Lower Limb Lengthening in Turner Dwarfism

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The aim of this study was to review our cases of lower limb lengthening to treat Turner dwarfism, and to speculate whether or not effective limb lengthening can be achieved in this rare condition. Twelve tibias and 2 femora were lengthened in 6 patients using the Ilizarov method for the tibia and a gradual elongation nail for the femur. The mean age at the time of surgery was 19 years, and the patients were followed up for a minimum of 2 years. The average gain in the tibial and femoral length was 6.2 cm and 6.9 cm, respectively. The average healing index of tibia and femur was 1.9 and 1.7 months. The average tibia-to-femur ratio improved from 0.68 preoperatively to 0.81 postoperatively, and leg-trunk ratios improved from 0.88 to 0.99. Seven segments (50.0%) had completed the lengthening protocol without complications. Two segments (14.3%) had an intractable pin site infection requiring a pin exchange, and four segments (35.7%) had twelve complications (a nonunition at the distraction site, premature consolidation, Achilles tendon contractures and planovalgus). The overall rate of complications was 100% for each bone lengthened. All the patients showing a nonunion at the distraction site had a reduced bone mass, which was less than 65% of those of the age-matched normal population. Despite the complications, all patients were satisfied with the results, and lower limb lengthening in Turner Dwarfism believed to be a valid option. However, it may require careful management in a specialist unit in order to prevent complications during the lengthening procedure. In addition, the osteopenia associated with an estrogen deficiency leading to problems in consolidation is a difficult issue to address.

**Key Words:** Turner dwarfism, lower limb lengthening

INTRODUCTION

Turner syndrome represents a spectrum of physical findings in female patients with one X chromosome. The most frequent karyotype is 45XO/46XY and the remainders are either mosaics (XO/XX) or have a partial deletion of the X chromosome. The incidence of this condition is 1 in 5000 live births, and the phenotypic characteristics include sexual infantilism, a webbed neck, cubitus valgus and a short stature. A study of Turner dwarfism documented a 15.5% percent decrease in height when compared with normal women, and the documented shortening in stature was not proportional because two-thirds of this condition occurred in the extremities as opposed to the trunk.\(^3\) Shortening was also greater in the tibia compared to the femur.

Limb lengthening may be useful for patients with a short stature due to skeletal dysplasias, endocrine abnormalities and idiopathic short stature. However, reports of limb lengthening have varied greatly in terms of the quality, the evaluation criteria, the patients’ characteristics (demographic data, diagnosis, and the bone lengthened), the technique of lengthening (unilateral; bilateral or ipsilateral; unifocal or bifocal; and distraction osteogenesis or callotasis), and the type of fixators used (monolateral and circular). Recently, two reports did not recommend lower limb lengthening using the callotasis method with a monolateral external fixator in Turner dwarfism due to the high complication rate.\(^2,3\) For the past fifteen years, this hospital has accumulated an extensive experience in limb lengthening via “distraction osteogenesis”. Therefore, the aim of this study was to review the results and complications of
lower limb lengthening in patients with a dispro-
portionate short stature due to Turner syndrome. A comparison of these results and complications with the available literatures on limb lengthening for other conditions was also made.

MATERIALS AND METHODS

Retrospective analysis was performed on six patients who underwent twelve tibial lengthening and two femoral lengthening procedures from April 1991 until July 1997. Tibial lengthening using the Ilizarov circular frame, and femoral lengthening using a gradual elongation nail were performed. The patients underwent an application of a Ilizarov tibial construct with either four trans-osseous tensioned wires in both the proximal and distal tibia, or with a half-pin proximally and distally, which substituted for the wire traditionally placed parallel to the medial surface of the tibia. The bifocal construct had two middiaphyseal wires affixed to an additional central ring. Tibial and fibular osteotomies were performed by using either an osteotome or a Gigli saw through a small incision with minimal periosteal disruption. Lengthening was usually commenced 5-7 days postoperatively at a rate of 0.25 mm four times daily at each distraction site. In femoral lengthening, the internal osteotomy with an intramedullary saw at a preselected site was undertaken in such a way as to preserve as much of the periosteum as possible. The bone was distracted at the site of the osteotomy before introducing the nail. Five millimeters of lengthening was obtained at the end of the procedure. The distraction was begun on the 7th postoperative day at a rate of 1 mm/day with five ratcheting, three times per day. At the end of the predetermined gain, the rotation and further elongation were automatically blocked by a fixed locking system, which had been set to prevent further movement when the desired lengthening had been achieved.

The preoperative data and lengthening parameters were recorded for each patient; the age at surgery, the number of days of distraction, the amount of time needed for callus maturation, the total times the fixators were used and the total treatment time (fixator use and subsequent immobilization). The occurrence of complications for each segment of a bone lengthened (i.e., femur or tibia) including neurovascular injury, premature consolidation, severe pin tract infection requiring intravenous antibiotics or pin removal, and a nonunion of the distraction site was recorded. In addition, the incidence of secondary surgical procedures was noted including osteoclasis for a premature consolidation, segment manipulation for angulation or fixator exchange, pin removal as a result of an infection, surgical stabilization and bone grafting for a distraction site nonunion, Achilles tendon lengthening for an equinus contracture, and surgery for foot deformities that developed during the lengthening procedures.

All radiographs used in this study were taken using a similar radiographic technique. The films were taken from the same distance using the same X-ray machine. The initial bone length was measured from the preoperative radiograph. The final amount of length gained was assessed from the first radiograph taken during the maturation period. All radiographs were serially examined for a coronal malalignment greater than ten degrees prior to the fixator removal. The radiographs were also reviewed to detect the presence of a fracture or deformation of the lengthened bone after removing the fixator. For each patient, the percentage of bone lengthening was determined by dividing the distance gained by the initial length. The healing index was calculated by dividing the total days of treatment by the total distance gained in centimeters. In the femur, the radiological consolidation endpoints were defined as occurring when the bone in the distraction gap had healed and three of the four cortices had been bridged on the anteroposterior and lateral radiographs. The indices were calculated and analyzed for the lengthened bones that had consolidated without internal stabilization and bone grafting. The follow-up clinical assessment involved an evaluation of the joint motion, the motor and sensory function and the vascular status. In three patients, bone densitometry was performed to evaluate the osteoporosis related to an estrogen deficiency that is often found in Turner syndrome.4

RESULTS

The mean age of the patients at surgery was 19 years (range, 12 to 24). The average height prior to surgery was 140.1 centimeters (range, 130.0 to 150.0) and all patients showed a disproportionate short stature with leg-to-trunk and tibia-to-femur ratios of 0.88 (range, 0.81 to 1.02) and 0.68 (range, 0.62 to 0.76), respectively. The patients were followed for an average of 43 months (range, 24 to 84), and achieved a lengthening with a mean increase in height of 7.9 cm (range, 3.0 to 13.4) and a mean final height of 148.0 centimeters (range, 138.6 to 156.0). The mean tibial and femoral length gained was 6.2 centimeters (range, 3.0 to 7.6), and 6.0 centimeters, respectively. The mean percentage of bone lengthening was 21% (range, 13 to 33) for the tibia and 15% for the femur. The mean leg-to-trunk and tibia-to-femur ratios improved to 0.99 and 0.81, respectively, and the mechanical axis deviation improved from 3.8 centimeters (medial deviation; range, 1.0 to 5.8) preoperatively to 0.6 (medial deviation; range, 0.3 to 1.7) postoperatively. A distraction was performed for an average duration of eighty-six days (range, 60 to 151). The average total time of fixator use was 254 days (range, 129 to 288), and the mean treatment time was 289 days (range, 164 to 343). The mean healing index of the tibia and femur was 1.9 months (range, 1.0 to 2.9) and 1.7 months (1.1, 2.2) per centimeter of length gained, respectively (Table 1). Two representative cases are presented in Fig. 1 and 2.

Complications

Seven segments (one femur and six tibiae, 50.0 percent) had completed the lengthening protocol without any complications (coronal malalignment before fixator removal, deformity of the lengthened bone without fracture after fixator removal, fracture of the lengthened bone, severe pin tract infection, a nonunion of the distraction site; severe pin tract infection, or equinus contracture). Two segments (two tibiae, 14.3 percent) had an intractable pin site infection requiring a pin exchange, respectively. Four segments (one femur and three tibiae, 35.7 percent) had twelve complications (4 nonunions at the distraction site, 4 premature consolidation of the tibia, two Achilles tendon contractures and two planovalgus). They were treated with autoiliac bone grafts, recorticonies, Achilles tendon lengthening and triple arthrodesis, respectively. The overall complication rate was 100 percent for each bone lengthened. All the patients showing a nonunion at the distraction site had 65 percent less bone mass of those of age-matched normal population. No cases of neurovascular injury or significant knee joint contractures were noted after the rehabilitation period.

DISCUSSION

Lower limb lengthening aimed at increasing stature is a topic of great interest and one that triggers a great deal of controversy among ortho-

Table 1. Data on the Patients with Turner Dwarfism

<table>
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<tr>
<th>Patient /Age</th>
<th>Preoperative height (cm)</th>
<th>Preoperative length (right, cm)</th>
<th>Preoperative length (left, cm)</th>
<th>Postoperative length (right, cm)</th>
<th>Postoperative length (left, cm)</th>
<th>Healing index (right, months/cm)</th>
<th>Healing index (left, months/cm)</th>
<th>Postoperative Height (cm)</th>
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<td>39.5 (F)</td>
<td>45.5 (F)</td>
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<td>156.0</td>
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<td></td>
<td></td>
<td>27.9 (T)</td>
<td>27.9 (T)</td>
<td>35.2 (T)</td>
<td>35.3 (T)</td>
<td>2.9 (T)</td>
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<tr>
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<td>29.7 (T)</td>
<td>29.7 (T)</td>
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<td>2.8 (T)</td>
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<td>31.0 (T)</td>
<td>1.1 (T)</td>
<td>1.0 (T)</td>
<td>138.6</td>
</tr>
</tbody>
</table>

*Femur; †Tibia.
Fig. 1. Radiographs of Case 1. (A) preoperative anteroposterior radiograph of the lower extremities showing a disproportionate stature, (B) single-level femoral lengthening using a gradual elongation nail. Note the new bone formation in the distraction site, (C) final anteroposterior radiograph showing a lengthened femora.

Fig. 2. Radiographs of Case 2. (A) preoperative anteroposterior radiograph of the lower extremities showing a disproportionate stature and tibia vara, (B) double-level tibial lengthening using the Ilizarov frame. Note the new bone
paedic surgeons.\(^5\) Lengthening for short stature can provide functional improvement and a cosmetic benefit for patients. Patients who live in societies that do not compensate for shorter people may achieve a greater functional benefit. Previously, Bidwell, et al.\(^2\) and Trivella, et al.\(^3\) managed to achieve limb lengthening with additional procedures besides the index operation, and did not recommend the lengthening procedure in Turner syndrome due to the high complication rates. This study documented the changes in the psychosocial development or health. Preoperatively, a short stature generated difficulties and problems for all of our patients in terms of social adaptation. However, the lengthening procedures could provide the patients with improved appearance and self-esteem.

This series recorded a predictable increase in bone length, for example, the tibiae were lengthened an average of 6.2 centimeters or 21 percent of the original length. This average increase in length is similar to previous reports of limb lengthening in short-statured patients due to other conditions.\(^6\)\(^-\)\(^7\)\(^-\)\(^17\) Although the patients were satisfied with the gains in bone length, this should be balanced by the extended time required for bony consolidation (average treatment time of 289 days and a healing index of fifty-four days for each centimeter of length gained). On the surface, our radiological results appear to be more than thirty-four to forty-two days per centimeter gained in tibial lengthening of other.\(^6\)\(^-\)\(^7\)\(^-\)\(^10\)\(^-\)\(^15\)\(^-\)\(^17\) Trivella, et al. reported longer healing times in their series, possibly due to an intrinsic hormonal imbalance.\(^7\) This study also encountered problems of bony development at the distraction site particularly in those patients whose bone mass was 65 percent of the normal population. In Turner syndrome, an estrogen deficiency and any associated osteopenia may contribute to the slow ossification and delay in the maturation of the cortex. Some commented that dysplasia of the lymphatics and lymph edema in the lower legs may also delay healing.\(^3\)\(^-\)\(^8\) It is believed that osteogenesis in Turner syndrome is abnormal, and the osteoporosis related to this rare syndrome should be treated preoperatively or during the lengthening procedure in order to enhance bone formation.

Previous studies that included the results of distraction osteogenesis for other conditions have reported complication rates ranging from 10.5 to 250 percent per segment of bone lengthened.\(^9\)\(^-\)\(^20\) These variations between publications may be due to differences in the terms of the disease entities, associated angular deformities and the distance that the bones were lengthened. Noonan, et al.\(^20\) noted that femoral and tibial lengthening using a monolateral external fixator, which was performed to treat a short stature of the endocrine and idiopathic etiology, is associated with the higher complication rates (150 percent), and required additional procedures than those performed to treat the short stature of the skeletal dysplasia. The overall complication rate of 100 percent (one complication per one segment of bone lengthened) in our series is quite low, and this was attributed to the use of a circular external fixator rather than a monolateral fixator. Ilizarov frame, which is more versatile in the simultaneous three-dimensional correction of malalignment and shortening, was used to lengthen the tibia. Age is another factor to consider before commencing the lengthening procedure. The femora and tibiae in patients who were fourteen years or older were associated with higher complication rates than those in patients who were younger than fourteen years old.\(^19\) In addition, obtaining the amount of length needed to produce a major increase in stature increases the prevalence of complications and additional procedures. This is a similar problem to that encountered in patients who had a limb-length discrepancy. In our series, there were few complications in the patients whose surgery was performed to correct a malalignment (varus deformities of the proximal tibia) rather than to lengthen the bone itself. Although the previously reported rate for an Achilles tendon contracture after tibial lengthening is quite high, the current rate was somewhat lower than those of most reports even in the two reports of lengthening in Turner syndrome.\(^12\)\(^-\)\(^17\)\(^-\)\(^19\)\(^-\)\(^20\) The differences in these rates might also reflect the different treatment regimes. In the patients with bifocal tibial lengthening, the tendo Achilles was elongated as a prophylactic measure. In addition, in contrast to the monolateral external fixator, the Ilizarov frame has the advantage of being constructed in order to prevent an ankle equinus.
during lengthening, which can enable the patients to participate in a rehabilitation program effectively during the lengthening procedures.

A mechanical intramedullary device, the gradual elongation nail (GEN), was designed as an alternative to an external fixator for the progressive lengthening of the femur. The theoretical advantages are that it is well tolerated, the infection rates are reduced, an axial deviation is avoided, and the internal osteotomy does not disturb the blood supply to the periosteum. We experienced one case of a consolidation defect in which an open osteotomy was necessary to complete the procedure after an incomplete internal osteotomy. In this case, the periosteum was also disrupted with the resulting total destruction of the endosteal and periosteal circulation. This complication may be due to our relative inexperience of using the instrument at the time of surgery as well as early point on the learning curve when using GEN. The periosteum is the major contributor to osteogenesis during a distraction because it is the main source of vascularity. Therefore its careful preservation is essential to bone healing. In our cases, there was no stiffness in the knee joint that is commonly reported after using an external fixator. This may be because the muscle is not transfixed.

In conclusion, lower limb lengthening in patients with dwarfism due to Turner syndrome can be used to improve the disproportionate short stature. However, this involves a lengthy procedure and there is the possibility of difficulties in bony consolidation. Further work needs to be done in order to reduce the complication rates in patients with osteoporosis. In addition, it is suggested that the lengthening procedure in Turner syndrome should be conducted in a specialist unit to reduce the high morbidity rate. This will provide more benefits to the patients when taking into account the costs associated with the concurrent complications.

REFERENCES