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Original Article

Bilateral double level tibial lengthening in dwarfism☆

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ABSTRACT

Purpose: Outcome assessment after double level tibial lengthening in patients with dwarfism.

Methods: Fourteen patients with dwarfism were analyzed after bilateral simultaneous double level tibial lengthening.

Results: Average age was 15.1 years. Average lengthening was 13.5 cm. The two levels were lengthened by an average of 7.5 cm proximally and 6.0 cm distally. Concomitant deformities were also addressed during lengthening. External fixation treatment time averaged 8.8 months. Healing index averaged 0.7 months/cm.

Conclusion: Bilateral tibial lengthening for dwarfism is difficult, but the results are usually quite gratifying.

1. Introduction

People born with various forms of dwarfing conditions may be afflicted with a variety of medical problems.[1] However, they also can suffer from psychological disturbances related to their short stature, and to limitations of routine daily activities such as require reaching high objects, using public restrooms, using pay phones, driving cars, or taking food from a salad bar. For these reasons, a certain segment of the dwarf population is interested in extended limb lengthening.[2]

There is an existing, limited, literature on limb lengthening for short stature.[3–18] Most of the studies cited have done relatively modest degrees of lengthening, and most do not include any type of evaluation scale to measure outcome.

Double level lengthening is particularly useful when there are angular deformities at the knee and/or ankle. Another potential benefit of double level lengthening is the ability to

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achieve more length in less time. We have been using double level tibial lengthening for selected cases of dwarfism. We now report our initial results with this technique.

2. Materials and methods

We reviewed the charts and radiographs of 14 consecutive skeletal dysplasia patients who underwent double level tibial lengthenings. Short stature patients who underwent single level tibial lengthenings were not included.

Radiographs were measured before surgery, at the time of removal and at follow-up. Measurements made included the following: preoperative length, postoperative length, medial proximal tibial angle (MPTA), lateral distal femoral angle (LDFA), joint line convergence angle (JLCA), lateral distal tibial angle (LDTA), posterior proximal tibial angle (PPTA), anterior distal tibial angle (ADTA), and mechanical axis deviation (MAD).

Outcomes were assessed by a grading scale that considered the following factors: ankle range of motion; lengthening achieved; gait change; clinical deformity; pain; and activity level. This was modified from a previous scale used by our group. A maximum of twenty-five points were awarded for excellent results in each of the first four categories, and a maximum of 30 points could be deducted for poor results in the last two categories. Thus, the maximum outcome was 100 points. An overall score was generated from these six categories that was graded as follows: excellent = 95 to 100 points, good = 75 to 94 points, fair = 40 to 74 points, and poor <40 points.

This grading system was adapted from a system we devised to study tibial lengthening outcomes. In each category, there is a possibility of rating as excellent, good, fair, or poor. For example, under the category "gait", an excellent result (25 points) means either no limp before or after lengthening, or a limp before lengthening that disappeared after lengthening. A good result (20 points) is improvement in a moderate limp to a mild limp, or no change in a mild limp. A fair result (10 points) is moving down a grade from no limp before surgery to a mild limp after lengthening, or from mild limp to moderate limp. A poor (0 points) result in the limp category is downgraded two levels. Similar scales were devised for the other categories (Details in Table 1).

3. Results

Fourteen patients with dwarfism underwent bilateral simultaneous double level tibial lengthening for stature. Average age was 15.1 years (range; 11.3–24 years). Diagnoses included achondroplasia (8), hypochondroplasia (3), metaphyseal chondrodysplasia (2), spondyloepiphyseal dysplasia (1). All were lengthened with the Ilizarov frame. In every case, we extended the tibial frame to include a heel ring to fix the ankle in neutral position. The fibula was cut at one level in 6/14 patients, and at two levels in 5/14. In 3/14 patients, the fibula was not cut, because it had been resected previously at another institution. (This is the Kopits procedure, used to prevent tibia vara in achondroplasia.[1]

Average lengthening was 13.5 cm (range 10–16 cm) and percent lengthening was 69% (range; 33–110%). The two levels were lengthened asymmetrically, by an average of 7.5 cm proximally and 6.0 cm distally as the consolidation distally tends to proceed in a slower rate (Choi1999). Nineteen tibias exhibited some preoperative angular deformities that were also addressed with the lengthening. 8/14 underwent intentional "pull-down" of the head of the fibula, to tighten the lateral collateral ligament.[19] The average distance these fibulas descended was 17 mm (range; 11–39). External fixation treatment time averaged 8.8 months (range; 5.5–14.6 months). Healing index averaged 0.7 months/cm (range; 0.5–0.9 months/cm). Follow-up time after frame removal averaged 2.0 years (range; 0.4–5.2 years).

Complications were many, and will be described by category.[20] Peroneal nerve signs or symptoms developed in 10 patients (20 tibias). Our first response was to slow down the rate of distraction, and this was successful in restoring normal nerve function in 4 tibias. The others (16 in 9 patients) all required surgical decompression of the peroneal nerve at the neck of the fibula, and into the anterior compartment. In the first half of the study, we were monitoring for nerve stretch injuries by clinical examination.

Confirmation of significant injury was obtained with near nerve conduction velocity measurements. In the latter half of the study, patients were monitored regularly (every two weeks) with quantitative sensory testing in the feet to detect early changes in static two-point discrimination, the "PSSD" (Pressure Specified Sensory Device). The specifics of these measurements have been reported elsewhere.[21] One patient, after acute corrections of bilateral 10° supramalleolar deformity, developed tarsal tunnel syndrome, which resolved with prompt tarsal tunnel decompression.

Other complications included premature consolidation of the tibia or fibula in two patients (three legs; fibula bilaterally in 1 patient, one tibia on another patient). This required repeat corticotomy in all three legs. Three other patients had "impending premature" consolidation. These potential precocious consolidations were thwarted by increasing the rate of distraction.

Knee contractures were generally temporary and mild, but in five patients required additional intensive physiotherapy to resolve, beyond the ususal amount. No patient lost knee motion at follow-up. Unanticipated angular deviation occurred during lengthening in 13 tibias, and was treated by frame modification and adjustment in the out-patient clinic. These included proximal procurvatum in 9 tibias, distal procurvatum in 4 tibias, proximal in 1 tibia, proximal valgus in 1 tibia, and distal valgus in 1 tibia (Some tibias had more than one deformity).

Pooling all patients together, the average MPTA pre-op was 87°, and post-op was 89°. The average LDTA went from 94° preoperatively to 88° postoperatively. The average PPTA changed from a pre-op value of 85°–84° at follow-up. The average ADTA went from 87° pre-op to 89° post-op. Our radiographic follow-up showed mild residual deviations in the LDTA in 11/28 tibias. Similar mild residual deviations were seen in the MPTA in 8/28 tibias. In the sagittal plane radiographs were available for review on 25/28 tibias and showed mild deviations from standard norms in the ADTA in 18/25 tibias and in the