



Information for Patients About Paget's Disease of Bone

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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?

An estimated 1 million people in the United States have Paget's disease, or about 1.3 people per 100 men and women age 45 to 74. The disease is more common in older people and those of northern European heritage. Men are about twice as likely as women to have the disease. 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.

What Other Medical Conditions May It Lead to?

Paget's disease may lead to other medical conditions, including:

- **Arthritis** – Long bones in the leg may bow, distorting alignment and increasing pressure on nearby joints. In addition, pagetic bone may enlarge, causing joint surfaces to undergo excessive wear and tear. In these cases, pain may be caused by a combination of Paget's disease and osteoarthritis.
- **Hearing loss** – Loss of hearing in one or both ears may occur when Paget's disease affects the skull and the bone that surrounds the inner ear. Treating Paget's disease may slow or stop hearing loss. Hearing aids also may help.
- **Heart disease** – In severe Paget's disease, the heart works harder to pump blood to affected bones. This usually does not result in heart failure except in some people who also have hardening of the arteries.
- **Kidney stones** – Kidney stones are more common in patients with Paget's disease.
- **Nervous system problems** – Pagetic bone can cause pressure on the brain, spinal cord, or nerves and reduced blood flow to the brain and spinal cord.
- **Sarcoma** – Rarely, Paget's disease is associated with the development of a malignant tumor of the bone. When there is a sudden onset or worsening of pain, sarcoma should be considered.
- **Loose teeth** – When Paget's disease affects the facial bones, the teeth may loosen. This may make chewing more difficult.
- **Vision loss** – Rarely, when the skull is involved, the nerves to the eye may be affected, causing some loss of vision.
- Paget's disease is not associated with osteoporosis. Although Paget's disease and osteoporosis can occur in the same patient, they are completely different disorders. Despite their marked differences, several medications for Paget's disease also are used to treat osteoporosis.

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. Of the six bisphosphonates currently available to treat Paget’s disease, the most commonly prescribed are the four most potent: Actonel,¹ Fosamax, Aredia, and Reclast. Didronel and Skelid may be appropriate therapies for selected patients, but are less commonly used. People with severe kidney disease should not use any of these drugs.

- **Actonel (risedronate sodium)** – tablet; 30 mg (milligrams) once daily for 2 months; patients should wait at least 30 minutes after taking before eating any food, drinking anything other than tap water, taking any medication, or lying down (patients may sit).
- **Fosamax (alendronate sodium)** – tablet; 40 mg once daily for 6 months; patients should wait at least 30 minutes after taking before eating any food, drinking anything other than tap water, taking any medication, or lying down (patients may sit).
- **Aredia (pamidronate disodium)** – intravenous; approved regimen 30 mg infusion over 4 hours on 3 consecutive days; more commonly used regimen 60 mg over 2 to 4 hours for 2 or more consecutive or nonconsecutive days. Generic pamidronate disodium for injection is also available. People with low levels of blood calcium or vitamin D should not be treated with Aredia until these levels have been corrected.

¹ Brand names included in this fact sheet are provided as examples only, and their inclusion does not mean that these products are endorsed by the National Institutes of Health or any other Government agency. Also, if a particular brand name is not mentioned, this does not mean or imply that the product is unsatisfactory.

- **Reclast (zoledronic acid)** – Intravenous; approved regimen 5 mg infusion given over 15 minutes. People with low levels of blood calcium or vitamin D should not be treated with Reclast until these levels have been corrected. Zoledronic acid used for treating Paget’s disease outside the United States is known as Aclasta; when used for certain cancer treatments, it is called Zometa.
- **Didronel (etidronate disodium)** – tablet; approved regimen is 200 to 400 mg once daily for 6 months; the higher dose (400 mg) is more commonly used; no food, beverages, or medications for 2 hours before and after taking; course should not exceed 6 months, but repeat courses can be given after rest periods, preferably of 3 to 6 months.
- **Skelid (tiludronate disodium)** – tablet; 400 mg (two 200 mg tablets) once daily for 3 months; may be taken any time of day, as long as there is a 2-hour period before and after eating, drinking, and taking medications.

Calcitonin is a naturally occurring hormone made by the thyroid gland. The medication may be appropriate for certain patients but is less effective than bisphosphonates and seldom used. The nasal spray form of this medication is not approved for the treatment of Paget’s disease.

- **Miacalcin (salmon calcitonin)** – administered by injection; 50 to 100 units daily or three times per week for 6 to 18 months; repeat courses can be given after brief rest periods.

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 National Academy of Sciences, everyone age 50 and older should get 1,200 mg of calcium and at least 400 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 600 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.

Resource

For more information about Paget's disease, contact:

The Paget Foundation
120 Wall Street, Suite 1602, New York, NY 10005
Tel: 212-509-5335 or 800-23-PAGET (free of charge)
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For Your Information

This publication contains information about medications used to treat the health condition discussed here. When this fact sheet was printed, 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 at 888-INFO-FDA (888-463-6332, a toll-free call) or visit its Web site at www.fda.gov.

For updates and questions about statistics, please contact the Centers for Disease Control and Prevention's National Center for Health Statistics toll free at 800-232-4636 or visit its Web site at www.cdc.gov/nchs.

Recognizing the National Bone and Joint Decade: 2002–2011