LENGTHENING THE ULNA IN PATIENTS WITH HEREDITARY MULTIPLE EXOSTOSES

JAMES W. PRITCHETT

From the Pacific Medical Center, Seattle

Deformity of the forearm is common in patients with hereditary multiple exostoses, producing cosmetic and functional impairment in which shortening of the ulna is a significant factor. The results of ulnar lengthening in 10 forearms of eight patients are reported.

Lengthening was performed by osteotomy of the shaft followed immediately by a bone graft and internal fixation, or by gradual distraction with an external fixator. In all patients the appearance was improved and the range of radial deviation at the wrist was increased. In most patients forearm movement and radial head stability were improved. Partial recurrence of the deformity was seen during the follow-up of skeletally immature patients, but in general ulnar lengthening was found to be a useful operation.

The condition of hereditary multiple exostosis, also called diaphyseal aclasis, or metaphyseal aclasis (Solomon 1963; Shapiro, Simon and Glimcher 1979; Fogel et al. 1984) is a developmental anomaly in which there is heterotopic proliferation of epiphyseal chondroblasts. It is characterised by defective metaphyseal remodelling, producing exostoses and growth retardation (Solomon 1961).

There are deformities of the forearm in 60% of patients with this condition (Shapiro et al. 1979). The ulna is usually shortened as a result of deformity or of early closure of its distal physis. The radius becomes bowed to accommodate the ulna but, as length discrepancy increases, cubitus varus or radial head dislocation may occur (Figs 1 and 5). At the wrist there may be an ulnar tilt of the radial epiphysis with ulnar deviation of the hand and translocation of the carpus towards the ulna (Figs 6 and 9). These complex deformities cause a loss of radial deviation at the wrist and of pronation-supination of the forearm. They may also be aesthetically disturbing.

Treatments recommended in the past have included excision of the exostoses, radial shortening, stapling half of the distal radial epiphysis and resection of a dislocated radial head (Siffert and Levy 1965; McCormack 1981; Fogel et al. 1984). These procedures, however, all leave the patient with significant residual deformity. This paper describes correction of the deformity by ulnar lengthening, which may be immediate, or by gradual distraction using an external fixator.

MATERIALS AND METHODS

Eight patients from 7 to 18 years old with hereditary multiple exostoses and forearm deformity including a shortened ulna were treated by lengthening the ulna; they have been followed-up for a mean period of 37 months (range 27 to 60 months) (Table I).

All available radiographs of the wrist, forearm and elbow were reviewed. The tilt of the radial articular surface and the amount of carpal slip in an ulnar direction were measured and recorded. Carpal slip was measured as the percentage of the lunate not in contact with the radius. The relative ulnar shortening and stability of the ulnar head were also determined. The ranges of wrist, forearm and elbow movement were measured preoperatively and at the latest follow-up.

The indications for ulnar lengthening were shortening of the ulna by 1.5 cm or more, carpal slip of more than 50%, instability of the radial head, and symptomatic loss of wrist, elbow or forearm movement.

Three patients had immediate lengthening; in two (Cases 1 and 2) this was performed by placing an accurately cut tricortical iliac crest bone graft between the distracted ends of the ulna after a transverse diaphyseal osteotomy. The position and lengthening obtained was then secured with an AO dynamic compression plate and screws. Postoperative immobilisation was not needed. In the third patient (Case 3) bilateral lengthening of the ulna was held with Rush rods.

In five patients six lengthenings were performed using external fixation. Three pins were placed in the proximal third of the ulna and three in the distal third; a transverse mid-diaphyseal subperiosteal osteotomy was then performed. The external fixation clamp was then applied to produce distraction of about 1 cm. Postoperatively the ulna was lengthened by a further 1 mm per day; the neurovascular status of the limb was carefully
Table I. Results of ulnar lengthening in 10 forearms of 8 patients with hereditary multiple exostoses

<table>
<thead>
<tr>
<th>Case number</th>
<th>Age at operation (yr)</th>
<th>Age at operation (m)</th>
<th>Sex</th>
<th>Side</th>
<th>Follow-up (months)</th>
<th>Tilt of radial articular surface (degrees)</th>
<th>Carpal slip (per cent)</th>
<th>Relative ulnar shortening</th>
<th>Gain in length (cm)</th>
<th>Radial head before operation*</th>
<th>Ulnar lengthening</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>15</td>
<td>1</td>
<td>M</td>
<td>R</td>
<td>28</td>
<td>Before: 35 After: 15</td>
<td>Before: 10 After: 0</td>
<td>2.5 Before: 0 After: 2.5</td>
<td>2.5 Stable</td>
<td>Immediate</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>18</td>
<td>6</td>
<td>F</td>
<td>L</td>
<td>27</td>
<td>Before: 40 After: 25</td>
<td>Before: 10 After: 10</td>
<td>3.0 Before: 1.0 After: 2.0</td>
<td>2.5 Subluxated</td>
<td>Immediate, RO</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>13</td>
<td>6</td>
<td>F</td>
<td>R</td>
<td>35</td>
<td>Before: 50 After: 20</td>
<td>Before: 10 After: 10</td>
<td>1.5 Before: +0.5 After: 1.5</td>
<td>4.0 Subluxated†</td>
<td>Immediate, RO</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>7</td>
<td>6</td>
<td>F</td>
<td>L</td>
<td>33</td>
<td>Before: 45 After: 20</td>
<td>Before: 10 After: 10</td>
<td>1.5 Before: 0.3 After: 1.2</td>
<td>4.0 Dislocated†</td>
<td>Gradual, RO</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>7</td>
<td>7</td>
<td>M</td>
<td>R</td>
<td>32</td>
<td>Before: 25 After: 5</td>
<td>Before: 30 After: 30</td>
<td>3.5 Before: 0.5 After: 4.0</td>
<td>4.0 Full</td>
<td>Gradual, RO</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>8</td>
<td>7</td>
<td>F</td>
<td>L</td>
<td>36</td>
<td>Before: 35 After: 20</td>
<td>Before: 10 After: 10</td>
<td>2.4 Before: 0.2 After: 2.5</td>
<td>4.0 Full</td>
<td>Gradual</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>11</td>
<td>4</td>
<td>F</td>
<td>R</td>
<td>60</td>
<td>Before: 40 After: 15</td>
<td>Before: 50 After: 10</td>
<td>2.4 Before: 0.5 After: 3.0</td>
<td>4.0 Subluxated</td>
<td>Gradual</td>
<td></td>
</tr>
</tbody>
</table>

* All became stable after operation except †, which was subluxated RO, radial osteotomy.

Table II. Ranges of movement in degrees before and at the latest follow-up after ulnar lengthening

<table>
<thead>
<tr>
<th>Case number</th>
<th>Side</th>
<th>Supination-pronation</th>
<th>Ulnar deviation-radial deviation</th>
<th>Flexion-extension at the elbow</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>R</td>
<td>60-45 60-45</td>
<td>20-0 20-10</td>
<td>Full</td>
</tr>
<tr>
<td>2</td>
<td>L</td>
<td>45-30 60-45</td>
<td>40-0 20-20</td>
<td>10-130 0-140</td>
</tr>
<tr>
<td>3</td>
<td>R</td>
<td>45-20 70-60</td>
<td>30-0 20-20</td>
<td>10-130 Full</td>
</tr>
<tr>
<td>4</td>
<td>R</td>
<td>15-15 35-15</td>
<td>40-0 30-10</td>
<td>10-130 0-140</td>
</tr>
<tr>
<td>5</td>
<td>R</td>
<td>30-0 40-30</td>
<td>30-0 20-20</td>
<td>20-130 10-140</td>
</tr>
<tr>
<td>6</td>
<td>L</td>
<td>35-25 35-25</td>
<td>30-0 20-10</td>
<td>20-120 10-130</td>
</tr>
<tr>
<td>7</td>
<td>R</td>
<td>30-10 30-10</td>
<td>30-0 20-20</td>
<td>Full</td>
</tr>
<tr>
<td>8</td>
<td>L</td>
<td>25-15 60-20</td>
<td>30-0 30-20</td>
<td>10-140 Full</td>
</tr>
</tbody>
</table>

observed. The osteogenic potential of the periosteal tube made bone grafts unnecessary (Figs 2 and 7). Union occurred uneventfully in all cases and there were no neurovascular complications.

RESULTS

These are given in detail in Tables I and II and three cases are shown in Figures 1 to 13. In all the patients the appearance was improved and the range of radial deviation increased. Three forearms showed no improvement in pronation-supination, but seven had an average of 25° improvement. Tilt of the radial articular surface and carpal slip were also improved in all 10 patients.

The ulna was lengthened by an average of 1.8 cm in the immediate distraction group (range 1.2 to 2.5 cm) and by 2.8 cm in the gradual distraction group (range 2.3 to 4.0 cm). Radial head stability was improved in five of the six forearms that had instability before operation (Cases 2, 3, 4, 5 and 8). In these five patients elbow movement was limited before operation but improved after it. One patient (Case 5) later required a proximal radial osteotomy but still had some instability of the radial head (Figs 1 to 4).

Deep infection occurred in one of the immediate lengthening group (Case 1). The plate and screws were removed and an external fixator was applied; the ulna then healed and the infection cleared rapidly. One ulnar
osteotomy failed to unite but was asymptomatic (Case 3). In this case immediate lengthening with stabilisation by Rush rod had been performed (Figs 9 to 13), and although the ulna was slightly overlengthened, no additional procedure was needed as the patient was satisfied with the result. There were no other complications in the series.

Ulnar shortening recurred with growth in all six operated forearms in the five skeletally immature patients. The rate of recurrence varied from about 0.4 cm/year at the age of 8 years (Figs 4 and 8) to approximately 0.25 cm/year at the age of 12 years.

DISCUSSION

The forearm deformity caused by shortening of the ulna relative to the radius in patients with hereditary multiple exostoses is related to three factors:

1. The cross sectional area of the distal ulnar physis is only one-quarter of that of the distal radius, so that any heterotopic loss of chondroblasts will result in loss of more longitudinal growth in the ulna than in the radius (Fogel et al. 1984).

2. The distal ulna is more commonly involved by the condition than the distal radius (Shapiro et al. 1979).

3. There is more longitudinal growth at the distal ulnar physis than at the distal radial physis.

In addition to the cosmetic deformity there is impaired function. The exostoses may mechanically disturb adjacent joints but the tethering effect which causes the radius to develop a medial concavity is more significant. The axis of rotation of the forearm normally runs from the distal ulnar physis through the centre of the radial head, and forearm deformity of more than 10° will cause loss of movement due to distortion of this axis (Matthews et al. 1982). With increasing deformity the superior and inferior radio-ulnar joints may become displaced and more movement will be lost. Subluxation or dislocation of the head of the radius may limit flexion and extension of the elbow (McCornack 1981), while the slowly progressive ulnar deviation of the wrist and translocation of the carpus contribute to diminished efficiency of the hand (Dal Monte, Andrisano and Capanna 1980).

Excision of exostoses has not been effective in controlling the progression of deformity (Fogel et al. 1984) nor has resection of the radial head proved satisfactory. Radial hemi-epiphyseal stapling, used alone or with ulnar lengthening, has been effective but causes shortening of the forearm which may be objectionable to some patients.
Case 4. Figure 5  Radiograph of the right forearm of a six-year-old girl with limited pronation-supination movement, showing bowing of the forearm. Figure 6  One year later the radial head has subluxated. Figure 7  The ulna was gradually lengthened by 2.5 cm, overcorrection of 0.8 cm being designed to offset subsequent recurrence of shortening, expected because of her bony immaturity. The radius was straightened by osteotomy. Figure 8  At 12 years of age the improved pronation-supination movement and radial deviation of the wrist have been maintained. The radial head is stable and there is no cosmetic deformity. The ulna is one cm shorter than the radius.

Case 3. Figures 9 and 10  Radiographs of the forearm of a 13-year-old girl who presented with severe bowing, limited forearm movements and ulnar deviation of the wrist. Figures 11 and 12  After osteotomy and plating of the radius the ulna was lengthened and straightened by double osteotomy, with a graft from the radius at the distal osteotomy. The shaft of the ulna was stabilised with a Rush rod, having been overlengthened by 0.5 cm. Figure 13  Three years later the proximal ulnar osteotomy remains ununited but there is no pain and both appearance and function have been improved.
The disadvantage of ulnar lengthening is that in young patients the deformity will recur and further lengthening may be needed. Some patients, particularly those with short arms or asymmetrical involvement, may accept the possible need for additional operations. However, initial overcorrection by 0.5 cm is well tolerated and is recommended for skeletally immature patients.

Two other reports available on ulnar lengthening in patients with hereditary multiple exostoses have confirmed its value. The rate of recurrence of deformity of 0.25 cm per year at age 12 years in this series is comparable to the 0.27 cm per year at age 11 years reported by Dal Monte et al. (1980). The average gain in pronation-supination was only 25° in our patients compared with 45° improvement in their series. Unlike Irani et al. (1982), we found that bone grafting was not required after distraction by an external fixator.

Conclusions. Lengthening of the ulna, particularly with the use of an external fixator, has given predictable results and is a useful method of treating forearm deformity in patients with hereditary multiple exostoses.

This technique reduces deformity by restoring the normal relationship of the radius and ulna as closely as possible. Wrist stability, joint function, radial head stability and appearance are all improved.

The author wishes to express his thanks to Dr John Ricker, Dr Herbert J. Louis and Dr Fritz P. Snyder.

REFERENCES