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Functional consequences of haemophilia in adults: the development of the Haemophilia Activities List

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Summary. Several instruments can be used to evaluate the functional status of patients with haemophilia, but none of these instruments is specific for haemophilia. We developed a haemophilia-specific self-assessment questionnaire to evaluate and monitor a patient's perceived functional health status: the Haemophilia Activities List (HAL). In three separate but interlinked substudies, the questionnaire was constructed and tested for face, expert, and convergent validity, as well as internal consistency and patient-evaluated relevance. Items for the questionnaire were collected by interviewing 162 patients, using the McMaster-Toronto Arthritis Patient Preference Disability Questionnaire (MACTAR). The items were combined to generate the first version of the questionnaire [HAL(1)]. This version was evaluated and commented on by two focus groups (patients and caregivers), and then the questionnaire was adapted on the basis of these comments, forming the final version, HAL(2). This version was then validated in a pilot study with 50 consecutive patients using the Dutch Arthritis Impact Measurements Scales 2 (Dutch-AIMS2) and the Impact on Participation and Autonomy (IPA) questionnaires. The HAL(2) showed good convergent validity (Pearson correlation 0.80–0.91; $P < 0.01$), and the internal consistency was good for six of the eight domains (Cronbach's α 0.83–0.95). Patients considered the content of the HAL to be more relevant to their situation than the content of the other questionnaires ($P < 0.01$). Three major factors (upper extremity function, lower extremity function, key activities/major problem activities) were identified by factor analysis. The questionnaire seems to be a useful tool to identify problematic activities as part of the functional health status of patients with haemophilia. The construct validity, test–retest reliability, and responsiveness of the HAL will be established in the future.

Keywords: activities, functional health status, internal consistency, questionnaire, validity

Introduction

Haemophilia is a relatively rare, sex-linked inherited bleeding disorder resulting from a deficiency of clotting factor. Affected individuals lack factor VIII or IX procoagulant activity (<1% in severe haemophilia, 1–5% in moderate haemophilia and 6–25% in mild haemophilia) and consequently suffer from repeated bleeding, most frequently located in major joints (elbows, knees and ankles) and muscles. As a result, affected individuals have progressively worsening joint and muscle impairments that may seriously affect their daily activities. For this reason, it is important to monitor the individual patient as well as groups of patients and evaluate the long-term effects of concomitant treatment such as on-demand or prophylactic factor substitution and arthroplasty. A review by De Kleijn *et al.* [1] identified 34 clinimetric instruments used in haemophilia research that cover quality of life aspects and all the domains mentioned in the International Classification of Functioning Disability and Health (ICF [2]).

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However, most of these instruments are used infrequently, and none was specifically designed for patients with haemophilia.

The aim of this study was to develop a disease-specific instrument to evaluate the functional health status of patients with haemophilia. The Haemophilia Activities List (HAL) is a patient self-assessment questionnaire and can be used as part of a test battery to evaluate a patient's functional health status. Knowledge of a patient's specific problems and symptoms may make it possible to tailor therapy.

Methods

This study comprised three separate but interlinking substudies. First, questionnaire items were selected on the basis of replies to the McMaster-Toronto Arthritis Patient Preference Disability Questionnaire (MACTAR [3]) administered to patients with haemophilia. The items were categorized according to the ICF [2] and then appropriate items were selected to form the first version of the HAL [HAL(1)]. This version was sent to two focus groups for evaluation. The two groups consisted of patients with haemophilia and physical therapists involved in haemophilia care. Both patients and physical therapists evaluated the contents and style of the HAL(1), using a structured evaluation form, and the patients were invited to participate in a focus group meeting to further discuss the evaluation. The HAL(1) was adapted on the basis of these comments to form the second, and final version, HAL(2). This questionnaire was then used in a pilot study to evaluate the clinical applicability of the HAL(2) and its convergent validity and internal consistency. Fifty patients with haemophilia took part in this study.

Methods 1: Development of the questionnaire

A physical therapist administered the MACTAR [3], which is routinely used in our clinic, to consecutive patients visiting the clinic for their regular medical checkup. During this semi-structured interview, each patient was asked to identify a maximum of 10 problematic activities that he would like to be able to perform without pain and/or difficulty, and then to rank the five most important. These activities were then categorized according to the ICF [2]. Item collection was discontinued when no new activities were mentioned by subsequent patients (saturation point).

Categorized items were included in the questionnaire when they were considered 'relevant' by two independent researchers. Items were 'relevant' when they related to the 'Activities' domain mentioned in the ICF ('d'-categories), but not to 'Participation' (such as work-related items). Moreover, activities that were categorized in a 'other specified' or 'unspecified' ICF category were excluded. The remaining items were classified in common domains, mostly based on their position in the ICF. All questions were formulated in a standard format: 'In the previous month, did you encounter any difficulty due to haemophilia with…'. The response options were 'Always', 'Mostly, 'Sometimes', 'Rarely', and 'Never', meaning a 5-point Likert scale [similar to the scoring-method of the Dutch Arthritis Impact Measurements Scales 2 (Dutch-AIMS2)] [4,5].

Results 1

In total 162 male patients (mean age ± SD, 40.8 ± 13.5 years; range: 18–76) with haemophilia participated in this substudy. Most had haemophilia A or B (140 and 16, respectively) and none had acute pathology (e.g. bleeding). Patient characteristics are given in Table 1.

A total of 622 ranked/prioritized activities were mentioned, which were grouped into 66 categories, from which 53 were selected. Items such as 'Pain in a body part' (mentioned by 19 patients; in the ICF 'Function' class) and 'Seeking employment' (in the ICF 'Participation' class) were excluded. The remaining items were divided into eight different domains, each representing an aspect of daily activities. Thus, the first version of the HAL covered the following domains: 'lying down/sitting/kneeling/standing'

Table 1. Patient characteristics.

	Severity*			
Disease	Severe	Moderate	Mild	Total
Item collection [age (years) – mean, SD (range)]: 40.8, 13.5 (18–76)				
Type A	99	15	26	140 (86.4%)
Type B	15	–	1	16 (9.9%)
Type B – Leyden	–	–	1	1 (0.6%)
von Willebrand	4	–	–	4 (2.5%)
Factor VII	–	1	–	1 (0.6%)
Total	118	16	28	162
Validity and patient preference assessment [age (years) – mean, SD (range)]: 44.9, 14.2 (18–70)				
Type A	37	2	3	42 (84.0%)
Type B	6	–	2	8 (16.0%)
Total	43	2	5	50

*Severity: severe, <1% clotting activity; moderate, 1–5% clotting activity; mild, 6–25% clotting activity.

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(9-items; e.g. 'Sitting down' and 'Kneeling/squatting'); 'functions of the legs' (9-items; e.g. 'Walking short distances' and 'Climbing up the stairs'); 'functions of the arms' (4-items; e.g. 'Lifting heavy objects' and 'Reaching above your head'); 'use of transportation' (4-items; e.g. 'Driving a car' and 'Using public transportation'); 'self-care' (10-items; e.g. 'Washing your hair' and Putting on socks and shoes'); 'household tasks' (8-items; e.g. 'Cleaning the house' and 'Preparing a meal'); 'leisure activities and sport' [6-items; e.g. 'Sports' and 'Going out (theatre/museum/movie theatre/bar)']; and 'other' (3-items; e.g. 'Writing').

Methods 2: Focus groups

The first version of the HAL was evaluated by two focus groups, consisting of patients with haemophilia ($n = 7$) and international physical therapists involved in the care of patients with haemophilia ($n = 16$). The patients received a Dutch version of the questionnaire whereas the physical therapists received an English version. The participants were asked to indicate whether they thought individual items should be included in the questionnaire and to rate the clarity of the description of each item. They were also asked to assess the style of the questionnaire regarding font size, number of items and pages, the arrangement of the items in the aforementioned eight domains, and the sequence of the domains and the items in each domain. The patients were asked to participate in discussion about the questionnaire. The HAL(1) was adapted according to the results of this evaluation.

Results 2

On a scale from 1 to 10 (1 = very bad; 10 = very good), the content and style of the HAL(1) were scored 8.0 (± 0.7) and 8.2 (± 0.4) by the patients and 7.7 (± 1.0) and 8.1 (± 0.8) by the physical therapists. It took the patients on average 7 min to complete the questionnaire. Thirteen physical therapists (81%) indicated that they wanted to use the HAL(1) in daily practice to evaluate the functional health status of their patients with haemophilia.

On the basis of the findings and the discussion, 4-items were removed and replaced by four new items (all of which were mentioned by several patients and physical therapists); four other items were split; and 7-items needed further clarification. Two additional questions relating to adaptations using the car and the use of a walking aid (e.g. cane) were added. An additional response option 'impossible' was added for all items, and the option 'not applicable' was added for 12-items. A scoring system was developed to quantify the functional health status of each respondent (scores for individual domains and sum scores). The second version of the HAL [HAL(2)] consisted of 57-items in eight domains.

Methods 3: Pilot study

Next, the convergent validity and the internal consistency of the HAL(2) were evaluated. The Medical Ethics Committee of the University Medical Centre Utrecht approved the study. Fifty consecutive patients with haemophilia were invited to participate during the annual medical checkup at the Van Creveldkliniek. They all gave their written informed consent to participation.

The participants were asked to complete the HAL(2). Two other questionnaires were administered to evaluate physical, psychological, and social aspects (Dutch-AIMS2 [4, 5]), and personal impact of illness on participation and autonomy and related experience of problems (the Impact on Participation and Autonomy questionnaire; IPA [6–9]). The three questionnaires were not administered in a specific order.

The Dutch-AIMS2 was originally designed for use in patients with rheumatoid arthritis [10] and evaluates physical, psychological, and social aspects of patients, but it has been used in patients with haemophilia [4, 5]. This self-administered questionnaire comprises 81-items, distributed over 12 health status scales (mobility level; walking and bending; hand and finger function; arm function; self-care; household tasks; social activities; support from family and friends; arthritis pain; work; level of tension; and mood) and five components (physical, affect, symptoms, social/interaction and role). We were mainly interested in the first six health status scales and the physical component. The questionnaire takes about 20 min to complete, and the response to each item is scored on a 5-point Likert scale. The component 'symptoms' was adjusted, to ask individuals about 'haemophilic' instead of 'rheumatic' pain [4]. Moreover, five questions (1, 6, 7, 8 and 9) were split, because they represented two very different activities and thus needed to be assessed separately (e.g. question 7: 'Walking several blocks or climbing a few stairs'). Additionally, one question was added which was related to the time the patient needed to adjust to his morning/starting stiffness ('How long does it take you to get going after lying down or sitting?').

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The IPA is a generic questionnaire addressing the personal impact of illness on participation and autonomy and related experience of problems [6, 8]. The self-administered questionnaire consists of five domains: autonomy indoors (7-items, e.g. getting around at home, self-care activities when one wants), family role (7-items, e.g. looking after the home, economic self-sufficiency), autonomy outdoors (5-items, e.g. visiting friends, spending leisure time the way one wants), social activities (6-items, e.g. quality of relationships, receiving respect), and work and educational opportunities (6-items, e.g. doing the job or education one wants). Perceived participation and problems are scored on a scale of 1–5 and 0–2, respectively. A higher score represents more restrictions in participation or worse problems [9].

To enable comparison, all scores were normalized to a score ranging from 0 (best functional health status) to 10 (worst functional health status). The formula used for all three questionnaire: $Score_{normalized} = (Score_{patient} - Score_{minimum}) \times [10/(Score_{max} - Score_{min})]$, where '$Score_{min}$' and '$Score_{max}$' are the lowest and highest possible score for a questionnaire, respectively.

After completing the questionnaires, all patients were asked to rate the individual questionnaires in terms of how the items related to their daily life, using a 1–10 scale (1 means very bad and 10 means very good). In this way, the relevance of each questionnaire to the patient's situation could be established [11].

Data were analysed with SPSS 10.1, using a significance level of $\alpha = 0.05$. First, all data were assessed for normal distribution using PP-plots, kurtosis/skewness analysis, and the Kolmogorov–Smirnoff test for normality. Descriptive statistics for all three questionnaires were calculated. The convergent validity of the HAL was determined by correlating the normalized sum scores for all three questionnaires, using Pearson's correlation coefficient. Two sum scores were calculated for the Dutch-AIMS2: the overall sum score and a 'physical' sum score, based on the first six health status scales of the Dutch-AIMS2 (i.e. 'Mobility Level', 'Walking and Bending', 'Hand and Finger Function', 'Arm Function', 'Self-care Tasks' and 'Household Tasks'). Thus, the HAL_{sum} was correlated with the IPA_{sum}, Dutch-$AIMS2_{sum}$ and Dutch-$AIMS2_{phys-sum}$.

The internal consistency of the HAL(2) was determined by calculating Cronbach's α for the whole questionnaire, as well as for each separate domain of the HAL(2). Differences in patient preference were calculated using a randomized block design (repeated measure analysis).

Explorative factor analysis with Varimax rotation was used to perform a preliminary analysis of the construct validity of the HAL(2) and to detect possible underlying constructs. Only factors with an eigenvalue of 1.0 or higher were considered for analysis and separate items were required to have a loading of at least 0.4 on a factor.

Results 3

Fifty patients who were not involved in the other substudies participated in this pilot study (see Table 1). All patients were males, 42 had haemophilia A, and eight haemophilia B. Forty-three patients had severe haemophilia (<1% clotting activity), two moderate haemophilia (1–5% clotting activity) and five mild haemophilia (6–25% clotting activity). Their average age was 44.9 ± 14.2 years (range: 18–70). None of the patients had acute pathology (e.g. bleeding) or recent bleedings.

All sum scores (both raw and normalized) were normally distributed. It took the patients on average 10 min (95% CI: 8–13; range: 3–45) to complete the HAL(2). The scores for each instrument are shown in Table 2. The average normalized sum score was 3.2 (SD: 1.8) for the HAL(2), 2.8 (SD: 1.4) for the overall Dutch-AIMS2, 2.4 (SD: 1.6) for the physical score of the Dutch-AIMS2 and 2.8 (SD: 1.8) for the IPA.

Both the raw and the normalized scores of the three questionnaires were correlated to assess the convergent validity of the HAL(2). Although all raw scores for the three questionnaires differed significantly (paired Student's *t*-test: $P < 0.01$), the raw sum scores of the HAL(2) correlated highly with the raw scores of both the Dutch-AIMS2 (0.89; $P < 0.01$) and the IPA (0.75; $P < 0.01$). The normalized sum scores showed similar correlations; the HAL(2) and the Dutch-AIMS2 were highly correlated (0.90; $P < 0.01$), whereas the correlation coefficient for the HAL(2) and the IPA was 0.80 ($P < 0.01$) (see Table 3). The internal consistency of the HAL was established by means of the Cronbach's α statistic. Values ranged from 0.43 (domain 'Other') to 0.95 (domain 'Functions of the legs'). For the overall questionnaire, Cronbach's α was 0.96. Table 4 shows the internal consistency for all domains of the HAL.

Patients considered that the HAL(2) better reflected the problems they experienced in daily life than did either of the other two questionnaires. On a

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Table 2. Sum scores and separate domain scores for each of the questionnaires.

			Raw scores		Normalized scores	
	n	Range	Mean (SD)	Range	Mean (SD)	Range
HAL						
Sum score (HAL_{sum})	57	57–342	137.9 (45.8)	58–279	3.2 (1.8)	0.1–7.8
Dutch-AIMS2						
Sum score (Dutch-$AIMS2_{sum}$)	77	81–383	155.2 (39.5)	97–276	2.8 (1.4)	0.7–6.7
Physical sum score (Dutch-$AIMS2_{phys–sum}$)	33	33–165	63.9 (21.1)	35–134	2.4 (1.6)	0.2–7.7
IPA						
Sum score (IPA_{sum})	31	0–124	31.2 (19.5)	0–77	2.8 (1.8)	0–6.8

HAL, Haemophilia Activities List; Dutch-AIMS2, Dutch Arthritis Impact Measurements Scales 2; IPA, Impact on Participation and Autonomy questionnaire.

Table 3. Convergent validity.

	Paired *t*-test		Pearson correlation	
	Mean difference	*P*-value	Correlation	*P*-value
Raw scores				
HAL_{sum} vs. Dutch-$AIMS2_{phys–sum}$	74.2	<0.001	0.88	<0.001
HAL_{sum} vs. Dutch-$AIMS2_{sum}$	−18.3	<0.001	0.89	<0.001
HAL_{sum} vs. IPA_{sum}	106.8	<0.001	0.75	<0.001
Normalized scores				
HAL_{sum} vs. Dutch-$AIMS2_{phys–sum}$	0.82	<0.001	0.91	<0.001
HAL_{sum} vs. Dutch-$AIMS2_{sum}$	0.38	0.003	0.91	<0.001
HAL_{sum} vs. IPA_{sum}	0.41	0.019	0.80	<0.001

HAL, Haemophilia Activities List; Dutch-AIMS2, Dutch Arthritis Impact Measurements Scales 2; IPA, Impact on Participation and Autonomy questionnaire.

Table 4. Internal consistency of the Haemophilia Activities List (HAL).

	Items (*n*)	Cases (*n*)	Cronbach's α
HAL overall	57	44	0.96
HAL domain			
Lying/sitting/kneeling/standing	11	48	0.91
Functions of the legs	9	49	0.95
Functions of the arms	5	50	0.89
Use of transportation	4	50	0.63
Self-care	11	49	0.94
Household tasks	7	50	0.90
Leisure activities and sport	8	47	0.83
Other	2	50	0.44

10-point scale, patients awarded the HAL(2) a score of 7.4 (SD: 0.9; range: 5.0–9.0), the Dutch-AIMS2 a score of 6.7 (SD: 1.1; range: 4.0–8.0), and the IPA a score of 6.5 (SD: 1.3; range: 2.0–8.5). The score for the HAL(2) was significantly higher than that for the other two questionnaires ($P = 0.003$).

Explorative factor analysis with Varimax rotation revealed eight factors (eigenvalues >1.0) that together accounted for 77% of the total variance. Three major factors (>10% variance explained) were identified (44% VAR_{expl}): 'upper extremity function' (19.6% VAR_{expl}), 'lower extremity function' (13.6% VAR_{expl}), 'key activities/major problem activities' (11.3% VAR_{expl}).

Discussion

The aim of this study was to develop a disease-specific instrument to evaluate problematic activities as a component of the functional health status of patients with haemophilia and to assess the validity and internal consistency of this instrument. Because the questionnaire was based on items mentioned by 162 patients, it has face validity. The expert validity of the questionnaire was established by asking patients with haemophilia and caregivers to evaluate the content of the HAL. Finally, the convergent validity of the HAL was established by its high correlation with the Dutch-AIMS2 0.91 ($P < 0.01$) and the IPA 0.80 ($P < 0.01$) (Fig. 1). Two domains of the HAL had a low internal consistency, namely, 'use of transportation' and 'other' (0.63 and 0.43, respectively), probably because of the limited number of items (4 and 2, respectively [12]) and the

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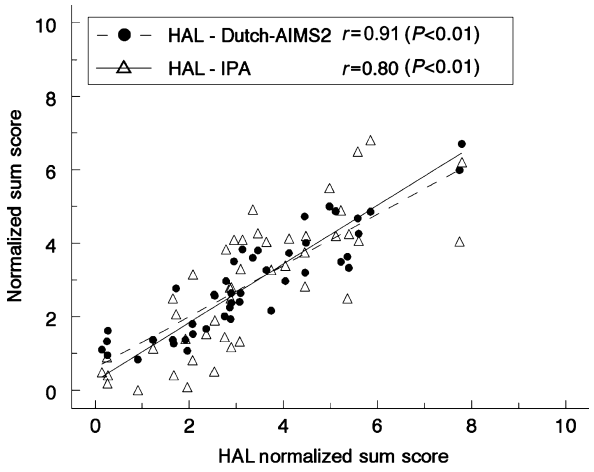

Fig. 1. Convergent validity of the Haemophilia Activities List (HAL) vs. the Dutch Arthritis Impact Measurements Scales 2 (Dutch-AIMS2) and the Impact on Participation and Autonomy questionnaire (IPA).

diversity of the items in these domains. Because of the high internal consistency of the other six domains, removal of one or more items from the questionnaire can be considered. Three major underlying components were identified, two of which clearly relate to activities relating to specific body regions (upper and lower extremities, respectively). Further research to address the implications of this finding is needed; in factor analysis a minimum of 10–15 cases per factor is needed, which means a minimum of 80 patients is required. The HAL proved to have a good clinical applicability in terms of time needed to complete it (approximately 10 min), and 81% of the physical therapists in the focus group indicated that they would like to use the HAL in daily practice. Moreover, the patients who participated in the pilot study indicated that the questions of the HAL reflected their own personal situation better than did the questions of the Dutch-AIMS2 and the IPA. Most of the patients included in the pilot study had severe haemophilia A, and it is to be expected that such patients experience the most problems in daily life. Thus, it has to be ascertained whether the HAL can be used for patients with mild or moderate haemophilia. Moreover, because of the small number of patients ($n = 50$), the results of the explorative factor analysis should be interpreted with caution and the conclusions may not be generalizable to other populations.

In an earlier study, Van Meeteren *et al.* [5] interviewed 67 patients using the Canadian Occupational Performance Measure (COPM [13]), an instrument that is closely related to the MACTAR in that patients are also allowed to mention up to five problematic activities. Although the COPM covered similar 'activities' as the MACTAR, it identified problems that were not covered by the MACTAR, namely, 'Working in a paid job/volunteering', 'Functioning in a job', 'Impairments' and 'Sleeping'. This is because during the MACTAR interview, the interviewer solely focused on 'Activities', whereas 'Working in a paid job/volunteering', 'Functioning in a job' belong to the 'Participation' domain, and 'Impairments' should be classified as such and were therefore not recorded. Nevertheless, 19 patients (12%) mentioned 'starting pain' as one of their main complaints. This item falls under the ICF as 'b2801 Pain in a body part' [2], and was therefore omitted in the activities list. Nevertheless, pain is an important item for patients with haemophilia and measuring its intensity and duration is of clinical importance [14]. An instrument to assess pain (duration, intensity) is an essential component of a future core-set of instruments to evaluate health status in patients with haemophilia.

Although the applicability of the IPA to patients with haemophilia has not yet been tested, it has been validated in very diverse patient populations, including rheumatoid arthritis and osteoarthritis, two diseases closely related to haemophilia [15]. Thus, the use of the IPA to validate the HAL would seem appropriate although the IPA and the HAL measure different ICF domains (Activities and Participation, respectively). Although several aspects of the validity of the HAL were addressed in this study, some psychometric properties, including the test–retest reliability and the responsiveness, were not. It is especially important to establish the responsiveness of the HAL, in order to use the instrument to evaluate interventions (e.g. joint surgery) that aim to increase the functional health status of patients with haemophilia. In order to obtain comprehensive insight into a patient's functional health status, it is important that caregivers are aware of the patient's subjective and actual abilities. Indeed, the importance of measuring components of functional health status, and 'Activities' in particular, seems to be underestimated in haemophilia care. During the XIXth Congress of the International Society on Thrombosis and Haemostasis, health-related quality of life (HRQoL) was addressed as an emerging principal measure of health outcome [16]. Szende *et al.* stated that HRQoL is broader than the aspects evaluated in currently used clinical instruments because of its multidimensional nature [17]. HRQoL is a cumulative outcome measure, and without insight into

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the problems that give rise to a diminished HRQoL, it is not possible to target interventions. For this reason, self-rating and performance instruments are needed [18]. The HAL could be included in a core-set of instruments for use in haemophilia [1], but only if complemented with one or more performance instruments. Thus, future research should focus on larger patient groups with a greater diversity of disease severity and measures should include both self-rating and performance instruments.

Conclusion and recommendations

The HAL has both face and expert validity and a good convergent validity when compared with the Dutch-AIMS2 and the IPA. It has a high internal consistency and its items are considered by patients to reflect their problems better than the items of the other two questionnaires. The HAL is therefore a valuable instrument to evaluate the functional health status of patients with haemophilia. Its reliability and responsiveness will be assessed in a larger group of patients. We recommend that the HAL be used together with one or more performance-based activity instruments to fully assess a patient's functional health status.

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