SURGICAL TREATMENT OF WRIST DEFORMITY IN HEREDITARY MULTIPLE EXOSTOSIS

T. U. JIYA1, J. E. H. Prujs3, J. W. Van der Eijken2

Wrist deformity represents a unique problematic entity in patients with Hereditary Multiple Exostosis (HME). We report our experience in the treatment of wrist deformities due to HME using three surgical procedures and the outcome of 12 wrist surgical corrections by comparing preoperative, postoperative and last follow-up radiographic values of the carpal slip, radial articular angle and ulnar shortening.

Eight out of 12 forearms did show a postoperative improvement of the radiographic parameters, although recurrence of the deformity occurred frequently. Two forearms showed no change in postoperative radiologic parameters. The average age at operation was 13 years, 4 months and the average duration of follow-up was 76.1 months.

Prevention and reducing the progression of deformity and functional disability is an important goal in the management of these patients. The type of deformity is the most important factor in deciding the type of surgery to be performed.

Keywords: exostosis; wrist; treatment.
Mots-clés: exostoses; poignet; traitement.

INTRODUCTION

Hereditary multiple exostosis (HME) is a benign autosomal dominant disorder of enchondral bone growth, characterised by multiple cartilage covered bony excrescences, accompanied by defective metaphysseal modelling and asymmetrical retardation of longitudinal bone growth.

Wrist deformity represents a unique problematic entity in patients with HME. Involvement of the wrist is seen in 30 to 60 percent of patients suffering from HME (2, 3, 9, 11, 14).

Several reports have appeared in the literature on the pattern of wrist deformity and the success of various surgical procedures in its management.

In this paper, we retrospectively review the postoperative outcome and follow-up of 12 surgical corrections of the forearm performed, due to wrist deformities associated with hereditary multiple exostosis.

MATERIAL AND METHODS

A total of 32 patients with HME visited the University Childrens Hospital in Utrecht between 1978 and 1993. One patient who visited the Emma Childrens Hospital Amsterdam in the same period was included in our series. Twenty two out of the 33 patients had wrist deformities at presentation, 10 (a total of 12 forearms) out of the 22 patients with wrist deformities had a surgical correction.

All available pre- and postoperative anteroposterior radiographs of the forearms were reviewed. The types of deformity were classified according to Masada et al. (4) (fig. 1).

The following radiographic parameters were measured (fig. 2) (Burgess et al.) (2):

— Linear axis (LA): This is a line drawn from the ulnar borders of the distal and proximal radial physeal plates. It defines the central axis of the forearm.

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The data and radiographs of 10 patients (12 forearms) operated on were further analysed postoperatively and at the last follow-up in order to evaluate the outcome of surgery and recurrence of deformities.

RESULTS

Sixty six percent (22 out of 33) of the patients with HME also had wrist deformities. Three patients presented with symptoms related to wrist deformity, one with paresthesia of the hand, another with decreased grip force, and a third one with difficulty in swimming due to poor supination/pronation function of the forearm. Nineteen patients presented with complaints due to exostosis at sites other than the wrist. All patients showed an increase in ulnar deviation and a decrease in radial deviation. Flexion and extension were little affected and wrist pain was not a common feature.

Three wrists with type III deformity were treated with excision of the exostosis alone. Recurrence of the mass was observed in one case (case 6), necessitating a second excision. CS and RAA remained unchanged and normal at the last follow-up (table 1).

One wrist with type I deformity was treated with excision of the exostosis alone (case 10). The US improved from -6 mm to -2 mm at 15 months postoperative follow-up. The CS got slightly worse but the RAA remained unchanged.

Recurrence of US was seen in all 4 wrists with type I deformity treated with a combination of ulnar lengthening, radial osteotomy and excision of the exostosis. The rate of recurrence per year ranged from 0.3 mm to 0.9 mm (follow-up range was 46-103 months). Both CS and RAA were significantly improved in all 4 wrists.

Recurrence of ulnar shortening did occur in one of the two wrists (both with type I deformities) treated with a combination of ulnar lengthening and radial osteotomy (case 7). In one patient with type I deformity (case 8), ulnar lengthening was combined with excision of the osteochondroma. The CS was not normalised postoperatively and only slightly improved during follow-up. No significant recurrence of US was seen.

— Radiological parameters. a = ulnar shortening; b = linear axis of the forearm; c = radial articular angle; d = carpal slip.
Table I. — Radiographic parameters of the 12 forearms operated on, measured pre- and postoperatively and at last follow-up

<table>
<thead>
<tr>
<th></th>
<th>case</th>
<th>side</th>
<th>type</th>
<th>preop</th>
<th>postop</th>
<th>l.f.</th>
<th>preop</th>
<th>postop</th>
<th>l.f.</th>
<th>preop</th>
<th>postop</th>
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<th>surgical operation</th>
<th>age at surgery</th>
<th>follow-up (ma)</th>
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<tr>
<td>1</td>
<td>R</td>
<td>I</td>
<td>38</td>
<td>28</td>
<td>36</td>
<td>15</td>
<td>22</td>
<td>-7</td>
<td>+5</td>
<td>0</td>
<td></td>
<td>u, r, c</td>
<td>4y9m</td>
<td>64</td>
<td></td>
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<tr>
<td>2</td>
<td>L</td>
<td>I</td>
<td>54</td>
<td>21</td>
<td>28</td>
<td>30</td>
<td>18</td>
<td>20</td>
<td>-2</td>
<td>+5</td>
<td>+4</td>
<td>u, r</td>
<td>4y1m</td>
<td>56</td>
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<tr>
<td>3</td>
<td>L</td>
<td>III</td>
<td>50</td>
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<td>15</td>
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<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td></td>
<td>c</td>
<td>8y9m</td>
<td>52</td>
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<td>4</td>
<td>R</td>
<td>I</td>
<td>55</td>
<td>15</td>
<td>33</td>
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<td>+1</td>
<td>-4</td>
<td>u, r, e</td>
<td>10y1m</td>
<td>86</td>
<td></td>
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<tr>
<td>5</td>
<td>L</td>
<td>III</td>
<td>25</td>
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<td>20</td>
<td>30</td>
<td>32</td>
<td>32</td>
<td>-11</td>
<td>-5</td>
<td>-8</td>
<td>e</td>
<td>10y</td>
<td>85</td>
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<tr>
<td>6</td>
<td>R</td>
<td>I</td>
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<td>noe</td>
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<td>48</td>
<td>30</td>
<td>32</td>
<td>-11</td>
<td>-11</td>
<td></td>
<td>u, r, c</td>
<td>6y7m</td>
<td>169</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>L</td>
<td>I</td>
<td>38</td>
<td>33</td>
<td>21</td>
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<td>-13</td>
<td>-11</td>
<td>-11</td>
<td>e</td>
<td>7y2m</td>
<td>46</td>
<td></td>
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<tr>
<td>8</td>
<td>R</td>
<td>III</td>
<td>44</td>
<td>34</td>
<td>9</td>
<td>9</td>
<td>21</td>
<td>5</td>
<td>-6</td>
<td>-2</td>
<td>-4</td>
<td>e</td>
<td>13y</td>
<td>191</td>
<td></td>
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<tr>
<td>9</td>
<td>R</td>
<td>I</td>
<td>100</td>
<td>81</td>
<td>88</td>
<td>42</td>
<td>30</td>
<td>24</td>
<td>-9</td>
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<td>-5</td>
<td>u, c</td>
<td>8y8m</td>
<td>17</td>
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<tr>
<td>10</td>
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<td>I</td>
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<td>42</td>
<td>70</td>
<td>42</td>
<td>23</td>
<td>39</td>
<td>-4</td>
<td>+3</td>
<td>0</td>
<td>u, r, c</td>
<td>8y8m</td>
<td>103</td>
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u = ulnar lengthening; r = radial osteotomy; e = excision; noe = no ossification centre; l.f. = last follow-up.

Fig. 3. — Radiographs of case 3 (the right side showing type I deformity and the left side showing type III deformity); a = preoperative; b = postoperative; c = at last follow-up; d = an example of type III deformity.

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Immediate ulnar lenthening was performed in all procedures (8) by means of a step-cut osteotomy, procuring lenthening with a retractor or a Wagner fixator, and achieving fixation with cortical screws.

Non-union occurred in one patient (both forearms) following a postoperative fracture of the immature callus at the site of the ulnar step-cut osteotomy (case 1). In both forearms the pseudoarthrosis was resected and the breaching gap was grafted with spongy bone from a radial donor site. There were no further complications recorded in this series.

**DISCUSSION**

The treatment of wrist deformities in HME is still a difficult problem. There is no general agreement on the indications for surgery and the prognosis after operation is uncertain. The timing of surgery remains of vital importance and should be preceded by a considerable period of observation (14). The younger the patient the greater the potential for remodelling, which should enhance surgical results (3) although with a greater risk of recurrence. Delay of surgery should however be contemplated when the degree of progression of deformity and functional impairment are considered to be minimal (14).

Various indications for surgical treatment have appeared in the literature. These include an ulnar shortening of more than 1.5 cm with or without a symptomatic subluxation or dislocation of the radial head, radial articular angle of more than 30 degrees, a carpal slip of more than 60%, and a vascular or neural compression in the region of the carpal tunnel. Less subtle indications are pain, cosmetic disturbance and prominence of the mass (3, 14).

The effectiveness of excision of osteochondromas in controlling progression of wrist deformity has long been disputed. Fogel et al. (3) reported that the procedure was not effective in controlling the progression of wrist deformity even when there was no recurrence. Shapiro et al. (9) however reported an effective prevention of progression of deformity after simple excision of the osteochondroma.

All patients treated with excision of the exostosis alone in our series, show no significant progression of deformity at last follow-up (follow-up range was 15-191 months). This observation in our type III deformities is in concordance with the report made by Shapiro et al. (9). Excision of osteochondroma is the treatment of choice in type III wrist deformities (4). Ulnar lenthening alone or in combination with some other surgical procedure have been widely applied in the treatment of wrist deformity due to HME. Pritchett reported correction of wrist deformity in 10 forearms of 8 patients by ulnar lenthening, either as an immediate procedure or by gradual distraction using an external fixator (7). He obtained good results, with normal relationship of the radius and ulna being restored as closely as possible, with improved wrist stability and joint function. In our series only one patient was treated with ulnar lenthening alone.

Although the number is too small to draw any conclusions, we observed that soft tissue tightness and significant bowing of the radius makes an initial overcorrection troublesome. The risk of neurovascular problems which may arise from ulnar lenthening could be significantly reduced by dividing the cord-like portion of the interosseous membrane (4, 14). No patient treated with ulnar lenthening in our series developed neurovascular problems even when the interosseous membrane was not incised.

Ulnar straightening, a more recent advancement, may be performed prior to ulnar lenthening. This can be achieved with multiple osteotomies and the use of a Rush pin to obtain fixation (6). Ulnar lenthening alone increases the support of the ulnar side of the wrist joint and corrects the ulnar translocation of the carpus. It is however believed not to affect the RAA (3). Therefore when there is an associated abnormality of the radiocarpal angle, causing subluxation of the lunate, a combination of procedures is indicated (3, 4). Corrective osteotomy of the distal radius when combined with ulnar lenthening gives satisfactory results. Radial osteotomy normalises the RAA, and carpal slip, and corrects bowing of the radius. Masada et al. reported excellent results in their series by using a combination of excision of
osteochondromas, ulnar lengthening, and corrective osteotomy of the radius (4). In all 4 patients who received the same combination of treatment in our series, the deformity was significantly corrected. When ulnar lengthening is combined with hemiepiphyseal stapling of the distal radius, the functional and cosmetic results obtained are better than with ulnar lengthening alone (3). Retardation of the radial side of the distal radial epiphysis allows growth of the ulnar side to become proportionately increased, so that the articular surface will become more perpendicular to the long axis of the forearm (3). The superiority of corrective osteotomy of the radius over hemiepiphyseal stapling has not been clearly established. Stapling causes shortening of the forearm which is a major disadvantage, also the final length discrepancy is difficult to predict (4). For these reasons corrective osteotomy may be preferable in combination with ulnar lengthening.

Rodgers et al. recently published the use of radioulnar fusion (the so-called one bone forearm) as a salvage procedure in recalcitrant cases, when the more conservative options fail, leaving patients with a painful and weak extremity. Although such instances are quite rare, when they do present, radioulnar fusion can relieve pain (8).

Although hereditary multiple exostosis does not shorten life expectancy except in patients with malignant degeneration, it does cause considerable morbidity (11, 12). Despite the various procedures employed in treating wrist deformity in HME, there is frequently need for repeated operations particularly in the skeletally immature patient, in whom the primary disease process remains active.

Preventing and reducing the progression of deformity and functional disability should remain the most important goal in the management of these patients. Also the type of deformity should be the most important factor in deciding the type of surgery to be performed.

REFERENCES


SAMENVATTING


Pols afwijkingen vormen een bijzondere problematiek in de behandeling van patienten met Multiple Hereditaire Exostose (M.H.E.). In dit artikel beschrijven wij onze ervaring in de chirurgische behandeling van pols afwijkingen bij 10 kinderen (12 polsen) met M.H.E. 3 chirurgische ingrepen werden toegepast alleen of in combinatie. Rontgenologische waarden van de „carpal slip” „radial articular angle” en ulnaire verkorting werden preoperatief, post-operatief en bij de laatste poliklinisch
bezoek vergeleken. 8 van de 12 polsen vertonen post-operatief een verbetering van de röntgen parameters, echter een recidief werd vaak gezien tijdens follow-up. 2 polsen met normale preoperatieve röntgen parameters lieten geen verandering zien op de postoperatieve röntgen foto's. De gemiddelde leeftijd van de patiënten ten tijde van de operatie was 13 jaar en 4 maanden en de gemiddelde periode van follow-up was 76,1 maanden.

Voorkomen van progressie van afwijkingen en het voorkomen van functionele beperkingen zijn belangrijke doelen bij de behandeling van pols afwijkingen bij patiënten met M.H.E. Het type afwijking is de belangrijkste factor voor het bepalen van de chirurgische ingreep.

RÉSUMÉ


Douze poignets sont examinés en pré- et post-opératoire et au dernier contrôle. Les paramètres radiographiques pour le glissement du carpe, l’angle radial articulaire et le raccourcissement du cubitus sont comparés. Les paramètres radiographiques étaient amélioré dans 8 des 12 avant-bras opérés bien que la déformation ait souvent récidivé. Dans 2 cas les paramètres radiographiques post-opératoires n’avaient pas changé. L’âge moyen à l’opération était de 13 ans et 4 mois et le recul moyen de 76 mois.

Le but principal du traitement de ces patients doit être la prévention et la correction de la déformation et de la limitation fonctionnelle du poignet.

Le choix de la technique opératoire dépend surtout du type de la déformation.