

Construct Validity of ILAR and EULAR Criteria in Juvenile Idiopathic Arthritis: A Population Based Incidence Study from the Nordic Countries

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ABSTRACT. Objective. New classification criteria (ILAR) have been proposed for juvenile idiopathic arthritis (JIA). They are more descriptive than those formerly used [American College of Rheumatology (ACR), European League Against Rheumatism (EULAR)], but require validation against classifications already in use. We validated the ILAR criteria in relation to the EULAR criteria in a prospective, incidence, and population based setting, and analyzed their feasibility.

Methods. Construct validity of ILAR and EULAR classification criteria refers to how closely the 2 instruments are related and how each of them operates in classifying subgroups/categories. Twenty doctors in 5 Nordic countries collected data from the incidence cases within their catchment areas during an 18 month period beginning July 1, 1997. Clinical and serological data from the first year of disease were collected.

Results. A total of 322 patients were included. Classification according to the ILAR criteria was possible in 321 patients; 290 patients had a disease duration ≥ 3 months and were classified according to the EULAR criteria. One child could only be classified according to the EULAR criteria. Thus, 31/322 (9.6%) children were classified according to the ILAR criteria only. Forty-eight of 321 (15%) patients did not fit into any category and 6% (20/321) fulfilled criteria for 2 categories. In the ILAR classification 5 out of 7 categories/subgroups have 2 to 5 specified exclusion criteria that highly discriminate the definition of each patient. In our study the exclusion criteria were fulfilled to only a small extent.

Conclusion. The EULAR and ILAR criteria differ concerning the operational definitions of the subvariables involved, which complicates their comparison. By using ILAR rather than EULAR criteria the number of cases with juvenile arthritis increased by 10%, considering the first half-year after onset. The validity of the ILAR criteria is low since they often exclude patients from subgroup classification and the possibility of having more than one diagnosis is not negligible. The specified exclusion criteria for some of the subgroups are difficult to fulfill in clinical work and variables involved could be questioned with regard to their consistency. (J Rheumatol 2001;28:2737-43)

Key Indexing Terms:
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CLASSIFICATION CRITERIA

Idiopathic arthritis of childhood involves a spectrum of clinical findings and onset is possible at any time before 16 years of age. The heterogeneity of childhood chronic arthritis is one explanation for classification difficulties.

Today, different classifications are used in different parts of the world, an indication that no classification system is considered good enough to gain universal acceptance.

We wish to stress the purposes of disease classifica-

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tion^{1,2}: Classification may serve an epidemiological function in supporting health care planning and giving clues to etiological factors. Another purpose is to facilitate scientific cooperation to find biologically homogenous groups. As long as we do not have a common classification system, we will have problems in comparing one study with another. A new classification system that functions well in everyday clinical work would facilitate prognostication, as well as judgments regarding medical treatment. With new drugs and new modes of treatment this is becoming even more important.

The American College of Rheumatology (ACR)³⁻⁵ criteria are most commonly used in the United States, while the criteria of the European League Against Rheumatism (EULAR) criteria⁶ are used in most European countries. In addition to using different terms [juvenile rheumatoid arthritis (JRA) in ACR criteria and juvenile chronic arthritis (JCA) in EULAR criteria) the time required for diagnosis is 6 weeks for ACR criteria versus 3 months for EULAR criteria. Another important difference is that the EULAR criteria include more conditions than the ACR criteria, such as probable juvenile ankylosing spondylitis, juvenile psoriatic arthropathy, and arthropathies associated with inflammatory bowel disease.

Neither the ACR nor the EULAR criteria have been prospectively validated. Importantly, a new classification for juvenile arthritis would need to be reliable and easy to interpret in clinical work.

In 1995 the Pediatric Standing Committee of the International League of Associations for Rheumatology (ILAR) proposed a new set of criteria, using the umbrella term “juvenile idiopathic arthritis” (JIA)⁷. These criteria were revised in 1997, and they have been called the “Durban criteria”⁸ (Table 1). They were meant to be more biologically relevant than the old ones, but before acceptance their construct and predictive validity need to be investigated in a population based setting. The ILAR criteria introduce a new classification aspect by specifying strict exclusion criteria, specific to 5 of the 7 categories/subgroups. These new criteria also need to be validated to determine their feasibility. We assessed the construct validity of the ILAR and EULAR criteria in a prospective, incidence and population based setting in the Nordic countries.

MATERIALS AND METHODS

Study population. The study population consisted of 322 children from 5 Nordic countries: Sweden, Norway, Finland, Iceland, and Denmark. All children are incidence cases identified prospectively between July 1, 1997, and December 31, 1998, a period of 18 months. The patients were followed up clinically at 6 and 12 months.

Catchment area. The catchment area and population was well defined in each country: Sweden, 5 counties and one health district; Denmark, Århus County and the eastern part of Denmark with the islands of Sjælland, Bornholm, Møn, and Lolland-Falster, but not the county of Copenhagen; Norway, 2 counties in the Tromsø region and 3 in the Trondheim region; Finland, the Helsinki University Central Hospital catchment area; and all of

Iceland. The number of children included from each country is shown in Table 2.

Inclusion criteria. Inclusion criteria were one or more of the following: (a) arthritis > 6 weeks; (b) inflammatory back pain and enthesitis; (c) EULAR criteria for systemic or possible systemic disease. Arthritis was defined as either a swollen joint or 2 of the following 3: (a) limitation of movement, (b) warmth, and (c) pain on passive or active movement. This means that joint pain itself is not enough for establishing the time of onset.

Time of onset. Time of onset is based on onset of arthritis (as defined above), on the first symptom of systemic or possible systemic disease, or on the time when a patient was diagnosed as having inflammatory back pain and enthesitis in combination with at least one more criterion for enthesitis related arthritis⁸.

Data collection. Twenty pediatricians with experience in pediatric rheumatology collected data from the incidence cases in their catchment areas. Letters were sent to all general practitioners, orthopedic surgeons, rheumatologists, and pediatricians in the area asking that children fulfilling the inclusion criteria for our study be referred to the local study center.

Children were examined as part of the study at 6 months of disease duration, with an acceptance of -1 to +2 months' deviation from the exact date. This time period was summarized by recording clinical findings, the results of antinuclear antibody (ANA) and rheumatoid factor (RF) analysis, heredity, pattern of affected joints at onset and during disease course, and EULAR group as determined by the local coordinator. Since several children were diagnosed late, ANA and RF results were accepted if analyzed during the first year (± 2 mo) of the disease.

Data were stored in a database, developed in 4D (ACI, Clichy, France). In Sweden the study coordinator visited the local study centers once every 6 months, to help collect data and to discuss patients for inclusion. All the Nordic country coordinators met regularly at least once every 6 months.

Classification. Each child was classified according to the EULAR criteria by the participating pediatrician at 6 months. The EULAR classification has long been the gold standard in Europe and it was natural for our participants to classify patients according to EULAR criteria themselves. The following categories were distinguished, according to our interpretation of the EULAR criteria: (a) systemic definite, with fever, rash, and arthritis; (b) systemic probable, where arthritis is absent but fever, rash, and 2 out of 3 of generalized lymph node enlargement, hepato- or splenomegaly, and serositis are fulfilled; (c) polyarthritis; and (d) oligoarthritis. Other forms include: (e) juvenile ankylosing spondylitis (JAS); (f) arthropathy associated with inflammatory bowel disease; (g) definite psoriatic arthritis (PsA); and (h) probable PsA. Since there currently is no commonly accepted classification of JAS⁹, we chose to define JAS either as arthritis with radiographic diagnosis of sacroiliitis or as a combination of arthritis, enthesitis, and clinical signs of sacroiliitis. For PsA the Vancouver criteria^{10,11} were used as a clarification in the way they are most commonly used and accepted today. According to those criteria, “definite juvenile psoriatic arthritis” was defined as arthritis beginning before age 16 and typical psoriasis or arthritis and 3 of: dactylitis, nail pitting, psoriasis-like rash, family history of psoriasis. “Probable juvenile psoriatic arthritis” was defined as arthritis and 2 of the minor criteria described above. In the original article the EULAR classification uses the term pauciarticular arthritis and not oligoarticular arthritis⁶. However, to avoid confusion from using both terms we chose to use only the term oligoarticular arthritis.

Classification of the patients according to the ILAR/Durban criteria⁸ was performed by the coordinator of the study, at 6 months duration or in some cases at one year (oligoarthritis, persistent or extended). In the ILAR classification system, strict exclusion criteria for each category are part of the classification. The number of specified exclusion criteria varies from one subgroup to another (Table 1). For the systemic category no exclusion list is given, although in clinical work this category is one of the exclusion criteria and they were discussed with participants in the study. The number of exclusion criteria fulfilled was studied for each child and each subgroup.

Table 1. ILAR criteria, definitions and exclusions.

SYSTEMIC ARTHRITIS

Definition. Arthritis with or preceded by daily fever of at least 2 weeks' duration, that is documented to be quotidian for at least 3 days and accompanied by one or more of the following:

- (1) Evanescent, non-fixed erythematous rash
- (2) Generalized lymph node enlargement
- (3) Hepatomegaly or splenomegaly
- (4) Serositis

Exclusions. Exclusions have not been listed.

OLIGOARTHRITIS

Definition. Arthritis affecting 1–4 joints during the first 6 months of disease. Two subcategories are recognized:

- (1) Persistent oligoarthritis: affects no more than 4 joints throughout the disease course
- (2) Extended oligoarthritis: affects a cumulative total of 5 joints or more after the first 6 months of disease

Exclusions.

- (1) Family history of psoriasis confirmed by a dermatologist in at least one first or second degree relative
- (2) Family history consistent with medically confirmed HLA-B27 associated disease in at least one first or second degree relative
- (3) Positive RF test
- (4) HLA-B27 positive male with onset of arthritis after 8 years of age
- (5) Presence of systemic arthritis as defined above

POLYARTHRITIS (RF negative)

Definition. Arthritis affecting 5 or more joints during the first 6 months of disease, associated with negative RF tests on 2 occasions at least 3 months apart.

Exclusions.

- (1) Presence of RF
- (2) Presence of systemic arthritis as defined above

POLYARTHRITIS (RF positive)

Definition. Arthritis affecting 5 or more joints during the first 6 months of disease, associated with positive RF tests on 2 occasions at least 3 months apart.

Exclusions.

- (1) Absence of positive tests for RF on 2 occasions at least 3 months apart
- (2) Presence of systemic arthritis as defined above

PSORIATIC ARTHRITIS

Definition.

- 1. Arthritis and psoriasis or
- 2. Arthritis and at least 2 of:
 - (a) Dactylitis
 - (b) Nailpitting or onycholysis
 - (c) Family history of psoriasis confirmed by dermatologist in at least one first degree relative.

Exclusions

- (1) Presence of rheumatoid factor
- (2) Presence of systemic arthritis as defined above

ENTHESITIS-RELATED ARTHRITIS

Definition.

Arthritis *and* enthesitis,

or

arthritis *or* enthesitis with at least 2 of:

- (a) Sacroiliac joint tenderness and/or inflammatory spinal pain
- (b) Presence of HLA-B27
- (c) Family history in at least one first or second degree relative of medically confirmed HLA-B27 associated disease
- (d) Anterior uveitis that is usually associated with pain, redness, or photophobia
- (e) Onset of arthritis in a boy after the age of 8 years

Exclusions.

- 1. Psoriasis confirmed by a dermatologist in at least one first or second degree relative
- 2. Presence of systemic arthritis as defined above.

OTHER ARTHRITIS

Definition.

Children with arthritis of unknown cause that persists for at least 6 weeks but that either:

- 1. Does not fulfil criteria for any of the other categories, or
- 2. Fulfills criteria for more than one of the other categories.

Exclusions. Patients who meet criteria for other categories.

Serology. For the ILAR criteria, neither the methods to be used for analysis of ANA and IgM-RF nor the cutoff point to define abnormal values are defined, but the definition of the local laboratory is used. In our study, each physician interpreted the results of ANA and IgM-RF analysis as normal or abnormal according to the reference values of the local laboratory. The number of IgM-RF analyzed in patients with polyarticular and oligoarticular arthritis were recorded, since the ILAR criteria require that 2 IgM-RF are analyzed at least 3 months apart to classify a child as RF positive or negative.

Informed consent was obtained from both parents and children. The Research Ethical Committees at each regional university gave their approval.

Statistical methods. The validity of diagnostic instruments involving observer based assessments refers to the quality of the diagnosis made by the instrument. A common approach to evaluating validity is to compare instruments intended to diagnose the same disorder. Our study concerns construct validity, which refers to how closely the 2 instruments, the ILAR and EULAR criteria, which have 2 different operational definitions of the same theoretical concept to be measured, are related¹². As the 2 instruments were developed from different perspectives, the evaluation will also concern the completeness of the instrument in the coverage of important areas. The comparisons of the criteria for diagnosis are described in the

tables. Median value and percentiles are used for description of the basic characteristics of the patients.

RESULTS

The study group consisted of 322 children, 203 (63%) girls and 119 (37%) boys. Table 2 shows the number of children included from each country. The median age at time of onset was 6.8 years (10–90 percentile, 1.6–13.5 yrs). A total of 321 patients qualified for classification according to the

Table 2. The number of children included from each country.

Country	No. of Children 0–15 years Included
Sweden	124
Finland	82
Norway	39
Denmark	70
Iceland	7
Total	322

ILAR criteria (Table 3), 290 of them had a disease duration ≥ 3 months and were also classified according to the EULAR criteria, after 6 months duration of the disease (Table 4). One child could be classified according to the EULAR criteria only.

One hundred sixty children fulfilled the ILAR criteria for oligoarticular arthritis, including the diagnosis of “oligo extended,” “oligo persistent,” and “oligoarticular arthritis not classified as persistent or extended,” as well as 9 patients with oligoarthritis, also fulfilling another category. In the EULAR classification 184 children were classified as oligoarticular. Twelve (12/321) children were classified as enthesitis related arthritis according to the ILAR criteria. In the EULAR classification 7/291 patients were considered to have JAS (Tables 3,4, 5).

Thirty-one of the 322 children (10%) had disease duration < 3 months and were classified according to the ILAR criteria only. The majority, 25/31 (81%), had oligoarthritis, 11 received steroid injection(s) associated with recovery. Nine of the 31 patients became symptomatic again during

the second 6 months; 6 of them had received steroid injection(s) during the first 6 months after onset.

In addition to the above categories in the ILAR classification, 2 new subgroups evolved in our study (Table 3): polyarthritis with only one or no RF analyzed during the first year after onset and oligoarthritis that could not be specified as persistent or extended because the children were included late or they dropped out from the followup (Table 3).

RF is an important discriminator in the ILAR criteria as it is an exclusion criterion for oligoarthritis, PsA, and for differentiation of polyarthritis. In 78 children classified as polyarticular by the ILAR criteria, 43/78 (55%) could not be defined as RF positive or negative, because RF was not analyzed twice. In 7 of those 43 patients RF was never analyzed and in the other 36, RF was analyzed only once. It should be noted (Table 3) that 11 children with polyarticular disease belong to the subgroup “fits more than one category.” For 160 patients classified as persistent, extended, or “oligoarticular arthritis not classifiable as persistent or extended,” 54/160 (34%) had RF analyzed twice.

Table 3. Results of ILAR classification in 321 patients and classification events in 20 patients fitting 2 categories.

ILAR Group	No. of Patients	Patients (20) Fitting 2 Categories. Distribution of the 40 Classification Events [†]
Systemic	13	0
Oligoarthritis, persistent	123	6
Oligoarthritis, extended	17	2
Polyarthritis, RF negative	22	7
Polyarthritis, RF positive	6	0
Psoriatic arthritis	10	2
Enthesitis related arthritis	12	18
Polyarthritis, not classified	39	4
Oligoarthritis, not classified	11	1
Fits no category	48	
Fits more than one category	20	
Total	321	40

[†] Some patients fit 2 categories in the ILAR criteria; each time they are classified, this is called a classification event.

Table 4. EULAR classification in 291 patients.

EULAR Group	No. of Patients
Systemic definite	11
Systemic probable	1
Juvenile ankylosing spondylitis	7
Arthritis associated with IBD	0
Juvenile PsA, definite	5
Juvenile PsA, probable	3
Oligoarticular	184
Polyarticular	76
Not classified	4
Total classified	291

PsA: psoriatic arthritis; IBD: inflammatory bowel disease.

Table 5. ILAR classification. Combinations of categories in 20 patients, fulfilling 2 categories each.

First Category	No. of Patients	Second Category	No. of Patients
Enthesitis related arthritis	18	Poly RF negative	6
		Poly not classified	4
		Oligo persistent	5
		Oligo extended	2
Psoriatic arthritis	2	Oligo not classified	1
		Poly RF negative	1
		Oligo extended	1

Poly: polyarthritis; RF: rheumatoid factor.

Of the 321 patients classified according to the ILAR criteria, 253 fulfilled the criteria for one category and 20 children satisfied 2 categories each, while 48 children did not fit into any category. Thus, 273 patients had 293 classification events. Of the 20 (6%) patients fulfilling the ILAR criteria for 2 categories, enthesitis related arthritis was classified in 18. For the remaining 2, PsA in combination with either polyarticular RF negative arthritis or oligoarticular extended arthritis was found (Table 5). No patient satisfied more than 2 categories. In Table 3 the 40 classification events for the 20 patients fulfilling 2 categories each are shown.

Forty-eight of the 321 (15%) patients did not fit any ILAR category (Table 6). All 48 patients initially fulfilled criteria for oligo persistent or oligo extended arthritis and 5/48 patients initially also fulfilled criteria for enthesitis related arthritis. Of 53 classification events, 40 had to be excluded because of psoriasis in a first or second grade relative, 8 because of one or 2 positive RF, and 5 because of HLA-B27 associated disease in a first grade relative.

Of the 273 patients who fulfilled one or 2 categories of the ILAR criteria (a total of 293 classification events), 280 classification events fell into subgroups with specified exclusion criteria. Table 7 presents the number of exclusion criteria for each category and the extent to which the information was complete for fulfilling them. Oligoarticular arthritis has 5 exclusion criteria and information to fulfill all of them was complete in only 45/160 (28.1%) of the patients.

DISCUSSION

Our study showed that the EULAR and ILAR criteria differ concerning the operational definitions of the variables involved, which complicates comparison between them. Therefore, the construct validity between the 2 classification systems is low. Another problem is that no gold standard is available against which to compare the different sets of criteria. We can only observe what happens when a population of children with chronic arthritis is classified according to one system or the other.

Arbitrary values, such as the number of affected joints, are difficult to avoid, but we also know that rheumatologists examine and grade joint inflammations differently¹³.

The EULAR criteria and the definition of each subgroup have kept their initial structure, but over time, through research and experience, the subgroups of PsA, for example, have become more detailed according to the Vancouver criteria^{10,11}. In spite of this change in the definition of PsA we still talk about it as part of the EULAR criteria. The EULAR criteria were (at least in parts) quite broadly defined from the beginning⁶ and therefore prone to change, such as the definition of PsA. In most studies an exact definition of the different subgroups/categories is seldom given. We cannot take for granted that the criteria used are exactly the same in different studies and the imprecise use of the EULAR and the ACR classifications is well known¹.

Onset type according to EULAR criteria is based on the first 6 months of the disease. The ILAR criteria are also based on this time period, but make exceptions for the oligo

Table 6. ILAR criteria. Analysis of the 48 patients excluded from 53 possible classification events and consequently not fitting into any category.

Possible Category	No. of Classification Events	Cause of Exclusion	No. of Classification Events
Oligo arthritis, extended or persistent	48	PsA, in 1st degree relative	15
		PsA, in 2nd degree relative	20
		One or two positive RF	8
		HLA-B27-associated disease, in 1st or 2nd degree relative	5
Enthesitis related arthritis	5	PsA, in 1st degree relative	3
		PsA, in 2nd degree relative	2

Table 7. ILAR criteria. Analysis of 280 classification events in the categories with specified exclusion criteria.

ILAR Category	No. of Exclusion Criteria	No. of Classification Events	No. (%) of Classification Events with Fulfilled Exclusion Criteria [†]
Oligoarticular	5	160	45 (45/160, 28%)
Polyarticular	2	78	36 (36/78, 46%)
Psoriatic arthritis	2	12	3 (3/12, 25%)
Enthesitis-related arthritis	2	30	26 (26/30, 87%)
Total		280	110 (110/280, 39%)

[†] Values in parentheses are percentages of fulfilled exclusion criteria of all classification events in the category.

extended and oligo persistent forms. In this study it was clear that the laboratory analysis necessary to classify according to ILAR criteria was not usually done during the first half-year, partly because patients were not always examined within the first 6 months of the disease. This observation shows that the arbitrary limit of 6 months for time of onset is not always feasible.

The ILAR criteria are the preliminary report from a board of experts in pediatric rheumatology (ILAR) who strived to find more homogeneous subgroups of children with chronic arthritis^{7,8}. The criteria evolved from the clinical experience of the group and scientific studies on the natural history of the disease, as well as from immunogenetic and immunopathological considerations.

The ILAR criteria obviously diagnose more children with chronic arthritis than the EULAR criteria, as the duration of arthritis needs only to be 6 weeks compared with 3 months for the EULAR criteria. In our prospective study with a longitudinal design, the 31 patients with short duration, who thus fulfill only the ILAR classification, are of special interest. Nine of them became symptomatic again during the second half-year after onset. Thus, by using the ILAR and not the EULAR criteria, the number of cases with juvenile arthritis increased by 10% (31/322), considering the first half-year after onset. This results in new epidemiological data and may have other consequences, such as an increased number of children with problems in being covered by health insurances.

Since the ACR criteria are also widely used, especially in the Americas, it would be of great interest to study the consequences of using the ACR and ILAR criteria in the same prospectively collected incidence based population.

RF is highly discriminating for 3 of the ILAR categories, and we feel there is a need to further study the role of RF in childhood arthritis. RF positive oligoarthritis, for example, is encountered only occasionally in large populations and is a subject of case reports^{5,14}. It is not known whether this is a homogenous group of patients. The low numbers of RF analyses in our study were explained by some of the participating physicians as depending on their experience that seropositive arthritis is mainly a disease in teenagers and that RF analysis in small children is more a waste of money than a diagnostic tool. The common opinion among the participants was that if one RF test was negative, the patient was considered RF negative. The study would have been much improved with RF analyzed to a fuller extent since RF can have serious prognostic implications. ANA positivity, which we clinically consider seriously and which we already know predicts for uveitis¹⁵, on the other hand, is not part of the ILAR criteria.

“Hereditiy” is another variable that is highly discriminating in the ILAR criteria. Fifteen percent of the patients did not fulfill the criteria for any category in our study. In the majority of these patients PsA in a first or second degree

relative was the cause. This is in accord with findings from other studies^{16,17}. Heredity for psoriasis varies between populations¹⁸, and this influences the result of ILAR classification. The importance of correctly assessing heredity raises the question of how reliable the subgroup classification according to ILAR will be¹⁹. Assessing heredity assumes that the information given is reliable and that the information from the adult rheumatologists, dermatologists, and gastroenterologists has been correctly understood. Some of the diagnoses used in heredity, although clearly defined, may not be used uniformly among adult rheumatologists. Ankylosing spondylitis is one such “difficult” diagnosis. Another is the term “HLA-B27-associated disease,” an umbrella term for 3 diagnoses, where the detailed definition of each diagnosis and to what extent HLA-B27 is truly involved remains to be determined.

In the process of finding important discriminators for classification of juvenile arthritis a research group from the UK presented results from subtyping juvenile idiopathic arthritis. They used a statistical method that explains observed relationships between clinical and laboratory variables. The method implies probabilities of different variables to be statistically independent. Clinical features like pattern of joint involvement and ANA positivity seemed more important in subtyping than findings such as fever, spinal pain, psoriasis, and RF²⁰.

One goal of the ILAR criteria is to keep the subgroups mutually exclusive. How this should be done is not defined, since the exclusion criteria do not mutually exclude each other except for some categories. Six percent of the patients were eligible for classification in 2 groups. Enthesitis related arthritis could be classified in the oligo persistent group, oligo extended group, or polyarticular RF negative group, since the combination of arthritis and enthesitis avoids all the exclusion criteria for oligoarthritis, and the polyarticular group does not have other exclusion criteria except positive RF or systemic arthritis. PsA could as well be classified as polyarticular RF negative arthritis, since the only exclusion criterion for this polyarticular group is RF positivity or systemic arthritis. PsA in combination with oligoarthritis is also possible, since heredity for psoriasis is not necessary for diagnosis of PsA.

To correctly classify a child with chronic arthritis according to the ILAR criteria, many variables must be registered. Our study shows that although this work was done by experienced colleagues, quite a few variables were missed. For the polyarticular group 2 RF tests, necessary for correct diagnosis, were analyzed in less than 50% of the patients, and for the oligoarticular type only 34% had RF analyzed twice. This indicates low feasibility for the ILAR criteria when used in clinical work. If these criteria are to be properly used and bring clarification, we should also consider all exclusion criteria for each classification event. For clinical practice it is also disturbing that 6% of the chil-

dren fulfilled more than one category. This makes it difficult to correctly assess to which category the patient belongs.

The ILAR criteria are already being used without having been validated. This was not the initial intention^{7,8}; moreover, neither the ACR nor the EULAR criteria have been prospectively validated.

The ILAR criteria raise new important questions, such as how to continue the work to find well functioning classification criteria. The EULAR and the ACR criteria are important since they have been used for a long time and we have learned a great deal from them. Continued analysis of prospective, longitudinal, population based studies of children with chronic arthritis is essential to improving classification and to obtaining a better idea of the extent to which clinical findings, ANA, RF, or heredity is important in evaluation of outcome, prognosis, and choice of medication. Ideally, classification criteria for patients with juvenile arthritis should be acceptable in clinical work as well as for scientific purposes. Future studies should aim to find groups as clinically homogenous as possible, but this will only be possible if we are stricter in our definitions, whatever the classification system used.

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