

Valgus osteotomy combined with intramedullary nail for Shepherd's crook deformity in fibrous dysplasia: 14 femurs with a minimum of 4 years follow-up

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Received: 31 March 2009 / Published online: 24 July 2009
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Abstract

Background and objectives To investigate the effectiveness of valgus osteotomy combined with intramedullary nail in treatment of Shepherd's crook deformity of fibrous dysplasia.

Method A retrospective study was performed in 13 patients (14 femora) of fibrous dysplasia who were treated in our hospital between August 2000 and February 2005. All patients had Shepherd's crook deformity. Six patients had monostotic disease, and seven had polyostotic disease. There were seven males and six females. The four-step procedure was performed orderly as valgus osteotomy, curettage lesion, massive impaction allograft, and insert intramedullary nail with neck cross pinning.

Results All patients were followed up from 4 to 7 years with an average duration of 75.3 months. The average neck–shaft angle was corrected from preoperative 75° (range 55°–100°) to postoperative 120° (range 95°–130°). The average extremity lengthening was 3.4 cm (range 2.0–4.5 cm). 19 location of osteotomy showed good union. There were no infection and recurrent fracture and progression of deformity.

Conclusion The valgus osteotomy can correct Shepherd's crook deformity, prevent recurrent fracture, and restore alignment, thus improve functioning of limb. The intramedullary nail with neck cross pinning should be the first consideration of internal fixation. Massive impaction allograft is the key technique to improve full incorporation of allograft and to prevent pathological fracture.

Keywords Shepherd's crook deformity · Intramedullary nail · Valgus osteotomy · Massive impaction allograft · Double-level osteotomy

Background and objectives

Fibrous dysplasia of bone (FD) is a disease with a wide spectrum of involvement and presentations. It can occur in one bone (monostotic) or in several bones (polyostotic) [1, 2]. Shepherd's crook deformity tends to occur during the first 4–5 years of age, particularly in the inter-trochanteric region [3]. The well-known progressive varus Shepherd's crook deformity in fibrous dysplasia is associated with limb shortening, limping, and occasionally chronic fatigue fractures with disabling walk [4]. In spite of aggressive orthopedic treatment, recurrent fractures and relentless progression of the varus deformity are still characteristic involvements in this disease [4–6].

Although various kinds of surgical treatment have been reported for this type of combined deformity, restoring the alignment and preventing re-fracture is the big challenge [7–12]. Some literatures have focused on this problem, but most of them have limited number of cases [9, 11, 12]. The purpose of this current study is based upon larger number of cases to evaluate the effectiveness of single/double-level osteotomy with intramedullary nail internal fixation to manage the Shepherd's crook deformity in fibrous dysplasia.

Materials and methods

A retrospective study was performed in 13 patients (14 femora) with histologically confirmed fibrous dysplasia who were treated in our hospital between August 2000 and February

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Table 1 Clinical data of the group

Case	Age at operation	Gender	Type of lesion	Pathological fracture	Pain	Length of follow-up (months)	Fate of lesion	Clinical course
1	21	F	M	Yes Femur neck	Yes	85	Smaller	Asymptomatic Fracture healed
2	14	F	M	No	Yes	84	Smaller	Asymptomatic
3	30	M	M	Yes #	Yes	84	Denser	Asymptomatic
4	14	M	M	No	Yes	80	Denser	Asymptomatic
5	18	M	M	Yes Femur neck	Yes	79	Smaller	Asymptomatic Fracture healed
6	39	F	M	No	Yes	76	Smaller	Asymptomatic
7	15	M	P	No	Yes	75	Denser	Asymptomatic
8	17	F	P	Yes Femur neck	Yes	75	Smaller	Asymptomatic Fracture healed
9	22	F	P	Yes Femur Shaft	Yes	73	Same	Asymptomatic Fracture healed
10	20	M	P	Yes Intertrochanteric	Yes	73	Denser	Asymptomatic Fracture healed
11	36	F	P	Yes Femur Shaft	Yes	68	Denser	Asymptomatic Fracture healed
12	15	M	P	No	Yes	59	Denser	Asymptomatic
13	34	M	P	Yes Femur Shaft	Yes	49	Denser	Asymptomatic

Case 3: in 12 years, case 3 sustained first time pathological fracture

2005. Six patients had monostotic disease and seven had polyostotic disease. There were seven males and six females. The mean age at the time of index procedure was 22.7 years (range 14–39 years). Among these cases, a total of ten times of hip or femur fracture had sustained, usually after minor trauma. Six cases had one fracture and two cases had two (case 3 and 11). Before the osteotomy with intramedullary nail fixation (our index procedure), eight patients had been performed at least one operative procedure, two had three procedures. The average neck–shaft angle and lower extremity discrepancy of 14 femoral was 75° (range 55°–100°) and 3.2 cm (range 2.0–4.5 cm), respectively. Six patients had trendelenburg gait preoperative. All cases had hip pain, four walked with bilateral crutches support, two with unilateral crutch support, and seven cannot walk due to pathological fracture. Further details on gender, primary complaint, clinical course, and fate of lesion were listed on Table 1.

Preoperative planning included the use of paper tracings and cutouts from the radiographs, as recommended by Sofield and Millar [13]. We used the full-length anteroposterior and lateral plain film with magnificent rate of 100% to make the template paper. The location, size, and angle of each wedge osteotomy and insert point of intramedullary nail were simulated on the template and were checked by intra-operative intensifier. The extent of exposure varied depending on the number of osteotomy that needed for correcting deformity.

All cases performed the index procedure. This procedure included four steps, orderly as valgus osteotomy, curettage lesion, massive impaction allograft and insert intramedullary nail with neck cross pinning. Simple varus deformity usually was performed single level valgus osteotomy at subtrochanteric region for the effort to ensure neck–shaft angle 120° (Fig. 1). Regarding the combined deformity of the femur neck and the subtrochanteric region, double-level valgus osteotomy was performed with the first level at subtrochanteric region and the second around the dome of the varus femur deformity. The second osteotomy site was determined by the host bone quality, therefore, to ensure the healing of osteotomy site. The goal of double-level osteotomy is to restore the neck–shaft angle at least to 90° and re-establish the alignment of femur. Additionally, if the bone quality of femur calcar is poor, the medial displacement valgus osteotomy should be considered. The preoperative preliminary osteotomy should ensure the intramedullary nail could through the intramedullary cavity without impinging the lateral cortex of femur.

The clinical outcome was assessed by a modified criteria described by Guille et al. [7]. Union of the osteotomy, the neck–shaft angle of the femur, limb length discrepancy, and mechanical axis deviation were compared before and after surgery.

The postoperative treatment included early isotope exercise of quadriceps and passive exercise of hip and knee.



Fig. 1 Case 11, a 36-year-old female, this patient had two pathological fractures and performed three procedures. **a** Preoperative pelvic plain film showed polyostotic disease, bilateral Shepherd's crook deformity and pathological fracture of subtrochanteric of left femoral; **b** preoperative plain film; **c** single-level medial displacement valgus osteotomy and massive impaction allograft, choose the intramedullary nail with neck cross-pin as implant. **d** Postoperative 3 months film showed slightly absorb of bone-grafts; **e** Postoperative 6 years showed good union of osteotomy site, alignment has been correct, no evidence of recurrence of lesion and re-progress of deformity

After 2 weeks, the patient began the straight elevate exercise. The partial weight-bearing support with cane is considered at 4–6 weeks postoperative. More activity exercise was determined by the radiography results, the number of osteotomy, and the rigid of internal fixation.

Results

Nine single level osteotomy were performed in seven unilateral femoral (Fig. 1a–e) and one bilateral femora, five double-level osteotomy in five unilateral femora (Fig. 2a–e). All patients were followed up from 4 to 7 years with an average duration of 75.3 months. Fourteen femoral mechanical alignments had been corrected radiologically. The average neck–shaft angle was corrected from preoperative 75° (range 55°–100°) to postoperative 120° (range 95°–130°). The average extremity lengthening was 3.4 cm (range 2.0–4.5 cm). 19 osteotomy location showed good union. There were no infection and recurrent fracture and progression of deformity. Regarding the implant failure, there were no evidence of implant loosen and broken at the last follow-up. Mechanical axis deviation was corrected from mean 33 (5–43) mm medialization before surgery to mean 9 (5–14) mm medialization after surgery. There were no neurovascular complications, infection, or loss of joint or muscle function due to limb lengthening.

All bone-grafts were absorbed slightly beginning at 3 months and 80 percent of bone-grafts markedly absorbed at 10–14 months postoperatively. At the last follow-up, 11 cases were painless, 2 cases had significance elimination of pain and all cases were able to walk, even though 2 cases walked with the aid of unilateral crutch. Only two patients had slightly trendelenburg gait postoperative. The clinical scores, according to the modified criteria of Guille et al. [7], were improved from an average of 3.5 (range 2–6) points preoperative to an average of 8.7 (range 7–10) points at the last follow-up. Five patients achieved excellent result, seven good, one fair, and no patient had a poor result. The details of clinical outcome were shown as Table 2.

The average volume of perioperative bleeding was 875 mm (range 600–3,000 mm), significant high in the patients underwent the multi procedures that involved extensive exposure and double-level osteotomy.

Discussion

Various kind of surgical treatment have been reported, but treatment for Shepherd's crook deformity are still being big challenge orthopedic surgeon faced [7, 9, 11, 12, 14–18]. Because of limited cases, there are no agreement in the

Fig. 2 Case 13, a 34-year-old male, this patient had femoral shaft pathological fracture. **a** Preoperative pelvic plain film showed unilateral Shepherd's crook deformity and pathological fracture of left femoral shaft; **b** preoperative full-length plain film of bilateral thigh; **c** postoperative film, double-level medial displacement valgus osteotomy and massive impaction allograft, choose the intramedullary nail with neck cross-pin as implant Preoperative film of left femoral; **d** postoperative 5 years showed good union of osteotomy site, alignment has been correct, no evidence of recurrence of lesion and re-progress of deformity; **e** postoperative 4 years showed good function of left hip joint

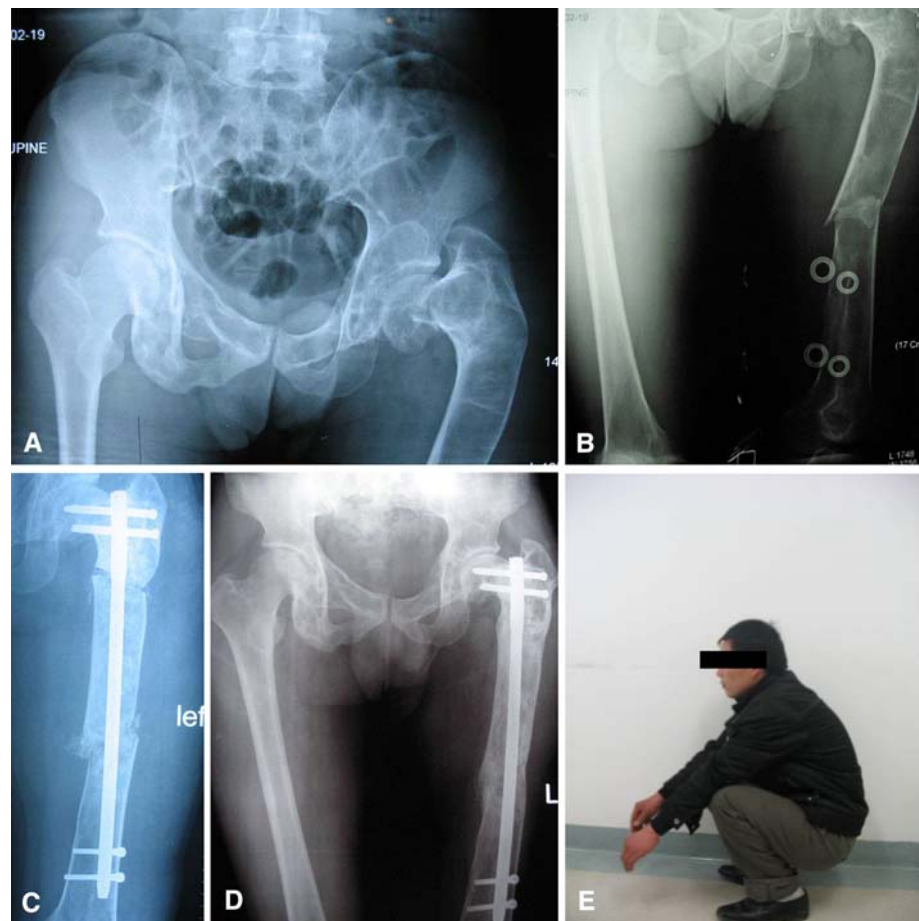


Table 2 The outcome comparisons of pre- and postoperative clinical results

Categories	Unsatisfactory		Average		Satisfactory	
	Pre-op	Post-op	Pre-op	Post-op	Pre-op	Post-op
Pain	13	0	0	2	0	11
Hip motion	12	0	1	1	0	11
Limping	13	0	0	1	0	13
Activities of daily living	10	0	2	2	0	11
Social activities	12	0	1	3	0	10

The number of femurs that belong to different categories, in a total of 13 patients with 14 femora. Clinical results, i.e., unsatisfactory, average, and satisfactory were scored as 0, 1, and 2. For a potential maximum total of 10 points, >9 points was defined as excellent, 7 or 8 points as good, 5 or 6 points as fair, and <5 points as poor

standard procedure for this combined deformity of the femoral neck and subtrochanteric region [9, 19, 20]. Early surgical management has included attempts to eliminate disease bone, internal fixation for the mechanical support of fractures or corrective osteotomy. Simple curettage and bone grafting had been proven ineffective in deformity of fibrous dysplasia [7]. Excision of en bloc intertrochanteric region with re-attachment of muscle insertion also had limited success.

Reviewed the literatures of surgical treatment of Shepherd's crook deformity, there were several aspects of this problem should to be addressed. The first question involves

osteotomy. Early in 1961, Depalma and Dodd [21] had concluded that medial displacement osteotomy was the choice procedure for patient who had Shepherd's crook deformity and that valgus osteotomy should be considered in cases who had coax vara. These authors also pointed out that medial displacement osteotomy could achieve more functional and serviceable weight-bearing relationship, alleviated stress and thus helpful to prevent pathological fracture, improved stability and gait, and decreased pain. In the study of Harris et al. [22], subtrochanteric osteotomy also can achieve good result in the patients with Shepherd's

crook deformity. In our series, a total of 19 osteotomy were performed in 14 femora, and clinical scores were significantly improved. This result is persisted with the opinion of DePalma and Harris. Although being middle–short term follow-up, the result also demonstrated that valgus osteotomy or medial displacement valgus osteotomy can restore neck–shaft angle and re-establish mechanical alignment to improve the function and can prevent recurrent fracture or pathological fracture.

The second problem relates to the determination of internal fixation implant. Guille et al. [7] have demonstrated that osteotomy with internal fixation led to good result. To prevent deformities and recurrent fracture, a number of internal fixation methods had been devised. Fixation with plate and screw has been applied most widely, however, the internal fixation with plate frequently allows recurrence of the deformity or re-fracture below the plate, and in many cases, it is difficult to get sufficient purchase on the soft host bone with screws [3] and get plates to fit the contour of a deformed femur [9, 11]. Long-term follow-up have shown unsuccessful results when using a simple intramedullary nail or a flexible nail with recurrence of the deformity [6, 9, 11, 23]. In addition, Connolly [9] and Freeman [11] also have demonstrated that the shortcoming of Zickel nail was that it only had one screw in the proximal and distal end to control the rotation of femoral head and distal of the femur shaft after surgery.

In 1959, Scofield and Millar [13] first used the device consisting intramedullary rod and a cross pin directed into the femoral head and neck. Unfortunately, they did not supply the follow-up result. In the classic report in 1962, Harris [22] pointed out that the feasibility and importance of stabilization of femoral neck by a fixation device that transverse the involved neck and has a rigid purchase in the femoral head. Recently, in the multi-center study of Ippolito et al. [14], intramedullary nail had led to a good result in prevent recurrent fracture after surgery. In 2006, Jung et al. [12] also demonstrated that using multiple osteotomies and two screws crossing the femoral neck could lead to good result. In the current study, we used the intramedullary nail with neck cross pinning also can improve the clinical outcome and effectively prevent recurrent fracture and progression of deformity. Although long-term follow-up and multi-center study need deserved, this may be suggested that the intramedullary nail with neck cross pinning to control the rotation of femur head had important influence on the result and deformity.

Last, but the most important issue is regarding the neurological and vascular problems caused by the limb lengthening and the bone-graft. Kempf et al. [24] had suggested that the maximum lengthening should not exceed 4.0 cm in the femur, theoretically. But, in the study of Murray et al. [25], 117 one-stage corrections of leg shortening all provided

correction of deformity with no neurovascular complications, infection, or loss of joint or muscle function. Up to 7 cm of length have been achieved when the shortening was mainly due to angulatory deformity. Hantes et al. [26] had demonstrated that the most serious complications occurred in patients with >30% bone lengthening. Patients with <15% lengthening had a significantly decreased complication rate. In the study of Jung et al. [12], limb-length discrepancy was corrected from mean 27 (8–41) mm before surgery to a mean of 6 (2–11) mm after surgery. But they did not reported whether there occurred neurological and vascular problems due to limb lengthening. In this current study, the average extremity lengthening was 3.4 cm (range 2.0–4.5 cm), only one patient's lengthening exceeded 4 cm. Lower limb lengthening of all cases did not exceed beyond the 15% of initial length. There were no neurovascular complications, infection, or loss of joint or muscle function due to limb lengthening. This result is in consistence with Murray [25]. According our experience, the Shepherd's crook deformity is a kind of combined angulatory deformity. The maximum lengthening can exceed 4 cm through complete release of the surrounding soft tissue.

Because allograft has the least and the slowest internal replacement by host bone, more of the grafts persist for longer. This makes fibrous dysplasia to be one of the few diseases for which allograft are biologically preferable to autogenous grafts [19]. According to our experience, the massive impaction allograft is the key to improve the incorporation between allograft and host bone and to prevent recurrence of lesion and varus deformity. Although massive impaction allograft has been widely accepted in revision joint surgery [27], this question should be paid more attention in treatment of fibrous dysplasia.

Limitation of the current study

It should be noted that this study has examined only middle–short term result of surgical treatment of Shepherd's crook deformity, not a long term follow-up. In addition, the number of cases are still limited, the results should not be taken as an evidence of long-term or criteria procedure. Notwithstanding its limitation, this study does suggest the importance of valgus osteotomy, intramedullary nail with neck cross pinning and massive impaction allograft in the management of this deformity.

Conclusion

This current study is, to our knowledge, the largest number report focus on the surgical treatment of the Shepherd's crook deformity of the fibrous dysplasia. The valgus osteotomy or medial displacement valgus osteotomy can via

restore neck–shaft angle and re-establish mechanical alignment to improve the function and prevent re-fracture. The intramedullary nail with neck cross pinning should be the first regard of internal fixation implant. Massive impaction allograft is the key technique to improve allograft incorporating fully and to prevent recurrence of deformity and pathological fracture.

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