American Research Journal of Orthopedics and Traumatology (ARJOT)
Volume 2016, 8 Pages

AMERICAN RESEARCH JOURNALS
An Academic Publishing House

Research Article Open Access

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

Adel Mohammed Salama, M. D.

Assistant professor, orthopedic surgery department, Faculty of Medicine, Zagazig University, Egypt
yousufmmkh@gmail.com

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.

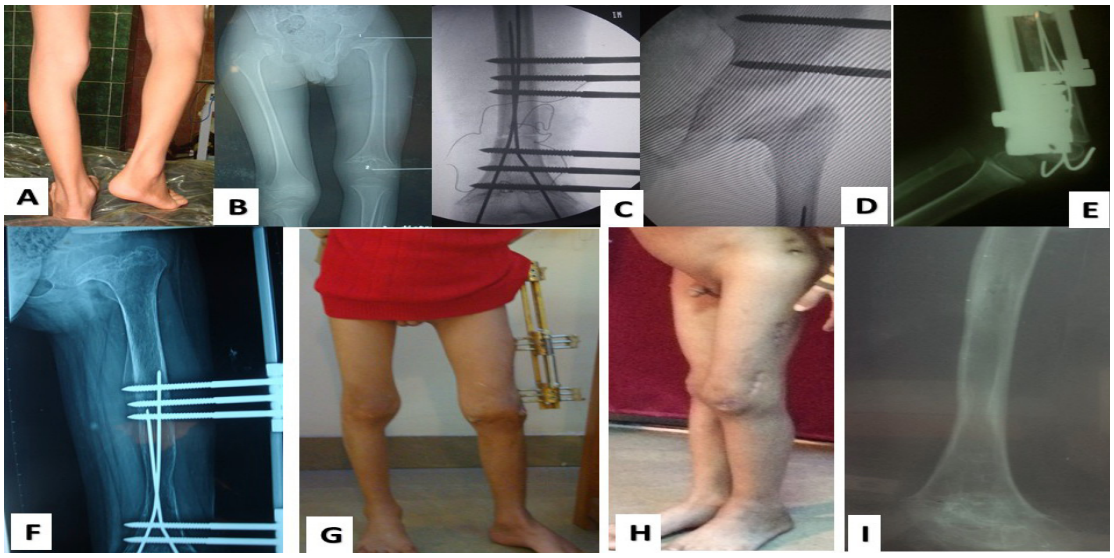

Fig1. *Male patient 6 years old with left congenital short femur A) preoperative photo showing shortening of left leg; B) preoperative scanogram with 5 cm femoral shortening; C & D) intraoperative radiology; E) knee subluxation during lengthening; F)X-ray7 months postoperative; G& H) photo of the patient with equalized limb length in external fixator; I) x-ray 2 years postoperative showing the femur after lengthening and union.*

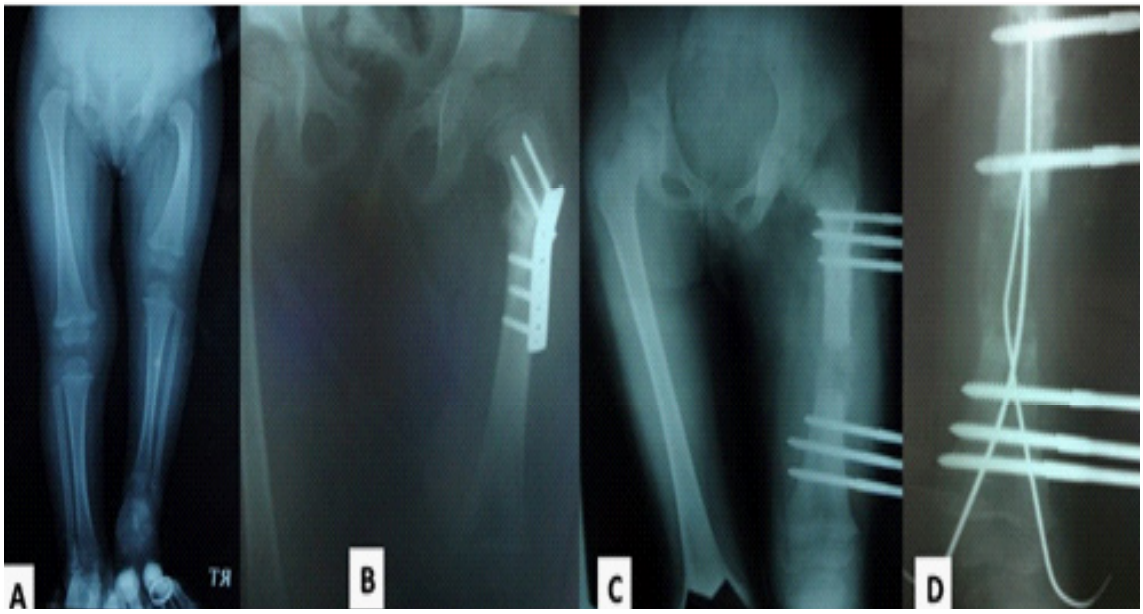

Fig2. *4.5 years old female with congenital short femur left side; A) preoperative x-ray showing left femoral shortening with coxa vara; B) x-ray after correction of coxa vara using plate fixation on valgus osteotomy of proximal femur; C) x-ray after second operation for lengthening with removal of the 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 7.5years (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.

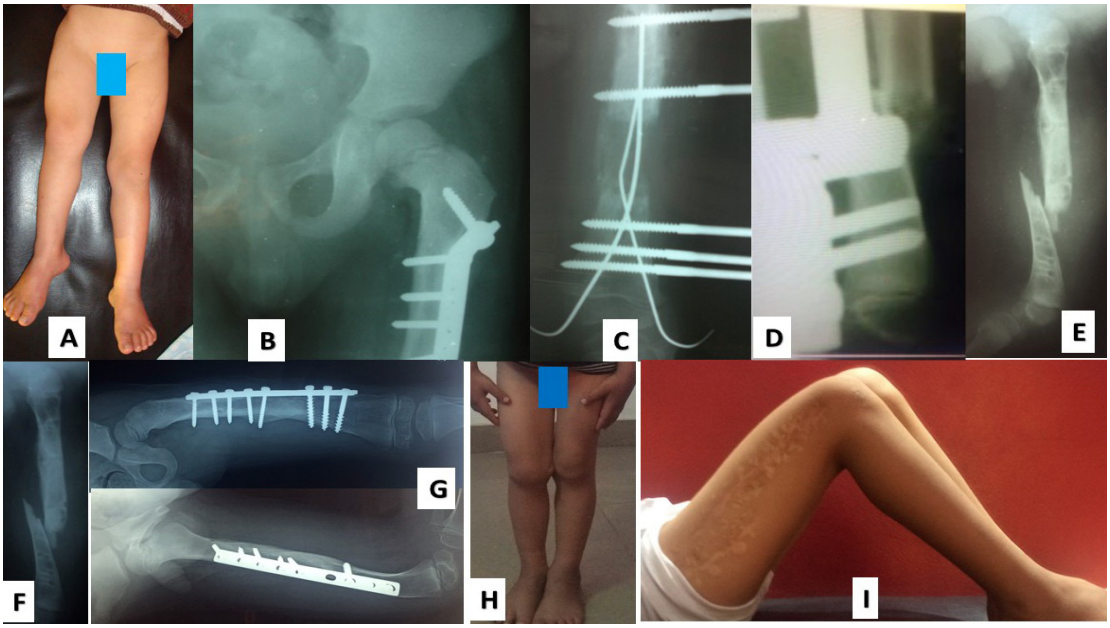

Fig3. *Male patient 5 years old with right side congenital short femur A) preoperative photo with short femur; B) x-ray after correction of coxa vara; C) x-ray 6 months postoperative with callus formation in the lengthened segment; D)X-ray photo after plate fixation due to fracture after removal of external 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.*

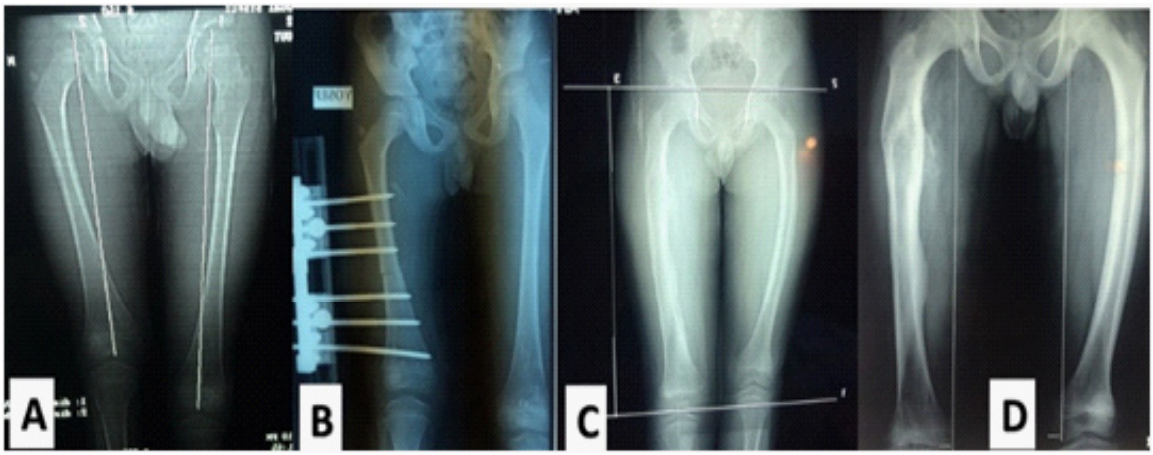

Fig4. *Male patient 6.5 years old with short right femur: A) preoperative scanogram with 4.5 cm shortening; B) x-ray after osteotomy and fixator; C) scanogram after finishing lengthening with2cm overlengthening; D) x-ray of femur after consolidation.*

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 3cm) 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*

PODCI category	Standardized score		Normative score	
	Mean ± SD	Range	Mean ± SD	Range
Upper extremity	96 ± 3	86 -100	54 ± 2	44 - 56
Basic mobility	95 ± 4	90 -100	50 ± 4	40 - 52
Sports and physical functioning	85 ± 9	75 – 100	44 ± 10	23 - 52
Pain / comfort	90 ± 11	55 -100	48 ± 8	25 -53
Happiness	91 ± 8	65 -100	51 ± 7	20 – 55
Global functioning	92 ± 5	80 - 100	49 ± 6	22 - 56

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*

Complication	Number of patients
Knee subluxation (readjustment of fixator)	2
Pin tract infection (conservative treatment)	5
Fracture after removal of fixator (plate fixation)	1

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

1. Aitken GT. Proximal femoral focal deficiency–definition, classification, and management. In: Aitken GT, ed. Proximal Femoral Focal Deficiency. A Congenital Anomaly. Washington, DC, USA: National Academy of Sciences; 1969:1–22.
2. Torode IP, Gillespie R. The classification and treatment of proximal femoral deficiencies. Prosthet Orthot Int. 1991; 15: 117–126.
3. Koman LA, Meyer LC, Warren FH. Proximal femoral focal deficiency: a50-year experience. Dev Med Child Neurol.1982; 24:344–355.
4. Herring JA, Birch JG. Congenital femoral deficiency. In: Herring JA, Birch JG, eds. The Child With a Limb Deficiency. Rosemont, IL, USA: American Academy of Orthopaedic Surgeons; 1998: 61–150.
5. Gillespie R, Torode IP. Classification and management of congenital abnormalities of the femur. J Bone Joint Surg Br. 1983; 65:557–568.
6. Amstutz HC, Wilson PD. Dysgenesis of the proximal femur (coxa vara) and its surgical management. J Bone Joint Surg Am. 1962; 44:1–24.
7. Ackman J, Altiok H, Flanagan A, Peer M, Graf A, Krzak J, Hassani S, Eastwood D, Harris GF. Long-term follow-up of Van Nes rotationplasty in patients with congenital proximal focal femoral deficiency. Bone Joint J. 2013; 95:192–198.
8. Van Nes CP. Methods of treating pseudoarthrosis of the femoral neck, and their indications. Arch Chir Neerl. 1959:11:327–342.
9. Westberry DE, Davids JR. Proximal focal femoral deficiency (PFFD): management options and controversies. Hip Int. 2009; 19 (Suppl 6):S18–25.
10. Simpson-White RW, Fernandes JA, Bell MJ. King's procedure for Aitken B/Paley 2a proximal femoral focal deficiency with 19year follow-up–a case report. Acta Orthop. 2013; 84:323–325.
11. Stephens MM, Hsu LC, Leong JC. Leg length discrepancy after femoral shaft fractures in children. Review after skeletal maturity. J Bone Joint Surg Br. 1989; 71:615–618.
12. Viehweger E, Pouliquen J-C, Kassis B, Glorion C, Langlais J. Bone growth after lengthening of the lower limb in children. J Pediatr Orthop B. 1998; 7:154–157.
13. Aston WJS, Calder PR, Baker D, Hartley J, Hill RA. Lengthening of the congenital short femur using the Ilizarov technique: a single-surgeon series. J Bone Joint Surg Br. 2009; 91:962–967.
14. Paley D. Problems, obstacles, and complications of limb lengthening by the Ilizarov technique. Clin Orthop Relat Res. 1990; 250:81–104.
15. Oostenbroek HJ, Brand R, van Roermund PM. Growth rate after limb deformity correction by the Ilizarov method with or without knee joint distraction. Acta Orthop. 2009; 80:338–343.
16. Oostenbroek HJ, Brand R, van Roermund PM. Lower limb deformity due to failed trauma treatment corrected with the Ilizarov technique. Acta Orthop. 2009; 80:435–439.
17. Hope PG, Crawfurd EJ, Catterall A. Bone growth following lengthening for congenital shortening of the lower limb. JPediatr Orthop. 1994; 14:339–342.

18. Popkov D, Journeau P, Popkov A, Pedeutour B, Haumont T, Lascombes P. Analysis of segmental residual growth after progressive bone lengthening in congenital lower limb deformity. Orthop Traumatol Surg Res. 2012; 98:621–628.

19. Sabharwal S, Paley D, Bhave A, Herzenberg JE. Growth patterns after lengthening of congenitally short lower limbs in young children. J Pediatr Orthop. 2000;20:137–145.

20. Prince DE, Herzenberg JE, Standard SC, and Paley D. Lengthening With External Fixation Is Effective in Congenital Femoral Deficiency. Clin Orthop Relat Res. 2015 Oct; 473(10):3261-71.

21. Paley D, Bhave A, Herzenberg JE, Bowen JR. Multiplier method for predicting limb-length discrepancy. J Bone Joint Surg Am. 2000; 82:1432–1446.

22. Paley D, Standard SC. Lengthening reconstruction surgery: for congenital femoral deficiency. In: Rozbruch SR, Ilizarov S, eds. Limb Lengthening and Reconstruction Surgery. Boca Raton, FL, USA: CRC Press; 2006:393–428.

23. Paley D, Tetsworth K. Mechanical axis deviation of the lower limbs. Preoperative planning of uniapical angular deformities of the tibia or femur. Clin Orthop Relat Res. 1992; 280:48–64.

24. Paley D, Tetsworth K. Mechanical axis deviation of the lower limbs. Preoperative planning of multiapical frontal plane angular and bowing deformities of the femur and tibia. Clin Orthop Relat Res. 1992; 280:65–71.

25. Paley D. Principles of Deformity Correction. 1st ed. New York, NY, USA: Springer; 2003:1–821.

26. Haynes RJ, Sullivan E. The Pediatric Orthopaedic Society of North America Pediatric Orthopaedic Functional Health Questionnaire: an analysis of normals. J Pediatr Orthop. 2001; 21:619–621.

27. Herzenberg JE, Scheufele LL, Paley D, Bechtel R, Tepper S. Knee range of motion in isolated femoral lengthening. Clin Orthop Relat Res. 1994; 301:49–54.

28. McCarthy JJ, Kim H, Saluan P, Karsky D, Davidson RS. The effects of limb lengthening on growth. JP ediatr Orthop B.2003; 12:328.

29. Nogueira MP, Paley D, Bhave A, Herbert A, Nocente C, Herzenberg JE. Nerve lesions associated with limb-lengthening. J Bone Joint Surg Am. 2003; 85:1502–1510.

30. Paley D, Herzenberg JE, Tetsworth K, McKie J, Bhave A. Deformity planning for frontal and sagittal plane corrective osteotomies. Orthop Clin North Am. 1994; 25:425–465.

Citation: Adel Mohammed Salama, "*Bone Lengthening for Management of Type 1a Proximal Femoral Deficiency with or without Varus Deformity". American Research Journal of Orthopedics and Traumatology; Volume 1, 2016; pp:1-8*