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CASE REPORT

Refracture of the tibia 6 years after bilateral tibia nail removal in a patient later diagnosed with osteopetrosis

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Joost H van Linge[1,2]
JHJM Gert Bessems[1]
P Koen Bos[1]

[1]Department of Orthopaedic Surgery, Erasmus MC, Rotterdam, the Netherlands; [2]Reinier de Grant Gasthuis, Ga Delft, the Netherlands

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Abstract: Osteopetrosis is a rare bone disease with a high fracture incidence and a risk for recurrent fractures. We describe a case report of an adult female with a refracture of her right tibia 6 years after routine bilateral tibia nail removal. The patient and 3 of her family members had a history of multiple (stress) fractures. Later we diagnosed autosomal dominant osteopetrosis in our patient and her family. In addition we give an overview of osteopetrosis and our considerations regarding routine removal of intramedullary implants in these patients.

Keywords: fracture, stress fracture, pathological fracture, intramedullary nailing

Introduction

Intramedullary nailing is widely accepted as standard treatment for closed, unstable tibial fracture, open tibial fractures, and tibial stress fractures.[1–3] There is no consensus, however, concerning absolute or relative indications for intramedullary nail removal after consolidation of the fracture. The decision to remove nails has largely been based on symptoms, routine treatment, or patient's preference.[4]

There is only one paper in literature describing serious peri- or intra-articular injuries following high velocity injuries in patients with a femoral nail in place.[5] But does this justify routine removal? Anterior knee pain may form an indication for tibial nail removal.[6] However, tibial nail removal for anterior knee pain may lead to disappointing results and can even provoke anterior knee pain in previous asymptomatic patients.[7,8]

There is a potential harm in routine removal of metal implants such as wound infections, deep venous thrombosis, excessive blood loss, nerve damage to the infrapatellar branch of the saphenous nerve, and refracturing.[8] Nail removal can be a difficult procedure due to bony overgrowth, broken screws, or unknown nails that were implanted elsewhere. To prevent further damage during extraction, nail retention has even been reported.[7,9,10]

We present a case of refracture of the tibia 6 years after removal of an intramedullary nail, originally implanted for a fractured tibia. The patient had a history of stress fractures and a family history of recurrent fractures. Although the increased risk for refracture in metabolic or hereditary brittle bone disease is well known, in the intramedullary nail literature, there is little attention for evaluation of bone diseases during decision making for nail extraction.

Case report

A 27-year-old woman was treated with an intramedullary nail for a fractured left tibia in an area of a stress fracture (Figure 1). Her right tibia was treated with a nail one year later for a symptomatic stress fracture (Figure 2). Because of her young age and

Correspondence: J H van Linge
Erasmus MC, Department of Orthopaedic Surgery,
Postbus 2040, 3000 CA Rotterdam, the Netherlands
Tel: +31-10 10 – 703 3537
Fax: +31-10 10 – 703 2001
Email j.vanlinge@erasmusmc.nl

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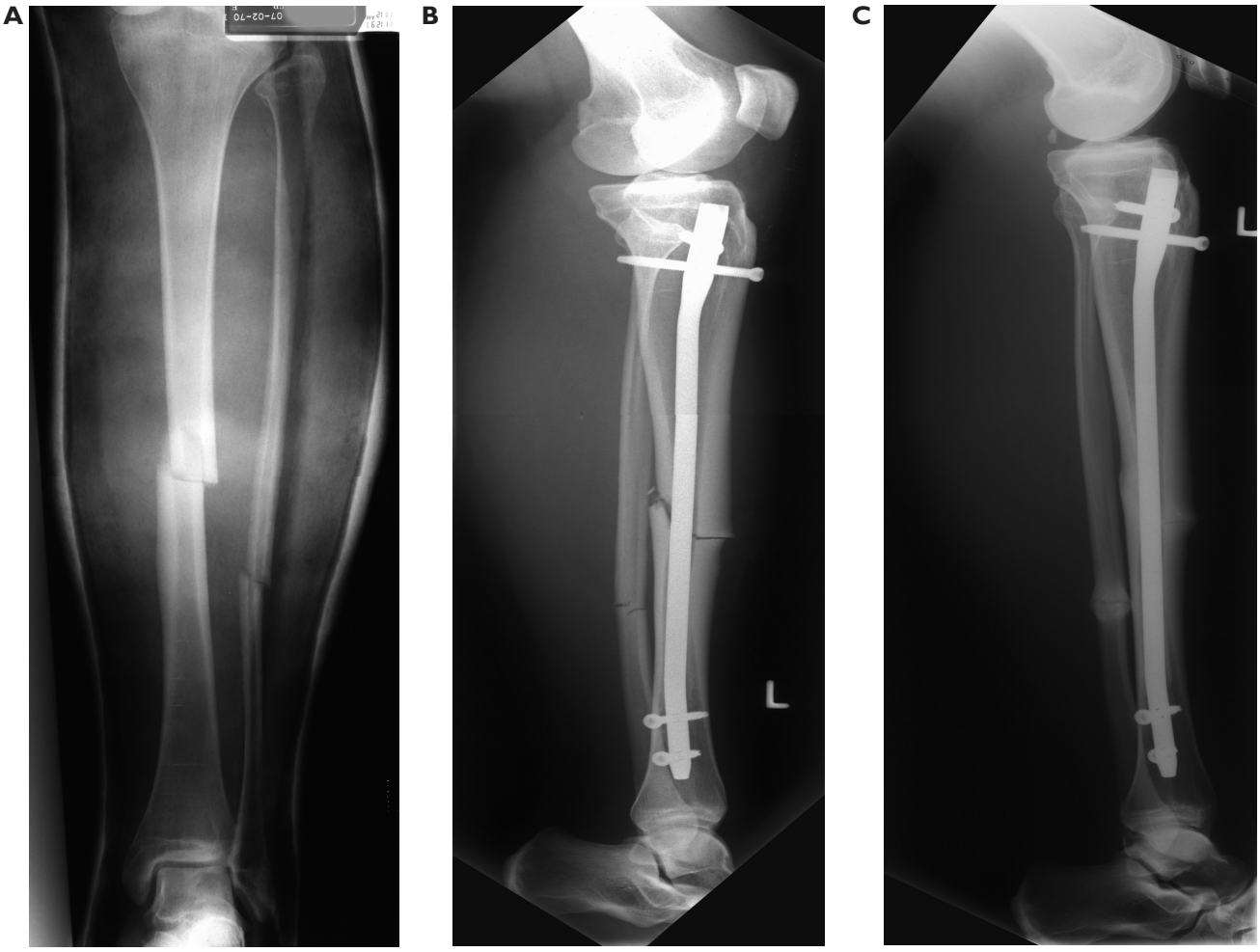

Figure 1 Fracture of left tibia and fibula at the site of a stress fracture 8 years prior to her presentation in 2005 **A**) before surgery, **B**) after intramedullary nailing, and **C**) 1 year after surgery showing tardy consolidation.

minor complaints, both implants were removed simultaneously after the fractures healed well.

Six years later, in 2005 at age 35 when she was 30 weeks pregnant, she was admitted following a minor fall in which she sustained a subtrochanteric femoral fracture on the left side and a refracture of her right tibia in the previous fractured area. The femoral fracture was stabilized with a long Gamma nail (Stryker, Mahwah, New Jersey, USA). This was a challenging procedure that took more than 3 hours because of complete obliteration of the femoral shaft. Because of her pregnancy and because we expected a demanding second procedure we decided to treat her tibia fracture conservatively with reposition and cast immobilization. Unfortunately, she developed a skin necrosis due to secondary angulation in the cast. 10 weeks later, after she gave birth to a son, the fracture of her right tibia was still not consolidated. We treated her fracture with a reamed intramedullary tibia nail (Figure 3). At the time of surgery we thought the infection healed but nevertheless it was complicated by a deep infection, probably due to the skin necrosis around the fracture site. This pre-tibial skin defect was treated by a fasciocutaneous flap according to Pontén. The fracture healed well but the nail had to be removed and the intramedullary infection was treated with intramedullary gentamicin beads. Because of the history of recurrent (stress) fractures and patient's preference to reduce the risk of future fractures, we decided to implant two reamed tibia nails. Recently, we treated her right femur with a long gamma nail because of a fracture after a minor fall (Figure 4).

Family history and counseling

With the fractures mentioned above and no significant trauma we decided to take a detailed history of the patient and her family. It appeared that she had suffered tibial stress fractures at age 22 and an insufficiency fracture of the foot at age 30. We also discovered that we treated her aunt some years ago because of multiple (stress) fractures. Despite the earlier efforts of the endocrinology and pathology department to

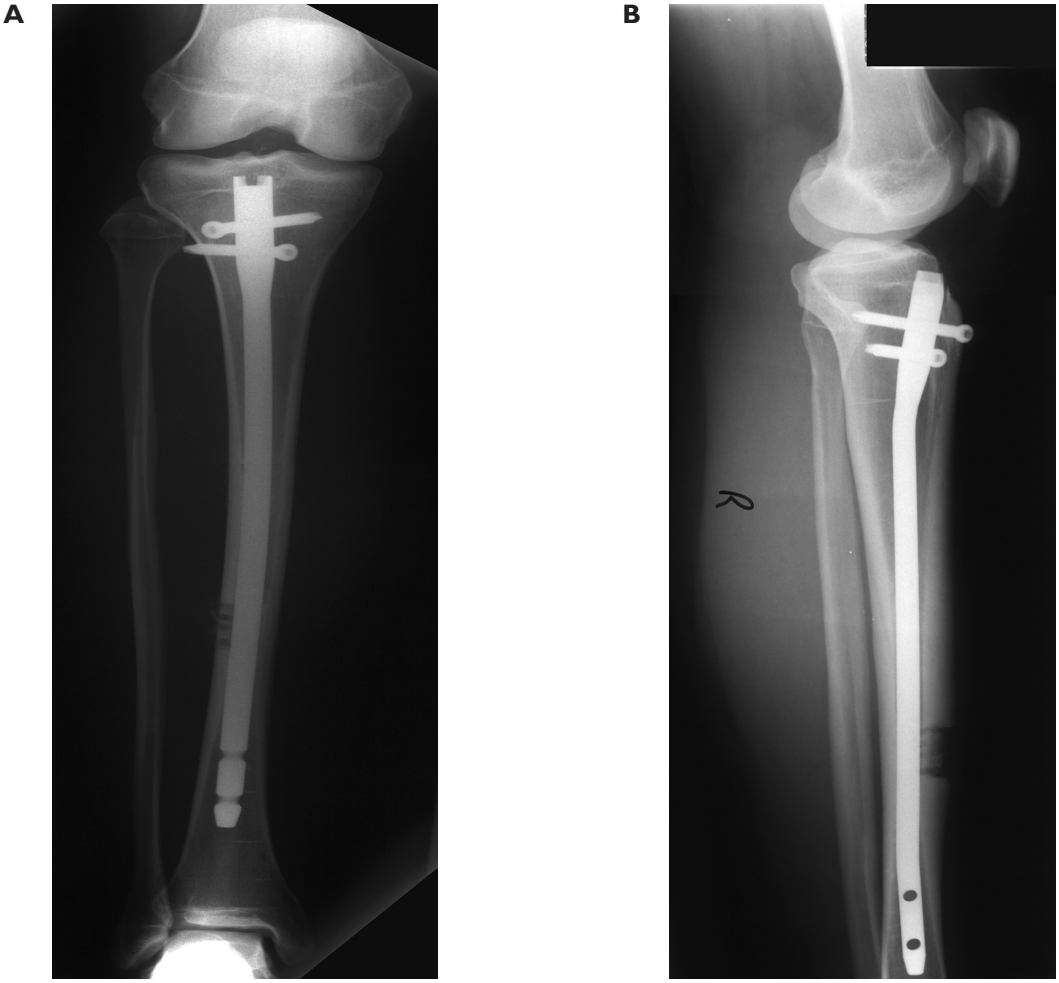

Figure 2 A) and **B**) Stress fracture of right tibia 1 year later after surgery. An intramedullary nail was placed and a biopsy was performed.

come to a diagnosis for her aunt, only an osteopetrosis-like disease was diagnosed mainly based on histology. The father and sister of our patient had multiple fractures as well. The patient's two brothers did not sustain any fractures.

Taking into account the patient's and family history with multiple fractures, we consulted the clinical genetics department. A detailed family history was taken, and additional X-rays were made to evaluate radiological signs. Medullary canal narrowing and subtle skull density was found, but no other typical radiological features were present. The Dutch Skeletal Dysplasia Group judged that there was enough evidence for diagnosing osteopetrosis with an autosomal dominant heritage pattern. Further evaluation did not reveal evidence for another cause of brittle bones (osteoporosis, endocrine pathology, or osteogenesis imperfecta).

Osteopetrosis

Osteopetrosis is a heterogeneous group of heritable conditions characterized by defective osteoclast resorbtion leading to hard and brittle bone.[11–14] Osteopetrosis is a rare bone disease (prevalence varies from 3.3 per million for the malignant form to 55 per million for the benign form); it is also known as Albers-Schönberg disease or marble bone disease.[11,12] The malfunction of the osteoclast's capacity to resorb bone leads to osteosclerosis and increased brittleness of the bones. Before identification of genes that affect the function of osteoclast, three types of osteopetrosis were classified based on clinical aspects: the first type is the infantile or 'malignant' osteopetrosis, an early autosomal recessive form characterized by presentation at a young age and severe symptoms. The obliterated medullary cavity leads to an inadequate hematopoietic capacity. The scull bones show serious abnormalities with cranial nerve dysfunction. These infants die at very young age. The second type is the intermediate autosomal recessive form which gives a higher fracture risk and mild hematopoietic disturbance and less cranial nerve dysfunction than in the malignant form. The third type is the 'benign' or late form. This type is autosomal dominant and the most common. There

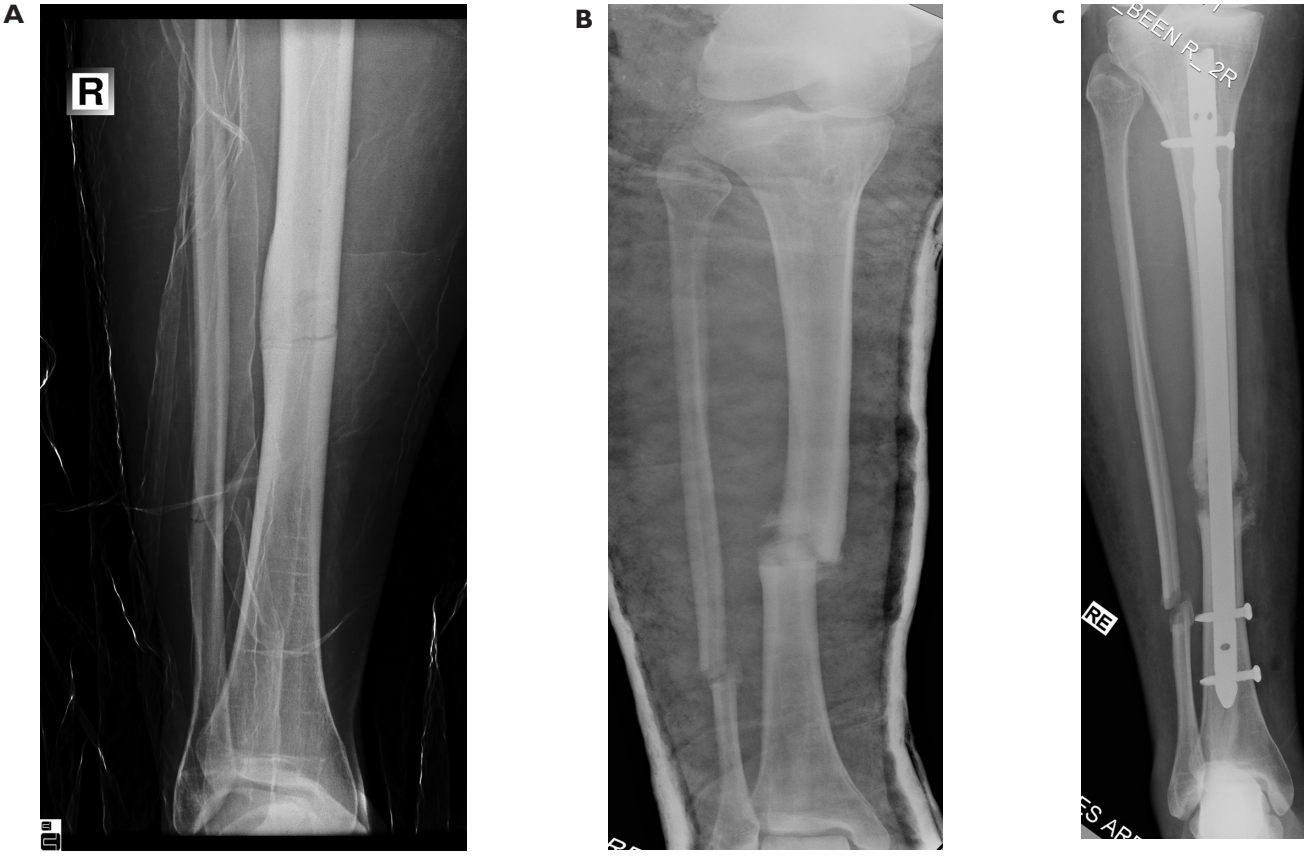

Figure 3 A) Right tibial refracture at presentation in 2005. **B**) Secondary dislocation in plaster cast 10 weeks later. **C**) After insertion of a reamed intramedullary nail.

are two subtypes: type I, with marked sclerosis of the vault but a normal fracture risk; and type II, with sclerosis of the base of the skull, rugger jersey vertebrae, and endobones in the pelvis. This subtype has a higher fracture risk.[15] Autosomal dominant osteopetrosis has a genetic basis; however, some individuals do not develop clinical features because there is a 75% penetrance.[12] Genetic abnormalities in the chloride channel gene, proton pump and carbonic anhydrase II were identified. They have different effects on the osteoclast physiology, but all cause a decreased bone resorbtion.

On histology, osteopetrosis bone shows lack of remodeling during bone development. Lack of osteoclastic bone resorbtion leads to sclerotic cancellous bone. Calcified cartilage and thickened trabeculae in diaphyseal bone explain the mechanical inferior bone quality and increased fracture risk. Clinically, patients present with frequent fractures (commonly stress fractures; life-long prevalence of fractures varies from 40%–66.6%), bone deformity, osteoarthritis, osteomyelitis, spondylolysis, and cranial nerve palsies (present in 20%–25%). Fracture healing has been described as prolonged.[12,16–18] Radiological findings include generalized sclerosis, bone-within-bone appearance (endobone), medullary canal narrowing, skull base thickening skull density, and vertebral endplate thickening (rugger jersey spine).

Surgical fracture treatment is complicated by extremely hard and brittle bone. Drilling and reaming during internal fixation or arthroplasty can be very challenging. Therefore, nonoperative treatment should be considered when reasonably possible. Although intramedullary fracture fixation of long bones may be a challenge due to medullary canal obliteration, it may be the treatment of choice because of the long-term strength and stability.

Discussion

As we discussed above, the need for routine removal of (intramedullary) osteosynthesis devices remains controversial. We have shown that removal of an intramedullary tibia nail, in a patient later diagnosed with autosomal dominant osteopetrosis, resulted in a refracture after 6 years. The risk of these refractures leaves us a difficult clinical decision whether to remove the intramedullary nails or not. Our patient was seriously invalidated during the last weeks of her pregnancy, and the subsequent fracture treatment resulted in prolonged hospital stay and multiple

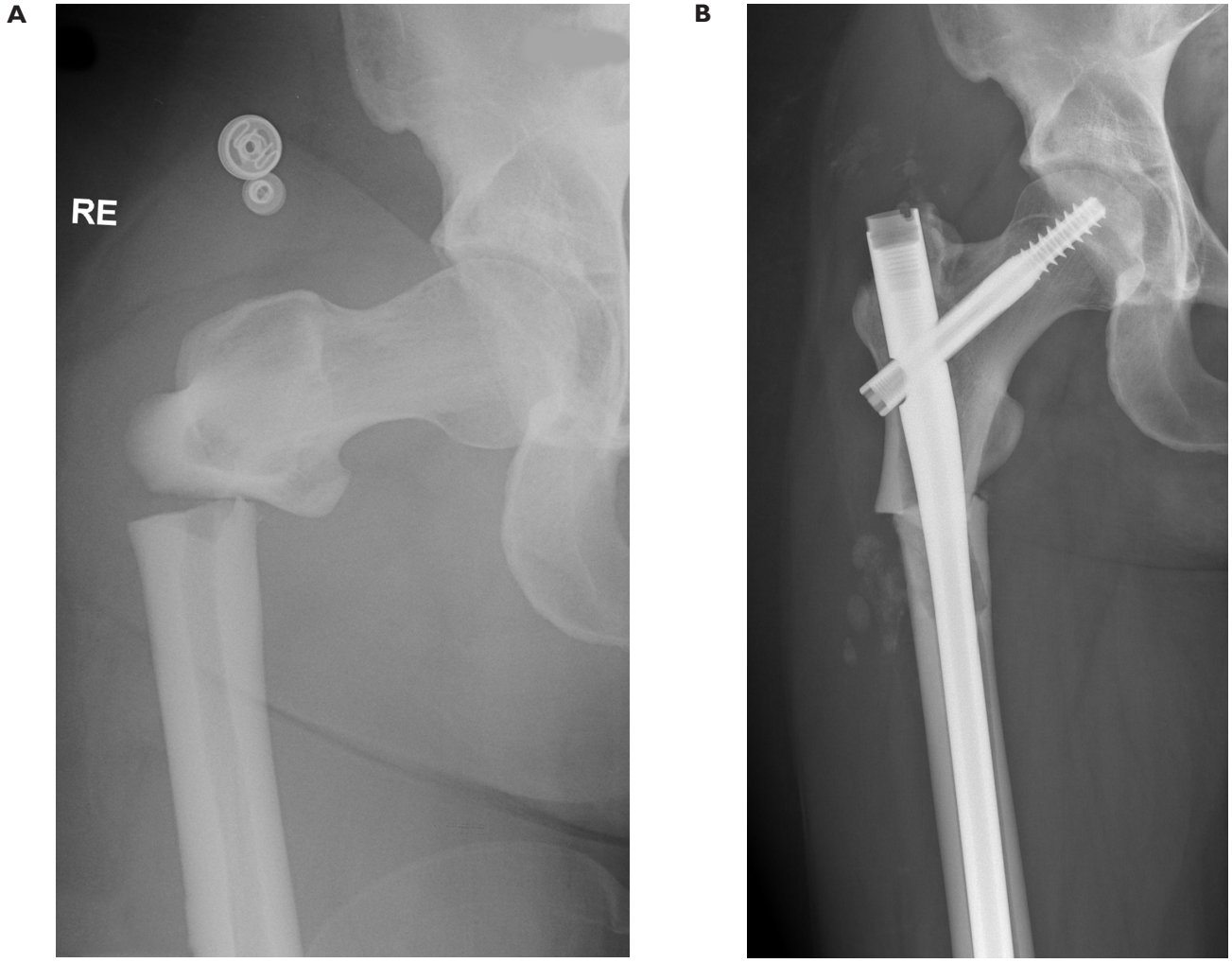

Figure 4 Right femoral fracture 3 years later: **A**) showing a narrow intramedullary canal and thickening of the cortex; and **B**) after surgery.

operations. In our department, the current attitude is that we don't routinely remove intramedullary nails. Relative indications for nail removal are pain related to hardware, young and active patients, and impending need for arthroplasty.

The increased risk for multiple fractures and refractures with brittle bone disease is well described. Until now, standard evaluation of possible presence of brittle bone disease (the risk for multiple fractures) was not emphasized in the intramedullary nail literature.

Conclusion

Based on this case history we advise to pay attention to the patient's and their family's fracture history before deciding to remove (intramedullary) implants. With a suspected metabolic or hereditary brittle bone disease we advise our patients to leave the device in situ to prevent recurrent fractures and subsequent morbidity.

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Disclosure

The authors report no conflicts of interest in this work.

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