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Paget's disease of bone

Paget's disease of bone affects the structure of parts of one or more bones. In most cases it causes no symptoms and does not need treatment. Symptoms develop in some cases and pain in the affected part of bone is the most common symptom. Affected bones are weaker than normal and may become deformed and may break (fracture) more easily than normal. Treatment with a bisphosphonate medicine is used in some cases to ease pain and to help treat complications.

Understanding normal bone

Bone is a living tissue. There are two main types of cells within the hard bone material. One type, called osteoblasts, makes and lays down bone material. Another type, called osteoclasts, dissolves (resorbs) particles of 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 correctly woven. There is a slow but constant turnover of bone.

What is Paget's disease of bone?

Paget's disease of bone was first described by Sir James Paget in 1877. (It is a different condition to Paget's disease of breast.)

This leaflet is only about Paget's disease of bone. The condition will be called 'Paget's disease' for the rest of this leaflet.

In Paget's disease, the bone-making process (bone turnover) becomes faster and out of control. Affected areas of bone form new bone material in an abnormal way. The newly formed bone is thicker than normal and the bone may become wider. However, the bone is not made properly and it is weaker than normal bone. This can cause deformity of the affected bone, particularly in weight-bearing bones such as the leg bones, which may bend. Affected bone is also more likely to break (fracture).

What causes Paget's disease of bone?

The underlying cause is a problem with the osteoclast bone cells. In affected areas of bone there are abnormal osteoclasts which are bigger than normal and more in number. It is thought that these are wrongly programmed in the way they mould bone. More bone than normal is dissolved (resorbed) by the abnormal osteoclasts. As a response to this, the osteoblasts increase in activity to make new bone material. But this increase in bone turnover leads to badly structured areas of bone that are wrongly woven.

How the osteoclasts in a localised area of bone become out of control is not known. One theory is that they may become infected with a virus which alters them in some way. There also seem to be some genetic factors. Your genetic makeup is the material inherited from your parents which controls various aspects of your body. Around 15 in 100 people with Paget's disease have a close relative (parent, mother, brother, sister) who also develops the disease.

Who develops Paget's disease of bone?

It mainly affects people aged over 50 years. The disease affects three men for every two women. It becomes more common with increasing age. Around 5 in 100 women and 8 in 100 men in their 80s are thought to have some degree of Paget's disease. However, most people with Paget's disease have no symptoms and will not be aware they have the condition. The UK has the highest rate of Paget's disease in the world. It is not clear exactly why this is but it may be due to the genetic and other factors discussed above.

What are the symptoms of Paget's disease of bone?

One or more bones may be affected. The bones most commonly affected are:

- The pelvis.
- The thigh bone (femur).
- The bones (vertebrae) of the spine.

- The skull.
- The shin bone (tibia).

Other bones are less commonly affected. The abnormality of bone formation starts in one point on a bone. It can then gradually spread along the bone. This may lead to one or more of the following.

In many cases there are no symptoms

More than 9 in 10 people with Paget's disease have no symptoms. The abnormal structure of bone is localised and may never cause any problems. The disease is commonly found by chance when an X-ray is taken for another reason.

Pain

This is the most common symptom if symptoms do occur. The pain is typically a deep-seated ache of the bone, which can be present both at rest and on 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

A deformity may develop, depending on the site of the disease and the size of the affected part of the bone. Bowing of the upper leg is the most common and can happen if the femur is affected. Bowing of the tibia can lead to a deformity known as sabre tibia. Another example is an odd shape to the head that may develop if the skull is affected. The skull can become enlarged and you may develop a prominent, wide forehead.

Fractures

Affected bones are more liable to break (fracture). 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. For example, neuralgic pain (a different type of pain to the more common bone pain of Paget's disease). Another quite common symptom if the skull is affected is deafness in one ear. This is caused by pressure on the ear nerve that passes through an affected skull bone.

Other possible problems include weakness of muscles supplied by a compressed nerve, or numbness of an area of skin supplied by a compressed nerve.

Joint inflammation (arthritis)

This may develop if an affected section of bone is next to a joint.

Some rare complications of Paget's disease of bone

The following are some possible, but rare, complications.

Heart failure

If large areas of bone throughout the body are affected then you need a lot of blood flow to the affected bone. This can cause problems for the heart, as it needs to deliver this extra blood flow and so [heart failure may develop](#).

Vascular steal syndrome

If the skull is extensively affected, the extra blood flow needed to supply the enlarged skull may be 'stolen' from the blood supply going to the brain. This may result in you becoming tired and listless. A similar effect can occur if a large area of spine is involved. Blood flow to the spinal cord may be 'stolen' and diverted to the affected bone. This can cause various symptoms such as numbness and weakness of parts of the body supplied by nerves branching from the affected part of the spinal cord.

Bone cancer

Rarely, abnormal bone turns cancerous. This is thought to occur in less than 1 in 1,000 people with Paget's disease. [Bone cancer may be suspected](#) if you develop worsening pain or swelling over an affected area of bone.

Other

The high bone turnover may cause high blood levels of various chemicals. Rarely, this leads to problems such as gout or a high blood calcium level which can cause various symptoms.

How is Paget's disease of bone diagnosed?

- **X-ray.** A bone with Paget's disease can usually be seen quite easily [on an X-ray](#). The X-ray may be done if the disease is suspected by your doctor (if you have pain, etc). More often, it is seen by chance when an X-ray is done for another reason.
- **Bone scan.** If an area of Paget's disease is found in one bone, [a bone scan may be done](#). The test involves an injection of a very small amount of a radioactive chemical (radioisotope). The radioisotope is taken up into bone but is much more readily taken up by areas affected by Paget's disease which have a high bone turnover. A scan then detects the level of radioactivity coming from each bone. High levels of radioactivity are found in affected bones. Therefore, this test can detect which bones are affected throughout the body. The radioactive chemical passes out of the body after a short time.
- **Blood tests.** A chemical in the blood called alkaline phosphatase can be measured in a blood sample. Levels rise with increasing bone turnover. The level of alkaline phosphatase gives some idea of the activity of the disease and how extensive it is throughout the body. This blood test is also useful to see how well treatment is working. The level falls when treatment is working and the rate of bone turnover falls. The blood test may be advised every so often if treatment is given, to check that treatment is continuing to work.

In most cases, the above investigations are enough to be able to diagnose Paget's disease. Rarely, [a sample \(biopsy\) of bone](#) is needed if the diagnosis is uncertain.

What is the treatment for Paget's disease of bone?

In many people with Paget's disease, it is found by chance, does not cause symptoms, and treatment may not be needed. Treatment may be advised if symptoms such as pain or bone deformity develop, or if complications develop. Also, treatment may be advised if you do not have symptoms but Paget's disease is in a site where it may cause problems as the disease progresses. For example, if you have an area of Paget's disease at the base of your skull. Progression of the disease here, if left untreated, may result in compression of the ear nerve and cause deafness.

Your specialist will advise on whether treatment is necessary in your case.

Bisphosphonate medicines

If treatment is advised, a [bisphosphonate medicine](#) is the usual treatment given. There are several types which include etidronate, pamidronate, [risedronate](#) and [zoledronic acid](#). Bisphosphonates work by reducing the abnormal bone turnover. This means that any new bone formed is more normal in structure. Bisphosphonates have an effect on existing disease to reduce pain, although this may take several months of treatment. In theory, it is also thought that bisphosphonates may help to prevent further progression of the disease and also complications.

A course of treatment usually lasts several weeks or months. This often has a knock-on effect for several months or years. Most of the bisphosphonates are taken as tablets; however, pamidronate and zoledronate are given by injection.

If you take bisphosphonate tablets, it is vital that you take them exactly as instructed. Read the leaflet that comes with the tablets for details on how to take the tablets. For example, one instruction will be to take the tablets on an empty stomach. This is because the absorption of bisphosphonates from your gut is affected by certain foods.

Like all medicines, bisphosphonate medicines may cause side-effects but generally they are well tolerated and work well. The leaflet that comes with the tablet packet gives a full list of possible side-effects. Tell your doctor if you have side-effects when taking a bisphosphonate, as an alternative one may be better.

One potential problem to consider with bisphosphonates

Bone destruction (osteonecrosis) of the jaw is a rare complication that affects a small number of people who are treated with a bisphosphonate. The main symptoms of osteonecrosis of the jaw include pain, numbness or a heaviness of the jaw, gum infections and loosening of teeth. It may eventually lead to disfigurement of the jaw. Osteonecrosis of the jaw mostly occurs in people who have been treated with a bisphosphonate for a number of years. The risk is increased if you are on chemotherapy as a treatment for cancer, if you are on steroid treatment, or if you have dental treatment.

Therefore:

- It is best to have any dental treatment done before you commence treatment with a bisphosphonate.
- If you are being treated with a bisphosphonate, tell your doctor or dentist as soon as possible if you develop any gum, tooth or jaw symptoms.
- Before having any dental work, tell your dentist that you are taking a bisphosphonate.
- Maintain good oral and dental hygiene.

Other treatments

Various other treatments may be advised, depending on your circumstances – for example:

- **Painkillers.** The painkillers used may be different for bone pain and neuralgic pain – the two different types of pain which may occur.
- **Calcium and vitamin D supplements** are needed by some people with Paget's disease if they are thought to have low levels of these.
- **Walking sticks and shoe raises** may be needed if there are specific problems such as a shorter limb or limb deformity due to Paget's disease.
- **Calcitonin** is a medicine that also reduces bone turnover. It was used before bisphosphonates were developed. It is less effective than these newer medicines and is not used so much now.
- **Surgery** may sometimes be needed if the disease has caused deformity, a break (fracture) or damage to a joint. For example, joint replacement surgery may be an option if a bone near to a joint is affected and has caused bad arthritis.

Further reading

- [Ralston SH, Langston AL, Reid IR](#); Pathogenesis and management of Paget's disease of bone. *Lancet*. 2008 Jul 12;372(9633):155–63.
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Authored by:	Peer Reviewed by: Dr Adrian Bonsall, MBBS	
Originally Published: 19/11/2023	Next review date: 24/02/2017	Document ID: doc_4582

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