

Paget's disease of bone

This booklet provides information
and answers to your questions
about this condition.

Arthritis Research UK produce
and print our booklets entirely
from charitable donations.

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.

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At a glance

Paget's disease

Paget's disease affects bone and can cause damage to nearby joints.

What is Paget's disease of bone?

Paget's disease of bone affects the way that bone is renewed and repaired. These processes are sped up and uncontrolled, 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 moved to 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 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 eating a balanced diet with enough calcium and vitamin D.



What is Paget's disease?

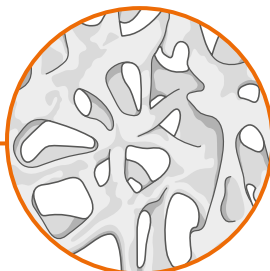
Paget's disease of bone is named after Sir James Paget, who first identified the condition in the late 19th century.

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 make sure that bone remains strong and healthy.

Matrix

(magnified)
collagen fibres
and other
substances,
which make
bone tough



Mineral

mainly calcium
and phosphate,
which make
bone rigid

Cells

osteoblasts
and osteoclasts,
which renew and
repair bone

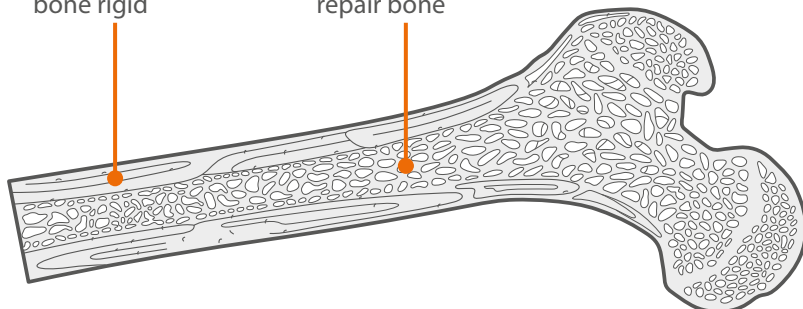


Figure 1
The structure
of normal bone

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

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 uncontrolled and increases by up to 40 times.
- The new bone is abnormal in shape and structure and is weaker than usual.

The weakened bones can become bent or deformed and 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).

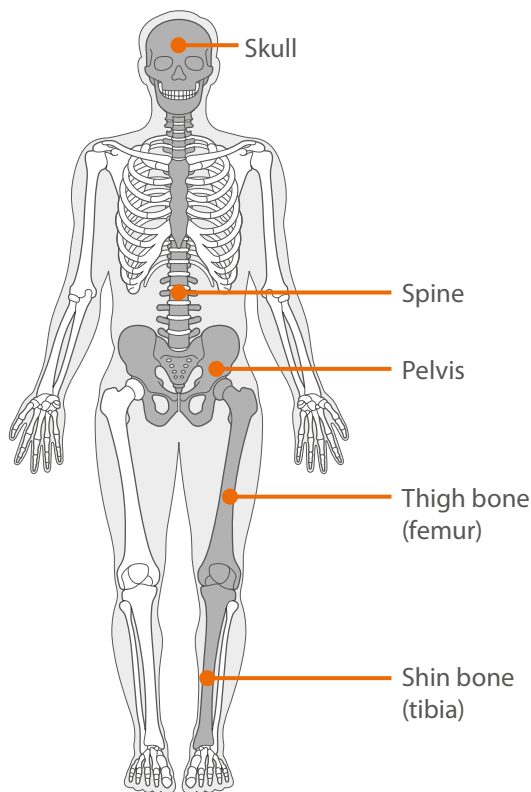


Figure 2
The bones commonly affected by Paget's disease

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.

i See Arthritis Research UK booklets

Back pain; Neck pain; Osteoarthritis; Pain and arthritis.

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 can be more serious:

Bone expansion – Bone that's affected by Paget's disease expands and may become deformed due to the unusual cell activity. 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, although it's estimated that this happens in less than 1 in 500 cases. 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 moved to in the past. Paget's is rare under the age of 40 but becomes increasingly more common with age. In the UK, it's been estimated that about 2–3 people in 100 over the age of 50 have Paget's disease in some part of their 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 cases the disease can be passed on from one generation to the next. But often 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 not everyone agrees with this.

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 clear 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 amount of radioactivity injected is far too small to cause any harm to the body. The isotope is concentrated in the areas of bone affected by Paget's disease so that they show up clearly when the body is scanned with a special camera. After this scan 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.

i 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 overactive renewal and repair process in Paget's disease and act mainly on the osteoclasts (the cells that break down bone) to slow down the fast bone turnover. Types of bisphosphonates include:

pamidronate	Series of weekly or fortnightly infusions (six in total) or as a single infusion
zoledronate	Single infusion
risedronate	Tablets given daily for two months
tiludronate	Tablets given daily for three months
etidronate	Tablets given daily for three to six months

An isotope bone scan is the most effective way of pinpointing where the affected bone is and how active the disease is.

The advantage of infusions 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.



Bisphosphonates can often help your symptoms within three to six months, while painkillers can help pain caused by osteoarthritis.

If you're taking bisphosphonate tablets, you should take them at around the same time each day. You shouldn't take them within two hours either side of a calcium supplement because calcium can reduce the amount of bisphosphonate absorbed into the body.

Bisphosphonates are often effective at easing bone pain caused by Paget's disease, but it may take between three and six months for symptoms to improve.

Bisphosphonates are well tolerated and unlikely to cause serious side-effects. The most common side-effect with pamidronate and zoledronate is flu-like symptoms that last one to two days. With risedronate, tiludronate and etidronate the most common side-effect is a mild stomach upset.

If your symptoms come back, you may need repeated courses of treatment with bisphosphonates, but their effect often lasts for at least one year and sometimes for much longer.

If you're unable to tolerate bisphosphonates, Paget's disease is occasionally treated with daily injections of a drug called calcitonin.

Most people also need 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.

i See Arthritis Research UK drug leaflets *Drugs for osteoporosis; Non-steroidal anti-inflammatory drugs (NSAIDs); Painkillers (analgesics).*

Physical therapies

A therapy assessment, generally by a physiotherapist, can be very helpful. They can, for example, 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.

i See Arthritis Research UK booklets *Everyday living and arthritis; Feet, footwear and arthritis; Keep moving; Looking after your joints when you have arthritis; Occupational therapy and arthritis; Physiotherapy and arthritis.*

Surgery

Surgery isn't usually needed, but sometimes if the bone breaks an operation will be needed to fix it, depending on the type of break and how bad it is.



New research into Paget's disease could give us new treatments in the future.

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.

i See Arthritis Research UK booklets
Hip replacement surgery; Knee replacement surgery.

Self-help and daily living

Diet and nutrition

It's important that people with Paget's disease eat a good diet with enough 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 1,000 milligrams (mg) of calcium, 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 lack (deficiency) of vitamin D 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.

i 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.

i See Arthritis Research UK booklet

Complementary and alternative medicine for arthritis.

Research and new developments

Arthritis Research UK are funding a number of research projects looking into Paget's disease. Researchers at the University of Edinburgh are hoping to develop a genetic test for people with the condition so that we can match patients with the best possible treatments for their bone disease.

Another research project at the University of Aberdeen is investigating how changes to proteins inside the cell may lead to disease development.



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 therapy – a therapy which uses a range of strategies and specialist equipment to help people to reach their goals and maintain their independence. It's given by a trained specialist (an occupational therapist) who gives practical advice on equipment, adaptations or changing the way you do things (such as learning to dress using one-handed methods following hand surgery).

Osteoarthritis – the most common form of arthritis (mainly affecting the joints in the fingers, knees, hips), causing cartilage thinning and bony overgrowths (osteophytes) 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.

Physiotherapy – a therapy given by a trained specialist (a physiotherapist) that helps to keep your joints and muscles moving, helps ease pain and keeps you mobile.

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 surgery*
- *Knee replacement surgery*

Self-help and daily living

- *Complementary and alternative medicine for arthritis*
- *Diet and arthritis*
- *Everyday living and arthritis*
- *Feet, footwear and arthritis*
- *Keep moving*

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Paget's disease of bone

- *Looking after your joints when you have arthritis*
- *Pain and arthritis*

Drug leaflets

- *Drugs for osteoporosis*
- *Non-steroidal anti-inflammatory drugs (NSAIDs)*
- *Painkillers (analgesics)*

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

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St Mary's Court
St Mary's Gate, Chesterfield
Derbyshire S41 7TD
Phone: 0300 790 0400
www.arthritisresearchuk.org

Related organisations

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

Arthritis Care

Floor 4, Linen Court
10 East Road
London N1 6AD
Phone: 020 7380 6500
Helpline: 0808 800 4050
Email: info@arthritiscare.org.uk
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
Email: reception@cot.co.uk
www.cot.co.uk

Paget's Association

323 Manchester Road
Walkden, Worsley
Manchester M28 3HH
Phone: 0161 799 4646
Email: helpline@paget.org.uk
www.paget.org.uk

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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, Copeman House, St Mary's Court, St Mary's Gate, Chesterfield, Derbyshire S41 7TD

A team of people contributed to this booklet. The original text was written by Prof. Stuart Ralston, who has expertise in the subject. It was assessed at draft stage by GP with a special interest in rheumatology Dr Tony Mitchell. 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 products from our online and high-street shops.

To get more **actively involved**, please
call us on **0300 790 0400**, email us at
enquiries@arthritisresearchuk.org
or go to
www.arthritisresearchuk.org



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