

Paget's Disease of Bone – A Clinical Guideline

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.

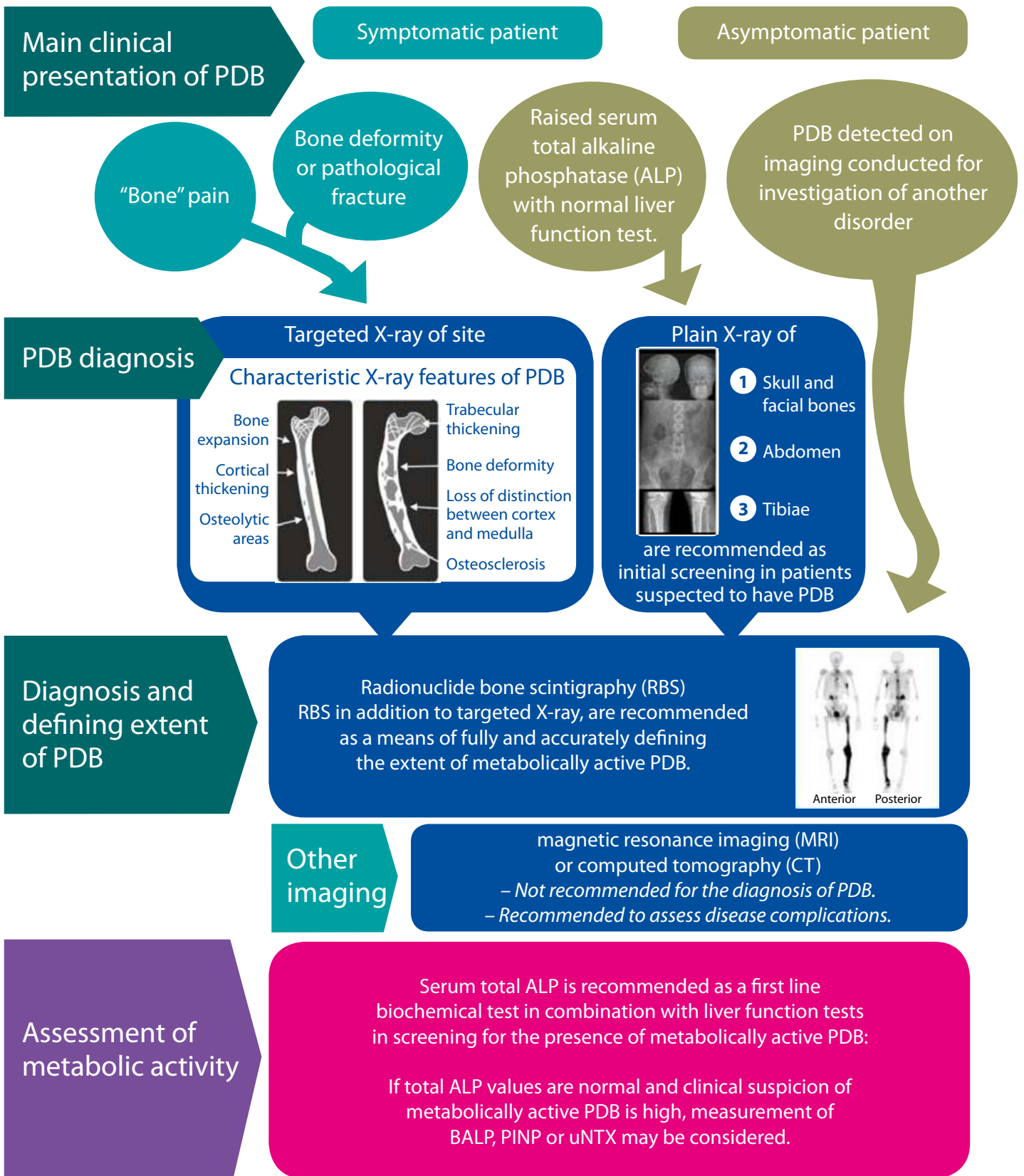
- 1) Radionuclide bone scans, in addition to targeted radiographs, are recommended as a means of fully and accurately defining the extent of the metabolically active disease in patients with Paget's Disease of Bone.
- 2) Serum total alkaline phosphatase (ALP) is recommended as a first line biochemical 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:*

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: Journal of Bone Mineral Research. Vol. 34, p 579-604.

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.














ALP: alkaline phosphatase, BALP: bone specific alkaline phosphatase, GGT: gamma-glutamyl transferase, PDB: Paget's disease of bone; PINP: procollagen type 1 amino-terminal propeptide; uNTX: urine N-terminal telopeptide.

*Reproduced from Ralston, S. H. et al, (2019), Diagnosis and Management of Paget's Disease of Bone in Adults: A Clinical Guideline. Journal of Bone Mineral Research. Vol. 34, p 579-604.

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*.

 <p>Bone pain</p>	<p>Bisphosphonates are recommended for the treatment of bone pain associated with Paget's disease</p> <p>Quality of evidence: MODERATE</p> <p>1st choice Zoledronic acid</p>	 <p>Established Paget's</p>	<p>Treatment aimed at improving symptoms is recommended over a treat-to-target strategy aimed at normalising total ALP in Paget's</p> <p>Quality of evidence: MODERATE</p>
<p>QoL</p> <p>Health related quality of life</p>	<p>There is insufficient evidence to recommend bisphosphonates as a means of improving quality of life to a clinically meaningful extent in Paget's disease</p> <p>Quality of evidence: VERY LOW</p>	<p>Treatment of PDB complications</p>	
 <p>Prevention of fractures</p>	<p>There is insufficient evidence to recommend bisphosphonates as a means of preventing fractures in Paget's disease.</p> <p>Quality of evidence: VERY LOW</p>	 <p>Neurological symptoms</p>	<p>→ A trial of bisphosphonate or calcitonin may be considered as part of the treatment package in patients with PDB who have evidence of spinal dysfunction.</p> <p>→ Spinal surgery may be considered for patients with Paget's disease who develop spinal stenosis and cord compression.</p> <p>Quality of evidence: VERY LOW</p>
 <p>Progression of osteoarthritis</p>	<p>There is insufficient evidence to recommend bisphosphonates as a means of preventing progression of osteoarthritis in Paget's disease.</p> <p>Quality of evidence: VERY LOW</p>	 <p>Treatment of fractures</p>	<p>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.</p> <p>Quality of evidence: VERY LOW</p>
 <p>Progression of deafness</p>	<p>There is insufficient evidence to recommend bisphosphonates as a means of preventing progression of hearing loss in Paget's disease.</p> <p>Quality of evidence: VERY LOW</p>	 <p>Joint replacement</p>	<p>Joint replacement surgery and osteotomy are recommended for the treatment osteoarthritis resistant to medical therapy in patients with Paget's disease</p> <p>Quality of evidence: VERY LOW</p>
 <p>Blood loss</p>	<p>There is insufficient evidence to recommend bisphosphonates as a means of reducing blood loss prior to elective orthopaedic surgery.</p> <p>Quality of evidence: VERY LOW</p>	<p>Suppressing metabolic activity in PDB</p>	
 <p>Bone deformity</p>	<p>There is insufficient evidence to recommend bisphosphonates as a means of preventing bone deformity in Paget's disease.</p> <p>Quality of evidence: VERY LOW</p>	 <p>Metabolic activity</p>	<p>Bisphosphonates are highly effective at reducing concentrations of biochemical markers of bone remodelling</p> <p>Quality of evidence: HIGH</p> <p>The clinical benefit of giving bisphosphonates with the primary aim of suppressing bone turnover is unknown</p> <p>Quality of evidence: VERY LOW</p>

*Reproduced from Ralston, S. H. et al, (2019), Diagnosis and Management of Paget's Disease of Bone in Adults: A Clinical Guideline. Journal of Bone Mineral Research. Vol. 34, p 579-604.

Information, Support and Research

The Paget's Association is a charity, which provides information and support to all those affected by Paget's Disease of Bone. The Association also funds quality research, raises awareness, and supports professionals to achieve excellence in care and research.

Booklets and Leaflets



Paget's Disease – The Facts

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



Paget's Disease and Pain



Paget's Disease – Investigations Explained



Paget's Disease of Bone – The Essential Facts

This leaflet is ideal for information stands.

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

Paget's
Association

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