Pediatr Radiol (2013) 43 (Suppl 1):S172–S180
DOI 10.1007/s00247-012-2586-9



# Juvenile idiopathic arthritis and enthesitis-related arthropathies

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Received: 3 August 2012 / Revised: 1 October 2012 / Accepted: 11 November 2012


**Abstract** Juvenile idiopathic arthritis (JIA) represents a spectrum of non-pyogenic inflammatory arthritides affecting children. The purpose of this pictorial review is to illustrate the imaging spectrum of JIA and the role of radiology in disease diagnosis and management.

**Keywords** Juvenile idiopathic arthritis · Radiography · MRI · Joint injections · Children

## Introduction

Juvenile idiopathic arthritis (JIA) represents a spectrum of non-pyogenic inflammatory arthritides that present in children younger than 16 years of age with at least 6 weeks of symptoms (Table 1) [1, 2]. The JIA classification system refined by the International League of Associations for Rheumatology replaces older classification systems including juvenile chronic arthritis and juvenile rheumatoid arthritis [3]. Psoriatic arthritis and enthesitis-related arthropathies are subcategories within the JIA classification.

Joint involvement in children with oligoarticular JIA includes knees, ankles, wrists and elbows [4]. Polyarticular JIA involvement tends to be symmetrical with preferential involvement of the wrist, hands and feet.

Juvenile psoriatic arthritis, unlike its adult counterpart, may present with arthritis prior to skin changes [5]. Children with juvenile psoriatic arthritis before age 5 are more likely to be female, ANA positive and have clinical similarities to oligoarticular and polyarticular JIA but also may present with dactylitis.

Enthesitis-related arthritis replaces the term juvenile spondyloarthritis. Older children with enthesitis-related arthritis are more likely to be male and have symptoms similar to spondyloarthritis, including axial involvement. The new terminology is meant to emphasize that pediatric disease is more likely to present with enthesitis [6]. Unlike adult spondyloarthritis, axial involvement is rare, particularly in children in their first decade. Hip and feet involvement is more common in children [4]. The enthesitis component of the disease tends to involve tendon and ligamentous insertion and origins about the pelvis, knee and calcaneal tuberosity.

## Imaging

There are three radiographic stages of JIA: early, intermediate and late [7]. Early in disease, periarticular osteopenia, effusions and juxtasynovial soft-tissue swelling may be seen. Radiographs are often normal early in disease. Intermediate changes include the development of joint space narrowing, cortical erosions and epiphyseal overgrowth (Fig. 1). Late changes include ankylosis, joint angular deformities, contractures and muscle atrophy (Figs. 1 and 2).

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**Table 1** Juvenile idiopathic arthritis, International League of Associations for Rheumatology classification [2]

| Subgroups | Joints | Other |
|---|---|---|
| Systemic arthritis | 1 or more | Fever for at least 2 weeks with one or more of the following: rash, lymphadenopathy, hepatosplenomegaly, serositis |
| Oligoarthritis | 1–4 during first 6 months of disease | - |
| Polyarthritis | 5 or more joints during first 6 months | RF negative and positive are subgroups |
| Psoriatic arthritis | Any number of joints | With 2 of the following: Dactylitis, nail pitting/oncholysis, psoriasis in a first-degree relative |
| Enthesitis-related arthritis | Any number of joints and enthesitis | With 2 of the following: sacroiliac/lumbrosacral pain, HLA-B27, onset of arthritis in male > 6 years of age, acute anterior uveititis, positive family history |
| Undifferentiated arthritis | Any number of joints | Does not fulfill any of the above criteria, or fulfills criteria in 2 or more of the above categories |

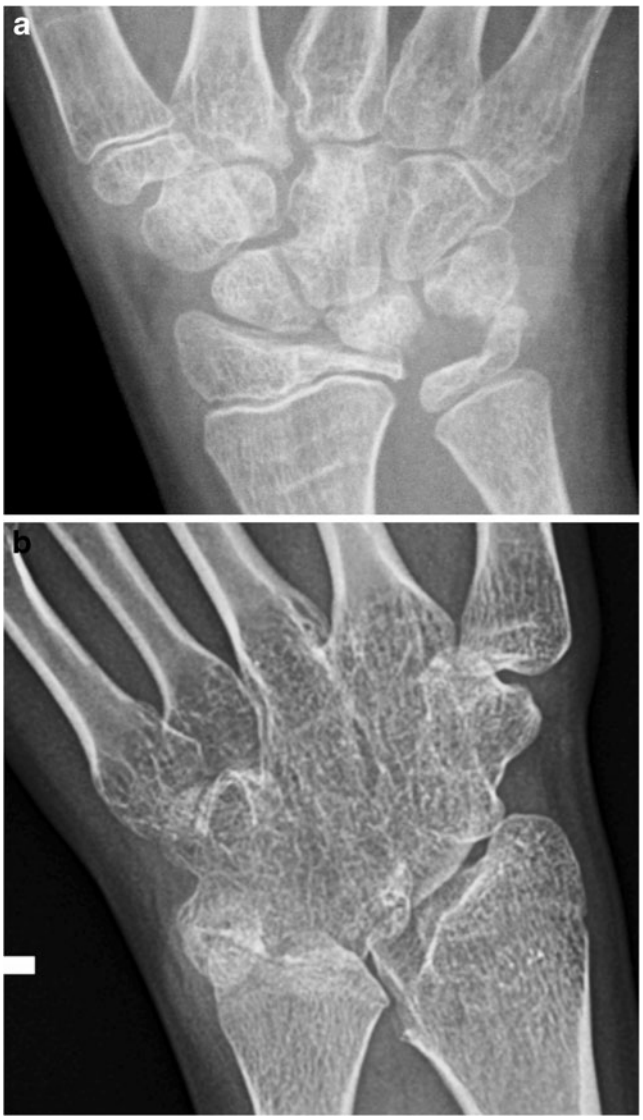


**Fig. 1** Wrist radiographs in a 7-year-old girl with JIA. **a** Note innumerable carpal and metacarpal erosions, joint space narrowing and periarticular osteopenia. **b** In the same patient at the age of 18, there is carpal and carpometacarpal ankylosis present and mild Madelung deformity

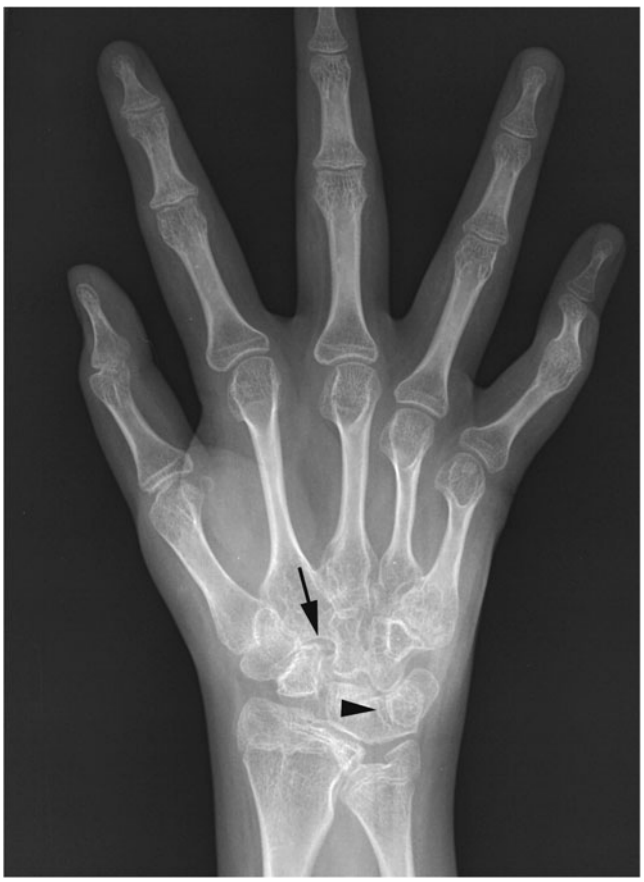

**Fig. 2** Hand radiograph of a 14-year-old girl with a JIA. Boutonniere deformity of fifth finger. Innumerable carpal and metacarpal erosions are present as well as periarticular osteopenia. Note the radial side capitate erosion (*arrow*) and the ulnar side lunate erosion (*arrowhead*) with extensive joint remodeling

Periostitis may be seen more commonly with JIA compared with adults with rheumatoid arthritis.

Erosions are less likely to be present in children, particularly in young children, not because the disease is less innocuous in youth, but because of the relative abundance of cartilage surrounding bone that must be destroyed before erosions would be visible radiographically. Adults have a single layer of overlying cartilage (articular cartilage), whereas children have three layers of overlying cartilage (articular, epiphyseal and spherical growth plate) (Fig. 3). Therefore, radiographic follow-up in children with an established diagnosis of JIA is limited in regard to assessment of joint damage, and MRI plays a more important role [8].

Erosions about the wrist should be differentiated from normal carpal undulations [9, 10]. Normal carpal undulations most commonly affect the capitate followed by the hamate (Fig. 4). Carpal undulations are common. In a study by Avenarius et al. [9] of 87 pediatric wrists, carpal undulations were found in 50 children (57%). MRI is useful in these situations. If a suspected carpal undulation is associated with marrow edema, cartilage destruction and synovial enhancement, erosion is favored (Fig. 5). Erosions also tend to be larger and more deforming compared with normal carpal undulations. Erosions may also be seen and preferentially involve the bare areas of intra-articular bone, such as the nonarticular portions of the metacarpal and metatarsal head and bases (Fig. 5). These intra-articular portions of bone are considered bare areas because there is no overlying cartilage. Because of the lack of overlying cartilage, erosions may

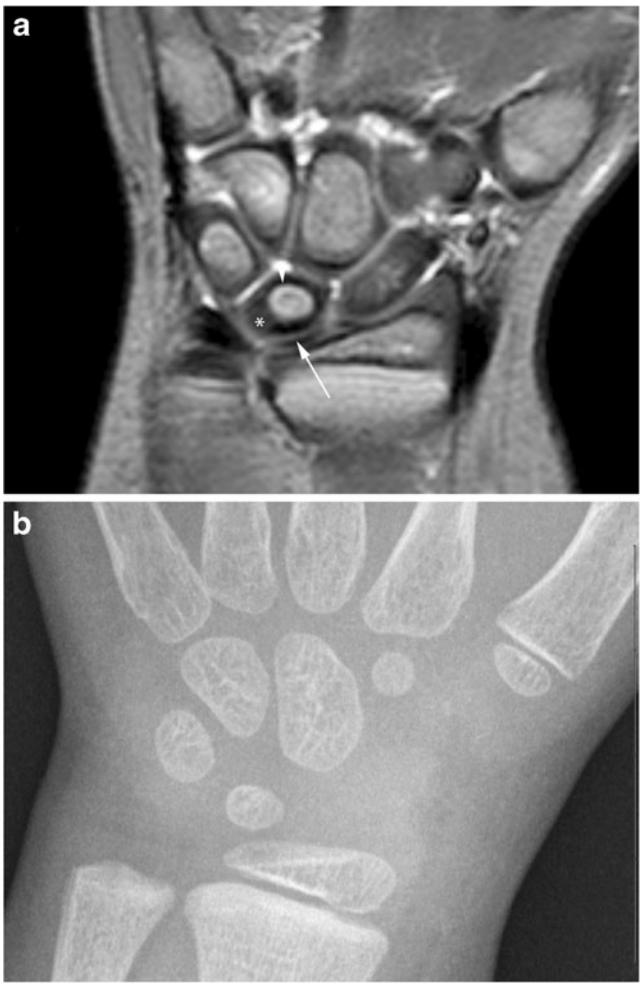


**Fig. 3** A 4-year-old girl's wrist. **a** A T2-W coronal MR image of the wrist shows articular (*arrow*), epiphyseal equivalent (*asterisk*) and spherical growth plate (*arrowhead*) surrounding the lunate bone. **b** PA wrist radiograph only delineates the bones and radiolucent cartilage occupies the majority of the intercarpal radiolucent space

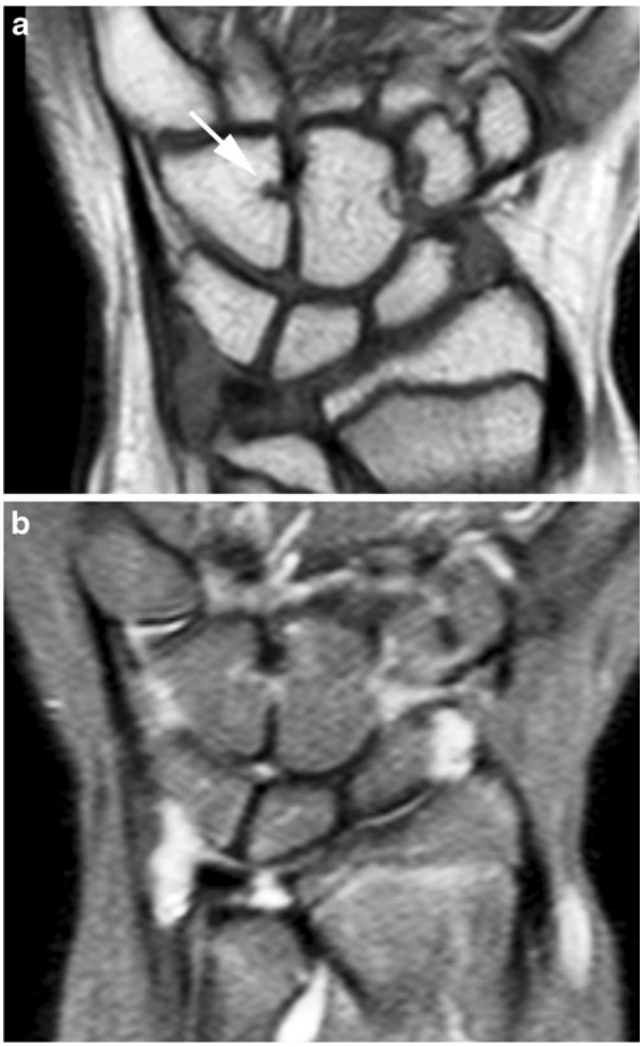


**Fig. 4** Normal hamate carpal undulation on MR in a 13-year-old girl. **a** A T1-W coronal image shows cortical undulation in hamate (*arrow*). **b** STIR coronal image shows no corresponding edema; therefore, this is a normal finding and should not be confused with an erosion

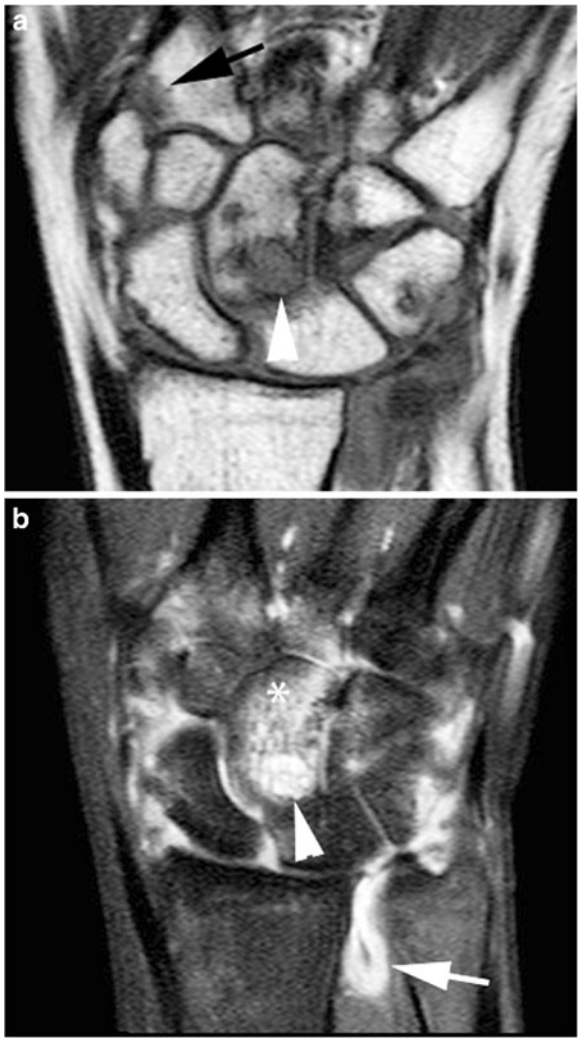


**Fig. 5** MR images in a 16-year-old with JIA. **a** T1-W coronal image demonstrates multiple carpal bone erosions, including bare areas of the metacarpal bases (*black arrow*). Note large capitate erosion (*arrowhead*). **(b)** A T1-W fat-saturated coronal MR image after intravenous gadolinium administration shows extensive marrow enhancement and multicompartmental synovial enhancement including the distal radioulnar joint (*arrow*). Note large capitate erosion (*arrowhead*) with extensive adjacent marrow edema (*asterisk*)

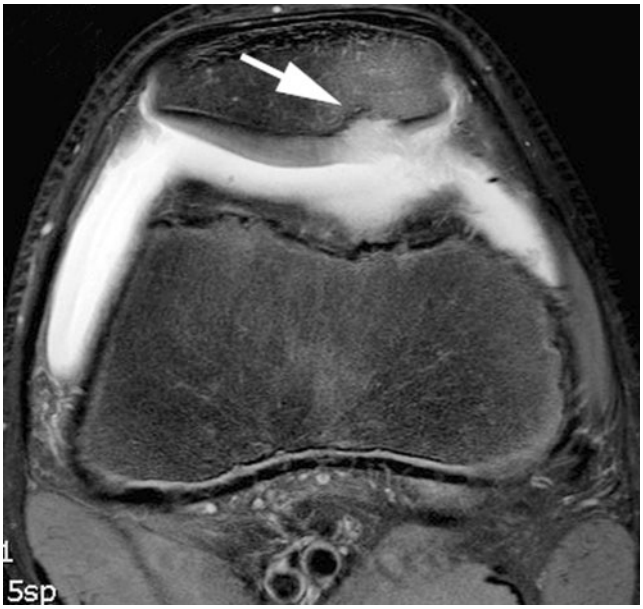


**Fig. 7** Images in a 15-year-old boy with JIA. Proton density fat-saturated axial MR image demonstrates a large knee effusion and medial patellar facet full thickness cartilage loss with early cortical destruction consistent with erosions (*arrow*). Note patellar bone marrow edema as well

preferentially occur in these locations before they occur elsewhere in the joint.

US is useful for identifying effusions in children with an established diagnosis of JIA. US is not useful for distinguishing pyogenic and non-pyogenic causes of inflammatory arthritis [11]. The role of US is limited in joints such as the elbow or knee because clinical evaluation is sufficient for identification of an effusion due to the superficial nature of these structures. The role of US is more important in identifying inflamed joints that are more challenging to evaluate, such as the hip joint (Fig. 6). Many conditions may mimic hip pathology. Precise localization of pathology specific to the hip joint rather than adjacent muscle or bone may be challenging

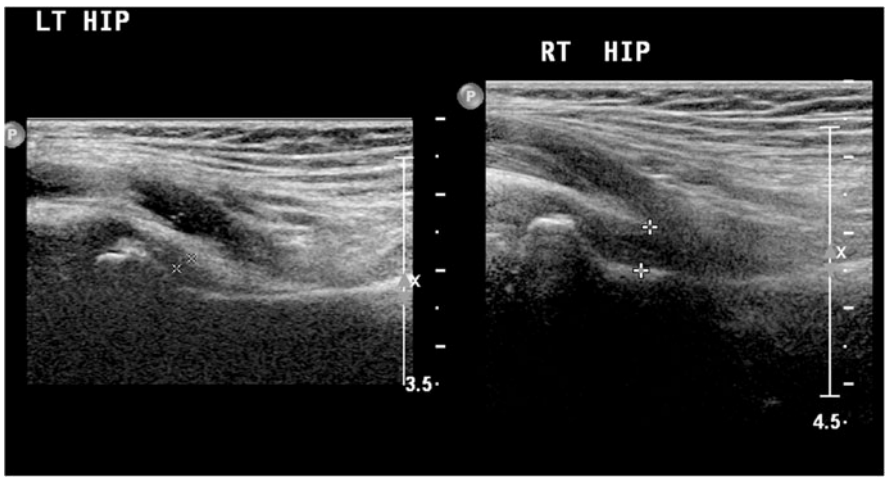


**Fig. 6** Longitudinal hip US in a 3-year-old girl with JIA.Calipers delineate the right hip effusion. A physiologically normal left hip effusion is used for comparison

clinically because of its deep location. Capsular distention of anechoic or hypoechoic fluid with debris is the primary finding by sonography of a joint effusion. Additional findings include regional hyperemia by color Doppler and synovial thickening.

The utility of MRI early in disease is to identify active inflammation before radiographs are abnormal. Early changes include synovitis, joint effusion and marrow edema. Synovitis is defined by the presence of synovial enhancement with synovial thickening in

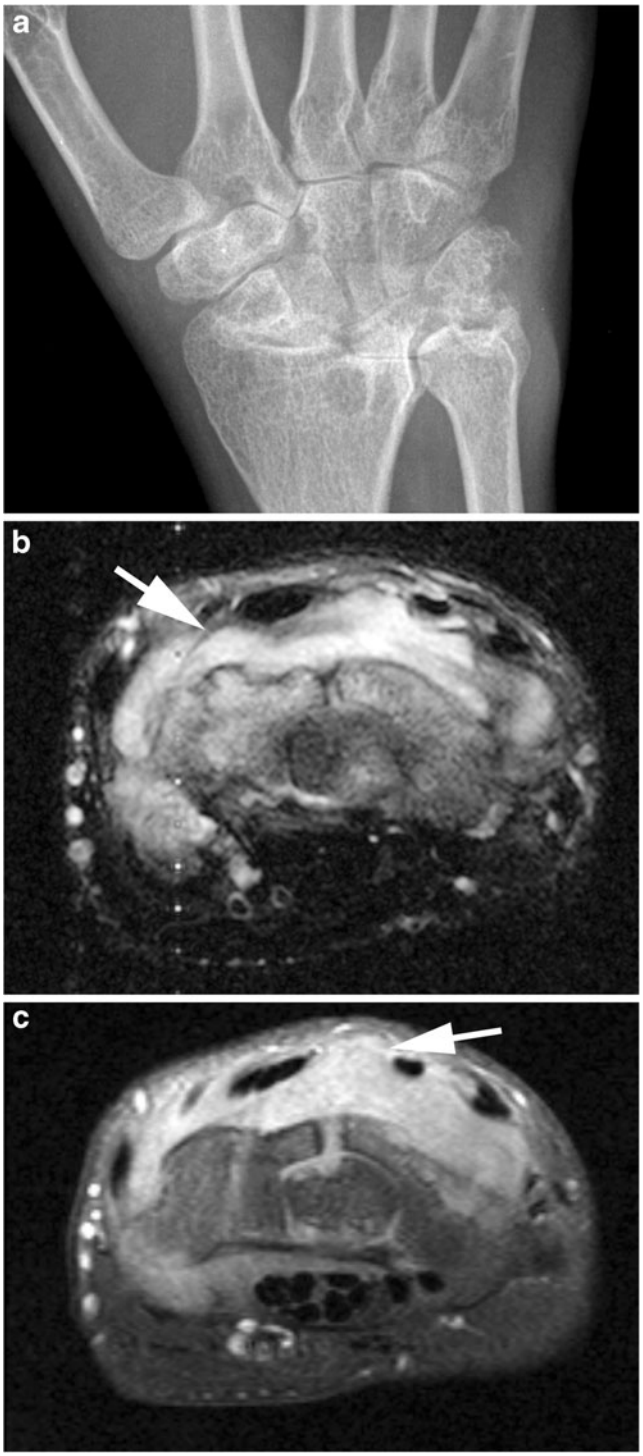


**Fig. 8** Images in a 16-year-old boy with JIA. **a** A wrist radiograph demonstrates severe joint space loss and multiple cortical erosions. **b** A T2-W fat-saturated axial MR image through the wrist shows thick mass-like dorsal intercarpal pannus (*arrow*). **c** A T1-W fat-saturated axial MR image after intravenous gadolinium administration demonstrates exuberant enhancement of the pannus (*arrow*)

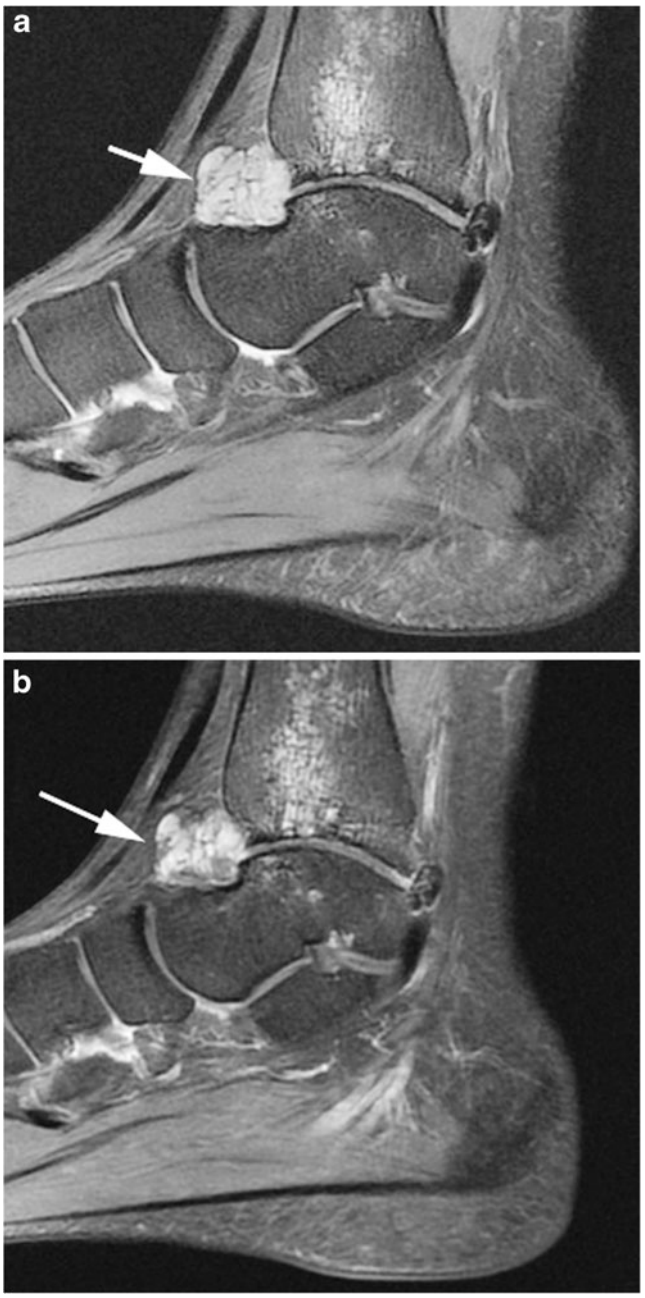


**Fig. 9** MR images in an 18-year-old woman with JIA. **a** A proton density fat-saturated sagittal image and (**b**) T1-W post-Gd fat-saturated sagittal sequence demonstrate tumefactive hypervascular pannus in the tibiotalar joint (*arrows*)

the setting of a joint effusion (Fig. 5). Synovitis should not be diagnosed when synovial enhancement is the only feature present. Normal synovial enhancement can be seen in approximately 9–44% of healthy non-rheumatologic adults [12, 13].

The presence of bone marrow edema on MRI in adults with rheumatoid arthritis has been shown to be a precursor for cortical erosions [14]. Mundwiler et al. [14] showed erosions arose in prior regions of marrow edema with a sensitivity of 50% at 6 months and 67% at 12 months.

Chronic inflammatory changes on MRI related to JIA include erosions, pannus formation and rice bodies (Figs. 7, 8, 9 and 10) [15]. T1-W sequences best delineate trabecular and cortical anatomy of bone; hence, it is the optimal sequence to use to characterize erosions (Fig. 5). Fluid-sensitive sequences lack the anatomical detail provided by T1-W sequences; distinguishing marrow edema and erosions is possible but more challenging (Fig. 5). Pannus represents mass-like synovial thickening. Pannus and inflamed synovium have similar morphological imaging features on fluid-sensitive sequences, and enhancement patterns after contrast administration. Pannus may superficially mimic diffuse pigmented villonodular synovitis, except hemosiderin staining is less likely to be present. Rice bodies represent fibrinous exudate. Rice bodies distribute throughout the joint, are small discrete particles and do not have mass-like properties of pannus. Rice bodies have been historically described in the setting of not only JIA, but also in the setting of tuberculous arthritis [15]. Rice bodies should be distinguished from synovial chondromatosis, a rare disease in children. Pannus and rice bodies may superficially mimic each other on fluid-sensitive sequences. Post-gadolinium sequences are helpful to distinguish these two entities. Pannus is usually hypervascular and will enhance, whereas rice bodies will not enhance (Fig. 9).

Additional findings seen on MRI related to JIA include meniscal hypoplasia about the knee [16], cartilage thinning without cortical erosions and tenosynovitis (Fig. 11). Osteonecrosis may sometimes be seen and may be related to complications related to corticosteroid use.

Enthesitis-related arthritis distribution of joint disease differs compared with oligo-and polyarticular JIA, but the individual imaging features of disease affecting the joints are similar. In addition to joint involvement, enthesitis-related arthritis may also involve tendon and ligament origins and insertions. On radiography, subtle thickening of tendons and ligaments may be seen. Tenosynovitis may be seen as well and is indistinguishable from tenosynovitis related to oligo- and polyarticular JIA (Figs. 11 and 12). Wisp-like periostitis develops along the osseous attachment of the tendon or ligament with advanced disease. On MRI, edema and thickening of the tendon or ligament is present with marrow edema at the bony attachment (Figs. 13 and 14). A sausage

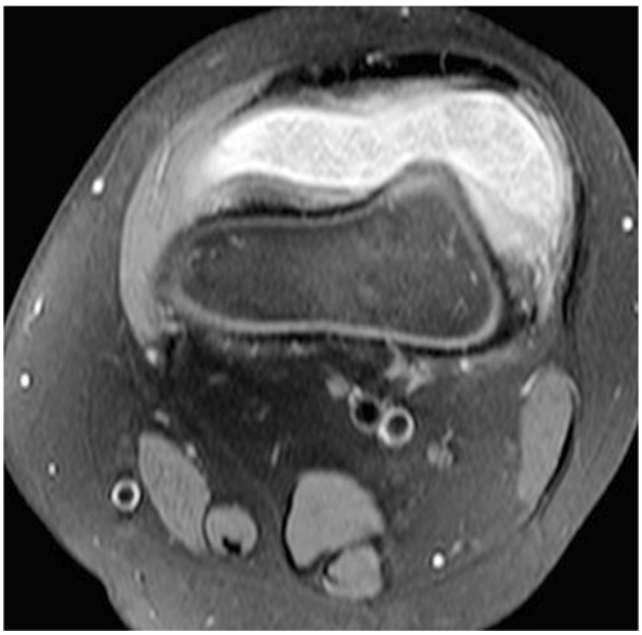

**Fig. 10** Rice bodies in a 4-year-old boy with JIA. A proton density fat-saturated axial MR image demonstrates innumerable hypointense particles within a large knee joint effusion, consistent with rice bodies

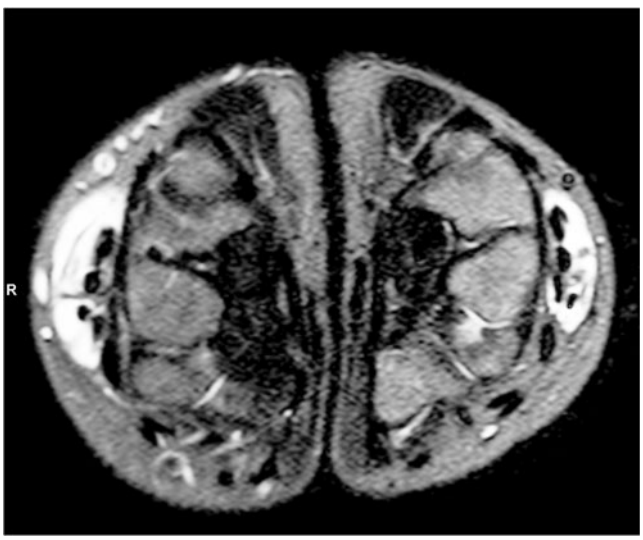


**Fig. 11** Tenosynovitis in a 10-year-old boy with JIA. STIR axial MR image through both wrists shows extensive bilateral extensor digitorum tenosynovitis

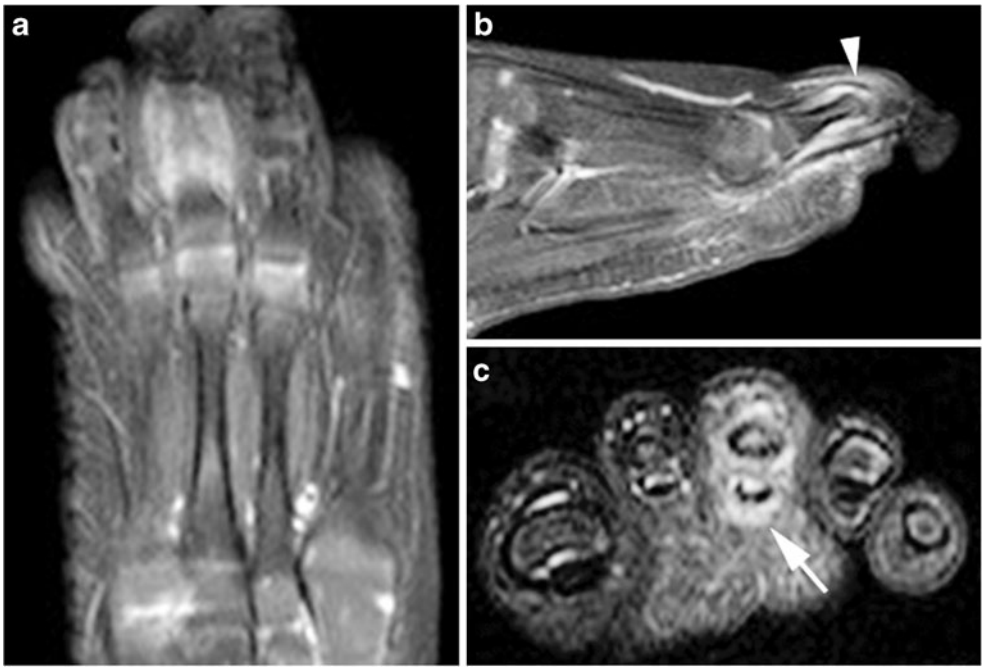


**Fig. 12** Sausage in a 3-year-old boy with sausage digit. **a** A STIR footprint demonstrates diffuse third toe soft-tissue swelling. **b** Post-Gd T1-W fat-saturated sagittal MR image shows proximal interphalangeal synovitis (*arrowhead*). **c** A post-Gd T1-W fat-saturated short-axis image also shows third digit flexor tendon tenosynovitis (*arrow*)

digit (dactylitis) may be seen with enthesitis-related arthropathies and juvenile psoriatic arthritis (Fig. 12). A sausage digit is generalized soft-tissue swelling of an affected toe or finger secondary to tenosynovitis and interphalangeal synovitis.

## Treatment

To minimize long-term orthopaedic complications of JIA, early diagnosis and pharmacological control of disease is important. Treatment options include systemic and targeted

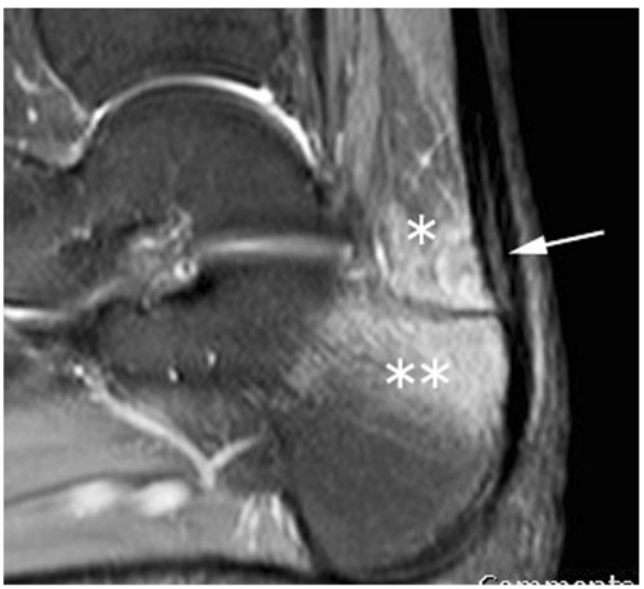

**Fig. 13** Enthesitis-related arthritis in an 18-year-old man. MRI shows Achilles tendon thickening (*arrow*) and edema, marrow edema at the calcaneal tuberosity at the Achilles insertion (*double asterisk*), and Kager's fat pad edema (*asterisk*), consistent with enthesitis

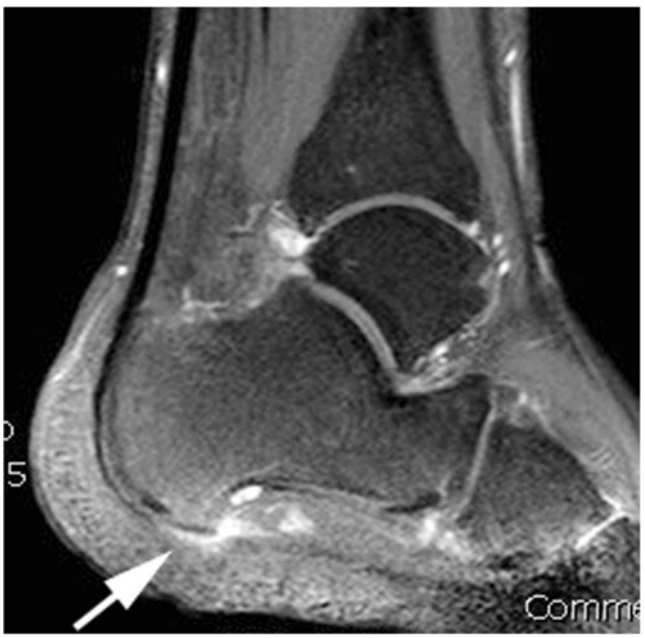

**Fig. 14** Enthesitis-related arthritis in an 18-year-old man. MRI shows that the plantar fascia origin enthesitis is present at the plantar calcaneal tuberosity (*arrow*)

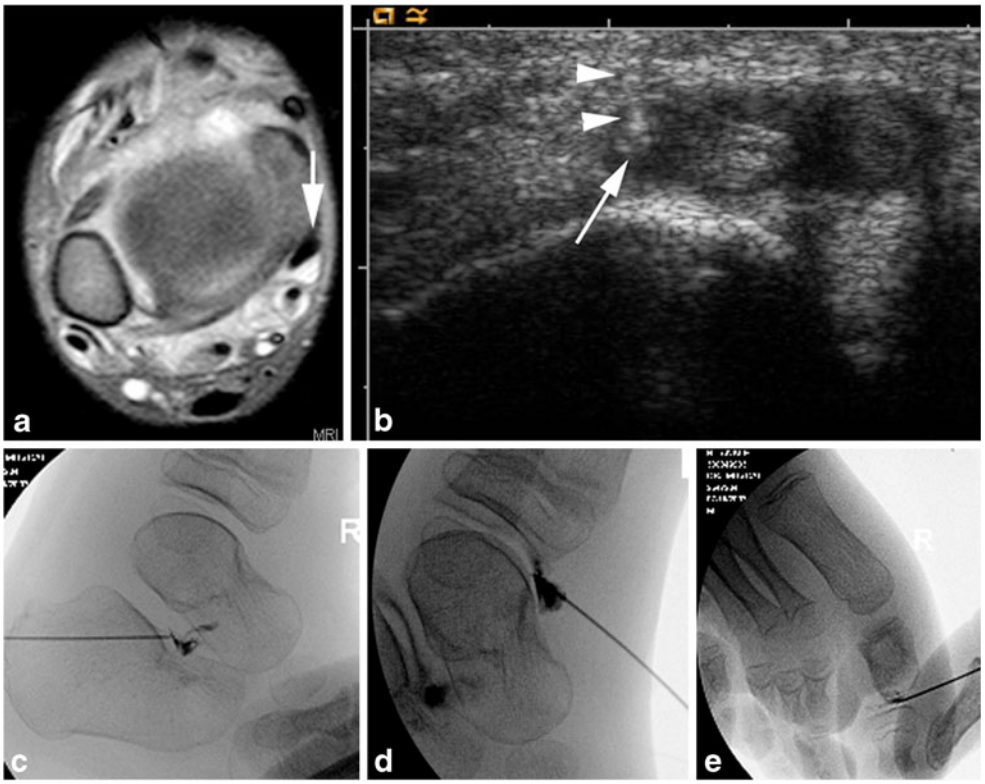


**Fig. 15** A 4-year-old boy with JIA. **a** A STIR axial MR image of the ankle demonstrates multicompartmental tenosynovitis, including the tibialis posterior (*arrow*) as well as multicompartmental arthritis about the foot (not shown). Subsequently, the child was referred for selective tendon sheath steroid injections and joint injections. **b** US-guided tibialis posterior tendon sheath injection is performed (*arrowheads* needle; *arrow* needle tip). Fluoroscopic contrast injections (**c**–**e**) are performed to confirm intra-articular needle location prior to injection of the subtalar (**c**), tibiotalar (**d**), and first IP (**e**) joints

local control. Systemic pharmacological therapy includes non-steroidal anti-inflammatory agents (NSAIDS), and disease-modifying antirheumatic drugs (DMARDS), such as methotrexate, biological therapy and systemic corticosteroids [4].

Targeted local control with intra-articular corticosteroids is usually performed as an adjunct to systemic therapy. The goal of targeted local control is to decrease the need for systemic therapy. MRI may be used as a road map to determine which individual joints to inject [17]. This is particularly useful in the hands and feet, where joints are in close proximity with each other and clinically distinguishing which individual joints are inflamed may be challenging. In addition, sometimes tenosynovitis may superficially mimic arthritis clinically and MRI may be particularly helpful distinguishing these two entities (Fig. 10). Fluoroscopy or US may be used to guide intra-articular or tenosynovial injections (Fig. 15). Our institution's preferred agent is triamcinolone hexacetonide (Aristospan; Sandoz, Princeton, NJ) at 20 mg/cc concentrate. Children are then sent home with joint immobilization to delay systemic distribution of intra-articular corticosteroids. Complications related to joint injections include infection, bleeding, soft-tissue calcifications and subcutaneous fat atrophy over the injection site.

## Conclusion

Juvenile idiopathic arthritis represents a constellation of arthritides that clinically and radiologically overlap. MRI plays an important role during disease surveillance, compared with radiography, to identify early damage to growing cartilage. The radiologist plays a key role in diagnosis, disease follow-up and image-guided treatment. A multidisciplinary approach with pediatric rheumatologists and radiologists is important to optimize patient care.

**Disclaimer** The author has no financial interests, investigational or off-label uses to disclose.

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