Evaluation of Revised International League of Associations for Rheumatology Classification Criteria for Juvenile Idiopathic Arthritis in Spanish Children (Edmonton 2001)

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ABSTRACT. ***Objective.*** To evaluate the revised (Edmonton 2001) International League of Associations for Rheumatology (ILAR) classification criteria for Juvenile Idiopathic Arthritis (JIA) in a cohort of Spanish children.

Methods. One hundred twenty-five patients with chronic arthritis categorized according to traditional criteria and to the first revision of ILAR JIA criteria (Durban 1997) were reclassified according to the second JIA criteria revision (Edmonton 2001).

Results. Edmonton criteria allocated 92% of the patients classified by traditional criteria in their corresponding ILAR categories. Most patients with systemic (94%), pauciarticular (91%) and polyarticular (88%) juvenile chronic arthritis as well as those with juvenile spondyloarthropathy (94%) were reclassified in the corresponding ILAR categories. Two children with probable psoriatic arthritis (PsA) were reclassified in the rheumatoid factor-negative (RF–) polyarthritis category, whereas only one of 2 children with definite PsA could be allocated to the ILAR PsA class. Ten patients (8%) constituted the undifferentiated arthritis group, 8 because of psoriasis in a first-degree relative, one because of the presence of RF in a girl with oligoarthritis, and another because of psoriasis in a boy who was HLA-B27-positive. In comparison with the Durban JIA criteria the Edmonton revision decreased the number of patients whose arthritis fulfilled criteria in no category or in 2 or more categories (from 19 to 10), and delineated better the population included in the RF– polyarthritis category.

Conclusion. The Edmonton criteria made the ILAR classification more transparent and easy to apply. Family history of psoriasis was responsible for most allocations to the undifferentiated arthritis category (8/10). (J Rheumatol 2005;32:559–61)

Key Indexing Terms:

CHILDHOOD ARTHRITIS
CLASSIFICATION CRITERIA
JUVENILE SPONDYLOARTHROPATHY
JUVENILE IDIOPATHIC ARTHRITIS
JUVENILE CHRONIC ARTHRITIS
JUVENILE PSORIATIC ARTHRITIS

In 1994 an International League of Associations for Rheumatology (ILAR) Taskforce on Classification of Childhood Arthritis was created with the aim of proposing "a unified, internationally acceptable and applicable set of classification criteria...and to facilitate more meaningful research and better patient care." The original set proposed in Santiago, Chile[1], was revised in 1997 in Durban, South Africa[2]. The umbrella term juvenile idiopathic arthritis (JIA) was coined at that time to indicate arthritis with onset before the 16th birthday, persisting for at least 6 weeks, of unknown cause. The ILAR classification criteria of JIA was updated in 2001 in Edmonton to make the classification "more transparent, consistent and easy to apply" and to achieve "relatively homogeneous, mutually exclusive categories of idiopathic childhood arthritis based on predominant clinical and laboratory features"[3].

We evaluated the revised ILAR criteria in a group of Spanish children with chronic arthritis previously classified according to a traditional set of criteria as well as according to Durban criteria.

MATERIALS AND METHODS

One hundred twenty-five patients previously classified according to the Durban criteria[4] were reclassified following the revised Edmonton JIA criteria. All patients had initially been categorized according to a traditional classification set of criteria, including European League Against Rheumatism criteria[5] for juvenile chronic arthritis (JCA), European Spondylarthropathy Study Group preliminary criteria for juvenile spondyloarthropathy (SpA)[6], and Vancouver criteria for juvenile psoriatic arthritis (PsA)[7].

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Accepted November 15, 2004.

RESULTS

Edmonton JIA criteria vs traditional criteria. The Edmonton criteria allocated 92% of the patients classified by traditional criteria in their corresponding ILAR categories (Table 1), including most patients with systemic (17/18, 94.4%), pauciarticular (54/59, 91.5%), and polyarticular JCA (22/25, 88%), as well as those with juvenile SpA (16/17, 94,1%).

Ten children diagnosed as either JCA or juvenile SpA by traditional criteria were classified in a different JIA category (Table 1). One 11-year-old HLA-B27-positive boy formerly classified as pauciarticular JCA was allocated to the enthesitis related arthritis (ERA) group. Another patient classified as pauciarticular JCA, a 9-year-old girl with oligoarthritis at disease onset, was excluded from the ILAR oligoarthritis category due to the presence of persistently positive rheumatoid factor (RF). She was classified in the undifferentiated arthritis group. The remaining 8 patients were not included in the corresponding ILAR categories because of a history of psoriasis in a first-degree relative (Table 2), including one patient with systemic JCA.

There were 4 patients categorized as juvenile PsA according to the Vancouver criteria. The 2 patients with probable PsA (arthritis plus dactylitis and family history of psoriasis in a second-degree relative) were classified by the Edmonton criteria in the polyarthritis (RF-negative) category. Only one of 2 children with definite PsA by the Vancouver criteria (arthritis and psoriasis) was classified in the ILAR PsA category. The other patient with definite PsA was a 7-year-old HLA-B27-positive boy whose B27 status precluded him from inclusion in the ILAR PsA category, whereas his psoriasis excluded him from the JIA ERA group. He was classified in the ILAR undifferentiated arthritis category.

Ten patients (8%) were allocated to the undifferentiated arthritis group (Table 2), 8 because of a history of psoriasis in a first-degree relative, one because of the presence of RF in a girl with oligoarthritis, and one because he fulfilled criteria in no category.

Edmonton vs Durban JIA criteria. The Edmonton revision improved the homogeneity of the RF-negative polyarthritis group and decreased the number of patients not classifiable in other categories (those included in the "undifferentiated" and "other" arthritis groups) from 19 to 10, respectively

Table 1. Classification of patients according to the proposed ILAR JIA and traditional classification systems.

	Edmonton Revision/ILAR Classification Criteria						
Traditional Criteria	Oligo, n = 54	Poly RF–, n = 24	Poly RF+, n = 2	Systemic, n = 17	ERA, n = 17	Juvenile PsA, n = 1	Undifferentiated Arthritis, n = 10
Pauci JCA, n = 59	54	—	—	—	1	—	4
Poly JCA, n = 25	—	22	—	—	—	—	3
Poly RF+, n = 2	—	—	2	—	—	—	—
Systemic JCA, n = 18	—	—	—	17	—	—	1
Juvenile SpA, n = 17	—	—	—	—	16	—	1
Juvenile PsA, n = 2	—	—	—	—	—	1	1
Probable juvenile PsA, n = 2	—	2	—	—	—	—	—

JCA: juvenile chronic arthritis; RF: rheumatoid factor; SpA: juvenile spondyloarthropathy; PsA: psoriatic arthritis; ERA: enthesitis related arthritis.

Table 2. Clinical and laboratory features of the patients classified in the ILAR JIA undifferentiated arhtirits category.

N	Sex	Initial Diagnosis	JIA Durban	Age at Onset, (yrs)	History of Psoriasis†	Disease Course	Small Joints of the Hand	ANA	RF	B27	Lumbar/SI Pain	Enthesitis	Dactylitis
1	M	PsA	PsA	7.6	Yes	Oligoarthritis (A)	No	+	–	+	No	No	No
2	F	Poly RF–	Poly RF–	10.5	Yes	Polyarthritis (S)	Yes	–	–	–	No	No	No
3	F	Poly RF–	Poly RF–	3.1	Yes	Oligoarthritis (S)	No	+	–	–	No	No	No
4	F	Pauci JCA	Other	7.7	Yes	Oligoarthritis (A)	Yes	+	–	–	No	No	No
5	F	Poly RF–	Poly RF–	1.5	Yes	Polyarthritis (A)	Yes	+	–	–	No	No	No
6	F	Pauci JCA	Other	9.5	Yes	Oligoarthritis (A)	No	–	–	–	No	Yes	No
7	M	Juvenile SpA	Other	12.9	Yes	Oligoarthritis (A)	Yes	–	–	+	Yes	Yes	Yes
8	M	Pauci JCA	Other	6.9	Yes	Oligoarthritis (A)	No	–	–	–	No	No	No
9	M	Syst	Syst	6.9	Yes	Oligoarthritis (A)	No	–	–	–	No	No	No
10	F	Pauci JCA	Other	9.2	No	Polyarthritis (S)	No	–	+	–	No	No	No

† History of psoriasis in the patient (Patient 1) or in a first-degree relative (Patients 2–9). (S): Symmetric arthritis (involvement of the same joint area on both sides of the body in 75% or more of the affected joints). (A): Asymmetric arthritis. Poly RF–: polyarticular rheumatoid factor (RF) negative juvenile chronic arthritis (JCA); Pauci: pauciarticular; SpA: spondyloarthropathy; Syst: systemic JCA or systemic JIA; PsA: psoriatic arthritis; ANA: antinuclear antibody; SI: sacroiliac.

Table 3. Classification of patients according to Durban and Edmonton revised ILAR JIA classification criteria.

Traditional Criteria	JIA Edmonton Oligo, n = 54	Poly RF–, n = 24	Poly RF+, n = 2	Systemic, n = 17	ERA, n = 17	PsA, n = 1	Undifferentiated Arthritis, n = 10
Oligo, n = 47	47	—	—	—	—	—	—
Poly RF– n = 28	—	24	—	—	1	—	3
Poly RF+, n = 2	—	—	2	—	—	—	—
Systemic, n = 18	—	—	—	17	—	—	1
ERA, n = 9	—	—	—	—	9	—	—
PsA, n = 2	—	—	—	—	—	1	1
Other arthritis, n = 19	7	—	—	—	7	—	5

RF: rheumatoid factor; PsA: psoriatic arthritis; ERA: enthesitis related arthritis.

(Table 3). In this regard, 14 of 19 children classified in the Durban JIA "other arthritis" category could be assigned to either the oligoarthritis (n = 7) or the ERA (n = 7) revised categories. Five children classified in several Durban JIA categories, however, had to be allocated to the Edmonton undifferentiated arthritis group (Table 3).

DISCUSSION

The Edmonton criteria allocated 115 of our 125 patients (92%) into distinct diagnostic categories, indicating that most patients categorized according to traditional criteria could be classified in their corresponding Edmonton revised ILAR categories. Among the children who could not be classified in the corresponding ILAR category there was a child with oligoarthritis and persistently positive RF, and an older HLA-B27-positive boy with psoriasis. In both cases the revised criteria achieved what they were supposed to, that is, "relatively homogeneous, mutually exclusive categories of idiopathic childhood arthritis." The other 8 patients classified in the undifferentiated arthritis group were excluded from other categories because of a family history of psoriasis. In most instances there was a possibility to develop PsA in the future, although this did not seem to be the case in our patient with systemic disease. It may be worthwhile to address this issue in future revisions of the criteria.

Compared to the former JIA criteria (Durban), the Edmonton revised criteria were clearly easier to apply. In addition, the updated criteria addressed and resolved the main problems reported during the validation of former ILAR criteria in Spanish children[4], the interference detected between the categories ERA and polyarthritis (RF-negative), and the exclusion of a significant number of children from other categories because of a family history of psoriasis in second-degree relatives. Accordingly, the application of the revised ILAR criteria resulted in a more precise definition of exclusion criteria for the RF-negative polyarthritis category, and a decreased number of patients who were not classifiable.

As we concluded 2 years ago[4], the ILAR proposal represents a much needed effort to develop internationally accepted criteria that would facilitate communication among patients, physicians, and scientists, and enable the identification of homogeneous groups of children with chronic arthritis. In light of this and other studies that have analyzed the performance of the ILAR JIA classification system, these criteria merit further evaluation and revision.

ACKNOWLEDGMENT

The authors thank Dr. Daniel Lovell for his review of the manuscript and helpful comments.

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