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Modified Juvenile Spondyloarthritis Disease Activity Index in the Childhood Arthritis and Rheumatology Research Alliance (CARRA) Registry

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ABSTRACT. ***Objective.*** To validate the Juvenile Spondyloarthritis Disease Activity Index (JSpADA), and modified versions thereof, in a North American cohort of patients with enthesitis-related arthritis (ERA).

Methods. We utilized the Childhood Arthritis and Rheumatology Research Alliance Registry database ERA cohort to validate the JSpADA and its modifications (JSpADA6-no Schober, no C-reactive protein [CRP]/erythrocyte sedimentation rate [ESR]; JSpADA7-no Schober; and JSpADA7-no CRP/ESR) using the Outcome Measures in Rheumatology principles of face validity, discriminative validity, and responsiveness to change.

Results. There were 51 subjects (64 visits) with complete JSpADA data with a mean age of 13.7 years and disease duration of 30.9 months. Subjects were predominantly White (84.3%), and 56.9% were male and 50% were HLA-B27 positive. The JSpADA showed high correlation with the clinical 10-joint Juvenile Arthritis Disease Activity Score (cJADAS10; $r = 0.81$), moderate-to-high correlation with physician global assessment (PGA; $r = 0.69$), and low-to-fair correlation with Childhood Health Assessment Questionnaire (CHAQ; $r = 0.22$). The modifications of the JSpADA (JSpADA7-no Schober; JSpADA7-no CRP/ESR; and JSpADA6-no Schober, no CRP/ESR) performed similarly with high correlation with cJADAS10 ($r = 0.81$, 0.79, and 0.80, respectively), moderate-to-high correlation with PGA ($r = 0.65$, 0.67, 0.64, respectively), and low-to-fair correlation with CHAQ ($r = 0.35$, 0.34, 0.39, respectively). All modified versions of JSpADA had good responsiveness to change. All versions of JSpADA had excellent discriminative validity.

Conclusion. We propose the term *modified JSpADA* for the modification of JSpADA with 6 elements (JSpADA6-no Schober, no CRP/ESR). This shorter disease activity index may improve implementation of JSpADA in both clinical practice and research trials.

Key Indexing Terms: disease activity score, juvenile idiopathic arthritis, spondyloarthropathy

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Enthesitis-related arthritis (ERA) is in the juvenile spondyloarthritis (JSpA) spectrum and is characterized by enthesitis, lower extremity arthritis, axial inflammation, symptomatic (acute) anterior uveitis, and association with HLA-B27.[1] Compared to patients with other categories of juvenile idiopathic arthritis (JIA), those with ERA tend to have poorer outcomes with more pain, lower quality of life, and lower rates of remission.[2-6]

There are several disease activity measures in JIA, most of which were not developed specifically for use in JSpA.[7] Disease activity measures developed for ankylosing spondylitis are not appropriate for use in children because of differences in disease phenotype.[8,9] The Juvenile Spondyloarthritis Disease Activity Index (JSpADA) is the first disease activity score developed specifically for JSpA and consists of the following equally weighted elements: active joint count, active enthesitis count, visual analog scale (VAS) for pain, erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP) related to JSpA activity, morning stiffness, clinical sacroiliitis, uveitis, and back mobility.[10] Each item is rated 0 or 1 for a maximum score of 8. JSpADA was validated in a retrospective multicenter cohort of 243 children in 2014, where

it had high or moderate correlations with the clinical 10-joint Juvenile Arthritis Disease Activity Score (cJADAS10), physician global assessment (PGA), patient global assessment, and the Childhood Health Assessment Questionnaire (CHAQ).[10] JSpADA was further validated in a prospective cohort of 127 children with ERA from India[11]; eliminating the Schober measure improved the performance of the modified JSpADA in this cohort. However, most of the patients in this prospective validation cohort were on nonsteroidal antiinflammatory drugs and/or disease-modifying antirheumatic drugs with none of the patients on biologic agents, limiting generalizability to other populations. As such, this project aims to validate JSpADA, and several modifications thereof, in a North American cohort of patients with ERA.

METHODS

Patients. Patients who were enrolled in the Childhood Arthritis and Rheumatology Research Alliance (CARRA) Registry and fulfilled the International League of Associations for Rheumatology criteria for ERA were eligible for inclusion. The CARRA Registry prospectively collects data in the US and Canada with a primary objective to evaluate safety of therapeutic agents in children with rheumatic diseases.[12] Subjects and/or parents provide consent upon enrollment in this registry. Institutional review board exemption was obtained at Children's National Hospital. Deidentified data were obtained for subjects with ERA, psoriatic arthritis (PsA), and undifferentiated JIA categories from July 2015 to February 2019. Only subjects with ERA who had data for all items of the original JSpADA (complete dataset) were included in the final analysis. Demographic data (age, sex, and race), disease duration, HLA-B27 status, and disease characteristics were gathered for each patient.

JSpADA. JSpADA score was calculated for each patient with complete data. Modified versions of the JSpADA were created and analyzed in different iterations by first eliminating the Schober measure (ie, JSpADA7-no Schober); second, by eliminating the inflammatory measures (CRP/ESR; ie, JSpADA7-no CRP/ESR); and finally, by eliminating both CRP/ESR and the Schober (ie, JSpADA6-no Schober, no CRP/ESR). PGA, CHAQ, and cJADAS10 for each patient were correlated with the original JSpADA and with the modified versions.

Statistical analysis. Statistical analyses were performed using SAS 9.4 (SAS Institute). Baseline demographic differences were assessed with descriptive statistics. Outcome Measures in Rheumatology (OMERACT) principles of truth, discrimination, and feasibility were used to test the validity of the modified JSpADA versions in the CARRA cohort.[13]

Truth. Face and content validity for the original JSpADA had been established through the modified Delphi questionnaire exercise.[10] Construct validity was tested using other JIA disease activity measures such as the cJADAS10, PGA, and CHAQ using Pearson correlation.

Table 1. Baseline characteristics.[a]

Variable	Complete JSpADA, n = 51	Incomplete JSpADA, n = 654	*P*
		Subject-Level Differences	
Age, yrs, mean (SD)	13.7 (3.3)	14.1 (3.1)	0.42
Male sex	29 (56.9)	382 (58.4)	0.83
Race/ethnicity			
White	43 (84.3)	556 (85)	0.89
Black	2 (3.9)	34 (5.2)	0.69
Hispanic	5 (9.8)	50 (7.6)	0.58
American Indian	1 (2)	6 (0.9)	0.47
Asian	1 (2)	25 (3.8)	0.50
Disease duration, mos, mean (SD)	30.9 (34.6)	39.0 (37.2)	0.13
HLA-B27 positive	19 (50)	249 (46.1)	0.64
Clinically active sacroiliitis	15 (31.3)	67 (11.2)	< 0.001
Sacroiliitis by imaging[b]	6 (37.5)	57 (39.6)	0.87
Sacroiliac joint damage by imaging[c]	3 (16.7)	48 (25.7)	0.40
	n = 64	Visit-Level Differences n = 1897	
Active joint count, median (IQR)	2 (0-4.5)	0 (0-2)	< 0.001
Active enthesitis count, median (IQR)	2 (1-4)	1 (0-4)	0.19
cJADAS10, mean (SD)	9.5 (6.8)	6.4 (5.9)	< 0.001
PGA, mean (SD)	2.8 (1.9)	1.6 (1.8)	< 0.001

Values are n (%) unless otherwise indicated. [a] Comparison of baseline characteristics of those with complete dataset for JSpADA to those without, both at subject and visit level. [b] Sacroiliitis on imaging is defined in the CARRA Registry as imaging evidence of active sacroiliac arthritis in the form of synovitis or bone marrow edema by MRI or CT. [c] Sacroiliac joint damage is defined in the CARRA Registry as imaging evidence of sclerosis, joint space narrowing, or ankyloses on radiograph, MRI, or CT. CARRA: Childhood Arthritis and Rheumatology Research Alliance; cJADAS10: clinical 10-joint Juvenile Arthritis Disease Activity Score; CT: computed tomography; JSpADA: Juvenile Spondyloarthritis Disease Activity; MRI: magnetic resonance imaging; PGA: physician global assessment.

Discrimination. PGA was used as the external criterion to determine discriminative validity, where VAS = 0 denoted inactive disease and VAS > 0 signified active disease. The 2-sample *t* test was used to compare groups with active and inactive disease. Responsiveness to change in clinical disease activity over time was calculated by comparing the mean change in the JSpADA between visits with improved or worsening of disease activity according to PGA. The analysis was performed between the first and last visits (≥ 6 months apart) for patients with > 2 visits using 2-tailed *t* test and the standardized mean difference (SMD) to clarify the effect size. An SMD of 0.2 was considered small, 0.5 medium, and 0.8 large.[14]

Feasibility. The original JSpADA used the modified Delphi exercise to develop the list of items that were intended to be easily measured, inexpensive, and presented minimal or no risk to the patient.[7] Since we did not add any new items to the score, a separate Delphi exercise was not performed.

RESULTS

Patients. Of 4058 total CARRA visits for patients with JIA with ERA, PsA, and undifferentiated arthritis, 1961 were for patients exclusively with ERA. Of these, 64 visits (n = 51 subjects) had complete JSpADA data (Supplementary Table S1, available with the online version of this article). There was high percentage of missingness for the Schober and inflammatory markers (74.1% and 55.1%, respectively) in the total ERA visits (Supplementary Table S2). Patients with a complete dataset had a mean age of 13.7 years; mean disease duration was 30.9 months (Table 1). The majority were White (84.3%), and 56.9% (19/38) were male and 50% were HLA-B27 positive. The median joint count was 2 (IQR 0-4.5), and the median enthesitis count was 2 (IQR 1-4). Clinically active sacroiliitis was present in 15/48 (31.3%) of patients, sacroiliitis was noted on imaging in 6/16 (37.5%), and sacroiliac joint damage in 3/18 (16.7%). The cohort of patients with ERA where complete JSpADA was calculable had more disease activity compared to those with incomplete JSpADA with notable differences in clinically active sacroiliitis (31.3% vs 11.2%; $P < 0.001$), active joint count (2 vs 1; $P < 0.001$), cJADAS10 (9.5 vs 6.4; $P < 0.001$), and PGA (2.8 vs 1.6; $P < 0.001$). Summary statistics of JSpADA, its modified versions, cJADAS10, PGA, and CHAQ are noted in Supplementary Table S3.

Table 2. Correlation of JSpADA score and the modified versions of JSpADA with the CHAQ, PGA, and cJADAS10.

JSpADA Versions (n)	Other Disease Activity Measures	Pearson Correlation Coefficient	*P*
JSpADA (64)	CHAQ	0.22	0.11
	PGA	0.69	< 0.001
	cJADAS10	0.81	< 0.001
JSpADA7-no Schober (242)	CHAQ	0.35	< 0.001
	PGA	0.65	< 0.001
	cJADAS10	0.81	< 0.001
JSpADA7-no CRP/ESR	CHAQ	0.34	< 0.001
	PGA	0.67	< 0.001
	cJADAS10	0.79	< 0.001
JSpADA6-no Schober, no CRP/ESR	CHAQ	0.39	< 0.001
	PGA	0.64	< 0.001
	cJADAS10	0.80	< 0.001

OMERACT principle of face value demonstrating correlation of various versions of JSpADA to other validated disease activity measures such as CHAQ, PGA, and cJADAS10. CHAQ: Childhood Health Assessment Questionnaire; CRP: C-reactive protein; cJADAS10: clinical 10-joint Juvenile Arthritis Disease Activity Score; ESR: erythrocyte sedimentation rate; JSpADA: Juvenile Spondyloarthritis Disease Activity; OMERACT: Outcome Measures in Rheumatology; PGA: physician global assessment.

Truth. Correlation with cJADAS10 was high for JSpADA ($r = 0.81$; Table 2). Correlation between cJADAS10 and the modified JSpADA scores was also high at 0.81, 0.79, and 0.80, respectively, for JSpADA7-no Schober, JSpADA7-no CRP/ESR, and JSpADA6-no Schober, no CRP/ESR. Correlation between PGA and JSpADA was moderate-to-high at 0.69 and was also moderate-to-high for the modified versions at 0.65 (JSpADA7-no Schober), 0.67 (JSpADA7-no CRP/ESR), and 0.64 (JSpADA6-no Schober, no CRP/ESR). Correlation with CHAQ was low-to-fair at 0.22 (JSpADA), 0.35 (JSpADA7-no Schober), 0.34 (JSpADA7-no CRP/ESR), and 0.39 (JSpADA6-no Schober, no CRP/ESR).

Discrimination. JSpADA and its modified versions had excellent discriminative validity in separating active and inactive disease (Figure). The mean scores for subjects with active and inactive disease were 3.31 and 0.94 for JSpADA ($P < 0.001$), 3.03 and 1.13 for JSpADA7-no Schober ($P < 0.001$), 2.77 and 1.13 for JSpADA7-no CRP/ESR ($P < 0.001$), and 2.45 and 0.65 for JSpADA6-no Schober, no CRP/ESR ($P < 0.001$), respectively.

The SMD between those who improved and those who worsened was 0.55 ($P = 0.55$), 1.15 ($P = 0.004$), 0.94 ($P = 0.048$), and 1.42 ($P < 0.001$) for JSpADA, JSpADA7-no Schober, JSpADA7-no CRP/ESR, and JSpADA6-no Schober, no CRP/ESR, respectively (Table 3).

Feasibility. Whereas only 3.2% of total ERA visits had a complete dataset to calculate the original JSpADA, the percent of visits where the modified versions of JSpADA could be calculated increased to 7.1% for JSpADA7-no CRP/ESR, 12.3% for JSpADA7-no Schober, and 25.8% for JSpADA6-no Schober, no CRP/ESR (Supplementary Table S4, available with the online version of this article). The other variables contributing to the missingness are noted in Supplementary Table S2.

DISCUSSION

In this study, our aim was to validate the JSpADA in ERA in the CARRA Registry. Further, we sought to validate modified versions of the measure with a reduced number of items, (ie, by first excluding the Schober [JSpADA7-no Schober], then excluding CRP/ESR [JSpADA7-no CRP/ESR], and then excluding both [JSpADA6-no Schober, no CRP/ESR]).

We chose to eliminate Schober and CRP/ESR as abnormality in Schober may be a late finding in ERA and patients may not have laboratory evaluations completed at every visit.[15] The high percentage of missingness in the CARRA database for the Schober and inflammatory markers further supports the need for simpler measures. Applying the OMERACT principles, we demonstrated that the original JSpADA and its modified versions are valid. The JSpADA and the modified versions

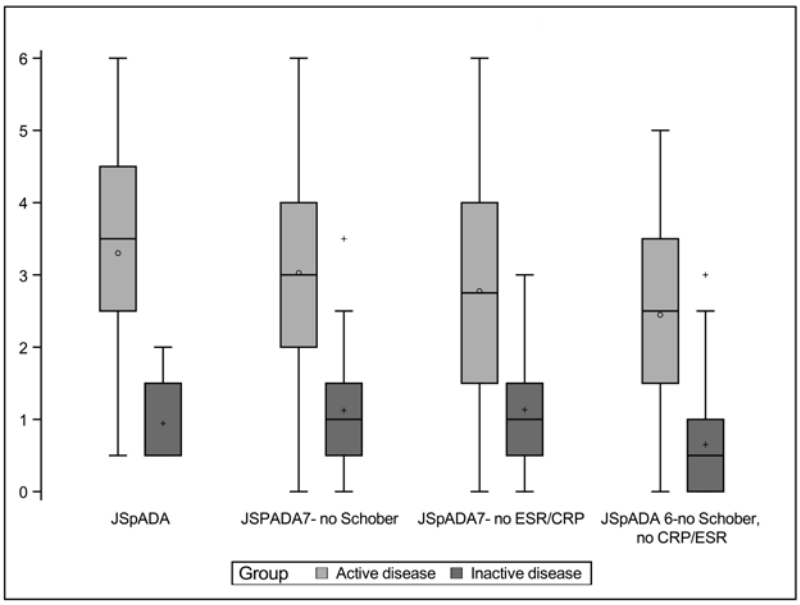

Figure. Distribution of JSpADA score by active disease. CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; JSpADA: Juvenile Spondyloarthritis Disease Activity.

Table 3. JSpADA responsiveness to change.

	Group (n)	Mean (SD)	Median (IQR)	SMD[a]	*P*
JSpADA	Stable (2)	−0.25 (2.47)	–	–	–
	Improved (5)	−1.60 (0.65)	–	–	–
	Worsened (3)	−0.33 (3.21)	–	0.55	0.57
	Improved (5)	–	−1.5 (−2.0 to −1.0)	–	–
	Worsened (3)	–	−0.25 (−2.00 to 1.50)	–	0.55*
JSpADA7-no Schober	Stable (7)	−0.36 (1.18)	–	–	–
	Improved (34)	−1.43 (1.35)	–	–	–
	Worsened (7)	0.36 (1.75)	–	1.15	0.004
JSpADA7-no CRP/ESR	Stable (2)	0.50 (0.71)	–	–	–
	Improved (12)	−1.25 (0.92)	–	–	–
	Worsened (6)	0.25 (2.07)	–	0.94	0.14
	Improved (12)	–	−1.00 (−1.75 to −0.75)	–	–
	Worsened (6)	–	0.5 (0.0 to 1.0)	–	0.048*
JSpADA6-no Schober, no CRP/ESR	Stable (17)	−0.09 (0.69)	–	–	–
	Improved (75)	−1.03 (1.17)	–	–	–
	Worsened (26)	0.67 (1.23)	–	1.42	< 0.001

[a] SMD was calculated between improved and worsened group. *t* test was used to compare JSpADA score between those that improved and those that worsened. Analysis was limited to subjects who had complete data for the original JSpADA. *Wilcoxon-Mann-Whitney *U* test. CRP: C-reactive protein; ESR: erythrocyte sedimentation rate; JSpADA: Juvenile Spondyloarthritis Disease Activity; SMD: standardized mean difference.

showed high correlation with cJADAS10, moderate-to-high correlation with PGA, and low-to-fair correlation with CHAQ. This could be because of the close constructs in cJADAS10, PGA, and JSpADA, whereas CHAQ uses functional measures and has a floor effect, making it difficult to measure changes in status as subjects approach normal physical function.[16]

All versions of JSpADA had excellent discriminative validity. JSpADA and its modified versions showed medium to large effect size in determining responsiveness to change in disease activity over time.

We confirmed the findings by Zanwar et al that eliminating Schober does not affect the performance of the JSpADA score.[11] We further demonstrated that shortening the score to 6 items by eliminating both the Schober and CRP/ESR does not negatively affect the performance of the scoring system.

There are several limitations to this study. While the CARRA Registry includes all JSpADA measures, we had to exclude several visits because of missing data. Notably, greater than half of the excluded visits were for missing Schober (74.1%) and/or inflammatory markers (55.1%), likely because data elements were not collected clinically, particularly if the patient was doing well. This resulted in a subcohort with complete data who were clinically different from the ERA subjects with incomplete data. The subjects with complete data had higher percentage of clinical

sacroiliitis, and higher active joint count, cJADAS10, and PGA. This could have biased the results and may affect the generalizability of our findings. The 8 items in the original JSpADA score were generated by international consensus and remain an ideal disease activity measure for JSpA. However, the modified version with 6 elements may serve as an alternate measure especially when missing data are frequent.

JSpADA and its modified versions perform well as disease activity measures for ERA in the CARRA Registry. We propose the term *modified JSpADA* for the modification of JSpADA with 6 elements (JSpADA6-no Schober, no CRP/ESR). This shorter disease activity index may improve implementation of JSpADA in both clinical practice and research trials.

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ONLINE SUPPLEMENT

Supplementary material accompanies the online version of this article.

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