A clinical Guideline*, for the Diagnosis and Management of Paget’s Disease of Bone in Adults, was published in 2019, on behalf of the Paget’s Association, the European Calcified Tissue Society (ECTS), and the International Osteoporosis Foundation (IOF).

Commissioned by the Paget’s Association, the Guideline is a result of work carried out by the Guideline Development Group (GDG), which was established in 2016 and coordinated by our Vice-Chairman, Dr Stephen Tuck. The GDG comprised of a group of experts in the field, with patient involvement, and has been endorsed by the ECTS, the IOF, the American Society of Bone and Mineral Research, the Bone Research Society (UK), and the British Geriatric Society.

The GDG identified six key questions which were used to form the basis of the Guideline. Based on these, a full literature search was performed, and the quality of the evidence was assessed using explicit criteria.

The key questions were:
1. Which measurements or tools are effective in the identification and diagnosis of Paget’s disease?
2. Which diagnostic measurements or tools are effective in predicting the clinical response to treatment?
3. What are the indications for drug treatment in Paget’s disease?
4. What are the effects of drug treatment in Paget’s disease?
5. For individuals prescribed pharmacological interventions, what is the optimal duration or mode of treatment?
6. What are the effects of non-drug treatment in Paget’s disease?

Further Research Needed
The GDG found that there was a lack of research on patient-focused clinical outcomes and so they were able to identify several areas where further research is needed. This will assist the Paget’s Association when consideration is given to applications for research funds.

Open Access
The Guideline was published in 2019, in the Journal of Bone and Mineral Research* and whilst access is often restricted to those who subscribe to the journal, the Association provided funds to ensure that the full document has unrestricted, online access. This allows everyone access to the full breadth of information within the Guideline, providing the opportunity for all health professionals to be fully informed, prior to discussing options with their patients.

Download the Document
The full Guideline is available to download from our website www.paget.org.uk

Recommendations
Several recommendations are made in the Guideline. 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.

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.

*The information has been reproduced from the following paper:
Diagnosis and Monitoring of Paget’s Disease of Bone

This diagram*, taken from the clinical Guideline, provides easy reference information to assist professionals in diagnosing and monitoring Paget’s disease.

Main clinical presentation of PDB

- “Bone” pain
- Bone deformity or pathological fracture

Symptomatic patient

Raised serum total alkaline phosphatase (ALP) with normal liver function test.

Asymptomatic patient

PDB detected on imaging conducted for investigation of another disorder

PDB diagnosis

Targeted X-ray of site

Plain X-ray of

Characteristic X-ray features of PDB

- Bone expansion
- Cortical thickening
- Osteolytic areas
- Trabecular thickening
- Bone deformity
- Loss of distinction between cortex and medulla
- Osteosclerosis

Skull and facial bones

Abdomen

Tibiae

are recommended as initial screening in patients suspected to have PDB

Radionuclide bone scintigraphy (RBS)

RBS in addition to targeted X-ray, are recommended as a means of fully and accurately defining the extent of metabolically active PDB.

Other imaging

- Other imaging: magnetic resonance imaging (MRI) or computed tomography (CT)
  - Not recommended for the diagnosis of PDB.
  - Recommended to assess disease complications.

Assessment of metabolic activity

Serum total ALP is recommended as a first line biochemical test in combination with liver function tests in screening for the presence of metabolically active PDB:

If total ALP values are normal and clinical suspicion of metabolically active PDB is high, measurement of BALP, PINP or uNTX may be considered.


### Management of Paget’s Disease of Bone

To aid professionals in their management of Paget’s disease, the authors of the clinical Guideline included this easy reference guide.*


<table>
<thead>
<tr>
<th>Symptom</th>
<th>Recommendation</th>
<th>Quality of evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bone pain</td>
<td>Bisphosphonates are recommended for the treatment of bone pain associated with Paget’s disease</td>
<td>MODERATE</td>
</tr>
<tr>
<td></td>
<td><strong>1st choice</strong></td>
<td></td>
</tr>
<tr>
<td>QoL (Quality of life)</td>
<td>There is insufficient evidence to recommend bisphosphonates as a means of improving quality of life to a clinically meaningful extent in Paget’s disease</td>
<td>VERY LOW</td>
</tr>
<tr>
<td>Prevention of fractures</td>
<td>There is insufficient evidence to recommend bisphosphonates as a means of preventing fractures in Paget’s disease.</td>
<td>VERY LOW</td>
</tr>
<tr>
<td>Progression of osteoarthritis</td>
<td>There is insufficient evidence to recommend bisphosphonates as a means of preventing progression of osteoarthritis in Paget’s disease.</td>
<td>VERY LOW</td>
</tr>
<tr>
<td>Progression of deafness</td>
<td>There is insufficient evidence to recommend bisphosphonates as a means of preventing progression of hearing loss in Paget’s disease.</td>
<td>VERY LOW</td>
</tr>
<tr>
<td>Blood loss</td>
<td>There is insufficient evidence to recommend bisphosphonates as a means of preventing bone deformity in Paget’s disease.</td>
<td>VERY LOW</td>
</tr>
<tr>
<td>Bone deformity</td>
<td>Treatment aimed at improving symptoms is recommended over a treat-to-target strategy aimed at normalising total ALP in Paget’s disease</td>
<td>MODERATE</td>
</tr>
</tbody>
</table>

#### Treatment of PDB complications

- **Surgery** is recommended for fixation of fractures in Paget’s disease but there is insufficient information to recommend one type of surgical treatment over another.
- **Joint replacement surgery and osteotomy** are recommended for the treatment of osteoarthritis resistant to medical therapy in patients with Paget’s disease.
- **Suppressing metabolic activity in PDB**
  - Bisphosphonates are highly effective at reducing concentrations of biochemical markers of bone remodelling.
  - The clinical benefit of giving bisphosphonates with the primary aim of suppressing bone turnover is unknown.

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Over the following months, this treatment often normalises the abnormal bone remodelling and one dose can be effective for many years. Pamidronate

Pamidronate is also an effective treatment but slightly less effective than zoledronic acid. Also given in the form of an infusion, the usual dose is 60mg, on three separate occasions. It can be used in people with borderline kidney function where zoledronic acid is not advisable.

Risedronate

Risedronate is given in tablet form. The usual dose is 30mg daily, taken for two months. The tablets need to be taken on an empty stomach. Risedronate is an effective treatment but is not quite as good as zoledronic acid at helping pain. Also the effect doesn't last quite as long. It is useful for people who don’t want to have injections and in those with borderline kidney function where zoledronic acid is not advisable.

Clinical Guideline

In 2019, a new clinical Guideline* for the diagnosis and management of Paget’s disease 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, which enable health professionals to discuss all options with the patient.


Further Information

This leaflet has been produced by the Paget’s Association, a charity dedicated to supporting those affected by Paget’s disease, as well as funding research into the condition. More detailed written information is available from the Association. The charity also provides a nurse helpline, support network, information events and a quarterly newsletter.

How do I Contact the Paget’s Association?

Email helpline@paget.org.uk

Telephone 0161 799 4646

The Paget’s Association

Suite 5, Moorfield House

Moorside Road, Swinton

Manchester, M27 0EW

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Paget’s Disease

This booklet reflects the current 2019 clinical Guideline. It is available to individuals and to professionals for their patients.

Paget’s Disease and Pain

This leaflet is ideal for information stands.

Paget’s Disease – The Facts

Paget’s Disease – Investigations Explained

Paget’s Disease of Bone – The Essential Facts

Paget’s Helpline

Contact our Nurse Helpline for information, advice and support, regarding any aspect of Paget’s disease.

Email helpline@paget.org.uk

Telephone 0161 799 4646

Information Events

Our information events provide an opportunity to hear presentations from experts in Paget’s disease, raise questions and meet with one another. Contact us for details.

Membership

We welcome both UK and overseas members. Health professionals, researchers, patients and carers can join the Paget’s Association online or request a membership application form using the contact details below.

Our membership pack includes information booklets, copies of our magazine, information sheets on various aspects of Paget’s disease and a complete list of the UK Centres of Excellence. In addition, our Paget’s Passport provides individuals with an opportunity to keep a record of test results, any treatment received and how Paget’s disease affects them.

For full details and a complete list of current membership benefits, please visit our website.

Paget’s News

Members of the Paget’s Association receive our quarterly magazine, ‘Paget’s News’, which contains information, personal experiences and networking opportunities, as well as research news and advice relating to Paget’s disease.

Research

The Paget’s Association funds and supports research into various aspects of Paget’s disease to help improve the quality of life for those with the condition and their families.

Information regarding our grants and student research bursaries can be found on our website.

Online

www.paget.org.uk

Get in Touch

Telephone 0161 799 4646  Email helpline@paget.org.uk

Write to: Paget’s Association, Suite 5, Moorfield House, Moorside Road, Swinton, Manchester, M27 0EW

Paget’s Association

Registered Charity No. 266071