Patterns of Joint Involvement at Onset Differentiate Oligoarticular Juvenile Psoriatic Arthritis from Pauciarticular Juvenile Rheumatoid Arthritis

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ABSTRACT. ***Objective.*** To compare the patterns of joint involvement of patients with oligoarticular onset juvenile psoriatic arthritis (Oligo-JPsA) and pauciarticular onset juvenile rheumatoid arthritis (Pauci-JRA) in order to estimate the predictive performance of specific patterns for the diagnosis of Oligo-JPsA.

Methods. Twenty-three children who fulfilled the diagnostic criteria for JPsA (Vancouver criteria) and who had fewer than 5 joints involved in the first 6 months of disease (Oligo-JPsA), and 64 children with Pauci-JRA (ACR criteria) were enrolled. Patients were also classified with respect to the ILAR criteria for juvenile idiopathic arthritis (JIA). Patient characteristics and clinical features at onset and during followup were determined. Patterns of joint involvement at onset of disease and their ability to differentiate between Oligo-JPsA and Pauci-JRA/Oligo-JIA were evaluated.

Results. Small joint disease (defined as involvement of any of the metatarsophalangeal or proximal or distal interphalangeal joints of the foot, or metacarpophalangeal or proximal or distal interphalangeal joints of the hand) was significantly more frequent in Oligo-JPsA than in Pauci-JRA at disease onset. The odds of patients with Oligo-JPsA having small joint disease or wrist disease within 6 months of disease onset were much higher than those with Pauci-JRA or Oligo-JIA ($p < 0.05$ or 0.001).

Conclusion. Small joint disease and wrist disease are suggestive of Oligo-JPsA. The use of a criterion consisting of small joint disease and/or wrist disease and/or dactylitis instead of dactylitis alone may increase the ability to differentiate Oligo-JPsA from Pauci-JRA or Oligo-JIA. (J Rheumatol 2002;29:1531–5)

Key Indexing Terms:
JUVENILE PSORIATIC ARTHRITIS CLASSIFICATION OF JUVENILE ARTHRITIS

Juvenile psoriatic arthritis (JPsA) has been characterized as an inflammatory arthritis occurring before the age of 16 years in association with the characteristic rash of psoriasis[1]. Because psoriasis may not appear until many years after the onset of arthritis, the early diagnosis of JPsA and its differentiation from juvenile rheumatoid arthritis (JRA) are difficult. The Vancouver criteria[2] facilitate the early diagnosis of JPsA and its differentiation from other forms of arthritis. Using these criteria, a diagnosis of JPsA can be made even in the absence of a psoriatic rash by relying on other clinical features: nail pitting, dactylitis, family history of psoriasis, and a psoriasis-like rash. A followup study[3] of patients with JPsA diagnosed by these criteria showed that 73% of patients have an oligoarticular onset (≤ 4 affected joints) and that even in those with a polyarticular onset, the median number of joints involved at onset was only 6 (range 5–10). This study noted that JPsA may present with arthritis of the small joints of the hands and feet. Small joint disease as a distinct finding in JPsA has been described in other studies, but no specific pattern of joint involvement was found to discriminate between JPsA and the other chronic arthritides. We compared the patterns of joint involvement at onset of disease in oligoarticular JPsA (Oligo-JPsA) and pauciarticular JRA (Pauci-JRA) and estimated the predictive performance of specific patterns for the diagnosis of Oligo-JPsA. The results indicate that recognition of specific patterns of arthritis should facilitate identification of children with JPsA.

MATERIALS AND METHODS

Definitions. JPsA was defined by both the Vancouver criteria[2] and those of the International League of Associations for Rheumatology (ILAR)[4]. A diagnosis of JPsA is made by the Vancouver criteria in a child with chronic arthritis who has psoriasis, or arthritis and 3 (definite) or 2 (probable) of 4 minor criteria: dactylitis (diffuse digit swelling extending beyond the margin of the joint capsule); nail pitting (2 or more pits on the fingernails at any examination); a psoriasis-like rash (historical or examination

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features suggestive of a psoriatic rash, but without conclusive evidence); or a family history of psoriasis in a first- or second-degree relative. JPsA as classified by the ILAR criteria[4] is defined as arthritis with psoriasis, or arthritis with a family history of psoriasis in a first-degree relative, plus dactylitis or nail abnormalities (pitting or onycholysis) in the patient. Rheumatoid factor (RF) must be absent. Pauciarticular JRA is defined by the criteria of the American College of Rheumatology (ACR)[5] as arthritis in 4 or fewer joints during the first 6 months of disease, providing other diseases are excluded. The ILAR definition of oligoarticular juvenile idiopathic arthritis (Oligo-JIA) is the same as that for the ACR criteria for Pauci-JRA, except that, in addition, children are excluded from this category if there is a family history of psoriasis or spondyloarthropathy, or if the child has RF. In all classifications, arthritis begins before 16 years of age and persists for at least 6 weeks.

Subjects. All patients attended the pediatric rheumatology clinics of British Columbia's Children's Hospital or the Mary Pack Arthritis Center, Vancouver, between 1973 and 1995. Some of the patients were reported in previous studies[2,3], but for this study inclusion criteria limited patients to those who (1) had been followed for ≥ 5 years, (2) met the diagnostic criteria (Vancouver criteria for JPsA or ACR criteria for Pauci-JRA), and (3) presented with oligoarticular onset of arthritis, in order to achieve homogeneity of the study group.

Patients with Oligo-JPsA. Twenty-three patients satisfying the Vancouver criteria for JPsA and presenting with oligoarticular arthritis (≤ 4 affected joints) in the first 6 months of disease were eligible for definite entry to the study. Eleven additional children eventually fulfilled the Vancouver criteria for JPsA, but did not do so within the first 6 months of disease. These patients were analyzed separately.

Patients with Pauci-JRA. Sixty-four patients meeting the ACR criteria for diagnosis of Pauci-JRA (≤ 4 affected joints in the first 6 months of disease) were randomly selected from the clinical database by choice of every third chart among consecutively seen patients. All records were reviewed retrospectively and the diagnosis reassessed using the ILAR criteria.

Laboratory studies. RF was detected by latex agglutination or nephelometry; a positive test result was defined as a titer ≥ 1:80. Antinuclear antibodies (ANA) were detected by immunofluorescence microscopy using HEp-2 cells; a positive result was defined as a titer ≥ 1:40. Major histocompatibility complex class I and II antigens were determined by standard microcytotoxicity assays.

Statistical methods. Data are expressed as the mean ± standard deviation (SD). Chi-square or Fisher's exact tests were used for comparison of percentages. Odds ratios (OR) were calculated, with their 95% confidence intervals (CI). P values < 0.05 were deemed statistically significant. Data analysis was performed using the SAS System, Release 6.1, statistical software package (SAS Institute, Inc., Cary, NC, USA).

RESULTS

Patient characteristics. The patient characteristics are shown in Table 1. Sex distribution, age at onset of arthritis, and frequency of seropositivity for ANA and RF were similar in both groups. The groups did not differ with regard to proportions of patients with HLA-B27 or monoarthritis at onset of disease. Sixteen patients had psoriasis prior to the onset of arthritis, with a mean interval between onset of psoriasis and arthritis of 5.7 ± 4.8 years. In 20 patients with Oligo-JPsA (87%) the disease remained oligoarticular during the observed course of disease; in 3 patients more than 4 joints became involved; they developed symmetrical polyarthritis of mainly large joints, metacarpophalangeal (MCP) joints of hands, and metatarsophalangeal (MTP) joints of feet from 5 to 15 years of followup. In the 64 patients with Pauci-JRA, 6 patients progressed to have polyarticular disease. Patients with JPsA who had skin lesions of psoriasis (n = 16) did not differ significantly from those without skin lesions (n = 7) with regard to pattern of arthritis at onset and disease course.

Comparison of classification criteria. The frequencies of major and minor diagnostic criteria for JPsA at onset are summarized in Table 2. At onset of disease, 17 of the 23 patients satisfied the Vancouver criteria for definite JPsA; 6 patients had probable JPsA. The definite and probable JPsA groups did not differ with respect to the relative frequencies of any of the minor diagnostic criteria. At onset of disease 20 of the 23 patients who met the Vancouver criteria satisfied the ILAR criteria for JPsA. Three patients (all diagnosed as having probable JPsA by the Vancouver criteria) who had the criterion psoriasis-like rash (2 patients) and with a history of psoriasis in a second-degree relative (one patient) remained unclassifiable by the ILAR criteria. All of the 11 patients who fulfilled the Vancouver criteria after the first 6 months of disease also eventually fulfilled the ILAR criteria. Thus 14 patients ultimately diagnosed as having JPsA by the ILAR criteria did not fulfill these criteria within the first 6 months of disease. None of the 64 children with Pauci-JRA satisfied ILAR criteria for JPsA, but 7 of these patients would have been excluded from a classification of Oligo-JIA (ILAR criteria) because of a family history of psoriasis.

Patterns of joint involvement at onset of disease. Joint involvement at disease onset in children with JPsA (Vancouver criteria), Pauci-JRA (ACR criteria), and Oligo-JPsA or Oligo-JIA (ILAR criteria) is summarized in Table 3. Arthritis in small joints of the hands, the wrist, and MTP joints of the feet was significantly more frequent in patients with Oligo-JPsA than in those with Pauci-JRA. When the ILAR criteria were applied, the findings were basically the same as those using the Vancouver or ACR criteria.

Comparison of associations of small joint and wrist involvement. The odds ratios (OR) associated with the development of JPsA in the presence of specific criteria are shown in Table 4. Comparing Oligo-JPsA and Pauci-JRA, the OR of a diagnosis of JPsA in a child with wrist disease is 11.13, and with small joint disease is 12.52. Using the ILAR criteria for Oligo-JIA and JPsA, the OR of a diagnosis of JPsA in a child with wrist disease is 9.98, and with small joint disease 14.25. Using either sets of criteria, OR for nail pitting or dactylitis is also high. A possible modification to the ILAR criteria for the diagnosis of JPsA in patients with oligoarticular onset could be replacement of "dactylitis" with a composite criterion: the presence of small joint disease and/or dactylitis and/or wrist disease. Evaluation of the composite criterion shows that it is strongly associated with JPsA diagnosed by either the Vancouver or ILAR criteria (Table 4); patients who fulfilled this criterion were 11.13 times as likely to have JPsA as Pauci-JRA, and 12.11

Table 1. Comparison of patient characteristics.

	Oligo-JPsA, n = 23	Pauci-JRA, n = 64	p
Female:male	16:7	48:16	NS
Age at onset of arthritis, yrs*	7.6 (1.3–14.1)	5.48 (1.1–15.5)	NS
Age at onset of psoriasis, yrs*	5.5 (0.9–13.1)	—	—
ANA test positive	8	32	NS
RF test positive	0	0	NS
HLA–B27 positive	3	9	NS
Chronic asymptomatic uveitis	3	9	NS
Monoarthritis at onset	7	21	NS
Progression to polyarthritis	3	6	NS

* Median (range). NS: nonsignificant.

Table 2. Comparison of frequencies of individual criteria* at onset of disease in patients with Oligo-JPsA classified by Vancouver and ILAR criteria.

	Vancouver Criteria		ILAR Criteria
	Definite JPsA, n = 17	Probable JPsA, n = 6	JPsA, n = 20
Dactylitis	2	2	3
Nail pitting	5	3	8
Psoriasis-like rash	1	2	—**
Family history of psoriasis			
1st degree relative	6	4	10
2nd degree relative	1	1	—**
Psoriasis	16	0	16

*Criteria present at onset: the number of patients in whom a listed criterion occurred during the first 6 months of the disease. ** Not accepted by ILAR criteria.

times as likely to have JPsA as Oligo-JIA ($p < 0.001$ for both comparisons). Application of the composite criterion to those patients who eventually fulfilled Vancouver or ILAR criteria for JPsA, but did not do so within the first 6 months of disease onset, indicated the value of this criterion in early diagnosis. Thus 5 of 11 children who did not meet the Vancouver criteria within the first 6 months of disease, and 4 of the 14 who did not meet the ILAR criteria, would have been classified within the first 6 months as having JPsA if the composite criterion was used. The use of the composite criterion would have allowed 82% of the total group of 34 patients to be diagnosed by the modified Vancouver criteria, and 70% to be diagnosed by the modified ILAR criteria, compared to 67% and 58%, respectively, using the unmodified criteria.

DISCUSSION

This study suggests that small joint disease and wrist disease at onset are helpful in differentiating children with Oligo-JPsA from those who have Pauci-JRA or Oligo-JIA. The pattern of joint involvement in children with PsA has been the subject of other studies, but none has shown a significant difference between children with PsA and those with oligoarticular JRA. In 1977 the JRA Criteria Subcommittee of the ACR[5] noted that PsA in children was similar to the disease in adults in that there was a high frequency of DIP joint involvement, although it is not clear what evidence was available to support this assertion. Lambert, *et al*[1] suggested that there was a specific pattern of joint involvement comparable to that seen in adults with involvement of the DIP joints of the fingers, the interphalangeal joints of the toes and/or asymmetrical involvement of the joints of one or 2 fingers. They found that JPsA presented most commonly with asymmetrical intermittent polyarthritis with tendon sheath effusions.

In the study by Sills[6] comparisons of JPsA and JRA subtypes revealed marked differences in the pattern of joint involvement with regard to small joint disease in the hands and feet. Patients with JPsA had hand involvement in 88% and foot involvement in 67%, but the authors felt that the pattern was not useful in discriminating between JPsA and JRA subtypes.

Shore and Ansell[7] described patients with JPsA as presenting with a pauciarticular onset in 73% of cases, most (87%) showing progression toward polyarthritis. In their patients, wrist disease, although rarely a presenting

Table 3. Comparison of the frequency of individual joint involvement at onset of disease.

	Oligo-JPsA (Vancouver), n = 23	Pauci-JRA (ACR), n = 64	p	Oligo-JPsA (ILAR), n = 20	Oligo-JIA (ILAR), n = 57	p
Affected joints, n (%)						
Knee	18 (78.3)	57 (89.1)	NS	16 (80)	51 (89.5)	NS
Ankle	8 (34.8)	23 (35.9)	NS	7 (35)	19 (33.3)	NS
Subtalar	4 (17.4)	3 (4.7)	0.07	4 (20)	2 (3.5)	0.03
Mid-tarsal	1 (4.3)	3 (4.7)	NS	1 (5)	2 (3.5)	NS
Hip	1 (4.3)	2 (3.1)	NS	1 (5)	1 (1.8)	NS
Foot						
MTP	4 (17.4)	1 (1.6)	0.016	4 (20)	1 (1.8)	0.015
PIP	3 (13)	1 (1.6)	0.05	3 (15)	1 (1.8)	0.05
DIP	3 (13)	1 (1.6)	0.05	3 (15)	1 (1.8)	0.05
Shoulder*	0 (0)	0 (0)	—	0 (0)	0 (0)	—
Elbow	0 (0)	2 (3.1)	NS	0 (0)	2 (3.5)	NS
Wrist	8 (34.8)	2 (3.1)	0.0001	7 (35)	2 (3.5)	0.001
Hand						
MCP	9 (39.1)	1 (1.6)	0.0001	6 (30)	1 (1.8)	0.001
PIP	7 (30.4)	3 (4.7)	0.003	6 (30)	2 (3.5)	0.003
DIP	3 (13)	0 (0)	0.017	3 (15)	0 (0)	0.016
TMJ	1 (4.3)	2 (3.1)	NS	1 (5)	1 (1.8)	NS
Cervical Spine	0 (0)	1 (1.6)	NS	0 (0)	0 (0)	—
Lumbar Spine	0 (0)	0 (0)	—	0 (0)	0 (0)	—
Sacroiliac joint	1 (4.3)	0 (0)	NS	1 (5)	0 (0)	NS
Any small joint**	18 (78.3)	4 (6.3)	0.0001	15 (75)	3 (5.3)	0.0001
Dactylitis	4 (17.4)	1 (1.6)	0.016	4 (20)	1 (1.8)	0.015
Composite criterion***	20 (87)	5 (7.8)	0.0001	17 (85)	4 (7)	0.0001

* Glenohumeral or sternoclavicular or acromioclavicular joints. ** MCP, PIP, DIP of fingers, MTP, PIP, DIP of toes. *** Small joint disease, dactylitis, or wrist joint disease. TMJ: temporomandibular joint.

Table 4. Odds ratios for different patterns of joint involvement at onset using different diagnostic criteria.

	Comparisons					
	Oligo-JPsA (Vancouver Criteria) and Pauci-JRA (ACR Criteria)			Oligo–JPsA (ILAR Criteria) and Oligo-JIA (ILAR Criteria)		
Criterion	OR	95% CI	p	OR	95% CI	p
Small joint disease	12.52	4.73–33.13	< 0.001	14.25	4.60–44.12	< 0.001
Wrist disease	11.13	2.55–48.62	< 0.001	9.98	2.26–44.11	< 0.05
Dactylitis	11.13	1.31–14.63	< 0.05	11.40	1.35–14.78	< 0.05
Composite criterion	11.13	4.73–26.21	< 0.001	12.11	4.63–31.72	< 0.001

problem, became a feature in almost two-thirds. PIP joint involvement was shown to be a frequent feature at onset (25–28%), whereas MCP involvement was more common during the course of arthritis (33%). DIP joints were affected in more than 40% of their patients. Half had tendon sheath involvement.

In 1989 Southwood, *et al*[2] suggested diagnostic criteria for JPsA (the Vancouver criteria). The authors reviewed the clinical and laboratory data of 35 patients who fulfilled the proposed definition. In 33 of 35 patients with JPsA the onset of arthritis was pauciarticular, but the disease followed a polyarticular course in 23 of 35. The most common affected joints at onset included those of the digits in 12 patients (34%) and, less frequently, the ankles (14%), hip (11%), and wrist (9%). During the course of arthritis an asymmetrical digital arthritis occurred in 24 (69%) and dactylitis occurred in 17 patients (49%). In a large series of patients with JPsA, Truckenbrodt, *et al*[8] showed that at disease onset, 16–23% had small joint disease of either hands or feet, and 30% had wrist joint disease. Only 8% of their patients had dactylitis at onset of disease. Those findings are consistent with the results of our study and the patterns of arthritis described in the clinical followup study of Roberton, *et al*[3]. Discrepancies in these studies may be accounted for by differences in criteria used to define JPsA. Limitations of this study include its retrospective design and the relatively small numbers of patients observed by several examiners, among whom there may have been differences in joint assessment.

A larger patient group might even have shown a higher frequency of subtalar joint involvement in patients with JPsA.

It is clear that the presence of arthritis of small joints of the hands or feet or of the wrist joints is much more common in children who fulfill currently used criteria for the diagnosis of JPsA than in children with Pauci-JRA. Recognition of this pattern of joint involvement should facilitate identification of children with JPsA, and should be considered as a criterion modification in the ILAR classification of JIA, perhaps as part of a composite criterion including the presence of small joint disease, dactylitis, and wrist joint disease. Further prospective evaluation of this suggestion in other populations is warranted.

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