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TIBIAL LENGTHENING IN PATIENTS WITH ACHONDROPLASIA BY ILIZAROV METHOD

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Abstract
Surgical lengthening of the limbs is invasive, dangerous, and long-term. It is associated with high risks. Moreover, it requires a special psychological approach on the part of the patients and their families.
However, it can be a reasonable option for patients with achondroplasia, not only for improving height but also improving self-esteem and quality of life.

Key Words: Achondroplasia, Ilizarov, Tibial Lengthening

INTRODUCTION

Achondroplasia is the most frequently encountered form of non-lethal skeletal dysplasia and is characterized by defective enchondral ossification owing to a defective gene encoding for fibroblast growth factor receptor 3. It is a type of rhizomelic dwarfism with an incidence of approximately 1 in 10,000 live births. [1]

Achondroplasia is associated with physical and psychological handicaps. [2] These patients may also have symptomatic mal-alignment of the lower limbs. [3] For these reasons many achondroplasts approaching maturity ask to have their limbs lengthened. [4]

Limb lengthening remains controversial in patients with achondroplasia, as it is associated with a high complication rate. Many authors suggest a goal of lengthening of a bone segment to 20% of its original length. [5,6] Improved techniques and understanding of distraction osteogenesis in limb lengthening has led to an increase in the number of limb lengthening procedures and more successful outcomes. [7, 8, 9, 10, 11]

PATIENTS AND METHODS

Patients

Between January 2017 and June 2018, a prospective study was done on 20 patients (40 tibiae) with achondroplasia who underwent bilateral tibial lengthening procedures. They were treated by bilateral tibial lengthening by Ilizarov method.

All patients were diagnosed with achondroplasia as proven by genetic evaluation. The average ages of the patients were 7.2 years (range: 5.5 – 14.5 years). The study population consisted of 6 males and 14 females. Any patient with achondroplasia who had joint pain related to abnormal limb alignment and functional deficit was included in our study.

Any patient with achondroplasia who had the following criteria was excluded from the study: ages less than 5 years, and patients who had previous injuries involving the lower extremities. The minimum follow-up period was 6 months after frame removal.

Preoperative evaluation

At the time of the initial visit, all patients showed short stature. Prior to lengthening, all patients underwent a thorough orthopaedic and neurologic evaluation with special emphasis on the joint range of motion and stability. On physical examination, height was clinically measured using a measuring tape. Preoperative radiological evaluation included anteroposterior and lateral radiographs of both lower limbs taken in standing position with patellae facing forwards. (Figure 1)

A **B**

Fig. 1: Frontal plane angles and normal values relative to the mechanical and anatomical axes (A), and evaluation of sagittal plane limb alignment (B).

Surgical technique

All procedures were performed under general anaesthesia, and patients were positioned in a supine position. A broad spectrum antibiotic was preoperatively administered via an IV route. Pre-assembly of the Ilizarov frame was done after matching the appropriate size of the Ilizarov rings to the size of the patient's leg. (Figure 2)

Fig. 2: A clinical photograph showing a pre-assembled Ilizarov frame.

The upper ring "reference ring" was mounted to the bone, at a level just distal to the upper tibial physis, by means of a K wire. This wire was a transfixing one that passed through the upper tibio-fibular syndesmosis. (Figure 3)

Fig. 3: A fluoroscopic picture showing the transfixing wire of the upper tibio-fibular joint.

Tibial osteotomy was performed just distal to the tibial tuberosity. The fibula was cut at the junction of the middle and lower fibular thirds, with excision of 0.5 - 1 cm of bone. (Figure 4)

Fig. 4: A fluoroscopic picture showing the tibial osteotomy and the fibular ostectomy.

After acute correction of the tibial deformities, the two distal rings were then sequentially connected to the tibia by means of K wires, and Schanz screws. (Figure 5) The distal tibio-fibular joint was transfixed with a K wire to prevent proximal migration of the distal fibula during lengthening. Closure of wounds was done using absorbable sutures. Dressings were put over the wounds and around the frame pins.

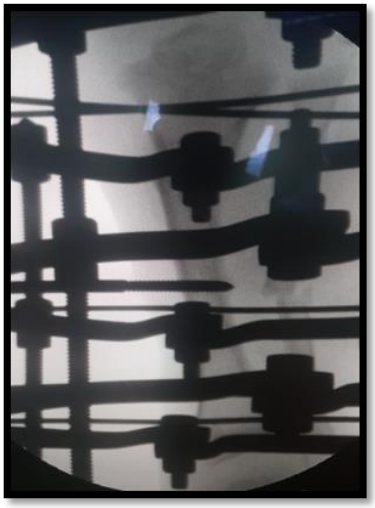
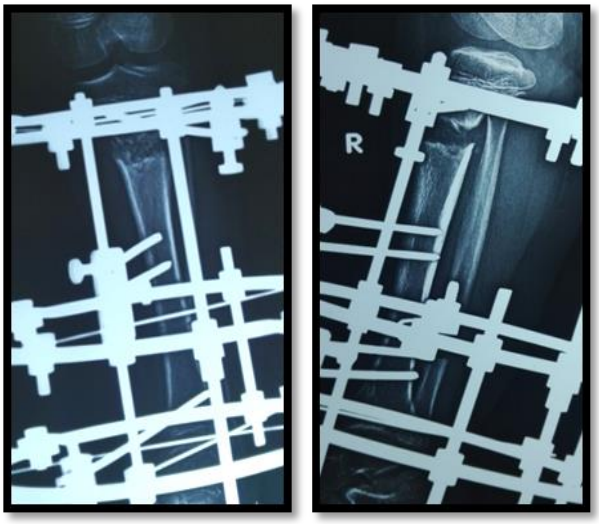


Fig. 5: A fluoroscopic picture showing completion of the tibial fixation.

Postoperative care and follow – up

Standard postoperative radiographs of both lower limbs were taken, and compared with the preoperative images. (Figure 6)



A **B**

Fig. 6: (A) AP and (B) lateral postoperative images, after 2 weeks of follow-up, showing the tibial osteotomy and good alignment of the tibia.

Postoperatively, oral antibiotics were administered, together with analgesics and anti-edematous medications. Full weight bearing was allowed from day 3 after surgery, and distraction began 7 days after the index procedure.

Patients underwent daily physiotherapy. Patients were routinely followed during the lengthening period, and then during the consolidation period. At the time of each visit, the amount of lengthening was measured.

All patients were examined for any signs of pin tract infection, ROM of adjoining joints, angulation or translation of the osteotomy site, and other complications that could occur during lengthening. The frame was removed when three continuous cortices on the radiographs were observed. (Figure 7)

Fig. 7: Follow-up radiograph showing good consolidation of the regenerate bone.

At the final follow up visit, a digital scanogram was done to obtain accurate data regarding final length, and mechanical axis alignment. The amount of lengthening, lengthening ratio (LR), and healing index (HI) were recorded. Final evaluation was performed using the Karlstrom - Olerud modified functional evaluation system [12, 13], and the Paley evaluation system [14].

RESULTS

Clinical measures

The mean amount of the initial tibial length was 14.25 cm (range: 10.2 to 20.1 cm). The mean amount of the regenerate length was 5.9 cm (range: 4.2 to 8.8 cm). The mean amount of the final tibial length was 20.1 cm (range: 14.2 to 28.1 cm). The mean lengthening ratio with respect to the original segment length was 45.1 % (range: 37 to 51 %). The mean healing index was 16.1 days/cm (range: 11 to 25.8 days/cm).

Complications

We had 52 complications, that were categorized as follows;
* 20 legs had ankle equinus deformity; where TA lengthening was done,
* 18 legs had superficial pin tract infection; which resolved after antibiotic therapy,
* 6 legs had valgus or varus deformities; that were treated by corrective osteotomy,
* 6 legs had deep infection; which resolved after thorough debridement,
* 1 leg had premature consolidation; that was treated by revision of the tibial osteotomy, and
* 1 leg had delayed maturation of the regenerate; that was treated by vitamin D supplements.

Functional outcome scores

According to the Paley system, Paley divided the results into functional results and bone results. For functional results, 12 patients had an excellent result, 6 patients had a good result, and 2 patients had a poor result. For bone results, 14 patients had an excellent result, 2 patients had a good result, 2 patients had a fair result, and 2 patients had a poor result.
According to the Karlstrom - Olerud system, 4 patients had an excellent result, 6 patients had a good result, 4 patients had a satisfactory result, 4 patients had a moderate result, and 2 patients had a poor result.

DISCUSSION

When talking about strengths of our study, we believe it provided deeper insights into the importance of assessment of the functional outcomes of the lengthening procedures, so that the benefits of such risky procedures should outweigh their potential hazards.

Furthermore, we applied the concept of resection of a segment of the fibula, to prevent its premature healing; that would render the distal tibio-fibular joint at risk of disruption during distraction.

CONCLUSION

Our study recommends that bilateral limb lengthening of the tibia should be followed by femoral lengthening. This might be a reasonable option for patients with achondroplasia.

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