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Original Article

Multiple valgus osteotomies combined with intramedullary nail for shepherd's crook deformity in polyostotic fibrous dysplasia: a case series and review of the literature

Xue-Lei Zhang*, Chun-Yu Chen*, Hong Duan, Chong-Qi Tu

*Department of Orthopedics, West China Hospital, Sichuan University, China. *Equal contributors.*

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Abstract: The aim of this study was to investigate the long-term clinical outcomes of valgus osteotomy combined with intramedullary nail for the treatment of shepherd's crook deformity of polyostotic fibrous dysplasia. We reviewed the clinical data of 22 patients with polyostotic fibrous dysplasia that was treated at our department between January 1995 and January 2010. All 22 of the patients (23 femurs) had shepherd's crook deformity. We performed a sequence of 3 procedures to treat shepherd's crook deformity: valgus osteotomy, massive impaction allograft with deep-frozen allogeneic cortical bone, and insertion of an intramedullary nail with neck cross pins. The mean follow-up time was 97 (range 60-155) months. The neck-shaft angle was corrected from a mean value of 75° (range 45°-100°) preoperatively to a mean value of 125° (range 95°-140°) postoperatively. The mean extremity lengthening was 28 mm (range 18-45 mm). Almost all of the osteotomy sites healed within 6 months postoperatively. The alignments of all of the 23 femurs were restored to normal. There was no recurrent fracture or deformity progression after a mean follow-up of 8 years. Valgus osteotomy can correct shepherd's crook deformity, restore the alignment of the femur, and restore the neck-shaft angle. The impaction allograft can provide enough initial stability for the intramedullary nail to help the union of the osteotomy site. The intramedullary nail with neck cross pins can maintain the corrected alignment of the femur and prevent both recurrent pathological fracture and progression of the deformity.

Keywords: Shepherd's crook deformity, surgical treatment, valgus osteotomy, massive impaction allograft, intramedullary nail

Introduction

Fibrous dysplasia is an uncommon skeletal disorder in which normal bone and bone marrow are replaced by fibro-osseous tissue. The etiology of fibrous dysplasia has been linked to an activating mutation in the gene that encodes the α subunit of stimulatory G protein (Gsα), located at 20q13.2-13.3 [1]. Fibrous dysplasia may involve 1 bone (monostotic fibrous dysplasia) or multiple bones (polyostotic fibrous dysplasia) [2]. The proximal femur is the region in which fibrous dysplasia is most prone to occur [3]. The lesion reduces the strength of the host bone. Subsequently, the weight of the body and the strong gluteal muscles act on the weakened proximal femur, resulting in pain, limping, deformity, and pathological fracture. The continuous mechanical stress and repeated fractures result in progressing varus and bowing, leading to the classical shepherd's crook deformity [4, 5]. This deformity usually occurs and progresses in adolescents [6, 7]. If this deformity is not treated early enough, secondary valgus deformity in the distal femur may occur, complicating the deformity correction [8]. Despite improvements in surgical techniques and orthopedic instruments, the constant reduction to the bone strength of the proximal femur makes it very difficult to both correct this deformity and maintain the correction. To date, the treatment of this disease has only been the focus of a relatively small number of studies with limited numbers of cases [9-17]. Further, genu valgum secondary to the correction of shepherd's crook deformity has never been dis-

Table 1. The patients' clinical characteristics

Patient no.	Age/ gender	Side	Pain	Pathological fracture (region)	Pre-op NSA (deg)	No. of Osteotomies#	Internal fixation	EBL* (ml)	Surg time (min)	Follow-up (month)	Post-op NSA** (deg)
1	14/M	L	Yes	Yes (femoral shaft)	75	2	PFRN	1030	155	82	135
2	15/M	R	Yes	No	80	2	PFRN	830	140	155	140
3	15/M	L	Yes	No	85	1	PFNA	400	105	99	135
4	16/F	R	Yes	No	70	1	PFRN	490	120	76	120
5	16/M	L	Yes	Yes (femoral shaft)	90	1	PFRN	320	105	60	120
6	16M	L	Yes	No	50	1	PFRN	315	110	74	130
7	17/F	R	Yes	Yes (femoral neck)	80	1	PFRN	405	95	102	130
8	17/M	R	Yes	Yes (trochanteric)	70	1	PFRN	290	90	84	125
9	20/M	R	Yes	Yes (femoral shaft)	70	1	PFNA	385	115	65	130
10	22/F	R	Yes	Yes (femoral neck)	85	1	PFRN	280	95	68	135
11	22/M	L	Yes	Yes (femoral neck)	60	2	PFRN	1280	135	95	120
12	23/M	L	Yes	No	60	2	PFRN	775	110	86	110
13	24/M	R	Yes	Yes (femoral shaft)	70	2	PFRN	1415+ 375	165+ 90	90	125
14	27/M	L	Yes	No	75	2+1	PFRN	975+ 360	135+ 85	132	95
15	28/M	L	Yes	Yes (femoral neck)	70	2	PFRN	710	120	126	130
16	32/F	L	Yes	No	70	2+1	PFNA	1750+ 455	120 85+	97	130
17	34/M	R	Yes	Yes (femoral shaft)	75	1	PFRN	290	90	109	125
18	34/M	L	Yes	Yes (femoral shaft)	70	2	PFRN	1350	120	110	130
19	36/F	L	Yes	Yes (femoral shaft)	70	1	PFRN	375	105	128	120
20	36/M	L	Yes	No	100	1	PFRN	415	90	69	135
21	36/F	R	Yes	No	60	1+1	PFNA	1165+ 315	115+ 75	147	120
		L	Yes	Yes (femoral shaft)	90	1	PFRN	1225	90		100
22	39/F	R	Yes	No	90	2+1	PFNA	1245+ 415	125+ 95	83	130

EBL* estimated blood loss, NSA** neck-shaft angle, PFRN proximal femoral restruction nail, PFNA proximal femoral nail antirotation. No. of Osteotomies# proximal femur +distal femur.

cussed. Accordingly, we decided to perform the present study of the long-term clinical outcomes of valgus osteotomy combined with intramedullary nail for the correction of shepherd's crook deformity. Our analysis includes the largest cohort of patients with shepherd's crook deformity that has been studied to date. Additionally, we demonstrated the surgical treatment of genu valgum secondary to the correction of shepherd's crook deformity in the proximal femur. Finally, we included a review of the literature on the surgical treatment of shepherd's crook deformity.

Materials and methods

Patient characteristics

We reviewed the clinical data of 22 patients with histologically confirmed fibrous dysplasia who were treated at our hospital between January 1995 and February 2010. Shepherd's crook deformities were seen in 23 femurs from the 22 patients, of whom 15 were men and 7 were women. The mean age at the time of surgery was 25 (range 14-39) years. Twenty of the patients experienced 1 to 5 fracture events in the proximal femur, usually after minor trauma. Thirteen patients had fresh fractures and the other 7 patients had fractures that had healed by the time that they presented at our department. The preoperative mean neck-shaft angle was 75° (range 45°-100°). The preoperative lower extremity discrepancy was 3.2 cm (range 2.0-5.0 cm). All of the patients had hip pain and abnormal gait. Four of the patients walked with a Trendelenburg gait, 2 patients walked with unilateral crutch support, 3 patients walked with support from bilateral crutches, and 13

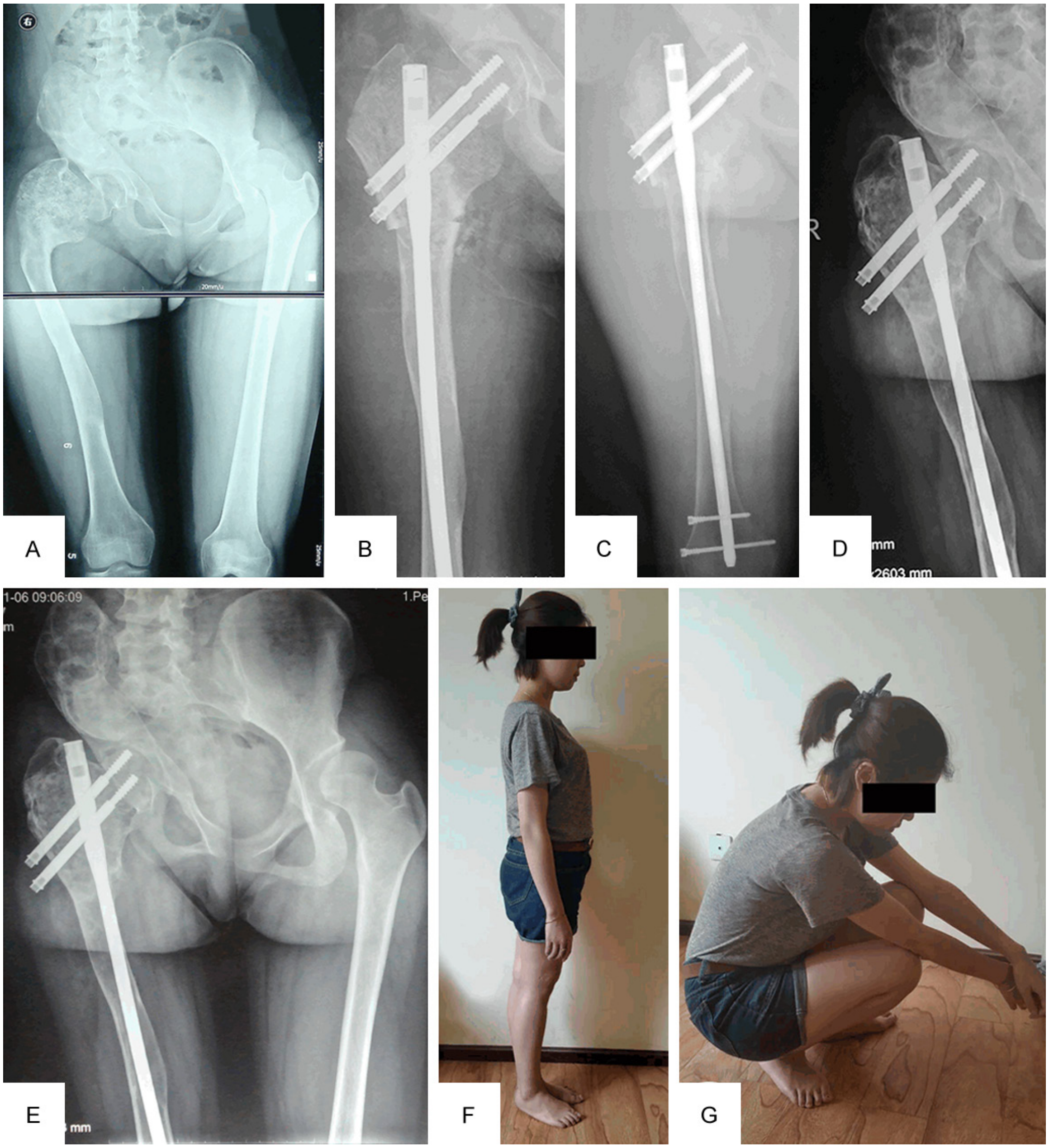

Figure 1. Case 4, a 16 years-old woman. Shepherd's crook deformity was present in the right proximal femoral. A: The neck-shaft angle was 70° and there was grade 2 hip osteoarthritis. B: The neck-shaft angle was corrected to 120°. C: The osteotomy site healed 4 months after surgery. D: Five years later, there was collapse of the femoral head, shortening of the femoral neck, and loosening of the neck cross pins. Further, there was grade 3 hip osteoarthritis. E: Twelve years postoperatively, there was no pathological fracture or deformity progression. The neck-shaft angle was 120°. F, G: Twelve years postoperatively, there was good function of the right hip joint.

patients could not walk due to pathological fracture. According to the Kellgren and Lawrence (KL) grading system, 6 patients had hip osteoarthritis (grade 2, n = 2; grade 3, n = 2). Further details on ages, internal fixation devices, surgery times, and other characteristics are listed in **Table 1**. This study was conducted in accordance with the Declaration of Helsinki and with approval from the Ethics Committee of West China Hospital (Chengdu, China). Written informed consent was obtained from all participants.

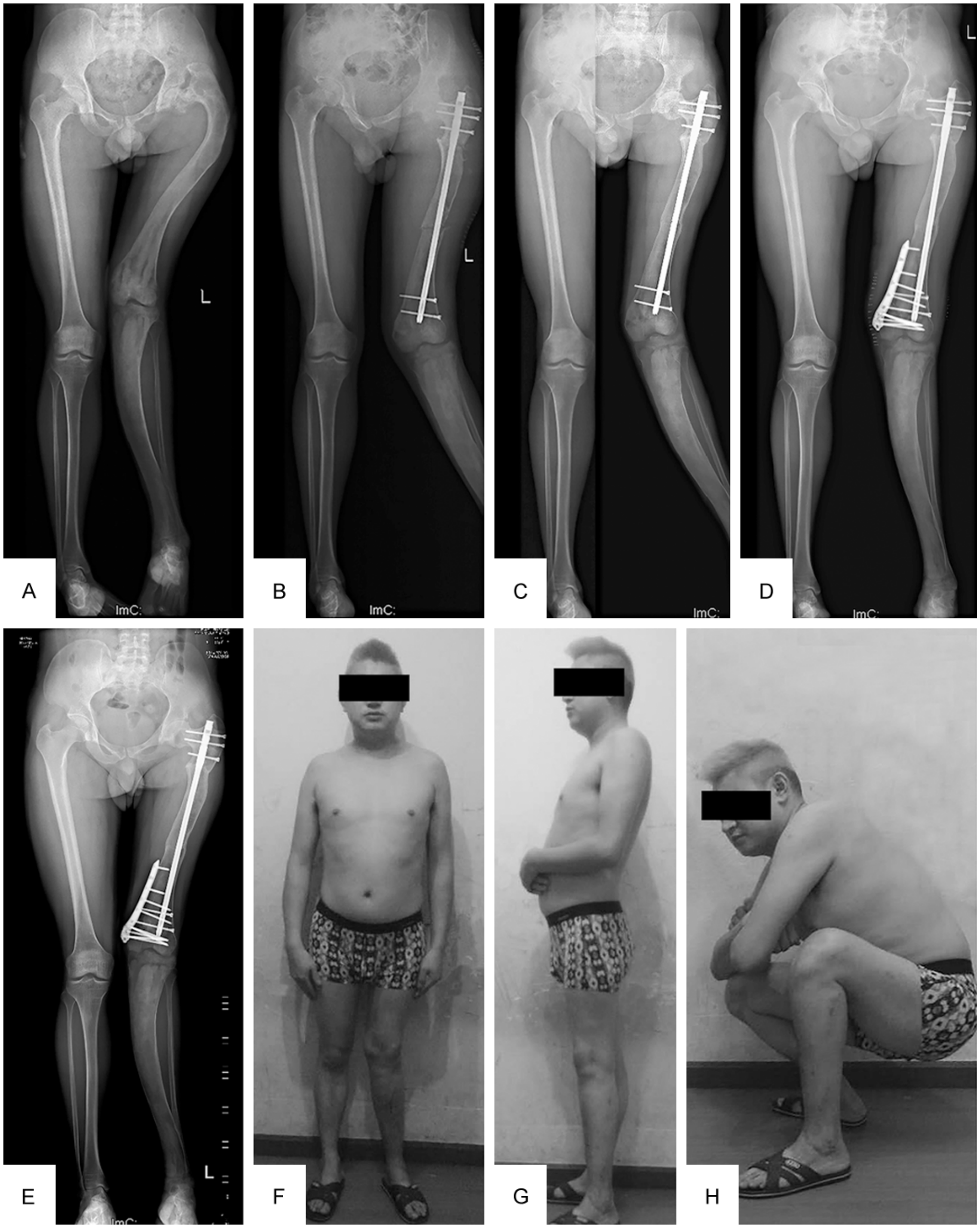

Figure 2. Case 14, a 27-year-old man. In this patient, the lower limb was involved. A: The neck-shaft angle was 75°. B: The shepherd's crook deformity was corrected with double-level osteotomy, and the neck-shaft angle was restored to 95°. C: The osteotomy site healed 6 months after surgery, but there was a secondary genu valgum and the femoral tibial angle was 30°. D: The alignment of the femur was corrected to normal and the femoral tibial angle was 10° after supracondylar femoral varus osteotomy. E: Eleven years postoperatively, there was no fracture or deformity progression. F-H: Eleven years postoperatively, there was good function of the left hip joint.

Surgical treatment

Preoperative preparation: Preoperative planning included the use of paper tracings and cutouts from radiographs, as recommended by Sofield and Millar [18]. The number, location, and angle of each valgus osteotomy and the insertion of the intramedullary nail were all sim-

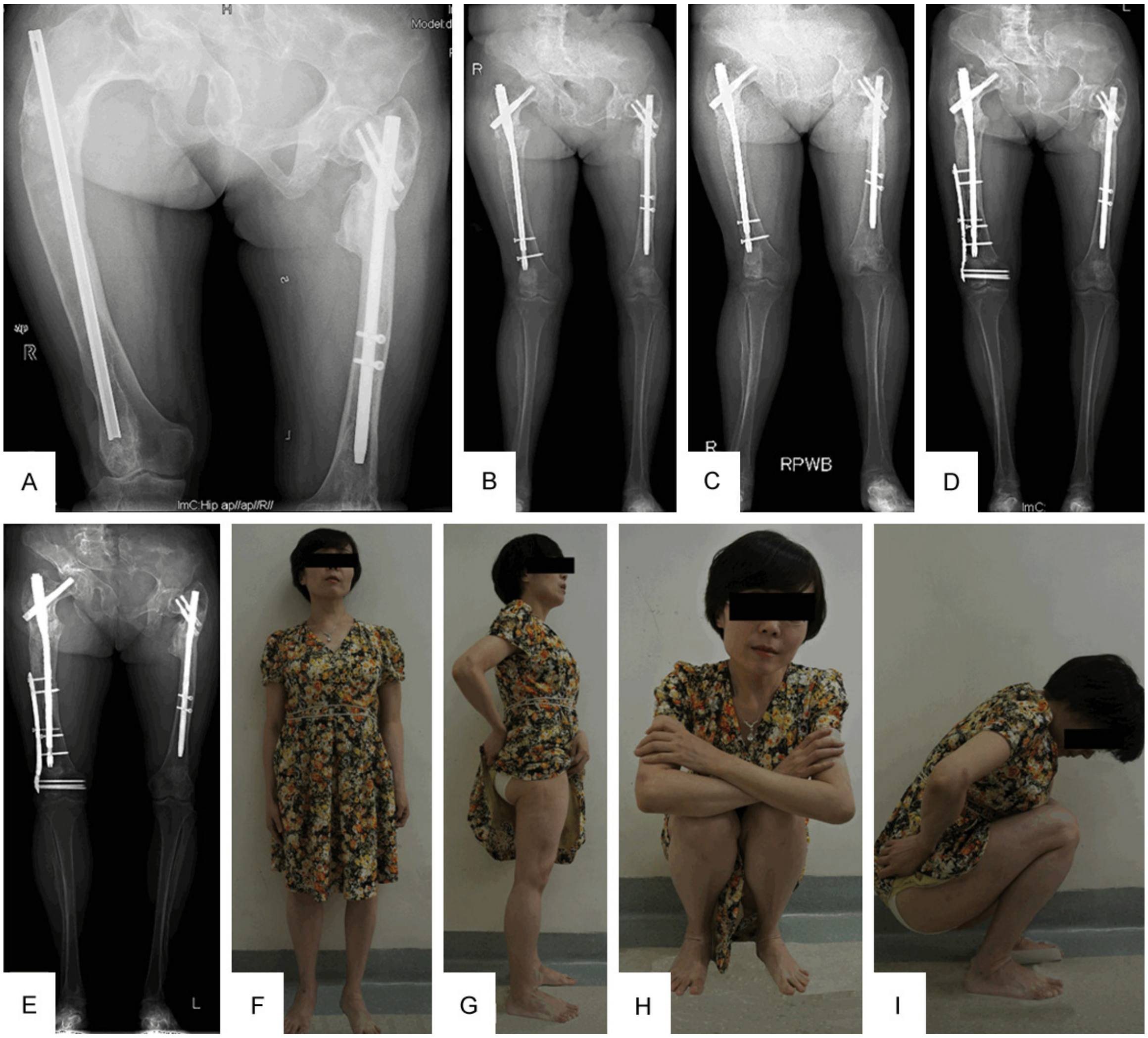

Figure 3. Case 22, a 36-year-old woman. In this patient, the bilateral femurs were involved. We treated the right side at this time; the left side had been treated 4 years previously. A: The neck-shaft angle was 60° and there was grade 2 hip osteoarthritis. B: The neck-shaft angle was corrected to 120° and the femoral tibial angle was 20°. C: The osteotomy site healed 6 months after surgery. D: The alignment of the lower limb was corrected to normal with supracondylar femoral varus osteotomy. The femoral tibial angle was 5°. E: Ten years postoperatively, there was no fracture or deformity progression. F-I: Ten years postoperatively, there was good function of the right hip joint.

ulated on the paper template preoperatively to ensure the passage of the intramedullary nail, the correction of the neck-shaft angle, and the alignment of the femur. The extent of exposure depended on the number of valgus osteotomies that were needed to correct the deformity.

Surgical technique: All patients were treated in 3 contiguous steps: valgus osteotomy, massive impaction allograft, and insertion of an intramedullary nail with neck cross pins. The primary goals of valgus osteotomy were to restore the neck-shaft angle and the alignment of the femur. Simple varus deformity was treated with single-level subtrochanteric valgus osteotomy with the objective of providing a neck-shaft angle of 120° (**Figure 1**). When there was a complex deformity that consisted of both coxa vara and varus deformities in the proximal femoral shaft, it was difficult to restore the neck-shaft angle and maintain the correction with single-level valgus osteotomy. We therefore tried to restore the neck-shaft angle and the alignment of the femur with double-level valgus osteotomy. The first valgus osteotomy was made at the intertrochanteric or the subtrochanteric region to restore the neck-shaft

Figure 4. Case 5, a 16-year-old man. Shepherd's crook deformity was present in the right proximal femoral shaft. A: The neck-shaft angle was 90°. B: The neck-shaft angle was corrected to 120°. C: The osteotomy site had not healed 9 months after surgery. D: The osteotomy site had not healed 16 months after surgery. E: The 2 neck cross pins were removed 16 months after surgery. F: The osteotomy site healed 12 months later. G: The alignment of the lower limb was normal on a full-length standing radiograph. H-J: Five years postoperatively, there was good function of right hip joint. Grade 2 hip osteoarthritis was present.

angle. The second osteotomy was made around the vertex of the varus deformity in the proximal femoral shaft. Further, the second osteotomy site should have good bone quality to ensure the stability of the correction. In patients with severe deformity and poor bone quality in the femur that was treated with double-level valgus osteotomy, it was sometimes difficult to restore the neck-shaft angle to a normal value and to maintain this normal value. Therefore, the goals of double-level osteotomy were to restore the neck-shaft angle to at least 90° and to re-establish the femur's alignment (**Figure 2**). Additionally, medial displacement valgus osteotomy should be performed to stabilize the osteotomy when the femoral calcar has been destructed. Further, juxta-articular valgus deformity might be present around the knee in patients with severe deformity in the proximal femur. In such circumstances, there might be a secondary genu valgum deformity after the correction of shepherd's crook deformity in the proximal femur. When the femoral tibia angle was more than 10°, an additional supracondylar femoral varus osteotomy was performed to correct the secondary genu valgum after the union of the osteotomy site in the proximal femur (**Figure 2**).

Postoperative management: The patients were asked to undergo early isotonic exercise of the quadriceps and passive exercise of the hip and

Table 2. Outcome comparisons for pre- and post-operative clinical results

Categories	Unsatisfactory		Average		Satisfactory	
	Pre-op	Post-op	Pre-op	Post-op	Pre-op	Post-op
Pain	17	1	5	3	0	18
Hip motion	10	1	4	2	8	19
Limping	17	2	3	4	2	16
Activities of daily living	13	1	4	3	5	18
Social activities	16	2	3	3	3	17

The number of femurs that belong to different types, in a total of 22 patients with 23 femurs. 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.

knee under the guidance of rehabilitation physicians, as soon as the plasma drainage tube had been removed. Two weeks later, the patients were asked to begin straight leg raise exercises. Four to 6 weeks later, the patients began partial weight-bearing exercise with the support of crutches or a walking frame. Further exercise was based on follow-up radiographs and the number of osteotomies.

Outcome evaluation

We used the follow-up radiographs to evaluate the union of the fracture and osteotomy, the change to the neck-shaft angle, the deviation of the alignment of the femur, and the development of hip osteoarthritis according to the KL grading system [19]. We used the modified criteria of Guille et al. to evaluate the clinical outcomes, including hip pain, limping, the motion of the hip joint, daily activity, and social activity [20].

Results

Thirty-three valgus osteotomies were conducted in the proximal part of the 23 femurs to correct shepherd's crook deformity: 13 femurs were treated with single-level valgus osteotomy and 10 femurs were treated with double-level valgus osteotomy. Nineteen femurs were fixed with proximal femoral reconstruction nail and 4 femurs were fixed with proximal femoral nail antirotation. The mean follow-up time was 8 (range 5-15) years. Thirty-two osteotomy sites healed within the 6 months following surgery. All 13 of the pathological fractures healed 3 to 6 months postoperatively. After the osteotomy site in the proximal femur had healed, we performed supracondylar femoral varus osteotomy in 8 femurs to correct secondary genu valgum (**Figure 3**). The alignments of the 23 femurs were restored to normal. The neck-shaft angle was corrected from a preoperative mean of 75° (range 45°-100°) to a postoperative mean of 125° (range 95°-140°). The mean lower limb lengthening was 31 mm (range 20-46 mm). The mechanical axis deviation was corrected from a mean 30 mm (range 5-45 mm) medialization preoperatively to a mean 8 mm (range 4-14 mm) medialization postoperatively. No infection, thromboembolism, bedsore, failure of internal fixation, pathological fracture, or recurrence of deformity occurred. No neurovascular complication, or loss of joint or muscle function due to limb lengthening was found during the follow-up. Nonunion was found in 1 osteotomy site 16 months after the operation. This osteotomy site healed 12 months after removing the 2 distal locking screws (**Figure 4**). Loosening of the neck cross pins combined with shortening of the femoral neck was found in 1 femur 5 years after surgery (**Figure 1**). The neck cross pins were not removed because the patient did not feel any discomfort. Of the 6 hips in which hip osteoarthritis had been diagnosed before surgery, 2 grade 2 hips progressed to grade 3 (**Figure 1**) and no hip progressed to grade 4. Two other hips were found to have grade 2 hip osteoarthritis after surgery (**Figure 2**).

At the last follow-up, 18 patients had no pain and 4 experienced pain that had been significantly alleviated. All patients could walk by themselves, although it was judged that 3 of the patients should walk with a unilateral cane and 6 of the patients had a limp gait. According to the modified criteria of Guille et al., the clinical scores had improved from a mean of 2.5 (range 2-6) points before surgery to a mean of 8.7 (range 6-10) points at the last follow-up [20]. Eleven patients achieved excellent results, 8 achieved good results, 3 achieved fair results, and no patient had a poor result. The details of the clinical outcomes are shown as **Table 2**. The mean estimated blood loss was 850 mm (600-3300 mm), which increased significantly

Table 3. A review of the literature on the surgical treatment of shepherd's crook deformity

Author (reference)	Pub-date	No. of Patients (femurs)	Age (yrs)	Sex (No.)	No.of osteotomies	Follow-Up (yrs)	Internal fixation	Pre-op NSA* (deg)	Post-op NSA (deg)
DePalma 13	1961	2 (2)	21, 50	F (2)	1 (2)	-	Plates	-	-
Connolly 9	1977	1 (2)	6	M (1)	L (1), R (2)	1	Zickel nail	-	-
Freeman 10	1987	1 (2)	6	F (1)	L (3), R (2)	4.3	Zickel nail	-	-
Dunstan 14	2005	1 (1)	12	M (1)	Prosthetic replacement	50	Customised polyethylene prosthesis	60	125
Chen 15	2005	2 (2)	30	F (2)	1	11	DHS	45	130
			31		1	2.1	DHS	115	115
Jung 11	2006	5 (7)	24 (13-47)	M (3)	multiple (7)	2.5 (2.1-4)	PFRN (7)	92	129
				F (2)					
Watanabe 17	2007	1 (1)	9	F (1)	2	3	TSF	100	141
Liu 12	2009	13 (14)	23 (14-39)	M (7)	1 (9), 2 (5)	6.3 (4-7)	PFRN (14)	75 (55-100)	120 (95-130)
				F (6)					
Ippolito 16	2015	10 (11)	14.3 (6-36)	M (5)	1 (2), 2 (9)	4.7 (1.3-8.2)	PFN (8)	83 (50-100)	130 (120-140)
				F (6)			PHN (3)		
Zhang (now)	2015	22 (23)	25 (14-39)	M (15)	1 (13), 2 (10)	8.1 (5-12.9)	PFRN (18)	75 (50-100)	125 (95-140)
				F (7)			PFNA (5)		

NSA* neck-shaft angle, DHS dynamic hip screw, PFRN proximal femoral restruction nailing, TSF three-ring Taylor Spatial Frame, PFN proximal femoral nail, PHN proximal humeral nail, PFNA proximal femoral nail antirotation.

in patients who had undergone extensive exposure and double-level osteotomy.

In addition to analyzing the 22 patients who had been treated at our department, we performed a review of the literature on the surgical treatment of shepherd's crook deformity. The studies included in our review are summarized in **Table 3**.

Discussion

Although some researchers have proposed various surgical methods for treating shepherd's crook deformity, there is still no widely accepted treatment for this deformity [9-17]. The present study was based on the largest cohort of patients with shepherd's crook deformity. We reviewed the treatments of 22 patients (23 femurs) with shepherd's crook deformity and followed up their cases for a mean of 8 years. Additionally, this is the first report of the surgical treatment of secondary genu valgum. After performing a comprehensive review of the literature, we identified 3 issues with the treatment of shepherd's crook deformity that should be discussed.

The first issue is osteotomy. In 1961, Dipalma and Dodd concluded that valgus osteotomy should be performed to correct coxa vara and that medial displacement osteotomy should be considered in cases with poor femoral calcar [13]. In 1987, Freeman restored the alignments of 6 femurs of shepherd's crook deformity with multiple valgus osteotomies [10]. In the present study, we performed 33 valgus osteotomies in 23 femurs. All of the femurs' alignments and most of the neck-shaft angles were restored to their normal states. There was no recurrence of fracture or progression of deformity. This result is consistent with the findings of Dipalma and Freeman's study. We therefore concluded that valgus osteotomy or medial displacement valgus osteotomy could effectively restore the neck-shaft angle and the alignment of the femur. Secondary valgus deformity might be present in the juxta-articular area around the knee [8]. The type of the valgus deformity in the juxta-articular area might change to secondary genu valgum after correction of the shepherd's crook deformity (**Figures 2B**, **3B**). After the correction of the deformity in the proximal femur, if the femoral tibial angle was more than 10°, an additional supracondylar femoral varus osteotomy was conducted. We treated secondary genu valgum successfully with supracondylar femoral varus osteotomy in 8 femurs. This procedure has not been reported by other researchers.

The second issue is the choice of internal fixation devices. After osteotomy, an appropriate fixation device should be used to stabilize the osteotomy site and prevent progression of the deformity and recurrence of fracture. Multiple internal fixation devices have been used to fix the osteotomy site [9, 10, 15, 18, 21, 22]. The plate and screw system has been widely used to treat deformity or fracture in the proximal femur, but is not suitable for treating fibrous dysplasia for the following reasons: First, it is difficult to provide sufficient stability with the plate and screws in the weakened bones [5, 6]. Second, it is difficult for the plate to fit the contour of a deformed femur [9, 23]. Third, fractures might occur easily because of the stress shielding effect of the distal part of the plate [10]. Fourth, the deformity might progress after removing the plate and screws [20]. Accordingly, increasingly, many orthopedists have chosen various intramedullary devices in recent years [6, 8, 10, 12, 16]. Long-term follow-up has shown that pathological fracture or recurrence of the deformity may occur when treating this disease with a flexible intramedullary nail or a simple intramedullary nail without neck cross pins [9, 10]. Connolly, Freeman, and Jung reported good results for the treatment of shepherd's crook deformity with multiple osteotomies and an intramedullary nail with neck cross pins [9, 10, 12]. The advantage of this device is that it stabilizes the area of the lesion along the entire length of the femur, in addition to stabilizing the femoral neck by fixation of the femoral head. However, a total of 12 patients were treated in these 3 studies. We treated 23 femurs (22 patients) with shepherd's crook deformity using 2 kinds of intramedullary nail with neck cross pins: 19 femurs were treated with proximal femoral reconstruction nail and 4 femurs were treated with proximal femoral nail antirotation. The mean neck-shaft angle was corrected from 75° preoperatively to 125° postoperatively. There was no deformity progression or recurrent fracture after a mean follow-up of 8 years.

The third issue is bone grafting. Bone grafting has been proved to be effective for treating monostotic fibrous dysplasia [4, 5, 23]. However, in polyostotic fibrous dysplasia, most of the grafted bone would be replaced by the dysplastic bone [7, 23]. Most femurs with shepherd's crook deformity also have poor bone quality and an enlarged intramedullary cavity. Without bone grafting, the intramedullary nail might move in the intramedullary cavity, especially when the patient participates in early weight-bearing activity. Therefore, massive impaction was very important for providing early stability for the intramedullary nail, to facilitate the union of the osteotomy. Some studies have demonstrated that dysplastic bone has the same healing ability as normal bone, with a healing time that is usually between 3 and 6 months [3, 6, 9]. In 2005, DiCaprio et al. stated that, in the treatment of fibrous dysplasia, cortical allogeneic grafts have the least and slowest internal replacement by host bone (as compared with various kinds of grafts) [23]. In 2009, Liu Yang et al. demonstrated that massive compaction allograft is the key to improving incorporation between the allograft and host bone, as well as for preventing recurrence of the lesion and varus deformity [12]. The results of our study and that of Liu Yang et al. are consistent. In our study, we used massive impaction allograft with deep-frozen allogeneic cortical bone to provide the initial stability for the intramedullary nail. All the osteotomy sites healed. There was no loosening or fracture of the intramedullary nail. We therefore concluded that massive impaction allograft with deep-frozen allogeneic cortical bone is an important step the treatment of shepherd's crook deformity.

Conclusion

To the best of our knowledge, the present study is the largest number report focus on the treatment of shepherd's crook deformity. Valgus osteotomy was able to both restore the neck-shaft angle and re-establish mechanical alignment. Massive impaction allograft with deep-frozen allogeneic cortical bone could provide sufficient initial stability for the intramedullary nail to facilitate the union of the osteotomy site or the pathological fractures. The intramedullary nail with neck cross pins was an ideal choice in the fixation of the osteotomy site, especially for patients whose epiphyseal plate had closed.

Disclosure of conflict of interest

None.

Address correspondence to: Chong-Qi Tu, Department of Orthopedics, West China Hospital, Sichuan University, China. E-mail: tuchongqi387@126.com

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