who had "a very high skull, flattened at the back and sometimes on the sides, while the upper frontal region bulges and syndactyly in all four limbs."¹ The Apert variety is the most common one seen by the pediatric orthopaedic hand surgeon.³

Although the condition is said to be inherited as an autosomal dominant trait, affected children almost always have a spontaneous genetic mutation, since they rarely have offspring. The lack of progeny is likely the social result of their severe facial anomalies, which until modern times could not be reconstructed. The condition occurs in 1 in 100,000 or more live births.³



A



B

FIGURE 14–125 A, Band revision for more proximal clefts gives an improved contour. B, Once the scars mature, they are cosmetically acceptable. The technique described in Figure 14–121 was used in this case.

Upton feels strongly that, at least initially, children with Apert's syndrome are normal mentally and that modern craniofacial reconstruction throughout childhood can yield a normal, functioning brain.³ Historically, many of these unfortunate people were institutionalized and their intellectual potential was never realized. Even today, about half require some special assistance in school.

Clinical Manifestations. There are striking and characteristic facial features caused by (1) premature closure of the basal portions of the coronal and frequently the lambdoidal sutures, (2) shallow orbits, causing a bulging-eyed appearance (exophthalmos), (3) failure of forward growth of the maxilla, and (4) a parrot-beaked nose, high-arched palate, and crowding of the maxillary teeth and tongue. The latter can lead to upper airway difficulties. Other than the musculoskeletal anomalies, there are no other associated abnormalities in these children.

The musculoskeletal anomalies are most apparent in the hands and feet. These deformities include symmetric, complex, and complicated syndactyly. Skeletal dysplasia of the glenohumeral joint and occasionally of the elbow joint can also limit the positioning of these severely deformed hands.

The hand deformity is variably severe, but two general hand patterns exist: the less severe, flat or "spadelike" hand and the more severe, cupped or "spoonlike" hands. Common to both patterns are (1) short, radially deviated thumbs with a delta-shaped proximal phalanx, (2) a complex osseous syndactyly of the index, long, and ring fingers, (3) symphalangism, with little or no interphalangeal motion of the fingers, (4) a simple cutaneous syndactyly in the fourth web space, and (5) the fifth ray as the most normal digit. The MP and DIP are usually functional but lack a functioning PIP joint. A proximal IV-V metacarpal synostosis is frequently present that limits the opposition of this "best digit" toward the thumb.

Treatment. The early treatment of hand deformities is essential for these children. It is most important to coordinate the efforts of the craniofacial team with the pediatric hand surgeon, since the treatment of both head and hand anomalies must begin in infancy. The long-term outcome for these children and their ability to function independently as adults depends on the protection of their brains with appropriate craniofacial surgery and the limited but important hand function that can be restored by carefully planned and timed hand surgery early in life. Psychological evaluations of adults with this condition show self-esteem, sense of mastery, and competence to be very related to hand function.²

The surgical reconstruction of the hands involves complicated decision making that centers on developing the best possible thumb-index web space, mobilizing the "best digit," always located on the ulnar border of these hands, and the optimum number of digits to reconstruct from the remaining II–IV synostotic digital mass. Hand surgery on these children should be done in centers where teams of surgeons can begin the reconstruction as early as possible and carry out bilateral hand procedures, often beginning before age 1. The challenging goal is to have useful hands, with most of the hand reconstruction completed by the time the child enters school at age 5. The details of this treatment are beyond the scope of this book and out of the realistic realm of most general pediatric orthopaedic and hand sur-

geons. These children should be referred to centers where their anesthetic, craniofacial, and hand surgery challenges are managed on a more routine basis. The interested reader is referred to a recent publication by Van Heest, House, and Reckling for details of decision making and an excellent bibliography for further reading.⁴

REFERENCES

Apert's Syndrome

1. Apert E: Apert syndrome. In Buck-Gramcko D (ed): *Congenital Malformations of the Hand and Forearm*, p 159. New York, Churchill Livingstone, 1998.
2. Belfer ML, Harrison AM, Murray JE: Body image and the process of reconstructive surgery. *Am J Dis Child* 1979;133:532.
3. Upton J: Apert's syndrome. In McCarthy J (ed): *Plastic Surgery*, vol 8, p 5293. Philadelphia, WB Saunders Co, 1990.
4. Van Heest AE, House JH, Reckling WC: Two-stage reconstruction of Apert acrosyndactyly. *J Hand Surg* 1997;22-A:315.

Juvenile Arthritis and Other Noninfectious Inflammatory Conditions

This section reviews the care of the hands and wrists of the juvenile arthritis patient. A discussion of important general principles is followed by a discussion of the treatments for specific joints. Finally, standard treatments for the less common inflammatory conditions of childhood, such as systemic lupus erythematosus, dermatomyositis, scleroderma (linear scleroderma), and psoriatic arthritis, are presented.

GENERAL PRINCIPLES

Juvenile arthritis and other inflammatory conditions of children are much more common than is appreciated by most of the lay public and many in the medical profession. The incidence of juvenile arthritis is the same as the incidence of juvenile diabetes, about 1 in 1,000.² The disease, historically called juvenile rheumatoid arthritis or JRA, is different from the adult counterpart in so many ways that it is more properly termed juvenile arthritis.

There are three basic forms of juvenile arthritis. In half of the patients only a few joints (defined as less than four) are involved. This particular form, especially in girls, is associated with serious ophthalmologic consequences. The remainder of juvenile arthritis patients have the polyarticular form (40 percent), in which five or more joints are involved. About 10 percent of these children have the systemic form of the disease and can be very ill. Surgical treatment is reasonable only when accompanied by an appropriate medical regimen. Optimum modern medication programs are complex and changing as new medications become available. The best care of these patients usually is provided by a team consisting of a pediatric rheumatologist, a pediatric orthopaedic surgeon, a hand surgeon, an ophthalmologist, the family pediatrician, a psychologist, family counselors, schoolteachers, and parents. When the entire team functions well and the disease can be controlled, as it usually can, academic achievement and social independence are possible in most cases. The rheumatology of the illness and other

inflammatory conditions are discussed in detail in Chapter 33, Arthritis.

The pediatric hand surgeon has a relatively small but valuable role to play in the treatment of most of these patients. At Texas Scottish Rite Hospital for Children, the hand service has focused on the following:

1. Educating the patient and parents about the basic mechanism of joint damage
2. Providing recommendations for proper splinting and exercise of affected joints
3. Injecting affected small joints with corticosteroid when the disease is unresponsive to medical treatments
4. Performing surgical interventions when indicated

Educating the Patient and Parent. Parents and older children are comforted by a simple discussion of the basic anatomy of the involved joint, the mechanism of injury caused by the disease, and the logic of proposed treatments. This discussion leads to better compliance with all treatments recommended. Despite some parents' copious but shallow knowledge gleaned from the latest Internet arthritis web sites, many patients and their families lack a basic

understanding of what a joint looks like, what articular cartilage is, and what synovium is and does. They rarely know the meaning of the word synovium and do not understand that *synovitis* means inflammation of a normal tissue. They frequently do not know that it is the inflamed synovium that causes the swelling and pain they see in their child. Finally, few realize the connection between synovitis and the destruction of cartilage, bone, and ligaments, or the long-term implications of such destruction. A few minutes spent sketching a normal joint and teaching this information when the patient is first seen is usually a good investment of the surgeon's time (Fig. 14–126). The reward is better compliance with treatment.

Appropriate Therapy and Splinting. A few words about the principles of therapy and splinting are in order. Too often patients and their families have a poor understanding of the proper use of splints and exercise. Some think that forced exercise must be done at all costs or the patient will be doomed to crippling. Sadly, the hidden belief here is that if one makes the child exercise enough, the arthritis will get discouraged and leave. Finally, the word "therapy" has

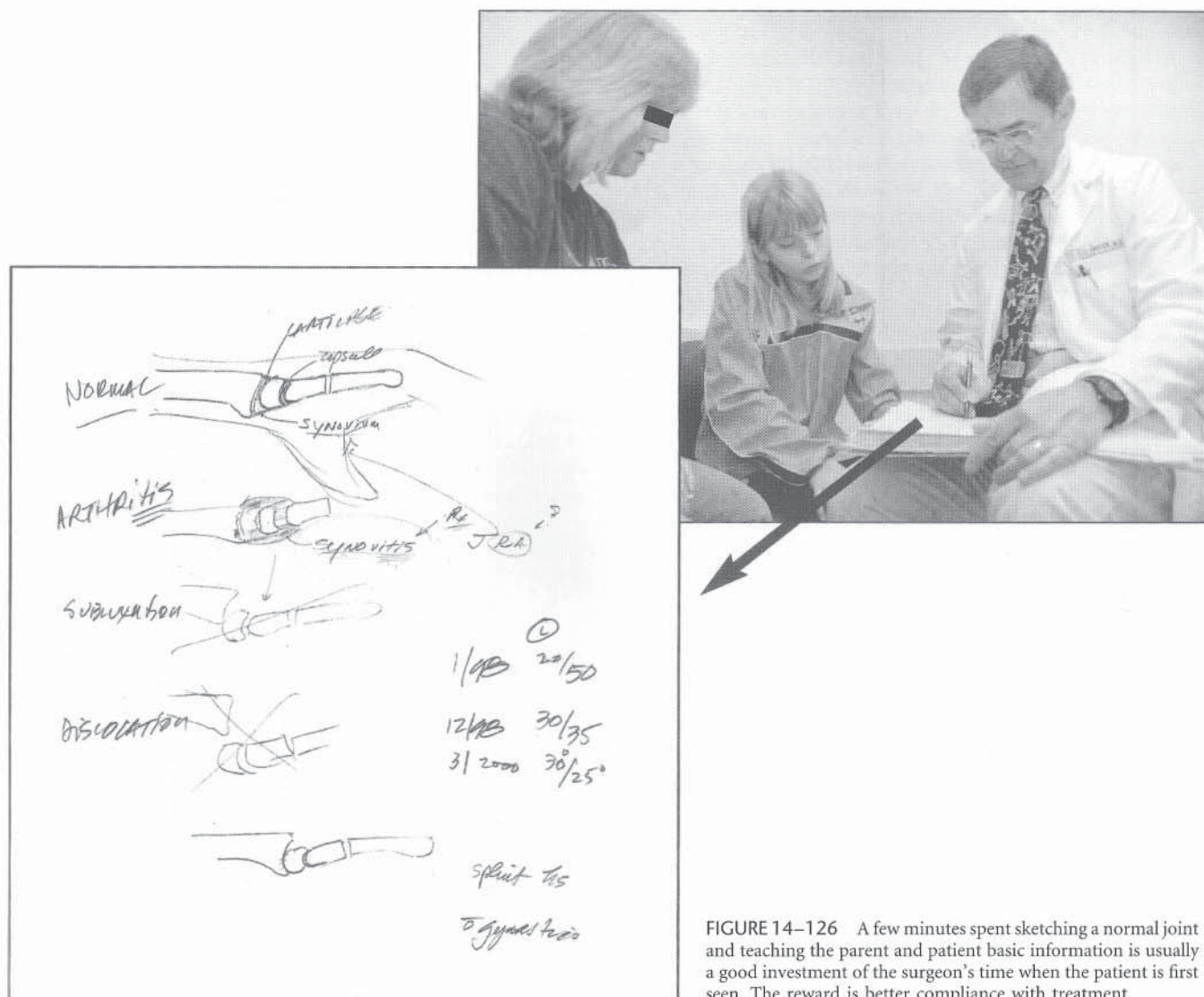


FIGURE 14–126 A few minutes spent sketching a normal joint and teaching the parent and patient basic information is usually a good investment of the surgeon's time when the patient is first seen. The reward is better compliance with treatment.

inappropriate magical connotations that feed the parent's guilt and anxieties. It is critical that these misconceptions be corrected.

Put simply, appropriate splinting can be summed up as follows: At those times when the arthritis is out of control and the joints are hot and painful, rest and splinting are required and exercise is avoided. When the disease is under control and the joints are less swollen and painful, exercise *within the limits of discomfort* is an important and useful part of the care of these diseased joints. The parent and child must know: *Pain is not gain. Pain is a warning sign.*

Intra-articular Steroid Injection. Judicious and skillful use of intra-articular steroid injection into the small joints of the wrist and hand is a critical adjunct to the medical regimen and an important clinical tool for maintaining function in patients with juvenile arthritis. Most rheumatologists and many general orthopaedists are not skilled in injecting the small joints of the fingers or wrist. For this reason, we frequently see patients at the request of the pediatric rheumatologist for this treatment. Tenosynovitis, carpal tunnel syndrome, and joint synovitis all may respond to this treatment when medical therapies have failed. The relief is usually dramatic and rapid, and the patient's function may be restored to near normal. Although the treatment is not curative, the effects can be surprisingly longlasting, the injection rarely has to be repeated. Although injection is not without some risk, the parent usually understands the risk-benefit analysis of injecting a joint that is out of medical control. This is especially true if the proper patient-parent education has been done. Proper technique in injecting steroids into these joints and tendon sheaths is important to minimize complications and maximize results.

THE WRIST

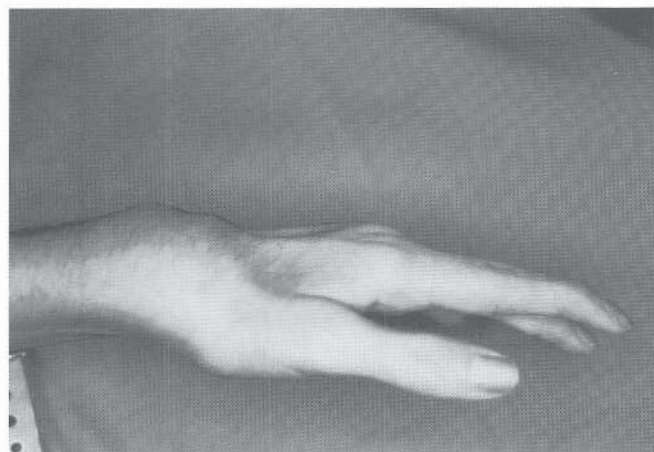
Wrist Joint Synovitis. The normal inclination of the radial carpal joint surface is an inherently unstable one of ulnar deviation and volar flexion. In the normal wrist, this unstable

condition is neutralized by a strong and complex set of anterior wrist ligaments that resist supination of the carpal bones on the distal forearm. In the inflamed wrist of a patient with juvenile arthritis (as in the adult counterpart), uncontrolled joint synovitis stretches these essential ligaments and eventually erodes cartilage and bone. When uncontrolled, this process results in a progressive shift of the carpus ulnarward and volarward. This movement of the hand-carpus unit leaves the ulna dorsally dislocated and also makes for a dorsal wrist step-off because of the subluxed carpus (Fig. 14–127). This is accompanied by loss of active and passive wrist extension. If the disease spreads to the distal radioulnar joint, as it often does, forearm rotation becomes limited.

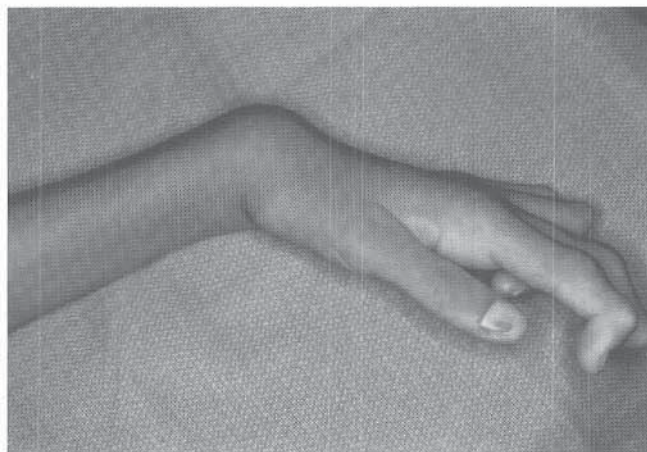
In the early stages, the wrist joint synovitis is clinically manifested by a relatively subtle fusiform swelling that does not move with finger flexion and extension. This is in contradistinction to tenosynovitis of the extensor tendons, in which the swelling is much more obvious and moves to and fro with finger motion.

Tendon Synovitis (Tenosynovitis). The long tendons of the extrinsic flexors and extensors are controlled by the retinacula, fibrous condensations at the wrist and digital levels. The mechanical requirements of this precision system necessitate a close fit of tendons in these sheaths. Because of the great distances these tendons glide, lubrication is required for unhindered motion. This lubrication is provided by the specialized synovium around tendons—the tenosynovium. The tenosynovium is not spared in arthritis, and tenosynovitis leads to unique clinical findings in the hand and wrist of patients with arthritis. When the size of the extensor tendon synovium exceeds the volume of the retinacular tunnel, crepitance, triggering, loss of active motion, visible movement of the mass, and finally rupture of the tendon may result.

The examiner can actually see the mass of tenosynovitis move back and forth over the dorsum of the wrist as the extensor tendons follow the motions of the digits. This



A



B

FIGURE 14–127 In the inflamed wrist of a patient with juvenile arthritis, uncontrolled joint synovitis stretches essential ligaments and eventually erodes cartilage and bone. When uncontrolled, this process results in a progressive shift of the carpus ulnarward and volarward. This movement of the hand-carpus unit leaves the ulna dorsally dislocated and also makes for a dorsal wrist step-off because of the subluxed carpus. This is accompanied by a loss of active and passive wrist extension. **A**, A hand with moderate disease. **B**, The result of prolonged, uncontrolled synovitis.

movement is usually painless, especially early, in distinct contrast to the joint synovitis at the wrist, which is more subtle in appearance but usually quite painful, especially in its early stages.

Anteriorly, the flexor tendons of the digits also pass through a wrist retinaculum that, unlike all others, contains a nerve. Here in the carpal tunnel, the swelling of tenosynovitis may lead to the pain and numbness of median nerve compression—carpal tunnel syndrome. Unlike on the dorsum of the wrist, the swelling is harder to see clinically owing to the deep position of the tendons in the bony arch of the carpus and the thick, restraining fascia of the forearm.

Triggering may be seen as a result of tenosynovitis at the wrist, but it is rare compared to the frequency of triggering caused by tenosynovitis in the miniaturized retinacula of the digits—the tendon sheaths.

Treatment of Synovitis and Tenosynovitis. The goals of treatment are to preserve alignment and motion of joints, maintain the free, smooth gliding of tendons within their retinacula, prevent tendon rupture, and relieve carpal tunnel syndrome. The nonoperative treatments include splints, heat, injections, and exercise.

SPLINTS. Wrist extension splints that support ulnar carpal “sag” help support the carpus on the forearm. Although limited in their ability to supply adequate force through the soft tissues, they are an important treatment, especially in the early stages, before significant deformity has occurred. The wrist splint should be simple and comfortable and should not impede digital motion but hold the wrist in neutral position or a few degrees of extension (see Fig. 14–35). Because all splints interfere to some degree with the patient’s ability to make the best use of the hand, we initially prescribe splints for nighttime use only.

HEAT. Moist heat and paraffin baths can be effective in controlling pain and helping to ease stiffness. When family compliance is adequate, this treatment is conveniently and effectively done at home.

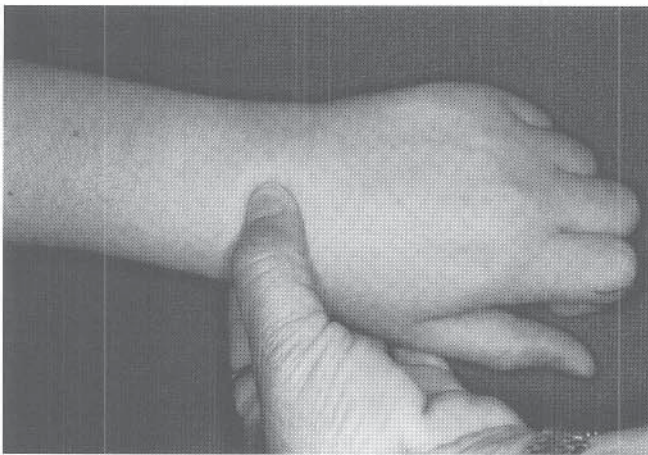
INTRA-ARTICULAR STEROIDS. When joint synovitis is active and not controlled with systemic medications, direct injection of the radiocarpal, midcarpal, and distal radioulnar joints is a very useful clinical tool (Fig. 14–128). Our usual injection is equal parts of triamcinolone, 40 mg/mL (Aristocort Forte), a triamcinolone timed-release preparation (Aristo-span), and Xylocaine. Although injections for wrist extensor tenosynovitis are less often done because there is some risk of inducing tendon rupture, the risk of rupture from the tenosynovitis is a real one. The risk-benefit analysis of injection is beneficial to the patient, and if the operator is careful to avoid injecting the tendon forcefully, the risk can be minimized.

A carpal tunnel injection must avoid injecting the median nerve, but otherwise it is a safe and effective tool for relief of pain and numbness (Fig. 14–129). In addition, relief of symptoms is a useful clinical confirmation of the diagnosis of carpal tunnel syndrome. Not infrequently a single injection is particularly helpful, especially until other medical control becomes adequate.

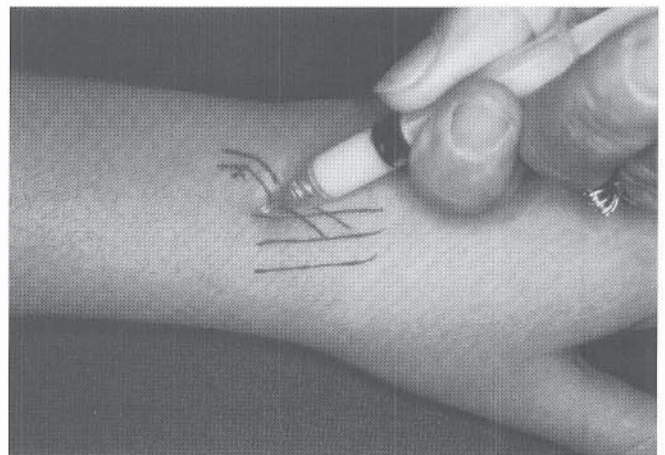
EXERCISES. Gentle active and active assisted motion within the limits of pain is appropriate and should be encouraged whenever the general activity of the disease permits. Exercise should not be painful and should not be done with hot swollen joints, a sign of active disease. Passive motion carried out by a “Big Swede” physical therapist with the attitude of “no pain, no gain” has no place in the management of the small joints of the wrist and hand. Pain is not gain but a warning sign of a need for rest.

OPERATIVE TREATMENT

General Principles. When medical treatment and the nonoperative surgical measures described above prove inadequate, we have found surgical treatment to be very helpful for certain children. However, as a general rule, the surgeon must be careful to treat the patient, not the radiograph. Many of these children may function very well in spite of severely abnormal wrist films. The simple radiographic presence of



A



B

FIGURE 14–128 When joint synovitis is active and not controlled with systemic medications, direct injection of the radiocarpal, midcarpal, and distal radioulnar joints is a very useful clinical tool. **A**, The location of the open area free of tendons. **B**, Injection in the open area into the radiocarpal joint and between the II compartment and the III compartment.

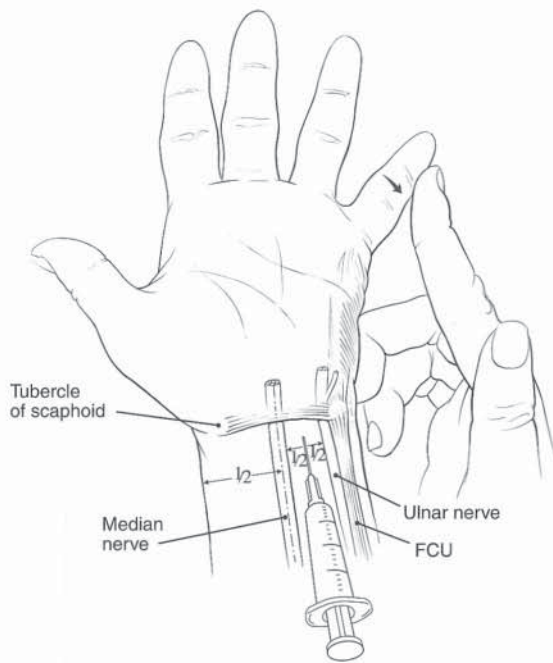


FIGURE 14–129 The FCU tendon is easily palpated when the patient abducts the little finger.

carpal coalitions, gross joint destruction, and even moderate joint subluxation may be associated with a relatively painless, functional arc of wrist flexion and extension. This less than normal wrist is often sufficient to cope with the arthritic child's reduced muscle strength, stiff, deformed fingers, and markedly restricted functional demands. The risk-benefit evaluation of operative intervention can be difficult in these children. The surgeon should be cautious about surgical intervention. The risk of general anesthesia in these children is especially serious when the mandible, temporomandibular joint, and cervical spine are abnormal. The airway can be very difficult to maintain and requires a prepared and skillful anesthesiologist.

As a general rule, we favor fusions and resection arthroplasties around the wrist joint and essentially never advocate implant arthroplasty in children. The goals are pain relief, joint alignment, and preservation of muscle tendon units by ensuring that tendons are not eroded over rough bony edges. The principles of decision making in these severely impaired patients are among the best examples of the outline presented earlier under Principles of Reconstruction.

Synovectomy. Synovectomy of multiple joints of the wrist is rarely useful in children with juvenile arthritis. In most cases, joint synovectomy has been replaced by steroid injections as the primary management tool. Later, more permanent correction may become appropriate by virtue of the patient's age or degree of joint destruction. However, in the unusual monoarticular case in which systemic medication and joint injection have failed to control synovitis and the joint architecture, including the joint space, is still essentially normal, synovectomy may be useful.

Wrist Fusion. Once the child has serious malalignment of the hand on the forearm, wrist fusion is a predictable method to restore alignment, reduce pain, and improve the appear-

ance and use of the affected wrist and hand. After wrist arthrodesis, these teenagers are among the most grateful in the hand clinic. The improvements in function and independence after wrist fusion may be very dramatic. It is ideal if fusion can be delayed until growth has stopped, but in severe cases these small children rarely grow to normal height and may benefit more from restoration of hand function than from an extra centimeter or two of growth at the distal radius. Our experience with so-called "chondrodesis" or attempted fusion in young children while sparing the radial epiphysis unfortunately has not been predictable.

Technique of wrist fusion: When significant deformity is present, we tend to use small plate fixation to ensure maintenance of alignment. Usually a semitubular 3.5-mm plate is aligned on the third metacarpal first and then fixed to the radius. Some of these very small children have such slender metacarpals as to defy screw fixation. In these cases a T-plate anchored into the bases of the metacarpals has been adequate (Fig. 14–130). Although the bone fuses very quickly, we supplement the internal fixation for a few weeks with a short-arm cast and later have the patient use a wrist splint when in public for another 6 weeks. Bone graft from a distant site usually is not required unless there is marked bone erosion. We often use the head of the ulna or the proximal carpal row as source of bone.

Many of these cases have severe, fixed flexion deformities, and the surgeon must be careful to resect enough carpal bone to allow neutral or straight alignment without putting excessive stretch on the anterior structures, especially the median nerve. Since bilateral involvement is common, we have sequentially fused both wrists in some cases. The historical orthopaedic concern that bilateral wrist fusions will make perineal care impossible has not been borne out in our patients and does not justify the use of implant arthroplasty. We position the fused wrist in neutral flexion-extension. In patients with poor finger extension, fusing the wrist in any amount of extension compromises the patient's ability to extend the fingers.

Darrach Resection. Resection arthroplasty of the distal radioulnar joint is very effective in adolescents with wrist destruction severe enough to need wrist arthrodesis. We often combine it with wrist fusion. Darrach's procedure is especially useful in these teenagers when they have lost forearm rotation. Improved independence postoperatively may be dramatic. For example, the Darrach operation has made it possible for patients to finally accomplish the simple task of opening the door to their own room. On the other hand, the surgeon should remember that resection of the ulna when the joint is unstable instead of stiff and when good range of motion is still present can destabilize the wrist. The symptoms of weakness as well as the painful snapping of the ulna can worsen, and deformity may increase in this setting.

Technique of Darrach Resection: The surgeon should take care to resect minimal ulna (just enough to allow rotation without crepitation). The dissection of bone is carried out subperiosteally, and careful repair of all remaining ligaments around the joint is important. Relocation of the extensor carpi ulnaris after the method of Clayton, using a flap of retinaculum to fashion a supporting loop, helps support the ulnar carpus that has sagged into supination on the forearm (Fig. 14–131). The frail, diseased tissue that must be used for reconstruction requires the protection of a short-

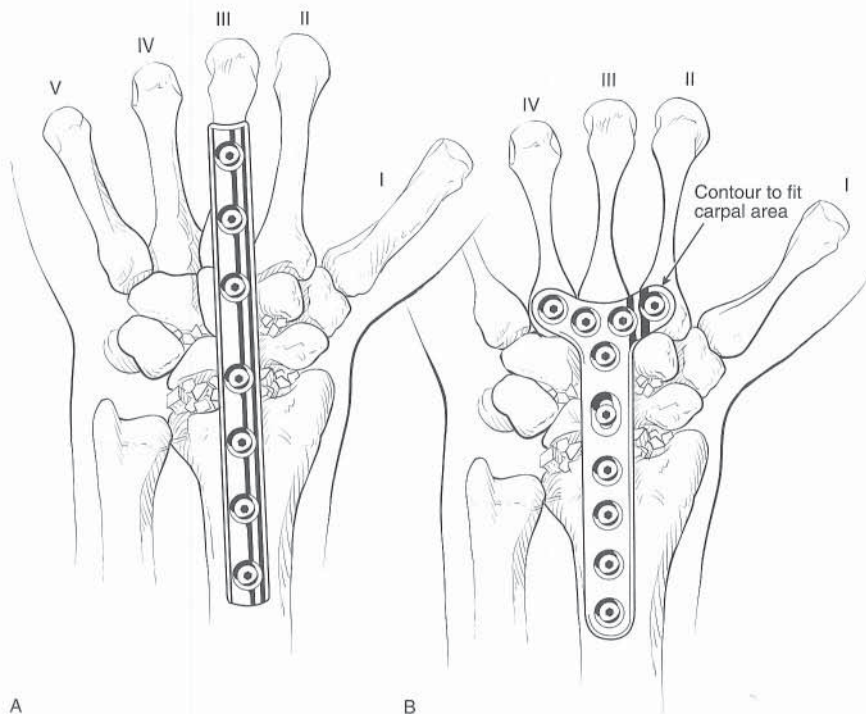


FIGURE 14-130 Technique of wrist fusion. When significant deformity is present, we tend to use small plate fixation to ensure maintenance of alignment. Usually a semitubular 3.5-mm plate is aligned on the third metacarpal first and then fixed to the radius. **A**, Some very small children may have such a slender metacarpal as to defy screw fixation. **B**, In these cases a T-plate anchored into the bases of the metacarpals has been adequate.

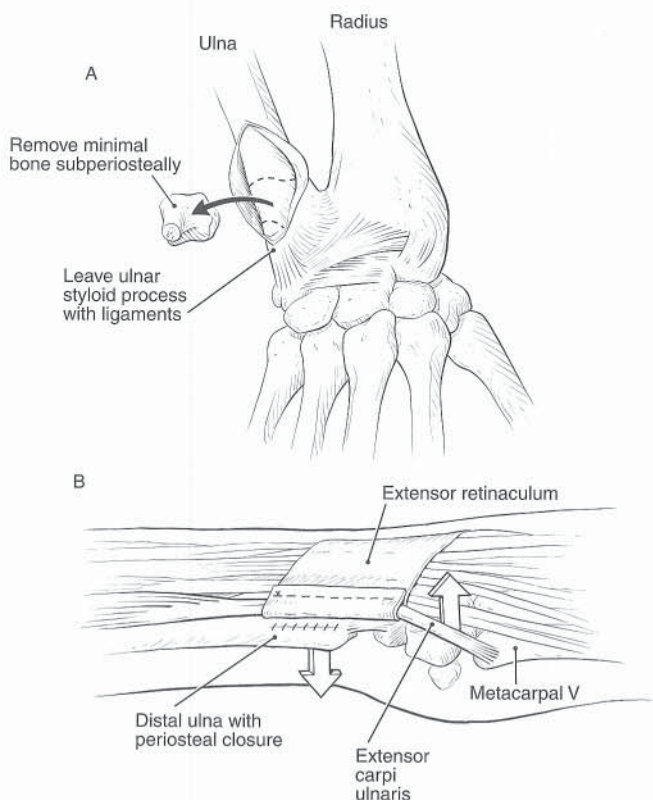


FIGURE 14-131 **A** and **B**, Technique of Darrach resection of the distal ulna. The surgeon should resect minimal ulna, try to do the dissection subperiosteally, and repair soft tissue structures around the joint. Relocation of the extensor carpi ulnaris after the method of Clayton, using a flap of retinaculum to fashion a supporting loop, helps support the ulnar carpus that has sagged into supination on the forearm.

arm cast postoperatively. This supports the carpus, limits rotation during the early healing period, reduces postoperative discomfort, and reduces the risk of late instability.

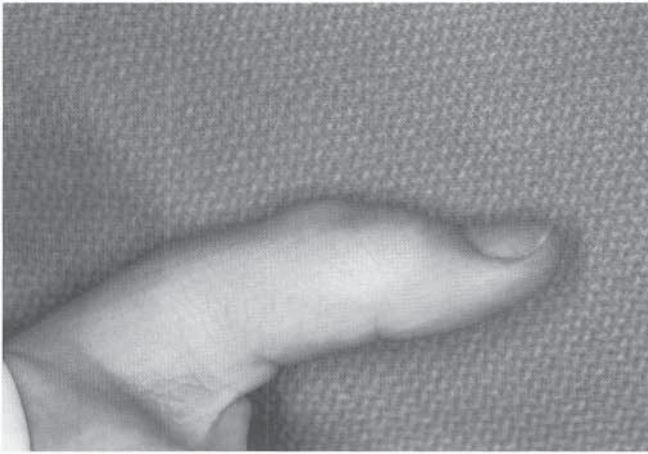
DIGITAL INVOLVEMENT

Synovitis of the Digits. Synovitis of the small joints of the digits leads to specific deformity (Fig. 14-132). The finger and thumb deformities are the result of the uncontrolled effects of the synovial inflammation on the ligaments and surfaces of the joint, the tendons moving these joints, and the sheaths or retinacula in which the tendons glide.

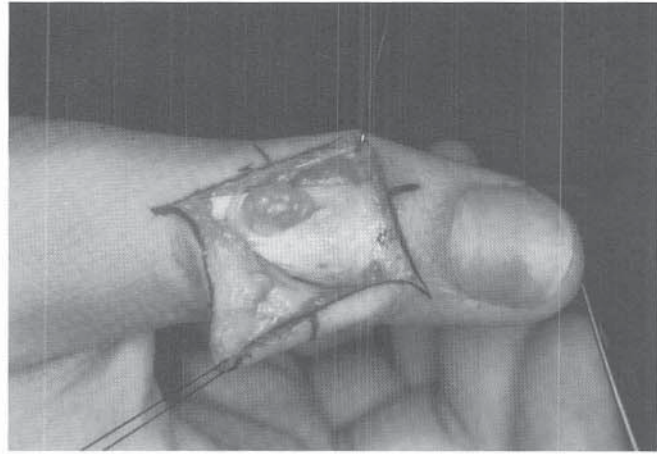
Finger Synovitis and Deformity. The typical deformity in the hands of children with juvenile arthritis is yet another striking contrast to the adult with rheumatoid arthritis. The adult tends to develop loose, unstable joints with loss of MCP flexion, ulnar drift, and swan neck deformities; the child has just the reverse of the typical adult deformity. In juvenile arthritis the joints are stiff, not loose. The MCP joints are stiff in extension and radial deviation. The fingers of a patient with juvenile arthritis may exhibit swan neck deformity, but more common is a flexion deformity of the PIP joint (boutonniere) (Fig. 14-133). Why the deformities are so different in adults and children is unknown and is yet another reason to call the arthritis of children juvenile arthritis, not juvenile rheumatoid arthritis.

Thumb Synovitis and Deformity. As in the adult, deformities of the thumb in children are varied and include adduction contracture as well as instability, deformity, and pathologic dislocation at all joint levels (IP, MCP, and CMC).

Tenosynovitis in the Digits. Triggering is rarely seen as a result of tenosynovitis at the wrist, but it is quite commonly associated with tenosynovitis in the miniaturized retinacula



A



B

FIGURE 14-132 Synovitis of the small joints of the digits causes a fusiform swelling. **A**, The inflamed synovium has penetrated the extensor tendon. **B**, The finger and thumb deformities are the result of uncontrolled synovial inflammation of the joint ligaments, tendons, and joint surfaces.

of the digits—the tendon sheaths. Initially the early presence of tenosynovitis in this area is best appreciated by having the patient open and close the fingers around the finger of the examiner. A crepitance will be obvious to the examiner and patient. In the next stage, the patient begins to lose active flexion of the digit. Early, passive flexion exceeds active flexion—the pathognomonic sign of flexor tenosynovitis in the digital flexors. In the intermediate stage, the patient may report or demonstrate the jerky movement of the finger called triggering. Occasionally the finger may be locked in flexion or extension by a nodule of tenosynovitis that cannot traverse the strong annular ligaments of the digital sheath. Tendon rupture, although less common in children than in adults, may finally occur. In this final and catastrophic stage,

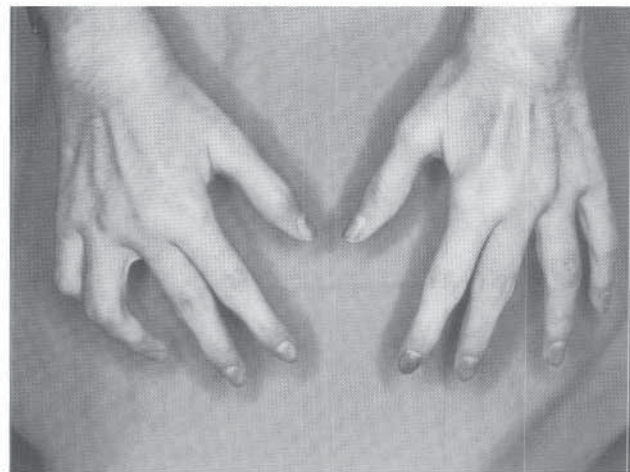
active control of the finger is lost and the digit becomes flail and useless.

TREATMENT OF HAND INVOLVEMENT

Splinting. Splinting of the fingers is complicated by the fact that finger splints interfere significantly with use of the hand. In addition, there is no evidence that splinting prevents finger deformities in the long term. For these reasons we use splinting in moderation. Dynamic splints to wedge the PIP joints of the fingers into extension are cumbersome, and we prescribe them for nighttime use only. In general, small, form-fitting static splints are better tolerated by patients (see Fig. 14-29). These splints are useful for resting



A



B

FIGURE 14-133 The typical deformity in the fingers of children with juvenile arthritis is yet another striking contrast to rheumatoid arthritis in adults. **A**, The adult tends to develop loose, unstable joints with loss of MP flexion, ulnar drift, and swan neck deformities. **B**, The child has just the reverse of the adult typical deformity. In juvenile arthritis, the joints are stiff, not loose. The MP joint is stiff in extension and radial deviation. The fingers of a patient with juvenile arthritis may demonstrate swan neck deformity, but more common is a flexion deformity of the PIP joint (boutonniere).

an acutely inflamed, tender, swollen joint and can be easily changed to gently wedge the joint out toward extension. Our goal is not a normal finger but an extensor lag in the range of 30 to 40 degrees. This can usually be accomplished in the compliant patient. The nuisance of finger splints makes compliance difficult for the patient, and an attentive, sympathetic therapist is very important.

Injections. Digital joint injections with steroids are a mainstay of our treatment program to supplement medical management. Injections are indicated for the painful, swollen joint that does not respond to systemic medication. The injection is facilitated by use of a small (26-gauge, 3/8-inch) needle (Fig. 14–134). It is essential to appreciate the exact topographical anatomy of the finger joint. There is a tendency, especially when the joint is extended, to expect the joint line to be more proximal than it is. Gentle traction on the finger will open the joint and make finding the joint with the needle easier (Fig. 14–135).

Synovectomy. Synovectomy is even less useful in digital joints than in the wrist joint and has been replaced by corticosteroid injection, especially when multiple joints are involved, as is usually the case in the hand. Synovectomy is indicated only in the rare patient with an isolated joint or only a few joints that remain inflamed after several injections. Synovectomy is not indicated when radiographs show joint destruction.

Joint Reconstruction

PIP JOINTS. Fortunately, the boutonniere deformity of the fingers, although unsightly, is usually very functional until it exceeds 60 degrees. With good splinting and careful medical management of the disease, this only occasionally occurs. Reconstruction of the extensor mechanism in early boutonniere deformities has little predictable long-term reward for the patient, especially if the disease is uncontrollable. Cosmetic reconstruction of mild boutonniere deformity is therefore of little benefit to the patient. However, as the extensor

lag worsens to around 90 degrees, the loss of opening of the hand becomes a severe functional impairment and interferes with dressing, bathing, and other activities of daily living (Fig. 14–136). In these cases, a PIP arthrodesis in slight flexion is functionally justified and improves the appearance as well. In these patients, arthrodesis in 20 to 40 degrees of flexion (less for the radial digits and more for the ulnar digits) can allow the patient to use the finger much more efficiently. We prefer a butt joint fusion held in place by K-wires for 6 to 8 weeks. The poor skin coverage makes the bulk of buried wires impractical in these small hands. We usually leave the wire out through the skin for easy and complete removal later. The small additional aggravation of the need to protect the wire with a cast and later a splint has proved rewarding for patient and surgeon alike. Although we prefer to wait until the epiphyses have closed, a few millimeters of growth in the middle of the digit must be weighed against improved function. The digit is also made relatively longer by fusion. Care must be taken at operation not to extend or stretch the anterior neurovascular structures, causing digital ischemia.

MCP JOINTS. MCP joint deformities in the fingers of patients with juvenile arthritis usually do not require surgical treatment unless they are very severe. We avoid implant arthroplasty of the fingers in children unless there are severely mitigating circumstances, such as markedly reduced function of the shoulder, elbow, and wrist. These patients have less loading on the breakable implant. MCP arthrodesis is usually the treatment of choice if there is adequate PIP motion, and especially in the index finger, where pinch forces are great and opening to the palm is critical. As in the PIP joints, butt joint fusion technique stabilized with percutaneous fixation is used. The position of fusion is tailored to optimal function, considering which finger is involved as well as the existing movement of the PIP joint distally. Volar plate interposition arthroplasty in selected cases is appropriate.

Thumb Deformities. One of the most functionally limiting deformities for these children is a fixed flexion deformity of the MCP joint of the thumb with a tight adduction contracture. These children then must use two hands to hold a drinking glass, and any function requiring thumb-index web opening is greatly compromised. MCP arthrodesis is very effective in providing stability and opening of the web, especially when combined with adduction contracture release. The goal is to restore a useful thumb-index web space, even at the expense of weakening some of the adductor and thenar muscles. This is particularly effective when the IP and CMC joints have been spared and have an adequate range of motion.

IP arthrodesis of the thumb can be considered when there is a marked deformity, usually radial or ulnar deviation. As with MCP fusion of the thumb, IP fusion is most effective if the other joints of the thumb are spared. In the child with a severely involved thumb (two or more joints involved), joint fusions do compromise function. However, even though functional restoration is much more limited than in the thumb with single-joint disease, fusion may give these severely involved children significant improvement in the daily activities of living.

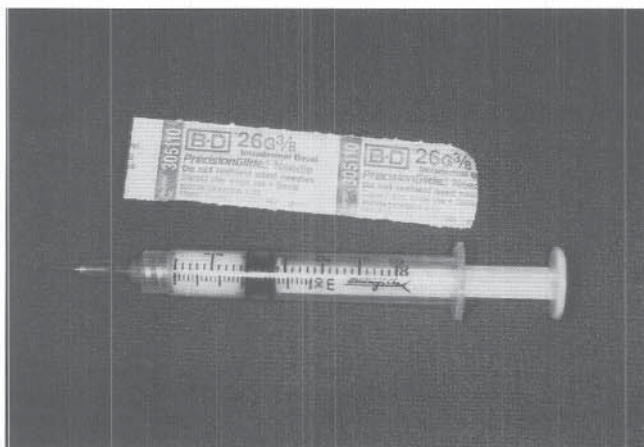
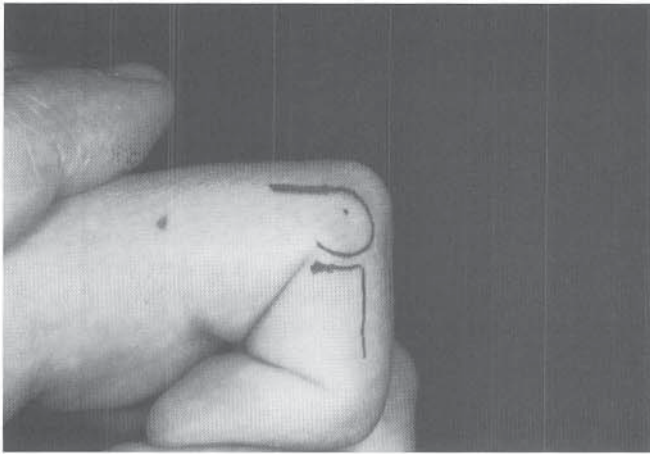
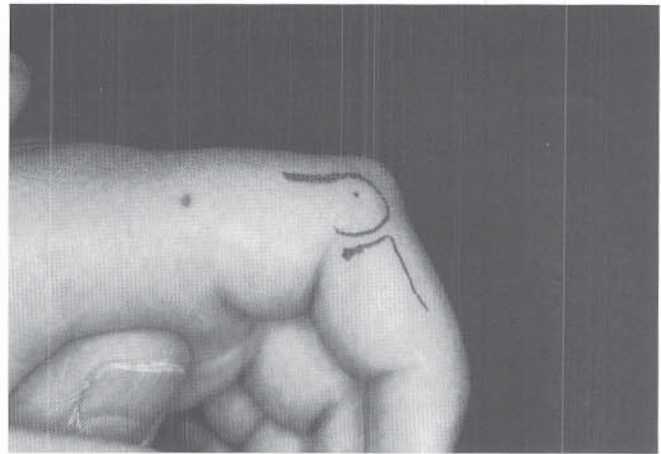


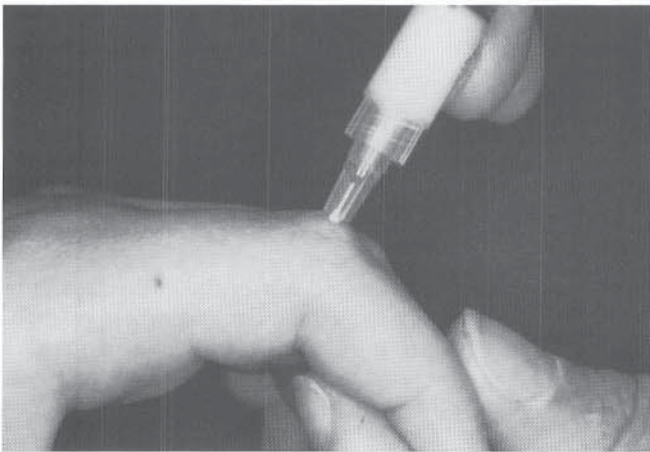
FIGURE 14–134 Joint injections with triamcinolone are a mainstay of our treatment program to supplement the systemic medication of the pediatric rheumatologist. These are quite effective for the painful swollen joint that is resistant to systemic medication. The injection is technically relatively simple if a small (26-gauge, 3/8-inch) needle is used and the exact topographical anatomy is appreciated.



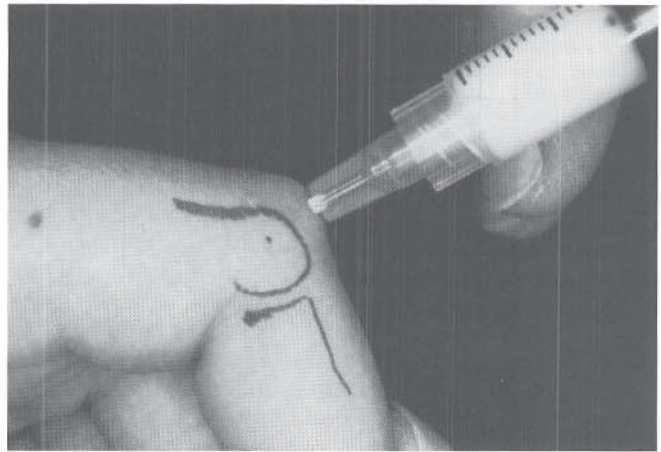
A



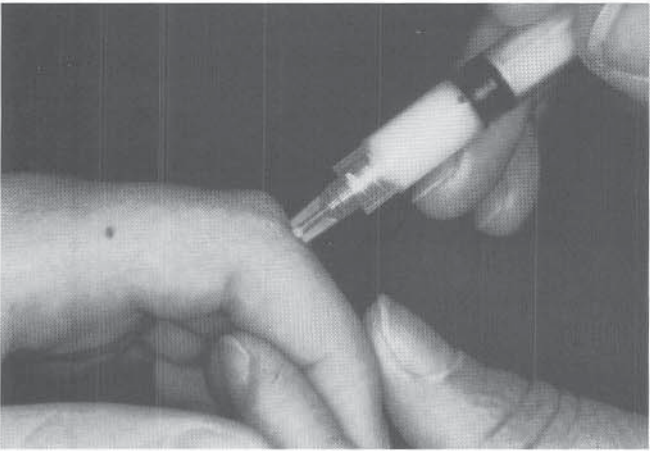
B



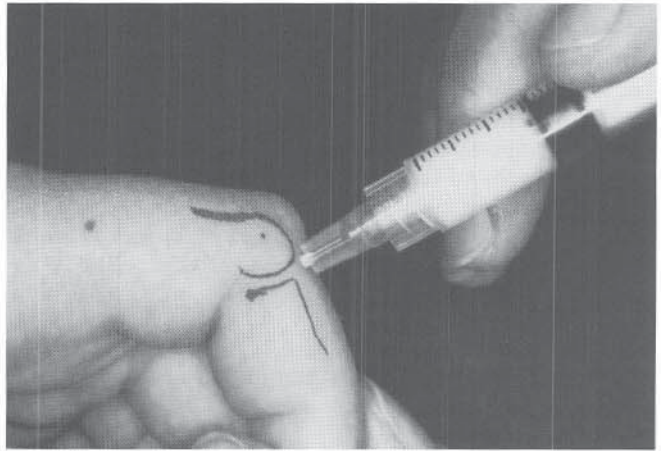
C



D



E



F

FIGURE 14-135 There is a tendency, especially when the joint is extended, to expect the joint line to be more proximal than it is. Gentle traction on the finger will open the joint and make finding the joint with the needle easier. **A and B**, The position of the joint between the proximal and middle phalanx. **C and D**, Incorrect insertion of the needle at the apex of the bend. In this position the needle impales the condyle of the proximal phalanx and fails to enter the joint space. **E and F**, Correct insertion of the needle inferior and distal to the apex of the bend of the finger.

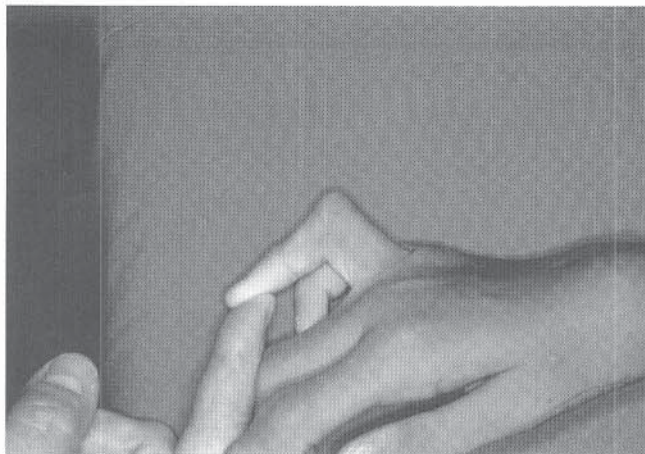


FIGURE 14-136 A severe fixed boutonniere deformity of the PIP joint. A boutonniere deformity of the fingers, although unsightly, is usually very functional until it exceeds about 60 degrees. As the extensor lag worsens toward 90 degrees, the loss of opening of the hand becomes a severe functional impairment and interferes with dressing, bathing, and other activities of daily living. In these cases, a PIP arthrodesis in slight flexion is functionally justified and improves the appearance as well. In these patients arthrodesis in 20 to 40 degrees of flexion (less for the radial digits and more for the ulnar digits) can allow the patient to use the finger much more efficiently.

NONINFECTIOUS INFLAMMATORY CONDITIONS

In patients with dermatomyositis, scleroderma (linear scleroderma), and psoriatic arthritis, the hands are very stiff and skin coverage is often poor. Joint fusions using the principles noted in the patient with juvenile arthritis are the mainstay of joint treatment.

Some of these patients have severe ischemia of their digits and often may lose tissue due to small vessel disease. Amputations can sometimes be forestalled with sympathectomy, both surgical and with nerve block. A series of intermetacarpal digital nerve or wrist blocks can be tried using marcaine 0.025%. Sometimes a block or two will break the vasospasm, relieve the patient's pain, and restore the color and temperature of the digit. If the regional anesthetic appears helpful but is only transient in effect, the technique of digital sympathectomy as described by Flatt has been useful.¹

When amputation finally becomes necessary, the general healing capacity of these patients is severely compromised, and deciding on a level that preserves function and still will heal is a problem. Many times in these tragic cases, dressing changes and supportive treatments are all that can be done.

REFERENCES

Juvenile Arthritis and Other Noninfectious Inflammatory Conditions

1. Flatt AE: Digital artery sympathectomy. *J Hand Surg* 1980;5-A:550.
2. Lovell DJ: Pediatric rheumatic diseases. In Klippel JH (ed): *Primer on the Rheumatic Diseases*, 11th ed, p 393. Atlanta, Arthritis Foundation, 1997.

Infections

Hand infections in children are not as common as in adults because behaviors that lead to adult infections and concomi-

tant medical problems are not as common in children. Work-related injuries, diabetes mellitus, and drug abuse are predisposing factors in the older population but not in the younger child.

All forms of infection can involve a child's hand. Children taking immunosuppressive medications may be susceptible to the same opportunistic infections as their older counterparts. Certain disease processes can mimic infection in the child and should be considered in the unusual presentation or when microorganisms cannot be cultured.

The arthritides, particularly psoriatic arthritis, can present with a clinical picture of septic joint or tendon sheath.

The most common hand infections in the otherwise normal child are fingertip infections, paronychia and felons, herpetic whitlow, and infection following a break in the skin. Treatment principles include identification of the infecting organism if possible; elevation, warm soaks, and antibiotics in the early phase; incision, drainage, and placement of a wick to facilitate drainage in the phase in which purulence has accumulated; and *never* close an infected wound.

PARONYCHIA

Paronychia is an acute or chronic infection of the periungual tissue. It often results from a pulled "hangnail" and presents as swelling at the lateral border of the nail plate, quickly extending as a "runaround" infection under the cuticle or paronychia tissue (Fig. 14-137). Persistence of the suppurative process undermines the nail plate. The bacterial flora is almost always *Staphylococcus aureus* or oral anaerobes.

Treatment of acute paronychia in the early stage is by warm soaks, elevation, and antibiotics. Treatment of established paronychia with purulence present is elevation of the nail fold, or incision and removal of the nail plate if it has loosened. Antibiotics should cover *Staphylococcus* and oral



FIGURE 14-137 Paronychia. Note the periungual swelling and purulence. Treatment of early paronychia includes warm soaks, oral antibiotics, and drainage by incision of the nail fold.



FIGURE 14-138 Felon. Note the palmar swelling, erythema, and acute tenderness. Treatment requires drainage and decompression of the vertical septa of the digital pulp.

anaerobes. Cephalexin, dicloxacillin, nafcillin, and clindamycin are appropriate choices.

Chronic paronychia is recognized by chronic induration and swelling of the nail fold. Mixed organisms, including fungi, should be suspected. Chronic paronychia is not as common in children as in adults. Treatment includes antifungal medication, nail plate removal, and marsupialization of the eponychial fold.

FELON

A felon is a pulp space infection in the distal segment of the finger. It is a deep infection within a tight septal compartment. Felons usually follow penetrating trauma and are recognized clinically by intense pain, erythema, and palmar swelling of the fingertip (Fig. 14-138). If the lesion is relatively superficial, the purulence may “point” palmarly, and is then known as an apical abscess. The usual organism is *S. aureus*, and treatment includes antibiotics appropriate for *Staphylococcus* species as well as surgical drainage of purulence.

The surgical approach to a felon is designed to preserve the tactile surface of the fingertip and to avoid damage to the digital neurovascular bundles. The approach depends on whether or not it is already pointing. If the lesion is pointing, an incision is placed over the apex of the lesion, through skin only, and then the septa are divided and a wick is placed into the depths of the abscess cavity. If the felon is not pointing, the appropriate incision is a J-shaped incision extending from the tip of the finger just under the nail plate and then laterally along the midaxial line to the proximal extent of the nail. The vertical septa in the pulp space are divided and the incision is held open with a wick of gauze that can be removed after 48 hours. The wound

is left open to heal by secondary intention. Complications of a felon include loss of pulp tissue, osteomyelitis, epiphyseal damage, and septic arthritis.

HERPETIC WHITLOW

Whitlow is the name given to a superficial skin infection caused by the herpes simplex virus. HSV-1 or oral herpesvirus is almost invariably responsible for pediatric whitlows. The clinical presentation is one of a prodrome of tingling, followed by pain and vesicle formation. The base of the vesicle is often very red and extends over a wider area than the vesicles. Initially the vesicles are clear, but they become cloudy over several days due to cellular response. Whitlow can be distinguished from paronychia and felon because there is no dorsal or volar swelling and the contour of the finger is normal except for the presence of vesicles (Fig. 14-139). Viral particles can be cultured from the vesicular fluid, but this requires 3 to 4 days. Characteristic findings on Tzanck stain will also confirm the diagnosis. The clinical findings are characteristic.

Treatment is conservative and consists of symptomatic care and preventing secondary infection or spread to other persons in contact with the child. The involved digit or hand can be covered and topical anesthetic preparations such as used for cold sores or teething pain applied to temporarily relieve the discomfort. Acetaminophen or ibuprofen is appropriate for the painful lesion. Oral acyclovir may be effective in shortening the course if given very early in the prodromal phase of the infection. Topical acyclovir is ineffective. The whitlow will run its course in 5 to 7 days. Immune compromise should be suspected in the child with repeated whitlow infections.

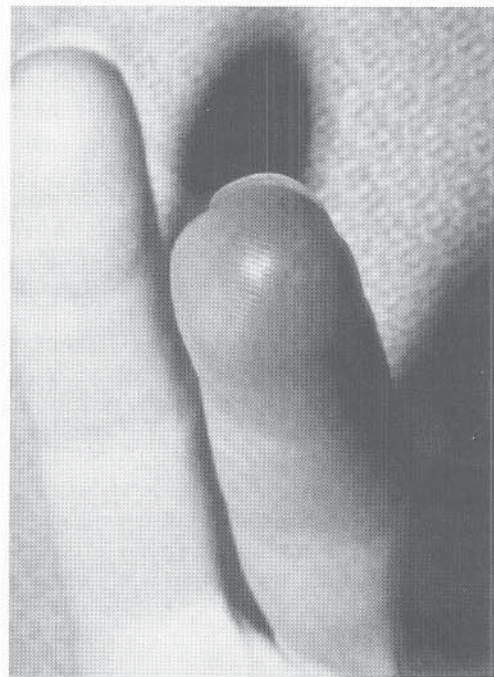


FIGURE 14-139 Herpetic whitlow. Note the vesicles on an erythematous base without general swelling of the digital tip. Secondary infection may occur; spread of the virus should be prevented.



FIGURE 14–140 Flexor tenosynovitis usually follows penetrating injuries with inoculation of the synovial sheath. Fusiform enlargement of the digit and pain on motion occur early in the course.

PYOGENIC TENOSYNOVITIS

Tenosynovitis is inflammation of the tenosynovium within the flexor tendon sheaths of the digits and palm. These are closed spaces in which a pyogenic infection spreads in a predictable fashion, into the thenar space or the radial and ulnar bursa with which the tendon spaces may communicate. The characteristic findings were described by Kanavel and include swelling of the flexor tendon sheath from the fingertip to the palm, tenderness over the flexor sheath, pain with passive extension of the finger (as extension increases the pressure in the compartment), and a flexed resting posture of the finger (Fig. 14–140). There is fusiform swelling of the digit, and there may be proximal lymphangitic signs. The child may also be febrile. Pyogenic tenosynovitis occurs within 12 to 24 hours following penetrating trauma. The usual organism is *S. aureus*.

Conditions from which pyogenic tenosynovitis must be distinguished include a subcutaneous abscess and septic arthritis. In the chronic case, especially where no organism can be cultured, psoriatic tenosynovitis may masquerade as pyogenic tenosynovitis.

Treatment for pyogenic tenosynovitis is elevation, intravenous antibiotics, and repeat clinical evaluation after 12 to 24 hours. If the characteristic signs are present after this initial treatment, drainage of the flexor sheath is indicated. The incision can be midaxial or multiple transverse incisions. Care must be taken to preserve the pulley system. Complications include stiffness, tendon rupture, and extension of the infection. Prior to recognition of the process and the availability of antibiotics, amputation was a frequent result of hand infections.

DEEP SPACE INFECTIONS

Deep space infections include thenar space and midpalmar space infections. The thenar space involves the base of the

thumb, radial to the long finger metacarpal, and around the adductor muscle. Involvement of this space results in a globular appearance of the hand with swelling and erythema. The midpalmar space infection involves the region of the palm ulnar to the long finger metacarpal. These infections follow penetrating trauma, and the usual organism is *S. aureus*. Treatment is initial elevation and parenteral antibiotics, with reassessment at 12- to 24-hour intervals until either resolution of the findings and symptoms or localization of the purulence, which must be surgically drained. Complications include skin necrosis, tendon rupture, scarring, and stiffness. Children may be toxic as a result of the infection.

BITE WOUNDS

Human Bite Wounds. In general, human bite wounds are a disease of adults. Children do occasionally sustain bite wounds, but the mouth flora of a child is less virulent than an adult's. The wound is usually in an area that is caught in the biter's teeth, and not that of the dorsal metacarpal "fight bite" seen in adults. The organisms to be wary of include the usual *Staphylococcus* and *Streptococcus* species, but also *Eikenella corrodens*, a small gram-negative rod that is a facultative anaerobe and is often difficult to culture. *Bacteroides* species are the most common anaerobes in the human mouth, and herpes simplex may also be inoculated.

Treatment depends on the timing of presentation. Early treatment includes splinting, elevation, and parenteral antibiotics (Timentin, Unasyn or penicillin and cefazolin) followed by oral augmentin, or penicillin and dicloxacillin. For established infection with cellulitis, lymphangitis, purulent drainage, or bone or joint infection, formal drainage and debridement followed by parenteral antibiotics is the treatment of choice. Complications include stiffness, growth abnormality, and necrosis of involved tissues.

Animal Bite Wounds. Children sustain bite wounds from animals more often than adults do because of their curiosity and initial lack of fear in approaching animals. The order of prevalence of biters is dogs, then cats, then rodents. The oral flora of dogs and cats often includes *Pasteurella multocida*, a small gram-positive coccus. The wound management and care are the same as for the human bite. Penicillin is effective against *P. multocida*, and a broad-spectrum antibiotic is appropriate for the other organisms.

Other bites from unusual domestic pets or wild or unknown animals may raise the suspicion of rabies, although this is extremely rare. CDC recommendations should be consulted for treatment. Human diploid vaccine and rabies immune globulin may be used in the treatment of established or strongly suspected cases.

SEPTIC ARTHRITIS

Septic arthritis of the hand in children almost always follows penetrating trauma. Signs of infection occur within 12 to 24 hours and include pain, loss of active range of motion, pain on passive range of motion, and discrete swelling of the joint segment rather than of the whole digit. Treatment is the same as for other early infections, and then joint drainage and debridement in the case where sepsis is identified. Aspiration and lavage of the joint in a child are probably

not indicated since an anesthetic is needed and the likelihood that the process would need repeating is high.

OSTEOMYELITIS

Osteomyelitis in the hand of a child is very rare. It is almost always associated with penetrating trauma and retention of a foreign body or devitalized tissue. Hematogenous osteomyelitis and joint sepsis tend not to involve the hand. The wrist and distal radius may be involved, especially with widespread sepsis.

OTHER INFECTIONS

Human Papillomavirus, Common Warts. Warts are common in children. Usually they are a nuisance and do not require treatment. The warts eventually will often spontaneously disappear. When treatment is sought, the goal of treatment is to eradicate the virus but preserve normal skin. Excision, cauterization, or freezing with liquid nitrogen all risk full-thickness loss of skin and scarring. Destruction of the periungual tissues is to be avoided.

A safe alternative treatment method is the injection of 1% plain Xylocaine at the base of the wart in the dermis, using a Luer-Lock type of needle until the base blanches. A second injection is sometimes needed if there is no regression of the wart after 3 weeks. A topical anesthetic cream lessens the initial discomfort of needle insertion. This method is also effective for the wart in the plantar location.

Atypical Mycobacterium Infections. Penetrating trauma with exposure to fresh or salt water may result in chronic infection with the atypical *Mycobacteria marinum* species. Other species abound, but infection is not as common as with *M. marinum*. Clinical presentation is a chronic, indolent swelling of synovial tissue, usually involving extrinsic tendons of the hand. Pain is often minimal and the complaint is limitation in range of motion combined with soft tissue swelling or thickening. Draining lymph nodes may be enlarged and should be considered as a source of biopsy material for culture. Systemic indicators of inflammation such as erythrocyte sedimentation rate, C reactive Protein, or leukocytosis are unusual. Acid fast stain of biopsy material may reveal organisms. Histology may show noncaseating granulomata. Diagnosis is established by culture. Specific instructions regarding suspected *M. marinum* should be conveyed to the laboratory so that material can be cultured at 30°C as well as at 37°. Treatment is organism specific and long-term.

CONDITIONS MIMICKING INFECTION

Although rare in children, a malignancy in the hand may mimic an infection. Tumor should be included in the differential diagnosis of lesions presenting as unusual or atypical infections. Inflammatory conditions may also mimic infectious tenosynovitis or septic arthritis. Dactylitis associated with sickle cell disease can mimic osteomyelitis.

These lesions should be biopsied as well as cultured, and appropriate blood studies should be done to look for immunologic or metabolic conditions.¹⁻⁹

REFERENCES

Infections

1. Eisenstein EM, Gesundheit B: Neonatal hand abscess, osteomyelitis and meningitis caused by *Streptococcus pneumoniae*. *Pediatr Infect Dis J* 1998;17:760.
2. Hoyer HA, Lacey SH, Graham TJ: Atypical hand infections. *Hand Clin* 1998;14:613.
3. Jebson PJ: Deep subfascial space infections. *Hand Clin* 1998;14:557.
4. Jebson PJ: Infections of the fingertip: paronychia and felons. *Hand Clin* 1998;14:547.
5. Karanas YL, Yim KK: *Mycobacterium tuberculosis* infection of the hand: a case report and review of the literature. *Ann Plast Surg* 1998;40:65.
6. Louis DS, Jebson PJ: Mimickers of hand infections. *Hand Clin* 1998;14:519.
7. Murray PM: Septic arthritis of the hand and wrist. *Hand Clin* 1998;14:579.
8. Newey ML, Magnussen PA: Deep hand infection complicating chickenpox in an otherwise healthy infant. *Br J Clin Pract* 1995;49:53.
9. Reilly KE, Linz JC, Stern PJ, et al: Osteomyelitis of the tubular bones of the hand. *J Hand Surg* 1997;22-A:644.

Traumatic Disorders

NEONATAL BRACHIAL PLEXUS PALSY

Birth-related upper limb paralysis was first mentioned in a midwifery text by Smellie in 1764.²⁶ Upper limb paresis was later described by Erb; however, not in infants. He localized the lesion to the point in the brachial plexus that now bears his name. He also credited Duchenne with describing the condition in infants, and the eponym Erb-Duchenne type of upper plexus palsy recognizes the contributions of the two men. Klumpke described lower plexus involvement.

The incidence, severity, and extent of brachial plexopathy in infants have been minimized by modern obstetric diagnosis and care. Induction of early labor when a large baby is anticipated, recognition of breech and other malposition by ultrasonography, and cesarian section delivery of these babies have certainly prevented cases of traumatic vaginal delivery. In spite of these measures, the risk and occurrence of neonatal brachial plexopathy have not been eliminated. The incidence remains at 0.3 to 2.5 per 1,000 live births.^{1,8,11,15,22,27,29,31} The trend toward larger birth weights as a result of better nutrition and general health of mothers and societal pressure to bring down cesarian section rates contribute to the number of cases still seen. However, the number of cases with severe involvement appears to have decreased.¹⁰

Etiology. Recognized risk factors include large birth weight, breech position, prior delivery of a child with a brachial plexopathy, shoulder dystocia, and a prolonged second stage of labor. At a fetal weight of approximately 3,500 grams, the cross-sectional area of the shoulders equals and then exceeds that of the head. Antenatal prediction of birth weight is notoriously inaccurate. Shoulder dystocia is recognized from the crowning and then recession of the head during the second stage of labor, reflecting failure of the shoulders to proceed into the birth canal. Labor may progress uneventfully until maternal factors determine the ease or difficulty of shoulder delivery.

The mechanism of injury is stretch across the plexus. The causes of this stretch include the forces of labor, especially in cases of shoulder dystocia, and extraction maneuvers.

Greater trauma occurs with forceful extraction maneuvers. Exactly how much stretch is needed to produce permanent injury in any infant is not known. A breech delivery focuses stretch at the lower plexus, with traction applied to the trunk with an abducted arm. A vertex delivery focuses stretch at the upper plexus, with lateral traction applied at the neck. The anatomic variation of the so-called prefixed plexus, with a greater contribution from the C4 root, may predispose some infants to tolerate less stretch across the upper plexus.

In experimental studies, Wickstrom and colleagues looked at the tensile strength of the plexus.^{32,33} Stronger connective tissue and a more oblique angle of traverse across the neck provided greater support to the upper plexus at the root level. A more transverse position and weaker proximal soft tissue support made the lower plexus more susceptible to disruption with less tensile force. Others have correlated the clinical findings of tendency for intraplexal disruptions of the upper plexus and root avulsions of the lower plexus with these findings.^{6,21} Root avulsions with injury at the cord level account for the occasional upper motor neuron lesions and lower limb findings in some of these children.

Classification. Classification by the part of the plexus involved allows comparisons of prognoses, treatments, and outcomes. Narakis recognizes four categories of anatomic plexus involvement.²⁰ In group 1 are upper plexus lesions or lesions of C5 and C6; they are recognized by weakness of the shoulder abductors, external rotators, elbow flexors, and wrist extensors. In group 2 are lesions of C5, C6, and C7, which also lack elbow extension and are associated with weaker shoulder adductors. In group 3 are the panplexus lesions. The fourth group is characterized by lower plexus involvement only; these lesions are rarely seen in isolation.

The types of nerve lesions were classified by Seddon according to terms described by Cohen.²³ *Neurapraxia* is paralysis in the absence of peripheral degeneration; the delay to recovery may be long, but recovery will be complete. *Axonotmesis* is damage to nerve fiber with complete peripheral degeneration but with intact external tissues to provide support for accurate spontaneous regeneration. Good recovery is anticipated, and no intervention can improve the outcome. In *neurotmesis* all essential structures, both neural and supporting tissues, have been disrupted. This category includes neuroma in continuity, division of nerve, and anatomic disruption.

Sunderland²⁸ also classified nerve lesions, as follows: A first-degree injury preserves all structure, but conduction is temporarily blocked. In second-degree injury wallerian degeneration occurs, but endoneurial integrity is maintained and recovery is complete. A third-degree injury adds endoneurial destruction and internal fascicular disorganization; recovery is poorer, with possible cross-regeneration. In fourth-degree injury there is complete internal disorganization, but some continuity of external structure remains. A fifth-degree injury involves complete disruption of all nerve structures.

Most stretch injuries of the brachial plexus involve a mixture of types of nerve lesions and may involve multiple sites of injury. Injury to the surrounding tissue may result in perineural fibrosis that further slows or precludes spontaneous recovery.

Clinical Features. The newborn with a brachial plexus injury has decreased spontaneous movement and asymmetry of infantile reflexes such as Moro's reflex or asymmetric tonic neck reflex. With involvement of the lower plexus, the grasp reflex may be absent. An ipsilateral Horner's syndrome consisting of ptosis, miosis, and enophthalmos, or a small pupil with a droopy eyelid, indicates injury to the T1 cervical sympathetic nerves. Phrenic nerve involvement is said to occur in up to 5 percent of upper plexus lesions.

Cephalic hematoma, laryngeal nerve injury with vocal cord paralysis, and facial nerve paralysis in cases of forceps-assisted delivery may be found and are evidence of birth-related trauma. Cord-level trauma due to root avulsion should be suspected in cases of lower limb weakness or spasticity. Fractures of the clavicle, humerus, and other long bones may also be seen. Ipsilateral clavicle fracture is actually a favorable finding in birth-related plexopathy as the fracture allows the shoulder girdle to compress, thus decreasing the overall traction on the plexus. Cerebral palsy associated with brachial plexopathy is rare, and if and when it is encountered, it would be the type with global involvement associated with hypoxia. Spastic hemiparesis is not associated with brachial plexopathy.

Upper plexus or Erb-Duchenne-type lesions initially present with a shoulder abductor and external rotator weakness and absence of elbow flexors. The upper limb is positioned in adduction, internal rotation, and elbow extension. The wrist will often be held in a flexed position. If C7 is also involved, the elbow extensors will also be weak and the elbow will be held in midposition.

In the globally involved limb, the hand will also be weak, cool, and maintained in a supinated and intrinsic minus position. The mother will be able to report whether sensation is present or absent. Sensory loss of the hand may be extensive in the lower plexus type.

Prognosis and Natural History. The natural history of neonatal brachial plexus palsy varies with the author according to medical specialty, referral patterns, and the age of patients at referral. Reported rates of complete recovery range from 7 to 95 percent.^{8,10,11,27,29} In patients whom we followed from birth, we found a 50 percent rate of full recovery. Gilbert reported in 1984 that infants who did not recover elbow flexion by age 3 months had a poor chance of full recovery.⁷ He based this conclusion on a study by Tassin in which 44 infants were followed from birth; 20 did not recover shoulder and elbow function by 3 months of age.³⁰ Since Gilbert's report appeared, there has been intense renewed interest in early surgical intervention in the plexal injury. Controversy continues over the timing of surgical intervention, the type of surgical procedure (neurolysis versus intraplexal or extraplexal nerve transfers or grafts), and the anticipated results from nerve procedures in comparison with the natural history of recovery. Later orthopaedic reconstructive procedures are reliable in augmenting function that has recovered poorly.

Differential Diagnosis. In the newborn, fracture of the clavicle or humerus or proximal humeral physeal separation manifest as diminished spontaneous movement. Fractures of the clavicle and humeral shaft are relatively common; proximal physeal separation is rare. In an infant with normal or unclear initial movement, septic arthritis of the shoulder,

acute osteomyelitis, or some other form of sepsis must be considered even in the absence of fever and generalized toxicity. Fracture or injury due to child abuse must also be ruled out. Loss of normal reflexes occurs in all of these conditions. Radiographs are indicated for further investigation. Tumors involving the spinal cord or plexus are rare but should be considered if there is deterioration in function. Congenital malformation of the plexus is also rare and can be verified by exploration. Gilbert has reported on four cases of aplasia of the cervical spinal roots,⁵ some associated with other malformations in the upper limb. Postinfectious plexopathy of the Parsonage-Turner type may occur and usually results in a flaccid paralysis of the muscles innervated by the involved nerves.

Initial Treatment. The involved upper limb should be protected initially by pinning the sleeve to the shirt or wrapping it to the body for the first several weeks. Gentle range-of-motion exercises must be initiated as soon as the child can comfortably tolerate this treatment. Muscle imbalance develops rapidly, and soft tissue contracture contributes to deformity and joint incongruity early in the neonatal period.

ASSESSMENT. During the first several months after birth, careful, repeated observations are needed to establish a pattern of recovery. Attention to the shoulder range of motion, especially maintaining passive external rotation, is critical. If there is a sudden loss of range of passive external rotation, the shoulder should be investigated with sonography or other imaging to determine whether it is subluxating or dislocating posteriorly. Closed or open reduction, contracture stretching or release, and casting are needed to treat posterior shoulder instability.

Usually by 4 months a decision is made to obtain electrodiagnostic studies to look for early evidence of reinnervation in muscles that have not yet recovered. The window for surgical intervention on the plexus is usually between 4 months and 1 year of age.

It is essential that the shoulder remain congruously reduced during this period of motor reinnervation.

Later Treatment. If motor recovery is not adequate to maintain shoulder muscle balance, early contracture release and muscle transfer to the external rotators should be considered before established joint deformity occurs. Residual deformity of the shoulder depends on the type of lesion, the degree of recovery, and, to a lesser extent, the diligence of the patient and family in doing the range-of-motion exercises.

If the shoulder joint is still congruously reduced and the humeral head has not flattened, muscle rebalancing procedures should be considered. If the shoulder has become “congruously incongruent,” meaning that the potential to increase a smooth range of motion is no longer present, extra-articular procedures to improve limb position are more effective.

Residual Deformities. Residual deformities of the shoulder have been classified by Zancolli into several main types.³⁵ The first general category includes those with contractures. The most common is the internally rotated, adducted shoulder. This can be seen with or without joint deformity. Affected children present with limited range of motion, often

augmenting their hand position with hyperlordosis and lateral trunk motion (Fig. 14–141). Tassin has described Mallet’s classification of the typical positions of upper limb function in this type of shoulder deformity.³⁰ Involvement of the suprascapular nerve paralyzes the supra- and infraspinatus muscles, resulting in loss of external rotation, weakness of posterior capsular support, and loss of steering capacity of that portion of the rotator cuff. A contracture of the anterior deltoid may also be present, making this a complex deformity that is difficult to reduce even with open treatment (Table 14–8).

Zancolli’s second most common type is the externally rotated abducted type. Affected patients exhibit winging of the scapula on attempts at internal rotation—Putti’s sign. Overhead function is usually good, but the cosmetic deformity of sloping shoulder and the fixed winging of the scapulae are sometime unacceptable (Fig. 14–142).

Loss of extension of the shoulder may be related to posterior subluxation of the humeral head from the glenoid and loss of the normal fulcrum and joint forces, or to loss of posterior deltoid function.

A second general category of upper limb deformity is characterized by flaccid paralysis of the upper limb. This is extremely unusual in neonatal plexopathy; it is usually seen after other types of plexus trauma or in the Parsonage-Turner syndrome.

ELBOW. Flexion contracture of the elbow occurs commonly in brachial plexopathy. It is somewhat paradoxical because the elbow flexors are initially flail. If anything, one would expect extension contracture because of the greater involvement of C5 and C6. One explanation is that the reinnervation is not accurate and that cocontraction of antagonistic flexors and extensors develops. Greater relative involvement of C7 weakens elbow extensors and also contributes to the contracture.

Muscle imbalance with flexor overpull followed by joint deformity makes the elbow flexion contracture difficult to treat. The contracture may progress relentlessly, reaching 70 degrees or greater by skeletal maturity. Diligent nighttime splinting may help limit the severity of the deformity and may offer partial correction, but must be carried out over many years.

Posterior subluxation or dislocation of the radial head was found in 27 of Aitken’s 107 cases and was thought to be due to treatment that rigidly splinted the elbow into flexion.² He postulated that spasticity in the elbow extensors led to muscle imbalance; however, spasticity is not present unless there has been upper motor neuron dysfunction.

Anterior dislocation of the radial head may be seen in plexopathy that involves C7 and C8 to a greater degree than C5 and C6. The fixed supination posture maximizes the anteriorly dislocating force of the biceps at the proximal radius. Attempts to stretch out the shortened biceps and contracted joint may increase the subluxating force and result in an anterior dislocation of the radial head.

WRIST. Depending on the level of plexus involvement and the degree of recovery, several deformities may be seen at the wrist. Lack of wrist extension occurs with C6 involvement. C7 innervation of the extensor carpi ulnaris may cause ulnar deviation of the wrist but does not provide extension. The extensor carpi ulnaris may be strong enough to be



FIGURE 14-141 Obstetric brachial plexus paralysis of the right upper limb in a 6-year-old boy. Note the limitation of active abduction and lateral rotation of the shoulder, and also the pronation contracture of the forearm.

TABLE 14-8 Classification of Shoulder Sequelae in the Upper Arm Type of Birth Palsy

Group	Subgroup*	Scapular Elevation Sign	Muscle Contracture		Initial Obstetric Lesion	Surgical Treatment
			Primary	Secondary		
Group 1: Joint contracture (90.2%)	1. Internal rotation-adduction contracture without joint deformity or dislocation (26.5%)	Negative	Subscapularis	Pectoralis major Coracobrachialis		Anterior shoulder release plus external rotation transfer
	2. Internal rotation-adduction contracture with joint deformity and posterior subluxation or dislocation (67.4%)	Positive	Subscapularis	Short head of biceps Anterior deltoid	Upper brachial plexus and scapulothoracic joint	External rotation osteotomy
	3. External rotation-abduction contracture with anteroinferior subluxation or dislocation (4.8%)	Positive	Infraspinatus, teres minor	Posterior deltoid		Posterior shoulder release or internal rotation osteotomy
	4. Pure abduction contracture (1.2%)	Positive	Supraspinatus	—		Supraspinatus lengthening
Group 2: Pure flaccid paralysis (9.8%)	Shoulder abduction and rotation paralysis and elbow flexion paralysis	Negative	—	—	Upper brachial plexus	Shoulder arthrodesis and elbow flexoplasty

*Statistics in this classification are based on the evaluation of 92 patients with surgical reconstruction of the shoulder. From Zancolli EA: Classification and management of the shoulder in birth palsy. *Orthop Clin North Am* 1981;12:433.

transferred into the extensor carpi radialis brevis to effect wrist extension.

Lower plexopathy allows unopposed supination and may cause instability of the distal radioulnar joint. A late change in the position of the wrist toward forearm neutral position or pronation may require osteotomy, decompression of the

distal radioulnar joint with resection of the ulnar head, or a Sauve-Kapandji procedure (Fig. 14-143).¹⁴

HAND. Involvement of the hand indicates a poor functional outcome because of sensory and motor impairment. Recovery of extrinsic and intrinsic muscle function is less likely

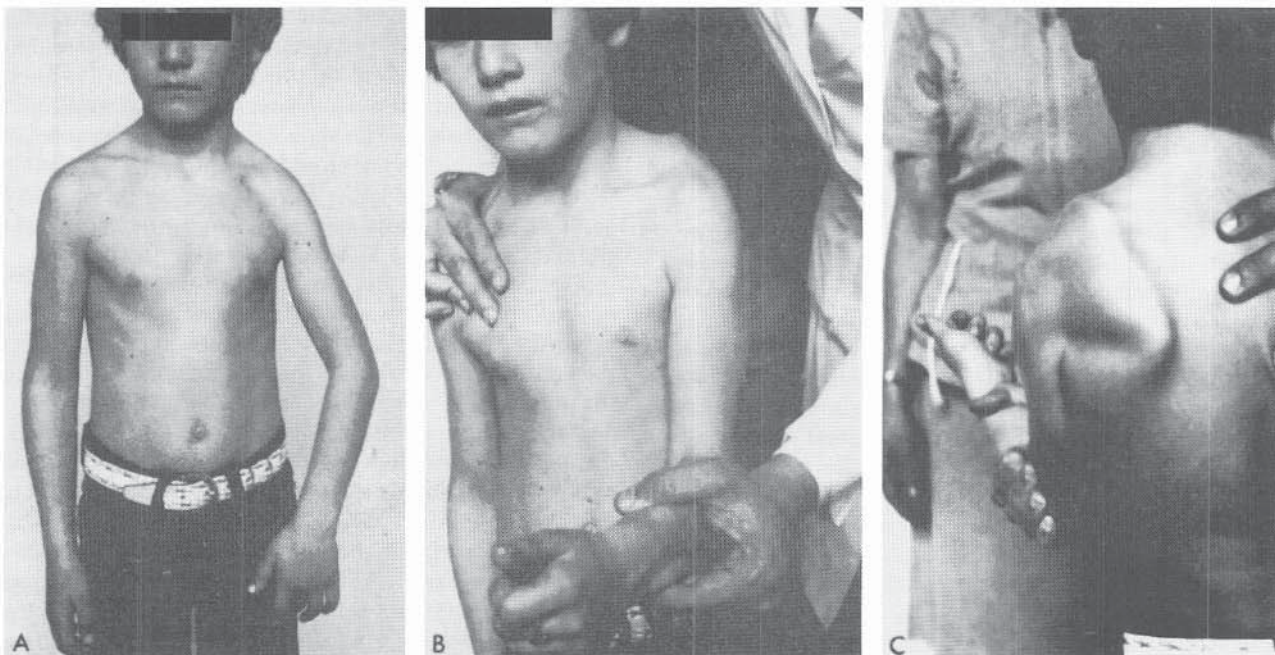


FIGURE 14-142 Putti's sign. A, Severe medial rotation contracture in a 10-year-old boy with brachial plexus paralysis. Note that the glenohumeral joint is in abduction. B, Passive adduction and lateral rotation results in a positive scapular sign. C, Putti's maneuver.

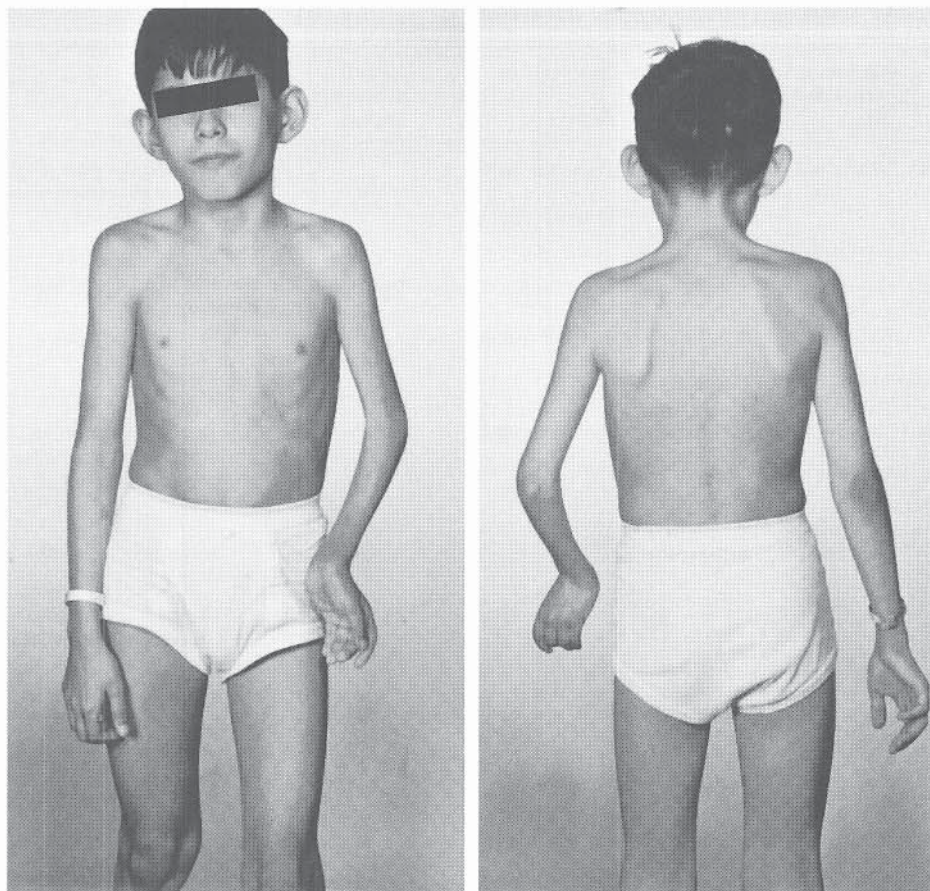


FIGURE 14-143 Supination contracture of the left forearm and flexion deformity of the elbow in the whole-arm type of brachial plexus paralysis. Note also the extension and *hypersupinated* position of the wrist.

because of the greater probability of root avulsion in lower plexus lesions and the distance from the lesion to the muscle or sensory end-organ.

Management of Residual Deformities

SHOULDER

Internal Rotation, Adduction Contracture of the Shoulder. The selection of shoulder procedures depends on the congruency of the shoulder, the residual plasticity and remodeling potential of the skeletal elements, the strength of active muscles, the condition of the hand, and the concerns of the individual patient and family. The initial goal is to keep the shoulder reduced during motor reinnervation. Contractures that are not responsive to stretching limit joint motion and cause progressive deformity of the growing cartilaginous joint configurations. While the joint is still congruous, or has the potential to remodel if the joint is reduced, soft tissue procedures to reduce and rebalance the shoulder are indicated. If recovery of external rotator muscles has not occurred, contracture will likely recur if muscle imbalance is not corrected at the time of contracture release. Once the shoulder has developed bony deformity, soft tissue procedures will not correct deformity, nor will they increase motion or relieve discomfort. Surgical procedures in the older child are limited to changing the position of the limb to improve appearance or function in a different zone of hand access.

The etiology of the contracture is muscle imbalance be-

tween strong external rotator and weak external rotator muscles. The adductors and internal rotators are the latissimus dorsi, the subscapularis, the pectoralis major and minor, and the teres major. The external rotators of the shoulder are the teres minor and the infraspinatus. Of the internal rotators, the latissimus and teres minor are available for transfer to rebalance the shoulder rotators. The subscapularis tendon is tightly adherent and contracted at the anterior part of the shoulder joint. The coracoid and acromion become beaked and close the vault of the anterior part of the shoulder joint as the humeral head slips posteriorly.

Operations have been described for correction of the internal rotation deformity. Fairbank approached the shoulder anteriorly and released the tight subscapularis, the upper portion of the pectoralis major, and the anterior capsule of the shoulder.⁴ Sever divided the entire subscapularis and pectoralis tendons but left the capsule intact in order to prevent anterior dislocation.^{24,25} L'Episcopo added a transfer of the teres major to an external rotator position,^{16,17} and Zachary added the latissimus dorsi to the external rotator transfer.³⁴ Transfer of the origin of the anterior portion of the deltoid to a posterior position was thought by Green to add external rotation.⁹ Lengthening of the subscapularis and pectoralis major with transfer of the latissimus and teres major to the external rotator position was the procedure used by Green and Tachdjian.⁹ The anterior procedures that lengthen the subscapularis tendon at the insertion all risk

anterior instability and leave the anterior shoulder markedly scarred. Section of the pectoralis major tendon destroys the contour of the anterior axillary fold and leaves a defect that is unsightly.

Others modified the manner of attachment of the tendon transfers, Zancolli by long Z-lengthening of the latissimus dorsi tendon and relative lengthening of the subscapularis by attaching it to the tendon of the pectoralis major.³⁵

Ingram lengthened the subscapularis at its origin on the anterior surface of the scapula and moved the insertion of the teres minor proximally by transferring it into the infraspinatus insertion.¹³ Hoffer and colleagues used the teres major and latissimus dorsi to transfer into the rotator cuff at the insertion of the supraspinatus to add both external rotation and abduction.^{12,20} Carliz and Brahimi combined release of the subscapularis with transfer of the latissimus dorsi and teres major to the rotator cuff.³

Attempts to rebalance the shoulder must preserve some internal rotation function or a fixed external rotation deformity may develop with subsequent inability to reach the midline and marked winging of the scapula.

Bony procedures that have been described in the older child include resection of the coracoid and acromioplasty⁹ and rotational osteotomy of the humerus.^{32,33} The goal of these procedures is to relieve discomfort and reposition the limb for better function in front of the body. Osteotomy of the humerus at a proximal level rotates the insertion of the deltoid from an anterior to a lateral position and can augment lateral abduction.

Author's Recommendation for Correction of Internal Rotation Shoulder Deformities. In young children less than 1 year old there is still a chance for recovery of the external rotator muscles. Electrodiagnostic studies to examine reinnervation potentials in the infraspinatus muscle can help the surgeon decide whether or not a transfer to augment external rotation is also needed. Consideration should be given to microsurgical exploration of the suprascapular nerve with decompression or neurolysis either at the same time as contracture release or prior to it.

If there is still a potential for recovery of the external rotator function and a fixed contracture develops, the shoulder is examined with the child under general anesthesia. If the shoulder is easily brought to a position of 60 degrees or more of external rotation *with the humerus at the child's side in an adducted position*, a cast is applied maintaining this position and worn for 4 to 6 weeks. Care must be taken not to cause a fracture at the proximal humerus as the shoulder is manipulated. The elbow should be flexed and the forearm supinated during the external rotation manipulation to protect the radial head and elbow joint. If a fracture occurs, the child is still casted in a position of 45 degrees of external rotation.

If the contracture is fixed and the shoulder is not completely dislocated, the open technique for contracture release is a sequential approach to the subscapularis, the latissimus dorsi, the coracoacromial ligament, and the pectoralis major. Release of the subscapularis at its origin is done as described by Carliz³ but through a *transverse* incision in the posterior axillary fold two to three fingerbreadths (the child's finger) proximal to the inferior pole of the scapula. Through this incision the latissimus dorsi muscle and tendon can be iso-

lated and developed for transfer if needed. The quality and contractility of the teres minor and infraspinatus can also be observed directly if desired. The lateral edge of the scapula anterior to the teres major muscle is identified, and under direct vision the entire subscapularis muscle can be elevated from the anterior aspect of the scapula. The serratus muscle and the scapular stabilizers attaching to the medial border of the scapula must not be detached. At this point the shoulder is gently brought into external rotation and checked for improvement. If the release is inadequate, or if there is residual posterior subluxation, the coracoacromial and coracohumeral ligaments are released through a small separate saber incision. Reduction and external rotation are again checked.

If the contracture still remains tight, intramuscular or Z-lengthening of the pectoralis major tendon is done through an anterior axillary incision to preserve the anterior axillary fold.

The postoperative cast is a long-arm cast with the elbow at 90 degrees, the forearm supinated, and the wrist in slight extension. The cast is affixed to a wide, well-padded lumbosacral band of plaster or fiberglass. The abdominal area is relieved during application of the cast to allow space for the abdominal contents when the child is upright, and to allow space for meals. Old wooden crutches make ideal struts to attach the arm cast to the trunk. The curve of the crutch contours nicely and the size is easily cut to fit.

If the shoulder is posteriorly dislocated and a posterior capsular redundancy is present on reduction of the shoulder, a posterior capsulorrhaphy is done through an approach between the infraspinatus and teres minor.

If a decision has been made to transfer to augment external rotation, I prefer using the latissimus dorsi, mobilizing its full length and taking the tendon off the proximal humerus. This can be done through the same transverse incision. The tendon of the latissimus is secured with multiple weave-type, nonabsorbable sutures before it is passed through a subcutaneous tunnel across the posterior axillary fold to the proximal humerus. The tendon can be placed into a bony trough or secured into drill holes along the lateral aspect of the humerus proximal to the deltoid insertion. Alternatively, the tendon can be secured to the middle or posterior third of the deltoid insertion.

Transfer of the latissimus dorsi and the teres major tendons into the supraspinatus tendon as described by Hoffer and colleagues¹² offers augmentation of both abduction and external rotation. Requisites for the procedure include a concentrically reduced shoulder without significant deformity, a normal passive range of motion, and at least 60 degrees of active abduction (Fig. 14-144, Plate 14-1).

The latissimus is developed as described above and passed superficial to the teres minor and the infraspinatus, through an opening in the posterior deltoid fascia, and under the posterior deltoid, then retrieved through a separate saber incision that exposes the supraspinatus tendon by detaching a portion of the middle and posterior deltoid from the acromion. The teres major tendon often is too bulky and too short to reach, and the latissimus tendon alone is transferred. The latissimus tendon is then woven into the tendon of the supraspinatus with nonabsorbable suture. The deltoid is reattached, and after surgery the limb is casted with a long-arm cast affixed to the lumbosacral cast with a strong

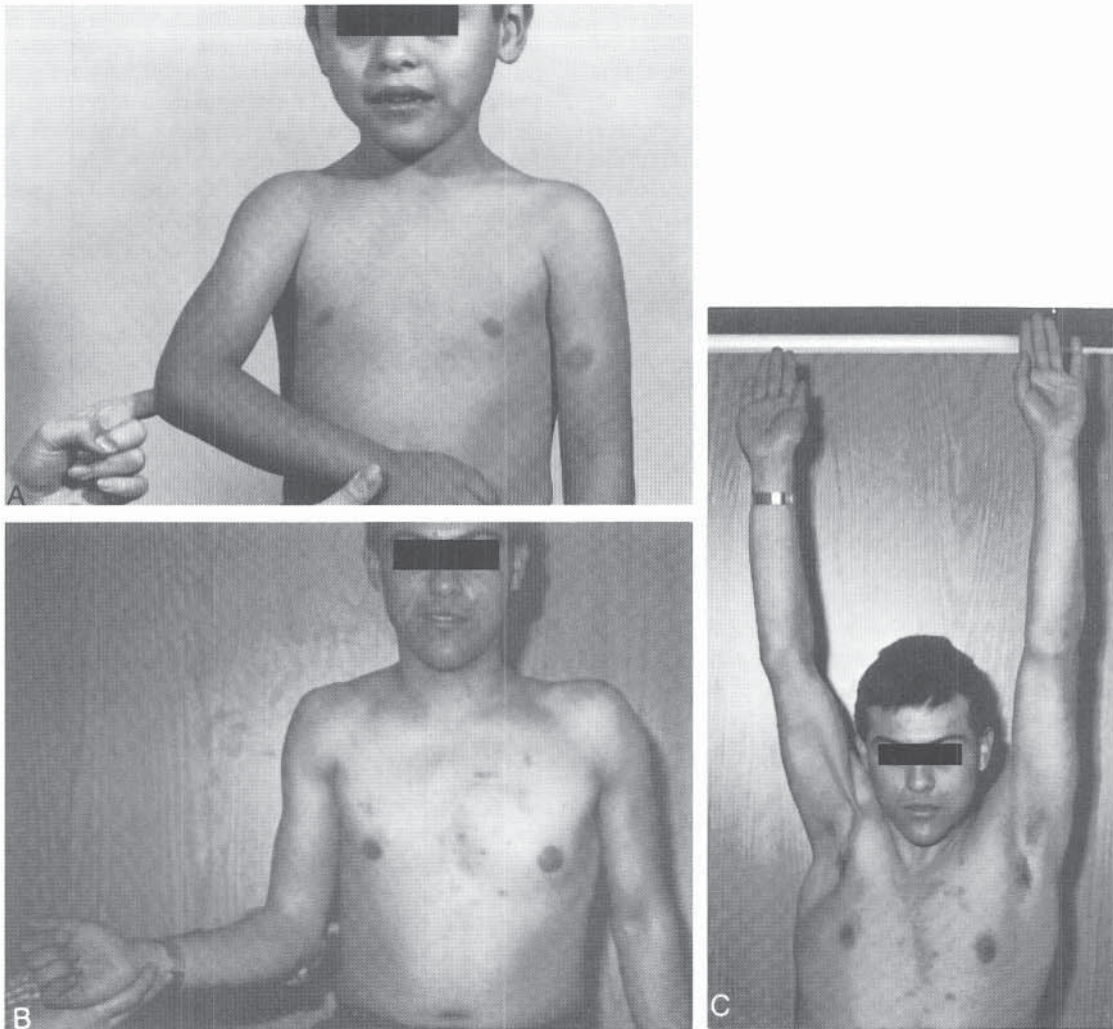


FIGURE 14-144 Results of transfer of the latissimus dorsi and teres major muscles to the rotator cuff in brachial plexus paralysis. A, Preoperative photograph of patient showing attempted abduction and lateral rotation. B and C, Postoperative photographs. Note the range of abduction and lateral rotation. (Courtesy of M. Hoffer, M.D.)

strut in a position of abduction and external rotation—the Statue of Liberty cast.

Later procedures in the presence of joint deformity are directed to changes in the position of the limb. A proximal humeral osteotomy above the deltoid insertion can be done through either an anterior or a posterior approach. Z-lengthening of the pectoralis major tendon allows preservation of the anterior axillary fold and also covers the plate on closure. The osteotomy is held with internal plate and screw fixation, and postoperatively the limb is protected with a sling until the osteotomy has healed (Plate 14-2).

Abduction and External Rotation Contracture. An abduction and external rotation contracture of the shoulder is often very functional, and the complaint is of fixed winging of the scapula. Overhead abduction and position of the hand in space are not functionally limiting in these children, unlike children with the internal rotation and adduction contractures. Zancolli has described anterior subluxation and dislocation of the shoulder in this type of deformity.³⁵ The posterior contracture can be released by dividing the teres minor at the insertion and the infraspinatus tendon at the

musculotendinous tendon. The tendon of the infraspinatus is sutured to the tendon of the teres minor after the shoulder is internally rotated (Figs. 14-145 and 14-146).

Abduction Contracture. Abduction contracture of the shoulder is due to shortening of the deltoid. The dilemma is that lengthening the deltoid will weaken it and may result in less functional range of motion. The complaint in this situation is cosmetic winging of the scapula. Patients and families must be counseled carefully about the potential for loss of range of motion. If a release is needed, the deltoid can be recessed from its origin, or the tendon of insertion can be brought proximally. Neither of these procedures is very satisfying.

ELBOW AND FOREARM. Flexion contracture of the elbow may develop relentlessly despite efforts at long-term splinting. Fixed extension contracture of the elbow is less functional than moderate flexion. In the absence of a strong triceps, the contracture will recur. Surgical release may result in greater stiffness and an overall loss of arc of motion and should be undertaken cautiously. Resection of bone, release

of capsule, and lengthening of tendons may be needed to effect extension.

A forearm deformity must be considered in the context of the function of the entire limb. A position of neutral to slight pronation is preferable to a position in either extreme of supination or pronation. Full active supination is usually limited in upper plexopathy and active pronation is weak in lower plexopathy. Attention to maintaining passive range of forearm rotation will usually achieve a functional range of motion. Unacceptable pronation can be addressed by lengthening or rerouting the pronator teres. A bony torsional deformity of the forearm may develop, and correction will not be possible by soft tissue procedures. Rotational osteotomy with an osteoclast technique is usually sufficient to adjust the position of the forearm.

Unopposed supination in which passive pronation is still possible can be addressed by Zancolli's technique, which involves a Z-lengthening and rerouting of the biceps tendon. The distal limb of the Z is passed between the radius and ulna and brought to the dorsal side of the radius, where it is secured to the proximal limb of the Z, thus creating a pronation moment for the biceps force (Plate 14–3). Fixed supination deformity is more awkward as it precludes a tenodesis effect for release of objects from the hand, which is usually severely affected by the lower plexopathy. The occasional hypersupination deformity will be associated with subluxation or dislocation of the distal radioulnar joint. Forced change in position of an incongruous joint will often result in a painful situation. Decompression of the joint is achieved with a distal ulnar resection or, preferably, the Sauve-Kapandji technique,¹⁸ in which a motion segment is created with an ulnar resection proximal to the joint and an arthrodesis of the distal radioulnar joint.

Anterior dislocation of the radial head may be minimized by transfer of the biceps tendon to the brachialis tendon and an attempt at reduction. Posterior dislocation of the radial head is best addressed at the time that symptoms develop, and can be treated with resection.

HAND. Motor and sensory impairment of the hand in lower plexopathy is profound. Reconstructive operations to improve the function of the hand are limited to the child who attempts to use the hand and who has a specific need, with dependable motor units available for tendon transfer. The surgeon who has experience with reconstruction of multiple peripheral nerve deficits and who understands the consequences of a surgical alteration in a learned pattern and its implications for the patient may be able to improve the condition of the hand in carefully selected patients with hand involvement in brachial plexopathy.

REFERENCES

Traumatic Disorders

- Adler JB, Patterson RL Jr: Erb's palsy: long-term results of treatment in eighty-eight cases. *J Bone Joint Surg* 1967;49-A:1052.
- Aitken J: Deformity of the elbow joint as a sequel to Erb's obstetrical paralysis. *J Bone Joint Surg* 1952;34B.
- Carlioz HN, Brahimi L: Place of internal disinsertion of the subscapularis muscle in the treatment of obstetrical paralysis of the upper limb in children. *Ann Chir Infant* 1971;12:159.
- Fairbank HAT: Birth palsy: subluxation of shoulder joints in infants and young children. *Lancet* 1913;1:1217.
- Gilbert A: Obstetrical brachial plexus palsy. In Tubiana R (ed): *The Hand*, vol 4. Philadelphia, WB Saunders Co, 1993.
- Gilbert A, Khouri N, Carlioz H: Birth palsy of the brachial plexus: surgical exploration and attempted repair in twenty-one cases. *Rev Chir Orthop Reparatrice Appar Mot* 1980;66:33.
- Gilbert A, Tassin J: Reparation chirurgicale du plexus brachial dans la paralysie obstetricale. *Chirurgie* 1984;110(1):70.
- Gordon M, Rich H, Deutschberger J, et al: The immediate and long-term outcome of obstetric birth trauma. I. Brachial plexus paralysis. *Am J Obstet Gynecol* 1973;117:51.
- Green WT, Tachdjian MO: Correction of residual deformities of the shoulder in obstetrical palsy. *J Bone Joint Surg* 1963;45-A:1544.
- Greenwald AG, Schute PC, Shiveley JL: Brachial plexus birth palsy: a 10-year report on the incidence and prognosis. *J Pediatr Orthop* 1984;4:689.
- Hardy AE: Birth injuries of the brachial plexus: incidence and prognosis. *J Bone Joint Surg* 1981;63-B:98.
- Hoffer MM, Wickenden B, Roper B: Brachial plexus birth palsies: results of tendon transfers to the rotator cuff. *J Bone Joint Surg* 1978;60-A:691.
- Ingram AJ (ed): *Campbell's Operative Orthopedics*, p 1722. St. Louis, CV Mosby Co, 1971.
- Kapandji IA: The Kapandji-Sauve operation: its techniques and indications in non rheumatoid diseases. *Ann Chir Main* 1986;5:181.
- Khatree MH, Gamsu HR, Rudd P, et al: Features predictive of brachial plexus injury during labour. *S Afr Med J* 1982;61:232.
- L'Episcopo JB: Restoration of muscle balance in the treatment of obstetrical paralysis. *NY State J Med* 1939;39:357.
- L'Episcopo JB: Tendon transplantation in obstetrical paralysis. *Am J Surg* 1934;25:122.
- Millroy P, Coleman S, Ivers R: The Sauve-Kapandji operation: technique and results. *J Hand Surg* 1992;17-B:411.
- Moore BH: A new operation for brachial birth palsy-Erb's palsy. *Surg Gynecol Obstet* 1935;61:832.
- Narakas A: Brachial plexus surgery. *Orthop Clin North Am* 1981;12:303.
- Narakas A: Indications and results of direct surgical treatment in lesions caused by stretching of the brachial plexus. I. Indications for direct surgical treatment. *Rev Chir Orthop Reparatrice Appar Mot* 1977;63:88.
- Rossi LN, Vassella F, Mumenthaler M: Obstetrical lesions of the brachial plexus: natural history in 34 personal cases. *Eur Neurol* 1982;21:1.
- Seddon HJ: Lesions of individual nerves: upper limb. In *Surgical Disorders of the Peripheral Nerves*. London, Churchill Livingstone, 1949.
- Sever JW: Obstetrical paralysis. *JAMA* 1925;85:1862.
- Sever JW: Obstetrical paralysis. *Surg Gynecol Obstet* 1927;44:5547.
- Smellie W: Collection of Preternatural Cases in Observations in Midwifery: Completing the Design of Illustrating His First Volume on That Subject, vol III, p 504. London, Wilson & Durham, 1764.
- Specht EE: Brachial plexus palsy in the newborn: incidence and prognosis. *Clin Orthop* 1975;110:32.
- Sunderland S: *Nerves and Nerve Injuries*. Edinburg, Churchill-Livingstone, 1978.
- Tan KL: Brachial palsy. *J Obstet Gynaecol Br Commonw* 1973;80:60.
- Tassin JL: Paralysies obstetricales du plexus brachial evolution spontanée resultats des interventions reparatrices precodes. *Faculte de Médecine VII, Université Paris*, 1983.
- Vassalos E, Prevedourakis C, Paraschopoulou P: Brachial plexus paralysis in the newborn. *Am J Obstet Gynecol* 1968;101:554.
- Wickstrom J: Birth injuries of the brachial plexus: treatment of defects in the shoulder. *Clin Orthop* 1962;23:187.
- Wickstrom J, Haslam ET, Hutchison B: The surgical management of residual deformities of the shoulder following birth injuries of the brachial plexus. *J Bone Joint Surg* 1955;37-A:27.
- Zachary RB: Transplantation of teres major and latissimus dorsi for loss of external rotation at the shoulder. *Lancet* 1947;2:757.
- Zancolli EA: Classification and management of the shoulder in birth palsy. *Orthop Clin North Am* 1981;12:433.

Tumors of the Upper Limb

A mass in the upper limb in a child is a cause for concern, even though only about 2 percent of lesions were malignant in a recently reported large series.³ In taking the history, the examiner should include questions about onset, presence at

Text continued on page 496

Latissimus Dorsi and Teres Major Transfer to the Rotator Cuff

OPERATIVE TECHNIQUE

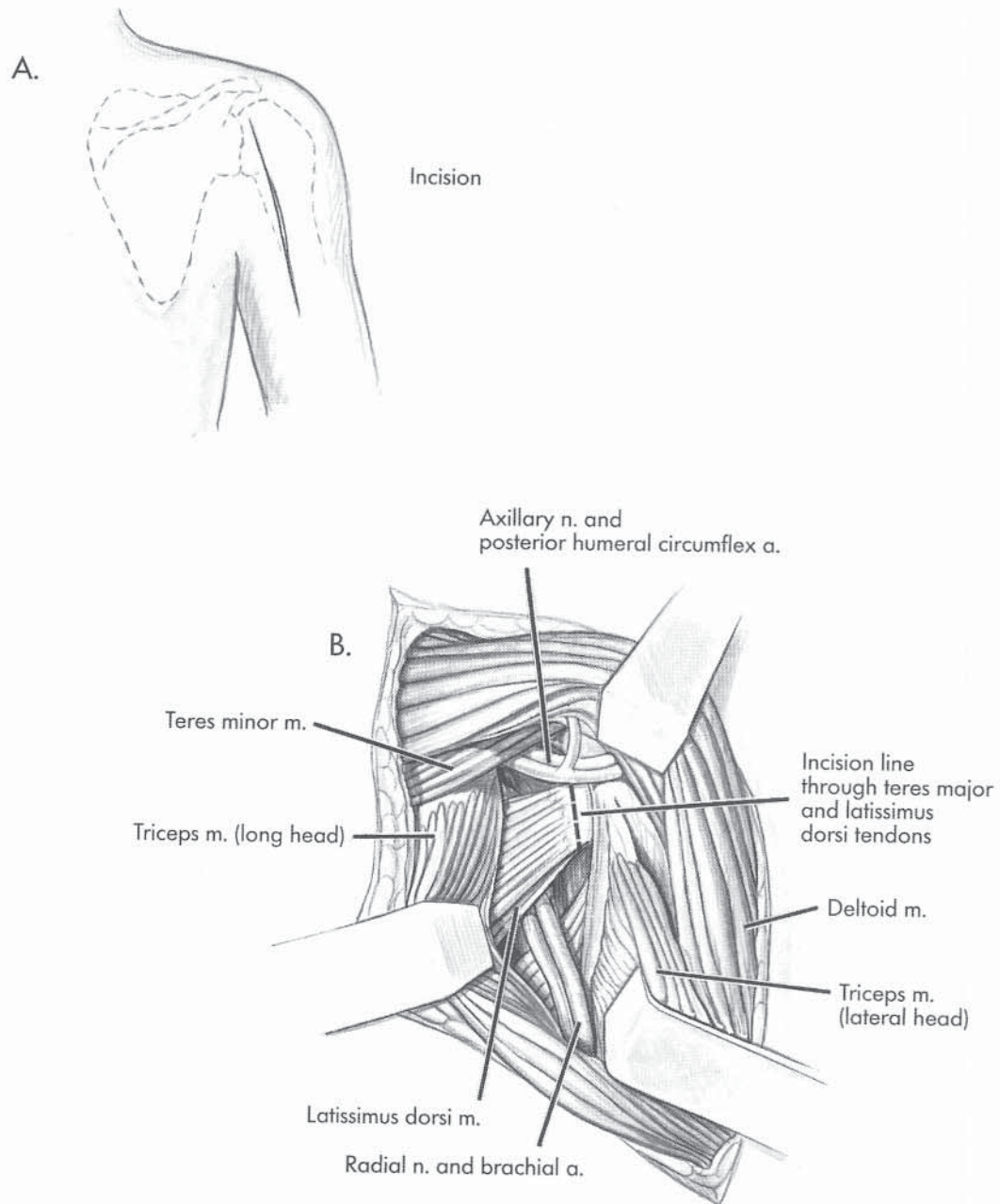
To save anesthesia time in the operating room, it is desirable to manufacture a bivalved shoulder spica cast with the shoulder and upper limb in appropriate position for postoperative immobilization. This is relatively simple in the cooperative patient.

The patient is positioned in a side-lying lateral posture, and the paralyzed upper limb, shoulder, and neck are prepared and draped. The upper limb should be draped free, and the sterile area of preparation should extend to the midline anteriorly and posteriorly. If there is persistent adduction-medial rotation deformity of the shoulder, the pectoralis major is sectioned at its insertion through a short anterior axillary incision. This author prefers to lengthen and not section the pectoralis major because the cosmetic appearance is much more pleasing and it saves power of medial rotation of the shoulder.

A, The arm is adducted across the chest, and a 7- to 8-cm-long incision is made over the deltoid-triceps interval. The incision should be proximal enough to expose the rotator cuff. The deltoid muscle is retracted anteriorly and the long head of the triceps is retracted posteriorly. The surgeon must avoid injury to the radial and axillary nerves and posterior circumflex humeral vessels.

B, Next, the tendinous insertions of the latissimus dorsi and teres major are identified, sectioned at their insertions, and passed posterior to the long head of the triceps.

PLATE 14-1. Latissimus Dorsi and Teres Major Transfer to the Rotator Cuff



Latissimus Dorsi and Teres Major Transfer to the Rotator Cuff

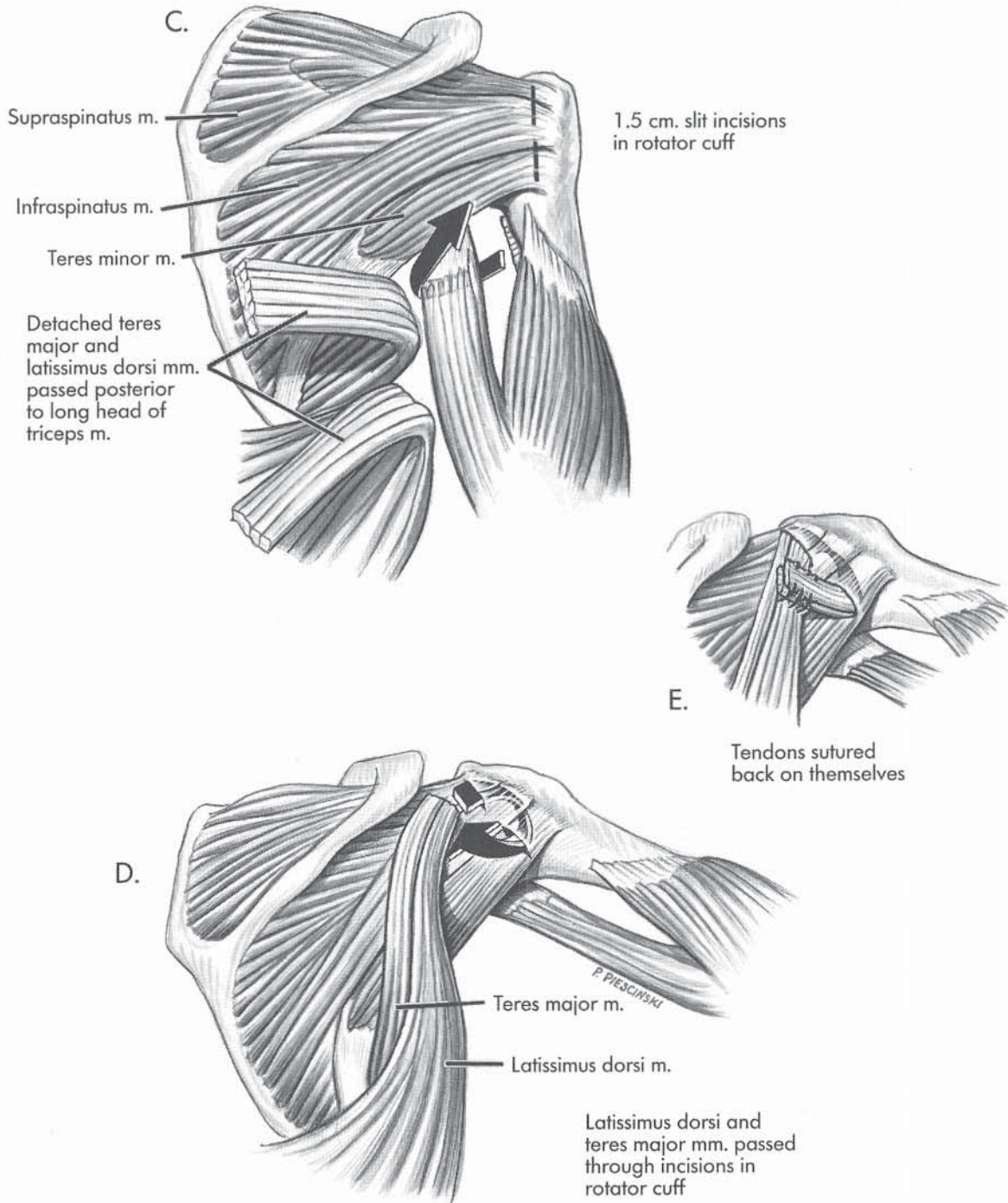
Continued

C to E, The interval between the posterior border of the deltoid and rotator cuff is then developed by blunt dissection. The shoulder is maximally abducted and laterally rotated, and the tendons of the latissimus dorsi with or without teres major are passed through two incisions in the rotator cuff and sutured to itself. The transferred latissimus dorsi and teres major function as lateral rotators instead of medial rotators of the shoulder. The wounds are closed in routine fashion. The preoperatively manufactured bivalved shoulder spica cast is applied for immobilization.

POSTOPERATIVE CARE

The spica cast is bivalved 4 weeks postoperatively and the tendon transfer is trained as a lateral rotator. Shoulder abduction and medial rotation exercises are performed to mobilize the elbow, forearm, and wrist. In between exercise periods the shoulder is maintained in abduction-lateral rotation until the transferred muscles are fair or better in motor strength. Thereafter, new shoulder splints are made, gradually adducting the shoulder. The shoulder splint is worn at night for a period of 6 months.

PLATE 14-1. Latissimus Dorsi and Teres Major Transfer to the Rotator Cuff



Lateral Rotation Osteotomy of the Humerus

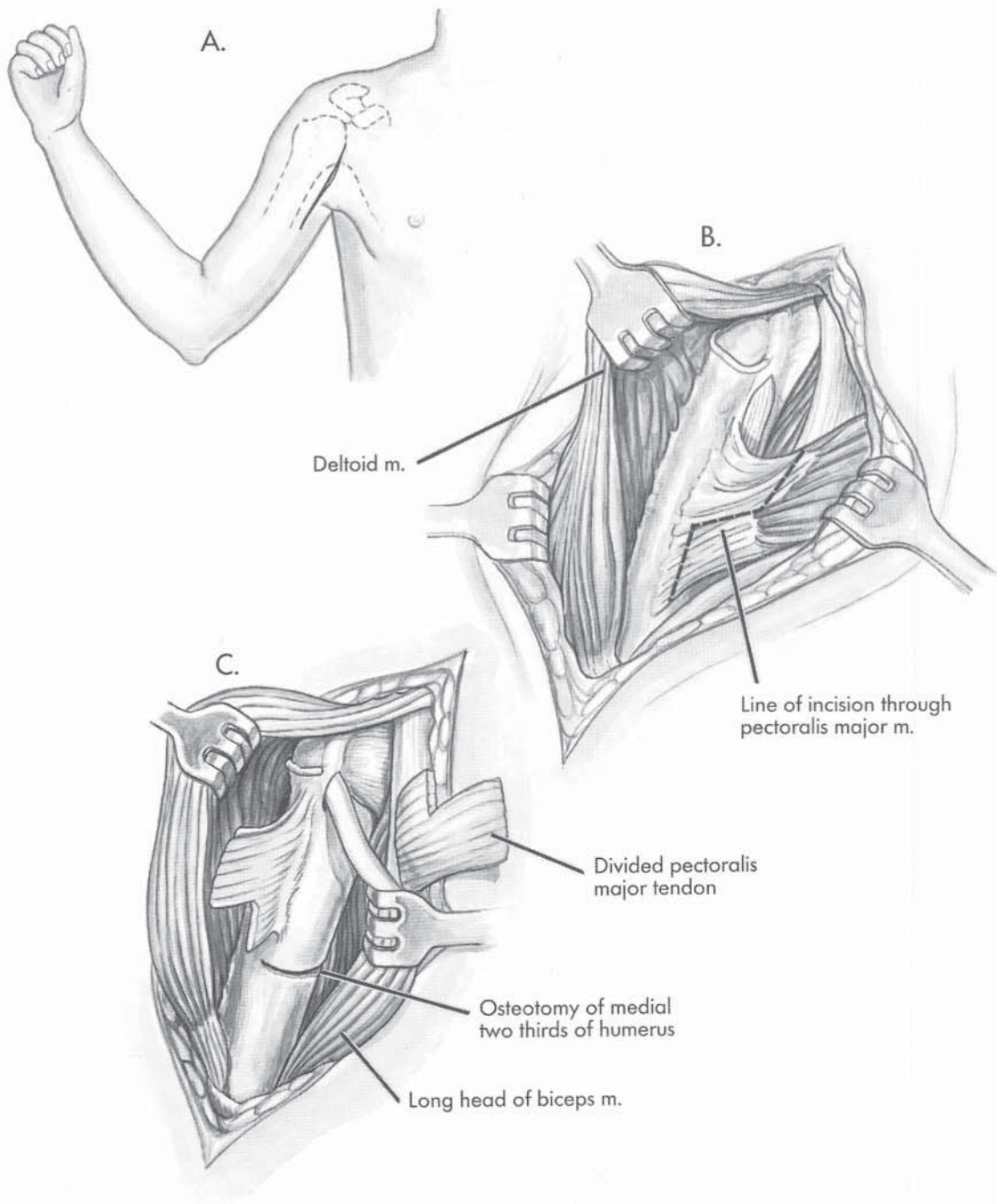
OPERATIVE TECHNIQUE

A, The skin incision begins at the coracoid process, extends to the middle of the axilla, and then curves distally on the medial aspect of the arm, terminating at its upper third. Surgical exposure of the proximal humerus by this axillary approach results in minimal visibility of the operative site.

B, The lateral skin margin is retracted laterally. With the shoulder in medial rotation, the upper humeral shaft is exposed. The surgeon must avoid injury to the cephalic vein and anterior humeral circumflex vessels. The proximal humeral physis should not be disturbed.

C, The level of osteotomy is distal to the insertion of the pectoralis major and proximal to the insertion of the deltoid.

PLATE 14-2. Lateral Rotation Osteotomy of the Humerus



Lateral Rotation Osteotomy of the Humerus *Continued*

D to F, This increases range of shoulder abduction and also facilitates exposure of the humeral shaft. This author recommends internal fixation with a four- or five-hole plate. First the surgeon performs an incomplete osteotomy of the humeral diaphysis three-fourths of the way through the anteromedial aspect. Second, the upper humeral segment is fixed to the plate with two screws (D). Then the osteotomy is completed with an electric saw, and the arm is rotated laterally to the desired degree and temporarily fixed with a bone-holding forceps (E). Next, passive range of shoulder rotation is tested. The ideal position of the shoulder is complete lateral rotation of the shoulder in 90 degrees of abduction. Then, with the shoulder in adduction, the hand should touch the anterior abdomen without elevating the scapula. The surgeon should avoid the pitfall of overcorrection, as it will produce lateral rotation-abduction contracture of the shoulder. Finally, once the desired degree of lateral rotation is obtained, internal fixation of the osteotomy is completed by insertion of the distal two or three screws (F). The wound is closed as usual. The shoulder is immobilized in a shoulder spica cast. To save operating room time, the shoulder spica cast may be manufactured preoperatively, bivalved, and fitted at the completion of surgery.

POSTOPERATIVE CARE

Six weeks after surgery the cast is removed and range-of-motion exercises of the shoulder and elbow are performed.

The results of lateral rotation osteotomy of the humerus are very satisfactory; the improved rotational posture of the shoulder increases range of shoulder abduction.

PLATE 14-2. Lateral Rotation Osteotomy of the Humerus

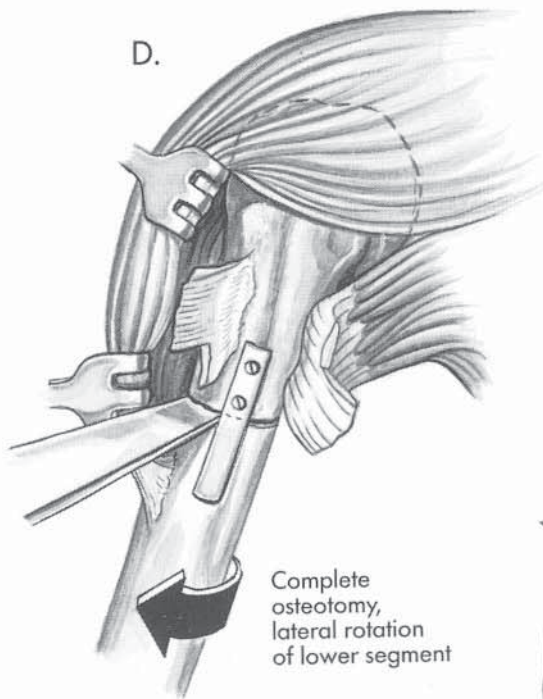
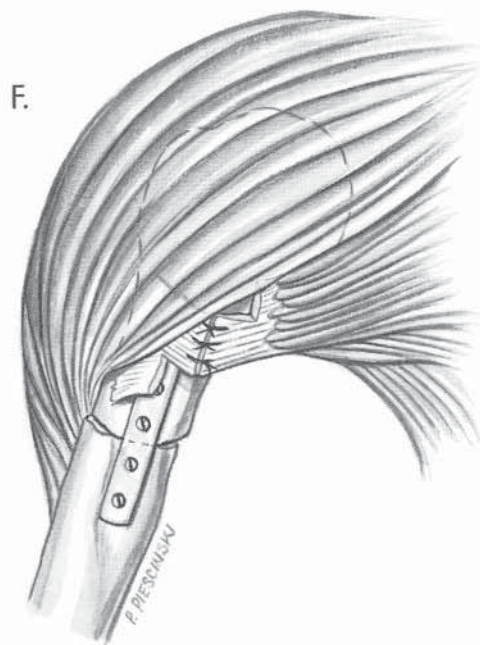
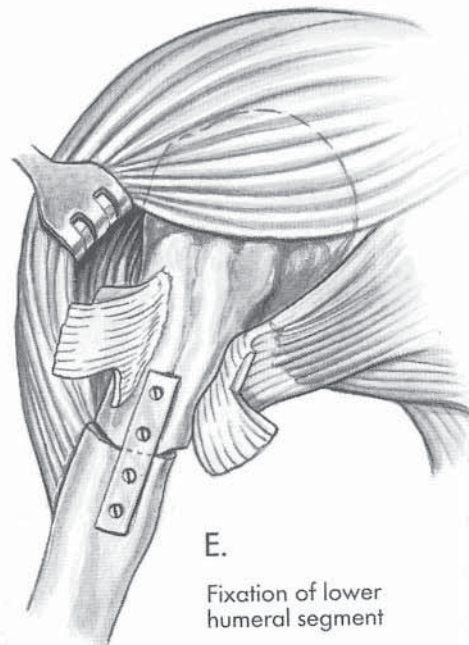


Plate and screws fixing upper humeral segment



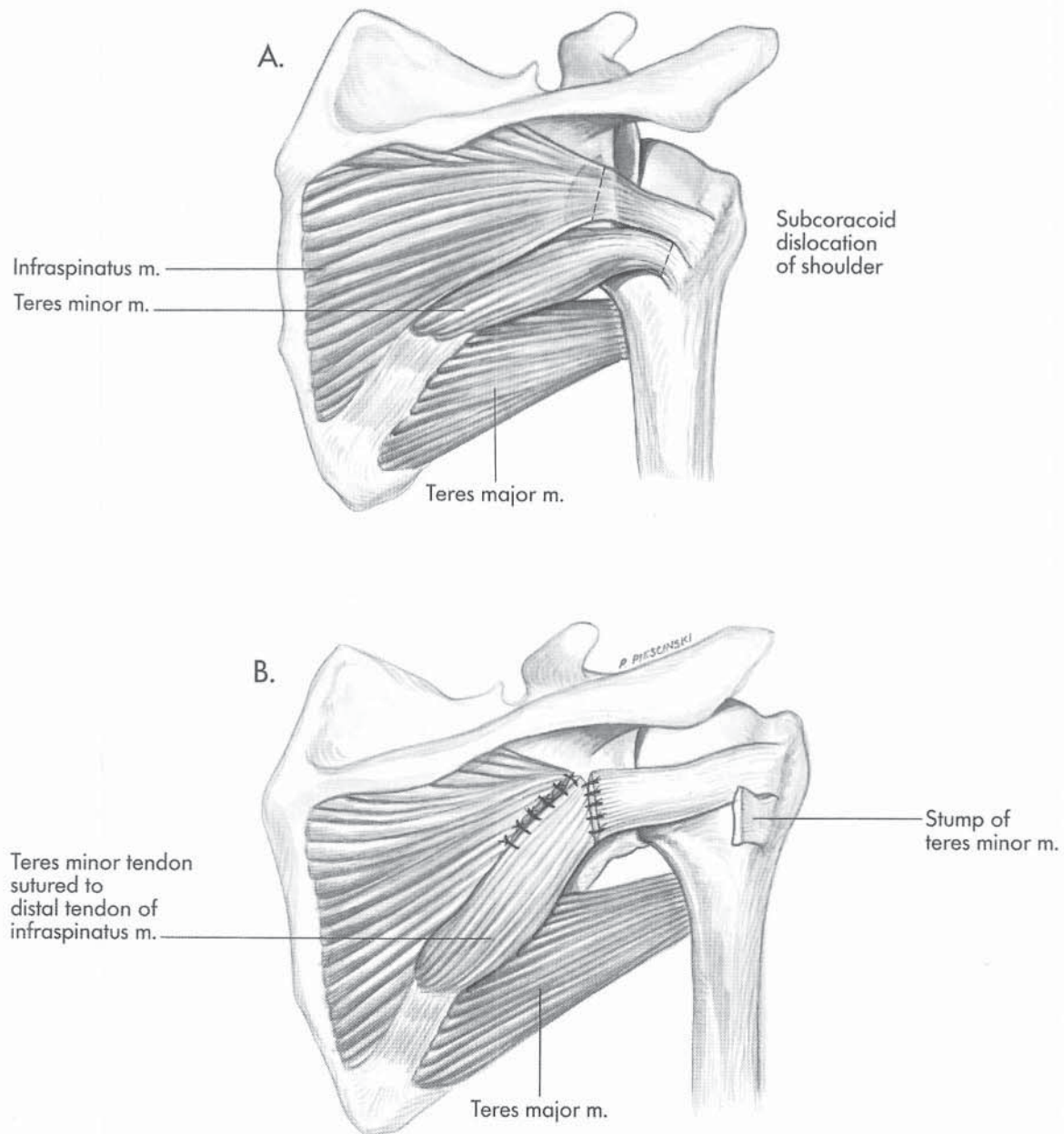


FIGURE 14-145 Lengthening of the lateral rotators of the shoulder. **A**, Lines of division of the infraspinatus and teres minor muscles near their insertion. **B**, The teres minor tendon is sutured to the distal tendon of the infraspinatus muscle and the proximal part of the infraspinatus is sutured to the teres minor.

birth, rapidity of growth, history of trauma, family history, and the child's general health. A useful approach to any lesion is first to determine the tissue involved. Careful examination should reveal whether the mass is bony or soft tissue, fixed or mobile with respect to adjacent tissue, and discrete or ill-defined. Any relationship to neurovascular structures should be noted. Old scars over or near the mass may indicate a retained foreign body. A mass that moves with a tendon may be synovial in origin. Multiple lesions may indicate a systemic condition such as osteochondromatosis.

IMAGING STUDIES

Imaging studies should include plain radiography of the affected part, with the exposure windowed to either bone

or soft tissue, as appropriate. Other imaging studies are selected based on specific questions to be answered. The evaluation of a primarily bony lesion is often best done by CT; soft tissue lesions are best studied with MRI.

NON-NEOPLASTIC MASSES

Many non-neoplastic masses can occur in the hands of children.

Synovial Cysts (Ganglions, Retinacular Cysts). Synovial cysts are the most common. These are benign, fluid-filled cysts that arise from supporting tissues around joints or synovial-lined spaces. Common locations for ganglion cysts are the dorsum of the wrist, the radioulnar wrist, the A-1

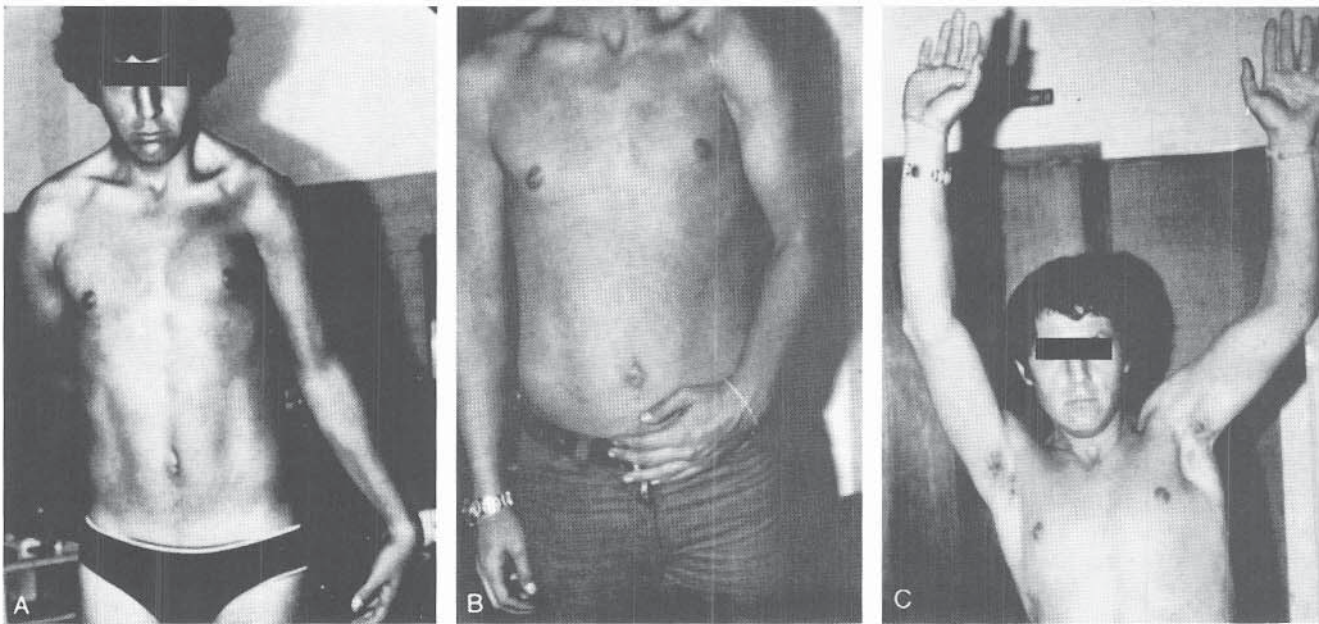


FIGURE 14-146 Abduction-lateral rotation deformity of the shoulder in brachial plexus paralysis in an 18-year-old man. **A**, Preoperative appearance of 18-year-old with marked abduction-lateral rotation contracture. Scapular elevation was severe, and he was unable to touch the anterior aspect of his body with his hand. **B**, Postoperative appearance after medial rotation osteotomy of the humerus. The patient regained medial rotation of the upper limb. **C**, Elevation of the shoulder was maintained. (From Zancolli EA: Classification and management of the shoulder in birth palsy. *Orthop Clin North Am* 1981;12:433.)

pulley in the digital flexor sheath, and the first dorsal compartment. The lesions are firm, single- or multiloculated, and fixed to underlying structures, as they arise from deep synovial-lined tissue. Transillumination with a penlight or otoscope is the easiest way to establish the diagnosis. If doubt exists, ultrasonography can define these masses as fluid-filled. Any solid mass around the wrist must be treated as an unknown and considered for biopsy.

The treatment for ganglion cysts, synovial cysts, or reticular cysts is usually observation. Most will spontaneously regress or rupture if not actively treated. If the cyst is particularly symptomatic, treatment can consist of aspiration, aspiration with injection of corticosteroid, or surgical excision. Even with surgery, the recurrence rate may be as high as 25 percent, and there are risks associated with each active treatment option. Most parents are reassured with the knowledge that the lesion is benign.

Epidermoid Inclusion Cysts. Epidermoid inclusion cysts are benign accumulations of sebaceous material that arise from implanted and still viable dermal and epidermal elements. A history of a prior puncture wound or laceration is sometimes found and confirmed by an overlying scar.

With time, the material becomes loculated and develops into a slow-growing, firm mass affixed to the overlying skin. Rupture of the cyst can cause an intense inflammatory reaction, and the clinical presentation in this scenario resembles that of infection. Treatment is by surgical excision, with care not to spill the contents of the cyst in the surgical field.

Foreign Bodies or Reactions. Foreign bodies or reactions to a foreign body occur relatively commonly in children. Metal fragments from pellet guns, glass fragments, or splinters that become embedded in soft tissue may produce symp-

toms because of the inflammatory response or because of movement of adjacent tissues. Ultrasonography may be useful in localizing the object or establishing the diagnosis of a foreign body surrounded by fluid. Treatment is by surgical removal. Antibiotic prophylaxis is indicated.

Aneurysms and Pseudo-aneurysms. Aneurysms and pseudo-aneurysms are possible complications of trauma to the child's hand. A true aneurysm occurs after blunt trauma and intimal damage to an artery. A pseudo-aneurysm occurs after penetrating trauma with the development of a "false" wall holding the hematoma. Clinically, an enlarging pulsatile mass is seen in both cases. Treatment is surgical excision and vascular reconstruction, if possible, in both cases.

Malformations. Malformations are conditions that arise from errors in embryologic development. These lesions consist of tissues that are histologically normal but occur in a size, quantity, or combination that is not normal. Malformations usually grow in proportion to the normal growth of the child.

Vascular Malformations. Vascular malformations differ from hemangiomas in that there is no early rapid endothelial proliferation and no evidence of later involution, as seen with hemangiomas. Mulliken and colleagues classified vascular malformations as capillary, venous, lymphatic, and combined lesions.⁶ The capillary malformation is marked by a port-wine stain of the skin that is pink in infancy and darkens in adulthood. These malformations are confined to the capillary bed and usually are not symptomatic. Venous lesions are the most common and can be extensive. These are the lesions likely to be intramuscular or deep to fascia. Lymphatic malformations present with lymphedema but no

Text continued on page 502

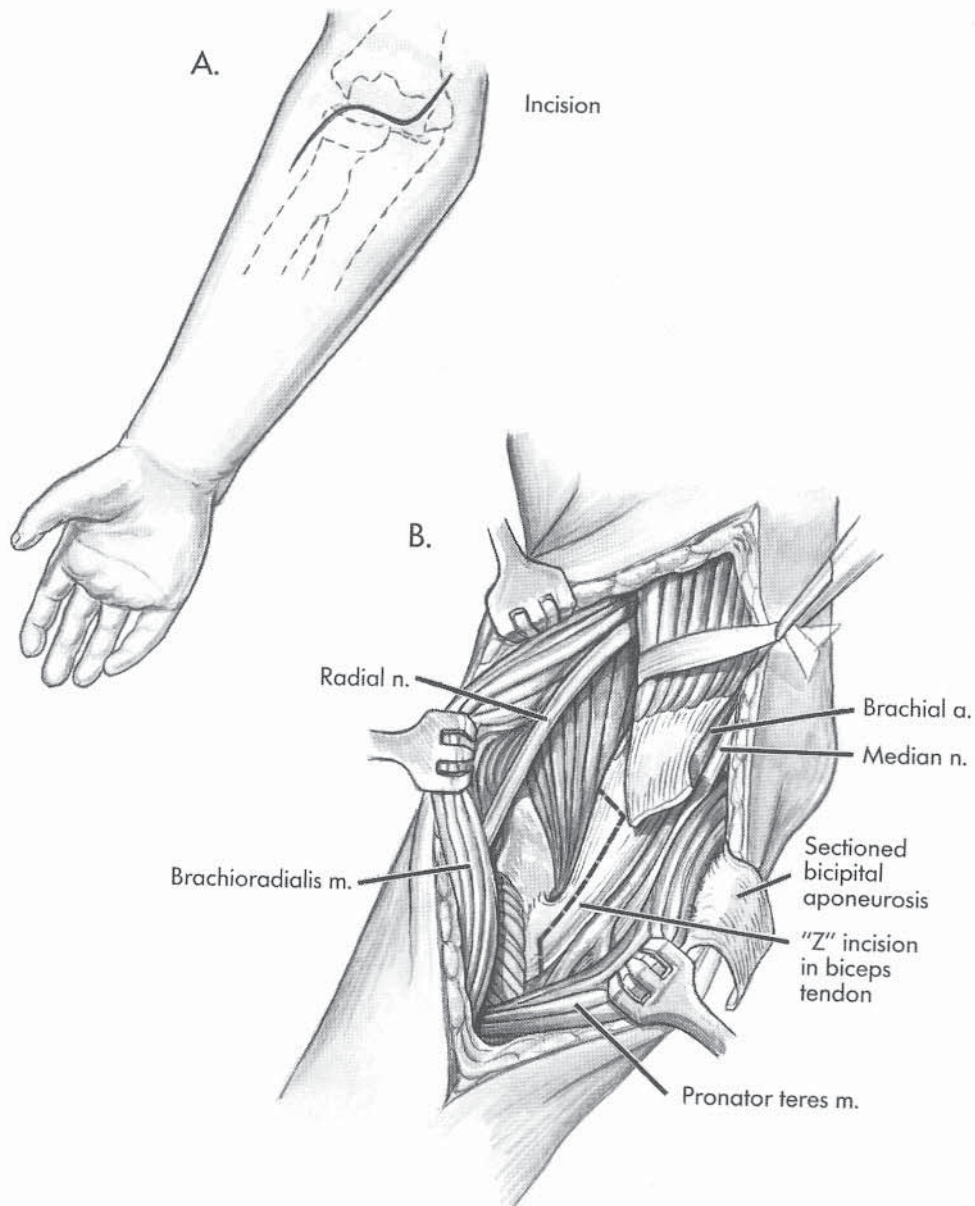
Rerouting of Biceps Brachii Tendon to Convert Its Motion from Supinator to Pronator of the Forearm (Zancolli Procedure)

OPERATIVE TECHNIQUE

A, An S-shaped incision is made on the volar surface of the elbow. The incision is begun 3 to 5 cm above the elbow joint and extended to the antecubital crease, then laterally to the radial head and distally into the forearm for a distance of 5 cm. The subcutaneous tissue and deep fascia are divided in line with the skin incision.

B, The biceps tendon is exposed and traced distally to its insertion to the bicipital tuberosity of the radius. The brachial vessels and median nerve are identified and traced.

PLATE 14-3. Rerouting of Biceps Brachii Tendon to Convert Its Motion from Supinator to Pronator of the Forearm (Zancolli Procedure)



Rerouting of Biceps Brachii Tendon to Convert Its Motion from Supinator to Pronator of the Forearm (Zancolli Procedure)

Continued

C, A long Z-plasty of the biceps tendon is performed.

D, The distal segment of the biceps tendon is rerouted around the neck of the radius, passing it mediolaterally.

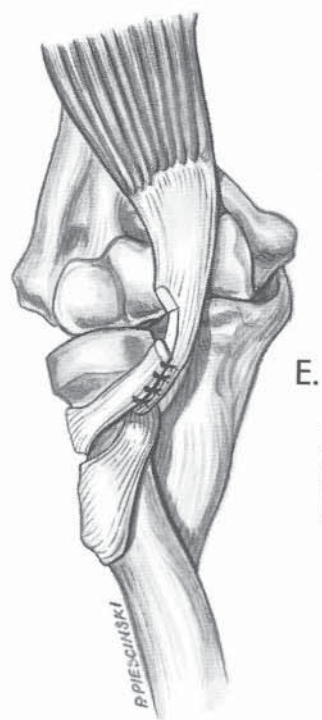
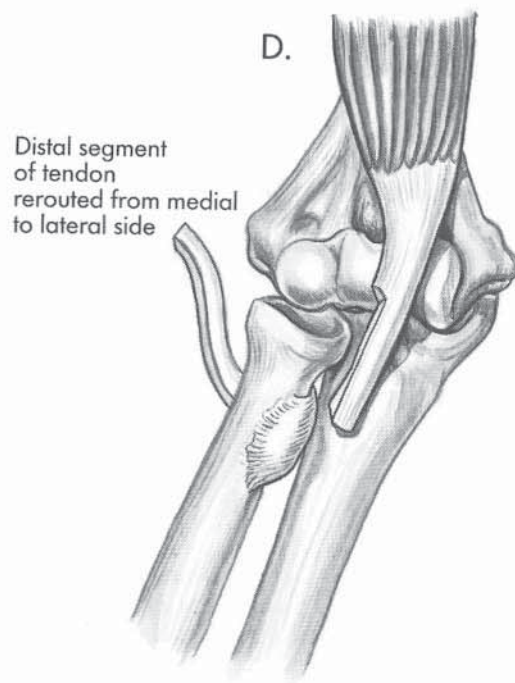
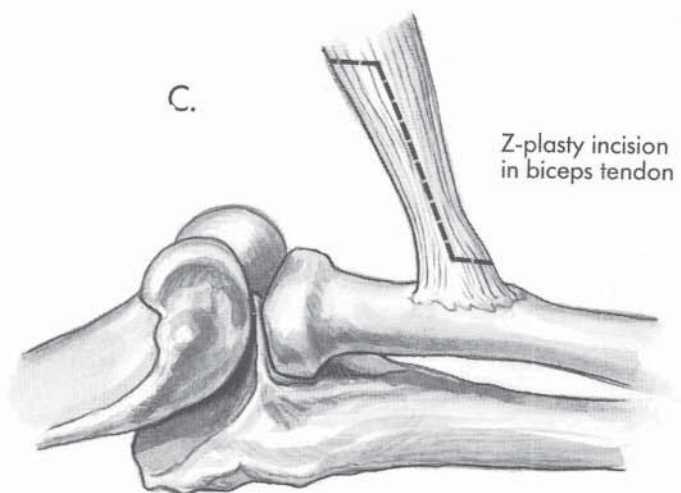
E, The divided biceps tendon segments are resutured side to side at a length that will maintain full pronation of the forearm and extension of the elbow.

The surgeon should avoid excessive tension on the tendon in young children and when the forearm is hyperflexible into pronation. The wounds are closed in routine fashion. An above-elbow cast is applied with the elbow in 30 degrees of flexion and the forearm in full pronation.

POSTOPERATIVE CARE

Four weeks after surgery the cast is removed, and active assisted exercises are performed three to four times a day to develop pronation and supination of the forearm and elbow flexion-extension. Gentle passive exercises are carried out to maintain full pronation and supination of the forearm and complete flexion and extension of the elbow. At night a plastic splint is worn, maintaining the forearm in full pronation and the elbow in 30 degrees of flexion.

PLATE 14-3. Rerouting of Biceps Brachii Tendon to Convert Its Motion from Supinator to Pronator of the Forearm (Zancolli Procedure)



color change. Many lesions are combined, with elements of more than one of these vascular structures.

Imaging of the larger lesions is best accomplished with MRI to define the location and the extent of the malformation. Laser treatment of the capillary lesions may help lighten the discoloration but may produce scarring. Complete excision of a large venous lesion may not be practical, but debulking may be of cosmetic value. Coagulopathy associated with the vascular malformations, or Kassebach-Merritt syndrome, is due to sequestration of platelets within the lesion. This thrombocytopenia is not directly related to the size of the lesion. Lymphatic lesions are often extensive and found in combination with the other vascular components. Hypertrophic scarring is common with the lymphatic lesions.

HAMARTOMA OF THE NERVE. Hamartoma of the nerve is a type of malformation. Presentation is often with enlargement within the nerve territory, or macrodactyly. Another presentation is with compression neuropathy, particularly of the median nerve in the carpal canal. The diagnosis is usually clear from the presence of enlarged digits and/or thickening and fullness over the course of the involved nerve. Tinel's and Phalen's signs may be positive. Treatment is by decompression of the nerve and debulking or epiphysiodesis of the digit.

NEOPLASMS

Soft tissue neoplasms may involve any soft tissue component of the limb. Soft tissue neoplasms can be divided into benign lesions, aggressive but benign lesions, and malignant lesions. Although the likelihood of a malignant lesion is small, attention to proper technique during biopsy is important so that tissue planes are not contaminated in case the lesion should prove to be malignant.

Benign Lesions

HEMANGIOMAS. Hemangiomas are benign vascular neoplasms that are usually not present at birth but appear shortly afterward, proliferate rapidly, and then spontaneously involute. Hemangiomas are differentiated from vascular malformations by the presence of endothelial proliferation and growth out of proportion to the rest of the child's growth. Treatment for these benign lesions is expectancy, as involution can be expected during the first decade.

GIANT CELL TUMOR OF THE TENDON SHEATH. Giant cell tumor of the tendon sheath (localized nodular tenosynovitis, xanthoma of the synovium) is not a common tumor in childhood but can occur. The lesion is histologically identical to a pigmented villonodular synovitis. It is found in close association with joints or a tendon sheath and is thought to arise from a synovial source. Large lesions may cause symptoms by limiting joint excursion or by compressing neurovascular structures. Treatment is excision of the entire lesion, including any part of the lesion that has extended along soft tissue planes. Recurrence should be prevented by complete excision.

GLOMUS TUMORS. Glomus tumors are benign lesions of vascular origin. The normal glomus structure is a histologically recognizable bundle of arterioles connecting directly with

veins and possessing an abundant nerve supply. The glomus body is normally found in the reticular layer of the dermis. Its role is to regulate blood flow in the dermis. The typical location for a glomus tumor is under the fingernail, although they may be found in other locations. Glomus tumors are characterized by marked hypersensitivity to cold, severe pain, and tenderness. MRI can be used to localize the lesion, but the clinical findings are usually diagnostic. Excision of the lesion is curative. The surgical technique can be by removal of the nail plate or through a lateral approach.

PYOGENIC GRANULOMA. Pyogenic granuloma is a benign lesion that usually occurs following trauma, especially on the glabrous skin surfaces. It may take the form of a capillary hemangioma. It appears as hypertrophic granulation tissue that is red, rapidly growing, and very friable. The characteristic appearance is diagnostic. Treatment for small lesions is by cauterizing with silver nitrate; larger lesions are better treated with surgical resection and either direct closure of the skin edges or application of a split-thickness skin graft.

Fibrous Tumors. Fibrous tumors of the hand in children include the juvenile aponeurotic fibroma (calcifying aponeurotic fibroma), the infantile digital fibroma (digital fibroma of childhood, or recurring digital fibroma), fibromas, and neurofibromas. Each has a characteristic clinical presentation, location, and prognosis. Aponeurotic fibromas and the digital fibromas tend to be locally recurrent but are still benign.

APONEUROTIC FIBROMAS. Aponeurotic fibromas are unusual lesions that involve the palmar aponeurosis. They tend to occur in preteens or teenagers rather than in young children. The lesions are ill-defined, involve skin, and extend along the aponeurotic connections of the palm. Treatment is total excision with a margin of normal skin. Recurrence is related to subtotal excision.

INFANTILE DIGITAL FIBROMA. Infantile digital fibromas occur in the young child. The lesions are often multiple and present as hard, enlarging masses on the fingers. Reports of intracytoplasmic inclusion bodies on histologic examination of these lesions raises the possibility of an as yet unidentified causative infectious agent. Treatment is excision with a margin of normal tissue and closure with local flaps or skin grafts. Recurrence is common.

SIMPLE FIBROMAS. Simple fibromas are unusual but may occur. These well-defined lesions present as hard masses, usually on the dorsum of the hand. They are not densely adherent or invasive. Treatment is by excision. An unusual type of fibroma is seen at the periungual region in children with tuberous sclerosis.

NEUROFIBROMAS. Neurofibromas are seen in association with neurofibromatosis, often as multiple lesions. Treatment is reserved for lesions that are large enough to cause symptoms.

LIPOMAS. Lipomas are rare in children. A soft tissue tumor that does not transilluminate must be further evaluated to establish a diagnosis. Fat density on plain radiographs or signal characteristics on MRI are diagnostic. Surgical excision is the treatment for lesions that are bothersome.

Malignant Soft Tissue. Solid lesions must be suspected of being the rare malignancy, even in childhood. Appropriate

evaluation, including examination of regional lymph nodes, imaging studies of the region and the chest, and blood work, must be coordinated with the multidisciplinary pediatric oncology specialist. The surgical emphasis is on accurate diagnosis and preserving the best functional outcome—life first, then limb.

MALIGNANT SCHWANNOMAS AND NERVE SHEATH TUMORS. Malignant schwannomas and nerve sheath tumors may occur in children and should be considered in the differential diagnosis of a solid lesion in the region of a peripheral nerve. Not all masses at the wrist are ganglions.

SUBUNGUAL MELANOMA. Subungual melanomas may be seen in children and adolescents as a malignant degeneration of a preexisting congenital nevus. Deeply pigmented lesions of the subungual and glabrous skin should be closely watched for any change indicating malignant transformation. Treatment depends on the location, and the procedure must obtain acceptable margins.

OTHER MALIGNANT TUMORS. Malignant fibrous histiocytomas, fibrosarcomas, synovial sarcomas, and other malignant tumors are rare in the child's hand but can occur. A treatment protocol exists for each tumor type, and they are best managed by the oncology team.

Benign Bone Tumors. Benign skeletal tumors are common in childhood and adolescence. Many occur as part of generalized skeletal developmental abnormalities. Aberrations in enchondral osseous development present as bony lesions in enchondromatosis and osteochondromatosis. More localized lesions are seen in dysplasia epiphysealis hemimelica or Trevor's disease.

SOLITARY ENCHONDROMA. A solitary enchondroma is usually identified after pathologic fracture or as an incidental finding. Usual locations are in the metacarpals or proximal phalanges, where the enchondroma is central and does not produce irregularities on the exterior of the cortex. Treatment is facilitated if the fracture is allowed to heal before the lesion is treated by curettage and bone grafting.

ENCHONDROMATOSIS (OLLIER'S DISEASE). Endochondromatosis, or Ollier's disease, is a more widespread defect in enchondral ossification. Multiple lesions are usually present in the small tubular bones of the hands. These cartilaginous tumors produce jagged irregularities in the external contours of the bones. The chondral lesions are firm and often expansive. The risk of malignant degeneration is present but rare in children, particularly in the hand. In Maffucci's syndrome (enchondromatosis associated with multiple hemangiomas), the risk of malignancy, chondrosarcoma, or nonsarcomatous malignancy is much greater. Treatment entails excision and curettage of the symptomatic lesions and bone grafting.

OSTEOCHONDROMATOSIS. Osteochondromatosis involves the hand and forearm in several typical sites. Lesions at the distal end of the forearm cause enlargement and tendon irritation. Disruption of longitudinal growth of the distal ulna results in a typical ulnar deviation and shortening of the ulnar side of the forearm. Lesions in the small bones in the hand are often small, but the subungual lesions disrupt nail formation and cause noticeable deformity. Treatment addresses the symptomatic lesions with excision. Correction

of the ulnar deviating deformity may require excision of the lesion and osteotomy, distraction, or resection of the distal ulna.

Detailed information on the diagnosis and management of benign and malignant upper limb masses in children can be found in the references.¹⁻⁹

REFERENCES

Tumors of the Upper Limb

1. Arms DM, Strecker WB, Manske PR, et al: Management of forearm deformity in multiple hereditary osteochondromatosis. *J Pediatr Orthop* 1997;17:450.
2. Cawte TG, Steiner GC, Beltran J, et al: Chondrosarcoma of the short tubular bones of the hands and feet. *Skeletal Radiol* 1998;27:625.
3. Colon F, Upton J: Pediatric hand tumors: a review of 349 cases. *Hand Clin* 1995;11:223.
4. Krajca-Radcliffe JB, Thomas JR, Nicholas RW: Giant-cell tumor of bone: a rare entity in the hands of children. *J Pediatr Orthop* 1994;14:776.
5. Mota CR, Marques L, Silva E, et al: Symmetrical enchondromatosis of the hands and feet in two sisters. *J Pediatr Orthop B* 1997;6:15.
6. Mulliken JB, Zetter BR, Folkman J: In vitro characteristics of endothelium from hemangiomas and vascular malformations. *Surgery* 1982; 92:348.
7. Peterson HA: Deformities and problems of the forearm in children with multiple hereditary osteochondromata. *J Pediatr Orthop* 1994;14:92.
8. Yaw KM: Pediatric bone tumors. *Semin Surg Oncol* 1999;16:173.
9. Yin Y, Gilula LA, Kyriakos M, et al: Giant-cell tumor of the distal phalanx of the hand in a child. *Clin Orthop* 1995;310:200.

Microsurgery

REPLANTATION (ATTACHMENT OF SEVERED PARTS)

Advances in microsurgery have allowed viable attachment of some amputated extremity parts. However, true measures of success of attachment of these severed extremities must include subsequent useful function of the part. Achieving useful function depends on surgical expertise, proper preoperative care, and appropriate indications for attachment of the amputated part.

Preoperative Care. In the emergency room, proper care of the amputated part(s) is critical. The part(s) should be wrapped in a saline-soaked gauze, then placed in a bag on ice. This step is critical, since it converts warm ischemia to cold ischemia time. Amputated digits can tolerate warm ischemia times of less than 12 hours, while a properly stored digit can tolerate cold ischemia times at least twice this long. In more proximal amputations with substantial muscle tissue, warm ischemia times should not exceed 6 hours.

Indications. The type of amputation best suited for replantation is a sharp, clean injury, and those least favorable are avulsion-type and dirty wound amputations. The site of amputation is also an important consideration. Replantation should be considered for thumb amputations, multiple digit amputations, partial hand amputations, and wrist or forearm amputations, because successful replantation usually results in a quality of function better than that afforded by amputation at these levels. An attempt should be made to replant amputated thumbs if technically possible, especially when the level of amputation is at or proximal to the proximal phalanx. When multiple digits

have been amputated, the more important digits are attached first. If the thumb is not suitable for replantation, an amputated index or middle finger may be placed in the thumb position. Success rates for replantation should be greater than 80 percent.

Controversy exists regarding the relative indications for and contraindications to replantation of amputated parts. Contraindications to replantation include severely crushed parts, multiple-level amputations, and prolonged warm ischemia times. In addition, patients with other serious injuries or disease should not be subjected to the prolonged operative times required for replantation. Mentally deficient or unstable patients are also poor candidates for replantation.

Relative contraindications are controversial but include reattachment of single-digit amputations, especially in zone II (the area between the distal palmar crease and the insertion of the superficialis tendon). Digits severed at this level have limited motion and are often bypassed in favor of other normal digits. Amputations that are proximal to the elbow have a less favorable risk-benefit ratio because of the high incidence of muscle necrosis and infection following reattachment. We therefore recommend that replantation be performed only for clean, sharp amputations proximal to the elbow with short preoperative ischemia times.

REVASCULARIZATION

Revascularization patients represent a special subset of amputation patients. Revascularization is defined as repair of a part that has been incompletely amputated. In these patients it is important not to sever the intact skin bridge, as this often provides important venous outflow for the revascularized part. Restorative procedures in these patients generally have a slightly higher success rate owing to the patent venous outflow tract.

FREE TISSUE TRANSFER

Free tissue transfer has made possible many advances in the treatment of both congenital and traumatic soft tissue deformities in children. In the past, surgeons were reluctant to use microsurgical procedures in the pediatric patient. The conventional wisdom was that the small size and severe spasm of a child's vessels led to increased failure rates. More recently, however, several authors have reported success rates approaching or exceeding those in adult patients.⁴

Inherent Advantages of Free Tissue Transfer. When large soft tissue defects are present or when padded skin is required to protect nerves, joints, or gliding tendons, reconstruction options are limited. Historically, the pedicle flap has been the primary tool for coverage of these types of defects, with the workhorse being the groin flap. Although the advantage of a shorter operating time cannot be ignored, pedicle flap coverage has important inherent disadvantages, particularly in the pediatric population. Restraint and understanding in children are either limited or nonexistent, and many children cannot be counted on to protect the surgeon's work. Even more significantly, the pedicle flap

must survive in a parasitic manner, on the ingrowth of the blood supply available at the recipient site. Such blood supply at the site of injury is often tenuous or nonexistent because the patient needing this type of coverage usually has a severely traumatized extremity with widespread devitalization of tissues. Frequently the recipient site requiring coverage is complicated by current or previous infection.

A free tissue transfer brings the blood supply in with the new tissue—a feat no pedicle flap can sustain after its pedicle is released. This important quality of the microvascular free flap potentially affords an improved milieu for treating infection as well as diminishing fibrosis during and after healing. In the upper limb, less cicatrix is especially critical when nerves, joints, and gliding tendons are nearby (Fig. 14–147). Even in the very young patient, less than 1 year old, free flap coverage has been successful, even though the anastomosis might have to be constructed in vessels of very small diameter.⁴

FREE MOTORIZED MUSCLE TRANSFER

Microsurgery can also be useful for reconstruction to restore extrinsic digital flexion in the patient with severe Volkmann's ischemia during the second stage of reconstruction. This can be effectively accomplished using a free microneurovascular transfer. The gracilis muscle is particularly suited for this reconstruction because it is thin, is an expendable donor, has a single motor nerve, and has blood vessels of suitable size, even in the young patient. We have used this type of reconstruction in patients less than 2 years old. The artery and vein are anastomosed in the antecubital area and the gracilis motor nerve is sutured to the remaining stump of the anterior interosseous nerve in the proximal forearm. Function of the transfer usually can be appreciated after about 6 months (Fig. 14–148).

FREE TOE TRANSFER

Another important use for microsurgical techniques, toe transfer involves reconstruction of absent digits in children with congenital band syndrome, traumatic amputation, or symbrachydactyly. The transferred toe does not improve the appearance of the hand, and therefore the procedure is used only when increased hand function can reasonably be expected. The procedure is appropriate in children with absent thumbs and multiple affected digits. Children with amputations secondary to congenital band syndrome or traumatic amputation are better candidates for free toe transfer because proximal to the amputation, the underlying tendons and nerves are more predictably normal. In contrast, the hands of patients with symbrachydactyly often have hypoplastic or even absent flexor tendons and nerves.

Selected patients can benefit from microsurgical transfer of one or two toes to improve the grasp of the hand. Lack of full development of flexor tendons in the recipient hand may make the final motion achieved in the transformed digit less than ideal. When a single existing digit has good motion but no post it can oppose, even a stiff digit can increase use of the hand (Fig. 14–149). Harvesting one or



A



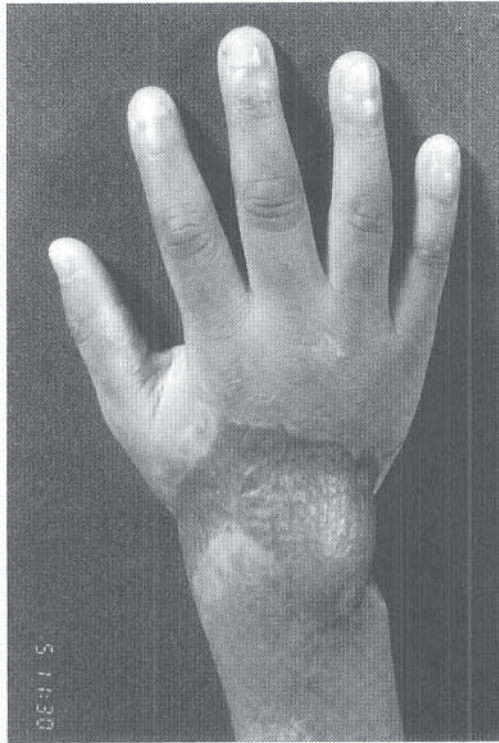
B



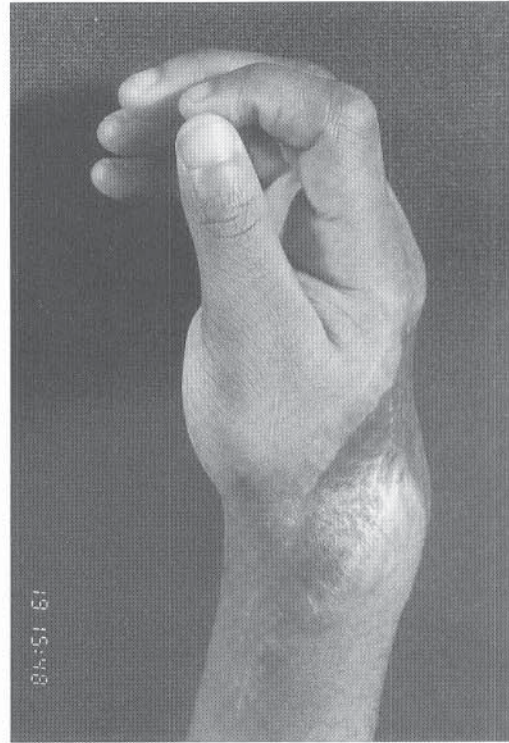
C

FIGURE 14-147 A and B, Severe shotgun wound to the right hand and wrist of a 6-year-old girl. The defect involved skin, soft tissue, tendon, and bone. C, The radiograph showed extensive bony involvement.

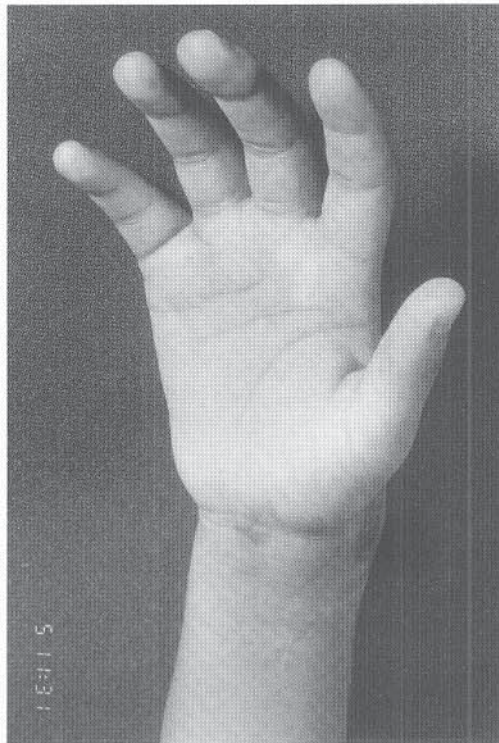
Illustration continued on following page



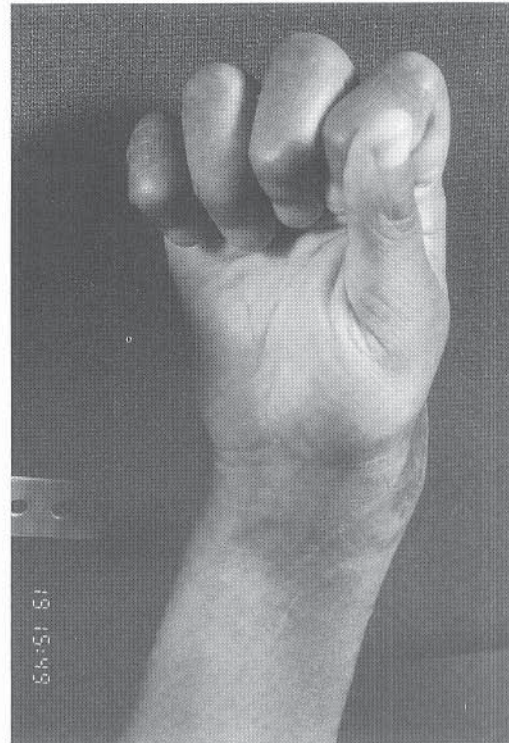
D



E

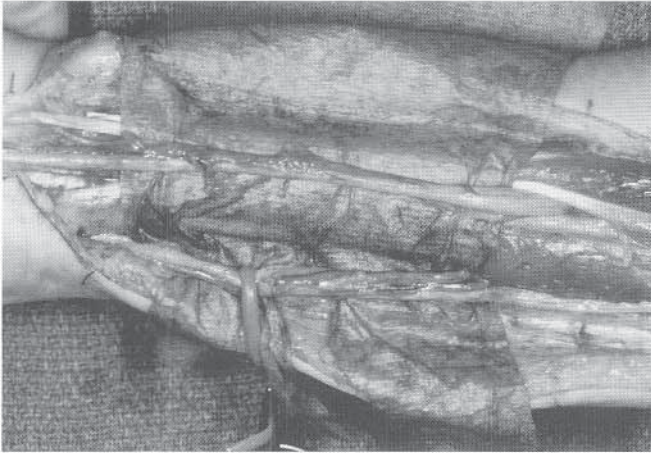


F

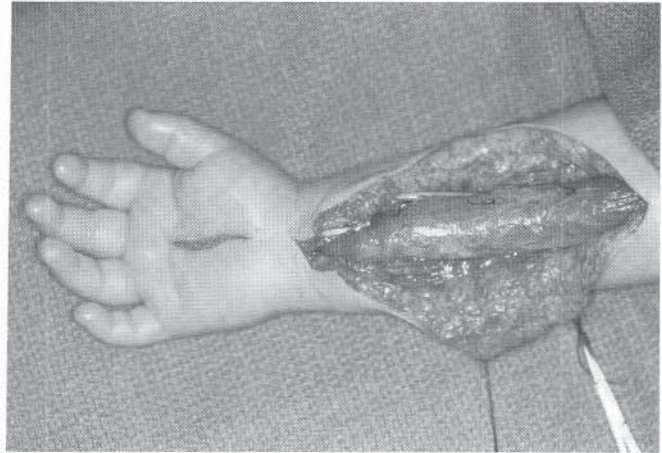


G

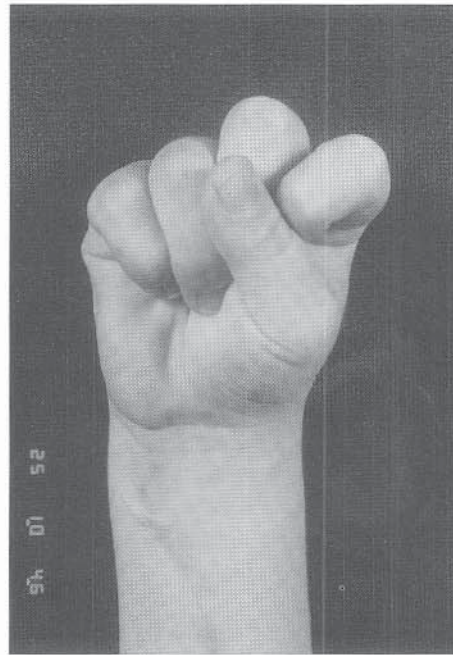
FIGURE 14-147 *Continued.* D to G, Reconstruction of bone, tendon, and soft tissue was required. A free serratus anterior muscle flap with a split-thickness skin graft was used for soft tissue coverage.



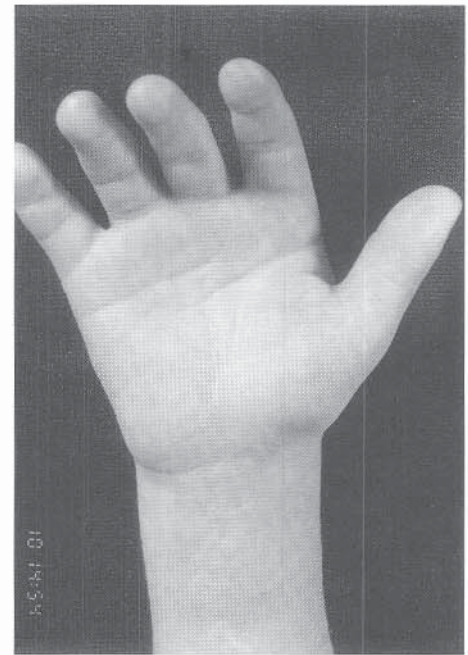
A



B



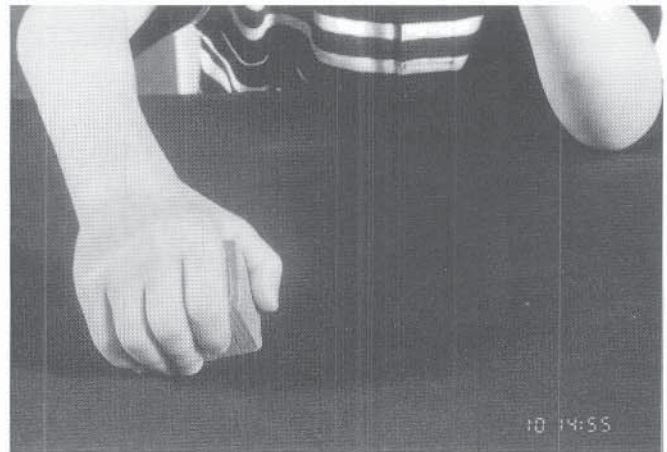
C



D



E



F

FIGURE 14-148 A, An infant boy with severe Volkmann's ischemic contracture and muscle infarct was treated with initial debridement and neurolysis. B, After return of sensation and intrinsic function (which took several months), a second operation to restore extrinsic flexor function was done using a free innervated gracilis transfer. C to F, Postoperative photographs showing the healed wound and function after the second-stage reconstruction.

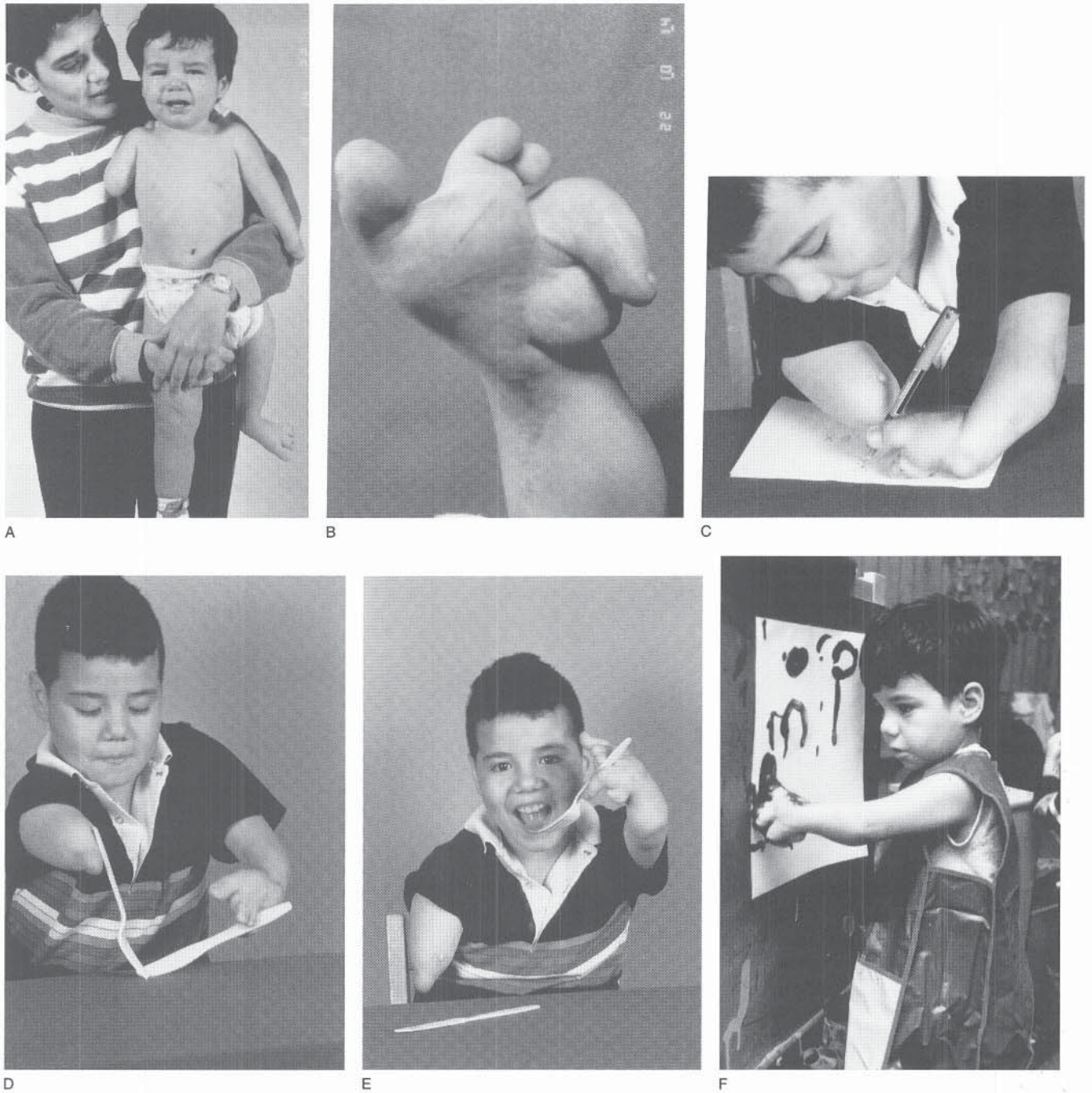


FIGURE 14-149 A, A child with severe proximal femoral focal deficiency. Amputation and transfer of a portion of the foot was required for prosthetic fitting. B, The medial three toes were transferred to the monodactylous and only hand. C to F, The hand is used by this patient.

two toes from the foot is well tolerated and alters mechanics in the lower extremity only slightly.^{1,2,6}

FREE VASCULARIZED BONE TRANSFERS

Pseudarthrosis of the forearm and leg has been successfully treated by microsurgical transfer of vascularized bone to

the area of pseudarthrosis (Fig. 14-150). The ability to transfer relatively large and vascularized bone segments makes a more complete excision of the abnormal tissue at the site of the pseudarthrosis possible. Free vascularized fibular grafts have a place in the treatment of noncongenital nonunions with severe soft tissue injury. Donor site morbidity has been acceptable when care is taken in harvesting the fibula correctly. Injury to the peroneal nerve in the

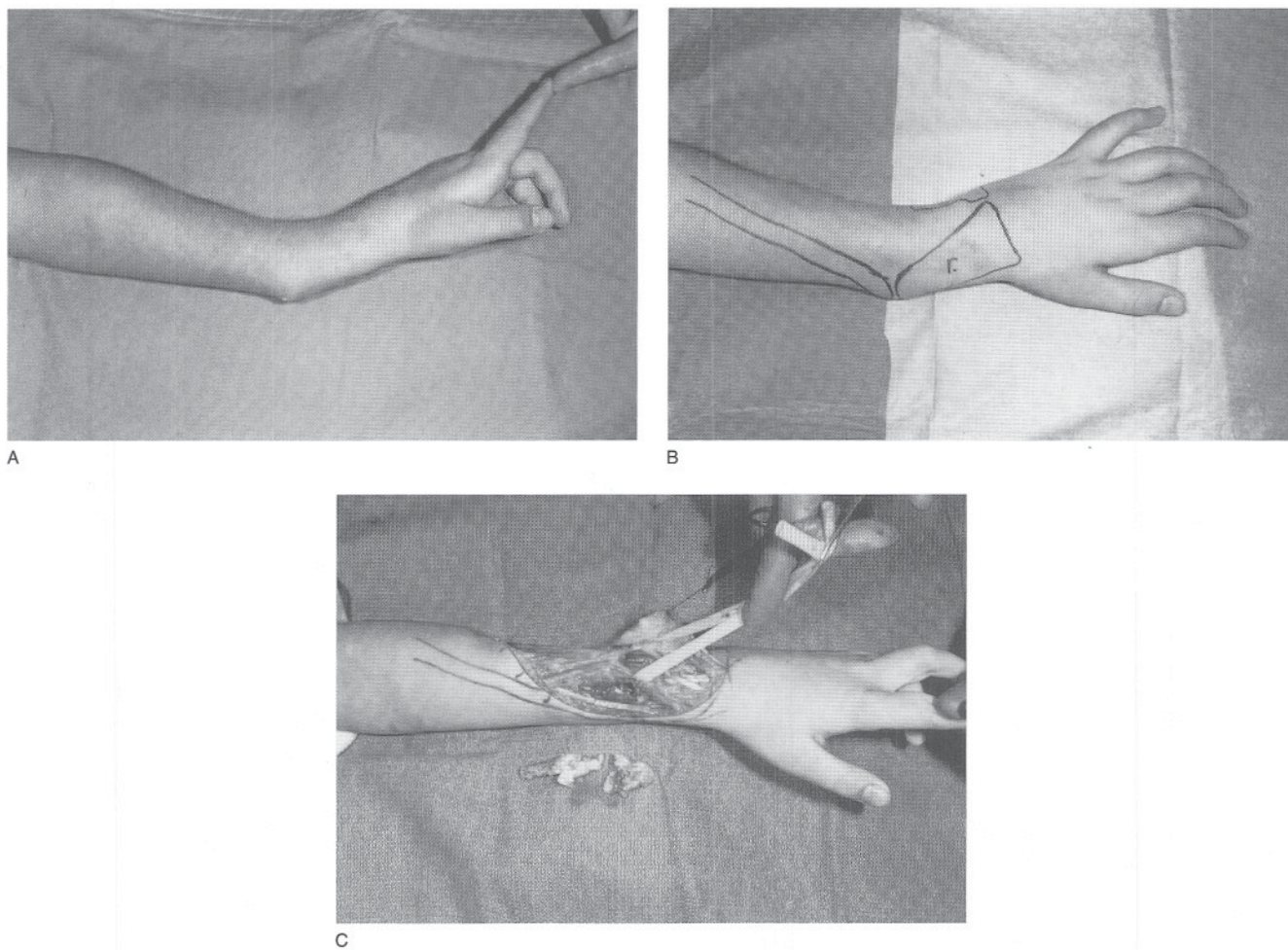
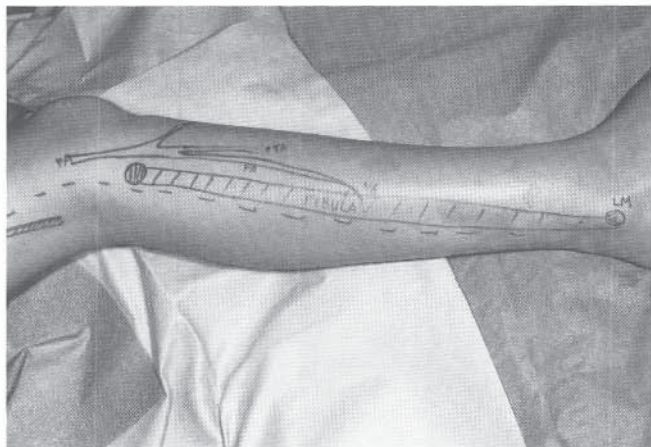
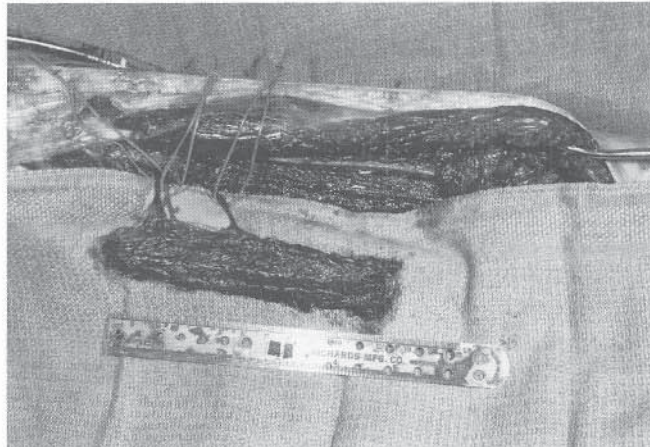


FIGURE 14-150 A and B, Congenital pseudarthrosis of the forearm. C, Because of the free fibular graft, a radical excision of the diseased area of the radius and ulna was possible.

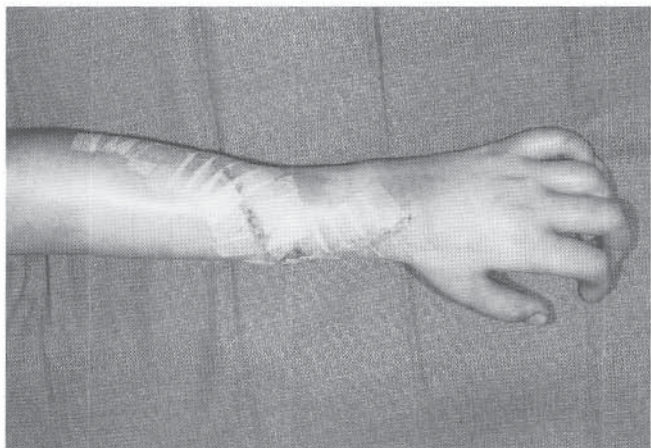
Illustration continued on following page



D



E



F



G

FIGURE 14–150 *Continued.* D and E, The vascularized free fibular graft was harvested as shown. F, Rigid fixation of the graft allowed restoration of the forearm by means of a one-bone forearm reconstruction. G, The patient is asymptomatic at 20-year follow-up. The x-ray shows solid union of the radius to the ulna and good alignment of the forearm. (The authors' use of this case is courtesy of Peter R. Carter, M.D., and John Tebbetts, M.D.)

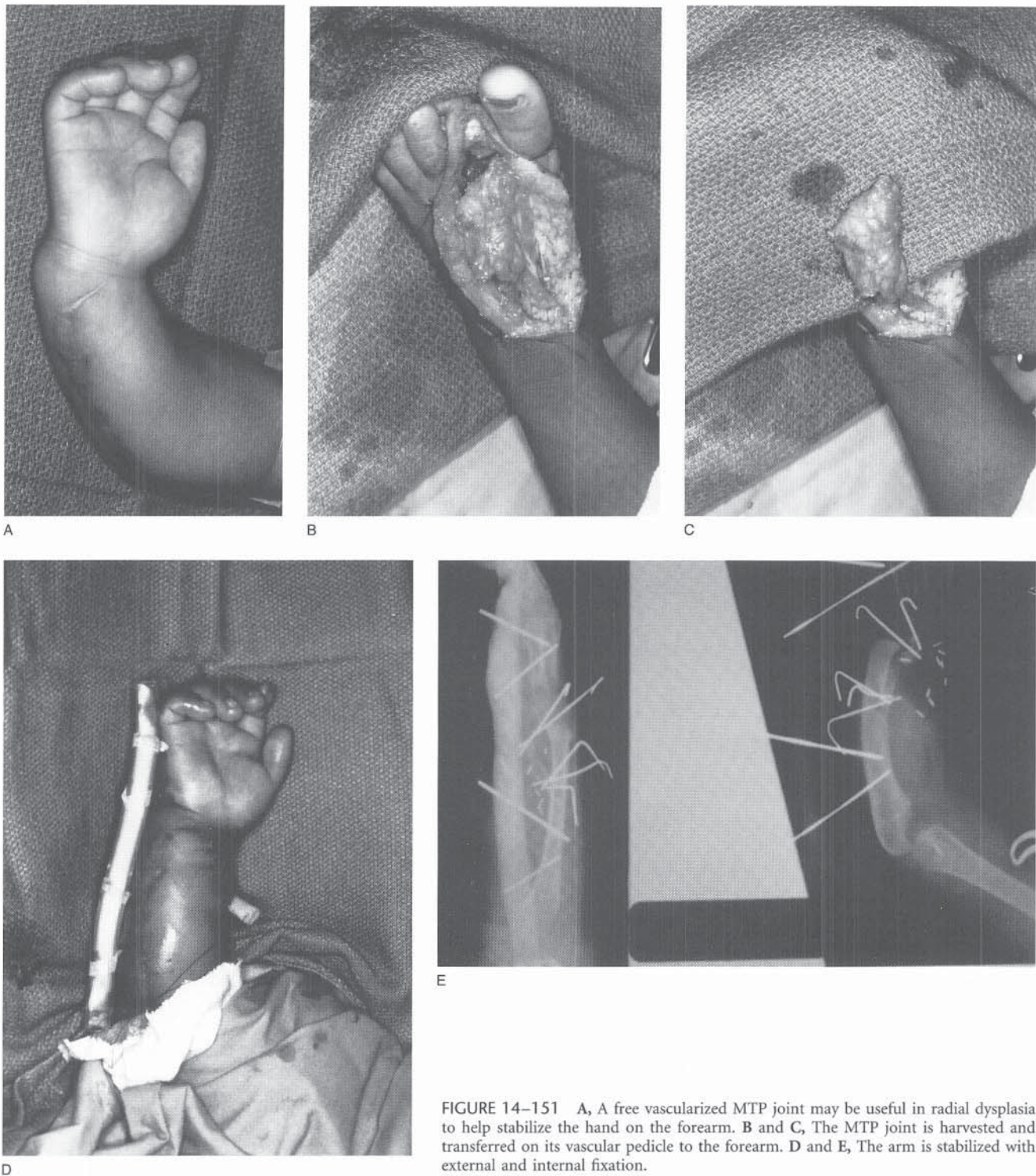


FIGURE 14-151 A, A free vascularized MTP joint may be useful in radial dysplasia to help stabilize the hand on the forearm. B and C, The MTP joint is harvested and transferred on its vascular pedicle to the forearm. D and E, The arm is stabilized with external and internal fixation.

proximal dissection must be avoided, and adequate fibular length must be maintained distally to prevent ankle instability.^{3,5}

FREE JOINT TRANSFER IN RADIAL CLUBHAND

A new development in pediatric microsurgery involves the use of a vascularized metatarsophalangeal (MTP) joint transfer in patients with radial hemimelia. In our experience, a pre-

liminary soft tissue release is needed to establish free passive correction of the radial clubhand deformity. Later, when the child is 2 to 3 years old, a free vascularized MTP joint (usually the second) is transferred to the radial side of the distal forearm. The MTP is anchored to the ulna and is inserted into the hand after passing the carpus. This gives support to the radial side of the wrist, as well as transferring an epiphysis to this area. In initial follow-up reports, the epiphysis appears to grow.⁷ Long-term follow-up is needed (Fig. 14-151).

REFERENCES

Microsurgery

1. Kay SP, Wiberg M: Toe to hand transfer in children. Part 1. Technical aspects. *J Hand Surg* 1996;21-B:723.
2. Kay SP, Wiberg M, Bellew M, et al: Toe to hand transfer in children. Part 2. Functional and psychological aspects. *J Hand Surg* 1996; 21-B:735.
3. Lee EH, Goh JC, Helm R, et al: Donor site morbidity following resection of the fibula. *J Bone Joint Surg* 1990;72-B:129.
4. Serletti JM, Schingo VA Jr, Deuber MA, et al: Free tissue transfer in pediatric patients. *Ann Plast Surg* 1996;36:561.
5. Strauch B, Yu HL: *Atlas of Microvascular Surgery: Anatomy and Operative Approaches*, p 218. New York, Thieme Medical Publishers, 1993.
6. Van Holder C, Giele H, Gilbert A: Double second toe transfer in congenital hand anomalies. *J Hand Surg* 1999;24-B:471.
7. Vilkki SK: Distraction and microvascular epiphysis transfer for radial club hand. *J Hand Surg* 1998;23-B:445.