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CASE REPORT

Ilizarov treatment for extreme bilateral genu recurvatum in a pseudoachondroplasia patient : A case report

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The history of a 17-year-old female patient with pseudoachondroplasia, with an extreme bilateral genu recurvatum due to tibial growth disturbance, is presented. She was treated by the Ilizarov method over a nine- month period for consecutive bilateral correction of the lower legs, leading to an excellent functional result. The authors advocate the Ilizarov method as the treatment of choice for severe juxta-articular osseous deformities, where the technical limits of a standard osteotomy with internal fixation are being exceeded.

Keywords : pseudoachondroplasia ; genu recurvatum ; osteotomy ; Ilizarov.

INTRODUCTION

Pseudoachondroplasia is a syndrome due to a mutation in the cartilage oligomeric matrix protein (COMP) gene. It usually presents with short stature, ligamentous laxity and incomplete physeal involvement with partial growth arrest, leading to rotational and axial deviations, which in the sagittal plane may give rise to severe genu recurvatum. The bony deformation may be localised in the distal femur, the proximal tibia or both, and may be associated with knee joint laxity. Whereas minor deformities in the sagittal plane can be treated conservatively with bracing, greater deviations require surgery. Most often, an opening wedge osteotomy with internal fixation is recommended, as by Chen *et al* (*1*) and van Raaij *et al* (*6*). However, when the deformity reaches dramatic proportions, with a 75° recurvatum as in this patient, standard osteotomies become insufficient and a gradual correction with external fixation, as in the Ilizarov method, is more appropriate.

CASE REPORT

A 17-year-old girl, diagnosed with pseudoachondroplasia, was referred for treatment of a severe bilateral genu recurvatum. At the age of 6 months she had undergone a right sided clubfoot release with two additional Achilles tendon lengthenings when she was 4 and 8 years old.

At 4 years of age, a tibial derotation of the right leg was performed because of excessive external tibial rotation. She progressively developed a recurvatum deformity of both knees, for which a tibial osteotomy with acute correction and external

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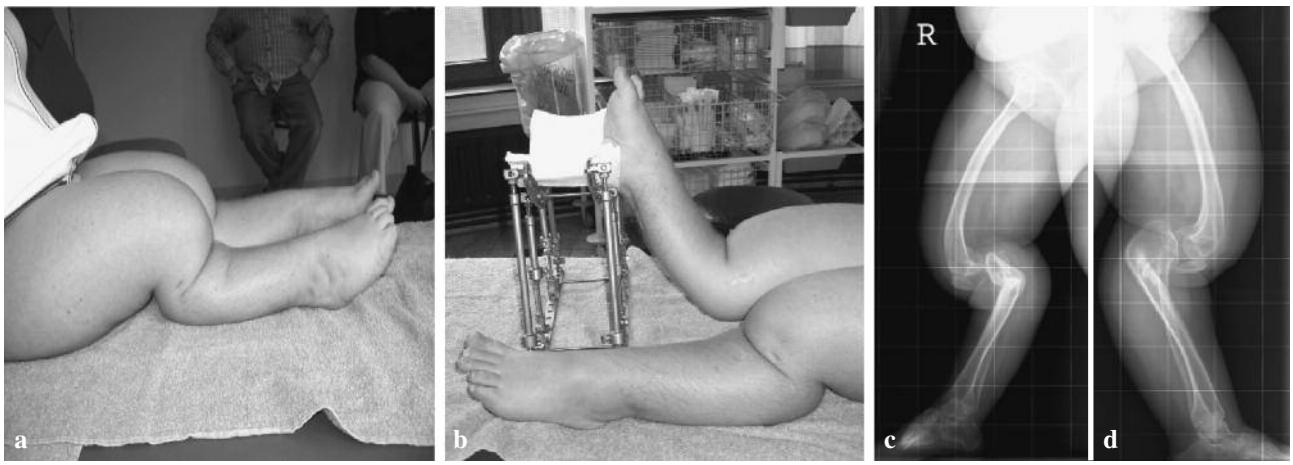

Fig. 1. — A, B. Clinical presentation of a pseudoachondroplasia patient with extreme bilateral genu recurvatum. When bending the knees, the legs appear straight (a). True recurvatum becomes apparent when the knee joint is fully extended (b). c, d. Lateral full leg radiographs of both legs showing hip dysplasia, femoral bowing, aberrant sagittal tilting of the condyles and an abnormal tibial slope. With legs apparently straightened, there is an almost vertical joint line.

fixation was carried out for the right and left knee at the age of 10 and 11 respectively. Despite her complex medical history, she could walk independently, maintained an interest in sports and was a swimmer of the Belgian paralympic team.

By the time of referral, 6 years after the last intervention she presented with a severe bilateral recurrence of the recurvatum deformity, with impaired ambulation and progressive bilateral knee pain. There was a severe functional limitation due to a bilateral hyperextension of both knees of approximately 75°. To keep an upright position, the patient had to bend the knees to the same extent, which gave an apparent straight leg (fig 1a & 1b). During ambulation this flexion was maintained throughout her gait pattern to prevent her from falling backwards. Bicycling had also become impossible because her range of motion was only 90° and full flexion only compensated for her recurvatum, simulating a straight knee. Moreover she suffered from a multidirectional knee joint laxity. Radiographs revealed a bilateral hip dysplasia, an excessive bowing of both femoral diaphyses, condylar dysplasia and a severe proximal tibial malformation with a 70° angle in the sagittal plane between the tibial plateau and the diaphysis (fig 1c & 1d).

Correction was initiated on the right side, using a standard Ilizarov frame fixed to the tibia by three 1.8 mm K wires and two 5 mm half pins per segment (fig 2). A proximal tibial – distal to the tuberosity – and distal fibular osteotomy were performed, and correction started after 5 days by anterior distraction around posterior hinges at a rate of 4×0.5 mm per day. Over a two-month period, her hyperextension disappeared and she obtained 90° of true flexion (fig 3a, 3b & 3c). The gradual adaptations were stopped and six weeks later there was enough callus to reduce the frame to a simple unilateral external fixator (fig 3d). At the same occasion, an identical Ilizarov procedure was started on the left side. Six months after initiation of the treatment, the right leg was completely consolidated and the fixator removed, whereas the circular frame on the left was reduced to a unilateral fixator. Another three months later the left leg was also completely healed and the frame was removed. During the 9 months period, she had a two months episode of peroneal nerve neurapraxia with temporary drop foot on the left side, probably provoked by difficult pin placement due to the very aberrant anatomy. At follow-up one year after removal of the last fixator, the patient was completely pain free, without any

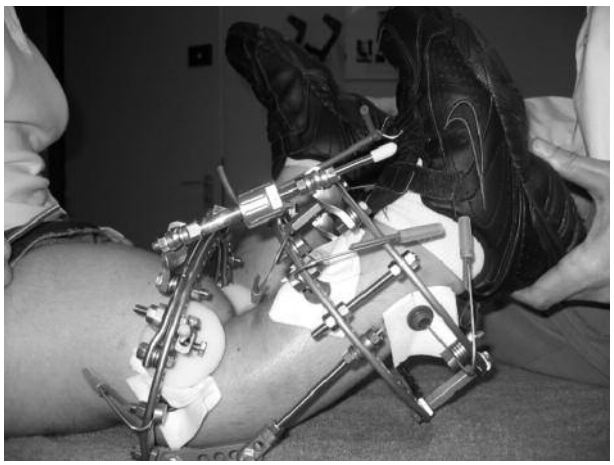

Fig. 2. — Clinical image of the right leg in the Ilizarov frame with the posterior hinges and the anterior distractor for a gradual correction in the sagittal plane.

hyperextension of the knees and a bilateral maximal flexion of 100° (fig 4). She ambulated without external aid and returned to her sports activities in the paralympics' swimming team.

DISCUSSION

Pseudoachondroplasia, due to a mutation in the cartilage oligomeric matrix protein, was first clearly delineated by Maroteaux and Lamy *(4)* in 1959 as 'pseudoachondroplastic spondyloepiphyseal dysplasia' and is phenotypically similar to multiple epiphyseal dysplasia, the latter usually presenting as a slightly milder clinical entity. It presents as a short-limb dwarfism but, unlike in achondroplasia, the head and facies appear normal. Epiphyses show irregular fragmentation and metaphyses are mushroom shaped. Brachydactyly, platyspondyly with kyphosis and scoliosis, general joint laxity and knee deformities in the coronal and sagittal plane are common and often very severe.

Genu recurvatum may cause knee pain by impingement of the fat pad, the anterior cruciate ligament or the anterior portion of the menisci, by patellofemoral problems or by overall soft tissue elongation. In this patient, angles of tilt of both tibial plateaus and femoral condyles were difficult to determine because of the general dysplasia but the recurvatum angle, as measured by Moroni *(5)*, was exceeding 45°. With the superimposed ligamentous laxity, the clinical hyperextension reached about 75°. Due to an almost vertical joint line in the sagittal plane, dysfunction and pain were probably caused by impaired load bearing conditions and the need for continuous knee flexion.

Although correction of the joint line is the primary goal – besides restriction of knee hyperextension – this was considered inappropriate owing to the extreme malformation. An accessory femoral

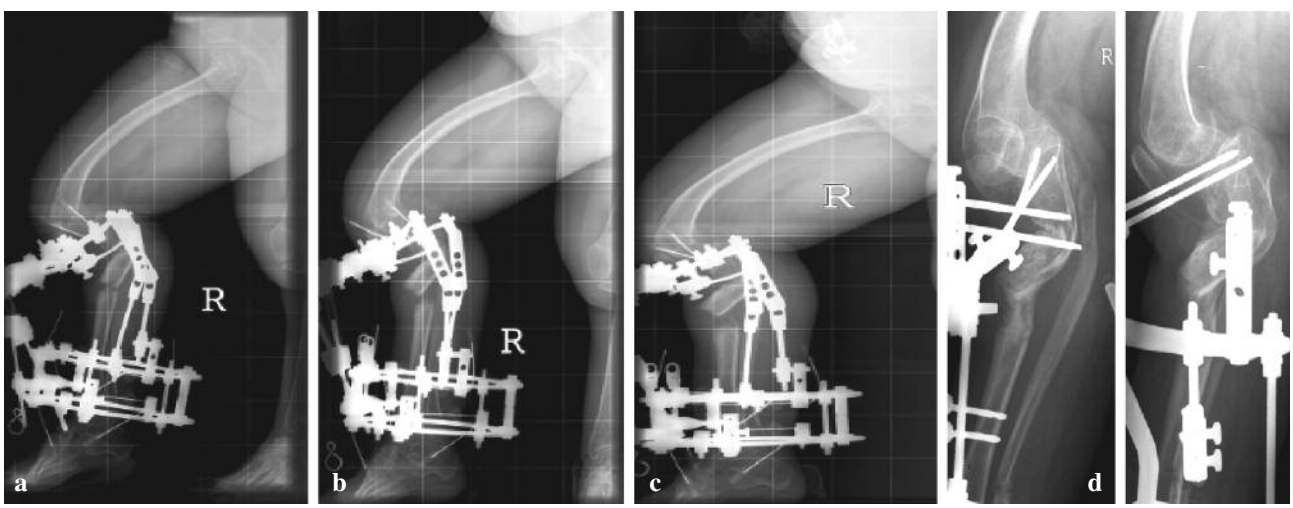

Fig. 3. — Radiographic image of the right leg at initiation of correction (a), after 2 weeks (b) and after 4 weeks of correction (c). Ten weeks later, the frame was reduced to a simple unilateral fixator (d).

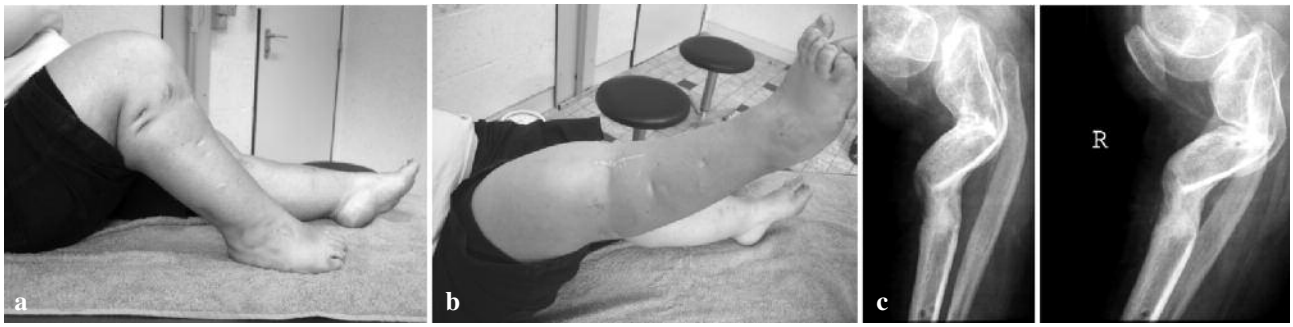

Fig. 4. — Aspect of both legs after removal of the frames. There is over 90 degrees of true flexion (a) and full extension (b) despite the tortuous aspect of the proximal tibia and the elevated tibial slope on the radiographs (c).

correction would have resulted into severe shortening of the extensor apparatus and patellofemoral hyperpression, probably resulting in anterior knee pain and impaired flexion. By altering the distal femoral shape, the quite normal appearance of the knee – with the upper leg somewhat anterior – would have been lost too, resulting in an unsightly bayonet shaped leg. Obtaining a stable femoral Ilizarov frame allowing progressive correction would have been difficult because of the relative obesity and short stature of the patient. Finally, femoral pin placement could have impaired knee flexion on a temporary or even definite basis.

Publications on proximal tibial opening wedge osteotomies concern patients with moderate recurvatum not exceeding 25°. They state that addition of 10° of tibial downslope by osteotomy proximal to the tuberosity can lead to patella infera and anterior knee pain. We therefore considered this type of surgery as inappropriate for the given case. Since neither closing wedge osteotomy nor total knee arthroplasty were valuable alternatives in this case, opening wedge callotasis was our treatment option of choice.

To avoid interference with the patellofemoral relationship we planned an osteotomy distal to the tuberosity, as previously described by Choi *et al* (*2*). This is in contrast to Moroni, who recommends to do an osteotomy proximal to the tubercle but with detachment and reinsertion of the patellar tendon. Choi *et al* published a technique of preoperative planning with plotting of the radiographs on the unaffected contralateral side. Because of bilateral involvement, this specific planning could not be carried out, but hinges were placed posteriorly at the osteotomy level. Anterior distraction rate was 4 times 0.5 mm daily, which is slightly higher than it would be based on the triangulation distraction method by Herzenberg and Waanders (*3*). Nevertheless, the callotasis went uneventfully with good bone formation.

After bilateral correction, hyperextension was corrected and maximal flexion was achieved. There was no residual malalignment of the lower limbs in the frontal or coronal plane. The gait pattern was markedly improved and the patient did not suffer from any discomfort around the knee.

CONCLUSION

The Ilizarov treatment of this 17-year-old patient with bilateral combined soft tissue and osseous recurvatum, with a clinical hyperextension of 75° enabled to fully correct the hyperextension deformity in a nine-month period. Remaining knee laxity, residual radiologic deformity and severe dysplasia of the femoral condyles do not prohibit a pain free status and participation in the Belgian paralympics swimming team. Despite the lack of evidence based data on treatment options for correction of extreme genu recurvatum, we advocate the Ilizarov method as the treatment method of choice in severe axial deformity.

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