CASE REPORTS

Congenital absence (ankylosis) of the knee

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A congenital, unilateral, fixed flexion deformity in a neonate was diagnosed as a congenital absence of the knee. A single cartilage mass, with fusion of the lower femoral and upper tibial ossification centres, was demonstrated by imaging studies. This condition has been reported in the literature only once before. Surgery on our patient, which was performed at the age of two years, consisted of separation of the fused cartilaginous anlage and gradual correction of the deformity using an Ilizarov frame.

Congenital ankylosis of the knee is an extremely rare condition which causes a flexion deformity of the joint. Only one case has been previously reported in the literature.1 We describe a two-year-old boy who was born with a rigid flexion deformity of his right knee. Radiographs of the knee demonstrated fusion of the lower femoral and upper tibial epiphyses. Surgical exploration revealed an absent tibiofemoral joint space with a single cartilaginous anlage.

The aetiology of this type of malformation of the knee is unknown. Its appearance as an isolated, skeletal malformation raises the possibility of a focal embryonic defect. The estimated morphogenic stage during embryonic development may be that of the cavity appearance phase, O’Rahilly stage 22.2

A distal femoral extension osteotomy has been recommended as a method of correction of a flexion deformity.1,3 We performed an osteotomy through the fused epiphyses followed by gradual correction of the deformity using an Ilizarov frame.

Case report

The patient was born after a normal pregnancy and spontaneous delivery. He was the first-born child of young, unrelated parents. No abnormalities were detected in systematic pre-natal sonographic surveys. The birthweight was 3245 g, and there was no family history of musculoskeletal or neurological conditions. Physical examination after birth revealed bilateral clinodactyly, bilateral simian lines, retrognathia and an 80° fixed flexion deformity of the right knee. The right femur was 2 cm shorter than the left. The boy had normal psychomotor development and growth curves showed his weight and head circumference to be within the 40th percentile and body length within the 90th percentile. No chromosomal abnormalities were detected. The radiograph of his right lower limb suggested continuity between the distal femoral and proximal tibial secondary ossification centres, a feature that was confirmed by CT and MRI studies (Fig. 1). MRI of the brain and spine, as well as electrophysiological studies of the lower limbs, were normal.

He underwent surgery at the age of two years and two months. Exploration of the knee confirmed the absence of joint space with a single cartilage anlage joining the femur and the tibia. The extensor mechanism was normal (Fig. 2). Osteotomy of the fused cartilaginous ossification centres was performed, the hamstring muscles were released and an Ilizarov apparatus applied. Distraction and extension continued uninterrupted for three months and two weeks until full extension was achieved. The frame was then changed to a plaster cast for six weeks and thereafter to a removable brace for three months.

On review at the age of four years and two months, he had a straight leg with a normal mechanical axis, a leg-length discrepancy of 3 cm and an expected discrepancy of 8.5 cm (Fig. 3). He had returned to full activities for a healthy child of his age.

Discussion

Human limb malformations occur in approximately one in 1000 neonates. The developmental timetable of the knee has been well documented in several studies.2,4 In summary, the knee cavity appears during O’Rahilly stage 22, initially as the patellofemoral joint. The proximal tibiofibular joint communicates with the lateral meniscotibial joint between the tenth and 11th week of development and separates at or after the 13th week. Apart from its growth and polarisation, the formation of the limb bud includes numerous events which are necessary for the determination of limb identity and morphogenesis. Multiple genes are involved, some of which have been implicated in human limb malformations, such as those encoding T-box transcription factors, bone morphogenetic protein, cartilage-derived morphogenetic protein and homeobox genes.5

The development of joints is customarily divided into three phases. In the first phase, the blastemal condensations of skeletal elements are formed, while
joint cavitation takes place in the second. The synovium and intra-articular structures are formed in the third phase. Anomalies of the limbs may be isolated or associated with other developmental defects according to the expression domain of the gene involved. The precise mechanism which underlies joint cavitation is unknown. Cell death and apoptosis have been suggested as possible mechanisms. Movement has been shown to be crucial for cavitation to proceed normally. The isolated dysmorphogenesis in the absence of other skeletal malformations in our patient suggests a focal disturbance during the joint cavitation process.

Our literature survey revealed one similar description of congenital ankylosis of the knee in which there was a concomitant absence or complete atrophy of the extensor mechanism. The author recommended a supracondylar extension osteotomy in order to correct the flexion deformity. We chose to correct the deformity at its origin, the joint line. We believe that this approach affords a better correction of the joint and better alignment of the limb. We used the Ilizarov technique for gradual correction of the angular deformity combined with joint distraction as has been reported. The leg lengthening procedure which we expect to perform for this patient in the future will be simpler due to the correction of the alignment.
Distal radio-ulnar joint subluxation following intramedullary nailing of the radius in a child

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We describe a patient with fractures of both bones of the forearm in whom flexible intramedullary nail fixation of the radius alone led to ulnar malunion and a symptomatic distal radio-ulnar joint subluxation. This was successfully treated by ulnar osteotomy.

The majority of diaphyseal fractures of the forearm in children are treated by closed reduction and plaster immobilisation. The indications for surgical intervention include instability, irreducibility and an open wound. Recent reports show that intramedullary nailing is gaining popularity over plate fixation. Distal radio-ulnar joint (DRUJ) subluxation following intramedullary nailing of the radius alone when both forearm bones are fractured is a rare complication. To our knowledge this has not been previously reported.

Case report
An 11-year-old girl presented with midshaft fractures of both bones of her right non-dominant forearm after a simple fall. On initial radiographs the radial fracture was displaced but the ulnar fracture was not (Fig. 1). The wrist and elbow joints were unaffected, either clinically or radiologically (Fig. 1b). She was treated by closed flexible intramedullary nailing (FIMN) of the radius. The ulnar fracture was treated conservatively as it was apparently undisplaced. The arm was immobilised in an above elbow plaster cast.

At six weeks the cast was removed, radiographs showed the radius to be in good alignment but there was malunion of the ulnar fracture with 12˚ of volar angulation (Fig. 2). At three months review she had wrist pain, which was worse on supination. On examination the range of pronation was 60˚ and supination, 40˚. Radiographs of the wrist showed volar subluxation of the DRUJ with the forearm in supination. The subluxation reduced spontaneously on pronation. At one-year review the wrist pain was more severe. New radiographs showed persistent DRUJ subluxation with no remodelling of the ulna. The radial nail was removed and a corrective osteotomy of the ulna was performed, closing a dorsally-based wedge and securing it with a plate. Subsequent radiographs confirmed the DRUJ to be reduced (Fig. 3). The osteotomy healed uneventfully. When reviewed one year later she was asymptomatic.

Discussion
Fractures involving the shafts of the radius and ulna are among the most common childhood injuries. The majority are treated by closed reduction and cast immobilisation. Indications for operative management of diaphyseal forearm fractures in children include failure to achieve a satisfactory reduction, persistent instability and open fractures. Recent reports on intramedullary fixation have reported excellent clinical results and promoted the advantages of this technique over plating in children.

These fractures present specific problems. If a good range of pronation and supination is to be