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SPECIAL ARTICLE

ICF Core Sets: how to specify impairment and function in systemic lupus erythematosus

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The World Health Organization's International Classification of Function (ICF) is a tool to characterize and illuminate better the full of array of problems a patient faces when affected by disease. Specifying these problems is a particular challenge in a disease like systemic lupus erythematosus (SLE) because of the wide variety in organ systems involved, its variable activity and severity, and considerable ethnic and local differences. The authors of this manuscript believe, however, that a broader understanding will prove essential for optimal patient care, and that there is sufficient experience now in defining ICF Core Sets to successfully complete core sets for SLE. Therefore, we will embark on an international project for developing ICF Core Sets for SLE, which we here delineate. This development will include two versions: 1) The Brief ICF Core Set for SLE will be a very focused list of categories essential for SLE clinical trials; and 2) The Comprehensive ICF Core Set will be much broader and useful for guiding multidisciplinary assessment in patients with SLE. Both Core Sets will be developed in a formal decision-making and consensus process of health professionals integrating evidence gathered from preliminary studies. The final definition of the Core Sets will occur at a consensus conference which will integrate: i) a systematic review of the literature regarding the outcome measures used in clinical trials and selected observational studies; ii) focus groups or semi-structured interviews with SLE patients; iii) a Delphi exercise with world wide involvement of experts; and iv) the evidence from empirical studies. The development of these SLE ICF Core Sets is designed to be an inclusive, open, worldwide process. We therefore invite both SLE clinical experts and SLE patients to participate actively. *Lupus* (2006) **15**, 248–253.

Key words: impairment; quality of life; systemic lupus erythematosus; World Health Organization

Introduction

Comprehending the problems faced by patients with a given disease is an essential aspect of medical care, although always a challenge. Health professionals, in general, and physicians, in particular, tend to focus on a limited number of parameters that can be readily measured. While this approach is appropriate for determining organ involvement and disease activity as well as making therapeutic decisions, it can fall short when evaluating over-all patient well-being or disease-related cost. Exploring the perspective of patients is particularly important in a heterogeneous disease such as systemic lupus erythematosus (SLE), because individual patient problems may vary greatly in severity and duration. Thus, ideally, an outcome measurement should provide a comprehensive picture of the different problems experienced by the individual patient, and should not only focus on biomedical outcome measures.

Achieving such a comprehensive view of SLE patients' problems will be a difficult task. Why then should rheumatologists take on such challenge? Since, despite considerable work, such endeavour might produce a useful outcome, what is there to gain? There are

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several answers to these important questions, which, we think, will stand in the end. One of the answers comes from occupational therapy, where function and disability are the main focus of attention. While physicians uncommonly focus on disability in their professional approach, but rather on stopping disease from causing disability, occupational therapists are trained to mainly see this other side of the patient. By doing so, however, occupational therapists and other health professionals will need our effort to set the stage for optimal patient care. It is the physicians and nurses, on the one hand, and the patients, on the other, who know intimately about the manifestations of SLE, and can therefore link the available information into a meaningful framework.

In addition to providing the information for a solid background for all health professionals, a comprehensive definition of the problems faced by SLE patients will be a powerful tool for dealing with both politics and decisions by health authorities. While very important on an emotional level, the example of single cases can never substitute for such data. If we do not understand the variety of problems our patients face, there is little we can do about them on either a medical level or on a political one. Moreover, applying comprehensive models of functioning and health will be the base for a true assessment of the burden of disease. These comprehensive models will then allow comparisons between different diseases, and lead political decision makers to informed decisions with regard to the allocation of resources, laws, and care provision.

Finally, such a comprehensive understanding will hopefully lead to better tools for daily practice. Fatigue scales for example, have found their way into lupus trials and care,[1] although derived from outside our own field. It is important that neither fatigue nor quality of life measures were found associated with SLE disease activity in cross-sectional studies,[2] suggesting that important aspects of the lupus patient's disease burden – potentially including treatable conditions – are not responding to treatment despite intensive medical care for internal organ disease. Along similar lines, we may want to develop a lupus skin score showing the patient's view on disease-related problems. Such a score could become be a valuable tool for guiding therapeutic efforts to control skin manifestations of SLE or cutaneous lupus. After all, it is the impaired function of a patient in daily life, and the ensuing loss in life quality, that will influence patient decisions about accepting the risks or side effects for specific treatments.

These concepts pertain for practically any given disease, and the World Health Assembly has therefore responded by approving the new International Classification of Functioning (ICF),[3] the first universally accepted way to classify and describe functioning, disability and health in persons with a condition. Following the WHO definition of health, the ICF framework is based on the bio-psycho-social model. The ICF models daily functioning of patients with different health conditions within the framework of a classification. Thus, the ICF offers a frame of reference to describe the functioning of patients with a certain health condition.

Technically, the ICF classification has two parts, each consisting of separate components. Part 1 covers functioning and disability and includes the components *body functions* (b) and *structures* (s) as well as daily *activities and participation* (d). Part 2 covers the context, comprising *environmental* (e) and *personal factors*. Each component is divided into several chapters, which represent health domains. Examples are the chapter *neuromusculoskeletal and movement-related functions* within the component *body functions* and the chapter *self-care* within the component *activity and participation*. Each chapter then consists of categories, which are the factual units of the classification.[3]

In total, the ICF classification includes more than 1400 categories, thus limiting use in clinical practice. It is possible, however, to derive practical tools from the classification, which are called ICF Core Sets.[4,5] ICF Core Sets are short lists of selected ICF categories, which are useful for modelling the function of patients with a specific disease. To date, ICF Core Sets have been developed for 12 chronic conditions.[6,7] The one example in rheumatology is the ICF Core Set for rheumatoid arthritis.[8] Core Sets for several other chronic conditions are currently being developed.

It is important to reiterate that the ICF is a framework, and the ICF Core Sets are practical tools to classify and describe patient functioning, using a classification of world-wide acceptance. However, at this point, the categories included in ICF core sets do not define the basis of measurement. Instead, the ICF Core Sets can serve as reference framework when selecting instruments for measuring symptoms and consequences of the disease, or when defining the elements covered by specific measures. The ICF and the ICF Core Sets may become the base for further developing such measures. In this way, the ICF is an important concept and ICF Core Sets are interesting tools for the further development of clinical (and rehabilitation) science.

The development of ICF Core Sets for SLE poses considerable complexities and even obstacles. SLE is prototypically a heterogenous disease, perhaps even more so than other multisystem disorders. Not only is there a wide range of both disease activity and severity, but therapy varies markedly in intensity as well as toxicity, all of which can influence patient outcomes. SLE involves essentially all organ systems, including

the CNS, and may induce various functional problems as well as damage in the brain. The various possible CNS manifestations of the disease, and their treatment, will further add to the heterogeneity of problems faced by SLE patients. On the other hand, there are typical symptoms which will afflict most SLE patients, regardless of their main organ manifestations. Therefore, ICF Core Sets for SLE will need to address the full spectrum of problems encountered both with SLE in general, and with the most common forms of SLE organ involvement in particular. We expect that there will be overlapping core sets with other diseases afflicting the same organ system, such as between SLE with lupus arthritis and rheumatoid arthritis, or between SLE renal disease and other forms of glomerulonephritis.

The most commonly used measures in SLE are activity scores, such as (in alphabetical order) the British Isles Lupus Assessment Group index (BILAG),[9] the European Consensus Lupus Activity Measurement (ECLAM),[10] the SLE Index Score (SIS),[11] the Systemic Lupus Activity Measure (SLAM),[12] or the SLE Disease Activity Index (SLEDAI).[13] All of these scores contain information on a variety of symptoms experienced by the patient (Table 1), in addition to a variety of laboratory and other parameters.

Regarding these symptoms, there are important differences among the organ systems affected. For example, general, mucocutaneous, or neurological symptoms receive prominent attention, whereas other important organ manifestations are scarcely considered (Table 1). In part, this relative imbalance is due to a lack of symptoms, such as often is the case in early renal disease, although the activity measures account for such manifestation by using laboratory evaluations instead. Also, the symptoms graded here serve the purpose of indicating activity; they do not weigh damage or well-being, and some are quite uncommon features.

In addition to the activity scales, which correlate well with each other,[14] there is one well-established SLE damage index, the Systemic Lupus International Collaborative Clinics/American College of Rheumatology (SLICC/ACR) damage index.[15] Moreover, several non-specific quality of life-measures have been successfully used in patients with SLE.[1,2] While useful in pointing to candidate ICF categories, these indices, however, will neither allow for functional measurement nor will they directly indicate the most relevant categories in understanding the functional disease burden of SLE.

To meet the need for new outcomes measures for SLE, we here propose a process to lead to comprehensive ICF core sets for patients with SLE. While this plan was first derived in a meeting conducted in Munich in November 2004, and bolstered after interactions with several other centres, we would like to invite worldwide participation in this effort. Only by cooperation and exchange can we can reach the goal of developing ICF core sets for SLE (and possibly including isolated cutaneous lupus) to depict comprehensively the functional problems faced by patients all over the world, including their many diverse and confusing variations.

Methodological approach to develop ICF Core Sets for SLE

Types of ICF Core Sets that will be developed

The development of ICF Core Sets for SLE will include two versions: The Brief ICF Core Set for SLE to be used in clinical trials. Thus, the Brief ICF Core Set for SLE will include a list of ICF categories with as few categories as possible to be practical, but as many as necessary to describe as comprehensively as possible the spectrum of problems in functioning of patients with SLE for clinical studies.

The Comprehensive ICF Core Set will be applicable for guiding multidisciplinary assessment in patients with SLE. Thus, more ICF categories will need to be included to adequately describe, in a multidisciplinary assessment, the typical spectrum of problems in functioning of any patient with SLE. Two additional add-on ICF Core Sets Lists will be developed. The first will be for mucocutaneous manifestations, while the second will be for musculoskeletal involvement, the latter based on the existing core set for rheumatoid artritis (RA).

Based on knowledge gained in the previous ICF Core Set developments, the following methodological approach for the Core Set development is projected.

Study design

As is the standard procedure for the ICF Core Sets, the SLE core sets will be defined at a consensus conference, which will integrate i) a systematic review of the literature regarding the outcome measures used in clinical trials and selected observational studies, ii) focus groups or semi-structured interviews with SLE patients, iii) a Delphi exercise with world wide involvement of experts, and iv) empirical studies.

With this study design, all relevant perspectives should be adequately addressed. The patient perspective, which is crucial to the success of the project, is addressed both in a qualitative way in focus groups and in a quantitative way in the empirical studies. In addition, we will try to initiate a Delphi exercise for patients. The investigator perspective is addressed in the systematic literature reviews. Finally, various

Table 1 Symptoms (not objective parameters) rated in commonly used SLE activity scores

Organ system	*Symptom*	*Score*	*ICF*	*Description*
Constitutional/general	Anorexia/nausea/vomiting	BILAG	b530	Weight maintenance functions
			b5350	Sensation of nausea Regurgitation
			b5106	and vomiting
	Fatigue	BILAG, ECLAM, SIS, SLAM	b130	Energy & drive functions
			b4552	Fatiguability
	Fever/pyrexia	BILAG, ECLAM, SIS, SLAM, SLEDAI	nd	
	Lymphadenopathy	BILAG, SIS, SLAM	s4201	Structure of lymphatic nodes
	Splenomegaly	BILAG, SLAM	s4203	Spleen
	Weight loss	BILAG, SLAM	b530	Weight maintenance functions
Mucocutaneous	Alopecia	BILAG, ECLAM, SIS, SLAM, SLEDAI	b850	Functions of hair
	Angio-oedema	BILAG	b4351	Hypersensitivity reactions
	Calcinosis	BILAG	s898	Skin and related structures, other specified: calcinosis
	Mucosal ulcers	BILAG, ECLAM, SIS, SLAM, SLEDAI	b810	Protective functions of the skin
	Panniculitis	BILAG	b820	Repair functions of the skin
	Sclerodactyly	BILAG	b810	Protective functions of the skin
			b820	Repair functions of the skin
	Skin rash	BILAG, ECLAM, SIS, SLAM, SLEDAI	b810	Protective functions of the skin
	Skin ulcers	BILAG, SIS, SLAM, SLEDAI	b810	Protective functions of the skin
Musculoskeletal	Arthralgias	BILAG, ECLAM, SIS, SLAM	b28016	Pain in joints
	Arthritis	BILAG, ECLAM, SIS, SLAM, SLEDAI	s7701	Joints
			b28016	Pain in joints
			b710	Mobility of joint functions
			b715	Stability of joint functions
	Aseptic necrosis	BILAG	s7700	Bones
			s7701	Joints
	Contractures	BILAG	b710	Mobility of joint functions
	Myalgias	BILAG, SIS, SLAM	b28018	Pain in body part, other specified: muscle
	Muscle weakness	SIS, SLAM, SLEDAI	b730	Muscle power functions
	Tendonitis/tendosynovitis	BILAG, SLAM	s73023	Ligaments and fasciae of hand
Neurological	Cerebellar ataxia	BILAG	b7602	Coordination of voluntary movements
	Chorea	BILAG, SLAM	b7650	Involuntary contractions of muscles
	Cranial nerve disorder	BILAG, SIS, SLAM, SLEDAI	s1106	Structure of cranial nerves
	Depression	BILAG, ECLAM, SIS, SLAM, SLEDAI	b152	Emotional functions
	Headache	BILAG, ECLAM, SLAM, SLEDAI	b28010	Pain in head and neck
	Mononeuritis multiplex	BILAG, SIS, SLAM	b280	Sensation of pain
			b265	Touch functions
			b270	Sensory functions related to temperature & other stimuli
	Organic brain syndrome	BILAG, ECLAM, SIS, SLAM, SLEDAI	b1102	Quality of consciousness
			b114	Orientation functions
			b152	Emotional functions
	Psychosis	BILAG, ECLAM, SIS, SLAM, SLEDAI	b1102	Quality of consciousness
			b114	Orientation functions
			b152	Emotional functions
	Seizures	BILAG, ECLAM, SIS, SLAM, SLEDAI	s1100	Structure of cortical lobes
			nd	

(Continued)

Table 1 Continued

Organ System	*Symptom*	*Score*	*ICF*	*Description*
	Stroke symptoms	BILAG, ECLAM, SIS, SLAM, SLEDAI	s110	
	Structure of brain		nd	
	Transverse myelitis	BILAG, SLAM	s120 nd	Structure of spinal cord
	Visual disturbance	BILAG, SLAM, SLEDAI	b210	Seeing functions
Pulmonary	Arrhythmias	BILAG	b4101	Heart rhythm
Cardiovascular	Dyspnoea	BILAG, ECLAM, SLAM	b440	Respiratory functions
	Phlebitis	BILAG	s4102	Veins
	Pleuropericardial pain	BILAG, ECLAM, SIS, SLAM, SLEDAI	b28011	Pain in chest
	Raynaud's	BILAG, ECLAM, SIS, SLAM	b415	Blood vessel functions
	Thromboembolism	BILAG, SIS	b430	Haematological system functions
Abdominal	Abdominal vasculitic crisis	BILAG, ECLAM, SLAM	b415	Blood vessel functions
	Pancreatitis	SLAM	s550	Structure of pancreas
	Peritonitis	ECLAM, SLAM	b28012	Pain in stomach or abdomen
Renal	Nephrotic syndrome	BILAG	b6100 b54500	Kidneys Water retention

expert opinions involving professionals from different backgrounds shape the Delphi exercise, as well as the final conference.

Consensus conference

The consensus conference is scheduled for Vienna in mid-2007. Experts in the field of SLE will work together actively to select the ICF categories for the Comprehensive and the Brief Core Set. Based on previous experience, it is proposed to form three groups with seven experts each, including different health professionals, who can work together in the spirit of the multi-professional and multi-disciplinary approach typical for care and research in SLE. The method selected to regulate the group dynamic and the teamwork during the conference is the 'Nominal-Group Technique'.[16]

Evidence from empirical studies

A systematic review of the concepts of outcome measures used in published studies[17] is being performed. The objectives of the systematic review are to identify outcome measures cited in published studies focusing on individuals with SLE, and to quantify the concepts contained in these measures. The concepts of the retrieved outcome measures will be linked to ICF categories using standardized linking rules.[18] The results from this systematic review of empirical studies should be available in June 2006.

Focus groups with patients

'Focus groups', ie, open interviews with patients in small groups of up to seven people, have been chosen as the most appropriate available qualitative approach for identifying the patient perspective of functioning and health.[19,20] As compared to individual interviews, focus groups are enriched by interactions in the group, and therefore usually generate additional information. Focus group studies will at least be performed in Europe (Austria) as well as in the US and East Asia. These groups should be completed by the end of August 2006, so that their results can be included in the Delphi exercise and empirical studies (below).

Delphi exercise

A Delphi exercise will encourage experts to identify problems they appreciate for their patients in a feedback procedure based on information-technology,[21] with the goal of shaping common consensus. Balanced involvement with regard to WHO regions and professions will be facilitated by directly approaching experts from both underrepresented areas (eg, Africa, South America) and fields (eg, psychologists, physiotherapists). Delphi exercises will start in September 2006, and should be finished by January 2007. We will try to also initiate an SLE patient Delphi exercise with a similar approach.

Empirical study

A world-spanning multi-centre cross-sectional study of individuals will identify ICF categories relevant for patients with SLE. The ICF Checklist,[22] the SF-36,[23] a questionnaire for socio-demographic variables, and additional items derived from the literature review will be used for this study. Health status measures identified in literature reviews will be linked to the ICF.[18] These empirical studies will likewise start in October 2006, with results available by February 2007.

Conclusions

In embarking on the project to define ICF Core Sets for SLE, we here have described the background and the approach for development of both a Comprehensive and a Brief ICF Core Set for SLE. It is interesting, in this regard, that SLE activity scores include important information on some features of the disease, which link to the components *body functions* and *body structures* of the ICF classification (Table 1). However, it is likewise obvious that there is no one-to-one linkage, demonstrating systematic differences in the approach, and that some symptoms could not be linked to an ICF category. Moreover, when considering the patient perspective, we may also need to assess the other components, namely the individuals' functioning in daily life and social participation. Since functioning and participation in daily living contribute significantly to the quality of life, they are highly relevant aspects for the comprehensive assessment of a patient with SLE.

Comprehensive assessment and understanding of individual daily problems requires a strong focus on patient perspectives, which most likely differ between cultures. The ICF includes the environment as a contextual factor, which also differs between countries and cultures. Therefore, this project will accommodate this international perspective both in the selection of locations for patient focus groups and the participation of additional experts for the Delphi exercise.

The development of the SLE ICF Core Sets will be an inclusive and open process. We therefore encourage both SLE clinical experts and SLE patient representatives to actively participate in the process. Individuals, institutions and associations, can be formally associated as partners of the project. Everybody interested should please contact the project coordinators (MA, TS, DP). We believe that it is time develop the tools to assess the life of patients with SLE with greater clarity precision and thereby promote treatments to enhance their quality of life.

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