Incidence of Juvenile Idiopathic Arthritis in the Nordic Countries. A Population Based Study with Special Reference to the Validity of the ILAR and EULAR Criteria

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ABSTRACT. ***Objective.*** To find the incidence of juvenile arthritis according to the ILAR and EULAR criteria within defined areas in the Nordic countries, and to study the validity of the ILAR and EULAR criteria from this perspective.

Method. A longitudinal, prospective, population based study with patients enrolled according to the ILAR and EULAR criteria. Twenty doctors in Iceland, Norway, Sweden, Denmark, and Finland collected data from the incidence cases within their catchment areas over a period of 1.5 years, beginning July 1, 1997. Clinical and serological data from the first year of the disease were collected.

Results. In the whole group of 315 patients, the incidence rate was 15 per 100,000 children/year (95% CI 13–17) according to the ILAR criteria, varying from 7 (1–13) in Iceland, 19 (7–31) and 23 (10–36) from 2 different regions in Norway, and 9 (5–12) and 16 (9–23) from 2 different areas in Denmark, to 15 (12–18) in Sweden and 21/100,000/year (15–26) in the Helsinki region in Finland. An early peak in distribution for age of onset was found in girls but not in boys. The number of antinuclear antibody (ANA) positive children in the whole group, made up of children who had undergone at least one analyzed ANA test, was 123/315 (39%). Girls were ANA positive in 83/197 (42%) and boys in 40/118 (34%). Uveitis developed in 27/315 (8.6%) children during the first 6 months of the disease.

Conclusion. Incidence rates of juvenile arthritis for areas within the Nordic countries were in accord with previous data. The ILAR criteria present slightly higher incidence rates, with a shorter disease duration for inclusion, compared to the EULAR criteria. Patients in one subgroup in either of the criteria sets do not necessarily belong to the expected subgroup in the other set of criteria; e.g., for juvenile ankylosing spondylitis (EULAR) and enthesitis related arthritis (ILAR). Our epidemiological findings are a reminder to be aware of possible new subgroups in children with juvenile arthritis. (J Rheumatol 2003;30:2275–82)

Key Indexing Terms:
JUVENILE RHEUMATOID ARTHRITIS EPIDEMIOLOGY CHILD INCIDENCE

Epidemiological studies of arthritis in childhood are important in describing the natural history and outcome in different disease entities and for the identification of possible etiologic factors. Differences in the occurrence of juvenile arthritis between regions and countries could generate a hypothesis regarding environmental and genetic factors that may influence the disease. Epidemiological studies may also predict early prognostic factors and may be

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valuable for evaluation of classification criteria[1]. Understanding how different subgroups of disease present in the population is vital for research and clinical work. Methods for patient retrieval, definition of the catchment population, and choice of classification criteria are of fundamental importance to the results. Incidence can be measured in different ways, but to get as close to the true incidence as possible a population based approach is necessary. A prospective setting raises the awareness of a certain condition and more complete patient retrieval can be accomplished according to the criteria chosen. In the Nordic countries, where health care systems are socialized and fairly homogenous, epidemiological studies are facilitated.

One of the primary purposes of the new classification criteria (International League Against Rheumatism, ILAR) for juvenile idiopathic arthritis (JIA) is to provide an organizational framework that facilitates communication among physicians and scientists[2,3]. The classification is based on clinical characteristics present during the first 6 months of the disease rather than on a laboratory based rationale, and is more descriptive than those used previously [i.e., the American College of Rheumatology[4,5] and European League Against Rheumatism (EULAR)[6] criteria]. The new classification criteria still require validation against the classifications already in use[7]. This prospective study presents the incidence of childhood arthritis over an 18 month period in well defined areas in the Nordic countries. It also gave us the opportunity to study the influence of the 2 sets of classification criteria applied to the patient population. This type of study is a prerequisite for "testing the proposed ILAR criteria, under appropriate epidemiological and statistical direction, against classifications already in use," as stated in the proposal for the ILAR criteria[8].

MATERIALS AND METHODS

Study design. A longitudinal, prospective, population based incidence study with patients diagnosed according to the ILAR and EULAR criteria. The health care systems in the Nordic countries include regular checkups at a child health center and later at school health care centers, all free of charge. A child with a long-standing complaint will probably be found and referred to a pediatrician. Pediatricians working locally and at the hospitals belong to the same organization, which facilitates cooperation.

Study period. The inclusion period was from July 1, 1997, to December 31, 1998. Data collection for that period continued until May 2000 to avoid missing cases that were referred late.

Inclusion criteria. Arthritis > 6 weeks, inflammatory back pain and enthesitis, or EULAR criteria for systemic or possible systemic disease as well as ILAR criteria for systemic arthritis. Arthritis was defined as either a swollen joint or 2 of the following 3: (1) limitation of movement, (2) warmth, and (3) pain on passive or active movement.

Time of onset. Defined as onset of arthritis (as above), the first symptom of systemic or possible systemic disease, or the time when a patient was diagnosed as having inflammatory back pain and enthesitis in combination with at least one other criterion for enthesitis related arthritis. Consequently, joint pain was not enough to establish the time of onset.

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 (except for the eastern part of Denmark), orthopedic surgeons, rheumatologists, and pediatricians in the area, asking that children who satisfied the inclusion criteria for our study be referred to the local study center. No patient refused to participate in the study.

Study population. The catchment area and population were well defined in each country (Figure 1). In Sweden, the catchment area consisted of 5 counties and one health district and 6 pediatric departments were thus involved. The eastern part of Denmark, comprising the islands of Sjaelland, Bornholm, Mön, and Lolland-Falster, but not the county of Copenhagen, was included. Seven pediatric departments were involved in this area. The main hospital in Copenhagen received patients from general practitioners, the local pediatric departments in the area, or any of the other pediatric departments mentioned. The county of Århus in Denmark was also part of the study. In this area, the pediatric department in Skejby Hospital received patients from general practitioners and from one local pediatric department in the county. In Norway, 2 counties in the Tromsø region (Troms and Finnmark) were included and one in the Trondheim region (Sør-Trøndelag). The Tromsø region has 2 pediatric departments and the Trondheim region one. Within these areas general practitioners and pediatricians had referred all children with arthritis to the 2 clinics. In Finland, the Helsinki University Central Hospital catchment area was included, which involves 4 pediatric departments besides the one at the University Central Hospital. Within this area, primary care doctors and pediatricians have a tradition of referring patients to the specialist clinic at Helsinki University Central Hospital. The whole of Iceland was also part of the catchment area. In the Icelandic health care system children are mainly taken care of by pediatricians and general practitioners and there are 2 pediatric departments, one in Reykjavik and one in Akureyri.

Classification. The term "juvenile arthritis" was used as a general description of cases, irrespective of what other term would fit for that patient ("juvenile chronic arthritis" in the EULAR criteria and/or "juvenile idiopathic arthritis" in the ILAR criteria). Each child was classified according to the EULAR criteria by the participating pediatrician at 6 months. The following categories were distinguished according to our interpretation of the EULAR criteria: (1) systemic definite, with fever, rash, and arthritis; (2)

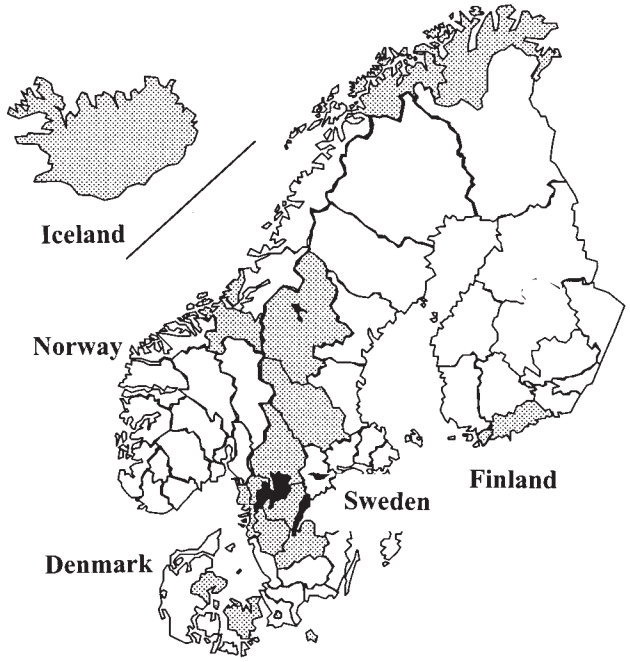

Figure 1. The Nordic countries and the geographical areas participating in the study (shaded).

systemic probable, where arthritis is absent but fever, rash, and 2 out of 3 of generalized lymph node enlargement, hepato- or splenomegaly, and serositis must be fulfilled; (3) polyarthritis; and (4) oligoarthritis. "Other forms" included (5) juvenile ankylosing spondylitis (JAS); (6) arthropathy associated with inflammatory bowel disease (IBD); (7) psoriatic arthritis (PsA) definite; and (8) PsA probable. JAS was defined either as arthritis with radiological diagnosis of sacroiliitis or as a combination of clinical signs of sacroiliitis, arthritis, and enthesitis. For PsA, the Vancouver criteria[9] were used as clarification in the way they are most commonly used and accepted today.

Classification of patients according to the ILAR /Durban criteria[3] was performed by the coordinator of the study, at 6 months' disease duration and with acceptance of –1 to +2 months' deviation from the exact date. Patients with oligoarthritis were classified at one year disease duration as oligoarthritis, persistent or oligoarthritis, extended.

Uveitis was studied only for the first 6 months of disease.

Serology. To define a patient as antinuclear antibody (ANA) or rheumatoid factor (RF) positive according to the ILAR classification system, 2 ANA or RF tests must be positive, with at least 3 months in between, during the first 6 months of the disease[3]. For reasons stated in an earlier article[10], we have included all serologies analyzed during the first year after onset for this subdivision of patients. In a few patients 4 ANA tests were analyzed during the first year of disease. In those cases, 2 consecutively positive ANA tests were regarded as positive in our opinion, regardless of their order in relation to the other analyses. One positive and one negative test or more was regarded as negative. If the only test examined was positive for ANA or RF, the result in the analyses was regarded as "missing" in the ILAR system.

Since traditionally we did not stress a necessity to have 2 ANA or RF analyzed before the ILAR classification was introduced, we also summarized how many patients had only one positive ANA or RF test.

In our study, each physician interpreted the results of ANA and IgM RF analysis as normal or abnormal according to the reference values of their local laboratory.

The Swedish laboratories in our study measured ANA in a uniform way using immunofluorescence (IF) on HEp-2 cells as the main method. At the other laboratories, IF on HEp-2 cells was used at all times, except the one in Tromsø, where they used an ELISA method (Pharmacia, Sweden), and Iceland, where they used IF on rat kidney cells. RF was determined by latex agglutination test at all the laboratories, except 2 that used an ELISA method.

Informed consent/assent was obtained from both parents and children. The Research Ethics Committee at each regional university gave their approval for the study.

Statistical methods. Median value and quartiles were used for a description of the basic characteristics of the patients. Ninety-five percent confidence intervals (CI) for incidence were calculated based on the Poisson distribution. An estimation of population at risk was calculated as follows: the mean value of the juvenile population (< 16 yrs) at the beginning of 1997 and 1998 was calculated. The mean value of this figure and the population at the beginning of 1999 then constituted the estimated population at risk. Incidence rates from each area were based on the number of children included during the 18 month period, divided by the estimated population at risk, divided by 1.5. Histograms and cumulative plots were used for a presentation of groups of patients. The chi-square test was used when comparing fractions, and the limit of significance was set at 0.05. Demographic statistics from 1997 to 1999 were obtained from Statistics Sweden, Norway, Denmark, Finland, and Iceland.

RESULTS

The study group consisted of 315 children, 197 (62.5%) girls and 118 (37.5%) boys. The numbers of children aged 0–15 years were: Sweden 123, Finland 82, Norway 33, Denmark 70, and Iceland 7. The median age at the time of onset was 6.8 years (25%, 75% quartiles, 2.8, 10.8 yrs). A total of 314 patients qualified for classification according to the ILAR criteria, 284 of whom had a disease duration ≥ 3 months, and were also classified according to the EULAR criteria after 6 months' duration of disease. One child could be classified according to the EULAR criteria only and a total of 315 patients were thus included in the study.

The number of children included from each catchment area, the population at risk, and the incidence rate for that area are presented in Table 1. Annual incidence rates according to the ILAR criteria varied between 7 patients/100,000 in Iceland and 23/100,000 in the Trondheim region of Norway. According to the EULAR classification, annual incidence rates were slightly lower, and varied between 7 and 21 patients/100,000. Even though the number of patients was slightly higher using the ILAR criteria, confidence limits overlapped.

The proportion of patients with oligoarticular disease from each catchment area varied from 45% to 76% (Table 1).

The distribution of the 197 girls according to age at the time of onset is shown in Figure 2A and for the boys in Figure 2B. An early peak in the distribution for the age of onset of juvenile arthritis in girls was found (Figure 2A). In boys the incidence of juvenile arthritis decreased slowly over time after one year of age (Figure 2B). No obvious peak in the incidence rate for boys was found. The peak incidence for girls corresponded to a high incidence of patients with oligoarticular as well as polyarticular disease.

In the group of patients for whom at least 2 ANA were analyzed (n = 197), 66 were positive and 131 negative. Data were missing for 118 patients. If only one positive ANA was accepted as "ANA positivity," 123 patients were regarded as ANA positive and 185 ANA negative, and data were missing for 7 patients. The groups of 83 girls and 40 boys with at least one positive ANA are presented by age at onset in Figure 3. The early peak of incidence in girls (< 3 years of age at onset) corresponded to an over-representation of ANA positive girls ($p = 0.0001$) compared to the rest of the girls.

In total, 27/315 (8.6%) children developed uveitis during the first 6 month period of the disease. The majority, 17/27 (63%), had at least one positive ANA during the first year of the disease, 12/27 (44%) had 2 positive ANA tests during that period, and 5 patients were HLA-B27 positive. The median age of onset of arthritis disease in the uveitis group was 3.9 years (25%, 75% quartiles, 2.3, 5.2 years) and 17/27 (63%) were girls.

Nine patients (3% of the total incidence group) were RF positive in 2 tests at least 3 months apart during the first year of the disease. The median age of this group of children was 13.2 years (range 9.1–14.7), and the majority were girls (8/9, 89%). Six patients belonged to the polyarticular RF+ subgroup and 3 belonged to the subgroup "other arthritis 1." The latter 3 patients all had oligoarticular persistent disease,

Table 1. The population at risk, number of patients studied, number of patients with oligoarticular disease, and the incidence rate from each catchment area.

	Population At Risk	No. of Patients, ILAR Criteria	No. of Patients, EULAR Criteria	Patients with Oligoarticular Disease n (%), EULAR Criteria	Incidence 100,000/yr (95% CI), ILAR Criteria	Incidence 100,000/yr (95% CI), EULAR Criteria
Sweden	536,457	122	109	72 (66)	15 (12–18)	14 (10–17)
Finland, Helsinki	264,134	82	71	54 (76)	21 (15–26)	18 (13–23)
Denmark, East	310,669	41	40	18 (45)	9 (5–12)	9 (5–12)
Denmark, Århus	123,218	29	28	20 (71)	16 (9–23)	15 (8–22)
Norway, Trondheim	54,788	19	17	12 (71)	23 (10–36)	21 (9–33)
Norway, Tromsø	49,060	14	13	7 (54)	19 (7–31)	18 (6–29)
Iceland	68,989	7	7	4 (57)	7 (1–13)	7 (1–13)
Total	1,407,315	314	285	187 (66)	15 (13–17)	14 (12–15)

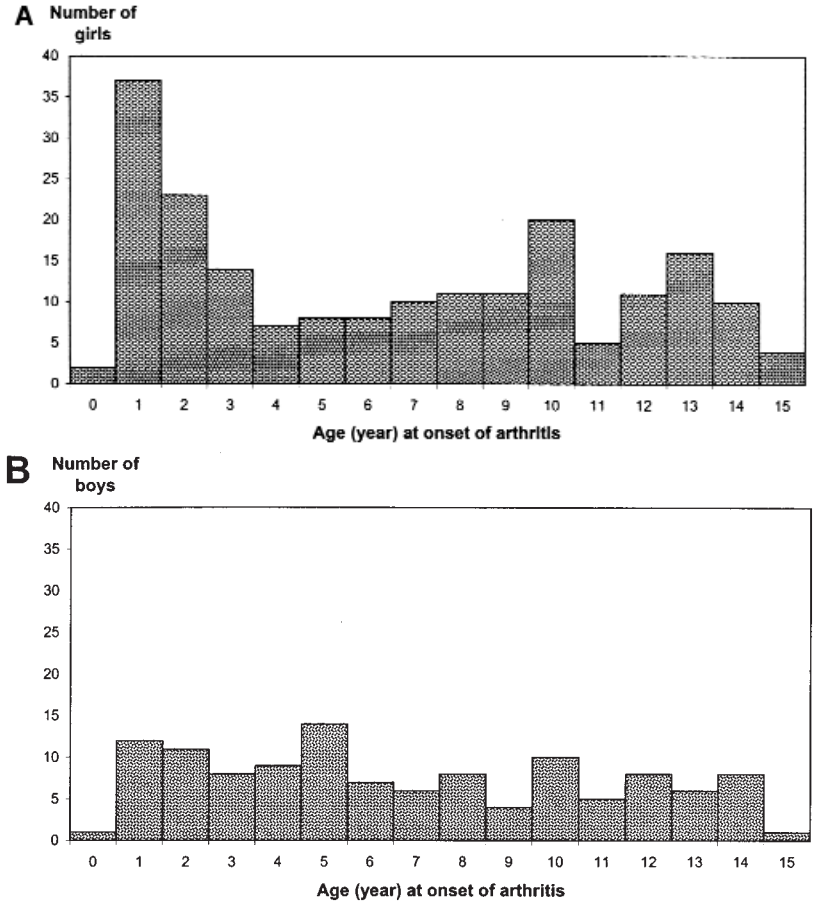

Figure 2. A. Age distribution in girls at time of onset of arthritis (n = 197). B. Age distribution in boys at time of onset of arthritis (n = 118).

but according to the ILAR criteria were excluded because of 2 positive RF tests.

Table 2 gives the number of patients and the proportion in each subgroup according to the ILAR as well as the EULAR criteria. Incidence rates for all subgroups according to the EULAR criteria are also presented. According to the ILAR classification, 12 patients were classified as enthesitis related arthritis. The corresponding EULAR subgroup for the 12 patients with enthesitis related arthritis was classified as JAS in 2 cases and oligoarticular disease in 9 patients, and one patient did not have a disease duration long enough to be classified according to the EULAR criteria, i.e. < 3 months. The 7 patients with JAS consequently corresponded to 2 patients with enthesitis related arthritis in the ILAR criteria and 5 with "other arthritis 2." The latter 5 patients all satisfied the criteria for enthesitis related arthritis as well as

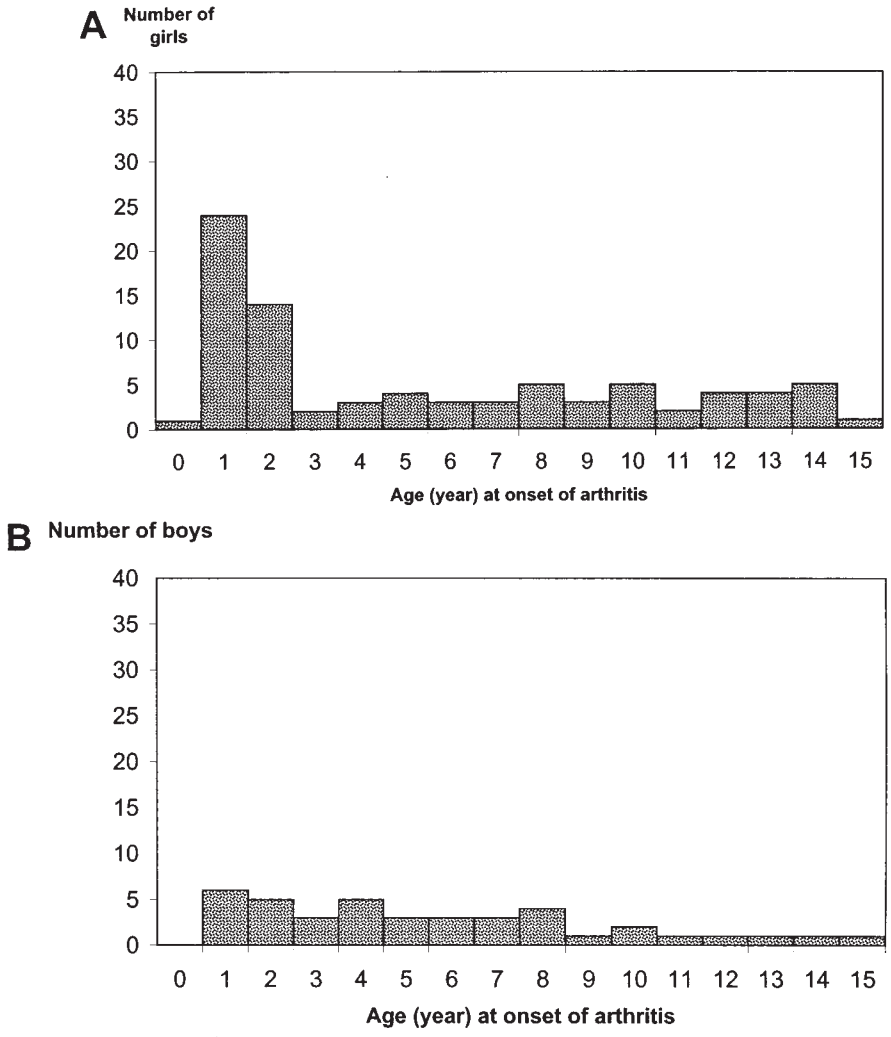

Figure 3. A. Age at onset of arthritis in 83 girls with at least one positive ANA during the first year of disease. B. Age at onset of arthritis in 40 boys with at least one positive ANA during the first year of disease.

the criteria for either RF negative polyarticular disease (n = 2), polyarticular not classified (2), or oligoarticular persistent disease (1).

DISCUSSION

The incidence rates in our study are in accord with earlier studies from the Nordic countries[11-15], except for Iceland, where previously no incidence data for juvenile arthritis existed (Table 1).

It is not surprising that we found a tendency to higher incidence rates with the ILAR criteria even though there was no statistically significant difference. We know from earlier studies that if a shorter disease duration is required for diagnosis, such as in American Rheumatism Association (ARA) criteria, this results in higher incidence rates compared to studies calling for a long disease duration[16]. The incidence rate from Sweden [14 (95% CI 10–17) using the EULAR criteria] is close to 10.9 per 100,000/year found in a prospective population based Swedish survey in 1984-88[12]. Our incidence figure from the Helsinki area of 18 per 100,000 (EULAR criteria) and 21 (ILAR criteria) is also close to an incidence rate of 19.6/100,000 in a prospective population based study using the ARA criteria[11]. A more recent retrospective study, using the ARA criteria in a large central district of Finland, presented an incidence rate of 19.5 per 100,000[13]. In Northern Norway a combined retrospective and prospective study over 15 years using the EULAR criteria gave incidence rates as high as 22.1–22.6[14,15]. The Tromsø population in our study covers the same study population as in that study[15]. During the short period of time for inclusion in our study (18 months) there was a tendency toward lower incidence figures

Table 2. Classification according to ILAR and EULAR criteria in 315 patients from catchment areas in 5 Nordic countries. The number (n) and proportion (%) of patients in each subgroup, as well as the incidence in each subgroup (the EULAR classification), are specified.

ILAR Subgroups, n = 314 No. of Patients (%)		EULAR Subgroups, n = 285 No. of Patients (%)		Incidence 100,000/yr (95% CI)
Systemic	13 (4)	Systemic definite (n = 11) and probable (n = 1)	12 (4)	0.6 (0.2–1.0)
Polyarticular RF +	6 (2)	Polyarticular	66 (23)	3.1 (2.2–4.0)
Polyarticular RF –	22 (7)			
Polyarticular not classified* (RF + or RF –)	38 (12)			
Oligoarthritis persistent	118 (38)	Oligoarticular	187 (66)	8.8 (7.3–10.4)
Oligoarthritis extended	17 (5)			
Oligoarticular not classified** (extended or persistent)	10 (3)			
Juvenile psoriatic arthritis	9 (3)	Juvenile psoriatic arthritis, definite (n = 5) and probable (n = 3)	8 (3)	0.4 (0.05–0.7)
Enthesitis related arthritis	12 (4)	JAS	7 (2)	0.3 (0.03–0.63)
		Arthritis associated with IBD	0	
Other arthritis 1†	49 (16)	Not classified	5 (2)	
Other arthritis 2††	20 (6)			

* Patients with polyarticular disease, not possible to classify as RF positive or negative because of missing data. ** Patients with oligoarticular disease, not possible to classify as extended or persistent as they dropped out of the followup after 6 months of the disease. † Patients who did not fit into any category. †† Patients who fit into more than one category.

(18/100,000). This is an example of how incidence rates for an uncommon disease and a relatively small population at risk need to be measured over a long time period to gain accuracy. The method employed for patient retrieval can strongly influence the results. Sullivan, *et al* experienced in their study that children referred from far away were sicker compared to children coming from areas closer to the referral center[17]. Population based studies generally present higher figures compared to those from referral centers[1]. Although we have not used different case ascertainment we have strived to get as close as possible to a population based approach. One theory is that a low incidence rate and a low percentage of oligoarticular patients indicate referral bias, since patients with milder localized disease might be missed more easily[1]. The 2 Danish catchment areas show a tendency toward a difference in incidence, even though it is not statistically significant, 9 patients per 100,000/year in the eastern area compared to 15 in the Århus area. With allowance for the short time for patient retrieval, the combination of a lower incidence rate and a lower proportion of oligoarticular disease in the eastern area compared to the Århus area raises the question whether the health care system is organized in such a way that cases of uncomplicated oligoarticular disease are identified and referred. Another source of error could be that information letters about the study were not sent to the general practitioners in the area. There is one retrospective study from Denmark in which cases were retrieved from general pediatric clinics. The incidence rate found was 6–8 cases per 100,000/year[18]. The Norwegian catchment areas (Tromsø, Trondheim) and also Iceland generated so few patients that they were not eligible for further analysis. However, the high proportion of oligoarticular patients indicates no referral bias of importance.

The question whether there is a north-south gradient in the incidence of juvenile arthritis in Europe has been raised[1]. The impression is that the most northerly countries have the highest incidence rates, although differences within the Nordic countries are not significant, neither for statistical nor methodological reasons. Patient retrieval methods have varied greatly between studies, although studies from other European countries show lower incidence rates compared to the Nordic countries[19,20] or rates similar to the lowest rates in our study[21,22]. Even if the short patient retrieval time has a strong influence on the results in areas of small population at risk, the highest incidence rates were found in the 2 areas in Norway and in the Helsinki area in this study and in others[11,13-15].

Two earlier Nordic studies have used a population based, prospective approach, one from Sweden[12] and one from Finland[11]. The pattern of age at onset in the Swedish study is quite similar to ours, with an early peak of onset for girls

but not for boys. In that study the early peak in girls was part of a bimodal distribution for time of onset. A bimodal distribution of time of onset was also found by others[16]. In our study we could not make an assumption of a bimodal pattern for girls or boys.

In the EULAR classification, the proportion of 4% systemic disease, 23% polyarticular disease, and 3% juvenile PsA is in agreement with the 7%, 25%, and 3%, respectively, found by Andersson Gäre and Fasth[12]. In their study, the mono- and oligoarticular patients constituted 61% of patients at onset, and we found 66%, using the same classification criteria. The EULAR classification in our study facilitates comparison with earlier studies. This was one reason we have presented incidence rates for each subgroup in the EULAR and not ILAR classification. Another reason was the complexity of the ILAR criteria. The figure for the incidence rate and the figure for the proportion of children in each subgroup are sometimes combined, which complicates any comparison between studies.

Little is known about the specificity of ANA in juvenile arthritis[23]. The clinical importance of ANA in predicting chronic anterior uveitis is well known[24]. Figure 3A shows the time of onset of arthritis in the 83 girls with at least one positive ANA. ANA positivity in girls was over-represented in the early peak of incidence mentioned above ($p = 0.0001$) (Figure 2A). This reinforces the impression that young ANA positive girls constitute a more biologically and immunologically homogenous group compared to those with a later onset. Girls were ANA positive in 42% of the patients and boys in 34%. The 39% ANA positivity in the whole group is in accord with results from other epidemiological studies in the Nordic countries (25–41%)[11,12,14]. The 34% ANA positivity in boys seems to be higher than reported earlier (12.5–25%)[12,14]. The majority of ANA positive boys had onset of arthritis before age 8 (Figure 3B) and 60% had oligoarticular onset according to the EULAR criteria. One explanation for high ANA positivity in boys could be that we collected a high proportion of patients with an oligoarticular disease and that the level of awareness of the disease occurring in boys was also high.

Around 9% of children with arthritis developed uveitis during the first 6 months of disease. The majority of them had at least one positive ANA during the first year of disease, while 44% had 2 positive ANA tests during that time. Children with uveitis in our study were young, with a median age at onset of arthritis of less than 4 years. Earlier studies have stated chronic uveitis to be particularly common in ANA positive girls with early onset of arthritis (< 4 yrs)[25]. In the same review the proportion of girls with uveitis varied between 56% and 92%. In our study, the sex distribution in children with uveitis was similar to that of the whole group with juvenile arthritis (63% girls). The result is in accord with a recent population based prospective survey of uveitis in juvenile arthritis[26]. This raises the question whether the previously stated increased risk for girls is due only to over-representation of girls. A longer followup of our patients is necessary to reveal the true incidence of uveitis during the disease course.

Since the group of RF positive patients is small in general, we chose to concentrate on those patients with 2 analyses at least 3 months apart, as suggested in the ILAR criteria. The group of 9 RF positive patients according to the ILAR criteria is strikingly homogenous, consisting mainly of girls with a late age of onset of arthritis. Six patients belonged to the polyarticular RF positive subgroup. The other 3 RF positive patients clinically had oligoarticular persistent disease. Due to their RF positive status they were excluded from the oligoarticular group and put into the "other arthritis 1" group. A positive RF (especially a high titer) is a major predictor of erosions in adults with RA[27]. It is well known that patients with juvenile oligoarticular disease are rarely RF positive. There have been descriptions of such patients having early erosive disease[28]. Perhaps RF positivity should be further considered in the classification of juvenile arthritis patients and not just in those with polyarticular disease. Further followup of the oligoarticular, RF positive patients in our study would be of particular interest.

The ILAR criteria set is a valuable tool for further classification studies. One problem is the mixture of onset and course type. The criteria are based on onset type, with the first 6 months as a basis, but at the same time they consider the course of the disease over an unknown time period thereafter to make a further distinction of patients with oligoarticular disease.

It must be emphasized that patients belonging to one subgroup in the ILAR classification sets do not necessarily belong to the expected subgroup in the EULAR criteria. For example, 64 of the 187 patients in the large group of patients with oligoarticular disease in the EULAR classification belong to the subgroups "other arthritis 1" (45 patients), "other arthritis 2" (6 patients), enthesitis related arthritis (9 patients), and juvenile PsA (4 patients) in the ILAR classification. Only 2 of the patients classified as having JAS by the EULAR criteria can be classified as enthesitis related arthritis according to the ILAR criteria. The other 5 belong to "other arthritis 2," where enthesitis related arthritis is combined with a polyarticular disease (RF negative or not classified) in 4 patients and with oligoarticular disease in one. The place of juvenile onset spondyloarthropathies in the newly proposed ILAR criteria needs further evaluation. Enthesitis as a clinical finding has a prominent role in the subgroup of enthesitis related arthritis in the ILAR criteria[3], which is supported by others[29].

The clinical presentation of the patient is the basis for whatever classification system we use, although there has been discussion about including immunological markers more in the ILAR criteria in order to explore homogeneity

within groups. Clinical homogeneity hopefully indicates, at least to some extent, etiologic and pathogenic homogeneity[7]. Incidence studies such as ours create valuable tools for further analysis of juvenile arthritis. The predictive validity of the ILAR criteria in a longitudinal followup will be of great interest.

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