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Achondroplasia (ACH) is the most frequently encountered form of non-lethal skeletal dysplasia and is characterized by defective enchondral ossification due to defective gene encoding for fibroblast growth factor receptor-3[1,2,3]. It is a type of rhizomelic dwarfism with an incidence of around one in 10,000 live births[2]. In addition to short stature, individuals with ACH often develop significant angular deformities of the limbs, spinal problems as well as other neurological and cranio-facial abnormalities[4,5].

Achondroplasic dwarfism results in considerable physical as well as psychological handicaps due to the disproportionate stature of the body as well as the difficulty in performing routine activities of daily living. These individuals often feel “different” in their families and friend-circles, often suffer from emotional disturbances and are prone to develop inferiority complexes[2]. They also have serious orthopaedic issues and symptomatic mal-alignment of lower limb eventually requires surgical correction to decrease pain and prevent early onset degenerative arthritis[3,6].

Limb lengthening in ACH by the Ilizarov method has been used extensively over the past few decades with varying amount of success[1,2,7-10]. The Ilizarov method, by its modularity and versatility can simultaneously address the multiple problems of Limb shortening as well as angular deformities commonly encountered in achondroplastics [1]. A number of previous reports have put forth the technique as well as the obstacles seen in the use of the Ilizarov technique for this disease. However this is the first study, to the best of our knowledge, that has explored the effect of limb lengthening on quality of life (QOL) parameters at a long-term follow-up.

Therefore we evaluated our operated patients clinically and radiologically at a mean follow-up of 4.9 years in an attempt to answer the following questions : (1) What are the complications associated with bilateral lower limb lengthening, and how common are they? (2) Does QOL increase after surgery in parameters like AAOS lower extremity score, SF-36 and the Rosenberg self-esteem scores compared with an equivalent group of patients who have not undergone limb lengthening? (3) Is there any correlation between the complication rate and QOL of the patients?
Methods and Materials

We performed a retrospective study on 22 patients (88 segments) with ACH all of whom underwent bilateral tibial lengthenings with Ilizarov ring fixator and bilateral femoral lengthening with monolateral external fixator between January 2002 to December 2005. The average age of the patients at the time of surgery was 12.7 years (range 8-25 years). There were 7 males and 15 females. All the patients underwent bilateral tibial lengthening first after which bilateral femoral lengthening was performed 6 to 22 months later. Average follow-up of patients was 4.9 years (range 4.5-6.9 years).

A thorough review of the medical records of all patients was done to obtain demographic data and other factors related to the treatment given in the form of age at surgery, height, scheme of lengthening used, number of days of distraction, number of days required for the maturation of the callus as well as the total duration of treatment. Complications were recorded in detail and need for secondary operations looked for. Various percentages and indices like the lengthening percentage, fixator index and bone healing index were calculated.

Operative treatment and post-operative protocol:

The Ilizarov ring fixator (U & I, Seoul, Korea) was used for the tibial lengthening and deformity correction. Three or four rings were used depending on whether unifocal or bifocal osteotomies were done at the center of rotation of angulation (CORA) for gradual correction of deformity and lengthening. Paired hinges were aligned with the apex of deformity and a single lengthening rod was placed opposite to them. We used 2 wires each at proximal and distal ring. Proximal and distal tibiofibular joints were transfixed with a wire each to prevent distal or proximal migration of the fibula, respectively. We inserted 2 additional half pins at proximal and middle ring. The osteotomy using multiple drill-hole method was done at the level of CORA.

A monolateral external fixator was used for the femoral lengthening. Three or four Schanz screws were inserted in proximal and distal end of the femur perpendicular to anatomical axis. Transverse osteotomy was performed at mid diaphyseal region in all cases after longitudinally incising the periosteum.

Lengthening with or without gradual correction of deformity was started after 7 days at a rate of 1 mm/day (0.25 mm every 6 hours). The rate was adjusted during follow-ups according to callus morphology in radiographs. Removal of the fixator was done when 3 cortices have shown satisfactory corticalization on radiographs. A long leg cast was applied after fixator removal, and was removed at 4 to 6 weeks. Postoperative radiographic measurements were based on immediate post-removal radiographs. All the radiographs were taken using StarPACS, Infinitt, PiView STAR, 5.0.6.0 with x-ray beams perpendicular to the center of the distraction site.
Charts were reviewed to determine the clinical outcomes and complications. During visits, patients were examined for any signs of pin tract infection, range of motion of adjoining joints, angulation or translation of the osteotomy site and other complications which can occur during lengthening. The complications were recorded according to the classification of Paley et al. (problems, obstacles, and sequelae) [11] (Table 1). The MAD (conventional or ground mechanical axis deviation) and tibia-femur (T-F) angle were measured on standard weight-bearing full-length lower-extremity radiographs to determine angular deformities of the knee. The conventional MAD (MAD-C) was measured as the distance between the centre of the knee and a line drawn from the center of the femoral head to the center of the talus. The ground MAD (MAD-G) was measured as the distance between the center of the knee and a line drawn from the center of the femoral head to that of the heel when in contact with the ground on long leg-standing radiographs [1]. The difference between MAD-C and MAD-G is the result of inclusion of heel varus in the measurement of ground mechanical axis deviation. Heel varus or valgus was measured with tibio-calcaneal angle on standing radiographs (Table 2).

Neutral alignment was defined when the mechanical axis line drawn from the center of the femoral head to the center of the calcaneus (MAD-G line) passed through the 30% lateral or medial plateau region measured between the center of the knee and medial or lateral edges of the medial or lateral plateau. Mal-MAD such as valgus or varus was defined when MAD-G line passed through more than 30% lateral or medial plateau region. Valgus or varus ankle was defined when the tibio-calcaneal angle on standing radiographs was more than -5 or +5 degrees. Additionally, various other radiographic indices including PTFD (proximal tibio-fibular distance), DTFD (distal tibio-fibular distance) and TFDR(tibio-fibular distraction ratio) i.e. the amount of fibular lengthening divided by the tibial lengthening, were measured preoperatively and postoperatively (Table 2).

All the patients were called up using the medical records and asked to visit the hospital. They were examined clinically by the senior author (HRS) and any complications and sequelae were looked for. Standard radiographs of both lower limbs were taken and were compared with the pre-operative and immediate post-operative ones. Quality of life assessment was done using four validated questionnaires: 1) American Academy of Orthopaedic Surgeons (AAOS) lower limb outcome questions [12], 2) Rosenberg self-esteem questions [13], 3) Short Form 36 (SF-36) questions [14] and 4) Paediatric Quality of Life assessment tool [15] (For children between 8 to 12 years of age). Informed parental consent was taken for children 16 years or below. This group of 22 patients was compared with 22 patients of same diagnosis, similar age and height but on whom surgery had not been performed.

**Statistical analysis:**

The data was entered using Microsoft Excel 2007 version and analysed using SPSS software. The results of the lengthening and the various indices were denoted as Mean ± standard deviation. The difference in results of the AAOS lower extremity scores, Rosenberg Self-esteem scores, SF-36 and PedsQL scores between the surgical and
non-surgical groups were analyzed using the Mann-Whitney, Wilcoxon, 2-tailed and Wilcoxon test. A p-value of less than 0.05 was considered significant. The operated patients were also divided into two groups for statistical analysis i.e. Group I (9 patients) who had four or less complications and Group II (13 patients) who had more than four complications. Two more groups were also made about whether there were complications without sequelae (Group A- 7 patients) or complications with sequelae (Group B- 15 patients).
Results

Lengthening and other indices:

The average gain in length was 10.21 + 2.39 cm in femur and 9.13 + 2.12 cm in tibia. The mean lengthening percentage (LP) was 35.5% (range 14-65%) in femur and 37.2% (range 15-67%) in tibia. The mean external fixator index (EFI) was 33 days/cm (range, 21-84 days/cm) in femur and 28 days/cm (range, 11-80 days/cm) in tibia. The mean healing index (HI) was 34 days/cm (range, 17-80 days/cm) in femur and 35 days/cm (range, 15-90 days/cm) in tibia (Table 2).

Complications:

A total of 123 complications were encountered in 88 segments. Joint complications were seen in 70 (57%) segments and bony complications in 47 (38%) segments (Fig.1). Other minor complications (problems) were seen in 6 (5%) segments and properly dealt with in the outpatient clinic.

Mal-alignment of the mechanical axis was one of the major bony complications seen in these patients. It was identified in all the phases of the procedure even after fixator removal. It was treated by a case-to-case basis according to the causes of the mechanical axis deviation seen in that particular case. At the final follow-up after bilateral tibial and femoral lengthening with/without gradual correction of deformity, mal-alignment of the mechanical axis was seen in 9 limbs.

The joint related complications included hip flexion contracture and equinus deformity of the ankle. Flexion contracture of the hip more than 30 degrees occurred in 30 (68%) of 44 femoral segments for which intramuscular recession of rectus femoris, Sartorius and the iliopsoas muscles and partial release of the iliobibial band during the consolidation phase was done to improve the range of movement (Fig.2). Assisted physiotherapy and mobilization started when pain was subsided. At the final follow up after soft tissue release, average hip flexion deformity was 9.5 degrees. Equinus deformity occurred in 26 (59%) of 44 tibial segments. Intramuscular recession of the gastrocnemius and soleus with/without foot frame for gradual correction was performed and the mean equinus deformity were 5.2 degrees (range, 0-9 degrees) at the final follow up.

Quality of life assessment:

Functional assessment of the patient was performed using the AAOS lower limb score[12] which measures the ability of the patient to walk on flat surfaces, lower limb stiffness, swelling and pain and the ability to ambulate with or without support. Analysis of this score in both groups of patients showed that the surgical group fared slightly better than the surgical group though the difference was not statistically significant(Table 3).
The Short-Form-36 (SF-36) questionnaire put forth some interesting findings. Both the groups had limitation in performing all physical activities like bathing and dressing though the non-surgical group fared marginally better than the surgical one. However the surgical group showed significantly better scores in the mental health component of the SF-36 forms. The combined scores therefore were statistically similar in both groups.

The Rosenberg self-esteem scores, however, showed that the lengthened group had higher score of 22, and the nonsurgical group had lower score of 19. The difference in self-esteem scores between the surgical and non-surgical groups were proven significant at p=0.000025. Group I with less than four complications showed higher Rosenberg self-esteem score of 23 and group II with more than five complications showed lower score of 20 (p=0.00175). This result suggested fewer complications could improve QOL after surgery. Also group A without the sequelae showed higher Rosenberg self-esteem score of 22 and group B with one or more sequelae showed lower score of 19.5 (p=0.000125).

There was no significant difference between surgical group and nonsurgical in PedsQL study (Table 3).
Fig. 0: Fig 1 is the radiograph showing refracture of femur after lengthening.

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**Fig. 0:** Fig. 2 is the latest radiograph after hemiepiphysiodesis. Malalignment was corrected but there was 2.5cm remained limb length discrepancy. Flexible nail was inserted due to refracture of the femur.

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Fig. 0: These are clinical photographs showing hip flexion contracture.

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Fig. 0: These are clinical photographs showing knee extension stiffness after femoral lengthening.

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Fig. 0: Fig.5 is a radiograph taken during tibia correction.

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Fig. 0: Fig. 6 is the latest radiograph showing good alignment of lower extremity.

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**Fig. 0:** Fig. 7 is a radiograph taken after fibula reosteotomy and additional half pin insertion at fibula. Fig. 4c is the latest radiograph showing good alignment after bone union. TFDR was increased to 0.92

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Conclusion

The efficacy of the Ilizarov technique in lengthening of both the lower limbs in achondroplasia and other causes of short stature has been effectively proven by several reports [7-10]. However, the basic concept of lengthening in ACH is still a contentious issue. While there is no doubt that the total height of the patient can be increased by this method, there was still no evidence about how this method would have an impact on overall function or quality of life. Also, the long arduous nature of the treatment as well as the significant potential for immediate and long-term complications makes it a controversial technique to apply[16].

With regards to bone or joint-related complications during the tibial lengthening, most common complications were premature consolidation of the fibula, valgus and procurvatum deformity of the tibia, fracture or deformation of the callus, and equinus deformity of the ankle. The valgus and procurvatum deformity of the proximal tibia during lengthening attributed to different resisting force based on different muscle bulk between medial, lateral, anterior, and posterior compartment(Fig.3). Another cause of the valgus tibia was less distraction amount of the fibula compared to the tibial distraction amount. In our study, all cases of the tibial lengthening showed less distraction of the fibula ranging from 1 cm to 5.2 cm than the tibial distraction even though the proximal and distal tibia-fibular joint was fixed with one Ilizarov wire. During large amount of lengthening, the proximal one wire which was inserted through the fibular head and crossed the tibia could not hold the fibula due to high resisting force. It had high incidence of cut out from the fibular head which in turn resulted in less distraction of the fibula, premature consolidation of the fibula and distal migration of the fibula. Authors performed the fibular osteotomy at the distal one third and the distance between the proximal wire and osteotomy site of the fibula was long. Hence the proximal wire did not distract the fibula sufficiently due to resisting force of the attached muscles. Therefore, insertion of additional 3 or 4 mm half-pins at the proximal fibula at the initial surgery can be recommended to prevent complications associated with the fibular shortening (Fig.4). The distal wire through the distal fibula and tibia could hold the distal fibula without proximal migration of the distal fibula during lengthening because the distance between the osteotomy and wire was short and the diaphyseal area of the distal fibula had more cortical area which provides more holding strength of the wire compared to the proximal fibular head which has the more metaphyseal area. However, the distal wire had high incidence of tethering the peroneal muscles which resulted in heel valgus deformity. Therefore, careful insertion of the wire is required to avoid this deformity. Another cause of heel valgus was the proximal migration of the distal fibula after removal of the fixator which resulted from the, subsidence of the fibular lengthening site. According to our results, the role of the fibula was important during the tibial lengthening.

During the femoral lengthening, common bone-related complications were varus angulation and delayed consolidation at the lengthening site. Joint-related complications
were flexion and adduction contracture of the hip and valgus deformity at the knee due to tight ilio-tibial band and lengthening along the anatomical axis without bony deformity at the distal femur (Fig.5).

The varus angulation occurred because the monolateral fixator was attached to the lateral side of the femur and more distraction force to the lateral side with less distraction force to the medial side which has bulkier muscles than the lateral side. This deformity could be corrected acutely with reassembly of the fixator during follow-up. Delayed consolidation was a very serious complication which was associated with fracture after the removal of the fixator.

Even with these complications, our series shows that serial lower limb lengthening is a good option for patients of achondroplasia in terms of good QOL scores. The lengthened patients scored well in the mental scoring in the SF-36 questionnaire as well as the Rosenberg Self-esteem scores. The physical and functional results of the operated patients were equivalent to the non-surgical group as shown by the AAOS lower extremity scores and the physical component of the SF-36 scores. Such a discrepancy was expected as a long and complicated procedure such as bilateral limb lengthening is bound to have some sequelae in the form of decreased range of motion and residual deformities and will thus cause a resultant decrease in the functional score. The physical as well as mental health scores were directly related to the number of complications seen as the Rosenberg as well as the SF-36 scores reduced significantly as the number of complications increased more than four. Our data thus shows that early recognition of complications and prevention of sequelae is extremely important in gaining an excellent functional result at the long-term.

Various studies have explored the causes of complications of extensive limb lengthening in achondroplasia and have tried to show their effect on patient satisfaction as well as function, though they have not explained the same in QOL terms. Lavini et al[17] and Aldegheri et al[18] reported that 85 % to 95% of their patients were satisfied with results without major complications and they had resumed a normal social life after lower limbs lengthening. Similarly Barretto et al[19] showed, in their report of twenty two patients with achondroplasia, that their results in these patients were as good as those in leg-length discrepancy and all the patients who had no or only minor complications were extremely satisfied in their results.

Our study thus suggests that bilateral limb lengthening can be safely tried in achondroplasia, provided complications are looked for and kept in check. We believe that our study would be a valuable addition to the current literature in this subject, which is surely lacking in regard to quality-of-life and functional assessment of these patients. However, further multi-center studies are still necessary for setting up objective guidelines for proper patient selection or protocols for evaluation of surgical outcome and QOL before this method attains widespread use.
Fig. 0: These are clinical photographs showing knee extension stiffness after femoral lengthening.

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Fig. 0: Fig 1 is the radiograph showing refracture of femur after lengthening.

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Fig. 0: Fig 1b is the radiograph showing refracture of femur after lengthening.

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Fig. 0: Fig. 2 is the latest radiograph after hemiepiphysiodesisis. Malalignment was corrected but there was 2.5cm remained limb length discrepancy. Flexible nail was inserted due to refracture of the femur.

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References


