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Radiological imaging in pediatric rheumatic diseases

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Summary

Radiological imaging plays a fundamental role in the diagnosis and monitoring of rheumatic diseases. The basic method of imaging is a classic X-ray picture, which for many years has been used as a single method for the recognition and evaluation of the effects of disease management. In today's modern day treatment of rheumatic diseases, ultrasonography and magnetic resonance are more commonly performed for early detection of inflammatory changes in the region of soft tissue, subchondral bone and bone marrow. In spite of their usefulness and fundamental role in the diagnosis, X-ray still remains an essential tool in the diagnosis of rheumatoid arthritis in children and is complementary to today's methods of imaging diagnostics. In clinical practice, X-ray imaging is still an important examination performed not only to recognize the disorders, but also to provide a differential diagnosis. It helps estimate disease progression and is used to monitor the effects of treatment and the development of possible complications. Differential diagnosis of rheumatic diseases is performed on the basis of localization and type of radiographic changes. The surrounding periarticular soft tissues, bone structures, joint space, with special attention to articular bone surfaces and epiphyses, are analyzed. The aim of this work is to describe characteristic inflammatory changes present on X-ray imaging typical for the most commonly diagnosed rheumatic diseases in children, such as juvenile idiopathic arthritis, systemic lupus erythematosus, systemic scleroderma, mixed connective tissue disease, juvenile dermatomyositis, juvenile spondyloarthropathy and systemic vascular disease.

Keywords: radiograms • arthritis in children • imaging • juvenile idiopathic arthritis • juvenile spondyloarthropathies

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Background

Rheumatic diseases in children are rare. In Poland, the incidence is approximately 6–7 children per 100,000 per year. Inflammatory involvement of the motor system during the period of rapid growth may result in some children entering adulthood as disabled individuals. In the era of modern therapy for rheumatic diseases that offers a possibility of sustained remission, early diagnosis is essential.

Despite the introduction of ultrasonography (USG) and magnetic resonance imaging (MRI) [1,2], radiographs (X-rays) remains the primary baseline study performed in any patient with clinical suspicion of rheumatic disease [3–14]. X-ray is performed to:

1. diagnose diseases, taking into account differential diagnosis;
2. assess disease severity;
3. monitor the effects of treatment, including complications.

In many cases, radiological signs are characteristic of the individual disease entity. However, only an analysis of the clinical picture combined with the results of additional tests help establish the diagnosis. Furthermore, a large number of rheumatic diseases, overlapping syndromes or

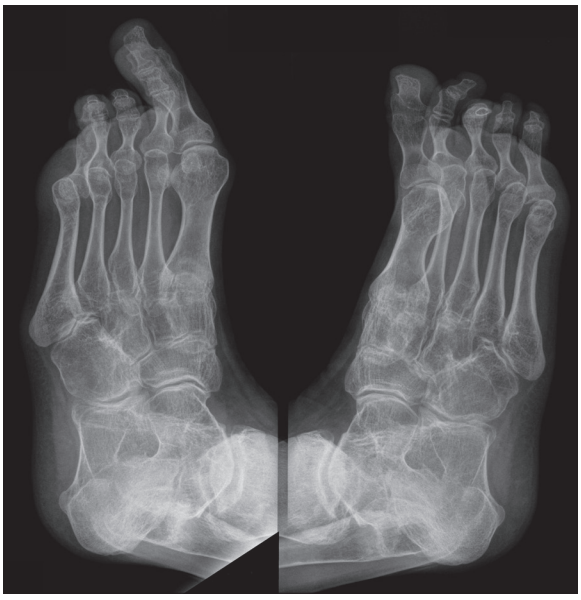

Figure 1. Oblique view of a child's feet: generalized osteoporosis, active disease process present in the tarsal and some of the metatarsophalangeal joints with joint space narrowing, geode at the base of the first proximal phalanx of the right foot, ankylosis of the first IP joint of the right foot, MTP joints within the norm.

coexisting diseases may require further imaging methods, including histopathology, for the final diagnosis.

In children, X-ray examination is most commonly performed in the diagnostic workup for:
– juvenile idiopathic arthritis (JIA);
– other inflammatory connective tissue disorders (juvenile systemic lupus erythematosus, systemic sclerosis, mixed connective tissue disease, juvenile dermatomyositis);
– systemic vasculitis (Granulomatosis with polyangiitis – GPA, formerly known as Wegener's granulomatosis – WG, Churg-Strauss syndrome purpura, Kawasaki disease);
– juvenile onset spondyloarthropathies (SpA), including psoriatic arthritis (PsA) and SAPHO-CRMO syndrome.

Radiologic assessment of the joints analyzes periarticular soft tissue, structure of bones forming the joint, including articular surface and joint space width.

Irregularities visible on plain radiographs include [6–14]:
- thickening of the periarticular soft tissue shadow;
- osteoporosis (periarticular/systemic);
- joint space narrowing or (rarely) widening;
- soft tissue calcifications;
- inflammatory cysts (geodes) and erosions;
- osteolysis;
- bone proliferation;
- periosteal reactions;
- osteophytes;
- bone deformities.

Juvenile Idiopathic Arthritis [6,7,9,11,15,16]

Juvenile idiopathic arthritis is a chronic inflammation of the joints with a frequency of approximately 3–10 per

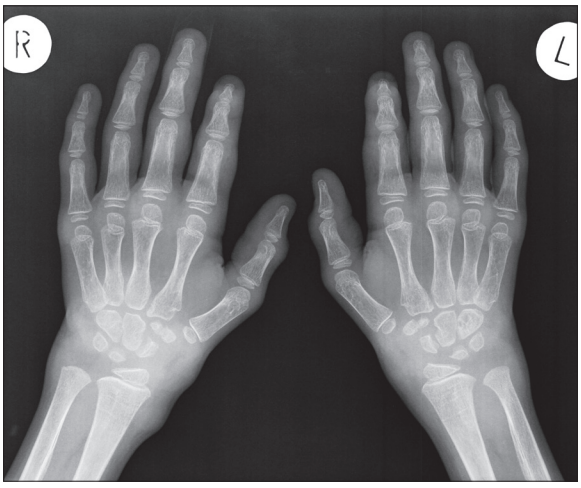

Figure 2. AP view of a child's hands: periarticular soft tissue swelling, generalized osteoporosis, periosteal reaction, uneven growth and development of wrist bones (abnormal modeling), few geodes are also visible.

100,000 children under 16 years of age. Changes occurring in JIA largely involve the musculoskeletal system, but may also include internal organs or the vision. Inflammatory changes are usually localized in the knee joints (often with monoarticular onset), wrist, ankle, small joints of the hands, hip joints, cervical spine, temporomandibular joints and rarely in metatarsophalangeal joints (Figure 1). In case of single knee joint involvement, radiographic analysis is difficult and must be differentiated from tuberculosis, trauma and hyperplastic process.

Periosteal thickening at the diaphysis of metacarpals, metatarsals and phalanges is a characteristic feature of JIA. Its presence can be likely attributed to the exposure of loosely adherent periosteum to inflammation and vascular congestion. Oftentimes, this may be the only inflammatory symptom found throughout the course of the disease. Although, periarticular osteoporosis and, in advanced cases, generalized or hypertrophic osteoporosis occurs most frequently. Furthermore, increased saturation of soft tissue periosteal shadow, including articular adipose tissue (e.g., Hoffa's fat pad) and accelerated or delayed development of ossification (Figure 2). Ultrasonography is used to differentiate from pachydermodactyly, or hypertrophy of the subcutaneous tissue surrounding proximal interphalangeal joints II–V, in the presence of appropriate hand bone structure (Figure 3). In children and adolescents, joint space narrowing presents late due to increased amount of cartilage. Erosions and geodes are also less common than in adults (Figure 4). There is also tendency for increased bone stiffness (osseous ankylosis) (Figures 5 and 6). Ankylosis may occur in any of the affected joints. Chronic inflammation of the cervical spine in the course of JIA can lead to stiffness of facet joints and suboccipital area. Stiffening within these joints is preceded by geodes and erosions of articular surface of upper facet joints (usually C2/C3), followed by the middle segment. Hypoplasia of vertebral bodies and intervertebral discs is rather a consequence of an ongoing disease process in the intervertebral joints (Figure 7). On the other hand, odontoid process damage and dislocation is not

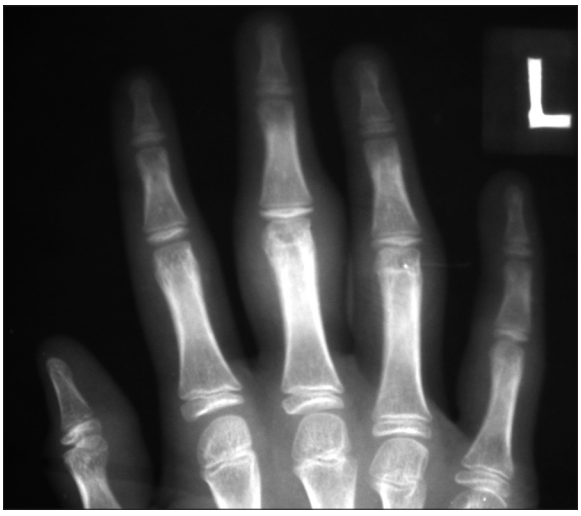

Figure 3. AP view of a child's left hand: periarticular soft tissue swelling of the PIP joints, mainly the III–IV in the course of JIA, differentiation with pachydermodactyly is at the discretion of the clinician.

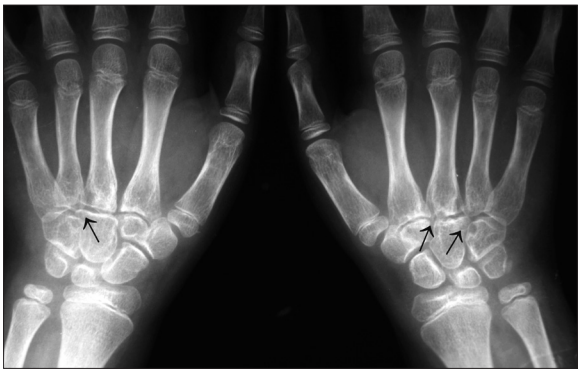

Figure 4. AP view of a child's hands: geodes and erosions in both wrist bones and the base of the metacarpal bones, joint space narrowing of the second CMC joint.

as common in children as it is in adults with rheumatoid arthritis (RA). JIA disease process disturbs a child's development. Accelerated or delayed ossification occurs in early childhood. As the disease progresses, growth disorders and bone modeling follow, including developmental disorders. Epiphyseal involvement leads to hypertrophy as a result of epiphyseal and growth plate congestion. There is also abnormal modeling and epiphyseal shaping, leading sometimes to their distension (Figure 8). Premature closure of growth plates leads to bone growth dysfunction and shorter bone length, even hypoplasia. Shortening of the long bones with considerable epiphyseal distortion resembles systemic osteochondrodysplasia, also called rheumatoid pseudochondrodystrophy. Chronic, active disease beginning early in life may lead to dwarfism. Nowadays, however, this complications is rare with the administration of proper therapy. Growth disorders occur most frequently in the form of systemic and polyarticular JIA. Some patients develop asymmetric disease, with epiphyseal overgrowth and shortening or lengthening of the limbs. Local developmental changes, such as lengthening or shortening of the affected limb, occur in cases that began with monoarticular disease or involvement of only a few joints. Some patients

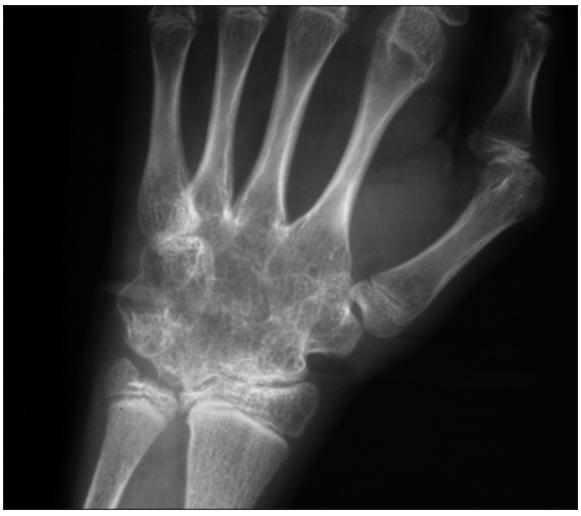

Figure 5. AP view of a baby's right wrist: ankylosis in wrist bones and CMC joints II–IV, active disease process present in the first MCP.

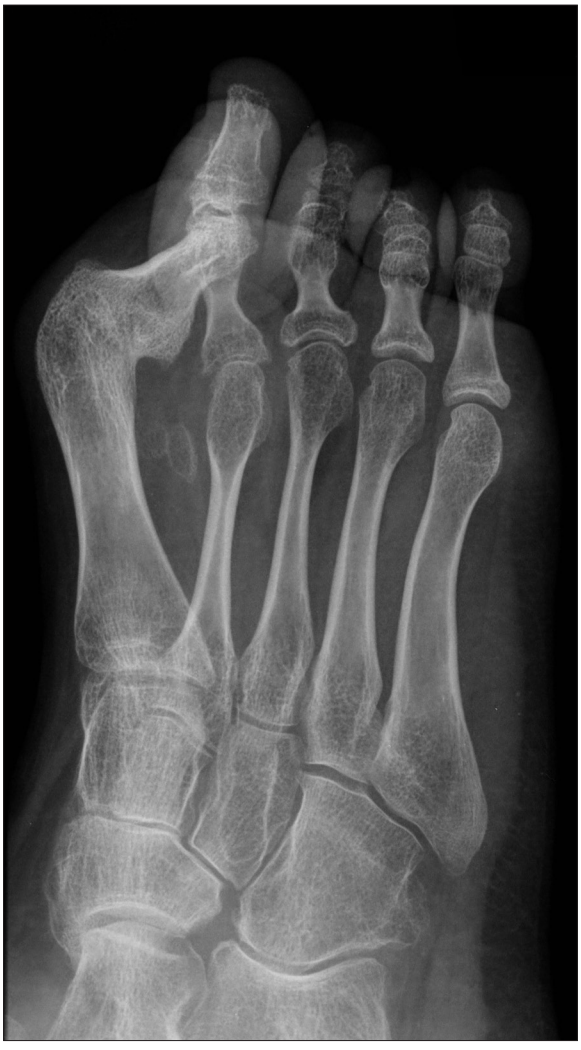

Figure 6. AP view of a child's right foot: ankylosis of the MTP joint, periarticular osteoporosis of the metatarsophalangeal joints, hypoplasia of the second metatarsal bone and joint space narrowing of the second MTP joint.

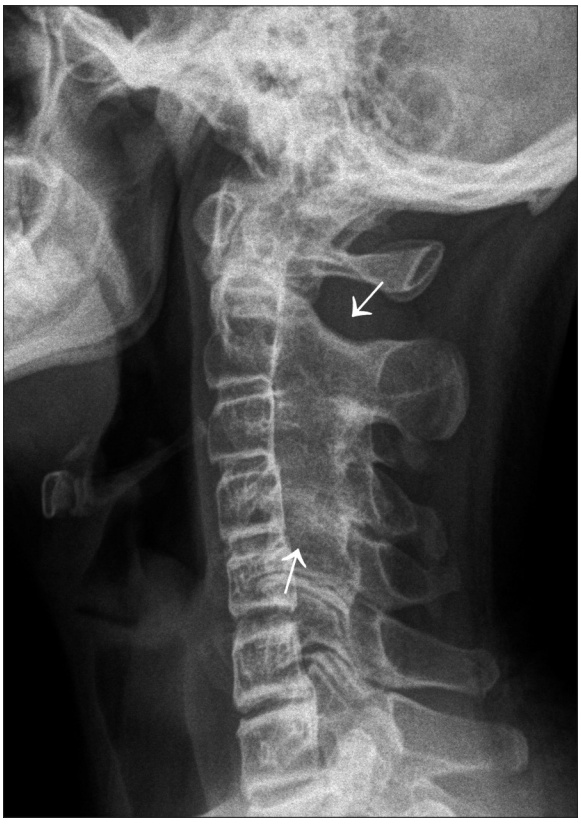

Figure 7. Lateral view of a child's spine: generalized osteoporosis, ankylosis of the facet joints C2–C5, hypoplasia of the vertebral bodies in the top-center of the cervical spinal column, apparent narrowing of the intervertebral disc C6/C7.

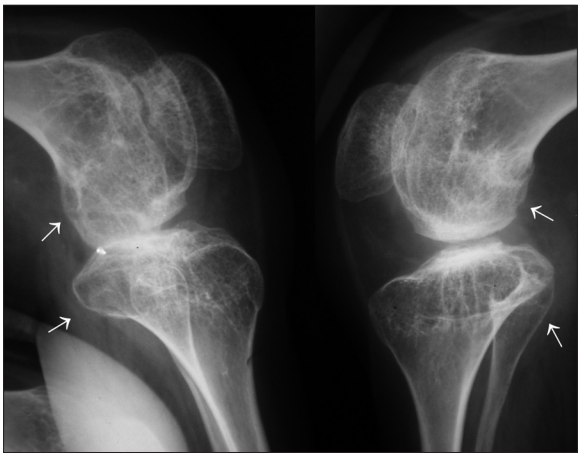

Figure 8. View of child's knees: generalized and hypertrophic osteoporosis, hypertrophy of the femoral and tibial epiphyses, hypoplasia of the femoral, tibial and fibular bones.

present with brachydactyly, or shortened phalanges of the hands. Local involvement may also lead to the shortening of only some metacarpals and phalanges (Figure 9). For as long as there has been no growth plate closure, the young body will be able to compensate, either partially or fully, for the inhibited growth.

An important diagnostic task, especially in terms of prevention of complications, is an early identification of

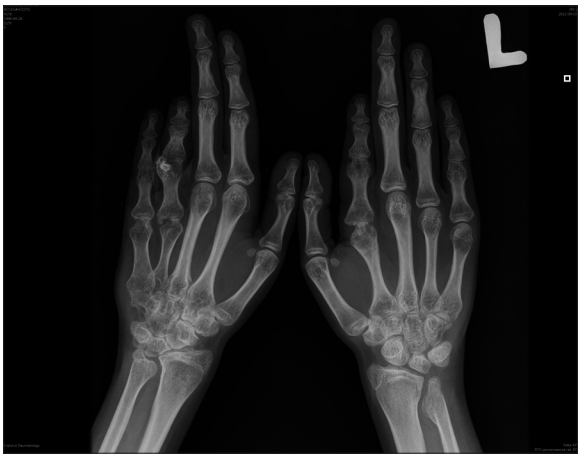

Figure 9. A-P view of hands: periarticular osteoporosis and of some of the joints of hands, mainly the right wrist, developmental malformations in the form of shortened phalanges of the IV and V finger of the right hand and II of the left hand, shortening of the IV and V right metacarpal bones and II left metacarpal bone, abnormal modeling of the distal epiphyses of the bones of the forearm and wrists, mainly right.

temporomandibular joint involvement provided by an MRI. Developmental damage of the mandibular condyle, which is a growth center, may lead to reduction in joint mobility (difficult opening and closure) (Figure 10A, 10B), including developmental disorders of the jaw (bird face) and secondary degenerative changes. The severity of bone growth disorders is greater with the younger age at onset of the disease.

Juvenile Spondyloarthropathies

Juvenile ankylosing spondylitis (JAS) [1,6,7,9,16]

The disease usually presents with an acute, subacute or chronic monoarticular inflammation, mainly in the lower limbs. Occasionally, the initial symptoms of JAS present in the form of ocular involvement. Changes in the X-ray involve large joints of the lower extremities (knee, hip and metatarsophalangeal joints) and sporadically joints of the upper extremities. Sternoclavicular joints may also be some of the first joints to become involved (Figure 11). Changes in the joints of the big toe and enthesopathy of Achilles tendon and plantar fascia are characteristic (Figure 12). Spine involvement during childhood may only be related to the sacroiliac joints (sacroiliitis), and occasionally, tend to be one-sided initially.

Radiologic joint assessment in children is difficult due to a large number of cartilaginous elements, as a result of which, joint space appears wider on an X-ray, as compared to adults (Figure 13). In the course of JAS, there is usually no complete stiffness of sacroiliac joints (sacroiliitis grade IV). It should be emphasized that early inflammatory changes, especially in the presence of normal X-ray findings of sacroiliac joints, are an indication for an MRI. MRI allows for visualisation of bone marrow edema in the proximity of the joints, corresponding to inflammation (osteitis/osteomyelitis) or less commonly inflammation of the

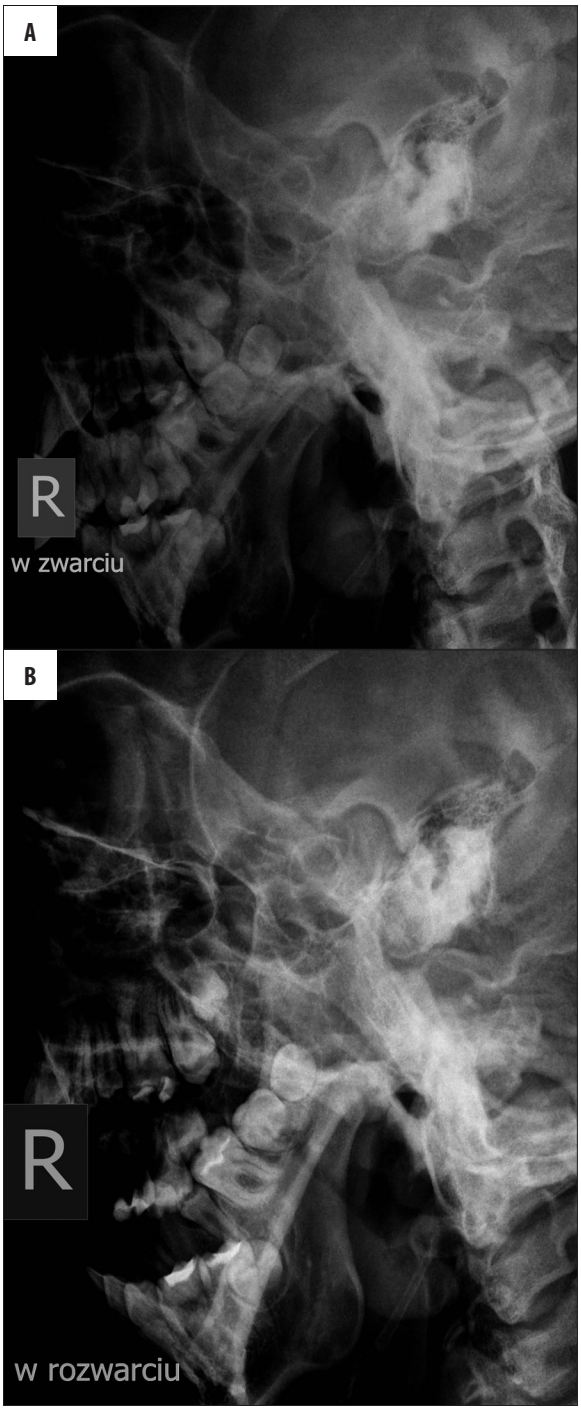

Figure 10. Transcranial projection of the temporomandibular joint (**A**) closed view and (**B**) the open view: loss of mobility of the temporomandibular joint.

attachment of articular capsule or ligaments around the sacroiliac joint (capsulitis and enthesitis) [17,18].

Features of vertebral body squaring and syndesmophytes, or destructive lesions in the vertebral body – intervertebral disc – vertebral body segments (spondylodiscitis), are very rarely observed in children. Contrary to adults, there is no stiffening of spinal segments which lead to "bamboo stick" appearance. Cervical spine involvement is also sporadic.

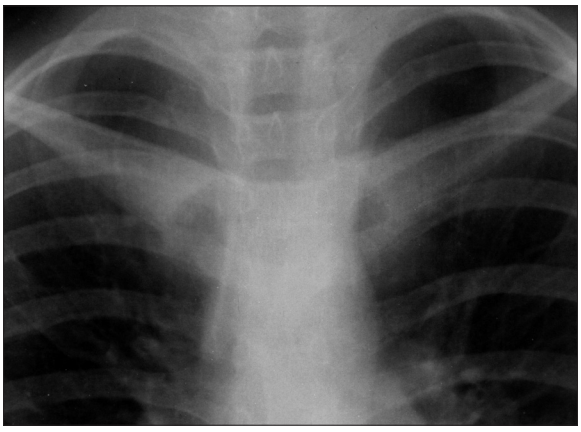

Figure 11. View of the sternoclavicular joints: soft tissue swelling, blurred, uneven margin of the sternal end of clavicles with loss of bone shadow, sclerosis.

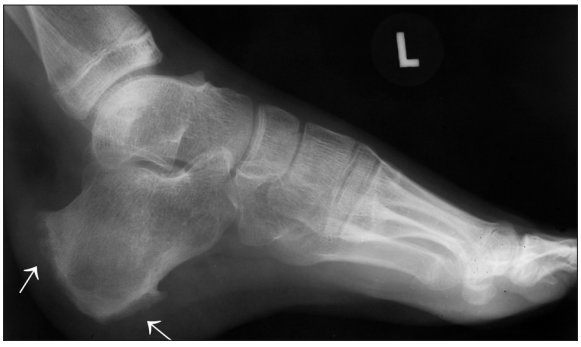

Figure 12. View of the left foot: radiological features of enthesopathic changes of the calcaneal tuberosity (arrows).

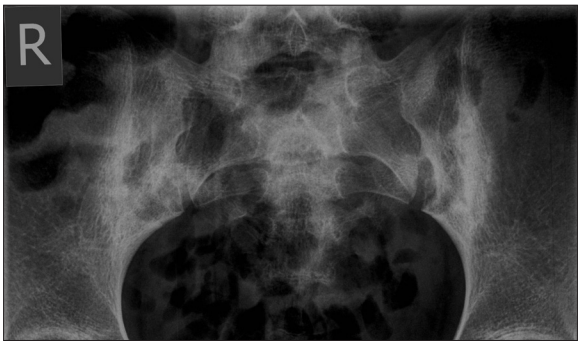

Figure 13. Sacroiliitis: sclerosis of both sacroiliac joints, mainly on the sides of hip bones with the presence of bone erosions.

Destruction of vertebral bodies in the cervical spine may occur, but it is followed by reconstruction at the later stages of the disease. Involvement of the spine may be only limited to the cervical segment.

Juvenile psoriatic arthritis (JPsA) [6,7,9,12,16]

Radiologic findings in psoriatic arthritis in adults and children are similar, although inflammation of the sacroiliac joints and the spine is less frequent in children than in adults, and full radiographic evidence of a psoriatic arthritis is also rarely observed in children. Boys are more likely to develop inflammation of the sacroiliac joints and the spine, while peripheral joints tend to be more frequently

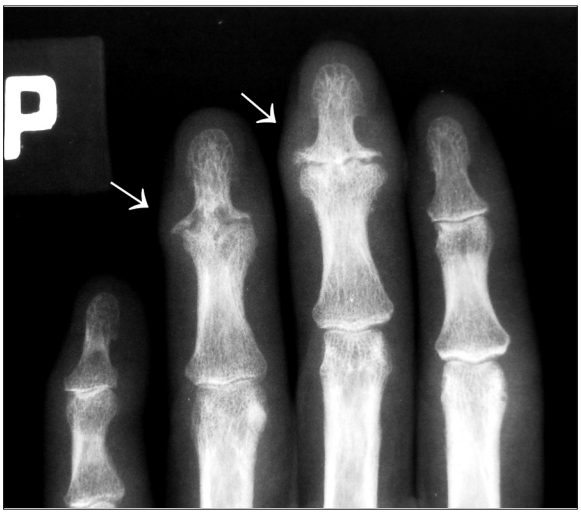

Figure 14. View of an adult hand (similar radiological picture may be present in the course of JIA): the coexistence of bone erosions and proliferative reactions in the third and fourth DIP joints (arrows).

affected in the girls. Juvenile psoriatic arthritis usually presents in the form of asymmetric arthritis. For a long time, enthesopathic changes in tendons and ligaments remain the only symptom arising from the musculoskeletal system. Inflammation of the distal interphalangeal joints of hands and feet with simultaneous destruction (erosions) and proliferation of bone (Figure 14), is characteristic. Also typical is a periosteal buildup in the shafts of phalanges, metacarpals and metatarsals. Acroosteolysis may develop further leading to osteolysis of the phalanges. Oftentimes, both osteolysis and ankylosis may be detected in a hand or a foot in the absence of osteoporosis. In the course of JpsA, inflammation of flexor tendon sheaths, similarly to the involvement of proximal and distal interphalangeal joints or soft tissue of the fingers, leads to the formation of sausage-shaped digits.

Enthesopathic changes lead to the formation of bone erosions and mineralized scars in the area of enthesis. X-ray appearance of mineralized scars near the enthesis of Achilles tendon and short flexors of the fingers (less frequently plantar fascia) is commonly referred to as a heel spur, upper and lower.

Reactive arthritis [6,7,9,14]

Reactive arthritis is rare in children. It is described with a classic triad of arthritis, conjuctivitis and urethritis. Reactive arthritis is considered to be a form of spondyloarthropathy associated with an immune response, usually following bacterial infection of the gastrointestinal tract, urogenital tract or the respiratory system. It presents most frequently with an asymmetric involvement of large joints of the lower limb (knee, ankle; rarely the hip). It sometimes affects small joints of the foot in the form of dactylitis/sausage-shaped digits (thickening, redness, swelling caused by inflammation of the finger's subcutaneous fat tissue, or tenosynovitis of the flexor tendon sheats, or arthritis of the finger's joints – similarly to JPsA discussed earlier). Enthesopathies of calcaneal tuberosity

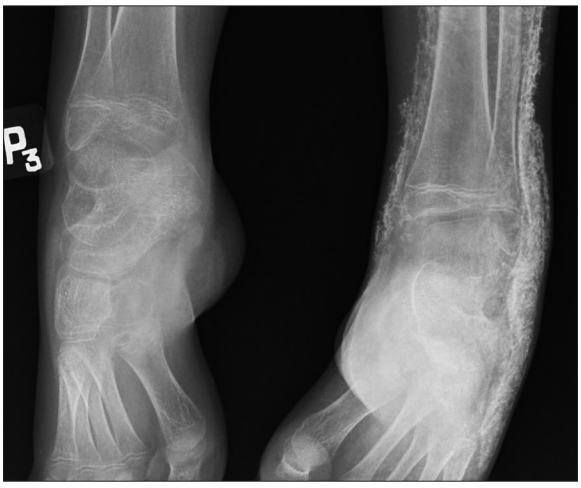

Figure 15. View of feet with visible calcifications in the soft tissues around the ankle of the left foot.

are characteristic. There is also a bilateral arthritis of the sacroiliac joints, asymmetric form is less frequent.

Enteropathic arthropathies in the course of ulcerative colitis and Crohn's disease [6,7,9,19–22]

Symptoms arising from the muscoloskeletal system are divided into axial and peripheral forms. Radiologic findings in the axial form are similar to those found in JAS. Sacroiliac joints are involved symmetrically, while spinal joints are affected less frequently with vertebral body squaring and syndesmophyte formation. Spinal disease begins with inflammatory foci within the vertebral bodies („shiny corners" on MRI), which will generate syndesmophytes, i.e. bony growths leading to spinal rigidity. Peripheral form usually involves large joints of the lower extremities, oftentimes asymmetrically. Typically, there is soft tissue swelling and periarticular osteoporosis and no evidence of bone destruction. This form is more common in patients with Crohn's disease than in ulcerative colitis. Arthritis may appear before the intestinal disease, but usually develops during the course of the disease.

Polymyositis and Dermatomyositis [6,9,10,16,23]

Juvenile polymyositis and dermatomyositis (JPM/JDM) is the most common inflammatory myopathy. Soft tissue calcinosis develops more frequently in children than in adults, occurring in 30–70% of cases (in subcutaneous tissue, muscles, joints, vessels and nerves) (Figure 15) and rare coexistence of JPM/JDM with cancer. Interstitial lung disease is also rare. Muscle dysfunction is mainly associated with proximal muscles of the extremities, especially shoulder and hip girdle, muscles of the back and the neck. Dysphagia and dysphonia are the result of muscle weakness in the esophagus, pharynx and larynx. Breathing difficulty may occur in the presence of intercostal muscle, diaphragm or airway smooth muscle involvement. Polymyositis with no evidence of skin lesions is rare. Frequently, joint symptoms precede the symptoms of dermatomyositis. Joint symptoms, such as pain and swelling, occur in about half of the children in the early stages of the disease. X-ray will only reveal osteoporosis, as destructive changes do not occur.

Systemic Scleroderma [6,9,13,24,25]

Systemic scleroderma is a connective tissue disease characterized by disseminated microangiopathy leading to fibrosis of the skin and internal organs. X-rays reveal osteolytic lesions of fingernails and toenails (acroosteolysis). Calcifications may occur in the phalanges, skin and subcutaneous tissue. Contractures and subluxation can occur as a result of scar tissue formation in the skin and tendons. Signs of destructive arthritis (articular surface erosions, subchondral geodes, osteoporosis, joint space narrowing) may occur in a small fraction of cases. Pediatric systemic scleroderma may involve the heart, lungs and esophagus. Changes occur in the form of interstitial pulmonary fibrosis and pulmonary arterial hypertension. Chronic interstitial lung diseases are characterized radiographically by the presence of linear reticulonodular infiltrates. Later presenting with a "honeycomb" appearance caused by cystic enlargement of the bronchioles and alveolar ducts. Changes usually occur bilaterally and symmetrically at the base and central lower lung fields. High-resolution computed tomography (HRCT) is the test of choice for confirmation of changes. Disorders of esophageal motility, such as weak peristalsis and impaired contractility, are present. Furthermore, there is a moderate degree of esophageal dilatation and partial smoothing of the mucosal folds. Presence of air in the esophagus is a result of hypotension and stiffness of the esophageal wall, as well as impaired contractility of the upper sphincter. These changes are irreversible.

Juvenile Systemic Lupus Erythematosus (JSLE) [6,8,10,24,26,27]

Joint pain and swelling are some of the most common symptoms of SLE and may also be the first symptoms of the disease. They are migratory and usually subside during the course of the disease. Joints most commonly affected include knees, ankles and wrists; less frequently elbow joints. Erosions may develop sporadically. In some cases, inflammation of periarticular tissues (ligaments, joint capsules) may lead to contractures or subluxations, also known as Jaccoud's arthritis. This form of the disease occurs less frequently in children, than in adults. Pericarditis may develop during the course of lupus. It presents with non-specific enlargement of the heart on chest X-ray. Myocarditis and endocarditis may also develop, leading to pleurisy with or without effusion. Moreover, aseptic necrosis of the femoral head and steroid-induced osteoporosis may develop during the course of the disease or as a result of therapy.

Conclusions

Imaging studies play a key role in the diagnosis of arthritis, its progress and the differential diagnosis. In light of the current knowledge concerning immunologic pathogenesis of rheumatic diseases, the role of X-ray imagery has been validated. Early stages of soft tissue and subchondral bone inflammation are visible on an ultrasound and MRI [1,2]. X-ray images of periarticular osteoporosis and soft tissue edema are not specific, hence the increasing popularity of ultrasound and MRI in the diagnostic workup of rheumatic diseases in children. However, X-ray examination still plays a crucial role in the initial staging of arthritis, exclusion of lesions other than inflammatory, especially neoplastic and traumatic conditions in children, as well as in the assessment of complications.

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