Bone Lengthening for Management of Type 1a Proximal Femoral Deficiency with or without Varus Deformity

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Abstract:

Background: The advances in bone lengthening for treatment of moderate congenital femoral deficiency have the advantages of preserving the limb function and decreasing the psychosocial burden on the patients.

Patients and Methods: sixteen patients 10 males and 6 females with unilateral Paley Type 1A unilateral proximal femoral deficiency were treated with distraction osteogenesis bone lengthening. The mean follow up period in this prospective study was 36 months (range: 30 – 48 months). The Pediatric Orthopaedic Society of North America Pediatric Outcomes Data Collection Instruments (PODCI) score was used for clinical and functional outcome of the patients. The calculation of growth inhibition or stimulation in this study was began just after removal of the external fixator until 2 years after its removal.

Results: The mean length gained at the end of the procedure was 6.5 cm (SD ± 3; range, 4 – 8.5 cm). The mean correction of femoral length discrepancy was 105 % (SD ± 45; range, 85% –165%) with overlengthening of 2-4 cm (average 3cm) in 6 patients (37.5 %). Ten patients (62.5 %) reached the equal femoral length in comparison to the normal femur. According to the PODCI score categories surveyed, the standardized and normative scores were accepted.

Conclusion: The surgical technique of femoral lengthening in patients with Paley Type 1A proximal femoral deficiency is a successful and well tolerated technique.

Keywords: Bone lengthening, proximal, femoral deficiency, deformity.

INTRODUCTION

Aitken was the first to describe congenital femoral deficiency as a congenital deformities affecting the lower limb with specific attention to the proximal femur [1]. The deformities of the limb associated with the condition vary from very mild cases with normal short femur, severe coxa vara, and pseudoarthrosis of the proximal femur, valgus deformity of the distal femoral, ligaments hypoplasia, muscles hypoplasia, and abnormal vascularity [2 – 7]. The patients with Paley Type 1a and 1b congenital femoral deficiency were treated by femoral lengthening, while patients with Paley Type 2a treated with knee fusion and prosthetic fitting, Syme amputation, or rotationplasty [8- 12]. The advances in bone lengthening for treatment of moderate congenital femoral deficiency have the advantages of preserving the limb function and decreasing the psychosocial burden on the patients [13, 14].

The tension on the soft tissues in bone lengthening surgery affects the adjacent physis which has an effect on bone growth [15, 16]. There is a controversy about the effect of limb lengthening on the growth of bone [12, 17-19]. In the literature, there is no well-established safe surgical technique for femoral lengthening in patients with proximal femoral deficiency. The clinical outcomes, patient function and psychosocial effects on the patients correlated to the lengthy surgical procedure are not well described in literature. The length which can be gained, the range of movements of the hip and knee joints, the effect of lengthening technique on the
femoral growth, and specific complications have not been reported clearly in literatures. A recent study on distraction osteogenesis for femoral lengthening and reported effective lengthening without loss of hip or knee functions in 38 patients with congenital femoral deficiency [20].

**Patients and Methods**

In the period from June 2009 to June 2015, 16 patients 10 males and 6 females with unilateral Paley Type 1A unilateral proximal femoral deficiency were treated with distraction osteogenesis bone lengthening. 14 from 16 patients had coxa vara which treated before lengthening by valgus osteotomy and plate fixation. Uniplanar external fixator was used for lengthening in all cases. The indications for this technique included stable hips and knees, unilateral cases, Paley type 1a, and limb length discrepancy more than 4 cm in comparison to normal leg. Four patients from 20 at the start of the study were excluded due to: one patient did not complete follow up, three patients previous stage of lengthening, so 16 patients completed the study a mean follow up period of 36 months (range: 30 – 48 months) in this prospective study. All procedures using the same technique were performed at our institution with recording the postoperative clinical and radiological data. The surgical complications, the Range of Motion (ROM) were documented until last follow up.

**Surgical Technique**

The surgical technique performed in all patients was distal femoral osteotomy with distraction osteogenesis using unipolar Limb Reconstructive System (LRS) external fixator and a mean distraction rate of 1 mm per day. The osteotomy was done at the distal metaphysis of the femur percutaneously through an anterolateral incision. Multiple drill holes were performed using 3.2 mm drill completed with an osteotome. If the hip subluxation was a risk before lengthening two schanz screws inserted in iliac bone to support the pelvis (Figure 1). Two 2mm crossing k-wires were inserted during application of external fixator in 12 patients and during follow up in 4 patients when noticed displacement in the osteotomy side with adjustment of the fixator (Figure 2). The physical therapy began soon after the operation by 1 hour daily until the ROM reaches an acceptable limits and the parents were taught to complete the therapy protocol at home. When tibiofemoral subluxation observed, the rate of lengthening was slowed or stopped temporarily and rate of physical therapy until regaining ROM or stop the lengthening.

**Fig1.** Male patient 6 years old with left congenital short femur A) preoperative photo showing shortening of left leg; B) preoperative scanogram with 5 cm femoral shortening; C & D) intraoperative radiology; E) knee subluxation during lengthening; F)X-ray7 months postoperative; G& H) photo of the patient with equalized limb length in external fixator; I) x-ray 2 years postoperative showing the femur after lengthening and union.
4.5 years old female with congenital short femur left side; A) preoperative x-ray showing left femoral shortening with coxa vara; B) x-ray after correction of coxa vara using plate fixation on valgus osteotomy of proximal femur; C) x-ray after second operation for lengthening with removal of the plate at the same sitting; D) x-ray after readjustment of fixator and insertion of 2 crossing k-wires for alignment.

During follow up plain x-rays were taken every 2 weeks during the period of lengthening, then monthly during consolidation. After osteotomy consolidation, the plain x-rays were done every 6 months until the end of follow up. Care of the pins with gentle cleansing, if dry gauze wrapping to prevent motion at the pin-skin interface. Report of complications, obstacles, and surgical interference for these complications during follow up was done. Digital and traditional films for all patients with a magnification marker were done measuring overall limb length discrepancy. The formulae derived by Paley et al [21 - 25] to predict overall limb length discrepancy, and growth stimulation or inhibition were calculated for the patients included in this study.

Serial radiographs at different time periods were used to measure the amount of growth inhibition or stimulation comparing that with the growth in the normal side as a percentage of normal growth. The calculation of growth inhibition or stimulation in this study was began just after removal of the external fixator until 2 years after its removal. The Pediatric Orthopaedic Society of North America Pediatric Outcomes Data Collection Instruments (PODCI) score was used for clinical and functional outcome of the patients [13, 26]. Poor outcome/worse (PODCI) score is calculated as “0” and the best possible outcome is “100” points. Multiple scales of the PODCI including the upper extremity, global function, basic mobility, sports and physical functioning. Added to this the pain/comfort and happiness are included.

The PODCI scores were recorded at least 2 years after completing the bone lengthening operation. The mean age of the patients at the time of lengthening was 7.5 years (range: 5–10 years). In 14 patients the lengthening was 10-14 months after correction of coxa vara (Figure 3). In 2 patients there was no prior hip surgery before lengthening (Figure 4). The goal of lengthening surgery was the temporary equalization of overall limb length expecting at least 4 cm length gaining or more by normal growth in 12 patients, and overlengthening of about 2 to 3 cm was done in four cases with cooperative child and parents and good tolerance.
Fig3. Male patient 5 years old with right side congenital short femur: A) preoperative photo with short femur; B) x-ray after correction of coxa vara; C) x-ray 6 months postoperative with callus formation in the lengthened segment; D) X-ray photo after plate fixation due to fracture after removal of external fixator; E & F) X-ray with fracture femur after removal of the LRS; G) X-ray after plate fixation of fracture femur; H & I) photo of the patient with equalized lower limbs 2 years after plate fixation.

Fig4. Male patient 6.5 years old with short right femur: A) preoperative scanogram with 4.5 cm shortening; B) x-ray after osteotomy and fixator; C) scanogram after finishing lengthening with 2 cm overlengthening; D) x-ray of femur after consolidation.

RESULTS

The mean duration needed to get the expected lengthening was 203 days (SD ± 47; range, 160 – 270 days). The mean length gained at the end of the procedure was 6.5 cm (SD ± 3; range, 4 – 8.5 cm). The mean correction of femoral length discrepancy was 105 % (SD ± 45; range, 85% – 165%) with overlengthening of 2-4 cm (average 3 cm) in 6 patients (37.5 %). Ten patients (62.5 %) reached the equal femoral length in comparison to the normal femur.
At the end of the bone lengthening procedure of the knee flexion ranged from 20 to 85 degrees (average 60 degrees) in a mean duration of 18 weeks. At the end of follow up, the flexion range of knee recovered to near normal range in all patients by a mean of 36 months (range: 30-48 months). The range of hip flexion decreased to 80 degrees (range, 35 – 95 degrees) at the end of the bone lengthening with recovery of normal range by the end of follow up. There was no significant reduction of hip ROM from preoperative to postoperative flexion or extension. The mean loss of knee extension by the end of follow up decreased by 10 degrees (5-15 degrees).

In two patients, knee subluxation and antero-posterior displacement of the osteotomy appeared 10 weeks postoperative solved by reduction of the knee and correction of the osteotomy with external fixator adjustment, then continued lengthening and 2 crossing k-wires for alignment (Figure 2). In those patients limited knee ROM of 30 degrees and needed quadriceps plasty after complete union of lengthening site (42 weeks). The gained bone lengthening was greater in female patients than male patients (6.5 cm; range, 4.5 – 8.5 cm in females and 5cm; range, 4 – 6.5 cm in males), (p =0.023).

**Clinically**

According to the PODCI score categories surveyed, the standardized and normative scores were accepted (Table 1). It was noted that in the patients with femoral lengthening greater than 6 cm the global functioning scores were lower than that in the patients with less than 6 cm lengthening (88; SD ± 8 versus 94; SD ± 2 ; p = 0.056). The pain/comfort scores were less in patients needed lengthening of more than 25% of preoperative femoral length (74; SD ± 20 versus 90; SD ± 6; p = 0.032).

**Table 1. the PODCI score for all patients after completing the lengthening procedure**

<table>
<thead>
<tr>
<th>PODCI category</th>
<th>Standardized score</th>
<th>Normative score</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean ± SD</td>
<td>Range</td>
</tr>
<tr>
<td>Upper extremity</td>
<td>96 ± 3</td>
<td>86 -100</td>
</tr>
<tr>
<td>Basic mobility</td>
<td>95 ± 4</td>
<td>90 -100</td>
</tr>
<tr>
<td>Sports and physical</td>
<td>85 ± 9</td>
<td>75 – 100</td>
</tr>
<tr>
<td>functioning</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pain / comfort</td>
<td>90 ± 11</td>
<td>55 -100</td>
</tr>
<tr>
<td>Happiness</td>
<td>91 ± 8</td>
<td>65 -100</td>
</tr>
<tr>
<td>Global functioning</td>
<td>92 ± 5</td>
<td>80 - 100</td>
</tr>
</tbody>
</table>

**Complications**

The rate of complications was 50% (8 of 16) (Table 2). Knee subluxation and antero-posterior displacement of the osteotomy treated with correction of the osteotomy with external fixator adjustment, then continued lengthening.

**Table 2. complications during follow up period**

<table>
<thead>
<tr>
<th>Complication</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Knee subluxation ( readjustment of fixator)</td>
<td>2</td>
</tr>
<tr>
<td>Pin tract infection ( conservative treatment)</td>
<td>5</td>
</tr>
<tr>
<td>Fracture after removal of fixator (plate fixation)</td>
<td>1</td>
</tr>
</tbody>
</table>

**Discussion**

The bone lengthening to correct the femoral length discrepancy is a procedure that may need a variety of surgical techniques and the published literature on heterogeneous groups of patients have mixed results and complications.
In a recent study conducted by Prince DE; et al 2015 they presented objective radiographic and clinical data reporting the patients function after performing lengthening with an external fixator plus intramedullary rodding and reported a mean lengthening of 6 cm and 1.3 months/cm lengthening index. Both hip and knee ROM, were maintained although 50% rate of complications during treatment.

Aston et al. in their study achieved mean 6 cm femoral lengthening [13]. They incorporated the use of intramedullary rod fixation surgery and lengthening over the rods. Other authors recommended the rods fixation after lengthening at the time of external fixation removal, they recommended the rodding after removal of fixator to give the chance of using larger intramedullary nail to protect the entire lengthened bone to prevent fracture and deformity through the regeneration [20, 22, and 25]. In this study 16 patients with congenital femoral deficiency were treated with bone lengthening using unilateral external fixator with 2 intramedullary 2mm crossing k-wires at the same time of applying the fixator in a mean follow up of 36 months. The postoperative knee flexion, complication rates, and the amount lengthening obtained were similar to those of published literatures using the same technique [20 - 25].

Physical therapy is one of the main methods to regain hip and knee function and ROM. In this study the physical therapy was performed in our institute using the same modalities and techniques with the record of results by physical therapists.

The serial x-rays were performed in radiology department of our institute using the same magnification markers to record the follow up changes. The reported healing index for every cm is 1.3 months in this study, and these results is comparable with published results of femoral lengthening for patients with congenital femoral deficiency Paley Type 1 [15, 17, and 18]. It is unclear to the authors why female patients have more length gaining than male patients (average 2 cm). It is claimed to be due to neurodevelopmental stage of each gender [20].

In this study, the range of motion (ROM) decreases during lengthening, but normal knee and hip ROM was regained at the end of follow up. These results are similar to Herzenberg et al study in a series of 25 patients who underwent lengthening of average 6 cm on isolated femoral shortening, in those patients knee ROM decreased to a minimum of 37 degrees at the end of lengthening, but increased to 69 degrees at last follow up [16].

Popkov et al. in their study using the Ilizarov technique for lengthening in 86 children (59 femoral and 55 tibial). The mean bone age was 8.5 years at time of lengthening for various congenital conditions. They published a comprehensive report on factors that influence postoperative growth after limb lengthening [18]. The average follow up period in their study was 4.5 years after fixator removal. The mean femoral lengthening was 5cm. Some reports in the literature found that in young patients there is a stimulatory effects [19], while others have found no change in growth rate in older patients [9, 15, 17, 28]. In this study we could not support the hypothesis of stimulation of growth in younger patients which may be due to the small number of cases and the relatively short follow up period.

The authors used the PODCI scores reported that to maintain higher function and less pain after lengthening you should avoid excessive lengthening beyond 6 cm or more than 25% lengthening of the femur [20]. The rate of complications after lengthening surgery for congenital femoral shortening in this study are within the range of published reports [16, 29, and 30]. Complications were found in 50% of patients. Oostenbroek et al. reported a complication rate of 69% when the limb length discrepancy is the only predictor for complications after surgery [16].

**Conclusion**

The surgical technique of femoral lengthening in patients with Paley Type 1A proximal femoral deficiency is a successful and well tolerated technique. The good results of the surgical technique of distraction osteogenesis
in younger patients, with 25% of the femur maximum lengthening is the key for preserving the knee and hip ROM. The qualified physical therapist doing and recording the intensive physiotherapy is one factor of gaining good function. The limitations of this study are the small number of the sample and short follow up period.

**REFERENCES**


