Femoral lengthening in achondroplasia

MAGNITUDE OF LENGTHENING IN RELATION TO PATTERNS OF CALLUS, STIFFNESS OF ADJACENT JOINTS AND FRACTURE


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Extensive limb lengthening may be indicated in achondroplastic patients who wish to achieve a height within the normal range for their population. However, increasing the magnitude of lengthening is associated with further complications particularly adjacent joint stiffness and fractures. We studied the relationship between the magnitude of femoral lengthening and callus pattern, adjacent joint stiffness and fracture of the regenerate bone in 40 femoral lengthenings in 20 achondroplastic patients. They were divided into two groups; group A had lengthening of less than 50% and group B of more than 50% of their initial femoral length. The patterns of radiological callus formation were classified according to shape, type and features. The incidence of callus features, knee stiffness and regenerate bone fracture were analysed in the two groups. Group B was associated with an increased incidence of concave, lateral and central callus shapes, adjacent joint stiffness and fracture. Statistically, the incidence of stiffness in adjacent joints and regenerate bone fracture was significantly associated with the magnitude of lengthening.

We suggest that careful radiological assessment of the patterns of callus formation is a useful method for the evaluation and monitoring of regenerate bone.

Achondroplasia is the most common genetic form of dwarfism with short limbs and trunk of normal length. In order to achieve height within the normal population range at maturity, extensive limb lengthening has to be considered. However, this is associated with complications particularly stiffness of adjacent joints and fractures leading to a poor outcome. Yun et al. reported rates of 20% and 26% for fractures and knee stiffness respectively when the limb was lengthened by 20% of its initial length. Karger, Guille and Bowen noted rates of 27% and 65% for fractures and joint contractures respectively when the limb was lengthened by 24% of its initial length. Many authors have recommended that the goal of lengthening in a bone segment should be limited to 20% to 30% of its initial length. However, to our knowledge there are no reports of the complications associated with the magnitude of femoral lengthening required in patients with achondroplasia. Paley recommended that refracture may be avoided by careful analysis of the regenerate bone in the distraction gap before removal of the apparatus and that the appropriate management of muscle contractures is essential during lengthening. Serial radiological evaluation and a classification of the radiological information regarding the distraction osteogenesis such as that of Li et al. are helpful in decision-making during the healing process.

In this study we reviewed the results of femoral lengthening in achondroplastic patients using a monolateral external fixator. We compared the incidence of shape, type and features of the callus, stiffness in adjacent joints and regenerate bone fracture between those who had lengthening of less than 50% and those with lengthening of more than 50% of their initial bone length. We also studied the relationship between the magnitude of femoral lengthening and the shape and features of the callus, stiffness of adjacent joints and fracture.

Patients and Methods

We undertook a retrospective study of 20 achondroplastic patients who, between 2002 and 2006 underwent bilateral femoral lengthening using a monolateral external fixator. There were 15 females and five males with a mean age at operation of 12.5 years (8.0 to 21.0). The mean follow-up was 3.9 years (2.1 to 6.7). There were 12 patients with lengthening of less than 50% of the initial bone length (group A) and eight patients with lengthening greater than 50% (group B). Only those patients with full medical records and radiographs were included.
Patients with systemic disease, previous injuries or operations and tumours were excluded. The nature of the procedure, duration of treatment, complications and the risks of surgery were explained fully to the patients and ethical approval was obtained from our institution.

Operative technique. All operations were undertaken by the senior author (HRS), using a monolateral external fixator (L-Standard type Dyna-extor, Joy-M, Seoul, Korea). Three Schanz screws were inserted in the distal femur parallel to the knee and perpendicular to the bony surface and a further three screws in the proximal femur perpendicular to the bony surface. A transverse osteotomy was performed in the mid-diaphyseal region through a circumferential incision of the periosteum and drill holes connected by a small osteotome. Completion of the osteotomy was confirmed under the image intensifier. The muscles were kept under stretch by flexing the knee while inserting the screws.

Daily physiotherapy to maintain range of movement (ROM) at the hip and knee was started one or two days post-operatively and toe-touch weight-bearing allowed after four to five days, followed by partial weight-bearing once the patient was comfortable. Distraction was started seven days post-operatively at the rate of 0.25 mm four times a day until the desired length was achieved or exceeded, or complications precluded further distraction. We adjusted the rate of distraction with or without compression during the distraction period, according to the appearance of the callus. All patients were evaluated clinically and radiologically weekly during the distraction phase and every four weeks during the consolidation. All radiographs were taken using StarPACS, PiView STAR, 5.0.6.0 (Infinitt, Seoul, Korea) with the x-ray beam perpendicular to the centre of the distraction site. At follow-up visits, patients were examined for signs of pin-track infection and the ROM of the adjoining joints was assessed. The external fixator was removed when three continuous cortices could be seen on the radiographs and dual-energy x-ray absorptionometry showed the bone mineral density to be 70% to 75% of normal. The limb was then protected either by a cast or brace depending on the type of callus seen on the radiograph.

All the radiographs and charts were analysed by three observers (HRS, KPV, JYY). The results were assessed on the basis of the classification described by Li et al,9 which assesses the callus according to shape, type and features. Shape is classified as fusiform (regenerate wider than the original bone), cylindrical (same width as the original bone), concave (narrower than the original bone), lateral (mainly on one side of the distraction gap) and central (a thin pillar) based on the width of callus compared with the original osteotomy site (Fig. 1). Type is based on four patterns of distraction osteogenesis (sparse, homogeneous, heterogeneous and lucent) and three densities (low, intermediate and normal). Callus features are divided into ten types (Fig. 2) as follows: type 1 (sparse low density) typically first seen in the distraction gap, type 2 (homogeneous low density) in which the newly formed bone appears to consist of longitudinal striped columns homogeneously bridging the two ends of the osteotomy; type 3 (heterogeneous low density) where newly formed bone appears heterogeneously, bridging the two ends of the osteotomy; type 4 (lucent low density) where a central radiolucent zone is present in the newly formed bone appears homogeneously bridging the two ends of the osteotomy; type 5 (lucent low density) where a central radiolucent zone is present in the newly formed bone; type 5 (lucent low density) where newly formed bone appears heterogeneously bridging the two ends of the osteotomy; type 6 (lucent low density) where a central radiolucent zone is present in the newly formed bone; type 7 (lucent low density) where a central radiolucent zone is present in the newly formed bone; type 8 (lucent low density) where a central radiolucent zone is present in the newly formed bone; type 9 (lucent low density) where a central radiolucent zone is present in the newly formed bone; type 10 (lucent low density) where a central radiolucent zone is present in the newly formed bone.
with continuous cortices; and type 10 (heterogeneous normal density) where normal density bone bridges the two ends of the distraction gap heterogeneously with irregular ossification. Furthermore, the different pathways to maturity of the regenerate are described. The homogenous pathway begins with a type 2 pattern progressing through type 6 to type 9. The heterogeneous pathway begins with type 3 progressing through type 7 to type 10. The lucent pathway has types 4 and 8 as the predominant patterns.

The incidence of shape, type and features, adjacent joint stiffness and regenerate bone fracture were analysed in both groups. The external fixator index, calculated by dividing the duration of external fixation in months by the total length of regenerate bone in centimetres, was measured in all patients.

**Statistical analysis.** Student's *t*-test was used to compare all matched variables between the groups. In order to check any significance of those variables among the different pathways and shapes, we used non-parametric analysis of variance (ANOVA) with the Kruskal-Wallis test. A p-value ≤ 0.05 was considered to be significant. The classification was tested for concurrence and reproducibility by interobserver studies. Intraobserver studies were not carried out. The data were analysed statistically to give intercorrelation coefficient values.

**Results**

The mean gain in length was 9.2 cm (4.5 to 13.0) and the mean percentage gain in length was 39.3% (14.0 to 65.0). The mean external fixator index was 1.3 months/cm (0.7 to 2.4) and the mean period of application of external fixator was 10.8 months (6 to 17). The mean ROM of the knee was 53.8° (25° to 70°) during the consolidation phase, 97° (65° to 110°) during removal of the external fixator and 114.1° (95° to 126°) at the final follow-up. The mean hip flexion deformity was 15.2° (7° to 20°) during lengthening and 1.5° (0° to 5°) at the final follow-up.

**Lengthening.** The mean age of the patients in group A was 12.9 years (9.0 to 21.0) and in group B 11.8 years (8.0 to 19.0). The mean gain in length was 7.8 cm (5.3 to 10.1, 29.9%) in
The fusiform shape showed the best external fixator index (1.0 months/cm) followed by the cylindrical shape (1.2 months/cm). Concave, lateral and central shapes had poorer external fixator indexes per centimetre of 1.47 months, 1.7 months and 1.9 months, respectively.

Range of movement at the knee and hip. All patients showed a reduced ROM at the knee in the early post-operative period probably because of interference with the extensor mechanism and the iliotibial track by the Schanz screws. This progressively decreased during the distraction phase. However, during consolidation, it gradually improved and a satisfactory, but not full ROM was regained after the removal of the fixator and at the final follow-up (Table I). The ROM of the knee in group A during the healing period was better than that in group B. In three segments with central and lateral shapes, the ROM at the knee was less than 40° after removal of the external fixator. A quadricepsplasty was performed in these patients and at final follow-up the ROM at the knee improved to 90°. The difference in ROM at the knee between group A and group B was significant during the consolidation phase (p = 0.011) and after removal of the external fixator (p = 0.011), but there was no significant difference at the final follow-up (p = 0.26; Table I). There was no statistical significance between the callus pathways and shapes during all the phases.

Flexion contracture of the hip was less in group A than group B. A flexion deformity of more than 30° occurred in three group A and ten group B lengthenings. We performed an intramuscular recession of rectus femoris, sartorius and iliopsoas and partially released the iliotibial band during the consolidation phase to improve the ROM and mobility in these patients followed by physiotherapy and mobilisation when the pain had subsided. At the final follow-up the mean flexion deformity was less than 5° in all patients. A significant difference was seen between group A and group B during lengthening (p = 0.0004), but not at the final follow-up (p = 0.3400; Table I).
Fracture of the regenerate bone. There were six fractures in the 40 regenerate segments (15%). They were more common in group B, heterogeneous pathway than in group A, homogeneous pathway. No fracture was associated with a lucent pathway or fusiform shape. Fractures in concave segments were managed by insertion of a Rush pin and re-application of the external fixator (Fig. 3). In central- and lateral-shaped segments in which there was no progressive formation of new bone during three to four months of the consolidation phase bone grafting was performed with re-application of the external fixator. In one segment with a concave shape and deformity of 35° in the coronal plane, corrective osteotomy with the insertion of a Rush nail and external fixator was necessary. A cylindrical-shaped segment with a stress fracture was managed with application of a long-leg cast. All fractures in the regenerate healed in three to five months with good consolidation. We had no problems with premature consolidation in any segment.

The interobserver agreement among the four observers of feature types was good, with a correlation coefficient between 0.6710 and 0.9690 and a confidence interval (CI) between 0.36805 and 0.9855. With regard to callus type, the correlation coefficient was between 0.3580 and 0.9660 and the CI between -0.0450 and +0.9856. For callus shapes the overall p-value was less than 0.023.

Discussion
Limb lengthening is a complex procedure with a high rate of complications. Formation of callus is influenced by periosteal and endosteal conditions and intrinsic factors such as age, systemic disease, the corticomedullary ratio, previous injury or operation, tumours and the vascularity of the surrounding tissues. Defective callus formation may be associated with intraoperative and post-operative problems such as a traumatic corticotomy, initial diastasis and too rapid distraction. In our study we tried to reduce these factors using a standard procedure and an atraumatic type of osteotomy.

Early restriction of joint movement remains a persistent problem during limb lengthening. Herzenberg et al reported a mean ROM at the knee of 37° during lengthening, 60° during consolidation and 122° at follow-up in isolated femoral lengthenings in non-achondroplastic patients. Our study compares favourably with that of Herzenberg et al, because achondroplastic patients show increased ligament and joint laxity, their muscle length exceeding bone length before lengthening thereby facilitating the lengthening process. Yasui et al reported a progressive increase in ROM at the knee from a mean of 60° during lengthening to 90° during dynamisation and 130° at final follow-up in femoral lengthening (mean 7.2 cm) in achondroplastic patients. They believed that knee contracture during femoral lengthening was related to the pins rather than the lengthening. However, in our study patients in group A showed a good ROM of the knee throughout the healing process when compared with those in group B. In our opinion the ROM at the knee is not simply related to the pins but also to the magnitude of lengthening. Therefore, a decreased ROM at the knee was associated with both a homogeneous and heterogeneous pathway and concave, lateral and at the central shapes. These patterns of callus formation were predominant in group B. With excessive lengthening, we believe that the thigh muscles are unable to keep pace with the lengthening, resulting in a decreased ROM at the knee and fixed flexion at the hip. Homogeneous and heterogeneous pathways showed good healing indices, but resulted in more stiffness and fractures when compared with a lucent pathway. This may be because of the reduced amount of lengthening achieved in the lucent pathway segments as a result of interventions such as halting the distraction and using acute compression and subsequent distraction which helps the muscles to keep pace with the lengthening. Overlengthening and then shortening can also be useful in preventing joint contractures.

Karger et al reported various soft-tissue procedures such as release of the fascia of vastus lateralis and hamstring...
therapy, splinting and fixation across joints to anticipate oral lengthenings for congenital deficiency of the femur and tendon lengthening to correct joint stiffness. They reported recommended weekly radiological examination, physiotherapy, careful adjustment of the distraction rate and soft-tissue releases when necessary during the healing process. Regular stretching and the maintenance of a good ROM can help to decrease the pain and muscle spasm during lengthening and allow the patient to cope more easily with the activities of daily living.16

Excessive lengthening was associated with a decreased ROM of the adjacent joints, decreased new bone formation and delayed weight-bearing. Knee movement helps to prevent muscle atrophy and adhesion of the muscles to the underlying bone regenerate.5,17 It is also said to increase the blood supply to bone and bone strength.18

There were six fractures in 40 femoral lengthenings (15%) in our series. Yun et al4 reported a femoral fracture rate of 20% and Karger et al7 of 27% (22 femora) when lengthening the femur for short stature in non-achondroplastic patients. The incidence of fracture is less in our study, but bone healing is known to be good in achondroplastic patients.15 Aldegheri et al6 reported six fractures in 117 femoral lengthenings with a mean lengthening of 7.8 cm (29.6%) in achondroplastic patients. In our study there was one fracture in 24 femora in group A which is similar to the results of Aldegheri et al.6 The incidence of fracture was higher in group B (5 fractures in 16 femora). There was a predominance of concave, lateral and central shapes in this group. In fractures associated with lateral and central shapes we used bone grafting together with reapplication of the external fixator to accelerate the healing process, since there was no progressive formation of new bone during the consolidation phase. We recommend halting distraction and applying compression when concave, lateral or central callus shapes appear in the regenerate bone until satisfactory regenerate is formed.

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