

Validation of the Childhood Health Assessment Questionnaire in the Juvenile Idiopathic Myopathies

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ABSTRACT. Objective. To examine the validity of the Childhood Health Assessment Questionnaire (CHAQ) in patients with juvenile idiopathic inflammatory myopathy (IIM).

Methods. One hundred fifteen patients were enrolled in a multicenter collaborative study, during which subjects were assessed twice, 7–9 months apart. Physical function was measured using the CHAQ. Internal reliability was assessed using adjusted item-total correlations and item endorsement rates. Construct validity was assessed by comparing predicted and actual correlations of the CHAQ with other measures of physical function and disease activity. Responsiveness was assessed by calculating effect size (ES) and standardized response mean (SRM) in a group of a priori defined “improvers.”

Results. Item-total correlations were high (r_s range = 0.35–0.81), suggesting all items were related to overall physical function. Manual muscle testing and the Childhood Myositis Assessment Scale correlated moderate to strongly with the CHAQ (r_s = –0.64 and –0.75, both $p < 0.001$). Moderate correlations were also seen with the physician global assessment of disease activity (r_s = 0.58, $p < 0.001$), parent global assessment of overall health (r_s = –0.65, $p < 0.001$), Steinbrocker function class (r_s = 0.69, $p < 0.001$), and global skin activity (r_s = 0.40, $p < 0.001$), while global disease damage and skin damage had low correlations (r_s = 0.13 and 0.07, $p \geq 0.17$). Responsiveness of the CHAQ was high, with ES = 1.05 and SRM = 1.20.

Conclusion. In this large cohort of patients with juvenile IIM, the CHAQ exhibited internal reliability, construct validity, and strong responsiveness. We conclude that the CHAQ is a valid measure of physical function in juvenile IIM, appropriate for use in therapeutic trials, and potentially in the clinical care of these patients. (J Rheumatol 2001;28:1106–11)

Key Indexing Terms:

JUVENILE IDIOPATHIC INFLAMMATORY MYOPATHY JUVENILE DERMATOMYOSITIS
OUTCOME ASSESSMENT PHYSICAL FUNCTION ASSESSMENT
VALIDATION MEASUREMENT

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Supported by funds from the intramural research programs of the Center for Biologics Evaluation and Research, FDA, to Drs. Miller, Lachenbruch, and Rider; and from the NIAMS, and the Clinical Center, NIH, to Drs. Miller, Hicks, and Rider. Dr. Feldman is a recipient of a Career Scientist Award from the Ontario Ministry of Health. Dr. Huber is supported by a Hospital For Sick Children Graduate Scholarship at the University of Toronto.

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Submitted June 7, 2000 revision accepted November 11, 2000.

The juvenile idiopathic inflammatory myopathies (IIM) are a group of rare, chronic illnesses of children, characterized by inflammation of muscle, as well as of skin and other organs. Juvenile dermatomyositis (DM) is the most common of these disorders, but other distinct entities such as juvenile polymyositis (PM) and overlap myositis (myositis associated with another connective tissue disease) are also recognized¹. The cause of juvenile IIM is unknown, although both genetic and environmental influences are felt to be important¹.

Since the advent of corticosteroid therapy for juvenile IIM, there has been a dramatic decrease in mortality to < 3%². Despite this, juvenile IIM is still associated with considerable longterm morbidity and functional disability, as documented in a recent followup study of outcome in juvenile DM³. In that study, when physical function was assessed 3–14 years after diagnosis, 28% of subjects had some degree of disability and nearly 10% were moderate to severely disabled. This physical disability was the result of several mechanisms, including weakness, arthritis, calcinosis, pain, and joint flexion contractures. Physical function is therefore an important and relevant outcome in children with juvenile IIM.

To date, a validated tool to measure physical function in this population, either for use in therapeutic trials or clinical practice, has not been available. However, the Childhood Health Assessment Questionnaire (CHAQ)⁴, originally developed to measure physical function in children with arthritis, recently underwent preliminary validation in a small cohort of children with juvenile DM⁵. That study suggested that the CHAQ had good construct validity in juvenile DM and was responsive to clinically important change. However, it was limited by a small sample size, and by the fact that most patients were at the start of their disease course, which is usually a time of rapid clinical change. This may have led to an overestimation of responsiveness. Thus, that study must be considered preliminary only, and does not provide definitive data regarding the validity of the CHAQ in juvenile IIM.

We believe that further examination of the measurement properties of the CHAQ is justified because there is a key difference between the CHAQ and objective measures of muscle strength, like manual muscle testing (MMT)⁶, or observational measures of function and endurance, like the Childhood Myositis Assessment Scale (CMAS)⁷. The CHAQ is a parent or self-report questionnaire that reflects the parent's or subject's perceptions of his/her physical abilities or limitations. It therefore provides information not available with other measures, like MMT or the CMAS, and may have distinct advantages over these other measures.

The goal of our study was to validate the CHAQ in a large cohort of children with juvenile IIM. Based on our results, we conclude that the CHAQ is a valid measure of physical function in patients with juvenile IIM, and is appro-

priate for use in therapeutic trials, and potentially in the day to day clinical care of these patients.

MATERIALS AND METHODS

Population. The Juvenile Dermatomyositis Disease Activity Collaborative Study Group conducted a multicenter study to develop and validate disease activity measures in juvenile IIM⁸. Consecutive subjects who met "probable" or "definite" criteria for DM or PM, as described by Bohan and Peter⁹, and who were < 18 years old at the time of diagnosis were enrolled between July 1994 and March 1997 at each of the 11 participating centers (Appendix). The characteristics of these patients are summarized in Table 1. Most subjects were studied > 1 year after disease onset, with a median disease duration at the first evaluation of 17 months. Most were female (68.7%) and Caucasian (73.9%).

Procedures. Approval from the institutional review boards of all participating centers was obtained. All subjects or their parents/legal guardians gave informed consent to participate in the study. Subjects were assessed at baseline (at any point in their disease course) and again 7–9 months later. At each assessment, a structured history, physical examination, laboratory investigations, as well as measures of disease activity and damage and physical function were recorded. Not all centers administered all measures. For parent or self-administered tools, such as the CHAQ, the parent/guardian usually completed the measure, except for 5 children > 10 years old, who completed the measure. Data from these 5 subjects were included to maximize the sample size.

Measures. The CHAQ is a 30 item parent or self-report questionnaire that examines physical function in 8 domains: dressing and grooming, arising,

Table 1. Characteristics of subjects at the time of their baseline assessment (n = 115).

Age at assessment, yrs, median (range)	8.9 (3–18.8)
Time since diagnosis, mo, median (range)	17 (1–137)
Sex, number (%) of subjects	
Female	79 (68.7)
Male	36 (31.3)
Ethnicity, number (%) of subjects	
Caucasian	85 (73.9)
African-American	12 (10.4)
Hispanic	7 (6.1)
Asian	3 (2.6)
Other*	8 (7.0)
Calcinosis, number (%) of subjects	24 (20.9)
Ulcerative disease course, number (%) of subjects	31 (27.0)
Clinical subset, number (%) of subjects	
Juvenile dermatomyositis	105 (91.3)
Juvenile polymyositis	6 (5.2)
Overlap myositis	4 (3.5)
Disease course, number (%) of subjects	
Chronic continuous	28 (24.3)
Chronic polycyclic	19 (16.5)
Monocyclic	17 (14.8)
Undefined [followed < 2 years] (%)	51 (44.4)
Muscle enzymes [proportion (%) of subjects with test value > upper limit of normal at first assessment]	
Creatinine kinase	21/112 (18.8)
Lactic acid dehydrogenase	41/80 (51.3)
Aspartate aminotransferase	36/108 (33.3)
Alanine aminotransferase	20/101 (19.8)
Aldolase	37/84 (44.0)

*4 Caucasian/Hispanic, 1 African-American/Hispanic, 1 Native American/Hispanic, 2 unknown.

eating, walking, hygiene, reach, grip, and activities⁴. Each domain has 2–5 questions, each describing an activity (e.g., Is your child able to stand up from a low chair or floor?). Potential responses range from 0 (able to do without any difficulty) to 3 (unable to do). A “not applicable” option is also available for patients not developmentally able to perform an activity. Each domain is scored as the highest item in that domain. If aids or assistance are required for an activity, the minimum score is 2 for the corresponding domain (i.e., if a child could perform an activity without difficulty, but only using an aid such as a cane, that domain would have a score of 2). The final score is the average of the answered domains, and ranges from 0 (no or minimal physical dysfunction) to 3 (very severe physical dysfunction).

Manual muscle testing (MMT)⁶ was done at 4 centers. Subjects > 4 years of age were tested by a single pediatric physical therapist or physiatrist at each center. Seven proximal and 5 distal muscle groups were assessed bilaterally, as well as 2 axial muscle groups. Assessors used either a 10 point Kendall¹⁰ or an expanded 5 point Medical Research Council¹¹ scale, with “plus” and “minus” subgrades for each level assigned based on clinical experience. To allow comparison between centers, we converted both scales to the expanded 5 point scale¹⁰. We then converted the scores to a continuous, adjusted total MMT score by adding the scores for each muscle group assessed, dividing by the maximum possible for that patient, depending on the number of muscles assessed, and multiplying by 100 to obtain a score between 0 (no strength) and 100 (normal strength). Several subscales of the total MMT score were also created using the same methods, but only including muscle groups relevant to that subscale (proximal, distal, axial, upper extremity, lower extremity).

The Childhood Myositis Assessment Scale (CMAS) is a 14 activity observational, performance based assessment of physical function, strength, and endurance⁷. The raw score ranges from 0 (very poor physical function) to 52 (normal physical function). Examples of activities that are assessed include situps, head elevation while in the supine position, and rising from a seated position. The CMAS was administered by a single pediatric physical therapist at one center, and by the attending pediatric rheumatologist at the remaining 9 centers. As with MMT, we standardized the scores by adding the scores for each item, dividing by the maximum possible for that patient, depending on the number of activities assessed, and multiplying by 100 to obtain a score between 0 (very poor physical function) and 100 (normal physical function). This gave the adjusted CMAS score.

Physician global assessments of disease activity, disease damage, skin activity, and skin damage were obtained using 10 cm visual analog scales (VAS), anchored by appropriate terms, and using standardized definitions⁸. Subject/parent global assessments of overall health, disease activity, and muscle symptoms were recorded using 10 cm VAS, anchored by relevant terms. This type of measure has been shown by our group to correlate well with physician assessments⁸.

Functional capacity was determined by the subject’s rheumatologist, using the Steinbrocker classification¹². To allow for comparison of laboratory investigations performed at different centers, we standardized all results by dividing by the upper limit of normal for the laboratory in which the test was performed. “Ulcerative disease” was defined as those subjects who, at any point of their disease, had gastrointestinal or cutaneous ulceration¹³. To determine disease course, subjects who had a disease duration of < 2 years had an “undefined disease course.” Those with a disease duration > 2 years were classified as follows: “monocyclic” if they had a full recovery within 2 years without relapse regardless of drug therapy, “chronic polycyclic” if they had a prolonged relapsing course with one or more relapses occurring between periods of inactive disease, and “chronic continuous” if they had persistent disease for longer than 2 years, which was never inactive despite drug therapy (after Spencer, *et al*¹⁴).

Analysis. The population was described using nonparametric statistics, as most variables were not normally distributed. We then examined the validity of the CHAQ in juvenile IIM in 3 ways. First, to assess internal reliability, we calculated adjusted item- and domain-total correlations as well as item and domain endorsement rates. Second, because there is no

gold standard of physical function against which to compare the CHAQ, we assessed construct validity of the CHAQ by comparing predicted and actual correlations of the CHAQ with other measures. Finally, we calculated responsiveness statistics (also called sensitivity to change) for the CHAQ. The details of these analyses are given below.

Internal reliability. We calculated item-total and domain-total correlations using Spearman’s correlation coefficient. Adjusted correlations were used to avoid inflating the correlations. The adjusted item-total correlation for the first item was determined by calculating the correlation of the first item with a rescored CHAQ that was calculated with the first item deleted. We repeated this procedure for all items and all domains. We then determined endorsement rates by calculating the number and percentage of subjects who gave a score > 0 for each item and domain. When calculating the item endorsement rates, subjects were excluded if the item in question was not developmentally applicable.

Construct validity. To test construct validity, we made a number of predictions about the correlations of the CHAQ with other measures. These predictions were made a priori, and were not influenced by knowledge of the data. Spearman’s correlation coefficients were used as the data were not normally distributed. Predictions were as follows: (1) The CHAQ would exhibit moderate correlations (defined a priori as 0.4–0.7) with the physician global assessment of disease activity, parent/patient global assessments of overall health, illness severity and muscle symptoms, the adjusted total MMT, and the adjusted CMAS. (2) The adjusted total MMT and adjusted CMAS would correlate more highly with the CHAQ than the global assessments, given that the MMT and CMAS measure similar things. (3) Correlations of the CHAQ with skin activity, Steinbrocker functional class, and MMT subscales would be moderate. (4) Correlations of the CHAQ with both skin and disease damage would be low (given that this population had relatively little damage to contribute to impaired physical function). (5) Correlations of the CHAQ with muscle enzymes would be low.

Responsiveness. To examine responsiveness, we calculated the effect size (ES) and standardized response mean (SRM), as described by Liang¹⁵. We defined a group of “improvers” as the population having > 3 cm improvement in the physician global assessment of disease activity. This was the primary analysis, but the analysis was repeated using a smaller improvement of 1 cm, which was felt to reflect the minimum difference that would be clinically relevant. For comparison, the analysis was also repeated for those subjects who failed to meet the 3 cm and 1 cm criteria for improvement. We chose absolute criteria for improvement because in general, the cohort was relatively mildly affected. If a relative criterion had been used (e.g., 30% improvement), this would have included subjects with clinically insignificant changes. The ES was calculated as the mean change divided by the standard deviation (SD) of the baseline scores. The SRM was calculated as the mean change divided by the SD of the individual change scores. As described by Cohen¹⁶, a large ES was 0.8, moderate was 0.5, and small was 0.2. After Beaton¹⁷, similar values were used for the SRM.

RESULTS

Outcome measures. Outcome measures at both assessments are summarized in Table 2. Most subjects were relatively mildly affected by their disease during the study. However, for most measures, there were subjects who represent both extremes of the range of possible values. The only exception to this was the adjusted total manual muscle testing (MMT), which had a range of 58 to 100 (where 0 is no strength). The medians of all measures improved from the first to the second assessment. Outcomes were not statistically different for subgroups of the population, including those based on sex, ethnicity, clinical subset, disease course, age, or presence of ulcerative disease (data not shown).

Table 2. Summary of outcome measures at baseline and followup assessments.

Measure	Possible Score	n	Baseline		n	Followup	
			Median	Range		Median	Range
CHAQ	0–3	115	0.25	0–3	90	0	0–2.6
PGA of disease activity	0–10	115	2.1	0–9.7	92	0.55	0–8.3
PGA of disease damage	0–10	115	0.5	0–10	92	0.4	0–8.2
Patient/parent global assessment of health	0–10	114	9.0	0.5–10	90	9.5	1.0–10
Patient/parent global assessment of illness severity	0–10	114	1.5	0–10	90	0.6	0–9.6
Patient/parent global assessment of muscle symptoms	0–10	113	1.3	0–9.7	90	0.6	0–9.6
Adjusted total MMT	0–100	57	91	58–100	42	93	68–100
Adjusted CMAS	0–100	115	97	0–100	90	100	25–100
PGA of skin activity	0–10	113	1.6	0–10	91	0.3	0–9.3
PGA of skin damage	0–10	111	0.5	0–10	90	0.1	0–7.9
Steinbrocker functional class							
I		57			63		
II		38			23		
III		15			5		
IV		5			1		

Internal reliability. In general, individual items of the CHAQ correlated with the total score (range of Spearman's $r = 0.35$ – 0.81 , mean = 0.66) and all correlations were statistically significant ($p < 0.0001$). Only 4 items, “write or scribble with a pen or pencil,” “lift a cup or glass to mouth,” “turn faucets on and off,” and “brush teeth,” had adjusted item-total correlations ≤ 0.50 . Each domain of the CHAQ also correlated well with the total CHAQ score (range of Spearman's $r = 0.59$ – 0.84 , mean = 0.77 , all $p < 0.0001$).

Individual items of the CHAQ received a score > 0 from 7–46% (mean 24.5%) of our subjects. Nine items were endorsed by $> 30\%$ of the cohort, 20 items were endorsed by more than 20% of the cohort, and only 2 items (“write or scribble with a pen or pencil” and “lift a cup or glass to mouth”) were endorsed by $< 10\%$. The endorsement rates were even higher for the domains of the CHAQ; all domains of the CHAQ were given a score > 0 by more than 25% (mean 37.5%, range 25.2–45%) of our subjects. Two domains, “activities” and “arising,” were the most highly endorsed (both by 45% of patients).

Construct validity. Spearman's correlation coefficients, as well as the 95% confidence intervals, used to assess construct validity are presented in Table 3. As predicted, we observed moderate correlations (0.4 – 0.7) between the CHAQ and the physician global assessment of disease activity, the patient/parent global assessment of overall health, illness severity and muscle symptoms, and the adjusted total MMT (prediction 1). The adjusted CMAS, an observational measure of physical function, was more strongly correlated (> 0.7) with the CHAQ. The correlations of the adjusted total MMT and the adjusted CMAS were similar to those for the other outcome measures, with confidence intervals largely overlapping (prediction 2). Moderate correlations were obtained for the physician assessment of skin activity and the Steinbrocker functional class (predic-

tion 3). Physician assessments of disease damage and skin damage were not significantly correlated with the CHAQ (prediction 4). Correlations of the CHAQ score with serum muscle enzymes were also low (prediction 5).

Responsiveness. Responsiveness statistics calculated using the population with a 3 cm improvement in physician global assessment of disease activity were considered the primary responsiveness outcome. Ninety subjects had CHAQ scores available at both assessments, 18 of whom met improvement criteria. In those subjects, the ES was 1.05 and the SRM was 1.20, both of which represent strong responsiveness. In the 72 subjects who did not improve by 3 cm, the ES was 0.20 and the SRM was 0.32. We repeated these calculations using a 1 cm improvement in physician global assessment of disease activity. Forty-four subjects met this criterion, with ES = 0.67 and SRM = 0.87 . Even with this much less stringent criterion for improvement, the CHAQ still showed responsiveness in the moderate to strong range. The 46 subjects who failed to improve by 1 cm had ES = 0.05 and SRM = 0.07 .

DISCUSSION

We have shown that the CHAQ, when used in juvenile idiopathic inflammatory myopathy (JIM), exhibits both internal reliability and construct validity. We have also shown that the CHAQ is strongly responsive in this population. Based on these results, we conclude that the CHAQ is a valid measure of physical function in children with juvenile JIM.

The CHAQ has undergone preliminary validation in a small, single center study of 37 children, most with newly diagnosed juvenile DM⁵. The current study is consistent with these previous results, but provides more definitive evidence of the validity of the CHAQ in juvenile JIM for several reasons. We studied a multicenter cohort that was

Table 3. Correlation of the CHAQ score with other potential measures of outcome used to examine construct validity of the CHAQ. Data are derived from the first assessment only. All values are Spearman correlation coefficients. All p values are < 0.0001, except those indicated in brackets, and are not adjusted for multiple comparisons.

Outcome Measure	n	Correlation	95% Confidence Interval
PGA of disease activity	115	0.58	0.43, 0.73
PGA of disease damage	115	0.13 (p = 0.17)	-0.05, 0.31
Patient/parent global assessment of health	114	-0.65 [†]	-0.79, -0.51
Patient/parent global assessment of illness severity	114	0.64	0.50, 0.78
Patient/parent global assessment of muscle symptoms	113	0.69	0.56, 0.82
Adjusted total MMT score	57	-0.64 [†]	-0.84, -0.44
Proximal	57	-0.60 [†]	-0.81, -0.39
Distal	57	-0.53 [†]	-0.75, -0.31
Axial	57	-0.51 [†]	-0.74, -0.28
Upper extremity	57	-0.62 [†]	-0.88, -0.41
Lower extremity	57	-0.60 [†]	-0.81, -0.39
Adjusted CMAS	115	-0.74 [†]	-0.86, -0.62
PGA of skin activity	113	0.40	0.23, 0.57
PGA of skin damage	111	0.07 (p = 0.44)	-0.12, 0.26
Steinbrocker functional class	115	0.69	0.56, 0.82
Creatine kinase	112	-0.02 (p = 0.78)	-0.21, 0.17
Lactic acid dehydrogenase	80	0.28 (p = 0.01)	0.07, 0.49
Aspartate aminotransferase	108	0.25 (p = 0.01)	0.07, 0.43
Alanine aminotransferase	101	0.14 (p = 0.16)	-0.06, 0.34
Aldolase	84	0.15 (p = 0.16)	-0.06, 0.36

[†]Negative correlations are expected for these values as subjects with greater degrees of disability have higher scores with the CHAQ, while these measures assign higher scores for normal health or physical function.
MMT: Manual muscle testing.

more than 3-fold larger, which allows us to be more certain of the relationships that we have described. We also studied a broader range of outcome measures, and demonstrated that the CHAQ correlates well with other measures of physical function, some of which have been specifically developed for use in juvenile IIM, with measures of disease activity and severity, and with subject's perceptions of their health and symptoms. This provides compelling evidence of the construct validity of the CHAQ in juvenile IIM. Furthermore, we studied subjects with a range of disease durations, which means our results should be applicable throughout the juvenile IIM disease course. This is particularly important for responsiveness, which may be overestimated when subjects are at the beginning of their illness, as in the previous validation study. Thus, the results from the current study provide new and convincing evidence of the validity of the CHAQ in juvenile IIM.

There are several other tools that could be used to measure physical function in juvenile IIM, including manual muscle testing, the Childhood Myositis Assessment Scale, and the Myositis Functional Index¹⁸. However, we feel that the CHAQ has an important advantage over these measures. Parents or patients complete the CHAQ based on their subjective perceptions of their child's/their disability. The CHAQ, therefore, reflects a broader range of experience than weakness or endurance alone. It may be affected

by other factors that may influence the ability to perform tasks, such as pain, fatigue, or cognitive or emotional disturbances. For this reason, a parent or self-report measure of physical function, like the CHAQ, may be the best way to assess patients with juvenile IIM, as laboratory or clinic based assessments of function may not adequately reflect the subject's ability to perform day to day activities.

The primary limitation of this study is that the CHAQ suffers from a considerable "floor effect." That is, as subjects approach normal physical function, it becomes difficult to measure changes in status, given that subjects are unable to have a score better than 0. This is a characteristic of the CHAQ, and is not limited to patients with juvenile IIM. We do not feel that the floor effect should discourage the use of the CHAQ in juvenile IIM because the CHAQ was still strongly responsive in this population. In the future, inclusion of items addressing advanced activities of daily living, as recently reported for the adult HAQ in patients with rheumatoid arthritis¹⁹, may be one approach to overcoming this problem.

We show that the CHAQ is a valid measure of physical function in juvenile IIM. We feel that it is an appropriate tool for research, and may be of potential value in the day to day clinical care of children with these illnesses. In the future, it may form a part of a composite measure of disease activity.

ACKNOWLEDGMENT

We thank Drs. Robert Sundel and Balu Athreya for participation in the Juvenile Dermatomyositis Activity Study Group, and Drs. Holly Cintas and Elizabeth Adams for critical review of the manuscript.

APPENDIX

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