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MADELUNG'S DEFORMITY

A Follow-up Study of 26 Cases and a Review of the Literature

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Madelung's deformity, the history, aetiology, pathogenesis and prognosis, is discussed, on the basis of the literature. The author's material of 26 cases has been followed up. Thirteen of them were treated by resection of the ulna head or shortening of the ulna, with or without wedge osteotomy of the radius. The result was an improvement in pain and a cosmetic improvement, but mobility was unaltered. Spontaneous improvement in the symptoms is common, so that a waiting policy, and observation of the patient until growth ceases, is recommended.

Key words: deformity of the wrist; dysplasia of bone; dyschondrosteosis; epiphysis; growth; Léri-Weill syndrome; Madelung's deformity

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In 1878, O. Madelung described a severely deformed wrist in a young woman. According to Madelung, this *spontaneous forward subluxation of the hand* (Figure 1) had already been described (Dupuytren 1834, Malgaigne 1855, Weber 1859, Nélaton 1847) and Madelung himself had at that time seen 12 cases. Madelung considered that the deformity was due to a disturbance of the growth of the joint, the result of overloading a joint already predisposed to malformation. The prognosis was good and Madelung advocated the term *manus valga* for the condition. Since then, the topic has fascinated numerous investigators, not least Scandinavians (Poulsen 1904, Natvig 1905, Solberg 1906, Björkroth 1932, Paus 1941, 1951, Bergan 1942, Schulstad 1971), and it was Poulsen (1904) who showed that Malgaigne and not Dupuytren should receive the honour of reporting the first

Figure 1. Illustration from Madelung's original study.

case of Madelung's deformity (MD). Since then, there has been uncertainty as to the aetiology and disagreement regarding the pathogenesis. The original picture of the disease has been widened and has become increasingly difficult to define, as evidenced by the 33 synonyms which can be encountered in the literature.

In the years 1929 and 1931, Léri & Weill and Léri et al. published two cases of genetic dwarfism called dyschondrosteosis, where MD was one of the charac-

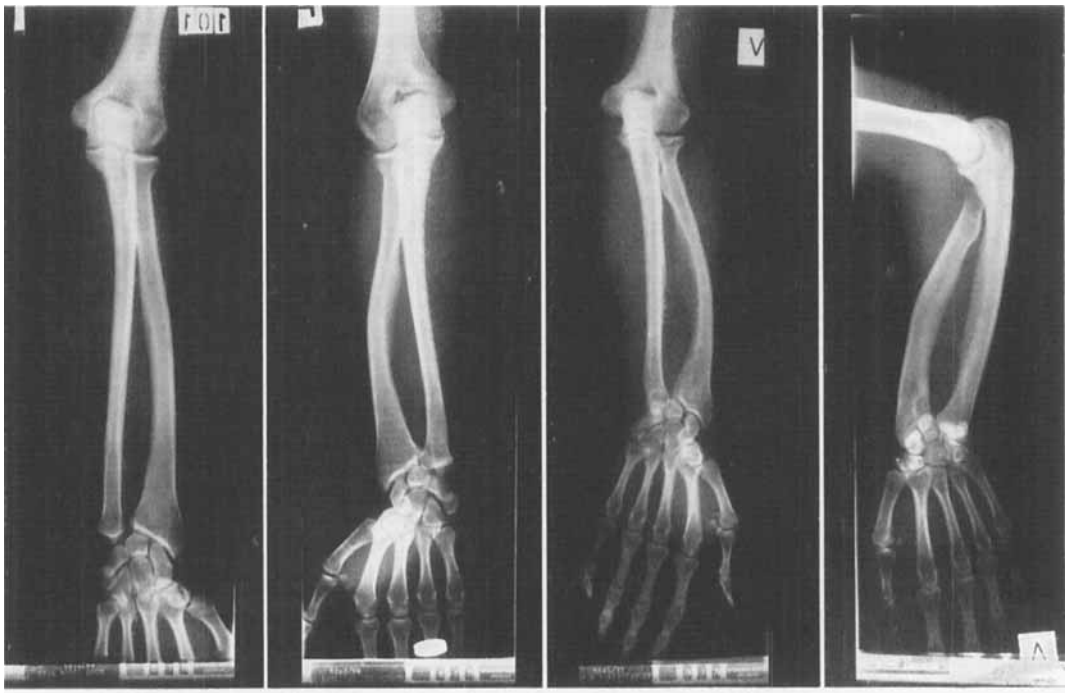

Figure 2. X-rays of the forearm in four different patients with Madelung's deformity showing an increasing degree of severity from left to right. The patient on the right shows a malformation of the head of the radius, which is never observed in patients with genuine Madelung's deformity.

teristic features. As a consequence of this, several authors have regarded these two diseases as being identical (Herdman et al. 1966), whereas others have clearly distinguished between them (Felmann & Kirkpatrick 1969). Symptomatic cases have also been described simulating Madelung's *spontaneous* subluxation: traumatic, neoplastic or infectious (Stetten 1909, Björkroth 1932, Anton et al. 1938, Stypa 1973), hormonal, biochemical or genetic disturbances (Thompson & Kalayjian 1939, Henry & Thorburn 1967) or in generalized dysplasia of bone (Table 1) (Nassif & Harboyan 1970, Nielsen & Galatius-Jensen 1971). According to Kelikian (1974) seven cases of reversed MD have been described.

It is maintained nowadays that MD is a genetic disorder found more frequently in women than in men, twice as frequently bilaterally and then often with the same degree of severity on both sides. The condition is localized to the distal epiphysis of the radius, which shows failure of growth, most often in the ulnar/volar portion. Clinically, it appears during the growth period with functional pain and troublesome prominence of the ulnar head, apparent subluxation of the wrist, and curvation and possible shortening of the forearm. The deformity varies from quite mild, symptomless forms to cases of more severe invalidism (Figure 2).

Table 1. Types of bone dysplasia associated with Madelung's deformity.

Dyschondrosteosis
Achondroplasia
Multiple exostoses
Multiple epiphyseal dysplasia
Dyschondroplasia (Ollier)

Madelung advised conservative treatment, and Poulsen (1904) reports that

Table 2. Sex distribution and age of onset of the 26 patients with Madelung's deformity (MD), grouped according to the classification of Table 5. The table shows the number of patients operated on, the cases with familial disposition, and the stature at follow-up examination.

Classification		No. of pts.	♀	♂	Age of onset	No. of ops.	Familial disp.	Stature at follow-up examination
Idiopathic MD	Genuine	13	13	0	6–15 (10½)	7**	2	157–172 cm (165 cm)
	Dysplasia *	10	9	1	11–16 (13)	5**	6	137–156 cm (150 cm)
Symptomatic MD		3	0	3	–	1	–	170–188 cm (180 cm)
Total		26	22	4	–	13	8	–

* 9 cases of dyschondrosteosis + 1 case of multiple exostoses.
** 1 patient in each group underwent operation on both sides (a total of 15 operations).

surgical intervention with tenotomy of the "contracted" flexors by the method of Busch (1864) was soon abandoned. The first osteotomy of the curved radius for MD was performed by Duplay (1885). Numerous surgical procedures have been recommended, in which either the deformity of the radius was attacked or that of the relatively elongated ulna; in some cases both were treated (Poulsen 1904, Stetten 1909, Darrach 1913, Burrows 1937, Kelikian 1974).

PATIENTS AND METHODS

A total of 26 patients (22 women and 4 men) with MD attended the Department of Surgery of the Hand, Orthopaedic Hospital, Copenhagen, during the years 1954 to 1974. The symptoms were pain and restricted movement of the wrist, which had their onset in the age group 6 to 16 years. Familial disposition was frequently encountered. Patients with bone dysplasia were short in stature (Table 2). MD coincided with other deformities (various exostoses, bone cysts, short fourth metacarpal, sacralisation and double kidney) in six out of the nine patients with dyschondrosteosis. This has also been found by other authors (Henry & Thorburn 1967, Nassif & Harboyan 1970, Solberg 1906, Stetten 1909, Kelikian 1974, Nagura 1971).

In half of the patients the symptoms were so pronounced that operation was indicated. The operated patients were between 14 and 57 years of age (average 22 years). Table 3 shows the operations employed.

Table 3. Fifteen operations for Madelung's deformity in 13 patients.

	No. of wrists
Excision of the head of the ulna	4
Excision of the head of the ulna + wedge osteotomy of the radius	4
Shortening of the ulna	2
Shortening of the ulna + wedge osteotomy of the radius	1
Resection in the distal ulna	1
Resection in the distal ulna + radio/ulnar arthrodesis distally	3
Total	15

In the case of excision of the head of the ulna a simple resection of the distal head was performed, except for the styloid process, whose connection with the radius via the articular disc and via the collateral ligament to the carpal bones ensures stability of the wrist. Shortening of the ulna was performed as a Z-shaped shortening osteotomy of the distal ulna which was immobilized by means of a plate and screws. Resection of part of the distal ulna (removal of a segment of the shaft of the ulna, one cm long, about one cm above the radio-ulnar joint distally) was performed to create a pseudarthrosis in order to permit free rotation in the forearm, especially in three severe cases,

Table 4. Follow-up examination of 26 patients with Madelung's deformity (13 treated operatively).

		Results of follow-up examination		
		Improved	Unchanged	Worse
A.				
13 patients operated on	Subjective	9	4	0
(7 MD, 5 D, 1 SMD) **	Rotation	1 *	10	2
B.				
13 patients not operated	Subjective	7	4	2
(6 MD, 5 D, 2 SMD)				

* $> 29°$ increased rotation in the forearm.
** MD = Genuine Madelung's deformity; D = dysplasia; SMD = symptomatic Madelung's deformity.
In Group A, 8 were working, 4 were at home/school, 1 had a disablement pension.
In Group B, 6 were working, 5 were at home/school, 2 had a disablement pension.

where the radius had a very steep joint surface (Figure 2, last film). Here, radio-ulnar arthrodesis was performed using a screw distally, to avoid ulnar dislocation of the carpus. Of the other 12 wrists, the radius deformity was found to be so pronounced in six cases, that in addition to one of the three surgical interventions on the ulna described above, a wedge osteotomy of the radius was performed, a radial skin incision being made and a subperiosteal wedge removed from the distal radius about two cm above the radio-ulnar joint. The base of the wedge was positioned more or less dorso-radially, depending on the steepness of the joint surface and the curvature of the radius. The osteotomy was immobilized with clamps, or bone or periosteal sutures. Reoperation was necessary in three cases; in two of them the ulna was shortened, and in the third a radio-ulnar re-arthrodesis was performed.

All patients in the group were followed up. The operated patients were seen on an average 8 years and 6 months after the surgical intervention (from 6 months to 20 years).

RESULTS

The results are shown in Table 4. Nine out of thirteen operated patients reported a subjective improvement of their condition in the form of reduced pains. Four patients found their condition unchanged. Physical examination showed that the mobility, especially rotation of the forearm, had been significantly improved (i.e. more than 29°) in only one patient; in ten patients the mobility was generally unchanged and in two patients, the range of movement had deteriorated.

Table 5. Classification of Madelung's deformity.

I) Idiopathic Madelung's deformity.
a) Classic genuine Madelung's deformity.
b) Dysplasia of bone + Madelung's deformity.
II) Symptomatic Madelung's deformity (Pseudo-Madelung's deformity).

DISCUSSION

The literature shows that numerous classifications of a more or less complicated nature have been attempted, based either on morphology or on "aetiology" (Anton et al. 1938, Henry & Thorburn 1967, Jaikaran 1969, Kelikian 1974, Matev & Karagancheva 1975). A new classification which fits in with the material presented here is shown in Table 5.

All idiopathic deformities were found to be bilateral, often unequal, with a considerable preponderance of symptoms on one side. This applied both to patients with genuine classic MD and to patients with dyschondrosteosis or some

other form of bone dysplasia, contrary to the findings of Matev & Karagancheva (1975) and other authors. In both groups there was a familial occurrence of the disease (Table 2), but this was more common in the latter group. More remarkable, however, is the fact that almost all patients in the group with idiopathic MD were women.

Already in his studies Madelung was aware that most often the trouble appeared between the ages of 8 and 14 years, often progressing from initially mild functional pain to fatigue and loss of strength, and finally reduced mobility. After some years the trouble often became stationary, and the pain decreased. Due to this fact a certain period of observation was required in order to be able to reach a decision as to whether or not operation should be performed.

The results show that on an average our patients were operated almost 10 years after onset of symptoms. The reason was our decision to delay operation until termination of growth and closing of the epiphysis of the radius had occurred at the age of 18 to 20 years.

Of the thirteen operated patients, nine felt that the operation had helped; they had less pain, better subjective function and a cosmetically more attractive wrist (Table 4). However, in only one patient was the mobility, especially rotation of the forearm, significantly improved. It had remained more or less unchanged in ten patients, and had deteriorated in two patients. The patient with improved rotation, however, did not experience any beneficial effect from the operation, while the two patients with more limited rotation postoperatively considered that the operation had been helpful. This agrees with the observation that limitation of movement is not a serious complaint. What the patients attach importance to, and what leads them to seek medical help, is the pain. This was the case in 23 out of the 26 patients in question. The last three patients sought treatment for cosmetic reasons due to the prominent head of the ulna.

Based on this investigation, operation cannot be considered of any significant value as regards mobility, but the functional pain decreases and the strength is evaluated subjectively as having increased. Ranawat et al. (1957) found similar results. Only a minority of patients require operation.

We have adopted an attitude of reserve, and a longer period of observation is required, at least during the period of growth, before surgical intervention is decided upon and choice is made as to the method preferred.

This material still provides no real aetiological explanation for MD, but some sort of "aetiological" classification as shown in Table 5 would seem rational and easy to grasp.

Conclusion

On the basis of the literature and this material, a new classification of MD is recommended.

Almost all patients included in the investigation with non-traumatic MD are women, and a familial incidence is common.

Because of a difference in the degree of severity, the complaint is often only one-sided. Unilateral deformity was found only in patients with so-called traumatic MD. The most severe cases are found in patients with other forms of bone dysplasia, especially dyschondrosteosis. It is common to find spontaneous improvement in pain and in impaired strength at the end of the growth period. Therefore a period of observation is recommended after closure of the epiphyseal line, before any decision is made to operate.

No specific surgical method can be recommended. The type of intervention must depend on the degree of severity of the deformity. Only a minority of pa-

tients require surgical correction, and this rarely improves mobility to a significant degree, although it reduces pain, improves subjective function and results in a cosmetically more attractive wrist.

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