Juvenile Idiopathic Arthritis
Oligoarthritis and Polyarthritis

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KEYWORDS

• Juvenile idiopathic arthritis • Oligoarthritis • Polyarthritis • Classification • Diagnosis • Treatment

KEY POINTS

- Juvenile idiopathic arthritis (JIA) is defined as arthritis of 6 weeks' or more duration in a child 16 years of age or younger with an unknown cause. It is a clinical diagnosis based on history and physical examination.
- Different categories of JIA have different arthritis patterns and demographics. Arthritis is divided into oligoarthritis (<5 joints) and polyarthritis, each composed of 2 subtypes.
- There is no specific diagnostic laboratory test or image, although the presence of antinuclear antibody and rheumatoid factor can help classify JIA and better guide prognosis and therapy.
- Early aggressive therapy leads to improved outcomes and prevents potential disease complications (eg, growth disturbances, joint contractures, vision loss).
- Patients should be followed by a pediatric rheumatologist for careful monitoring of medication toxicity, adverse events, and infection in all patients receiving systemic immunomodulatory medications.

INTRODUCTION

Juvenile idiopathic arthritis (JIA) broadly refers to a group of heterogeneous diseases that share the common feature of chronic inflammatory arthritis of unknown cause lasting longer than 6 weeks with onset before 16 years of age. Historically, the terms *juvenile rheumatoid arthritis* (JRA) and *juvenile chronic arthritis* were used by clinicians in accordance with the publication of classification criteria as proposed by the American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR), respectively.[1] The preferred term, *JIA*, was first introduced in 1995 by the International League of Associations for Rheumatology (ILAR) as part of the classification criteria aimed at unifying the ACR and EULAR's criteria and eliminating confusion while being

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more broadly inclusive of all forms of chronic childhood arthritis. Although the ILAR's criteria originally served to standardize research, they have been adopted by rheumatologists as a clinical diagnostic approach toward children with chronic arthritis.[1,2]

The ILAR's criteria further divide JIA into 7 relatively homogenous categories: oligoarthritis, rheumatoid factor (RF)–negative polyarthritis, RF-positive polyarthritis, systemic arthritis, psoriatic arthritis, enthesitis-related arthritis, and undifferentiated arthritis (**Table 1**). Each category differs with respect to clinical presentation and anticipated disease course. Classification is based on several factors, including the number of joints affected with arthritis, RF and HLA-B27 positivity, medical and family history, and associated extra-articular manifestations. Unlike previous classification systems, the ILAR's classification set includes exclusion criteria in an attempt to prevent overlap between categories.[1] This article focuses on oligoarthritis and polyarthritis (RF-negative and

Table 1
International League of Associations for Rheumatology's classification

ILAR Category	Definition
Oligoarthritis	Arthritis affecting 1–4 joints during the first 6 mo of disease Persistent: affecting 4 or fewer during disease course Extended: affecting more than 4 joints after first 6 mo *Exclusions: a, b, c, d, e*
RF-negative polyarthritis	Arthritis affecting 5 or more joints during the first 6 mo of disease plus a negative RF *Exclusions: a, b, c, d, e*
RF-positive polyarthritis	Arthritis affecting 5 or more joints during the first 6 mo of disease plus 2 or more positive RF tests at least 3 mo apart *Exclusions: a, b, c, e*
Systemic arthritis	Arthritis in one or more joints with or preceded by fever of at least 2-wk duration documented to be daily quotidian for at least 3 d and accompanied by at least one of the following: evanescent erythematous rash, generalized lymphadenopathy, hepatomegaly and/or splenomegaly, serositis *Exclusions: a, b, c, d*
Psoriatic arthritis	Arthritis and psoriasis OR arthritis and at least 2 of the following: dactylitis, nail pitting or onycholysis, psoriasis in a first-degree relative *Exclusions: b, c, d, e*
Enthesitis-related arthritis	Arthritis and enthesitis OR arthritis or enthesitis and at least 2 of the following: sacroiliac joint tenderness (present or historical) and/or inflammatory lumbosacral pain; presence of HLA-B27 antigen; onset of arthritis in a boy older than 6 y; acute symptomatic anterior uveitis; history of ankylosing spondylitis, enthesitis-related arthritis, sacroiliitis with inflammatory bowel disease, Reiter syndrome, or acute anterior uveitis in a first-degree relative *Exclusions: a, d, e*
Undifferentiated arthritis	Arthritis that fulfils criteria in no category or in more than 1 of the above categories

Exclusions: a: Psoriasis or a history of psoriasis in patients or first-degree relative; b: Arthritis in an HLA-B27–positive boy beginning after the 6th birthday; c: History of or a first-degree relative with ankylosing spondylitis, enthesitis-related arthritis, sacroiliitis with inflammatory bowel disease, Reiter syndrome, or acute anterior uveitis in a first-degree relative; d: Presence of immunoglobulin M RF on at least 2 occasions at least 3 months apart; e: Presence of systemic JIA.

From Petty RE, Southwood TR, Manners P, et al. International League of Associations for Rheumatology classification of juvenile idiopathic arthritis: second revision, Edmonton, 2001. J Rheumatol 2004;31(2):390–2; with permission.