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Original Article

Bilateral double level tibial lengthening in dwarfism

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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. However, they also can suffer from psychological disturbances related to their short stature, and to limitations of routine daily activities such as requiring 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. There is an existing, limited, literature on limb lengthening for short stature. 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...
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 (LDTA), 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 (20 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.)

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.11 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 precarious 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 usual 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

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tibias and in the PPTA in 9/25 tibias. Note should be made that significant deviations were present pre-operatively in most patients.

Pin tract infections were noted in all fourteen patients, but none required operative treatment or hospitalization. All resolved with either oral antibiotics, minimal pin debridement in the out-patient clinic, or pin removal (for those that occurred late in the treatment, and could be removed without substitution and without jeopardizing the stability of the frame. Fracture after frame removal occurred in 3 tibias. All occurred within the first month in all three.

Complications involving ankle function were by far the most vexing, leading to downgrading of results in two patients (both fair). Unilateral rupture versus surgical injury of the tibialis anterior tendons occurred in one patient. It is not clear exactly how and when this happened, but may have been related to the Gigli saw osteotomy that was used to cut the distal tibia. Another possibility is spontaneous rupture during lengthening. At the time of her frame removal, percutaneous pin releases were performed in the distal tibias, and it is conceivable that the tendons could have been surgically cut at that time. This patient was treated with tendon reconstruction, but remains with a partial drop foot, compensated for with the Extensor Hallucis Longus. Despite this, she ambulates with no limp, and has no sensory deficits.

The other patient with bilateral ankle sequelae, a 14 year old girl with hypochondroplasia, may have had compression of her ankle joints despite protection with a foot frame after relatively modest 10 cm lengthenings. She has good strength (despite a TAL), but has less than half of her pre-operative ankle motion. She has somewhat less function now than pre-operatively (she has given up some sports activities), and recently underwent reconstructive surgery to restore mobility. At surgery she had an anterior ankle release, debridement of anterior osteophyte that was blocking dorsiflexion, and a supramalleolar 10° extension osteotomy. (She had developed some procurvatum of her ankles; ADTA = 88°–90°, compared to pre-op values of 84°–86°. At her most recent surgery follow up, ankle total arc of motion improved from 20° to 60°.

While nearly all patients had strong triceps surae by manual muscle grade testing (5/5), three had persistent weakness as tested by the Jacqueline Perry single stance toe rise test. Three of these patients are less than 18 months post-operative, and are improving. Persistent push off weakness of the gastrocsoleus two years after frame removal was seen in both of the only two patients who had heel cord lengthenings done at the time of frame removal. We no longer recommend heel cord lengthening for stature lengthening in dwarfism. We observed similar weakness (by Perry test) in three patients who were relatively non-compliant with their physical therapy program. These patients are all less than 18 months after frame removal, and may still improve.

One 13 year old girl with metaphysal chondropatplasia developed unilateral mild anterior ankle subluxation, despite protection with a foot frame. The subluxation was treated by acute adjustment of the ankle frame, and she remains well reduced and mobile, one year after frame removal, although she has decreased gastrocsoleus strength after her 15.5 cm lengthenings.

One 16 year old achondroplastic patient did well for one year after a 15 cm lengthening, with an excellent result. She had an elective spinal procedure at another hospital for thoracolumbar kyphosis, unrelated to her lengthening. Unfortunately, that surgery was complicated by permanent paraplegia.

Grading of outcomes was done with a modified instrument that we devised for evaluation of results in tibial lengthening. By this scale, we had nine excellent, three good, one fair, and one poor result. By patients’ subjective reporting, 13/14 were happy with the outcome. The only patient that was not overly happy is the achondroplastic girl, currently age 18, who wound up with stiff ankles and occasional pain. She has given up sports, and underwent surgery to improve her ankle

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motion. Eleven of the 14 patients have been offered to undergo additional lengthenings, and so date, 9 of 11 have returned to undergo additional lengthenings done (humerus, femur). The other two are planned.

4. Discussion

Extended limb lengthening in dwarfism is controversial, and the patient support group Little People of America, does not condone the procedure (http://www.lpaonline.org/index.php?option=com_content&view=article&id=72). Nonetheless, a limited number of centers around the world have employed various techniques for stature lengthening in achondroplasia and other forms of dwarfism.\(^3,6,8,10,13,14,16,18,22\) The older methods of Wagner\(^4\) have been supplanted by the newer techniques of Di Bastiani and Ilizarov.\(^24,25\)

Trivella et al\(^15\) reported on callus distraction (callotasis) and epiphysseolysis (chondrodiatasis) as techniques for lengthening in dwarfism. The performed 49 tibial and femoral lengthenings by callotasis, and 52 tibial femoral lengthening by chondrodiatasis. Their mean lengthenings were relatively modest (and all single level), at 7.8 cm (26%) for the femur with callotasis, 7.6 cm (34%) for the tibia with callotasis, 7.7 cm (33%) for the femur with chondrodiatasis, and 7.2 cm (36.3%) for the tibia with chondrodiatasis. While they dutifully reported their complications, they did not have any outcome measurement or grading system.

The same group published another analysis of lengthenings in 61 achondroplasia and hypochondroplasia patients in which the femur and tibia of the same limb were lengthened simultaneously with unilateral fixators. They compared four techniques: transverse osteotomy, oblique osteotomy, callotasis of the shaft, and chondrodiatasis of the epiphysis. They concluded that chondrodiatasis of the femur and callotasis of the tibia gave the fewest complications. As in most reports, there is no grading scale or outcome measurement.

The same group\(^6\) reported on a group of 16 Turner’s syndrome patients that underwent 32 femoral and tibial lengthenings, using the crossover method of ipsilateral tibia and contralateral femur lengthening. Average length gain was 6 cm (17%) in the tibia and 7 cm (25%) in the femur. Serious complications included AVN of the femoral head in one, and stiff ankle in another. The healing index in Turner’s patients was relatively long at 1.5 months/cm.

Price\(^12\) attempted to reproduce the Italian experience in North America, and became disenchanged after three patients, and abandoned it, concluding that “… the functional benefits may not justify the expense, morbidity, and risk of the procedure.”

Vilarrubias in Spain\(^17\) lengthened 104 patients (208 tibiae and 156 femora). In his protocol, all patients had percutaneous Achilles tenotomy. Lengthenings were in the range of 15–17 cm in the tibia and 15–17 cm in the femora. Regrettably, there is once again, no outcome scores or grading system. He did report a smattering of complications, including reduction of ankle motion (6), valgus deviation (15), varus deviation (1), and procurvatum (8). Interestingly, he found that the hyperlordosis spontaneously corrected during femoral lengthening, presumably from the pull of the hamstrings on the ischium. Yasi and co-workers\(^18\) lengthened 35 patients with achondroplasia and 7 hypochondroplastics using unilateral fixators. Mean age was 14.5 years (range; 10–18) and follow up period 3.2 years (range; 2.0–5.8 years). Average lengthening was 7.1 cm (range; 4.5–13 cm) in the tibia. To his credit, Yasi tested function by physical strength test testing. Strength was rated well in spite of many mechanical axes that were not necessarily good. This group had four ankle contractures, no nerve palsies, and 5 fractures.

Cattaneo et al\(^11\) used the Ilizarov in 23 achondroplasia patients age from 10 to 22 years. They did 44 lower extremity lengthening and 4 humeral lengthening, achieving 14–18 cm in the legs. Complications included bowing and valgus deformity of the tibia, and equinus deformity of the foot. Unlike most other studies, they did have functional outcome scale, albeit loosely defined.

Ganel et al\(^3\) presented some very intriguing data on the effect of age on lengthening in achondroplasia. He lengthened 7 girls (age; 6.5–13.5 years; ave. 9.5 years), and 5 boys (age; 3.5 to 13.0; ave. 8.0 years). 6/7 girls had growth retardation, whereas 4/5 boys had growth acceleration! They concluded that achondroplasts should not be lengthened before age 8 years because they cooperate poorly in physiotherapy, and that lengthening in girls should be delayed until near skeletal maturity.

In a similar technique to our study Choi et al\(^26\) performed double level tibial lengthenings (not generally bilateral). He addresses mainly the factor of reducing treatment time by double level lengthening and the importance to distract the distal site at a slower rate.

In a more recent study, Venkatesh et al\(^16\) reported their results of 40 femoral lengthenings in 20 achondroplastic patients. They focused on differences in callus formation in patients who had more or less than 50% lengthening of their initial femoral length. They found a significant correlation between increased incidence of joint stiffness and regenerate bone fracture and the amount of lengthening.

Novikov\(^17\) from the Ilizarov center in Kurgan published a paper in 2014 with a different approach. He focused on the complications and divided them into soft tissue related, bone related, and functional and subjective clinical complications. He included 131 patients with bilateral limb lengthening. He identified 48 patients with a total of 59 complications, 37 related to soft tissue complications (17/37 needed surgical intervention), and 22 bone related (16/22 needed surgical intervention). Regarding the outcome, he found excellent results for 72 patients (55%), good for 52 (40%), satisfactory for six (4.5%), and poor for one patient (0.77%). Novikov concludes that limb lengthening with the Ilizarov external fixator is a possibility for carefully selected compliant patients with awareness of the risks associated with this technique.

The issue of peroneal nerve injury is an important one to discuss. Most series do not mention this as a problem, or only sporadic cases. In our series, 10/14 patients had some clinical signs of peroneal nerve stretch injury, ranging from dysesthesias to weakness. We have taken a very aggressive approach in the diagnosis and management of these problems, and have been able to complete our goals of lengthening despite the risk to the nerve. We have employed Wagner’s concept of nerve decompression by incising the fascia over the nerve at the neck of the fibula, into the anterior and lateral

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compartment.22 We are not completely alone in this thinking. Galardi et al24 followed five achondroplasts who underwent bilateral Ilizarov tibial lengthening electrophysiologic testing. They found electrical, if not clinical evidence of nerve stretch injury and muscle damage in all 10 limbs. As with our series, the onset of the signs are not directly related to amount lengthened. Polo et al12 did a similar study and found electrical changes in the peroneal nerve in 14 dwarves undergoing crossover lengthenings with unilateral fixators on the femur and tibia. Not all completely recovered. Young et al23 found similar findings in tibial lengthenings in children.

Lee et al50 found changes in somatosensory-evoked potentials in limb lengthening in rabbits tibiae. Strong et al51 also studied an animal model and documented nerve stretch injuries all the way up to the lumbar plexus in canine femoral lengthening. MRIs of the spinal cord were normal, however.

In summary, we found that bilateral tibial lengthening for dwarfism is difficult, but the results are usually quite gratifying. Utmost attention must be directed to factors affecting ankle strength and motion, as this was the commonest cause of diminishment in outcome scores. We advise against prophylactic heel cord lengthening, as both patients in our series who had this developed persistent weakness in the triceps surae. The high incidence of peroneal nerve stretch injuries demands vigilence, but does not preclude achieving the goals of lengthening, provided appropriate nerve decompressions are done. All told, 13 of 14 patients in our series are very happy with the results. The un-happy patient has experienced a functional decline after surgery related to ankle stiffness. This may be salvaged by recent reconstructive ankle surgery, but it is too soon to know the outcome.

More recently, we have abandoned the double level tibia strategy unless there are distinct proximal and distal tibial deformities that demand correction. Instead, we have opted for bilateral single level tibial lengthening, with simultaneous bilateral single level femoral lengthening. The results of this strategy will be reported in a future work, but our impressions are that we see much less peroneal nerve problems with the newer strategy. The same length may be achieved in one treatment, but spread out over the entire limb (tibia + femur), rather than focused solely on the tibia.

Conflicts of interest
All authors have none to declare.

Uncited reference

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REFERENCES


