# key points

SELECTED BY

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**Paget's disease of bone is a metabolic disease in which** focal abnormalities of bone remodelling occur in one or more skeletal sites. The affected bones enlarge and may become deformed and this can lead to complications including bone pain, pathological fractures, secondary osteoarthritis, deafness and nerve compression syndromes.

**Paget's disease is seldom diagnosed below the age of 50** but the incidence doubles every 10 years thereafter to 5-7/10,000 population per year by the ninth decade in the UK. Many people with Paget's disease do not experience symptoms and it is estimated that only 7-15% of people with X-ray evidence of Paget's disease come to medical attention. Men are affected more often than women in a ratio of 1.4:1. The three main risk factors are age, male gender and family history. People with a first-degree relative with Paget's disease have a 7-fold increased risk of developing the disease.

**The most common presenting features in those that do** present clinically are bone pain (52.2%), bone deformity (21.5%), deafness (8.9%) and pathological fractures (8.5%). Some patients experience pain at rest which is dull and localised to an affected site. Others can have pain which is worse with weight bearing or more localised because of pseudofractures. Paget's disease may be picked up by an incidental finding on blood tests or by imaging carried out for another reason. Rarely, Paget's disease may present with osteosarcoma (0.3% of patients) which usually manifests with a local increase in swelling and pain at an affected site most commonly the pelvis, femur or humerus.

**A diagnosis of Paget's disease can be confirmed by X-ray** which shows the characteristic features of osteolysis alternating with osteosclerosis, cortical thickening, bone expansion and bone deformity. A radionuclide bone scan is the most sensitive diagnostic investigation. However, if this is unavailable, plain X-rays of the abdomen (including the ribs and femoral heads), both tibias and the skull (including facial bones) will detect 93% of cases. Routine biochemistry can also be helpful in the diagnosis of Paget's disease that is metabolically active; typically patients will have an isolated elevation of ALP. However, when Paget's disease affects a single bone, ALP values can be normal.

**Patients who have bone deformity or symptoms that** might be due to Paget's disease should be referred to secondary care. The main indication for medical treatment is bone pain localised to an affected site where there is evidence of increased metabolic activity. Bisphosphonates are the treatment of choice and risedronate, zolendronic acid and pamidronate are currently licensed for the treatment of Paget's disease in the UK. Vitamin D deficiency should be corrected prior to initiating bisphosphonate therapy to reduce the risk of hypocalcaemia. Treatment is monitored both clinically and by serial ALP measurements. Orthopaedic surgery is required in around 10% of patients. The most common reasons are fracture repair, hip or knee arthroplasty or spinal surgery to correct spinal stenosis.