Radial Club Hand

A CONTINUING STUDY OF SIXTY-EIGHT PATIENTS WITH ONE HUNDRED AND SEVENTEEN CLUB HANDS

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ABSTRACT: Based on a review of the embryology, genetics, and anatomy of radial club hand, it is suggested that damage to the apical ectoderm on the anterior aspect of a developing limb bud leads to the deformity. Study of the families of thirty-five children with radial club hand suggested that the condition is not genetically patterned. The anatomical findings and associated congenital abnormalities in the cases known to be related to thalidomide and in those in which thalidomide was not a factor were similar except that the incidence of other skeletal deficiencies was higher in the thalidomide group. Thirty-one of the 117 radial club-hand deformities (in sixty-eight patients) under my personal supervision were treated by centralization of the carpus on the ulna with satisfactory improvement of the deformity. In three cases wrist deformity recurred mainly in a volar direction, apparently the result of muscle imbalance. No significant impairment of ulnar growth occurred and straightening of the wrist did not affect function adversely. Pollicization of the index finger was done on twenty-eight occasions. Although problems developed in the early cases, these can be avoided using the methods described and the operation can improve both function and appearance. A scheme of management is recommended.

Radial club hand has been described under at least twelve different names since 1733, but the one used here is practical and descriptive of the unsightly deformity which is so difficult to control. Pulvertaft in his Watson-Jones Lecture to The British Orthopaedic Association in 1967, prefaced his remarks by stating that over the previous ten years no subject in hand surgery had so exercised his judgment or caused him more concern in management.

This report is based on experiences over the past fifteen years with sixty-eight patients, whose average age at the time of review was twelve years (range, three to sixteen years). Three children had died in infancy: two soon after birth and one at the age of three years. Most of these children were from Scotland, which has a population of five million, but nearly one-third came from other parts of the British Isles. Since 1959, they have been part of a continuous program of supervision and review. With the exception of the three children who died and four who were living overseas, all were reviewed by me in August 1974. At the same time, an occupational therapist, who had been involved in their management for many years, carried out a final functional assessment of activities of daily living. All children had been seen regularly: at intervals of three months or less when operative or other treatment had been instituted and at intervals of not more than six months when active treatment was not considered advisable. Many of these children had been under observation for ten to twelve years and none for less than a year since treatment was started. The vast majority had been seen regularly over periods ranging from six to ten years.

Embryology

Numerous theories of etiology have been propounded over the past 100 years, but few stand up to critical evaluation. The work of Saunders suggested the most probable cause of this deformity. By removing part of the apical ectodermal ridge in the developing wings of chick embryos, he produced anomalies similar to the human deformity. His work confirmed that the limb develops in an orderly proximal-to-distal sequence. Warren had previously shown that excision of the anterior half of the upper limb
bud suppressed the growth of the proximal and anterior part of the humerus and of the preaxial border of the forearm. Saunders' more selective local excision of the apical ectoderm alone produced the same effect. Unless there is an intact, healthy apical ectoderm, differentiation of the underlying mesenchymal tissue does not take place.

Genetics

Birch-Jensen found the frequency of this defect to be one in 30,000 births. Warkany stated that the mortality rate of infants with radial defects at birth is high, and that the children who do live have a high mortality rate up to the age of twenty. While I have no information relative to the first statement, the second certainly proved not to be correct in this series. Two patients died in the early weeks of life; both were found to have severe ventricular septal defects and other cardiac lesions as well as agenesis of one kidney and hydrenephrosis of the other. A third child died at the age of three years following surgery for tetralogy of Fallot. To date, all other patients remain alive and well. In most instances this condition is not genetically determined, although a few such sporadic cases were reported by various authors. Wynne-Davies reviewed the cases of thirty-five children with radial club hand from my clinic and found that in twelve of them the mother was known to have taken thalidomide. In all thirty-five cases the family was investigated to at least the third-degree relatives and in no case was the condition found to be genetically determined. The seventy parents, seventy-one siblings, 380 second-degree relatives, and 443 third-degree relatives were all normal.

Considering the entire series, twenty-six of the sixty-eight mothers had ingested thalidomide and fourteen had taken some unidentified medication during the early months of pregnancy, leaving twenty-eight in whom there was no known significant etiological factor and whose pregnancies had been entirely normal.

Absence of the radius is also found in association with the Holt-Oram syndrome (atrial septal defect), which is of dominant inheritance; with Fanconi pancytopenia (of recessive inheritance); and with the chromosomal abnormality trisomy 18. None of these conditions was represented in the present series.

Associated Anomalies

Radial club hand has been described in association with almost every possible congenital deformity. This is hardly surprising, since many organs develop at the same time as the upper limb buds. In a particular patient, the important thing is to be aware of this association and to exclude any severe anomalies that would make surgery inadvisable. Other anomalies were found in seventeen (25 per cent) of the sixty-eight patients in this series: in seven associated with thalidomide and in ten with no known clues as to etiology. The lesions of the genito-urinary system and even of the heart (especially patent ductus arteriosus) were often asymptomatic.

In 1973, Gillespie reviewed fifty-four of the children in this series to determine whether the type of radial club hand and the frequency of associated anomalies differed in the patients whose lesions were known to be the result of thalidomide (twenty-six patients) as opposed to the group in which there were no known clues as to etiology (twenty-eight patients). In the thalidomide group, seven of the twenty-six patients showed longitudinal deficiencies in the limbs, including varying degrees of dysplasia of the proximal end of the humerus as well as bilateral gross deficiency of the femur and absence of the tibia in four of these seven patients. The associated anomalies in the thalidomide group, eleven in all, were: patent ductus arteriosus, one; imperforate anus, two; esophageal atresia, one; absent external ears, one; nerve deafness, one; congenital scoliosis, one; and idiopathic scoliosis, four.

In the non-thalidomide or idiopathic group of twenty-eight patients, there were no other limb deficiencies, but ten of them had a total of sixteen associated anomalies: ventricular septal defect, four; tetralogy of Fallot, one; coarctation of the aorta, one; patent ductus arteriosus, one; right renal agenesis and left hydrenephrosis, two (these two children also had ventricular septal defects and died in early infancy); congenital scoliosis, one; idiopathic scoliosis, three; agenesis of the upper lobe of one lung, one; and a pedunculated encephalocele with communicating hydrocephalus, two.

Anatomy

The anatomy of radial club hand has been described and reviewed by many authors as a usual abnormality, but in this series the ulna was nearly normal, being normally situated but small.

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mity due to soft-tissue contracture at the deformed wrist. Similarly, a fibrous anlage of the radius is often described as being present but this was found at operation in only two of the thirty-one forearms operated on in this series.

It must be appreciated that this is not a simple skeletal deficiency on the preaxial side of the extremity. There are also abnormalities of muscles, nerves, and joints. A common finding is absence or deficiency of the muscles arising from the common extensor origin (especially the extensor carpi radialis longus and brevis) and from the radius. The most important muscle in this group, which arises normally from the volar aspect of the radius, is the index profundus. Significant deficiency or absence of this muscle may influence the decision regarding subsequent pollicization of the index finger.

The radial side of the carpus is nearly always deficient. In the present series, the scaphoid was absent in 98 per cent and the trapezium, in 100 per cent of the 117 wrists. In O'Rahilly's series the scaphoid was absent in 82 per cent. As the wrist approaches skeletal maturity, various intercarpal fusions commonly become evident, particularly fusion of the trapezoid to the capitate.

Function of the joints of the hand is always deficient, the defect ranging from complete stiffness of the joint similar to that in severe arthrogryposis to minor stiffness with almost normal movements. In most cases the defect is somewhere between these extremes, the severity of involvement becoming progressively less going from the radial to the ulnar side of the hand.

Growth and Function of the Limb

Before surgical treatment is contemplated, it is necessary to know how the affected limb will develop and the function it will achieve.

To determine the growth pattern of the forearm in this condition, growth in length of the ulna was measured on serial roentgenograms of twenty-two forearms not treated surgically and on similar roentgenograms of fifteen forearms treated by centralization of the carpus on the ulna.

These measurements were made on the roentgenograms with a measuring rod marked off in centimeters. The two groups were comparable with respect to age and type of deformity. Six of these patients had bilateral deformity and operation had been done only on one side, so that the two sides could be compared. The length of the ulna was found to range from forty to fifty millimeters at birth and from 100 to 140 millimeters at twelve years of age, or one-half to two-thirds as long as the normal ulna of a twelve-year-old child. In the limbs in which the humerus was normal, the opportunity was also taken to compare the length of the humerus with that of the ulna during the various stages of growth. The normal ratio of the length of the humerus to that of the ulna is about 1:1. In the cases of an absent radius with a normal humerus, the ratio of the humerus to the ulna was 3:2, or even as high as 2:1. Thus, even in untreated limbs the ulna is much shorter than normal. The degree of shortness of the forearm was the same in Heikkel's series as in this one, and he noted that the distal ulnar epiphysis appeared later and closed earlier than in normal children. If no attempt is made to control the deformity during growth, the curvature of the ulna increases, and with it the apparent shortening of the forearm.

Since the ulna will be so short anyway, it becomes somewhat easier to recommend surgical procedures which might interfere with growth of the distal ulnar epiphysis. Placing the hand centrally over the end of the ulna improves the appearance and increases the apparent length of the forearm considerably.

The function of the limb and hand is, of course, of paramount importance, and in this series meticulous functional assessments were carried out by experienced occupational therapists. All functions of daily living were accurately assessed and the ranges of movement of all the joints of the upper limb were recorded, as well as any changes that occurred during development.

Since the structure and function of the digits are much poorer on the radial than on the ulnar side of these hands, the radial deviation has the definite advantage that the radial border of the hand is positioned so that it is towards objects to be manipulated (Fig. 1). This advantage must be weighed against the functional gains achieved when the wrist is straightened. In the present study, the therapist charted function both with the wrist in the deviated position and with it splinted as straight as possible. In those cases in which the wrist could be corrected fully by a splint, there was no loss of function with the wrist straight and in many instances function was improved.

The change of position of these wrists had little effect on the range of movement of the joints of the hand, and it is assumed that the improvement was due to the gain in stability of the wrist and the improved line of pull of the long tendons.

So-called ulnar-oriented hands. The more functional ring and little fingers are used for prehension. One of these twins has no elbow flexion, both elbows being held stiffly in extension, while the other child has normal elbow mobility. Since these boys with bilateral radial aplasia were first seen at the age of seven years and function was quite good, no operation was considered.
The assessment of function was based on simple functional tests, including self-care activities in dressing, toilet, washing, and feeding, and on a number of simple manipulative tasks to test the function of the hand by itself and in bimanual activities. A system of points was used to assess the functional capacity of each child on a percentage basis (100 per cent being normal function). In the patients with a unilateral lesion there was no impairment of function, but in those with a bilateral lesion there was considerable difficulty in fastening buttons, cutting meat, combing hair, and putting on socks, although most self-care activities were possible. The ability to be independent in toilet care depended primarily on the length of the limb. Based on this point system, the functional assessments in the patients with a bilateral lesion ranged from 46 to 95 per cent, with an average of 73 per cent.

The ranges of movement of the joints of the fingers were measured and recorded and, as already stated, there was considerable diversity in the amount of motion. Typically, the metacarpophalangeal joints had excessive hyperextension and limited flexion. Metacarpophalangeal-joint flexion ranged from 0 to 75 degrees (average, 36 degrees) in the index finger, from 0 to 80 degrees (average, 35 degrees) in the long finger, from 0 to 85 degrees (average, 42 degrees) in the ring finger, and from 0 to 90 degrees (average, 52 degrees) in the little finger.

The proximal interphalangeal joints had fixed flexion deformities, some of them associated with skin webbing. In the fifty-four patients whose proximal interphalangeal joints were examined carefully, the average flexion contractures were: in the index finger, 29 degrees; in the long finger, 37 degrees; in the ring finger, 25 degrees; and in the little finger, 8 degrees. In all fingers there was marked restriction of motion and the amount of motion tended to increase progressively from the index to the little finger. Active flexion ranged from 0 to 53 degrees (average, 24 degrees) in the index finger; from 0 to 72 degrees (average, 35 degrees) in the long finger; from 0 to 82 degrees (average, 57 degrees) in the ring finger; and from 0 to 86 degrees (average, 78 degrees) in the little finger.

The roentgenographic appearance of these joints was normal and the few joints explored at surgery also had normal configurations. The limitation of motion, therefore, appeared to be related to extra-articular causes. This impression was reinforced by the finding that in all fingers with sufficient mobility of the proximal interphalangeal joints to permit testing, the flexion of these joints increased as the metacarpophalangeal joint was hyperextended (Figs. 2-A and 2-B). This finding suggested that there was some abnormality of the extensor mechanism.

Instability of the wrist, a regular feature of untreated radial club hand, was associated with the development of two deformities, one or the other predominating. The first of these, radial displacement, is well known and is seen when progressive displacement of the carpus and increasing soft-tissue contracture are allowed to develop. The second, less well recognized deformity is volar displacement of the carpus (Figs. 3-A and 3-B); it is caused partly by the unopposed action of the wrist flexors, notably the flexor carpi radialis, which is the dominant deforming influence, and partly by the finger flexors, particularly the superficialis, which span the unstable wrist and flex it so that much of the power of finger flexion is lost.

Stiffness of the elbow in extension is frequently associated with radial club hand and was present in twenty-seven (23 per cent) of the 117 elbows. The presence of a stiff elbow in extension is an important consideration. If the stiffness cannot be corrected it is a definite contraindication to any surgical correction of the wrist deformity. It is difficult to ascertain what factors contribute to this limitation of movement. Certainly, the weak elbow flexors are...
overwhelmed by the powerful elbow extensors, which are nearly always normal. Roentgenograms of the elbow are of little help in a young child, but as the child grows older, abnormalities of the structure of the elbow joint may become visible.

The wrists of the limbs with stiff elbows were splinted as straight as possible, so that active elbow flexion to bring the hand to the face was encouraged. By this maneuver, the range of active elbow flexion improved to 90 degrees in twenty of the twenty-seven elbows. It was found that elbow flexion could improve over a period of more than a year with normal activities if the wrist deformity was controlled by splinting.

In one girl with bilateral radial aplasia and both elbows fixed in extension, the right elbow was mobilized so that it gained 90 degrees of active flexion over a period of two and a half years, following which centralization was carried out, while the left elbow only gained 40 degrees of active flexion and hence centralization was not performed. In only three of the remaining seven limbs in which the elbow remained stiff in extension or gained flexion to only 40 degrees or less was function of the hand considered good enough to attempt to improve active elbow flexion by a posterior capsular release and transfer of the triceps to the front of the elbow. In two of these three limbs, the procedure proved satisfactory. In the first, a boy eight years old whose completely stiff elbow was fixed in extension, active flexion increased to 90 degrees and remained at this level four years later. In the second, a girl whose elbow was completely stiff in extension before operation was done at the age of six years, active flexion increased after operation to 45 degrees at one year and to 90 degrees at eighteen months. Seven years after operation, the elbow still flexed to 90 degrees. In the third child, 30 degrees of active flexion was present after operation, but seven years later her elbow was again stiff in extension.

Treatment

There is considerable difference of opinion as to whether any treatment is justified, perhaps because most surgeons have limited personal experience in this area and their opinions are based on series collected from the experience of many surgeons using a variety of procedures, some of them ill conceived. For many years, textbooks also compounded the difficulties by describing several operations, most of them tried on only a few occasions and not evaluated by adequate follow-up.

Four general methods of surgical correction have been described over the past eighty-seven years (Fig. 4).

1. Soft-tissue releases with or without ulnar osteotomy are done to correct existing deformity. They are also performed as a preliminary to other types of surgery. By themselves, however, release and osteotomy produce only temporary improvement and relapse is inevitable. Before 1900, several types of ulnar osteotomy designed to give permanent correction were proposed by Hoffa, Sayre, Bardenheuer, and Romano, but they all proved ineffective.

![Fig. 3-A and 3-B: This seven-year-old boy had a volar-displacement deformity at the wrist. The elbow had very restricted movement and operation on this wrist was not indicated. Fig. 3-A: Normal position of wrist at rest. Fig. 3-B: Maximum active dorsiflexion.](image)

![Diagrammatic review of the operative procedures on bone described since 1890 for radial club hand.](image)
2. Replacement of the missing radius by a bone graft was recommended by Albee, who used the tibia; by Ryerson, who used the ulna; and by Starr and Roried, who used the proximal fibular epiphysis. These procedures also failed to maintain alignment.

3. Centralization of the hand over the ulna was used by Sayre, who sharpened the distal end of the ulna to fit into a carpal notch. This technique is now considered inadvisable and the method described by Lidge is recommended.

4. Arthrodesis of the wrist, usually reserved as the final definitive procedure after cessation of growth, is done to correct the deformity and to stabilize the wrist, with the realization that mobility in the carpal area will be lost. While this procedure does correct the ugly deformity, it should not be performed before growth of the ulnar epiphysis has ceased. The main disadvantage of this procedure is that it is carried out after the age of twelve and completely changes the pattern of prehension that the child has been using for many years. While arthrodesis may be indicated in a patient with a previously untreated unilateral deformity, it is not recommended for the treatment of bilateral deformity. The results of arthrodesis in bilateral cases compare unfavorably with those of centralization of the carpus, which can be carried out at an early age, allowing a satisfactory pattern of prehension to develop with the deformity corrected and the wrist stabilized.

These different procedures, for the most part, have been used only in a limited way by any one surgeon. Also, in the cases reported in the literature the follow-ups were often poor, the rates of recurrence of the deformity were considerable, and the function of the hands after operation was seldom described adequately. Another procedure, described more recently (Fig. 4), which involves subperiosteal dissection of the distal end of the ulna and displacement of the ulna to the radial side of the carpus, can, I believe, be expected to cause quick cessation of ulnar growth. I have therefore not considered it.

Although the bone-replacement procedures are theoretically very attractive, particularly the transfer of the growing proximal fibular epiphysis, they have proved to be ineffective and should be abandoned. Carroll followed eighteen patients with a fibular transfer for an average of eighteen patients with a fibular transfer for an average of

3. Function may be seriously compromised by straightening the wrist, with the inevitable associated stiffness.

Such a functional loss was not verified in this series, provided that prior to centralization the patient had active finger flexion to at least 90 degrees and a usefully functioning hand, an observation that applied in both unilateral and bilateral cases.

In the present series, 107 operations were performed on sixty-eight patients with 117 deformed extremities. These procedures could be divided into three general categories: correction of wrist deformity, sixty-eight; procedures to mobilize stiff joints; five; and operations to improve thumb function or to eliminate a useless thumb remnant, thirty-four.

The sixty-eight procedures to correct wrist deformity included thirty-one centralizations, which are discussed later in this report, and thirty-seven other procedures, as follows: There were seventeen soft-tissue releases and nine ulnar osteotomies, of which two releases and five os-
teotomies were performed preliminary to, in association with, or subsequent to centralization. The other fifteen releases and four osteotomies were performed as isolated procedures and all were followed by recurrence. Three fibular transplants were also followed by recurrence of deformity.

Five tendon transfers (transfer of the flexor carpi radialis to the dorsum of the carpus) were done to eliminate a strong deforming factor, two such transfers being performed at the time of centralization. In two other centralization procedures, after the wrist deformity was corrected the fingers were acutely flexed because of tightness of the superficialis muscle, and this muscle had to be lengthened at its musculotendinous junction. In only one patient was arthrodesis used to correct the wrist deformity.

Five procedures were performed to mobilize stiff joints, three at the elbow and two at finger joints. The procedures at the elbow included a posterior capsulotomy and transfer of the triceps tendon, as previously described.

In one of the finger operations, an exploration of a stiff metacarpophalangeal joint, the extensor tendon and hood were found to be replaced by a flat sheet of amorphous fibrous tissue which had very limited gliding and appeared to cause the lack of flexion. The collateral ligaments were resected on the chance that they were contributing to the loss of flexion, but following this procedure there was no improvement in the range of joint movement.

In the other finger procedure, an exploration of a contracted proximal interphalangeal joint, no obvious abnormality was found and after the procedure motion of the joint did not improve.

Of the thirty-four operations on the thumb, twenty-eight were index-finger pollicizations (discussed later in this report), three were removals of a useless thumb, and three were reconstructive procedures. The reconstructions were shortening of a pollicized index finger, a tendon transfer to provide opposition, and plastic procedures on a distally placed hypoplastic thumb, including proximal transfer and deepening of the thumb-index finger web space.

**Technique of Centralization**

Two important technical points must be kept in mind. For one of these I am indebted to MacConaghy for his views on the mechanical requirements for successful centralization (Fig. 6).

![Diagram showing the forces (F₁ and F₂) that act after centralization and tend to displace the hand in direction M by rotating it around point X.]

1. To ensure stability of the ulna in the carpus, the depth of the slot in the carpus must equal the diameter of the ulna (Figs. 7-A, 7-B, and 7-C). It is a well recognized engineering principle that mechanical stability is only achieved when the depth of insertion of one component into another is at least equal to the diameter of the inserted component. Using measurements made on a preoperative roentgenogram, it is usually evident that the whole of the
Figs. 7-A, 7-B, and 7-C: Technique of centralization. These drawings were made from roentgenograms of the limb of a child with radial aplasia and, atypically, a thumb.

Fig. 7-A: Position of deformity.

Fig. 7-B: Slot removed from the center of the carpus.

Fig. 7-C: Ulna inserted into the slot in the carpus and held by a Kirschner wire inserted across the center of the distal ulnar epiphysis into the shaft of the third metacarpal. Note that the ulna is inserted to depth A, which is equal to the transverse diameter of the ulna.

capitate must be removed to achieve this degree of stability.

2. The ulna must fit snugly but easily into the carpal defect.

At operation, a sinuous incision is made on the dorsum of the hand; it starts at a distal point on the radial side over the second metacarpal, goes across the hand to a point on the ulnar side near the prominence of the ulnar head, then deviates radially along the dorsal carpal crease, and finally passes around the radial side of the wrist, whence it extends up the volar aspect to the proximal part of the forearm (Figs. 8-A and 8-B). This incision provides an extensive view of the dorsal, radial, and volar aspects of the hand, wrist, and forearm. On the dorsum of the hand and wrist, great care is taken to avoid damage to the veins. As the incision is deepened on the radial side, it must be appreciated that the median nerve is usually lying superficially and radially and should be picked up immediately subjacent to the deep fascia.

In seven of the thirty-one wrists treated by centralization, the median nerve divided into two large terminal di-
visions about seven centimeters proximal to the wrist. The most radial division continued along the radial side of the carpus and hand, presumably supplying the innervation normally provided by the radial nerve, while the other branch passed to the front of the wrist and through the carpal tunnel. In the first wrist in which this anomaly was encountered, the carpal branch was isolated and protected but the radial division was inadvertently divided. This was immediately repaired; postoperatively there was some impairment of sensation in the index finger but sensation subsequently was recovered completely.

Once the median nerve has been isolated and protected, dissection on the radial side is continued, dividing the tight soft-tissue structures and elevating the periosteum and capsule as a distally attached flap from the dorsum of the carpal bones by sharp dissection until the carpus can be visualized. It is often difficult to identify specific carpal bones, particularly in very young children, but it can be assumed that the trapezium and scaphoid are absent.

The distal end of the ulna is identified and dissected out extraperiosteally, starting from the dorsal aspect and doing as little soft-tissue stripping as possible. The size of the distal end of the ulna is assessed and a recess is prepared in the carpus to allow the end of the ulna to be inserted into it without difficulty. If the deformity has not been allowed to progress too far and the distal end of the ulna has not been allowed to grow out of contact with the carpus for too long a time, operative correction of the deformity and placement of the ulna in a carpal notch is usually quite easy, and the carpus is stable on the ulna. The flap of soft tissue and periosteum previously raised from the dorsum of the carpus is closed carefully over the ulna. This may provide sufficient stability but, if not, a Kirschner wire is inserted in a proximal-to-distal direction through the shaft of the third or fourth metacarpal and out through the metacarpal head. It is then drilled proximally through the center of the distal ulnar epiphysis and up the medullary cavity of the ulna. This procedure can be technically difficult but is usually easy when the ulna is not curved. If there is marked curvature of the ulna, osteotomy should be carried out, if need be at multiple levels. If necessary, the superficialis can be lengthened or transferred, and if there appears to be a strong deforming force produced by the flexor carpi radialis it can be transferred to the dorsum of the wrist. At the conclusion of the procedure, the tourniquet is released and hemostasis is obtained. A firm compression bandage is applied with a dorsal plaster slab holding the elbow flexed 90 degrees. Invariably, there is considerable swelling; it is recommended that a complete plaster cast not be applied immediately postoperatively and that the hand be elevated for several days. When the swelling has subsided, which may take up to a week or more partly due to the poor finger movement, a new, complete plaster cast from the metacarpal heads to above the elbow is applied and maintained for six weeks. Following this, if a Kirschner wire has been inserted and is in good position, no other immobilization is required and

The wire can be left in place for several months unless it extrudes (Fig. 9). Should this happen, or if a wire was not used, a carefully molded plastic splint is used for a period of not less than six months after operation.

In twenty-five of the thirty-one centralizations performed in this series, a Kirschner wire was used to maintain the position. In two cases, the wire was left in for just over five years, as it had shown no signs of extruding and did not appear to be impairing the epiphyseal growth. In the remainder, the wire had to be removed because it was backing out either proximally or distally. In most cases, the wire remained in for six to twelve months. In two, the wire broke at the level of the wrist and the distal part of the wire was removed nine and thirteen months after operation, respectively.

Results of Centralization of the Carpus

Thirty-one centralization procedures were carried out on twenty-seven patients, of whom four had a bilateral operation. The age at operation ranged from two to thirteen
years (average, six and one-half years). In August 1974, the duration of follow-up since operation ranged from one to twelve years, with an average of five years.

The average preoperative radial deviation measured 78 degrees (range, 45 to 110 degrees). At follow-up, the average angle of deviation was 22 degrees (range, 0 to 37 degrees).

Considerable attention was paid to the range of elbow movement. In all but one patient, the preoperative range of motion of the elbow had been retained. The loss of motion occurred in the one patient who had less than 90 degrees of active elbow flexion before centralization was carried out. Her preoperative range of elbow flexion was 85 degrees, but after centralization at the age of five her elbow flexed only to 70 degrees. Prior to operation, she had a 90-degree radial club-hand deformity on the non-dominant side, which was corrected to 30 degrees. Following operation, she was unable to reach her face with this hand. In the other twenty-six patients there was a temporary reduction of elbow flexion, but in all of them the preoperative range was regained within a year of operation. In no patient was there any increase in the range of elbow flexion after operation.

As has already been indicated, the range of flexion of the digits is limited. The range of motion in each joint of every digit was assessed and charted in all limbs in which centralization was carried out both before and after operation. To simplify this presentation, these ranges are described as summations of the active flexion of the three joints of each digit before and after operation. The average ranges of flexion in each digit before and after operation were: index finger, 98 and 88 degrees; long finger, 101 and 102 degrees; ring finger, 185 and 155 degrees; and little finger, 210 and 212 degrees. The range of movement was thus not significantly altered except in the ring finger, but that decrease caused no loss of function. The prehension pattern was not changed unless pollicization of the index finger was carried out subsequently.

After three of the centralizations a second operation was required because of increasing deformity, apparently due to muscle imbalance. At the second procedure the flexor carpi radialis was transferred to the dorsum of the carpus, and thereafter satisfactory correction was maintained.

In general, the results in this series indicate that centralization of the carpus should provide good correction of the deformity without loss of function of the limb (Fig. 10). Lidge 19, in the twenty-five children in his series, found that the wrist was stable and retained some useful movement. He thought that the optimum time for operation was two to three years of age. In our series, an opinion as to the quality of the result was sought from the parents and from each child who had a centralization procedure. All patients thought that the operation had improved the appearance of the deformity very satisfactorily and, with one exception, all parents thought that surgery had been of considerable cosmetic benefit. No parent or child had noted any impairment of function following operation, but the general opinion was that any improvement that had occurred was minimum.

In particular, the four patients who had bilateral centralization had no decrease in their self-care functional assessments. One of them, a boy (Fig. 10) who had bilateral centralization and pollicization on the right side, became totally independent in toilet and dressing functions after operation, which had not been the case preoperatively. This finding is contrary to the experience of Goldner and associates 10; in a review of the results of thirty-two centralization operations, they found considerable stiffness of the wrists and advised against doing the procedure bilaterally. A very important point was whether there was any premature cessation of growth of the distal ulnar epiphysis after centralization. The expected ulnar growth pattern, as determined in twenty-two untreated limbs, was compared with the ulnar growth following centralization of the carpus in fifteen limbs (Fig. 5). Eight of these fifteen ulnae were in patients who had the operation between the ages of two and five years. Their growth pattern was normal after centralization. In the other seven patients, whose operations were carried out at a later age (between eight and twelve years), only a little growth occurred at the distal ulnar epiphysis after operation.
In summary, therefore, one may anticipate that after centralization there may be a slight reduction in the range of motion both of the elbow, usually temporary, and of the fingers, but that this loss is seldom of functional significance; that the improvement in appearance will be very acceptable; and that, if the procedure is carried out correctly and supplemented by tendon transfers as necessary to ensure balance, the correction will be maintained. Suppression of growth of the distal ulnar epiphysis appears to be a distinct possibility after operation, but in this series it occurred only when the operation was carried out in older children at a time when growth was about to cease spontaneously.

Pollicization

No discussion of the surgical treatment of radial club hand would be complete without some comment on the role of pollicization. After the wrist deformity has been corrected, consideration should be given to transposition of the index finger if its muscle and joint structures are sufficient to provide good function as a thumb. Many believe that this procedure should not be carried out in cases of unilateral deformity and that in bilateral cases it should be done only on one side, usually the dominant one. I do not share this belief, and think that the function of the hand can be so improved by pollicization that it should be carried out either unilaterally or bilaterally whenever the over-all condition of the limb and hand justifies confidence that functional improvement will be possible. Considerable benefit can be achieved by providing full independence which was previously lacking. Twenty-eight pollicization procedures in twenty-one patients were carried out in this series. The operations were done at ages ranging from one to thirteen years (average, seven years). At the time of review in August 1974, the time from operation ranged from one to twelve years (average, five years). In seven patients, the operation had been carried out on both sides.

The technique used (Figs. 11-A and 11-B) combines the incision described by Barsky, the procedure for the isolation of the neurovascular bundle and transfer of the digit suggested by Littler, and the rearrangement of the first dorsal interosseus to function as the abductor pollicis and of the first volar interosseus to serve as the adductor pollicis proposed by Riordan. The second metacarpal is removed except for its head, which is fixed in the transposed position by soft-tissue suture. In some of the early cases, a hyperextension deformity developed at the base of the new thumb because of the hyperextensibility of the metacarpophalangeal joint often present in these hands, as already noted. If hyperextension develops, it may increase any flexion contracture at the proximal interphalangeal joint of the pollicized index finger, resulting in an unsightly and less effective thumb. This fault was rectified by rotating the metacarpal head so as to tighten the volar capsule.

Particular attention must be paid to any neurovascular anomalies which may be present. It is important to separate the nerve to the second cleft into its component parts which go to the index and long fingers, freeing both components well back to the base of the hand so that the index-finger neurovascular bundle is not stretched or kinked when the digit is transposed. The index finger must be rotated about 150 degrees to get the new thumb into correct position (Fig. 10). The operative procedure converts a three-jointed index finger into a two-jointed thumb, with the head of the metacarpal becoming the new trapezium (Fig. 12). The rearranged intrinsics should be attached to the extensor mechanism in the region of the proximal interphalangeal joint. If there is a good flexor digitorum profundus before transfer, it soon tightens up, and the patient gains excellent flexion. Extensor power is less satisfactory and reinforcement of the intrinsics is an important step in getting the best possible extension. This technique was introduced after the first eight operations...
The second metacarpal is removed apart from the head, which is rotated through 150° and fixed in position by soft tissue suture (and by K-wires if necessary) in order to try to prevent hyperextension. (Buck-Gramcko)

The intrinsics are rearranged and attached to the extensor mechanism at PIPJ level. (Riordan)

Diagram showing how the index finger is transferred into the position of the thumb after removal of all but the head of the second metacarpal.

Recommendations for Over-all Management

Based on the experience gained in this series, the following program is recommended.

1. Splinting is continued from birth. Usually the initial deformity is easily correctable, and the correction should be maintained. When the child is old enough to start using the hands, the splint is worn only at night. If the deformity has become fixed it can often be corrected by gentle corrective splinting using a ratchet-type splint (Figs. 13-A and 13-B).

2. Full activity in all play and other functions is allowed during the day. If the splint is used regularly at night, no contracture will develop. During this period, careful functional assessment is carried out at regular intervals.

3. If there is limited flexion of the elbow, attempts are made to mobilize it, as already described.

4. Usually, at about the age of three to three and one-half years, the carpus starts to displace off the distal end of the growing ulna either in a radial or a volar direction, or, as is usually the case, in a radiovolar direction. At this stage, splinting is no longer effective in controlling the deformity and centralization is carried out utilizing the technique described. After the age of three years, the distal ulnar epiphysis broadens considerably, making the operative procedure more difficult. In the series described

and improved the subsequent results greatly. The rearranged intrinsics also reproduce the appearance of the thenar muscles and make the transposed digit look more like a thumb and less like a finger.

An early fault was to make the transposed digit too long. Not enough of the second metacarpal was removed in the belief, often mentioned in the literature, that the transposed digit does not grow at the same pace as the rest of the hand. Such impairment of growth was not observed in this series. However, any excess length is easily rectified. Experience has shown that removal of all but the head of the second metacarpal provides a thumb of satisfactory length.

Figs. 13-A and 13-B: Ratchet-type splint. First seen at the age of two years, this little girl had had no splinting previously and passive correction could not be obtained beyond the position shown in Figure 13-A. Fig. 13-A: Deformity when the splint was applied.
by MacCon, the later the surgical correction, the poorer the functional result.

5. Postoperative splitting is continued for a minimum of six months (Fig. 14).

6. Careful assessment of function is then carried out to determine the advisability of pollicizing the radial digit. Provided there is active flexion at the terminal joint and the structure of the joints is satisfactory, pollicization can be carried out about six months after centralization.

References


