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Editorial

Newly Proposed Classification for Juvenile Idiopathic Arthritis

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Juvenileid iopathic arthritis (JIA) is a clinically heterogeneous group of arthritides categorized by the International League of Associations for Rheumatology (ILAR). JIA begins before the age of 16 years, persists for more than 6 weeks, and is of unknown cause [1]. The ILAR classification was proposed to resolve the ambiguities associated with, and compensate for the differences in an earlier classification system. The ILAR classification was based on clinical findings, family history, and laboratory test results. Six distinct categories of JIA are defined according to specific inclusion and exclusion criteria for the maintenance of homogeneity within each category and to avoid overlap [2]. About 20% of children with chronic arthritis do not meet the criteria for any of the six categories of JIA; thus, a seventh category, undifferentiated arthritis, can be used to capture these patients, as well as those who fit into more than one category [2,3]. However, The ILAR classification has the limitation of not being informed by clinical data, instead being based on expert consensus [4].

Several investigators have evaluated the ILAR classification system. In one study, a homogeneous group of patients characterized by the presence of antinuclear antibody (ANA), early onset, female predominance, asymmetric arthritis, and high incidence of iridocyclitis was classified into three different JIA categories: oligoarthritis, rheumatoid factor (RF)-negative polyarthritis, and psoriatic arthritis. Moreover, the number of joints affected during the first 6 months of the disease, in addition to the presence of psoriasis, did not constitute useful criteria for identifying homogeneous disease entities [5]. In a subsequent study, ANA-positive patients grouped into the categories persistent oligoarthritis, extended oligoarthritis, and RF-negative polyarthritis were very similar in terms of age at presentation, female-to-male ratio, frequency of asymmetric arthritis, and incidence of iridocyclitis [6]. Based on these similarities, it has been proposed that the ILAR classification should be revised so that persistent oligoarticular, extended oligoarticular, RF-negative, and ANA-negative polyarthritis are all within the same category [6,7]. Furthermore, one study that strictly applied the ILAR classification criteria found that up to 30% of the JIA population should be classified as undifferentiated arthritis [8].

In a previous issue of this journal, Kwon et al. [9] proposed that JIA patients should be reclassified according to RF and ANA to identify homogeneous disease entities, while the number of involved joints and the presence of psoriasis were recommended to be excluded from the ILAR classification. The medical records of 262 JIA patients were investigated retrospectively and the patients were reclassified into six categories using the new classification system proposed by Martini at the 23rd Paediatric Rheumatology European Society Congress (2016) [9]. The new classification system recognizes the following six categories, in which the number of joints criterion was replaced by positivity for RF and ANA or anti-cyclic citrullinated peptide antibody (anti-CCP Ab): systemic JIA, RF-positive JIA, early onset ANA-positive JIA, enthesitis/spondylitis-related JIA, other JIA, and unclassified JIA. Patients not meeting the criteria for the first four categories, or those fitting into more than one category, were classified as other JIA or unclassified JIA [9]. RF-positive JIA, early onset ANA-positive JIA, and enthesitis/spondylitis-related JIA were significantly different in terms of the gender ratio, age of disease onset, and the cumulative number and type of involved joints. However, 24.8% of

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the patients did not meet the criteria for one of these four categories and were reclassified as other JIA, which is not a homogeneous disease entity in the new classification. Kwon et al. [9] stated that a lower rate of ANA positive might result in a higher proportion of other JIA cases. In one multi-ethnic cohort, patients of Asian origin had the lowest rate of early onset ANA-positive arthritis (18%), while those of native North American origin had the highest rate (40%); however, the difference was not significant [10]. Although this new classification system may offer clarity regarding the categorization of JIA, it is not the case for others.

The goal when developing a new JIA classification system is to define homogeneous groups of patients, especially those who might not meet more rigid classification criteria, and to facilitate the development of better and more specific therapies by JIA category through an improved understanding of the pathophysiology of the disease [2,3]. Thus, it is necessary to revise some of the present JIA classification systems to better-identify homogeneous patient populations. Rapid advances in genomics and gene expression analysis may provide an opportunity to modify the JIA classification system and thus resolve the current controversies [2].

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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