"Radial clubhand" is a convenient but inaccurate label. It is commonly used to include a large variety of developmental abnormalities occurring along the preaxial border of the upper limb in association with a hand deviated at the wrist. The term “radial dysplasia” is useful to differentiate the deformity from an acquired clubhand but does not accommodate the significant anatomical deformities of joints, muscles, nerves, and vessels that are found in the forearm, elbow, and shoulder.

The spectrum of abnormalities varies from a minor degree of thumb hypoplasia to total absence of the thumb, its metacarpal, the scaphoid and trapezium, and the whole radius. In a small percentage of severe cases the developmental failure may trespass on the second ray and involve the scaphoid, trapezoid, second metacarpal, and index finger. The soft tissue structures of the hand and forearm are also involved, and variations of great surgical significance are common in the muscles, nerves, and vascular supply. Despite the fact that this condition is considered a preaxial affliction, the ulna is almost invariably involved. In moderate and severe cases the ulna is usually only about 60% of its
normal length at birth and this discrepancy is never corrected during subsequent growth. Radial clubhand is slightly more common in males than in females and in whites than in other races. Its occurrence has been variously estimated between 1 in 30,000 live births and 1 in 100,000 live births. The lower figure may be nearer the truth since it is based on Birch-Jensen's study of birth records in a Danish population of 4 million.

**ETIOLOGY**

The etiology of this defect is obscure. The essential cause must be a dysplastic factor acting on the apical ectoderm of the limb bud during the first few weeks of fetal life. Some theories claim that the deformity is produced by environmental factors such as compression during intrauterine life (Dareste theory), inflammatory processes (Virchow theory), maternal nutritional deficiencies, roentgen rays, and drugs such as insulin and thalidomide. Other theories explain the deformity as phylogenetic in origin, but some physicians consider a genetic factor more likely. No single theory is strong enough to resist reasoned criticism.

The genetic basis for the deformity is poor since hereditary tendencies are not common. Radial defects are unilateral in about half of the patients so affected, and the great majority of these defects occur sporadically. In unilateral cases the right side is affected almost twice as frequently as the left side. Bilateral involvement has been recorded in identical twins, and varying views have been expressed as to the degree of dominant or recessive heredity. In my series of 127 radial clubhands in 2758 hands and in those reported in the literature, I have found no real inheritance of radial clubhand from one generation to the next.

Radial clubhand is frequently associated with other malformations, and there are a great number of syndromes in which radial clubhand usually occurs.

**ASSOCIATED ABNORMALITIES**

It is essential that a child born with any degree of radial dysplasia have a complete physical examination by a competent pediatrician. Deficiencies along the preaxial border are frequently associated with other anomalies and syndromes; Goldberg has emphasized that a radial clubhand can point to far more serious but initially occult malformations elsewhere. He has listed over 20 syndromes associated with the condition. There are a few cases of isolated radial clubhand which have a sporadic or nongenetic etiology, although a few published pedigrees may suggest otherwise. The fact that the deformity is not part of a recognizable syndrome does not mean that other malformations may not be present and certainly does not justify the omission of a complete physical examination.
The following is a list of some of the syndromes associated with radial clubhand; they are described in more detail in the Syndrome Index.

**Syndromes with blood dyscrasias**
- Fanconi anemia
- TAR syndrome
- Aase syndrome

**Syndromes with congenital heart disease**
- Holt-Oram syndrome
- Lewis upper limb cardiovascular syndrome

**Syndromes with craniofacial abnormalities**
- Nager acrofacial dysostosis
- Juberg-Hayward syndrome
- Baller-Gerold syndrome
- Rothmund-Thomson syndrome
- Duane-radial dysplasia syndrome
- IVIC syndrome
- Levy-Hollister (LARD) syndrome

**Syndromes with congenital scoliosis**
- VATER association
- Goldenhar syndrome
- Klippel-Feil syndrome

**Syndromes with mental retardation**
- Seckel syndrome (rare)

**Radial aplasia and chromosome aberrations**
- Trisomy 18
- Trisomy 21
- Trisomy 13
- Chromosome 4

The blood dyscrasias and heart abnormalities are most important for the general health of the child and must be checked for in the physical examination. The three blood dyscrasias can be distinguished by a physical examination: in Fanconi anemia the thumb is absent, in the TAR syndrome it is present, and in the rare Aase syndrome a triphalangeal thumb is nonopposable and associated with a hypoplastic distal radius. A confirmatory peripheral blood study is always needed. In Fanconi anemia there is often a subclinical progressive pancytopenia which does not show up until midchildhood; the long-term outlook is dismal. In TAR syndrome the thrombocytopenia begins in the neonatal stage but the platelet count improves with time, and conservative treatment is proper until surgery can safely be done. Some prefer to think of this as the TARK syndrome because the knee is abnormal in over half the patients. I have never seen a child with Aase syndrome; its clinical features are outlined in the Syndrome Index.

An international Fanconi Anemia Registry is maintained by the Rockefeller University, New York, N.Y.
Cardiac abnormalities are particularly common in these patients, and the incidence of congenital heart disease has been recorded as between 10% and 13% in various series. There is a strong relationship between forearm anomalies and ventricular septal defects. It is interesting that the shaft of the radius begins development in the fifth week of fetal life, which is exactly the time that ventricular septum development begins. The Holt-Oram syndrome, which includes atrial septal defects, results from a dominant hereditary trait and shows a high association with radial aplasia. All these children show bilateral involvement, with the left limb always being more affected. The thumb is variably affected but is usually hypoplastic, nonopposable, and lying in the same plane as the palm. The Lewis upper limb cardiovascular syndrome is probably not a separate entity but a more severe example of radial and cardiac involvement.

The craniofacial abnormalities are described by a number of syndromes, each with a distinctive craniofacial problem. Some are rare, and the appropriate ones are listed in the Syndrome Index. Congenital scoliosis associated with radial clubhand occurs in three general types: the VATER association, the Goldenhar syndrome, and Klippel-Feil syndrome. The VATER association includes vertebral abnormalities, anal atresia or imperforation, tracheoesophageal fistula, esophageal atresia, and renal defects—all of which may not necessarily occur at the same time as the radial dysplasia. VATER has now become VACTERLS in the minds of some because about half the patients have a cardiac abnormality, 20% will have a lower limb abnormality, and a single umbilical artery occurs in many. The external genitalia of both males and females may be malformed.

With all these potential problems it is reasonable to expect these children to show some delay in the normal developmental stages but the long-term outlook is excellent. Development will catch up, the intelligence is normal, and reconstructive procedures have a lot to offer.

With all these potential problems it is reasonable to expect these children to show some delay in the normal developmental stages but the long-term outlook is excellent. Development will catch up, the intelligence is normal, and reconstructive procedures have a lot to offer.

The Klippel-Feil syndrome occasionally includes a patient with radial dysplasia as does the Goldenhar syndrome.

ANATOMICAL VARIATIONS

The morphological variations from the standard upper limb anatomy are numerous. Some are only of academic interest but may have important surgical implications. The first detailed anatomical study was published over 250 years ago when Petit reported the autopsy findings in an infant with bilateral radial clubhands. Since this time scattered anatomical reports have appeared, and in 1969 Skerik and I published a detailed study of the anatomical variations associated with these deformities. There follows a somewhat detailed account of the anatomical variations associated with varying degrees of involvement of the preaxial border. I have included it because I believe a surgeon needs to be aware of these anomalies. The impatient may ignore it but perhaps at their
patients' peril because scattered in these pages are practical comments on the surgical problems created by these alterations in normal anatomy.

The major anatomical defects are illustrated in cross sections in Fig. 16-1. The differences in the anatomical relationships at midforearm level are best seen between the normal and hypoplastic hands and between the hypoplastic and partially aplastic hands. The cross sections at wrist level accentuate the differences between partial and total aplasia of the radius. The flexor digitorum superficialis tendons to the index and small fingers and the extensor digitorum communis tendon to the index are frequently absent. Less frequently, but not uncommonly, the flexor digitorum profundus to the index is also absent.

Bones and Joints

Both O’Rahilly and Heikel have published detailed descriptions of the skeleton in radial dysplasia, and only the major defects of the skeleton are reviewed here.

Bones normal in more than two thirds of cases. The capitate, hamate, triquetrum, and the ulnar four metacarpals and their phalanges are the only bones of the upper extremity that are present and free from defect in nearly 100% of cases of radial clubhand. The trapezoid, lunate, and pisiform are also usually normal; however, they are more frequently involved (10%) than the others. Abnormalities of these carpals tend to be hypoplasia, fusion, or delayed ossification rather than total absence.

Bones present but abnormal

Short Humerus. In radial clubhand the humerus is generally considerably shorter than in a normal limb. In bilateral cases the humeri are usually of equal length except in cases in which the degree of involvement is much greater on one side. Other humeral defects are absence of the capitulum, the coronoid fossa, the intertubercular sulcus, the medial condyle, or the entire distal end of the bone. A small or poorly formed trochlea or olecranon fossa is a common defect. The proximal epiphysis is rarely mentioned in radial dysplasia, but the distal humeral epiphysis may demonstrate varying degrees of involvement, which seem to relate to the severity of the radial dysplasia.

Ulna. The ulna is usually curved, shortened, and thickened. It may be displaced backward on the humerus, its olecranon tuberosity may be defective, and its styloid and coronoid processes may be missing. Heikel reports, “There seems to be a tendency toward a delay in appearance of the distal epiphyseal nucleus of the ulna and possibly also toward too early a fusion of the distal ulnar epiphyseal line in partial and total aplasia of the radius.”

Wrist joint. A well-developed articulation between the ulna and carpus has been demonstrated in only a few cases. In most cases there is only a fibrous connection, although a flat cavity sometimes lined with hyaline cartilage has

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Fig. 16-1. Cross sections of the forearm and wrist in different degrees of radial dysplasia. R, radius; U, ulna; a, abductor pollicis longus; b, extensor pollicis brevis; c, extensor carpi radialis longus; d, extensor carpi radialis brevis; e, extensor pollicis longus; f, extensor indicis proprius; g, extensor digitorum communis; h, extensor digiti minimi; i, extensor carpi ulnaris; j, pronator quadratus; k, flexor carpi ulnaris; l, ulnar nerve; m, ulnar artery; n, flexor digitorum superficialis; o, flexor digitorum profundus; p, palmaris longus; q, median nerve; r, flexor carpi radialis; s, flexor pollicis longus; t, radial artery; v, pronator teres tendon; w, brachioradialis. (See Credits.)
been observed on the radial aspect of the distal ulna. The bones are usually bound together by tough fibrous tissue and are not covered with articular cartilage.

The patients in our series had only half the degree of active wrist motion demonstrated by the patients in Heikel's series. Average wrist motion in flexion and extension was 45°, and average active wrist motion in radial and ulnar deviation was 17°. Heikel reported averages of 83° and 28°, respectively [Fig. 16-2].

**Digital joints.** The most significant clinical feature of the finger joints is their lack of complete range of motion. Heikel found the index and long fingers more limited in flexion at the metacarpophalangeal joints than the other fingers, but hyperextension of the joint was often possible. Dissection showed that the joint surfaces of the interphalangeal joints of the index and long fingers were flat and irregular. He found flexion contractures most frequently at the proximal interphalangeal joints.

In contrast to Heikel's findings the metacarpophalangeal joints could not fully flex in 9 of the 11 limbs in our series. Of the two who could flex their joints, one had normal motion and one lacked 10° of full motion. These two patients with normal or near normal motion were the youngest in the study. Three patients were able to hyperextend at the metacarpophalangeal joints, and all patients were able to actively extend their fingers to neutral at the metacarpophalangeal joints.

The average ranges of digital motion in all three finger joints are recorded in Table 16-1.

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Fig. 16-2. *Active wrist motion.* **A,** Comparison between the range of radial and ulnar deviation reported by Heikel (left) and that recorded in the University of Iowa series (right). **B,** A comparison between the range of flexion and extension reported by Heikel (left) and that recorded in the University of Iowa series (right). [See Credits.]
Bones absent in more than one half of cases

**Radius.** If the radius is not totally absent, the proximal portion of the bone varies in shape and size and can be represented by a fragment that may or may not be fused with the ulna. Several cases have been described in which the proximal portion was absent and the distal radius present. This condition is very uncommon and, according to Steindler, not prone to produce deformity. Surprisingly, the literature lacks detailed descriptions of the radius in cases of partial aplasia or hypoplasia, but several cases have been reported in which the lower epiphysis of the hypoplastic radius was intersected with fibrous tissue. This tissue band provided attachment for some of the muscles that ordinarily originate or insert on the radius.

**Scaphoid.** Four of 21 cases reviewed had a normal complement of carpal bones; the scaphoid was missing in the remaining 17 cases. If the scaphoid is present, it is usually normal since it is rarely described as either rudimentary or fused with other carpal bones.

**Trapezium.** The trapezium, like the scaphoid, is frequently absent. In this review it was missing in 14 of the 21 cases. When it was present, it was rudimentary more often than the scaphoid.

**First metacarpal and its phalanges.** O'Rahilly states that the thumb, including the metacarpal and its phalanges, is absent in more than 80% of the cases of partial and total aplasia of the radius: It was absent in 15 of the 21 cases under review, but as previously mentioned, four of these cases had a hypoplastic radius. A rudimentary thumb is not uncommon. When this exists, the thenar eminence is usually absent and the thumb is a fleshy stump, sometimes having a small bone chip but no muscles.

In our series of 12 limbs there were two normal thumbs, six hypoplastic thumbs, two rudimentary thumbs, and two absent thumbs.

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Table 16-1. Average ranges of digital motion in all three finger joints

<table>
<thead>
<tr>
<th>Finger</th>
<th>Average Metacarpophalangeal Flexion*</th>
<th>Average Metacarpophalangeal Flexion†</th>
<th>Average Proximal Interphalangeal Flexion*</th>
<th>Average Distal Interphalangeal Flexion*</th>
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<td>Index</td>
<td>35°</td>
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<tr>
<td>Average</td>
<td>38°</td>
<td>54°</td>
<td>57°</td>
<td>22°</td>
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*From 0° extension.
†From hyperextension.
The hypoplastic thumbs were able to adduct and flex the first metacarpal but were severely limited in their ability to extend, abduct, and rotate the first metacarpal. All thumbs showed total absence of flexion and extension at the interphalangeal joint. The rudimentary thumbs had no functional capacity.

**Surgical implications.** The lack of normal skeletal support is the fundamental problem in the restoration of function to the limb. In the past varied attempts were made to create an internal support for the carpus and thereby retain some degree of wrist motion. In general the follow-up studies showed that these operations were not successful. The surgical pendulum then swung back to methods of stabilization of the carpus on the distal end of the ulna. Recently it has swung back again to attempts to provide some degree of controlled wrist motion.

The amount of growth potential in the distal ulna is poor since its epiphysis always appears late and closes early in comparison with the normal limb. Since at best the ulna achieves only 60% of its normal growth, to delay surgery until the epiphysis is closed is to wait too long. The profound changes that would occur are far worse than the loss of 1 to 2 cm of added growth [Fig. 16-3]. The bones of the affected limb are always shorter than normal, and nearly all will be sufficiently short to require alteration of ready-made clothing.

Lack of digital joint motion is one of the most severe handicaps suffered by these patients, and surgery has little to offer in the restoration of finger motion. Arthrogryposis is a poor diagnosis for these stiff fingers since the lesion lies in the joint and capsular tissues rather than in the extrinsic and intrinsic muscles. In some patients the stiffness can be extreme, but true symbrachydactyly is rare.

Those patients with a markedly curved ulna are sometimes subjected to correction osteotomies. I have tried this procedure and have abandoned it, ex-

![Fig. 16-3. Untreated radial dysplasia in an adult. The dislocation of the hand proximally along the ulna is profound. The distal ulna protrudes, and no length advantage has been gained in the limbs by delaying surgical correction.](image-url)
cept in extreme curvature, because I was unable to demonstrate any significant functional improvement. Usually a closing wedge osteotomy is done and rapid healing occurs; however, Wolff's law continues to operate and the curvature continues. Even multiple osteotomies have not helped, and I do not believe that the cosmetic improvement alone is sufficient to justify the procedure.

**Muscles**

There are numerous muscle abnormalities and deficiencies in these patients, and they vary in proportion to the skeletal defects. The postaxial muscles arising from the medial humeral epicondyle, such as flexor carpi radialis and ulnaris and the pronator teres, may be well defined in the proximal forearm but distally have many abnormal insertions. The preaxial muscles arising from the lateral epicondyle are more profoundly affected and show many variations.

These muscle aberrations mean that standard surgical approaches around the wrist are of little use. They are discussed in some detail because the variations around the wrist can be utterly confusing to those not experienced with this problem.

**Muscles normal in more than two thirds of cases.** All muscles inserting on the humerus, except the pectoralis major and the coracobrachialis, are usually normal.

In the forearm the origin of the flexor carpi ulnaris is always normal, but occasionally the insertion is aberrant.

If the extensor carpi ulnaris is abnormal, its origin or insertion or both may deviate somewhat from normal or it may be fused with the extensor digitorum communis or with the flexor carpi ulnaris.

The extensor digiti minimi is occasionally abnormal. When anomalous, it is usually fused with either the extensor carpi ulnaris or the extensor digitorum communis. The insertion, then, may be normal, or it may be in common with the muscle to which it is attached.

Within the hand the lumbricals have been described as part of the flexor digitorum profundus. Their innervation frequently varies. Total ulnar innervation is common, or the first lumbrical may be innervated by the median. If a lumbrical is missing, the one most frequently lacking is the lumbrical to the index.

Although the interossei are usually normal, the first dorsal interosseus, and occasionally others, can be absent. They can also be rudimentary, atrophied, or present but undifferentiated.

The hypothenar muscles are almost always normal, but like the interossei, they can occur as an undifferentiated mass in which the individual muscles cannot be isolated.

**Muscles frequently abnormal**

**Muscles absent in part.** The abdominal origin of the pectoralis major from the aponeurosis of the external oblique is often missing. Less frequently it may have an abnormal insertion, usually into the capsule of the shoulder joint, or it
may have an abnormal connection with other muscles, particularly the deltoid.

The long head of the biceps is almost always absent, but if present it originates from the anterior diaphysis of the humerus, the crest of the greater tuberosity of the humerus, or the capsule of the shoulder joint. When it is present in total absence of the radius, it usually inserts into the lacertus fibrosus of the biceps.

The short head of the biceps is always present but rarely normal. It is usually fused with another muscle, either the coracobrachialis at its origin or, more distally, the muscles of the forearm. The combined muscles usually originate from either the epicondyle or the forearm flexors. A terminal tendon is often absent, and the insertion is into the joint capsule and the brachial fascia. In the absence of the long head the short head frequently divides into two terminal tendons that insert either on the radial rudiment, medial epicondyle, or into the joint capsule. The biceps and all the muscles of the anterior compartment are frequently innervated by the median nerve.

The extensor digitorum communis is rarely absent but is frequently fused with neighboring extensors, primarily with the extensor carpi radialis longus, the extensor digiti minimi, or both.

The finger flexors usually show more variations in their superficial component. The flexor digitorum superficialis is commonly present but often incomplete, atrophied, or fused with the deep flexors. The radial head of origin is almost always absent, and the tendon to the index is most frequently missing. Insertions are normal but thin and tight or absent to the index or small fingers or both. The flexor digitorum profundus is more frequently normal than the superficialis, but like the superficialis, the tendon to the index is missing more often than any other tendon. Abnormal insertion sites for the profundus have been described on the bases of the proximal or middle phalanges of the digits.

**Muscles rudimentary or fused with other muscles.** The coracobrachialis frequently originates with the short head of the biceps as a single fused muscle mass, and it is often innervated by the median nerve.

The brachialis can be normal, rudimentary, fused, or absent. Most frequently it is fused with the biceps at its origin and has no specific site of insertion but becomes continuous with the muscles originating from the common flexor site. It can also insert into the tuberosity of the radius, into the joint capsule, or into the lateral epicondyle.

The pronator teres is frequently fused with the biceps-brachialis mass, with the palmaris longus, or with the flexor carpi radialis and usually inserts into the rudiment of the radius or into the intermuscular septum.

The brachioradialis is described by some as usually absent and by others as missing only in total absence of the radius. If the radius is present, the brachioradialis inserts on it; otherwise it inserts on the carpus. If it is fused to another muscle, it usually inserts with that muscle.

The extensor carpi radialis longus is often missing but is more often rudimentary or fused with adjacent extensors, primarily the extensor carpi radialis
he deltoid.
its origin, it originates from the lateral tuber-
cus of the humerus. It is a muscle of the deltoid.
Muscles in origin or, occasionally, from the lateral flaps.
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Table 16-2. Status of thumb muscles in 22 cases

<table>
<thead>
<tr>
<th>Muscle</th>
<th>Absent</th>
<th>Abnormal</th>
<th>Normal</th>
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<tbody>
<tr>
<td>Flexor pollicis longus</td>
<td>14</td>
<td>5</td>
<td>3</td>
</tr>
<tr>
<td>Abductor pollicis longus</td>
<td>14</td>
<td>5</td>
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<tr>
<td>Extensor pollicis longus</td>
<td>13</td>
<td>4</td>
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<td>Extensor pollicis brevis</td>
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<td>2</td>
<td>2</td>
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<tr>
<td>Flexor pollicis brevis</td>
<td>12</td>
<td>8</td>
<td>2</td>
</tr>
<tr>
<td>Abductor pollicis brevis</td>
<td>13</td>
<td>7</td>
<td>2</td>
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<tr>
<td>Opponens pollicis</td>
<td>16</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>Adductor pollicis</td>
<td>16</td>
<td>5</td>
<td>1</td>
</tr>
</tbody>
</table>

If the flexor carpi radialis is present, it is usually so highly anomalous that it is hard to identify with certainty.

When the palmaris longus is present, it is frequently fused with the flexor digitorum superficialis or the other flexors and takes a more ulnarward insertion than usual, attaching to the pisiform, to the fifth metacarpal, or into the soft tissue over the third and fourth metacarpals.

The pronator quadratus is usually totally absent. Occasionally it may be represented by a mass of muscle surrounding the distal extremity of the ulna or may be found inserting into the radial side of the carpus or into the second metacarpal.

In partial and total absence of the radius the extrinsic and intrinsic muscles of the thumb are rarely normal whether or not a thumb is present. In more than 50% of the cases of radial hypoplasia the muscles are abnormal or absent, with the intrinsic muscles especially affected. When the intrinsic muscles are present, they are usually grossly abnormal, having neither a normal origin nor a normal insertion. A common insertion site is into the metacarpal of the index or long finger. This aberrant insertion occurs even in cases with a rudimentary or hypoplastic thumb.

The status of thumb muscles in 22 cases reported in the literature is listed in Table 16-2.

**Surgical implications.** The many aberrations of the muscles make textbook surgical approaches to the area around the wrist impossible. The abnormal muscle bellies and the aberrant insertions utterly confuse the tyro seeking normal muscle planes. There are normal fascial planes between the abnormal muscles, but fusions between muscle bellies frequently prevent the usual proximal soft tissue spreading and separation.

Multiple insertions into both sides of the carpus, but particularly on the
The extrinsic flexor tendons are frequently undifferentiated at the wrist, and occasionally tendons will branch off to carpal or metacarpal insertions while the main tendons pass on into the fingers. The main tendons to each finger should be identified, and their free motion should be demonstrated against passive resistance of the fingers. Free-running tendons are vital if the full power of the muscles is subsequently to be used in mobilization of the fingers.

Nerves

Disturbances of the normal pattern of nerve distribution may be profound and can be found as far proximal as the root of the brachial plexus. Although generally considered normal, some of the main branches of the plexus contain fibers
from a higher cervical segment than usual. Figs. 16-5 and 16-6 illustrate the nerve supply to the upper limb in several cases of radial dysplasia.

Normal nerves. The axillary nerve and the ulnar nerve are usually present and normal.

Absent nerves. The musculocutaneous nerve is most frequently missing. If present, it is usually anomalous, either joined with or substituted by the median nerve. The radial nerve usually ends just above the lateral epicondyle after innervating the triceps. The median nerve provides sensitivity to the radial side of the hand and anastomoses with the sensory branch of the ulnar nerve on the dorsum of the hand.

Abnormal nerve

Median. The vitally important median nerve is always present, but its distribution may be altered, depending on the status of the other nerves. It supplies the muscles of the anterior compartment of the arm in the absence of the musculocutaneous nerve. It substitutes for the terminal distribution of the radial nerve by dividing into two branches of which the more radial branch takes over radial sensory function. In a quarter to a third of the patients this division occurs within the forearm. The remainder occur within the plexus or arm.

The course of the median nerve is subject to many strange variations, most of which have been described by Stoffel and Stumpel, who traced its course in 16 cases. Its route varied in eight ways.

1. It traveled very superficially along the radial edge of the brachioradialis until it entered the palm.
2. It coursed through the forearm on the underside of the palmaris longus and flexor digitorum superficialis to the carpal ligament.
3. It traveled through the forearm along the lateral edge of the flexor digitorum superficialis to the hand.
4. At the elbow it dipped beneath the flexors and supplied them, then reappeared superficially in the middle of the forearm between the brachioradialis and the extensor digitorum communis.
5. After sending strong branches to the flexors of the arm, it went under the pronator teres to the undersurface of the flexors, supplied this area, then came to the surface at the flexor aspect of the wrist, and gave off a dorsal branch to the hand.
6. In two cases it supplied the flexors of the arm and came to the surface between the flexor digitorum superficialis and the brachioradialis, traveling to the radial side of the palm.
7. After branching to the flexors of the arm, it pierced and supplied the brachioradialis. It then followed a normal course in the forearm except for piercing the pronator teres and locating itself between the flexor digitorum superficialis and the flexor digitorum profundus.
8. It supplied the arm flexors and then sent a sensory branch to the brachioradialis, which it pierced to supply the area normally innervated by the absent lateral antebrachial cutaneous nerve.
illustrate the unusual variety of aberrant nerve supply that can occur in radial clubhand. The thumb is usually present but its sensory supply is often aberrant. If the superficial radial nerve is not present, the musculocutaneous nerve is typically missing. If the humeral epicondyle is not ossified, the radial side of the brachioradialis muscle is supplied by the median nerve on the ulnar aspect of the forearm.

In the presence of a radial clubhand but a normal thumb, its normal sensory supply is from the median nerve. This is due to the absence of the superficial radial nerve and the musculocutaneous nerve. In patients with a radial clubhand, nerve distribution of the radial branch of the lateral antebrachial cutaneous nerve is variable; in many patients this nerve is distributed almost solely in the ulnar area. Some patients have a normal plexus or a normal radial nerve. Other variations, most frequently observed, are the aberrant course of the median nerve.

To the flexor digitorum profundus, then re supplied the flexor digitorum superficialis on the preaxial side. The brachioradialis is supplied by the median nerve on the ulnar side, then re supplied by the ulnar nerve on the lateral side.

Aberrant nerve supply—When the thumb is present but the superficial radial nerve is absent, the thumb's sensory supply comes solely from the median nerve through its normal and supplementary branches. Note absence of the musculocutaneous nerve. (See Credits.)

Aberrant nerve supply—When the thumb is absent, the median nerve still supplies supplementary cutaneous branches along the preaxial border. Note absence of the musculocutaneous nerve. (See Credits.)
Surgical comments. Despite the fact that disturbances of the standard neurological pattern occur as far proximal as the roots of the plexus, the distal peripheral course of the nerve is of great surgical significance.

There is great variation in the sensory pattern, but invariably there is good coverage; areas of anesthetic skin are not known to occur. Variations in standard motor patterns also occur but do not have clinical significance.

It is the median nerve above all others that presents a significant surgical challenge. The nerve is thicker than is usually seen in a normal arm of the same age because it carries additional sensory fibers normally distributed by the radial nerve. The course is almost invariably aberrant. It is consistently preaxial and usually lies immediately beneath the fibers of the deep fascial cylinder. The nerve represents a strong and unyielding bowstring of the radially bowed forearm and hand.

If the common pattern of the two-stage corrective operation is used, the nerve may cause great trouble in the first, or soft tissue, corrective stage. After the Z-plasty flaps of the skin approach have been mobilized, the deep fascia will have to be split widely up the forearm and the peripheral distribution of the nerve identified (Fig. 16-7).

Release of abnormal muscle insertions and other soft tissue attachments will usually provide a much greater degree of correction than can be tolerated by the nerve. It is hard to judge how much stretching the nerve can withstand and yet provide sensory and motor conduction. I usually err on the side of greater tension and have not yet seen any persistent evidence of anesthesia or paralysis, although temporary minor degrees of disturbances have been detected.

I do not repair the deep fascia because to do so would only restore the original restraint on the nerve. Even the skin lengthening supplied by the Z-plasty is usually not adequate, and the nerve bowstrings beneath the flaps as the hand is corrected on the ulna at the end of the operation. Serial plaster casts usually provide full correction without much difficulty, and only very rarely have I had to do a second soft tissue release operation to obtain a satisfactory positioning of the hand on the single forearm bone.

Occasionally the nerve breaks up into its final small branches well proximal in the forearm, making it both difficult and tedious to dissect out all these important branches (Fig. 16-8). More often the nerve retains its astonishingly large size almost to the level of the wrist crease, and its large size and subcutaneous course have frequently led to its resection by a surgeon under the mistaken impression that it represents the fibrous anlage of the radius (Fig. 16-4).

Vessels

The brachial artery is usually normal. However, it may divide into two branches high in the upper arm or may not divide at all at the elbow. The deep brachial artery may come off the posterior humeral circumflex. Blauth and Schmidt have described 11 arteriograms done on nine children with radial dys-
plasia and have shown that the vascular anomalies correspond closely to the extent of radial dysplasia. The greater the involvement, the more likely the radial artery and palmar arterial arches are to be involved. The ulnar artery is usually present and normal, but it may be anomalous concomitant with the absence of the radial artery. Heikel points out that the vessels, as well as the nerves and tendons in the forearm, course radially at the distal end of the ulna (Fig. 16-9).

The interosseous arteries are usually well developed and may replace the radial or ulnar arteries or both. In severe cases the palmar digital arteries may be of small caliber and even absent on the radial side of the index finger.
Surgical comments. Vascular anomalies cause little problem for the surgeon. The tendency for a radialward deviation of the vessels does mean that the normal relative positions of the radial and ulnar arteries and median and ulnar nerves are disturbed. During a first-stage soft tissue release operation it is usually easy to find the ulnar artery, which is frequently enlarged. I deliberately look for and protect this artery because it is frequently the sole—or at least the major—arterial supply for the limb. The second-stage operation is done through a dorsal approach and must place at hazard any branches of the interosseous arteries that enter the hand over the dorsum of the metacarpals. If the ulnar artery is damaged during the first-stage operation and the radial artery is rudimentary or absent, the blood supply of the hand can be seriously compromised by the second-stage dorsal approach. If the index finger is to be pollicized, the absence of its radial digital artery demands a very careful protection of its ulnar digital artery during the operation.

FUNCTIONAL IMPLICATIONS

A radial clubhand is not a normal hand set on an abnormal wrist; rather, it is a profoundly abnormal hand joined to a poor limb by a bad wrist. The fully developed radial clubhand is a hideous deformity and a functional liability (Fig. 16-10). Lesser degrees of involvement imply a smaller loss of function, but in fact dexterity and skilled activities are severely inhibited. Most children with unilateral involvement become functionally independent, but in those with bilateral deformity functional ability is often less than might be anticipated.

Lamb has carefully followed over 68 patients and stresses that in the unilateral case the other normal arm dominates function and the affected limb is simply used as an aid.
Children with bilateral deformities often have significant difficulties washing, dressing, and feeding themselves. These everyday tasks become virtually impossible when there is associated stiffness of the elbows. Lamb has recorded a high incidence of elbow stiffness in the first and second years of life. Fortunately, as the child grows, motion usually returns to the joint. Full elbow range is virtually never achieved, but eventually nearly 90° of flexion has been obtained in many patients.

The ulnar two digits in these patients are nearly always normal, and the children tend to grip with these fingers (Fig. 16-11). This ulnar prehension is common because the two radial fingers invariably show some degree of stiff-
The index is particularly involved and is often held flexed, and many index fingers even show hypoplasia and significant flexion contracture.

We studied the functional activities of nine individuals with radial dysplasia chosen to represent the spectrum of digital involvement. Three patients had bilateral involvement: one with normal thumbs, one with hypoplastic thumbs, and one with rudimentary thumbs. Three unilateral cases had no thumbs, two had hypoplastic thumbs, and one had a rudimentary thumb.

In defining maldevelopment of the thumb we have used Heikel's definition of rudimentary as a thumb with no active motion and attached by a soft tissue pedicle. Hypoplastic thumbs are those that are not rudimentary but do deviate from the normal in size, shape, and position. Hypoplastic thumbs were present on half of the hands studied, but they did not in general significantly increase the functional level of the hand over those hands without thumbs. Factors that contributed to this were:

1. Active finger flexion was limited, and opposition between the thumb and the fingers was impaired.
2. Thumb motion frequently was limited. Motion ranged from indifferent carpometacarpal “wiggles” to a fair representation of most motions. Complete active range of motion at each joint was not present in any thumb. Interphalangeal flexion and extension were severely limited in all thumbs. Abduction and extension motions were in most cases far more limited than flexion and adduction motions.

The only significant difference between hands with and without thumbs was the ability of the patients whose hands had thumbs to touch their thumb to the ring and small fingers, which in most cases displayed a greater range of interphalangeal motion than was present in the index and long fingers. Since the thumb was not able to abduct and rotate to a great degree at the carpometacarpal joint, a lateral-type pinch was used (Fig. 16-12). This usually worked for fine pinch, but when strength was needed, the index was substituted for the thumb.

Although the hands could in some way perform most activities, they did not demonstrate the generally accepted normal functional patterns. Patterns of use could be identified, and it was found that patients with thumbs used a lateral pinch. A true tip- or palmar-type pinch is rarely possible for a hand with congenital radial dysplasia.

Spherical grip was preferred by all patients (Fig. 16-13). Adduction and abduction between the index and long fingers were developed to a greater range of motion than in the normal hand, allowing the index to substitute nicely for a thumb (Fig. 16-14). The index also frequently demonstrated a significant degree of pronation so that its palmar surface faced ulnarward, further enhancing its ability to substitute for the thumb. With a spherical grip the fingers are positioned in a tripod configuration, allowing a relatively strong and dexterous grip.

Cylindrical grip was less used, probably because the thumb could not ab-
and many others.

Radial dysplasia patients had thumbs, two thumbs, and many thumbs.

Definition of the thumb is as follows: soft tissue and bone that deviate in a manner present in any motions. That is limited in cases far

They did not use a spherical grip. Since the carpometacarpal joints worked for the

Fig. 16-12. *Lateral pinch.* Whether or not a thumb is present, a lateral pinch is the type of pinch preferred by all patients with radial dysplasia. (See Credits.)

Fig. 16-13. *Spherical grip.* Note the incomplete abduction at the carpometacarpal joints of the thumbs when spherical grip is used by hands with total aplasia of the radius. (See Credits.)

Fig. 16-14. *Tripod configuration of the fingers.* In congenital radial dysplasia with total absence of the thumb, the index passes beneath the long and ring fingers to oppose the pulp of the small finger. (See Credits.)
duct sufficiently and the index was not in a position to substitute for a thumb (Fig. 16-15). Power grip was usually impossible. A good fist could not be made because of lack of full flexion of the fingers. To our surprise hook grip was the least preferred, even though it is the only form of prehension that does not actively use the thumb. Lack of full finger flexion and weakness of the finger flexors must be the reasons for the general disfavor toward hook grip.

When the thumb is absent, the index tends to deviate ulnarward and the other fingers radialward. Thus in flexion the central two fingers tend to overlap the converging border fingers (Fig. 16-16).

![Fig. 16-15. Lateral pinch between two fingers. An attempt to perform a cylindrical grip results in the most prevalent form of pinch used by patients with radial dysplasia. (See Credits.)](image1)

![Fig. 16-16. Rotation of border fingers. In the absence of the thumb the rotated border fingers tend to lie beneath the central digits during flexion. (See Credits.)](image2)
Two types of prehension are seen in these hands that are uncommon in the normal hand: a lateral pinch between any two fingers and what appears to be a variation of spherical grip—the index and small fingers are placed on the sides of an object to be picked up and the long and ring fingers on top of the object. A combination of adduction and flexion of the fingers is then used to pick up the object.

Thus, although these patients do not use the more formal or standardized types of prehension, they can do almost anything they wish. Their hands are clumsy and lack skill, but they also lack too many of the necessary anatomical components for surgery to produce dramatic functional improvement.

CLASSIFICATION

Treatment for a condition carrying such a variety of anatomical and functional possibilities must be based on some form of classification. Heikel's original classification into three types has been usefully expanded by Bayne into four types based on skeletal size. He points to the direct relationship between the amount of radial deficiency and the degree of clinical deformity of the radial clubhand.

Type I: Short Distal Radius

This is the second most common type. In it the distal radial epiphysis is present but delayed in appearance; thus the growth of the distal radial epiphysis is decreased, causing a normal appearing but short radius. There is little radial deviation of the hand because of adequate radial carpal support. The radius has normal proximal epiphyseal growth. Thumb hypoplasia is almost always present (Fig. 16-17).

Type II: Hypoplastic Radius

Both proximal and distal epiphyses are present, but growth is defective in both. This is the rarest type and is essentially a radius in miniature. Growth of the radius proceeds at a decreased rate; the ulna is bowed and the carpus unsupported (Fig. 16-18).

Type III: Partial Absence of Radius

The radius is partially absent. The defect has been reported in the proximal, middle, or distal third; the most frequent is the absence of the distal one or two thirds. The hand is radially displaced. The ulna is thickened, shortened, and bowed radially, and the carpus is unsupported (Fig. 16-19).
Fig. 16-17. *Type I radial deficiency*. A, Hypoplasia of the right radius and thumb. There is an inhibition of the development of the distal epiphysis of the radius. Observe the thickness of the right ulna. B, Clinically there is only a discrete degree of radial deviation and a visible hypoplasia of the thumb on the right. [See Credits.]

Fig. 16-18. *Type II radial deficiency*. Although both proximal and distal epiphyses are present, defective growth has produced a miniature radius. [See Credits.]
A, Hypoplasia of the right distal epiphysis of the radius of discrete degree.

Fig. 16-19. Partial aplasia of the radius. Note absence of the distal radius, extreme hypoplasia of the thumb, and curvature of the ulna.

Fig. 16-20. Total aplasia of the radius. There is hypoplasia of the whole hand, absence of the thumb, and absence of several carpal bones. The hand is dislocated proximally on the radius.

Type IV: Total Absence of Radius

Most believe this is the most common type of radial deficiency. It comprised 66% of Bayne's series of 101 radial deficiencies and was nearly three times more common than Type I. The hand is unsupported and is severely radially displaced. A radial anlage is rarely present (Figs. 16-20 and 16-21).

The care of these four types of involvement is very different and includes both conservative and surgical measures.
PRINCIPLES OF TREATMENT

Treatment of this condition is difficult because of the balance that must be made between function and appearance. The fact that adults with fully developed bilateral deformities can work full time, marry, and rear children is hardly a justification for surgical nihilism in the growing child. All the adults I have known with this condition have perforce adjusted to it, but none were happy with their lot. Commonly, adults with this deformity demonstrate adequate function and adaptation of their limb to ordinary activities. They often choose occupations appropriate to their limitations, and appeals to improve their cosmetic appearance should be very carefully analyzed. Virtually any surgery might compromise function, and function must remain the principal aim of treatment (Fig. 16-3).

Children born with Type I and mild Type II aplasia can be treated by manipulation, plaster casts, stretching, and splinting. Serial casting will be necessary, and splintage may have to be maintained until bone growth ceases.

However, when the wrist is unstable as in the more severe Type II and in Types III and IV, surgery will be necessary. The obvious aim of treatment is to position the hand on the single forearm bone in such a manner that motion, growth, and good appearance are obtained. When one limb is normal, the function of the affected side becomes less important and surgical correction can tend toward appearance more than ability. I agree with Lamb that in bilateral
cases both sides can be operated on, particularly when the planned procedure for the less functional limb may provide better appearance, even at the expense of some function.

Before any surgery can be undertaken, motion must be available in the elbow. Although infants with a stiff elbow can place their hand in the mouth, adults cannot. Stiffness of the elbow tends to improve with time and is greatly helped by manipulation.

Many surgical procedures to permanently maintain a centralization of the hand have been reported over the last 100 years. These have been extensively reviewed by others, and Urban and Osterman have recently summarized the history of this period. Centralization was usually accomplished after stretching or division of the deforming soft tissues, osteotomy of the ulna, and excision of the carpal bones. Fixation of the hand in the corrected position was achieved by a variety of means using silk, kangaroo tendon, gold wire, chromic catgut, and stainless steel wire and pins.

Another approach has been to attempt to replace the absent or dysplastic radius by a bone graft. Tibial, ulnar, and fibular grafts have been used to stabilize the hand in proper alignment on the distal end of the forearm. In 1965 Riordan reported his 15 years' experience using the upper end of the fibula. He abandoned the procedure and practices early implantation of the distal ulna into the carpus. Modern microsurgical techniques have prompted new attempts to transfer the upper end of the fibula with vascular anastomosis to the epiphyseal blood supply. No reports of a significant series of this new trial have yet been published.

Splitting the ulna longitudinally to broaden its distal end is another method that has been described to maintain the hand over the distal end of the ulna. Muscle, silk, and ivory pegs have been placed between the two halves of the split distal end of the ulna in an effort to keep the hand in the correct position. In 1966 Define described a technique in which results were satisfactory after 5 years. The periosteum is elevated from the distal third of the ulna and the hand displaced ulnarward so that the distal end of the ulna is in contact with the radial side of the carpus. A periosteal tube is then formed on the ulnar side of the ulna with the expectation that a bone peg will be produced. The hand is then supported by the original distal end of the ulna on its radial side and by the newly formed bone peg on its ulnar border. The common theme in all these reports has been the problem of maintaining the stability of the wrist joint.

Wrist stabilization improves the appearance but more importantly improves function by abolishing the waste of the power of the extrinsic digital muscles over a floppy wrist. Finger motion is thereby improved and grasp increased.

For years the common operation has been to square the distal end of the ulna and place it in a commensurate slot created in the carpus aligned with the third metacarpal. The improvement in function and in appearance is substantial, and the long-term follow-up studies show that the improvement is main-
tained. Some surgeons felt that placing the ulna into a carpal slot during early infancy or childhood could arrest growth of the distal ulnar epiphysis. This is true but not functionally significant.

Most descriptions of radial aplasia include curving of the ulna as an integral part of the deformity (Figs. 16-19 through 16-21). This is especially true of untreated cases. Lamb believes, and I agree, that in most cases this curvature is caused by soft tissue tightness on the radial side. In many patients the ulna can be coaxed to grow straight if properly splinted and if soft tissue surgical release is done when appropriate. If the ulna is already bowed at birth, this is probably indicative of the presence of an anlage of the radius creating a tight bowstring (Fig. 16-4). Splinting or osteotomy will not help these cases, and early surgical release is imperative. Osteotomy of the curved ulna is an attractive idea and may well improve the cosmetic appearance, but it does little to gain length. Mathematical studies show that even under optimum conditions the percent-age gain in length is tiny.

In recent years several successful procedures have been devised to stabilize the hand and provide some degree of controlled wrist motion by appropriate tendon transfers. These operations are invariably preceded by a period of splint- ing and stretching the tight radial structures.

A few surgeons are now applying the Ilizarov system to the short forearm. Long-term results of these attempts are not yet published, and anecdotal reports vary from enthusiasm to concern over the complication rate. A recent report from Lecco, Italy, states that six patients had:

... ulnar lengthening in the presence of complete radial agenesis. ... All had a successful lengthening, gaining from 4 to 13 cm. However, the procedures were prolonged (7 to 25 months) and all patients experienced complications. ... The follow-up averaged 45 months. ... These cases suggest a cautious approach. ... This is a long, arduous, painful process that requires a psychologically robust patient.

CONTRAINDICATIONS TO SURGERY

There is general agreement that surgery is not indicated in certain patients (although several surgeons voice polite dissent). These patients include the following:

1. Children with severe associated anomalies which significantly decrease life expectancy.
2. Very young patients—under the age of 6 months. These patients rarely need surgery; it is much better to allow time for blood dyscrasias and major organ defects to be detected.
3. Adults with firmly established functional patterns. I do not believe this is an absolute contraindication to correction and still operate on the

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occasional adult. However, societal pressures are now such that few children are left untreated.

4. Patients with such severe soft tissue contractures and neurovascular deficiencies that correction is not possible. This is really a post-hoc situation discovered at surgery, and one is faced with doing nothing or further shortening the ulna to avoid circulatory complications.

5. Patients with Type I and mild Type II deformities. These patients can usually be treated satisfactorily by conservative means.

TREATMENT

Whatever the degree of radial deficiency, treatment should start at birth with a thorough physical examination. If all is well, then manipulation and splinting of the limb into the corrected position should be started immediately. This may be all that is necessary in treatment of the patient with hypoplasia of the radius; in more extensive involvement it is a necessary preliminary to permanent operative correction.

Types I and II Radial Deficiencies

**Manipulation.** In the Type I and in milder degrees of Type II deficiencies manipulation of the wrist and elbow may be all that will be required. A stiff elbow can often be improved by repeated gentle manipulation, and the mother must be taught to do this for short periods many times a day. Parents do not relish the idea of hurting their infant, but they have to be taught that to inflict discomfort is virtuous in this instance. A fussing infant is acceptable, but a bawling and restless baby is in pain, and the degree of correction being obtained is too great for tolerance. As the elbow is being bent, the hand must also be corrected on the forearm both by distraction and ulnar deviation.

The key to success is to apply persistent mild stretching rather than repeated quick "bendings"; I have seen a fractured ulna result from the latter type of "manipulation." Ideally, one should hope for 90° of flexion, but lesser flexion is acceptable; in my series the average preoperative flexion was 70°.

**Splinting.** If the parents cannot bring themselves to inflict discomfort on their infant, then serial casts will have to be tried. Such casts or splints are hard to apply and hard to keep in place; manipulation is a far better treatment. The only practical method to use is a lightly padded plaster of Paris cast extending well proximal to the flexed elbow. The cast must be placed on the infant's limb in three parts in a similar fashion to the application of a cast for clubfoot. The hand itself must be enclosed in plaster; the thumb should be excluded, but the fingers may be included in the early casts if by this means a good purchase is obtained along the radial border of the hand. The hand is then correctly placed on the forearm, and the wrist and lower forearm are enclosed in plaster. Finally, the cast is extended high up on the arm over the elbow, which is flexed to at least 90°. It is not easy to apply such a cast, and great care must be taken not to
make pressure sores from plaster creases. It is impossible to maintain such a
cast on the chubby straight arm of an infant, and if the elbow is stiff in exten-
sion, a cast cannot be used.

The cast will need to be changed to gradually improve the position of
the elbow and hand, but it can usually be left on for several weeks between
changes. An alternative to a plaster cast is some form of bracing. I have had
poor results from bracing an infant’s hand, principally because of the difficulty
in obtaining a proper fit.

Although bracing will encourage soft tissue relaxation or stretching, it does
not always provide the essential ingredient to proper alignment—namely, place-
ment of the hand across the distal end of the ulna. Too often the external device
tends to tilt the ulnar border of the carpus on the distal ulnar epiphysis. Re-
peated mild distraction manipulation is usually necessary to separate the car-
pus from the ulna; the hand should then be displaced ulnarward to line up the
long finger metacarpal with the shaft of the ulna. When this has been achieved,
a brace can be applied. The carpus can be correctly centered over the distal ulna
without full correction of the radial deviation. This does not matter since it is
this type of forearm that responds readily to the ratchet-type splint advocated
by Lamb (Fig. 16-22).

When, after treatment in a cast or brace, the hand can be readily centered
on the forearm, a bivalved, plastic, gutter-type splint with Velcro strapping,
which is easily applied by the parents and can be readily cleaned, is used.
Cooperation by the parents is essential in the early days of this treatment, and
they must be taught to correctly place the center of the carpus over the end of
the ulna every time they reapply the splint. I prefer to leave the digits free to
move but usually recommend that the splints be worn continuously in the first
few months of life. When full correction can be readily maintained with only
the slightest ulnarward pressure, the splints can be left off in the daytime but
should be applied during rest periods and at nighttime. Manipulation supple-
mented by night splinting will be needed at least until growth has ceased.

In the more severe Type II deficiency, growth will make it increasingly
difficult to retain the carpus on the distal end of the ulna. Eventually external
splinting can do no more, and surgery will have to be undertaken. I have found
that one is forced into a surgical decision well before the child reaches school
entry age and frequently before the age of 2 years.

When a fibrous anlage of the radius is present at birth, if the ulna is mark-
edly curved or if the soft tissue contracture is so severe that correction cannot
be obtained, then an early surgical release of the radial soft tissues is essential.

Surgical release. The approach to unyielding radialward contractures has to
be through the concave radial side using a Z-plasty to provide adequate expo-
sure. The deep fascia has to be opened, and portions have to be excised; it is not
resutured at the end of the operation (Fig. 16-23).

More often than not, the median nerve is lying immediately beneath the
depth fascia (Fig. 16-7), and on occasion the fibrous anlage of the radius will be
found on a deeper plane (Fig. 16-4). Proximal and distal mobilization of the
median nerve and release of the anlage may allow full correction.
tain such a stiff in extension.

The position of the carpals between the head of the radius. I have had some difficulty
in obtaining, it does not necessarily place-
line up the radial styloid. When achieved, dislocation of the distal ulna
is almost certain since it is not advocated
in this way.

If the carpus can be correctly centered over the distal ulna, then a ratchet-type brace will be useful in obtaining full correction of the radial deviation. A, The deformity before correction. B, Photograph of same child after relief of fixed deformity by gradual correction. (See Credits.)

Fig. 16-23. Deep fascia of the forearm. The deep fascia is a tough, unyielding layer that must be opened and excised to obtain full correction. The median nerve penetrates the fascia proximal to the wrist and is subcutaneous in the rest of its course.
When the carpus cannot be fully corrected, both normal and abnormal muscle attachments to the carpus must be released. This release must be ruthless; there is no point in inflicting the operation on an infant if full relaxation of the contracture is not obtained. If the detached muscles can be reattached to the dorsoulnar portion of the carpus after release, they should be held in place with a few fine nonabsorbable sutures.

Often the median nerve will still be the tightest structure after all soft tissue release has been done. It should be placed under only mild tension, and the appropriate alignment of the hand in relation to the forearm must be determined before the Z-plasty flaps are transposed and the skin is closed with interrupted mattress sutures of fine catgut. The arm and forearm are covered by a lightly padded plaster of Paris cast, which is left on for about 2 weeks. After this the plaster can be removed, the forearm measured for a splint, and a new plaster cast applied while the splint is being made. Since correction of the carpus on the ulna has now been obtained, the future aftercare is similar to that for the Type I and mild Type II deformities.

Types III and IV Radial Deficiencies

In these more severe deficiencies early manipulation and splinting are helpful but not curative. The hand will have to be maintained on the distal ulna by surgical means. Controlled motion of the elbow is essential before stabilization, and it is rare for early manipulation and casting not to have yielded sufficient elbow range.

Only very rarely would one have to consider doing a posterior elbow capsulotomy. Even more uncommon would be tendon transfers to aid the usually weak flexors in overcoming the strength of the normal elbow extensors.

The choices available for stabilization are arthrodesis, which yields no wrist motion, or the more recently developed operations, which allow some wrist motion. The latter are preferable, but there are still indications for ulnar implantation into the carpus.

Ulnar implantation. Lamb has written extensively about this procedure, and his results are impressive. The fact that an arthrodesis is to be done does not exclude the need to eliminate deforming forces by transferring aberrant tendon insertions to the ulnar side of the hand.

How early should this “early” surgery be done? Certainly there is general agreement among those who are caring for significant numbers of these children that to delay until after 3 years of age is late. At this time the operation becomes technically more difficult because of the width of the ulna. Implantation of the ulna should probably be done during the first year of life, and the timing is dictated more by the failure of manipulation and splinting than by the calendar. The results of operations done before the age of 3 are far better than those done at a later age. Most surgeons find the first birthday a reasonable compromise between the size of the structures to be dissected and the tensions on the radial side of the limb.
Details of operation. Several skin approaches can be used, depending on the extent of the planned operation. If an extensive amount of soft tissue release and several tendon transfers are anticipated, the incision used by Lamb is appropriate. The approach extends from the dorsal base of the index over the ulnar side of the wrist and across the flexor aspect of the forearm to its radial border. An S-shaped dorsal skin incision gives good access to the dorsum, but one cannot approach the flexor aspect of the wrist.

If releasing incisions and tendon transfers has already been done, then a good approach is through the protuberant bulge on the ulnar side of the wrist joint. I usually excise this redundant skin and outline my incision in an elliptical fashion but incise only the distal border at the beginning of the operation. The dorsal and palmar apices of the ellipse are placed over the center of the carpus.

The incision is developed down to the level of the tendons on both the dorsal and palmar surfaces. In doing this, I like to identify the ulnar nerve, the ulnar artery, and its venae comitans. With these structures localized and protected, I feel more comfortable with the further dissection that is necessary among all the aberrant structures on the radial side of the wrist.

The advantage of this ulnar approach is that the radial dissection only has to be extended until the distal end of the ulna is delivered into the wound and the proximal curved surface of the carpus is mobilized. Despite this advantage I make it a practice to identify the median nerve and make sure that its branches, which spread onto the dorsal and palmar surfaces of the hand, are free of tension when the carpus is corrected.

The distal end of the ulna is usually covered with a thickened false joint capsule that should be carefully dissected up and left attached on the dorsoulnar side so that it can be sewn back over the top of the ulna after it has been implanted. Often aberrant insertions of wrist tendons are found joining this capsular tissue. The epiphyseal end of the ulna is a firm oval-shaped mass of tissue in which it may be hard to identify the plane of the epiphysis. This mass has to be trimmed into a squared-off end, and this should be done by gentle paring with a scalpel; osteotomes and other impact tools may damage epiphyseal growth. When the size of the distal ulna has been established, the carpus is brought into the corrected position and score marks are made on the carpal bones around the end of the ulna. These marks define the sides of the slot that will have to be cut in the lunate and capitate. Lamb has stressed that to obtain a secure union the sides of the carpal slot must be as long as the width of the ulna. Ideally, the carpus should be positioned on the distal ulna in such a fashion that the line of the third metacarpal is perpendicular to the growth plate. If the end of the ulna extends more distally and lies over the whole capitate or even the base of the metacarpal, further soft tissue release will have to be done until the proper amount of distraction is obtained.

The next thing to be decided is the orientation of the hand in relation to the forearm. The single forearm bone precludes pronation and supination, wrist flexion and extension, and even lateral deviation. Assuming that reasonable
shoulder and elbow motions are present, the hand should be placed in a position about halfway between neutral and full pronation. Thus the slot in the carpus is not necessarily cut in a plane at right angles to the dorsal surface of the carpus.

After the ulna and the carpal slot have been properly matched, the position is maintained by driving a K wire through the medullary canal of the long or ring finger metacarpal, across the capitate, into the center of the squared-off distal surface of the ulna, and up into the main shaft of the bone (Fig. 16-24).

The size of the K wire used is subject to some controversy. Delorme recommends using the largest wire or rod that the medullary canal of the metacarpal will accept, but most surgeons use a smaller diameter wire. The diameter of wire selected must vary with the size of the patient's hand, but in children under the age of 1 year I have found a .045- or .062-inch diameter satisfactory. It is important that both ends of the wire be pointed. Long K wires that are pointed at both ends may be hard to come by, and I usually sharpen them myself before surgery. Be careful to avoid making a flattened spear end on the

Fig. 16-24. Implantation and K-wire pinning. After the ulna has been slotted into the carpus, the position is maintained by a K wire passing through the ulna, the capitate, and the long or ring finger metacarpal. The lower illustration shows that it is not always easy to transfix a metacarpal down its medullary shaft.
The hand should be held with the fingers flexed at the metacarpophalangeal joints while the wire is passed retrograde through the remnants of the capitate, into the base of the metacarpal, through its shaft, and out through the skin.

The wire can be felt as it penetrates the metacarpal head, and the overlying extensor tendon is pushed to one side before the wire end is brought out through the skin. The drill chuck is now replaced on the end protruding through the metacarpal and the wire withdrawn until its tip just shows in the proximal surface of the capitate. After the elbow has been flexed, the tip of the wire is then inserted into the exact center of the ulna and passed up into the ulna to eventually exit either along the subcutaneous border of the bone or through the olecranon. The chuck is once again removed and placed on the proximal wire end so that the wire may be withdrawn proximally until the long finger metacarpophalangeal joint can be freely moved passively without grating on the wire still protruding through the metacarpal head.

Once the wrist is stabilized, the hand position must be checked again to ensure that its orientation is the best for the shoulder and elbow motion available in the limb. When the proper position has been obtained, I often place several fine absorbable sutures into the dorsal fascia of the ulna and sides of the carpal slot to get good apposition of the cut cartilaginous surfaces. Sutures can be safely passed through the cartilage if this is necessary to snug up the cut surfaces. The soft tissue flap is then sutured over the carpal slot and the end of the ulna.

Any necessary tendon transfers should now be done before the skin is closed. I usually shorten the effective length of the extensor carpi ulnaris by lifting and reattaching it to the base of the fifth metacarpal. I usually move the origins of the hypothenar muscles more proximally onto the ulna and also bring the pisiform and flexor carpi ulnaris more proximally onto the ulnar, rather than the flexor, aspect of the forearm.

Now that the hand is corrected on the forearm, the extent of resection of the redundant ulnar skin can be readily judged. Some degree of absorption will occur, and the amount excised does not have to be so great as to make tension on the skin edges. Interrupted fine catgut sutures should be used, and I do not usually use a drain.

Postoperative care. There is little motion in these tiny fingers, and postoperative swelling to a marked degree is common, particularly after the ellipse of ulnar skin is excised. Because of this, I often apply a firm compression dressing of Dacron fluff and Kling bandage and elevate the limb. A posterior gutter splint of plaster of Paris is almost always necessary to maintain elbow flexion after traction is applied. The flexed position is more comfortable for the child and is needed to take tension off the radial soft tissue structures, particularly the median nerve. The swelling is usually gone within a week, at which time a complete plaster cast is applied with the elbow held in flexion. Since
The Hand

Fig. 16-25. Recurrent radial deviation. A, This child was operated on at the age of 3 years; the intermetacarpal ligament between index and long finger metacarpals was resected, and the ulna was implanted in the carpus. B, Six years later the space between the index and long fingers was considerably wider and some radial deviation had recurred at the level of the carpometacarpal joints. No tendon transfers were done in this hand.

catgut sutures have been used in the skin, this cast can be safely left on for about 5 or 6 weeks.

Long-term results. I have used this procedure for years and am generally pleased with the results. I became more satisfied as my experience increased and I ceased to expect perfection. Problems occur but do not negate the general improvement gained by the child.

Radial deviation does recur, more particularly at the carpometacarpal joints and sometimes at the intercarpal joints (Fig. 16-25).

On the whole, the additional motion caused by the recurrence does not cause loss of function, although it does detract from the appearance of the limb. Premature arrest of the distal ulnar epiphysis has certainly occurred in some of my patients, and I am sure that the same problem has been seen by all surgeons practicing implantation of the ulna. I am not persuaded that occasional early epiphyseal arrest in an area of unpredictable growth potential is too high a price to pay for satisfactory hand stability.

Stability of the hand provides the greatest potential for digital movement, and early stabilization should allow the maximum development of grasp in partially stiff hands. Follow-up for long periods of time is possible under the University of Iowa records system, and representative results are shown in Figs. 16-26 through 16-28.
Fears; the t, and the and long zel of the t on for enerally tcreased general al joints does not e limb. some of surgeons al early to high a movement, grasp in der the in Figs.

Fig. 16-26. *Ulna implantation—long-term result.* A, At birth. B, Age 6 years, before ulna implantation by Dr. A. Steindler. C, Age 13 years. D and E, In adult life full stabilization and excellent grip have developed.
Fig. 16-27. Ulna implantation—long-term result. Unilateral implantation patient followed up into adulthood. A, Before and after implantation at age 5 years. B, X-ray film 12 years after implantation showing vestigial proximal radius and solid union of carpus and ulna. C and D, Photographs 43 years later showing hypoplasia, as compared with the normal hand, and slight recurrence of radial deviation.
Followed up 2 years after.

Fig. 16-28. Bilateral ulna implantation followed up into adulthood. A and B, Before implantation at age 10 years. C and D, Result at age 15 years. This patient was seen at age 37 but declined to be photographed; her stabilization remains sound.
I have no doubt that arthrodesis of the wrist produces a happier individual with improved hand function as compared with the preoperative state. It is impossible to measure attitudes with any accuracy, and I can only report that I believe the brave front put on by the unoperated individual routinely conceals a sad soul who resents the very visible deformity. I will continue to recommend surgical stabilization for suitable patients.

The indications have been greatly reduced by recently introduced operations which provide some wrist motion. Implantation should now be reserved for the acutely distorted wrist, which does not yield full correction after soft tissue releases. The other indication is its use as a fall-back position when centralization has failed to produce satisfactory stability and motion. This is not common, but some long-term studies are showing a disturbing tendency for increasing palmar dislocation of the hand on the ulna.

Centralization. The purpose of centralization without arthrodesis is to stabilize the hand on the ulna and yet provide motion—a lofty ideal yet hard to achieve. Compromise is necessary. The operation does not have to produce a range of motion comparable to that of a normal wrist. In the normal individual the usual useful range of motion is only 10° of flexion, 35° of extension, and 10° of radial and 14° of ulnar deviation. These figures are comparable with the published figures of those reporting on this procedure.

"Recurrence" of radial deviation is quoted as a complication in some reports. Academically this may be correct, but radial deviation is a necessary motion in many actions, and the hand does not have to always sit straight in the line of the ulna.

The operative details vary among the descriptions of Bora, Buck-Gramcko, and Bayne, but a principle common to all is the need for tendon transfers to balance the hand on the single-bone forearm. Some remove carpal bones to allow repositioning of the hand; others try not to remove any bone but trim the cartilage interfaces at the wrist.

All agree that there must be adequate soft tissue release done either as a preliminary stage or at the time of operation. Another important feature of all these procedures is a careful reefing of the capsular tissues. I have referenced these authors' detailed descriptions and offer here a few additional comments.

I believe two skin incisions are necessary: a Z-plasty on the tight radial side and a transverse wedge incision over the prominent distal ulna. The former incision supplies skin lengthening but may not be needed if a preliminary release has already been done. The latter incision gives a good exposure to the ulnar side of the wrist and allows removal of excess fibrofatty tissue and skin.

A distally based capsular flap containing the ulnar collateral ligament should be developed when exposing the ulna, and the blood supply to its epiphysis should be preserved. Be very careful to protect the ulnar nerve, its dorsal cutaneous branch, and particularly the ulnar artery because the radial artery is often absent. The extensor retinaculum should be released; usually the radial extrinsic extensors and flexors are found to have a common muscle mass, and their insertions must be released from the carpal bones.
Radial Clubhand

When exploring through the Z-plasty, beware of the white, cordlike structure tenting up under the deep fascia—it is the median nerve. Occasionally in these more severely affected limbs a cartilaginous “anlage” will be found and must be excised. In my experience an “anlage” is found more commonly in ulnar “clubhands” than in these patients. The palmar capsule usually needs to be released, but after this the hand should be free enough to be placed on the end of the ulna. A slight shaving of the cartilage on the end of the ulna and on the opposing carpal surface to produce a plane joint is sensible.

Buck-Gramcko, who published his concept of radialization in 1985, is more radical in placing the ulna in the line of the second metacarpal; others choose the third metacarpal as the line for the ulna. The value of Buck-Gramcko’s procedure is that it increases the moment arm or leverage of the ulnar transposed muscles. A strong, double-pointed K wire is placed retrograde through the metacarpal, brought out through its head, and then down proximally through the center of the ulnar epiphysis and up into the ulnar shaft. If the ulna is straight, the K wire should come out at the olecranon; in a curved ulna the K wire will appear at about its midpoint. It should then be withdrawn from the ulna until the second metacarpophalangeal joint is freely movable. An x-ray check of the K-wire’s position is wise. The most common cause of a poor result is a misplaced wire and inadequate reduction.

Stabilization is achieved by reinforcing the supports of the ulnar side of the hand. The released radial flexors and extensors should be reattached to the ulnar carpus. The extensor carpi ulnaris should be placed back in its sheath on the ulna and reattached more distally on the shaft of the fifth metacarpal; the flexor carpi ulnaris can also be moved. The hypothenar muscle mass can also be detached from its origin and reattached to the ulna if it can reach, which is unlikely unless carpal bones have been removed. The distally based capsule must be firmly reattached.

Bora supplements these procedures by taking the superficialis tendons of the long and ring fingers, passing them subcutaneously around the ulnar border of the forearm, wrapping them around the index and long metacarpal shafts, and sewing the ends back to their tendons.

The operation is completed by skin closure and application of a padded cast from palm to axilla with the elbow flexed. In general the K wire is removed at about 6 to 9 weeks, and a short arm plastic splint which allows freedom for the fingers is made. I believe this should be worn long term, certainly up to about the age of 6 to protect the centralization during growth. After this it should be used as a night splint.

Long-term results. Not all these procedures are successful, but the number of complications is small and the degree not severe. The range of motion achieved is in the useful range; Bora’s series averages out at about 26°. Buck-Gramcko quotes a range of “up to 20 to 30 degrees of extension,” and Bayne tells me he is pleased with a range of 30° to 40°.

When a well-balanced hand uses this range during growth, the distal ulna tends to broaden, giving more support to the carpus; I have seen some that
almost resemble a distal radius. Some radial deviation does occur, both in arthrodesis and in centralization, some of it may be an actual recurrence, but the deviation most often occurs at the carpometacarpal joints.

The best results are obtained in children operated on before the age of 3; when done between the ages of 5 and 10, the results are not as satisfactory. The quality of the result is significantly dependent on the ability of the transferred muscles to maintain the centralization; if they are weak and of poor quality, recurrence is more likely.

When bowing of the ulna continues, repeated osteotomies may be necessary; preferably they should be done in the distal third since healing is better. Osteotomy should be delayed as long as possible; if done near the time of skeletal maturity, it will last.

PROVISION OF A THUMB

All combinations of defects may occur in the thumb in cases of radial dysplasia. Sixty-five percent of those patients with radial agenesis have no thumb, and in another 10% the thumb is so small as to be useless [Fig. 16-29]. When a reasonably sized thumb is present but weak and of little use, its power of opposition can be improved by an abductor digiti minimi opponensplasty. Manske and McCarroll reported 21 of these procedures in patients with radial deficiencies. There are three indications for this transfer: weak opposition, isolated thenar aplasia, and thenar aplasia with other anomalies.

In 20 of the 21 operations the transferred abductor digiti minimi muscle pulled the thumb into opposition between 6 and 8 weeks after the operation. Dexterity, strength, and usefulness of the thumb as well as its appearance were improved. The one failure was due to a postoperative flexion contracture caused by multiple factors. The technique and pitfalls of this operation are well described in the published article.

When the thumb is useless or absent, there is a tendency for the index finger to undergo spontaneous pollicization in the sense that it tends to move into abduction away from the rest of the hand and show varying degrees of pronation. It therefore seems logical to complete the process and formally construct a thumb out of the index finger.

The concept is admirable, execution is often difficult. The actual transfer of the index on a neurovascular pedicle is a well-established procedure with a high rate of success. The lack of a radial digital artery is not vital. Transfer on the ulnar digital artery alone is possible and satisfactory. The problem in achieving a good functional result is in providing a carpometacarpal joint and in substituting for the absent extrinsic and intrinsic thenar muscles.

Establishing the correct indications for pollicization in these patients is difficult. There is little doubt that in bilateral thumb aplasia pollicization on at least one side can significantly increase functional capacity. Index transfer in unilateral lesions and in the second limb in a bilateral case is less clearly indicated. Eaton has commented that boys, in order to cope with the challenge of
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occupational competition, have a somewhat greater need when the condition is unilateral. The scars, however minimal they may be, tend to somewhat discourage pollicization in girls with unilateral aplasia. I personally do not consider this a significant contraindication because when the operation is carefully performed, gratifying functional and cosmetic results can be obtained.

I have found that in bilateral cases most parents request that the second pollicization be performed but that in unilateral cases they are usually not enthusiastic. I do not press the issue in these cases but have on occasion introduced them to patients (and their parents) with bilateral pollicizations so that they can compare functional abilities. The theoretical objections to bilateral pollicization are the risk of loss of a digit and the narrowing of the palm. The former is a real possibility but rare, and the latter is more apparent than significant. Breadth of the hand is always important, but these patients are unlikely to adopt heavy manual occupations, and dexterity is probably more important to them.

Fig. 16-29. The thumb in radial aplasia. A, Absence of the thumb and deformity of the index. B, Hypoplastic thumb and a well-developed space between index and long fingers used for grasp. C, The rudimentary thumb is attached by a skin pedicle and has no voluntary motion. Note how the border fingers have rotated in an attempt to provide some degree of opposition. (See Credits.)
In patients with fixed Type IV hands, pollicization of the most ulnar digit can be of use. Over 20 years ago Harrison reported such a case and tells me that he has subsequently done further cases. Wood reported a similar operation on a child with the VATER complex in 1988. I have never done this operation, but I believe that any procedure that will improve function in these severely handicapped children is appropriate. It is essential to use the anatomical parts available to the child’s best advantage, even if it means supplementing function on the ulnar rather than the radial side of the hand.

The indications for this operation are narrow. It is best used in a child with bilateral Type IV deficiencies in whom the wrists are fixed in severe flexion and radial deviation, the thumbs are absent, the humeri are short, and the elbows stiff in extension. The technique is essentially a reversal of the usual index finger pollicization, but Wood’s article contains some excellent pointers on the steps of the operation.