LENGTHENING FOR CONGENITAL SHORT FEMUR

RESULTS OF DIFFERENT METHODS

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We have reviewed the results in 37 patients with unilateral congenital short femur (Pappas classes III to IX), treated by different lengthening procedures. The increase in the length of the femur varied from 15.6% to 142%, excellent or good results being obtained in 32 patients (86%).

There was an average of 1.9 complications per case, most being seen earlier in the series when the Wagner technique was used. With the Orthofix and the Ilizarov techniques, we used callus distraction in all cases. We found that the proximal diaphysis of the congenitally abnormal femur healed less well, and we now prefer to perform corticotomy and callus distraction of the distal metaphysis. The Ilizarov method gave the best results, offering the possibilities of the simultaneous use of a Hoffmann fixator across the hip and the treatment of knee dislocation and instability.

Congenital short femur is a rare condition, ranging from absence of the entire bone with abnormal development of the pelvis to the least severe form where a femur of normal shape is smaller than that on the other side. The usual classification is that of Pappas (1983) who distinguished nine classes, of which I is the most severe (see Fig. 1). The condition may be unilateral or bilateral and is often associated with other congenital anomalies. The incidence of this complex defect has been reported by Rogala et al (1974), from the Edinburgh Register of the Newborn, to be about 1 in 50 000 of the population (0.2 per 10 000).

At birth the exact characteristics of the deformity cannot be determined, but by about two years of age classification is possible by clinical and radiological examination (Fixsen and Lloyd-Roberts 1974; Hamanishi 1980; Pappas 1983). Recently the use of CT scans and MRI have proved to be of great diagnostic and prognostic value.

It has been shown that the relative lengths of the femora and the tibiae remain almost unchanged during growth (Ring 1959; Amstutz 1969; Westin, Sakai and Wood 1976; Pappas 1983), and this enables the final discrepancy in leg length to be predicted from early radiographs. Careful interpretation of the growth data makes it possible to determine the correct combination of lengthening, shortening, and epiphyseodesis which will be needed to achieve equality of leg length. Treatment then consists of a complete series of operations, not only to achieve leg length equality, but also to improve the stability of the knee and hip and to correct any associated foot deformity.

Because of the need for numerous operations, entailing long periods in hospital, and the high risk of a poor outcome in classes III to VII (Pappas 1983) amputation has, until recently, been advocated for severe cases (Gillespie and Torode 1983; Grogan, Love and Ogden 1987).

We have reviewed our experience of leg lengthening in order to evaluate different methods in terms of the elongation achieved, healing time, fixation, period in hospital and the incidence and management of complications.

PATIENTS AND METHODS

From 1979 to 1988, a total of 40 patients with unilateral congenital short femur were seen at the Speising Hospital, Vienna or the Bulovka Hospital, Prague. Patients with isolated congenital or infantile coxa vara, a lesion which is usually restricted to the femoral neck, were not included. Three patients (one in class II and two in class...
III) were not treated, leaving 11 boys and 26 girls with 20 right and 17 left femora affected.

Pre-operative assessment included clinical examination and standard, standing and orthoradiographs of both legs with views of the hips, knees and ankles to determine stability, femoral neck retroversion and any other anomalies. Growth predictions were made from the orthoradiographs, using films of the hand and wrist (Green and Anderson 1960), a Moseley (1977, 1987) graph and the Pappas (1983) growth chart for congenitally abnormal femora.

Femoral lengthening was considered for some class III and class IV cases, when the parents could not accept ablative procedures, and in all patients in classes VII, VIII and IX. A prerequisite in all these cases was the probability of satisfactory function of both the hip and the knee.

Lengthening procedures were performed at ages ranging from four to 20 years, earlier in patients in classes III, IV and VII because of the severity of their deformities. Patients with class VIII and IX deformities usually had their operations between nine and 13 years of age. Before lengthening, the femora were from 35% to 87% of the length of the contralateral normal bones (10.5 to 37.7 cm).

**Surgical methods**

**Class III and class IV.** The four patients with class III and IV deformities (Fig. 2) lacked bony continuity between the femoral head and the shaft: correction is therefore performed in three stages by the technique of one of the authors (PD).

1) A unilateral fixator is fixed to the ileum and to either the femur or the tibia. Continuous distraction is then used to bring the proximal end of the femoral shaft to the level of the acetabulum. During this period of distraction healing of a subtrochanteric pseudarthrosis was seen in one case (Figs 2b and c).

2) At the next stage, the pseudarthrosis at the level of the femoral neck is resected, allowing fusion between the femoral head and the shaft (King 1973).

3) At the third stage, lengthening of the short femur is achieved by using an Ilizarov fixator ring below a corticotomy of the distal femoral metaphysis (Figs 2d and e). To protect the femoral head and maintain hip stability, the Ilizarov ring is connected to a Hoffmann frame (ETS Jaquet Freres, Geneva) placed between the ilium and the shaft of the femur to provide pelvifemoral fixation (Fig. 2c).

When the planned femoral lengthening has been achieved and healing is adequate, the external fixators are removed and replaced by a hip spica for three months. Walking and partial weight-bearing is then allowed in a specially fitted splint.
Classes VII, VIII and IX. We used four different techniques of lengthening for the 33 patients in these classes, with three different designs of external fixator: the Wagner (Synthes, Paoli, Pennsylvania), Orthofix (EBI Medical Systems, Parsippany, New Jersey), and the Ilizarov (Richards Medical Company, Memphis, Tennessee).

The Wagner distraction device was used for 10 femora after transverse diaphyseal osteotomies performed with an oscillating saw. Immediate distraction of 1 cm was followed by a daily distraction of 1 to 2 mm. When adequate length had been obtained, the gap between the bone ends was filled with autologous graft from the iliac crest and stabilised by a special AO plate designed for this procedure. In the first six cases the plate was fixed laterally, in the later four cases it was placed on the posterior side of the femur to provide better stability against lateral bending forces.

Five other patients had the Wagner device and a step-cut osteotomy. Distraction of the callus was within a periosteal sleeve, and additional stabilisation during and after lengthening was provided by a Küntscher nail of small diameter in the medullary cavity. The distraction was by 0.5 mm twice in each day. No grafting was necessary (Bost and Larsen 1956; Westin and Gunderson 1969).
The Orthofix device popularised by DeBastiani et al (1987) was used in 14 cases. After corticotomy of the proximal femur and seven to 14 days delay, callus distraction was started at a rate of 0.25 mm four times daily. When lengthening was complete the apparatus remained in situ until there was adequate new bone in the distraction gap. Axial weight-bearing (dynamisation) was then allowed to stimulate remodelling. When corticalisation of the new bone was seen in the whole of the distraction gap the fixator was removed. No grafting was necessary.

The original Ilizarov distractor and a distal femoral corticotomy were used in eight cases, four of them in classes III and IV as described above. One case in class VII also had additional pelvifemoral stabilisation with a Hoffmann frame. The regimen for callus distraction was the same as that described for the Orthofix device. We used distraction epiphysesoty for only one of our patients (Grill 1984).

Correction of anatomical and functional alignment is also very important. Angular deformities of the proximal femur such as coxa vara (class VII) were corrected by valgus osteotomy, on average at about 1.5 years after complete consolidation of the lengthened bone. This osteotomy can provide a further femoral lengthening of 1.5 to 2.5 cm (see Figs 3f and g). Shortening of the contralateral normal leg was performed for 12 patients, using distal femoral and proximal tibial epiphysesoties or shortening osteotomies.

A total of 239 operations were performed on the 37 patients we report, an average of 6.5 procedures for each case.

RESULTS

The evaluation of results has to take into account both the severity and class of the original deformity and the technique used for lengthening. Femoral lengthening varied from 15.6% to 142% of the original length (Table I), with a daily gain of 0.37 mm to 0.73 mm. A value of 0.5 mm per day seems to be an acceptable compromise between speed and optimum bone formation. The mean elongation using the Wagner system was 5.2 cm, with the Orthofix system it was 8.9 cm and with the Ilizarov system 11.7 cm (Table II). The complexity of the course of treatment and the number of operations was least in class IX patients.

Excellent results, with both femurs of equal length or both legs equal and normal hip and knee function, were obtained in 19 patients (51%), all originally in classes VIII or IX (Table III). In 13 patients (35%) only partial correction of the deformity was possible. Five patients (14%), all in classes VII or VIII, gained no correction: these patients had worse problems than before operation. All other patients had good function and were able to attend normal schools or employment. Complications. There were 64 complications with an average of 1.9 per case, with significantly fewer problems in class IX patients (Table IV).

We distinguished between problems and two types of complications. A problem is a difficulty arising during treatment which can easily be reversed by a minor non-surgical intervention. Minor complications are those which require surgical intervention but do not prejudice

### Table I. Details (number and mean values) of 37 patients treated for congenital short femur by bone lengthening

<table>
<thead>
<tr>
<th>Class of deformity (Pappas 1983)</th>
<th>III</th>
<th>IV</th>
<th>VII</th>
<th>VIII</th>
<th>IX</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>3</td>
<td>1</td>
<td>10</td>
<td>9</td>
<td>14</td>
</tr>
<tr>
<td>Age at start of treatment (yr)</td>
<td>3.2</td>
<td>2.3</td>
<td>4.0</td>
<td>5.5</td>
<td>6.0</td>
</tr>
<tr>
<td>Age at start of lengthening (yr)</td>
<td>5.5</td>
<td>6.0</td>
<td>8.5</td>
<td>13.0</td>
<td>12.1</td>
</tr>
<tr>
<td>Femur</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Original length (cm)</td>
<td>13.3</td>
<td>10.5</td>
<td>34.2</td>
<td>33.7</td>
<td>37.7</td>
</tr>
<tr>
<td>Relative shortening of femur (cm)</td>
<td>16.3</td>
<td>19.5</td>
<td>10.8</td>
<td>9.5</td>
<td>5.9</td>
</tr>
<tr>
<td>Percentage of normal femur</td>
<td>45</td>
<td>35</td>
<td>76</td>
<td>78</td>
<td>87</td>
</tr>
<tr>
<td>Total elongation (cm)</td>
<td>16.7</td>
<td>15.0</td>
<td>7.2</td>
<td>8.9</td>
<td>5.5</td>
</tr>
<tr>
<td>Percentage elongation</td>
<td>125</td>
<td>142</td>
<td>32</td>
<td>27</td>
<td>15.6</td>
</tr>
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</table>

### Table II. Mean values for treatment of congenital short femur by lengthening

<table>
<thead>
<tr>
<th>Pappas class</th>
<th>III</th>
<th>IV</th>
<th>VII</th>
<th>VIII</th>
<th>IX</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of operations</td>
<td>9</td>
<td>8</td>
<td>7.3</td>
<td>6.8</td>
<td>5</td>
</tr>
<tr>
<td>Period of elongation (days)</td>
<td>227</td>
<td>280</td>
<td>191</td>
<td>200</td>
<td>111</td>
</tr>
<tr>
<td>Stay in hospital (days)</td>
<td>525</td>
<td>449</td>
<td>243</td>
<td>207</td>
<td>141</td>
</tr>
<tr>
<td>Number of admissions</td>
<td>8.66</td>
<td>9</td>
<td>8.2</td>
<td>7.6</td>
<td>5</td>
</tr>
<tr>
<td>Gain in bone length (mm/day)</td>
<td>0.73</td>
<td>0.53</td>
<td>0.37</td>
<td>0.42</td>
<td>0.50</td>
</tr>
</tbody>
</table>

### Table III. Results of treatment for congenital short femur

<table>
<thead>
<tr>
<th>Result</th>
<th>Pappas class</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>III</td>
</tr>
<tr>
<td>Femurs equal*</td>
<td>0</td>
</tr>
<tr>
<td>Legs equal*</td>
<td>0</td>
</tr>
<tr>
<td>Improved†</td>
<td>3</td>
</tr>
<tr>
<td>Failed</td>
<td>0</td>
</tr>
</tbody>
</table>

* satisfactory function
† partial correction of length discrepancy, adequate function
Figure 3a – Femur of a boy of 16 years, showing an Orthofix device and corticotomy. Figure 3b – Callus distraction to give 11 cm lengthening. Figures 3c and d – Even after a long period of dynamisation the bone shows the so-called 'chewing-gum effect'. Figures 3e and f – After stabilisation by a Rush pin there is consolidation of the bone. Figure 3g – Varus deformity of the upper femur was corrected by an intertrochanteric valgus osteotomy, giving another increase in length.

Table IV. Complications of treatment for congenital short femur

<table>
<thead>
<tr>
<th>Pappas class</th>
<th>III</th>
<th>IV</th>
<th>VII</th>
<th>VIII</th>
<th>IX</th>
<th>All</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>3</td>
<td>1</td>
<td>10</td>
<td>9</td>
<td>14</td>
<td>27</td>
</tr>
<tr>
<td>Pin track infection</td>
<td>2</td>
<td>1</td>
<td>8</td>
<td>7</td>
<td>9</td>
<td>27</td>
</tr>
<tr>
<td>Deep infection</td>
<td>–</td>
<td>–</td>
<td>2</td>
<td>1</td>
<td>–</td>
<td>3</td>
</tr>
<tr>
<td>Femoral shaft fracture</td>
<td>3</td>
<td>1</td>
<td>9</td>
<td>3</td>
<td>6</td>
<td>22</td>
</tr>
<tr>
<td>Neural palsy</td>
<td>–</td>
<td>–</td>
<td>1</td>
<td>–</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Dislocation of knee</td>
<td>2</td>
<td>1</td>
<td>3</td>
<td>2</td>
<td>–</td>
<td>8</td>
</tr>
<tr>
<td>Dislocation of hip</td>
<td>–</td>
<td>–</td>
<td>1</td>
<td>1</td>
<td>–</td>
<td>2</td>
</tr>
<tr>
<td>Total number of complications</td>
<td>7</td>
<td>3</td>
<td>24</td>
<td>14</td>
<td>16</td>
<td>64</td>
</tr>
</tbody>
</table>
the final result. Major complications hinder the achievement of the original goal or cause permanent problems which may make the deformity worse (Paley 1988).

Fracture of the femur in the lengthened zone was common: there were 22 such fractures in our series, 18 of them in the 15 patients treated by the Wagner technique. Fractures usually occurred after removal of the plate but some were seen with the plate in situ. One patient in class VII had four fractures over a period of two years. In classes VII and IX, fractures always reduced the lengthening obtained and required treatment by operative osteosynthesis, usually with Rush pins.

The four class III and IV patients, all treated by a combination of the Hoffmann and Ilizarov techniques, all had secondary femoral shaft fractures as a result of minor trauma. All these fractures healed well with callus formation providing a stronger bone and thus improving the end result.

Proximal diaphyseal lengthening by callus distraction after corticotomy, with either the Wagner or the Orthofix apparatus, caused increasing atrophy and rarefaction of the elongated shaft (the chewing-gum effect) in several patients (Fig. 3). This effect seemed to depend not only on the duration of the lengthening procedure but also on the duration of external fixation itself. Once established, these changes continued to develop during the periods of neutralisation and dynamic weight-bearing with the Orthofix device, and could be reversed only by removing the fixator, stabilising the very atrophic femur with a Rush pin (Fig. 3e) and encouraging partial weight-bearing. This allowed the femur to consolidate and develop a normal shape over several months (Fig. 3f).

Boden et al (1989) described a defect in the proliferation and maturation of chondrocytes of the proximal growth plate in a 21-week foetus with unilateral proximal femoral focal deficiency. The proximal growth plate failed to migrate away from the central diaphysis and showed abnormal architecture in every zone, including lack of normal preparation of matrix by the hypertrophic cells. Consequently the timing and occurrence of mineralisation, vascular invasion and endochondral growth in the proximal femur were greatly disturbed. These findings may help to explain some of the complications of management by bone lengthening.

Neurological complications were seen in two cases. One girl with a class IX femur developed a complete sciatic nerve palsy on the seventh day of lengthening. There was full recovery after a year. The second case showed irritation of the sciatic nerve, probably caused by an excessively long transfixing pin. The symptoms settled when the pin was removed.

Deep infection necessitating revision and drainage was seen in only three cases, all treated by the Wagner method.

Considerable, but usually transient, knee stiffness was found at some stage in 34 of the 37 cases, and flexion contracture of the hip in 21. These restrictions of joint movement occurred most often during rapid lengthening and in patients being treated out of hospital. Three patients already had a flexion contracture of the knee as part of their deformity, and all 14 patients in classes III, IV and VII had a radiologically dysplastic knee. In four of them aplasia of the cruciate ligaments was diagnosed arthroscopically. We treated simple limitation of movement by physiotherapy, continuous passive motion, and a few days interruption in the leg-lengthening process. Extension contracture of the knee was not a major problem with regard to the final result, the range of movement always improved, but in some cases this took more than a year. Normal ranges of both hip and knee movement were achieved in only a few class IX patients (Table V).

Table V. Limitation of range of hip and knee movement after treatment for congenital short femur

<table>
<thead>
<tr>
<th>Pappas class</th>
<th>III</th>
<th>IV</th>
<th>VII</th>
<th>VIII</th>
<th>IX</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of cases</td>
<td>3</td>
<td>1</td>
<td>10</td>
<td>9</td>
<td>14</td>
<td></td>
</tr>
<tr>
<td>Limitation of knee movement</td>
<td>3</td>
<td>1</td>
<td>10</td>
<td>11</td>
<td></td>
<td>34</td>
</tr>
<tr>
<td>Limitation of hip movement</td>
<td>3</td>
<td>1</td>
<td>10</td>
<td>5</td>
<td>2</td>
<td>21</td>
</tr>
</tbody>
</table>

Subluxation or dislocation of the knee, caused or aggravated by lengthening, was seen in eight of the 14 patients with knee dysplasia. Treatment of this severe complication was by soft-tissue release, including the iliotibial band, in three; by extended use of the Ilizarov method in three (Fig. 4); and by simple traction in the other two. At the latest review, seven of these cases had satisfactory reduction, but all eight showed some loss of movement compared with their pre-operative range.

Dislocation of the hip was seen in two cases. In one four-year-old patient with coxa vara, femoral distraction caused the femoral head to dislocate, but this was corrected after adjustment of the apparatus. The second dislocation occurred in a class VIII patient and was the result of a dysplastic acetabulum and retroversion of the femoral neck. Derotation osteotomy, posterior arthroscopy and a Chiari pelvic osteotomy were performed, but one year later displacement recurred and the patient's parents did not consent to a further operation.

Soft-tissue complications are best avoided by extreme care not to transfix muscle by pins or screws, especially at the level of the distal femur. When using the Ilizarov device the knee should be flexed when wire is inserted from the anterior side and extended before the pin is drilled through the posterior cortex. During an
operation the mobility of the knee should be tested after any pin or screw insertion.

DISCUSSION
We have been unable to find a paper, in English or German, which reports the indications, results and complications of different lengthening procedures for unilateral congenital short femur and, in particular, we found no previous references to leg-length equalisation in patients with class III or class IV deformities.

The prevalent opinion is that length equalisation in class VII, VIII and IX patients is best achieved by contralateral epiphysodesis or a shortening osteotomy, and that lengthening of the femur or the tibia or both should rarely be considered (Panting and Williams 1978; Koman, Meyer and Warren 1982; Gillespie and Torode 1983; Pappas 1983; Grogan et al 1987). Length equalisation is said to be indicated only where the length of the short femur is over 60% of that on the normal side and the predicted discrepancy is less than 17 cm (Koman et al 1982).

However, there is general agreement (Blauth and Hepp 1978; Koman et al 1982; Epps 1983; Gillespie and Torode 1983; Pappas 1983) that children with class III to VIII deformities are likely to develop a length discrepancy of from 20 to 35 cm; and that this is too great a discrepancy to correct surgically. For this reason, some form of amputation is the most common treatment. If the eventual use of a below-knee prosthesis is planned then a Van Nes rotation plasty is performed. If an above-knee level is chosen a Syme amputation (Syme 1843) can be performed with a view to knee fusion and possible epiphysodesis to give the correct length of stump (Kostuik et al 1975; Kritter 1977; Epps 1983; Grogan et al 1987).

There are few reports of the results of lengthening procedures for congenital short femur. Gillespie and Torode (1983) report eight patients lengthened by the Wagner technique, three of them in preparation for a Van Nes procedure. The complications included posterior dislocation of the knee in five patients, femoral fracture in two, and transient footdrop in one. They gave no data on the amount of lengthening. Koman et al (1982) mention 13 leg-lengthening procedures in nine of 62 patients, but give no details. Pappas (1983) considered that the abnormal short femur was less likely to heal well than a normal bone, and that this could result in delayed union, failure to achieve the anticipated length gain, and a high incidence of complications due to the presence of abnormal tissues.

In our series, the least satisfactory results and the most complications were seen with the original Wagner technique. This confirms the experience of Jones and Moseley (1985) and Mosca and Moseley (1986) who reported 142 complications (60 serious) in 63 lengthenings. Only eight cases had no complications. These disappointing results can be explained by the rate and
frequency of distraction, and the failure to preserve periosteal and marrow tissues and the nutrient blood vessels at the level of the osteotomy. In the five cases in our series in which the Wagner distractor was combined with Kuntscher nailing and callus distraction without subsequent grafting and plating, the results and complications did not differ from the Orthofix group.

In the Orthofix and Ilizarov groups, distraction was performed after open corticotomies and the onset of callus formation, using the tension stress effect for the genesis and growth of the tissues (Ilizarov 1988, 1989a.b). Distraction was by 0.25 mm twice daily: a 50% slower rate than that suggested by Ilizarov (4 times 0.25 mm per day) for normal bones. Despite this difference we saw no premature consolidation of the lengthening bone, possibly because a congenital short femur has less competence in bone healing.

In the Orthofix group the fixator was in place for over 60% longer than in the Ilizarov group and seven of the 14 cases showed no stable bone consolidation during or after lengthening. This 'chewing-gum effect' was not observed in the Ilizarov group and there were fewer fractures. The fractures which occurred in the Hoffmann–Ilizarov group were compression fractures of the newly formed bone. The difference can be explained by the fact that in this group the corticotomies, except in one case, was in the distal femur and not proximally in the region of the most severe structural changes, where the cortex was thicker and there was narrowing of the medullary cavity. An additional factor may be the biomechanical difference between the Ilizarov and Orthofix systems, the former allowing more cyclic axial micromotion (Fleming et al 1989).

Nevertheless, the Orthofix fixator is more easily mounted and has high patient acceptance (Grill 1989). We had no complications in class IX patients with the Orthofix system, and, in this type of deformity, we still recommend it. However, for class VII and VIII patients we prefer the Ilizarov system, for the above-mentioned advantages and because of the development of new apparatus for the fixation of the proximal femur (Catagni 1989), and an oblique support connection which allows more comfortable fixation in the proximal area. Another advantage of the Ilizarov method is that it allows for an articulation across either hip or knee; slow reduction of a joint deformity is possible (Fig. 4).

For patients with class III and IV deformities the advantages, disadvantages and alternatives to the lengthening procedure require detailed discussion with the parents. The average healing time after a Van Nes rotationplasty (Van Nes 1950) is about 40 days (Kritter 1977); treatment by bone lengthening takes six months to one year, it involves several operations, some pain and intensive physiotherapy. Even after successful leg-length equalisation there is likely to be early osteoarthritis of either hip or knee or both. However, our four class III and class IV children and their parents were all satisfied: they could walk without orthoses or crutches using only shoe lifts; they all have functional ranges of knee and hip movement.

Of our 37 patients who had lengthening procedures for congenital short femora, all but five had improved function and were able to be active with a higher level of quality of life.

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


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