Paget's Association

Paget's Disease THE FACTS

Version: 4
Revised: 2019

Paget's Disease – The Facts

Contents

Welcome

Welcome to 'Paget's – The Facts', in which we provide useful information and answer some of the questions which you may have about Paget's Disease of Bone. We hope this booklet will assist you to participate in relevant discussions with your doctor. It has been reviewed by both medical and lay Trustees of the Paget's Association.

The Paget's Association is the only charity in the United Kingdom (UK) dedicated solely to adult Paget's disease. The Association raises awareness of the condition and provides high quality information and support for patients, carers and health professionals. In addition, the Association funds quality research into Paget's disease.

Should you decide to become a member of the Association, you will receive our quarterly newsletter, a range of information booklets, a Paget's Passport, and an invitation to join our Paget's Support Network. You will find further details on page 27.

Our Nurse Helpline is available to anyone who requires support or has questions regarding Paget's disease. You can contact the Helpline by email, telephone or post. Please see the contact details on page 25.

Diana Wilkinson RGN BSc (Hons)
Paget's Association

How did 'Paget's disease' get its name?

figure 1

James Paget was born in Great Yarmouth on 11th January 1814. He became both a surgeon and pathologist. He is considered one of the founders of scientific medical pathology. His family motto was 'Labor ipse voluptas', meaning 'Work itself is a pleasure'. In 1858, James Paget was appointed Surgeon to Queen Victoria and in 1872 she conferred on him a baronetcy, which he accepted with pride.

Sir James Paget *(figure 1)* studied Paget's disease extensively and had his first paper about it published in 1877. His work is held in high esteem and the condition he studied and wrote about in great detail came to be known as Paget's Disease of Bone.

Whilst Sir James Paget's name was also given to other conditions, such as Paget's disease of the breast, these have no connection to Paget's Disease of Bone.

Sir James Paget's great, great grandson, Sir Henry Paget, is a Patron of the Paget's Association.

Paget's Information Events

The Paget's Association's Information Events are a wonderful opportunity to learn from the experts and meet others with the same condition (see page 26).

What is Paget's Disease of Bone?

Throughout life, normal bone is renewed and repaired through a process called bone remodelling. Paget's disease is characterised by abnormalities in this process and the disease can occur in one or several bones. The affected bone is renewed and repaired at an increased rate, causing abnormalities in the bone's structure. Pagetic bone can appear enlarged and misshapen.

The normal bone remodelling process

To understand Paget's disease, it is necessary first to appreciate how normal bone behaves. Bone is living tissue, which is renewed and replaced throughout life. This process is known as bone remodelling and is important in maintaining a healthy skeleton by ensuring that old or damaged bone is removed from the skeleton and replaced with new healthy bone tissue. This balance of the breakdown of bone (resorption) and the production of new bone (formation) is carefully regulated.

There are three main types of bone cell involved in the remodelling process, osteoclasts, osteoblasts and osteocytes. The osteoclasts break down (resorb) old or damaged bone. The osteoblasts then lay down new bone. Some osteoblasts become buried in the newly formed bone and become osteocytes. Osteocytes play a key role in regulating the activity of osteoclasts and osteoblasts. Under normal circumstances, the amount of bone resorbed is exactly balanced by the amount of new bone that is formed (figure 2). Over time, this bone is mineralised, forming a rigid and strong skeleton.

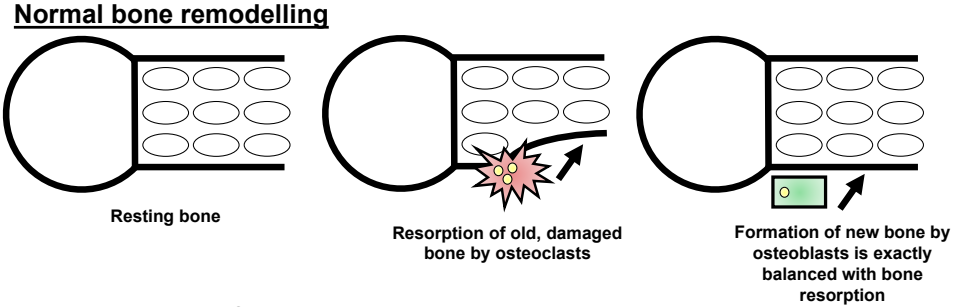


figure 2

How does Paget's disease affect bone?

In Paget's disease, the processes of bone resorption and bone formation are markedly increased. The osteoclasts are larger than normal and break down bone faster than normal. The osteoblasts respond to this by depositing new bone at an increased rate. This dysregulation of bone turnover in Paget's disease (figure 3) results in abnormalities of the bone structure, weakening of the bone and enlargement or deformity of the affected bones. Pagetic bone often appears misshapen and enlarged.

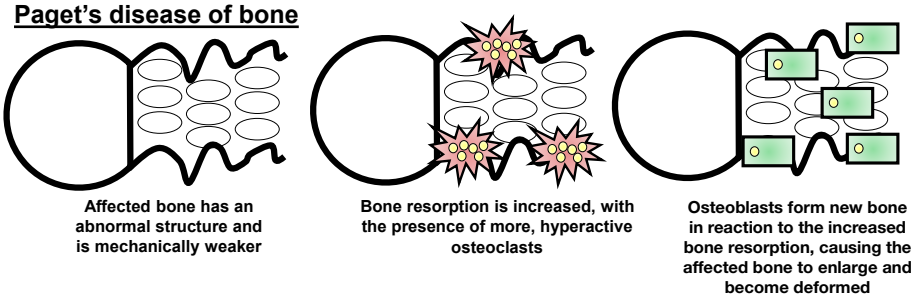


figure 3

Illustrations for figures 2 and 3 provided by Dr D. Green from the University of East Anglia

Which bones can be affected?

- Paget's disease may affect only one bone when it is known as monostotic Paget's disease. If more than one bone is affected, it is known as polyostotic Paget's disease.
- Whilst any bone can be affected by Paget's disease, common sites are the pelvis, spine, thigh bone (femur), shin (tibia) and skull.
- It is unclear why only certain bones are affected. It is possible that it may be related to mechanical stresses, which are placed on the skeleton at specific sites or differences in the blood supply to these bones.

Who develops Paget's disease?

Paget's disease is the second most common metabolic bone disease after osteoporosis. Approximately 1% of people in the UK, over the age of 55 years, are thought to be affected.

The condition is also common in other European countries such as France, Spain and Italy and in people of European descent who have emigrated to other regions of the world, such as Australia, New Zealand, the United States of America and Canada.

What causes Paget's disease?

There are still some uncertainties about the causes of Paget's disease, but it is generally believed to be due to a combination of genetic and environmental influences.

Genetic factors play a key role in predisposing to Paget's disease. It is thought that people who develop Paget's disease inherit variations in one or more genes that regulate osteoclast activity, causing bone resorption to be increased. This is thought to be responsible for the increased bone remodelling that is typical of the disease. The most important predisposing gene for Paget's is called Sequestosome 1 (SQSTM1). Abnormalities of SQSTM1 have been identified in 40% to 50% of people with a family history of Paget's disease and also in 5% to 10% of people who have no family history.

Environmental factors also play a role in Paget's disease, as evidenced by the fact, that over the last few decades, the frequency and severity of the disease have declined in many countries, and is most marked in regions that previously had a high prevalence, such as the UK. Various environmental triggers have been suggested, including dietary calcium or vitamin D deficiency, exposure to environmental toxins, repetitive mechanical stress on the bone, skeletal trauma and slow viral infections. Despite this, researchers have yet to discover which environmental factors influence the development of Paget's disease.

Are my children at risk of developing Paget's disease?

Yes. The children of people with Paget's disease are about seven times more likely to develop Paget's disease than people who don't have a family history. Genetic tests have been developed which can help assess someone's risk of developing Paget's disease. These are not widely available in routine clinical practice at present but can be performed by some specialist research centres.

Can I do anything to avoid Paget's disease?

Unfortunately, as we do not know exactly what triggers Paget's disease, we do not know how individuals who are predisposed to it, can avoid it.

General advice for everyone is to try to lead a healthy lifestyle. This includes a healthy diet, regular exercise and maintaining a healthy weight. You will find information on the NHS website which can help you achieve healthy lifestyle goals: **www.nhs.uk**

What symptoms might I have, and could there be any complications?

- The symptoms of Paget's disease vary widely.
- Many people who have Paget's disease do not have symptoms and never develop complications. In many cases, individuals are unaware that they have the condition.
- Of those who present with symptoms, pain is the most common. Pain may arise from the affected bone itself, or from the altered biomechanics as the result of limb deformity; for example, a bowed leg alters the way you walk and puts stress on the joints and soft tissues.
- The blood flow to active areas of Paget's disease increases and this can lead to a feeling of warmth over the affected bone.
- The affected bone may become enlarged and misshapen.
- The potential for complications will depend on which bones are affected by Paget's disease (figure 4).
- Complications may require medical or surgical intervention.

(continued overleaf)

figure 4

Affected bone	Examples of potential symptoms/complications
Pelvis	Pain Osteoarthritis in the hip joint
Skull	Headaches Change in shape/size i.e. wide forehead/enlarged head Hearing loss Ringing in the ears (tinnitus)
Spine	Pain Curvature of the spine Pressure on nerves may cause tingling, sciatica, weakness and/or numbness in the legs
Thigh	Local pain at the site of disease Pain related to osteoarthritis of the hip joint Fissure (partial) fractures in the bone may lead to a complete fracture
Shin	Pain The affected bone may feel warm The leg may look bowed Osteoarthritis in the knee joint Fracture
Many people do not have symptoms and will never develop any complications	

Deformity
Paget's disease can cause bone to become enlarged and misshapen. For example, if the skull is affected, the enlarged bone is sometimes first noticed when the individual becomes aware that their head is larger and their forehead may be wider than normal.

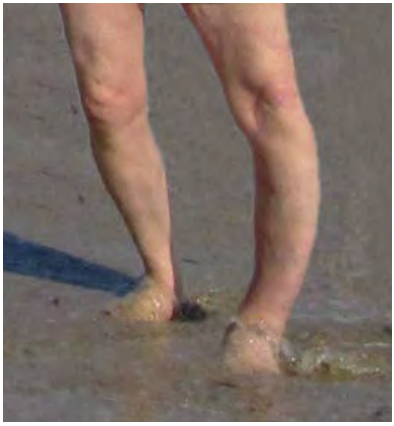
Long-standing disease over many years may cause the weight-bearing bones of the leg to develop a bowing deformity (figure 5).

Deafness
If the skull is involved, hearing loss can occur.

Fracture
There is an increased risk of fracture, particularly in the long bones of the arm and leg. Fractures may initially be incomplete, (stress fractures or fissure fractures), which are at high risk of complete fracture. Fissure fractures predominantly, but not exclusively, affect weight-bearing bones, such as the thigh bone (femur).

Osteoarthritis
Paget's disease can predispose to the development of osteoarthritis at adjacent joints.

figure 5



This patient's left leg has been bowed by Paget's disease

Neurological complications
Neurological complications can occur, often because of bony overgrowth leading to compression. For example, enlargement of vertebrae in the spine can produce pressure on nerves causing pain, leg weakness or sciatica.

Increased vascularity
Should the bone fracture or surgery be undertaken, active Paget's disease has the potential to result in excessive blood loss. This is because blood flow increases to areas where Paget's disease is active.

Heart disease
Paget's disease does not directly affect the heart but if it is in many bones, is highly active and uncontrolled, the heart may have to work harder to pump extra blood to the affected bones. Whilst heart failure due to this increased blood flow has been reported, it is extremely rare.

Osteosarcoma
An unusual and very rare complication of Paget's disease is a type of bone cancer called osteosarcoma. This occurs in less than 1:1,000 people with the disease.

Should I approach my GP if I am concerned about Paget's disease?

If you are concerned that you may have Paget's disease, you should see your GP who will assess your symptoms and may carry out initial investigations such as blood tests. You should tell your GP about any history of Paget's disease in your family. It may be helpful to take this booklet with you. Your GP can refer you to a consultant for a full assessment.

Paget's Disease - Key Facts

- Paget's disease affects the normal repair and renewal process of bone (remodelling).
- The clinical presentation and severity of Paget's disease varies widely.
- Paget's disease is common in the UK, although the frequency and severity of the disease has declined.
- Genetic factors play an important role.
- Environmental triggers probably play a role in the disease, but the identity of these triggers is unclear.
- Many people who have Paget's disease do not have symptoms and will never develop complications.

Assessment

When Paget's disease is suspected, it is important that there is a detailed assessment process, ideally carried out by a hospital consultant who understands the condition. The consultant will consider symptoms, enquire about any family history of the condition, and ensure that appropriate investigations are carried out.

Many rheumatologists and endocrinologists have the expertise to treat patients with Paget's disease and it is likely that most hospitals in the UK have a specialist who can deal with the condition. If there is no specialist locally, then referral to one of the Paget's Association Centres of Excellence might be beneficial.

Centres of Excellence

The Paget's Association has developed a Centre of Excellence Award, which recognises hospital (NHS) and university departments, within the UK, which demonstrate excellence in both the treatment of and research into Paget's disease.

At the time of publication of this booklet, there are twelve Centres of Excellence recognised by the Paget's Association (figure 6). It is anticipated that additional centres will be identified in the future. An up-to-date list can be found on our website **www.paget.org.uk**

figure 6

The Paget's Association's Centres of Excellence

1 Edinburgh
2 Newcastle upon Tyne
3 Middlesbrough
4 Liverpool
5 Manchester
6 Salford
7 Sheffield
8 Nottingham
9 Norwich
10 Stanmore
11 Llandough/Cardiff
12 Southampton

Diagnosis

- The majority of people are over fifty when they are diagnosed with Paget's disease.
- The condition may be identified by an x-ray, blood test or bone scan.
- In many cases, Paget's disease is found by chance when tests are carried out for another reason.
- It has been estimated that less than 10% of patients with x-ray evidence of Paget's disease come to medical attention.

Blood tests

A common blood test in general practice is to measure liver function. Included in this test is an enzyme called alkaline phosphatase (ALP). This is present in many cells within the body, but particularly in liver and bone cells (osteoblasts). If there is overactivity of the osteoblasts due to Paget's disease, alkaline phosphatase is released into the bloodstream and can be measured.

When Paget's disease is active, the ALP level will often, but not always, be raised. A raised ALP can stand out as being the only abnormal result. If there is co-existent liver disease, it may be necessary to perform further blood tests to identify the source of the elevated ALP.

Serum total alkaline phosphatase (ALP) is recommended as the first-line test

Serum total alkaline phosphatase (ALP) is recommended as the first line blood test, in combination with liver function tests to detect Paget's disease. If the total ALP values are normal and clinical suspicion of Paget's disease is high, then measurement of more specialised tests such as bone alkaline phosphatase (BALP) or N-terminal propeptide of type I procollagen (P1NP) may be required.

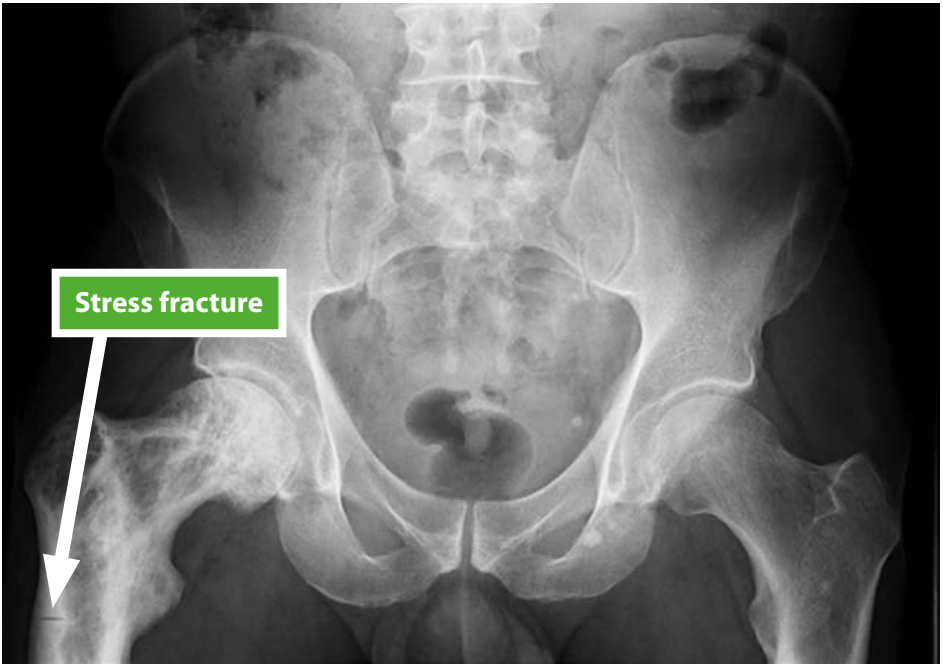
X-ray

When an x-ray is taken of a bone affected by Paget's disease, characteristic features can often be seen (figure 7).

Research has shown that plain x-rays targeted to several sites, the abdomen, skull and facial bones, and both tibiae (shins), are likely to detect 93% of bones affected by Paget's disease, compared with a single x-ray of the abdomen, which detected less (79%).

A single x-ray cannot give information about the many bones which can be affected by Paget's disease. A radionuclide bone scan, as described on page 16, is the best way of fully evaluating the extent of Paget's disease.

figure 7.



This x-ray, of the upper part of the hip, shows a patient with normal bone on one side and bone affected by Paget's disease on the other (seen on the left side of the picture). The affected bone is enlarged and the pattern of the bone is abnormal. The x-ray also shows a small stress fracture (indicated by the arrow), which reflects the fact the bone is weak and can be a cause of pain.

Bone scan

There are two types of bone scan in common use. Dual-energy x-ray absorptiometry (DEXA) scans, which are used in the diagnosis of osteoporosis, and radionuclide bone scans, used in the diagnosis of other bone conditions.

Radionuclide bone scans, also known as scintigrams, isotope bone scans or nuclear medicine bone scans, are the most helpful in Paget's disease. A scan is recommended to determine which bones have Paget's disease and how active the disease is.

The radionuclide scan involves the use of a small amount of a radioactive tracer, which is injected into a vein in the arm. After some time, the tracer collects in the bones and pictures can be taken by a scanner (gamma camera).

A radionuclide bone scan is the best way of determining the extent of Paget's disease

Bone biopsy

A bone biopsy is a procedure in which a small sample of bone is taken and examined under a microscope. This is seldom required but can be useful if there is uncertainty about the diagnosis.

For a more detailed explanation of tests, please see our booklet *"Paget's Disease - Investigations Explained"*

Treatment

Paget's disease does not always cause symptoms and not everyone needs treatment.

The main reason for treatment is if the affected bones are painful. If the pain is directly from Paget's disease, it often improves with treatment. Pain, however, can arise from complications (see page pages 9 and 10).

Bisphosphonates

Bisphosphonates are drugs which work by inhibiting the bone remodelling process (detailed on page 6). These drugs reduce abnormal bone destruction by the osteoclasts, thereby restoring a more normal remodelling process.

The main treatment goal is to control bone pain. The clinical benefit of giving bisphosphonates to patients who have a raised level of alkaline phosphatase in their blood, but do not have symptoms, is unknown.

It can take several months for bisphosphonates to have their full effect and for the individual to feel the maximum benefit.

Zoledronic acid

For those who require treatment, the current first-line bisphosphonate, due to its potency and prolonged duration of action, is zoledronic acid. It is the bisphosphonate most likely to relieve pain from active Paget's disease. It is usually given in hospital, as an outpatient. A single dose of 5mg is given through an infusion (a drip) directly into the bloodstream (intravenous), over 15 minutes. Over the following months, this treatment often normalises the abnormal bone remodelling and one dose can be effective for many years.

The current first-line bisphosphonate used to treat Paget's disease, is **zoledronic acid**

Alternatives to zoledronic acid

Bisphosphonates can also be given as tablets; these are slightly less effective than an infusion of zoledronic acid, at reducing bone remodelling, and the effect doesn't last quite as long.

Risedronate

The most commonly used oral treatment is 30mg of risedronate sodium, taken daily, for two months. If necessary, the course can be repeated.

(continued overleaf)

For risedronate to be effective, it is important that you do not take it with food, other medicines or drinks (other than plain water). If taken at the same time, medicines containing any of the following lessen the effect of risedronate: calcium, magnesium, aluminium (e.g. some indigestion mixtures) or iron. To ensure risedronate is absorbed properly, it should be taken with a full glass of water, on an empty stomach, first thing in the morning. These tablets can irritate your oesophagus (the tube that takes food into your stomach) therefore, sit upright or stand for at least 30 minutes after taking it. Wait at least 30 minutes before eating or drinking (other than water), and before taking other medication.

Pamidronate

Pamidronate is an effective treatment but has largely been superseded by zoledronic acid which lasts longer and is easier to administer. Pamidronate is given in several doses, intravenously (an infusion into the bloodstream), and repeated when necessary, dependant on symptoms. Doses can vary, but commonly 60mg is given by an infusion over a period of four hours and this is repeated on three consecutive days.

Calcitonin

In cases where bisphosphonates are not recommended, calcitonin injections may be considered to treat bone pain in Paget's disease.

What about side effects of bisphosphonates?

A comprehensive review on the effects of bisphosphonates in the treatment of Paget's disease, published in 2017, concluded that serious side effects were rare and that the most common side effects experienced were a flu-like illness in people given bisphosphonates by infusion, and stomach upset in those given tablets. Severe side effects causing treatment discontinuation were rare.

The most common side effect with zoledronic acid is a flu-like reaction that lasts a day or two in most people who experience it. It is usually of mild or moderate severity, but more severe reactions can sometimes occur. Pamidronate can also cause a flu-like reaction but there is less information on the frequency with which this occurs or its severity. If a flu-like reaction does occur with either drug, it can usually be controlled with paracetamol or ibuprofen.

Both Zoledronic acid and pamidronate may cause a decrease in calcium levels. The risk of this is reduced in people who have a good dietary calcium intake and those with normal levels of vitamin D. Sometimes vitamin D levels (25-hydroxy vitamin D) might be measured before treatment to determine if supplements are required. Depending on the results, calcium and vitamin D supplements may be given prior to treatment as a preventative measure.

The most common side effect of taking risedronate is stomach upset.

On rare occasions, bisphosphonates are associated with osteonecrosis of the jaw (ONJ). This is a condition which may present after dental surgery when exposed bone fails to heal. ONJ is rarely seen in those with Paget's disease and the risk of ONJ is greater if bisphosphonates are given for cancer. As a precaution, however, if possible, complete any extensive dental treatment prior to bisphosphonate treatment. If you have to undergo extensive dental procedures, it is important to inform your dentist that you are having, or have had treatment with a bisphosphonate.

You should discuss your individual circumstances with your doctor, but if bisphosphonate treatment is required, the benefits often outweigh the risk of any potential side effects.

When should bisphosphonates be avoided?

The decision to give treatment may be modified if you have another medical problem which could be aggravated by bisphosphonates. As bisphosphonates are excreted by the kidneys they cannot be used if there is significant kidney disease. If you are being considered for bisphosphonate treatment, the doctor will usually organise a blood test to check kidney function, to see if it is safe to go ahead. Although bisphosphonates are rarely given to younger people, they should be avoided during pregnancy as their effects on the foetus are unknown.

Follow up

An assessment of the response to treatment should take place between 3 and 6 months after the course of treatment has been completed.

Other medication

Analgesics

Although bisphosphonates can help reduce bone pain, some patients require a painkilling drug (analgesic) for maximum pain relief, particularly if there is significant osteoarthritis in major joints. Analgesics which are commonly used include paracetamol, co-codamol and dihydrocodeine or non-steroidal anti-inflammatory drugs such as ibuprofen.

For additional information, see our booklet *"Paget's Disease and Pain"*

Calcium and vitamin D

Calcium is vital for the development of healthy bones, whilst vitamin D is required to help regulate the way your body uses calcium, and to ensure your bones, muscles and teeth remain strong. Calcium is in foods such as milk and cheese, broccoli and cabbage, and soya beans. Additional calcium may be required if an individual has a low intake of dairy produce.

Vitamin D is mostly obtained by the action of sunlight on your skin, but it is also in foods such as oily fish. In the UK from October to March, we cannot make vitamin D in our skin therefore, some people might not have enough to last through the winter and may need a supplement to boost their diet. Advice given by Public Health England in 2016, was for everyone to have a daily supplement of 10 micrograms (which is equal to 400 International Units) of vitamin D. This was a general recommendation for everyone, as it is difficult to know who receives enough sunlight exposure to produce vitamin D, and also to cover those at risk of deficiency. Most supplements contain vitamin D3 (cholecalciferol) which may be slightly more effective than vitamin D2 (ergocalciferol).

As discussed on page 19, treatment with zoledronic acid or pamidronate may cause some patients to have low levels of calcium in their blood. Calcium and/or vitamin D supplements may be prescribed prior to treatment.

Diagnosis and Treatment - Key Facts

- Most people are over 50 when they are diagnosed with Paget's disease.
- The condition may be identified by an x-ray, blood test or radionuclide bone scan.
- A radionuclide bone scan is the best way to determine the distribution and extent of Paget's disease.
- Paget's disease does not always cause any symptoms and not everyone needs treatment, but if there is pain which is thought to be caused by Paget's disease, treatment with bisphosphonates should normally be given.
- A detailed assessment is key, since there are many potential causes of pain in people who have Paget's disease.
- The current first-line treatment for pain caused by Paget's disease is zoledronic acid, since it is the one most likely to relieve pain from active Paget's disease, but other bisphosphonates can also be effective.
- If pain is present, usually the benefits of treatment outweigh the risk of any potential side effects.

Is surgery required?

Surgery may be required to treat complications of Paget's disease such as fractures, osteoarthritis and when bone enlargement is thought to be compressing the nerves in the spine.

Prior to surgery, it is usual for bisphosphonate treatment to be administered in the hope that this might reduce blood loss, which can occur in Paget's disease, due to the increased vascularity of affected bone.

(continued overleaf)

Surgery may be required under the following circumstances:

- If an affected bone fractures (breaks), an operation is usually necessary to stabilise the fracture (figure 8).
- When Paget's disease leads to marked damage of the joints (osteoarthritis) and if symptoms and disability from this become severe, joint replacement surgery may be required (figure 9).
- When there is marked bone deformity, usually seen in the lower leg, an osteotomy can be carried out. This involves breaking the bone and realigning it to correct the shape.
- If nerve compression occurs, surgery may be necessary, e.g. Paget's disease in the spine can press on the spinal cord, causing a narrowing that occasionally needs to be corrected surgically, if medical treatment is unsuccessful.

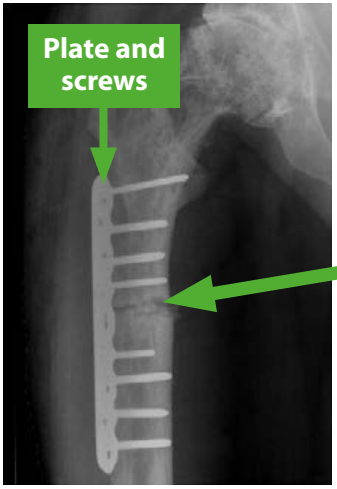


figure 8

An x-ray of a femur (thigh bone) affected by Paget's disease, which has had a fracture repaired using a plate and screws

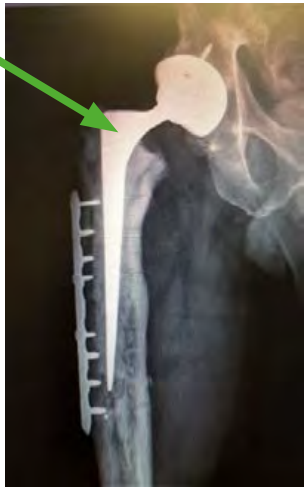


figure 9

This x-ray is from the same patient as in figure 8, who some years later, has undergone a successful hip replacement for osteoarthritis of the hip

Exercise

Regular exercise is important. It helps to keep the joints mobile, control weight and maintains muscle tone, which helps protect bones.

Whilst most forms of exercise and physical activity are good for general health to prevent falls, the most effective exercise should focus on both strength and balance. These exercises are often performed as part of a group in the community.

To avoid excessive stress on Pagetic bone, any extreme forms of exercise should only be undertaken following discussion with your doctor.

Physiotherapy

Some people with Paget's disease benefit from physiotherapy. This can involve exercises and techniques to help reduce pain and improve movement. It can also help to reduce the risk of injury and falls.

Occupational therapy

Occupational therapists can provide support to improve an individual's ability to carry out everyday tasks and restore their ability to function as independently as possible. Therapists can help with special equipment like mobility aids and help adapt people's homes to accommodate their individual needs.

Orthotics

An orthotic device is something that is externally fitted to support a part of the body. It may help reduce pain, improve function and improve mobility. Examples of such products are lumber supports, leg braces, footwear and insoles. An orthotist is a professionally qualified person who works as part of a multi-disciplinary team, ensuring that the orthotic prescription reflects the most appropriate solution to meet the individual's needs.

Further information

A clinical Guideline

In 2019, a new clinical Guideline, for the Diagnosis and Management of Paget's Disease of Bone in Adults, was published on behalf of the Paget's Association, the European Calcified Tissue Society, and the International Osteoporosis Foundation. The Guideline provides evidence-based recommendations for the diagnosis and management of Paget's disease. It allows health professionals to consider the available evidence and discuss options with the patient. For further information, please contact the Paget's Association (see page 25).

The following recommendations* were highlighted by the Guideline Development Group as the most important:

1. Radionuclide bone scans, in addition to targeted radiographs, are recommended as a means of fully and accurately defining the extent of the metabolically active disease in patients with Paget's Disease of Bone.
2. Serum total alkaline phosphatase (ALP) is recommended as a first line biochemical screening test, in combination with liver function tests, in screening for the presence of metabolically active Paget's Disease of Bone.
3. Bisphosphonates are recommended for the treatment of bone pain associated with Paget's disease. Zoledronic acid is recommended as the bisphosphonate most likely to give a favourable pain response.
4. Treatment aimed at improving symptoms is recommended over a treat-to-target strategy aimed at normalising total ALP in Paget's Disease of Bone.
5. Total hip or knee replacements are recommended for patients with Paget's Disease of Bone who develop osteoarthritis, for whom medical treatment is inadequate. There is insufficient information to recommend one type of surgical approach over another.

**Reproduced from Ralston et al (2019), JBMR, DOI: 10.1002/jbmr.3657 (full reference on page 30).*

Additional information regarding bisphosphonates

A Cochrane Review of Bisphosphonates for Paget's Disease was published in 2017. The Cochrane Database of Systematic Reviews is a trusted resource for systematic reviews in health care. Visit **www.cochranelibrary.com**

Paget's Association

The founder,
Ann Stansfield MBE

The Paget's Association is a registered charity, focusing solely on Paget's Disease of Bone. Also known as The National Association for the Relief of Paget's Disease, the charity was founded in 1973, by the late Mrs Ann Stansfield MBE, following her personal experience of the lack of understanding of Paget's disease by those caring for her husband.

Here to help

The Paget's Nurse Helpline

The Paget's Association has a Helpline managed by an experienced Registered Nurse. The Helpline is available to anyone who needs support or who has a question regarding Paget's Disease of Bone. The Helpline can be accessed by telephone, email or letter.

How to Contact the Paget's Association

General and Membership Enquiries
Email **membership@paget.org.uk**

Telephone 0161 799 4646

Nurse Helpline
Email **helpline@paget.org.uk**

Telephone 0161 799 4646 and ask to speak to the Nurse

Via our website
www.paget.org.uk

Support through membership

When you become a member of the Paget's Association, you join many others affected by Paget's disease. Membership provides support and information in various ways and all members receive a Paget's Information Pack, as well as our quarterly Paget's Newsletter.

What is in the Paget's Membership Pack?

The contents of the Information and Membership Pack will vary to reflect the current information available. For example, the pack usually includes:

- A booklet about Investigations for Paget's disease.
- The booklet: Paget's Disease and Pain.
- A Paget's Passport.
- Two newsletters containing information, news and discussion relevant to those who have Paget's or have an interest in Paget's disease.
- Information sheets on various aspects of Paget's disease.
- A complete list of the UK Centres of Excellence.
- An invitation to join our Support Network, which will put you in touch with others.

Members also benefit from:

- Receiving the quarterly Paget's Newsletter, which is free from advertising.
- Free attendance at our UK Paget's Information Events, for you and a carer/spouse. These provide an opportunity to listen to presentations from experts, raise questions and meet with one another.
- Access to the members' area of the Paget's Association's website, where you will find previous newsletters, personal experiences of those with Paget's disease and much more.

- The right to vote at our Annual General Meeting.
- Opportunities to take part in member consultations i.e. regarding research.

How do I become a member?

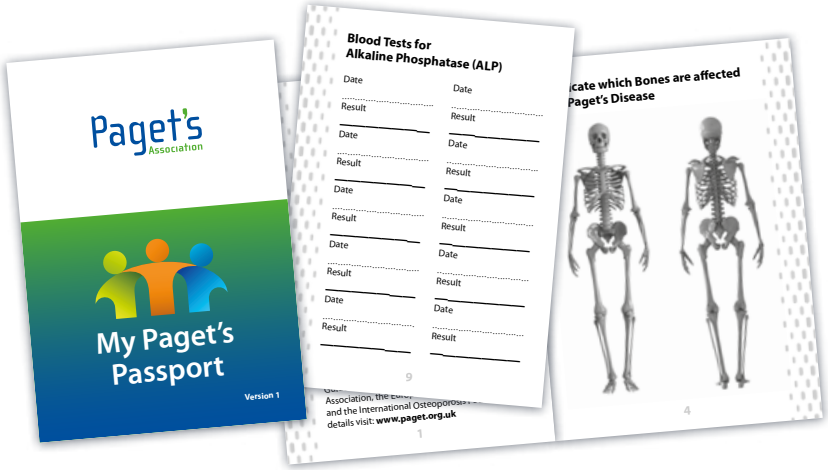
Patients, family members, carers and health professionals are welcome to join the Paget's Association. For full details of how to do this please, visit our website, **www.paget.org.uk**. Alternatively, email **membership@paget.org.uk** or telephone our office on 0161 799 4646.

Paget's Support Network

The Paget's Support Network is a free mutually supportive network to enable members of the Paget's Association to talk with or write to others, who either have Paget's disease, or who care for someone who has the condition. To join, please contact the Paget's Association.

Patient Passport

The Paget's Passport is a document in which to record a summary of how Paget's disease affects you, including test results and any treatment you have had. Ideally completed by your consultant, it can be taken to all your health-related appointments. The Paget's Passport will help you explain your condition to those who are unfamiliar with it. It is also useful to take with you when you are away from home, in case you need medical assistance. The Passport is available free of charge to members of the Paget's Association and is also available from NHS hospitals who have been awarded Paget's Association Centre of Excellence status (see page 13).



Online support

- Our website has details of our Information Events, news and links to our Paget's Forum and social media.
- Our newsletters can be downloaded from the area of our website that is dedicated to members, where you can also read detailed experiences of others.
- Full details regarding membership of the Paget's Association can be found on our website.

www.paget.org.uk

Research into Paget's disease

We know that research is important to those who have Paget's disease. The Paget's Association funds high-quality research projects, which result in advances in the understanding and treatment of Paget's disease.

Paget's Awareness Day

Add the 11th January to your calendar

The 11th January is Paget's Awareness Day so why not join us in doing something, however large or small it may be, to help us raise awareness of Paget's disease?

How can I donate to the Paget's Association?

The Paget's Association relies on voluntary contributions, legacies and fundraising activities, to develop and continue its work. Here are some ways that you can donate to our Charity:

TELEPHONE
0161 799 4646

STANDING ORDER
Request a form
Email **sue@paget.org.uk**
or call 0161 799 4646

POST
Post a cheque payable to 'Paget's Association' *(to the address on our website)*

ONLINE
www.paget.org.uk

THANK YOU FOR YOUR SUPPORT

Other sources of support and information

Carers UK
0808 808 7777
advice@carersuk.org
www.carersuk.org

Age UK
0800 055 6112
www.ageuk.org.uk

Pain Concern
0300 123 0789
info@painconcern.org.uk
www.painconcern.org.uk

Disabled Living Foundation
0300 999 0004
info@dlf.org.uk
www.dlf.org.uk

Versus Arthritis
0800 5200 520
helpline@versusarthritis.org
www.versusarthritis.org

Arthritis Care
0808 800 4050
helplines@arthritiscare.org.uk
www.arthritiscare.org.uk

References

Paget's Guideline
Ralston, S. H., Corral-Gudino, L., Cooper, C., Francis, R. M., Fraser, W. D., Gennari, L., Guañabens, N., Javaid, M. K., Layfield, R., O'Neill, T. W., Russell, R. G., Stone, M. D., Simpson, K., Wilkinson, D., Wills, R., Zillikens, M. C. and Tuck, S. P. (2019), Diagnosis and Management of Paget's Disease of Bone in Adults: A Clinical Guideline. Journal Bone Mineral Research. doi:10.1002/jbmr.3657

References regarding other sources of information used for this booklet are held by the Paget's Association. Should you wish for further information on these, please contact us.

Tell us what you think

To tell us what you think of this booklet or any of our information and support services, please send your views to the Paget's Association using the contact details on page 25.

If you require this information in another format, such as larger text, please get in touch.

Notes

Paget's Association

www.paget.org.uk
Tel: 0161 799 4646

Registered Charity No. 266071