We present a retrospective review of a single-surgeon series of 30 consecutive lengthenings in 27 patients with congenital short femur using the Ilizarov technique performed between 1994 and 2005.

The mean increase in length was 5.8 cm/18.65% (3.3 to 10.4, 9.7% to 48.8%), with a mean time in the frame of 223 days (75 to 363). By changing from a distal to a proximal osteotomy for lengthening, the mean range of knee movement was significantly increased from 98.1° to 124.2° ($p = 0.041$) and there was a trend towards a reduced requirement for quadricepsplasty, although this was not statistically significant ($p = 0.07$). The overall incidence of regenerate deformation or fracture requiring open reduction and internal fixation was similar in the distal and proximal osteotomy groups (56.7% and 53.8%, respectively). However, in the proximal osteotomy group, pre-placement of a Rush nail reduced this rate from 100% without a nail to 0% with a nail ($p < 0.001$). When comparing a distal osteotomy with a proximal one over a Rush nail for lengthening, there was a significant decrease in fracture rate from 58.8% to 0% ($p = 0.043$).

We recommend that in this group of patients lengthening of the femur with an Ilizarov construct be carried out through a proximal osteotomy over a Rush nail. Lengthening should also be limited to a maximum of 6 cm during one treatment, or 20% of the original length of the femur, in order to reduce the risk of complications.

Congenital short femur is a rare congenital bone dysplasia with an incidence of one case per 52,029.1 There is a spectrum of severity ranging from simple hypoplasia to proximal femoral deficiency. It was first classified by Aitken² based on the development of the proximal femur and hip. Since then, other systems have been proposed based on the spectrum of hip stability³ and on anatomical variance.⁴ More recently, Paley⁵ has described a classification system taking into account factors that influence the reconstruction of congenital short femur. The aetiology of the condition remains uncertain, although milder forms may be multifactorial. Statistically significant epidemiological factors include male gender, first-born child and summer birth.¹ More severe forms have occurred in association with environmental factors, such as thalidomide ingestion in pregnancy affecting the fetus in the fourth to eighth weeks of gestation.⁶

Congenital short femur is associated with other abnormalities apart from isolated shortening of the bone. These include distal femoral valgus, abnormal soft tissues, knee instability due to absence or hypoplasia of the anterior cruciate ligament,⁷ abnormal morphology of the femur and an externally rotated position of the leg. Reconstruction of the congenital short femur is challenging because of these associated abnormalities, and also because the formation of the regenerate during callotasis bone lengthening is variable and complications are frequent.⁸⁻¹⁰

In this study we describe the use of a circular fixator which we believe to be versatile and able to resist the deforming forces during treatment, although lengthening can also be carried out with a monolateral fixator or hybrid fixation, which may have the advantage of better patient comfort.

**Patients and Methods**

This is a retrospective review of 30 consecutive femoral lengthenings for congenital short femur in 27 patients. The patients were treated by the senior author (RAH) over a period of 11 years between 1994 and 2005. They were identified from a prospective database of all Ilizarov treatments using circular frames, and the medical and physiotherapy records and radiographs were reviewed. The radiological
review was conducted independently by two observers (WJSA, DB). Further information was gained from clinical review or by telephone interview.

Of the 27 patients, three had the femur lengthened twice. There were 18 males and nine females with a mean age at initial lengthening of 9.25 years (5.6 to 14.7). The mean age at second lengthening was 13.1 years (11.5 to 14.5). The left femur was lengthened in 14 limbs, and the right in 16, with a mean duration of hospital stay of 8.83 days (6 to 17). The mean follow-up for first lengthening was 6.75 years (1.3 to 10.4) and for the second lengthening 2.9 years (1.5 to 4.5). All patients were classified as Paley type 1 (intact femur, mobile hip and knee) and were in Pappas grades VII, VIII and IX. The main deformity was hypoplasia of the femur, as in type IX, with coxa valga in type VIII and coxa vara in type VII.

Initial patient selection was from referrals to a tertiary clinic. Patients selected for limb reconstruction had an anticipated leg-length discrepancy at maturity of between 5 cm and 20 cm using the multiplier method as described by Paley et al. All had clinical evidence of insufficiency of the anterior cruciate ligament, but were asymptomatic and had a reasonable lower segment suitable for weight-bearing. The hip was assessed radiologically and in all cases judged to be sufficiently stable, with adequate cover of the femoral head. All patients were reviewed in a multidisciplinary pre-admission clinic which included a psychological assessment and discussion of the possible need for two or three reconstructive procedures.

During the course of this series there was an evolution in technique from a distal femoral osteotomy for lengthening, with or without acute correction of deformity, to a proximal osteotomy with acute derotation over a pre-placed Rush pin left in situ during treatment (Figs 1 and 2). In the most recent procedure, hemi-epiphysiodesis by stapling was used to correct distal femoral valgus (Fig. 1). The development of the technique was in response to the incidence of complications, particularly knee stiffness and fracture or deformation of the lengthened segment after removal of the Ilizarov frame (Smith and Nephew, Memphis, Tennessee). In virtually all cases the knee was spanned by the frame as protection against subluxation or dislocation, but in the more recent operations a multi-axial hinge was incorporated into the frame to permit controlled knee movement. A threaded rod between the distal femoral ring and the tibial ring permitted the knee to be locked in extension; this was removed for knee flexion exercises.

In cases with a distal osteotomy, the frame consisted of one or two rings distal to the osteotomy depending on the size of the patient, connected by distraction rods to a complete ring if there was sufficient room, or directly to two Italian arches for proximal fixation. Fixation was obtained distally with fine-tensioned wires and proximally with half pins. A tibial ring was attached to the distal femoral ring to span the knee.
In cases of proximal osteotomy, the frame consisted of a proximal Russian arch angled as shown in Figure 2, attached by three or four distraction rods to a complete distal femoral ring block consisting of one or two rings. Although all wire fixation was initially used proximally, the anterolateral to posteromedial wires were difficult for patients to manage, and fixation was changed to one or two anteromedial to posterolateral wires with additional fixation with half pins inserted in a lateral to medial direction. When Rush nails (Orthosolutions, Maldon, United Kingdom) were used the bone was pre-drilled at the level of the proposed osteotomy prior to insertion of the nail. The external frame was applied with the nail in situ. The proximal and distal ring blocks were then disconnected to permit completion of the osteotomy with an osteotome around the nail. Acute derotation not exceeding 30° was then performed and the ring blocks reconnected. Care was taken with the wire insertion technique, in the Russian manner by ensuring the quadriceps muscle was stretched by flexing the knee during introduction of the wire and recruiting soft tissue into the lengthening area. The knee was hinged using Russian-style multiaxial hinges made up from Ilizarov components, and positioning of the hinges was adjusted until there was free flexion of the knee.

The post-operative regimen was standardised. Distraction was started on day five, at an initial rate of 1 mm/day in four increments. The rate of distraction was modified during follow-up depending on radiological evidence of bone formation. Patients mobilised fully weight-bearing with or without crutches as comfortable. They followed a pin site protocol with chlorhexidine in spirit dressings kept in place with clips to provide pressure to the wounds. These were changed at 48 hours and then daily until the oozing stopped. Subsequently they were kept covered for five days or until fully dry, at which point they were exposed to the air. Indications for antibiotics were discharged pus, spreading erythema and increasing pain related to a wire or half pin.

Patients were seen for outpatient review every two weeks during the distraction phase and every four to six weeks during the consolidation phase. Clinical and radiological assessment was carried out to monitor formation of the regenerate and to identify hip or knee subluxation.

All patients had regular physiotherapy, including pre-clinic hydrotherapy, review by a specialist physiotherapist in outpatients, and community physiotherapy monitoring and treatment, concentrating on weight-bearing and range of movement exercises for both the hip and the knee during and after lengthening. Patients were provided with a reducible shoe raise to facilitate weight-bearing. If the knee had been spanned by the frame, the threaded rod between the distal femoral ring and the tibial ring was removed to permit controlled knee flexion exercises at least two to three times daily, but otherwise kept in place during the lengthening phase. The frame was removed under general anaesthetic when the regenerate was felt to have consolidated sufficiently. This was determined by seeing three of four cortices at least 2 mm thick on anteroposterior and lateral views of the femur. Once the frame was removed, patients were advised to mobilise partially weight-bearing with crutches for six weeks.

We compared differences in the healing index, range of knee movement, incidence of quadricepsplasty, regenerate deformation and open reduction and internal fixation (ORIF) for fracture following removal of the Ilizarov frame between proximal and distal lengthenings. Statistical differences were sought using the Mann-Whitney U test and chi-squared test as appropriate. A p-value of < 0.05 was considered significant.

Results

In all, 28 femoral Ilizarov frames were applied for unifocal lengthening; two patients had ipsilateral femoral and tibial frames for bifocal lengthening. Of the 28 femoral constructs, 24 initially spanned the knee, and of the four that did not, two were later extended across the joint. One frame integrated a Taylor Spatial Frame (Smith and Nephew) within the construct.

A total of 17 distal and 13 proximal osteotomies were performed for lengthening. Of the 13 proximal osteotomies, seven were lengthened over a Rush nail. The osteotomies were performed using a drill and an osteotome in 18 cases (12 proximal and six distal), with a Gigli saw in 11 (one proximal and ten distal), and with a power saw for one distal osteotomy.

A total of 17 patients also had a corrective osteotomy for angular or rotational deformity, nine of which were distal and eight proximal. In the distal group, four underwent acute lateral translation and four were gradually translated laterally with the frame construct; two of these involved wedge resections performed for 30° fixed flexion with 20° of valgus and 20° fixed flexion, respectively. One proximal lengthening had a distal corrective osteotomy and one had a distal hemi-epiphysiodysis.

Of eight corrective osteotomies performed proximally, five were derotated by 30° and three had angular corrections of 6°, 10° and 18°, respectively. Of these eight osteotomies, seven were at the site of lengthening and performed over a Rush nail. The 18° angular correction was plated acutely and subsequently lengthened distally.

The mean increase in length was 18.65% (9.7% to 48.8%). The mean length gained was 5.8 cm (3.3 to 10.4). The mean healing index for the whole group was 39.97 days per centimetre (22.6 to 76) and the mean range of movement of the knee at latest follow-up was 105° (15° to 140°). The mean time with the frame in situ was 223 days (75 to 363).

One patient in the proximal osteotomy group was excluded from the range of movement analysis because of loss to follow-up after removal of the frame (Table I). There was no significant difference between the ORIF rate at the
two sites of lengthening (chi-squared test, \( p = 0.79 \)). However, analysis of the ORIF rate in the proximal osteotomy group, on the basis of whether they were lengthened over a nail or not, showed significant differences (chi-squared test, \( p < 0.001 \), Table II), and comparison between a distal osteotomy for lengthening and a proximal one over a Rush nail also showed significance for fracture rate requiring ORIF (chi-squared test, \( p = 0.04 \), Table III).

### Complications
In all, nine patients had a poor functional range of knee movement with flexion of less than 90° which was refractory to physiotherapy. Two patients improved with a manipulation under anaesthesia, and seven required a quadricepsplasty one year after removal of the Ilizarov frame. Of these, six were in the distal osteotomy group, giving a quadricepsplasty rate of 35.3% (6 of 17), compared with a rate of 7.7% (1 of 13) in the proximal osteotomy group (chi-squared test, \( p = 0.077 \)).

Joint subluxation was seen in ten patients (33%), involving the knee in nine, eight of which were corrected with frame adjustment and one by slowing the rate of distraction during lengthening. One hip subluxed and lengthening was stopped.

Bone deformation in the frame was seen in six patients and required adjustment of the frame under general anaesthetic in four patients to correct angulation. The other two fractured through the proximal pin sites and underwent revision of the proximal fixation. The overall rate of bone deformation while in a frame was 20% (6 of 30 femora).

Neurovascular complications occurred in three patients. All were nerve traction injuries and recovered. Two involved reduced sensation to the foot and occurred in the two patients who had ipsilateral femoral and tibial frame lengthening. There were no vascular injuries.

There were no episodes of premature consolidation requiring lengthening to be accelerated or stopped, but five
cases of delayed consolidation were seen, three of which had cysts within the regenerate. These three had a mean lengthening of 7 cm. Two were managed successfully with an injection of bone marrow aspirate to the cyst, and one required ORIF as described below.

There were no episodes of deep infection. There was a 73.3% (22 of 30 patients) incidence of pin-site infection and two patients needed half pins replaced with wires because of infection. Breakage of half pins occurred in seven of the 30 patients, two of whom required revision of the fixation. There were no wire breakages.

After frame removal there were problems with the regenerate requiring ORIF. Fractures occurred in ten patients (33%) (Fig. 3), varus or anterior bowing of the regenerate in five (16.7%). There was cyst formation in one and one had an inadequate regenerate. The overall ORIF rate was 56.7% (17 patients) after frame removal.

Information on patients’ activity levels after treatment was obtained for 17 of the 27 patients. One patient had died from unrelated causes. In all, 11 patients were running, playing sport or dancing, three reported no functional problems, two were riding their bicycles and one was ‘happy’ with the outcome.

Further analysis of the complications was performed. Cut-off points for total length gained were taken at 5 cm or below and over 5 cm; 6 cm or below; and over 6 cm. Cut-off points for percentage length gained were taken at 15% or less or over 15%; and 20% or less and over 20% of the original length of the bone.

Significant differences were found in the delayed consolidation group and those with fracture of the regenerate. Patients lengthened more than 6 cm were more likely to have delayed consolidation ($p = 0.028$), and those lengthened more than 20% of the original length were more likely to suffer from a fracture of the regenerate ($p = 0.011$).

There were no significant differences in the analyses of length gained for the patient having one or more complications, quadricepsplasty, subluxation of the knee, neurological injury, half pin fracture and ORIF.

**Discussion**

To date, this is the largest published series of lengthening for congenital short femur using the Ilizarov technique by a single surgeon in the English literature. The overall results compare favourably with those of previously published series. Grill and Dungal$^8$ reported eight lengthenings of congenital short femur using the Ilizarov technique in a series of 37 lengthenings by various methods. Stanitski et al$^9$ reported the results of seven lengthenings; Danziger, Kumar and De Weese$^{10}$ reported six cases. However, these series give little information about detailed complications apart from fracture rates, making comparisons difficult. Regarding the severity of the deformity, Grill and Dungal$^8$ stated that four of their eight cases were Pappas grade III/IV and one was grade VII, but gave no information about the other three. Stanitski et al$^9$ had six hypoplastic femora and one proximal femoral focal deficiency. Danziger et al$^{10}$ provide no information about the severity of the deformity. In this series we treated ten patients with Pappas grade VII, 12 with grade VIII and eight with grade IX.

Lengthening a congenital short femur is difficult, with a risk of complications such as joint subluxation, poor regenerate formation, fracture after frame removal and knee stiffness. Careful clinical and radiological follow-up is mandatory to identify and anticipate complications. Joint subluxation is a significant risk, and in our series nine patients had subluxation of the knee. This emphasises the need for good-quality anteroposterior and lateral radiographs of the knee during follow-up, as well as radiographs of the regenerate site. It was our experience that patients with knee subluxation may present clinically with pain and swelling of the knee, with the skin over the knee having a shiny appearance. The likely causes of knee subluxation include pre-existing instability due to hypoplasia of the anterior cruciate ligament,$^7$ and increasing tension in the hamstring muscles as lengthening proceeds. The knee can be bridged by the frame to reduce this risk, and in 26 of 30 femoral lengthenings this was done from the outset, with two further patients subsequently having the knee bridged. After an initial review of the first group of 11 primary and three secondary lengthenings,$^{12}$ the senior author reviewed his technique of distal corticotomy to try to reduce the rate of complications, in particular knee stiffness necessitating...
quadricepsplasty and fracture after frame removal. The technique was changed to the knee being hinged with a Russian-style multi-axial hinge as well as improving the technique of insertion of the wire, and changing to a proximal osteotomy above the bulk of the quadriceps to reduce muscle scarring and adherence to the regenerate. These measures were effective in reducing the incidence of knee stiffness, but owing to the manner of development of our technique it is not possible to isolate the relative contribution of these changes. However, there was a significant increase in the mean range of movement of the knee from 98.1° to 124.2° (p = 0.041) and a trend towards reduction in the need for quadricepsplasty from 35.3% to 8.3% (p = 0.077). In the nine cases where knee subluxation did occur, it was recognised promptly and successfully managed by frame adjustment, physiotherapy, and in one case by slowing the rate of distraction. There was one case of hip subluxation necessitating cessation of lengthening.

The rate of bone deformation while in the frame was 20% and required further intervention. Regenerate formation is sometimes asymmetrical and subject to deformation (Fig. 2), possibly because there is a varus and posterior bending force on the bone due to the abnormal soft tissues and because of the alignment of the frame. Although it is important to recruit as much tissue as possible into the lengthening segment by having a good distance between the rings, stable fixation is also important. It is valuable to have a frame that can be adjusted to compensate for regenerate deformation. Lengthening over a Rush pin helps to maintain alignment with lengthening along the anatomical axis.

Of the five cases of delayed consolidation, three were noted to be due to cyst formation within the regenerate, which occurred in patients with above-average lengthenings in terms of both percentage (25.4%) and actual length gained (7 cm). Accordingly, over-ambitious lengthening in this condition should be avoided.

In the previous series the fracture rate after removal of the frame has been up to 50%. In our series one-third of the lengthenings fractured at this stage. However, once the insertion of the intramedullary Rush nail was added to the technique of proximal osteotomy, no further fractures occurred.

Comparison of the original technique of a distal osteotomy and lengthening over a Rush nail, shows a statistically significant reduction in the fracture rate at the site of lengthening requiring ORIF (p = 0.043).

Lengthening of a congenitally short femur is extremely challenging. From our findings, we recommend that when using a circular frame lengthening should be carried out through a proximal osteotomy over a Rush nail, with the frame hinged at the knee to reduce the complication rate of knee stiffness, fracture after removal of the frame and development of angular deformity. A second distal osteotomy for acute or gradual correction of distal femoral valgus can be performed if appropriate, although we currently prefer distal hemi-epiphysiodesis. Lengthening should also be limited to a maximum of 6 cm during one treatment, or 20% of the original length of the femur.

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