

ENTITLEMENT ELIGIBILITY GUIDELINES **PAGET'S DISEASE OF BONE (OSTEITIS DEFORMANS)**

MPC 01309
ICD-9 731.0

DEFINITION

Paget's Disease of Bone (Osteitis Deformans) is a disease of bone marked by repeated episodes of increased bone resorption followed by excessive attempts at bone formation, resulting in weakened and deformed bones of increased mass.

DIAGNOSTIC STANDARD

Diagnosis by a qualified medical practitioner is required. Results of x-rays and alkaline phosphatase blood test are required. A bone scan is often helpful.

ANATOMY AND PHYSIOLOGY

Paget's Disease represents an imbalance of bone formation and resorption. It typically begins with excessive bone resorption followed by excessive bone formation. The main disturbance is an exaggeration of osteoclastic bone resorption, initially producing a localized bone loss. The disorder is usually not recognized until the subsequent bone formation response is so pronounced that enlarged and deformed bones result. This excessive resorption and formation culminates at the tissue level in an abnormal mosaic pattern of lamellar bone associated with extensive vascularity and increased fibrous tissue deposition in adjacent marrow spaces.

Early in the disease process, osteolysis (loss of bone) is accompanied by some level of repair. The repair usually occurs in local areas near the regions of excessive resorption. Newly deposited bone is both woven and lamellar bone. The hematopoietic marrow, which is closely opposed to the area of increased resorption, is replaced by a hypervascular loose fibrous connective tissue. These changes occur in both cancellous and cortical bone. The typical mosaic pattern of Paget's Disease results from the abnormal deposition of lamellar bone. In some circumstances, the rate of resorption decreases, but new bone formation may continue, resulting in an increased mass of bone per unit volume which is termed the osteoblastic phase. In this phase, the bone becomes increasingly sclerotic and brittle.

Paget's Disease can affect any bone in the body, and may involve several bones at the same time. The spine, skull, pelvis, femora, and tibia are most commonly involved.

Fractures as a result of this condition are common, especially in weight-bearing long bones.

Some data suggests a significant genetic component; however, the results may be confounded by the fact that family members may be exposed to the same environmental factors.

While there is some indication of a viral etiology for Paget's Disease, there is a lack of sound medical evidence at this time to implicate any external agent, disease or environmental influence in the development of Paget's Disease of the bone.

The disease is virtually non-existent in Japan and other parts of eastern Asia.

CLINICAL FEATURES

Paget's Disease may occur in only one bone (monostotic Paget's Disease) or in multiple bones (polyostotic Paget's Disease). It varies in severity from isolated asymptomatic bone lesions to crippling deformities of multiple bones. The clinical presentation of Paget's Disease and the radiographic abnormalities may resemble some neoplasms, especially metastatic carcinoma.

Paget's Disease is rare before age 20, with most patients being older than 50 years.

Clinical presentation is dependent on which bone is involved. The most common sites of involvement include the spine, pelvis, skull, femur and tibia. If the disease is widespread, part of every bone may be involved. Typically, the disease is asymptomatic and may be discovered on a screening x-ray for other purposes. The disorder may become clinically evident when bone involvement results in pain, gross deformity, compression of roots or spinal cord, fracture of an involved bone, or alteration of joint structure and function leading to osteoarthritis or locally increased vascularity. When it affects the long bones, the overlying skin may be warm and hyperemic, possibly due to increased blood flow to the entire region.

Skull involvement may produce enlargement of the head characterized by more evident frontal bossing and dilated superficial cranial muscles. If the facial bones are involved as well, a typical leonine appearance may be noted. Conductive and/or sensorineural hearing loss may result from disease of the temporal bone or ossicles. Compression of nerve fibres may also occur. Pagetic involvement at the base of the skull may lead to basilar invagination or platybasia with consequent compression of structures in the posterior fossa, spinal cord or, rarely, cerebellar tonsillar herniation. Symptoms of this process may manifest as ataxia, weakness, or respiratory compromise.

Paget's Disease affecting the spine may produce pain directly or as a result of nerve root irritation or compression. Pressure of the spinal cord is unusual. Limb pain may be due to the lesion itself or result from nerve root compression. If the Pagetic lesion is near a joint, mechanical changes can influence the development of osteoarthritis, e.g. arthritis in the knees may stem from disease of the distal femur or patella. Hip disease due to involvement of the femoral head or acetabulum is common.

The x-ray appearance of Pagetic bone reflects the underlying process.

Radiographically, it proceeds through a purely lytic phase, a mixed lytic and blastic phase, and a blastic phase.

Pagetoid bone lacks the strength of normal bone. As a result, it deforms and fractures easily. Persons with Paget's Disease may develop primary bone malignancies in the Pagetoid bone, including osteogenic carcinomas that spread rapidly and have an extremely poor prognosis.

PENSION CONSIDERATIONS

A. CAUSES AND/OR AGGRAVATION

THE TIMELINES CITED BELOW ARE NOT BINDING. EACH CASE SHOULD BE ADJUDICATED ON THE EVIDENCE PROVIDED AND ITS OWN MERITS.

1. Inability to obtain appropriate clinical management

Excessive progressive bone resorption can be slowed by medication. Inability to obtain such management may lead to aggravation of the condition.

B. MEDICAL CONDITIONS WHICH ARE TO BE INCLUDED IN ENTITLEMENT/ASSESSMENT

C. COMMON MEDICAL CONDITIONS WHICH MAY RESULT IN WHOLE OR IN PART FROM PAGET'S DISEASE AND/OR ITS TREATMENT

- Osteoarthritis from damage to cartilage of affected joints, most commonly:
 - (1) spine - cervical osteoarthritis
 - thoracic osteoarthritis
 - lumbar osteoarthritis
 - (2) ankle
 - (3) knee
 - (4) hip
- Headaches (from skull involvement)
- Conductive and/or sensorineural hearing loss (from temporal bone involvement)
- Vertigo (from temporal bone involvement)
- Tinnitus (from temporal bone involvement)
- Neurological consequences of particular Pagetic lesions (eg: thoracic spinal cord or nerve root compression)
- Sarcoma in area of Paget's disease
- Benign tumours in area of Paget's disease
- Fractures at the site of Paget's Disease

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