

Удлинение конечностей при синдроме Turner

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Мы сообщаем о результатах и осложнениях у восьми пациентов, которым производили удлинение обеих голени по поводу низкого роста при синдроме Turner. Применялся метод distractionного остеосинтеза монолатеральным наружным фиксатором. Голень удлиняли в среднем на 9,2 см или на 33% от ее исходной длины. Общий период лечения в среднем составлял 268 дней; осложнения отмечались в 169%, что потребовало в среднем по 1,7 дополнительных вмешательств на каждом сегменте. В семи случаях (44%) потребовалось удлинение ахиллова сухожилия, а в 9-ти случаях (56%) развилась угловая деформация до или после снятия фиксатора; для 6-ти из этих сегментов потребовалась корригирующая остеотомия из-за осевой деформации. В двух случаях (12,5%) отмечалось несращение в зоне distraction, в результате чего потребовалась фиксация пластиной и костная трансплантация. При лечении данной серии пациентов мы пришли к выводу, что удлинение голени методом distractionного остеосинтеза можно использовать при непропорционально низком росте у пациентов с синдромом Turner. Однако, преимущества косметического увеличения роста не компенсируют высокий процент осложнений по сравнению с ранее опубликованными результатами distractionного остеогенеза у пациентов с разницей в длине нижних конечностей или дисплазией.

Ключевые слова: голень, низкий рост, синдром Turner, distractionный остеосинтез.

INTRODUCTION

Limb lengthening via distraction osteogenesis can be utilized for the treatment of limb length discrepancy for differences in length that are greater than five centimeters at maturity. In addition, limb lengthening can be useful for patients with short stature due to skeletal dysplasias, endocrine abnormalities and short stature. In the later cases, lengthening for short stature can provide a functional improvement for patients. Greater functional benefit may be received by those patients who live in societies that do not compensate for the short statured person. Increased stature in affected individuals will facilitate the use of many conveniences such as public restroom's, telephones and automobiles. In addition, limb lengthening may potentially provide a cosmetic benefit in patients with short stature due to disproportionate shortening.

Turner syndrome represents a spectrum of physical findings in those female patients with one X chromosome. The incidence of this condition is 1 in 5000 live births [20]. Phenotypic characteristics in a female include web neck often with a low hairline, coarctation of the aorta, primary amenorrhea, and

short stature. A study of Turner dwarfism in ten patients documented a decrease in height of 15.5 per cent when compared with normal women [3]. The documented shortening in stature was not proportional as two thirds occurred in the extremities as opposed to the trunk. Shortening was also greater in the tibia as compared to the femur [3].

For the last seventeen years, the Clinica Universitaria of Navarra (C.U.N.) in Pamplona, Spain has accumulated an extensive experience in limb lengthening via distraction osteogenesis. At this institution, limb lengthening has been utilized for European and South American patients with limb length discrepancies as well as for those patients with short stature. At the C.U.N. several patients have undergone tibial distraction osteogenesis for disproportionate short stature due to Turner syndrome. The purpose of this study is to review the results and complications of tibial lengthening in patients with disproportionate short stature due to Turner syndrome. Comparison of these results and complications with the available literature for tibial distraction osteogenesis for other conditions will be made.

MATERIALS AND METHODS

Retrospective analysis was performed on eight patients that underwent sixteen tibial lengthenings at the C.U.N. from November 1987 until September 1991. For each patient we recorded preoperative and lengthening parameters including age at surgery, diagnosis, waiting period, number of days of distraction, the amount of time needed for callus

maturation, total time of fixator use and total treatment time (fixator use and subsequent immobilization). We recorded the occurrence of complications including neurovascular injury, premature tibial or fibular consolidation, severe pin tract infection requiring intravenous antibiotics or removal, and nonunion of the distraction site. Additionally we

noted the prevalence of secondary operative procedures including osteoclasia for premature tibial or fibular consolidation, segment manipulation for angulation or fixator exchange, pin removal for infection, operative stabilization for fracture of the lengthened bone, operative stabilization and bone grafting for distraction site nonunion, achilles lengthening for equinus contracture, knee manipulation or arthrotomy for contracture or subluxation, and delayed corrective osteotomy for unacceptable residual malalignment.

All radiographs used in this study were taken with similar radiographic technique from the same distance with the same X-ray machine. The initial bone length was measured from the preoperative radiograph. The final amount of length gained was measured from the first radiograph taken during the maturation period. All radiographs were serially examined for coronal axial malalignment greater than 10 degrees produced before fixator removal, sagittal malalignment was not assessed due to the overlapping projection of the fixator on lateral radiographs. Radiographs were also reviewed to detect the presence of fracture or deformation of the lengthened bone after fixator removal. Fractures were distinguished from deformation on the presence of a visible fracture line.

For each patient, we determined the percentage of tibia lengthened from the distance gained divided by the initial tibial length. The rate of distraction in millimeters per day was calculated by dividing the distance gained by the number of days of distraction. The healing index [8] was calculated by dividing the total days of treatment (fixator use and subsequent immobilization) by the total distance gained in centimeters. Indices were only calculated and analyzed for lengthened tibias that consolidated without internal stabilization and bone grafting for distraction site nonunion.

Surgical Procedure and Lengthening Protocol

All cases were managed by members of the Department of Orthopaedics and Traumatology of C.U.N. with the same general protocol. No instances of bifocal lengthening were performed. In each procedure, the fixator was placed prior to osteotomy on the anteromedial aspect of the tibia. In the first twelve cases the Wagner external fixator was used, and in the later four cases the Monotube external fixator (Howmedica, Rutherford, New Jersey) was used. In all instances, two pins were placed proximally and distally. Percutaneous tibial osteotomy was performed as previously described [4]. All osteotomies were performed in the proximal metaphyseal region, no osteotomies were performed in the distal 3/4's of the tibia. The distal fibula was

always stabilized either from the tibial side with an external fixator pin or with a screw placed from the lateral aspect of the leg. A portion of fibula was routinely resected for one to three centimeters at the junction of the distal and middle one third of the fibular diaphysis.

The lengthening protocol closely followed that as previously described [8]. After three to five days of hospitalization, patients were discharged and encouraged to ambulate with partial weight bearing when possible. Following the waiting period, distraction was initiated at the rate of one millimeter per day in two 0.5 millimeter increments. Daily pin care was performed with soap and water. Occasionally inflamed and painful pins that were resistant to local care required short courses of oral antibiotics. In some cases, pin infection necessitated intravenous antibiotics or surgical removal and replacement. During the distraction period, patients returned to the clinic every two to three weeks for routine physical and radiographic examination. All patients were directed to keep their knee extended as much as possible; in addition, patients were uniformly prescribed an ankle foot orthosis to be worn at night. These measures were recommended to decrease the risk of knee subluxation or flexion contracture and achilles contracture. No attempts were made to promote knee flexion or aggressive ankle range of motion during the distraction period.

Lengthening continued until the desired distance was obtained or an additional procedure was required secondary to complications. When the final length was obtained, the fixators were locked and patients were encouraged to increase their activities and weight bearing. At this point, patients were prescribed range of motion exercises to begin regaining knee and ankle motion that was lost during the distraction stage. To additionally improve bone maturation, external fixators were dynamized in those segments treated with Monotube devices. Fixators were removed when good cortical continuity was seen in three out of four cortices on anteroposterior and lateral radiographs. The devices were routinely removed under anesthesia and twelve segments had further immobilization in a cast or splint. The later segments were immobilized for an average of sixty-two days (range, thirty to eighty days). Following the lengthening protocol and complete healing of the distraction site; all patients were followed for secondary problems such as residual joint stiffness, malalignment or continued inequality.

A representative case example is presented in Figure 1.



Figure 1. Tibia lengthening in Turner syndrome: a) left side, 3 weeks post-op, b) right side, 3 weeks post-op, c) left side, end of lengthening, d) right side, end of the lengthening phase.



e

Figure 1 (continue). Tibia lengthening in Turner syndrome:
e) 1 year after the external fixation removal.

RESULTS

Sixteen tibias from eight female patients were simultaneously lengthened (symmetric lengthening). The mean age of the patient at surgery for each segment was 16.1 years (range, thirteen to twenty years). The average waiting period before onset of distraction was 11 days (range, eight to fifteen days). Distraction was performed at an average rate of 1.0 millimeters per day (range, 0.4 to 1.4 millimeters per day) for an average duration of the distraction period lasting ninety-four days (range, sixty to 146 days). The average total time of fixator use was 215 days (range, 170 to 261 days); and the mean total treatment time was 268 days (range, 227 to 300 days). All patients were followed for an average of 4.3 years (range, 2.7 to 5.8 years). The mean distance gained was 9.2 centimeters (range, 6.5 to 12.0 centimeters) and the average percentage of length gained was 33 per cent of the original tibial length (range, 19.7 to 72.7 per cent). The mean healing index was thirty-one days of total treatment time per centimeter of length gained (range, twenty-three to forty days of total treatment time per centimeter of length gained).

Complications

Three segments (18.8 per cent) had completion of 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; nonunion of the distraction site; severe knee joint contracture or subluxation; and equinus contracture]. Two segments (12.5 per cent) had one complication, eight segments (50 per cent) had two complications, and three segments (18.8 per cent) had three complications. The overall rate of complications was 169 per

Table 1

Total Complications		
	Frequency	Percentage
Axial Malalignment *	7	43.8
before Fixator Removal		
Fracture of Lengthened Bone	2	12.5
Callus Deformation without Fracture after Fixator Removal	2	12.5
Severe Pin Site Infection	7	43.8
Nonunion of Distraction Site	2	12.5
Equinus Contracture	7	43.8
Total Complications	27	169

*All cases of malalignment consisted of valgus angulation. no cases of varus angulation were noted

cent for each tibia lengthened via distraction osteogenesis. The specific frequency for each complication is presented in Table 1. All cases of coronal tibial malalignment produced prior to fixator removal were due to valgus deviation, no cases of varus malalignment were noted. No cases of fibular consolidation or tibial premature consolidation were noted. No cases of neurovascular injury were recorded in these eight patients. All fractures of the lengthened bone and the two cases of callus deformity without fracture after fixator removal were treated with cast immobilization. No significant knee joint contractures were noted after the normal rehabilitative period.

Three segments (18.8 per cent) had completion of the treatment protocol without any associated operative intervention (excluding fixator removal). Two segments (12.5 per cent) required one additional operative procedure, eight cases (50 per cent) required two additional procedures, and three segments (18.8 per cent) required three additional procedures. The overall rate of associated proce-

dures was 169 per cent for each segment lengthened. The rates of each surgical intervention is additionally presented in Table 2. All seven cases which had had achilles lengthening required this during or after the distraction period, no prophylactic lengthenings were performed.

Table 2

Total Associated Procedures		
	Frequency	Percentage
Pin Removal for Infection	7	43.8
Manipulation for Axial Malalignment	5	31.3
Corrective Osteotomy for Residual Malalignment	6	37.5
Achilles Lengthening	7	43.8
Internal Fixation and Bone Graft for Nonunion	2	12.5
Total Associated Procedures	27	169

DISCUSSION

In the current study: we review the results, complications and additional procedures from a consecutive series of sixteen tibias from eight patients with Turner syndrome lengthened via distraction osteogenesis. The indications for surgery in these patients reflect cultural and societal differences at odds with the peoples of North America. In European and South American countries, life for the short stature patient is more difficult as society does little to accommodate to their needs [15, 19]. Previously, Trivella et al reported the results of lengthening in patients with Turner dwarfism and concluded that lengthening in these patients corrected the disproportionate length discrepancies and improved appearance and self-esteem [22]. In the current study we did not attempt to document any changes in psychosocial development or health, however we did record a predictable increase in tibial length. In this series, tibias were lengthened an average of 9.2 centimeters or 33 per cent of the original bone length. The average increase in length is similar to previous reviews of tibial lengthenings in patients with short stature due to achondroplasia which report increases in length of 14.5 to 58 per cent [1, 8, 16]. Although gains in length are impressive, these results must be balanced by the extended time required for bone consolidation (average treatment time of 268 days) and the presence of complications and associated procedures.

In the current investigation of sixteen tibial lengthenings we noted twenty-seven complications consisting of malalignment before fixator removal; deformity of the lengthened bone without fracture after fixator removal; fracture of the lengthened

bone; severe pin tract infection; nonunion of the distraction site; and equinus contracture. On average, each tibia had a complication 169 per cent of the time. Previously published complications rates from papers that include tibial distraction osteogenesis report prevalence of complications of 10.5 to 250 per cent [2, 5-7, 9, 10, 12-14, 17, 18, 21, 22]. Variations between the current and previous publications are most likely due to differences in classification of complications, patient diagnosis, associated limb deformities and the distance that tibias were lengthened. Additional procedures for the treatment of complications or delayed corrective osteotomies for any residual malalignment were documented in twenty-seven instances. On average, each tibia lengthened required 1.7 additional operations to optimize the outcome of the limb.

Published papers have also recorded the prevalence of ankle or knee contractures requiring operative intervention in 8.5 to 69 per cent of cases [5-7, 9, 10, 12-14, 17, 18, 21, 22]. In this series, we document a high rate of achilles contracture; approximately 44 per cent of tibias lengthened required an achilles lengthening. Although the previously reported range for these problems is quite large; the current rate is somewhat higher than most reports and is likely due to the relatively larger distances gained. Differences in these rates may also reflect different treatment philosophies. At the C.U.N., achilles lengthening is considered a relatively minor procedure that permits greater tibial lengthenings and allowing plantigrade foot positioning.

Previously, Trivelli et al reported the results of femoral and tibial lengthenings in sixteen patients

with Turner dwarfism due to Turner syndrome, mixed gonadal dysgenesis and Noonan syndrome. In the prior study, they found longer healing times possibly due to an intrinsic hormonal imbalance. In the current study the healing index was thirty-one days of total treatment time for each centimeter of length gained. On the surface, this value may seem equal to rates of thirty-four to forty-two days per centimeter gained in other studies of tibial distraction osteogenesis [10, 13, 17, 18, 21]. However we noted problems of bony development with fracture, callus deformation or nonunion of the distraction site in 37.5 per cent of patients. These findings confirm the conclusions reached by Trivella et al who found a high complication rate in patients with Turner Dwarfism that underwent distraction osteo-

genesis [22].

In conclusion, tibial distraction osteogenesis in patients with dwarfism due to Turner Syndrome can be used to improve the disproportionate discrepancy. In the current report we lengthened tibias 33 per cent of the original length at the expense of high rates of complications and associated procedures. Further work needs to be done to reduce the rates of these problems in all patients who undergo limb lengthening. In particular efforts should be made to determine the psychosocial and functional benefits of limb lengthening in patients with short stature. The later will allow a more critical analysis of the benefit of these procedures in light of the cost associated with concurrent complications.

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