Juvenile Idiopathic Arthritis Overview and Involvement of the Temporomandibular Joint

Prevalence, Systemic Therapy

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KEYWORDS

• Rheumatoid arthritis • Juvenile idiopathic arthritis • Juvenile rheumatoid arthritis
• Juvenile chronic arthritis • Temporomandibular joint • JIA • TMJ

KEY POINTS

- In the past decade there have been significant advances in the care of patients with inflammatory arthritis.
- Arthritis of the temporomandibular joint (TMJ) has benefited from these advances; yet recognition, evaluation, and management of TMJ arthritis differ significantly between adult and pediatric rheumatologists.
- Orthopantomogram and computed tomography scans have a role in evaluation of TMJ disease and are easily obtained; MRI with and without contrast using a TMJ coil is the preferred modality to determine active versus chronic disease.
- Intra-articular steroids, arthrocentesis, biologic therapies (intra-articular and systemic), and surgery have a role in preventing long-term adverse effects.

The temporomandibular joint (TMJ) is one of the many joints involved in the inflammatory arthritides. As imaging of joints has developed, so have the data regarding extent and prevalence of TMJ involvement in these diseases. TMJ disease is especially prevalent in juvenile arthritis. The adult and pediatric inflammatory arthritides share common pathophysiology but are still markedly different. The preponderance of TMJ arthritis research exists in juvenile arthritis.

CLASSIFICATION OF JUVENILE IDIOPATHIC ARTHRITIS

Arthritis in children affects approximately 300,000 in North America. Approximately half of these children have juvenile idiopathic arthritis (JIA). The prevalence of JIA in developed countries varies from 10 to 150 per 100,000. JIA is the most common chronic rheumatologic disease in the pediatric population. JIA was formerly known as juvenile rheumatoid arthritis (JRA) or juvenile chronic arthritis (JCA) in North America and Europe, respectively (**Table 1**).[1,2]

The clinical features of JIA are morning stiffness, joint swelling, and joint tenderness with changes in range of motion. It often has an insidious onset in the younger population, and its inflammatory nature, as well as the medications used to treat it, contributes to the morbidity and mortality of the disease. The current biologic medications have helped to change the extent of disability and

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Table 1
Juvenile arthritis aomenclature

Classification	ACR (1972)	ILAR (1997)
Nomenclature	Juvenile rheumatoid arthritis (JRA)	Juvenile idiopathic arthritis (JIA)
Arthritis onset	<16 y old	<16 y old
Arthritis duration	≥6 wk	≥6 wk
Subtypes, %	Pauciarticular	Oligoarticular (50%) Extended Persistent
	Polyarticular	Polyarticular RF negative (15%–25%) RF positive (5%–10%)
	Systemic	Systemic (5%–10%) Enthesitis-related arthritis (5%–10%) Psoriatic arthritis (5%–10%) Undifferentiated (10%)

Abbreviations: ACR, American College of Rheumatology; ILAR, International League of Associations for Rheumatology; RF, rheumatoid factor.

need for major surgeries and joint replacement in JIA; however, the full extent of improvement in morbidity and mortality is not well defined and is a moving target, given the growth of biologic and small molecule treatments.

The term "rheumatoid" in JRA meant a swollen joint, but was interpreted by some as adult rheumatoid arthritis (RA). The term "chronic" was introduced to help alleviate some of the issues with the American College of Rheumatology (ACR) classification of JRA, but the JCA classification system had its own limitations.[3] To help alleviate some of the confusion and shortcomings of the JRA and JCA classification, the International League of Associations for Rheumatology (ILAR) developed the JIA classification system that we use today. This classification system points out exclusion and inclusion criteria, while demonstrating the unique nature of each subtype of JIA.

The JIA nomenclature includes the latest clinical, laboratory, and genomics data; however, it is not without its own limitations.[4,5] As more advances occur in genomics, proteomics, and metabolomics, the utility and need to modify current classification schemes for autoinflammatory and autoimmune conditions will be clearer.

JIA in the ILAR classification system indicates individuals with disease onset before 16 years of age in whom the arthritis is persistent for at least 6 weeks. The ILAR JIA classification system delineates 7 onset categories: oligoarticular (persistent and extended), polyarticular rheumatoid factor (RF)-negative, polyarticular RF-positive, systemic, psoriatic arthritis, enthesitis-related arthritis, and undifferentiated. The undifferentiated category refers to patients who do not meet criteria for any subtype or meet more than one subtype.

The most common of the subtypes is oligoarticular JIA, making up approximately 50% of patients with JIA. The peak age of onset is 2 years with a female predilection of 5:1 female:male. By definition, patients with oligoarticular JIA have 1 to 4 joints affected in the first 6 months of disease. After the first 6 months, 2 categories exist to define the type: extended oligoarticular JIA or persistent oligoarticular JIA. Extended oligoarthritis encompasses individuals with more than 4 affected joints after the first 6 months of disease. Persistent oligoarthritis is involvement of no more than 4 joints throughout the disease. Children are excluded from this classification if they have psoriasis or a first-degree relative with psoriasis; are HLA-B27 positive or RF positive; or have ankylosing spondylitis, Reiter syndrome, sacroiliitis with inflammatory bowel disease, or systemic JIA features. The antinuclear antibody (ANA) test is positive in up to 80% of oligoarticular JIA. The oligoarticular JIA population does not typically have a positive RF. Up to 30% of oligoarticular JIA children develop uveitis, which is sometimes called iritis or iridocyclitis.

Patients with polyarticular JIA make up 4 in 10 children with JIA. Polyarticular JIA indicates arthritis in 5 or more joints during the first 6 months of disease. Five percent to 10% of children with JIA have RF-positive polyarticular JIA. A positive RF must be demonstrated 2 or more times at least 3 months apart in the initial 6 months of disease onset.

Children with RF-positive polyarticular JIA are the subtype most similar (genotype and