

## Paget's disease

Paget's disease of the bone is the most common form of bone inflammation, or osteitis. The bones become weakened, soft and abnormally enlarged. Any part of the skeleton can be affected, but the most common sites include the skull, spine, pelvis, thigh bone, shin and the bone of the upper arm.

Paget's disease tends to affect people over the age of 40 years, with around three per cent of this age group affected. The rate increases to about 10 per cent in people over 80 years of age. Men are more commonly affected than women.

The cause of Paget's disease is not yet fully understood. Some theories suggest people with a genetic predisposition to the disorder may become infected with an unidentified virus, which then triggers the bone abnormalities.

Paget's disease of the bone is often confused with an unrelated skin disease that also bears Paget's name and is usually associated with an underlying cancer.

### **Bone growth mysteriously accelerates**

Healthy bone tissue is maintained by cells called osteoblasts and osteoclasts. The osteoblasts build new bone, while the osteoclasts help to dispose of old bone.

In a person with Paget's disease, the balance between these two groups of cells is disturbed. The osteoblasts become overactive and too much bone tissue is produced, leading to enlargement. The abnormal growth means that the new bone tissue is weak and unstable.

The reason for this accelerated bone growth is unknown. Genetic and environmental factors are suspected. It appears that an unidentified virus may trigger the disease in those people with a family history.

### **Risk factors**

The cause of Paget's disease is unknown, but the risk factors seem to include:

- **Race** - people of Anglo-Celtic background are more likely to develop the disorder, particularly those living in Britain, Australia and Germany.
- **Age** - the disorder becomes more common with increasing age.
- **Genetics** - there is a family history in up to 30 per cent of cases.

### **There may be no symptoms**

In 95 per cent of cases, a person with Paget's disease presents no symptoms. In other cases, the symptoms may develop gradually over a long period of time or may quickly become severe and debilitating. Pain is often caused by the enlarged bone putting pressure on surrounding nerves.

Symptoms may include:

- Stiffness in the joints
- Constant fatigue
- Painful aching in the bones
- Aches and pains becoming more acute during the night
- The bone appears bent or thickened
- The affected site is significantly warmer than the rest of the body
- Bowing of the leg bones (if the legs are affected)
- Headache and increased head size if the skull is involved
- Bone fractures.

## Possible complications

Paget's disease can lead to further complications, including:

- **Osteoarthritis** - a form of arthritis that can occur in joints nearby to bones affected by Paget's disease.
- **Broken bones** - bones that tend to break of their own accord.
- **Deafness** - caused by pressure on the auditory nerves.
- **Disorders of the inner ear** - such as tinnitus (ringing in the ears) or vertigo.
- **Numbness or paralysis** - caused by deteriorating vertebrae pinching the nerves in the spinal cord.
- **Increased workload on the heart** - due to an increased number of blood vessels in the affected bones, leading to increased blood flow through the bones. This usually does not result in heart failure except in some people who already have heart disease.
- **Cancer of the affected bone tissue** - occurs in about one per cent of cases.

## Diagnosis is often accidental

Paget's disease is often discovered by accident during x-rays taken for some other reason. The diagnosis can be confirmed by further x-rays, bone scans or by a particular blood test that checks for an enzyme crucial to bone growth, called alkaline phosphatase.

## Treatment

There is no cure for Paget's disease. There are treatments available to ease some of the symptoms. Treatment options include:

- **Drugs acting on the bones** - such as bisphosphonates and calcitonin (used rarely), to slow the progression of the disorder.
- **Aspirin and non-steroidal anti-inflammatory drugs (NSAIDs)** - to alleviate pain.
- **Calcium and vitamin D** - a diet rich in calcium, safe levels of sun exposure to ensure adequate vitamin D levels or supplements of both may be prescribed for overall bone health. It is best to discuss your calcium and vitamin D requirements with your doctor; high amounts of calcium in the blood are common in Paget's disease and need to be carefully monitored to avoid causing other medical problems.
- **Surgery** - to relieve pinched nerves or bone fractures, or to replace a joint severely affected by arthritis.
- **Exercise** - to maintain skeletal health and maintain mobility in the joints and strength in the surrounding muscles. However, as bones are weaker and more likely to fracture, certain forms of exercise will be inappropriate for those with Paget's disease. It is best to consult a physiotherapist for an individually prescribed exercise program.

## Paget's can also be a skin disease

Paget's name is also given to a skin disease. Paget's disease of nipple, breast, armpit or anal or genital areas is a skin disease. It is usually associated with an underlying cancer.

The symptoms include redness and crusting of the skin, and may be similar to dermatitis or eczema. Diagnosis of Paget's disease of the skin may require a biopsy. Treatment may include surgery, radiation therapy or chemotherapy.

## Where to get help

- Your doctor
- Endocrinologist - specialist in hormonal and metabolism disorders
- Rheumatologist - specialist in joint and muscle disorders

## Things to remember

- Paget's disease of the bone causes the abnormal enlargement and weakening of bone.
- Commonly affected sites include the skull, pelvis, spine and the long bones of the arm and thigh.
- The cause is not fully understood.
- Paget's disease of the bone should not be confused with an unrelated skin disease that also bears Paget's name

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## Arthritis Victoria incorporating Osteoporosis Victoria

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