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CONGENITAL RADIO-ULNAR SYNOSTOSIS

Report of 37 Cases

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Congenital synostosis of the proximal ends of the radius and ulna is a rare malformation which often completely prevents pronation and supination of the forearm. According to Wilkie (1914) the first case was described in 1793 by Sandifort in his *Museum Anatomicus*. Perusing the literature up to 1964, Jeanty found that so far a total of about 220 cases had been published. During the period 1912–1967, a total of 124 patients with congenital malformations of the forearm were seen in the Orthopaedic Clinic in Munich. Among these patients Albrecht (1967) found 7 with congenital radio-ulnar synostosis.

PRESENT MATERIAL

At a brief interval we saw 2 patients with congenital proximal radio-ulnar synostosis. Perusal of the diagnostic files from 10 orthopaedic departments in Denmark revealed that during the 10-year period 1958–1968 this deformity had been diagnosed in 37 cases. The patients were evenly distributed over the country.

RESULTS

Of these 37 patients 16 were females and 21 males. In 28 cases the diagnosis had been made between the ages of 1 and 12 years, and the remaining 9 patients were between 16 and 52 years. Twenty-eight were referred to an orthopaedic out-patient department because members of the family had noticed reduced mobility of the forearm, 2 were detected by chance when examined for something else, and 7 applied because of difficulty in performing their work.

In 5 instances there was a familial predisposition. All the familial cases were bilateral, transmitted through the paternal line. In one of

the families the disease was present in the patient's paternal grandmother, father, and 3 paternal uncles.

In 12 cases the anomaly was right-sided, in 6 left-sided, and in 19 bilateral.

Several authors have classified the radio-ulnar synostoses into two types. In one type (the so-called primary radio-ulnar synostosis) the proximal end of the radius is imperfectly formed and is fused to the ulna for a length of 3–6 cm. The normal contour of the radial head is completely obliterated, and the radial shaft is distinctly forward arched, longer, and considerably stouter than the ulna which is more slender than usual. In the other type synostosis is less marked, and the radial head is dislocated forward or backward. In the present material, 27 cases were of the former type and 6 of the latter. In four of the bilateral cases, type I was found on one side and type II on the other.

In all the patients the hand was more or less pronated. In 12, it was fixed in maximum pronation. In type I cases pronation and supination were practically abolished, while in type II mobility was from a few degrees up to approximately half the normal range. About one-third of the patients had an extension defect of 10–30° in the elbow, whereas the others had free mobility in this joint.

No patient showed congenital malformations other than the synostosis.

It is apparent from the case records that in a number of the cases there had been doubt as to whether the patients were to be advised to have operation of the synostoses. Several of the children were followed with a view to possible operation when they had grown up. Only two of the patients underwent operation. These two cases, as well as a typical case history, will be reported below.

CASE REPORTS

1. A 16-year-old girl was referred to an out-patient department because of reduced mobility of the right forearm. The complaints were mild, manifesting themselves mainly in difficulty of writing and sewing. The symptoms had been present from birth. There were no similar cases in the family.

On physical examination the right forearm was found to be fixed in a position of extreme pronation. Pronation-supination was completely abolished. Otherwise, mobility of the elbow joint and wrist was free. On the left there was also a tendency to pronation of the forearm. On this side the range of pronation-supination was about half the normal. X-rays (cf. Figure 1) showed on the right a 4½ cm area of total synostosis between the proximal end of the radius and ulna. The radius was very stout and distinctly forward arched. On the left there was a very

small synostosis combined with upward and backward dislocation of the radial head.

The patient was advised against operation.

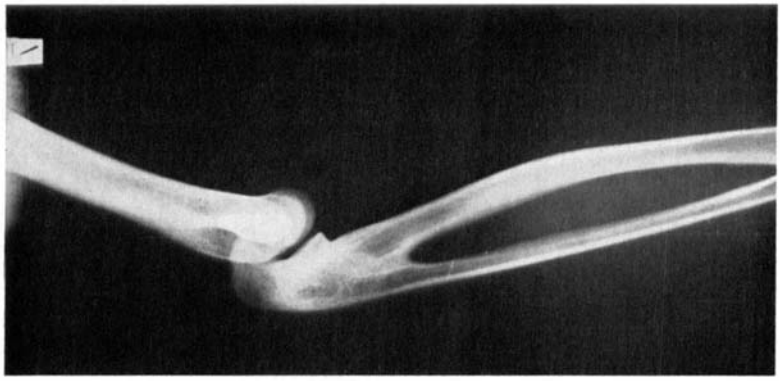

Figure 1. Congenital radio-ulnar synostosis in a girl, aged 16 years. Right arm.

2. A 16-year-old girl with bilateral proximal radio-ulnar synostosis was referred because of abolished rotation in both forearms. Partial resection of the left radius was performed, removing 2½ cm of the bone just below the synostosis. In spite of intensive training, active rotation of the forearm was not attained. At X-ray follow-up 18 months later there was again osseous contact between the resected bone ends. Supination-pronation was still abolished. Subjective complaints had been somewhat reduced, as the hand, previously in maximum pronation, was now fixed in 30° pronation. The patient was not interested in an operation on the right arm.

3. A 16-year-old boy was admitted with right-sided congenital proximal radio-ulnar synostosis. The hand was in maximum pronation, and supination could not be performed. As the patient was going to be an artisan, he needed better function of the hand. Osteotomy was done at the junction of the distal two-thirds and the proximal one-third of the radius. The hand was fixed in 30° pronation. During the postoperative course the function of the radial nerve was affected, but this improved considerably after myotensor therapy. Three months after the operation the osteotomy had healed. Pronation-supination was still abolished, and the subjective complaints were practically unchanged.

DISCUSSION

According to the present study, congenital radio-ulnar synostosis is somewhat more common than would be expected according to previous investigations.

The symptoms in these cases consist of reduced or entirely abolished

supination and pronation of the forearm. Apparently, the hand is always fixed in pronation. This is fortunate, since the patients would be much worse off with the hand in supination. The restriction of mobility may to some extent be compensated for by movements of the shoulder. In unilateral cases the affected forearm is often thinner and somewhat shorter than the good one. The explanation why the patients seldom complain of the restricted mobility is perhaps habit, as they have never known free use of the forearms.

From the literature it is apparent that the condition is bilateral in more than 80 per cent of the cases. In our series close to 50 per cent were bilateral. The malformation appears to be equally common in both sexes. In a few cases there is a family history of synostosis (Freyer 1966, Mercer 1950, Wilkie 1914). Among our patients, about 13 per cent had a familial predisposition. These cases, apparently always bilateral, were transmitted mainly through the paternal family.

Radio-ulnar synostosis has been described in Klinefelter's syndrome with an XXYY chromosomal pattern. Among 6 patients with radio-ulnar synostosis, Cleveland et al. (1969) found two to be of an XYY karyotype. The relationship of the bony abnormality to the presence of an extra Y chromosome was not clear. A chromosomal study was not performed on the present material.

As already mentioned, many authors have divided the synostoses into 2 separate types. In 4 of our cases the condition was type I on one side and type II on the other. It seems reasonable to assume, therefore, that there is not a question of 2 separate varieties of malformation, but of a difference in degree.

It is well known that synostosis between the radius and ulna may arise as a consequence of fractures. It is extremely rare for this acquired form to be mistaken on the X-ray film for the congenital form. Fielding (1964) has reported the case of an 11-year-old boy in whom traumatic epiphysiolysis of the radial head was followed by radio-ulnar synostosis of the same configuration as the congenital variety. He was unable to find reports of similar cases in the literature. Congenital and traumatic dislocation of the radial head in children has been reported several times. Schubert (1965) published a case arising from a difficult breech delivery. The dislocation did not lead to radio-ulnar synostosis, as it was recognized immediately and treated by reduction and fixation in plaster. It cannot be ruled out that some of the unilateral synostoses are not congenital but due to injuries sustained at or shortly after birth.

It has been reported that patients with congenital radio-ulnar synostosis frequently have other malformations too, e.g. dislocation of the hip, anomalies of the knee joint, club-foot, flat feet, Madelung's deformity, syndactyly, and polyactyly (Freyer 1966, Mercer 1950, Wilkie 1914). Among our 37 patients 2 had Scheuermann's kyphosis and 2 were intellectually impaired, but there were no cases of other malformations.

Operations and autopsies have revealed, apart from the osseous changes, atrophy and fibrosis of the supinator muscles as well as of the pronator teres and pronator quadratus. The interosseous membrane is thickened and very tight (Hohmann 1962, Mercer 1950, Wilkie 1914).

The results of surgical treatment have often been extremely unsatisfactory (Bier et al. 1958, Jeanty 1964, Mercer 1950, Wilkie 1914). Even after radical resection of the synostosis with interposition of fat, muscle, or a piece of plexiglass, and after passive pronation and supination of the forearm has been rendered completely free, considerable or total restriction of movement usually recurs (Bier et al. 1958). Hohmann (1962) published the case of a 6-year-old boy whom he had treated by cutting the shaft of the radius in the middle and turning the forearm from maximum pronation to an intermediate position. Thereby he obtained considerable improvement of hand function. Kelikian & Doumanian (1957) designed a stainless steel swivel which they inserted into the medullary cavity after resecting a piece of the radius, so that the bone could rotate on the cylindrical prosthesis. This prosthesis has been used successfully in 4 cases of traumatic radio-ulnar synostosis. Although they had not tried it, they believed that the swivel was well suited for the congenital cases too. However, the result is not likely to be favourable, as the congenital synostoses consist not only in osseous changes, but also in changes of the muscles and interosseous membrane. Therefore, even though free passive pronation-supination is obtained, active mobility will not be significantly improved.

On the basis of the literature and the above case histories (2 and 3), it may be concluded that it is advisable to refrain from operation in most cases of congenital proximal radio-ulnar synostoses. If, in a rare case, the complaints are so severe that an intervention is required, it seems reasonable to restrict it to osteotomy on the forearm, placing the hand in an optimum position for function.

SUMMARY

In 10 Danish departments of orthopaedic surgery 37 cases of congenital radio-ulnar synostosis were diagnosed during the 10-year period 1958–1968. Of these patients 16 were females and 21 males. 12 cases were right-sided, 6 left-sided, and 19 bilateral. Five of the patients had a family history of radio-ulnar synostosis. These cases, all bilateral, had been transmitted through the paternal family. In the literature the synostoses are divided into 2 types. In 4 of the present cases we found type I on one side and type II on the other. Very probably, therefore, there is not a question of two different forms of malformation, but of a difference in degree.

Two of the patients had undergone operation without obtaining any essential improvement of their condition. It is concluded that congenital radio-ulnar synostosis should in most cases not be treated surgically.

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