CLINICAL ARTICLE

The West China Hospital radiographic classification for fibrous dysplasia in femur and adjacent bones: A retrospective analysis of 205 patients

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Objective: This study aims to investigate the reliability and clinical outcome of a newly developed classification system for patients with fibrous dysplasia (FD) of the femur and adjacent bones, optimizing its evaluation and management.

Methods: A total of 205 patients (121 female and 84 male) with FD in the femur and adjacent bones were included in this retrospective study. All affected femurs were measured and treated based on this classification at our institution between 2009 and 2019. Based on previous studies and extensive clinical follow-up, we cautiously proposed the West China Hospital radiographic classification for FD in femur and adjacent bones following corresponding treatment options. There are five types with five radiographic features, including proximal femur bone loss, coxa vara, femoral shaft deformity, genu valgum, and hip arthritis. The intraobserver and interobserver reproducibility of this classification was assessed by four observers using the Cohen kappa statistic. The clinical outcome was evaluated using the criteria of Guille.

Results: At a median follow-up of 60 months (range 6–120), 205 patients (median 34.8 years old, range 18–73 years old) were categorized into the following five types: Type I 31.7%, Type II 30.2%, Type III 20.5%, Type IV 10.2%, and Type V 7.3%. The mean interobserver and intraobserver kappa scores were 0.85 (range 0.77–0.89) and 0.85 (range 0.79–0.92), respectively. For clinical outcomes, there was no significant difference in the postoperative Guille score for Type I patients (mean 9.01 ± 1.22). There was a significant increase in the postoperative Guille score in Type II, III, IV, and V, compared to the preoperative values ($P < 0.01$). For complications, two Type III patients reported pain, and one Type III patient had mild-to-moderate Trendelenburg gait. One Type IV patient had a mild Trendelenburg gait. And two Type V patients still had mild limping.

Conclusion: This classification is reproducible and serves as a tool for evaluating and treating FD in the femur and adjacent bones. Therefore, we recommend this classification for the diagnosis and treatment of FD-related deformities in the femur and adjacent bones.

Key words: Classification; Fibrous dysplasia; Radiograph; Treatment

Introduction

Fibrous dysplasia (FD) is a common skeletal disorder (monostotic or polyostotic) that results in pathological fractures, deformity, limping, and pain. FD was first identified by McCune and Bruch in 1937, which combined with precocious puberty, pathologic pigmentation of the skin, and hyperthyroidism.[1] Then, FD was characterized by a benign intramedullary fibro-osseous lesion.[2] The mutation of the GNAS-gene decreases the GTPase activity of the stimulatory G-protein, which increased the intracellular levels of cyclic adenosine monophosphate (cAMP) and interleukin-6 (IL-6) secretion.[3] The increased intracellular cAMP content and increased IL-6 secretion result in an increased numbers of osteoclasts and bone resorption. Primitive bone fails to transform into the mature lamellar bone and realign in response to mechanical stress.[4] The diagnosis of most FD can be made clinically after a complete radiological evaluation. And biopsy with histological evaluation was required in questionable cases and patients with a high suspicion of malignancy.[5] Generally, radiologic features of FD include a grayish "ground-glass" appearance, endosteal scalloping, shepherd's crook deformity, and intramedullary expansible lesion with a smooth sclerotic margin.[6]

The lower extremity is frequently affected by deformity, fracture, leg length discrepancy (LLD), and limping.[7–10] 10 Given the numerous surgical options and complicated deformities in the femur and adjacent bones, it can be challenging for orthopedic surgeons to evaluate the severity of deformities and select an appropriate treatment strategy. The surgical strategy is aimed at bone pain relief,[11] restoring normal femoral alignment,[8] gait normalization,[9] correcting LLD,[8] and preventing pathological fracture[5,7] for FD patients. Over the past decade, numerous publications have explored the issues mentioned above and suggested internal fixation and precise osteotomy.[8,9,12] With advancements in navigation and computer simulation, correcting femoral alignment and LLD became more accurate.[13,14]

To provide a uniform standard for surgical management, a clinical classification of FD is critical. Then, Ippolito et al.[15] developed a classification system that characterized only femoral deformities of polyostotic FD in six patterns, which served to guide the prediction of progression and surgical treatment. Type 1 was the normal neck-shaft angle with the altered shape of the proximal femur; Type 2 isolated coxa valgus with neck-shaft angle >140°; Type 3 was characterized by isolated coxa vara with neck-shaft angle <120°; Type 4 had lateral bowing of the proximal half of the femur associated with normal neck-shaft angle; Type 5 combined Type 4 and coxa valgus; Type 6 combined Type 4 and coxa vara.[15] However, this classification ignored monostotic FD patients and the adjacent joints of the femur, including hip and knee. Additionally, hip osteoarthritis and genu valgum resulting in severe pain and limping are also seen in polyostotic FD patients. Sierra et al.[11] first reported that total hip arthroplasty (THA) provided long-lasting pain relief and functional improvement for FD patients with a relatively

high complication rate. Moreover, monostotic and polyostotic FD lesions can vary in severity and require different surgical strategy. Therefore, clinical classification in FD is challenging with multiple barriers.

There are two existing systematic categories[15,16] that classify a variety of proximal femoral deformities in FD (Table 1). To correct the alignment of the lower limb and comprehensive LLD, both hip and knee should be taken into consideration. However, both classifications failed to guide the most appropriate strategy for treatment. According to our experience in assessing deformities and deformity-related change with pairing surgical strategies,[9,14,16] we proposed a radiographic classification with five types following corresponding treatment options. There are five following features, including proximal femur bone loss, coxa vara, femoral shaft deformity, genu valgum, and hip arthritis. Therefore, the purpose of the present study is: (i) to develop a reproducible and generalizable classification with corresponding surgical strategy for FD patients with deformity of femur and adjacent bones; (ii) to investigate the interobserver reliability and intraobserver reliability of this classification; and (iii) to evaluate the outcomes of surgical strategy based on this classification system.

TABLE 1 Present classification systems for fibrous dysplasia in low limb

Classification system	Details	Limitations
Ernesto Ippolito et al.[15]	Type 1: normal neck-shaft angle with the altered shape of the proximal femur Type 2: isolated coxa valga with neck-shaft angle ›140° Type 3: isolated coxa vara with neck-shaft angle ‹120° Type 4: lateral bowing of the proximal half of the femur associated with normal neck-shaft angle Type 5: type 4+ coxa valga Type 6: type 4+ coxa vara	(1) No description of proximal femoral strength (2) No description of limb alignment (3) No description of hip degeneration (4) Classification possible only for the level
Zhang et al.[16]	Type 1: without sever deformity Type 2: Reduction in the proximal femoral strength Type 3: Coxa vara ± type 2 Type 4: Varus deformity in the proximal femoral shaft ± type 2 Type 5: Coxa vara+ type 4 ± type 2	(1) Classification possible only for the proximal femoral level (2) No description of limb alignment (3) No description of hip degeneration

Methods

Explanation of this classification system

For radiographic analysis, the following features were determined in the lower extremity radiographs and axial computed tomography (CT) scans complex deformities based on standing limb alignment[6]: (i) focal thinning of cortical bone and involvement of the calcar in the proximal femur, which was measured on axial CT[17]; (ii) coxa vara, in which the neck-shaft angle was ≤120°; metaphyseal or femoral shaft deformity, in which varus or valgus malalignment was detected; (iii) genu valgum, in which the mechanical femorotibial angle (MFA) was ≥10°[18]; and (iv) hip arthritis, in which was detected according to the Kellgren–Lawrence grading system.[19]

Type I lesion was defined as none of the five features mentioned above (Fig. 1A). Type II lesion was defined as extensive bone loss of proximal femur (focal thinning of cortical bone and/or involvement of the calcar) (Fig. 1B). Type III deformity included coxa vara and femoral shaft deformity, either alone or in combination. Additionally, hip internal or external rotation was measured (Fig. 1C). Type IV deformity was characterized by severe genu valgum (Fig. 1D). Genu valgum can be caused either by FD lesions in the femur and/or tibia or by valgus osteotomy of femoral deformity. For the latter, FD patients who had normal mechanical femorotibial angles preoperatively often had increased mechanical femorotibial angles more than 10° postoperatively. And Type IV was to be alone or in combination with Type III. Type V deformity was defined as any type associated with severe hip arthritis (Fig. 1E). To eliminate LLD, preoperative planning set the intact side (monostotic cases) or relatively short side (polyostotic cases) as standard. The opening- and closing-wedge osteotomy were provided for preventing LLD intraoperatively.

Demographics

We retrospectively analyzed FD patients who were treated between January 2009 and January 2019 in our institution. We only included: (i) skeletally mature patients, who were not less than 18 years old[20]; (ii) radiological diagnosis was made according to the features, including a grayish "ground-glass" appearance, endosteal scalloping, shepherd's crook deformity, and intramedullary expansible lesion with a smooth sclerotic margin[6]; (iii) histological evaluation is only required to questionable cases or if malignancy is suspected; and (iv) all cases of Type II-V were histologically proven FD postoperatively. The study was approved by the Institutional Ethics Committee of West China Hospital (Chengdu, China, No. 2019342), and the study protocol adhered to the guidelines stipulated in the World Medical Association Declaration of Helsinki.

Perioperative Management

For corresponding treatment of each type, Type I only involved monitoring every 6 months.[5] And the indication for surgical treatment in Type II-V patients included the following: mechanical/weight-bearing bone pain, hip and/or knee stiffness, walking with a limp, fracture, and severe LLD.[5] Therefore, Type II was treated with internal fixation (IF) following simple curettage.[7] Type III was treated with the IF following single or multiple level valgus osteotomies and simple curettage.[8,9] Type IV was treated with a tibial osteotomy (TO)[21] or distal femoral osteotomy (DFO),[22] followed by femoral malalignment
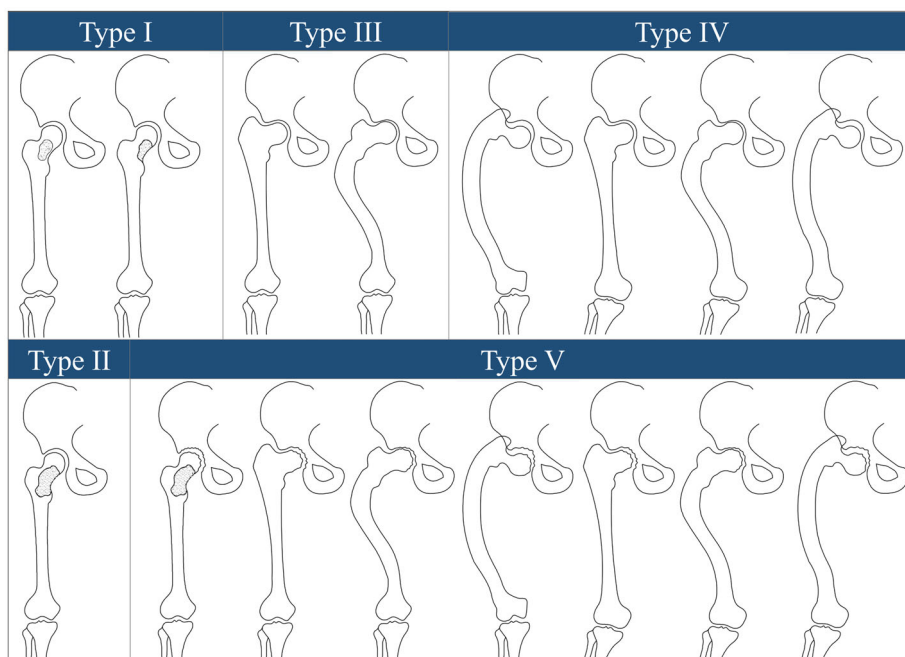

Fig. 1 The classification is indicated by Roman numerals, starting with I and ending with V.

correction. Type V was treated with THA and lesion curettage,[11] if necessary single-level valgus osteotomy was performed to fit the femur stem[14] (Table 2). Additionally, based on age, non-steroidal anti-inflammatory drugs and bisphosphonate were administrated to patients, who suffered from moderate to severe pain. Moreover, curettage was only performed in monostotic FD patients.

Type I patients followed up with a semi-annual assessment. Early weight-bearing protocols varied in Type II–IV. Type II patients were allowed full weight-bearing immediately after surgery. Type III and IV patients were allowed toe-touch weight-bearing within 6 weeks, and partial weight-bearing at 6–12 weeks postoperatively. Then progressive weight-bearing was permitted thereafter.[23] For Type V patients who underwent valgus osteotomy, the weight-bearing protocol was the same as that of Type III and IV. Without valgus osteotomy, the protocol for Type V patients was the same as that for Type II patients.[14] For the preoperative and postoperative clinical outcomes, all patients were assessed by the criteria of Guille et al.[9,24] The criteria included five factors, including pain, hip motion, limping, activities of daily living, and social activities. Each aspect was scored as 0 (unsatisfactory), 1 (average), or 2 (satisfactory). In all, >9 points were defined as excellent, 7 or 8 points as good, 5 or 6 points as fair, and <5 points as poor.

Agreement Analysis

For testing intraobserver and interobserver reproducibility, the principal investigator randomly chose 60 patients (12 per type) from our database. Four observers, including one radiological specialist and orthopedic specialist, and two residents, categorized the radiographic images based on this classification system. Four raters were blinded to each other and performed a two-round classification independently through picture archiving and communication systems with an interval of 6 weeks, which concealed the clinical information of the patients.

In polyostotic FD patients involving the bilateral lower limbs, we record the severer side for agreement analysis to avoid confusion. A mistake was made by the observers in distinguishing Type III from Type IV. Type IV was characterized by severe genu valgum, and Type III was defined as coxa vara and/or femoral shaft deformity. Some Type III patients were detected genu valgum in some degree postoperatively. The genu valgum, secondary to femoral osteotomy, was excluded.

Statistical Analysis

The intraobserver and interobserver reproducibility were evaluated with the Cohen kappa statistic and the criteria of Landis and Koch.[25] The standard answer was frequently chosen by the four observers. The normality of the continuous data was verified by the Shapiro–Wilk test. The homogeneity variance of data was tested by the Levene test. Normally and abnormally distributed parameters were assessed by the paired-samples t-test and the Mann–Whitney–Wilcoxon test, respectively. A P-value of <0.05 was determined to be

TABLE 2 Radiographic classification system of fibrous dysplasia in femur and adjacent bones

Indexes	Type I	Type II	Type III	Type IV	Type V
Extensive bone loss of proximal femur	–	+	+/–	+/–	+/–
Coxa vara	–	–	+/–	+/–	+/–
Femoral shaft deformity	–	–	+/–	+/–	+/–
Genu valgum	–	–	–	+	+/–
Hip arthritis	–	–	–	–	+
Intervention	Monitoring	Type II m IF +/– Curettage / Type II p IF	Type III m Single/ Multiple-level osteotomy + IF +/– Curettage / Type III p Single/ Multiple-level osteotomy + IF	Type IV m Single/ Multiple-level osteotomy + IF + DFO +/– Curettage / Type IV p Single/ Multiple-level osteotomy + IF + TO	Type IV* Single/ Multiple-level osteotomy + IF + TO	Type V m THA +/– Single-level osteotomy +/– Curettage / Type V p THA +/– Single-level osteotomy

Abbreviations: *, genu valgum secondary to femoral valgus osteotomy; DFO, distal femoral osteotomy; IF, internal fixation; m, monostotic; p, polyostotic; THA, total hip arthroplasty; TO, tibial osteotomy.

TABLE 3 Patient characteristics.*	
	No. of patients (%)
Age (years)	Median 34.8 (range 18–73)
Gender	
Female	121 (59.0)
Male	84 (41.0)
Category	
Monostotic	55 (26.8)
Polyostotic	141 (68.8)
McCune–Albright syndrome	9 (4.4)
Prevalence of different types	
Type I	65 (31.7)
Type II	62 (30.2)
Type III	42 (20.5)
Type IV	21 (10.2)
Type V	15 (7.3)
Follow-up (months)	
Total	Median 60 (range 6–120)
Type I	Median 63 (range 6–120)
Type II	Median 48 (range 12–120)
Type III	Median 72 (range 60–120)
Type IV	Median 60 (range 24–120)
Type V	Median 55 (range 23–65)

Note: *N = 205 patients (221 affected femurs).

statistically significant. Data analyses were performed using SPSS 21.0 software (IBM Corporation, Armonk, NY, USA).

Results

Classification of the Population
A total of 205 adult patients with FD in the femur and adjacent bones were enrolled in our institution. Of the included 205 patients, 55 had monostotic FD, 141 had polyostotic FD alone, and ten patients had McCune–Albright syndrome. All femurs could be categorized by this radiographic classification. The classification including five types of FD was noted (Table 3).

Agreement Analysis
In the specialist group, the mean intraobserver and interobserver kappa scores were 0.85 (range 0.80–0.89) and 0.87

(0.79–0.92), respectively. In the resident group, the mean interobserver and intraobserver kappa scores were 0.80 (range 0.77–0.81) and 0.83 (0.80–0.84), respectively. In total, the mean interobserver and intraobserver kappa scores were 0.85 (range 0.77–0.89) and 0.85 (0.79–0.92), respectively. The highest percentage of erroneous classification was observed in Type III and Type IV.

Clinical Outcome
At a median follow-up of 60 months (range 6–120 months), all patients were evaluated using the criteria of Guille et al.[9,24] There was a significant increase in the postoperative Guille score in Type II, III, IV, and V, compared to the preoperative values (P < 0.01) (Table 4). Regarding the variations of the functional outcome before and after surgery, Guille scores of Type III and V patients were sharply increased (P < 0.01). In the Guille score of Type III patients, there was a sharp increase in pain and hip motion postoperatively than those preoperatively. In the Guille score of Type V patients, all five factors, including pain, hip motion, limping, activities of daily living, and social activities, had a significant improvement postoperatively when compared to those preoperatively (P < 0.01).

Complication
Two Type III patients still complained of pain, and one Type III patient had a mild-to-moderate Trendelenburg gait. One Type IV patient had a mild Trendelenburg gait. After 6-month rehabilitation, the Trendelenburg gait of these patients was relieved. And two Type V patients still had mild limping. Physical therapy was administrated to those patients with Trendelenburg gait by strengthening the abductor muscles.

Discussion

Classification of the Population
In this retrospective study, we only included skeletally mature patients older than 18 years old, because the treatment of pediatric FD patients often differs from mature patients. Concerning in growth plate, intramedullary nail

TABLE 4 Preoperative/postoperative clinical scores evaluated by the modified criteria of Guille				
Indexes	Type II (n = 68)	Type III (n = 45)	Type IV (n = 21)	Type V (n = 15)
Pain*	0.33 ± 0.44/1.73 ± 0.42	0.13 ± 0.32/1.67 ± 0.55	0.24 ± 0.44/1.48 ± 0.68	0.00 ± 0.00/1.67 ± 0.72
Hip motion*	0.85 ± 0.68/1.62 ± 0.54	0.19 ± 0.38/1.57 ± 0.51	0.67 ± 0.66/1.33 ± 0.73	0.00 ± 0.00/1.73 ± 0.46
Limping*	0.66 ± 0.87/1.73 ± 0.44	0.24 ± 0.60/1.51 ± 0.57	0.81 ± 0.87/1.29 ± 0.64	0.13 ± 0.35/1.60 ± 0.63
Activities of daily living*	0.51 ± 0.82/1.70 ± 0.46	0.84 ± 0.35/1.58 ± 0.54	0.71 ± 0.72/1.43 ± 0.51	0.20 ± 0.41/1.87 ± 0.35
Social activities*	0.32 ± 0.65/1.55 ± 0.52	0.21 ± 1.44/1.51 ± 0.58	0.43 ± 0.51/1.05 ± 0.59	0.00 ± 0.00/1.60 ± 0.63
Total*	2.66 ± 2.43/8.38 ± 1.37	0.65 ± 0.98/7.81 ± 0.96	2.86 ± 1.98/6.57 ± 2.09	0.33 ± 0.49/8.47 ± 1.68
P value	<0.01	<0.01	<0.01	<0.01

Notes: Clinical outcomes were scored as 0 (unsatisfactory), 1 (average), or 2 (satisfactory). For a potential maximum total of 10 points, >9 points were defined as excellent, 7 or 8 points as good, 5 or 6 points as fair, and <5 points as poor; * Mean score.
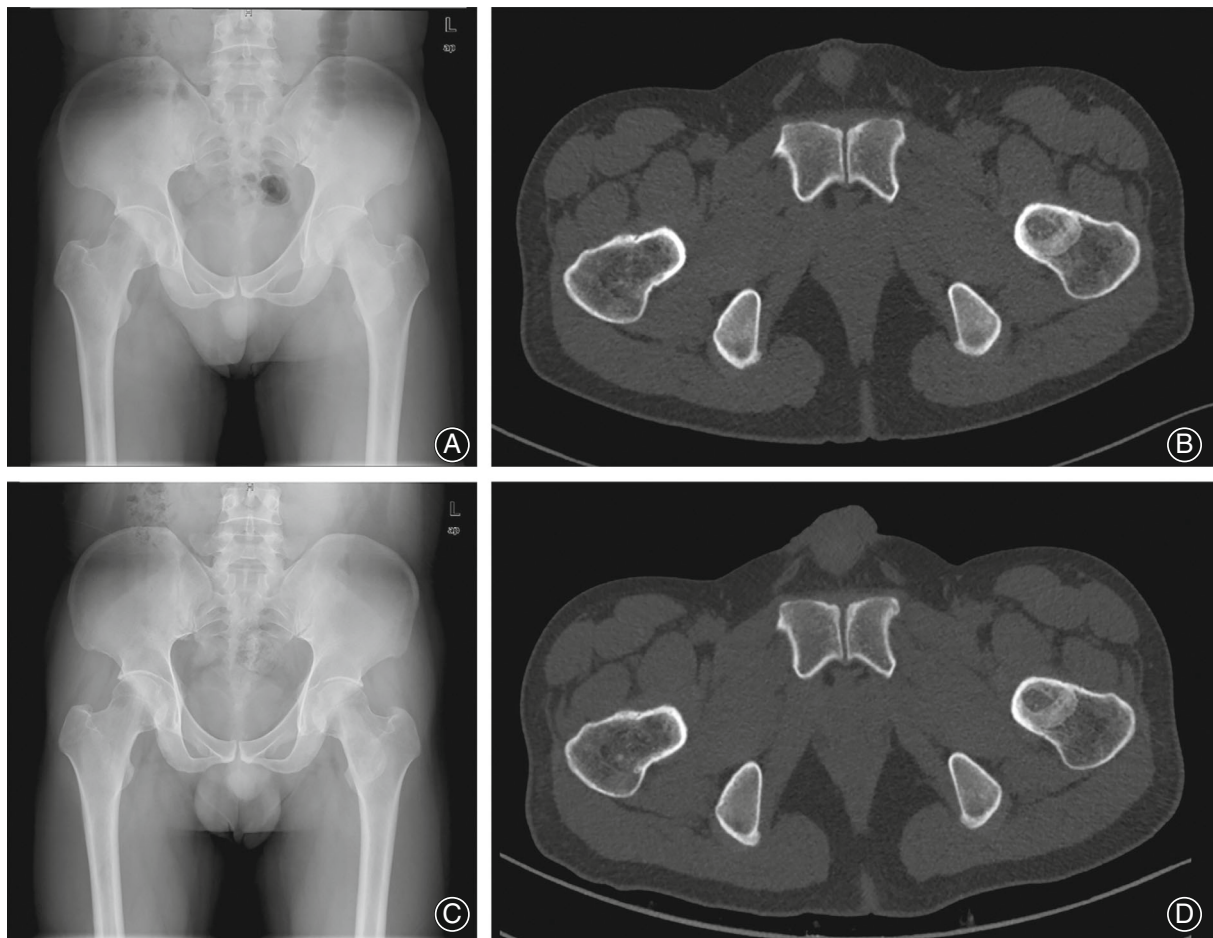
Fig. 2 Type I in a 33-year-old female. The lesion was present in the left femoral neck with intact cortical bone and calcar on CT (A, B). Type I lesion was stable at the 24-month follow-up (C, D).

and THA are not recommended. Although surgical procedures may be different in monostotic or polyostotic FD patients, including curettage and bone graft, both patients follow the same principle in correcting the deformity. The management of femoral FD include five strong points, including the range of affected bone and bone strength, the number and site of deformities, LLD, adjacent joint degeneration, and the timing of surgery.[5,10] This classification considered all five points mentioned above. Additionally, patients in each type had considerable clinical outcomes following corresponding treatment. For outcome-oriented strategy, all patients had appropriate diagnosis and intervention. Therefore, this radiographic classification was proposed to treat FD in the femur and adjacent bones, according to the clinical outcomes, corresponding treatment, and radiographic features.

Agreement Analysis
In Ippolito et al.'s[15] study, three orthopedic surgeons and one pathologist evaluated FD femurs on two occasions

with an interval of 6 weeks. The intraobserver (0.855) and interobserver (0.833–0.871 range) agreement were both excellent. And they reported the highest percentage of mistakes was for types 1, 2, and 3 (in cases with mild shepherd's crook deformity) versus types 4, 5, and 6. In our primary classification system,[16] two senior orthopedic surgeons evaluated the cases for two rounds with a 6-week interval. The intraobserver and interobserver agreements were both excellent. In this study, the mean interobserver and intraobserver kappa scores were 0.85 (range from 0.77 to 0.89) and 0.85 (range from 0.79 to 0.92), respectively. Although the agreement was excellent, the dispute was focused on the genu valgum between Types III and IV. The definition of genu valgum is MFA $\geq 10°$.[18] If the MFA of Type III patients increased over $10°$ after femoral osteotomy in preoperative planning, these patients were then classified as Type IV. Then, surgical treatment is inevitable for improving MFA. Because of the consistent one-to-one match between radiographic image and intervention, surgeons must be prepared to predict the genu
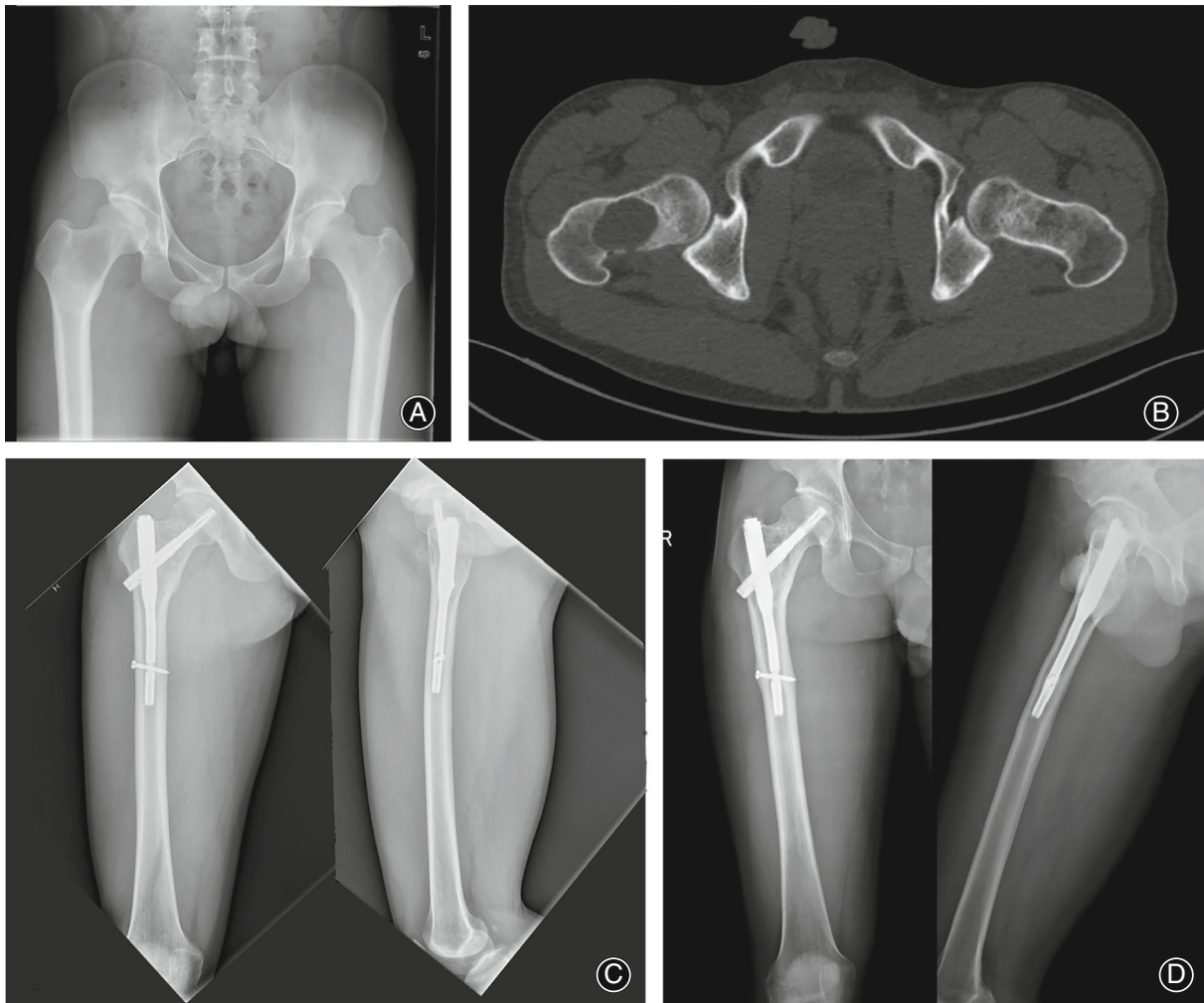
Fig. 3 Type II lesion in a 29-year-old male. The lesion was located in the right proximal femur, and the neck-shaft angle was normal without coxa vara deformity (A). The extensive bone loss of proximal femur with focal thinning of cortical bone (B). Postoperatively, the 7- and 68-month follow-up showed a slight absorb of bone grafts without recurrence of lesion or fracture (C, D).

valgum secondary to femoral osteotomy. Therefore, we recommend classifying these patients as Type IV for correspondence surgical options.

Surgical Management and Clinical Outcome in Type I

Type I lesions have no focal thinning of cortical bone and involvement of the calcar in the proximal femur, and we recommend conservative treatment with monitoring every 6 months (Fig. 2). Bone pain in FD should be discreetly assessed. Focal or weight-bearing pain may indicate an imminent or impending fracture.[26] Physiotherapy and pain medication can be administered, including opioids and nonsteroidal anti-inflammatory drugs.[5] Besides, intravenous bisphosphonate is proposed for moderate to severe levels of persistent pain, even in children and adolescents.[12,27] For patients with persistent pain after the pharmacological treatment, an intramedullary nail is necessary. Additionally,

surgical interventions are recommended when conservative treatment fails or if there are symptoms of an impending fracture. During follow-up, no severe complications were detected in Type I patients.

Surgical Management and Clinical Outcome in Type II

Type II lesions are characterized by focal thinning of cortical bone and/or involvement of the calcar, without other femoral deformities. Type II patients often have mechanical or weight-bearing bone pain, which is a sign of stress or an impending fracture.[5,28] Therefore, curettage, bone graft, and internal IF are recommended for monostotic FD patients (Fig. 3). The efficacy and complications of bone graft are still controversial.[5,29] Stephenson et al. reported that skeletally mature patients with monostotic FD had a more satisfactory result of bone graft than immature patients. In addition, small and solitary lesions had a good response to curettage
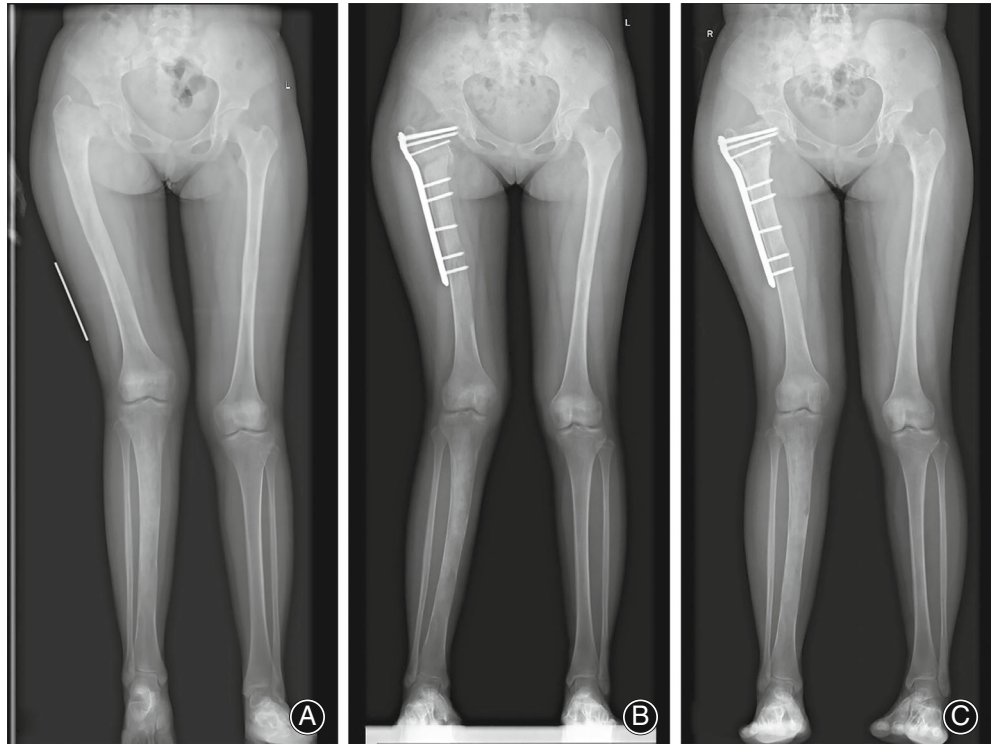
Fig. 4 Type III lesion in a 16-year-old female. The lesion was present in the intertrochanteric region with coxa vara (A). Postoperative 7- and 24-month films showed a good union of the osteotomy site. The coxa vara has been correct without recurrence of lesion or re-progress of deformity (B, C).

and bone graft.[20] Cortical allografts are slowly resorbed and simultaneously replaced by the host bone.[4,8] To provide persistent structural supports, cortical allografts are recommended for monostotic FD patients. While curettage and bone graft are not necessary for polyostotic FD patients. The relative procedure often results in significant blood loss,
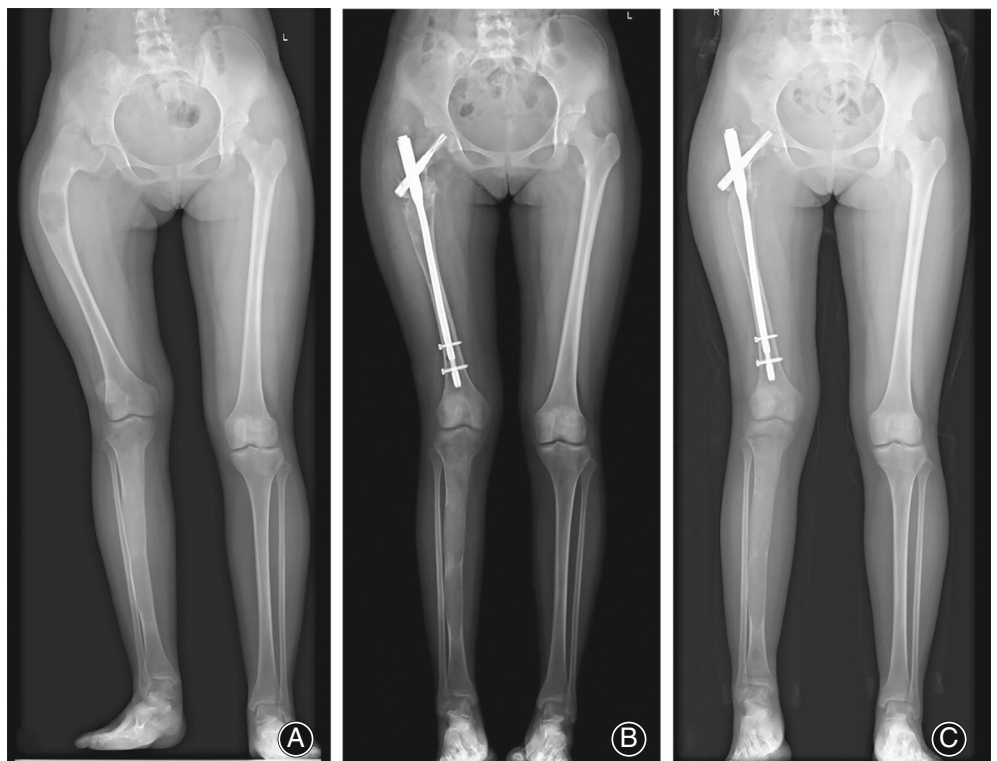
Fig. 5 Type III lesion in a 16-year-old female. The lesion was present in the right femoral shaft (A). The subtrochanteric osteotomy was performed in the dome of the deformity. The radiograph of 2- and 24-month follow-up showed good union of osteotomy site, and alignment has been corrected without recurrence of lesion or re-progress of deformity (B, C).

and the polyostotic FD lesion usually remodels the graft into FD tissue over time.[30] However, there are limited indications for the use of bone graft in polyostotic FD patients in selected cases. First, the bone graft is recommended in conjunction with internal fixation for providing primary stability. Second, strut allograft is used as reconstruction associated with allograft prosthetic composite surgery. Third, the bone graft is provided for the small bones with repeated fractures, including phalanx and metacarpus.[30] In Type II, the intramedullary lesion should be adequately bridged by IF including dynamic hip screw (DHS), anatomical plate, or intramedullary nail.[7,30,31] In this study, only one Type II patient had mild-to-moderate pain postoperatively.

Surgical Management and Clinical Outcome in Type III
Type III patients refer to the deformity of coxa vara and/or femoral shaft associated with bone pain. We recommend either single-level osteotomy or double-level osteotomy. For the single-level osteotomy, the subtrochanteric region[8,9,32] (Fig. 4) and the dome of the deformity[9,33,34](Fig. 5) are recommended for osteotomy. However, a double-level osteotomy is strongly considered for correcting severe deformity.[9,33,35] After osteotomy, orderly curettage, massive impaction allograft, and IF are

performed for monostotic FD patients.[7,36,37] In polyostotic FD patients, intramedullary nail fixation is recommended by following osteotomy. The choice of IF is still controversial. Previously, some authors suggested the longer DHS rather than intramedullary nailing, because of its ability in correcting varus, avoiding rotational deformities of the femoral neck, and simplifying the procedure.[8,32,34] However, some studies reported that intramedullary nails provided better biomechanical support than DHS.[9,36] In general, we recommend intramedullary nails for the following reasons. First, it provides sufficient stability that prevents stress fracture and screw loosening or pullout, especially in polyostotic FD patients.[8,31] Second, it accommodates multiple-level osteotomy.[33] To increase the initial stability, the transversal surface of the femur after osteotomy should be entirely matched for locking each other. Additionally, the intramedullary lesions with sclerotic rim have sufficient bone mass, which provides adequate stability for nail fixation. Therefore, the sclerotic bone should be discreetly preserved when curettage and canal reaming is used. Moreover, rotational deformities of the femur could be gradually corrected. In this study, two Type III cases still complained of pain, remained in pain, owing to mild hip joint degeneration. One Type III patient had a mild-to-moderate Trendelenburg gait. Preoperatively, the neck-shaft
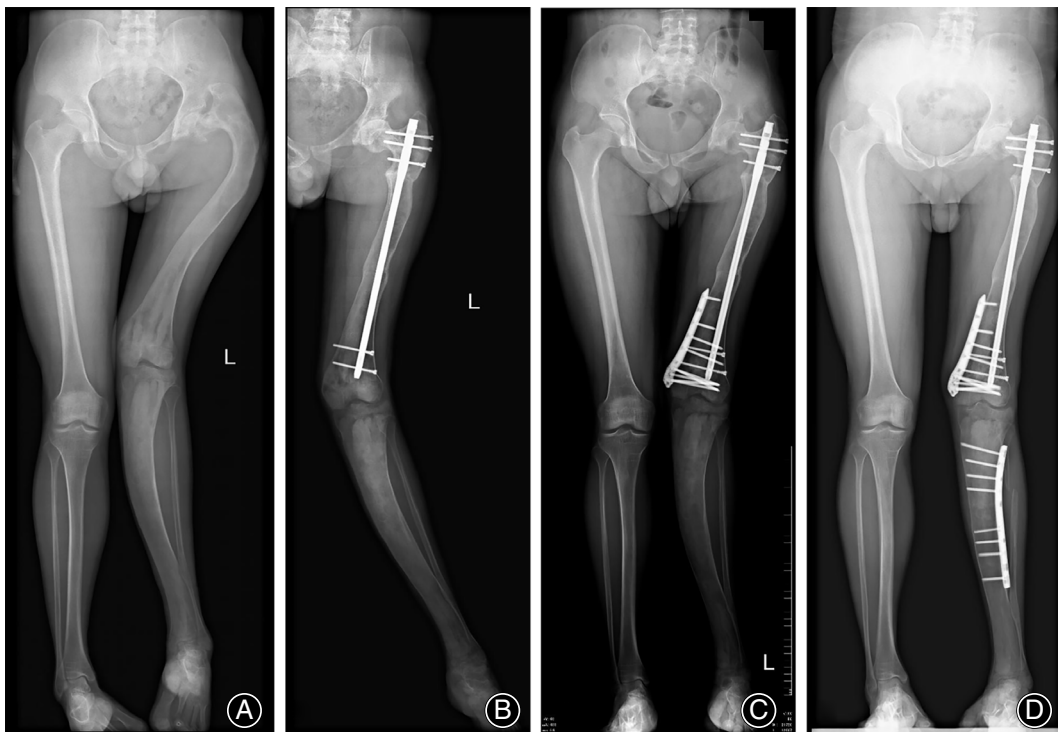

Fig. 6 Type IV lesion in a 25-year-old male. Type IV lesion was associated with coxa vara, femoral shaft deformity, and genu valgum (A). The radiograph of 9-month follow-up after the double-level osteotomy showed a good union of osteotomy site and alignment correction (B). Then the distal femoral osteotomy was carried out, and radiographs showed bone union and femoral alignment correction at 21 months after the double-level osteotomy and 10 months after distal femoral osteotomy (C). After that, tibial valgus osteotomy was performed, and radiographs showed good lower limb alignment and bone union at 80 months after the double-level osteotomy, 69 months after distal femoral osteotomy and 59 months after tibial valgus osteotomy (D).
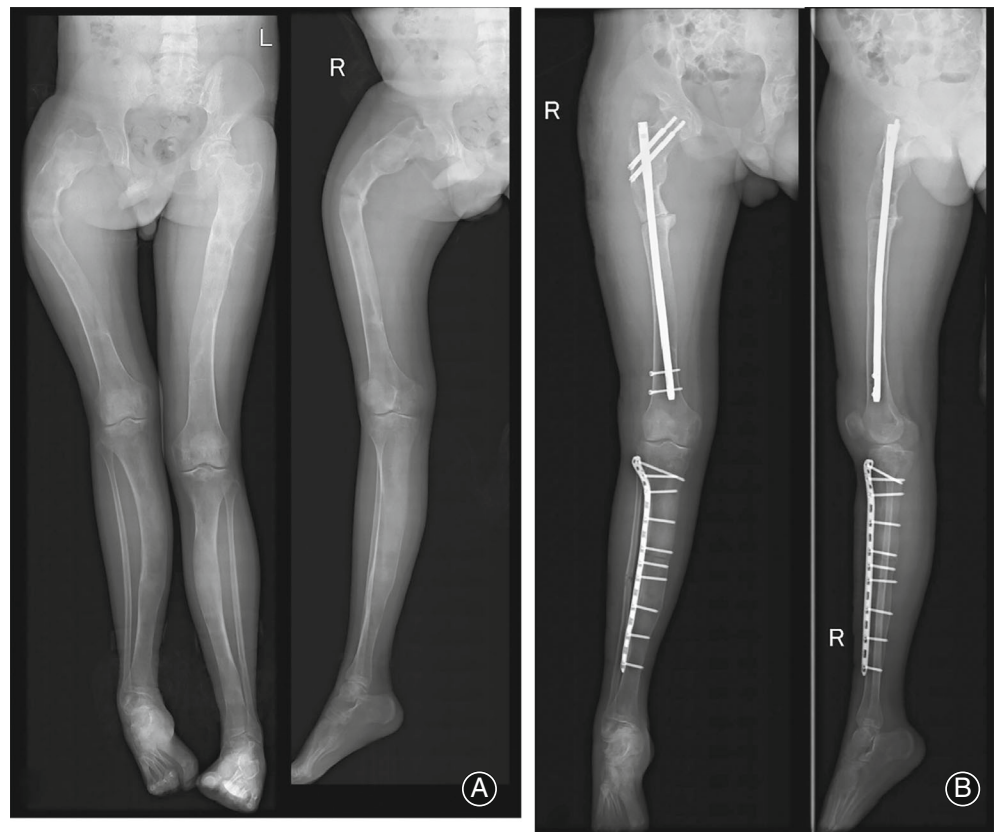
Fig. 7 Type IV lesion in a 24-year-old male. A fracture was present in the dome of the deformity with severe genu valgum (A). The tibial valgus osteotomy was performed 3 months after the femoral valgus osteotomy, and the lower limb alignment was corrected (B).

angle of this patient was only 75°. Over-tensioning of the gluteus medius was inevitable postoperatively after the correction of coxa vara.[38]

Surgical Management and Clinical Outcome in Type IV

Type IV lesions are detected in FD patients with severe genu valgum. When combined with Type III deformity, a two-stage treatment was recommended. Complex femoral deformities with or without lesion curettage were corrected first. After 6 months of rehabilitation, the second-stage procedure was performed for patients, who still reported typical symptoms. Therefore, we suggest TO or DFO for achieving a satisfactory appearance, correcting limb alignment, and relieving pain.[22,39] DFO is recommended for monostotic Type IV patients, in which FD lesions only involve the femur. While TO is suggested for polyostotic FD patients because the genu valgum is mainly caused by the proximal tibia (Fig. 6). Moreover, TO is also recommended for polyostotic FD patients with genu valgum secondary to femoral valgus osteotomy (Fig. 7). Type IV lesions are uncommon and the most challenging type and patients with this type of deformity have a lower Guille score, compared to other types of patients. In the current study, only one Type IV patient had a mild Trendelenburg gait, because of the over-tensioning of the gluteus medius.

Surgical Management and Clinical Outcome in Type V

Type V lesions are found in FD patients with severe hip arthritis, combined with other types (II/III/IV). The deformities of the femoral shaft, coxa vara, and genu valgum, are significantly associated with the degeneration of the hip and knee. Additionally, polyostotic FD is found to be more prevalent in hip arthritis than monostotic FD.[40,41] Although a small fraction of FD patients with mild deformity may be classified into Type V, the relatively low Guille score was preliminary evidence to guide treatment. Sierra et al.[11] first reported THA in patients with FD. When hip arthritis causes severe symptoms, total hip arthroplasty was suggested for Type V patients. For surgery, a cemented stem was suggested to obtain a lower revision rate when compared to an uncemented stem. However, our institution reported that the long uncemented stem showed reliable fixation at mid-term follow-up[14], using Mimics V17.0 Software (Materialise, Leuven, Belgium), precise preoperative planning, and simulation of the osteotomy. Furthermore, implantation of the prosthesis stem in a three-dimensional reconstructive model is essential. For the femoral component, we recommend a long-tapered stem with a full coating, which engages in normal diaphyseal bone bypassing the lesion areas at least 2 femoral canal diameters to decrease the risk of postoperative fractures[11] (Figs 8 and 9). Stem
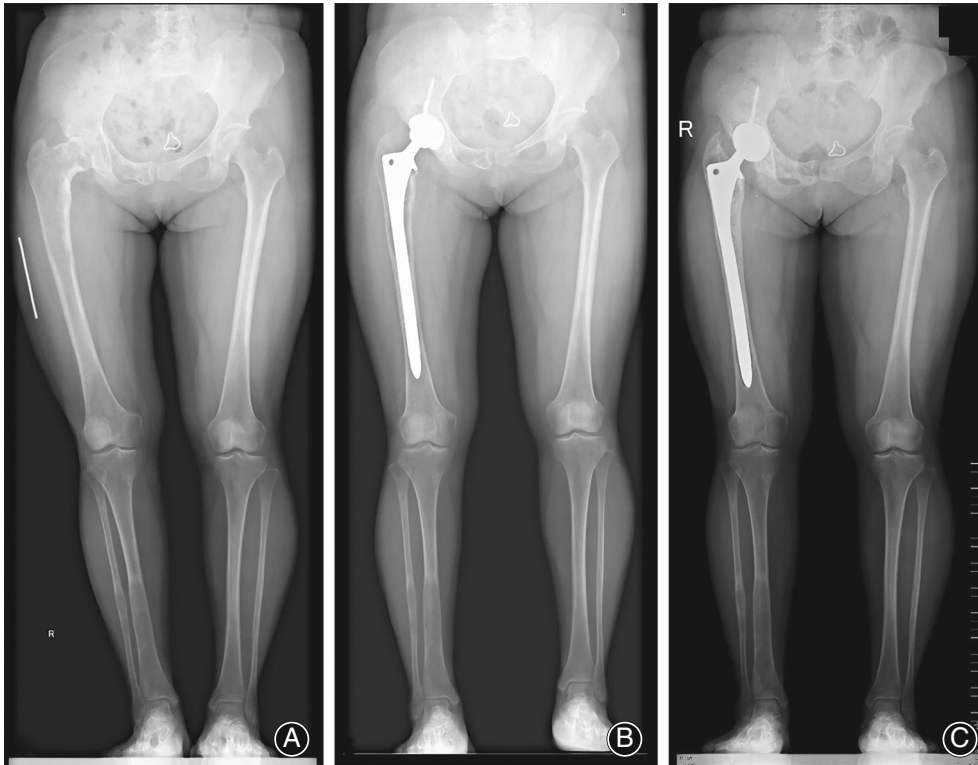
Fig. 8 Type V lesion in a 42-year-old female. Radiographs showed right hip osteoarthritis associated with coxa vara (A). Postoperatively, 1- and 35-month follow-up showed no recurrence of lesion or prosthesis loosening (B, C).

implantation is a crucial point. First, the femoral head is cut off after internal fixation removal to prevent further fracture. Then lesion curettage is done in the proximal femoral canal. Second, the precision osteotomy is assisted by a patient-specific instrument, using a micropendulum saw. After osteotomy, the lesion is evacuated in the rest of the femoral canal. During a thorough evacuation, all the processes of the femoral cavity are preserved, especially in the isthmus. Third, the femoral canal is gradually reamed by rasps until an optimal endosteal contact is obtained.
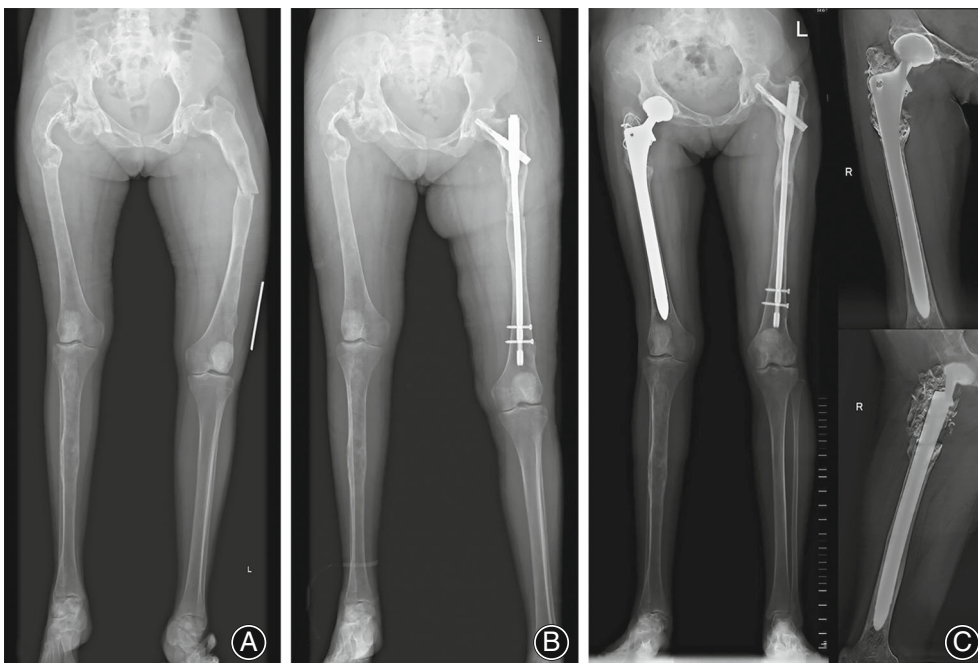

Fig. 9 Type V lesion in a 38-year-old female. A fresh fracture was detected in the left femoral shaft, while an old fracture was present in the right femoral neck secondary to the lesion in the right proximal femur (A). At 6 months after internal fixation, she went to our department for total hip arthroplasty (B). Postoperatively, a 16-month follow-up showed no recurrence of lesion or prosthesis loosening (C).

Fourth, a massive autograft is impacted in the femoral canal from the femoral head bone. Although autograft is not recommended for polyostotic FD, autogenous femoral head bone graft provides temporary augmentation for the stem.[30] Finally, the long-tapered stem with a full coating is inserted into the canal bypassing the lesion areas until rigid fixation. Two Type V patients had mild limping, because of LLD after THA.

Limitation of the Study

There were several limitations to this study. First, the classification was based on a retrospective clinical study that included both monostotic and polyostotic FD. The agreement analysis and surgical outcomes were evaluated by a relatively small sample size. Therefore, the quality of the evidence is insufficient to support a strong recommendation for this classification. Second, the follow-up time was significantly different among the five groups. Thus, multicenter studies with a longer follow-up time are needed to reach sufficient conclusions on this classification, especially for Type V. Third, surgical strategies and classifications are appropriate only for adult patients since most patients in our department were adults (median age 34.8 years, range 18–73). Finally, we have treated some patients with pathologic fractures, but those patients were excluded by this classification system.

Conclusions

We developed a new classification system for FD in the femur and adjacent bones, which was built on a review of the literature and clinical outcomes. Agreement analysis of the classification showed that this classification system is reproducible. Furthermore, follow-up evaluations showed pain relief and gait improvement in most FD patients. Therefore, we believe that this classification system can serve as a tool for evaluating and treating these deformities.

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