What Is Paget's Disease of Bone?

Paget's disease is a chronic disorder that can result in enlarged and misshapen bones. The excessive breakdown and formation of bone tissue causes affected bone to weaken—resulting in bone pain, misshapen bones, fractures, and arthritis in the joints near the affected bones. Paget's disease typically is localized, affecting just one or a few bones, as opposed to osteoporosis, for example, which affects all the bones in the body.

Scientists do not know for sure what causes Paget's disease. In some cases, the disease runs in families, and so far two genes have been identified that predispose affected people to develop Paget's disease. In most cases, however, scientists suspect that environmental factors play a role. For example, scientists are studying the possibility that a slow-acting virus may cause Paget's disease.

Who Is Affected?

The disease is more common in older people and those of northern European heritage. Research suggests that a close relative of someone with Paget's disease is seven times more likely to develop the disease than someone without an affected relative.

What Are the Symptoms?

Many patients do not know they have Paget's disease because they have no symptoms. Sometimes the symptoms may be confused with those of arthritis or other disorders. In other cases, the diagnosis is made only after the patient has developed complications.

Symptoms can include:

- **pain**, which can occur in any bone affected by the disease or result from arthritis, a complication that develops in some patients
- **headaches and hearing loss**, which may occur when Paget's disease affects the skull
- **pressure on nerves**, which may occur when Paget's disease affects the skull or spine
- **increased head size**, bowing of a limb, or curvature of the spine, which may occur in advanced cases
- **hip pain**, which may occur when Paget's disease affects the pelvis or thighbone
- **damage to cartilage of joints**, which may lead to arthritis.

Any bone or bones can be affected, but Paget’s disease occurs most frequently in the spine, pelvis, legs, or skull. Generally, symptoms progress slowly, and the disease does not spread to normal bones.

### How Is It Diagnosed?

Paget’s disease is almost always diagnosed using x-rays but may be discovered initially by either of the following tests:

- **Alkaline phosphatase blood test.** An elevated level of alkaline phosphatase in the blood can be suggestive of Paget’s disease.

- **Bone scans.** Bone scans are useful in determining the extent and activity of the condition.

If a blood test or bone scan suggests Paget’s disease, the affected bone(s) should be x-rayed to confirm the diagnosis.

Early diagnosis and treatment are important to minimize complications. Siblings and children of people with Paget’s disease may wish to have an alkaline phosphatase blood test every 2 or 3 years starting around the age of 40. If the alkaline phosphatase level is higher than normal, a bone scan may be used to identify which bone or bones are affected and an x-ray of these bones is used to verify the diagnosis of Paget’s disease.

### What Is the Prognosis?

The outlook for people diagnosed with Paget’s disease is generally good, particularly if treatment is given before major changes have occurred in the affected bones. Treatment can reduce symptoms but is not a cure. Osteogenic sarcoma, a form of bone cancer, is an extremely rare complication that occurs in less than 1 percent of all patients with Paget’s disease.

### Who Treats It?

The following types of medical specialists are generally knowledgeable about treating Paget’s disease:

- **Endocrinologists.** Doctors who specialize in hormonal and metabolic disorders

- **Rheumatologists.** Doctors who specialize in joint and muscle disorders

- **Others.** Orthopaedic surgeons, neurologists, and otolaryngologists (doctors who specialize in ear,
nose, and throat disorders) may be called on to evaluate specialized symptoms.

How Is It Treated?

Drug therapy. The Food and Drug Administration has approved several medications to treat Paget’s disease. The medications work by controlling the excessive breakdown and formation of bone that occurs in the disease. The goal of treatment is to relieve bone pain and prevent progression of the disease. People with Paget’s disease should talk to their doctors about which medication is right for them. It is also important to get adequate calcium and vitamin D through diet and supplements as prescribed by your doctor, except for patients who have had kidney stones.

Bisphosphonates are a class of drugs used to treat a variety of bone diseases. Several bisphosphonates are currently available to treat Paget’s disease. Calcitonin is a naturally occurring hormone made by the thyroid gland. The medication may be appropriate for some patients.

Surgery. Medical therapy before surgery helps decrease bleeding and other complications. Patients who are having surgery should discuss pretreatment with their doctor. Surgery may be advised for three major complications of Paget’s disease:

- **Fractures.** Surgery may allow fractures to heal in better position.
- **Severe degenerative arthritis.** Hip or knee replacement may be considered if disability is severe and medication and physical therapy are no longer helpful.
- **Bone deformity.** Cutting and realigning pagetic bone (a procedure called an osteotomy) may reduce the pain in weight-bearing joints, especially the knees.

Complications resulting from enlargement of the skull or spine may injure the nervous system. However, most neurological symptoms, even those that are moderately severe, can be treated with medication and do not require neurosurgery.

Diet and exercise. There is no special diet to prevent or help treat Paget’s disease. However, according to the Institute of Medicine of the National Academy of Sciences, women age 50 and older and men age 70 and older should get 1,200 mg of calcium and at least 600 IU (International Units) of vitamin D every day to maintain a healthy skeleton. People age 70 and older need to increase their vitamin D intake to 800 IU. People with a history of kidney stones should discuss calcium and vitamin D intake with their doctor.

Exercise is important because it helps preserve skeletal health, prevent weight gain, and maintain joint mobility. Patients should discuss any new exercise program with their doctor before beginning, to avoid any undue stress on affected bones.

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For Your Information

This publication contains information about medications used to treat the health condition discussed here. When this publication sheet was developed, we included the most up-to-date (accurate) information available. Occasionally, new information on medication is released.

For updates and for any questions about any medications you are taking, please contact the Food and Drug Administration toll free at 888–INFO–FDA (463–6332) or visit its website at www.fda.gov. For additional information on specific medications, visit Drugs@FDA at www.accessdata.fda.gov/scripts/cder/drugsatfda. Drugs@FDA is a searchable catalog of FDA-approved drug products.

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