Original article

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

C. Romana, a, b, G. Ciais, a, F. Fitoussi, a, b, *

a Service de chirurgie orthopédique et réparatrice de l’enfant, UPMC université Paris 06, hôpital Trousseau, Paris, France
b Centre de référence des malformations des membres, hôpital national de Saint-Maurice, Saint-Maurice, France

ARTICLE INFO

Article history:
Received 14 August 2014
Accepted 24 February 2015

Keywords:
Radial club hand
Distraction
Centralization

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°.

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 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
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 K-wire through the metacarpus of the index or middle finger. Ulnar

Table 1

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

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.
osteotomy was associated in 8 cases, depending on the degree of curvature (Fig. 2). Postoperative immobilization was imposed only in case of associated ulnar osteotomy.

In 3 patients, two distractors were positioned in the convexity, due to the severity of the deformity. In 1 patient, the articulated fixators were replaced by non-articulated mini-rails (M 103) once alignment had been achieved, to improve carpal descent (Fig. 3).

The angulation and translation of the hand with respect to the ulna were measured on preoperative and post-distraction radiographs. Angulation was measured following Manske et al. [7]: the axis of the hand is represented by the longitudinal axis of the metacarpus of the middle finger; the distal ulna axis is represented by the perpendicular to the middle of the distal epiphyseal region (Fig. 4). The angle between the two represents the angulation of the hand. Translation is measured by the distance D between the base of the metacarpus of the middle finger and the prolongation of the distal ulna axis (Fig. 5) [12]. Preoperatively, mean angulation was 52.3° and translation 15.2 mm.
3. Results

Table 3 presents the results for the 13 patients (11 boys, 2 girls). Mean age at surgery was 3 years (9–120 months). The older ages were due to late referral. Mean distraction time was 53.2 days (range, 26–90 days). Mean post-distraction angulation was 11.2°, with 41.1° mean correction (p < 0.05). Mean postoperative translation was 5.8 mm, with 9.4 mm mean correction (p < 0.05). On Kanojia’s criteria, 9 results were satisfactory, 3 good and 1 poor.

There were 2 early complications: one prominent K-wire inducing skin ulceration at the elbow, and 1 threaded wire replacement during distraction. One patient had a poor result on Kanojia’s criteria, due to insufficient coronal and sagittal correction.

4. Discussion

Severe radial club hand is difficult to treat. The main objective is to stabilize the carpus in alignment with the forearm, to improve both esthetics and also function by enhancing flexor digitorum force (Fig. 6). Centralization is employed in severe forms, not to reconstruct the radius but rather using the ulna as forearm growth axis [14]. The original technique involved extensive soft-tissue release in the concavity and the creation of a notch in the carpus to receive the ulnar head, achieving better stability but at the cost of further shortening of the forearm.

4.1. Efficacy of distraction

Some authors therefore recommended a distraction step ahead of centralization or radialization [8,10–13,15]: lengthening both concavity and palmar soft tissue avoids extensive skin and joint release; moreover, a large carpal descent avoids further shortening of the forearm by resection of the carpus [6,11]. Series, however, were small and, despite the correction achieved after distraction, the principal issue of maintaining long-term correction remains.

The present study confirmed the efficacy of distraction using two mini-rails, probably due to force applied in the concavity of the deformity, reducing mean axial deviation from 52.3 to 11.2° and translation from 15.2 to 5.8 mm. Articulated mini-rails were used, being easier to deploy, but we imagine that circular fixators, well-adapted for multplanar correction, could also be adopted. The distractors we used had only one articulation, whereas the deformity is in several planes. We positioned them in relation to the rotational center of the wrist seen on AP view (Fig. 7). Even so, we consider concavity soft-tissue distraction to be the key point to enable easy carpal reduction at the ulnar head during centralization. The forearm K-wires should be inserted via a minimally

Table 2

| Coronal hand/forearm angle | 2 |
| Sagittal hand/forearm angle | 1 |
| Ulnar growth | 0 |

Final assessment adopted the modified criteria described by Kanojia et al. [11], taking account of forearm/hand angle, flexion contracture of the wrist and ulnar growth (Table 2). Pre- versus postoperative data were compared on Student t test.

Table 3
Patient data.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Gender</th>
<th>Age at surgery (months)</th>
<th>Bayne type</th>
<th>Preoperative angulation (degrees)</th>
<th>Preoperative translation (mm)</th>
<th>Distraction time (days)</th>
<th>Postoperative angulation (degrees)</th>
<th>Postoperative translation (mm)</th>
<th>Ulnar osteotomy</th>
<th>Results (Kanojia)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>20</td>
<td>IV</td>
<td>30</td>
<td>12</td>
<td>90</td>
<td>10</td>
<td>5</td>
<td>No</td>
<td>Satisfactory</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>19</td>
<td>IV</td>
<td>63</td>
<td>13</td>
<td>26</td>
<td>8</td>
<td>5</td>
<td>No</td>
<td>Satisfactory</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>15</td>
<td>IV</td>
<td>70</td>
<td>14</td>
<td>60</td>
<td>20</td>
<td>7</td>
<td>Yes</td>
<td>Poor</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>96</td>
<td>IV</td>
<td>39</td>
<td>13</td>
<td>50</td>
<td>13</td>
<td>8</td>
<td>No</td>
<td>Good</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>42</td>
<td>IV</td>
<td>84</td>
<td>16</td>
<td>30</td>
<td>16</td>
<td>3</td>
<td>Yes</td>
<td>Satisfactory</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>32</td>
<td>III</td>
<td>43</td>
<td>18</td>
<td>40</td>
<td>5</td>
<td>16</td>
<td>Yes</td>
<td>Satisfactory</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>30</td>
<td>IV</td>
<td>N/A</td>
<td>N/A</td>
<td>60</td>
<td>13</td>
<td>5</td>
<td>Yes</td>
<td>Good</td>
</tr>
<tr>
<td>8</td>
<td>M</td>
<td>120</td>
<td>IV</td>
<td>52</td>
<td>15</td>
<td>N/A</td>
<td>16</td>
<td>5</td>
<td>No</td>
<td>Good</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>19</td>
<td>IV</td>
<td>N/A</td>
<td>N/A</td>
<td>68</td>
<td>8</td>
<td>4</td>
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<td>Satisfactory</td>
</tr>
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<td>106</td>
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<td>60</td>
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<td>Yes</td>
<td>Satisfactory</td>
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<tr>
<td>11</td>
<td>M</td>
<td>11</td>
<td>IV</td>
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<tr>
<td>12</td>
<td>M</td>
<td>12</td>
<td>IV</td>
<td>16</td>
<td>14</td>
<td>N/A</td>
<td>11</td>
<td>6</td>
<td>No</td>
<td>Satisfactory</td>
</tr>
<tr>
<td>13</td>
<td>M</td>
<td>35</td>
<td>IV</td>
<td>43</td>
<td>15</td>
<td>34</td>
<td>5</td>
<td>2</td>
<td>Yes</td>
<td>Satisfactory</td>
</tr>
</tbody>
</table>

N/A: information not available.
invasive approach, to spare vascular and neural structures. In case of severe associated hypoplasia of the thumb, subsequent pollicization should be taken account of in inserting the metacarpal wires, on an out-in route, sparing the dorsal venous structures and palmar vasculo-neural pedicles of the index. Seven patients subsequently underwent pollicization, with no particular complications.

4.2. Stabilization during growth

To improve wrist stability and maintain correction, some authors recommended dorso-radial muscle transfer toward the ulnar side of the carpus [3]. However, this seems not to provide lasting correction, especially in severe forms [16,17]. The causes of recurrence of radial deviation found in the literature comprise residual retraction on the radial side of the wrist, premature ablation of the stabilization wire, and poor quality of transferred muscle [5,18].

We did not use tendon transfer; our technique was exclusively percutaneous, relying more on the centralization wire than on transferred muscle quality (which was often poor) to maintain correction during growth [14]. Even so, K-wire stabilization is not free of complications: wire migration or breakage, skin ulcer, and iterative changes following growth. Despite such repeated revision, correction loss is acceptable and the fixation finally stabilizes over growth. The problem of lengthening the short forearm remains to be dealt with at the end of growth, in a multidisciplinary consultation, notably including the occupational therapists.

4.3. Study limitations

The study was retrospective, with a small series. Radiographic assessment of axial correction was only in the coronal plane, whereas the deviation also involved palmar flexion; however, sagittal deviation was assessed clinically.

5. Conclusion

The present study confirmed the usefulness of initial soft-tissue distraction in the treatment of severe radial club hand. The distraction technique was uniplanar but was simpler than and at least as effective as other reported techniques. The distraction force applied by fitting a mini-fixator in the concavity provided satisfactory correction in terms of angulation and translation. The remaining problem is to maintain correction during growth. Inserting a centralization K-wire, despite numerous inconveniences, stabilizes the carpus during axial growth of the ulna.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References