

Growing Pains: The ILAR Classification of Juvenile Idiopathic Arthritis



Classification is a necessary, if a sometimes perplexing, process. This is nowhere more evident than in rheumatology, where attempts at classification of the vasculitides, the sclerodermas, and childhood arthritis have helped bring clarity to realms of medicine that depend to a great extent on accurate clinical observation rather than on illumination provided by the laboratory. The International League of Associations for Rheumatology (ILAR) classification of juvenile idiopathic arthritis (JIA) recognizes 8 categories: systemic arthritis, oligoarthritis, extended oligoarthritis, polyarthritis [rheumatoid factor (RF) positive], polyarthritis (RF negative), enthesitis related arthritis, psoriatic arthritis, and “other arthritis.” These criteria, initially proposed in 1994 (the Santiago criteria)¹ and subsequently revised in 1997 (the Durban criteria)² have now been endorsed by the World Health Organization (1999). Their adoption by much of the pediatric rheumatology world is both encouraging and anxiety provoking: Encouraging because the use of a common classification in order to communicate accurately is a step in the right direction; anxiety provoking, because there is concern that the criteria are being applied as approximations rather than in a disciplined manner, with the result that the information yielded may yet be biased by individual perceptions. If this is the case, the gain is small, and the original intent of the classification is undermined.

The ILAR criteria require further evaluation and revision. Such an evaluation has been presented by Hofer, Mouy, and Prieur in this issue of *The Journal*³. Hofer and his colleagues have carefully applied the revised ILAR criteria to 194 patients with juvenile onset of chronic arthritis not due to other causes. They describe in some detail the reasons that prevent or complicate appropriate categorization; they then offer several suggestions for revision aimed at decreasing the number of patients fitting the category “other arthritis” without diluting the homogeneity of the population in each category:

1. That the category Polyarthritis, Rheumatoid Factor

Positive be changed to Oligo or Polyarthritis, Rheumatoid Factor Positive

2. That a new diagnostic category, Probable Psoriatic Arthritis, be added to include patients with a family history of psoriasis, but neither dactylitis nor nail changes

3. That cervical spine arthritis alone be insufficient evidence of spinal involvement to satisfy a criterion for Enthesitis Related Arthritis

4. That a diagnosis of RF negative polyarthritis be excluded under the following circumstances:

- the presence of enthesitis or sacroiliitis in a HLA-B27 positive boy over 8 years of age at onset of disease
- psoriasis in the patient, or a family history of psoriasis

5. That psoriasis in the patient be an exclusion criterion for Enthesitis Related Arthritis.

Application of these suggested changes reduced the number of children who fell into the category “other arthritis” dramatically — from 39 to 4. The determination to minimize the number of patients in the category inelocquently termed “other arthritis” is a recurring theme among many of the published studies or abstracts concerning the ILAR criteria. Although the result appears neater, there is a danger that pursuit of this goal will compromise the search for homogeneity within groups, enunciated as one of the principal guidelines for the ILAR process. Is it more important, for example, to group all children with rheumatoid factor in one category than to group children with oligoarthritis separately from those with polyarthritis? The authors point out that assessment of the frequency of HLA-DR4 in oligoarticular onset RF positive patients might help to clarify the appropriateness of this change.

The next revision of the ILAR criteria will have to consider both housekeeping and substantive changes. Housekeeping changes include increased precision and consistency in definition of the criteria. Thus, there are some inconsistencies within the classification itself, particularly with reference to the presence of psoriasis or a family

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history of psoriasis. A family history of psoriasis in a first or second degree relative is reason to exclude a patient from the oligoarthritis and enthesitis related arthritis categories, but not from polyarthritis RF positive or negative categories or from the systemic arthritis category. The requirement that psoriasis must be diagnosed by a dermatologist is very difficult to apply in practice, and, one suspects, is seldom strictly adhered to by those using the classification. Whether the requirement for 2 tests for RF at least 3 months apart in the first 6 months of disease is either practical or important requires evaluation. Substantive changes will require extensive discussion, and should be based on evidence from the clinic and the laboratory. The proposals made by Hofer and colleagues deserve serious consideration, but should not be applied unless and until they are endorsed by the ILAR Pediatric Standing Committee, lest we witness a proliferation of local modifications of the criteria, a situation that would set back the quest for a universally used set of criteria by decades.

In another approach, the British Paediatric Rheumatology Study Group employed latent class analysis to identify subtypes of childhood arthritis and compare them to the ILAR criteria⁴. Using a combination of 10 different clinical and serologic characteristics, 175 different profiles were observed among 572 patients. The latent classes correlated strongly only with the polyarthritis, RF positive category of the ILAR classification, however. A number of other studies have been presented at the European Pediatric Rheumatology Society meeting and the American College of Rheumatology meeting in 2000. Most recommended

changes similar to those suggested by Hofer and colleagues. The limits of clinical evaluation of the criteria are approaching, however. It is time to evaluate the ILAR criteria in a biologic context. If the classification is to have meaning beyond the semantic, it will be borne out in studies of genetics, molecular biology, therapeutic response, and outcome. Such studies are eagerly anticipated.

ROSS E. PETTY, MD, PhD, FRCPC,
Professor and Head,
Division of Rheumatology,
Department of Pediatrics,
University of British Columbia, and
British Columbia's Children's Hospital,
4480 Oak Street, Room 1A21,
Vancouver, BC, Canada V6H 3V4.

Address reprint requests to Dr. Petty.

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