SURGICAL TREATMENT OF WRIST DEFORMITY IN HEREDITARY MULTIPLE EXOSTOSIS

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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 shorthening.

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 metaphyseal 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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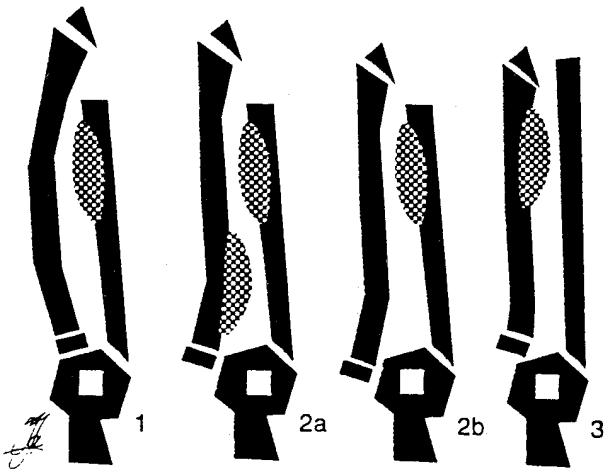


Fig. 1. — The types of wrist deformities (4). Type 1 : shortening of the ulna relative to the radius ; Type 2a : dislocation of the radial head caused by exostosis at the proximal end of the radius ; Type 2b : dislocation of the radial head in the absence of an exostosis at the proximal end of the radius ; Type 3 : shortening of the radius relative to the ulna.

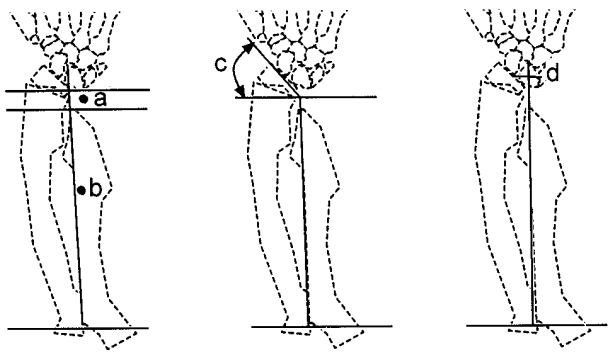


Fig. 2. — Radiographic parameters. a = ulnar shortening ; b = linear axis of the forearm ; c = radial articular angle ; d = carpal slip.

— *Radial articular angle* (RAA) : This is the angle between a line along the articular surface of the distal radius, and the linear axis of the forearm.
— *Carpal slip* (CS) : This is defined as the percentage of the lunate on the ulnar side of the linear axis. Normally the linear axis bisects the lunate.
— *Ulnar shortening* (US) : This is the distance between two perpendicular lines, one drawn from the LA of the forearm to the distal end of the ulna, and the other drawn from the LA of the forearm to the ulnar border of the distal radial epiphysis.

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 I).

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

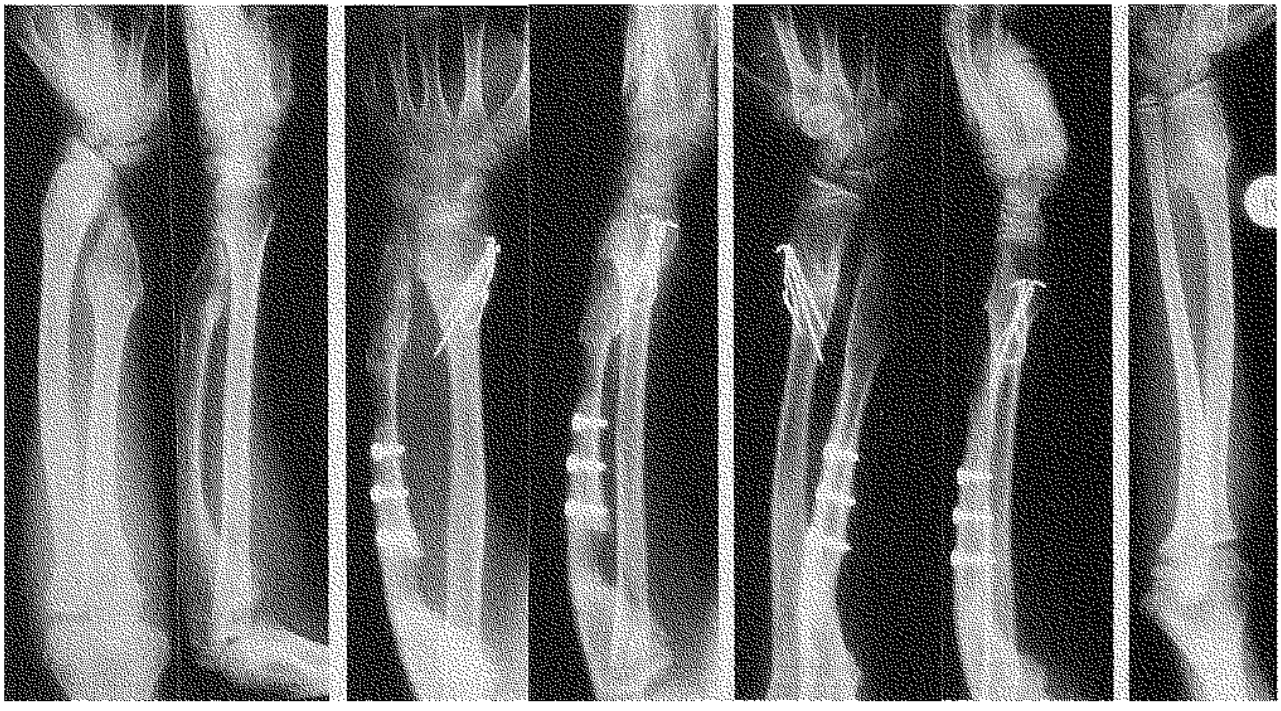
Recurrence of US was seen in all 4 wrists with type I deformity treated with a combination of ulnar lenghtening, radial osteotomy and excision of the exostosis. The rate of recurrence per year ranged from 0.3 mm to O.9 mm (follow-up range was 46-103 months). Both CS and RAA were significantly improved 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 lenthening 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.

Table I. — Radiographic parameters of the 12 forearms operated on, measured pre- and postoperatively and at last follow-up

			carpal slip (%)			radial articular angle (degrees)			ulnar shortening (mm)					
												surgical operation	*age at surgery*	*follow-up (mo)*
case	side	type	preop	postop	l. f.	preop	postop	l. f.	preop	postop	l. f.			
1	R	I	38	28	36	29	15	22	-7	+5	0	u, r, e	14y9m	64
	L	I	54	21	28	30	18	20	-2	+5	+4	u, r	14y1m	56
2	L	III	50	50		15	15		0	0		e	5y9m	52
3	R	I	55	15	33	34	13	20	-15	+1	-4	u, r, e	10y1m	86
	L	III	25			20			+2			e	10y	85
4	L	I	noc	noc	90	48	30	32	-11	-5	-8	u, r, e	6y7m	109
5	L	I	38	33	21	38	31	36	-13	-11	-11	u	7y2m	46
6	R	III	44			34			+13			e	13y	191
7	R	I	70	27	54	30	9	21	-6	-2	-4	u, r	11y6m	90
8	R	I	100	81	88	42	30	24	-9	-4	-5	u, e	8y8m	17
9	R	I	100	42	70	42	23	39	-4	+3	0	u, r, e	8y8m	103
10	L	I	30	30	50	30	30	30	-6	-6	-2	e	10y2m	15

u = ulnar lengthening ; r = radial osteotomy ; e = excision ; noc = no ossification centre ; l. f. = last follow-up.



a b c d

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.

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 breeching 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 lenghtening 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 lenghtening, 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 lenghtening 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 lenghtening could be significantly reduced by dividing the cord-like portion of the interosseous membrane (4, 14). No patient treated with ulnar lengthening 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 lengthening. This can be achieved with multiple osteotomies and the use of a Rush pin to obtain fixation (6).

Ulnar lenghtening 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 lenghtening 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 lenghtening, 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 lenghtening is combined with hemiepiphyseal stapling of the distal radius, the functional and cosmetic results obtained are better than with ulnar lenghtening 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 lenghtening.

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.

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SAMENVATTING

T. U. JIYA, J. E. H. PRUIJS, J. W. VAN DER EIJKEN. Chirurgische behandeling van polsafwijkingen met Multiple Hereditaire Exostose.

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 postoperatief een verbetering van de rontgen parameters, echter een recidief werd vaak gezien tijdens follow-up. 2 polsen met normale preoperatieve rontgen parameters lieten geen verandering zien op de postoperatieve rontgen foto's. De gemiddelde leeftijd van de patienten ten tijden 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 patienten met M.H.E. Het type afwijking is de belangrijkste factor voor het bepalen van de chirurgische ingreep.

RÉSUMÉ

T. U. JIYA, J. E. H. PRUIJS, J. W. VAN DER EIJKEN. Traitement chirurgical des déformations du poignet dans le syndrome des exostoses multiples héréditaires.

La déformation du poignet représente un problème spécifique chez les patients atteints du syndrome des Exostoses Multiples Héréditaires (E.M.H.). Notre expérience du traitement des déformations du poignet secondaire aux E.M.H., utilisant 3 techniques opératoires, sera discutée.
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 avants-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 fonctionelle du poignet.
Le choix de la technique opératoire dépend surtout du type de la déformation.