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Limb Lengthening in Achondroplasia

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Abstract：Most patients with achondroplasia have a functional disability due to their extremely short legs and arms. Limb lengthening can restore the normal proportion of the body and improves the quality of life(QOL)of these patients. We would like to share our experience in limb lengthening in 78 patients with achondroplasia between 2000 and 2010 using the distraction osteogenesis method. The mean age at first lengthening procedure was 11 years and 6 months. Our strategy for lengthening depends on the horizontal method. In 10 cases the horizontal method was applied followed by the vertical method to gain 40 cm in total of stature. Single level, but sometimes double level lengthening was applied. The duration of treatment from initiation of lengthening to removal of the apparatus was 12.8 months in the tibia, 7.8 months in the femur, and 8.3 months in the humerus. The consolidation index(days per centimeters of lengthening) was 27.5 days in the tibiae, 23.8 days in the femora, and 30.6 in the humeri. The average height gained was 15.7 cm(range from 14 to 19 cm)from tibial lengthening and 9.8 cm(range from 8 to 12 cm)from femoral lengthening. While the average humeral length gained was 9.9 cm(range from 8 to 12 cm). A total bone lengthening of up to 104 cm was reached in 10 cases of bilateral tibial lengthening, bilateral humeral lengthening, bilateral femoral lengthening and further longitudinal tibial and femoral lengthening. Despite the significant complication rate, we feel that the handicap due to short stature suffered by patients with dwarfing conditions warrants continued treatment using the Ilizarov technique. None of the patients in this series regretted their decision to undergo limb lengthening. The Ilizarov apparatus allows significant flexibility, allowing correction of axial deviation without surgical intervention. With appropriate motivation of the patient and skill of the surgeon, it is possible to overcome the difficulties presented by these long and potentially risky procedures.

Introduction

Achondroplasia is the most frequent chondrodysplasia of congenital origin and occurs in one out of every 10,000 births and is characterized by defective enchondral ossification owing to Mutations in FGFR3(fibroblast growth factor receptor 3)[4)11)13)]It is a type of rhizomelic dwarf-

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ism that results in considerable physical and psychological handicaps owing to the disproportionate stature of the body and difficulty in performing routine daily activities. These individuals often feel different from their families and circle of friends and often suffer from emotional disturbances and are prone to have inferiority complexes[4)11)].

In addition to short stature, individuals with achondroplasia often have substantial angular deformities of the limbs, spinal problems; other neurologic and craniofacial abnormalities develop[3)8)]. Limb lengthening by the Ilizarov method has been used in patients with achondroplasia by many authors during the past few decades, but with varying amounts of success[1)2)4)5)7)12)16)]. Owing to its modularity and versatility, the Ilizarov method can simultaneously address limb shortening and angular deformities in these individuals[9)19)]. In the last Twenty five years we have used Ilizarov's method in a number of patients affected by achondroplasia, with satisfactory results. The results in 78 cases, followed up for two years or more after the end of treatment, are presented here.

Method

Tibial lengthening

The pre-construction of the apparatus consists of a proximal block(a half-ring and a complete ring, connected by hexagonal sockets of 2 or 3 cm)and a distal ring, in addition to an intermediate ring positioned between the previous two (Fig. 1-a). The osteotomy of the fibula must be performed at an intermediate level(Fig. 1-b). For fixation of the apparatus to the leg, we introduce a first reference wire to the proximal tibia and a second wire distally, with transverse direction. The inferior tibio-fibular syndesmosis must be fixed with one of the Kirschner wires. Then the wires are connected to the rings. At this point the apparatus is centralized on the leg and the axis of the assembly is aligned with the anatomical axis of the tibial. To complete stabilization of the apparatus to the tibia more wires and Schanz screws are inserted(Fig. 1-c). After fixation, the tibial osteotomy is done subperiosteally using a Gigli saw.

Humeral lengthening

The device is constituted by a proximal arc (the smallest of those of the apparatus Catagni-Cattaneo)and a distal half ring connected via Angular supports. The first reference wire is inserted to the distal end, perpendicularly to the longitudinal axis of the humerus, directed from posterolateral to anteromedial, at the level of the epicondyle. Be sure that the humerus is fully centralized. Three half pins are inserted proxi-

a) b) c)

Fig. 1. Tibial lengthening a)the pre-construction of the apparatus b)fibular osteotomy c)the apparatus after fixation to the tibia on both sides

a) b) c) d)

Fig. 2. Humeral lengthening a) the pre-construction of the apparatus b, c) fixation of the apparatus to the humerus d) Callotasis

mally. Distal fixation is completed by an additional wire and a half pin. The osteotomy is performed distal to the insertion of the deltoid, preferably with multiple drill technique (Fig. 2).

Femoral lengthening

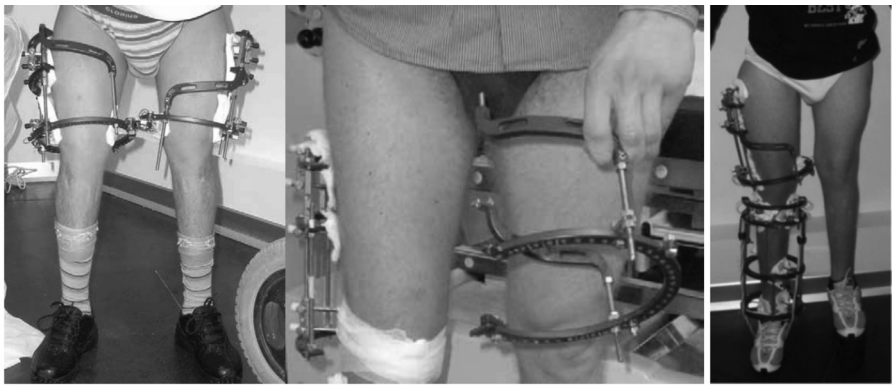
The configuration currently used consists of a distal ring, a proximal block consists of two arcs of the same diameter connected to each other through two hexagonal sockets 6 cm and connected with the distal ring by means of two threaded rods and two oblique supports (Fig. 3-a). The apparatus centered on the thigh, we introduce a transverse Kirschner wire with a diameter of 1.8 mm in the distal metaphysis of the femur. This wire is connected to the distal ring and tensioned to 110 kg. 3 or 4 half pins 6 mm in diameter are used to fix the proximal block, sometimes covered with hydroxyapatite. 3 half pins are applied to the distal ring, namely a medial, proximal to the rim, with an inclination of about 45° with respect to the transverse Kirschner wire, with direction from the posteromedial to the anterolateral and two lateral, proximal to the ring, inclined at 45°, with the direction from posterolateral to anteromedial (Fig. 3-a). This configuration offers a great stability to the system avoiding transfixation of the vastus lateralis and medialis, ensuring the full functionality of the quadriceps and allowing a better range of motion of the knee, with the prevention of possible stiffness. We proceed to the distal metaphyseal osteotomy with a Gigli saw subperiosteally. At the end of elongation and when bone regeneration is consistent, you can remove the medial components of the apparatus and the medial half pin. In this way, the configuration is only lateral, but with the half pins inserted in different planes (Fig. 3-b).

Distraction

Currently, the strategy[9)18)] applied at the Lecco Hospital is the following: 1. Simultaneous lengthening of the legs, with bifocal technique. The rate of elongation is never greater than 1/4 mm 3 times per day for each level of osteotomy. The lengthening of the Achilles tendon is performed only if necessary and, usually, after a few inches of stretch.

2. Simultaneous lengthening of the humerus, with one level technique, with the rate of 1/4 mm 3 to 4 times a day.

3. Simultaneous lengthening of the two femurs (applying the new hybrid configuration, the tol-



a) b) c)

Fig. 3. Femoral lengthening a) fixation of the apparatus to the femur b) remove the medial components of the apparatus and the medial half pin c) vertical lengthening in some patients

erance of the apparatus allows a bilateral application), with one level technique, with rate of 1/4 mm 3 to 4 times a day. For very young patients, if necessary; a further lengthening is performed, in this case vertical (femur and leg), so as to gain additional 10–12 cm (Fig. 3-c).

Patients and results

78 Achondroplastic patients, aged between 10 and 20 years at operation, who have been followed up for 24 months. The mean age at first lengthening procedure was 11 years and 6 months. Our strategy for lengthening depends on the horizontal method. In 10 cases the horizontal method was applied followed by the vertical method to gain 40 cm in total of stature. Single level, but sometimes double level lengthening was applied. The results comprise lengthening of 126 legs, 82 arms and 57 thighs; with a total of 265 lengthening procedures. The duration of treatment from initiation of lengthening to removal of the apparatus was 12.8 months in the tibia, 7.8 months in the femur, and 8.3 months in the humerus. The consolidation index (days per centimeters of lengthening) was 27.5 days in the tibiae, 23.8 days in the femora, and 30.6 in the humeri. The average height gained was 15.7 cm (range from 14 to 19 cm) from tibial lengthening and 9.8 cm (range from 8 to 12 cm) from femoral lengthening. While the average humeral length gained was 9.9 cm (range from 8 to 12 cm). A total bone lengthening of up to 104 cm was reached in 10 cases of bilateral tibial lengthening, bilateral humeral lengthening, bilateral femoral lengthening and further longitudinal tibial and femoral lengthening. There have been complications in 5 cases, the most frequent being pin tract infection and equinus deformity of the foot. The complications were treated with good results. We have not observed any fracture of a regenerated bone.

Discussion

Ilizarov's ingenious idea of influencing the growth of the bone regenerated after compactotomy by taking advantage of its pliability, makes it possible to lengthen the bone and at the same time to correct any deformity present[4)7)9)]. Limb lengthening is a complex procedure and has a high complication rate[10)]. The limb must be lengthened safely without causing deterioration in function. In the lower limb there

seems to be an increasing number of complications when the lengthening exceeds 20％.This phenomenon does not occur in the humerus[10)17)20)]. Achondroplastic patients tolerate lengthening well because of ligament and soft tissue laxity, and their muscle length exceeds their bone length before lengthening[14)]. Since one of the basic principles of Ilizarov's method is the application of load to the limb by weight bearing or exercise, the cooperation of the patient is essential. Treatment should not start until the child is capable of walking even when experiencing some pain. However, there may be more advantageous to plan limb lengthening when the patients are fully aware of their handicap. They are then willing to cooperate and more able to cope with the discomfort caused by wearing the apparatus. There is no contraindication to carry out a further lengthening of a bone after an interval of some years, since the regenerated bone is normal in structure. Limb lengthening can be started even after bone growth has ceased. After the second decade of life, Achondroplastic subjects lose interest in improving their physical aspect; at this age the risk of complications from treatment also increased[4)]. Surgical and postsurgical complications we have encountered were classified as minor, moderate, and severe. Minor complications were those which required only modification of the apparatus during treatment. Twenty-three percent of the lengthenings required some modification of the device during treatment. Moderate complications were those which required additional procedures during lengthening (correction of angular deformities by means of hinges, replacement of Kirschner wires). Forty-two percent of patients fell into this category. Finally, severe complications were those which required another surgery following treatment or left permanent lasting sequelae on the patient. Twenty-one percent of patients fell into this category. The most common complication was equinus contractures of the ankle which required treatment by tendo-achilles lengthening. Two pulmonary emboli were sustained following percutaneous tendo-achilles lengthening. One patient died as a result of this complication. In one case of bilateral tibial elongation through proximal osteotomy, there was a complete absence of the regenerate, and we had to proceed with distal osteotomy and bone transport with success[6)15)].

Conclusion

Despite the significant complication rate, we feel that the handicap due to short stature suffered by patients with achondroplasia warrants continued treatment using the Ilizarov technique. None of the patients in this series regretted their decision to undergo limb lengthening. The Ilizarov apparatus allows significant flexibility, allowing correction of axial deviation without surgical intervention. With appropriate motivation of the patient and skill of the surgeon, it is possible to overcome the difficulties presented by these long and potentially risky procedures. The surgical procedure that we applied with the circular Ilizarov apparatus has allowed us to improve the quality of life of our patients suffering from achondroplasia.

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