



FIGURE 74-2 Typical radiologic abnormalities Paget's disease. **A**, Compare the normal skull (*top*) with the skull (*bottom*) with the classic cotton-wool appearance, an enormously expanded calvarium, and osteosclerosis of the petrous bones. **B**, Classic asymmetrical involvement of the pelvis with a mixture of lytic and blastic lesions. **C**, Bowing deformity of the femur with a markedly thickened cortex. **D**, A whole body radionuclide scan demonstrates polyostotic Paget's disease.

patients, elevated concentrations of total serum alkaline phosphatase is an adequate and sensitive indicator of disease activity. However, the serum level of bone-specific alkaline phosphatase may be a more sensitive marker than serum total alkaline phosphatase for assessing disease in patients with low levels of disease activity. A bone scan at the time of diagnosis can define the location and extent of lesions (see Fig. 74-2D). Radiographs of the affected areas confirm Paget's disease, and they are useful for evaluating complications and local disease progression (Fig. 74-2A-C).

The two major goals of therapy are to relieve symptoms and prevent complications. Indications for treatment include alleviating symptoms (e.g., bone pain, headache, neurologic complications), decreasing blood flow preoperatively to minimize bleeding during elective surgery on a pagetic site, managing hypercalcemia in an immobilized patient with severe Paget's disease, and preventing future complications of progressive local

disease, such as bowing deformities of the long bones, hearing loss as a result of temporal bone involvement, and neurologic complications resulting from foramen magnum or vertebral involvement. Treatment of Paget's disease usually involves a combination of nonpharmacologic therapy (i.e., physical therapy) and pharmacologic therapy, including antiresorptive agents and analgesics for pain management.

The mainstay of therapy is bisphosphonates, including potent aminobisphosphonates such as zoledronate, which decrease bone resorption at pagetic sites by inhibiting osteoclasts. Calcium and vitamin D supplementation is recommended for patients taking the more potent bisphosphonates to prevent hypocalcemia or secondary hyperparathyroidism.

Orthopedic surgery may be indicated when a complete fracture occurs through pagetic bone. Surgery also is indicated for realignment of a severely arthritic knee and for total joint arthroplasty in a severely affected hip or knee.