

# Paget's disease 2: exploring diagnosis, management and support strategies

This article provides advice on diagnosing Paget's disease and outlines management options

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This is the second of a two-part unit on Paget's disease. Part 1 outlined the epidemiology, pathophysiology, causes and clinical features of the condition. This part outlines advice on diagnosis, drug therapy, surgery, other management options and emotional support.

## INTRODUCTION

Patients with Paget's disease may be managed in primary care or as outpatients. However, inpatient care may be necessary if surgery is needed, there are doubts about the diagnosis or a patient has marked pain.

Primary care nurses and those in relevant specialties should have basic knowledge of diagnosing the condition, commonly prescribed drug therapy and surgical options.

## DIAGNOSIS

Paget's disease may present with obvious signs or symptoms or may be an incidental finding of investigating other conditions.

The diagnosis can usually be confirmed by X-ray showing bone enlargement, cortical thickening and sclerotic changes in affected bones. X-rays can also show fractures and provide information on joint changes. Isotope bone scans are more sensitive in detecting the disease and may also be helpful in defining its extent.

Patients can be reassured that repeat X-rays or scans will usually be unnecessary unless new symptoms develop or symptoms become increasingly severe.

Since Paget's disease is associated with increased bone turnover, it is expected that markers of bone turnover will be raised in active disease. Serum alkaline phosphatase is usually elevated in 85% of patients with untreated Paget's disease and generally this level will be highest when

## LEARNING OBJECTIVES

1. Know the different ways of diagnosing Paget's disease.
2. Understand drug treatments for Paget's disease and how they are given.

several sites are involved (Eastell, 1999).

Elevated alkaline phosphatase may be associated with liver disease and, if this is present, it may be appropriate to measure bone-specific alkaline phosphatase. In specialist centres, urine tests may be used to assess disease activity. Once treatment has been given, alkaline phosphatase level decreases. Repeating the blood test is a way of measuring response to therapy.

On rare occasions a bone biopsy may be performed, particularly if malignant changes are suspected.

## DRUG TREATMENT

Treatment is recommended when patients have bone pain localised to an affected site. In the absence of pain, Selby et al (2002) suggested that treatment should be considered to prevent further complications but there is a lack of consensus on this.

Bisphosphonates, oral or intravenous, are commonly used. These bind to the surface of hydroxyapatite crystals within bone and trigger the death of osteoclasts, which cause the increased bone turnover. Oral therapy licensed in the UK includes risedronate, tiludronic acid and etidronate, with risedronate being the most potent and usually the most effective (Siris and Roodman, 2006). Pamidronate and zoledronic acid (the most recent drug) are given intravenously. Randomised comparative studies have reported no significant difference between the effects of different bisphosphonates on pain control (Miller et al, 1999; Siris et al, 1996). However, one recent study has shown that zoledronic acid was slightly more

effective than risedronate (Reid et al, 2005).

Both zoledronic acid and pamidronate can cause acute phase reactions. Oral treatment can be prescribed in primary care but may lead to gastrointestinal problems and some patients find the dosing regimen restrictive.

Nurses have a key role in explaining drug effects and encouraging adherence.

Common side-effects of oral treatment include indigestion and nausea and patients may have problems complying with dosing instructions. Oral drugs are sometimes prescribed for a set period of time, for example risedronate 30mg daily for two months. Taking the drugs continuously could impair bone mineralisation.

Risedronate in a different dose is prescribed for osteoporosis, so it is vital to check the right regimen is used. For details on oral and IV drugs, see Tables 1 and 2 in Portfolio Pages at [nursingtimes.net](http://nursingtimes.net).

In response to treatment, the alkaline phosphatase level generally decreases, with a more rapid and marked reduction in patients treated with zoledronic acid (Reid, 2006). It may be useful to perform a blood test after six months to reassure patients that treatment has been successful. Drug effectiveness can also be measured in terms of pain reduction, which usually occurs about three months after starting treatment.

Although bisphosphonates are very effective in reducing bone pain in Paget's disease, patients may have complications such as deformity and osteoarthritis that do not respond well to them. These patients may require analgesic and anti-inflammatory drugs and advice and encouragement to take them regularly. Non-pharmacological approaches, such as acupuncture, physiotherapy and transcutaneous electrical nerve stimulation (TENS), are used for pain control, but effectiveness of this is based on anecdotal reports, not rigorous assessment.

## SURGERY

Although few patients need surgery, surgical management of severe orthopaedic complications has improved quality of life.



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The most common indication for surgery is joint replacement for associated osteoarthritis. Other indications include fracture fixation, osteotomy to correct bone deformity, and surgery to correct spinal stenosis or to resect a malignant tumour.

Symptomatic arthritis of the hip needing arthroplasty occurs in approximately 10% of patients with Paget's disease (Graham and Harris, 1971). Most studies on outcomes of total joint arthroplasty have reported similar results to those achieved in patients without the condition but surgery at this site may be more technically challenging because of deformity and altered bone quality (Parvizi et al, 2006). In addition to normal surgical risks, there is a slight increased risk of heterotopic ossification (formation of bone outside the skeleton) and non-union of the trochanter.

Fracture surgery may be more complex in patients with Paget's disease because of the size and structure of the bone and healing may be more protracted. In femoral neck fractures treated with internal fixation, non-union is common so the best option in these patients is prosthetic replacement.

Osteotomy is occasionally used if there is marked deformity, fissure fractures and pain that has not responded to medical management. Advances in the field of external fixation, such as spatial frames and Ilizarov bone fixation technique, have

allowed correction of complex deformities.

Patients who develop neurological complications associated with Paget's disease in the lumbar spine should be treated medically. On rare occasions, spinal decompression may be performed to relieve pain and improve mobility.

A malignant lesion in a pagetic bone is a rare yet devastating consequence of the condition. Surgical resection aiming for wide margin is recommended but the outcome is usually compromised because of the high grade of the tumour, delay in diagnosis, patient age and the ineffectiveness of adjuvant chemotherapy.

Parvizi et al (2006) suggested that the vascularity of pagetic bone may mean increased blood loss during surgery. To minimise this risk, patients are usually given bisphosphonate treatment before surgery. It is vital that fluid balance and blood loss are monitored carefully post-operatively.

#### OTHER MANAGEMENT OPTIONS

Patients report using a wide range of options including acupuncture, TENS, aromatherapy and massage. Some of these appear beneficial but they have not been systematically researched.

Specific problems such as limb shortening and deformity may be helped by shoe raises and walking aids.

Following surgery, patients may also need to consider adaptations in the home, such as a raised toilet seat and handrails.

#### EMOTIONAL SUPPORT

While Paget's disease is rarely life-threatening, it can show some of the classic hallmarks of long-term conditions. It is inveterate, caused by non-reversible pathology, may leave residual disability and can need a long period of supervision and observation.

Some patients may be confused about the nature of the condition and concerned about the possibility of future complications including fracture and deformity. Some are frightened by perceived or actual side-effects of medication or may have unrealistic expectations of the outcome of therapy.

Pain is a common feature of Paget's disease and, if this becomes chronic, it may lead to anxiety, social isolation and failure of coping mechanisms.

As part of a wider healthcare team, nurses may assume the key role of counsellor to

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