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Treatment of severe radial club hand by distraction using an articulated mini-rail fixator and transfixing pins



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ABSTRACT

Introduction: Treatment of severe radial club hand is difficult. Several authors have emphasized the importance of preliminary soft-tissue distraction before centralization.

Hypothesis: Treatment of severe radial club hand by articulated mini-rail allowing prior soft-tissue distraction improves results.

Material and methods: Thirteen patients were treated sequentially, with an initial step of distraction and a second step of centralization. The first step consisted in fitting 2 mini-fixators, one in the concavity and the other in the convexity of the deformity. Four transfixing wires through the ulna and metacarpal bone connected the 2 fixators. After this preliminary distraction, the fixator was removed and a centralization wire was introduced percutaneously, with ulnar osteotomy if necessary. Sagittal and coronal correction was measured on the angle between forearm and hand.

Results: Mean age at treatment was 37.5 months (range, 9–120 months). Mean distraction time was 53.2 days (26–90 days). Ulnar osteotomy was required in 8 cases (61%). There were no major complications requiring interruption of distraction. Sagittal and coronal correction after centralization reduced mean residual forearm/hand angulation to $<12^{\circ}$.

Discussion: Soft-tissue distraction in the concavity ahead of centralization is essential to good correction, avoiding extensive soft-tissue release and hyperpressure on the distal ulnar growth plate. There have been several studies of distraction; the present technique, associating 2 mini-fixators connected by threaded K-wires, provided sufficient distraction in the concavity of the deformity to allow satisfactory correction in all cases. Subsequent complications (breakage or displacement of the centralization wires) testify to the complexity of long-term management.

Conclusion: The present study confirms the interest of a preliminary soft-tissue distraction step in treating severe radial club hand.

Level of evidence: IV.

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1. Introduction

Radial club hand is a rare abnormality, with incidence of 1/300,000 live births [1]. It involves the lateral part of the upper limb, with radial aplasia or hypoplasia. It comprises not only bone abnormalities but also cutaneous/muscular and neurovascular abnormalities of the lateral part of the upper limb, generally sparing the ulnar structures. Clinically, there is radial inclination of the hand, associated with palmar flexion-pronation. Apart from the

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severe esthetic defect, radial club hand induces functional deficits due to the shortened forearm, unstable wrist and reduced extrinsic tendon course [2].

Radial club hand is graded in 4 types according to the severity of hypoplasia [3], the more severe forms being associated with greater carpal displacement with respect to the distal ulna. Several treatments have been reported to achieve lasting alignment of the hand with respect to the forearm: replacing the radius by vascularized epiphysis transfer from a toe [4], radialization to position the carpus facing the distal ulna [5], or centralization [2,3,6,7].

The degree of shortening of the concavity structures in severe forms requires extensive soft-tissue release during the centralization procedure, often associated to carpal resection so as to house the ulnar head, leading to further shortening of an already short forearm. Kessler, however, demonstrated that centralization

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could also be achieved by soft-tissue distraction [8]. Subsequently, several authors reported the benefits of distraction ahead of centralization or radialization [9–13], but used various different fixators, with small series.

The present study sought to assess the technique and efficacy of uniplanar soft-tissue distraction ahead of centralization, associating transfixing K-wires and two articulated mini-rails, in a series of severe radial club hand.

2. Material and methods

Thirteen patients were included, presenting severe radial club hand with total Bayne type-4 (n=11) or subtotal type-3 (n=2) radial agenesis. Clinical examination, preoperatively and during successive follow-up, assessed shoulder and elbow range of motion, wrist deformity and digital chain range of motion. Six patients with elbow stiffness in extension received preliminary rehabilitation and orthosis in flexion to achieve 90° preoperative flexion. Complete assessment, performed by geneticists, screened for associated abnormalities (Table 1). Surgical indications were confirmed in multidisciplinary consultation, with occupational therapy assessment in some cases to ensure that centralization would not impair the child's functional capacities.

All patients were managed by centralization after prior distraction. Under general anesthesia, two articulated Orthofix (M122) distractors were employed, one in the concavity and the other in the convexity of the deformity. They comprised a mini-rail articulated

at the rotational center of the wrist. An AP view was taken under fluoroscopic control, with the wrist in traction. A radio-opaque marker was positioned to identify the theoretic center of the carpus, which is not ossified at the age of 1 year, and the corresponding skin mark was made, using a dermographic pen, to position the distractors. Two parallel threaded or unthreaded transfixing K-wires were placed in the ulna and two in the metacarpus to connect the distractors (Fig. 1). In case of subsequent pollicization, the metacarpal K-wires were positioned so as not to damage the dorsal or palmar structures of the index. Due to the metacarpal arch, only 2 or 3 metacarpi were fixed by the wires. Distraction was initiated immediately upon fixation, at a rate of 2-4 quarter turns per day, 1 full turn achieving 1 mm lengthening. Pin care was performed daily, and rehabilitation of the elbow and digital chains was performed throughout distraction. Lengthening was faster in the concavity (4 quarter turns per day) than in the convexity (2 per day), so as to progressively realign the wrist. Distraction was performed under day care, and efficacy was checked on successive radiographs taken during weekly follow-up. If K-wire tension appeared excessive, distraction was slowed down so as to allow the skin and capsule-ligamentous structures to relax progressively. Once correction of the radial deviation and wrist flexion was achieved (usually within 3–4 weeks), the fixator was left in position for a few days before centralization was performed, to allow the soft tissue to relax. Centralization was performed under fluoroscopic control, with no surgical approach to the wrist, using a proximal-to-distal Kwire through the metacarpus of the index or middle finger. Ulnar

 Table 1

 Associated abnormalities, preoperative elbow range of motion, and thumb surgeries.

Patient	Association	Range of passive elbow flexion (degrees)	Thumb
1	Cardiac (VSD), hearing loss, contralateral thumb hypoplasia	110	Index pollicization
2	Cardiac (VSD, persistent arterial canal), psychomotor retardation	90	-
3	Hypospadias, large vessel malpositioning, thumb hypoplasia	70	Index pollicization
4	Isolated	120	Index pollicization
5	Spine (binucleate T10 vertebra)	110	Index pollicization
6	Testicular ectopia	70	-
7	VACTERL	100	
8	Dextrocardia, cervical spine deformities	N/A	Index pollicization
9	Spinal, cardiac	60	Index pollicization
10	Cardiac (VSD), renal (incomplete duplicity, mega-ureter), low ears	90	Index pollicization
11	Isolated	80	
12	TRA, G6PD deficit	N/A	
13	TRA, contralateral thumb hypoplasia	120	

VACTERL: association of at least 3 of the following: vertebral anomalies, anal atresia, cardiac defects, tracheoesophageal fistula and/or esophageal atresia, renal anomalies and limb defects; VSD: ventricular septal defect; TRA: thrombocytopenia+radial aplasia; N/A: information not available.





Fig. 1. A. Bayne type-4 severe radial club hand. B. Positioning 2 mini-fixators connected by transfixing K-wires.

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