Residual deformity in congenital radial club hands after previous centralisation of the wrist

ULNAR LENGTHENING AND CORRECTION BY THE ILIZAROV METHOD

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We used the Ilizarov method in seven patients with severe congenital radial club hands who had had previous wrist surgery, to correct residual shortening and bowing of the ulna together with recurrent wrist deformity.

The mean age at operation was 6.5 years. The mean ulnar shortening was 5.3 cm and the mean angular deformity 42°.

The mean length gained was 51% of the original ulna. The mean healing index was 46.9 days (29.8 to 64.0). The ratio of the length of the lengthened ulna to the normal side improved on average from 64% to 95%. The angular deformity was initially completely corrected in six out of seven patients. The length ratio, however, decreased to 83% at the final follow-up. In four patients, the angular deformity partially recurred.

We recommend correction of congenital radial club hand by staged procedures. The first is centralisation and stabilisation of the wrist and the second lengthening of the ulna and correction of the angular deformity using the Ilizarov method.

J Bone Joint Surg [Br] 1998;80-B:762-5.

Received 17 February 1998; Accepted 24 March 1998

Congenital radial club hand represents a "failure of formation of parts" (category I) in the classification of congenital limb malformations of Swanson, Swanson and Tada.¹ The hand without the bony support of the radius shows marked radial deviation at the wrist. There is also shortening and bowing of the ulna. Current methods of treatment are concerned largely with correction of the radial deviation at

the wrist and pay little attention to the ulnar hypoplasia.²⁻⁷ Recurrence of the radial deviation has been commonly reported leading to unsatisfactory functional and cosmetic results.

The introduction of the Ilizarov method has dramatically changed the possibilities for treatment for many congenital and acquired conditions. We have used this procedure to correct severe residual shortness and bowing of the ulna and recurrent radial deviation at the wrist in congenital radial club hands in which a previous wrist operation had been performed.

Patients and Methods

We treated seven congenital radial club hands in seven patients by the Ilizarov method (Table I). One patient had bilateral radial club hands but only one side was included in our study. The contralateral radius in the remaining six patients was normal but the thumb was hypoplastic in four. All the patients had undergone previous centralisation of the wrist or radialisation of the ulna. The indications for the Ilizarov operation were residual shortening and bowing of the ulna with recurrent radial deviation of the hand causing functional and cosmetic problems for the patient and his/her family. The mean age at operation was 6.5 years (4 years 9 months to 12 years). The mean deficiency in length of the ulna was 5.3 cm (4.3 to 7.1) and the mean percentage length, which was defined as the ratio in length to the contralateral normal ulna, was 64%, excluding one bilateral case. The mean angular deformity was 42° (0 to 65). The mean follow-up was 3.6 years (2.1 to 5.6).

The exact surgical technique and postoperative management have been previously reported.⁸ Briefly, the Ilizarov apparatus consisted of two full rings (Fig. 1). Each ring was fixed to the ulna by two 1.5 mm Ilizarov smooth wires and one 3 mm half-pin or one wire and two half-pins. The two rings were connected by three threaded rods. The two rods on the convex side had a hinge at the level of the apex of the angular deformity. A 1.5 mm Ilizarov olive wire, which was inserted into the metacarpals from the radial to the ulnar side was connected to the distal ring by two posts. The metacarpal wire fixed the wrist in maximal dorsoulnar deviation and prevented radial deviation during lengthening. We performed an ulnar osteotomy subperiosteally

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0301-620X/98/58839 \$2.00

Table I. Details of seven patients with congenital radial club hands

Case	Age (yr/mth)		Sex	Preop			Postop*			Length gained (cm/%) [‡]	Time in frame (days)	HI [§] (days)	Follow-up (yr)	Ulnar length		Deformity (degrees) [†]	Complications
				cm	%	(degrees) [†]	cm	%	(degrees) [†]					cm	%		
1	12	0	M	14.5	71	65	20.5	100	25	6.0/41	204	34.0	5.6	21.8	97	40	
2	6	9	F	9.3	57	47	17.3	105	0	7.3/78	314	64.0	4.9	18.4	83	35	Pin-track infection, delayed consolidation
3	4	11	M	8.7	64	62	11.6	85	0	2.9/33	201	57.0	3.4	12.0	68	35	Early consolidation
4	4	9	F	7.0	56	25	10.5	85	0	3.8/54	167	43.9	3.5	11.6	76	0	
5	5	4	M	8.0	NA	55	13.0	NA	0	5.0/63	247	49.4	2.1	12.6	NA	0	Callus fracture
6	5	5	M	8.6	67	45	11.8	91	0	3.2/37	181	50.3	3.3	12.9	82	25	
7	6	0	M	9.4	69	0	14.3	104	0	4.7/50	140	29.8	2.6	13.7	94	0	Pin-track infection

* at four weeks after pin removal

† the sum of ulnar bowing and radial deviation of the hand at the wrist

‡ gain ratio to preoperative length

§ healing index

through a short incision using an osteotome. No soft-tissue releases or tendon transfers were performed.

Lengthening and correction of the angular deformity were started at seven days on the concave side using the threaded rod. Lengthening was performed at 0.25 mm per 12 hours. Correction continued until the forearm-hand unit appeared straight. After correction had been completed, bone lengthening was continued until the ulna was the same length as the contralateral side, provided that there was good callus formation. Radiographs were taken weekly to confirm the quality of the callus and the speed of lengthening was adjusted to avoid premature consolidation or poor formation of callus. When the callus had matured, we removed all the pins and wires without anaesthesia and applied a long-arm cast for four weeks. A night splint was then worn for at least one year.

Results

Satisfactory lengthening of the ulna was achieved in all patients. Details of the results are given in Table I. The mean period of lengthening was 124 days (101 to 140). The mean length of time in the frame was 208 days (140 to 314). The mean healing index was 46.9 days (29.8 to 64.0). The mean gain in length was 4.7 cm (2.9 to 7.3) which was 51% (33 to 78) of the original ulna. In the unilateral cases, the mean percentage length improved to 95% at the end of immobilisation in the cast. Complete angular correction was obtained at the time of pin removal and only the first patient who did not have immobilisation in a cast showed immediate deterioration within four weeks of pin removal.

The percentage length had decreased to 83% at a mean follow-up of 3.6 years. The smallest decrease was in a boy (case 1) who was 12 years old at the time of operation and who was followed until the end of growth. Bowing of the ulna and radial deviation of the hand showed a tendency to recur in four out of seven patients. The mean angular deformity was 19° at final follow-up, i.e., 57% of the

Fig. 1

The Ilizarov apparatus. The frame consists of two full rings each of which is fixed by two 1.5 mm Ilizarov smooth wires and one 3 mm half-pin. The wrist is stabilised by one olive wire.

correction had been maintained. Toileting and self-feeding activity were improved due to the increase in space accessibility of the hand.

There were no major complications. Pin-track infections, seen in two patients, responded well to oral antibiotics. There were no cases of deep soft-tissue or bone infection. Although flexion contracture of the elbow and fingers was seen during the lengthening period, it was transient and improved rapidly after pin removal. The arc of movement of the wrist did not change significantly, although it shifted radially as a result of the correction. Pinch power was improved in some patients. There were no neurological complications. One patient showed delayed consolidation of callus. The ulnar length did not equal that of the

Fig. 2a

Fig. 2b

Fig. 2c

Fig. 2d

Fig. 2e

A six-year-old boy with a left radial club hand had had centralisation with tendon transfers of the radial wrist flexor and extensor to the ulnar wrist extensor at the age of four years. His left thumb was hypoplastic but was not treated. The wrist was well stabilised with an active range of movement of 70° flexion, 0° extension, 50° radial deviation, and 0° ulnar deviation. The elbow flexed to 125° and extended to -20°. Radiographs show a) the extremely short ulna (left) before operation with the hand in a good position, b) immediately after operation, c) completion of the correction of the angular deformity, d) completion of lengthening of 4.9 cm and e) equalisation of the length of the ulna (left) with that of the contralateral side. The range of movement increased in ulnar deviation and extension, while flexion did not change and radial deviation decreased to 25°. The range of movement of the elbow did not deteriorate.

contralateral ulna in three patients. Two showed poor callus formation and the other early consolidation. There was one fracture of the callus after pin removal which was successfully treated by immobilisation in a cast. All the patients tolerated the Ilizarov apparatus well and they and their families with one exception (case 2) were satisfied with the results. An illustrative case is shown in Figure 2.

Discussion

The goal of treatment of radial club hand is a functional and cosmetically acceptable upper limb. A stable mobile wrist and sufficient forearm length are important prerequisites for achieving this. Bora et al³ stressed the importance of forearm length, although the final length of the ulna in their series was only 50% of normal. They also accepted a mean angular deformity of 35° and concluded that their centralisation procedure was satisfactory. Bayne

and Klug⁶ did not recommend corrective osteotomy of the ulna if the angular deformity was less than 30°.

In our series using the Ilizarov method we achieved a mean ulnar lengthening of 51%. The affected ulna was nearly equal to the contralateral normal side at the end of lengthening. At final follow-up, however, the percentage length had deteriorated to 83%. This may be because the ulnar physis in this disease has an intrinsic poor growth potential. Bone lengthening may also give rise to an excessive compression force on the ulnar physis which further reduces growth. To avoid such an adverse effect it may be wise to postpone lengthening until growth stops. If, however, lengthening is performed after maturity a gain in length of nearly 100% will be required for equalisation since the natural history of this disease has shown that the final shortening is as much as 40% of the normal ulna.⁹ The percentage length gain in the ulna in the patients of Cattagni, Szabo and Cattaneo,¹⁰ who were treated at 12 to 23

years of age, was less than 23% except in one. There were considerable complications in this series. Our patients had no complications involving nerves or joints probably because they were much younger (five to 12 years of age).

We believe that the cause of recurrent angular deformity in our patients was muscle imbalance which was exaggerated by the lengthening. Catagni et al¹⁰ advised arthrodesis of the wrist but this should be avoided if possible because of the loss of movement. In our series, the recurrence of angular deformity was less in a wrist with well-balanced muscle forces (Fig. 2). In this respect, the first centralisation procedure is of great importance in relation to the Ilizarov operation.

We have experienced one hand, not included in this series, in which centralisation had not been previously done. The Ilizarov procedure was performed as the first operation to correct the entire deformity. Complete recurrence occurred due to persisting muscle imbalance and instability of the wrist.

In the arm, limb-length discrepancy is not as critical as in the leg. In radial club hand, however, the length of the ulna on the affected side may be nearly half that of the normal and the cosmetic handicap is severe enough to justify limb lengthening. We recommend the following surgical strategy for the treatment of congenital radial club hand. Before the age of one year a stable, well-balanced, and mobile wrist is achieved using standard centralisation techniques.⁴⁻⁶ The

first Ilizarov operation is performed at five to six years of age and the second at 12 years. This divides the bone lengthening into two stages, reducing the amount of lengthening at each operation and protecting the soft tissues and physes from adverse effects.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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