INTRODUCTION — Benign childhood bone tumors range from static lesions, such as nonossifying fibromas, which remain essentially unchanged throughout childhood, to locally aggressive lesions, such as aneurysmal bone cysts, which continue to expand until treated. (See 'Nonossifying fibroma' below and 'Aneurysmal bone cyst' below.)

Most benign bone tumors have characteristic radiographic features and can be diagnosed with plain radiographs. It is important to be familiar with the radiographic appearance of the most common benign bone tumors. Benign bone tumors often are discovered incidentally, and recognition of benign lesions on plain radiographs can avoid unnecessary advanced imaging and invasive diagnostic studies.

An overview of the presentation, clinical and radiographic features, and management of the most common benign pediatric bone tumors will be presented below. Malignant bone tumors (Ewing sarcoma and osteosarcoma) are discussed separately. (See "Clinical presentation, staging, and prognostic factors of the Ewing sarcoma family of tumors" and "Osteosarcoma: Epidemiology, pathogenesis, clinical presentation, diagnosis, and histology".)

OVERVIEW

Clinical evaluation — Benign bone tumors often are asymptomatic and discovered incidentally during evaluation for trauma or another condition [1]. When they are symptomatic, benign bone tumors may present with localized pain, swelling, deformity, or pathologic fracture. In most cases, the differential diagnosis of these lesions can be narrowed based upon the age of the child, the involved bone, the location of the lesion within the bone, and other general radiographic characteristics (table 1) [2].

History — Certain aspects of the history may be helpful in narrowing the differential diagnosis of a benign-appearing bone tumor. These include (table 1) [1]:

- **Age** – Most benign bone tumors typically present during the second decade. However, ossifying fibroma (osteofibrous dysplasia) typically presents in the first five years of life, and Langerhans cell histiocytosis of bone presents in the first decade of life.
- **Pain** – Pain that quickly resolves (within 20 to 25 minutes) with aspirin or other nonsteroidal antiinflammatory medications is characteristic of osteoid osteoma. Nonaggressive benign bone tumors (eg, unicameral bone cyst, nonossifying fibroma) usually are asymptomatic but may cause pain in association with pathologic fracture, bursa formation, or neurovascular compression [1]. Aggressive benign bone tumors (eg, aneurysmal bone cyst, chondroblastoma, chondromyxoid fibroma) may cause pain that is mild, dull, slowly progressive, and worse at night [3]. The pain associated with malignant
bone tumors may awaken the child from sleep but is more rapidly progressive than the pain of aggressive benign bone tumors [3].

- **Systemic symptoms** – Associated systemic symptoms (eg, fever, malaise) may indicate an underlying generalized disorder or osteomyelitis [3].

**Examination** — Important aspects of the examination in a child with a bone tumor include [1,3]:

- **Growth parameters** – Patients with hereditary multiple osteochondromas may have short stature.
- **Head, eyes, ears, nose, and throat** – Signs of chronic/refractory otitis media (eg, scarring of the tympanic membrane) and proptosis may indicate Langerhans cell histiocytosis. (See "Langerhans cell histiocytosis (eosinophilic granuloma) of bone in children".)
- **Musculoskeletal** – During the musculoskeletal examination, it is important to determine the location and size of the lesion(s) and to assess bone tenderness, bone or joint swelling, deformity (patients with hereditary multiple osteochondromas or enchondromas may have angular deformities of the upper or lower extremities), joint range of motion, neurologic function, and vascular function (neurovascular compromise may indicate an aggressive tumor). Osteochondromas and periosteal chondromas may be palpable.
- **Skin** – Café-au-lait macules (picture 1) may indicate polyostotic fibrous dysplasia (McCune-Albright syndrome) or neurofibromatosis. Soft tissue myxomas may indicate Mazabraud syndrome [4]. Hemangiomas may suggest Maffucci syndrome (a subtype of enchondromatosis). Overlying erythema, warmth, and soft tissue swelling may suggest underlying infection (eg, osteomyelitis).
- **Lymph nodes** – Regional lymphadenopathy may indicate infection or a systemic process.

**Radiologic evaluation** — Collaboration between the clinician and radiologist is essential [5]. Most benign bone tumors have characteristic radiographic features and can be diagnosed with plain radiographs. It is important to be familiar with the radiographic appearance of the most common benign bone tumors (table 1). Benign bone tumors often are discovered incidentally, and recognition of benign lesions on plain radiographs may avoid unnecessary advanced imaging (eg, computed tomography, magnetic resonance imaging) and invasive diagnostic studies (eg, biopsy).

Plain radiographs — Plain radiographs are the best initial modality for evaluation of primary bone lesions [1,2]. The evaluation should include views in two planes [1]. The diagnosis of benign bone tumor can be made with plain radiographs alone in 80 to 90 percent of cases [3]. If the lesion is clearly benign on plain films, cross-sectional imaging should not be necessary [2].

Tumors of the pelvis or scapula can be difficult to see on plain films. Advanced imaging techniques (eg, computed tomography, magnetic resonance imaging) may be necessary to fully characterize bone tumors in these locations [3].

Plain radiographs should be reviewed systematically by considering the following questions [1,6]:

- Where is the tumor? (Long bone or flat bone?; epiphysis, metaphysis, or diaphysis?; medulla, cortex, or surface?) (figure 1)
- What is the tumor doing to the bone? Is the tumor destroying or replacing existing bone? If so, what is the pattern?
- What is the bone doing to the tumor? Is there periosteal or endosteal reaction? If so, is it well-developed? Is it sharply defined? What is the type of periosteal reaction: reinforcing, spiculated, solid, interrupted?
- Are there intrinsic characteristics that suggest histology? Is there bone formation? Calcification? Is the lesion completely radiolucent?

Radiographic characteristics of benign bone lesions include [2,7]:
Well-defined or sclerotic border
- Sharp zone of transition
- Small size or multiple lesions
- Confinement by natural barriers (e.g., growth plate, cortex)
- Lack of destruction of the cortex
- Lack of extension into the soft tissue

Radiographic characteristics of more aggressive lesions include [2,7]:

- Poor definition
- Cortical destruction ("moth-eaten" or permeative pattern)
- Spiculated or interrupted periosteal reaction; however, the absence of these findings does not exclude an aggressive lesion [8]
- Extension into the soft tissue
- Large size

Advanced imaging — Advanced imaging techniques (computed tomography [CT], magnetic resonance imaging [MRI], scintigraphy) may be necessary for bone lesions that are not clearly benign on plain radiographs. Advanced imaging also may be necessary to detect tumors in the spine, scapula, ribs, or pelvis [3].

CT may be helpful in evaluating areas of the body that are difficult to see with plain films (e.g., pelvis, scapula, spine) [2]. Compared with plain film, CT can better define the location of the lesion within the bone (e.g., periosteal, cortex, medullary), more accurately evaluate changes in the cortex (e.g., focal destruction or endosteal scalloping), and can detect more subtle matrix mineralization. It also may be helpful in guiding therapy (e.g., localization of the nidus in osteoid osteoma). However, the benefit of the supplementary information must be weighed against the additional radiation exposure.

MRI is the modality of choice when a malignant bone tumor is suspected [2]. It is the most sensitive modality for evaluation of medullary changes and defining the extent of the lesion and provides the best contrast resolution for demonstrating soft tissue masses and invasion of adjacent structures. Sedation may be required for MRI.

Bone scintigraphy may be helpful in evaluating metastatic disease or a multifocal process [2]. However, it is nonspecific, which limits its use in the initial evaluation of a single bone lesion.

Management — Most benign bone tumors are managed with observation (e.g., serial examination and radiographs). Symptomatic or aggressive benign tumors (osteoblastoma, periosteal chondroma, chondroblastoma, chondromyxoid fibroma, giant cell tumor, and aneurysmal bone cyst) usually are treated with curettage and bone grafting or excision. The management of specific tumors is discussed below.

Referral indications — Most children with bone tumors should be referred to and followed by an orthopedic surgeon familiar with these tumors, such as a tumor or pediatric specialist. Small fibromas, fibrous cortical defects, and asymptomatic osteochondromas that are detected incidentally are exceptions and may not require referral.

CLASSIFICATION — Bone tumors can be classified into several categories according to the matrix, or substance, that they produce:

- Osteoid- or bone-forming tumors, including osteoid osteoma and osteoblastoma (see 'Bone-forming tumors' below)
- Cartilage-forming tumors, including osteochondroma (exostosis), chondroma (enchondroma, periosteal chondroma), chondroblastoma, and chondromyxoid fibroma (see 'Cartilage-forming tumors' below)
• Fibrous lesions, including fibrous dysplasia, ossifying fibroma (osteofibrous dysplasia), and nonossifying fibroma (see 'Fibrous lesions' below)
• Cystic and vascular lesions, including unicameral bone cyst and aneurysmal bone cyst (see 'Cystic tumors' below)

BONE-FORMING TUMORS — Osteoid osteoma and osteoblastoma are benign bone-forming tumors.

Osteoid osteoma — Osteoid osteoma is a benign bone-forming tumor that is characterized by a small radiolucent nidus (usually <1 to 1.5 cm in diameter) [3]. The nidus produces high levels of prostaglandins [9]. In addition to prostaglandins, there is some evidence that osteoid osteomas may secrete osteocalcin [10].

Clinical features – Osteoid osteoma typically presents during the second decade [11,12]. The lower extremity is most frequently affected; the proximal femur is the most common site. Other common locations are the tibia, the remainder of the femur, and the spine. Boys are affected two to three times as often as girls [12,13].

Patients with osteoid osteoma typically complain of progressively increasing pain that is worse at night and unrelated to activity [3]. The pain is relieved by aspirin or other nonsteroidal antiinflammatory medications (ie, prostaglandin inhibitors), usually within 20 to 25 minutes [12,14,15]. Lack of relief by nonsteroidal antiinflammatory agents should prompt consideration of other diagnoses [3].

Children with lower-extremity lesions may present with limp, swelling, muscular atrophy, leg-length discrepancy, bone deformities, muscle contractures, and local point tenderness at the site of the lesion [9,11,12]. (See "Approach to the child with a limp".)

Children with spine lesions may present with limp, scoliosis, localized tenderness, restriction of motion, and/or spasm of paravertebral muscles [12]. Children and adolescents with new-onset, painful scoliosis should be evaluated for osteoid osteoma. (See "Clinical features; evaluation; and diagnosis of adolescent idiopathic scoliosis", section on 'History'.)

Radiographic findings – On plain radiographs, osteoid osteoma appears as a small, round lucency (nidus) with a sclerotic margin [16]. There may be a central ossification (image 1).

Approximately 25 percent of osteoid osteomas are not obvious on plain radiographs, either because of their location (eg, in the spine) or because cortical thickening obscures the nidus (eg, in the shaft of a long bone such as the tibia or femur), and require computed tomography (CT) or magnetic resonance imaging (MRI) for identification (panel A of radiograph) (image 2) [17-20].

Differential diagnosis – The differential diagnosis of osteoid osteoma includes stress fracture, infection (eg, osteomyelitis, bone abscess), and osteoblastoma. These conditions generally can be distinguished by their characteristic clinical and radiographic features.

• The pain of stress fractures usually worsens with activity and is relieved with rest; on plain radiographs, stress fractures typically are linear and run perpendicular or at an angle to the cortex, rather than parallel to it [15]. (See "Overview of stress fractures".)
• Bone infections may have a tract that extends from the lesion toward the nearest growth plate [15]. (See "Clinical features of hematogenous osteomyelitis in children".)
• The pain of osteoblastoma is more generalized and chronic and less responsive to nonsteroidal antiinflammatory medications than that of osteoid osteoma. It typically has a larger nidus, although this may not be visible. (See 'Osteoblastoma' below.)
Treatment – The treatment of osteoid osteoma depends upon the presence of symptoms. Lesions with symptoms that are tolerable or can be controlled with nonsteroidal antiinflammatory agents may be observed with serial examinations and radiographs every four to six months.

Treatment options for symptomatic lesions (eg, intolerable pain, limp, scoliosis) include surgical resection, which may be aided by CT-guided needle localization, or radiofrequency ablation (in certain institutions) (image 2) [21]. Treatment options may be limited by proximity to vital structures.

Prognosis – Untreated osteoid osteoma spontaneously resolves over the course of several years [14,22]. Removal of the nidus generally results in resolution of pain. Recurrence is possible if the nidus is not completely removed.

Osteoblastoma — Osteoblastoma is a rare benign bone-forming tumor of unknown etiology.

Clinical features – Osteoblastoma typically presents during the second decade, but may be seen at any age. Boys are affected more often than girls.

The most common location of osteoblastoma is the posterior column of the spine (the spinous process, lamina, and pedicles). Other common locations are depicted in the figure (figure 2). Tumors in the spine may be difficult to identify on plain radiographs [23].

Patients with osteoblastoma typically complain of chronic pain. The pain is less responsive to nonsteroidal antiinflammatory agents than that of osteoid osteoma [6]. Osteoblastoma uncommonly may cause systemic symptoms [24]. Children with spine lesions may present with limp or neurologic symptoms secondary to cord or nerve root compression. Children with lower extremity lesions may present with limp.

Radiographic findings – The radiographic findings of osteoblastoma are variable, and advanced imaging (eg, CT or MRI) often is required for identification. Osteoblastoma may appear similar to osteoid osteoma but is usually larger (>2 cm in diameter); it may appear as an expansive lesion (image 3), similar to an aneurysmal bone cyst, or it may have aggressive features, mimicking a malignant neoplasm [15]. Unlike more aggressive tumors, osteoblastomas rarely extend into the soft tissues.

Differential diagnosis – The differential diagnosis of osteoblastoma includes:

- Stress fracture (see "Overview of stress fractures")
- Infection (eg, osteomyelitis, bone abscess) (see "Clinical features of hematogenous osteomyelitis in children")
- Osteoid osteoma (see 'Osteoid osteoma' above)
- Osteosarcoma, a malignant bone tumor (see "Osteosarcoma: Epidemiology, pathogenesis, clinical presentation, diagnosis, and histology")
- Aneurysmal bone cyst (see 'Aneurysmal bone cyst' below)

These conditions may require advanced radiographic imaging (eg, CT or MRI) for identification.

Treatment – Treatment for osteoblastoma generally entails curettage and bone grafting. En block excision may be warranted for more aggressive lesions or in regions that permit excision of the bone (eg, fibula). Radiation may be required for spinal lesions when the tumor cannot be completely resected [25].

Prognosis – Untreated osteoblastoma continues to enlarge and may damage the bone and adjacent structures [25]. It may cause progressive neurologic symptoms if it abuts the spinal canal or neural foramina. The prognosis is good if the lesion can be completely removed. The rate of recurrence is up to 20 percent if the lesion has expanded outside the bone [22].
CARTILAGE-FORMING TUMORS — Benign cartilage-forming tumors of childhood include osteochondroma (exostosis), chondroma (enchondroma, periosteal chondroma), chondroblastoma, and chondromyxoid fibroma.

Osteochondroma and hereditary multiple osteochondromas — An osteochondroma (osteocartilaginous exostosis) is a bony spur arising on the external surface of a bone (image 4). A cartilaginous cap overlies the bony spur and is the source of growth (picture 2). The cartilage cap is thick in the child, narrows during adolescence, and generally is <1 cm in the adult [5].

Hereditary multiple osteochondromas (HMO, also known as hereditary multiple exostoses [HME, MIM #133700, MIM #133701, and MIM #600209]) is characterized by two or more exostoses in the appendicular and axial skeleton (image 5). Most cases are caused by autosomal dominant inheritance of a germline mutation in the tumor suppressor genes EXT1 or EXT2 [26]. However, spontaneous mutations also occur [5]. The prevalence of HMO in the general population is approximately 1:50,000 [27-29].

Clinical features – Osteochondromas and HMO typically present during the second decade. Males are affected more often than females [5,6].

Osteochondromas usually occur around the knee or the proximal humerus. The distal femur is the most common location.

Osteochondromas typically present as a painless mass near a joint or on the axial skeleton, or as a painful mass associated with local trauma. Osteochondromas near the ends of long bones are palpable. Deep osteochondromas (eg, in the axial skeleton) may be an incidental radiographic finding. Osteochondromas can cause pain, functional problems (decreased range of motion), deformity, and pathologic fracture.

Osteochondroma can affect nearby growth plates. Patients with HMO may have short stature and angular deformities (ie, varus or valgus deformities) of the upper or lower extremities. The deformities in children with involvement of the wrist and forearm may be severe. Hemiepiphysiodesis can be performed in some cases to allow gradual improvement. (See "Approach to the child with knock-knees", section on 'Other causes'.)

Osteochondroma can involve the vertebra and may encroach on the spinal canal. One group recommends screening magnetic resonance of the spine at least once during the growing years for children with HMO [30].

Radiographic findings – Radiographic features include a bony spur (sometimes large) that arises from the surface of the cortex and usually points away from the joint (image 4). The cortex of the spur is continuous with the cortex of the underlying bone.

The cartilage cap is thick in the child (may be >2 cm), narrows during adolescence, and generally is <1 cm in the adult [5]. If the cartilaginous cap is >1 cm in an adult, malignant transformation to chondrosarcoma is a concern. However, in a review of 67 osteochondromas and 34 secondary chondrosarcomas in adults, no chondrosarcomas had cartilaginous caps <2 cm, and 18 percent of (benign) osteochondromas had a cartilaginous cap >1 cm [31]. Biopsy and removal of the entire osteochondroma may be warranted for lesions with a cap ≥2 cm thick.

Differential diagnosis – The differential diagnosis of osteochondroma includes parosteal osteosarcoma (a low-grade surface osteosarcoma). Whereas the medullary canal of osteochondromas is always continuous with that of the bone, this is usually not the case with parosteal osteosarcomas. (See "Osteosarcoma: Epidemiology, pathogenesis, clinical presentation, diagnosis, and histology", section on 'Surface (juxtacortical) osteosarcomas'.)
Treatment – Most osteochondromas can be observed without treatment. Patients should be examined and may have radiographs taken yearly [3].

Indications for excision may include local irritation or deformity and concern for malignant transformation (cartilage cap ≥2 cm thick in an adult; increase in size after skeletal maturity; growth disturbance; new onset of symptoms; lesions of the spine, scapula, pelvis, or proximal femur) [3,5,32-34].

Prognosis – Osteochondromas grow throughout childhood. They generally stop growing when the physes (growth plates) close and remain static throughout adulthood. There is a moderate risk of recurrence if osteochondromas are incompletely removed before the physes close [35]. Positive prognostic factors (more benign presentation and less functional limitations) include female sex, involvement of <5 sites, and HMO caused by EXT2 mutations. None of these factors was predictive of malignant transformation [36].

Osteochondromas may cause local irritation. Lesions of the proximal femur may cause arthritis of the hip joint [37].

There is a small lifetime risk of malignant transformation to chondrosarcoma, which occurs during adulthood and most commonly in patients with HMO (in as many as 5 percent of cases) [26,32,36]. Malignant transformation may be heralded by a change in size of an osteochondroma after skeletal maturity or new onset of symptoms [3,5,32-34]. Osteochondromas of the spine, scapula, pelvis, and proximal femur are particularly prone to malignant transformation. (See "Chondrosarcoma", section on 'Osteochondroma'.)

Enchondroma — Enchondromas are benign cartilage-forming tumors that develop in the medulla (marrow cavity) of long bones (image 6).

Enchondromatosis (Ollier disease, MIM #166000) is defined by multiple enchondromas, often with a unilateral predominance (image 7) [3,6,26]. The estimated prevalence is 1 in 100,000 [38]. Maffucci syndrome is a subtype of enchondromatosis that is characterized by multiple enchondromas and soft tissue hemangiomas (image 8). Most cases of enchondromatosis and Maffucci syndrome are sporadic and associated with somatic mutations in the isocitrate dehydrogenase-1 and isocitrate dehydrogenase 2 genes (IDH1 and IDH2) [39,40].

Clinical features – Enchondromas typically present during the second decade but can present at any age. Enchondromatosis usually presents in children younger than 10 years [38].

Enchondromas usually occur in the long bones, particularly the long bones of the hand, followed by the humerus and femur (figure 3). They generally are central, metaphyseal lesions [6]. Enchondromas occur with equal frequency in males and females.

The signs and symptoms vary depending upon the anatomic site, extent, and distribution of involvement. Most enchondromas are asymptomatic unless a fracture is present [3]. They often are incidental findings. When symptomatic, clinical manifestations may include widening of the bone, angular deformity, and limb-length discrepancy [9]. In patients with enchondromatosis, enchondromas may arise from the skull base; clinical features include headache and cranial nerve palsy [41].

Radiographic findings – Radiographic features of enchondromas include an oval, well-circumscribed, central lucent lesion, with or without matrix calcifications (image 6) [42]. Matrix calcification usually is not seen in children [3,6]. There may be expansion of the surrounding cortex, especially when the lesion is in the hand or foot (image 7 and image 9). Multiple lesions may be present.

Differential diagnosis – The differential diagnosis of enchondromas includes bony infarcts (image 10) and low-grade chondrosarcoma (image 11), which must be excluded in patients with enchondroma who have pain without fracture. (See "Chondrosarcoma", section on 'Diagnostic and staging work-up'.)
Treatment – The treatment of enchondromas depends upon the presence of symptoms and size. Those that are asymptomatic and small enough not to increase the risk of pathologic fracture may be observed. The risk of fracture is increased when the lesions occur in a weight-bearing bone, are >25 mm in diameter, and involve >50 percent of the diameter of the cortex. The frequency of follow-up depends upon the size, location, and number of lesions.

Symptomatic chondromas are treated with curettage and bone grafting; low-grade chondrosarcoma must be excluded in patients with pain in the absence of fracture. Fractures should be permitted to heal before curettage.

Prognosis – Solitary enchondromas usually are self-limited. However, they may continue to grow. Recurrence after curettage and bone graft is rare [6].

Enchondromas, particularly those of the long bones and pelvis, may be complicated by malignant transformation to chondrosarcoma (image 11), which usually occurs after skeletal maturity and may be heralded by the development of pain [43]. Malignant transformation of a solitary enchondroma is extremely rare (<1 percent), but has been described [43]. The risk of malignant transformation is increased (as high as 50 percent) in patients with enchondromatosis (Ollier disease) or Maffucci syndrome [26,38,44-47]. Enchondromatosis and Maffucci syndrome also are associated with nonsarcomatous neoplasms, including brain tumors [3,48,49].

Periosteal chondroma — Periosteal chondromas (juxtacortical chondromas) are rare, benign, cartilage-forming tumors that arise from the surface of the cortex, deep in the periosteum, and erode into the cortex [3,6,50].

Clinical features – Periosteal chondroma occurs in children and adults [6]. The most common site is the proximal humerus; the other long bones and small bones of the hands and feet also may be involved [3,50]. Clinical features may include pain at the site of the lesion and a palpable nontender hard mass that is fixed to bone.

Radiographic features – On plain radiographs, periosteal chondromas appear as small, scalloped, radiolucent lesions on the outer surface of the cortex in the metaphysis or diaphysis (image 12) [3,6,50-53]. There is a rim of sclerotic bone [6]. Calcification is present in approximately one-third of cases [51]. Periosteal reaction is minimal.

Differential diagnosis – The differential diagnosis of periosteal chondroma includes [52]:

- Nonossifying fibroma (see ‘Nonossifying fibroma’ below)
- Soft-tissue tumors, secondarily eroding into the cortical bone
- Chondrosarcoma, a malignant tumor (see "Chondrosarcoma")
- Osteosarcoma, a malignant tumor (see "Osteosarcoma: Epidemiology, pathogenesis, clinical presentation, diagnosis, and histology")

Treatment – Periosteal chondroma usually is treated with extended curettage or en block excision to minimize the risk of local recurrence [3,6].

Chondroblastoma — Chondroblastoma is a benign cartilage-forming tumor that usually arises in the epiphyses or apophyses of long bones [42].

Clinical features – Chondroblastoma typically presents during the teenage years. The most common sites are the epiphysis of the proximal humerus (image 13), distal femur (image 14), and proximal tibia (image 15); 30 percent occur around the knee (figure 4) [54]. Chondroblastoma is approximately 1.5 times more common in boys than in girls. Symptoms of chondroblastoma include low-grade joint pain (constant, unrelated to activity) and swelling [9,22].
Radiographic findings – On plain radiographs, chondroblastomas appear as small, well-defined lesions with a sclerotic border that may cross the physis (growth plate) (image 16). Matrix calcification may be seen [22].

Differential diagnosis – The differential diagnosis of chondroblastoma includes:

- Giant cell tumor, a benign but locally aggressive skeletal tumor that occurs near the growth plate in young adults. (See "Giant cell tumor of bone".)
- Chondromyxoid fibroma. (See 'Chondromyxoid fibroma' below.)
- Avascular necrosis, an abnormality of subchondral bone in which pain is activity related. In contrast, in chondroblastoma, subchondral bone is normal, and pain is constant, unrelated to activity [9]. (See "Osteonecrosis (avascular necrosis of bone")
- Aneurysmal bone cyst. (See 'Aneurysmal bone cyst' below.)
- Osteomyelitis. (See "Clinical features of hematogenous osteomyelitis in children".)
- Clear cell chondrosarcoma. (See 'Chondrosarcoma', section on 'Clear cell chondrosarcoma'.)

Treatment – Chondroblastoma is treated with curettage and bone grafting. Reconstruction may be difficult if chondroblastoma involves the articular surface.

Prognosis – The prognosis for patients with chondroblastoma generally is good. Chondroblastoma that involves the articular surface may result in arthritis. Recurrence rates of up to 20 percent are reported [6]. In such cases, en bloc resection may be warranted.

Chondromyxoid fibroma — Chondromyxoid fibroma is a rare, benign, cartilage-forming tumor of the tubular long bones.

Clinical features – Chondromyxoid fibroma usually presents in the teens or 20s. Approximately one-quarter of cases occur in the proximal tibia (image 17), with the distal femur and calcaneus being the next most common sites. Males are affected approximately 1.5 times as often as females [55]. Symptoms of chondromyxoid fibroma may include pain and swelling.

Radiographic findings – Chondromyxoid fibroma is an eccentric, intramedullary, lobulated or bubbly lesion in the metaphysis; it has a sclerotic border (image 18). It typically is lucent, with a rare chondral matrix [22,42].

Differential diagnosis – The differential diagnosis of chondromyxoid fibroma includes:

- Nonossifying fibroma (see 'Nonossifying fibroma' below)
- Aneurysmal bone cyst (see 'Aneurysmal bone cyst' below)
- Chondroblastoma (see 'Chondroblastoma' above)
- Osteomyelitis (see "Clinical features of hematogenous osteomyelitis in children")
- Fibrous dysplasia (see 'Fibrous dysplasia' below)

Treatment – The treatment of chondromyxoid fibroma is curettage and bone grafting.

Prognosis – The prognosis of chondromyxoid fibroma generally is good. There is a 20 percent risk of recurrence, which may require en bloc resection [6].

FIBROUS LESIONS — Benign fibrous lesions of childhood include fibrous dysplasia, osteofibrous dysplasia (ossifying fibroma), and nonossifying fibroma.

Fibrous dysplasia — Fibrous dysplasia is a lesion in which portions of the bone are replaced by fibrous connective tissue and poorly formed trabecular bone [6]. The process originates in the medullary cavity. It is
caused by a postzygotic mutation in the guanine nucleotide stimulatory protein (GNAS1) gene. It is more of a skeletal dysplasia than a true neoplasm.

Fibrous dysplasia may occur in single or multiple bones (monostotic and polyostotic fibrous dysplasia, respectively). The polyostotic form of fibrous dysplasia is known as McCune-Albright syndrome (or Albright syndrome, MIM #174800) and is associated with endocrine abnormalities and café-au-lait spots (picture 1). Mazabraud syndrome is characterized by fibrous dysplasia and soft tissue myxomas; it overlaps clinically with McCune Albright syndrome [4].

Clinical features – Fibrous dysplasia most commonly presents in the teens or 20s. It may occur in any bone but is most common in the proximal femur (image 19), tibia, ribs, and skull (figure 5) [5]. Fibrous dysplasia affects slightly more males than females.

Most patients with fibrous dysplasia are asymptomatic [5]. However, fibrous dysplasia may be painful or cause swelling. It can cause repeated pathologic fractures or severe bone deformity, such as the "shepherd's crook" varus deformity of the proximal femur (image 20) [6].

Radiographic findings – On plain radiographs, fibrous dysplasia appears as a lytic lesion in the metaphysis or diaphysis with a "ground glass" appearance (image 21). There is expansion of the bone and possible bowing. The cortical bone is thinned with a scalloped, undulating pattern due to endosteal erosion [9]. Periosteal reaction usually is absent unless there is a pathologic fracture.

Differential diagnosis – The differential diagnosis of fibrous dysplasia includes:

- Nonossifying fibroma (see 'Nonossifying fibroma' below)
- Unicameral bone cyst (see 'Unicameral bone cyst' below)
- Aneurysmal bone cyst (see 'Aneurysmal bone cyst' below)
- Chondromyxoid fibroma (see 'Chondromyxoid fibroma' above)

Treatment – The treatment of fibrous dysplasia depends upon the presence of symptoms. Asymptomatic patients may be observed every six months with serial radiographs. Children with large lesions or lesions in the proximal femur or other weight-bearing bones are observed more frequently.

Curettage, bone grafting, and stabilization may be warranted for fibrous dysplasia that is associated with symptoms (pain, deformity) or fracture; however, there is a high rate of recurrence. Autograft should not be used because it will be resorbed. Bisphosphonate therapy is another alternative for symptomatic patients [56,57].

Prognosis – The deformity of fibrous dysplasia may progress with skeletal growth [6]. Fibrous dysplasia usually is static after growth ceases but may be reactivated with pregnancy [58,59]. Fibrous dysplasia often recurs after curettage and bone grafting.

Ossifying fibroma — Ossifying fibroma (also called osteofibrous dysplasia, intracortical fibrous dysplasia, Jaffe-Campanacci syndrome) is not a tumor, per se, but a deformity-inducing fibro-osseous lesion of the tibia and/or fibula [60,61]. The process originates in the cortex [6].

Clinical features – Ossifying fibroma occurs in children younger than 10 years of age [60,61]. It generally affects the tibia and fibula. Clinical features include swelling and/or anterolateral bowing of the lower leg. Osteofibrous dysplasia is painful only if it associated with a pathologic fracture.
Radiographic findings – Radiographic features of ossifying fibroma include a lytic thinning of the diaphyseal cortical bone with interspersed sclerosis, causing anterior or anterolateral bowing. There is a sharply circumscribed margin [6].

Differential diagnosis – The differential diagnosis of ossifying fibroma includes monostotic fibrous dysplasia (which originates in the medulla rather than the cortex), adamantinoma (a low-grade malignant bone tumor), and nonossifying fibroma. (See ‘Fibrous dysplasia’ above and ‘Nonossifying fibroma’ below.)

Treatment – The treatment of ossifying fibroma is usually observation. Asymptomatic patients may be observed every six months with serial radiographs. Children with large lesions or lesions in the proximal femur or other weight-bearing bones are seen more frequently (eg, every three to four months). Excision, bone graft, and correction of bony deformity may be warranted for lesions that are symptomatic (ie, with pain or deformity) after skeletal maturity.

Prognosis – Ossifying fibroma is noninvasive. However, it will recur if it is excised before skeletal maturity. Excision after skeletal maturity is usually successful [62,63].

Nonossifying fibroma — Nonossifying fibroma is a benign fibrous lesion that is also known as metaphyseal cortical defect, fibrous cortical defect, and benign metaphyseal bone scar. It is a developmental defect in which areas that normally ossify are filled with fibrous connective tissue [6].

Clinical features – Nonossifying fibroma usually is an incidental radiographic finding in teenagers. It occurs most commonly in the distal femur, followed by the distal tibia (image 22), and the proximal tibia (image 23). Girls are affected as often as boys [64].

Nonossifying fibroma usually is asymptomatic and discovered incidental to trauma [5]. Large lesions may be associated with pathologic fracture [1].

Radiographic findings – On plain radiographs, nonossifying fibromas appear as small, well-defined, eccentric, expansile, lytic lesions located in the metaphysis with scalloped sclerotic borders [1,6]. Multiple lesions may be present.

Nonossifying fibromas may have an atypical appearance as they fill with normal bone before they disappear [5,6]. When there is a concomitant pathologic fracture, nonossifying fibroma may have a more aggressive radiographic appearance, but it should not be mistaken for malignant tumor [5].

Differential diagnosis – The differential diagnosis of nonossifying fibroma includes:

- Chondromyxoid fibroma (see ‘Chondromyxoid fibroma’ above)
- Fibrous dysplasia (see ‘Fibrous dysplasia’ above)
- Langerhans cell histiocytosis (see "Langerhans cell histiocytosis (eosinophilic granuloma) of bone in children")

Treatment – Small, asymptomatic nonossifying fibromas that are discovered incidentally do not require any further follow-up. Parents are simply counseled to bring their child in if the area becomes painful. In younger children, the lesions may grow relative to adjacent bone, increasing the risk of fracture [5].

Curettage and bone grafting may be warranted for lesions causing pain or to prevent pathologic fracture if the lesion is greater than 50 percent of the diameter of the bone or is in a high-stress area (eg, distal femoral metaphysis) [1,5].
Prognosis – The prognosis for nonossifying fibroma generally is excellent. They usually fill in during adolescence [5]. The risk of recurrence is lower than for other benign tumors.

CYSTIC TUMORS — Benign cystic tumors include unicameral bone cysts and aneurysmal bone cysts.

Unicameral bone cyst — Unicameral bone cysts (simple bone cysts, solitary bone cysts) are fluid-filled lesions with a fibrous lining [1].

Clinical features – Unicameral bone cysts generally occur in the first 20 years of life. The proximal humerus and femur are the most common locations. Unicameral bone cysts occur with equal frequency in boys and girls [5].

Unicameral bone cysts commonly present with a pathologic fracture. However, they may be an incidental radiographic finding. Symptoms may include localized pain, limp, or failure to use the extremity normally [1,5].

Radiographic findings – On plain radiographs, unicameral bone cysts (UBCs) appear as well-marginated cystic lesions of the metaphysis or metadiaphysis without reactive sclerosis [1,5] (image 24). The lesion usually involves the full diameter of bone, with expansion of the cortex [6].

UBCs with pathologic fractures may be indicated by the "fallen fragment" or "fallen leaf" sign, in which a fragment of bone falls to the bottom of the cyst (image 25) [9,65].

Differential diagnosis – The differential diagnosis of UBC includes aneurysmal bone cyst and fibrous dysplasia. (See 'Aneurysmal bone cyst' below and 'Fibrous dysplasia' above.)

Treatment – The treatment of UBCs is observation with serial radiographs every four to six months. Activity restrictions are often necessary to avoid pathologic fracture. Alternatively, UBCs may be aspirated and injected with methylprednisolone [66,67]. Although it has never been conclusively proven, some authors believe that glucocorticoid injection may hasten the resolution of the cyst and thus allow patients to return to normal activities. Numerous substances have been injected into UBCs, including bone marrow, growth factors, and demineralized bone graft. None have been proven to be as effective as or more effective than steroid injection [68].

Curettage and bone grafting rarely are required for large lesions that compromise the structural integrity of the bone [69,70].

Prognosis – UBCs spontaneously resolve in all patients. However, resolution may not occur until after skeletal maturity.

Aneurysmal bone cyst — Aneurysmal bone cysts are benign expansile vascular lesions that consist of blood-filled channels [6,9]. They may grow rapidly and destroy bone. Aneurysmal bone cysts generally are solitary [1]. They may be primary or related to other benign bone lesions (eg, giant cell tumor, osteoblastoma, chondroblastoma) [2,5,6].

Clinical features – Aneurysmal bone cysts generally occur in adolescents. They may be found in any bone but are most common in the posterior spinal elements, femur, and tibia (figure 6) [71]. Aneurysmal bone cysts are slightly more common in girls than in boys [5].

Aneurysmal bone cysts typically cause localized pain. They may present with pathologic fracture, limp, or swelling as the lesion increases in size [1,5]. Lesions in the spine may be associated with neurologic symptoms [72]. Lesions that cross the growth plate may cause growth arrest [9].
Radiographic findings – On plain radiographs, aneurysmal bone cysts appear as aggressive, expansile, lytic metaphyseal lesions with an "eggshell" sclerotic rim (image 26). Pathologic fracture or periosteal reaction may be present (image 27). The lesions are sharply circumscribed. They may have a "soap bubble" appearance secondary to the reinforcement of the remaining trabeculae that support the bone structure [6]. The cortex is usually intact, although it may be thin.

Differential diagnosis – The differential diagnosis of aneurysmal bone cyst includes:

- Unicameral bone cyst (see 'Unicameral bone cyst' above)
- Giant cell tumor, a benign but locally aggressive skeletal tumor that occurs in young adults (see "Giant cell tumor of bone")
- Osteosarcoma, a malignant bone tumor (see "Osteosarcoma: Epidemiology, pathogenesis, clinical presentation, diagnosis, and histology")
- Osteoblastoma (in the spine) (see 'Osteoblastoma' above)
- Chondroblastoma (if they cross the growth plate) (see 'Chondroblastoma' above)

Treatment – Aneurysmal bone cysts are treated with excision, curettage, and bone grafting [70,73]. Chemical cauterization or cryotherapy may be required.

Prognosis – Aneurysmal bone cysts are locally aggressive and destructive [6]. They continue to expand until treated and may recur after excision (in 10 to 50 percent of cases) [1,5].

OTHER BONE TUMORS

Langerhans cell histiocytosis of bone — Langerhans cell histiocytosis (LCH) of bone is a benign form of LCH that is localized to bone. Patients may present with a solitary lesion (monostotic) or multiple sites of involvement (polyostotic). LCH of bone is discussed separately. (See "Langerhans cell histiocytosis (eosinophilic granuloma) of bone in children").

Giant cell tumor — Giant cell tumor of bone is a relatively rare, benign, but locally aggressive osteolytic skeletal neoplasm of young adults. It is discussed separately. (See "Giant cell tumor of bone").

SUMMARY

- Benign bone tumors often are discovered incidentally during evaluation of trauma or another condition. Symptoms of benign bone tumors, when they occur, may include localized pain, swelling, deformity, or pathologic fracture. (See 'Clinical evaluation' above.)
- Most benign bone tumors can be diagnosed with plain radiographs. Characteristic clinical and radiographic features are summarized in the table (table 1). (See 'Radiologic evaluation' above.)
- Most benign bone tumors are managed with serial examinations and radiographs. Symptomatic or aggressive benign tumors may be treated with curettage and bone grafting or excision. (See 'Management' above.)
- Benign bone tumors of childhood can be classified according to the matrix, or substance, that they produce: osteoid- or bone-forming tumors, cartilage-forming tumors, fibrous lesions, and cystic tumors. (See 'Classification' above.)
- Osteoid- or bone-forming tumors include:
  - Osteoid osteoma (image 1) (see 'Osteoid osteoma' above)
  - Osteoblastoma (image 3) (see 'Osteoblastoma' above)
- Cartilage-forming tumors include:
• Osteochondroma (image 4) and hereditary multiple osteochondromas (image 5) (see 'Osteochondroma and hereditary multiple osteochondromas' above)
• Solitary enchondroma (image 6), enchondromatosis (Ollier syndrome) (image 7), and Maffucci syndrome (image 8) (see 'Enchondroma' above)
• Periosteal chondroma (image 12) (see 'Periosteal chondroma' above)
• Chondroblastoma (image 16) (see 'Chondroblastoma' above)
• Chondromyxoid fibroma (image 17) (see 'Chondromyxoid fibroma' above)

• Fibrous lesions include:
  • Fibrous dysplasia (image 19) and polyostotic fibrous dysplasia (McCune-Albright syndrome) (see 'Fibrous dysplasia' above)
  • Ossifying fibroma (see 'Ossifying fibroma' above)
  • Nonossifying fibroma (image 22) (see 'Nonossifying fibroma' above)

• Cystic and vascular lesions include:
  • Unicameral bone cyst (image 25) (see 'Unicameral bone cyst' above)
  • Aneurysmal bone cyst (image 26) (see 'Aneurysmal bone cyst' above)

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