MADELUNG DEFORMITY: SURGICAL PROPHYLAXIS (PHYSIOLYSIS) DURING THE LATE GROWTH PERIOD BY RESECTION OF THE DYSCHONDROSTEOSIS LESION

D. VICKERS and G. NIELSEN

From the Royal Children's Hospital, Brisbane, Australia

The majority of cases of Madelung deformity are caused by hereditary dyschondrosteosis at the wrist. The principal lesion in the ulnar zone of the distal radial physis retards growth asymmetrically, especially in late childhood. Resection of this zone and its replacement with autologous fat (Langenskiöld procedure, or physiolysis) restores growth and minimizes deformity. The resection of an abnormal ligament tethering the lunate proximally may assist carpal advancement. A series of 17 patients (24 wrists) treated over a 12-year period is presented, with sufficient follow-up for evaluation of 11 patients (15 wrists). The results of this prophylactic procedure are encouraging, and, if it is performed early, the authors believe that Madelung deformity may be preventable, or at least controllable.

Journal of Hand Surgery (British Volume, 1992) 17B: 401–407

The description by Madelung (1878) of the deformity which now bears his name predated X-rays. The lesion producing primary Madelung deformity was subsequently termed "dyschondrosteosis" by Leri and Weill (1929). A dominant gene with incomplete penetrance usually transmits dyschondrosteosis, which in its most severe form includes mesomelic dwarfism (Langer, 1965). Sporadic (idiopathic) cases of dyschondrosteosis are less common. Reverse Madelung deformity is apparently inherited in the same way, as we have seen various mixtures of classic Madelung deformity and reverse Madelung deformity in the same family on two occasions, and one patient demonstrated both deformities.

19 years' experience with Langenskiöld's procedure for excision of bone bridges of traumatic origin has shown that this procedure restores growth in the inherited disorder of delta phalanx (Vickers, 1987). Preliminary reports of a similar procedure for Madelung deformity have been published (Vickers, 1980, 1989; Tachdjian, 1990a).

Natural history

Madelung deformity is not apparent in infancy or early childhood, but from X-rays it is clear that dyschondrosteosis has a long-standing and progressive effect upon the growth of the radius from an early stage of development. The radius is short and bowed with a radial and dorsal convexity. A growth spurt in late childhood, perhaps combined with a premature fusion across the physis, further aggravates the situation, giving rise to the Madelung deformity.

Pathology

From our operative experience the principal pathodynamics appear to depend upon two lesions, one bony, one ligamentous. The bony lesion (dyschondrosteosis) causes the abnormal inclination of the distal radius which is responsible for the visible deformity, as the ulna grows relatively straight. An abnormal volar ligament is implicated in the distortion of the carpus.

The dyschondrosteosis lesion in the distal radius occupies the ulnar third of the physis. The lesion not only fails to grow, but also acts as a tether. The usual volar disposition of the lesion causes classical Madelung deformity, whereas a dorsal disposition will cause reverse Madelung deformity. A mid-ulnar zone lesion causes carpal triangularization without visible deformity (chevron carpus). Premature bony fusion across the physis was only observed in a few patients who presented late. The epiphysis narrows progressively in the ulnar half, and trails "around the corner" to lie in a longitudinal plane. Growth in this zone can only add width to the bone, but not length.

An abnormal thick volar ligament up to 5 mm in diameter, tethering the lunate to the radius proximal to the physis, has been evident on a number of occasions, (Fig. 1). Since it occupies an identical position in both Madelung and reverse Madelung deformity it is unlikely to be a cause of the radial deformity. It appears to firmly tether the lunate in a proximal position between or below the radius and ulna. This ligament may also cause thinning of the radial epiphysis by compression. The abnormal pronator quadratus muscle described by Linscheid (1979) has not been encountered.

Ten operative specimens were submitted for histological examination. The chondrocytes appeared normal, but the usual orderly columns of the physis were not seen. Endochondral ossification appears "confused" in orientation. In one case the appearance was similar to achondroplasia. Two different specimens of the abnormal ligament were examined, and the tissue was confirmed to be mature ligament.

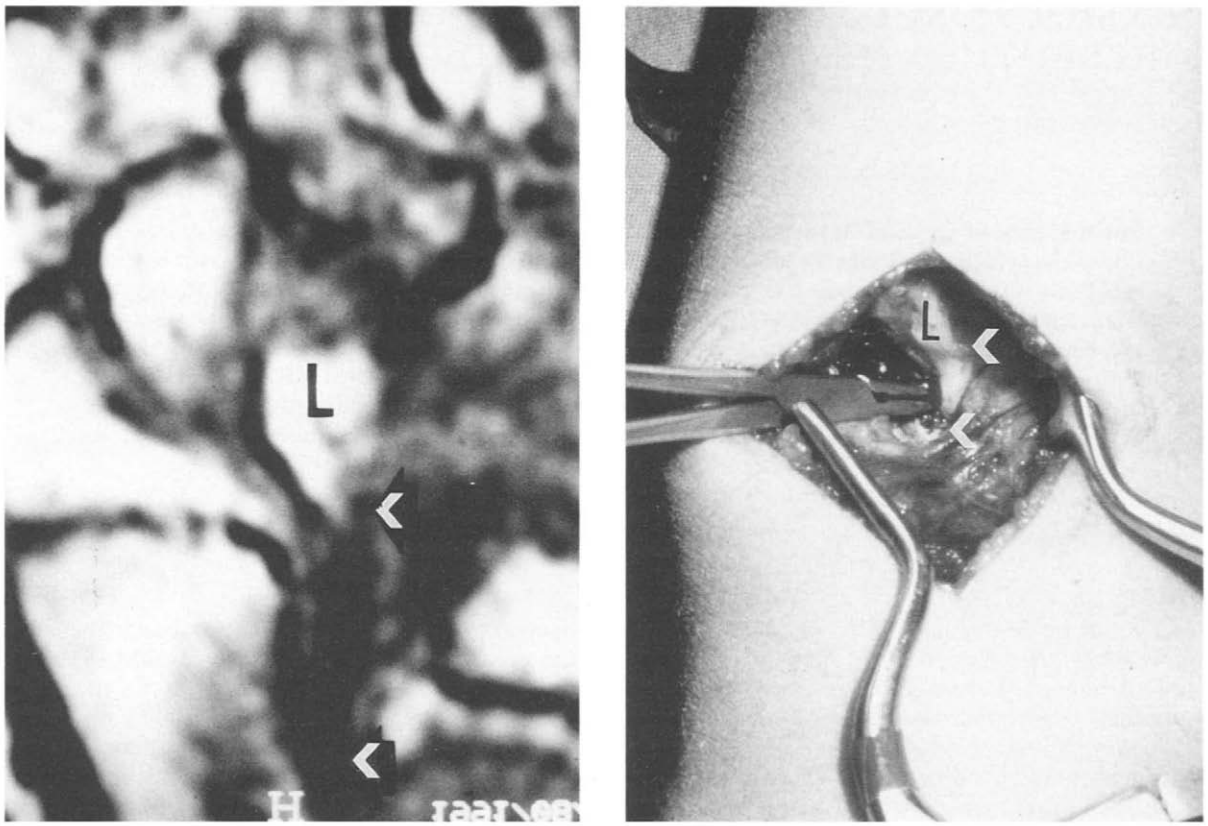

Fig. 1 MRI and operative images of the abnormal volar radio-lunate ligament in a reverse Madelung deformity.

Patients and methods

The Madelung deformities in this series were all of primary origin (dyschondrosteosis) and the skeleton was immature at the time of surgery. In no case was a repeat of the surgical physiolysis necessary. There were 17 patients, seven of whom had surgery on both wrists, giving a total of 24 operations. One reverse Madelung deformity is included. There were 15 female and two males. A family history of Madelung deformity or features of inherited mesomelia were present in 13 patients. The average age at presentation was just over 12 years, with a range from 11 years to 14 years 6 months. At the time of writing, nine patients had reached maturity. Pain and deformity were the presenting features in all except one patient who had little deformity but a painful chevron carpus. 14 patients requested surgery mainly for pain—only three believing that appearance was more important.

11 patients were assessed by the authors, two have been lost to follow-up, and four are too recent for significant post-operative evaluation. Accordingly an analysis was made of 11 patients (15 wrists), nine of whom are mature, and two are within six months of maturity. The follow-up period ranges from a maximum of 12 years to a minimum of 15 months.

Clinical evaluation of the deformity was difficult. Anteroposterior X-rays of the wrist were used for measurement, as there was too much radiographic variation in the lateral views for these to be of value. Because the radius is bowed, the longitudinal and transverse axes cannot be determined. The ulna is usually straight providing a more reliable longitudinal axis. A line at right angles to this axis passing through the most radial extent of the radial physis became the "transverse line" (Fig. 2). The physeal line of the radius was first identified in the post-operative X-ray. This line joins the most radial and most ulnar extents of the physis. The angle subtended by the transverse line and the physeal line is named the "physeal angle". It is now possible to mark the physeal line on the pre-operative X-ray by joining the radial extent of the physis to the ulnar physis at a similar point to that already determined on the post-operative X-ray. The surgical spike is a post-operative feature in the cortex.

Metaphyseal growth and lunate advancement can be

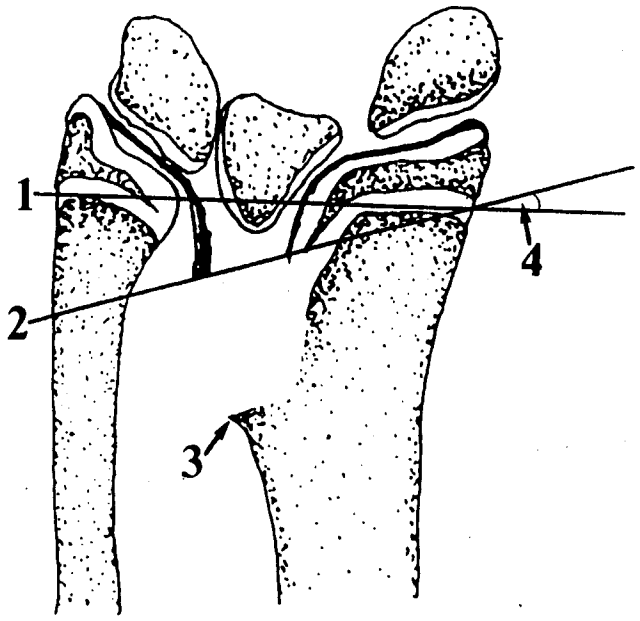

Fig. 2 Arbitrary (1) transverse and (2) physeal lines used as reference for growth and correction of the distal radius and carpus, (3) indicates the surgical defect in the cortex and (4) the physeal angle.

measured using the transverse line and the surgical spike. The change in the physeal angle indicates reorientation of the radial epiphysis.

Operative technique

Under tourniquet control, a volar transverse incision is made 1.5 cm proximal to the most proximal wrist crease. The approach passes ulnar to flexor carpi radialis and palmaris longus, but occasionally it is more convenient to pass to the radial side of these tendons. The median nerve and radial artery are protected. The approach continues radial to the mass of digital flexor tendons to locate the distal edge of the pronator quadratis muscle, some of which is mobilized at the radial end. An osteotome is used to make the initial longitudinal osteotomy in the radius, parallel to the long axis of the forearm, about 5 mm from the radio-ulnar joint. In cases of extreme volar subluxation of the carpus care is needed not to mistake the lunate for the underlying radius. The small fragment of the distal radius is reflected ulnarwards with the osteotome to preserve what exists of the flimsy connections between it and the ulna, and to leave some support for the lunate. A sagittal section of the distal radius is now visible. Magnification is recommended and knowledge of the Langenskiöld procedure is essential. If the initial osteotomy is too shallow, a featureless white sheet of fibrous tissue and cartilage is seen. Parallel osteotomy cuts 1 mm thick are repeated until the physis is clearly identified. When first seen it is thin and wavy and the epiphysis is significantly narrowed. When the physeal cartilage is clearly defined, some further bone is carefully removed from the metaphyseal side with a gouge or burr so that the profile of the cartilage is proud of the bone, and is intact and tidy from the dorsal periosteum to the volar periosteum. This prevents a new bar of bone forming from the bony margins of the physis. An abnormal volar ligament tethering the lunate to the radius should be sought and excised. The tourniquet is deflated and haemostasis is secured, using bone wax if necessary. The tourniquet is reinflated and the cavity is then washed out with normal saline to remove all bone chips and blood. A generous quantity of fat is obtained from the proximal forearm medially and inserted to more than completely fill the surgical cavity. This fat must make intimate contact with the entire length of the physeal cartilage, isolating the bony epiphysis from the bony metaphysis. Soft tissues fall together to hold the fat in place, and the skin is sutured. A short arm volar slab, or a crepe bandage, is applied for two weeks, depending upon the degree of deformity.

In reverse Madelung deformity a dorsal approach is better. The incision extends proximally from Lister's tubercle for 6 cm. A limited incision is made in the extensor retinaculum, and the plane between extensor pollicis longus and the wrist extensors provides a direct approach. During closure the extensor retinaculum has been used to stabilize extensor pollicis longus tendon and hold the fat graft in place.

Results

Pain, deformity and range of motion were assessed clinically in 15 wrists. Changes in the deformity were measured on X-ray in 14 of the 15 wrists (Table 1). There were no long-term complications of the physiolysis procedure itself, but in one case a cut was inadvertently made in the lunate with an osteotome, and this healed spontaneously.

Pain

All patients experienced a significant reduction in pain within the first six months after surgery. Previously restricted activities such as tennis were resumed with little or no pain. Only one patient returned complaining of pain which was described as an ache in the distal ulna. The pain occurred without provocation and lasted less than one week. The patient whose wrist is shown in Figure 3 complained of pain playing cricket and writing. Now pain is only "occasional". Although all patients had reduced pain, only four claimed to be totally pain-free and able to do all physical activities. One works as a carpenter.

Table 1—Radiological assessment of physiolysis

Case	*Metaphyseal growth (mm)*	*Lunate advancement (mm)*	*Initial angle of physis*	*Improvement in angle of physis*
1	+5	−2	27°	+9°
2	+5	−1	20°	Static
3	+7	+3	28°	+13°
4	+2	+4	38°	+28°
5	+11	−5	12°	Static
6	+7	−8	30°	+12°
7	Static	−4	22°	+10°
8	Static	Static	20°	Static
9	+3	+9	18°	+11°
10	+2	+7	48°	+19°
11	+2	+2	21°	+10°
12	+3	Static	21°	+7°
13	+3	−1	12°	+7°
14	Static	+3	0°	Static
Average	+3.5	N/A	23°	+10°

Case 15 does not appear here as early X-rays are not available for measurement.

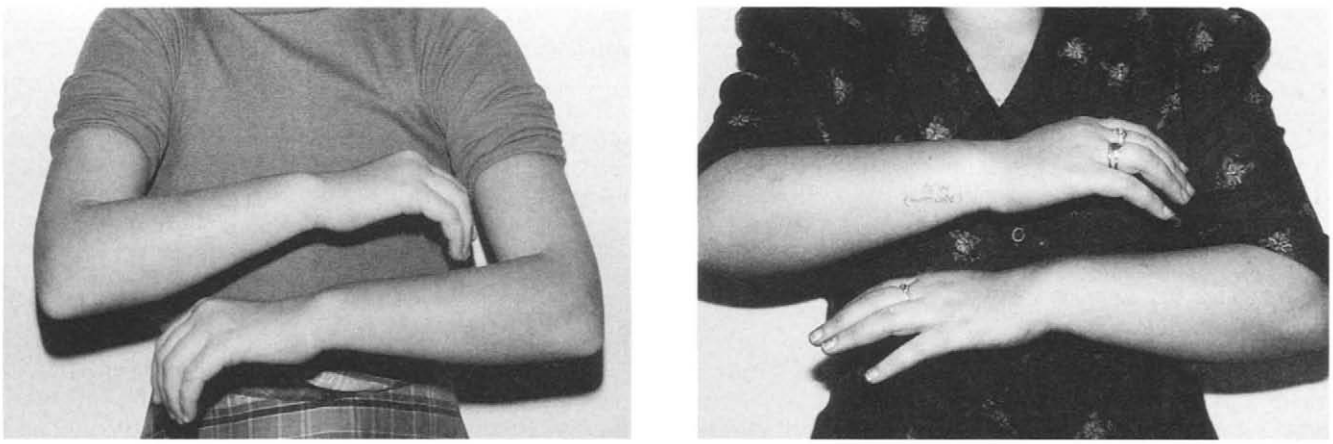

Fig. 3 Clinical progress to maturity of both wrists over an eight-year period after physiolysis.

Deformity

No patient displayed progression of the deformity after surgery. Slight improvement was observed in most cases. Although serial photographs were taken of all patients, radiological methods are more objective. Growth of the radius and improvement of the inclination of the radial physis and epiphysis is apparent in Figures 4 and 5, and in Table 1. Positive metaphyseal growth was measured in 11 wrists, static growth in three wrists, and there were no negative findings. The maximum metaphyseal growth was 11 mm, with a minimum of zero. The average growth in 14 wrists was 3.5 mm. Improvement of the angle of the physis and epiphysis was measured in ten wrists (eight of which also had positive metaphyseal growth); four were static and there were no negative findings. The maximum improvement in the physeal angle was 28° with a minimum of 0°, producing an average of 10° of improvement. The position of the proximal pole of the lunate relative to the transverse line showed positive improvement in six cases, was static in two cases and negative advancement was measured in six cases.

A most important accidental observation was made retrospectively in one case from a remote area. The unoperated wrist, which had been the better one, deteriorated seriously over a two-year period. Conversely, growth and correction were noted on the operated side which regained 60° of supination and became pain-free (Fig. 4).

The patient in Figure 5 presented with a mild Madelung deformity but considerable pain (case 12, Table 1). She was in the habit of strapping her wrist at school and

Fig. 4 The right wrist (a, b), already significantly deformed at presentation, improved over two years after surgery. Meanwhile the lesser affected and untreated left wrist (c, d) deteriorated.

Fig. 5 Progress radiographs from (a) pre-operative, (b) five months and (c) 18 months after operation illustrating radial growth and improvement in the shape of the radio-carpal joint.

during sporting activities. Fortunately she presented early enough for a physiolysis procedure to effect good growth, with correction in the distal radius. At her final review she had no pain, did not strap her wrist any more and displayed a full range of motion. Her mother describes the deformity as "a lot less". Three patients required a radial osteotomy because their deformity at initial presentation was too advanced for correction to occur in the residual growth period.

Range of motion

Supination was the movement most restricted pre-operatively and caused most of the activity-related problems. The average loss of supination was 31°, with a range from 0 to 90°. Post-operatively supination rapidly improved by an average of 23°, and was usually maintained. All other ranges of motion improved and no patient lost motion in any direction. Late in the growing period some patients lost some of the movement they had regained, but maintained a net gain overall.

Discussion

Surgery for established Madelung deformity is difficult and only rarely indicated, since pain is usually transient and deformity accepted (Dobyns, 1988; Lamb, 1988; Fagg, 1988; Tachdjian, 1990b). The rationale for prophylactic surgery is completely different and, we believe, may have an important place in the early management of Madelung deformity. The most difficult question which can only be answered by inference is "what would have been the outcome without surgery?". Clues to this dilemma came from two cases where the more deformed wrist treated by physiolysis demonstrated recovery while the non-operated wrist proceeded to increased deformity. Also, joint surfaces have been seen to improve, and this has not been observed following salvage surgery.

Initially pain was the only indication for surgery on the immature skeleton. Considerable experience with Langenskiöld's procedure led us to believe that the procedure should be safe. Post-operatively all the patients had less pain. Denervation could be a factor, but this is not an objective of the surgery. Deformity mostly improved, some remained static, and no patient with initial mild deformity required radial osteotomy or ulnar shortening after physiolysis. In contrast, Nielsen (1977) assessed 26 wrists affected by Madelung deformity at maturity, and 13 required salvage surgery in his opinion.

An improved range of motion was universally observed, with no patient losing range in any direction. Once again this is in contrast to Nielsen's report of 13 patients after salvage surgery, only one of whom displayed improved mobility, with two having decreased mobility. Early surgery is more effective, and since many cases are familial, early detection is possible. Surgery is not routinely recommended, but the family history, pain, bone age, and progress of deformity after a reasonable period of observation influence the decision. The later cases in the series had a more conservative resection with a better result. The physiolysis procedure to restore growth in the distal radius has been shown to improve the outcome of Madelung deformity.

MRI screening of young children in a family with the causative gene should allow early detection of the abnormal ligament, before the appearance of the osseous lesion or deformity. A timely excision of this ligament alone may have significant prophylactic potential.

References

DOBYNS, J. H. In: Green, D. P. (Ed). *Operative Hand Surgery*, 2nd Edn. Edinburgh. Churchill Livingstone, 1988: 512.

FAGG, P. S. (1988). Wrist pain in the Madelung's deformity of dyschondrosteosis. The Journal of Hand Surgery, 13B: 1: 11–15.

LAMB, D. (1988). Editorial: Madelung deformity. The Journal of Hand Surgery, 13B: 1: 3–4.

LANGER, L. O. (1965). Dyschondrosteosis, a hereditable bone dysplasia with characteristic roentgenographic features. American Journal of Roentgenology, 95: 178–188.

LERI, A. and WEILL, J. (1929). Une affection congénitale et symétrique dévelopment osseux la dyschondrostéose. Bulletin et Société Medicale Hôpitaux De Paris, 53: 1491–1494.

LINSCHEID, R. L. (1979). Madelung's deformity. Correspondence Newsletter No. 24, Correspondence Club. American Society of Surgery of the Hand.

MADELUNG, O. (1878). Die Spontane Subluxation der Hand nach norne. Archiv fur Klinische Chirurgie, 23: 395–412.

NIELSEN, J. B. (1977). Madelung's deformity. A follow-up study of 26 cases and a review of the literature. Acta Orthopaedica Scandinavica, 48: 379–384.

TACHDJIAN, M. D. (1990a). *Pediatric Orthopedics* (2nd Edn.) Volume 1. Philadelphia. W. B. Saunders, 214–215.

TACHDJIAN, M. D. (1990b). *Pediatric Orthopedics* (2nd Edn.) Volume 1. Philadelphia. W. B. Saunders, 213.

VICKERS, D. W. (1980). Premature incomplete fusion of the growth plate: causes and treatment by resection (physiolysis) in 15 cases. Australia and New Zealand Journal of Surgery, 50: 393–401.

VICKERS, D. W. (1989). Epiphysiolysis. Current Orthopaedics, 3: 41–47.

VICKERS, D. W. (1987). Clinodactyly of the little finger: a simple operative technique for reversal of the growth abnormality. Journal of Hand Surgery, 12B: 3: 335–342.

Accepted: 12 February 1992

Associate Professor David Vickers, 225 Wickham Terrace, Brisbane 4000, Australia.

This paper won the prize for the best paper presented at the Annual Scientific Congress of the Australian Hand Surgery Society in 1992.