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Fibrous dysplasia: Clinical features, deformities, management, and outcomes

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Fibrous dysplasia (FD) is a benign bone lesion characterized by the replacement of normal bone with fibro-osseous tissue due to a developmental disorder of bone.[1-4] First described by Lichtenstein[5] in 1938, FD has remained a subject of clinical and academic interest for more than eighty years. Fibrous dysplasia arises from a developmental failure in the remodeling of primitive bone into mature lamellar bone. As a result, the normal cancellous bone in the medullary canal is replaced by immature fibro-osseous tissue.[1-4,6-9] As this immature bone undergoes incomplete remodeling, proper mineralization does not occur, leading to reduced mechanical strength and predisposing the bone to microfractures and deformity.[1,2,4,8]

Fibrous dysplasia is most frequently diagnosed during the first three decades of life, particularly in late childhood and adolescence, although it may occur at any age.[4,8] The disease can involve a single

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ABSTRACT

Objectives: The aim of this study was to evaluate the rate of proximal femoral involvement and the incidence of shepherd's crook deformity in patients with fibrous dysplasia (FD) and to identify possible factors associated with the development of shepherd's crook deformity.

Patients and methods: Between January 1990 and December 2022, a total of 158 consecutive patients (78 males, 80 females; mean age: 23.8 ± 15.0 years; range, 3 to 71 years) diagnosed with FD were retrospectively analyzed. Presenting symptoms, lesion location, radiological findings, treatment modalities, development of deformity, postoperative complications, and the need for secondary surgery were recorded.

Results: The mean follow-up was 64.2 ± 38.4 (range, 24 to 210) months. Among the patients, 125 had monostotic and 33 had polyostotic lesions. Of the 215 lesions identified, 83 were treated surgically, while 132 were managed with clinical observation. Shepherd's crook deformity was observed in nine of 75 proximal femoral lesions. The deformity was significantly more common in polyostotic than in monostotic lesions (25.0% *vs*. 5.9% respectively; $p = 0.026$). McCune-Albright syndrome (MAS) was present in two patients, both exhibiting shepherd's crook deformity, whereas the rate was 9.6% among patients without MAS. Valgus osteotomy was performed in all nine patients with shepherd's crook deformity, improving the mean neck-shaft angle from 90.5° (range, 85 to 105°) preoperatively to 125.6° (range, 120 to 130°) postoperatively. In six of these patients, no marked loss in the postoperative neck-shaft angle was observed, while three patients required secondary surgery due to recurrence of the deformity.

Conclusion: Most lesions can be managed successfully with observation, while surgery is indicated for the eradication of symptomatic lesions, prevention of pathological fractures, and correction of deformities. Shepherd's crook deformity seems to be more common in patients with polyostotic lesions and MAS. Although valgus osteotomy can achieve radiographic improvement, recurrence of deformity and the need for multiple surgeries are not uncommon.

Keywords: Bone deformities, fibrous dysplasia, McCune-Albright syndrome, pathological fracture, shepherd's crook deformity, surgical treatment.

bone (monostotic) or multiple bones (polyostotic) and may be associated with extraskeletal manifestations, including hyperfunctioning endocrinopathies, known as McCune-Albright syndrome (MAS).[8-13] The monostotic type is more common, typically asymptomatic, and often detected incidentally. In contrast, the polyostotic form involves multiple bones, tends to grow more rapidly, and may continue to expand even after skeletal maturity. The risk of deformity development and pathological fractures is higher in this patient group, and they may also present with associated MAS or intramuscular myxomas (Mazabraud syndrome).[4,8,14,15] The clinical presentation of FD varies depending on the lesion's location, size, and type. It may be asymptomatic and identified incidentally, or it may present with symptoms such as pain, limping secondary to limb-length discrepancy, deformity, or pathological fractures.[4,8,16] The true incidence and prevalence still remain unknown, as most cases are asymptomatic.[4] However, these lesions are not uncommon and account for approximately 5 to 7% of all benign bone tumors.[1,4,17]

Persistent mechanical strain and repeated occult pathological fractures can lead to deformities in patients with FD.[2,4] The most characteristic deformity is coxa vara accompanied by lateral bowing of the proximal femur, known as the shepherd's crook deformity. These deformities are often symptomatic and may cause pain, limitation of hip motion, limb shortening, limping, and a Trendelenburg gait.[1,2,4,8,11,15]

There is no consensus on the treatment algorithm for FD. Most lesions are asymptomatic and can be managed with clinical observation. Surgery is indicated for the eradication of symptomatic lesions, prevention of pathological fractures, and correction of deformities.[2,4,8,13] Various surgical techniques have been described; however, the management of FD remains challenging, particularly in the correction of deformities.[1,2,4,8,15,18,19] Given the heterogeneous clinical presentation and variable progression patterns of FD, long-term institutional experience can provide meaningful insights into its natural course and treatment strategies. In the present study, we, therefore, aimed to evaluate the rate of proximal femoral involvement and the incidence of shepherd's crook deformity in patients with FD, to identify possible factors associated with the development of shepherd's crook deformity, and to examine the feasibility of treatment modalities in these patients.

PATIENTS AND METHODS

This single-center, retrospective study was conducted at Ankara University Faculty of Medicine, Department of Orthopedics and Traumatology between January 1990 and December 2022. All patients diagnosed with FD were screened. The bone tumor archive of our center was used to identify these patients. The diagnosis of FD was primarily based on characteristic radiographic features, such as a "ground-glass" appearance, referring to a homogeneous, radiolucent lesion without a visible trabecular pattern on radiographs. Histological confirmation was indicated in cases with atypical imaging findings. Pathology reports were reviewed for patients who underwent biopsy due to inconclusive radiological evaluation, as well as for those who underwent surgical treatment. All patients diagnosed with FD based on radiological or histological findings during the specified time period were included in the study. Patients with a follow-up period of less than two years (n = 12) and those with incomplete radiological data (n = 8) were excluded from the study. Finally, a total of 158 consecutive patients (78 males, 80 females; mean age: 23.8 ± 15.0 years; range, 3 to 78 years) diagnosed with FD were included. A written informed consent was obtained from each patient. The study protocol was approved by the Ankara University Faculty of Medicine Ethics Committee (Date: 17.02.2025, No: İ01-83-25). The study was conducted in accordance with the principles of the Declaration of Helsinki.

Symptoms at the time of the initial outpatient clinic evaluation were obtained from the medical records. Imaging studies, including radiographs, computed tomography (CT), magnetic resonance imaging (MRI), and bone scans, were reviewed. The size, number, and location of lesions were determined based on a review of radiological images. Patients with involvement of a single bone were classified as monostotic, whereas those with involvement of multiple bones were classified as polyostotic. The presence of pathological fractures or deformities was primarily evaluated using radiological images. The femoral neck-shaft angle was measured in patients with coxa vara deformity to assess the severity of the deformity. Radiological measurements were performed using DiCOM viewer software (RadiAnt DICOM Viewer; Medixant, Poznań, Poland). For this study, imaging assessments were performed by two blinded orthopedic specialists experienced in musculoskeletal oncology. Intraclass correlation coefficients (ICCs) were calculated for all radiological

TABLE I
Patient demographics

	n	%	M/F	Mean ± SD	Median	Min-Max
Age (year)				23.8 ± 15.0	20	3-71
Sex						
Male	78	49.4	0.97			
Female	80	50.6				
Presenting symptoms						
Incidental	69	43.7				
Pain	62	39.2				
Pathologic fracture	21	13.3				
Swelling	4	2.5				
Limping	2	1.3				
Type of involvement						
Monostotic	125	79.1				
Polyostotic	33	20.9				
Syndromes						
Mc Cune-Albright	2	1.3				
Mazabraud	2	1.3				

SD, standard deviation.

measurements to determine interobserver reliability. The interobserver ICC was 0.89, indicating good reliability (ICC between 0.75 and 0.90).

Lesions were classified according to the treatment approach as either surgically treated or conservatively managed. Surgical procedures including curettage, grafting, cementing, corrective osteotomy, plate fixation, intramedullary nailing, and excision were documented by reviewing the operative reports in hospital records. Postoperative complications, such as surgical site infection, implant failure, or pathological fracture, were recorded. Lesions involving the proximal femur were specifically evaluated for the presence of shepherd's crook deformity. Patients who developed shepherd's crook deformity were evaluated with respect to the surgical methods employed, choice of hardware, postoperative outcomes, and the need for secondary surgery. Endocrinology consultation notes were also reviewed for accompanying endocrinopathies, including MAS.

Statistical analysis

Statistical analysis was performed using the IBM SPSS version 22.0 software (IBM Corp., Armonk, NY, USA). Descriptive data were presented in mean ± standard deviation (SD), median (min-max) or number and frequency, where applicable. The Shapiro-Wilk test was used to assess normality of the data. The chi-square or Fisher exact tests were used to compare categorical variables between groups. The Mann-Whitney U test was performed to analyze the non-parametric data between groups. A p value of <0.05 was considered statistically significant.

RESULTS

The mean follow-up was 64.2 ± 38.4 (range, 24 to 210) months. Localized bone pain was present in 62 cases as the initial presenting symptom. Twenty-one patients were diagnosed after experiencing a pathological fracture. Localized swelling was the initial symptom in four cases. Two patients were diagnosed during the evaluation for limb-length discrepancy. Meanwhile, 69 patients were incidentally diagnosed on radiographs (Table I).

Of the 158 patients, 125 had monostotic lesions, while 33 had polyostotic lesions. A total of

TABLE II
Characteristics of lesions

	Number of patients		Total number of lesions	
	n	%	n	%
Monostotic	125	79.1	125	58.1
Polyostotic	33	20.9	90	41.9
Total	158		215	

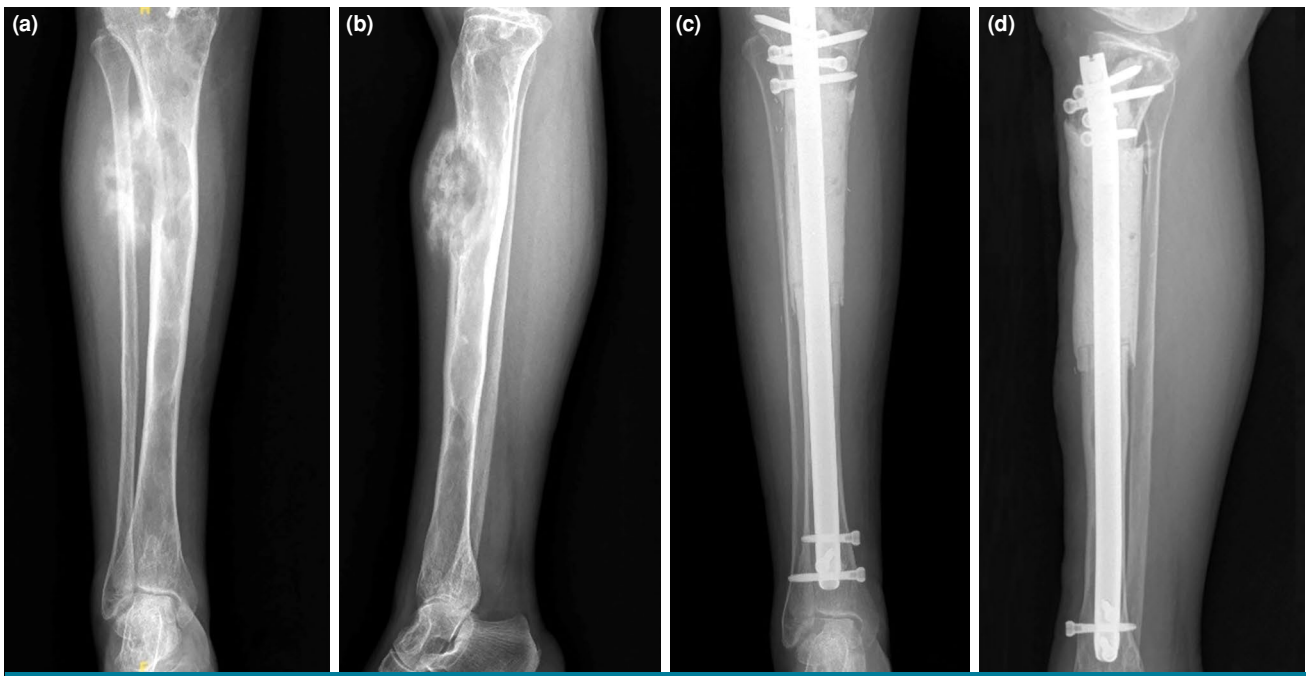

FIGURE 1. Malignant transformation in a 71-year-old female with fibrous dysplasia of the tibia. A patient with an 14-year history of fibrous dysplasia under follow-up at another center was referred to our institution for a newly developed expansile lytic lesion in the metadiaphysis of the right tibia. **(a, b)** Radiographs show an expansile lytic lesion with cortical destruction on a ground-glass matrix background, raising suspicion for malignant transformation. The biopsy confirmed osteosarcoma adjacent to an area of fibrous dysplasia. **(c, d)**. Resection, intramedullary nailing, and cementing were performed.

215 lesions were identified. Among these lesions, 125 were monostotic and 90 were polyostotic (Table II). McCune-Albright syndrome (MAS) was observed in two patients. Mazabraud syndrome, characterized by the coexistence of FD and intramuscular myxomas, was identified in another two distinct patients.

Surgical treatment was performed for 83 out of the 215 lesions, while the remaining 132 lesions were managed nonoperatively. Among the 83 surgically treated patients, 13 underwent surgery due to deformity and 15 due to pathological fractures. Surgical treatment was prophylactically performed in 54 patients due to the high risk of pathological fracture or deformity, determined by lesion location, size, and the presence of severe symptoms. In one patient, FD located in the tibial diaphysis transformed into osteosarcoma 14 years after the initial diagnosis. This patient underwent surgery due to malignant transformation (Figure 1). Table III summarizes the treatment methods and surgical indications. Case examples of surgical treatments are presented in Figures 2, 3, and 4.

The anatomical locations of the lesions are presented in Table IV. The most commonly affected sites were the femur (n = 86), tibia (n = 53), humerus (n = 24), and pelvis (n = 20). Surgical treatment was most frequently performed at the proximal femur and the tibial diaphysis (Table 4).

Pathological fractures were observed in 21 patients. Of these, 13 were located in the femur, four in the tibia, and four in the humerus. Conservative treatment was performed in six

TABLE III
Treatment methods

	Number of lesions		Total
	n	%	n
Clinical observation	132	61.4	215
Surgical procedures	83	38.6	
Surgical indications			
Deformity correction	13	15.7	
Pathologic fracture	15	18.1	
Prophylactic surgery			83
Fracture/deformity prevention Severe symptoms	54	65.0	
Malign transformation	1	1.2	

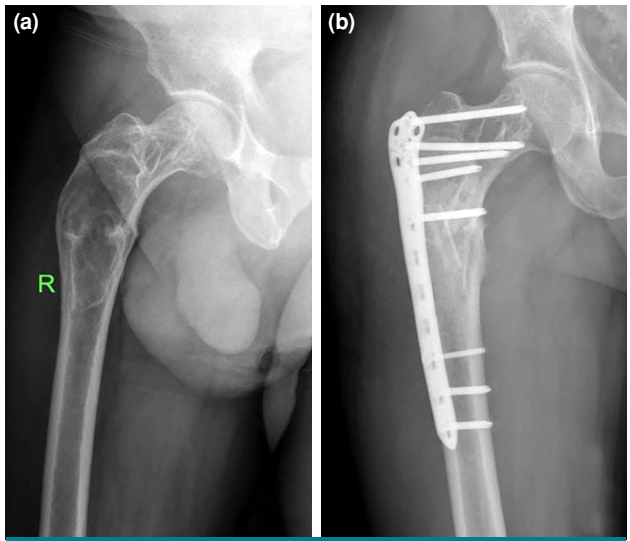

FIGURE 2. Fibrous dysplasia of the proximal femur in a 23-year-old male. **(a)** Preoperative radiograph shows the typical "ground-glass" appearance. **(b)** Curettage, iliac crest autografting, and internal fixation with a locking plate were performed.

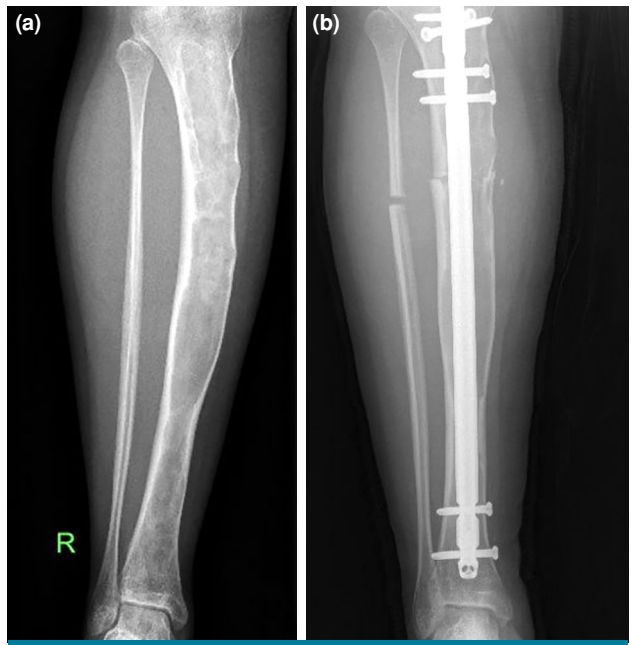

FIGURE 3. Fibrous dysplasia of the tibia in a 21-year-old male. **(a)** Preoperative radiograph shows anterior tibial bowing. **(b)** Corrective osteotomy followed by intramedullary nailing was performed.

patients, while 15 patients underwent surgery due to pathological fractures. Among the lesions with pathological fractures, five were polyostotic, all of which underwent surgical treatment. Among the 15 monostotic lesions, six were managed conservatively and nine underwent surgical treatment. Union was achieved in all cases.

Patients' characteristics with shepherd's crook deformity, along with their surgical treatments, are presented in Table V. Patient age at the time of deformity was 23.4 ± 14.9 (range, 9 to 54) years. Union at the osteotomy site was achieved in all cases at the

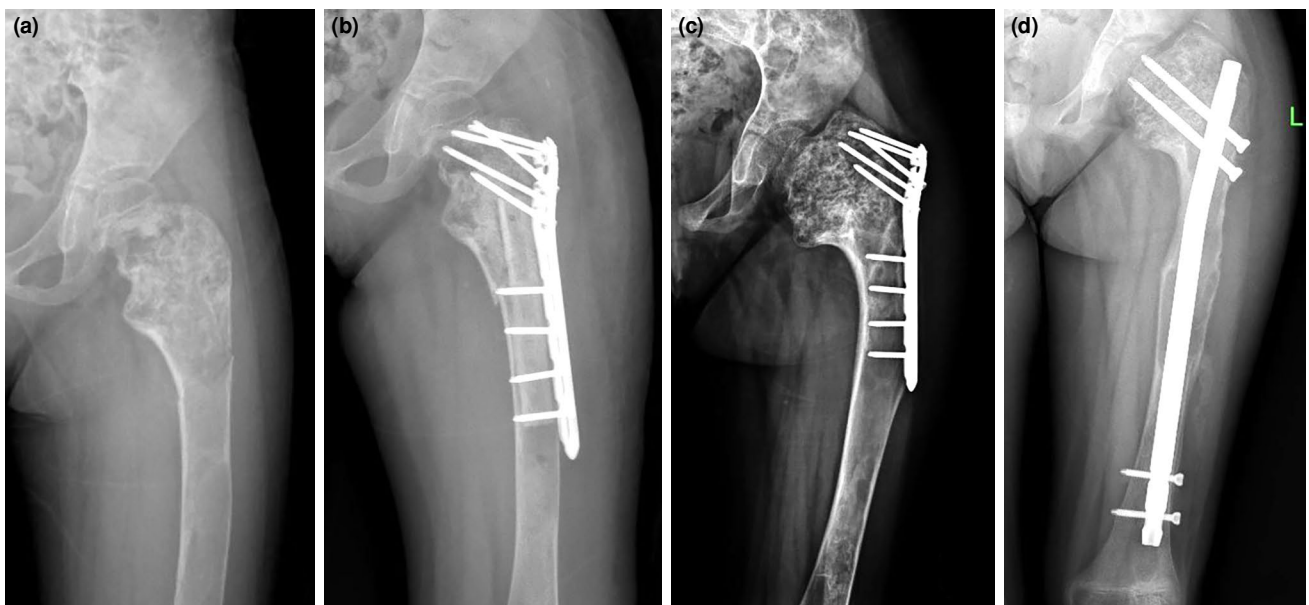

FIGURE 4. Fibrous dysplasia of the proximal femur in a seven-year-old female. **(a)** Preoperative radiograph shows the typical "ground-glass" appearance. **(b)** Curettage, non-vascularized fibular autografting, and internal fixation with a locking plate were performed. **(c)** Shepherd's crook deformity developed in the second postoperative year. **(d)** Corrective osteotomy, curettage, and intramedullary nailing were performed.

TABLE IV
Locations and treatment methods

	Surgical treatment		Nonoperative treatment		
	n	%	n	%	Total
Femur	45	52.3	41	47.7	86
Proximal	41	54.7	34	45.3	75
Diaphysis	2	28.6	5	71.4	7
Distal	2	50.0	2	50.0	4
Tibia	20	37.7	33	62.3	53
Proximal	4	28.6	10	71.4	14
Diaphysis	16	42.1	22	57.9	38
Distal	0	0	1	100	1
Humerus	4	16.7	20	83.3	24
Proximal	2	18.2	9	81.8	11
Diaphysis	2	20.0	8	80.0	10
Distal	0	0	3	100	3
Pelvis	6	30.0	14	70.0	20
Ilium	5	38.5	8	61.5	13
Ischium	0	0	1	100	1
Pubis	0	0	2	100	2
Acetabulum	1	33.3	2	66.7	3
Sacrum	0	0	1	100	1
Radius	5	31.2	11	68.8	16
Proximal	4	57.1	3	42.9	7
Diaphysis	0	0	7	100	7
Distal	1	50.0	1	50.0	2
Fibula	1	14.3	6	85.7	7
Ulna	1	100	0	0	1
Others	1	12.5	7	87.5	8
Scapula	0	0	2	100	2
Maxilla	0	0	2	100	2
Mandible	0	0	2	100	2
Lumbar vertebra	0	0	1	100	1
Calcaneus	1	100	0	0	1
Total	83	38.6	132	61.4	215

final follow-up. Preoperative measurements showed a mean neck-shaft angle of 90.5° (range, 85 to 105°), which increased to 125.6° (range, 120 to 130°) following surgical correction. At a mean follow-up of 74 ± 47.3 months (range, 30-140 months), no marked loss of correction was observed in six patients, and the mean neck-shaft angle at the final follow-up was 119.2°. In three patients, a marked loss in the neck-shaft angle was observed. The mean postoperative neck-shaft angle decreased from 126.7° to 93.3° at the final follow-up. These three patients underwent secondary surgery, which involved a repeat valgus osteotomy (Table V).

Among the patients in our study, 75 proximal femoral lesions were identified, and shepherd's crook deformity was observed in nine of them (12.0%). Table VI presents the comparison of patients with and without shepherd's crook deformity among those with proximal femoral lesions. Shepherd's crook deformity was more frequently observed in polyostotic lesions than in monostotic lesions

TABLE V

Demographics and clinical characteristics of patients with shepherd's crook deformity and their surgical treatments

No.	Sex	Side	Type	Previous surgery and age at procedure	Age at Shepherd's crook deformity	Preop angle	Surgery	Postop angle	Union of osteotomy	Last follow-up: Duration & angle	Secondary surgery & postoperative neck-shaft angle
1	F	Right	M	Curettage+ allograft + locking plate fixation / 12 years old	19	90°	Curettage + valgus osteotomy + intramedullary nail	125°	Yes	140 months, 80°	Curettage + valgus osteotomy + locking plate fixation / 130°
2	M	Right	M	None	54	95°	Curettage + allograft + valgus osteotomy + locking plate fixation	125°	Yes	30 months, 120°	None
3	F	Right	M	None	15	105°	Curettage + cortical autograft + valgus osteotomy + locking plate fixation	130°	Yes	125 months, 105°	Curettage + valgus osteotomy + locking plate fixation / 135°
4	M	Left	P	Curettage + cortical autograft + locking plate fixation / 12 years old	14	85°	Curettage + cortical autograft + valgus osteotomy + locking plate fixation	125°	Yes	35 months, 95°	Valgus osteotomy + intramedullary nail / 120°
5	F	Left	P	None	28	90°	Valgus osteotomy + intramedullary nail	125°	Yes	45 months, 120°	None
6	F	Left	P	Curettage + cortical autograft + locking plate fixation / 7 years old	9	95°	Curettage + valgus osteotomy + intramedullary nail	125°	Yes	54 months, 120°	None
7	M	Right	P	Curettage+ allograft + locking plate fixation / 10 years old	15	80°	Multiple valgus osteotomies + curettage + intramedullary nail	125°	Yes	25 months, 120°	None
8	M	Right	P	None	41	90°	Curettage + cortical autograft + valgus osteotomy + angled blade plate fixation	120°	Yes	136 months, 115°	None
9	F	Right	P	None	16	85°	Curettage + allograft + valgus osteotomy + angled blade plate fixation	130°	Yes	76 months, 120°	None

F, female, M, male, M, monostotic; P, polyostotic; Preop, preoperative; Postop, postoperative.

TABLE VI
Comparison of patients with and without shepherd's crook deformity in proximal femoral lesions

	Shepherd's crook deformity						
	Present (n = 9)			Absent (n = 66)			
	n	%	Mean ± SD	n	%	Mean ± SD	*p*
Age at initial diagnosis			17.6 ± 9.3			24.1 ± 15.3	0.281*
Sex							0.730**
Male	4	10.3		35	89.7		
Female	5	13.9		31	86.1		
Type							**0.026****
Monostotic	3	5.9		48	94.1		
Polyostotic	6	25.0		18	75.0		
Mc Cune-Albright syndrome							**0.013****
Present	2	100.0		0	0		
Absent	7	9.6		66	90.4		

SD, standard deviation; This table includes data from 75 lesions located in the proximal femur; * Mann-Whitney U test for numeric values and the ** Fisher exact test for categorical values were used to assess the potential differences between groups. Differences were considered statistically significant for *p* value < 0.05.

(25.0% *vs.* 5.9%, respectively; $p = 0.026$). Among the two patients with MAS, shepherd's crook deformity was observed in both, whereas the incidence was 9.6% among patients without MAS ($p = 0.013$). The mean age at the time of initial diagnosis was 17.6 ± 9.3 years in patients with shepherd's crook deformity and 24.1 ± 15.3 years in those without the deformity. Although the mean age at initial diagnosis was lower in the shepherd's crook deformity group, this difference was not statistically significant ($p = 0.281$). There was also no statistically significant difference in sex distribution between the groups ($p = 0.730$) (Table VI).

Among the 83 patients who underwent surgical treatment, only two (2.4%) developed postoperative superficial surgical site infections. Debridement was performed in both cases. In one patient, recurrence of shepherd's crook deformity resulted in intramedullary nail breakage at 140 months postoperatively. Revision surgery, including valgus osteotomy and fixation with a locking plate was performed. A total of seven implant failures were observed due to progression of the deformity.

DISCUSSION

In the present study, we evaluated the rate of proximal femoral involvement and the incidence of shepherd's crook deformity in patients with FD and identified possible factors associated with the development of shepherd's crook deformity. Our study results suggest that shepherd's crook deformity is more common in patients with polyostotic lesions and McCune-Albright syndrome. Although valgus osteotomy can achieve radiographic improvement, recurrence of the deformity and the need for multiple surgeries are not uncommon. Treatment should be tailored to each patient, taking into account factors such as age, lesion location and size, biological characteristics, and the presence of significant structural deformity or fracture.

Fibrous dysplasia is a benign bone lesion characterized by cortical thinning and the replacement of normal bone marrow with fibro-osseous tissue.[4,8-13,20,21] The proximal femur is the most commonly affected site in FD. Involvement of this region often leads to progressive deformities caused by persistent mechanical stress and repeated occult pathological fractures. These deformities include coxa vara, increased femoral neck anteversion, lateral bowing, and rotational changes of the femoral shaft, ultimately leading to the characteristic shepherd's crook deformity.[1,2,4,8,15] It is one of the most severe and undesirable clinical manifestations of FD, and its treatment is technically challenging.[2,4,15,18,19] In their review of FD, DiCaprio and Enneking[4] reported that diffuse polyostotic involvement of large weight-bearing bones led to bowing deformities that could progressively worsen with age and skeletal growth. Ippolito et al.[22] examined coronal plane femoral deformities and investigated predictors of deformity progression with growth in a study involving 27 children with

polyostotic FD. They identified younger age at the time of diagnosis and the presence of MAS as the most significant clinical predictors of progressive femoral deformity.[22] Consistent with the literature, shepherd's crook deformity in our series was more commonly associated with polyostotic disease and McCune-Albright syndrome (MAS).

McCune-Albright syndrome is a rare genetic disease classically characterized by a triad of polyostotic FD, precocious puberty, and café-au-lait skin macules. Over time, the clinical spectrum has expanded to include various endocrine abnormalities, such as hyperthyroidism, growth hormone excess, and Cushing's syndrome.[4,23,24] The clinical management of MAS should be individualized and implemented through a multidisciplinary team approach.[3,4,10-12,23,24] Evaluation by an endocrinologist is recommended for all young patients diagnosed with polyostotic FD to assess for endocrine dysfunction and initiate early treatment of associated abnormalities.[3,4,12,24] Endocrine disorders are typically managed with medical therapy.[23-25] However, skeletal lesions in patients with MAS tend to be larger, more persistent, and associated with higher complication rates than those in patients without the syndrome.[4] Therefore, Stanton et al.[3] recommended performing a technetium-99m labeled methylene diphosphonate (99Tc-MDP) bone scan during the initial evaluation of children with FD to determine the presence and extent of polyostotic disease, as supported by the international consensus statement. They also emphasized the importance of endocrinology consultation and the early initiation of treatment. In our case series, imaging revealed that 33 patients had polyostotic involvement, who were subsequently referred to the endocrinology department for further evaluation. Two of these patients were diagnosed with MAS based on the presence of hyperfunctioning endocrine disorders and subsequently received medical treatment. Both patients developed shepherd's crook deformity and underwent surgery for deformity correction.

In the management of FD, surgery may be performed to eradicate symptomatic lesions, correct deformities, and prevent or treat pathological fractures.[3,4,8,13] Most lesions are asymptomatic and can be managed with clinical observation.[4,10,21] However, there is no consensus regarding which lesions are suitable for observation alone and which may require orthopedic surgery. Treatment should be individualized. The location, size, and type of the lesion (monostotic or polyostotic), the severity of pain, and the patient's age are key factors influencing the choice of the most appropriate treatment method.[3,4,13]

Monostotic and polyostotic lesions are known to exhibit different biological behaviors. Monostotic lesions are typically asymptomatic and tend to remain active only until skeletal maturity.[3,4,10,11,21,26] Non-surgical conservative management is usually sufficient for asymptomatic monostotic lesions. Regular clinical follow-up with serial radiographs is recommended, until the lesion becomes biologically inactive and mechanically insignificant.[3,4,10] In severe symptomatic cases where conservative treatment is insufficient, monostotic lesions can be managed with conventional orthopedic procedures, similar to the treatment of other benign bone tumors.[1,3,4,13] On the other hand, polyostotic lesions exhibit more rapid growth and may continue to progress even after skeletal maturity.[3,4,8,21,26] Management of polyostotic disease is often challenging. Due to the increased risk of fractures and deformities in this patient group, regular clinical follow-up twice a year is recommended, even for asymptomatic patients.[3] The primary goal of management is to prevent deformity or pathological fractures; once these develop, surgical intervention becomes necessary.[3,4,13] In line with the literature, most incidentally diagnosed and asymptomatic monostotic lesions in our case series were successfully managed with conservative treatment. Clinical observation was sufficient for 132 lesions, whereas 83 lesions required surgical management. Among the surgically treated lesions, 28 underwent surgery due to deformity or pathological fracture, while 54 underwent prophylactic surgery due to severe symptoms. The literature lacks definitive criteria for identifying patients at higher risk for pathological fracture or deformity, and the decision to perform prophylactic intervention is multifactorial. In our case series, most lesions requiring prophylactic surgery were located in the weight-bearing bones of the lower extremity and occupied more than 50% of the bone diameter, which likely contributed to the increased risk of fracture and deformity.

Various surgical techniques have been described in the literature, and recommendations have evolved over time. Traditionally, intralesional curettage and bone grafting were considered the primary treatment method.[1,2,4,8,26,27] Early graft resorption has been reported in studies

evaluating the use of cancellous bone grafts. Guille et al.[27] investigated the outcomes of curettage and autogenous cancellous bone grafting in a series of 22 patients with FD of the proximal femur and reported favorable clinical outcomes; however, radiographic evaluation demonstrated complete resorption of all autogenous cancellous bone grafts, and none of the lesions had been eradicated at the final follow-up (mean 15 years; range, 2 to 41). Due to the high rate of resorption of autogenous cancellous bone grafts and the persistence of dysplastic lesions, cortical grafts began to be used as an alternative treatment option.[1,28] Enneking and Gearen[28] also reported favorable outcomes with the use of fibular strut allografts for stabilizing proximal femoral FD lesions. Cortical bone grafts are incorporated into host bone at a slow rate and, therefore, tend to be retained for a longer period.[4,28] In particular, cortical allografts undergo minimal internal remodeling, allowing a substantial portion of the graft to remain structurally intact over time. Consequently, FD is one of the few conditions in which allografts are biologically preferable to autogenous grafts.[4] Several studies have examined potential factors associated with the poor outcomes following bone grafting.[26,29] Stephenson et al.[29] reported unsatisfactory outcomes in patients under 18 years of age with lower extremity FD treated with grafting procedures and therefore advocated the use of internal fixation in this group. Leet et al.[26] evaluated patients with polyostotic FD who underwent bone grafting and found that 75% of the grafts had resorbed by the end of a mean follow-up period of 19.6 years. They reported that bone grafting had limited value in young patients with polyostotic FD.

In the light of the literature, we avoided the use of cancellous autografts in patients with FD who underwent curettage and grafting in our study. Instead, we preferred cortical autografts, allografts, or vascularized fibular grafts in these patients. The lesions treated with curettage and grafting without internal fixation were generally monostotic and located in the upper extremity. We used grafting to provide temporary augmentation for internal fixation in selected patients with polyostotic lesions and lesions located in the weight-bearing regions of the lower extremity. Additionally, in selected large lesions, cementing was employed to fill the defect and to provide local control or mechanical augmentation.

The treatment of shepherd's crook deformity is one of the major challenges in the management of FD.[1-4,11] The primary aim at managing FD involving the proximal femur is to prevent deformity. However, once deformity develops, appropriate correction is required.[2,11,15] Surgical treatment is recommended even in the absence of weight-bearing pain if the neck-shaft angle is less than 120 degrees.[3,30] Corrective osteotomy and internal fixation have been advocated in the literature to realign the deformity, restore normal gait mechanics, and relieve pain.[1-4,18] There is an ongoing debate in the literature regarding the optimal implant for internal fixation following surgical correction. Various studies have recommended the use of intramedullary nails, locking plates, or angled blade plates.[1-4,8,15,18,19] In our cohort, shepherd's crook deformity occurred in nine of 75 proximal femoral lesions. Four of these lesions underwent surgery prior to the development of the deformity. All nine patients were administered valgus osteotomy and internal fixation. Postoperatively, six patients maintained their neck-shaft angle, whereas three required additional surgery due to recurrence of the deformity. In the treatment of proximal femoral lesions, we believe that it is essential to inform both the patient and their parents about the potential development or recurrence of varus deformity, as well as the possibility of requiring multiple reconstructive surgical procedures in the following years.

Nonetheless, this study has several limitations. First, the retrospective design of the study and the heterogeneity of the patient population limited the ability to directly compare the treatment modalities. In addition, treatment decision-making was not standardized, as the choice of intervention varied according to patient age, lesion location, the biological behavior of the disease, and surgeon preference. Due to this heterogeneity, each subgroup ideally needs to be evaluated separately, which would require a much larger sample size. Although our study includes a large cohort on FD, multi-center, prospective studies with broader participation and standardized treatment algorithms are needed to compare different treatment options and to develop an approach for determining the timing and type of surgical intervention for specific patient groups. Finally, the extensive timespan of 35 years may have resulted in variations in diagnostic and management strategies, potentially influencing the comparison between early and late cases. However, to the best of our knowledge, this study is among the largest series on FD in a single tertiary center and provides valuable information about diagnosis and treatment.

In conclusion, treatment should be individualized, with the choice of intervention based on factors such as the patient's age, the lesion's location, size, biological nature, and the presence of any significant structural deformity or fracture. Most lesions can be managed with clinical observation, whereas surgery is indicated for of symptomatic lesions, prevention of pathological fractures, and correction of deformities. The risk of developing shepherd's crook deformity is higher in patients with polyostotic lesions and those with MAS. In patients with polyostotic lesions, the possibility of associated endocrinopathy should not be overlooked, and an endocrinology consultation should be obtained.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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