Lengthening and Deformity Correction in the Upper Extremities

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Indications for lengthening the upper extremities are more specific than those for lengthening the lower extremities. In the human non-weight-bearing upper extremity, a moderate discrepancy in length does not produce significant functional deficit. Before the introduction of the Ilizarov technique, humeral lengthening was reserved for patients with severe shortening because of the risks associated with the Wagner technique of bone grafting and plating. The Ilizarov technique broadens the indications for upper extremity lengthening and deformity correction.

With the Ilizarov technique, an external fixator is applied and gradual mechanical distraction is achieved at a controlled rate and rhythm. Length is restored and deformity eliminated by bone regeneration, rather than bone grafting. Gradual mechanical distraction with the Ilizarov apparatus has also been used to facilitate centralization and to correct radial club hand deformity.

HUMERAL LENGTHENING AND DEFORMITY CORRECTION

Before the introduction of the Ilizarov technique, humeral lengthening was reserved for patients with severe shortening. Reports on the use of the Ilizarov technique in the upper extremities have discussed lengthening of the humerus and have supported expanding the indications. Currently, lengthening of the humerus is performed for limb length discrepancy and in conjunction with lengthening for stature. Discrepancies greater than 5 cm affect appearance and can cause functional loss. Patients complain of difficulty reaching and carrying, discomfort sitting in chairs that have two arm rests, and the need to alter sleeve lengths. In some patients with dwarfism, rhizomelia of the humerus causes the humerus to be disproportionate to the forearm and the upper extremity to be disproportionate to the trunk. Upper limb lengthening restores proportion to the lower limbs and to the...
trunk if the lower limbs are lengthened 6 inches or more. Bilateral arm lengthening is functional in patients with dwarfism, because it allows increased ease for personal hygiene, which becomes difficult as these patients grow older and less flexible. It also makes reaching without the need for compensatory mechanisms much easier. For example, reaching to turn on a faucet or across a hot stove can be difficult and dangerous with short arms.

Premature physeal arrest, occurring secondary to infection or because of trauma, tumor, or radiation, is the most common cause of humeral shortening. Septic growth arrest of the proximal humerus in an infant after osteomyelitis can result in large humeral length discrepancy. Gradual distraction, using either a monolateral or a circular external fixator, can be used to lengthen the humerus.

Techniques of Humeral Lengthening

Monolateral External Fixator

The patient is positioned on a radiolucent table (diving board) that has no metal edges. The broad part of the image intensifier acts as the hand table. The patient is moved as close to the edge of the table as possible. The shoulder is rotated 90° to obtain anteroposterior and lateral views. When there is no deformity, the reference for alignment is the medial border of the humerus (Fig. 1A).

Figure 1. A. Anteroposterior view of the humerus shows that the medial cortex corresponds to the mid-diaphyseal line of the bone. B. Lateral view shows the distal curvature of the humerus, which is exaggerated in cases of achondroplasia. The circles represent the planned location of the half-pins.
The external fixation pins are inserted perpendicular to the medial border of the humerus. The first pin is the distal most pin. This is placed just proximal to the olecranon fossa as posterior as possible in the humerus because of its distal curvature (Fig. IB). For accuracy of placement, the cannulated drill technique is used. This involves first inserting a 1.8-mm K wire into the desired location. If the K wire is in the right plane and orientation, then a 4.8-mm cannulated drill bit is used to drill over the top of it, and a 6-mm threaded half-pin is inserted. If the orientation of the K wire is not accepted, then it can be repositioned easily before reaming with the 4.8-mm drill bit. This technique is very precise and avoids erroneous pin placement.
The distal most pin is used to orient the external fixator. Our experience has been primarily with the Orthofix (Orthofix, Richardson, TX) pediatric rail system (Fig. 2).

Figure 2. A, Bilateral Orthofix pediatric rail fixators were applied for lengthening of both humeri in a boy with achondroplasia. B, These fixators do not limit shoulder motion.
Circular External Fixator

An external fixation apparatus is preconstructed with a proximal, small, $90^\circ$ arch or segment of a half-ring (one can cut the carbon fiber half-rings using a Gigli saw) positioned anterolaterally and a distal $5/8$ ring opened anteriorly to facilitate elbow flexion (Fig. 4).

![Figure 4. Model of Ilizarov apparatus for lengthening of the humerus.](image)

Three distraction rods are placed between the arch and the $5/8$ ring: one from the midpoint of the arch to the most anterior and lateral hole of the ring, one connected with an oblique support from the most anterior hole of the arch to the most anterior and medial hole of the ring, and one connected with an oblique support from the most posterior hole of the arch to the midpoint of the ring posteriorly. The arch and the ring are placed parallel to each other and are spaced apart by the distance determined to be appropriate by preoperative radiographic and clinical measurements. Two 5- or 6-mm half-pins, inserted from anterior and lateral directions, are used to achieve proximal fixation.

With the patient positioned supine on a radiolucent table, an image intensifier is directly laterally introduced. The extremity is placed in normal resting position (i.e., $40^\circ$ of internal rotation). The first half-pin (5 to 6 mm) is placed perpendicular to the humeral shaft, distal to the surgical neck to avoid injury to the axillary nerve, and is inserted directly laterally. The apparatus is slipped over the extremity and applied to the half-pin. The limb is centered and aligned within the frame, and the
half-pin is fixed to the proximal surface of the arch. The 5/8 ring should then be at the level of the proximal edge of the olecranon fossa. A transverse, smooth, 1.8-mm wire is placed at that level in the coronal plane. Caution must be used to avoid injuring the ulnar nerve. The apparatus is centered relative to the arm and elbow, and the wire is fixed and tensioned.

A second half-pin, from anterior to posterior directions on the distal surface of the arch, is used to achieve proximal fixation. The half-pin must be centered on the humerus and should not impinge on medial soft tissue.

Two obliquely oriented, smooth, 1.8-mm transfixion wires are used in the coronal plane to achieve distal fixation. The wires are inserted obliquely upward from the proximal margin of the medial and lateral epicondyles and are connected by posts suspended from the 5/8 ring proximally and distally. The elbow needs to be taken through a range of motion, and the skin needs to be observed for tension. If the wire is tenting the skin, it should be retracted beneath the soft tissue and the skin should be repositioned. The wires can then be fixed at one end and simultaneously tensioned to 90 kg.

Threaded half-pins can be used distally as an alternative to using wires on the circular frame. One lateral and one oblique half-pin are inserted using the cannulated drill technique. Both are inserted from the lateral side. The oblique pin goes up the lateral column. This avoids protruding posts and pins that impinge on the chest wall and breast.

We prefer to use the external fixator when gradual deformity correction is required or when acute deformity correction proximal to the deltoid tuberosity is needed. Proximal to the deltoid tuberosity, it is advantageous to use a fixator that can easily achieve fixation in a multiplanar fashion.

Techniques of Humeral Deformity Correction

For acute correction of a deformity at the level of the surgical neck, the humeral head should be placed in an anatomic position relative to the glenoid. Two half-pins are inserted into the humeral head, from anterior and lateral directions, and are connected to a small arch. The extremity is repositioned along the thorax, and the standard distal humeral fixation is completed. A percutaneous osteotomy at the level of the surgical neck is performed from an anterolateral direction. The proximal and distal blocks of fixation are acutely positioned parallel to each other. Threaded rods are connected, and fixation is secured.

Diaphyseal angular deformities are corrected gradually using a hinged distraction rod construct. Distal humeral fixation is reinforced by using olives on both oblique wires. Other deformities, including supracondylar deformities and derotation, can be corrected acutely or gradually. To prevent stretch lesions of the skin and soft tissue, the skin should be recruited into areas where tension will be greatest before pin insertion.

Corticotomy of the Humerus

Corticotomy refers to a percutaneous, low energy, subperiosteal osteotomy. Corticotomy is performed either proximal or distal to the insertion of the deltoid. If it is performed proximal to the insertion of the deltoid, it will increase pressure on the humeral head as lengthening progresses. If it is performed distal to the insertion of the deltoid, it will have little effect on the shoulder.

A proximal humeral corticotomy is performed through an anterior incision, and a mid-humeral corticotomy is performed through an anterolateral incision. The bone underlying the incision is cut with a 5-mm osteotome, and the cortex is also cut medially and laterally. After proceeding around the cortex in both directions, the
osteotome is twisted to crack the remaining cortex. (Alternatively, the cortex can be perforated at the intended level with multiple drill holes placed obliquely in different directions, and an osteotome can then be used to complete the corticotomy.) Gentle rotational osteoclasis can verify completeness of the corticotomy.

Caution must be used in the distal humerus to avoid injuring the radial and ulnar nerves. A posterior 5-mm incision is made, the triceps fibers are split, and a small periosteal elevator is used to raise the lateral and medial periosteum. The same maneuver is performed on the medial side, and the corticotomy is performed under the protection of the periosteal elevator.

Case Reports

Ollier's Disease. A 13-year-old boy presented with Ollier's disease, which produced a valgus extension deformity of the proximal humerus and 10 cm of shortening. Radiographs revealed a 90° valgus extension angulation (Fig. SA).
Figure 5. A, Anteroposterior and lateral radiographs of a shortened deformed humerus in a 13-year-old boy with Ollier's disease. B, Two-week postoperative radiograph shows that the angulation of the hinges corresponds to the bony angulation. There is early separation of the distal lengthening osteotomy. C, Six-week postoperative radiograph shows the hinges and bone to be straight, with good bone regeneration at both distraction sites. D, Final radiograph obtained after 10 cm of lengthening and 90° deformity correction.

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Extensive enchondromatous involvement of the posterolateral aspect of the proximal humeral physis was seen. The patient was treated with a double-level osteotomy at age 13 years. The proximal osteotomy was made intentionally through the enchondroma site. In our experience, this has led to consolidation of the enchondroma. It does not stop new enchondromas from budding off the growth plate. The opening wedge hinge was oriented in the true plane of deformity so that the valgus and procurvatum could be simultaneously corrected. The distal osteotomy was used for lengthening.

The 2-week postoperative radiograph (Fig. SB) showed the angulation and lengthening. The 6-week postoperative radiograph (Fig. SC) showed the distraction gaps at the bottom and top, with good bone being formed at both locations and correction of angulation proximally. Five centimeters of lengthening was achieved at each end, so that all 10 cm of shortening was corrected. The final radiographs showed excellent consolidation and alignment (Fig. SD), with a normal carrying angle. Clinical photographs obtained before (Fig. SE) and after (Fig. SF) the procedure showed that the arm was lengthened and the deformity corrected. The patient's function and strength became equal on both sides (Fig. SG).
Septic Growth Arrest of the Humerus. A 14-year-old girl presented with an extreme case of septic growth arrest of both ends of the humerus (Fig. 6A).

Figure 6. A, A 14-year-old girl with sequelae of septic growth arrest of the humerus, including a 17-cm shortening, 45° internal rotation deformity of the shoulder, and flexion deformity of the elbow. B, Maximum extension radiograph shows flexion deformity of the elbow. C, Clinical photograph, obtained near the end of lengthening, shows the applied Ilizarov apparatus. D, Ten-week postoperative radiograph reversed to appear like arm seen from behind.

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Figure 6 (Continued). At the end of the gradual rotation correction, the same amount of internal (E) and external (F) rotation could be achieved with the device on.

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Figure 6 (Continued). G. At the end of 17 cm of lengthening, the arm was protected in a functional brace. H. After a stress fracture occurred, the bone hypertrophied. Before and after radiographs are shown for comparison. Clinical photographs obtained before (I) and after (J) the lengthening.
A previous nonunion of the shaft of the humerus had been plated and united. This patient had a 45° internal rotation deformity of the shoulder and a 17-cm shortening of the right arm. A maximum elbow extension radiograph (Fig. 6B) revealed a flexion deformity of the elbow.

The Ilizarov apparatus (Fig. 6C) was applied to the arm, with three distal wires placed in the frontal plane where the bone is very narrow. Two half-pins were used for proximal fixation. The frame consisted of one arch, one 5/8 ring, and three telescopic lengthening rods. After achieving the lengthening, the frame was modified to perform gradual derotation, from internal to external. Two derotation rings were centered on the bone. One ring was linked to the upper construct and one to the lower construct. The arm was then rotated gradually. Figure 6D shows the 10-week postoperative radiograph. At the end of rotation, the same amount of internal (Fig. 6E) and external (Fig. 6F) rotation was present with the device on. An extreme amount of lengthening, 17 cm, was achieved in this patient. She remained in the device for 2 years to complete consolidation (Fig. 6G). It would have been preferable to perform two lengthenings rather than one, but because of socioeconomic factors, this was not possible. After removal of the device, the bone was thin, and an expected stress fracture occurred inside the coaptation brace, which allowed the bone to hypertrophy (Fig. 6H). The bone thickened up, and the patient's shoulder and elbow returned to the preoperative level. A comparison of the arm before (Fig. 6I) and after (Fig. 6J) treatment using the Ilizarov technique is shown.

Septic Growth Arrest of the Humerus with Pseudarthrosis. A 10-year-old boy presented with a previous septic growth arrest of both the upper and lower humerus (Fig. 7A and B). Osteomyelitis had also produced a mid-diaphyseal nonunion. This patient was treated by osteotomy at the level of the surgical neck together with bone transport using a single longitudinal transport wire. The Ilizarov device was fixed to the acromion and scapula and to the humeral head proximally and to the distal humerus distally (Fig. 7C). Excellent bone was generated by distraction. The docking site was opened and bone grafted with successful union. In addition to repairing the bone defect, the arm was lengthened (Fig. 7D). The final result is shown in Figure 7E.
Figure 7. A and B, Photograph and radiograph of a 10-year-old boy with growth arrest of both the upper and lower humerus and a diaphyseal pseudarthrosis. C, Radiograph shows bone transport from proximal to distal.

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Septic Growth Arrest of the Humerus Secondary to a Unicameral Bone Cyst. This 30-year-old woman underwent multiple previous operations to heal her recurrent proximal humerus unicameral bone cyst. Union finally was achieved with a nonvascularized fibular graft (Fig. 8A). The proximal humerus healed with varus, internal rotation, and extension. It is not clear whether this deformity was caused by a malunion or by the associated growth arrest of the proximal humerus. This deformity pattern is typical for the proximal humerus. Preoperatively, the patient lacked flexion and abduction and external rotation of the arm and had 11 cm of shortening. The Ilizarov apparatus was applied with two half-pins in each segment, and two osteotomies were performed (Fig. 8B). Proximally, an acute flexion, abduction, and external rotation osteotomy was performed at the level of the surgical neck, as described above. A second osteotomy distal to the deltoid tuberosity was performed for gradual lengthening. Active and passive exercises of the shoulder and elbow were performed at physical therapy sessions (Fig. 8D and 8E). This Ilizarov frame is multi planar but essentially unilateral because all half-pins are lateral or anterolateral without the need for wires. Because there is no medial part to the frame, it is more comfortable against the chest. At the end of correction, both sides were of equal length (Fig. 8E). The total amount of lengthening was 11 cm. This patient’s active abduction was limited to 70° preoperatively (Fig. 8F) and improved to 135° postoperatively (Fig. 8G). Similarly, her flexion improved from 80° preoperatively to 135° postoperatively. The patient is shown before (Fig. 8H) and after (Fig. 8D) treatment.
Figure 8. A. Radiograph of a patient with 11 cm of shortening caused by growth arrest of the humerus secondary to a recurring unicameral bone cyst. There is a varus extension internal rotation deformity of the proximal humerus. B. Radiograph obtained during lengthening. Clinical photographs show extension (C) and flexion (D) of the elbow with the device on.

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Figure 8 (Continued). E, Radiographs obtained after lengthening show that both arms are of equal length. F, Preoperatively, abduction was limited to 70°.

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Figure 8 (Continued). G, Postoperatively, abduction improved to 135°. Clinical photographs show comparison of shoulder flexion before (H) and after (I) treatment.
Bilateral Humeral Lengthening in Achondroplasia. For bilateral humeral lengthening in patients with dwarfism, a monolateral fixator generally is used (see Fig. 2A and B). A percutaneous osteotomy is performed just distal to the deltoid tuberosity, the fixator is applied, and lengthening is attained gradually (Fig. 9A-C).

Some of these patients have a flexion deformity of the elbow. Although the apex of deformity is more distal, some extension correction can be achieved at the diaphyseal level by placing the distal pins in line with the curvature of the distal humerus. When all pins are aligned with each other, the osteotomy will extend. This was not necessary in the illustrated case.

Proximal Humeral Deformity Correction Without Lengthening. This 20-year-old woman with renal rickets developed a proximal humeral deformity that manifested clinically as limited flexion (Fig. 10A) and abduction (Fig. 10B). To correct the deformity, an acute correction was performed using the Ilizarov device. The proximal external fixation pins were inserted into the humeral head with the arm placed so that the humeral head was in the position that it would be in at the end of the correction: adduction, extension, and internal rotation. The distal pins were inserted with the arm placed at the side. The osteotomy was performed percutaneously using image intensifier control. The proximal arch, which is perpendicular to the neutral position of the humeral head, is then manipulated and made parallel to the distal arch, which is perpendicular to the shaft of the humerus (Fig. 10C and D). Before the correction, the pins appear crossed to each other (Fig. 10E), whereas after the correction, they appear parallel on the same radiographic view (Fig. 10F). This patient also had secondary hyperparathyroidism. Note the brown tumor distal to the surgical neck. This had to be curetted and bone-grafted. The final radiograph shows the realignment of the humeral head relative to the glenoid and the humeral shaft (Fig. 10G).
Figure 10. Before treatment, this patient had limited flexion (A) and limited abduction (B). C. Intraoperative photograph shows arches 90° to each other before correction. D. The two arches were made parallel after the osteotomy.

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Figure 10 (Continued). E and F, Radiographs obtained before and after the osteotomy with pins in place. G, Final radiograph obtained after correction and removal of the fixator.
LENGTHENING AND DEFORMITY CORRECTION IN THE UPPER EXTREMITIES

DEFORMITIES OF THE RADIUS AND ULNA

Paley's Classification

Length discrepancies of the forearm can be classified using Paley's classification (Fig. 11). Paley's system classifies discrepancies of the radius and ulna into five types.

Type 1: Radial shortening only
Type 2: Ulnar shortening only
Type 3: Absent radius with ulnar shortening
Type 4: Shortening of both the radius and ulna disproportionately
Type 5: Shortening of both the radius and ulna proportionately

Figure 11. Paley's classification of forearm length discrepancies: Type 1, shortening of the radius alone; Type 2a, shortening of the ulna alone; Type 2b, shortening of the ulna alone with dislocation of the radial head; Type 3, shortening of ulna with absent radius; Type 4, disproportionate shortening of the radius and ulna; Type 5, proportionate shortening of the radius and ulna.
Paley's Type 1: Shortening of the Radius Alone

Distal Radius Malunion. The most common cause of Type 1 discrepancy is distal radial fracture malunion. The typical malunion has deformities of dorsal tilt, loss of radial tilt, translation in the frontal or sagittal planes, shortening of the distal radius, and incongruity of the distal radioulnar joint (Fig. 12A). The angular and translation corrections can be accomplished acutely or gradually after a percutaneous osteotomy of the distal radius. The Ilizarov frame is applied first, and then the correction is performed. With acute correction, translation precedes angulation and then gradual lengthening is performed. With gradual correction, angulation and length are corrected together and then translation is performed (Fig. 12B - D). There is no need for a bone graft, a plate, or a long incision. Wrist flexion and extension and forearm supination and pronation usually are improved together with radiographic realignment (Fig. 12E - I).

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Figure 12. A. Radiograph shows a distal radial malunion. B and C. Photograph and radiograph of the apparatus at the beginning of treatment before gradual angular correction.

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Figure 12 (Continued). D, Apparatus at the end of the angular correction and during the gradual translation correction. E, Final anteroposterior and lateral radiographs obtained after correction show realignment of angulation and length of the distal radius.

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Figure 12 (Continued). F and G, Clinical photographs show final wrist extension and flexion. 

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Figure 12 (Continued). H and I, Clinical photographs show final forearm supination and pronation.
Distal Radius Growth Arrest. The most common cause of Paley's Type 1 discrepancy in children is distal radial growth arrest. Such discrepancies can be divided into those with and those without angular deformity. With distal radial growth arrest, the radius is short and the ulna is of normal length. A two-ring construct Ilizarov frame or a mono lateral fixator is fixed to the radius without fixation to the ulna. With the Ilizarov device, the rings are parallel to each other and are spaced to fit the length of the radius. Distally, one wire (1.5 mm) is used from the radio-volar side, going through and exiting the radius ulna-dorsally. This wire enters the bone just radial to the radial artery and exits dorsally without interfering with the distal radioulnar joint. This wire should be oriented approximately 23° to the joint surface. The wire is fixed to the distal ring of the frame. Distally, either a 3- or 4-mm half-pin is inserted into Lister's tubercle, approximately perpendicular to the wire that has been inserted. In the mid-radius, two half-pins are preferred in place of wires that used to be used in the proximal rings. Half-pins are safer, and there is less risk of injuring neurovascular structures. The proximal ring is positioned over the mid-forearm, immediately distal to the mobile wad. This two-ring construct apparatus is a very simple frame with no fixation to the ulna. If there is angulation in the distal radius, the distal ring is oriented at the angle of the magnitude of angulation, and the hinges are placed at the level of the center of rotation of angulation, as determined by the preoperative planning.
Case Reports

Complete Growth Arrest of the Distal Radius with Shortening, Without Angulation.
Post-traumatic distal growth arrest of the distal radius resulted in a 3-cm shortening of the radius compared with the ulna (Fig. 13A). A distal radial lengthening without any angular correction was performed (Fig. 13B). An Ilizarov frame was applied (Fig. 13C), and the radius was gradually restored to the length of the ulna (Fig. 13D).

Figure 13. A, Complete distal radial growth arrest with 3-cm shortening of the radius relative to the ulna. B, Distal radial lengthening without angular correction was required. C, Two-ring Ilizarov frame was used for this patient. D, Radiographs obtained after lengthening. The radius is out to the length of the distal ulna.
Partial Growth Arrest of the Distal Radius with Oblique Plane Angulation and Shortening. The distal radius developed a partial growth arrest on the dorsal and radial side. The distal radius kept growing on the ulnar and volar side, leading to malorientation of the articular surface (Fig. 14A and B). The angular deformity measured 42° on the lateral view and 38° on the anteroposterior view (Fig. 14C). This information can be plotted on a graph, with the anteroposterior angle on the x-axis and the lateral on the y-axis (Fig. 14D). By resolving the vector, the oblique plane of angulation is oriented 47° away from the frontal plane. The total magnitude of the deformity is 57° and its apex is anteromedial (ulno-volar) for the right hand. An Ilizarov device was constructed to correct this deformity (Fig. 14E-H). Using the Pritchett graphs for prediction of growth remaining in the ulna and radius, the amount of growth remaining in the ulna is calculated to be approximately 3 cm. The radius is then over-lengthened by approximately 3 cm relative to the ulna. With this construct, at the beginning of the correction, the hand and wrist are put in the same deformity position as the distal radius. During gradual correction, the hand comes to a neutral position, moving the hand together with the distal radius. For any lengthening of the distal radius of more than a couple of centimeters, the external fixation is usually extended to include the hand (Fig. 14E); otherwise, contractures can occur. It is important to fix the hand both to prevent contractures and to increase the efficiency of the physical therapy. With the hand fixed, the therapist can concentrate on the finger joints, because the wrist is stabilized by the construct. The therapist can work the finger joints without causing the wrist to move. Radiographs (Fig. 14I and J) show that the radius was over-lengthened by 3 cm relative to the ulna, in anticipation of the ulna's growth. There were three treatment choices for this patient: perform two lengthenings; lengthen the radius to the current length of the ulna and stop the growth of the ulna, accepting a 3-cm-short forearm; or over-lengthen the radius and wait for the ulna to catch up. Radiographs (Fig. 14K and L) show that the length of the ulna caught up to that of the radius, confirming that our prediction was accurate.

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Figure 14. A and B. Lateral and anteroposterior radiographs show deformity and shortening of the distal radius after partial distal radial growth arrest. C. Planning diagrams show a 42° deformity on the lateral view and a 38° deformity on the anteroposterior view. D. This information is then plotted on a graph. 1 mm = 1°; 42 mm on the y axis = the sagittal plane; 38 mm on the x axis = the frontal plane. Resolving the vector, the oblique plane angulation lies in a plane oriented 47° away from the frontal plane. The total magnitude of deformity is 57° (length of the line from the origin to the point 38,42). The apex is anteromedial for the right hand.

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Figure 14 (Continued). E, The Ilizarov device was applied with each ring perpendicular to its bone segment. The rings are angulated 45° to each other. The hand is kept in a neutral position relative to the distal radius. F and G, The radius was over-lengthened by 3 cm relative to the ulna, in anticipation of the continued ulnar growth.

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Figure 14 (Continued). H, At the end of the angular correction, the two rings are parallel and the hand lies in a neutral position. I, Final radiograph obtained after the removal of the apparatus shows that the distal radius is 3 cm longer than the ulna. J, The ulna continued to grow and to gradually catch up with the radius.

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Figure 14 (Continued). K, The length of the ulna caught up to that of the radius at skeletal maturity. L, Comparison radiographs of both wrists after skeletal maturity. Our prediction of future growth remaining in the ulna was accurate. This boy regained full supination and pronation.
Madelung's Deformity. Another cause of an isolated shortening of the radius is Madelung's disease. In patients with Madelung's disease, multiple deformities exist, including increased radial tilt, increased volar tilt, and ulno-volar depression of the lunate facet with proximal and volar subluxation of the lunate relative to the radius (Fig. 15A and B). Disruption of the proximal carpal row and subluxation of the distal radioulnar joint also are usually present in advanced cases. If this condition is longstanding, a diaphyseal angulation of the distal radius usually is present. Anatomic correction of this deformity complex is complicated, because there are so many deformities to address (Fig. 15C-F).

**Figure 15.** Madelung's disease. Anteroposterior (A) and lateral (B) radiographs show increased radial tilt, increased volar tilt, ulno-volar depression of the lunate facet, proximal subluxation of the lunate relative to the radius, and ulno-dorsal distal radioulnar joint dislocation. There is also a diaphyseal procurvatum deformity. C and D, Photograph and radiograph of Ilizarov device in place with extension to the hand and dorsal half-pin fixation of the lunate. E, After the lunate was reduced by distraction, an osteotomy was made to elevate and bone graft the lunate facet. An osteotomy to correct the diaphyseal deformity was also performed. F, Final anteroposterior and lateral radiographs show complete realignment of the radius and reduction of the lunate and distal radioulnar joint.

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Figure 15 (Continued).
The extra-articular deformity, either diaphyseal or metaphyseal, is corrected by an osteotomy of the distal radius. For some patients that is all that needs to be done, especially in the dorsal type (secondary to mesomelic dwarfism) (Fig. 16A-C).

Figure 16. A. Dorsal Madelung's deformity in a girl with mesomelic dwarfism. B. Oblique radiograph shows that the lunate and scaphoid facets are congruous with each other and can therefore be realigned using an extra-articular metaphyseal osteotomy. C. Final anteroposterior and lateral radiographs show reorientation of the wrist and distal radius.
For patients in whom the lunate is herniating and the distal radioulnar joint is dislocated, an extra-articular correction is insufficient. To reduce the subluxation requires an intra-articular osteotomy to elevate the lunate facet. Because the lunate is in a fixed volar and proximally migrated position, it needs to be reduced relative to the scaphoid facet before the lunate facet can be elevated by an osteotomy. We reduce the lunate by distraction. To distract the lunate, a half-pin is inserted from the dorsum of the wrist in addition to the metacarpal fixation. If the lunate is not fixed, it will separate from the rest of the carpus when the distraction is performed because of the strong contracted radiolunate ligament. Once the lunate is reduced, an osteotomy is performed and the bone under the lunate is elevated, creating support for the wrist. Two osteotomies, therefore, are performed: one in the diaphyseal region, frequently performed to correct the diaphyseal deformity, and one intra-articularly, which is a hemi-epiphyseal osteotomy performed to elevate the bone and support the lunate. Figure 16 shows a patient with Madelung's disease after such treatment.

Paley's Type 2: Shortening of the Ulna Alone

Ulnar growth arrest is a common cause of isolated shortening of the ulna with a normal length radius. With distal ulnar growth arrest, two problems can occur. The first is that when the distal ulna stops growing, the distal radius starts curving toward the ulna, creating a deformity of the distal radius with ulnar deviation of the hand. The second potential problem is that dislocation of the radial head can occur if the condition is left untreated long enough.

Figure 17 shows a patient with deformity of the distal radius, with shortening of the ulna. To treat this condition, we need a construct that will transport the ulna to the length of the radius and correct the deformity of the radius. When performing lengthening of the ulna, the radius always needs to be fixed proximally. Otherwise, the interosseous membrane between the radius and ulna will pull the radius out of its joint, transporting the radius distally. When performing lengthening of the radius alone, the ulna does not need to be fixed, because the ulna has such a stable joint at the elbow.

Case Example

Distal Ulna Growth Arrest with Deformity of the Distal Radius. Figure 17A shows a patient with a distal ulna growth arrest, with deformity of the distal radius. The ulna was transported distally using a proximal osteotomy (Fig. 17B). The radial deformity was corrected with a distal radial osteotomy. At the end of the correction, the distal radius was realigned and the ulna was out to length relative to the radius (Fig. 17C). After skeletal maturity, there is some residual discrepancy, because the distal radius continued to grow (Fig. 17D). Clinical photographs of the patient as an adult show that excellent function was achieved and maintained (Fig. 17E-H).
Figure 17. A, Radiograph shows distal ulnar growth arrest with secondary ulnar deviation of the distal radius. The radial head is not dislocated. B, Ulnar transport and gradual opening wedge correction of the distal radius were performed. C, Radiograph shows final alignment at the end of correction. D, Distal radius continued to grow, slightly overgrowing the ulna again, which could have been prevented by performing a distal radial epiphysiodesis. This is shown compared with the opposite normal side.

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Figure 17 (Continued). E and F, Clinical photographs show final wrist extension and flexion. G and H, Clinical photographs show final forearm supination and pronation.
Paley’s Type 3: Shortening of the Ulna in the Absence of a Radius

This type of shortening refers to the short forearm in patients with radial club hand.\textsuperscript{14,21} Distraction treatment of radial club hand should be initiated when the patient is between the ages of 1 and 2 years.\textsuperscript{6,26,30} After distraction centralization and radialization of the wrist, tendon transfer is performed, Distraction of the wrist in radial club hand can avulse the distal ulnar epiphysis. To avoid this complication, an ulnar incision is made and a circumferential capsulotomy from the ulnar side of the wrist is performed. The extensor carpi ulnaris and the extensor digiti minimi tendons need to be identified and retracted out of the way. The dorsal cutaneous branch of the ulnar nerve needs to be identified and protected. All of the other structures are retracted out of the way at the capsulotomy site. During the same surgery, an Ilizarov device consisting of one ring on the proximal forearm, with two half-pins on the ulnar side, and one ring going around the hand, with two wires through the four metacarpals, inserted with a translation hinge is applied.\textsuperscript{\textdagger} The hinge is located at the intersection of the second metacarpal and the shaft of the ulna. It is placed on the convex opening wedge side of the ulna so that when distraction takes place, both angulation and translation can be achieved, but the cartilage surfaces do not compress together. The capsulotomy is performed in an effort to prevent distal ulnar physiolysis. If a capsulotomy is not performed, distraction of the hand away from the ulna will pull the epiphysis of the distal ulna off (physiolysis), potentially damaging the growth plate. Distraction is then performed gradually until the hand is overcorrected. This means that the second metacarpal becomes in line with the shaft of the ulna, which is the Dieter Buck-Gramcko radialization position.\textsuperscript{\textdagger} The term radialization indicates that once the carpus is on the ulnar side of the ulnar epiphysis, it produces lateral pressure from the ulnar side onto the ulnar epiphysis, causing the distal ulna to remodel and look like a distal radius.

The first step, distraction centralization and radialization, obtains correction gradually (Fig. 18A - L).
Figure 18. A and B, Left and right hands of a 2-year-old boy with bilateral radial club hand. C and D, Clinical photographs obtained after distraction centralization and tendon transfer of the left hand. The right hand had not yet been treated. E, Clinical photograph obtained after pollicization on the left hand and distraction centralization on the right.

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Figure 18 (Continued). F and G, Preoperative anteroposterior and lateral radiographs of the left hand.

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Figure 18 (Continued). H, Radiograph obtained on the day of surgery. A total capsulotomy was performed between the ulna and carpal bones. The fixation consists of one ring around the metacarpals with two wires and one ring around the proximal ulna with two half-pins. The hinges are located on the bisector line of the angle between the second metacarpal and ulnar diaphyseal lines. I, Radiograph of the wrist obtained at the time of the tendon transfer. J and K, Radiographs of the hand and forearm obtained after removal of the apparatus. The hand was held in a splint for these radiographs. The splint was used after removal of the apparatus for 3 months day and night and then for 3 months at night only. L, Three months after the apparatus was removed, the index finger was pollicized. This radiograph was obtained 6 months after the pollicization. The same sequence of treatment was also performed on the right side.
To maintain the correction, the muscle pull on the hand needs to be balanced. This is accomplished by performing tendon transfers. We like to transfer the flexor carpi ulnaris to the dorsum of the wrist. The flexor carpi ulnaris is always present and is a major flexion-deforming force. It can be changed from a volar flexion force to a dorsiflexion force. The wrist needs to be balanced so that there is an ulnar pull and, even more importantly, a dorsiflexion pull on the wrist. Therefore, by taking the flexor carpi ulnaris and making it a wrist dorsiflexor, it still maintains its ulnar direction of pull and then becomes a dorsiflexor. Transfer the tendon from the volar side at the pisiform bone, taking with it a fleck of cartilage from the pisiform bone and suturing it to the site at which the extensor carpi ulnaris tendon inserts at the base of the fifth metacarpal. The extensor carpi ulnaris also is rerouted, and because it is on the ulnar side, it becomes loose from the distraction correction. Therefore, leave it attached and pull it taut in a more central direction, suturing it to the fourth or even the third metacarpal so that it becomes more of a centralized dorsiflexor. Two tendon transfers, therefore, are performed: the flexor carpi ulnaris to the base of the fifth metacarpal and the extensor carpi ulnaris usually to the base of the fourth metacarpal, without detaching it from the base of the fifth. All of this is performed with the external fixator in place. We used to remove the external fixator and pin the ulna, but we found that doing so was difficult and was traumatic to the epiphysis. We now prefer to use the frame, which can hold the wrist completely stable and protect the tendon transfer while waiting for the transfer to heal. The initial distraction for correction usually takes 4 weeks, and the frame is left in place for an additional 2 weeks. The tendon transfer is then performed, and the frame is left in place for another 6 weeks after transfer. After the transfer is mature (approximately 3 months after the initial surgery), the entire frame is removed. A splint is then made to hold the wrist in an ulnarly deviated, slightly dorsiflexed position. Physical therapy is started immediately to train the transfers to work.

Three to 6 months later, the patient undergoes pollicization. Once the pollicization is completed/ the patient is trained to use the substitute thumb. At that point, the child usually is between the ages of 2 and 4 years. No treatment is administered for the next few years, and then, when the patient is between the ages of 6 and 8 years, the first lengthening is performed. Performing the lengthening before the child reaches that age is not advised, because the bone is too small and the percentage of length that can be attained is limited. When the patient is between the ages of 6 and 8 years, 6 to 8 cm of lengthening of the ulna can be attained.
In some patients with radial club hand, the ulnas are bowed. If the ulna is significantly bowed, the bowing is corrected at the time of the initial application of the Ilizarov apparatus, when the patient is between the ages of 1 and 2 years. Correction of the bowing is accomplished by using four half-pins instead of two half-pins on the frame, with an osteotomy performed proximally and an acute correction (Fig. 19A-D).

Figure 19. A, Radiographs of a 2-year-old child with unilateral radial club hand with marked ulnar bowing. B, Apparatus was applied as described in the legend to Figure 18H. Four proximal pins were used, with an extension osteotomy between the two pairs of pins. C, Radiograph obtained at the time of the tendon transfer after completing the distraction centralization. D, Anteroposterior radiograph obtained after realignment of the ulna and hand.
It is important to correct the bowing initially. Otherwise, it is not possible to get the tendons on the correct side of the center of rotation of the wrist, which is in the ulnar head. The tendons sublux to the radial side, and the deformity tends to recur. If the bowing is minimal, it can be corrected at the time of the lengthening through a proximal osteotomy.

Lengthening is performed using the Ilizarov apparatus. The construct consists of two half-pins in the proximal ulna, two half-pins in the distal ulna, and one or two wires in the metacarpals. The device has a ring on the proximal ulna, a ring on the distal ulna, and a half-ring for the hand. The hand is always stabilized. If a pollicization has been performed, then a wire or small half-pin is inserted into the base of the metacarpal (Fig. 20A-G).

Figure 20. A, Clinical photograph of a 6-year-old girl who had undergone previous acute centralization and pollicization shows the length discrepancy between her left and right forearms. B, Clinical photograph obtained after 7 cm of lengthening of the left ulna.

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Figure 20 (Continued). C, Radiographs comparing the length of the ulna before and after the first lengthening. D and E, Patient’s function after the first lengthening was improved compared with preoperatively.

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Figure 20 (Continued). F, At the age of 11 years, the patient underwent a second lengthening of the ulna. As with the previous lengthening, the hand and thumb were stabilized with the Ilizarov pins and apparatus. G, Radiographic comparison of the length of the ulna initially (top), after the first lengthening (middle), and after the second lengthening (bottom).
In any patient for whom a pollicization has been performed, the thumb must be considered an unstable joint that needs stabilizing during lengthening. Otherwise, it will sublux or dislocate. The fingers become tight during the distraction, and it is the role of physical or occupational therapy to maintain finger range of motion, particularly extension, while obtaining more flexion because of the increased tension of the muscles. If the therapist and patient work hard, motion can be maintained and additional motion can be obtained. Because the fingers flex down, one often increases the range of flexion of the fingers without losing extension. One to 2 hours of physical or occupational therapy per day along with splinting of the fingers in extension is essential.

The second lengthening is performed when the patient is older than 10 years, usually between the ages of 12 and 14 years, when an amount of lengthening between 8 and 10 cm can likely be attained (Fig. 20F and G). A circular fixator usually is used, but a mono lateral fixator is used occasionally, especially if there is no deformity and a very stiff or fused wrist. If the hand needs to be stabilized because there is wrist motion or because a pollicization has been performed, then the circular fixator is a better choice because it can be extended to the hand and thumb with wires. The need for deformity correction, especially gradual deformity correction, is an indication for a circular fixator as opposed to a mono lateral fixator. If there is a radius that needs to be fixed proximally, that is another indication for a circular fixator, which can fix both bones.

Proximal lengthenings are preferred to distal lengthenings in the ulna if the wrist is centralized and corrected. If there is a recurrent wrist deformity and the patient is older than 6 years, then a distal osteotomy and realignment are preferred, especially if the growth plate is closed.” If the growth plate is closed, an operation composed of a distal osteotomy, lengthening, and realignment is a good choice because the distal ulna will not remodel. If the growth plate is wide open and an osteotomy is performed, the growth plate is maloriented, facing sideways, and will grow the wrist back into a deformed position. Therefore, if the growth plate is open and healthy, distal osteotomy realignment should not be performed. If the growth plate is closed and the wrist is stiff, distal realignment through a distal osteotomy with lengthening is preferred (Fig. 21A - E).
Figure 21. A and B, Radiographs of the hand and forearm of an 8-year-old girl with recurrent radial deviation flexion deformity obtained after previous acute centralization. The deformity is a bony one. The distal ulnar physis is closed as a complication of the previous centralization. Therefore, there is no further remodeling potential of the distal ulna. The index finger was previously pollicized. C, Ilizarov apparatus was applied and a distal ulnar osteotomy performed. D and E, Lateral and anteroposterior view radiographs obtained during distraction, after 4 cm of lengthening.
Paley's Type 4: Disproportionate Shortening of Both Bones of the Forearm

When both bones are short, but one is shorter than the other, the main indication for treatment is equalization of the length of both bones and correction of associated deformities such as distal radial angulation and proximal radial head dislocation. Multiple osteochondromas are a typical cause of disproportionate shortening of both the radius and the ulna. The problems associated with multiple osteochondromas include dislocation of the radial head, deformity of the distal radius, and, usually, a short ulna relative to the radius. If there are any large osteochondromas, they should be resected. At the proximal radius, there is usually a valgus deformity of the neck of the radius, and even when the radius is pulled out to length while lengthening the ulna, an osteotomy of the neck of the radius usually is needed to get the head of the radius back into joint.

Treatment involves a three-step process. step 1 is to resect any large osteochondromas that are blocking forearm rotation; step 2 is to lengthen the ulna with the radius and realign the distal radius with a distal radial osteotomy; step 3 is to perform an osteotomy of the neck of the radius proximally and reduce the radial head into the radiocapitellar joint. The result is a fully reduced elbow and a radius and ulna that are out to length (Fig. 22).
Figure 22. A, Anteroposterior radiograph of a 13-year-old boy with multiple osteochondromas. The radial head is subluxed, and there is a valgus deformity of the neck of the radius. The ulna is short relative to the radius. The hand is ulnarly deviated because of the increased distal radial tilt. B, Anteroposterior radiograph of the elbow after lengthening of the ulna with distal transport of the radius. An open reduction of the radial head into the joint was performed together with an acute varus osteotomy of the radial neck. C, Final anteroposterior and lateral radiographs obtained several years after the correction. There are large kissing distal osteochondromas that need to be removed because they are blocking forearm rotation.
Paley Type 5: Symmetrical Shortening of Both Bones of the Forearm

When the forearm is short overall without a discrepancy between the two bones, the decision for lengthening depends on the function of the forearm. If there is normal supination and pronation, then lengthening of both bones risks losing some forearm rotation. Therefore, functional gain from a longer forearm is overshadowed by greater functional loss from loss of rotation. The only indication for lengthening in such a situation is when the forearm is extremely short.

References

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