

# Surgical Treatment Results of Fibrous Dysplasia and Effects of Using Grafts

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## BACKGROUND/AIMS

Fibrous dysplasia (FD) is a skeletal system disorder characterized by replacement of normal bone and bone marrow with fibro-osseous tissue and proliferation of immature osteogenic cells. There is no consensus regarding the timing of orthopedic surgery, the choice of implants, and use of grafts owing to its variable clinical appearance and wider age range. This study aimed to contribute to the literature by performing a retrospective study involving patients with FD.

## MATERIAL and METHODS

This retrospective study included 41 patients with FD. Patients were evaluated for the first-visit and follow-up graphs. Patients were examined for the effects of intramedullary nailing and plate fixation. The use of grafts during surgery was noted and examined in terms of increase in deformity during the operation time and follow-up.

## RESULTS

Surgical intervention was performed on 45 bone lesions in 41 patients. Plate was used in 7 patients with pathological fracture, and intramedullary nail was used in 19 patients. There was a significant difference between the groups ( $P=0.011$ ). No deformity was detected in 16 of the 26 patients with pathological fractures. In 20 patients in whom grafts were used, cortical thickening was observed on the basis of the preoperative graphs. Graft survival rate was reported to be 83.3%.

## CONCLUSION

The presence of pathological fractures affects the choice of implants by surgeons. Cortical thinning is the most important parameter than deformity in terms of the risk assessment of pathological fractures for surgical planning. Allograft survival rate is high for 4 years, on average.

**Keywords:** Fibrous Dysplasia of Bone, pathological fracture, allograft, monostotic fibrous dysplasia, polyostotic fibrous dysplasia

## INTRODUCTION

Fibrous dysplasia (FD) is a skeletal disorder characterized by replacement of normal bone and bone marrow with fibro-osseous tissue and proliferation of immature osteogenic cells, leading to significant pain in patients (1). FD can clinically affect one or multiple bones. Attention should be given to pain, deformity development, and pathological fracture formation in the patients followed. Surgical procedures can be used for biopsy, correction of deformity, prevention of pathological fractures, or elimination of symptomatic lesions (2, 3). Cases detected because of pain or coincidentally should be followed in terms of both pathological fracture and deformity. Surgical procedures in patients without deformity are more successful.

FD is most often located in the proximal femur. Diseased bones have low resistance; therefore, long bones can easily develop bending and pathological fractures (4). Varus deformity, also called Shepherd's crook deformity, develops owing to localization of the lesion in the proximal femur and the bone structure deterioration. It is important to detect the varus deformity of the proximal femur early and correct it with valgus osteotomy. In polyostotic form, it can cause scoliosis in the spine. There is no consensus on surgical interventions in FD. Factors such as patient's age, presence of deformity, and localization and size of the lesions affect the choice of treatment of surgeons. Plates and intramedullary

fixation implants can be used in the surgery for FD. In the literature, it is argued that intramedullary implants are superior to plates in terms of implant selection (5-7). There is no consensus on the use of grafts in the surgical treatment of FD. A good graft must have osteoinductive and osteoconductive properties and have mechanical support when the bone cortex becomes thinner, such as in FD. Long-term graft resorption rates are high, especially in young patients for treatment using grafts alone. Mechanical implant support and stable fixation should be the priority of surgical treatment (8). If a bone graft using is planned, cortical grafts should be preferred instead of spongy bone because of their physical superiority (3). The use of bone grafts alone, including both allograft and autograft, has limited value in the treatment of FD lesions. This study aimed to investigate the effectiveness of surgical treatment strategy and graft use in FD.

## MATERIALS and METHODS

This study included 41 patients with a pathological diagnosis of FD. Ethical approval was obtained from the hospital ethics committee (approval number and date: 237/16/07/2020) for this study. Surgical consent was obtained from patients before any surgical procedure. All the procedures were in accordance with the ethical principles of Helsinki Declaration.

Patient data included age, sex, type of FD, surgical techniques, bone graft use, cortical thickening, and time to union. Incomplete patient data, patients who were not operated on, and patients who were operated for any reason in an external center were excluded from the study. The inclusion criteria included patients diagnosed with FD and those who were operated on. A total of 63 patients were operated in our hospital, and 41 patients who met the inclusion criteria were included in the study.

Patient's first-visit graphs and follow-up graphs were examined. They were grouped according to the number of bone lesions; monostotic FD (with a single bone lesion) and polyostotic FD (with more than one bone lesions). Follow-up patients admitted with pain and diagnosed with FD and patients with a pathological fracture without an initial diagnosis of FD were identified. Angulations that impair joint movements in any plane were accepted as deformities. Surgical indication in patients included pathological fractures, severe pain, and deformities that disrupt the movements. Patients were compared for the effects of using intramedullary nailing and plates, regardless of the use of grafts. During the surgery, radiography and surgery notes were checked for the use of grafts. The proper-

ties of the graft used were recorded, and in these cases, graft survival, cortical thickening, surgery timing, and recovery times were compared between patients in whom a graft was not used. Patients who had union in the pathological fracture area and who did not increase deformity during follow-up were considered as healed.

All patients were operated under general or regional anesthesia, in supine position, accompanied by scopy, according to the anatomical region and general medical condition. The patients were operated by 3 surgical teams. Curettage was applied by opening a window in the bone area held in all patients. Patients for whom grafts usage was planned, curettage was performed through the opened bone window, fibular strut allografts were placed, and then plate or intramedullary nailing was applied. Only surgical curettage and grafting were performed in patients in whom location and size of the lesion did not cause mechanical instability. Patients with mechanical instability were treated with plate or intramedullary nailing, depending on the location of the lesion and the preference of the surgical team. On the basis of the bone area involvement, some patients were treated with only curettage and grafts. Patients who had first-visit graphs, follow-up graphs, postoperative early graphs, and 1st, 3<sup>rd</sup>, 6<sup>th</sup> months; 1st and 2<sup>nd</sup> years control graphs were included in the study. Patients with incomplete files and follow-up graphs were excluded from the study.

## Statistical Analysis

The effectiveness of monostotic and polyostotic FD were analyzed and compared statistically using Student's *t*-test and Mann-Whitney U test. Patients were divided into 2 groups on the basis of the first-arrival complaints—complaints such as chronic pain or pathological fracture. Patients were also divided into groups on the basis of whether grafts were used and whether the implants used were plates or intramedullary nailing. Chi-square test and Fischer's exact test were used to find if the groups had any significant differences with respect to bone union and deformity increases in the patients' final control graphs. All data analyses were carried out using IBM SPSS software version 23 (IBM SPSS Corp.; Armonk, NY, USA).

## RESULTS

Of the 41 patients, 21 were women and 20 were men, with an average age of 17.4 years (5–38 years). The average follow-up period was 51.3 months (24–86 months). In total, 30 (73.6%) patients had monostotic FD and 11 (26.4%) had polyostotic FD. It was determined that 23 patients with monostotic FD had only femur involvement, 4 had only humerus involvement, and 3 had only tibia involvement. The localization of the lesions was examined in patients with polyostotic FD. Lesions in femur and tibia in 6 patients; both femur and tibia in 3 patients; and humerus, femur, and tibia in 2 patients were detected. When all the patients were examined, 57 bone areas were involved in 41 patients in total, including humerus, femur, and tibia. Surgical intervention was performed in 45 bone areas of these patients. Curettage and grafting were used in 3 surgeries, plate-screw system in 18 surgeries, and intramedullary fixation in 24 surgeries. Allograft was used in 23 patients (56.1%) (Table I).

### Main Points:

- Cortical thinning was found to be a more valuable follow-up parameter in terms of pathological fracture risk.
- Intramedullary fixation was found to give more successful results.
- It was found that the use of grafts led to cortical thickening in the lesion area.
- The patients with allografts had a mean follow-up time of 46.7 ( $\pm 19.7$ ) months, and the graft survival rate was 83.3%.

Surgical intervention was performed in 26 (57.8%) of the patients after pathological fracture. In total, 20 patients presented with primary pathological fracture. Six patients with pathological fractures were followed up with conservative treatment. Plate was used in 7 patients with pathological fracture, and intramedullary nail implants were used in 19 pa-

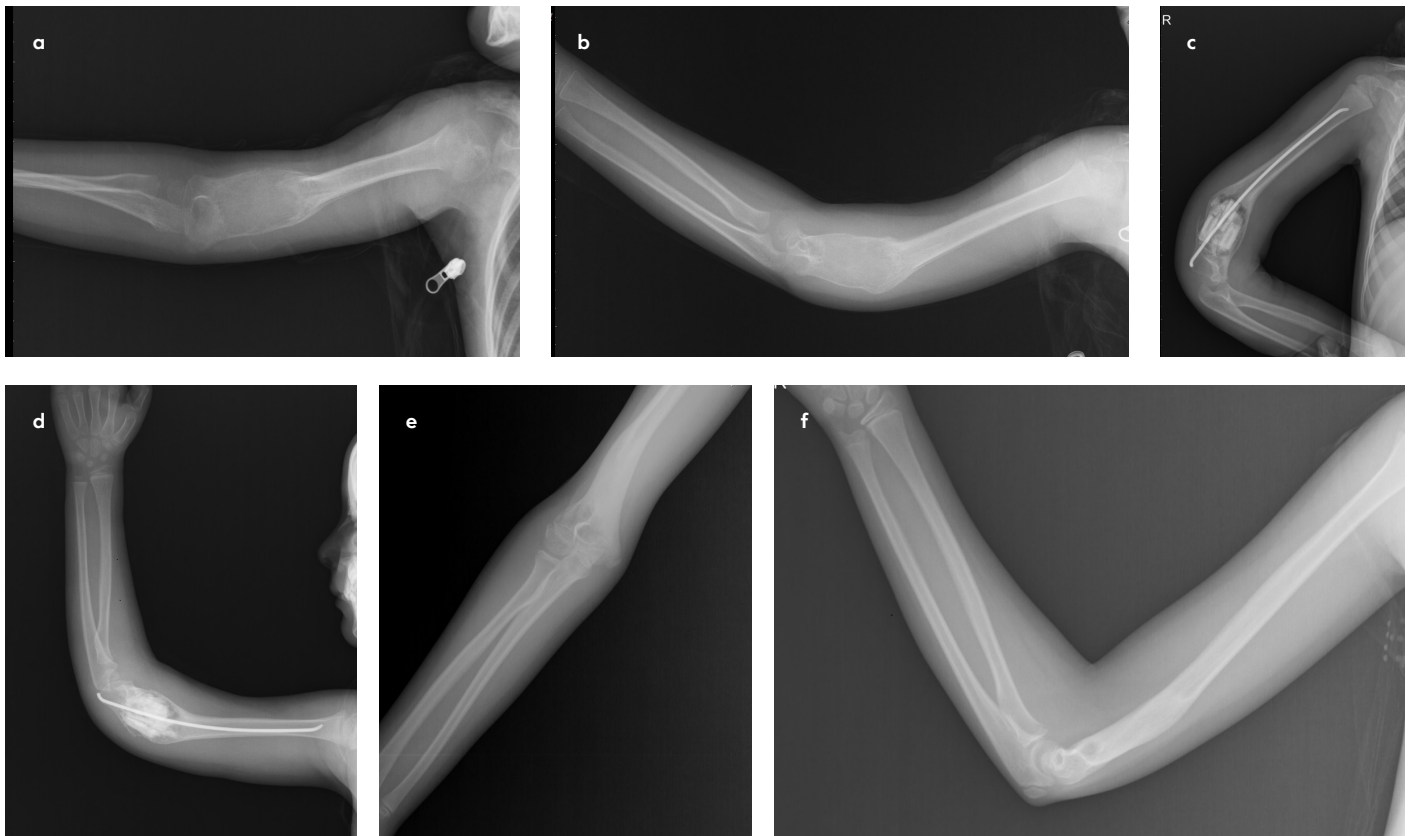
tients. There was a significant difference between the groups ( $p=0.11$ ). Grafts were used in only 14 patients with pathological fractures. There was no significant difference between the groups ( $p=0.522$ ). Although deformity was detected in 10 patients with pathological fractures, 16 patients had pathological fractures without any deformity. There was no significant difference between the groups ( $p=0.179$ ). The mean union time was 4 months (3–6 months) in patients operated for pathological fractures.

Deformity was found in 14 patients (31.1%); plate fixation was used in 6 patients, and intramedullary fixation was applied in 8 patients. There was no significant difference in the choice of fixation in these patients ( $p=0.627$ ). Allograft was used in 8 patients with deformity. There was no significant difference between the groups in terms of graft use ( $p=0.298$ ). Two of the patients were found to have Shepherd's crook deformity. The patients were treated with valgus osteotomy.

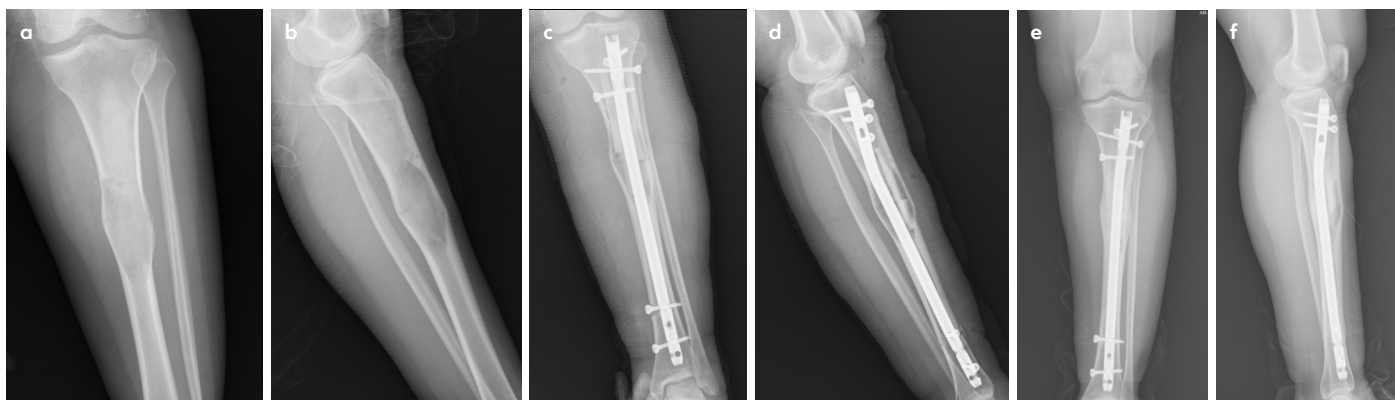
In 20 of the 24 surgical procedures using allografts, cortical thickening was detected according to the preoperative graphs (Figure 1). There was a significant difference in cortical thickening between the group with allograft and the group without allograft ( $p=0.000$ ). The mean follow-up period of patients using allograft was 46.7 ( $\pm 19.7$ ) months. Allograft survival rate was found to be 83.3%. It was found that there was cortical thickening in patients using allograft, regardless of patient age and remodeling capacity (Figure 2).

TABLE I. Demographic Data	
Demographics	Data
Gender	21 F/20 M
Type of FD	Monostotic FD 30/41 Polyostotic FD 11/41
Lesion localization	Isolated femur 23/41 Isolated humerus 4/41 Isolated tibia 3/41 Femur and tibia 9/41 Femur, tibia, and humerus 2/41
Number of surgeries	45
Type of surgery	Curettage and grafting: 3/45 Plate-screw fixation: 18/45 Intramedullary nailing: 24/45
Pathologic fracture	26/45 (first clinical diagnosis in 20 patients)
Use of allograft	24/45
Deformity	14/45

FD: fibrous dysplasia.



**Figure 1. a-f.** A 9-year-old girl with a right distal humeral fracture resulting from a simple fall. (a, b) Preoperative X-rays; (c, d) Postoperative X-rays; (e, f) X-rays 56 months after surgery



**Figure 2. a-f.** A 20-year-old girl with severe pain; left tibial lesion. (a, b) Preoperative X-rays; (c, d) Postoperative X-rays; (e, f) X-rays 60 months after surgery

## DISCUSSION

FD is a tumor-like lesion of bone and constitutes 5% of the benign bone lesions. FD has been associated with an activating mutation in the gene that encodes the alpha subunit of the stimulating G protein on the 20th chromosome. FD is able to hold all bones in the body. Its involvement in a single region is called monostotic FD, and its involvement in more than one region is called polyostotic FD. The polyostotic type is less common and can occur as part of McCune–Albright syndrome. FD, café-au-lait spots, and endocrine anomalies, such as puberty precoc, may accompany McCune–Albright syndrome. Polyostotic FD tends to occur mostly on one side of the body (2). Most lesions are monostotic, asymptomatic, detected coincidentally, and can be followed up by clinical observation.

It shows similar distribution among genders. There is no consensus on surgical procedures owing to a wide age range and variable clinical presentation. Patients can appear with severe pain or pathological fracture. In some patients, it can be detected coincidentally on radiographs taken for another reason. In the patients included in this study, the monostotic FD rate was 73.6%. Femur involvement was detected in 34 of the 41 patients (82.9%). Although the long bones of the lower extremity are typically involved with FD lesions, other bones may also be involved. FD lesions are most commonly observed in the femur and tibia. It can also be observed in the skull, ribs, and more rarely, in the pelvis (3, 5). In a study in which 173 patients were examined for the extracranial located skull and scalp pathologies, only 1 patient was found to have FD located in the skull bones (9).

Surgical intervention is performed in case of deformity increase or pathological fracture on the follow-up of patients. In surgical intervention, it should be aimed to obtain a painless and sufficiently stable bone. Surgical interventions include curettage, curettage and grafting, and implant fixation of bone with or without allografts. If more than two-thirds of the bone is involved with the lesion, curettage alone may not be sufficient because the bone is likely to be unstable (10). One of the most important points that we should emphasize in our study is that it was observed that only 10 of the 26 patients with pathological fractures developed deformity and subsequently pathological fractures occurred. In other words, pathological fracture may occur before deformity develops. Most patients with pathological fractures developed a fracture without deformity. The authors would like to emphasize that cortical thinning may be a more important sign in terms of patho-

logical fracture. In this study, intramedullary fixation was selected in 19 of the 26 patients who developed pathological fractures by 3 different surgical teams. There was a significant difference compared with the use of plates ( $p=0.011$ ). There are supporting studies in the literature that report that intramedullary fixation gives more successful results than plate-screw systems. In a study performed by Demiralp et al. (11), prophylactic intramedullary nails were applied to patients with monostotic FD without the use of allografts, and successful results were obtained.

No significant difference was found between both the implant selection and the use of allografts among 14 patients with deformity. Shepherd's crook deformity, which is a progressive varus deformity especially in the proximal femur, is the most common deformity. Patients with FD with involvement of the proximal femur develop post-stress recurrent occult fractures in the proximal femur. Over time, varus, anteversion, and anterior bowing occur, resulting in Shepherd's crook deformity. Patients come with antalgic walking and limb length discrepancy. Supporting evidence in the literature state that early treatment is more successful in patients with progressive deformity. In its treatment, it can be corrected by valgus osteotomy (3, 12, 13). Shepherd's crook deformity was detected in 2 of our patients. The patients were treated with valgus osteotomy.

The use of allografts is one of the controversial issues regarding surgical treatment of FD. In the study by Lindner et al. (14), high recurrence rates were observed in patients using iliac crest autograft, and it was emphasized that better results were obtained from fibrous cortical allografts. There are publications defending that a high rate of resorption develops in long-term follow-ups, especially when used at a young age. In this study, the mean follow-up time of 24 patients where allografts were used was 46.7 ( $\pm 19.7$ ) months, and graft survival rate was 83.3%. Fibular strut allograft was used in 22 patients and chips allograft in 2 patients. Despite the disadvantages of allograft, such as risk of immune rejection, disease transmission potential, and development of infection, it has osteoconductive properties but has limited osteoinductive potential (15). Leet et al. (8) found that graft survival rate was less in young patients with FD who were followed for an average of 19.6 years. In the average half of the patients, grafts were observed to be completely resorbed. In this study, the high graft survival may be due to the short follow-up period. In this study, 51.3 months (24–86 months) of follow-up was a limitation in terms of graft survival.

Another point that should be emphasized in patients with graft is the cortical thickening detected around the lesion. Although no cortical thickening was detected in any of the patients without graft in this study, significant cortical thickening was observed in the group using the graft ( $p < .001$ ). Kuznetsov et al. (16) observed that stem cells carrying mutant genes in the skeletal system decreased in time with apoptosis in young patients. This explains the progress in lesions with age and the inverse correlation with graft resorption. In polyostotic and young patients, the use of grafts is more unsuccessful than older patients with monostotic FD, and therefore, it will be more appropriate to decide for graft use on the basis of the age of the patient and the characteristics of the lesions. The limitations of the study are the wide age range of the patients, the presence of involvement in different bones, and the examination of different FD types. Long-term follow-up is required for allograft survival in larger patient series.

In conclusion, the presence of pathological fractures affects the choice of implants by surgeons. Cortical thinning is most important than deformity in terms of the risk assessment of pathological fracture for surgical planning. Intramedullary fixation in patients with FD gives more successful results. The use of grafts should be decided based on the age of the patient and localization of the lesions. Although the average survival of fibular cortical strut allografts has decreased over the years, cortical thickening in the lesion area is one of its important advantages.

**Ethics Committee Approval:** Ethics committee approval was received for this study from the ethics committee of Dicle University Medical Faculty (approval number and date: 237/16/07/2020).

**Informed Consent:** Written informed consent was obtained from patients who participated in this study.

**Peer-review:** Externally peer-reviewed.

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