Combined technique for the correction of lower-limb deformities resulting from metabolic bone disease

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We present the results of the surgical correction of lower-limb deformities caused by metabolic bone disease. Our series consisted of 17 patients with a diagnosis of hypophosphataemic rickets and two with renal osteodystrophy; their mean age was 25.6 years (14 to 57). In all, 43 lower-limb segments (27 femora and 16 tibiae) were osteotomised and the deformity corrected using a monolateral external fixator. The segment was then stabilised with locked intramedullary nailing. In addition, six femora in three patients were subsequently lengthened by distraction osteogenesis. The mean follow-up was 60 months (18 to 120). The frontal alignment parameters (the mechanical axis deviation, the lateral distal femoral angle and the medial proximal tibial angle) and the sagittal alignment parameters (the posterior distal femoral angle and the posterior proximal tibial angle) improved post-operatively. The external fixator was removed either at the end of surgery or at the end of the lengthening period, allowing for early mobilisation and weight-bearing. We encountered five problems and four obstacles in the programme of treatment.

The use of intramedullary nails prevented recurrence of deformity and refracture.

Patients and Methods

Informed consent for participation in a clinical study was obtained from all patients and the Institutional Review Board approved the study. Between 2001 and 2008, 43 lower-limb segments involving 27 femora and 16 tibiae in 17 patients (12 female, five male) with a mean age of 25.6 years (14 to 57) underwent correction of deformity through multilevel osteotomies with the assistance of a monolateral external fixator, followed by stabilisation with an intramedullary locked nail. No patients were lost to follow-up. Aetiological factors were hypophosphataemic rickets in 15 patients and renal osteodystrophy in two. All patients were treated in the endocrinology department, and were operated on only after normal serum levels of calcium (2.20 mmol/l to 2.60 mmol/l) and phosphate (0.81 mmol/l to 1.45 mmol/l) were achieved. Endocrinological monitoring continued post-operatively.

The pre-operative radiological assessment included anteroposterior (AP) and lateral orthogonal radiographs. The extent of the deformity was evaluated and the amount of mechanical axis deviation and the lateral distal femoral angle, the medial proximal tibial angle, the posterior distal femoral angle, the posterior proximal tibial angle, and the centre of rotation of angulation were measured and...
recorded. If there was more than one centre of rotation of angulation, as in multi-apical deformity, then surgery was planned accordingly. Pre-operative planning also included calculation and ordering of the custom-made holes in the intramedullary nail to be used as needed. Deformities were first corrected with the use of a monolateral fixator (Orthofix LRS, Verona, Italy) and then stabilised with an intramedullary nail (Ortopro Retrograde Femoral and Tibial Nail 4G, Istanbul, Turkey) under the same anaesthetic.

The mean shortening was 7.2 cm (6 to 8). The deformities consisted of genu varum in nine patients, genu valgum in six, and windswept deformity (genu varum on one side and genu valgum on the other) in two. In varus knees, the mean mechanical axis deviation was 74 mm (11 to 120); the mean lateral distal femoral angle was 110° (95° to 135°), and the mean medial proximal tibial angle was 78° (69° to 84°). In valgus knees the mean mechanical axis deviation was -53 mm (-112 to -20), the mean lateral distal femoral angle was 76° (56° to 83°), and the mean medial proximal tibial angle was 94° (93° to 110°). The mean angular deformity in either the frontal or the sagittal plane was 28.7° (10° to 50°). In three patients with short stature (six femora) lengthening was performed after acute deformity correction. After the desired lengthening was achieved, the intramedullary nail was locked statically at its opposite end in a second session, and the external fixator was removed. Full weight-bearing was allowed once three or four cortices were consolidated on AP and lateral views.

The criteria used to evaluate bone healing were the bone healing index (days/cm), the duration of external fixation (months) and the external fixation index (days/cm). The clinical results were assessed via the clinical and functional scoring system described by Paley et al. Complications related to treatment were evaluated using Paley’s classification for complications.

**Surgical technique.** The patient was placed supine on a radiolucent table and the lower limb was checked for visualisation on the radiography table from the hip joint to the ankle joint on both AP and lateral views. In patients with genu valgum, open release of the peroneal nerve was performed prior to correction of the deformity to avoid traction injury. Two Schanz screws were placed into the proximal and distal segments above and below the osteotomy for adequate stability, perpendicular to the anatomical axis, taking care to leave enough space for the inserted intramedullary nail without making contact with the screws. The level of osteotomy was chosen and performed percutaneously either using the multiple drill hole technique for the femur or a Gigli saw for the tibia. Upon completion of the osteotomies, the deformity was corrected using a monolateral external fixator. The correction was confirmed by obtaining AP and lateral long radiographs. At this step, the electrosurgery cable was also useful for checking the alignment under fluoroscopic control. If the desired correction was not achieved, it was adjusted and confirmed with new radiographs. Once the surgeon was satisfied with the correction, the intramedullary nailing was performed through a 2 cm transverse incision over the ligamentum patellae. The standard ligament split approach was used to open the tibial or femoral canals under fluoroscopic control. Over a guide wire, the medullary canal was over-reamed 1 mm larger than the diameter of the nail to allow sliding of the nail during distraction. Poller interference blocking screws were placed before or after nail insertion to reduce the larger diameter of the medullary canal at the metaphyseal level to prevent pendular movement of the nail.
The nail was inserted slowly to reduce the risk of fat embolism, and locked both proximally and distally if no lengthening was planned. Otherwise, the nail was locked only at the insertion site, and the opposite end of the nail was locked on completion of the lengthening period in another session. The positions of the nail and the Schanz screws and the deformity correction were once again confirmed under fluoroscopic control, and then the fixator was removed. In lengthening cases, the distraction test was performed with the external fixator: acute distraction of 2 mm was performed to confirm the completeness of the osteotomy under fluoroscopic control. The distraction was then re-compressed.

Isometric exercises were started after surgery in the recovery room and partial weight-bearing was allowed with crutches as tolerated. The lengthening was started on the seventh post-operative day, at a rate of $4 \times 0.25$ mm per day. The patients returned for follow-up every two weeks during lengthening, and every month during the consolidation phase. Full weight-bearing was allowed only after achieving the consolidation of three of four cortices on AP and lateral radiography. An example of the deformities and treatment is shown in Figures 1 and 2.

**Results**

The mean follow-up was 60 months (18 to 120) and the mean lengthening was 7.2 cm (6 to 8). In addition, the mean mechanical axis deviation after operation was 7 mm (0 to 29) in varus knees and -6 mm in valgus knees (-20 to +7) (Fig. 3).

The mean post-operative lateral distal femoral angle was $87^\circ$ ($80^\circ$ to $92^\circ$) in valgus knees, and $90^\circ$ ($87^\circ$ to $109^\circ$) in varus knees. The mean post-operative medial proximal tibial angle was $88^\circ$ ($86^\circ$ to $90^\circ$) in varus knees and $87^\circ$ ($85^\circ$ to $91^\circ$) in valgus knees. The sagittal parameters, the posterior distal femoral angle and posterior proximal tibial angle, improved by $16^\circ$ ($5^\circ$ to $32^\circ$) and $11.8^\circ$ ($1^\circ$ to $31^\circ$), respectively. The mean external fixation time was 78.9 days (67 to 87) and the mean external fixation index was 14.34 days/cm (7.8 to 25.1). The mean bone healing index was 38.32 days/cm (16.7 to 48.2). The mean time to achieve consolidation of the osteotomies without lengthening was 3.1 months (two to five).

The clinical results according to Paley’s scoring system were very good in eight patients and good in nine. In addition, the functional results were very good in seven patients and good in ten. Applying Paley’s classification of complications, where ‘problems’ could be treated without surgical intervention, ‘obstacles’ required surgical intervention, and ‘sequelae’ persisted after the treatment, there was a total of nine complications. These included five pin-track infections (five problems) in two patients, all of which responded to oral antibiotics and local wound care, and screw cut-out in two patients and loss of correction in two tibiae (four obstacles). There were no sequelae. The losses of tibial correction were encountered in one patient who received bilateral femoral and tibial correction under a single anaesthetic. Three days after the initial surgery we revised the tibiae, and upon achievement of the desired correction we used Poller blocking screws to protect it.

**Discussion**

The clinical manifestation of hypophosphataemic rickets is commonly genu varum or valgum, or in rare cases a windswept deformity.3-10 Our series had the clinical and laboratory findings consistent with hypophosphataemic rickets in 16 patients. Prior to any surgical intervention, all hypophosphataemic patients require appropriate metabolic management, and surgery should not be performed until the laboratory parameters have returned to normal.3-6,10 All patients in this study received phosphate and calcitriol treatment for at least six months before their operation.

Untreated patients with hypophosphataemic rickets exhibit severe deformities which in the long term can result in arthritis.14-16 Sharma et al15 reported that in the knee, varus malalignment accelerates medial compartment arthritis and valgus malalignment accelerates lateral compartment arthritis. In the absence of any control group with the same diagnosis, we cannot conclude that our corrective realignment has prevented the subsequent development of arthritis in the knee.

The pathological change caused by hypophosphataemic rickets occurs very close to the growth plate, leading to marked deformity and severe malalignment. Thus, pre-operative assessment usually reveals multiple centres of rotation of angulation, so several osteotomies are required to correct the deformity. Many methods of treatment have been described.5-10,17-20 The most common involve acute or gradual correction, using either Ilizarov or monolateral external fixators, which can secure accurate correction of...
the deformity and elimination of the limb-length discrepancy. However, these methods are uncomfortable for the patient, especially when both legs are involved. There are further disadvantages, such as the need for daily adjustments, weekly follow-ups, a high rate of pin-track infection, and a long duration of external fixation.3-6,9,10,17-21

Sofield and Millar22 first described the method of intramedullary nailing following multiple osteotomies for the treatment of bone deformities. Later, the same method was used to treat patients with hypophosphataemic rickets using different intramedullary fixation techniques.6,7,9

We used the fixator-assisted nailing technique described by Paley, Herzenberg and Bor23 and Paley and Herzenberg,24 with osteotomies created at the centre(s) of the deformity, followed by correction using a monolateral fixator and stabilisation with locked intramedullary nailing. Removal of the external fixator at the end of the operation reduces post-operative discomfort and avoids pin-track infections, except where patients require additional post-operative lengthening over the nails.1,2,25,26 However, this technique does not allow re-correction or adjustment post-operatively, therefore careful analysis of the deformity and pre-operative planning are crucial.1,2,12,23,24 In addition, the surgeon must be familiar with both external fixation and intramedullary nailing techniques, and the combined technique is technically demanding.

Paley et al12 reported an external fixation index of 15 days/cm and a bone healing index of 42 days/cm in their series of femoral lengthening over a nail. In our series, three patients who underwent lengthening procedures exhibited a mean bone healing index of 38.32 days/cm. These results are very similar to those of Paley et al, and superior to those obtained using circular external fixation alone to correct the deformity and control lengthening, or even correcting the deformity in isolation.3,5,10 However, in one of the three patients with lengthening, the external fixation index was longer than usual (25.1 days/cm) owing to poor regenerate formation, and distraction had to be reduced to 0.75 mm/day.

Among the problems encountered in the treatment of hypophosphataemic rickets is subsequent loss of correction or recurrence of the deformity.4,6 However, Choi et al3 reported a recurrence in only one of 14 corrective osteotomies. Song et al9 reported 20 patients with hypophosphataemic rickets who underwent corrective osteotomies of 55 segments with six different techniques for osteosynthesis. Recurrence or re-fracture occurred most commonly in the patients who were treated by external fixation only (48%), and least often following acute correction using external fixation, and then stabilisation using an intramedullary nail (5%). We attribute the absence of these complications in our series to the stability in three orthogonal planes provided by the intramedullary locking nails and the blocking screws.
The surgical technique is demanding. In four cases the mechanical axis deviation was outside the normal limits at the end of the treatment (Fig. 3). Three of these patients were treated during our learning curve.

It is recommended that after the corrective osteotomy at the metaphyseal level another proximal osteotomy should be made to permit distraction if lengthening is necessary.1,2 This is especially important when Poller blocking screws are used to narrow the medullary canal to prevent the nail from sliding. In three of our patients who underwent femoral lengthening following correction of the deformity, multiple osteotomies had been required because of the multi-apical deformity. In two patients the lengthening was through the multiple osteotomies, but in the remaining patient through only one osteotomy bilaterally, at a proximal site on one side and through the distal osteotomy contralaterally.

Performing all the surgery at a single operation as we did could be considered too onerous for both the patient and the surgeon. A correction of all four lower-limb segments takes approximately six hours in our experience. However, all of our patients tolerated this amount of surgical intervention well, probably because they were young, with a mean age of 25.6 years (14 to 57). This programme shortened the length of treatment and reduced the cost.

We recognise the shortcomings of this study. There is no control group of patients treated using another technique. Also, the follow-up time was short to moderate, so it is not possible to make a long-term evaluation. However, at a mean follow-up of five years we saw no recurrence of deformity. We believe the technique we describe to be safe and effective in these patients with severe lower-limb deformity, and we recommend it.

Supplementary material
A table showing the pre- and post-operative deformity assessment angles is available with the electronic version of this article on our website at www.jbjs.org.uk

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References

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