



ARTHRITIS FOUNDATION

Registered Nonprofit Organisation - No. 002-847 NPO

Helpline No: 0861 30 30 30

PAGET'S DISEASE EXPLAINED

This information leaflet is published by the Arthritis Foundation
as part of our continuing education programme
for all people with arthritis.



The Bone and Joint Decade is a global campaign to improve the quality of life for people with musculoskeletal conditions and to advance understanding treatment of those conditions through research, prevention and education.

ABOUT THIS LEAFLET

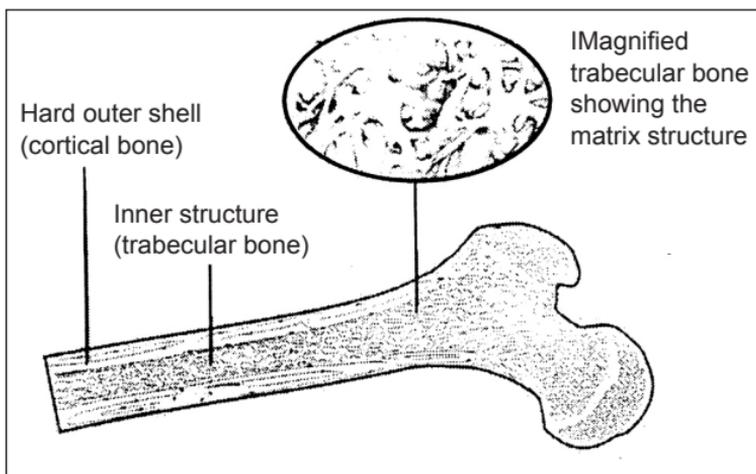
This leaflet has been produced for anyone suffering from Paget's disease of bone. We want to explain as much as we can about this condition; how it is diagnosed and how it can be treated. Unfortunately, we cannot hope to answer all your questions, because everyone is different and this leaflet is NOT intended to replace an individual consultation with doctor.

WHAT IS PAGET'S DISEASE OF BONE?

In order to understand this it helps to know a little about normal bone. Some people think that bone is a 'dead', inactive material. Nothing could be further from the truth. In a living person, bone is an active tissue made up of three main components:

- mineral (mainly calcium)
- matrix (tough collagen fibres and other substances, which give bone it's resilience)
- cells: osteoblasts (bone forming cells) and osteoclasts (bone absorbing cells)

Bones change constantly. 'Old' bone is continually removed by the action of osteoclast cells. It is replaced by new bone – made by osteoblast cells. This turnover or renewal of bone is carefully balanced. If it weren't, you could suffer from bone 'fatigue' (rather like metal fatigue in old machines). If that happens your bones would be more likely to break.

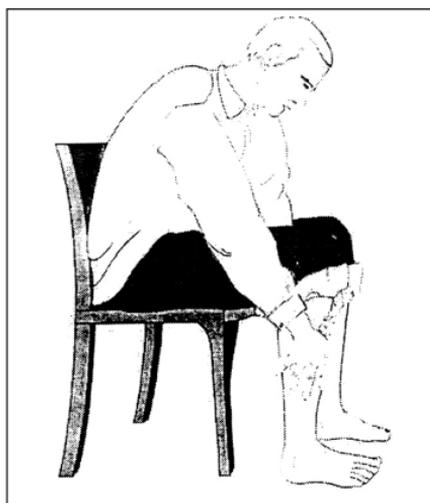


The structure of normal bone. This diagram shows a cross-section through part of the thigh bone (femur).

Paget's disease affects bone growth. The abnormality occurs in the bone cells. The regular, ordered process of bone loss and formation doesn't take place properly. Instead, the bone cells increase in number, become larger and also more active. Bone turnover increases by up to 40 times, so it is surprising that the new bone which is made is disorganised, abnormal in shape and weaker than usual. The increased activity of the bone cells also increases the blood flow through the bone. This is why you have bone affected by Paget's which is close to the skin (the shin, for example) it can feel warm to the touch.

HOW COMMON IS PAGET'S DISEASE?

Paget's disease is not rare. It is most common in the United Kingdom and in countries where large number of British people have emigrated in the past. It is most likely to affect older people. About 5 out of every 100 people over 50 years of age in the UK have Paget's disease in some part of the skeleton, although it is often unnoticed and may not cause symptoms. Paget's disease is even more common in Lancashire where up to 8% of the over 50s have the disease. Younger or non-white people are less likely to have Paget's, though it can occur in any race.



Bone which is near to the surface can feel hot to the touch when affected by Paget's disease.

WHAT CAUSES PAGET'S DISEASE?

Because of the racial and geographical pattern we have just mentioned, some doctors have concluded that the disease may be inherited. It is certainly true that some families have

several members who are affected by Paget's disease, this is quite rare. However, there is some evidence that Paget's may be caused by a 'slow virus' infection of bone cells. It is called 'slow' because the virus may settle in the bone cells early in life and only cause the disease many years later.

WHAT ARE THE SYMPTOMS?

Sometimes none at all

It is quite common for the disease to be discovered by accident on an x-ray which has been asked for for a quite different purpose. Similarly it may be detected after a blood test taken for another reason.

Pain

This is the most common symptom and is usually felt in the bone itself. Paget's disease can sometimes cause osteoarthritis in the joints nearby and this can also be painful. The increase blood flow through the bone which is a feature of Paget's disease also stimulates nerve fibres in the bone which send out pain signals.

WHAT ARE THE COMPLICATIONS OF PAGET'S DISEASE?

Bone expansion

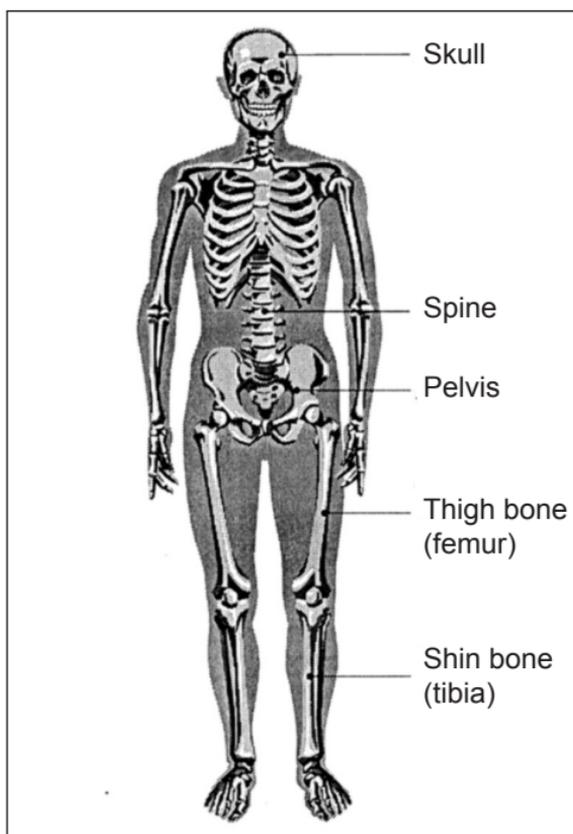
Bone which is affected by Paget's disease expands and may cause deformity due to the disorganized cell activity. Any bone in the skeleton can be affected but the most common are pelvic bones, the spine, the thigh and shin bones, and the skull. Long bones can curve, so that some people may end up with one leg effectively shorter than the other.

Fractures

Although bone affected by this condition usually expands, is weaker than usual and is more likely to break than normal healthy bone. Fortunately, however, the bone heals just as well as normal bone.

Nerve compression

When Pagetic bone expands it can sometimes squeeze nearby nerves. This can happen in the skull, where it can cause deafness, or in the spine, which can lead to weakness and tingling in the legs.



Bones commonly affected by Paget's disease.

Arthritis

If the disease reaches to the end of the bone, it can lead to arthritis in the joint. This is the same as osteoarthritis, and it can cause pain and stiffness on the moving joint. (see A.F. booklet 'Osteoarthritis').

Gout

People with Paget's disease have an increased risk of developing gout. Gout causes pain and swelling in a joint, often the big toe joint. If this happens the usual treatments for gout are effective (see A.F. booklet 'Gout').

Heart Failure

When the Paget's disease is very extensive the increased blood flow through the bone can put a strain on the heart. Fortunately, with the newer, more effective treatments for Paget's this complication is rarely seen nowadays.

Tumours

In a very small number of people with longstanding Paget's

disease tumours may develop in the bone, which can sometimes be cancerous. The first signs of this is increased pain and swelling at the site of the tumour. Fortunately this is a very rare complication.

HOW IS PAGET'S DISEASE DIAGNOSED?

Your doctor would have listened to your symptoms and examined you. If the bone is deformed in a way that is typical of the disease, it is an easy diagnosis. Often, however, x-rays and blood tests will be necessary to confirm it.

Sometimes the doctor will recommend a isotope bone scan. This is the most effective way of pinpointing where the Pagetic bone is and how active it is. A tiny dose of radioactive isotope (a radioactive substance which 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 will show how many bones are affected by the disease.

Once the diagnosis has been made, most patients will be referred to a specialist clinic for assessment and treatment.

HOW IS IT TREATED?

Paracetamol and/or non-steroidal anti-inflammatory drugs (NSAIDs) may help to reduce the pain in some people although they are not always very effective. Simple measures are sometimes useful. If, for example, one leg has effectively been shortened because the thigh and shin bones have grown curved, a built-up insole in the shoe can reduce the feeling of 'lop-sidedness'.

Treatment of the causes of Paget's disease will usually be with a group of drugs called bisphosphonates, which are very effective. They act primarily on the osteoclasts (the bone-eroding cells) to slow down the rapid bone turnover. The choice falls between pamidronate, a drug which is injected into a vein, and two oral drugs, risedronate and tiludronate. Pamidronate is given either as a series of weekly or fortnightly injections (6 in total) or, in some centres, as a single injection which may be repeated after 6 months if necessary. The advantage of the injection is that the drug is all absorbed into the body. Risedronate

is a 2-month course of tablets and tiludronate is a 3-month course. Some people may prefer tablets to injections and the choice available. Further courses may be given if necessary.

The pain in the bone is relieved within 3-6 months and sometimes more quickly than this. Most people have few side-effects from the drugs, although pamidronite occasionally causes flu-like symptoms lasting 1-2 days, and risedronate and tiludronate can sometimes cause indigestion.

WILL SURGERY BE NECESSARY?

Surgery is not normally necessary, but sometimes if the bone breaks, an operation will be needed to fix it. This depends on the type and severity of the break, as with any fracture. If osteoarthritis results from the disease then a new joint may be necessary – treatment with bisphosphonates prior to the operation helps to reduce the blood loss that can occur with this operation.

FINALLY ...

We do not yet have a cure for Paget's disease. However, there are treatments which can suppress the disease, and delay or even prevent the complications, and research continues which we hope will lead to even more effective treatments.

NATIONAL OFFICE

709 Tulbagh Centre
HansStrifdom Avenue
CapeTown 8001
PO Box 6775
Roggebaai 8012

Tel: (021)425-2344
Fax: (021)421-7330
Website: <http://www.arthritis.org.za>
E-mail: info@arthritis.org.za
National Helpline: 0861303030

Johannesburg Branch

POBox87360,Houghton 2041
Tel: (011)485-0199
Fax: 0867295321
E-mail: nadine@arthritis.org.za

Soweto COPE

Tel: (011)982-6315

Pretoria Branch

PO Box 145,
Menlyn 0063
Cell: 0735804199
Tel: (012)998-7203
Fax: 0866957117
E-mail: patvdv@arthritis.org.za

Bloemfontein Branch

POBox20269,Willows 9320
Cell: 0829214234
Fax: (051)447-0451
E-mail: corrie@arthritis.org.za

Eastern Cape Branch

POBox7740
NewtonPark 6055
Tel: (041)365-1419
Cell: 0825760382
E-mail: jeanette@arthritis.org.za

Western Cape Branch

705Tulbagh Centre
HansStrifdom Avenue
CapeTown 8001
POBox6775
Roggebaai 8012

Tel: (021)425-4759
Fax: (021)421-7330
E-mail: julie@arthritis.org.za

Kwazulu-Natal Branch

POSTNET#309
P/BX04,Dalbridge 4014
Tel/Fax:(031)201-8812
E-mail: revor@arthritis.org.za



ARTHRITIS FOUNDATION

Registered Non-profit Organisation Number 002847NPO

MEMBERSHIP APPLICATION/ANNUAL RENEWAL FORM

Please tick relevant boxes:

- | | | |
|--|---|------------------|
| <input type="checkbox"/> New application | <input type="checkbox"/> Renewal | |
| <input type="checkbox"/> Individual membership | <input type="checkbox"/> International membership | R 100 R 300 |
| <input type="checkbox"/> Corporate membership | <input type="checkbox"/> Other | R 5000 R |
| <input type="checkbox"/> Donation | | R |
| | Total | R |

(For details of Associate membership (individuals joining through the community outreach programme. Please contact your local branch).

My payment is made by:

<input type="checkbox"/> Crossed cheque	<input type="checkbox"/> Debit my Credit Card	Expires	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
<input type="checkbox"/> Electronic Payment to:	Card number, on front of Credit Card		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Standard Bank on _____ Date			<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Branch Code: 02090900			<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
Account No.: 070965226			<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	Plus card verification code (last 3 digits at BACK of your card →)		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>
	Signature: _____		<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>

Contact Details: Title (Mr., Mrs., Ms., Dr., Professor, other): _____

Surname: _____ First Names _____

Preferred form of address for correspondence ie. Dear _____

Home Address (number, name of dwelling, street, suburb, city/town/district, postal code): _____

Postal Address (if different): _____