

Leading Article

Juvenile idiopathic arthritis-a new classification

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Juvenile idiopathic arthritis (JIA) is defined as a chronic inflammatory disease characterised predominantly by idiopathic arthritis with onset before the 16th birthday, in the absence of a defined diagnosis (e.g. systemic lupus erythematosus, rheumatic fever, septic arthritis, neoplasia, immune deficiency etc).

This is the commonest rheumatic disease of children. Although the primary purpose of classification of Juvenile Idiopathic Arthritis is scientific, criteria developed for classification are commonly used for diagnostic purposes also. This is particularly so in rheumatology because of the slow evolution of signs and symptoms, absence of specific laboratory tests and considerable overlap between these diseases.

Nomenclature and classification

The term juvenile rheumatoid arthritis (JRA) is been used extensively in United States and Canada as the preferred term. The American College of Rheumatology (ACR) published its criteria for the diagnosis of JRA in 1973 and modified it in 1977.

ACR criteria for JRA

- Onset less than 16 years of age
- Persistent arthritis for over 6 weeks
- Types of onset:
 - Pauciarticular: less than 5 joints
 - Polyarticular: more than 4 joints
 - Systemic: characteristic fever and rash

The European League Against Rheumatism (EULAR) coined the term Juvenile Chronic Arthritis (JCA) and published its criteria for the diagnosis of JCA at the same time. The term JCA is used in England and Europe. Both terms JRA and JCA however are used interchangeably.

EULAR criteria for JCA

- Onset less than 16 years
- Persistent arthritis for over 3 months
- Types of onset:
 - Pauciarticular: less than 5 joints
 - Polyarticular (RF negative): more than 4 joints
 - Systemic
 - JRA (RF positive polyarticular)
 - Ankylosing spondylitis
 - Psoriatic arthropathy

Therefore these two definitions of childhood onset arthritis use different sets of criteria for classification of idiopathic arthritis of childhood.

In order to unify the language and define disease in a universally accepted manner; a task force was established by the International League Against Rheumatic diseases (ILAR). This group proposed a new set of criteria in 1995. In this classification each subset is clearly defined with description of the required characteristics, exclusions and descriptors. This proposed system specifies that only a six week period of persistent arthritis is required before the diagnosis can be made, eliminating a JCA/JCR discrepancy. It also coined the new term Juvenile Idiopathic Arthritis (JIA) to define this group of children with inflammatory arthritis.

Each of the subgroups given below are named and defined according to the proposed ILAR classification, formulated by the WHO Task Force for Classification Criteria at the initial meeting in Chile in 1995 and substantially revised at the second meeting in Durban in 1997. The following is the 1997 version.

Systemic arthritis

Arthritis with or preceded by fever of at least 2 weeks duration documented to be quotidian for at least 3 days, and accompanied by 1 or more of the following;

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- Evanescent, non-fixed, erythematous rash
- Generalised lymph node enlargement
- Hepatomegaly or splenomegaly
- Serositis

It is responsible in about 10 to 15% of patients with JIA.

Polyarthritis: Rheumatoid factor negative

Arthritis affecting 5 or more joints during the first 6 months of the disease.

Polyarthritis: Rheumatoid factor positive

Arthritis affecting 5 or more joints during the first 6 months of the disease, associated with positive rheumatoid factors tests on 2 occasions at least 3 months apart. This is a true childhood counterpart of adult Rheumatoid arthritis. A family history of psoriasis is a specific exclusion for this group.

Oligoarthritis

Arthritis affecting 1 to 4 joints during the first 6 months of the disease. Children with a family history of psoriasis or with a positive rheumatoid factor are excluded from this group.

A. Persistent oligoarthritis

Arthritis affecting 1 to 4 joints.

This is associated with HLA _A2, DR 3 or 8 and DPB1.0201.

B. Extended oligoarthritis

Arthritis affecting 1 to 4 joints during the first 6 months of disease, but progressing to a cumulative total of 5 joints or more after the first 6 months of the disease. This is associated with HLA DR 1.

Enthesis related arthritis

Arthritis and enthesitis, or:

Arthritis or enthesitis with at least 2 of:

- Sacroiliac joint tenderness and/or inflammatory spinal pain
- HLA B27
- Family history in 1st or 2nd degree relatives consistent with HLAB27 - associated disease medically confirmed
- Anterior uveitis that is usually associated with pain, redness or photophobia
- Male older than 8 years at the onset of arthritis.

At present, children with ANA, rheumatoid factor, or inflammatory bowel disease are specifically excluded.

Psoriatic arthritis

Arthritis and psoriasis or

Arthritis and family history of psoriasis in parents or siblings, in addition to dactylitis in the patients or nail abnormalities (pitting or onycholysis) in the patient. Children with a positive rheumatoid factor are excluded from this group.

This nomenclature is yet not complete. There are children who do not fit into any of the above categories, or who develop more than one category. The inability to satisfactorily place the child into one of these categories does not exclude the presence of arthritis or the ability to make a diagnosis. Rather, these children should be told that they have JIA.

This nomenclature will be further refined in the future, to allow better characterisation of children with idiopathic arthritis.

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