

# Bilateral Tibial Fibrous Dysplasia in a Pediatric Patient treated with Intramedullary Nailing

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**Fibrous dysplasia is a benign developmental disorder of bone in which fibrous connective tissue containing abnormal bone with irregular trabeculae replaces normal cancellous bone. It may affect 1 (monostotic) or multiple bones (polyostotic). Polyostotic disease is the less common of the 2, occurring in only 20 to 25% of fibrous dysplasia patients and tending to affect those who are younger than 10 years of age; patients having this form tend to experience bone enlargement beyond normal skeletal maturation, which can cause pain, progressive damage, and increased risk of pathological fracture. There are limited reports of bilateral involvement in fibrous dysplasia, with said involvement presenting most frequently in early childhood. We describe the rare case of an adolescent presenting with bilateral fibrous dysplasia in the tibial diaphysis (in the exact same location) that was successfully treated with bilateral intramedullary nailing of both tibias. [P R Health Sci J 2018;37:58-61]**

*Key words: Fibrous dysplasia, Polyostotic, Intramedullary nailing*

**F**ibrous dysplasia (FD) is a disorder consisting of a developmental defect in osteoblast differentiation and bone maturation that typically develops during childhood and early adulthood, manifesting with the appearance of tumor-like lesions. FD can be classified into 2 types: monostotic, involving a single bone, and polyostotic, consisting of multiple lesions in different bones (1). Polyostotic cases tend to be rarer than monostotic ones and can present with gradual enlargements that may result in bone bowing, fracture, and deformity, until full growth is achieved (2–4). In cases of long bone involvement, FD commonly occurs in the diaphysis and metaphysis, with some rare cases involving the epiphysis of the bone.

There are limited reports of bilateral involvement in FD, with most such cases presenting in early childhood (5–7). We describe the rare case of an adolescent presenting with bilateral FD of the tibia diaphysis (in the exact same location) that was successfully treated with bilateral intramedullary nailing of both tibias.

## Case Report

This is the case of an active 15-year-old male basketball player who presented initially with the chief complaint of bilateral leg pain. The patient denied any prior history of trauma or fractures in the involved areas. A physical exam of both legs did not reveal any significant bowing or swelling. Bilateral imaging studies of both legs were ordered and demonstrated osteolytic bone lesions that appeared as masses in the diaphysis of both tibias at the same height (Figure 1). Based on imaging studies, the diagnosis of bilateral FD was made and the patient was referred to the Orthopedic service for further management.

A full body bone scan was ordered to determine whether there were any additional masses in the body; the results were negative for additional masses. A subsequent MRI confirmed that the masses were in the medulla of both tibias, with swelling of the surrounding tissue (Figure 2). There was no endocrine abnormality such as abnormal pigmentation or precocious puberty that would otherwise have suggested McCune–Albright syndrome.

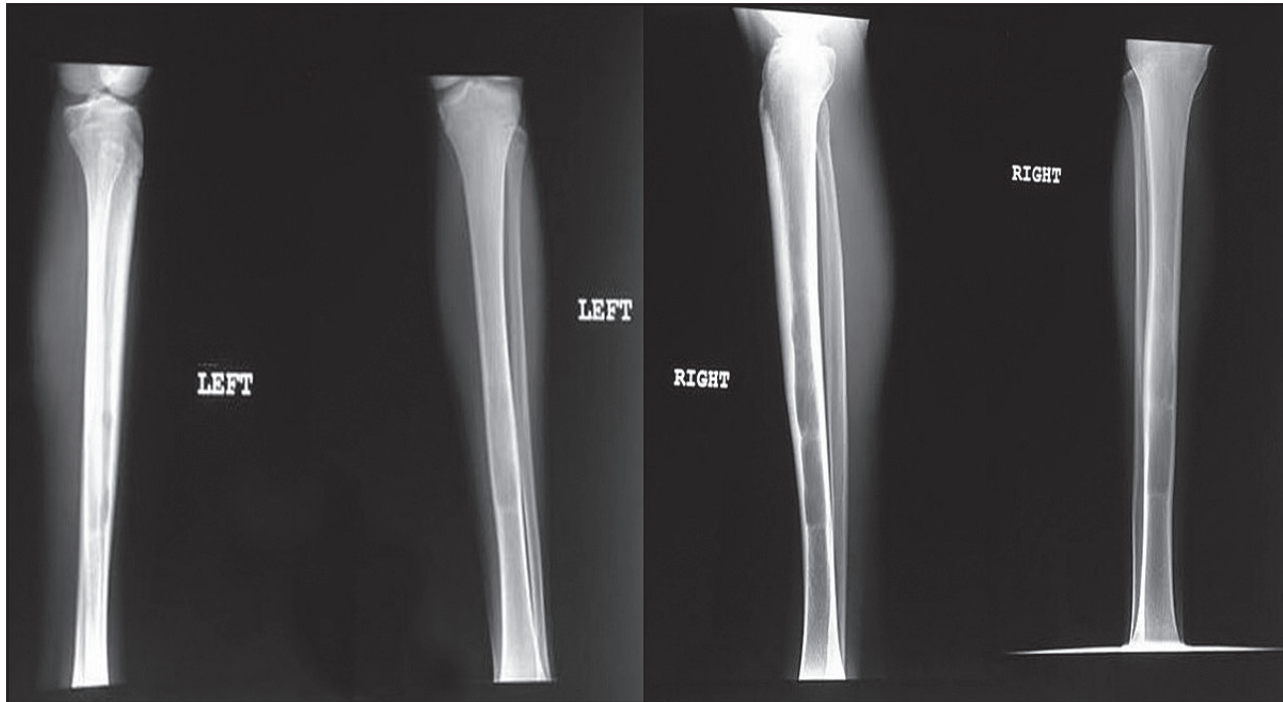
After analysis of the patient's history and plain radiographs, surgical intervention was recommended to prevent pathologic fracture of the tibia and/or bone bowing. Bilateral surgical reaming and placement of an intramedullary nail was performed in both tibias. Reaming of the intramedullary canal was performed until contact with the cortices was made and a thorough cleansing of the canal attained; an intramedullary sample was obtained for pathology analysis. Histologic analysis of the sample showed multiple fragments of brown-tan bony tissue on its gross appearance. Subsequent post-operative follow-up showed that the patient went on to heal properly. Tibia x-rays demonstrated exuberant cortical intramedullary bone formation, with the disappearance of the scalloped osteolytic lesions on the inner wall of the

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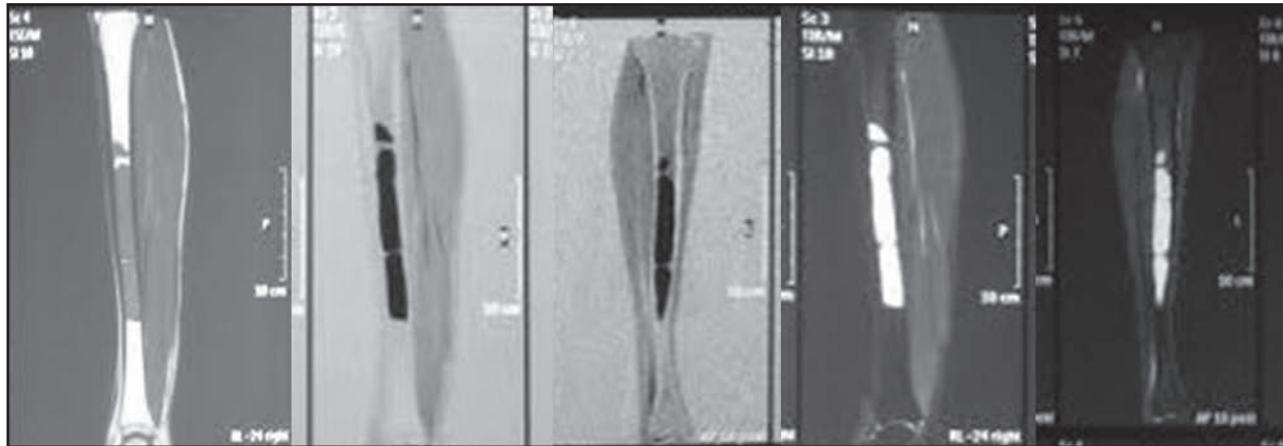
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**Figure 1.** Anteroposterior and lateral views of both tibias. Radiographs show scalloped osteolytic bone lesions in the intramedullary canals of both tibial diaphyses.



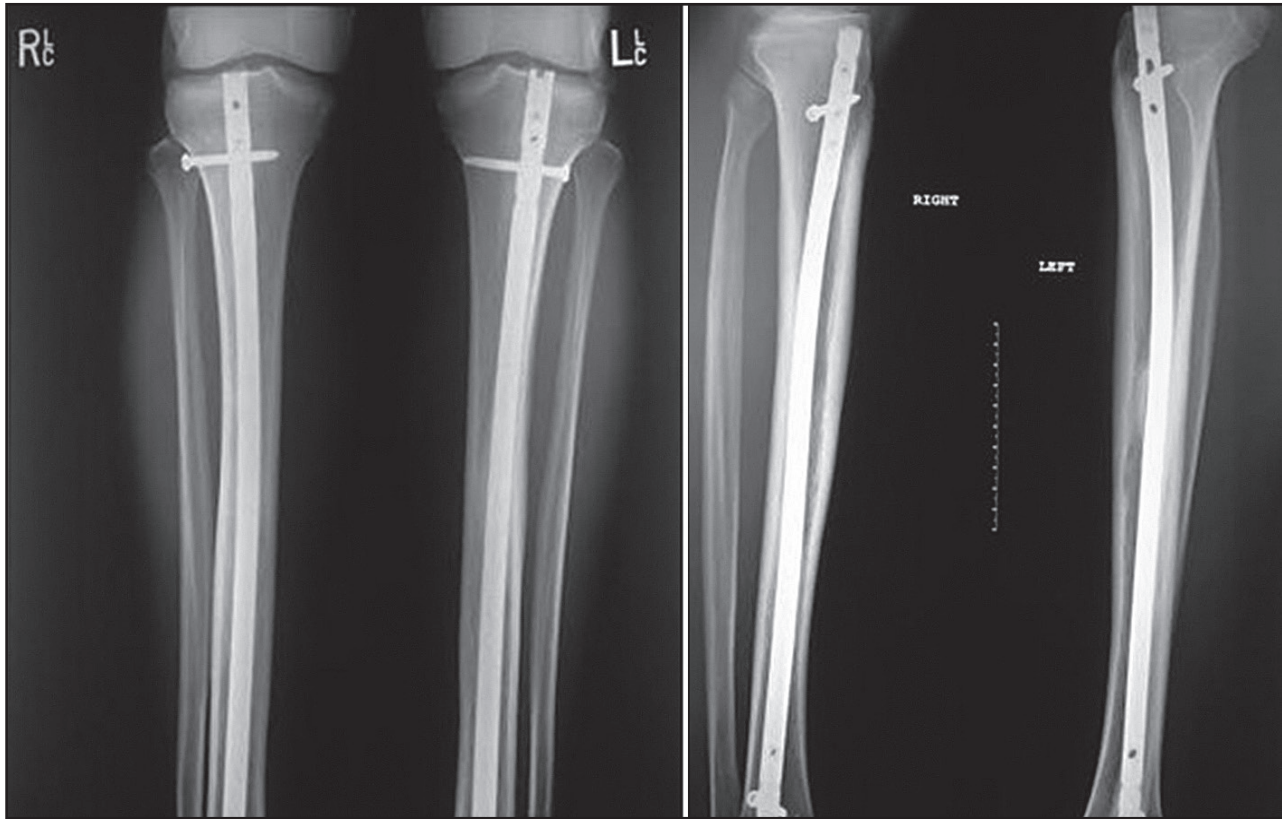
**Figure 2.** Magnetic resonance imaging (MRI) coronal views of both tibias. T1-weighted images show low intensity intramedullary lesions in both tibias. T2-weighted images show high intensity intramedullary lesions in both tibias. These radiographic findings are common in fibrous dysplasia.

tibial cortex (Figure 3). Clinically, the patient’s symptoms disappeared, giving him the opportunity to return to contact sports without limitation.

### Discussion

Fibrous dysplasia is considered a relatively common and well-known skeletal disorder. It consists of a single or multiple foci of fibrous tissue composed of irregular bone trabeculae that can lead to weakness or pathological fracture of the affected bones. The etiology of the disorder is a mutation in

the GNAS1 gene at chromosome 20q13.2-13.3, which encodes for the alpha-subunit of a stimulatory G protein (1, 3). This mutation occurs after the fertilization of somatic cells; therefore, it is not inherited. The disorder results in the production of abnormal matrix, bone trabeculae, and collagen orientation by undifferentiated mesenchymal cells (8). Monostotic FD is the most common presentation of fibrous dysplasia; it consists of a single bone lesion that usually arrests after bone maturation and is found mostly in the femur or ribs. On the other hand, polyostotic fibrous dysplasia is far less common, happening mostly in children under 10 years of age and tending to grow



**Figure 3.** Anteroposterior and lateral views of both tibias at follow-up. The proper placement of both intramedullary nails is seen, and there is no evidence of osteolytic lesions in either of the tibias.

beyond skeletal maturation, which can cause progressive damage and a greater tendency to suffer pathological fractures (4, 8). Plain radiograph findings of fibrous dysplasia are not specific for this disorder. However, lesions in long bones generally appear in the metaphysis as an intramedullary expansion with a thinning of the bone cortex. FD lesions are typically described as having a “ground glass” appearance but may vary from a radiolucent to a sclerotic appearance, depending on the degree of calcification and the development of the lesion. Based on the radiographs, the differential diagnosis may include intramedullary osteosarcoma, osteofibrous dysplasia, adamantinoma, and solitary bone cysts.

The proper management of polyostotic fibrous dysplasia varies, depending on the onset, severity, and location of the disorder. Since most polyostotic cases tend to become more severe, surgical management is recommended to prevent the possibility of considerable damage being done to the bone. Treatment options are closed fixation in the case of fractures, internal fixation, or curettage and bone grafting. The use of closed methods in the lower extremities has been associated with unsatisfactory results in skeletally immature patients (<18 years) because of recurrent pathologic fractures. In contrast, internal fixation has resulted in superior outcomes when managing FD in long bones (1, 4, 9–11).

Reports of bilateral FD are scarce in the literature, with only 8 having been reported in non-adolescent patients (5–7).

These cases all presented early in the patients’ lives and were successfully managed before they turned 10. Our report describes the case of an adolescent patient diagnosed with bilateral symmetric FD in the tibias, the first such presentation in the literature. The principles of FD management were applied, and intramedullary nails were placed in both tibias. Treatment outcomes were favorable, with the cessation of pain and full return to physical activity.

## Resumen

Displasia fibrosa es un desorden benigno del desarrollo de hueso, en donde el hueso normal es reemplazado por un tejido fibroso inmaduro y hueso trabecular inmaduro. Este desorden puede afectar un solo hueso (monostótico) o múltiples huesos (poliostótica). La forma poliostótica es rara, 20-25% de los pacientes, usualmente son pacientes de menores de 10 años y tiende a aumentar su tamaño pasado la maduración del hueso en donde puede causar dolor y una alta tendencia a fracturas. Los reportes en la literatura sobre displasia fibrosa son escasos, con la mayoría presentando a temprana edad. En este trabajo describimos el caso raro de un adolescente que presentó con displasia fibrosa bilateral de la diáfisis de la tibia (exactamente en el mismo lugar) que fue manejado exitosamente con clavos intramedulares en ambas tibias.

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