Paget’s disease of bone

This booklet provides information and answers to your questions about this condition.
What is Paget’s disease of bone?

Paget’s disease of bone affects the way that bone develops and renews itself, causing the affected bone to become weaker than normal. In this booklet we’ll explain what Paget’s disease is, what causes it and how it’s diagnosed. We’ll also look at what can be done to treat it and suggest where you can find out more about Paget’s disease.

At the back of this booklet you’ll find a brief glossary of medical words – we’ve underlined these when they’re first used in the booklet.
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What is Paget’s disease of bone?
Paget’s disease of bone affects the way that bone is renewed and repaired. These processes are accelerated and chaotic, resulting in the production of bone that’s abnormal in shape and structure. The affected bone is weaker than normal, can become bent or deformed and may break more easily. Paget’s disease can also cause damage to the joints close to the affected bone.

What are the symptoms?
Many people have no symptoms at all, but the most common one is pain, particularly in:

- the affected bone
- the joints near the affected bones
- the back, neck and legs, caused by an enlarged bone pressing on a nerve.

The bones most commonly affected are:

- pelvis
- spine
- thigh
- shin
- skull.

Who gets it?
Paget’s disease is most common in the UK, but it also occurs in people from western and southern Europe as well as Australia and New Zealand (where many Europeans have emigrated in the past). About 2–3 people in 100 over the age of 50 have Paget’s disease in some part of their skeleton.

What treatments are there?
Paget’s disease is often treated with painkillers and a group of drugs called the bisphosphonates. Bisphosphonates may be given as infusions (injections directly into the blood stream) or as tablets.

It may be recommended for you to try the following:

- physiotherapy
- occupational therapy
- surgery (needed for joint replacements and to repair fractures).
What are the possible complications?

In time, Paget’s disease can lead to a number of complications, including:

• bone deformity (bending)
• bone enlargement
• fractures
• compression of nerves
• deafness
• osteoarthritis
• tumours (this is very rare).

How can I help myself?

You can help yourself by maintaining a balanced diet with enough calcium and vitamin D.
What is Paget’s disease?

Paget’s disease of bone is named after Sir James Paget, who first identified the condition in the late 19th century. Paget’s disease of bone affects the way that bone develops and renews itself. To understand this, it helps to know a little about normal bone. Normal bone is made up of three main parts (see Figure 1).

Bone is a living, active tissue that’s constantly being renewed and repaired. Old and damaged bone is removed by cells called osteoclasts, while new healthy bone is produced by cells called osteoblasts. This process is finely tuned and is carried out in an orderly and balanced way to ensure that bone remains strong and healthy.

**Figure 1**
The structure of normal bone

This diagram shows a cross-section through part of the thigh bone (femur).

- **Matrix**
  (magnified)
  collagen fibres and other substances, which make bone resilient

- **Mineral**
  mainly calcium and phosphate, which makes bone rigid

- **Cells**
  osteoblasts and osteoclasts, which renew and repair bone
In Paget’s disease, the process of renewal and repair is disrupted:

- Bone cells increase in number and become larger and more active.
- The renewal and repair of bone becomes chaotic and increases by up to 40 times.
- The new bone is abnormal in shape and structure and is weaker than usual.

The weakened bones become bent or deformed and can cause damage to the joints.

The increased activity of the bone cells also increases the blood flow through the bone. The bones most commonly affected are the thigh bones, shin bones, pelvis, spine and skull (see Figure 2).

**Figure 2**
The bones commonly affected by Paget’s disease

- Skull
- Spine
- Pelvis
- Thigh bone (femur)
- Shin bone (tibia)
Paget’s disease can cause pain but often there are no symptoms at all. It’s often diagnosed when an x-ray or blood test is taken for an unrelated purpose.

There are many possible causes of bone pain in people with Paget’s disease.
What are the symptoms of Paget’s disease?

Paget’s disease can cause pain but often there are no symptoms at all. It’s not unusual for Paget’s to be diagnosed by chance when an x-ray or blood test is taken for a completely unrelated purpose.

For those who do have symptoms, pain is the most common problem and is usually felt in the bone itself or in the joints near the affected bones. There are many possible causes of bone pain in people with Paget’s disease:

- Pain in the affected bone can occur because of increased blood flow or because the nerve fibres surrounding the bone become stretched as a result of bone enlargement or bending. Because of the increased blood flow, the parts of the body affected by Paget’s disease often feel slightly warmer to the touch, especially if the bone is close to the skin, for example in the shin.
- Osteoarthritis, which is a common complication of Paget’s disease, often causes pain in the joints near the affected bones.
- Sometimes enlarged bone can press on nerves, causing pain in the back, neck and legs.

What are the possible complications of Paget’s disease?

In time, Paget’s disease can lead to a number of other symptoms or complications that are potentially more serious:

**Bone expansion** – Bone that’s affected by Paget’s disease expands and may become deformed due to the unusual cell activity. This is most common in the pelvic bones, the spine, the thigh and shin bones, and the skull. Long bones can curve so that one leg may end up shorter than the other.

**Fractures** – Although bone affected by Paget’s disease expands, it’s weaker than usual and is more likely to break than normal healthy bone. Occasionally it may take longer than normal for the bone to heal.

**Nerve compression** – When the bones expand they can sometimes squeeze nearby nerves. This most commonly happens in the spine, which can lead to weakness and tingling in the legs.

**Deafness** – If bones in the head have been affected by Paget’s it can result in a loss of hearing or deafness. This seems to be caused by thickening of the bones around the ear.

**Osteoarthritis** – If the disease reaches to the end of the bone it can lead to osteoarthritis in the joint, causing pain and stiffness when the joint is moved.

**Tumours** – Very rarely, a cancerous tumour can develop in a bone affected by Paget’s disease. The first signs of this are increased pain and swelling at the site of the tumour.
Who gets Paget’s disease?
Paget’s disease is most common in the UK. It also occurs in people from western and southern Europe and also in Australia and New Zealand, where Europeans have emigrated in the past. Paget’s is rare under the age of 40 but becomes increasingly more common with age. In the UK, it has been estimated that about 2–3 people in 100 over the age of 50 have Paget’s disease in some part of the skeleton. However, it often goes unnoticed and may not cause any symptoms. Paget’s is most common in white people, although it can occur in any race.

What causes Paget’s disease?
It’s not yet known exactly what causes the unusual pattern of bone renewal in Paget’s disease but genetic factors seem likely – about 10–15 in every 100 people with Paget’s disease have relatives who also have the condition, and in these circumstances the disease can be passed on from one generation to the next. Frequently, however, there’s no family history of the condition.

There’s some evidence that lifestyle factors play a part in developing Paget’s disease. For example, poor diet or bone injury early in life may act as triggers for the later development of Paget’s in people who also have genetic risk factors. Another theory is that Paget’s disease may be linked to a viral infection early in life, though this is controversial.

How is Paget’s disease diagnosed?
If the bone is deformed in a way that’s typical of the disease, your doctor may be able to diagnose Paget’s from your symptoms and a physical examination. Often, however, x-rays and blood tests are needed to confirm it. Some blood tests will point to Paget’s disease even though there are no apparent symptoms.

A blood test may show a raised level of an enzyme called alkaline phosphatase. There are many reasons why this blood test may be taken and many conditions besides Paget’s disease can cause the level of alkaline phosphatase to rise, so further tests will be needed before a definite diagnosis of Paget’s disease can be made.

Sometimes the doctor will ask for an isotope bone scan. This is the most effective way of pinpointing where the affected bone is and how active it is.

A tiny, well-tolerated dose of radioactive isotope that can be detected by the bone scan is injected into a vein and the whole skeleton is scanned several hours later. The radioactive material quickly passes out of the body in the urine. An isotope scan is important as it’ll show how many bones are affected by the disease.

Once the diagnosis has been made, you may be referred to a specialist clinic for assessment and treatment.

See Arthritis Research UK booklet
Meet the rheumatology team.
What treatments are there for Paget’s disease?

Drugs
Paget’s disease is often treated with a group of drugs called bisphosphonates. These drugs are often used in the treatment of osteoporosis but at a different dose and for a different length of time. They work by damping down the renewal and repair process that Paget’s disease causes to speed up. These drugs act primarily on the osteoclasts (the bone-eroding cells) to slow down the rapid bone turnover. Drugs in this group include:

- **pamidronate**: Series of weekly or fortnightly injections (six in total) or as a single injection
- **zoledronate**: Single injection – may be effective for up to 2 years
- **risedronate**: 2-month course of tablets
- **tiludronate**: 3-month course of tablets
- **etidronate**: 3-month course of tablets

The advantage of injections is that the drug is all absorbed into the body, but the condition can also be treated with tablets, which are more convenient for most people to take. These must be taken on an empty stomach and food should be avoided for at least 30 minutes afterwards, otherwise they won’t be absorbed into the blood stream.

An isotope bone scan is the most effective way of pinpointing where the affected bone is and how active the disease is.
If you’re taking bisphosphonates orally you shouldn’t do so within 2 hours either side of a calcium supplement because calcium can reduce the amount of bisphosphonate absorbed into the body. Bisphosphonates are often effective at relieving bone pain caused by Paget’s disease, but it may take several weeks for symptoms to improve. Further courses of bisphosphonates may be necessary if symptoms come back.

If a person is unable to tolerate bisphosphonates, Paget’s disease can also be treated with a drug called calcitonin, which can be given as a nasal spray or a daily injection.

Most patients also require painkillers such as paracetamol and/or non-steroidal anti-inflammatory drugs (NSAIDs), which are especially effective at reducing pain caused by osteoarthritis and nerve compression.

Physical therapies
A therapy assessment, generally by a physiotherapist, can be very helpful, for example, to identify whether one leg has been shortened because the thigh and shin bones have become curved. In this case, a built-up insole in the shoe can reduce the feeling of lopsidedness. Physiotherapists can also advise on muscle-strengthening exercise that may be useful, while an occupational therapist can offer advice on walking aids and other aspects of daily living.

Surgery
Surgery isn’t usually necessary, but sometimes if the bone breaks an operation will be needed to fix it, depending
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Paget’s disease of bone
on the type and severity of the break. If osteoarthritis develops from the disease, then joint replacement surgery may sometimes be necessary. Back surgery is occasionally needed if a bone in the back has become enlarged and is causing pressure on the nerves in the spine.

See Arthritis Research UK booklets *Hip replacement; Knee replacement.*

**Self-help and daily living**

**Diet and nutrition**

It’s important that people with Paget’s disease maintain a good diet with adequate amounts of calcium and vitamin D.

**Calcium**

The best sources of calcium are:

- dairy products such as milk, cheese, yogurt – low fat ones are best
- calcium-enriched varieties of milks made from soya, rice or oats
- fish that are eaten with the bones (such as sardines).

We recommend a daily intake of calcium of 1,000 milligrams (mg), possibly with added vitamin D if you’re over 60. Skimmed and semi-skimmed milk contain more calcium than full-fat milk. If you don’t eat many dairy products or calcium-enriched substitutes, then you may need a calcium supplement. We recommend that you discuss this with your doctor or a dietitian.

**Vitamin D**

Vitamin D is needed for the body to absorb and process calcium and there’s some evidence that arthritis progresses more quickly in people who don’t have enough vitamin D.

Vitamin D is sometimes called the sunshine vitamin because it’s produced by the body when the skin is exposed to sunlight. A slight deficiency is quite common in winter. Vitamin D can also be obtained from the diet (especially from oily fish) or from supplements such as fish liver oil. However, it’s important not to take too much fish liver oil.

If you’re over 60, dark-skinned or don’t expose your skin to the sun very often and are worried about a lack of vitamin D, you should discuss with your doctor whether a vitamin D supplement would be right for you.

See Arthritis Research UK booklet *Diet and arthritis.*
Complementary medicine
There’s no scientific evidence that suggests any form of complementary medicine helps to ease the symptoms of Paget’s disease. Generally speaking, though, complementary and alternative therapies are relatively well tolerated if you want to try them, but you should always discuss their use with your doctor before starting treatment. There are some risks associated with specific therapies.

In many cases, the risks associated with complementary and alternative therapies are more to do with the therapist than the therapy. This is why it’s important to go to a legally registered therapist, or one who has a set ethical code and is fully insured.

If you decide to try therapies or supplements, you should be critical of what they’re doing for you, and base your decision to continue on whether you notice any improvement.

Research and new developments
We don’t yet have a cure for Paget’s disease, but there are treatments that can suppress the abnormal bone turnover that causes the disease. Recent research has identified particular gene abnormalities that may contribute to the development of Paget’s disease. Studies such as the PRISM-EZ clinical study aim to find out if early treatment with bisphosphonates could help to prevent complications of the disease developing. The ZiPP study is looking into preventing the development of the condition in people who have a genetic abnormality that has been linked to Paget’s by treating them with a single dose of zoledronate.

See Arthritis Research UK booklet Complementary and alternative medicine for arthritis.
**Glossary**

**Bisphosphonates** – drugs used to prevent the loss of bone mass and treat bone disorders such as osteoporosis and Paget’s disease. They work by reducing high levels of calcium in the blood and by slowing down bone metabolism.

**Non-steroidal anti-inflammatory drugs (NSAIDs)** – a large family of drugs prescribed for different kinds of arthritis that reduce inflammation and control pain, swelling and stiffness. Common examples include ibuprofen, naproxen and diclofenac.

**Occupational therapist** – a therapist who helps you to get on with your daily activities (e.g. dressing, eating, bathing) by giving practical advice on aids, appliances and altering your technique.

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**Osteoarthritis** – the most common form of arthritis (mainly affecting the joints in the fingers, knees, hips), causing cartilage thinning and bony overgrowths (osteofytes) and resulting in pain, swelling and stiffness.

**Osteoporosis** – a condition where bones become less dense and more fragile, which means they break or fracture more easily.

**Physiotherapist** – a therapist who helps to keep your joints and muscles moving, helps ease pain and keeps you mobile.

**Physiotherapy** – a therapy that helps to keep your joints and muscles moving, helps ease pain and keeps you mobile. It’s carried out by a physiotherapist.

**Where can I find out more?**

If you’ve found this information useful you might be interested in these other titles from our range:

**Conditions**
- Back pain
- Neck pain
- Osteoarthritis
- Osteoporosis

**Therapies**
- Meet the rheumatology team
- Occupational therapy and arthritis
- Physiotherapy and arthritis

**Surgery**
- Hip replacement
- Knee replacement
Self-help and daily living
- Complementary and alternative medicine for arthritis
- Diet and arthritis
- Everyday living and arthritis
- Feet, footwear and arthritis
- Keep moving
- Looking after your joints when you have arthritis
- Pain and arthritis

Drug leaflets
- Drugs for osteoporosis
- Non-steroidal anti-inflammatory drugs
- Pamidronate

You can download all of our booklets and leaflets from our website or order them by contacting:

Arthritis Research UK
PO Box 177
Chesterfield
Derbyshire S41 7TQ
Phone: 0300 790 0400
www.arthritisresearchuk.org

Related organisations
The following organisations may be able to provide additional advice and information:

Arthritis Care
18 Stephenson Way
London NW1 2HD
Phone: 020 7380 6500
Helpline: 0808 800 4050
www.arthritiscare.org.uk

Chartered Society of Physiotherapy
14 Bedford Row
London WC1R 4ED
Phone: 020 7306 6666
www.csp.org.uk

College of Occupational Therapists
106-114 Borough High Street
Southwark
London SE1 1LB
Phone: 020 7357 6480
www.cot.co.uk

Paget’s Association
323 Manchester Road
Walkden, Worsley
Manchester M28 3HH
Phone: 0161 799 4646
www.paget.org.uk
We’re here to help

Arthritis Research UK is the charity leading the fight against arthritis.

We’re the UK’s fourth largest medical research charity and fund scientific and medical research into all types of arthritis and musculoskeletal conditions.

We’re working to take the pain away for sufferers with all forms of arthritis and helping people to remain active. We’ll do this by funding high-quality research, providing information and campaigning.

Everything we do is underpinned by research.

We publish over 60 information booklets which help people affected by arthritis to understand more about the condition, its treatment, therapies and how to help themselves.

We also produce a range of separate leaflets on many of the drugs used for arthritis and related conditions. We recommend that you read the relevant leaflet for more detailed information about your medication.

Please also let us know if you’d like to receive our quarterly magazine, Arthritis Today, which keeps you up to date with current research and education news, highlighting key projects that we’re funding and giving insight into the latest treatment and self-help available.

We often feature case studies and have regular columns for questions and answers, as well as readers’ hints and tips for managing arthritis.

Tell us what you think of our booklet

Please send your views to: feedback@arthritisresearchuk.org or write to us at: Arthritis Research UK, PO Box 177, Chesterfield, Derbyshire S41 7TQ.

A team of people contributed to this booklet. The original text was written by Prof. Stuart Ralston, who has expertise in the subject. An Arthritis Research UK editor revised the text to make it easy to read, and a non-medical panel, including interested societies, checked it for understanding. An Arthritis Research UK medical advisor, Prof. Anisur Rahman, is responsible for the content overall.
Get involved

You can help to take the pain away from millions of people in the UK by:

- Volunteering
- Supporting our campaigns
- Taking part in a fundraising event
- Making a donation
- Asking your company to support us
- Buying gifts from our catalogue

To get more actively involved, please call us 0300 790 0400 or e-mail us at enquiries@arthritisresearchuk.org

Or go to:
www.arthritisresearchuk.org

Providing answers today and tomorrow