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The website for the answer to all your Orthopaedic Questions

- **Orthopaedic Opinion Online** is a website designed to provide information to patients who have orthopaedic and musculoskeletal problems and are undergoing treatment.
- **Patient information** is provided in the form of downloadable information sheets.
- **Orthopaedic advice** and second opinions can be provided by our expert internationally renowned Consultant Orthopaedic Surgeons.
- **Online review** of patients' X rays or MRI scans can also be provided and any proposed treatment plans reviewed.
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Paget's Disease of Bone

Paget's disease is a condition that affects the integral structure of one or more bones. Most commonly this occurs in the bone of the leg resulting in bone pain. Affected bones are very hard but brittle and may become deformed. Various treatment modalities are available including Biphosphonate medication.

Normal bone metabolism and control

Bone is a living tissue. There are two main types of cells within the bones. One type called osteoblasts makes and lays down bone material. Another type called osteoclasts resorbs or dissolves bone. These cells are active throughout life. They work in a balanced way to make and mould bone, repair damage and keep the bone structure in tune with the structural demands experienced by the activity of the person. There is a natural slow but constant turnover of bone.

In a situation of inactivity or aging the osteoclasts slowly dominate to reduce the amount of chemical substance in the bones. This is similar to the process which produces osteoporosis or a weak structure in the bone. It is not similar to osteomalacia with which it is often confused. In Osteomalacia there is a deficiency of Vitamin D or calcium which results in a failure of the bone structure to be properly mineralised and hard bone. Thus osteomalacia is a qualitative deficiency of bone mineral rather than the quantitative deficiency of mineral structure which occurs in osteomalacia.

In the situation of injury, bone fracture or unaccustomed activity or sports training, the osteoblasts become more active and dominate laying down additional mineral structure in the bone which results in strengthening of the bones in response to the increased activity.

Paget's disease

Sir James Paget in 1877 first described this condition of bone, which since has been known as Padget's disease. In Paget's disease the bone turnover becomes out of control and accelerates and gets faster. Affected areas of bone form in an abnormal way. The newly formed bone is thicker and the bone may be larger, expanded and appear wider. However, the bone is not made with the proper structural arrangement and as a result is weak. This can cause a slow or progressive deformity of the affected bone, particularly weight bearing bones such as the tibia or femur bones that may slowly bend. The affected bone is also brittle and is also more likely to fracture or break.

Aetiology of Paget's disease

A problem with the osteoclasts (the bone forming cells) is thought to be the root cause of the imbalance. In affected areas of bone there are abnormal numbers of osteoclasts which are often bigger than normal. It is

thought that these are wrongly programmed in the way they mould bone. More bone than normal is resorbed or dissolved by the abnormal osteoclasts but this bone is then replaced by bone that is wrongly woven. This results in increased bone turnover and badly structured areas of bone. How the osteoclasts in a local area of bone become 'out of control' is not clear. Some evidence suggests that they become infected with a virus which results in some alteration.

There also seems to be some genetic factor in the condition. Up to 40% of people with Paget's disease have a close relative (parent, mother, brother, sister) who also develop the disease. It may be that the genetic make up makes you prone to this disease but it is then triggered by a certain virus.

Paget's disease mainly affects people aged over 40 and becomes more common with increasing age. Men are affected 3 times more often than women. One study found that up to 8% of people in the UK over the age of 55 had some X-ray appearance of Paget's disease on one or more of their bones. However, most people with Paget's disease have no symptoms at all and are unaware that they have the condition. It is much more common in some parts of the world including the UK, as compared to others. The condition is rare in Asia and the Far East.

One or more bones may be affected. The bones most commonly affected are the pelvis, the femur or thigh, the tibia or shin bone and the vertebrae of the spine.

Symptoms
Padget's disease usually starts in one point on a bone. It then gradually spreads along the bone at up to 12mm per year. Whilst 90% of people with Paget's disease have no symptoms, the condition may lead to one or more of the following problems.

- **Pain -** is the commonest symptom if symptoms occur. The pain is typically a deep seated ache of the bone that is usually not affected by rest or exercise. It is commonly worse at night. It may be mild but can become severe. Shooting pains from the affected area may also occur.

- **Deformity** - may occur depending on the site of the disease and the size of the bony overgrowth. Bowing of the upper leg is the most common when the femur or thigh bone is affected. The shin or tibia is also commonly affected. Another example is an odd shape to the head that may develop if the skull is affected.

- **Fracture** - affected bones are more liable to break. A fracture after a minor fall or injury may be the first indication that Paget's disease has developed.

- **Nerve compression -** abnormal overgrowth of a bone may press on nearby nerves. This can cause a variety of symptoms. The commonest is deafness of one ear due to pressure on the auditory or inner ear nerve going through an affected skull bone. Other possible problems include nerve pain and weakness of muscles supplied by a compressed nerves from the spine.

- **Arthritis -** may occur if an affected bone is next to a joint. Alternately mechanical deformity produces accelerated wear of an affected joint which leads to premature arthritis.

- **Metabolic problems -** the abnormal turnover of bone may cause high blood levels of various minerals. This may rarely lead to problems such as gout, high calcium levels and kidney problems. Alternately the high blood flow through the bone which results can lead to cardiac overload. Rarely, a bone cancer develops in an affected bone.

Diagnosis
- **X-ray -** a bone affected by Paget's disease can usually be noticed quite easily on an X-ray. The X-ray may be done if the disease is suspected by your doctor, but more often, it is seen by chance when an X-ray is done for another reason.

- **Bone scan -** this may be done if an area of Paget's disease is found in one bone. This test involves an injection of a radioisotope which is particularly designed to be taken up into active bone. A scan then detects the level of radioactivity coming from an affected bone. Therefore this test can detect which bones are affected throughout the body. The dose of radioactivity involved in this test is very small.

- **Blood tests –** These can easily measure a chemical in the blood which is associated with increased bone turnover. The Alkaline-Phoshatase level is elevated in people with a high bone turnover. The level of this chemical gives an indication of the activity of the disease, and how extensive it is throughout the body. The test is also used to monitor the effect of treatment: The level falls when treatment is effective and bone turnover falls.

Treatment
The main aims of treatment are to reduce pain and to prevent complications.

No treatment
This may be an option if the disease was found by chance and is not causing symptoms. However, some people with no symptoms are offered treatment if it is likely that the progression of the disease will lead to problems. For example, if an area of Paget's disease is found in an important part of the skeleton such as at the base of the skull or hip region. Progression of the disease here if left untreated may result in compression of nerves, deafness or in the case of the hip; arthritis.

Treatment
The treatment of Paget's disease has improved greatly over the last 10-15 years.

- **A biphosphonate drugs** - is now the usual treatment. These medicines include Etidronate, Tiludronic acid, Risedronate and Pamidronate. They work by interfering with the control of the osteoclast bone cells. These medicines reduce the abnormal bone turnover and cause any new bone formation is more normal in structure. They have an effect on existing disease to reduce pain although this may take several months of treatment. Treatment is good at preventing further progression of the disease. Like all medicines, they may cause side effects but generally they are well tolerated. With these medicines the outlook for people with Paget's disease much better than in the past, with control of the disease readily being maintained.

- **Calcitonin –** This drug also reduces bone turnover. It was used before biphosphonates were developed. It is less effective than these newer medicines and is therefore now not generally the preferred treatment.

- **Painkillers -** may also be needed if the disease is causing pain.

- **Surgery –** Surgery such as structural straightening of the bone may be needed if the disease has caused an ugly or mechanically unacceptable deformity. Joint replacement surgery is commonly necessary for a joint affected by the condition or one that becomes arthritic as a result of the condition in a nearby bone.

Summary

- Paget's disease is common in the UK.
- Many people have no symptoms and it is often found by chance.
- Progression of the disease may lead to bone pain, deformity and complications such as fractures and nerve compression.
- Biphosphonate drugs may prevent progression of the disease and in time may ease pain from existing areas of affected bone
- Other treatments such as painkillers and surgery also may be needed.

Further information
National Association for the Relief of Paget's Disease
323 Manchester Road, Walkden, Worsley, Manchester, M28 3HH
Tel: 0161 799 4646 Web: www.paget.org.uk