

PAGET'S DISEASE: THE SIMPLE FACTS

This information is intended to offer basic information regarding Paget's disease of bone. It provides sufficient information to aid discussion regarding management of Paget's disease. Further information can be obtained from the Paget's Association (details at the end of this leaflet).

What is Paget's disease?

- Paget's disease is a bone disorder in which the normal repair process is disrupted. It often causes no symptoms and may be found by chance.
- For those with symptoms these may include pain, deformity and fracture.
- Occurring more commonly with advancing age, it rarely occurs in those under 50.
- In the UK, it presents in approximately 8% of men and 5% of women, by the age of 80 years.
- Over the past 25 years both the prevalence and severity of Paget's disease has decreased.
- Either single or multiple bones may be affected with common sites being the spine, skull, pelvis and thigh (femur).
- Treatment with drugs (bisphosphonates) will control the disease.

How does Paget's disease affect bone?

Bone is constantly renewed by cells called osteoclasts and osteoblasts. This is known as bone remodelling. Osteoclasts breakdown old damaged bone and osteoblasts lay down new bone. Under normal circumstances the amount of bone removed is balanced by the amount of bone laid down. In Paget's disease the osteoclasts are larger than usual and break down bone more rapidly. The osteoblasts respond to this by depositing new bone at an increased rate. This activity leads to an increase in bone size which has an irregular and weakened structure.

Causes

- There is still some uncertainty about the causes of Paget's disease but there is a general understanding that it is due to a combination of inheritance and environmental influences.
- It is thought that genetic factors account for about 86% of the risk of developing Paget's disease.
- Exposure to certain viruses and other environmental factors may also influence the development of Paget's disease.

Common symptoms

- Pain in the affected bone is the most notable symptom.
- Pain is not relieved by rest or exercise and may be most severe at night.

- A fracture after minor injury to an affected bone could be the first indication of Paget's disease
- Deformity may develop, depending on the site and severity of the disease e.g. if the shin (tibia) is involved this may become bowed and if the skull is affected this can become enlarged

How is Paget's disease diagnosed?

- A simple blood test may show an increased alkaline phosphatase level that could indicate Paget's disease
- X-ray
- An isotope bone scan is used to determine the extent and activity of the disease

TREATMENT

- **It is important that you are seen by a hospital consultant for a full assessment of your Paget's Disease**
- In many cases Paget's disease is found by chance, does not cause any symptoms and requires no treatment
- Treatment may be recommended if the affected bones are painful
- Sometimes treatment is given if Paget's disease affects a site that might be expected to cause complications such as the skull or a weight bearing bone, although it is not yet known if complications can be prevented by treatment

Treatment with Bisphosphonates

Bisphosphonates are a group of drugs that reduce abnormal bone turnover, leading to a more normal bone structure. They control existing disease and help to reduce pain caused by active disease. Bisphosphonates have a long lasting effect so that a course of treatment may last for months or years. They are either given as tablets or directly into the bloodstream (intravenous), sometimes together with calcium and vitamin D supplements, as adequate intake is necessary for the treatment to work.

Intravenous Preparations

- Zoledronate (i.e. Aclasta): a single dose given into bloodstream may be effective for up to 6 years
- Pamidronate (i.e. Aredia): several doses given into bloodstream, repeated when necessary dependant on symptoms

Treatment is usually given as a hospital outpatient. These drugs may cause "flu like" symptoms 24-48 hours after treatment. It is important to notify your GP or hospital should lasting side effects occur.

Oral Preparations

- Risedronate (i.e. Actonel) is the most commonly used oral preparation: one tablet daily for 2 months

Prior to starting treatment it is essential to read the instructions on how to take the tablets. They need to be taken whilst fasting, and it is important not to lie down for at least 30 minutes after taking them. There are a number of possible effects to be aware of. These include heartburn, stomach discomfort, and on rare occasions, joint pains and skin irritation. Should side effects occur it is important to notify your GP. Further treatment may be given after 6 months.

Other Medication

- Painkillers may be required.

Surgery

- As Paget's disease near a major joint may increase the risk of osteoarthritis, joint replacement surgery may be required
- If a fracture cannot be stabilised with a plaster cast, surgery may then be required
- Surgery is occasionally required to correct marked deformity

Further information

The Paget's Association: Here to help

The Paget's Association is the only UK charity to focus solely on Paget's disease of bone. It acts as a resource for those with the condition, the public and health professionals, offering high quality information and support when necessary.

Information

Information available from the Paget's Association includes newsletters and booklets. Information is also available via the Association's website: www.paget.org.uk

Information booklets available:

Paget's Disease -The Facts

Paget's Disease & Pain

Paget's Disease – Investigations Explained





Paget's News

Our quarterly newsletter contains useful information regarding Paget's disease. To receive this and access other services provided by the Paget's Association you can join as a member either online or by post.

Membership of The Paget's Association

To join the Paget's Association call 0161 799 4646 and request an application form or register free online at www.paget.org.uk

The Paget's Helpline

The Paget's Association has a Helpline managed by an experienced Registered Nurse.

Email: helpline@paget.org.uk

Write Telephone: 07713568197 or call our offices on 0161 799 4646 and ask to speak to the Specialist Paget's Nurse.

Write to: The Specialist Paget's Nurse, Suite 5, Moorfield House, Moorside Road, Swinton, Manchester, M27 0EW.

General and membership Enquires

Please call the office on 0161 799 4646

Paget's Support Network

The Paget's Support Network is available free to members of the Paget's Association, enabling communication by telephone, email or letter, with others who have Paget's disease. Contact the Association using the details above.

Paget's Online Forum

You can share your experiences or ask a question via the Paget's Online Forum. This can be accessed either from our website www.paget.org.uk or www.healthunlocked.co.uk



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Paget's Association

UK Guidelines

The Paget's Association is producing Guidelines for Paget's Disease – this leaflet will be reviewed in line with them, when they become available. (April 2016)

Whilst this information is intended to offer you information on Paget's disease, it is not designed to replace specific guidance you may receive from a health professional, with respect to your individual care.

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