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TECHNICAL NOTE

Distraction osteogenesis to improve limb function in congenital bilateral humeroradioulnar synostosis

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Congenital bilateral humeroradioulnar fusion of the elbow is rare. Many patients tolerate unilateral elbow stiffness very well, but bilateral fixed elbows are very disabling. We report the management of a 13-year-old patient using the Ilizarov technique. The left elbow was ankylosed in 70 degrees flexion. It was gradually lengthened through an osteotomy across the fused elbow and flexed through the regenerate to 110 degrees. This has improved the function of the left arm.

Keywords : congenital humeroradioulnar fusion ; distraction osteogenesis.

CASE REPORT

A thirteen year-old boy with right hand dominancy presented with bilateral congenital humeroulnar and radial fusion. His birth history was normal, and there was no family history of skeletal malformations. On examination both elbows were ankylosed. The right elbow was in 30° of fixed flexion, and the left elbow was in 70° of fixed flexion deformity. Pronation and supination movements were absent. Brachial and radial pulses were normal, and there was no neurological deficit in either forearm. The patient's biceps and triceps muscles were reduced in bulk. On further clinical examination, no specific syndromic features were noted. He had mild stiffness of the interphalangeal joints of both thumbs, but there was no radiological evidence of fusion across the interphalangeal joints.

Radiographs showed a bilateral humeroradial synostosis with ulnar ray hypoplasia (fig 1A and 1B). Although there was a humeroulnar articulation, it was not well formed. A skeletal survey did not reveal any skeletal malformations. Magnetic resonance imaging (MRI) scans of the left elbow confirmed normal neurovascular anatomy (fig 2).

Functionally, the patient was unable to reach his mouth with his hands. He was unable to join two hands in the midline because the elbows were fixed at different angles. His mother used to help him with washing his face and to feed him. His other concern was that he could not bring his mobile phone to his ear. He could reach his perineal region with his right hand because the elbow was fixed in 30° of flexion. On the left the elbow was fixed in 70° flexion making it impossible for him to reach the perineum or mouth. We decided to leave the

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Fig. 1A–B. — (A) Lateral and (B) AP radiographs show humeroulnar and radial synostosis with ulnar ray hypoplasia.

Fig. 2. — MRI scan of the elbow showing normal neurovascular anatomy.

Fig. 3. — Radiograph of the elbow after osteotomy with application of Ilizarov frame.

right elbow undisturbed and elected to treat the left elbow deformity to improve function.

Under general anaesthesia and tourniquet control the ankylosed elbow was approached through a medial and lateral approach. On the medial side the ulnar nerve was identified and isolated. An osteotomy was performed at the level of the ankylosed 'elbow', through the lateral approach. A three-ring Ilizarov construct – one ring on the humeral side and two rings on the forearm side, with hinges at the osteotomy level, was applied to the left arm (fig 3). After a lag period of one week, gradual distraction was performed to separate the ends of the osteotomy by approximately 1.5 cm. Then a posterior push and an anterior pull rod were applied (fig 4) to gradually increase the degree of flexion until the desired flexion position of 110° was achieved. At this angle the child was able to reach his mouth with his left hand and was happy to stop the correction (fig 5). We left the frame *in situ* until at least three cortices were seen, to prevent deformation of the regenerate after removal of the frame. The total duration in frame was 18 weeks. The elbow was then immobilized in a plaster cast for 6 weeks to protect the regenerate bone (fig 6).

There was substantial improvement in the level of function in the elbow after removal of the plaster. Preoperatively he was unable to reach his mouth with either left or right hand. After changing the flexion deformity from 70 to 110 degrees he was able to reach his mouth with the left hand. He

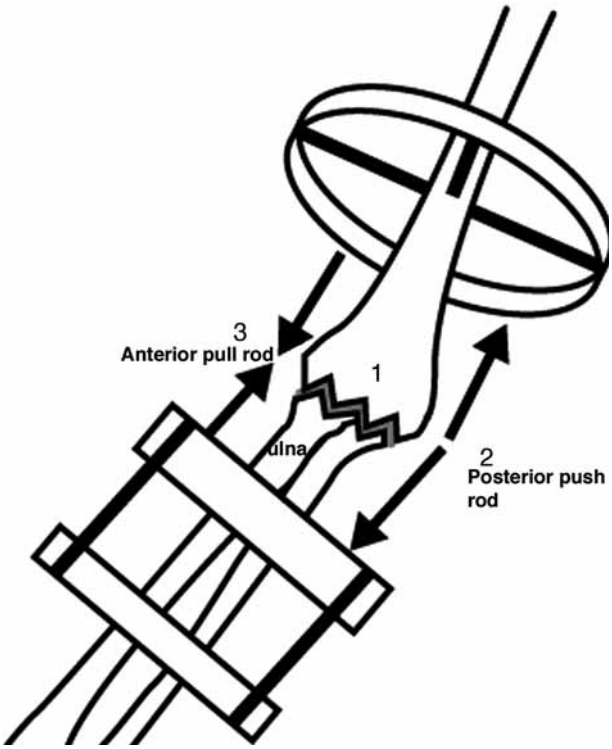

Fig. 4. — Line diagram showing the construct of the frame : Osteotomy (1), posterior push rod (2), and anterior pull rod (3).

Fig. 5. — Clinical photograph of the functional position of the left elbow.

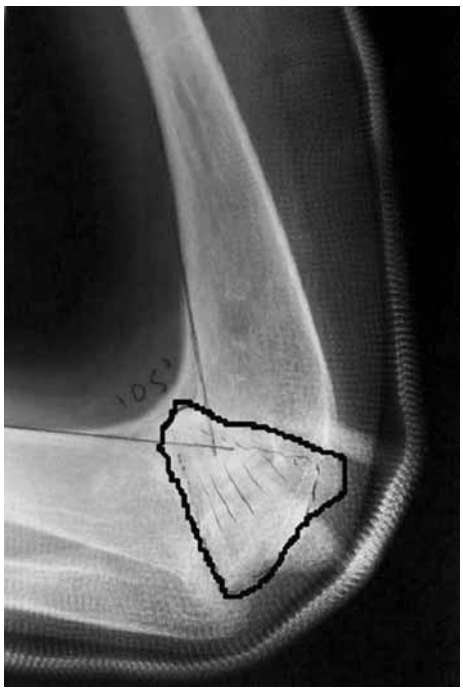

Fig. 6. — A postoperative radiograph of the regenerate

was also able to touch the back of his head and bring the hand close to his ear (while using the telephone). There were no complications during the frame treatment or after removal. At one year follow-up he maintains the achieved angle and improvement in function of the left upper extremity.

DISCUSSION

Congenital humeroradioulnar synostosis is a rare condition (*6*) and is generally unilateral. Bilateral involvement is even rarer. It is thought to occur from a failure of differentiation that may occur sporadically or as an extremely rare inheritable disorder (*2*). The current classification divides the synostosis into Class I (fixed in extension with ulnar ray hypoplasia) or Class II (fixed in flexion without hypoplasia). Class II may be associated with multiple systemic anomalies presenting as multiple synostosis syndromes (*5*).

Bilateral fused elbows can be extremely disabling if they are not fused in functional position, which is different on each side. It is recommended that one elbow should be placed in 100° of flexion to permit reaching the mouth and the other should be placed in 65° to aid in personal hygiene (*1*). Our patient's right elbow was in 30° of flexion, which was adequate for reaching the perineum. The left side was in 70° of flexion deformity and could not reach the mouth or perineum. We decided to improve the flexion angle of the left elbow so the patient could reach his mouth.

Various surgical options were considered. Elbow excision or arthroplasty were not suitable because the anatomy of the elbow was completely lacking. It was obvious from clinical examination that the biceps and triceps muscles were lacking in bulk and would have been inadequate to mobilize an excision arthroplasty. This was not entirely surprising because the muscles and tendons are directly related to the normal functioning of the joint *in utero* (*4*). An anterior closing wedge osteotomy would have involved taking an anterior based wedge in close proximity to the vessels, risking vascular injury. Moreover it would have shortened the forearm proportionate to the wedge taken. In theory, such shortening would have required more flexion angle to reach the mouth. Acute flexion of that degree can cause kinking of the vessels in the cubital fossa, similar to a supracondylar fracture of the humerus. We decided to achieve the flexion by first lengthening the forearm by gradual distraction and then gradually deforming the regenerate into flexion. The technique of Distraction Osteogenesis was described by Ilizarov and is extensively used in lengthening and correction of bone deformities (*3*). We haven't come across any literature reports of application of this technique to a case of congenital elbow fusion. We have successfully used the Ilizarov technique to gradually lengthen the forearm through an osteotomy at the elbow and were able to flex the regenerate bone to achieve the desired correction.

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