Editorial

Juvenile idiopathic arthritis in our orthopaedic practice

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NK Sferopoulos
Department of Pediatric Orthopaedics, "G. Gennimatas" Hospital, Thessaloniki, Greece

Correspondence: N. Sferopoulos, P. Papageorgiou 3, 546 35, Thessaloniki, Greece, Tel 00302310963270, Fax No: 00302310968265

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Editorial

This editorial reviews the most common initial clinical findings in patients referred to our pediatric orthopaedic outpatient clinic who were ultimately diagnosed with juvenile idiopathic arthritis (JIA). It also presents illustrative cases involving children younger than 14 years.

Types of JIA

Chronic arthritis in children was initially termed juvenile rheumatoid arthritis (JRA), the most common chronic arthropathy in pediatric populations. JRA was classified into three subtypes: pauciarticular (oligoarticular), polyarticular (polyarthritis), and systemic (adult-onset Still's disease). Oligoarticular disease most often affects the knees, elbows, and ankles. Polyarticular disease primarily affects the small joints of the hands and feet, as well as the wrists, major weight-bearing joints, and may involve the cervical and mandibular joints. The term JIA was introduced to encompass rheumatoid factor (RF) negative and positive polyarticular disease, systemic JIA, enthesitis-related arthritis (ERA), juvenile psoriatic arthritis (JPsA), and undifferentiated arthritis.[1,2]

Joint and tendon involvement

JIA causes persistent synovial inflammation in joints and tendon sheaths. Joint symptoms include morning stiffness, swelling, warmth, chronic pain, and limited motion. Lower limb involvement may lead to limping or walking difficulty, especially in younger children. The knee is most often affected. Tenosynovitis is common in JIA and may appear independently of joint inflammation, often affecting the tibialis posterior and peroneal tendons in the ankle or the extensor tendons in the hand.[3,4]

The value of clinical assessment in children

Differential diagnosis of childhood musculoskeletal complaints is broad and relies primarily on clinical assessment. Inadequate pediatric assessment skills can delay JIA diagnosis and referral, worsening outcomes.[5]

The hip-knee neural connection

The hip joint's deep location makes clinical signs difficult to detect, and hip pain is often felt in the knee and inner thigh, due to shared nerve pathways.[6,7]

Our trends in hip clinical examination

To prevent misdiagnosis, a thorough hip examination is essential for any child presenting with a limp, knee pain, or thigh pain. Pediatric orthopaedic surgeons are traditionally advised, to first rule out hip pathology in young children with knee or thigh pain. For younger children, the clinical examination is performed while they are naturally asleep. Both sides are assessed, beginning with the healthy side. Painful restriction of internal hip rotation is the most sensitive indicator of pediatric hip pathology and should be evaluated in both supine and prone positions. Clinical examination remains the most valuable initial approach and should not be replaced by imaging, as skipping steps may result in errors.

Hip JIA

Hip joint involvement is common in JIA, particularly in the systemic and polyarticular subtypes, as well as in ERA; however, it is rarely the first joint affected. Diagnosis is often delayed because early symptoms are vague or non-specific (Figure 1). Early hip involvement predicts a poorer outcome. Conventional hip radiography does not detect early pathological changes. Therefore, advanced imaging modalities such as ultrasound, computed tomography, and magnetic resonance imaging (MRI) are required. Severe hip damage may result from disease progression, inadequate treatment, or iatrogenic factors, most often after corticosteroid use. MRI is also critical for differentiating JIA-related hip synovitis from trauma, infection, avascular necrosis, leukemia, and other malignancies.[8–12] For example, we diagnosed leukemia in a young child with hip synovitis after an MRI revealed marrow infiltration in the pubic ramus.

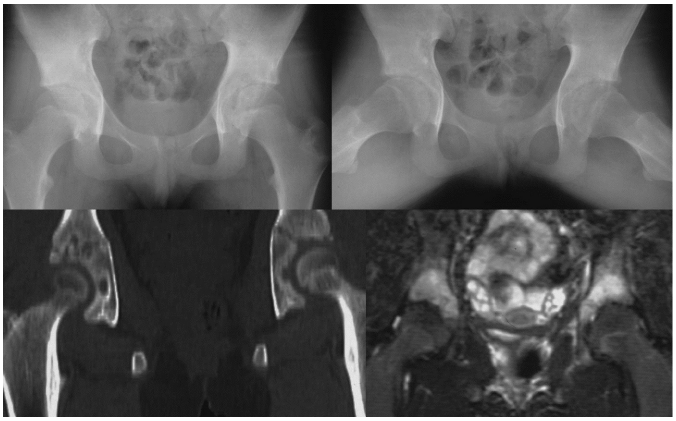

Figure 1 Imaging of an 11-year-old girl with polyarticular JIA showed bilateral hip involvement, including joint space narrowing, femoral head and acetabular erosions, and acetabular reactive subchondral edema.

Cervical spine JIA

Children with polyarticular or systemic JIA may develop cervical spine arthritis, usually as a late complication, more frequently than in oligoarthritis. Cervical spine involvement may rarely present as the initial symptom of JIA. Clinicians should consider this diagnosis in children with persistent torticollis, limited range of motion, or pain (Figure 2).[13–15] Case reports have described cervical spine involvement as the first sign of RF-negative polyarthritis.[16,17] Plain radiographs may be normal or show anterior atlanto-axial subluxation. MRI is useful for detecting inflammatory changes, including synovial proliferation, joint effusions, bone marrow edema, and cartilage-subchondral bone erosion. MRI typically shows pannus formation at the anterior and posterior margins of the odontoid, sparing the apex and alar ligament attachments (Figure 3a), which may lead to erosive changes. Ankylosis of the zygapophyseal joints at multiple levels is also a common radiographic finding in the cervical spine.[18–20]

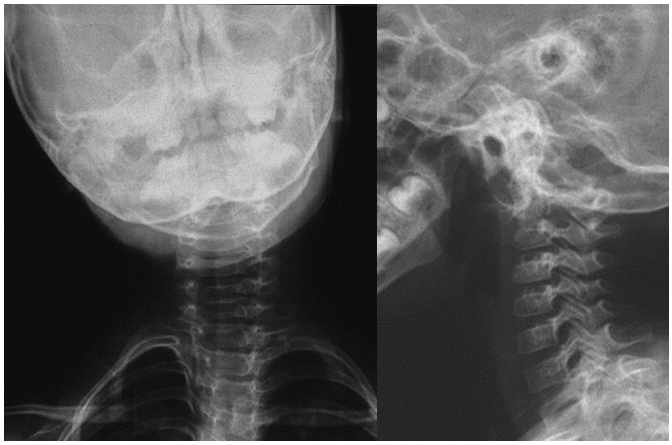

Figure 2 Cervical spine radiographs of a 3-year-old girl with a three-month history of torticollis.

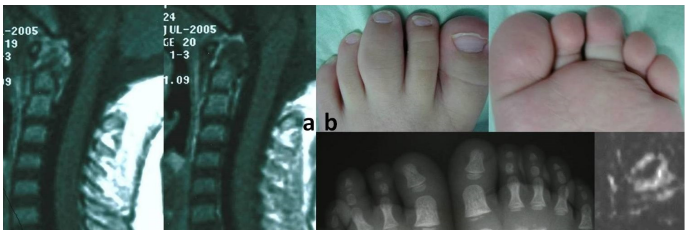

Figure 3 A 3-year-old boy presented with a six-month history of torticollis and diffuse swelling of the left third toe. Sagittal T1-weighted MRI with fat saturation and contrast showed intense enhancement and synovial hypertrophy in the atlanto-dens interval without widening (a). Radiographs demonstrated osseous overgrowth of the middle phalanx of the left third toe, and axial MRI indicated flexor-tendon synovitis. Knee involvement developed six months after the initial presentation. Medical treatment for JIA was initiated, resulting in the rapid resolution of toe swelling.

Hand and Foot JIA

Hand or foot dactylitis is the most common JIA abnormality observed in our practice and is a defining feature of JPsA. This complex inflammatory process involves tenosynovitis, usually of the flexor tendons, and is characterized by diffuse subcutaneous edema, enthesitis, and joint synovitis. Chronic, non-tender dactylitis features joint synovitis that develops after the acute phase has resolved. Dactylitis may affect one or more digits, either asymmetrically or simultaneously, resulting in a sausage-like appearance. Swelling can be localized or involve the entire digit. JPsA is a distinct JIA category and is also classified within the broader group of JSpA (juvenile spondyloarthropathies), but is distinguished by characteristic skin lesions, nail changes, and family history (Figure 4). JPsA may involve large joints, small joints, or the spine before a skin rash appears, which differs from adult presentations. When diagnosing a sausage-shaped digit (Figure 3b, 5, 6, 7), JIA, all forms of JSpA, infections, and other types of dactylitis with bone involvement should be considered. Dactylitis may persist as the only clinical manifestation of JSpA in HLA-B27-positive children (Figure 8).[21–23] We also observed localized, painful swelling of the great toe's soft tissues, without joint or tendon involvement, in a girl with Peutz-Jeghers syndrome (Figure 9).

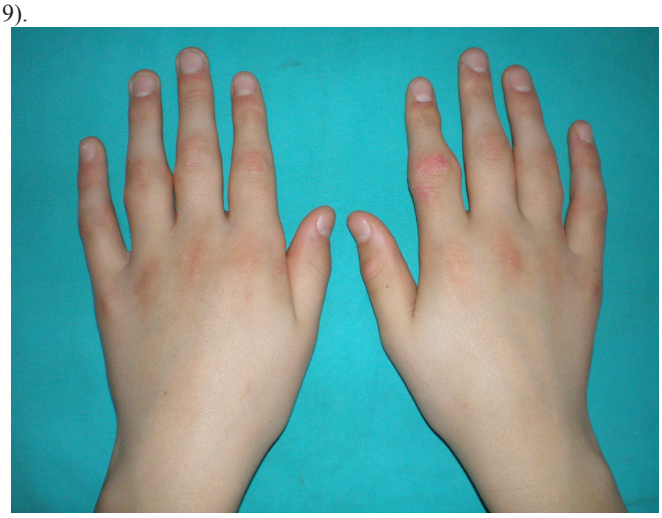

Figure 4 A 13-year-old boy with a five-year history of hand polyarthritis presented with bilateral arthritis of the metacarpophalangeal and proximal interphalangeal joints, along with a skin rash on the right index finger, but with no fever. He was diagnosed with JPsA.

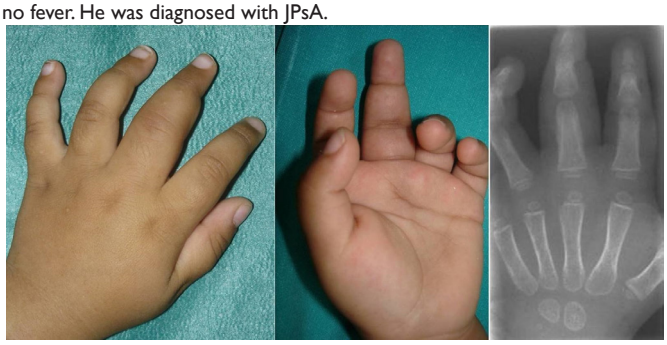

Figure 5 A 2-year-old girl presented with a four-month history of diffuse swelling and restricted movement of the left middle finger. She was afebrile and showed no systemic symptoms. Radiographs revealed periostitis of the proximal phalanx, and laboratory results were normal except for a positive antinuclear antibody titer of 1:320. Knee involvement developed after one month.

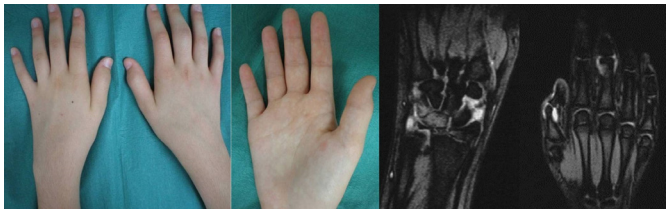

Figure 6 A 12-year-old girl presented with a six-month history of swelling and restricted movement of the right middle finger and one month of similar symptoms in the left wrist. She was afebrile, and laboratory results were normal except for a positive antinuclear antibody titer of 1:80. MRI showed inflammatory joint fluid in the left wrist, metacarpophalangeal joint of the thumb, right proximal interphalangeal joint of the middle finger, and the interphalangeal joint of the thumb.

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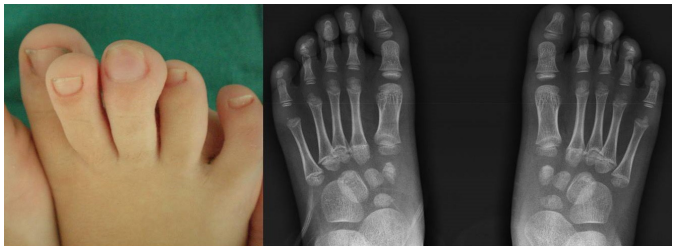

Figure 7 A 5-year-old boy presented with two months of distal swelling in the right middle toe. Radiographs showed osseous overgrowth of the distal phalanx, bilateral pseudoepiphysis of the first metatarsal, an incomplete transverse well-corticated lucency at the distal aspect, and bilateral symmetrical secondary ossification centers in the lesser metatarsals. Inflammation from JIA can disrupt the normal sequence of ossification centers in the foot, leading to premature appearance and fusion, abnormal morphology, and overgrowth.

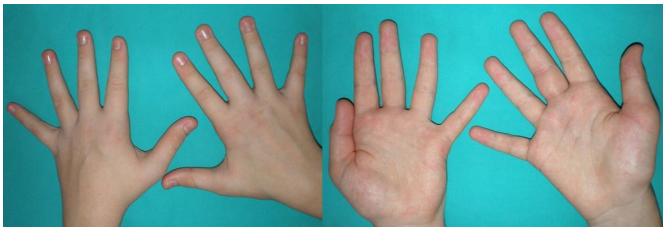

Figure 8 A 14-year-old boy presented with six months of painful, symmetrical finger swelling. HLA-B27 was positive. He received methotrexate, cortisone/prednisolone, ibuprofen/Nurofen, and sulfasalazine/Salazopyrin. He was referred for dactylitis of the right middle finger. Ultrasound showed flexor tenosynovitis.

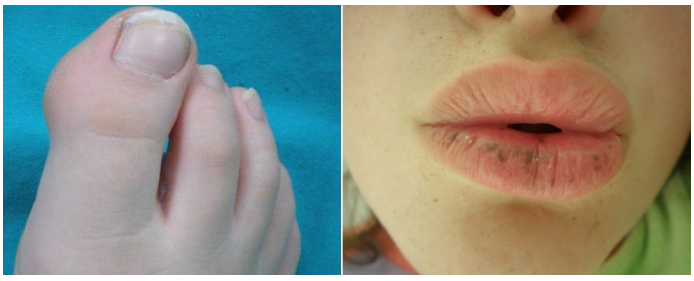

Figure 9 An 11-year-old girl presented with a six-month history of a soft tissue lesion of the right great toe. The lesion had recently enlarged and had been painful for two months, especially at night and when wearing shoes. Melanocytic macules were present on her face and lips. She had a family history of Peutz-Jeghers syndrome.

Baker's (popliteal) cyst

Baker's cyst is uncommon in children and is typically idiopathic. It usually presents as a painless mass that becomes more noticeable with full knee extension (Figure 10). Rarely, it may be the first sign of JIA, presenting with a subclinical active knee joint effusion. Popliteal cysts are common in children with JIA and active knee arthritis. Ultrasound is the preferred initial imaging technique, showing hypoechoic fluid collections in the medial popliteal fossa with internal septations that suggest posttraumatic debris or chronic synovial inflammation (Figure 11). Invasive treatment is not recommended for either children or adults. In pediatric patients, the cyst usually resolves within one to two years. In JIA, resolution occurs with treatment of the underlying disease.

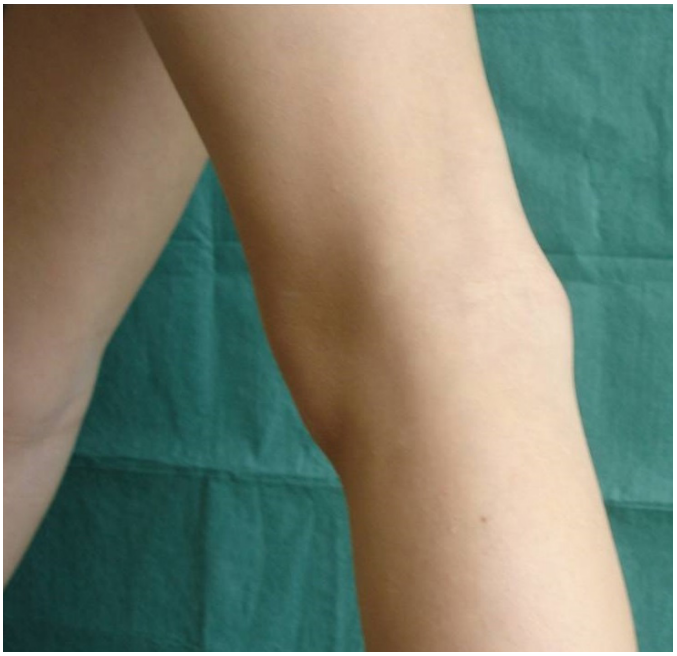

Figure 10 A 7-year-old girl was referred for a Baker's cyst.

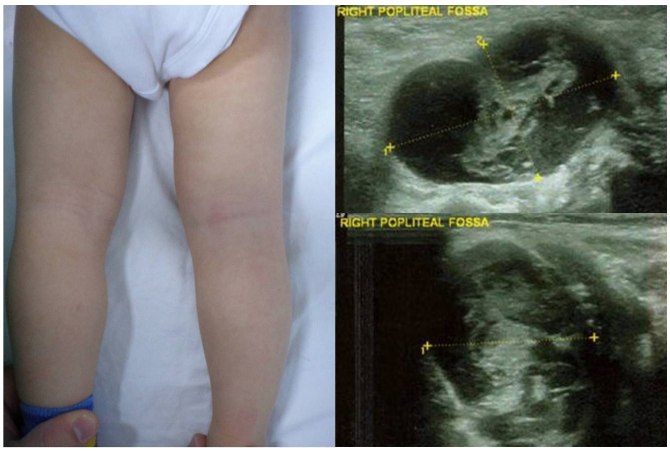

Figure 11 A 21-month-old boy presented with a two-day history of a nontraumatic popliteal cyst. Doppler ultrasound revealed heterogeneous internal echogenic tissue without vascularization.

Flat feet in JIA

A rare but serious deformity of JIA is the painful, rigid flat foot. While previously called 'spastic pronated flatfoot' or 'peroneal spastic flatfoot', the preferred term in children with JIA is 'arthritic flatfoot with peroneal spasm'.[24] Peroneal spasm (Figure 12) may be due to restricted subtalar movement and is commonly detected in patients with tarsal coalition, a congenital disorder, though most remain asymptomatic. Peroneal spasm can also result from painful tarsal joint disorders, including inflammatory (Figure 13) or degenerative arthritis, trauma, infection, and tumors. Acquired tarsal coalitions due to rheumatic diseases may also cause a painful, rigid flatfoot, with or without peroneal spasm.[25]

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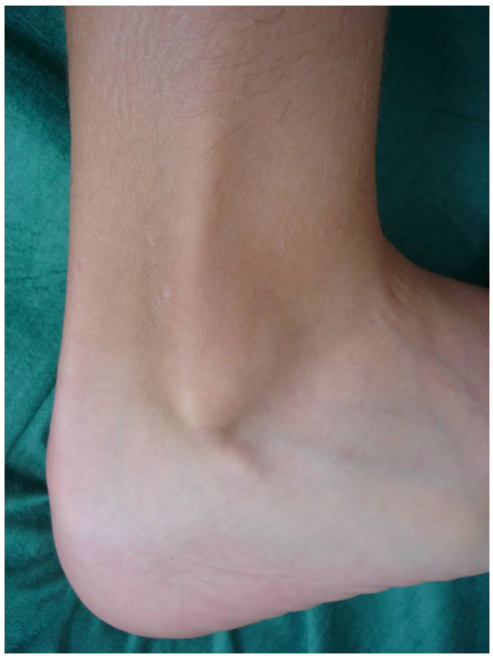

Figure 12 A 13-year-old boy presented with a two-year history of painful, rigid flat feet and peroneal spasm. The peroneal tendons appeared tight and prominent as they passed behind the lateral malleolus. Imaging revealed no coalition.

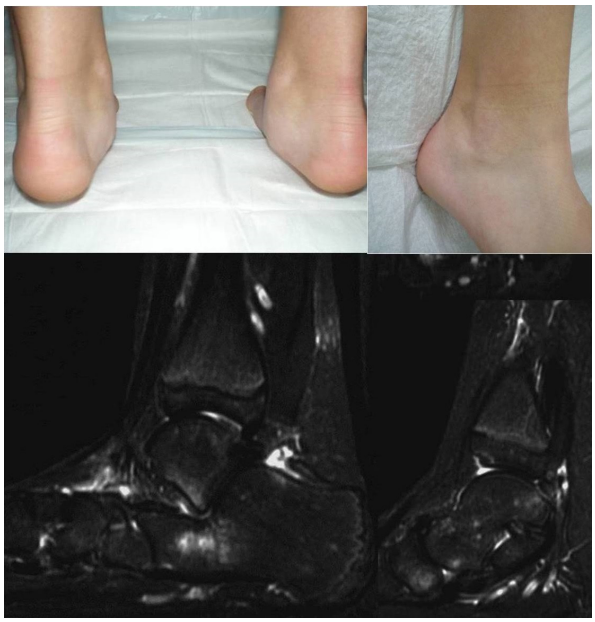

Figure 13 An 11-year-old girl had a two-year history of painful, rigid flat feet and peroneal spasm. T1-weighted MRI with fat saturation and contrast showed tenosynovitis, effusion, and synovial enhancement in the ankle, subtalar, and Chopart joints, and subarticular bone marrow edema.

In our practice, juvenile plantar dermatosis, a non-infectious inflammatory condition affecting the soles of children's feet, especially weight-bearing areas such as the forefoot, has sometimes been misdiagnosed as JIA (Figure 14). Although it may appear severe, with red, dry, shiny skin and painful cracks, it is not caused by fungi or bacteria. The condition is primarily a mechanical and environmental skin reaction, often triggered by sweating and friction inside shoes. While it is a chronic dermatitis, it is not autoimmune and should be distinguished from psoriasis or juvenile dermatomyositis.

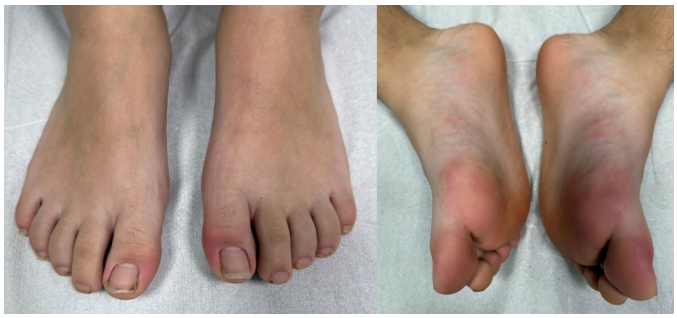

Figure 14 A 12-year-old girl was referred for pain when walking or wearing shoes. Signs of bilateral inflammation were present in the anterior third of the sole and great toes, with unaffected web spaces. She had difficulty flexing and extending her toes. The initial diagnosis of JPsA was later revised to juvenile plantar dermatosis.

Conclusion

Children with JIA involving the hands or feet should receive a thorough examination for cervical spine or large joint involvement, and vice versa. To prevent permanent impairment and achieve optimal outcomes, patients with suspected JIA should be referred promptly for pediatric or rheumatological consultation. Orthopaedic residents should avoid invasive diagnostic or treatment procedures to prevent triggering the inflammatory process, which is most aggressive in the early stages and may result in joint deformities.

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None.

Conflict of interest

The author certifies that he has no commercial associations (such as consultancies, stock ownership, equity interest, patent/licensing arrangements, etc) that might pose a conflict of interest in connection with the submitted article. The author received no financial support for this study.

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