CONGENITAL PSEUDARTHROSI S OF THE ULNA DUE TO NEUROFIBROMATOSIS

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Two cases of congenital pseudarthrosis of the ulna due to neurofibromatosis are reported. Similar radiographic changes in the ulna were found, with distortion of the capitulum and part of the trochlea and bowing of the radius. In one patient the head of the radius was dislocated and in the other it dislocated readily during pronation. A possible mechanism to explain the findings was suggested. On the basis of these and a review of the literature it is recommended that the main aim of treatment should be maintenance of the normal relative lengths of the radius and ulna by early excision of the pseudarthrosis to remove the restraining effect of the abnormal ulna. This will allow normal development of the lower end of the humerus and radius and prevent dislocation of the radial head.

Isolated pseudarthrosis of the ulna due to neurofibromatosis is rare. Nine cases have been reported in children (Moore 1949; Madsen 1956; Cobb 1968; Shertzer, Bickel and Stubbins 1969; Sage 1980; Allieu et al. 1981). The present study describes two patients with the condition who have been followed into adult life and draws attention to the effect upon the elbow.

CASE REPORTS
Case 1. An abnormality of the left forearm was noticed in a boy aged nine months. Increasing deformity prompted referral to this hospital at the age of five years.

The patient had multiple café-au-lait spots typical of neurofibromatosis although there was no family history of the condition. Varus deformity of the elbow was 30 degrees, pronation was limited by 10 degrees but supination was full. Radiological examination showed a pseudarthrosis in the middle of the ulna with atrophy of the distal half, although the distal epiphysis was present, and bowing of the radius with dislocation of the head (Figs 1 and 2).

The pseudarthrosis and the fibrous remnant of the distal ulna were excised leaving the epiphysis intact. The defect was replaced by a segment of the patient’s fibula and secured by a Kirschner wire (Figs 3 and 4). Histological examination of the excised tissue revealed woven bone at the proximal end and distally this bone abruptly changed into fibrous tissue containing abundant collagen fibres enclosed in a fibrous periostem.

Eight months after the operation the graft had totally reabsorbed but despite this the forearm regained excellent function.

When seen for review 20 years later the patient was working as a surveyor and had no complaint related to his left arm. Examination revealed there was still a 30-degree varus deformity at the elbow and an obvious dislocation of the head of the radius. The range of flexion at the elbow was 20 to 120 degrees and both pronation and supination were 0 to 45 degrees. Apart from a slight objective weakness of the power grip in the left hand no other functional abnormality was found in the limb.

Radiographs (Figs 5 and 6) confirmed the presence of dislocation of the radial head with flattening of the capitulum and part of the trochlea. Interestingly the distal ulna had reformed and the wrist joint appeared normal.

Case 2. A female shop assistant was referred because of deformity of her right forearm (present since birth) and a complaint of pain in the forearm on exertion.

There was a 25-degree varus deformity at the elbow (Figs 7 and 8) but no restriction of movement. Pronation was 0 to 60 degrees, supination 0 to 100 degrees and the hand functioned normally. The head of the radius could be readily dislocated during pronation. She had no café-au-lait spots or any other sign or family history of neurofibromatosis. Radiological examination revealed a similar appearance to the first case, associated with marked bowing of the radius, and was characteristic of neurofibromatosis.

As she was to be married she requested improvement in the appearance of the forearm. The deformity was corrected by osteotomy of the radius and maintained with an AO compression plate. Satisfactory correction of the deformity and union of the osteotomy were obtained (Figs 9 and 10). At the final follow-up nine months after surgery she was shown to have regained almost the full range of forearm movements seen before operation and was delighted with the cosmetic improvement.

DISCUSSION
The radiological features of these two patients resemble closely those seen in previously reported cases (Moore 1949; Madsen 1956; Cobb 1968; Sage 1980; Allieu et al. 1981).

We suggest that normal growth of the radius with tethering of the ulna results in increased pressure on the lower humeral epiphysis causing impaired development of the capitulum and part of the trochlea, making the radiohumeral joint unstable. During growth the discrepancy in length between the radius and ulna causes bowing
of the radius and eventually dislocation of its head. Review of the published radiographs and those of our two patients confirm the above changes.

In Case 2 the head of the radius was in place but could be readily dislocated. The gross bowing of the radius may have been less had the radial head dislocated. The patient showed no evidence of generalised neurofibromatosis, although the radiological appearance of her ulna was characteristic. Pseudarthrosis of the ulna (like that of the tibia) is not invariably associated with generalised neurofibromatosis (Aegerter and Kirkpatrick 1975).

**Treatment.** Treatment is difficult. Madsen (1956) suggested that conservative treatment suffices because there

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**Figure 1** - Pseudarthrosis in the middle of the ulna with atrophy of the distal half (16 November 1959).

**Figure 2** - Three months after operation (21 March 1960).

**Figure 3** - At final follow-up (15 April 1981).

**Figure 4** - Pseudarthrosis of the middle of the ulna with atrophy of the distal half (19 December 1980).

**Figure 5** - At final follow-up nine months after operation (9 October 1981).
is no significant functional loss. Surgical treatment ranges from simple excision to repeated bone grafting using various techniques, but success has been inconsistent (Moore 1949; Madsen 1956; Shertzer, Bickel and Stubbins 1969; Sage 1980; Allieu et al. 1981). Results have been difficult to assess as the number of cases is small and the follow-up short. We have no experience of vascularised fibular bone grafts (Allieu et al. 1981). On the basis of review of the literature and these two additional cases we recommend that the main aim of treatment should be maintenance of the normal relative lengths of the radius and ulna by early excision of the pseudarthrosis to remove the restraining effect of the abnormal ulna. This will allow normal development of the lower end of the humerus and radius and prevent dislocation of the radial head.

We are sincerely grateful to Mr D. W. Lamb for his guidance and for allowing us to review his patient.

REFERENCES


