Limb lengthening and correction of
deformity in the lower limbs of children with
osteogenesis imperfecta

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We performed limb lengthening and correction of deformity of nine long bones of the lower limb in six children (mean age, 14.7 years) with osteogenesis imperfecta (OI). All had femoral lengthening and three also had ipsilateral tibial lengthening. Angular deformities were corrected simultaneously. Five limb segments were treated using a monolateral external fixator and four with the Ilizarov frame. In three children, lengthening was done over previously inserted femoral intramedullary rods.

The mean lengthening achieved was 6.26 cm (mean healing index, 33.25 days/cm). Significant complications included one deep infection, one fracture of the femur and one anterior angulation deformity of the tibia. The abnormal bone of OI tolerated the external fixators throughout the period of lengthening without any episodes of migration of wires or pins through the soft bone. The regenerate bone formed within the time which is normally expected in limb-lengthening procedures performed for other conditions.

We conclude that despite the abnormal bone characteristics, distraction osteogenesis to correct limb-length discrepancy and angular deformity can be performed safely in children with OI.

Traditionally, multiple osteotomies and intramedullary nailing are used to treat angular deformities of the long bones in children with osteogenesis imperfecta (OI) as described by Sofield and Millar. Although this method helps to reduce the frequency of further fractures, limb-length discrepancy cannot be treated in this way. An increased incidence of complications has been reported when lengthening has been performed on limbs with underlying bone disorders, such as dysplasias and metabolic bone diseases. There have been few studies of limb lengthening in OI, probably because of the concern that bone of small diameter containing abnormal collagen may not tolerate external fixation for long enough to allow adequate formation of regenerate. Ring et al described the correction of deformities of the lower limb in adults with OI and suggested that these procedures should not be undertaken on patients who continue to suffer frequent fractures. We have previously reported limb lengthening in children with OI using monolateral fixators.

We now present the results of further experience using monolateral as well as Ilizarov circular frames to lengthen and correct deformities of the lower limbs in children with OI.

Patients and Methods

Between February 1988 and December 2000 we treated nine long bones of the lower limb in six children, three boys and three girls with OI for limb-length discrepancies and deformities using external fixators. There were four children (cases 1 to 4) with type-I and two (cases 5 and 6) with type-IV OI as classified by Sil- lence. Their mean age was 14.7 years (14 to 16). All had femoral shortening with a mean of 5.1 cm (1.5 to 8). Three (cases 1, 5 and 6) also had ipsilateral tibial shortening with a mean of 4.7 cm (1.5 to 8.5). Angular deformities included varus in three femora (cases 3, 5 and 6), valgus in one femur (case 4), valgus in one tibia (case 6) and valgus with procurvatum in one tibia (case 5). Four children (cases 1 to 4) were treated using the monolateral Orthofix limb reconstruction system (Orthofix SRL, Verona, Italy) and two (cases 5 and 6) using the Ilizarov external fixator system (Smith & Nephew Health Care Ltd, Cambridge, UK).

In patients in whom the monolateral fixator was used, a metaphyseal corticotomy as described by DeBastiani et al was performed. Lengthening was started on the seventh postoperative day until the desired length was achieved and the fixator was locked off during the consolidation phase. The fixator was dyna-
<table>
<thead>
<tr>
<th>Case</th>
<th>Gender</th>
<th>Age</th>
<th>Affected family members</th>
<th>Previous fractures treated</th>
<th>Limb segments treated</th>
<th>Pre-treatment shortening (cm)</th>
<th>Pre-treatment deformity</th>
<th>Fixator used and reconstruction technique</th>
<th>Length and deformity correction achieved (cm)</th>
<th>Bone healing index (days/cm)</th>
<th>Complications and their treatment</th>
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<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>13</td>
<td>Father, twin brother, elder brother, two aunts, paternal grandfather</td>
<td>&gt;70 #s of both femora and tibiae</td>
<td>Two staged – right femur followed by right tibia</td>
<td>Right femur 8, right tibia 8.5</td>
<td>None at the time of fixator application</td>
<td>Uses walking frame and caliper with shoe raise on right side</td>
<td>Orthofix monolateral fixator, De Bastiani monofocal callotasis</td>
<td>Femur 8, Tibia 8.5</td>
<td>Femur 24.75/Tibia 17.1</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>16</td>
<td>This boy is the elder brother of case 1</td>
<td>Multiple #s of both femora and right tibia</td>
<td>Left femur</td>
<td>5</td>
<td>None at the time of fixator application</td>
<td>Walking with short limb gait in spite of 2 cm shoe raise</td>
<td>Orthofix monolateral fixator, De Bastiani monofocal callotasis</td>
<td>5</td>
<td>37.8</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>16</td>
<td>None</td>
<td>Multiple #s of both femora. Last # at 14 years of age</td>
<td>Left femur</td>
<td>7</td>
<td>Varus of proximal femur</td>
<td>Occasional use of crutches, gets pain in left foot while walking due to holding foot in equinus</td>
<td>Orthofix monolateral fixator, De Bastiani monofocal callotasis</td>
<td>8</td>
<td>48.25</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>14</td>
<td>None</td>
<td>Multiple #s of both femora and tibiae</td>
<td>Left femur</td>
<td>4</td>
<td>9˚ valgus of distal femur</td>
<td>Walking with shoe raise and caliper on left side to control knee varus. Mobilising in a wheelchair</td>
<td>Orthofix monolateral fixator, De Bastiani monofocal callotasis</td>
<td>5.5</td>
<td>57.27</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>14</td>
<td>Brother and maternal grandmother</td>
<td>Multiple #s of left femur, tibia, fibula, one # each of right femur, humerus and 4th lumbar vertebra. Last # was that of right femur, occurred 4 weeks after lengthening of left leg</td>
<td>Simultaneous lengthening of left femur and tibia</td>
<td>Total shortening of left lower limb 3. Left femur 1.5, left tibia 1.5</td>
<td>20˚ varus bowing of left femur and varus with procurvatum of left tibia resulting in restriction of ankle dorsiflexion</td>
<td>Ilizarov method. Bifocal callotasis of left femur and tibia</td>
<td>Ilizarov method. Bifocal callotasis of left femur and tibia</td>
<td>3.8</td>
<td>26.1</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>15</td>
<td>None</td>
<td>Multiple #s of left proximal femur. one # of right femur</td>
<td>Simultaneous lengthening of left femur and tibia</td>
<td>Total shortening of left lower limb 8.5, most of it in left femur with fixed pelvic obliquity and scoliosis</td>
<td>Varus of femur and bifocal valgus of left tibia</td>
<td>Walking with crutches and wheelchair for outdoor mobilisation</td>
<td>Ilizarov method. Monofocal (distal femoral) callotasis of left femur and bifocal callotasis of left tibia</td>
<td>Lengthened by 5. Full correction of true shortening could not be done due to fixed pelvic obliquity and scoliosis. Femoral varus and tibial valgus corrected</td>
<td>21.5</td>
</tr>
</tbody>
</table>
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...mised or a plaster cast was applied when there was evidence of the formation of regenerate of good quality. Although weight-bearing was discouraged during the lengthening phase, partial weight-bearing was allowed during the neutralisation and dynamisation phases. In two children in whom the Ilizarov technique was used, monofocal or bifocal metaphyseal percutaneous corticotomies were performed. Lengthening began on the fourth post-operative day and the fixator was locked off until good formation of regenerate allowed removal of the fixator and application of a cast brace. Weight-bearing was encouraged throughout the period of treatment. Specific modifications were made on an individual basis. Regular physiotherapy was given to maintain active and passive movements and to improve muscle power. Two tibial lengthenings (cases 5 and 6) were performed simultaneously with ipsilateral femoral lengthening using the Ilizarov fixator and one tibial lengthening (case 1) was performed after ipsilateral femoral lengthening using a monolateral fixator.

One child (case 6) was undergoing treatment with intravenous bisphosphonate by the metabolic bone unit and hence received two cycles of pamidronate before lengthening.

Results
Details of the patients and the results of limb reconstruction are given in Table I. The mean follow-up was two years and one month (ten months to four years and one month). The mean lengthening achieved was 6.26 cm (3.8 to 8.5 cm). Limb-length discrepancies were corrected to within 1.5 cm of the length of the contralateral limb in five children (cases 1 to 5) (Fig. 1). In one child (case 6) with fixed pelvic obliquity and scoliosis, although the true leg-length discrepancy was 8.5 cm, the limb was lengthened by 5 cm in order to achieve correction of the functional leg-length discrepancy, resulting in a reduction of pelvic tilt and improvement of spinal and shoulder balance (Fig. 2). The bone healing index was calculated by dividing the number of days from the time of application of the fixator to its removal by the total number of centimetres of lengthening. The mean bone healing index was 33.25 days/cm of lengthening (17.1 to 57.27). There were no fractures through the new bone regenerate. The external fixators remained stable throughout the procedure in all cases with no evidence of loosening of pins or migration through the soft bone. All six patients regained the pretreatment range of movement of the hip, knee and ankle and one (case 5), who had no dorsiflexion of the ankle because of a procurvatum deformity of the tibia, gained 10° of dorsiflexion of the ankle. All patients were independently mobile without orthoses after the treatment and expressed satisfaction with function and body image.

Complications were classified according to Saleh and Scott as class I (no long-term functional or anatomical significance), class II (correction requires anaesthesia or operation but has no long-term significance), class III (significant functional or anatomical complication which improves spontaneously or is correctable by surgery) and class IV (irremediable by conventional treatment).

In class I, there were four cases of superficial pin-site infections (cases 1, 2, 3 and 5) which required oral antibiotics and one episode of pain and paraesthesia in the distribution of the lateral popliteal nerve (case 6) on the 12th post-operative day. This subsided in two days after which lengthening was continued.

In class II, there were three complications. Half-pin breakage in one child (case 6) required another procedure to insert additional half-pins and wires. Later, a stress fracture appeared through the pin track of the broken half-pin; it healed well. During the course of lengthening, the distal end of the extensible femoral intramedullary rod cut out of the site of the osteotomy, but the lengthening was continued and the regenerate consolidated in spite of the rod protruding into soft tissues. In one child (case 1), the proximal end of the extensible femoral rod migrated into the buttock during lengthening but it was easily repositioned.

In class III, there were also three complications. One child (case 4), in whom the intramedullary rod had been removed before the application of the fixator, sustained a fracture through the midshaft of the femur away from the site of lengthening, four months after removal of the fixator. This was successfully treated by the reinsertion of a Sheffield intramedullary rod. One child (case 1) had lengthening of the ipsilateral tibia after removal of the tibial rod but following removal of the fixator, developed 30° of anterior angulation of the proximal tibia. This was initially treated by closed osteoclasis and application of a plaster-of-Paris cast. Six months later, an extensible rod was inserted to give increased stability. In one child (case 2), infection of the distal pin track was initially controlled by antibiotics but a discharging sinus appeared five months after removal of the fixator. The discharge persisted in spite of excision of a ring sequestrum at the pin site and the rod had to be removed seven months later. There were no cases of class-IV complications.

Of the six femoral lengthenings, five (cases 1 to 4 and 6) had extensible femoral intramedullary rods inserted previously. In three of these (cases 1, 2 and 6), the rods were left in situ during the course of femoral lengthening since there was sufficient metaphyseal bone at each end and they had sufficient extending capacity to remain functional throughout the procedure. In the other two (cases 3 and 4) the rods had to be removed before the application of external fixators because of the presence of considerable bony deformities. Of the three femora in which the lengthening was done over the rod, one rod (case 1) migrated proximally into the buttock and had to be repositioned, one (case 2) had to be removed because of infection; and one (case 6) cut out of the distal femur during lengthening but no intervention was required since the bone remained stable. Of the two femora from which the rods had to be removed before lengthening, one (case 4) sustained a fracture of the midshaft four...
Case 5. A 14-year-old boy with type-IV OI; radiography showing a) varus bowing and 1.5 cm shortening of the left femur with valgus and shortening of the left tibia of 1.5 cm before reconstruction; b) application of an Ilizarov fixator and bifocal corticotomies of the left femur and tibia; c) good formation of regenerate in the left femur and (d) the left tibia on the 56th post-operative day; and (e) complete correction of limb-length discrepancy and deformity after treatment.
months after removal of the fixator and the rod had to be reinserted. In the other (case 3) prophylactic reinsertion of an extensible rod was performed after removal of the fixator in order to give the bone increased stability. One child (case 5) had no intramedullary rod in the femur at the time of treatment and did not undergo insertion of a rod after the completion of lengthening.

All three tibial lengthenings were done without rods in situ. In one child (case 1) a previously inserted extensible tibial rod had to be removed before the lengthening since the metaphyseal ends of the tibia were not large enough to accommodate fixator pins around it. After the completion of lengthening and removal of the monolateral external fixator, the tibia developed an anterior angulation which was treated by osteoclasis and subsequent reinsertion of an extensible intramedullary rod. Two other tibiae (cases 5 and 6) had no rods in them at the time of treatment and did not undergo insertion of a rod after the completion of lengthening.

Table II gives a summary of the intramedullary rods used in all the children.

**Discussion**

OI is a heterogeneous group of conditions resulting from one of several genetic defects, which produce either a reduction in the production of normal type-I collagen or the synthesis of abnormal collagen as a result of mutations in the type-I collagen genes. Increased fragility of bones results in an increased tendency to recurrent fractures, eventually leading to deformities of the long bones, often with limb-length discrepancy. In 1959, Sofield and Millar popularised the operation of multiple osteotomies and fixation by intramedullary rods to correct the deformity and reduce the incidence of fractures. The introduction of the extensible intramedullary device in 1963 by Bailey and Dubow reduced the number of further operations when compared with using rods of a fixed length and the use of the Sheffield rod system has further reduced the number of complications. Several authors have found that the use of extensible intramedullary rods in OI reduced the incidence of fractures, prevented deformities and improved walking capability whereas others have found that rodding has no major influence on motor development and walking status.

When associated with angular and rotational deformities, leg-length discrepancy can be an important contributing factor to the child's difficulty in walking and may also increase the mechanical forces acting on fragile bones, thereby increasing the risk of further fracture. Most potentially ambulatory patients with OI have a leg-length discrepancy. The latter may be caused by either malunited diaphyseal fractures or traumatic epiphyseal injury causing
growth arrest\textsuperscript{20} or epiphyseal tethering caused by the intramedullary rods. In children with severe OI, the growth rate is greatly reduced before the age of six or seven years and growth almost stops thereafter.\textsuperscript{21} Therefore, surgical correction by limb-lengthening is preferable to the restriction of growth potential by epiphysiodesis.\textsuperscript{4}

Although limb-lengthening procedures are increasingly performed for correction of leg-length discrepancies, there is little information in the English literature about the efficacy and safety of such lengthening through abnormal bone. Naudie et al\textsuperscript{2} compared the results in patients who had lengthening for a discrepancy secondary to an underlying bone disorder with those in patients who had lengthening through histologically normal bone. They reported a higher rate of complications in the former. In their group of eight children with a discrepancy due to an underlying bone disorder, there was only one child with OI. Ring et al\textsuperscript{3} reported the results of leg lengthening using the Ilizarov technique in six adults with Sillence type-I OI, and found that the newly regenerated bone has a radiographic appearance which was similar to that of the adjacent host bone and could withstand normal functional loading. However, they suggested that the fixator may need to be left in place for a longer period until the regenerated bone clearly appears radiographically mature and that the limb should be protected after the frame had been removed by an external support such as cast brace. The mean age of their patients at the time of leg lengthening was 31 years and they considered these procedures to be contraindicated in patients who have a greater degree of fragility of bones, as judged by the age at the time of first fracture and the total number, frequency, distribution and circumstances of the fractures, and in those who continue to sustain fractures. However, our patients belonged to Sillence type-I and type-IV OI groups and all had sustained multiple (>70) fractures. Some had had their first fracture in infancy and their mean age at the time of the primary procedure was 14 years. All had sustained at least one fracture in the previous three years and one boy (case 5) had a supracondylar fracture of the contralateral femur only four weeks before the primary procedure. We believe, as previously reported by Fern et al,\textsuperscript{4} that leg-length discrepancy can be safely corrected towards the end of the period of growth as the incidence of fractures in OI decreases after puberty. Correction of deformities and leg-length discrepancy in the latter stages of adolescence instead of adulthood may help in the early realignment of the mechanical axis of the lower limb resulting in a reduced risk of plastic deformation and micro-fractures of fragile bone, in the improvement of mobility, and in the prevention of the development of further compensatory changes in joints and soft tissues.

In all our cases, the abnormal bone tolerated the external fixator throughout the period of lengthening. There was only one stress fracture through a pin track 14 days after the removal of a half-pin. The fixation of pins in all cases remained stable, with no evidence of loosening or migration. Three of our initial four cases in which limb lengthening was done using a monolateral external fixator have been previously reported.\textsuperscript{4} Placement of half-pins to obtain stable fixation is difficult in more severe cases of OI in which the diameter of the bone is less than three times the diameter of the half-pin. In four limb segments of our last two cases (cases 5 and 6), which were more severe, the Ilizarov circular frame was used with wires in the tibiae. The proximal femora required half-pins for fixation but they were spanned and half-pins of appropriate diameter were used to negotiate around the intramedullary rod. During the application of the Ilizarov fixator in these cases, we
noticed that the bone was so soft that the wires could be passed into them by hand. We did not, however, have any instance of wires cutting out through the soft bone when distraction forces were applied to the fixator during gradual lengthening. In addition, use of the Ilizarov circular fixator allowed us to lengthen simultaneously both the femoral and tibial segments, correct multplanar deformities and allow weight-bearing throughout the period of treatment.

Formation of new bone and soft-tissue elongation during limb lengthening in OI appear to be similar to those which occur in other clinical conditions. The mean bone healing index of 33.25 days/cm in our cases suggests that the regenerate bone formed within the time which is normally expected in limb-lengthening procedures performed for other conditions. No fractures occurred through the new bone regenerate, although there was plastic deformation of the tibia in our first case (case 1) in which the monolateral fixator was removed early because of the fear of stress fractures occurring through pin tracks in the soft bone. The bone healing index in this patient was 17.1 days/cm. In the next three cases (cases 2 to 4) the monolateral fixators were used for a longer period resulting in higher bone healing indices (mean 47.8 days/cm). The children who were treated using an Ilizarov circular frame (cases 5 and 6) had lower bone healing indices (mean 23.8 days/cm) and maintained stability after removal of the frame. This may reflect the mechanical principles of elastic fixation of the Ilizarov circular frame and biological preservation when percutaneous corticotomy are done. Continuing mobilisation during lengthening also has a beneficial effect on the formation of regenerate and rehabilitation.

We have successfully performed limb lengthening in the presence of an extensible intramedullary rod when there was sufficient metaphyseal bone around the rods for the insertion of pins. Although there is a risk of infection, it is beneficial to retain the previously inserted intramedullary rod during lengthening since it gives stability to the bone both during and after the lengthening procedure. Removal of intramedullary rods before lengthening entails anarthotomy and further surgical insult compromising the range of movement of the knee. After the completion of limb lengthening, we have also safely performed further insertion of rods after the medullary canal has formed, to increase the stability of bone.

We conclude that despite the abnormal bone, limb lengthening and correction of deformity in lower limbs can be achieved in children with OI. The circular frame is preferable to the monolateral fixator especially in more severe cases of OI in which the diameter of the bone is too thin for fixation of half-pins and also when there is an indication for simultaneous lengthening of both femoral and tibial segments and correction of multplanar deformities.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References