CASE REPORT

The management of leg-length discrepancy in Ollier’s disease with a fully implantable lengthening nail

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Ollier’s disease is characterised by severe deformity of the extremities and retarded growth because of multiple enchondromas. For correction of deformity, the Ilizarov method has been used although it has many complications. A 17-year-old boy with Ollier’s disease had a limb-length discrepancy of 17.4 cm, with a valgus deformity of the right knee and recurvatum of the femur of 23°. He had undergone three unsuccessful attempts to correct the deformities by using external fixators. We used a fully implantable, motorised, lengthening and correction nail (Fitbone) to achieve full correction of all the deformities without complications. We decided to carry out the procedure in three stages. First, we lengthened the femur by 3.6 cm and the tibia by 4 cm. We then exchanged the femoral nail for a longer implant and achieved a further 6 cm of length. This reduced the shortening to 3.8 cm. When the boy has finished secondary school we will adjust the remaining discrepancy.

Ollier’s disease, multiple enchondromatosis, is a rare skeletal disorder (1:100 000) characterised by circumscripted masses of cartilage arranged in a linear fashion. The metaphyses and epiphyses of the long bones and the cartilage of the joints of the arms and legs are mostly affected. Limb deformity and retarded limb growth are seen and pathological fractures occur. Patients with Ollier’s disease must be monitored carefully as between 10% and 30% of enchondromas may progress to chondrosarcoma.

External fixators are commonly used to lengthen the femur or the tibia and to correct severe deformities. The Ilizarov technique with circular fixators allows complex and extended lengthening but has a high incidence of complications. In Ollier’s disease the bone may be relatively soft and the transosseous pins may cut out, necessitating removal of the external fixators.

Baumgart, Betz and Schweiberer have developed an intramedullary distraction nail (Fitbone; Wittenstein, Igersheim, Germany) with an enclosed motor drive which is available in two versions. The Fitbone Slide Active Actuator nail has a slide hole and an external diameter of 13 mm and is available in lengths between 260 and 520 mm. Lengthening of up to 85 mm and bone transport of up to 200 mm may be achieved. The Slide Active Actuator system is locked with three distally-placed screws. Daily distraction may be undertaken at intervals or even at night. The surrounding soft tissue is stretched simultaneously. After achieving the desired length the bone is stabilised by the nail while consolidation of the new bone takes place. Load bearing of up to 20 kg is allowed during this period.

Case report

A 17-year-old boy with Ollier’s disease of the right leg, diagnosed in 1991, had shortening with axial and rotational deformity of the right
Fig. 1
Standing radiographs showing Ollier’s disease of the entire right leg with 17.4 cm of shortening, valgus deformity and 23° recurvatum of the femoral shaft.

Fig. 2
Diagrams showing the planning of the correction of the deformity and of lengthening in the first stage.

Fig. 3
Radiographs of simultaneous distraction of the femur and tibia during the first stage.
Fig. 4

Standing radiographs showing correction of deformity of the femur and tibia and lengthening of 7.8 cm after the first stage.

Fig. 5a, Fig. 5b, Fig. 5c, Fig. 5d

Radiographs showing distraction of the femur during the second stage of treatment a) one week post-operatively, b) two weeks post-operatively, c) five weeks post-operatively and d) eight weeks post-operatively.
femur and tibia. At the age of four years, he had correction of a varus deformity of the right knee followed by lengthening procedures in 1992, 1995 and 2000. The operation in 1995 attempted elongation of the right femur with the Ilizarov technique. He had gained 3 cm when the external fixator loosened and the lengthening had to be stopped. During the third operation in 2000, a further attempt to lengthen the femur with an external fixator was undertaken but it also had to be stopped because of loosening.

When he was first seen at our centre long leg radiographs were taken with the limbs weight-bearing and the short leg equalised by standing on appropriate blocks. The centre of the beam was focused on the patient’s knees with the patella orientated forward. The difference in leg length and the angles of the deformities were measured. A CT scan assessed rotation at the hip and ankle joints. The total shortening was 17.4 cm. Shortening of the femur was 13.4 cm and associated with a recurvatum deformity of the femoral shaft of 23°. Shortening of the right tibia was 4 cm. The mechanical axis lay 13 mm laterally at the level of the right knee joint and 5 mm medially on the left (Fig. 1). The valgus deformity on the right was mainly explained by a medial proximal tibia angle of 95°. There was a greater angle of anteversion of the right femoral neck and the right ankle joint compared with the left side. Scintigraphy was performed to rule out neoplastic change.

Treatment was planned in three stages. First, the genu valgum, recurvatum and torsional deformities of the femur were treated. The operation was carefully planned with technical drawings to treat every deformity (Fig. 2) taking into account the anatomical axis and length discrepancy. In the tibia lengthening and varus displacement were undertaken and varisation in order to obtain an anatomical angle of 87° at the knee. In the femur a Fitbone Slide Active Actuator nail was used and the bone lengthened by 3.6 cm as allowed by the tension of the soft tissues. The tibia was lengthened by 4 cm and its axis corrected using a Fitbone Telescope Active Actuator nail with a diameter of 12 mm proximally and 10 mm distally. Lengthening of both bones of 1 mm each day was achieved without complications (Fig. 3). The patient was mobile on crutches and had physiotherapy to the knee and ankle joints. He was followed up weekly and by three months was able to fully bear weight (Fig. 4).

One year later, the second stage was carried out with further lengthening of the femur. A further osteotomy was performed and the Fitbone Slide Active Actuator distraction

Fig. 6a  Fig. 6b  Fig. 6c  Fig. 6d

Photographs of the clinical appearance before (a,b) and after the second stage (c,d).
nail was changed for a longer version. After 60 days of distraction another 6 cm were gained (Fig. 5). No problems arose and full joint movement was maintained (Fig. 6).

For the final stage, the remaining 3.8 cm of length will be regained in the femur when the new bone has fully consolidated and the patient has finished his secondary education.

One of the authors has contributed in the development of the fully implantable distraction device with the Wittenstein Company.

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References