Radioulnar fusion for forearm defects in children – a salvage procedure

MN Rasool
Department of Orthopaedics, Nelson R Mandela School of Medicine, KwaZulu-Natal

Abstract
Eight children aged 1-14 yrs with defects in the forearm were treated with the one-bone forearm procedure and followed up for 1-11 yrs. The defects were due to pyogenic osteomyelitis (n=3), osteochondroma (n=3), neurofibromatosis (n=1) and ulnar dysmelia (n=1). The radius was fixed to the ulna shaft with an intramedullary pin in six cases, and two children had centralisation of the radial metaphysis onto the ulna for “radial club hand” type deformity with Kirschner wires. All forearms united in 3-6 months. Shortening ranged from 1-10 cm. Fixed flexion deformity of the elbow (20°) resulted in one child and cubitus valgus (20°) occurred in another. One child had a radial articular tilt of 45°. The procedure achieved stability at the wrist and elbow. There was cosmetic and functional improvement in all patients.

Introduction
The treatment of forearm defects in children is challenging. When one growing forearm bone is destroyed by disease, develops imperfectly or abnormally, secondary structural changes are likely to follow. The defects in the growing radius and ulna can be caused by trauma, infection, congenital abnormalities, bone dysplasias and tumours. With defects of the radius the relative overgrowth and bowing of the ulna results in instability of the distal radioulnar joint with radial deviation of the hand and carpus. A cubitus valgus deformity can result. With ulna defects, there is radial overgrowth and bowing with resulting dislocation of the radial head and ulnar deviation of the radius and carpus. A cubitus varus deformity and forearm shortening result. Forearm defects result in several problems such as weakness of grip, instability of the wrist and elbow, bowing of the forearm, cosmetic deformity and pain at either joint. The choice of treatment of such defects lies between replacement grafting, bone lengthening, vascularised grafting and radio ulnar fusion. Radioulnar fusion or transposition is a salvage procedure in selected cases and produces a one bone forearm with loss of elbow rotation. Rotation at the shoulder compensates for this adequately. Correction of the deviation of the hand on a solid forearm gives a much stronger grip, and improves elbow and wrist movements. Longitudinal growth continues and improvement in cosmesis occurs.

The operation for radioulnar fusion was first devised by Hey Groves in an adult for traumatic defect of the radius. Greenwood described success in a child with osteomyelitis of the distal radius. Since then several case reports have appeared on the production of a one-bone forearm for patients with a partial defect of the ulna, or partial defects of the radius, or both the radius and the ulna. Various methods were used by surgeons to hold the radius to the ulna including catgut sutures, cerclage wires, side to side cross union with screws or Kirschner wires, plating and intramedullary pinning through both shafts. A series of six children was reported by Lowe for defects due to osteomyelitis, trauma, osteochondroma and congenital problems. Ono reported on nine children, but these were all distal procedures for “radial club hand deformity” following osteomyelitis.
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<td>Osteomyelitis radius</td>
<td>&quot;Radial club hand&quot;, deformity, shortening, radioulnar joint subluxation</td>
<td>Defect of whole radius, residual epiphysis</td>
<td>1</td>
<td>Full flexion extension of elbow, good rotation and forearm shortening 4 cm. Grip P3, stiff MPJ hand, stiff wrist</td>
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<td>2</td>
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<td>R</td>
<td>Osteomyelitis radius</td>
<td>&quot;Radial club hand&quot;, radioulnar joint subluxation</td>
<td>Defect of whole radius, residual metaphysis and epiphysis</td>
<td>2</td>
<td>Full flexion extension of elbow. Good forearm rotation, shortening 3 cm. Grip P4</td>
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<td>R</td>
<td>Osteomyelitis ulna</td>
<td>Radial head dislocation, shortening, pain at elbow, cubitus varus</td>
<td>Absent distal two thirds ulna, dislocated radial head</td>
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<td>Neurofibromatosis</td>
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<td>Osteochondroma distal ulna (Masada IIb)</td>
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<td>1.5</td>
<td>Full flexion extension elbow. Good rotation forearm. Grip P4, shortening 1 cm. (surgery to right side only)</td>
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<td>Osteochondroma prox. radius and distal ulna (Masada Ila)</td>
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<td>Short ulna, dislocated radial head, osteochondroma prox. radius and distal ulna</td>
<td>1</td>
<td>Full flexion, extension at elbow. Good rotation forearm. Grip P4, shortening nil</td>
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<td>8</td>
<td>F</td>
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<td>R</td>
<td>R</td>
<td>Osteochondroma distal ulna (Masada IIb)</td>
<td>Osteochondroma ulna. Marked radial bowing, dislocated radial head. Cubitus varus</td>
<td>Large osteochondroma ulna. Radial bowing, dislocated radial head</td>
<td>1.2</td>
<td>Full flexion extension elbow. Good rotation forearm. Grip P4.5, shortening 4 cm</td>
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The aim of this report is to discuss the results of radio-ulnar fusion for forearm defects in the radius and ulna by intramedullary fixation.

**Materials and methods**

The case records and radiographs of eight children treated by radioulnar reconstruction for forearm defects were reviewed retrospectively from 1990 to 2006 (*Table I*). Two children were first seen in the neonatal period and the remainder presented between 1 and 14 yrs. There were two males and six females. The dominant side was involved in three children. Six children had right-sided involvement, one left-sided, and one had bilateral involvement. The latter child had surgery to one side. There were three children with defects resulting from primary pyogenic osteomyelitis following sequestrectomy. Two of these had radial defects, and in one, the ulnar was involved. The former two had distal radioulnar instability with a “radial club hand” type deformity (*Figure 1a*), and the latter one had dislocation of the radial head.

There were three children with osteochondroma of the ulna. Two had multiple exostoses together with ulna involvement and one had localised ulna involvement. One of these had in addition an exostosis of the radial neck with radial head dislocation (Masada type Ila) and two had Masada IIb deformity (ulnar exostosis alone with radial head dislocation) (*Figure 2a*). One child had neurofibromatosis with numerous café au lait spots, pseudarthrosis of the ulna with a dysplastic and bowed radius and marked ulna deviation of the hand (*Figure 3a*). The radial head was clinically unstable. The remaining child had congenital longitudinal deficiency of the ulna with radial bowing and ulna deviation of the carpus and radial head dislocation (*Figure 4a*).

All patients had progressive deformity, complained of shortening and instability with weakness of grip and marked limitation of rotation. Those with radial head dislocation had a fixed flexion deformity of 30°-40° and cubitus varus of 20°-30°. Elbow pain was the presenting feature in three patients.

**Operative technique**

Through a posterolateral incision, the anconeus was reflected and the radial head and neck was excised in six children. In the two patients with “radial club hand” the radial head was absent due to sequestration following osteomyelitis.

Through two separate dorsal incisions, in four patients, and a single dorsal incision in four others, the radius and ulna were exposed at the appropriate level. The osteochondroma was excised in all three patients with distal ulna involvement and in the one with radial exostosis the tumour was excised with the radial neck. The radius and ulna were transected at the same level transversely and the proximal radius shaft was excised subperiosteally. The
The distal radius was passed through a small defect in the interosseous membrane and united to the ulna with an intramedullary pin through the olecranon in six children and from the radial styloid in two children with “radial club hand” deformity. The ulna was segmented in two children and the radius in two others to correct the curvature of the shaft and the radial articular tilt. Bone chips from the resected neck and shaft were placed around the osteotomy site. The wound was closed over a suction drain and immobilised in a plaster cast in neutral rotation and 90° of elbow flexion, and the wrist at 20° dorsiflexion.

Postoperative care
Immobilisation was continued for two more months after plaster change at 2 weeks postoperatively and thereafter for a further 3 months in a moulded splint. Physiotherapy to the shoulder, elbow and wrist was commenced during splintage.

Results
The patients were assessed clinically and radiologically. Bone healing was complete by 6-8 weeks (Figures 1b, 2b, 3b, 4b). Remodelling of the shaft continued to occur thereafter. There were no nerve palsies or wound break-down. One child had slight scar hypertrophy. Follow-up ranged from 1 year to 11 years (average 4 years). Subperiosteal new bone formation occurred in the resected radius in five patients and the distal ulna in two. The radial head reformed partially in one child and flexion was not impaired. One child had a fixed flexion deformity of the elbow of 20° and another had cubitus valgus of 20° without ulnar neuritis. No instability of the elbow could be elicited. Elbow flexion was beyond 120° in all patients and forearm rotation occurring at the shoulder, was full in all. The grip power ranged from P3 to P4.5, shortening ranged from 2 cm to 10 cm. One child had a stiff wrist and stiff metacarpophalangeal joints of all the fingers; this was present pre-operatively.
One child had a radial articular angle of 45° (Figure 3b) and there was no evidence of carpal slip in any patient. All parents reported improved function and cosmetic appearance.

**Discussion**

Partial defects of the forearm in growing bones can result in deformities and disabilities due to secondary structural changes. Stability is provided at the elbow by the ulna and at the wrist by the radius. Stress is usually transmitted from the radius to the ulna through the interosseous membrane.

Defects, especially of the distal half of the ulna, with continued radial growth result in dislocation of the radial head with lateral instability due to the discrepancy in the length between radius and ulna. Cubitus varus deformity and bowing of the radius occurs resulting in a shortened forearm. Ulna deviation occurs at the wrist with weakness of grip and pain on elbow motion. Lack of full extension and rotation occurs. Deficiency of the radius especially the proximal half, results in instability of the distal radioulnar joint, bowing of the ulna and cubitus valgus deformity. Radial club hand type of deformity can occur with entire diaphyseal deficiency.

For these deformities and impairments, a stable elbow and forearm can be established by synostosis of the proximal part of the ulna and distal part of the radius following resection of the proximal radius and distal ulna. The procedure creates a one bone forearm that may have growth potential, allows correction of ulna or radial bowing, provides elbow and wrist stability and improves function and cosmetic appearance.

Common causes of these forearm defects are trauma, congenital abnormalities of the ulna, chronic osteomyelitis with pseudarthrosis or following sequestrectomy, benign tumours especially osteochondroma and pseudarthrosis of the forearm bones due to neurofibromatosis.

In traumatic defects of a single forearm bone, autogenous bone grafting can restore the architecture when the graft and internal fixation devices are well covered by soft tissue and when the defect is small. When the remaining ends of bone are too small to receive a graft, or in growing bone with epiphyseal involvement, radioulnar fusion offers a better solution to the problem.

Hey Groves’ first reported the use of the one-bone forearm in adults for trauma. Subsequently, Murray used the technique in two adults by achieving cross union of the radius to the ulna with transverse screws. Haddad first reported its use in children for traumatic defects.

Primary pyogenic osteomyelitis of the forearm bones is rare. The radius is more commonly involved with the acute process occurring in the distal metaphysis resulting in pathological fracture, sequestrum formation and growth disturbance (Figure 1a). The commonest manifestation is a “radial club hand” type of deformity with a small distal radial fragment, radial deviation of the carpus and a prominent ulna styloid due to dorsal subluxation.

Several cases of bone defects resulting from osteomyelitis of the forearm bones have been reported, mainly in the radius. Dugdale used interpositional corticocancellous grafts successfully in small defects. The results of strut grafting in large defects have been unsatisfactory. Centralisation of the carpus has been described for large radial defects with epiphyseal destruction.

Griffiths used the one bone forearm technique for radial defects in two children following osteomyelitis. The follow up was short (2 months). Netrawichien described two children treated with AO plating and autogenous bone grafting. The defect was lengthened immediately and an open reduction of the subluxed distal radioulnar joint combined with ulna shortening was performed. However, limitation of rotation was still a problem. Ono used an external fixator to distract the radial remnant, with centralisation of the carpus on the distal ulna, and subtotal proximal row carpectomy in two patients.
Several problems were encountered including pin tract infection, median nerve paralysis, recurrent ulnar bowing, non-union and wound infection. Repeat bone grafting had to be performed.

In neurofibromatosis, bone grafting of defects in the forearm are rarely described in the literature and the management is more difficult due to the abnormal nature of the bone. In congenital pseudarthrosis of the ulna the bone ends are tapered with sclerosis and obliteration of the medullary canal (Figure 3a). The epiphysis of the ulna is usually present, but may not be seen clearly on radiographs. With continued growth the radial head dislocates.21–28 The distal radioulnar joint is usually intact, but there is a loss of wrist alignment.3 The radius may also be involved with bowing or pseudarthrosis. Historically union is difficult to obtain; corticocancellous grafting has achieved union of ± 35% in the forearm bones.29–31 Fabry32 used the Ilizarov technique to obtain union. The procedure is resource-intensive and few reports are available. Its use in the case where the greater part of the distal ulna is involved is difficult to envisage. Free vascularised fibular graft (FVBG) has a high success rate in pseudarthrosis. Follow-up in the forearm bones is short. Problems are distal fixation of the graft, limitation of rotation, refracture and delayed union.33–35

A stable elbow and forearm can be established by synostosis of the proximal part of the ulna and distal part of the radius following resection of the proximal radius and distal ulna

The procedure has been combined with Ilizarov lengthening.22 The limitations of these procedures are: availability of expertise, duration of the procedure and complications including nerve palsies, non-union and loss of motion. The one bone forearm procedure was used on several occasions for neurofibromatosis in children with successful functional outcome.34,35,36,37,38,39 Congenital longitudinal deficiency of the ulna is a rare upper limb abnormality.36 Various terms are used to describe the defect: ulna club hand, ulnar dysmelia and paraxial ulna hemimelia (Figure 4a).

Three types have been described by Ogden.9 Type I is a hypoplastic ulna with a distal epiphysis; type II – partial absence of the ulna with radial head dislocation; and type III – total absence of the ulna. A fibrocartilaginous band (anlage) is usually present in type II and attaches the distal ulna to the carpus or radial epiphysis and contributes to the progression of radial bowing and ulna deviation of the wrist resulting in radiohumeral dislocation and instability.

The one-bone procedure combined with resection of the fibrocartilaginous anlage gave good results in several case reports.22,35,36,37,38 In this study one patient was seen in whom the distal ulna regenerated after subperiosteal resection and gave support to the carpus. The anlage was not completely excised.

In multiple hereditary exostosis, significant forearm deformity occurs in 30% to 60% of patients.40,41 The commonest presentation is distal ulna involvement with a “carrot-shaped” or “licked candy stick” distal end.19

The ulna becomes shortened and fails to keep pace with the growing radius. The radius bows, and in progressive cases the radiohumeral joint dislocates, the carpus and the hand deviate ulna-ward. Forearm rotation and grip strength are compromised.19,30–32

Shortening occurs because the ulna contributes more to longitudinal growth than the radius, and the ulnar growth plate is one-quarter the size of the radial physis.33

Masada42 et al have classified forearm deformity due to osteochondroma into three types. Type I and III involve the distal ulna and distal radius respectively and are not associated with dislocation of the radial head.

Type Ia and IIb are less common and are associated with radial head dislocation.

Masada43 suggested excision of the osteochondroma and correction of the radius with osteotomy and gradual ulna lengthening with an external fixator to reduce the radial head. Problems encountered were radial nerve paralysis and proximal radioulnar synostosis.

Fogel44 et al used immediate ulna lengthening by osteotomy and interposition grafting, and internal or intramedullary fixation after excision of the osteochondroma, coupled with radial epiphyseal stapling as a preferred method of treatment.

Dal Monte45 et al reported ulnar lengthening either by immediate lengthening through osteotomy and plating or by gradual lengthening with an external fixator. Reduction of the radial head was achieved in two patients only. Pritchett,46 in a similar study in eight children, had no improvement of rotation in three. Other problems were recurrence and infection.

The one-bone forearm procedure was used successfully in several reports for treatment of osteochondroma of the ulna.19,30–33

This method is a less demanding technique than lengthening and vascularised grafting procedures. The function of the wrist and elbow are improved allowing growth of the forearm to continue. There is improvement in cosmetics with stability of the wrist and elbow.

The prognosis depends on the amount of distal radius that was injured by the infective, or dysplastic process or tumour involvement. The duration, extent of defect, age of patient and amount of initial shortening usually dictate the outcome. In this series, one patient had a stiff hand due to the initial underlying infection in the neonatal period. Another had an abnormal radial articular angle due to the severe initial deformity as a result of neurofibromatosis. The procedure is a method of salvage when other methods are not possible. End-to-end intramedullary rodding is technically easier than cross-union of the radius to the ulna which requires more extensive stripping of the interosseous membrane and periosteum. Segmentation and rodding allows realignment of the shaft and corrects radial tilt.
Subperiosteal excision allows new bone formation for reattachment of forearm muscles and maintains forearm width.

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References