



Paget's Disease & Pain

By Anne Sutcliffe - Healthcare & Education Officer
Paget's Association
Registered Charity No.: 266071

Revised
April 2011
Review: April 2013





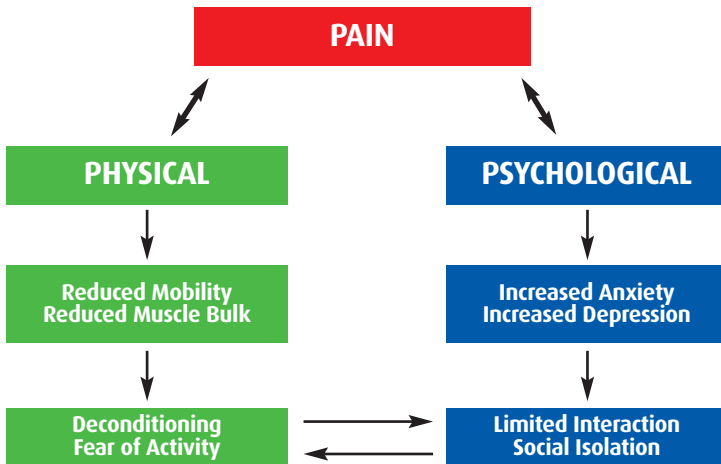
PAGET'S DISEASE AND PAIN

- Paget's disease is a chronic bone disorder occurring in approximately 3% of men and women over the age of 50 years.
- It is caused by an abnormality of bone remodelling which results in disorganised bone structure that is weaker than normal.
- Symptoms range from none to pain, fracture and deformity at affected sites in the skeleton
- The disease can be effectively controlled by medication.

PAIN IS THE COMMONEST PRESENTING SYMPTOM OF PAGET'S DISEASE WITH STUDIES ESTIMATING THAT IT OCCURS IN 60-80% OF THOSE WITH THE CONDITION

- Pain in Paget's disease may be directly related to the disease itself as a consequence of abnormal bone turnover. Indirect pain also occurs as the result of deformity of the bones, damage to the adjacent joints (osteoarthritis) or pressure on the nerves.
- Pain in the muscles, joints and bones, known as musculoskeletal pain, is common amongst the general population. It is caused by a variety of disorders and may be acute or chronic. Acute pain is related to direct damage caused by many factors including fracture and inflammation; it is self limiting and usually settles when healing is complete. Chronic pain is pain that will not go away and outlasts the normal healing process. Many things, including physical damage, genetic predisposition and social and emotional factors will impact on the development and intensity of chronic pain.
- There are diverse approaches to pain management including the evaluation of pain, use of medications, physical therapies such as physiotherapy and Trans electrical nerve stimulation (TENS) and the introduction of self management strategies.

EFFECTS OF MUSCULOSKELETAL PAIN



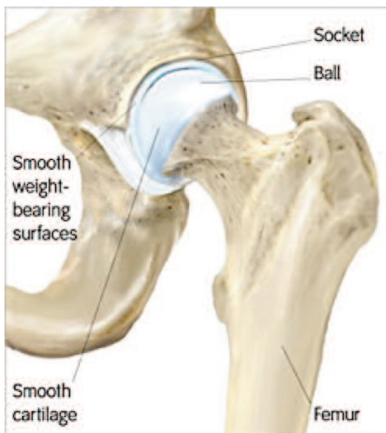
WHAT IS BONE PAIN?

- Bone pain at a specific site is detected by specialised transmitters located on the bone surface which then pass “messages” through the nervous system to the brain where the signals are recognised as pain.
- Pain in Paget’s disease may arise from the affected bone and is often described as “persistent and nagging”; it is usually present at rest, often at night, is not relieved by exercise and may respond poorly to pain killers.
- It is not fully understood why pain arises from pagetic bone but it may be caused by:
 - Increased blood supply to the affected area.
 - Stretching of the lining of bone
 - Fissure fractures that can occur in deformed weight bearing bones
 - Complete fracture
 - The extremely rare occurrence of a type of bone cancer, called sarcoma at the site of Paget’s disease

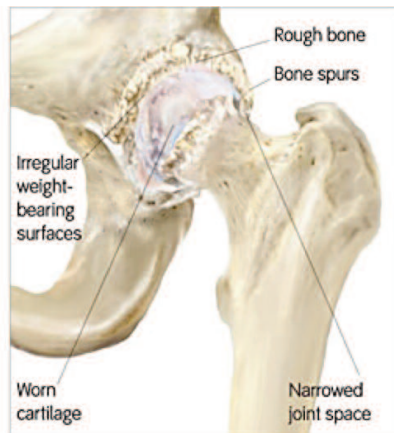
WHAT IS JOINT PAIN?

- Osteoarthritis is a common condition and those with Paget's disease are more prone to develop it at joints adjacent to pagetic bone due partly to the abnormal mechanical stresses placed on these joints. For example if a tibia (shin) becomes bowed this will cause shortening of the leg resulting in gait problems that can place extra mechanical stresses on the ankle and knee joints.
- When there are marked arthritic changes in a joint the joint surface becomes damaged, the underlying bone thickens, the cartilage becomes worn and eventually bone surfaces will rub against each other. This leads to increased stiffness and pain which is usually worse on moving the affected joint or on weight bearing.

NORMAL HIP JOINT



OSTEOARTHROTIC HIP JOINT



BACK PAIN

- Back pain is very common and in many instances is likely to be related to degenerative (“wear and tear”) changes throughout the spine and not just Paget’s disease.
- Non specific aches may arise from enlarged pagetic vertebrae and vertebral fractures can occur resulting in acute pain that may be felt in the spine and may also move round to the ribs.
- Occasionally an enlarged vertebra may press on the spinal nerves leading to pain felt down the legs and possible loss of function.

SKULL PAIN

- Paget’s disease can be associated with several symptoms including headaches and a band like tightness around the head resulting in an unpleasant sensation.

WHAT MEDICATION IS AVAILABLE FOR TREATING PAIN ASSOCIATED WITH PAGET'S DISEASE?

BISPHOSPHONATES

- Bisphosphonates are effective in reducing bone turnover and when pain is associated with active Paget's disease they will provide a degree of pain relief. The drugs commonly used in the UK are:
 - Risedronate 30mg (Actonel). Taken orally for 2 months (timing of repeat courses will depend on response to treatment).
 - Pamidronate (Aredia) given intravenously (directly into the bloodstream). Size and frequency of dose will depend on response to treatment.
 - Zoledronate 5 mg (Aclasta) given intravenously. Given annually or possibly less frequently (timing of repeat infusion will depend on response to treatment).
- In a small number of people bisphosphonates may initially cause a slight increase in bone and muscle pain but this usually subsides after a few days. Pain related specifically to Paget's disease usually responds well to treatment with bisphosphonates after a few months though response varies between individuals.
- Studies have shown that risedronate, pamidronate and zoledronate all lead to a reduction in bone pain 6 months after treatment.
- It is difficult to estimate the longer term effects of bisphosphonates on bone pain but one study has shown that pain attributed to Paget's disease remained reduced for two years after treatment.

WHAT OTHER TYPES OF MEDICATION MAY BE REQUIRED?

- Most people with Paget's disease require painkillers as well as bisphosphonates to control pain, especially if the disease has led to damage to the bones and joints. Examples of commonly used treatments are:
 - Paracetamol. One of the safest options which may prove beneficial.
 - Combination therapies e.g. co-codamol containing paracetamol and codeine may be used if paracetamol does not give sufficient relief.
 - Tramadol. Used for more severe pain that has not been helped by paracetamol or co-codamol
 - Non steroidal anti inflammatory drugs (NSAIDS). There is a wide variety of these including ibuprofen, diclofenac, naproxen, and a group of drugs known as Cox-2 selective inhibitors e.g. celecoxib and etoricoxib.
 - Pain killers are taken by mouth and in some cases applied as a cream to the painful area.

Although paracetamol and ibuprofen can be purchased without a prescription, it is very important that their use is discussed with a doctor or pharmacist, particularly if taken on a frequent and long term basis. The use of NSAIDS should be avoided in people who have previously suffered heart attacks, strokes or stomach ulcers.

- Pain in Paget's disease may also be associated with nerve damage and may respond to treatment with drugs such as gabapentin and amitriptyline that help this type of pain. These treatments may take several weeks before they begin to have an effect. Gabapentin may be associated with dry mouth, dizziness, diarrhoea and nausea and amitriptyline can cause blurred vision, dry mouth, low blood pressure and dizziness.

- Occasionally pain associated with Paget's disease can be so severe e.g. following fracture that a stronger opiate drug, such as morphine is recommended. Whilst very effective for severe pain this would be used for short periods only. It may be associated with nausea, vomiting, constipation and dependence.
- In the past calcitonin was frequently used to treat Paget's disease; although it is now rarely given as a standard treatment it is sometimes recommended for pain relief.

TAKING MEDICATION CORRECTLY

- Bisphosphonates and other medications will only help to reduce pain if they are taken at the correct time and in the correct way.
- Oral bisphosphonates need to be taken with water only on an empty stomach and food avoided for at least 30 minutes. To avoid problems with indigestion it is recommended to avoid lying down after taking the medication. It is important to take the full course of treatment prescribed.
- Other types of pain killers should be taken as recommended and it is particularly important to take paracetamol regularly (up to 8 tablets daily) and not just when the pain becomes intolerable.
- If any type of medication causes side effects it is vital to discuss this with a doctor, nurse or pharmacist.

IS SURGERY NEEDED TO TREAT PAIN ASSOCIATED WITH PAGET'S DISEASE?

- Few people with Paget's ever need surgery, but successful surgical management of severe complications can reduce pain and improve quality of life.
- Operations include:
 - Hip and knee replacements for associated osteoarthritis. This type of surgery can be very successful but the operation may be more technically challenging due to deformity and the altered bone quality. In addition to normal risks associated with an operation there is a slight increase in the risk of heterotopic ossification (formation of bone outside the skeleton) and non union of the bone.
 - Fracture surgery which may be more complex in those with Paget's disease because of the size and structure of the bone and in some cases healing may take longer.

Hip Replacement



Surgery to Fractured Tibia



- Osteotomy when a bone is cut to shorten, lengthen or change its position is occasionally used if there is marked deformity, fissure fractures or pain that has not responded to painkillers.

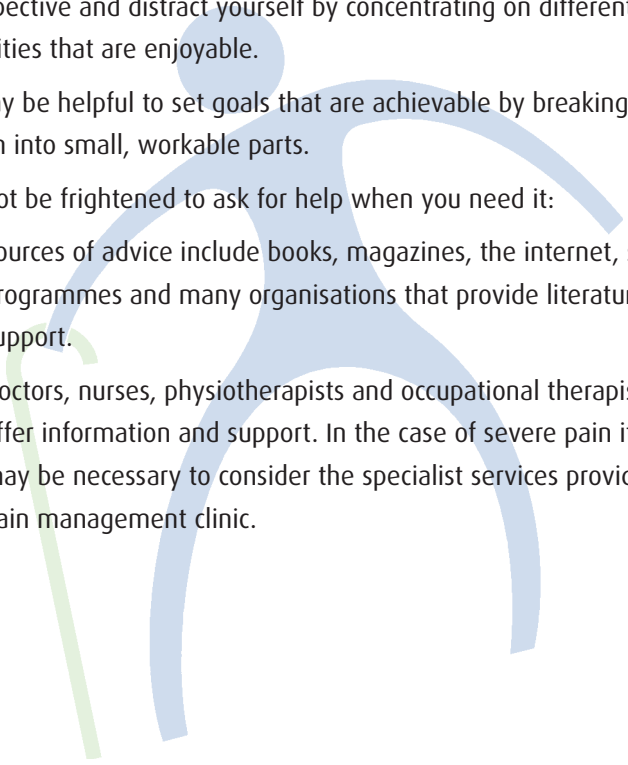
- Surgery to relieve pressure from the spinal cord and nerves caused by Paget's disease in the spine. This is usually relieved by drug treatment and an operation would rarely be required.
- The removal of a malignant tumour associated with Paget's Disease. This occurs very rarely.

WHAT OTHER METHODS MAY BE USED TO TREAT PAIN ASSOCIATED WITH PAGET'S DISEASE?

- Although there is limited evidence on other methods used to treat pain the following may be helpful:
 - Use of a Trans electrical nerve stimulation (TENS) machine
 - Physiotherapy
 - Heat and cold pads
 - Specialised footwear to balance any deformity
 - Use of walking aids
 - Massage
 - Acupuncture

WHAT CAN I DO MYSELF TO COPE WITH THE PAIN ASSOCIATED WITH PAGET'S DISEASE?

- Each individual perceives and subsequently copes with pain differently but the most important thing is to try and control the pain rather than letting it control you.
- It is important to try and focus away from the pain, keep it in perspective and distract yourself by concentrating on different activities that are enjoyable.
- It may be helpful to set goals that are achievable by breaking them down into small, workable parts.
- Do not be frightened to ask for help when you need it:
 - Sources of advice include books, magazines, the internet, self help programmes and many organisations that provide literature and support.
 - Doctors, nurses, physiotherapists and occupational therapists can offer information and support. In the case of severe pain it may be necessary to consider the specialist services provided by a pain management clinic.



This booklet has been reviewed by lay and medical trustees of The Association. Whilst it is intended to offer you information on Paget's disease and associated pain it is not designed to replace specific guidance you may receive from a health professional with respect to individual care.

References on specific sources of information are held by The Association and should you wish for further information on these please feel free to contact us.



WHAT OTHER ORGANISATIONS CAN PROVIDE SUPPORT?

Pain Concern

P.O. Box 13526

Haddington

EH41 4YD

Tel: 01620 822572

www.painconcern.org.uk

Provides a helpline and information for those who live with pain

The British Pain Society

Third Floor

Churchill House

35 Red Lion Square

London

WC1R 45G

Tel: 020 7269 7840

www.britishpainsociety.org

Largest UK multidisciplinary organisation involved in pain management.

Provides publications for those with pain.

Pain Relief Foundation

Clinical Sciences Centre

University Hospital Aintree

Liverpool

L9 7AL

Tel: 0151 5295820

www.painreliefoundation.org.uk

Information service to pain sufferers

NOTES





The Paget's Association, 323 Manchester Road, Walkden, Worsley, Manchester, M28 3HH
Tel.No: 0161 799 4646 Fax. No: 0161 799 6511
www.paget.org.uk E-mail: director@paget.org.uk

Registered Charity No.: 266071