

Leg lengthening for short stature in Turner's syndrome

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We describe ten patients with Turner's syndrome (karyotype 45, XO) who had leg lengthening for short stature. A high incidence of postoperative complications was encountered and many patients required intramedullary fixation as a salvage procedure. We discuss the reasons for this and highlight the differences between our findings and those of a similar series recently reported. In view of the considerable difficulties encountered, we do not recommend leg lengthening in Turner's syndrome.

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Turner's syndrome is named after Henry Turner who, in 1938, described seven girls displaying the same phenotype of short stature, sexual infantilism, webbed neck and cubitus valgus. Nearly two-thirds of these patients have an XO karyotype, the remainder are mosaics (XO/XX) or have partial deletion of the X chromosome.

Patients with Turner's syndrome are usually active and asymptomatic with a low incidence of mental retardation and a normal life expectancy. The short stature, however, is severe with a final mean loss of height of 25 cm (15.5%) compared with that of the normal population.¹ About two-thirds of this loss is due to shortening of the lower limbs, and although there is no associated functional deficit, limb lengthening is sometimes offered to improve the appearance of these patients.

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Patients and Methods

Between 1989 and 1996 we treated ten patients with the Turner syndrome karyotype. Their mean age was 18 years (13 to 22) and their mean final natural height was 136 cm (134 to 138).

The callotasis method² was used in all, with five patients having bilateral tibial lengthening, one bifocal bilateral tibial lengthening, and four femorotibial cross-lengthening. In one, the tibia was lengthened using an Ilizarov frame, but in the remainder a unilateral leg lengthener such as the Orthofix device (Orthofix, Bussolengo, Italy) was used.

The operative technique consisted of application of the fixator followed by subperiosteal corticotomy. In the cases of tibial lengthening, a segment of fibula was excised and a diastasis screw inserted across the distal tibiofibular joint. The patients with bifocal tibial lengthenings had elongation of tendo Achillis as a prophylactic measure. There was, on average, a delay of eight days before starting distraction. Lengthening was carried out at a mean rate of 1 mm/day, with four turns each day until the required length had been gained, unless complications dictated a variation. Once the target length had been achieved, the frame was locked until regenerate bone of good quality was seen radiologically. The fixator was then dynamised, either by progressive dismantling of the Ilizarov frame or by release of the locking bolt on the Orthofix fixator. All patients underwent vigorous physiotherapy from the first postoperative day to minimise soft-tissue complications and to maintain an adequate range of movement of the joint.

Results

The ten patients had a total of ten femoral and 18 tibial lengthenings. The mean period of treatment was 27 months (9 to 72). The mean increase in height achieved was 9.15 cm (7 to 13.5). In the femur the mean length gained was 5.8 cm (2.3 to 10), an increase of 18%, and in the tibia 7.2 cm (2.7 to 10), an increase of 27%.

Complications were encountered in 90% of the femora and 33.3% of the tibiae (Table I). Nine of the ten lengthened femora fractured, all within one week of removal of the fixator (Fig. 1). In each case, sound bony union was judged to have occurred clinically and radiologically. Six of

Table I. The complications encountered in the ten patients who had leg lengthening for Turner's syndrome

Complication	Femora (n = 10)	Tibiae (n = 18)
Fracture	9	-
Premature fusion of corticotomy	2	-
Plastic deformation of regenerate	1	4
Axial deviation	-	1
Equinus foot	-	5
Subtalar deformity	-	1
Nerve traction injury	-	1
Stiff knee	2	-
Tendon impingement	-	1

these fractures were treated by intramedullary nailing which was delayed until the pin tracks had healed, and three by immobilisation in a spica. All united satisfactorily.

In the tibiae, plastic deformation of the bony regenerate after removal of the external fixator required corrective osteotomy in four patients (Fig. 2). Lengthening of tendo Achillis was required in five.

Discussion

Leg lengthening is being offered increasingly to various groups of patients to correct short stature or inequality of length,³ using either the callotasis method or the Ilizarov technique. These procedures have been reported as being successful giving good correction of length, but with an incidence of complications ranging from 5% according to Ilizarov⁴ to 45% as reported for the method of Wagner.⁵

In Turner's syndrome, patients who are otherwise in good health can expect to be substantially shorter than their peers and leg lengthening may be considered.

Trivella, Brigadoi and Aldegheri⁶ recently described the results of leg lengthening in 16 patients with a Turner-syndrome phenotype. They achieved an overall mean increase in height of 13 cm with a rate of complications of 21% for the femur and 40% for the tibia. The methods used and the complications encountered were similar to ours. While they point out that these cases do have a significant rate of complications and that the patients should be managed in a specialist unit, they recommend short stature in Turner's syndrome as an indication for leg lengthening.

The results from our series have been poor. There were significant complications in 90% of the femora for a similar mean increase in height. These included fractures, deformation of regenerate bone of poor quality and soft-tissue problems associated with prolonged use of the external fixator.

Osteogenesis in patients with the Turner karyotype appears to be abnormal. Our group of patients all had the



Fig. 1a



Fig. 1b

Radiographs a) at the conclusion of lengthening 19 months after application of the fixator, and b) one week later when the femur had fractured. It was then nailed.



Fig. 2a



Fig. 2b

Radiographs a) at the conclusion of lengthening of the tibia and b) two weeks after removal of the frame showing increasing deformation.

true Turner syndrome whereas only nine of the patients described by Trivella et al⁶ were so affected. This would explain the difference in results.

While ultimately successful in providing an increase in height, leg lengthening in patients with Turner's syndrome is a very lengthy procedure with a high incidence of complications and morbidity. We do not recommend it.

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