

Case Report

Double-level osteotomy and one-stage reconstruction with long intramedullary femoral nail to correct a severe proximal and diaphyseal femur deformity in a patient with polyostotic fibrous dysplasia: case report and literatures review

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Abstract: Proximal femur is often involved with varus and retroversion deformity in polyostotic fibrous dysplasia (PFD). Multiple corrective osteotomies with intramedullary nails in two stages is recommended procedure as some authors described. We report a case using double-level osteotomy and one-stage reconstruction with intramedullary nail in a patient with painful proximal femur and diaphysis varus deformity, the neck-shaft angle was corrected from 95° pre-operatively to 125° post-operatively, the patient was free of pain and no evidence of recurrence at the 24-month follow up. The operative design and method were described, and a review of related literatures about the treatment alteration for PFD and relevant operative selection were also performed.

Keywords: Polyostotic fibrous dysplasia, Shepherd's deformity, osteotomy, internal-fixation

Introduction

Polyostotic fibrous dysplasia (PFD) is one of the subtypes of fibrous dysplasia result from somatic gene mutation of Gs- α [1-7], the proximal femur is often involved associated with Shepherd's deformity, limb shortening, limping and sometimes pathologic fracture. Although PFD can't be thoroughly treated by surgery, the increasing deformity, continuous pain, and pathological fracture are indications for operation as a common sense to this day [8]. For fixation with plates and screws is difficult to fit the contour of deformed femur, and may induce secondary deformities [9, 10], multiple corrective osteotomies with intramedullary nails in two stages is recommended procedure for a severe varus deformity [11-13]. We report a case using double-level osteotomy and one-stage reconstruction with a long femur interlocking intramedullary nail in a patient with severe varus deformity in both proximal and diaphysis of left femur, and continuous pain, who had pathological stress fracture history.

The peri-operative, operative steps and follow up result were described. Related literatures about the treatment and relevant operative methods for PFD were also reviewed.

Case report

Patient information and diagnosis

The patient was a 24-year-old woman suffering from PFD with left femur varus deformity, who had pathological fracture at left femur diaphysis and received operation at the age of 17-year-old. She complained of pain at the anterior region of left thigh at walking for 1 year. Limp and 4cm shortening of left leg could be seen at physical examination (**Figure 1A**). Pre-operative roentgenograms and 3-dimensional computed tomography (**Figure 1B, 1C**) revealed severe varus deformity of the left femur, the appearance of "ground glass" in the proximal region of femur, with widening of the medullary cavity. Bone scintigraphy showed abnormal uptake of the entire left lower extremity from pelvis to

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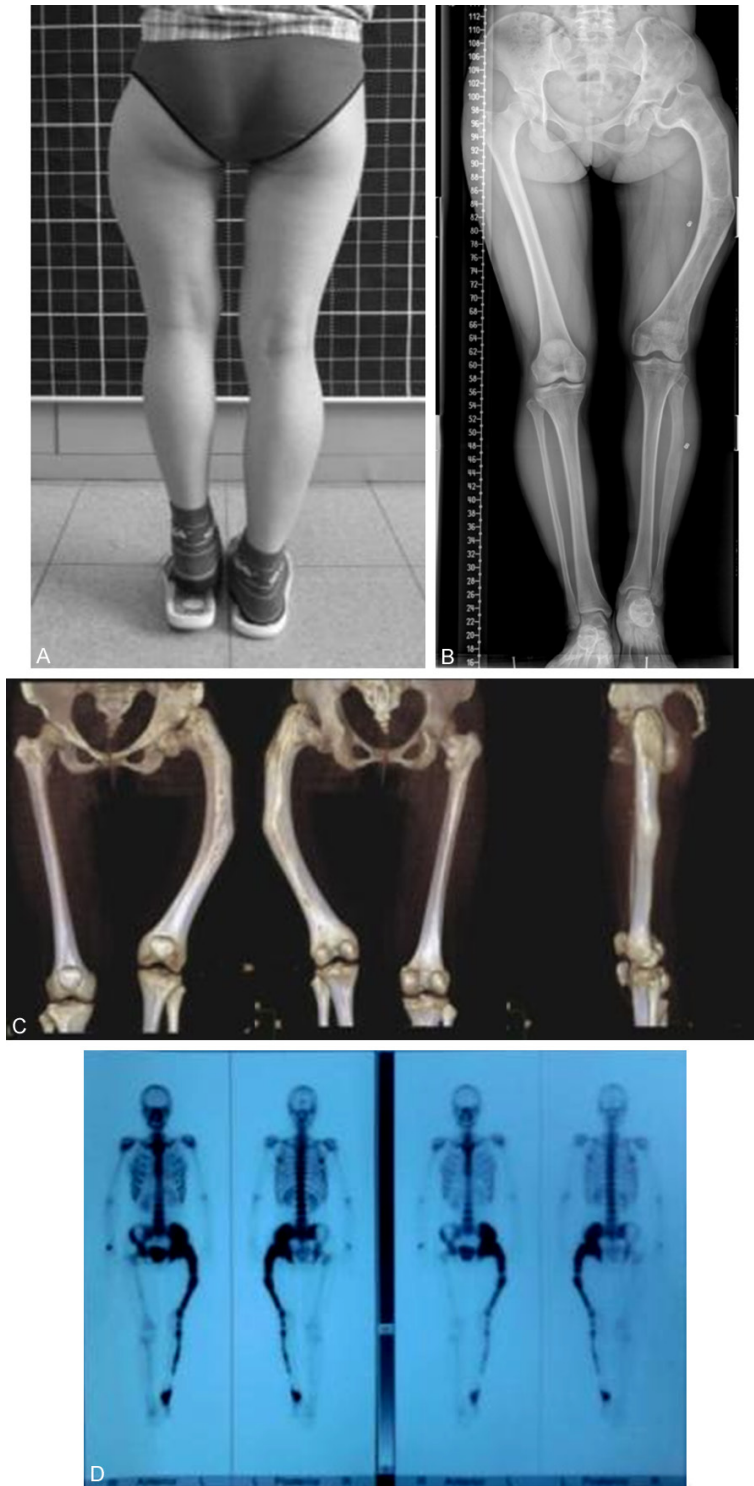


Figure 1. Preoperative (A) posterior appearance showed lateral bow deformity and shorter of the left lower extremity. (B and C) Pre-operative anterior-posterior lower extremity whole length roentgenograms and 3D-CT revealed severe varus deformity of the left femur, the appearance of “ground glass” in the proximal region of femur, with widening of the medullary cavity, varus apex were at subtrochanter and middle diaphysis. (D) Bone scintigraphy showed abnormal uptake of the entire left lower extremity from pelvis to ankle, confirmed the nature of PFD.

ankle which confirmed the nature of the lesion of PFD (**Figure 1D**).

Pre-operative planning

The radiographs were used to measure and determine the appropriate angle and location for osteotomy (**Figure 2A**), the length of left leg was 40 mm shorter than the right one on total length lower extremity radiograph, and the discrepancy happened in femur. The left neck-shaft angle was 95° , which was 125° on the right side. The varus apex happened at the subtrochanteric region and middle of the diaphysis where the previous fracture happened, the angle was 155° and 155° respectively. Lateral closed wedged osteotomy was decided, the osteotomy level was decided to be at the apex of varus, which meant subtrochanter and middle of femur diaphysis, the angle was calculated to be 50° at subtrochanter apex in order to correct both the varus of neck-shaft angle and proximal femur, and 25° at middle diaphyseal apex respectively (**Figure 2B**). The preplanned osteotomy site was drawn on tracing paper (**Figure 2C**). Since the total length of left femur was no shorter than the right side, and even 20 mm longer, the discrepancy could be corrected from 40 mm to 15 mm as calculated, bone graft was not taken into account.

Intra-operative management and follow up result

The patient was placed in lateral position under an image intensifier. Intraoperatively, lateral closed wedge osteotomy was performed where the

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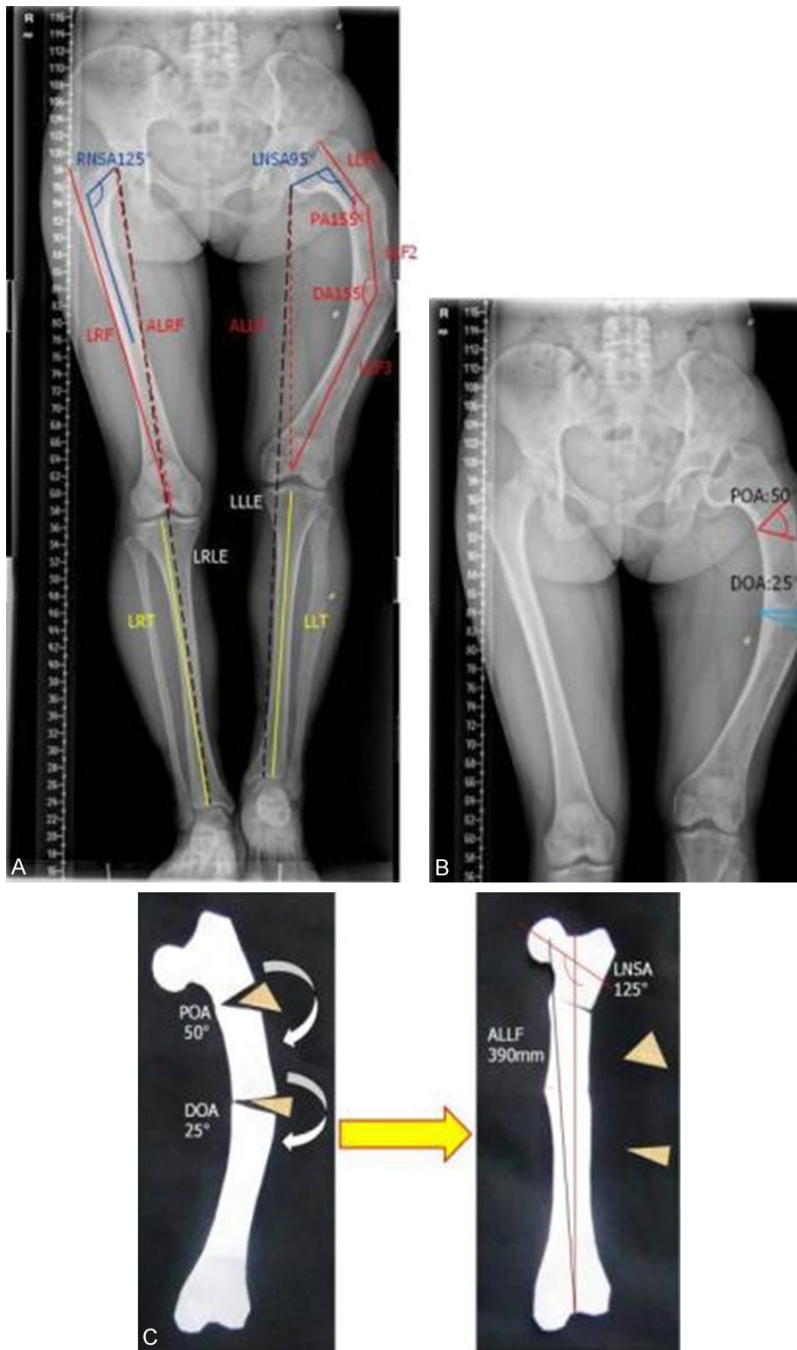


Figure 2. Preoperative measuring and planning (A) RNSA (right neck-shaft angle) 125°, LNSA (left neck-shaft angle) 95°, LRF (length of right femur) 410 mm, LLL1 (length of left femur) +LLF2+LLF3=430 (100+100+230) mm, even 20 mm longer than the right, ALRF (absolute length of right femur) 405 mm, ALLF (absolute length of left femur) 365 mm, which means 40 mm shorter than that of the right, LRT and LLT (length of right tibia and length of left tibia) the same length, 340 mm, LRLE (length of right lower extremity) 755 mm, LLE (length of left lower extremity) 715 mm, 40mm shorter than right one, which happened in femur. (B) Calculated angles POA (proximal osteotomy angle) 50°, DOA (distal osteotomy angle) 25°. (C) Diagram of osteotomy, after the double-level lateral closed osteotomy, the LNSA could be corrected to 125°, and ALLF be corrected from 355 mm to 390 mm, which is 15 mm shorter than that of right (405 mm).

guide pin and cortex were touching, and confirmed to be the site preplanned on image intensifier. Two-level osteotomy was performed at the subtrochanter and femur diaphyseal as preplanned (**Figure 3**). During reaming, removal of condensed fibrous tissues from the intramedullary cavity was performed. After reaming, a long femoral interlocking nail (Φ 11 mm, L 340 mm, Synthes Expert A2FN, Switzerland) was inserted and standard interlocking was performed. The fibrous tissues from intramedullary cavity were sent for pathologic examination and confirmed to be fibrous dysplasia. Postoperative radiograph showed that the neck-shaft angle was corrected from 95° to 125°, the limb length discrepancy was corrected from 40 mm to 15 mm (**Figure 4**). The patient was allowed for partial weight bearing 2 weeks after operation, and walking time with full weight bearing was 3 months based on follow-up radiograph. At 12-months post-operation follow-up, the proximal transverse screw was taken out, and there were no infection or other complications, the patient had no pain on the left leg, no limp was found and the osteotomy healed at the 24-month follow up (**Figure 5**).

Discussion

Fibrous dysplasia is a benign intramedullary fibroosseous lesion first de-

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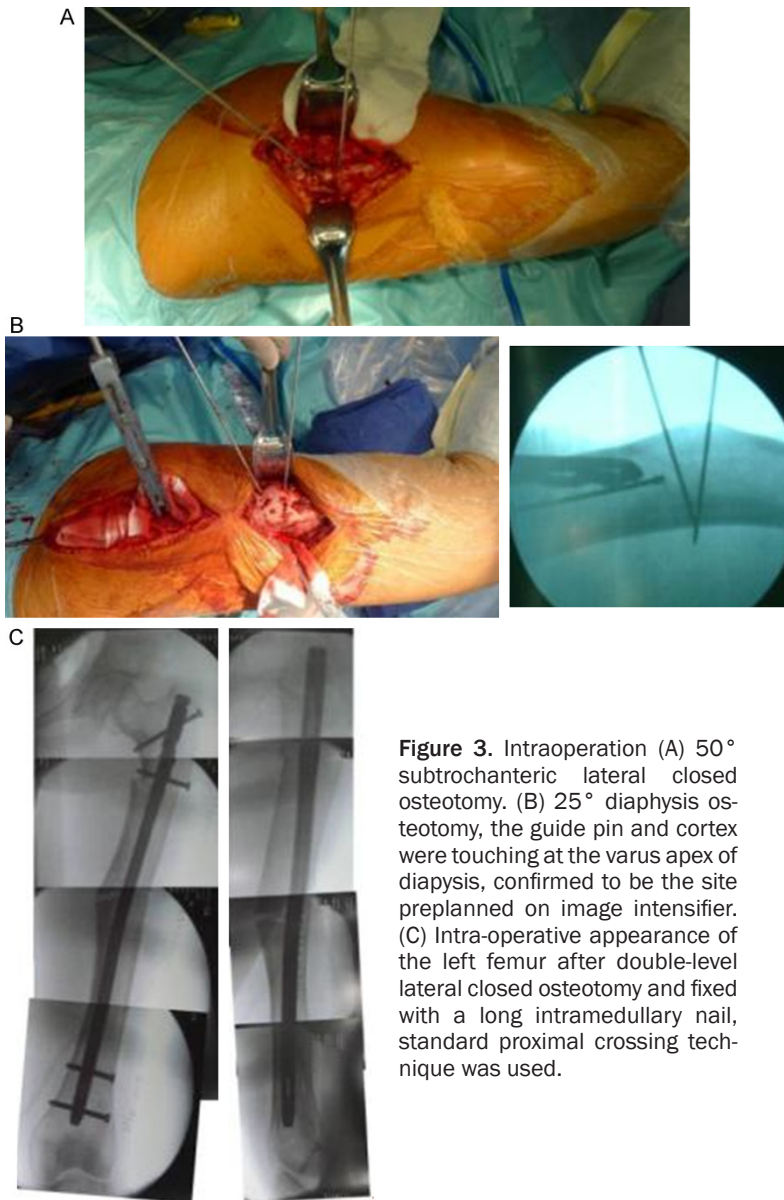


Figure 3. Intraoperation (A) 50° subtrochanteric lateral closed osteotomy. (B) 25° diaphysis osteotomy, the guide pin and cortex were touching at the varus apex of diaphysis, confirmed to be the site preplanned on image intensifier. (C) Intra-operative appearance of the left femur after double-level lateral closed osteotomy and fixed with a long intramedullary nail, standard proximal crossing technique was used.

scribed by Lichtenstein in 1938 [14], and can be classified into monostotic, polyostotic, and others (McCune-Albright disease). The etiology is linked with a mutation in the *Gsα* gene that occurs after fertilization in somatic cells [1-7]. Unlike monostotic disease which will cease after skeletal maturity, deformities in PFD may continue to progress in adult patients. Shepherd's crook deformity of the proximal femur is classic deformity of PFD, which is a result of intermittent fatigue fractures deforming from normal mechanical forces [8].

PFD is a genetic disease, which may be solved with the breakthrough of genetic re-engineer-

ing in the future, but to this day, only medical management and surgery can be used to control this condition. Though we didn't use it in the current case, bisphosphonate is recommended for symptomatic polyostotic cases [3, 4, 15-17], and reported to decrease bone pain intensity, improve radiographic manifestation with thickening of cortices and/or ossification of radiolucent areas.

Surgical intervention is indicated for correction of deformity, prevention or treating of pathologic fracture, and/or eradication of symptomatic lesions [2, 8]. Osteotomies and fixation devices and/or cortical grafts are recommended to achieve the goals of deformity correction and restoration of function, and it is better to postpone treatment until skeletal maturity to reduce the risk of recurrence with subsequent growth [8]. In our current case, the patient was 24 years old with skeletal maturity with definite pathological fracture history, and severe varus deformity both in the proximal femur and diaphysis, contin-

uous anterior thigh pain, which indicated for surgical intervention. The concentrations involved in the operation include the lesion curettage, bone graft or not, osteotomy, and selection of fixation device, which will be discussed as follows.

As for the lesion, traditional simple curettage and cancellous bone-grafting maybe useful in those monostotic disease in adults, but can't correct deformities or relieve symptoms for those active lesions in PFD cases [10, 18-21]. Simple curettage is associated with high risk of recurrence. Funk and Wells [20] indicated that polyostotic lesion involving proximal femur was

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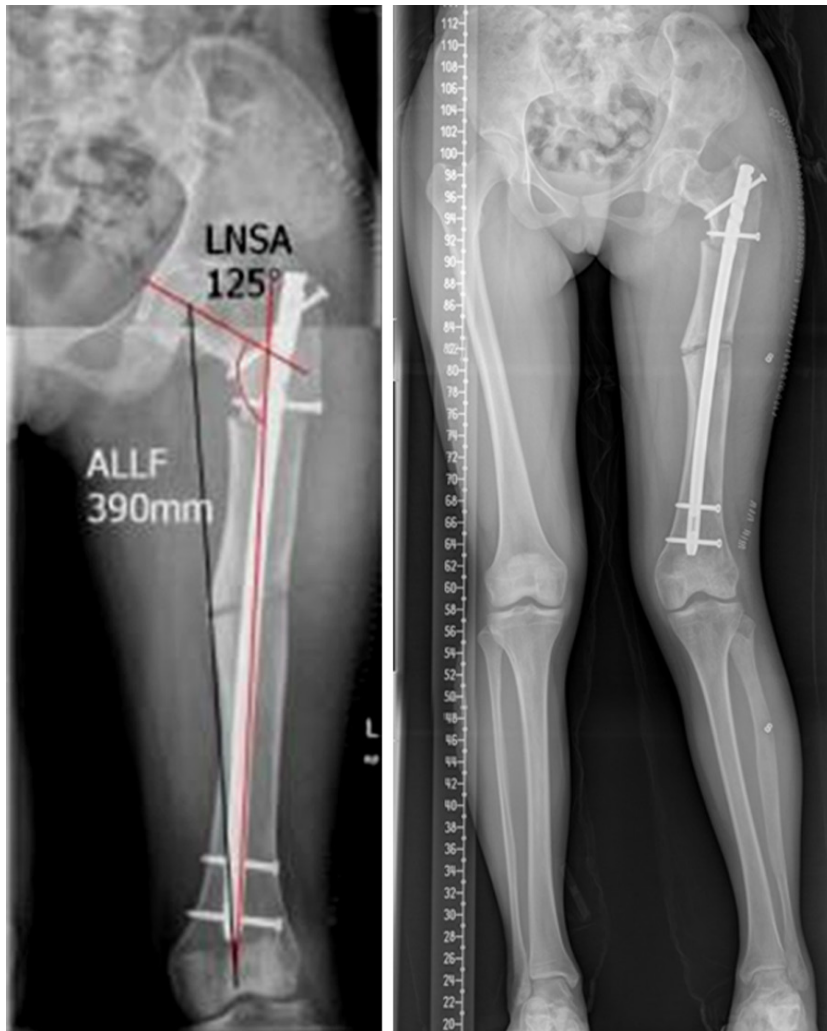


Figure 4. Postoperative left femur and lower extremity whole length radiograph (AP), the LNSA (left neck-shaft angle) was corrected to 125° , the ALLF (absolute length of left femur) was 390 mm, 15 mm shorter than the right one, which was consistent with preoperative planning.

complex than monostotic lesion and difficult to correct. Enneking [22] et al. reported their experience of using autogenous cortical bone-grafting to treat symptomatic lesion of femoral neck, slow resorption of the initial graft happened in two cases. DiCaprio [8] et al. recommended allogeneic cortical bone or vascularized bone graft for treatment of PFD if necessary. Guille [10] et al. reported on a larger series of lesions treated with curettage and autogenous bone grafting, results showed complete resorption of all autogenous cancellous bone graft, none of the lesions had been eradicated or decreased in size radiographically. Watanabe [23] concluded that curettage and bone grafting do not appear to be efficacious in the treat-

ment of fibrous dysplasia. Stanton [2] considered it unnecessary to use cortical grafts in treating of proximal femur deformity in PFD. Based on these studies, combined with the current case with no structural bone defect in the involved region, we didn't use any bone graft in this case, but repeated removal of the condensed fibrous tissues was performed during the operation.

Osteotomy is the major objective in correction of the deformity for PFD. Since the involved region of shepherd's crook deformity is at the proximal femur of neck-shaft and trochanter, correction osteotomy level is often at the subtrochanteric region, and closed wedge osteotomy is recommended [10-12, 24, 25]. Guille et al [10] advocated a valgus osteotomy and internal fixation early in the course of PFD with varus deformity of the proximal femur. Medial

displacement valgus osteotomy and overcorrection is recommended if the calcar of femoral neck is involved to provide a mechanically favorable position for healing of the microfracture [2, 10, 26]. The diaphyseal deformity may occur after pathologic fracture and sometimes need additional osteotomy. Yamamoto [27] et al. introduced the methods and advantage of oblique wedge osteotomy used in a case with fibrous dysplasia who had a three-dimensional diaphyseal deformity in left femur, two years of follow-up result showed good union and alignment of the femur and no progression of the varus deformity. In the current case, because deformity of the femur was complicated with both proximal and diaphyseal varus, the neck-

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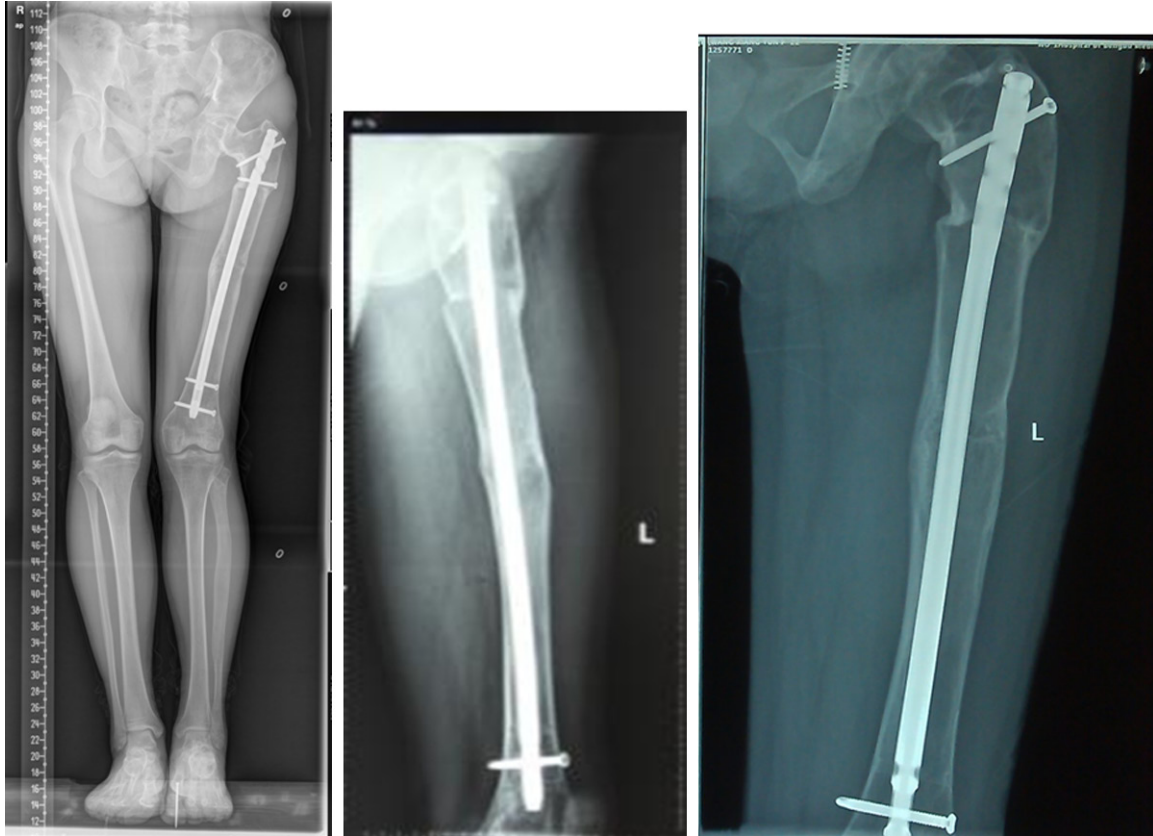


Figure 5. 12-month (left) post-operation, the proximal transverse screw was taken out, and 24-month (right) follow up radiograph showed healing of the osteotomy and nearly equivalent length of the lower extremities, no varus progress in the left proximal femur, and no limp was found.

shaft angle was 95°, double-level osteotomy was taken into account, as measured and designed on the preoperative radiograph as mentioned above, double-level of lateral closed osteotomy was performed as preplanned, the result of correction was satisfied as expected.

Exact fixation is important to promise the correction of Shepherd's deformity and preventing of refracture. Fixation with plate and screw has been widely used. Breck et al [28] reported treatment of fibrous dysplasia by total femoral plating and hip nailing. Chen [25] et al. reported two cases of shepherd's crook deformity of PFD treated with corrective osteotomy and dynamic hip screw (DHS), no recurrence or refracture was found with 2-11 years of follow-up. But other reports indicated that concentration of stress on the lower portion of the plates may induce secondary deformities, also it is difficult to fit plates conforming to the contour of the deformed femur [9, 10]. Stanton [2] had sug-

gested "Do not use plates and screws" in his "six do NOT work" procedure in the surgery for PFD.

Intramedullary fixation was reported in the correction of deformity after osteotomy. Several decades ago, Connolly [29] reported using of Zickel nail to treat shepherd's crook deformities of polyostotic fibrous dysplasia after osteotomy in a 6-year-old boy, long-term of follow-up showed that the nail maintained correction of the femoral deformities. O'Sullivan et al [28] used intramedullary rodding and bisphosphonate to treat PFD. For those severe varus retroversion deformities, corrective osteotomies using intramedullary devices in two stages is recommended procedure [11-13]. Nagda [12] et al. recommended subtrochanteric osteotomies with intramedullary fixation in the first stage and intertrochanteric osteotomies in the second stage with nail plate fixation, and considered it having the advantage of avoiding pro-

longed immobilization and preventing the recurrence of deformity. Some authors prefer one-stage operation for correction of shepherd's deformity in adult patient. Jung et al [11] recommended the use of multiple osteotomies and intramedullary nail to correct shepherd's deformity, they reported 7 femurs treated with this method, the result showed that neck-shaft angle was obtained from 92° preoperatively to 129° after surgery, no recurrence or refracture occurred. In their report, proximal crossing screws were crossed to the femoral neck in all cases, the entry point of nail was piriform fossa, which made the approach difficult because of the varus deformity of proximal femur. Kataria et al [24] reported a case where a subtrochanteric osteotomy and intramedullary nailing used to correct severe shepherd's deformity of the proximal femur with pathological fracture in a patient with monostotic disease, the neck-shaft angle was corrected to 125°, and no pain or limp at 57-month follow-up. In his case, the osteotomy was one level (subtrochanter) and the nail was also crossing to the femoral neck. Yamamoto [27] used Huckstep nail to fix the femur diaphysis after osteotomy, which is recommended for fixation of mechanically deficient femoral bones and allows the use of multiple transverse screws and providing adequate stability. The anterograde intramedullary nail we used in the current case was preferred because (1) the entry point is at the greater trochanteric tip, which is rather easier to insert than from the piriform fossa; (2) it has long central lever arm to hold in the distal diaphysis to promise the distal stability; (3) multiple selections for proximal crossing. As for the bone structure of femoral neck and intertrochanter is good and stable enough, the standard crossing was selected finally. The result of follow-up showed no progressive varus or fracture in the neck-shaft and intertrochanteric region. Nonetheless, the reaming process was difficult because of the dense fibrous tissue as other authors have reported.

Some authors [23, 26] considered it difficult to use internal fixation in correction of shepherd's deformity, especially in those complicated deformities needing multiple osteotomies, and recommended the use of external devices. Watanabe [23] et al. reported a successful case of double-level correction with a Taylor Spatial Frame (TSF) for varus retroversion

deformity of the proximal femur in a nine-year-old patient with PFD. Sakurakichi [26] et al. introduced Ilizarov technique to correct the shepherd's crook deformities in two cases. The authors concluded that the advantages of using external devices in correction of shepherd's deformity is the ability to modify the correction, lengthen limbs, minimize invasion, and early weight bearing. Despite advantages as authors described, deformity correction with external fixation is technique demanding, the inconvenience for sleeping, difficult postoperative care and relevant postoperative complications should be taken into account.

In conclusion, although surgery will never cure polyostotic fibrous dysplasia, appropriate surgical intervention can and should be considered early for correction of deformity, prevention or treating of pathologic fracture, and/or eradication of symptomatic lesions. Combined with the literatures and our limited experience, we believe that double-level osteotomy and one-stage reconstruction with an long interlocking nail is effective and practical method for treating the severe symptomatic varus deformity of PFD, at the same time, meticulous preoperative measurement and designing is crucial to promise the correction of alignment, equal length and the full contact of osteotomy surface, appropriate internal nailing fixation can provide exact stability and maintain the alignment of the femur.

Disclosure of conflict of interest

None.

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