# Miscellaneous Conditions with Manifestations in the Hand and Wrist

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#### Abstract

In any textbook dealing with conditions affecting a particular anatomical site, there will be some which do not fit conveniently under the standard chapter headings. The purpose of this chapter is to review these miscellaneous localised and systemic disorders that may affect the hand and wrist.

# 1 Introduction

In any textbook dealing with conditions affecting a particular anatomical site, there will be some which do not fit conveniently under the standard chapter headings. The purpose of this chapter is to review these miscellaneous localised and systemic disorders that may affect the hand and wrist.

## 2 Paget's Disease

Paget's disease of bone is a benign disorder affecting 3–4% of the population over 40 years of age predominantly in Caucasian races (Smith et al. 2002). There is recent evidence of decreasing prevalence (Cundy 2006). The abnormality in Paget's disease is excessive and abnormal remodelling of bone. Three pathologic phases have been described: the lytic phase, the mixed phase and the blastic phase. Initially there is osteolysis, followed by trabecular and cortical thickening and enlargement in the mixed phase of the disease, followed by sclerosis in the blastic phase. Paget's disease is usually polyostotic and asymmetric. **Fig. 1** Paget's disease. There is bony sclerosis and expansion of the proximal phalanx of the middle finger



Polyostotic disease (65–90%) is more frequent than monostotic disease.

Involvement in the hand and wrist has been reported infrequently, although bone scintigraphy studies reveal hand localisation varying from 6.4 to 11.6% (Friedman et al. 1982). Clinically, Paget's disease in the hands is usually asymptomatic; however, several cases with pain in the hands have been reported (Friedman et al. 1982). When Paget's disease occurs in the hand, there is usually a polyosinvolvement (Haverbush et al. totic 1972). Monostotic localisation in the hand is less frequent (De Smet et al. 1994; Rodriguez-Peralto et al. 1994; Calif et al. 2007). It typically affects the phalanges and metacarpals, with carpal involvement rare and distal phalangeal involvement almost unknown. The most common radiographic pattern of disease in the hand is sclerosis (Fig. 1; Haverbush et al. 1972) and is seen in 45% of cases (Friedman et al. 1982). However, all patterns as found elsewhere in the body (cortical destruction, bone expansion, coarsened trabeculae and bone sclerosis) may be seen in the hand (Friedman et al. 1982). Sarcomatous transformation is the most sinister complication of Paget's disease and is rarely seen in the hand. A review by Lopez et al. (2003) looked at 340 cases of Paget's sarcoma in the literature and identified no cases in

the hand and wrist. Isolated cases of malignant transformation have, however, been recorded (Friedman et al. 1982). The MR imaging appearances of Pagetic marrow can be variable (Smith et al. 2002) but a useful diagnostic sign is the preservation of the hyperintense marrow signal on T1-weighted images (Fig. 2; Boutin et al. 1998; Kaufmann et al. 1991; Sundaram et al. 2001; Vande Berg et al. 2001).

# 3 Sarcoid

Sarcoidosis is a granulomatous disorder of unknown cause that affects multiple organ systems. It is characterised by the presence of multiple non-caseating granulomas, most commonly seen in the lungs, lymph nodes, skin and eyes. The diagnosis is made by a combination of clinical, radiological and histological features.

Skeletal involvement has been reported in 1-13% of patients with the disease. Radiographic evaluation is often limited to the hands or feet, which demonstrate predominantly osteolytic lesions. The hand is the commonest site of skeletal sarcoidosis. The lesions may be unilateral or bilateral, but symmetrical involvement is unusual. Lesions in the hand are most commonly seen in the middle and distal phalanges, and less often in the proximal phalanges and metacarpals (Van Linthoudt and Ott 1986). There are several types of lesion that are seen. The characteristic lesion is the alteration of the trabecular lattice resulting in a honeycomb lytic appearance (Fig. 3). Cystic lesions may also occur leading to a punchedout appearance that may be located centrally or eccentrically (Fig. 3). Osseous destruction may appear rapidly if these lesions are associated with cortical erosion, which can lead to pathological fracture (Neville et al. 1976, 1977). Periosteal new bone formation is uncommon.

Acro-osteolysis (terminal phalangeal resorption) has been reported in sarcoid involvement of the hands (Fig. 4). The finding is non-specific. In both the hand and the wrist, tenosynovitis is a recognised complication of sarcoidosis (Katzman et al. 1997; Moore 2003). Other soft tissue changes can be seen on MR imaging. These include focal and diffuse muscle lesions, soft tissue infiltration and masses (Moore 2003). MR imaging may also be useful in the

**Fig. 2** a Radiograph and b coronal T1-weighted MR imaging of Paget's disease of the distal radius. The radiograph was initially misinterpreted as showing a giant cell tumour but the preservation of the hyperintense marrow signal in b is highly suggestive of Paget's disease





Fig. 3 Sarcoidosis. There is involvement of the proximal and middle phalanges of both hands. The appearances are more punched-out on the *left* with honeycomb osteolysis on the *right* 



**Fig. 4** Sarcoidosis. At presentation, **a** showing eccentric soft tissue swelling and lacy osteolysis of the middle phalanx of the middle finger; 14 years later, **b** showing soft tissue swelling of both index and middle fingers and progression of disease with terminal phalangeal resorption and further destruction of the middle phalanx of the middle finger. There is now also involvement of the proximal phalanges of both fingers

differential diagnosis, for example in the evaluation of soft tissue nodules distinguishing gout from sarcoidosis. Intermediate-weighted MR images typically show tophi as hypointense, whereas sarcoid nodules will usually be hyperintense.

Lofgren syndrome is a well-recognised manifestation of sarcoidosis, with patients exhibiting arthralgias, erythema nodosum and bilateral hilar lymphadenopathy. Polyarthritis involving the interphalangeal joints, wrists, elbows, knees or ankles is common. Monoarthritis and effusion are uncommon. Ten to thirty-five percentage of sarcoidosis patients will also experience joint symptoms due to granulomatous arthritis with a chronic transient or relapsing arthropathy. Sausage-like dactylitis of the fingers can also occur (Fig. 4). Pain is usually not severe. The uptake of FDG in PET scanning from sarcoid tissue is non-specific. However, Brudin et al. (1994) have described how the degree of FDG uptake correlates with disease activity, and although FDG PET is not useful for initial diagnosis, it could be used for evaluating the extent of active disease and monitoring response to therapy (Love et al. 2005).



**Fig. 5** Reflex sympathetic dystrophy. There is intense juxtaarticular osteopenia mimicking an acute inflammatory arthropathy

## 4 Reflex Sympathetic Dystrophy

Reflex sympathetic dystrophy (RSD) is a distinct condition that occurs in a variety of clinical situations. There are several terms that are used to describe the same condition including: causalgia, algodystrophy, Sudeck's atrophy, post-traumatic osteoporosis and complex regional pain syndrome. Although the cause is often idiopathic, many visceral, musculoskeletal, neurological and vascular conditions can cause RSD with trauma the most common. The most frequent site for RSD is in the hand. Clinical symptoms include stiffness, pain, tenderness and weakness. Radiographic changes range from soft tissue swelling with juxta-articular osteopenia (Fig. 5) to a more generalised osteopenia (Fig. 6). The former can mimic the



Fig. 6 Reflex sympathetic dystrophy. In this chronic case affecting the right hand, there is intense generalised osteopenia with multiple intracortical lucencies. Note the normal bone density of the unaffected left hand

presentation of an acute inflammatory arthropathy but in RSD there is no joint space narrowing or erosions. Both three-phase bone scintigraphy and MR imaging can be used to assess the "activity" of this disorder over time (Park et al. 2009).

# 5 Haematological and Marrow Disorders

This section covers the imaging manifestations in the hand and wrist of haematological and marrow disorders including haemophilia, sickle cell anaemia and thalassaemia.

## 5.1 Haemophilia

Haemophilia and related bleeding disorders are a group of conditions that are characterised by abnormality in blood coagulation caused by deficiency in certain plasma clotting factors. The two most common types associated with intraosseous and intraarticular bleeding are haemophilia A (deficiency in clotting factor VIII) and haemophilia B (deficiency in clotting factor IX). Haemarthrosis occurs in 75-90% of people with haemophilia and in time leads to a haemophilic arthropathy. This begins between the ages of 2-3 years and increases in frequency till the ages of 8-13 years. The joints most commonly affected include the knee, elbow, ankle, hip and shoulder. Usually, a single joint is involved in each episode, though eventually multiple joints become affected in this disease. Intraarticular bleeding is rare in the hand and wrist. A well-recognised but rare complication occurring in approximately 2% of patients with severe haemophilia is the so-called haemophilic pseudotumour. These result from chronic recurrent haemorrhage that may be intraosseous, subperiosteal or arise in the soft tissues. The bones most frequently involved, in order of decreasing frequency, are the femur, pelvis, tibia and the small bones of the hand

**Fig. 7** Haemophilic pseudotumour proximal phalanx of the thumb. The age of the patient and the radiographic appearances mimic an aneurysmal bone cyst



**Fig. 8** Sickle cell disease. A young child with severe hand–foot syndrome showing medullary lucencies and periosteal new bone formation involving all the long bones of the hand and the distal ulna. The visible mineralised carpal bones are not involved



Fig. 9 Thalassaemia. The marrow hyperplasia is causing severe generalised osteopenia, trabeculation and minor expansion of the long bones

(Park and Ryu 2004). Cases have also been reported affecting the distal forearm bones (Ahlberg 1975; Shaheen and Alasha 2005). Cases involving the hand and wrist are usually of intraosseous origin, whereas pseudotumours arising in the lower limbs and pelvis tend to be extraosseous with secondary pressure erosion of adjacent bones. In the long bones of the hand, intraosseous haemophilic pseudotumour shows an expansile benign-appearing lytic lesion mimicking amongst other pathologies an aneurysmal bone cyst (Fig. 7; Shaw and Wilson 1993).

## 5.2 Sickle Cell Disease

Sickle cell disease is one of the hereditary haemoglobinopathies characterised by an abnormal haemoglobin chain (HbS). Clinical manifestations of the disease include painful crises affecting the bones and joints of the extremities. In children with sickle cell anaemia, 30% will present with hand-foot syndrome



Fig. 10 Two different cases of neurofibromatosis. a Multiple soft tissue lumps due to subcutaneous neurofibromas and bone erosions in the index and middle fingers. b There is focal gigantism and sclerosis of the index and middle metacarpals and fingers

between the ages of 6–24 months with pain and swelling of the hands due to marrow infarction (Watson et al. 1963). The radiographic changes are those of a dactylitis with osteolysis and periosteal new bone formation (Fig. 8; Bohrer 1970). Hand–foot syndrome is rare after 6 years of age because the red marrow recedes from the distal tubular bones of the hand and is replaced by yellow marrow with reduced oxygen requirements. Severe dactylitis may cause premature growth plate fusion thereby causing relative shortening of one or more long bones of the hand (Cockshott 1963). Ischaemic changes in the carpal bones are uncommon (Lanzer et al. 1984).

#### 5.3 Thalassaemia

Thalassaemia is a hereditary haemoglobinopathy with an abnormality in the globin chain production of the red blood cell. This can involve a deficiency in alphaglobin chain synthesis or beta-globin chain synthesis. In severe untreated cases, there is generalised osteopenia and the tubular bones of the hand are expanded secondary to marrow hyperplasia (Fig. 9; Caffey 1957; Middlemiss and Raper 1966). There is also cortical thinning with a coarse, trabeculated appearance. When MR imaging is used in the assessment of the tubular bones of the hand, the findings may include an indistinct physeal–metaphyseal junction, altered signal intensity in the metaphysis, physeal widening and metadiaphyseal and epiphyseal lesions. Severe radiographic changes are not often seen in the developed world these days as most cases are treated from early infancy with repeated blood transfusions thereby reducing the marrow hyperplasia.

#### 5.4 Other Marrow Disorders

The radiographic appearances in the hands of other marrow and deposition disorders such as Gaucher's disease and Niemann-Pick disease are relatively minor when compared with changes elsewhere in the skeleton. The authors have reported a single case of Erdheim–Chester disease presenting with a lytic lesion in a meta-carpal mimicking a giant cell tumour or metastasis (Davies et al. 2010). Marrow malignancies are covered in chapter "Tumours and Tumour-like Leisions of Bone".



Fig. 11 Tuberous sclerosis. Multiple cyst-like radiolucencies involving the first and fifth metacarpals and all the fingers

#### 6 Amyloidosis

Amyloidosis is the deposition of abnormal insoluble proteins in soft tissues and bone. It is not a single disease entity with to-date over 20 variants identified. The World Health Organization has proposed a classification based on the precursor fibril protein (Westermark et al. 2002). This includes primary systemic, secondary systemic, familial, dialysis-related, etc. Soft tissue amyloid deposition produces nodules that are often prominent over the joints of the hand and wrist. Similar deposits in the carpal canal may lead to carpal tunnel syndrome. Ten to thirty percentage of patients with primary amyloidosis have carpal tunnel syndrome. Typically, a bilateral



**Fig. 12** Macrodystrophia lipomatosa in a child. Focal gigantism of the index and middle fingers with bony overgrowth and soft tissue syndactyly

distribution is seen, and signs and symptoms related to carpal tunnel syndrome often precede other manifestations of the disease. Amyloid deposition has also been documented in the wrists of patients on dialysis for chronic renal failure with carpal tunnel syndrome. The radiographic findings of amyloidosis in the hand and wrist include asymmetric soft tissue masses, periarticular osteoporosis, widening of the articular spaces and subchondral cysts and erosions. The appearance differs from rheumatoid disease of the hands and wrist as there is preservation of the joint space with soft tissue nodular masses. Extensive joint destruction is occasionally encountered in amyloidosis resulting from neuropathic arthropathy or osteonecrosis of the epiphyseal surfaces. On MR imaging, the amyloid deposits tend to be iso- or slightly hypointense with respect to muscle (Cobby et al. 1991; Kiss et al. 2005).



**Fig. 13** Macrodystrophia lipomatosa in an adult: focal gigantism of the index and middle fingers with periarticular exostoses

## 7 Haemochromatosis

Haemochromatosis results from excessive iron storage in the soft tissues. It may be primary, inherited as an autosomal recessive disorder, or secondary to chronic anaemias with multiple transfusions and dietary iron overload. In the hand and wrist, calcium pyrophosphate dihydrate (CPPD) crystal deposition causes chondrocalcinosis of the triangular fibrocartilage in up to 60% cases. In chronic cases, particularly the primary form, a structural arthropathy develops with a predilection for the second and third metacarpo-phalangeal joints with joint space narrowing and hook-like exostoses on the radial aspect of the metacarpal heads (Schumacher 1964).

#### 8 Phakomatoses

The phakamatoses are a group of neurocutaneous syndromes typified by the development of benign tumours and malformations, especially in organs of

Table 1	Diseases	associated	with	hypertroph	nic osteoart	hrop-
athy (m	odified from	n Gibson e	et al. 2	.008)		

Pulmonary	
Bronchogenic carcinoma	
Metastases	
Mesothelioma	
Pulmonary abscess	
Tuberculosis	
Bronchiectasis	
Cystic Fibrosis	
Cardiac	
Cyanotic heart disease	
Bacterial endocarditis	
Gastrointestinal	
Inflammatory bowel disease	
Biliary atresia	
Cirrhosis	
Miscellaneous	
Nasopharyngeal carcinoma	
Hodgkin's lymphoma	
AIDS	

ectodermal origin. The two such conditions that may manifest in the hands are neurofibromatosis and tuberous sclerosis. In neurofibromatosis, this includes soft tissue masses, bone erosion from adjacent neurofibromas and localised gigantism (macrodactyly; Fig. 10). In tuberous sclerosis, there may be multiple cyst-like radiolucencies, periosteal thickening and localised gigantism (Fig. 11; Bernauer 2001).

## 9 Macrodystrophia Lipomatosa

This is a rare congenital form of localised gigantism due to overgrowth of the mesenchymal tissues particularly hypertrophy of adipose tissue. This results in enlargement of one or more adjacent digits (Fig. 12; Hildebrandt et al. 1993). Most of the reported lesions are present at birth and are associated with a high incidence of local anomalies including syndactyly, polydactyly and clinodactyly. In the adult, the bony hypertrophy can be associated with periarticular exostoses (Fig. 13). The condition may also be associated with carpal tunnel syndrome (Ranawat et al. 1968).



**Fig. 14** Hypertrophic osteoarthropathy. Child with longstanding, untreated cyanotic heart disease. There is a singlelayer lamellar periosteal reaction along the distal radius and ulna, the metacarpals and to a lesser extent the phalanges. There is also evidence of digital clubbing

# 10 Hypertrophic Osteoarthropathy

Originally called hypertrophic pulmonary osteoarthropathy, this condition was first described in the French literature at the end of the nineteenth century (Marie 1890). Clinical presentation is with pain and tenderness of the extremities with digital clubbing. Although most commonly associated with lung diseases, classically bronchogenic carcinoma, the term pulmonary has been dropped in recognition of the nonpulmonary associations (Table 1). The typical radiographic appearance is symmetrical periosteal new bone formation along the tubular bones. This occurs in the diaphyses of the radius and ulna, and less frequently in the metacarpals and phalanges. The severity can vary



**Fig. 15** Hypertrophic osteoarthropathy. Adult with known bronchogenic carcinoma. There is symmetrical coarse periosteal new bone formation along all the metacarpals and to a lesser extent the phalanges

from simple elevation of the periosteum (Fig. 14), to a lamellated "onion-skin" appearance, to irregular periosteal "cloaking" with an undulating contour (Fig. 15; Ali et al. 1980; Pineda et al. 1987). The differential diagnosis for hypertrophic pulmonary osteoarthropathy includes pachydermoperiostitis sometimes referred to as primary hypertrophic osteoarthropathy. This is a rare, often familial, lesion predominantly of males with a predisposition for Afro-Caribbean populations. Clinically, there is clubbing of the fingers, thickening of the skin and hyperhydrosis. Compared with hypertrophic pulmonary osteoarthropathy, it is relatively pain free. The bones most commonly affected are the radius and ulna, followed by the tubular bones of the hands. The periosteal reaction is similar to hypertrophic pulmonary osteoarthropathy but is more solid and spiculated and also involves the epiphyses to produce outgrowths around joints (Fig. 16).



**Fig. 16** Pachydermoperiostosis in an adult of Afro-Caribbean ethnic origin. Digital clubbing with coarse periosteal new bone formation along the distal radius and proximal phalanges

## 11 Congenital Insensitivity to Pain

Congenital insensitivity to pain, first described in 1932 (Dearbon 1932), is a rare hereditary sensory and autonomic neuropathy characterised by the congenital inability to register pain or temperature changes (Rahalkar et al. 2008; Reilly 2009). The principal orthopaedic manifestations are recurrent fractures, neuropathic joints and osteomyelitis (Silverman and Gilden 1959; Siegelman et al. 1966; Karmani et al. 2001). The changes in the hand include acro-osteolysis, amputations and secondary osteomyelitis (Fig. 17; Gwathmey and House 1984). In children, epiphyseal separation and growth plate fragmentation may occur at the wrist.



**Fig. 17** Congenital insensitivity to pain. There is extensive terminal phalangeal resorption (acro-osteolysis) secondary to the underlying neuropathy. The appearances are non-specific and may be seen in other causes of neuropathy such as leprosy or following "trauma" such as frostbite

# 12 Toxic Agents and Pharmacological Agents

Toxic effects manifested in the hand may be localised or systemic. An example of a localised toxic effect is soft tissue necrosis and contractures secondary to snake bite venom (Huang et al. 1978). Snake and scorpion venom have also both been reported as causing acro-osteolysis (Qteishat et al. 1985). Osteolysis resulting from secondary infections due to animal bites is well recognised. The animals include domestic pets, rodents, exotic animals such as camels (Al-Boukai et al. 1989) and of course humans (Resnick et al. 1985).

Systemic toxic effects include lead poisoning seen in the past in children who had ingested leadbased paints. Radiographs show a radiodense line paralleling the growth plates in the metaphyses of



Fig. 18 Lead poisoning. An infant with a radiodense "lead line" in the distal radial and ulnar metaphyses

the distal radius and small tubular bones of the hand (Fig. 18; Leone 1968; Sachs 1981). Multiple lead lines may be seen if the exposure to the lead has been repeated. The differential diagnosis of radiodense lines in the distal radial metaphysis include other forms of heavy metal poisoning (e.g. bismuth and phosphorus) and the healing phase of rickets and scurvy and treated leukaemia. A toxic cause of more generalised bony sclerosis is fluorosis that is endemic in certain areas of India. In children, this may be associated with rickets-like changes (Christie 1980). An unusual form of occupational acro-osteolysis has been described in polyvinylchloride (PVC) poisoning (Gama and Meira 1978). The typical radiographic feature is a band of osteolysis through the terminal phalanges (Fig. 19a). With removal of exposure to the PVC manufacturing process, there is healing of the defects with shortening and widening of the terminal phalanges (Fig. 19b). A similar appearance may be seen with familial acro-osteolysis (Hajdu-Cheney syndrome) and has also been reported in a guitarist (Destouet and Murphy 1981).







**Fig. 20** Teratogenic effects of thalidomide therapy during pregnancy. The child has been born with proximal phocomelia and severe skeletal abnormalities of the hand



Fig. 21 Dialysis-related aluminium bone disease. Osteomalacic pattern with Looser's zone in the 2nd and 3rd metacarpals



Fig. 22 Radionecrosis. Historical case of a seamstress who underwent protracted fluoroscopy for the removal of a sewing needle embedded in the soft tissues. This caused skin ulceration and contractures and radionecrosis of the metacarpals (from the collection of Dr Philip Jacobs—deceased)

Prolonged and excessive exposure to numerous pharmacological agents may cause skeletal abnormalities identifiable on radiographs of the hand. These range from the devastating teratogenic effects of thalidomide (Fig. 20) to the rickets-like or osteomalacia pattern associated with phenytoin, phenobarbital, deferoxamine, diphosphonates and dialysisrelated aluminium toxicity (Fig. 21). Hypervitaminosis A can cause generalised periosteal reactions (Caffey 1951) and hypervitaminosis D sclerotic metaphyseal bands (De Wind 1961).

# 13 Radiation Changes

It was not long after the discovery of X-rays in 1895 by Wilhelm Roentgen that the deleterious effects were being reported. An eminent British Radiologist,



Fig. 23 Idiopathic Multicentric Osteolysis (Carpal-Tarsal Osteolysis). **a** Child showing marked resorption of the carpal bones with involvement of the bases of the 2nd and 3rd metacarpals. **b** Same case as an adult showing disease progression with further involvement of the metacarpals

John Hall-Edwards, working at the General Hospital in Birmingham was one of the first to give a comprehensive description of the adverse effects of X-rays on the bones of the hand-using his own hands for case material (Hall-Edwards 1908). Complications radiation-induced dermatitis include with an increased risk of malignant transformation, growth plate damage in children and radionecrosis of the bones (Fig. 22; Hartwell et al. 1964; De Smet et al. 1976). Radiation-induced damage to the hands from diagnostic X-rays is mercifully rare in this day and age but modern radiologists do need to be aware of the potential increase in radiation dose to the hands when undertaking interventional procedures under either fluoroscopic or CT control. Radiation-induced tumours of the hand following radiotherapy are rare (Libshitz and Cohen 1982). It is possible that the incidence of radiation-induced osteochondromas in the hands may rise due to the increasing use of whole-body irradiation in children as part of the preparative regimen for haematopoietic stem cell (bone marrow) transplantation.

## 14 Massive Osteolysis

Bone destruction is a non-specific feature of numerous different bone conditions. There is a rare group of idiopathic disorders causing significant bone destruction that can be categorised under the title of primary osteolysis syndromes (Resnick 2002). The commonest is known as Gorham's disease or Massive Osteolysis characterised by uncontrolled, destructive proliferation of vascular or lymphatic vessels within bone and the surrounding soft tissues (Gorham and Stout 1955). Radiographs show dramatic, progressive resorption of bone that can cross joints and involve adjacent bones. The shoulder and the pelvis are the most common sites

of involvement but cases have been reported in the hand (Patel 2005). Another rarer form of primary osteolysis is Idiopathic Multicentric Osteolysis also known as Carpal-Tarsal Osteolysis. Radiographs show progressive carpal bone resorption with in time involvement of the metacarpals (Fig. 23). Cases may be associated with a nephropathy (Warady et al. 1991).

## 15 Key Points

- the hand is the commonest site of bone involvement with sarcoid.
- the hand is the commonest site for reflex sympathetic dystrophy (complex regional pain syndrome).
- dactylitis may be seen in numerous diseases including sarcoidosis and sickle cell anaemia.
- hypertrophic osteoarthropathy may be associated with both pulmonary and non-pulmonary disorders.

# References

- Ahlberg AK (1975) On the natural history of hemophilic pseudotumor. J Bone Joint Surg Am 57A:1133–1136
- Al-Boukai AA, Hawass NE, Patel PJ, Kolawole TM (1989) Camel bites: report of severe osteolysis as late bone complications. Postgrad Med J 65:900–904
- Ali A, Tetalman MR, Fordham EW et al (1980) Distribution of hypertrophic pulmonary osteoarthropathy. AJR Am J Roentgenol 134:771–780
- Bernauer TA, Mirowski GW, Caldemeyer KS (2001) Tuberous sclerosis: II. Musculoskeletal and visceral findings. J Am Acad Dermatol 45:450–452
- Bohrer SP (1970) Acute long bone diaphyseal infarcts in sickle cell disease. Br J Radiol 43:685–697
- Boutin RD, Spitz DJ, Newman JS, Lenchik L, Steinbach LS (1998) Complications in Paget disease at MR imaging. Radiology 209:641–651
- Brudin L, Valind S, Rhodes C et al (1994) Fluorine-18 deoxyglucose uptake in sarcoidosis measured with positron emission tomography. Eur J Nucl Med 21:297–305
- Caffey J (1951) Chronic poisoning due to excess vitamin A. AJR Am J Roentgenol 65:12–26
- Caffey J (1957) Cooley's anemia: a review of the roentgenographic findings in the skeleton. Hickey Lecture. AJR Am J Roentgenol 78:381–391
- Calif E, Vlodavsky E, Stahl S (2007) Ivory fingers: monostotic Paget's disease of the phalanges. J Clin Endocrinol Metabolism 92:1590–1591
- Christie DP (1980) The spectrum of radiographic bone changes in children with fluorosis. Radiology 136:85–90
- Cobby MJ, Adler RS, Swartz R, Martel W (1991) Dialysisrelated amyloid arthropathy: MR findings in four patients. AJR Am J Roentgenol 157:1023–1027

- Cockshott WP (1963) Dactylitis and growth disorders. Br J Radiol 36:19–26
- Cundy T (2006) Is Paget's disease of bone disappearing. Skeletal Radiol 35:350-351
- Davies AM, Colley SP, James SLJ, Sumathi VP, Grimer RJ (2010) Erdheim–Chester disease presenting with destruction of a metacarpal. Clin Radiol 65:250–253
- De Smet AA, Kuhns LR, Fayos JV, Holt JF (1976) Effects of radiation therapy on growing long bones. AJR Am J Roentgenol 127:935–939
- De Smet L, Roosen P, Zachee B et al (1994) Monostotic localization of Paget disease in the hand. Acta Orthop Belg 60:184–186
- De Wind LT (1961) Hypervitaminosis D with osteosclerosis. Arch Dis Child 36:373–380
- Dearbon GV (1932) A case of congenital general pure analgesia. J Nerv Ment Dis 75:612
- Destouet JM, Murphy WA (1981) Guitar player acro-osteolysis. Skeletal Radiol 6:275–277
- Friedman A, Orcutt J, Madewell J (1982) Paget disease of the hand: radiographic spectrum. Am J Roentgenol 138(4): 691–693
- Gama C, Meira JB (1978) Occupational acro-osteolysis. J Bone Joint Surg Am 60A:86–90
- Gibson MS, Jennings BT, Murphey MD (2008) Hypertrophic osteoarthropathy. In: Pope TL Jr, Bloem HL, Beltran J, Morrison WB, Wilson DJ (eds) Imaging of the musculoskeletal system, vol 2. Saunders Elsevier, Philadelphia, p 1612
- Gorham LW, Stout AP (1955) Massive osteolysis (acute spontaneous absorption of bone, phantom bone, disappearing bone): its relation to haemangiomatosis. J Bone Joint Surg Am 37A:985–1004
- Gwathmey FW, House JH (1984) Clinical manifestations of congenital insensitivity of the hand and classification of syndromes. J Hand Surg [Am] 9A:863–869
- Hall-Edwards J (1908) The effects upon bone due to prolonged exposure to X-rays. Arch Roentgen Ray 13:44
- Hartwell SW Jr, Huger W Jr, Pickrell K (1964) Radiation dermatitis and radiogenic neoplasms of the hands. Ann Surg 160:828–834
- Haverbush T, Wilde A, Phalen G (1972) The hand in Paget's disease of bone: report of two cases. J Bone Joint Surg Am 54A:173–175
- Hildebrandt JW, Olson P, Paratainen H, Griffiths H (1993) Macrodystrophia lipomatosa. Orthopedics 16:1075–1077
- Huang TT, Blackwell SJ, Lewis SR (1978) Hand deformities in patients with snake bite. Plast Reconstr Surg 62:32–36
- Karmani S, Shedden R, De Sousa C (2001) Orthopaedic manifestations of congenital insensitivity to pain. J R Soc Med 94:139–140
- Katzman BM, Caligiuri DA, Klein DM, Perrier G, Dauterman PA (1997) Sarcoid flexor tenosynovitis of the wrist: a case report. J Hand Surg Am 22:336–337
- Kaufmann GA, Sundaram M, McDonald DJ (1991) MR imaging in symptomatic Paget's disease. Skeletal Radiol 20:413–418
- Kiss E, Keusch G, Zanetti M, Jung T, Schwarz A, Schocke M, Jaschke W, Czermak BV (2005) Dialysis-related amyloidosis revisited. AJR Am J Roentgenol 185: 1460–1470

- Lanzer W, Szabo R, Gelberman R (1984) Avascular necrosis of the lunate and sickle cell anemia: a case report. Clin Orthop Relat Res 187:168–171
- Leone AJ Jr (1968) On lead lines. AJR Am J Roentgenol 103:165–167
- Libshitz HI, Cohen MA (1982) Radiation-induced osteochondromas. Radiology 142:643–647
- Lopez C, Thomas DV, Davies AM (2003) Neoplastic transformation and tumour-like lesions in Paget's disease of bone: a pictorial review. Eur Radiol 13:L151–L163
- Love C, Tomas M, Tronco G et al (2005) FDG PET of infection and inflammation. Radiographics 25:1357–1368
- Marie P (1890) De l'osteoarthropathie hypertrophiante pneumonique. Rev Med (Paris) 10:1–36
- Middlemiss JH, Raper AB (1966) Skeletal changes in the haemoglobinopathies. J Bone Joint Surg Br 48B:693–701
- Moore S (2003) Musculoskeletal sarcoidosis: spectrum of appearances at MR imaging. Radiographics 23:1389–1399
- Neville E, Carstairs LS, James DG (1976) Bone sarcoidosis. Ann N Y Acad Sci 278:475–487
- Neville EW, Carstairs LS, James DG (1977) Sarcoidosis of bone. Q J Med 46:215–217
- Park JS, Ryu KN (2004) Hemophilic pseudotumor involving the musculoskeletal system: spectrum of radiologic findings. AJR Am J Roentgenol 183:55–61
- Park SA, Yang CY, Shin YI, Oh GJ, Lee M (2009) Patterns of three-phase bone scintigraphy according to the time course of complex regional pain syndrome type 1 after a stroke or traumatic brain injury. Clin Nucl Med 34:773–776
- Patel DV (2005) Gorham's disease or massive osteolysis. Clin Med Res 3:65–74
- Pineda CJ, Martinez-Lavin M, Goodbar JE et al (1987) Periostitis in hypertrophic osteoarthropathy: relationship to disease duration. AJR Am J Roentgenol 148:773–778
- Qteishat WA, Whitehouse GH, Hawass N (1985) Acroosteolysis following snake and scorpion envenomisation. Br J Radiol 58:1035–1039
- Rahalkar MD, Rahalkar AM, Joshi SK (2008) Case series: congenital insensitivity to pain and anhidrosis. Indian J Radiol Imaging 18:132–134
- Ranawat CS, Arora MM, Singh RG (1968) Macrodystrophia lipomatosa with carpal tunnel syndrome: a case report. J Bone Joint Surg Am 50A:1242–1244
- Reilly MM (2009) Classification and diagnosis of inherited neuropathies. Ann Indian Acad Neurol 12:80–88

- Resnick D (2002) Osteolysis and chondrolysis. In: Resnick D (ed) Diagnosis of bone and joint disorders, 4th edn. WB Saunders Co., Philadelphia, pp 4920–4944
- Resnick D, Pineda CJ, Weisman HM, Kerr R (1985) Osteomyelitis and septic arthritis of the hand following human bites. Skeletal Radiol 14:263–266
- Rodriguez-Peralto J, Ro J, McCabe K et al (1994) Case report 806: monostotic Paget's disease of the hand. Skeletal Radiol 23:55–57
- Sachs H (1981) The evolution of the radiologic lead line. Radiology 139:81–85
- Schumacher HR Jr (1964) Hemochromotosis and arthritis. Arthritis Rheum 7:41–50
- Shaheen S, Alasha E (2005) Hemophilic pseudotumor of the distal parts of the radius and ulna: a case report. J Bone Joint Surg Am 87A:2546–2549
- Shaw JA, Wilson SC (1993) Multiple hemophilic bone cysts in the hand. J Hand Surg Am 18:262–264
- Siegelman SS, Heimann WG, Manin MC (1966) Congenital indifference to pain. AJR Am J Roentgenol 97:242–247
- Silverman FN, Gilden JJ (1959) Congenital insensitivity to pain: a neurologic syndrome with bizarre skeletal lesions. Radiology 72:176–190
- Smith S, Murphey M, Motamedi K et al (2002) Radiologic spectrum of Paget's disease of bone and its complications with pathologic correlation. Radiographics 22: 1191–1216
- Sundaram M, Khanna G, El-Khoury GY (2001) T1-weighteds MR imaging for distinguishing large osteolysis of Paget's disease from sarcomatous degeneration. Skeletal Radiol 30:378–383
- Van Linthoudt D, Ott H (1986) An unusual case of sarcoid dactylitis. Br J Rheumatol 25:222–224
- Vande Berg B, Malghem J, Lecouvet F et al (2001) Magnetic resonance appearance of uncomplicated Paget's disease of bone. Semin Musculoskelet Radiol 1:69–77
- Warady BA, Haug SJ, Lindsley CB (1991) Multicentric osteolysis: an infrequently recognized renal-rheumatologic syndrome. J Rheumatol 18:142–145
- Watson RJ, Burko H, Megas H, Robinson M (1963) The handfoot syndrome in sickle cell disease in young children. Pediatrics 31:975–982
- Westermark P, Benson MD, Buxbaum JN et al (2002) Amyloid fibril protein nomenclature. Amyloid J Protein Folding Disord 9:197–200