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Weighting improves the information provided by joint counts on the severity of arthritis and its impact on patients' well-being in juvenile idiopathic arthritis

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***Objective*. To develop a scoring system for juvenile idiopathic arthritis (JIA) in which joints are weighted to reflect their relative importance to children's function and to examine whether weighting increases the correlation of joint counts with subjective and laboratory outcome measures.**

***Methods*. A weighted joint score was devised by a panel of experienced paediatric rheumatologists, who assigned a weight from 1 (not very important) to 10 (essential for key functional activities) to each joint based on its functional importance to children's physical and daily activities. The associations of simple and weighted counts of swollen, tender, limited and active joints with the physician's global assessment of overall disease activity, the parent's global assessment of the child's overall well-being and intensity of pain, the Childhood Health Assessment Questionnaire (C-HAQ), the Child Health Questionnaire (CHQ) and the erythrocyte sedimentation rate were compared using Spearman's correlation analysis in 60 unselected patients seen in the clinic and in 61 consecutive patients with disease duration ≥5 yr.**

***Results*. Weighted counts of swollen and active joints yielded greater correlation with the physician's global assessment than did simple counts. The correlation of weighted counts of swollen, painful and active joints with the parent's assessment of overall well-being and intensity of pain was superior to that provided by simple counts. Weighting increased most of the correlations between joint counts and the C-HAQ score and the physical component of the CHQ.**

***Conclusion*. Weighting improves the information provided by joint counts on the severity of arthritis and its impact on patients' well-being.**

KEY WORDS: Juvenile idiopathic arthritis, Juvenile rheumatoid arthritis, Juvenile chronic arthritis, Joint counts, Articular scores, Weighting.

The severity of joint disease in patients with juvenile idiopathic arthritis (JIA) is traditionally assessed by counting the number of joints with swelling, tenderness/pain on passive motion (TEN/POM) and limited range of motion (LROM), and by calculating, through a combination of these parameters, the number of active joints. A severity score can also be obtained for the three articular indices and the overall joint disease by grading symptoms in each joint on a categorical scale and summing the scores obtained in all joints [1, 2]. In both these scoring systems, however, each joint is considered as a single unit and, thus, a small joint in a hand or a foot is weighted as a large joint, such as a hip, a knee or a shoulder. Although this approach provides an objective measure of the spread of arthritis, it may not give a reliable picture of the patient's status. Since involvement of different joints may have different impacts on physical and daily activities, a child whose hips or knees are affected is regarded as having a more severe disease than if only two interphalangeal joints in the hand or foot were involved. This limitation of the simple joint count is particularly relevant in JIA, which is characterized by a widely variable distribution of joint disease, in terms of both number and type of affected joints [3].

The objectives of the present study were to develop a new scoring system in which joints are weighted to reflect their relative importance to children's function and to examine whether weighting increases the association of joint counts with the subjective and laboratory measures of JIA severity, including the physician's global assessment of disease activity, the parent-reported outcomes, and the erythrocyte sedimentation rate (ESR).

Patients and methods

Development of the weighted joint score

Six experienced paediatric rheumatologists working at the study units (A.R., N.R., S.M.M., A.B., A.L., A.M.) were asked to assign independently a weight from 1 to 10 to each joint of the body based on its functional importance to children's physical and daily activities (1 = not very important; 10 = essential for key functional activities). All investigators agreed to evaluate each joint as a single unit and not as a category (i.e. to assign a weight to one proximal interphalangeal joint and not to the group of interphalangeal joints

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TABLE 1. Comparison of unweighted and weighted joint scores

Joint	Unweighted score	Weighted score
Temporomandibular	1 (2)	3 (6)
Cervical spine	1 (1)	6 (6)
Shoulder	1 (2)	8 (16)
Elbow	1 (2)	5 (10)
Wrist	1 (2)	6 (12)
Hand metacarpophalangeal	1 (10)	2 (20)
Hand proximal interphalangeal	1 (10)	2 (20)
Hand distal interphalangeal	1 (8)	1 (8)
Hip	1 (2)	10 (20)
Knee	1 (2)	8 (16)
Ankle	1 (2)	7 (14)
Foot metatarsophalangeal	1 (10)	1 (10)
Foot interphalangeal	1 (10)	1 (10)

For each joint category, the maximum possible score that can be obtained by summing all joints present in the body is given in parentheses.

that are present in one or both hands). After review of each investigator's indications and extensive discussion, a consensus was reached among the members of the panel about a final set of joint weights (Table 1). Joint weights were agreed upon by the two physiotherapists who work in the study units. Furthermore, joint weights were developed after having conducted an inquiry within the members of the Italian Pediatric Rheumatology Study Groups, who were asked to provide their suggestion about the more suitable weight for each joint (data not shown).

Study data sets

Two different cohorts of patients followed at the study units and fulfilling the International League of Associations for Rheumatology (ILAR) [4] were studied: the first was composed of 60 patients seen consecutively between April and June, 2004; the second by 61 consecutive patients with a disease duration $\geq$5 yr seen between September 2003 and March 2004 and included in a study aimed at developing and validating a new damage tool for JIA [5]. Due to the potentially confounding effect of enthesalgia, patients with enthesitis-related arthritis were excluded.

Clinical assessments

The medical charts of each patient were reviewed for the following information: sex, age at disease presentation, JIA category, age at study visit, and disease duration. At the time of the study visit, a total of 67 joints (those that are included in the standard clinical evaluation; see Table 1) were assessed in each patient for the presence of swelling, TEN/POM and LROM; a joint was defined as active if it was swollen or, if no swelling was present, if it had TEN/POM + LROM. For purposes of analysis, the ankle and subtalar joint were considered as a single unit. The results of articular examinations were recorded in standardized forms. Joint assessments were performed by three investigators (A.R. or A.B. in Genova and S.M.M. in Pavia), who used the same method throughout the study. The following clinical variables were also recorded at the study visit: physician's global assessment of the overall disease activity on a 10-cm visual analogue scale (VAS) (0 = no activity; 10 = maximum activity); parent's global assessment of the child's overall well-being on a 10-cm VAS (0 = very well; 10 = very poor); parent's assessment of the child's intensity of pain on a 10-cm VAS (0 = no pain; 10 = very severe pain); Childhood Health Assessment Questionnaire (C-HAQ) disability index (0 = no disability, 3 = maximum disability), Italian version [6]; and ESR (Westergren method). In the cohort of patients with $\geq$5 yr of disease duration, clinical assessments included the evaluation of the health-related quality of life (HRQL) using the Child Health Questionnaire (CHQ), Italian version [6]. Briefly, the CHQ is a generic health instrument designed to capture the physical and psychosocial functioning of children 5 yr of age and older. It measures 15 health concepts through 50 items/questions and yields two summary scores: the physical summary score (PhS) and the psychosocial summary score (PsS). Scores for each subscale range from 0 to 100, higher scores reflecting better health status. The mean $\pm$ S.D. norm-based score for both PhS and PsS is 50 ± 10.

Statistics

In all analyses, non-parametric statistics were used to account for the non-normal distribution of the articular indices and most of the clinical and outcome variables. Based on the results of the articular examination, the following simple joint counts were obtained for each patient: number of swollen joints; number of joints with TEN/POM; number of joints with LROM; and number of active joints. The weighted scores were then calculated for each joint count by assigning to each involved joint its correspondent weight. The level of correlation of simple and weighted joint counts with the physician's global assessment of the overall disease activity, the parent's global assessment of the child's overall well-being, the parent's assessment of the child's intensity of pain, the C-HAQ disability index, the PhS and PsS of the CHQ, and the ESR was compared using Spearman's rank correlation analysis. Differences in the magnitude of correlation between the unweighted and weighted joint counts were interpreted qualitatively. The statistical package used for all analyses was Statistica (StatSoft, Tulsa, OK, USA).

Results

The weighted joint scores are presented together with the simple scores in Table 1. The hip, the knee and the shoulder were assigned the highest weights, whereas the distal interphalangeal joints of the hands and the small foot joints received a weight of 1, as in the simple joint count. The metacarpophalangeal and proximal interphalangeal joints of the hands were weighted by 2 points, but the total score that would be provided by the simultaneous involvement of all joints in each group in both hands is equal to that yielded by the involvement of both hips.

Table 2 shows the demographic and clinical features of the two patient data sets. Altogether, the patient cohorts were representative of the whole spectrum of severity of the JIA patients who are usually seen in a paediatric rheumatology clinic. Compared with the unselected cohort, patients with disease duration $\geq$5 yr were younger at disease onset and were judged, on average, to have a lower overall level of disease activity by the physician, and to have better well-being and less intense pain by the parents; however, the median counts of swollen, painful and active joints were comparable in the two cohorts. As expected, patients with longer disease duration tended to have a greater number of joints with LROM. Notably, the median C-HAQ score was higher in the cohort of patients with shorter disease duration, perhaps reflecting the prominent influence of joint pain on children's functional ability.

Spearman's correlations of simple and weighted joint counts with the physician's global assessment of disease activity, the parent-reported outcomes, and the ESR in the two patient cohorts are shown in Tables 3 and 4. In both data sets, weighted counts of swollen and active joints yielded greater correlation with the physician's global assessment than did simple counts. Likewise, the correlation of weighted counts of swollen, painful and active joints with the parent's assessment of overall well-being and intensity of pain was higher than that provided by the simple counts.

TABLE 2. Demographic and clinical features of the study patients

	Unselected patients seen in the clinic ($n=60$)	Patients with disease duration $\geq$5 yr ($n=61$)
No. males/females	14/46	14/47
Disease subtype (no.)		
Systemic	7	11
Polyarticular	9	14
Oligoarticular persistent	25	15
Oligoarticular extended	16	21
Psoriatic	3	–
Age at onset (yr)	3.7 (0.4–15.3)	2.5 (0.5–14.2)
Disease duration (yr)	3.6 (0.5–21)	7.7 (4.3–24.5)
Physician's global assessment of overall disease activity[a]	5.1 (0–10)	2.6 (0–10)
Parent's global assessment of child's overall well-being[a]	2.8 (0–10)	0.7 (0–9.4)
Parent's assessment of child's pain[a]	1.8 (0–10)	1.0 (0–9.9)
Childhood Health Assessment Questionnaire score[b]	0.25 (0–2.25)	0.12 (0–1.5)
No. of swollen joints	2 (0–24)	2 (0–30)
No. of swollen joints, weighted	10 (0–83)	13 (0–84)
No. of joints with tenderness/pain on motion	2 (0–41)	1 (0–28)
No. of joints with tenderness/pain on motion, weighted	14 (0–107)	8 (0–78)
No. of joints with limited range of motion	1 (0–16)	2 (0–61)
No. of joints with limited range of motion, weighted	8 (0–71)	12 (0–152)
No. of active joints	2 (0–30)	3 (0–32)
No. of active joints, weighted	14 (0–99)	16 (0–120)
Child Health Questionnaire—Physical summary score[c]	–	52.9 (18.3–58.3)
Child Health Questionnaire—Psychosocial summary score[c]	–	50.1 (34.8–62.3)
Erythrocyte sedimentation rate[d]	19 (4–130)	21 (2–108)

Values are median and ranges (in parenthesis) unless otherwise indicated. [a]Range of 0 (best) to 10 (worst); [b]range of 0 (best) to 3 (worst); [c]range of 0 (worst) to 100 (best) (mean $\pm$ S.D. norm-based score for both summary scores, 50 ± 10); [d]normally <15 mm/h.

TABLE 3. Correlation of simple and weighted joint counts with JIA severity measures in 60 unselected patients seen in the clinic

	MD global	Parent global	Parent pain	C-HAQ	ESR
No. of swollen joints	0.46[c]	0.05	0.14	0.06	0.05
No. of swollen joints, weighted	0.57[d]	0.18	0.26[a]	0.18	0.24
No. of joints with TEN/POM	0.54[d]	0.39[b]	0.39[b]	0.45[c]	0.31[a]
No. of joints with TEN/POM, weighted	0.48[d]	0.43[c]	0.43[c]	0.50[d]	0.39[b]
No. of joints with LROM	0.38[b]	0.23	0.19	0.43[c]	0.43[c]
No. of joints with LROM, weighted	0.35[b]	0.24	0.17	0.43[c]	0.42[c]
No. of active joints	0.46[c]	0.14	0.16	0.21	0.12
No. of active joints, weighted	0.54[d]	0.24	0.23	0.32[a]	0.29[a]

MD global, physician's global assessment of the overall disease activity; Parent global, parent's global assessment of the child's overall well-being; Parent pain, parent's assessment of the child's pain. [a]$P<0.05$; [b]$P<0.01$; [c]$P<0.001$; [d]$P<0.0001$ (P values designate the level of statistical significance of the Spearman's correlation for the respective joint count with each individual parameter).

TABLE 4. Correlation of simple and weighted joint counts with JIA severity measures in 61 patients with disease duration $\geq$5 yr

	MD global	Parent global	Parent pain	CHAQ	ESR	CHQ–PhS	CHQ-PsS
No. of swollen joints	0.59[d]	0.35[b]	0.53[d]	0.31[a]	0.37[b]	−0.18	−0.22
No. of swollen joints, weighted	0.65[d]	0.40[b]	0.55[d]	0.30[a]	0.33[b]	−0.20	−0.19
No. of joints with TEN/POM	0.34[b]	0.43[c]	0.48[d]	0.56[d]	0.35[b]	−0.40[b]	−0.17
No. of joints with TEN/POM, weighted	0.39[b]	0.48[d]	0.53[d]	0.57[d]	0.37[b]	−0.44[b]	−0.17
No. of joints with LROM	0.15	0.33[b]	0.33[b]	0.60[d]	0.16	−0.38[a]	−0.02
No. of joints with LROM, weighted	0.16	0.36[b]	0.33[b]	0.63[d]	0.16	−0.41[b]	−0.02
No. of active joints	0.45[c]	0.40[b]	0.51[d]	0.55[d]	0.35[b]	−0.37[a]	−0.11
No. of active joints, weighted	0.55[d]	0.49[d]	0.60[d]	0.55[d]	0.36[b]	−0.38[a]	−0.14

MD global, physician's global assessment of the overall disease activity; Parent global, parent's global assessment of the child's overall well-being; Parent pain, parent's assessment of the child's pain. [a]$P<0.05$; [b]$P<0.01$; [c]$P<0.001$; [d]$P<0.0001$ (P values designate the level of statistical significance of the Spearman's correlation for the respective joint count with each individual parameter).

Weighting also increased most of the correlations between the joint counts and the C-HAQ score and, in the cohort with longer disease duration, the CHQ-PhS. All correlations between joint counts, either simple or weighted, and the CHQ-PsS were low and none of them was statistically significant.

Discussion

Nearly 50 yr ago, Lansbury and Haut [7] pointed out that a simple count of the number of affected joints cannot provide sufficient information on the clinical status of an adult patient with rheumatoid arthritis. Considering that involvement of joints of

TABLE 5. Comparison of unweighted and weighted active joint counts in the seven study patients who had five active joints

	Involved joints	Unweighted active joint count	Weighted active joint count
Patient 1	3 hand MCP, 2 foot IP	5	8
Patient 2	5 hand PIP	5	10
Patient 3	1 wrist, 2 ankles, 2 foot IP	5	22
Patient 4	1 elbow, 2 wrists, 1 knee, 1 ankle	5	32
Patient 5	1 elbow, 1 hand PIP, 2 hips, 1 knee	5	35
Patient 6	2 wrists, 1 hip, 2 knees	5	38
Patient 7	1 hand MCP, 2 hips, 2 knees	5	38

different size (i.e. large *vs* small joints) may produce a different amount of arthritis, they devised a new index for measuring the severity of articular inflammation, in which joints were weighted for their size. This index was subsequently found to yield higher correlations with clinical and outcome parameters than did simple joint counts [8, 9]. Weighting of joint counts may be particularly appropriate for JIA, which is characterized by widely heterogeneous and frequently asymmetrical joint involvement, and may thus present with many different combinations of articular patterns [3]. However, the approach followed by Lansbury and Haut is not applicable to JIA patients because joint size in children varies with age and body growth.

Bearing in mind that certain key joints are more relevant to functionality than others, we developed a scoring system in which joints were weighted to reflect their relative importance to children's physical and daily activities. The score was developed by a panel of paediatric rheumatologists who have more than 5 yr of experience in the assessment of patients with JIA, thus ensuring face and content validity. The weighting method is simple and easy to apply in the clinical setting. After joint assessment in a patient and the recording of its findings on a standardized form in which the weights for each joint are already printed, calculating the weighted counts takes only a few minutes to complete.

To investigate whether weighting improved the clinical information about patient status provided by joint counts, we compared the association of weighted and simple joint counts with the subjective and laboratory measures of JIA severity, including the physician's global assessment of disease activity, the parent-reported outcomes and the ESR. We found that weighting improved consistently the correlation of swollen and active joint counts with the physician's global assessment. This finding is clinically relevant because swelling (which is a major determinant of the definition of joint disease activity) is generally viewed as the most objective measure of joint synovitis. However, because most physicians will take into account how significant the affected joints are when assigning their overall score, weighting the joints for functional importance may make the measure more similar to the physician's global assessment, leading to redundancy between the measures. Weighting also yielded a substantially greater correlation of swollen and active joint counts with the ESR (in the unselected patient cohort) and with most of the parent-reported outcomes. These may be considered more independent external standards than the physician's global assessment and, in the case of parent-derived parameters, more relevant standards because they directly reflect the impact of the disease on the patient. In contrast to what was seen for swollen and active joint counts, there was no overall increase in the correlation of weighted tender and limited joint counts with the physician's global assessment. Joint TEN/POM is a semi-objective measure of joint inflammation that may be influenced by the patient's/parent's perception. The subjective component of tender joint count may explain the superior correlation of its weighted version with the parent's assessment of well-being and pain intensity, the physical disability measure (the C-HAQ) and the physical component of the HRQL tool (the CHQ-PhS), which are determined not only by the level of impairment but also by subjective factors, including the parent's perception of the child's pain. Likewise, the weighted count of joints with LROM yielded a slightly higher correlation with the parent-reported measures of physical functioning (the C-HAQ and the CHQ-PhS) compared with the unweighted count, although only in the subset of patients with longer disease duration. The lack of correlation of joint counts, either simple or weighted, with the psychosocial component of the CHQ is not surprising because many other factors, beside joint symptoms, combine to determine the child's psychosocial well-being. Altogether, these results suggest that it is worthwhile to weight joint counts on the basis of the functional importance of each joint to children's physical and daily activities.

Differences in the pattern of joint disease may have important implications for clinical trials in JIA. To ensure equivalence in disease severity among treatment arms, patient groups are compared for a number of clinical measures at baseline, including joint counts. However, assuming that patients with the same number of affected joints have equally severe arthritis may be misleading. To provide an example, we compared the type of involved joints and calculated the weighted score in the seven study patients who had five active joints, which is the minimum requirement for inclusion in trials of second-line agents in JIA [10, 11] As shown in Table 5, these patients had a different pattern of joint disease and a widely variable weighted active joint count, which ranged from 8 to 38. The active joint count and the count of joints with LROM are part of the six core-set variables of the ACR Pediatric 30, which is the standard definition of improvement used in JIA clinical trials [12]. It remains to be demonstrated whether weighting enhances the responsiveness to change in joint counts and the discriminative power of response criteria. Future analyses of clinical trial data might compute both weighted and unweighted indices.

We acknowledge that, since the joint weights we developed were based on experts' opinions, they may have been affected by subjectivity. A statistical approach, such as Rasch modelling [13], might have generated more objective weightings, although it might have led to a loss of face validity. To make the weighted score simple and easy to apply, the ankle and subtalar joints were lumped together and the tarsal joint of the feet was not included. However, the weighting does not imply that the joints not included in the score do not need to be treated to achieve full remission of disease, or that they have a lesser impact on function or are less important in defining the burden of the disease. Another potential limitation of the weighting is that it may raise problems when the pattern of affected joints changes drastically over time.

In summary, we present a practical and easy-to-use weighted joint score that reflects the relative importances of the different joints to children's physical and daily activities. Weighting improved the association of joint counts with the subjective and laboratory measures of JIA severity, including the parent-reported child's well-being, pain intensity, functional ability and HRQL.

This index may provide useful information that can complement other clinical measures when assessing the level of disease activity, the effectiveness of treatment regimens, and disease progression in patients with JIA.

Rheumatology	Key messages
	• A weighted joint score is presented that reflects the relative importance of each joint to children's physical activities. • Weighting improved the association of joint counts with measures of disease severity.

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