Congenital Hand Anomalies: Principles of Management

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Congenital anomalies of the hand present as challenges to the hand surgeon. To handle these problems well, the surgeon must analyze the complexity of the problem, set the right priorities, choose the right timing for intervention, apply a multitude of techniques, and plan ahead for rehabilitation. Any reconstruction must not do any harm. Reconstructions have their limitations; either too optimistic or too conservative would bring inferior results. The functional consideration is far more important than structural restoration. With the admirable ability of the small children to adapt to structural defects, one expects reasonable functional achievements even with limited structural restorations. The diversity of problems, the complexity of planning, and the demand for surgical skills in the management of congenital anomalies are well illustrated in two examples, Thumb Deficiency and Radial Club Hand.

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Congenital anomaly of the hand is an area that not every hand surgeon engages in routinely. It depends very much on the circumstantial situation whether the surgeon would have the experience to deal with the congenital problems. How do hand surgeons set priorities when dealing with congenital anomalies of the hand?

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Congenital anomalies were often complicated structural abnormalities so that the question of priority would really deserve close scrutiny. Those who just performed the surgery according to what appeared convenient would not be able to provide the best results. On the contrary, an analytical prioritization before every surgical intervention would help significantly with functional restoration. Because we were dealing with children who presented with hand problems related to structures, there were obviously 3 areas of concern, namely, hand for surgery, children for surgery, and specific problems.

General Principles of Hand Surgery

The relationship between structure and function of the hand is so intimate that in any structural correction of the hand, the justification should be on
a functional restoration, rather than a simple structural correction. In other words, with the exception of pure cosmetic corrections under very special circumstances, functional restoration or improvement overrules structural corrections.

Functions of the hand should not be defined vaguely because they have long been identified and labeled. These functions include 3 different types of grip: power, diagonal, and hook; 3 different types of pinch: tip pinch, side pinch, and palp pinch, fulfilling different demands on physical activities, sensory feeling, and, lastly, expression. All surgical planning should pay full attention to the functional goal to be achieved.

Although the physical ability of the individual has a lot to do with the functional achievement based on intact hand structure, one still has to observe that structural integrity is not everything. By that we are talking about the length and position of the digits, the mobility and stability of the joints, the strength of the components, and the sensibility of the tactile parts. Without a proper length and favorable position, no digit could function. Without stability and mobility, no joint could function. Without reasonable strength, no hand could function properly, and, without sensation, a hand would not be able to protect itself, not to mention precision performance. These components need to be considered together. For example, good length without stability or mobility is meaningless.

The importance of functional considerations in replantation surgery always has been a practical problem of concern. Although replanting severed parts of digits were technically straightforward and feasible under most circumstances, before engaging in the surgical procedure one has to seriously consider whether after the restoration of the structure the functional region was sufficient to justify the demanding procedures of replantation.

Some recent reports on replantation surgery well supported the discussions. Cases of hand/finger replantation repeatedly were reported respectively: functional restorations were emphasized at the very beginning and details of actual functional return were given. Single digital replantation was always a controversial indication. The German experience of single digital replantation also supported the choice when functional restoration was predictably favorable. On the contrary, if otherwise, such complicated structural restoration might not be justified.

In other words, the days in which replantation of digits was considered to be always indicated were over because poor functional return of the reconstructed unit might even affect the overall function of the hand.

All these practical relationships between structure and function should be considered during any surgical planning for the restoration of function in congenital anomalies (Fig 1).

As far as surgical procedures are concerned, hand surgeons have worked out the order of priority to get the most ideal postoperative outcome. Different tissues deserve different amounts of attention. The order of priority should be:

- Vascular tissues for survival;
- Skin for rapid surface healing to allow other tissue healing and training;
- Bone for maintenance of basic functional structure so that healing and training may be facilitated;
- Nerve for maintenance of sensibility;
- All other tissues do not received special priority attention because remedial measures are possible.

In congenital anomalies of the hand, the vascular status of the distal organ was uncertain, and not infrequently, even after imaging investigations, not much additional information became available. Sometimes the conventional vascular supply was just missing (ie, the normal vascular bundle could not be found). Under such circumstances it would not be appropriate to insist on identifying the normal blood supply. When a normal vascular bundle was not found, the dissection at the area involved was kept at a minimum so that circulatory disturbances could be

![Figure 1](image-url)
limited. In situations in which tissue transfer was required (eg, in digital transposition or pollicization), a thick bend of soft tissue, together with its adjacent skin flap, could be mobilized as a relatively reliable pedicle providing the blood supply.

When a substantial amount of tissues were taken as a pedicle, closure of skin edges would be a problem. Immediate split skin grafting then became mandatory so as to give the best effects of vascularization. As a general rule, all tissues should be carefully repaired to gain the best possible results.

**PRINCIPLES GOVERNING PEDIATRIC MANAGEMENT**

It would be wrong to consider children as small adults. The special considerations required for management of children arise from the fact that their bodies are still growing.

**Timing of Surgery**

Performing structural correction as early as possible is a theoretical consideration that does not deserve serious support. The accepted principle should be as early as practical because too early is too difficult and might induce harm, whereas too late might mean too much growth damage and psychologic impairment. A good compromise is before 4 years unless severe growth disturbances do not allow the 3 to 4 years of deferral. Congenital anomalies in the hand should not be considered as anomalies in vital organs that affect survival. Growth in the hand also is expected to be slower than areas such as the maxillofacial area. Cleft lip and palate, for instance, would tend to shift open, if not repaired before 18 months. Hand anomalies, on the other hand, are not challenged by such early requirements. Moreover, the complexity of congenital anomalies also rules out the choice of having one major surgical endeavor to give the best chance of normal growth in the planning for correction.

**Difficult Diagnosis**

When the diagnosis is straightforward, surgical planning is simple. However, when the diagnosis is uncertain, planning will be very much affected. Thus, a hemangioma does not always require excision similar to other harmful tumors. Likewise, when diagnosed some chronic infections warrant special treatment. Longitudinal arrests are known to have a lot of coexisting soft-tissue anomalies: if not recognized, they naturally would lead to poor results (Fig 2). Cleft hands may have the most complicated anomalies of bone and soft-tissue components.

Although old uncertainties and confused issues remained unsolved (eg, over the different types of cleft hands), new forms of developmental syndromes with hand involvements are being reported. Japanese experts were particularly keen in the exposure of controversies existing in different forms of clefting. Cleft hands do have multiple varieties: missing longitudinal rays, total or partial; extra rays, complete or partial; abnormal joints and soft tissues; unstable to flail joints; and even hypoplasia. So in the overall diagnosis and planning of cleft hand corrections, comprehensive and thorough management requires a more academic and philosophic outlook to give the most reasonable outcome. However, some new modalities are as yet unknown to most surgeons (eg, brachydactyly type C), and in the past 5 years there were different reports on a variety of unfamiliar syndromes that consisted of hand anomalies. Examples included the limb-mammary syndrome, fragile X syndrome, spondylo-metaphyseal dysplasia, and de Bary syndrome. The varieties made corrective planning most difficult.

Swanson tended to take a simplistic view that his classification of anomalies was still practical and comprehensive so that after a quick match of the pathologic condition, the most likely correct diagnosis could be made and then the management could be planned accordingly. The simple approach worked
well for the simple situations in which a simple pathology was obvious. However, when multiple pathologies were present (e.g., the coexistence of longitudinal defects, joint instability, rigidity, and tissue duplications), and when none of Swanson’s clinical classifications fit well, clinical management became really difficult. Under such circumstances, identifying the most critical area and working out a staged policy would be a practical compromise.

**Growth Considerations**

Carelessly performed surgery leads to epiphyseal damages, which leads to growth arrests and deformities. Correction of deformities needs to take into consideration the time dimension, not only the structural dimensions.18

Although growth plates and growing epiphysis were vulnerable to injury, it did not mean that they could not be touched. In fact as long as the epiphysial plate was untouched, the epiphysis could be cut flat and stitched to its adjacent component in an attempt to achieve linear fusion, trying not to affect the optimal growth.19 Similarly, enlarged or abnormally faceted epiphyses could be shaved to give more congruent articulating surfaces without affecting the ultimate growth of the components handled. Tada and Yonenobi20 used this technique extensively in the Duplex thumb to bring about more satisfactory alignment of the retained component after sacrificing the unwanted component. We have adopted this technique since the 1980s and have had very gratifying results.21

**The Uncooperative Child and Psychologic Influences**

Unlike an adult, young children do not tolerate pain and limitations of activities. Henceforth, dressing has to be double safe and better protected, pinning has to be secure and hidden, and casts need to go up one proximal joint (e.g., including the elbow) so as to avoid loosening.22 Unfortunately, this issue, which has been considered essential, tends to be widely ignored. Softening of casts and losses of stability despite long casts still occur commonly. The more manpower-dependent supervision of weekly or more frequent evaluations should be endorsed.

Taking care of the postoperative child should include paying attention to his psychologic needs. The hand often reflects the mind. The hand is an important body image only second to the face. It is therefore easy to understand how a structurally abnormal hand could significantly affect a child. If at all possible, correction should be completed by school age.23 It would be wrong to assume that severe psychologic disturbances exist in the child with severe anomalies whereas for those who are not so seriously affected (e.g., duplication or syndactylyy), psychologic considerations are not necessary. In fact, it depends on environmental conditions, individual circumstances, and immediate family support for the actual outcome of possible psychologic disturbances. It might be logical for every child suffering from a hand anomaly to receive a proper assessment from the psychologist.

**Inaccurate Imaging**

With the growing child and unossified epiphyses, radiologic pictures need to be interpreted with extra caution and with a sound knowledge of ossification centers, otherwise fractures and displacements are misdiagnosed. Likewise, correct interpretation about length problems cannot be accurately given.24

The varieties of imaging have expanded extensively in recent years so that congenital anomalies could acquire much benefits. Although the conventional radiography and angiogram still served routinely for structural identification and delineation of vascular pattern, disappointments were not uncommon.

Ultrasonic examinations have benefited a lot of inflammation conditions such as tendonitis. The same device and technique should be useful to help identify thickened soft-tissue structures and to differentiate tissue planes between the normal and abnormal tissues.25 Conventional tomography and computed tomography of the radioulnar joint and the wrist are used in patients with persisting complaints or doubtful findings on plain radiographs and difficult anatomic situations.26 Suspected ligamentous injuries of the wrist including tears of the triangular fibrocartilage complex are evaluated by wrist arthrography or magnetic resonance imaging, the latter requiring a highly skilled imaging and interpretation technique. Magnetic resonance imaging is the method of choice for the detection of osteonecrosis.27 In one study, 75 surgical soft-tissue lesions of the hand were examined prospectively by experts on ultrasound and were correlated with surgical and pathologic findings. The normal ultrasonic anatomy of the hand was described. The use of real-time sonography allowed a reliable
diagnosis of the cystic or solid nature of soft-tissue tumors, and accurate estimation of their volume and their precise 3-dimensional localization. Sonography facilitates the location of foreign bodies, and appears as a new promising technique for the evaluation of tendons.25

**Special Surgical Considerations**

In the child, the neurovascular bundle is much more stretchable than in the adult. However, structural anomalies could well be associated with neurovascular anomalies. Awareness is much more important than special investigations, which cannot be taken as routine.

In the correction of alignment, very often joint surfaces and epiphyseal articulations have to be dealt with. It is important to remember that as long as the growth plate is not touched, growth disturbance will not occur. If desired, therefore, the cartilage end could be shaved.

Surgery these days tends to become gadget driven and gadget dependent. Surgical procedures should be followed closely, step by step. However, surgery performed for children with anomalies demand extra flexibilities on the operating table. Immediate decisions have to be made whether to stretch the neurovascular tissues or not; whether to end the surgery for a 2-staged maneuver or to continue on to completion; whether to attempt perfection or to accept some defects; whether to use a fixation device or rely on casting alone; and so forth.

**Limitations and Promises**

Surgeons used to be very conservative with congenital anomalies because they realized the limitations of reconstruction whereas a child born with many deficiencies could still learn to achieve good function.28 In contrast, there are those who are optimists and explore a means of reconstruction to provide a better baseline for adaptation. They are supported by modern techniques such as microsurgery, which has opened up new areas of reconstruction (Fig 3).19 New surgical implants, likewise, have created more options and versatilities for reconstruction.29

Microsurgery is a new technique applied by a number of proactivists in the reconstruction of deficient anomalies. The French group led by Foucher and Medina30 has been one of the most energetic groups who have used microsurgical transplantation of the toes to restore better function. A recent report gave very positive results in which 58 patients with congenital hand abnormalities underwent 65 toe-to-hand transfers. Symbrachydactyly (51 cases) was the most frequent indication. Forty-seven second toe-to-hand transfers were performed in 44 patients. The mean follow-up time was 5.2 years. Two failures occurred in cases in which only one artery was anastomosed; no failures were noted when more than one artery fed the transfer. Two patients with a single second-toe transfer presented with lateral instability of the transferred metatarsophalangeal joint. The mean active range of motion was 38°, with a mean extension lag of 25°. The mean 2-point discrimination was 5 mm. Forty-one patients used the transferred toe well when performing activities of daily living and playing games. Toe-to-hand transfer, before the establishment of the grip pattern, facilitated integration of the transfer.29

The report made promises. In our own experience we did witness promises, but we observed limitations at the same time. In my personal series of 13 cases, only 5 patients had reasonable results whereas 8 others did not show sufficient improvement, which we had expected (Fig 3). The transplanted toes could be so poorly placed, so rigid or so weak that little functional gain resulted. The poor positioning was because of the original deficiency that dictated the site of transplant. The rigidity and weakness were caused by the lack of appropriate motor unit in the recipient site. We would conclude after my early mistakes that possibly
proximal transfer of the abnormal finger unit, with the humble intention of achieving just a tip to forearm pinch, might be a much more practical way of correction in the single digit anomalies.31

However, although attempts on reconstruction might not be providing satisfactory functional restoration, the patient might still be happy. This reminds us of Bunnell’s31 wise comment, “when there is nothing, a little is a lot.” A realistic balance between the promises and limitation should converge into a careful, practical planning of action.32

**Adaptation**

The conservatives tend to stress the ability of the child to adapt under varieties of adverse structural hardship. These observations dissuade them from offering surgical intervention. However, the same argument could be the support for creative moves. Now that rehabilitation sciences have been enjoying many advances, they solidly support children for reconstructive surgery on a more disciplined pathway of adaptation—rehabilitation and training in the pursuit of better function.33 Under the new postoperative situation, rehabilitation training after functional correction should provide more real and adaptive value.

Indeed hand rehabilitation in recent years has advanced greatly under the leadership of the occupational therapists.34-36 Therapists were not only leaders and supervisors providing functional training and providing supervision in adaptation, but they made innovative hand splints and braces, started electronic devices, biofeedbacks, continuous passive movement devices, force-feedback gloves, and organized multiple disciplinary collaboration. They included other therapists, nurses, psychologists, musicians, and parents. Although more specialized groups such as those giving special service to the elderly and rheumatoid patients were becoming more active, more specialized groups would be expected to serve the congenital anomalies.37,38

**Growth Consideration**

Although carelessly planned surgical procedures undoubtedly affect the management outcome adversely, sometimes even producing a disastrous outcome, a better understanding of the growth components facilitates more versatile management. When a reasonably thin K-wire is passed longitudinally through the growth centers of the metacarpals and phalanges, with the intention of maintaining the favorable alignment for extra-long periods of time, the growth hazards usually are acceptable. Lamb and Smith39 would use an intramedullary K-pin to go through the second or third metacarpal bone(s) of the hand, through the carpus to the ulnar bone, with the aim of maintaining the wrist alignment after the procedure of centralization for radial club hand. The price to pay for possible disturbance of bone growth by the single longitudinal pin is far less than the loss of centralization to the wrist.40

**Case Illustrations**

To summarize, 2 difficult examples of congenital anomaly are used to show some of the important general and specific principles already discussed. The difficult conditions chosen are a thumb deficiency and radio club hand.

**FIGURE 4.** (A) A 14-month-old child with type 3B hypoplasia of the thumb and only minimal hypoplasia of the radius. (B) Pollicization of the index finger was performed.
THUMB DEFICIENCY

The classic treatment for thumb deficiency is pollicization. Pollicization of the thumb is a useful technique that has been applied to various forms of hypoplasia of the thumb with or without deficiency of the radius (Fig 4). Over the years the technique of pollicization of the index finger has been more or less standardized under the extensive work of Buck-Gramcko and others, and several principles may be generalized: a reduction in length of the digit to be pollicized, a rotation and repositioning of the pollicized digit to provide opposition, and soft-tissue reconstruction. The design and mobilization of skin flaps is important to achieve a well-covered and deep first web, and also to assist with stabilization of the pollicized digit to maintain the position in opposition. Long tendons and intrinsic muscle reconstruction also are of importance.

Except for special considerations, most of the time the most radial digit is pollicized as the thumb. The long-term outcome usually is gratifying and reasonable, with roughly 50% of range of movement and strength of the reconstructed thumb as compared with normal (Fig 5). However, pollicization for radial deficiency usually did poorly.

Shortening of the Pollicized Digit

Because a finger has 3 phalanges and a thumb only has 2, it has been a common practice to remove the entire second metacarpal through the physeal plate down to the base, to make use of the original proximal phalanx as the first metacarpal, and the middle phalanx as the proximal phalanx. The second metacarpal epiphysis is retained and positioned as a carpal bone, with the original metacarpophalangeal joint serving as the new carpometacarpal joint (Fig 6).

It should be realized that because the third digit is long, removing the entire second metacarpal sometimes would make the newly reconstructed thumb too short. At the same time, because the metacarpals are converging, placing the new digit close to the base of the third metacarpal will significantly narrow down the transverse arch of the hand. A better approach would be to retain part of the proximal shaft of the metacarpal so that the length of the new thumb can reach beyond the middle of the proximal phalanx of the middle finger. The presence of this proximal bone stalk also will help to provide a more abducted angle of the thumb. The second metacarpal epiphysis is rotated in the manner described by Buck-Gramcko and positioned and fixed to this proximal bone stalk (Fig 7).

Position of the Thumb

Buck-Gramcko described 2 important steps to ensure a better functioning thumb: the abducted and pronated position of the thumb, and a stable carpometacarpal joint. To achieve a stable carpometacarpal joint, the second metacarpal epiphysis is flexed to its end range of flexion and fixed in its new position (Fig 6). In other words, the original metacarpophalangeal joint is fully extended when placed in its new position. In this manner the new carpometacarpal joint will be stable, have less of a tendency toward hyper-
FIGURE 6. Drawings to show position of the metacarpal head during pollicization. (A) The metacarpal head is flexed fully and placed in the remnant of the base of the second metacarpal, which may be left long according to need. (B) A K-wire may help to keep the extended relationship between the metacarpal head and the phalanx and to position the new thumb in its new site. However, there is no fusion between the bones. (C) Radiograph of the same patient in Figure 5. Ossification of the transplanted metacarpal head (arrow) can be seen easily in the extended position and is well seated on the outer border of the base of the original metacarpal(*).
extension, and can provide good flexion, which is crucial for opposition.

The second metacarpal epiphysis also should be pronated 140° from its original plane into the new position and about 40° palmar abduction.

To assist with the positioning, a fine K-wire first may be passed through the second metacarpal epiphysis and then used as a lever to flex it, and then passed retrogradely through the base of the proximal phalanx and antegradely into the remnant of the second metacarpal. Sutures are then applied to the corners of the second metacarpal epiphysis to the second metacarpal remnant. Usually 2 sutures, diagonally set, are enough, and one does not aim for a solid union.

Maintaining a fibrous union between the 2 bones is advantageous for some movement in the joint to facilitate opposition in the future (Fig 6D).

The design of skin also is important to ensure a well-covered fist web. Final position of the skin flap also facilitated pronation and abduction of the transplanted digit. Pronation of the new thumb cannot be overdone, which may reduce the subsequent extension (Fig 8).

Skin Flap and Soft-Tissue Reconstruction

To ensure an adequate first web with minimal scarring, the design of skin flaps during pollicization is of vital importance. There are several commonly used incision. Basically a form of transposition and

![Image of a hand with a thumb.]  
**FIGURE 7.** Part of the second metacarpal was retained in this patient, adding length to the new thumb and at the same time providing a divergent angle between the first and second rays.

![Image of a hand with a thumb and a fingernail showing excessive pronation.]  
**FIGURE 8.** (A) There was excessive pronation of this thumb as shown by the fingernail. The palmar insetting point for the skin flaps (Buck-Gramcko technique; arrow) was too medial. (B) Although opposition was very good, extension of the thumb was restricted.
Z-plasty is designed (Fig 9). The limit of the skin incision around the proximal phalanx of the index finger (to be transposed) will mark the depth of the future first web and is governed by the length of the digit. The medial limit of the palmar incision governs the degree of abduction and pronation. The Buck-Gramcko design in addition extends 2 triangular flaps proximally from this limit. Insetting of these flaps in the new position helps pronation and abduction of the new thumb. Great care is required in dissecting these flaps because they need to be separated from the dorsal veins, and to expose the intrinsic tendons, the flaps may be undermined excessively. However, with careful techniques, necrosis of these flaps has been uncommon. The advantage of the Buck-Gramcko design is a direct exposure of the palmar neurovascular bundle early on during surgery. He recognized a high frequency of neurovascular anomalies and this approach will safeguard these and ensure a high success rate of pollicization.

After transposition of the digit, reconstruction of soft tissues is of great importance. The need to tighten the long extensor tendon is unanimously agreed. There is, however, a slight difference in opinion on the extent of intrinsic reconstruction. The trend is to restore both intrinsic muscles, although few would transfer the origin of the first dorsal interosseous to the hypothenar fascia as suggested by Kleinman and Zancolli. Most people leave the flexor alone for spontaneous adaptation.

**Alternatives to Pollicization**

Although pollicization can provide quite a promising aesthetic and functional outcome, there is an occasional excessive narrowing down of the arch of the hand, discrepancy in the length and size of the new

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**FIGURE 9.** Drawings of skin flap designs by different surgeons (A marks the palmar insetting point of the base of the digit). (A) Zancolli, (B) Corroll, (C) Buck-Gramcko (the dorsal skin flap may be divided at D or slightly skewed at D), and (D) Smith.
thumb, and stiffness (Fig 10). At the same time, many parents request alternative management, especially in the Asia-Pacific region.

Alternative techniques to reconstruct a hypoplastic thumb in radial deficiency can be quite difficult because the hand as a whole is quite hypoplastic. A useful technique is angulation and rotation osteotomy of the most radial digit to improve the pinch.\(^40,41\)

For isolated thumb hypoplasia without associated radial deficiency, an alternative technique is to transplant a free bone graft or toe phalanx.\(^42,43\) We have used a free metatarsal epiphyseal bone graft, together with a second-stage muscle and tendon transfer. The result was very acceptable (Fig 11).

Radial Club Hand/Radial Deficiency of the Forearm

Radial deficiency of the forearm, or radial club hand, is uncommon. Management has to follow several well-established principles to provide a satisfactory outcome (Fig 12).\(^41,44-47\) The hand is relocated over the distal end of the ulnar, known as centralization, and reconstruction of the hypoplastic or absent thumb is performed at a second stage, when a satisfactory result could be expected. Centralization usually results in reduction in movement or actual stiffness of the wrist joint, therefore, an important prerequisite before centralization is good mobility in the elbow. There are alternative approaches for stabilization of the wrist without sacrifice of motion, including vascularized or nonvascularized bone grafts.\(^48,49\) However, experience is still limited and more long-term results need to be seen before these techniques could be fully evaluated. Another new approach is the use of a distractor for soft-tissue distraction before centralization. Early results indicated that better motion preservation could be achieved by using this technique.\(^50,51\)

Centralization and Radialization

The technique of centralization involved a release of the contracted structures on the radial aspect of the wrist and distal forearm, relocation of the carpus over the distal end of the ulnar, and reinforcement of the capsule over the ulnar side of the wrist joint.\(^32\) Excision of carpal bones is frequently required or was considered routine by some surgeons. The trough created should be as deep as the width of ulnar it had to accommodate. Buck-Gramcko\(^45\) modified this without bone excision by performing a more thorough soft-tissue release, and brought the carpus over the distal end of the ulnar. The entire flexor-extensor muscle mass on the radial side may be detached, and subsequently transferred to the ulnar wrist extensor. He also relocated the ulnar head to the radial aspect of the carpal bones, a procedure he termed radialization (Fig 13). This would redistribute the force over the wrist joint and prevent recurrence. This was difficult to accomplish, but has become much easier with the aid of soft-tissue distraction.

The planning of the skin incision also is much discussed. There are a few different designs of skin incisions and skin flaps during centralization. They are a matter of personal preference and work equally well except for a slightly higher complication rate observed with the bilobed technique.\(^48\)

Soft-Tissue Distraction Before Centralization

Soft-tissue tightness has made it difficult sometimes to centralize the wrist without carpal bone excision. However, bone excision frequently results in stiffness of the wrist and may cause reduction of growth. The Ilizarov technique of gradual distraction may be applied to this situation to provide gradual soft-tissue distraction before surgery to overcome the soft-tissue contracture.\(^49,50\) By using an external fixator, the wrist joint is gradually distracted to open up the joint space and to partially correct the radial deviation. Some recent reports have highlighted the benefits and what could be achieved with this ap-

FIGURE 10. Another case of unilateral pollicization showing the excessively deep first web and narrowed arch of palm, although the size of the new thumb was quite good.
FIGURE 11. (A) A patient with type 3B hypoplastic thumb. (B) An epiphysis bearing hemimetatarsal bone graft was transplanted from the foot. This was followed 1 year later by tendon and muscle transfer. (C) Five years after surgery. Showing a good development of the thumb. (D) The 5-year radiograph showed good hypertrophy of the bone graft and viability of the transplanted epiphysis (arrow).
We have applied the technique in 3 patients. An Ilizarov frame was used in the first and an ulnar uniplanar frame was used for the 2 subsequent patients. The Ilizarov type of frame required a ring to be constructed over the hand and the distal forearm and this restricted the use of the hand as well as caused scarring after subsequent removal of the frame is less conspicuous.

FIGURE 12. Radial deficiency of the forearm with marked radial deviation of the hand and absent of thumb. (A) Dorsal view. (B) A patient with bilateral centralization showing satisfactory results.

FIGURE 13. (A) Radiograph of a patient with complete absence of the radius before surgery. (B) Radiograph of another patient showing that the carpus was located well over to the ulnar aspect of the distal ulnar. This was radialization (Buck-Gramcko).
impingement to elbow flexion. This was not the best construct. For the ulnar uniplanar frame, 2 pins were passed through the metacarpals, and the proximal pins were placed in the ulnar (Fig 14). An incision was made over the ulnar side of the wrist, and the release of tight structures over the radial side was carefully performed. Distraction was performed over 3 to 4 weeks. The aim was to attain a roughly 1-cm gap between the distal ulnar and the carpal bones (Fig 15).

When distraction was completed, the patient was taken to the operating room again, and, on removal of the external fixator, the wrist joint was carefully released through the original incision and the distal

![FIGURE 14.](image1)

(A) A young child with hypoplasia of the radius, absent thumb, and radial subluxation of the wrist. Manipulation has caused a pseudoreduction of the wrist. But despite ulnar sweeping of the metacarpals as shown, the wrist remained subluxed radially. (B) Soft-tissue distraction using an ulnar uniplanar frame was performed before centralization.

![FIGURE 15.](image2)

(A) Radiograph taken at the time of application of the external distractor. The wrist was dislocated radially and these was significant contracture. (B) Radiograph at the end of 3 weeks of distraction showing a much-increased gap between the ulnar and carpus, although it was not attempted to correct the radial deviation. Centralization was performed at the end of 4 weeks.
ulnar was placed to the radial side of the axis of the carpus. The joint capsule was then plicated, and the tendons were relocated, or, if present, the radial muscle mass (the joined flexor and extensor mass) was transferred to the ulnar side and sutured to the extensor carpi ulnaris tendon.

For the 3 patients in whom we have applied this technique, centralization at the second stage was made much easier after distraction, and no bone removal was required. However, to reduce the concavity of the distal ulnar or the proximal end of the carpus, we had to gently shave away some cartilage on either side in all 3 cases. Nevertheless, we found that there was quite a lot of fibrosis around the wrist joint during the second exploration, making identification of tendons difficult, and risk adhesion and inadvertent injury to them possible.

We therefore recommend only a minimal exploration of the wrist joint during the initial exploration and minimal release of the radial aspect of the wrist joint, and to leave the definitive release and tendon transfer for the second stage. Although the distraction technique is actively being developed, one has to emphasize that the ultimate outcome should be perfect, stable centralization of the wrist, which is essential for hand function.

**The Stiff Elbow**

The stiff elbow in radial deficiency constitutes a major functional deficiency (Fig 16). Unless the entire limb is not so short that the deviated hand can still reach the mouth, the patient will not be able to self-feed and the functional activities will be very limited. The incidence of stiff elbow in radial deficiency can be as high as 20%.⁵¹,⁵²

The management of a stiff elbow in radial deficiency is difficult and uncertain. Most investigators considered this as a contraindication to centralization of the wrist. In other words, there is no certain way of helping these patients, and they may end up having a major functional limitation of their upper limbs. Some patients may respond to a period of splinting of the wrist, therefore, encouraging flexion in the elbow to occur. Solutions to this problem may come from consideration of other congenital causes of elbow stiffness.⁵³

Extension deficiency of the elbow may present as pterygium cubitale, which occurs in isolation or as part of Larsen’s syndrome. Besides soft-tissue contrac-

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**FIGURE 16.** (A) A 10-month-old child with radial deficiency. There was no active flexion and the elbow was in extension. (B) There was elbow stiffness with limited flexion, which could not be corrected by splintage and physiotherapy.
Similarly, we adopted a 2-stage approach (Fig 17). In the first stage, the elbow was explored and any tight or obstructing structures were removed to regain passive range of flexion of the elbow. If this was successful, a second-stage latissimus dorsi muscle transfer was performed.

The elbow was approached through a lateral approach. Both anterior and posterior aspects of the elbow were accessed through appropriate soft-tissue dissection. The anterior muscle group (biceps and brachialis) was dissected off the anterior aspect of the humerus and extended carefully distally up to the radial head, taking care not to damage any radial nerve that may be present. The elbow joint capsule was released and any bony or soft-tissue obstruction to flexion could be cleared.

Posteriorly, the posterior muscle group (triceps) was dissected off the posterior aspect of the humerus and distally mobilized toward the olecranon, just posterior and lateral to the lateral epicondyle, with care not to detach the lateral collateral ligament. The triceps muscle insertion on the olecranon was preserved. The posterior elbow joint capsule was released and any bony or soft-tissue tethering preventing flexions were cleared. The aponeurosis of the triceps muscle was also released by multiple transverse releasing cuts. The triceps muscle was quite well developed in these situations.

We performed 2 such explorations in stiff elbows of radial-deficiency patients. In both cases we found only tethering of the capsule to the joint surface, minimal hyperplasia of articular cartilage and flattening of joint surfaces, in particular the olecranon or trochlea fossae, and contracture of the triceps muscle. There were no major incongruities or fusions between the joint surfaces. After soft-tissue release as described, together
with gentle and superficial shaving of the articular cartilage to provide more concave surfaces, the elbow joints could be manipulated to about 90° of flexion. These were very encouraging observations.

The first case subsequently relapsed back into stiffness in extension because there was no active elbow flexor, and the parents refused further surgical intervention. The second case was well maintained with elbow flexion range of up to 90°, and was waiting for the second-stage reconstruction with a latissimus dorsi muscle transfer. The theoretical advantage in radial deficiency compared with arthrogryposis is the presence of a normal latissimus dorsi muscle for transfer, and the functional result is expectedly better.

The Older Patient

For various reasons, it is not unusual in our area to have patients presenting at an older age or even in their teenage years with radial deficiency of the upper limbs. Despite the fact that these patients are usually quite well adapted to their limb impairments, they have social and psychologic reasons to request treatment. Management of these patients is not simple. Expectations are high and self-image and psychologic preparedness are low. It is utterly important to have a good understanding between the surgeon and the patient and family, and, if deemed necessary, a psychologic assessment and counseling before surgery is warranted.

The main request for reconstruction is usually the quest for a straight wrist and better-looking limb. Surgery in these patients is difficult because of the usually advanced and well-established soft-tissue contracture, and the increased risk for damage to neurovascular structures by surgery. Figure 18 shows the management of such a patient. The proximal carpal row was removed and a tension-band wire loop and intramedullary Steinman pin were used for a stable wrist fusion. Despite an actual shortening of the bone, functionally the hand could reach out more and cosmetically the hand was much improved, and activities performed by the hand were more aesthetically and socially acceptable. On the whole, it was a worthwhile procedure to perform. In a skeletally mature patient,

![FIGURE 18. A 15-year-old boy came with a neglected radial deficiency of his left forearm. (A) Palmar view, (B) dorsal view, (C) radiograph, and (D) radiography at the end of 1 year.](image-url)
there is less chance for the deformity to recur. With present day technology, the limb can be lengthened at a second stage.

**Residual Deformity and the Short Limb**

**Distraction Lengthening**

It has been realized that, with growth, a high proportion of patients had gradual and partial recurrence of their deformities.\(^{56,57}\)

Many patients also found their limbs too short, which made it difficult for them to care for personal hygiene. With the advent of limb-lengthening technology, these problems may be corrected by gradual limb lengthening. Residual limb bowing and radial deviation of the wrist may be corrected at the same time.\(^{58}\)

Figure 19 shows one such patient we have treated. The patient had bilateral type IV radial deficiency and
was treated with bilateral radialization at 1 year of age and pollicization at 2 years of age. There was residual radial deviation of the left wrist and the parents also complained that because of the shortness of the limbs, the child could not manage her perineal hygiene well. Two distraction procedures were performed on the left forearm and wrist. The first stage used a hybrid frame. Two wires were passed through the metacarpals for an Ilizarov type of distraction and gradual correction of radial deviation of the wrist. A uniplanar frame was incorporated and used for the proximal ulnar lengthening. The bowing was corrected acutely during osteotomy of the ulnar. The 2 procedures were performed simultaneously but at different rates. The ulnar was lengthened by 2 cm or roughly 15%, and the wrist deviation was largely corrected. However, soft-tissue distraction of the wrist was the more difficult part. The patient experienced tendon tightness and also mild paresthesia of the fingers during distraction early on, and the rate of distraction had to be kept at a low rate. At a second procedure, an ulnar uniplanar frame was used for soft-tissue distraction of the left wrist. This was followed by soft-tissue stabilization. The wrist was quite well centralized.

Early results of distraction correction of residual deformities from several recent series noted a high complication rate, but the results were, in general, satisfactory. However, a tendency toward partial recurrence of the angular deformities was noted in a sizable proportion of patients. Careful attention to avoid excessive traction pressure on the physis and reconstruction of a good muscle balance were suggested as important steps for sustainable long-term results.

**CONCLUSION**

Management of both thumb deficiency and radial club hand well show the principles being used for the treatment of congenital anomalies. First, the timing of surgical correction: too early will not only be too difficult technically because the structures are too small, but if more time is allowed, growth will expose more tendency of development that would allow overcorrection in the appropriate direction or avoid undercorrection. The compromise for surgical timing is still before school age. Of course, one more reason for not intervening too early is because of difficulties getting accurate information about vascular details. When special surgical considerations are being given (eg, when local skin flap coverage has to be raised) damage to vascular structures have to be avoided. Under such circumstances, bigger structures are more favorable.

For both pollicization and radial club hand, positioning is of vital importance for the restoration of function. With a good pollicized position, one does not need a long length, and even with one finger deficient, training could achieve so much that the overall hand function would not be deficient. With a good centralized wrist position, even though the radial club hand is abnormal, so much adaptation will be successful and the deficient hand will learn new ways of functional achievement despite missing parts, deficient lengths, rigid joints, deformities, and weakness.

Despite an ambitious outlook, one should realize that reconstructing a deficient hand that was born defective is a loser’s game. One must do no harm. One must not make things worse. One must not be overly ambitious. The target and a practical goal must be set at the very beginning. Too many attempts always create problems whereas multiple problems cannot be solved with one attempt. The way to deal with this complexity, therefore, is to allow a realistic compromise.

With our present experience and ability in helping children born with congenital anomalies, irrespective of whether surgery is indicated or not indicated, hand surgeons should be confidently engaged in positive, supportive moves in attempts to improve the deficiency. As long as a realistic goal is set, a good timing given, a practical plan designed, and requirements for rehabilitation followed, the child should enjoy a better functional outcome.

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