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Pathologic Fractures in Children

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Fractures through bone tumors are often difficult to treat. We reviewed our combined experience with this problem in children, as well as the existing literature, to formulate management guidelines. For this study, prospective databases (1987 to 2002) from three referral centers were screened for pathologic fractures occurring under the age of 14 years. One hundred five patients presented with fracture through unicameral bone cyst, nonossifying fibroma, fibrous dysplasia, aneurysmal bone cyst and osteosarcoma. Seventeen patients were excluded. The most common primary locations were the proximal humerus and proximal femur. Pathologic fracture through nonossifying fibroma had the best outcome; union occurred with nonsurgical treatment in all cases. Unicameral bone cyst required surgical treatment to avoid persistence of the cyst and refracture. However fracture healing was predictable without surgical treatment. Proximal femoral lesions tended to heal in malunion if not fixed surgically. Aneurysmal bone cyst required surgical treatment for the lesion to heal and to allow the fracture to heal as well. Percutaneous sclerotherapy may be the treatment of choice for many of these lesions. Fibrous dysplasia allows fracture healing with nonoperative therapy. Progressive deformity requires followup and surgical correction. Malignant lesions presenting a pathologic fracture are best managed by initial nonoperative therapy during investigation and neoadjuvant therapy when possible, followed by definitive treatment.

A pathologic fracture is one that occurs through abnormal bone and occurs because the bone is weak and lacks its normal biomechanical properties. Such fractures result from intrinsic and extrinsic processes.[12] Intrinsic processes include metabolic bone diseases (osteopenia from osteogenesis imperfecta) or bone tumors that replace healthy bone. Extrinsic processes decrease the structural integrity of bone and arise from surgery (such as internal fixation that is inadequate or removed prematurely, or a bone defect from surgery such as biopsy, or en bloc resection of osteoid osteoma) or from external radiation therapy.

The healthy bone of a child has greater plasticity than that of the adult; and therefore, a greater loss of normal mineral content or architecture may be necessary to generate a fracture than in the same adult bone. Additionally, immature bones are surrounded by a thick sheath of periosteum, which can impart some stability to a fracture.[5] The failure of abnormal bone may manifest itself as repeated microfractures or as a complete fracture.

Most microfractures occur in trabecular bone, such as in the metaphysis of long bones or in the vertebral body. Many microfractures are undisplaced, heal well, and in fact, some are unrecognized. Successive microfractures can result in deformity, for example, the shepherd's crook deformity of the proximal femur in patients with fibrous dysplasia. Greater forces can produce a complete fracture, and symptoms are more intense and similar to a normal fracture. A complete fracture might occur whether a lesion is present or not, but a lesion or bone disease allows fracture to occur with less traumatic energy than in normal bone. Also, the location of the fracture tends to be related to the stress riser caused by the bone lesion (for example a fracture through a previously known fibrous cortical defect).

A variant of the pathologic fracture is the stress fracture.[32] A stress fracture occurs when an exceptional repetitive force is exerted on bone that fails to remodel. The bone may be healthy or weakened by some pathologic process. The march fracture of a metatarsal bone in the military recruit is the classic example of stress (or fatigue) fracture through healthy bone. Patients being treated with chemotherapy for malignancies are repeatedly bedridden and receive medications such as high-dose corticosteroids

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and other agents that cause osteopenia; therefore stress fractures are not uncommon in these patients when the improvement in their health allows them to increase their activity levels.

Determining the underlying diagnosis is critical for planning the correct treatment in children with pathologic fractures. The history and physical examination and radiological and laboratory studies are useful in determining the probable diagnosis. Most lesions of bone can be diagnosed correctly from their radiological appearance and their location in the bone.[46] The laboratory findings are important in metabolic bone disease and osteomyelitis and less important in primary bone tumors.[24]

The benign tumors of bone that are associated with pathologic fracture in childhood include unicameral bone cyst, nonossifying fibroma, fibrous dysplasia, and aneurysmal bone cyst. We focused on these entities because they are the most frequent. The problem of pathologic fracture complicating osteosarcoma is not as common but merits discussion because of the severe consequences associated with mismanagement.

In the light of the diagnosis, optimal fracture treatment can be selected, usually orthopedic immobilization or open reduction and internal fixation, with or without curettage and bone grafting. Surgical treatment must be individualized and based on factors such as fracture displacement, stability, anatomic location and expected response to medical treatment (eg, neoadjuvant chemotherapy in the case of osteosarcoma).

We wanted to determine which scenario would be adequately treated with orthopedic treatment, which conditions would definitely require some form of surgical treatment and what the optimal timing for surgery might be. The classification proposed by Dormans et al[14] in this context is useful because it groups the lesions according to their capacity for spontaneous fracture healing.

MATERIALS AND METHODS

In this retrospective study, prospective databases from three referral centers were screened for pathologic fractures in children. The centers were Hopital Maisonneuve-Rosemont in Montreal (Canada), Hospital Universitario del Valle in Cali (Colombia) and Fundación Hospital Alcorcón in Madrid (Spain). The approach to these patients was similar, based on the authors' training (all trained at the Orthopedic Oncology Unit of the Massachusetts General Hospital, Harvard Medical School). Identified patients then were reviewed to establish adequacy of followup. The age and gender of the child, the anatomic location of the tumor, treatment strategy, complications, and outcome were recorded and analyzed for each diagnosis. Data were collected with a uniform questionnaire (Appendix 1) and entered in a standard database (Microsoft Access, Microsoft Corporation, Bellevue, WA)

We included patients with a minimum of 24 months of followup. The study was limited to patients aged 14 years or less, presenting with pathologic fractures through unicameral bone cyst, nonossifying fibroma, fibrous dysplasia, aneurysmal bone cyst, and osteosarcoma. From 1987 to 2002, 105 cases of pathologic fracture were identified in patients younger than or equal to 14 years old and 88 cases were included for analysis (Fig 1). Seventeen patients were excluded because the diagnosis was unclear or because data were incomplete. The diagnosis was confirmed histologically only when clinically indicated and otherwise was based on clinical and radiological characteristics and clinical outcome (eg, histologic diagnosis is not required for treatment of pathologic fracture through unicameral bone cyst; aspiration of fluid at the time of percutaneous treatment of the lesion and treatment outcome offer diagnostic confirmation).

Nonsurgical treatment methods included various forms of immobilization (sling, cast, or traction), as well as various forms of analgesic medication. The use of nonsteroidal anti-inflammatory medication was infrequent.

Surgical methods of treatment for the fracture included closed reduction and pinning or open reduction and internal fixation, with or without grafting.

RESULTS

Eighty-eight cases were retained for analysis: 35 unicameral bone cysts, nine aneurysmal bone cysts, 14 fibrous dysplasia, 17 nonossifying fibromas, and 13 osteosarcomas. The median age of the boys (56.9%) and girls (43.1 %) at the time of the pathologic fracture was 10 years (range, 2–14 years; Fig 2). The patients were followed up for an average of 6.6 years (range, 24 months–11 years). The median age by diagnosis with pathologic fracture was 9 years for unicameral bone cyst and aneurysmal bone cyst, 11 years for nonossifying fibroma and fibrous dysplasia, and 13 years for osteosarcoma.

The most common primary tumor locations were the proximal humerus in 34 patients (39%), the proximal femur in 18 patients (20%), and distal femur in 10 patients (11%). Of 34 pathologic fractures occurring through uni-

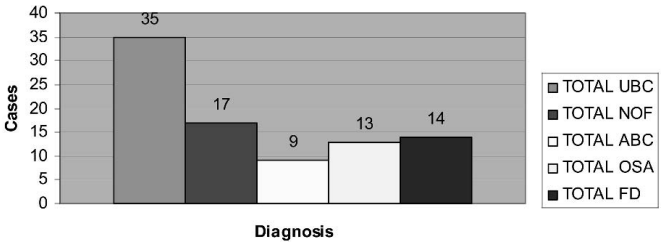


Fig 1. The figure shows the number of cases of each diagnosis. There were 35 cases of unicameral bone cyst (UBC), 17 cases of nonossifying fibroma (NOF), nine cases of aneurysmal bone cyst (ABC), 13 cases of osteosarcoma (OSA), and 14 cases of fibrous dysplasia (FD).

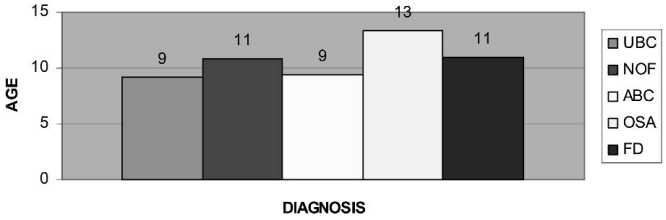


Fig 2. The figure shows the mean age of the patients with each diagnosis. UBC = unicameral bone cyst; NOF = nonossifying fibroma; ABC = aneurysmal bone cyst; OSA = osteosarcoma; FD = fibrous dysplasia.

cameral bone cysts, 24 were located in the humerus (71%), six in the proximal femur (19%), and one each in the tibia, pubic ramus, and radius. Although all the fractures healed, in 17 patients (50%) the unicameral bone cysts did not heal. Because the risk of fracture persisted, we thought further treatment (Scaglietti technique[41]) was justified in 13 of the 17 patients. For one patient with unicameral bone cyst in the pubic ramus and another in the proximal femur, intralesional resection and autologous or allogenic bone graft was performed.

All six of the fractures involving unicameral bone cyst of the proximal femur healed. Five were treated with hip spica casting. In the remaining patient, internal fixation was indicated to address displacement and instability, and was combined with curettage and bone grafting. Additional treatment was required for two patients. One of the six patients had a steroid injection and another had an osteotomy because of varus angulation.

In this series it is interesting to note that the rate of healing of the cyst after fracture was much higher when located in the proximal femur, as opposed to those lesions in the proximal humerus.

Of the nine cases of aneurysmal bone cysts, seven were treated with curettage and bone grafting. In two of these patients, the fracture did not heal. The other cases were one that involved the pubic ramus and was resected without reconstruction and one that was located in the humeral diaphysis and was associated with such widespread destruction of bone that wide resection and reconstruction with fibular autograft was done after biopsy confirmed the diagnosis.

The local recurrence rate in this series was two of nine patients (22%). Treatment consisted of repeat curettage and bone grafting.

Of the 17 cases of pathologic fracture through a nonossifying fibroma, nine were in the tibia, one was in the humerus, six were in the femur, and one was in the fibula. In all patients, the fracture healed and the nonossifying fibromas progressively resolved. Surgery was done in two patients for fracture displacement and in one a biopsy (followed by curettage) was indicated to confirm the diagnosis of nonossifying fibroma.

Fourteen fractures were associated with fibrous dysplasia, and 10 were located in the proximal femur. Seven patients were treated with hip spica casting or traction, and developed coxa vara for which corrective osteotomy was required in all seven. In the other three, open reduction and internal fixations were done as initial fracture treatments. None of the patients who underwent primary surgical treatment developed deformity. One patient with a lesion in the ulna was treated with open reduction and internal fixation. Three patients with lesions in the humerus were treated with a sling. All of these healed, but one developed a varus angulation that did not require further treatment.

Thirteen fractures occurred through osteosarcoma. Five involved the distal femur, two involved the femoral diaphysis, four involved the humerus and two involved the tibia. Nine of the 13 fractures healed with orthopedic immobilization; eight patients healed during neoadjuvant chemotherapy.

Five patients did not receive chemotherapy after the pathologic fracture. In two, the fractures were located in the distal femur with a large soft tissue mass and intractable pain, in addition to presenting widespread lung metastasis. Various attempts to achieve stability and pain control by nonoperative means failed, and we elected for hip disarticulation to achieve adequate resection margins and to relieve their pain. Two patients had wide resection and limb salvage reconstruction before adjuvant chemotherapy. Interestingly, the fifth patient had a fracture through an unrecognized lesion in the distal humerus that was treated by his primary care physician with a sling. The patient was later seen by an orthopedic surgeon for followup at which point the fracture was found to be healed, but an osteolytic lesion was diagnosed and biopsy revealed osteosarcoma. Neoadjuvant chemotherapy was initiated before definitive resection. (We did not report this patient with the cases showing healing using chemotherapy because he healed before initiation of chemotherapy.) All patients received chemotherapy that was in accordance with the standards of modern chemotherapy at the time of their treatment. At latest followup, six of the 13 patients (46%) had died of their disease.

DISCUSSION

When a child has a fracture after minimal trauma or when the radiographs suggest an abnormal process in the bone or an inappropriate site or pattern for the fracture, pathologic fracture should be suspected. Primary care givers and families need to be aware of the implications of pathologic fractures and how they affect treatment.

Frequently, pain is the earliest and most important symptom of the pathologic process in bone. Questioning the patient for painful symptoms before fracture may help establish the pattern of underlying disease. If the pain only was present with activity, this could suggest an impending fracture in bone weakened by an indolent process, whereas persistent and progressive pain and night pain are of considerable concern. In these scenarios, malignant tumor has to be ruled out. The past medical history should be reviewed for the occurrence of previous fractures, gastrointestinal or kidney disorders, or recent infections, to screen for metabolic or infectious bone disease. Physical examination, in addition to evaluation of the fracture, skin, joint and neurovascular status, should include assessment of possible limb deformity or limb-length inequality.

It is important to define the etiology of the fracture. For example, when a previous procedure (for example, decompression of an osteonecrotic femoral head,[35] open resection of the nidus of osteoid osteoma on a long bone) has left an otherwise healthy bone weakened by a residual defect, the fracture may be treated as any other fracture at that site in healthy bone. If, however, some primary bone abnormality is present, the treatment of the fracture may be of secondary importance to other considerations that may require evaluation in a multidisciplinary setting. Pathologic fractures that appear during or after treatment for malignancy (eg, radiation for Ewing's sarcoma) could be associated with local recurrence, progression of disease, or even a second malignancy.

After clinical radiological and laboratory evaluation, potentially benign aggressive or malignant processes should be biopsied. As amply described in several review articles on the subject of diagnostic evaluation of bone tumors,[24,31,32,37] the biopsy must be done through a small longitudinal easily excised incision, through a single compartment, with meticulous hemostasis. The orthopaedic surgeon and the bone radiologist should concur on location of the biopsy to select the most representative area. In particular it is important not to take the biopsy from callus, as this may lead to the erroneous diagnosis of osteosarcoma, or fail to diagnose another tumor.[45] An experienced musculoskeletal pathologist should be available for interpretation of the frozen section specimen at the time of biopsy to ensure that accurate and representative tissue is obtained. Cultures should be obtained at the time of biopsy. It is important to take precautions to prevent pathologic fracture, potentially caused by the lesion and by the stress riser related to the biopsy itself.

When diagnosis of a malignant tumor is made, staging is crucial to establish prognosis and to determine the correct treatment. A treatment plan then is devised taking into account the fracture and its location, as well as the underlying condition and its stage.[45]

The method of treatment depends on the diagnosis of the underlying lesion, its location, and the type of fracture.[9,31,37,45] If the tumor is benign and surgery is indicated for fracture stabilization, it is essential to select the type of resection (intralesional or marginal), the optimal filling method for the tumor defect, the reduction of fracture, an appropriate neutralization of forces across the bone, and the type of subsequent immobilization, if any. The filling material should be associated with minimal morbidity.

In light of the final diagnosis, optimal fracture treatment (simple immobilization, closed or open reduction with internal fixation), can be chosen. Optimal definitive treatment of malignant or possibly malignant tumors generally requires a multidisciplinary approach. Surgical treatment must be individualized and based on factors such as fracture displacement, stability, expected response to chemotherapy and radiation, and the perceived ability to achieve a wide resection that includes potential dissemination of the tumor by fracture hematoma.

Dormans et al[14] have proposed grouping the treatment of pathologic fractures. The four types of fracture treatments include: 1) no treatment—the fracture and lesion may heal spontaneously; 2) treating the fracture first and then the lesion; 3) treating the fracture and lesion simultaneously; and 4) treating only the underlying process—the fracture can heal with treatment of the underlying process.

Type 1

When the fracture occurs through a nonossifying fibroma, the fracture and the lesion usually will heal with nonsurgical treatment, and satisfactory results will be obtained unless the fracture is displaced and unstable (Fig 3). Almost all fractures associated with unicameral bone cysts healed; however, in 50% of our patients, the cyst persisted with a risk of refracture. According to the literature, only about 10% will heal completely after a fracture.[5,21] They should, therefore, probably be classified with Type 2.

Type 2

Some pathologic fractures will heal with a nonsurgical procedure, but require surgical treatment of the underlying lesion. This generally is true for aneurysmal bone cysts. However, if the fracture is displaced or unstable, particularly in a weightbearing bone (for example, the femur), open reduction and internal fixation often is required. It should also be noted that most cases of aneurysmal bone cyst are active and may progress during nonoperative treatment. If observation is elected, careful clinical and

Fig 3A–D. A pathologic fracture occurred in nonossifying fibroma, was treated with a cast, and a good result was obtained. Note of the small size of the lesion. (A) Pathologic fracture through nonossifying fibroma of the proximal metaphysis of the tibia in a 6-year-old boy is shown. (B) A lateral plain radiograph taken at the same time as Figure 3A is shown. (C) An AP radiograph after cast treatment shows the healed fracture and nonossifying fibroma 6 months after fracture. (D) A lateral radiograph taken at the same time as Figure 3C is shown.

radiographic follow up (4 to 6 weeks) of these lesions is mandatory to permit early recognition of disease progression and appropriate subsequent management. Some telangiectatic osteosarcomas may be mistaken for aneurysmal bone cyst. As stated above, unicameral bone cysts should probably be treated in this group, with immobilization being a reliable method to obtain fracture healing and some form of treatment (such as methylprednisolone injection) used to decrease the risk of refracture from persistence of the cyst.

Type 3

Some fractures complicating low-grade malignant or benign aggressive lesions are best treated by simultaneous treatment of the fracture and lesion. The appropriate surgical oncological treatment is followed by surgical recon-

struction or stabilization (eg, adamantinoma or other low grade sarcomas for which adjuvants are not indicated).

Type 4

In cases of pathologic fractures associated with high-grade malignant primary tumors, we recommend first treating the underlying process if effective medical treatment is available (eg, chemotherapy in osteosarcoma or Ewing's sarcoma). The fracture usually will heal with simple immobilization either with a cast, a brace, or with traction. Surgery, if indicated, follows neoadjuvant therapy in these patients. Definitive surgical treatment should proceed only if the orthopaedic oncologist, radiologist, and pathologist are certain of the diagnosis after careful analysis of the clinical, radiographic, and histologic features.

The anatomic location may dictate some aspects of treatment. For example, most of the lesions occurring in the upper extremity involve the proximal part of the humerus. A simple sling often will suffice as initial treatment for this site. In contrast, in the weightbearing bones of the lower extremities, particularly the proximal femur, surgical treatment will more often be required because of the relative difficulty in obtaining adequate immobilization, susceptibility for further fracture and the potential for deformation, as well as avascular necrosis.[20,35,36,49] Nevertheless, in malignant tumors, such as osteosarcoma, it is probably safer to immobilize the proximal femoral fracture using traction until the effect of chemotherapy allows mobilization with crutches and functional bracing. Fractures of the head and neck of the femur in healthy children are rare compared with such fractures in adults. Unlike in adults, most of these fractures result from severe high-velocity trauma. A minority of the proximal femoral fractures occurring in children are pathologic fractures and occur with trivial trauma (such as those in patients who have a unicameral bone cyst or fibrous dysplasia).[25] Accurate closed or open reduction and internal fixation usually are recommended for femoral neck fractures in healthy children as a means of minimizing the problems that were mentioned above.[8] Although the risk of avascular necrosis after femoral neck fractures is high in patients who have had high-velocity traumatic injuries, the risk is low after pathologic fracture.[26,43,44] This probably is because of the low energy of the trauma associated with pathologic fractures.[25]

Internal fixation of fractures through lesions of the proximal femur can be a challenge due to extensive loss of bone in the femoral neck. It is not uncommon to find residual healthy bone only in the epiphysis of the femoral head, which limits the efficacy of fracture implants.[43] Considering the important mechanical loads that characterize this region, it is often a better choice to revert to nonoperative methods of fracture treatment (such as traction or spica cast)[22] or to use "low-tech" methods such as those used by Wagner, using multiple K-wires into the epiphysis, fixed to the proximal femoral diaphysis by simple cerclage. (We were unable to find a reference for this technique apparently passed on by generations of Wagner trainees.)

Unicameral Bone Cyst

Little is known about the etiology and pathogenesis of unicameral bone cysts. A unicameral bone cyst may be characterized as a solitary cavity containing serous fluid originating in the metaphysis of growing children adjacent to the metaphyseal aspect of the growth plate. In children who are 3 to 5 years old, these cysts they typically increase with gradual growth of the diameter of the metaphysis, and during early adolescence the lesions tend to become less active. As the epiphysis grows away from the lesion, its location will become more diaphyseal. After skeletal maturation, the lesions slowly ossify and infrequently persist beyond the third decade of life.[5]

Although the cysts traditionally have been described as active if they are adjacent to the epiphyseal plate and latent if the lesion is greater than 0.5 cm from the physis,[34] this concept has not been validated by more recent observations.[39] That study defined a cyst as active if serial radiographic studies reveal an increase in length or width of the lesion exceeding 25%, if the patient had functional pain, if the patient had sustained multiple pathologic fractures without resolution of the cyst, or if the cyst was associated with cortical thinning (suggesting an impending fracture).

Seventy five percent of the patients with unicameral bone cysts present with a pathologic fracture[3,29]; before the fracture they usually are asymptomatic. The fractures can be expected to heal in a reasonable time frame, usually within 6 weeks (Fig 4), but the cyst most often persists.[36] Only about 10% will heal completely after a fracture[5,21]; however, our series show that the cyst heals in a higher percentage after fracture (50%), but still this is a problem. The displacement of the fractures associated with unicameral bone cysts usually is minimal and the overall treatment is rarely affected.

Simple closed reduction and adequate immobilization of the fracture are recommended for 4 to 6 weeks.[12,13] However, depending on the instability and displacement of fractures of the proximal femur, internal fixation is often indicated for this site.[30]

Specific treatment of the cyst is delayed until the fracture is well healed. Repeated fractures that commonly occur in an untreated cyst may well result in physeal injury as well.[10] Prolonged observation with the potential for

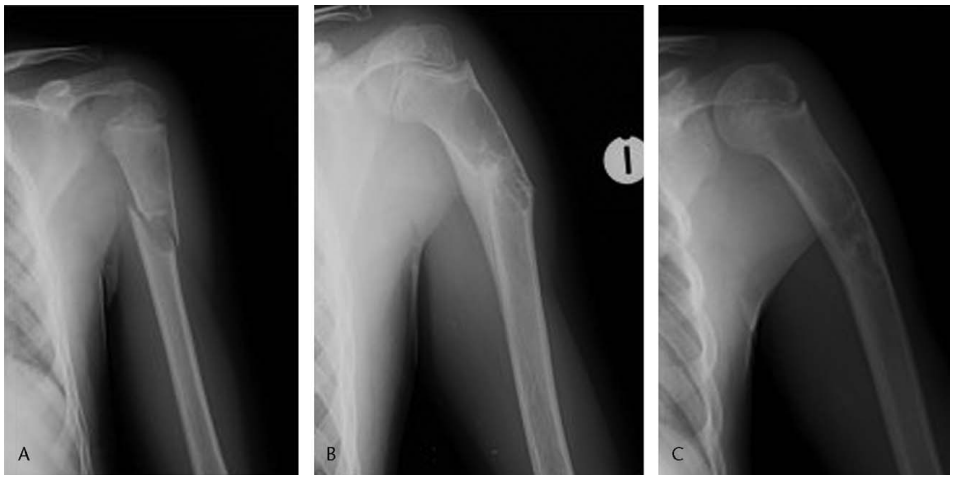


Fig 4A–C. These figures show a good outcome of the unicameral bone cyst and fracture of the proximal humerus treated with sling and cast in a 9-year-old girl who had a pathologic fracture. The fracture and unicameral bone cyst healed without a surgical procedure. (A) A pathologic fracture through well delimited osteolytic lesion compatible with unicameral bone cyst is shown. (B) The fracture healed in 3 months after treatment with immobilization. (C) The cyst healed 1.5 years after the fracture.

repeated fracture is stressful to patients and parents and adversely restricts children from normal childhood activities. It is therefore reasonable to advocate a 6-week to 12-week delay after fracture followed by definitive treatment of the cyst.[10]

The aim of treatment is to prevent a repeated fracture and the occasional physeal injury that may follow (Fig 5). Different authors[1,33] have attempted to quantify the risk of fracture using radiological measurements but to our knowledge these methods have not been validated. Many forms of treatment have been recommended for the unicameral bone cysts but discussion of this controversial subject exceeds the scope of this article.[1,13,28,34,40,41,42,44]

Nonossifying Fibromas

Nonossifying fibroma is a hamartomatous defect in the metaphyseal cortex of skeletally immature adolescents (5 to 15 years old). It is considered to be a developmental error. Once formed, the lesion gradually may expand and involve a substantial proportion of the metaphysis. As skeletal maturity approaches, spontaneous involution is usual, and the lesion will heal gradually.

Although only a minority of nonossifying fibromas is associated with pathologic fracture, some of the larger lesions will predispose to fracture. Previous reports suggest that absolute size parameters correlate directly with the risk of pathologic fracture.[2,16] The larger the lesion, the

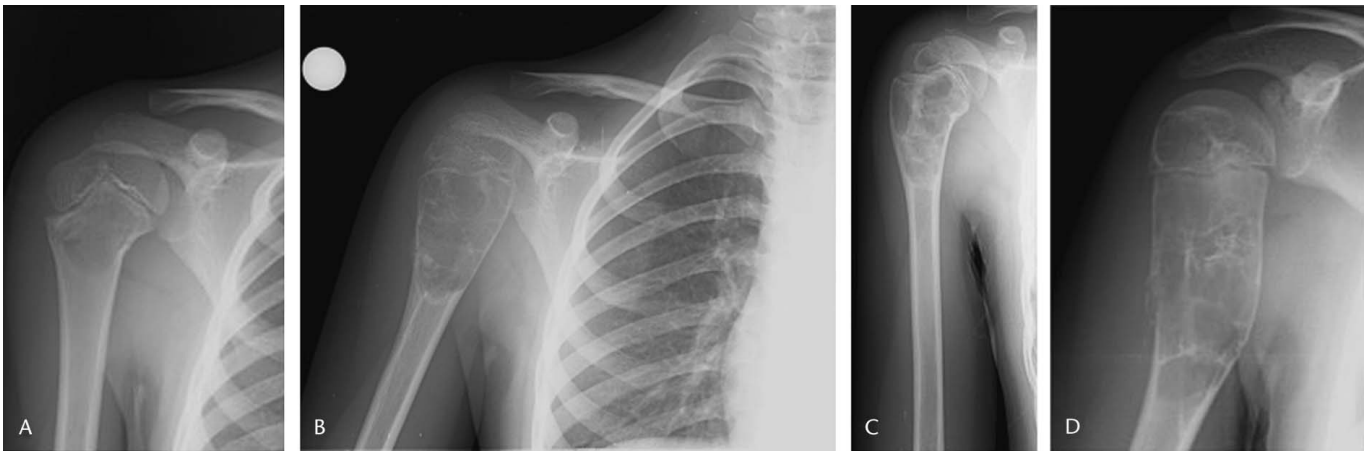


Fig 5A–D. These figures show a poor outcome of the unicameral bone cyst and fracture of the proximal humerus treated with sling and cast. Typical radiographs of unicameral bone cyst of the proximal humerus are shown. An 8-year-old boy had an incidental finding of unicameral bone cyst in 1999. He was observed for the next 3 years; the cyst increased in size and he had three pathologic fractures and epiphyseal involvement. (A) An AP radiograph of the proximal humerus in 1999 is shown. (B) The first pathologic fracture in 2000 is shown; the boy was treated with immobilization (sling). (C) An AP radiograph after treatment with sling shows healing of the fracture, an increase in density of the lesion, and initial healing of the unicameral bone cyst. (D) A radiograph 3 years after the new pathologic fracture and epiphyseal involvement is shown.

greater the mechanical defect, and therefore, it is assumed, the greater the chance for fracture. The "large lesions" have been defined as showing > 50% cortical involvement on AP and lateral radiographs and a height measurement of > 33 mm by Arata et al,[4] and these authors suggested that "prophylactic curettage and bone grafting be considered if there is a reasonable chance of impending fracture." Easley et al[16] suggested that although absolute size parameters are helpful, 59% of the "large" nonossifying fibromas lesions in their series did not fracture and therefore this did not support prophylactic curettage and grafting based solely on the dimension of the lesion. Fractures through nonossifying fibromas have excellent healing potential.

Unless curettage and bone grafting are done, the lesion usually persists after healing of the pathologic fracture. However, the incidence of documented refracture is low.[14] The data from our study support this finding. Because most of the fractures through nonossifying fibromas are stable, nonoperative treatment with adequate immobilization is recommended. Surgery is indicated according to the characteristics of the fracture configuration and displacement, when the size of the lesion could predispose to refracture or when the diagnosis is unclear.

Aneurysmal Bone Cysts

Aneurysmal bone cysts are expansile pseudotumors of reactive hemorrhagic tissue arising in bone. The etiology of a primary aneurysmal bone cyst is unknown and probably it is the result of a reactive reparative process. Approximately ½ of aneurysmal bone cysts occur primarily; the other ½ occur in association with other lesions, usually neoplasms, and are termed secondary aneurysmal bone cysts.[17]

Pathologic fracture is an infrequent event, occurring in approximately 10% of patients. In the differential diagnosis, the histologic appearance of aneurysmal bone cyst can be difficult to distinguish from unicameral bone cyst after pathologic fracture because of the presence of hemorrhage, fibrin, reactive fibroplasia, and granulation tissue. Pathologic fracture also can cause a fluid-fluid level in a simple cyst that can mimic aneurysmal bone cyst, but the presence of fluid-fluid levels and internal septations on radiographic studies makes the diagnosis of aneurysmal bone cyst more likely.[45,46]

Although not a neoplasm, the response of aneurysmal bone cyst to various surgical procedures is similar to that of benign neoplasms. Inactive, or latent, lesions have a negligible recurrence after curettage. Active lesions have a recurrence rate of more than 30% after curettage, especially in a younger child, and a recurrence rate of 5 to 10% after en bloc marginal excision.[17]

The goal of surgical treatment is to eradicate the lesion, avoid recurrence and restore bone integrity to prevent subsequent pathologic fracture or deformity. These goals often are difficult to achieve and may require multiple intervention, often with considerable morbidity. Sclerotherapy, or the intralesional injection of agents such as ethanol or zein (a vegetable protein) has been shown to be an effective method of treatment associated with low recurrence and little morbidity.[15] Some of the patients in our study might have benefited from this recent form of therapy.

Fibrous Dysplasia

This is a benign, nonfamilial disorder characterized by the presence of expanding intramedullary fibro-osseous tissue in one or more bones, particularly in the diaphyseal regions. Malignant transformation occurs infrequently and occurs more often in patients with polyostotic lesions or after radiation therapy.[7]

Pathologic or stress fractures may cause pain. Undisplaced fatigue fractures are common in areas of stress concentration within dysplastic lesions. The most common site is the femoral neck and calcar area. Most of the monostotic lesions are incidental findings and present little risk for pathologic fracture or deformity and clinical observation is the treatment of choice.

Surgical treatment is indicated to prevent or treat fractures or deformity and to alleviate pain secondary to loss of mechanical integrity of bone. Surgical indications can be summarized for severe or progressive disease, nonunion, persistent pain, or fracture of a weightbearing bone. Progressive deformity is characteristic of these cases and is the main focus of surgical correction. In essence, the aim of treatment is not to eradicate the disease but to correct the poor biomechanics of the bone by realignment and internal fixation. All cases located in the proximal femur initially treated with nonsurgical methods eventually had surgery to correct a varus deformity.

The surgical treatment of this disease is difficult because the fractures do not heal at the same rate as nondysplastic bone regardless of the fact that the patients are children, who have a greater capacity to heal quickly than adults do. If fixation is the choice, the construct should be strong. Consideration should be given to supporting the construct with strong cortical bone resistant to fibrous dysplasia invasion, for example autogenic or allogenic fibula strut graft. Even with such grafts, the patient must be monitored closely because continued resorption from disease progression can occur; particularly in the proximal femur, this can lead to recurrence of the deformity.[6,18] (Figs 6, 7). In light of our results, our recommendation is to surgically reduce and stabilize pathologic fracture

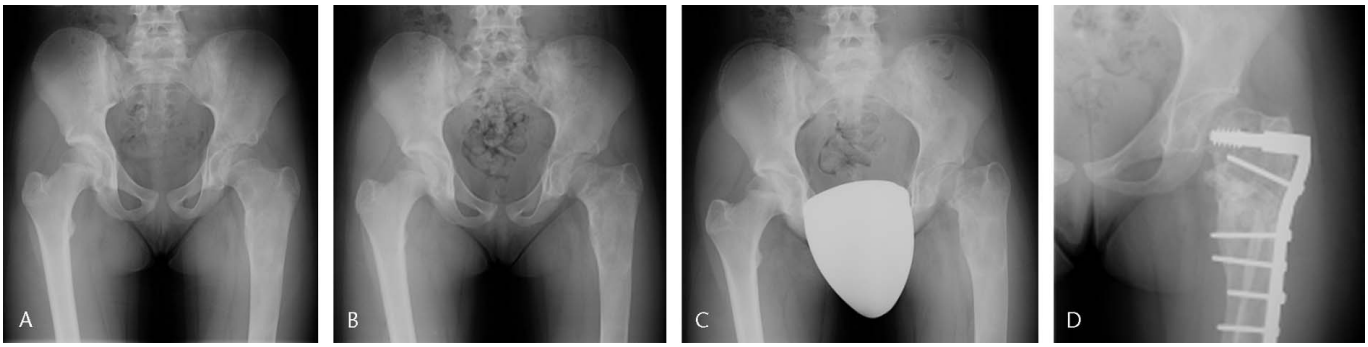


Fig 6A–D. A 13-year-old girl with a polyostotic fibrous dysplasia and neck fracture in the right femur developed a shepherd's crook deformity of the proximal femur, secondary to successive microfractures. (A) An AP radiograph of the pelvis shows the lesion in the proximal femur in 1998. (B) An AP radiograph of the pelvis in 2000 shows a linear fracture in the neck of the femur and coxa vara. (C) The patient has not had surgery and was treated with rest and traction. Successive radiographs showed an increased coxa vara deformity. (D) A plain radiograph taken 2 years after osteotomy, curettage, and intramedullary intercalary allograft shows a good outcome.

through fibrous dysplasia of the proximal femur to avoid malunion and later surgery. Some of these cases may nevertheless be easier to address surgically after malunion, depending on the anatomy of the fracture and related problems with fixation.

Malignant Bone Tumors

The presence of pathologic fracture through a malignant tumor raises the concern of tumor dissemination by the fracture hematoma. In the past, limb-sparing surgery was considered to be contraindicated in patients with malignant bone tumors who sustain a pathologic fracture.[19] This matter was based on theoretical concerns of local dissemination of tumor cells by fracture hematoma and the associated difficulty of doing an adequate resection. Pathologic fracture is also thought to result in tumor dissemination by hematogenous emboli liberated at the time of fracture that imparts a worse prognosis for the patient.[11] It may be, however, that more aggressive tumors cause more local bone destruction and metastasize early, and such lesions fracture early and the fracture may be the manifestation of a more aggressive lesion rather than the cause of a less favorable outcome.

Naturally, inappropriate treatment of the fracture can also increase tumor dissemination by surgical contamination of additional anatomic compartments. Inappropriate observation or nonoperative therapy also is suboptimal and delays the initiation of appropriate therapy.

If a fracture occurs through a malignant bone tumor during or after chemotherapy or radiation therapy, it is important to rule out disease progression or recurrence.[38] If the fracture occurs in a site previously irradiated, secondary sarcoma should be ruled out, especially when the latency period exceeds 10 years. Pathologic fracture at initial presentation historically has been considered an ominous prognostic factor, but recent studies have shown that some if not most of the fractures are able to heal during neoadjuvant chemotherapy.[4,27,47,48]

A Musculoskeletal Tumor Society study[48] showed that a pathologic fracture was significantly associated with a decreased rate of 5-year disease-free survival and an increased rate of local recurrence. However multivariate analysis revealed that although pathologic fracture was a

Fig 7A–B. A 13-year-old boy with a fibrous dysplasia had two pathologic fractures in his femur after nonsurgical treatment. (A) The lateral view of the pathologic fracture is shown. (B) After union of the pathologic fracture, a new pathologic fracture was discovered in the proximal metaphysis.

notable independent risk factor for local recurrence, it was not a risk factor for death. These results contrast with the Rizzoli Institute data.[4] In a review of 735 patients with high-grade osteosarcoma of the extremities treated with five different protocols of neoadjuvant chemotherapy, the rate of amputation was significantly higher in patients with a pathologic fracture than in the patients without a pathologic fracture. However, the outcome for the patients with pathologic fracture was not different from the patients without a pathologic fracture and there were no differences in the rate of local recurrence (4.3% compared with 4.8%). They suggested that a possible explanation for discordance was preoperative chemotherapy. In the study by Scully et al,[48] only 33 patients with a pathologic fracture received preoperative chemotherapy, whereas nineteen had an immediate amputation or limb salvage. In contrast, all of the Rizzoli patients received chemotherapy before surgery. It is possible that the lower rate of local recurrence that they observed was caused by the preoperative treatment. However, our short series (13 patients) showed that 46% of the patients who developed a pathologic fracture died despite fracture treatment and chemotherapy.

The treatment of pathologic fractures cannot be summarized as a recipe for a specific disease but rather should reflect a philosophy of treatment, taking into account the particular aspects of the lesion, its location, and the fracture itself.

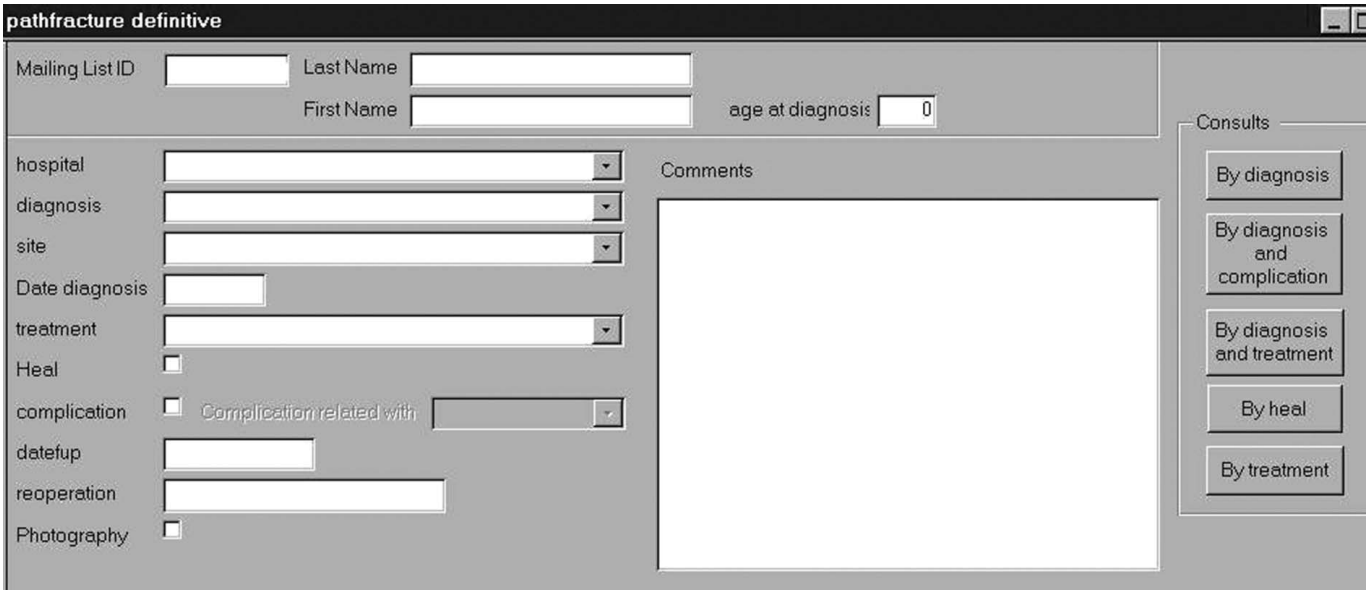
The first and most important step would seem to be the recognition of the pathologic nature of the fracture—the history of minor trauma resulting in a disproportionate bony injury and possibly the presence of pre-existing symptoms. Secondly, the nature of the underlying lesion requires careful consideration.

When in doubt, the physician should defer surgery until investigation is completed. In this context, simple immobilization is recommended as a first-line treatment. When the diagnosis is unclear, biopsy should be done after careful examination of the available data and using careful planning, taking into account the impact of the biopsy on the ultimate treatment.

When fractures occur through benign lesions of the proximal femur, often they require surgical fixation to avoid or correct deformity and facilitate nursing care. (Again, careful biopsy should be done when appropriate before proceeding to definitive surgical treatment.) More distally in the lower extremity, orthopedic treatment less often is associated with deformity; treatment should proceed based on the principles outlined earlier.

Pathologic fractures located in the upper extremity often can be treated by simple immobilization depending on the nature of the lesion and delayed treatment of the tumor once the fracture has healed.

A fracture through a malignant tumor is no longer considered an absolute contraindication to limb-salvage surgery. Treatment should be individualized based on the tumor extension as shown by modern imagery and the apparent response of the tumor to neoadjuvant chemotherapy.



Appendix 1. The questionnaire in which we collected our data is shown.

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