LEG LENGTHENING IN TURNER DWARFISM

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We have reviewed 16 patients treated by leg lengthening for various forms of Turner dwarfism with regard to the long period of healing and the complications. We consider that Turner dwarfism is a suitable indication for leg lengthening because of the moderate length deficit and the morphological appearance of the patients, and have introduced an improved programme of management to deal with the problems encountered.

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Short stature or ‘Turner dwarfism’ in the adult is a feature of syndromes caused by chromosomal abnormalities, notably the Turner syndrome, the mixed gonadal dysgenesis syndrome, the male Turner syndrome and the Noonan syndrome.

The Turner syndrome is relatively common (1/5000 live births) (Smith 1982), mostly affects females, and is the result of the presence of one rather than two X chromosomes (45,XO karyotype). The mixed gonadal dysgenesis syndrome, the male Turner syndrome and the Noonan syndrome arise when both X chromosomes are present but are abnormal (female XX karyotype with mosaicism, deletion and isochromosomes). The symptoms are less pronounced and all intermediate stages between normality and the classical Turner syndrome are possible in terms of both sexual development and stature.

Mixed gonadal dysgenesis can be considered as a form of hermaphroditism, with ambiguous external genitalia. The most frequent karyotype is 45.XO/46.XY mosaicism (Sohval 1963). The phenotype is almost always female and shortness of stature is not severe.

The male Turner syndrome (Ferrier and Ferrier 1967) is caused by partial testicular dysgenesis and has a normal male karyotype and phenotype. There is marked dwarfism.

The Noonan syndrome (Noonan 1968; Smith 1982) affects both males and females and is characterised by short stature and various forms of somatic anomaly, not always readily distinguishable from normal. Clinical evidence of the syndrome is seen in 1/1000 subjects and minor degrees may be present in 1/100 (Neri and Zelante 1990). There are no gonadal or karyotype abnormalities.

A study of Turner dwarfism was carried out by Aldegheri, Agostini and Antoniazzi (1992) in ten females in whom the mean final loss of height was 25 cm (15.5%) when compared with equivalent normal women (Aldegheri and Agostini 1993). The loss of height was not proportional since one-third occurred in the trunk and two-thirds in the legs. Here, shortening of the tibia was greater than in the femur with a tibial length of 77% of femoral length, compared with 84% in normal subjects.

We reviewed 16 patients with Turner syndrome in whom leg lengthening had been performed and discuss the results and complications.

PATIENT AND METHODS

All 16 patients were female with a mean age of 18 years (15 to 22) at the time of operation. Nine had the typical Turner syndrome (45,XO karyotype), three had mixed gonadal dysgenesis (45.XO/46.XX), and four had the Noonan syndrome (46,XX). All had a normal IQ. The average height before operation was 138 cm (132 to 153) and all showed disproportionate short stature with leg-to-trunk and tibia-to-femur ratios of 0.80 (0.71 to 0.86) and 0.78 (0.70 to 0.82), respectively (Fig. 1).

Both legs (32 femora, 32 tibiae) were lengthened by the callotasis method (Aldegheri 1993) using the Orthofix lengthener (Orthofix, Bussolengo, Italy). In the first eight patients we applied the fixator and performed an osteotomy. In the remaining eight we modified the technique to include fasciotomy of the fascia lata, the application of three fixation pins to the proximal osteotomy fragment in both femur and tibia, anterior application of the tibial fixator, fixation of the distal tibiofibular syndesmosis with one AO screw, percutaneous tenotomy of tendon Achilles and immobilisation in a below-knee cast throughout the period of distraction (Trivella et al 1992).

We used the cross-over procedure (Trivella and Alde-
gheri 1988) with lengthening of the right femur and left tibia, followed by lengthening of the left femur and right tibia after removal of the first two fixators. In five patients the second stage was performed once distraction of the first two segments was complete, which required the use of four fixators simultaneously for a mean period of three months (Fig. 2).

After operation we assessed the gain in length in centimetres and as a percentage of the preoperative lengths of the tibia and the femur as measured on radiographs. The
healing time was expressed as the healing index which is the number of days required for the complete healing of 1 cm of lengthened bone (De Bastiani et al 1987). We noted complications, including those which did not affect the planned final result and those which caused permanent damage, and measured joint function, the leg-to-trunk and tibia-to-femur ratios and the final height.

RESULTS

All patients achieved lengthening with a mean increase in height of 13 cm and with a mean final height of 151 cm (145.5 to 159). The leg-to-trunk and tibia-to-femur ratios were improved from 0.8 to 0.96 (normal value 0.9) and 0.76 to 0.81 (normal value 0.84), respectively, and the type of abnormality did not influence the result.

Table I shows the results of femoral and tibial lengthening. The mean lengthening in the 32 femora was 6 cm (4.5 to 9 cm) equivalent to a mean of 16.5% (12 to 23) of the initial length. All femora healed spontaneously with a healing index of 46.5 days (28 to 77). The mean lengthening in the 32 tibiae was 7 cm (4.5 to 10), equivalent to a mean of 25% (14.5 to 37) of the initial length. The mean healing index was 46 days (24 to 102).

Table II. Femoral and tibial complications of lengthening in 16 patients

<table>
<thead>
<tr>
<th>Complications</th>
<th>Femur</th>
<th>Tibia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fusion of osteotomy</td>
<td>3</td>
<td>-</td>
</tr>
<tr>
<td>Failure to consolidate</td>
<td>-</td>
<td>1</td>
</tr>
<tr>
<td>Fracture</td>
<td>3</td>
<td>-</td>
</tr>
<tr>
<td>Secondary axial deviation</td>
<td>-</td>
<td>7</td>
</tr>
<tr>
<td>Equinus foot</td>
<td>-</td>
<td>8</td>
</tr>
<tr>
<td>Subtalar deformity</td>
<td>-</td>
<td>1</td>
</tr>
</tbody>
</table>

Complications were seen in 21% of the femora and 40% of the tibia (Table II). One patient had necrosis of the head of the femur after subtrochanteric corrective osteotomy for residual femoral varus, and another was left with permanent stiffness of the tibiotalar joint. Three femora had early fusion of the lengthening osteotomy which was treated by a further osteotomy. Another three fractured after removal of the fixator; treatment was by intramedullary nailing in one and immobilisation in a plaster cast in two. In the latter patients a residual varus deformity required valgus osteotomy and immobilisation in an external fixator. There was one pin osteolysis which was treated by resiting of the pin.

There were 22 complications in 13 tibiae. Five tibiae each had three complications. Failure of consolidation in one required a corticocancellous graft and reapplication of the fixator. Seven had residual valgus deformities of more than 10° each requiring osteotomy and further use of an external fixator. Osteolysis required resiting of the pins in five cases and lengthening of tendo Achillis was necessary in eight. Another patient had a flat foot with a valgus deformity due to subtalar subluxation which resolved after soft-tissue surgery.

In all the remaining cases there was some limitation of joint movement for three months after removal of the fixator.

DISCUSSION

The healing time for callotasis is longer than that for other methods of treating length discrepancy (De Bastiani et al 1987) or other forms of short stature (Aldegheri et al 1988).

In patients with Turner dwarfism bone matures late and sometimes incompletely. Oestrogen deficiency and any associated osteopenia may contribute to the slow ossification and delay in maturation of the cortex. Dysplasia of the lymphatics and lymphoedema in the lower legs may also delay healing. More than half of our patients had lymphoedema during lengthening but it always resolved.

In view of the risk-to-benefit ratio involved in this procedure, a number of special considerations are required. The time during which the patient wears external fixators is protracted. In 11 patients the healing time for the first two bones was 11 months. The other two bones were then operated on three months later which took another 11 months before the fixators could be discarded. In the remaining five patients the second lengthening stage was commenced during the final healing stage of the first reducing the total time to 18 months.

The complication rate is high. In the first eight patients problems were encountered in all the tibiae, but in the remainder in whom the modified protocol was used this was reduced to 44%. The high number of complications is explained by the high percentage of lengthening required in the tibia to restore the leg-to-trunk and tibia-to-femur ratios, and the long healing times.

In recent years we have used only the Orthofix slide lengthener (Limb Reconstruction System) (Aldegheri 1993) which has advantages over the traditional lengthener fixator. It is more stable, allows fixation of very short segments with distal and proximal pins close together, ensures a safe distance from the distal femoral pins to the knee and in major lengthenings avoids any need for replacing a short....
lengthener with a longer model. Callotasis is a reliable technique; the lengthened segments heal spontaneously, although the long healing time requires prolonged external fixation. It is therefore important that the technique be applied with great care and precision. In future we will use the Limb Reconstruction System with fixation of bone segments with three proximal and three distal pins and, in the tibia, employ an epiphyseal proximal fixator clamp which allows a more proximal osteotomy (Pouliquen et al 1993). Slowing the distraction rate in the tibia will help to ensure more regular ossification and ease soft-tissue tension.

All our patients were followed up for a minimum of one year after fixator removal. Except for the two with permanent disability, all were satisfied with the final result. They appreciated the improvement in appearance, and said that they had greater self-esteem, with a better approach to their social life. The leg-to-trunk and tibia-to-femur disproportionate was completely corrected. In some cases the normal leg-to-trunk ratio was increased to 0.96 and the tibia-to-femur ratio to 0.80, close to the ideal value.

Patients with Turner dwarfism have a moderate degree of short stature with no other major skeletal deformity. Lengthening of the legs by 15 to 20 cm gives a considerable improvement in both appearance and morale. The delay in ossification and the high rate of complications, however, require careful management in a specialist unit.

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REFERENCES