Full Length Article

The *Health Assessment Questionnaire Disability Index (HAQ-DI)* as a valid alternative for measuring the functional capacity of people with haemophilia

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A R T I C L E   I N F O

A B S T R A C T

Background: There are several scales for evaluating the functional capacity of people with haemophilia (PWH).
Objective: To demonstrate the value and simplicity of the "Health Assessment Questionnaire Disability Index" (HAQ-DI) for making a functional assessment of PWH in those contexts in which the specific physical assessment scales cannot be used [Functional Independence Score in Haemophilia (FISH) and Haemophilia Activities List (HAL)]. The HAQ-DI is a validated generic self-administered questionnaire that is completed in <5 min.
Methods: Data was collected on the physical and functional status of 62 adult PWH having haemophilia. Their average age was 34.7 years. The impairment of the PWH was assessed using the generic self-administrable functionality questionnaire (HAQ-DI) and the following scales: The "World Federation of Haemophilia Physical Examination Score" (WFH-PES) and the Haemophilia Joint Health Score 2.1 (HJHS 2.1). We evaluated the correlation between generic HAQ-DI score and physical assessment scores WFH-PES, HJHS 2.1 (correlation analysis).
Results: The correlation between WFH-PES and HAQ-DI showed a Spearman's correlation coefficient of $r = 0.804$ ($p < 0.05$). The correlation between HJHS 2.1 and HAQ-DI showed a Spearman's correlation coefficient of $r = 0.823$ ($p < 0.05$). A positive and fairly strong correlation was found between them.
Conclusions: The presented associations established the application of HAQ-DI as a PROXY clinical indicator of functional status. HAQ-DI is a valid alternative for assessing functional capacity in adult PWH, especially in cases in which, for some reason, it is not possible to use other tools that are specific for haemophilia. Clinical practice needs a less time demanding assessment tools.

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1. Introduction

Haemophilia is a hereditary disease that is linked to the X chromosome and causes a deficiency in clotting factor VIII or IX. Depending on the clotting factor level, it is classified as severe (<1%), moderate (1–5%) or mild (>5%) [1]. Haemophilia is a haematological disease, but is fundamentally orthopaedic at the clinical level, since its principal symptom is the bleeding episodes which most frequently affect the joints (ankles, knees and elbows), as well as several muscle groups [2,3]. Haemophilia is treated by administering the deficient clotting factor when a bleed occurs (on-demand modality) or regularly (prophylactic modality) [4]. Prophylactic treatment is currently accepted by the scientific community to be the gold standard during childhood [5] - its use for adults being more questioned [6,7]. It is well known that bleeds cause

deterioration of joint and muscle function, causing a degree of disability that seriously impacts the patients' ability to carry out everyday activities and their quality of life [8]. It is therefore of fundamental importance to clinically monitor people with haemophilia (PWH) individually and find out how these changes impact their daily activities.

In a musculoskeletal context, the assessment and therapeutic approach must be based on four fundamental aspects: clinical examination, imaging test, functional capacity and quality of life. The clinical examination must be full, and include the patient's locomotion capability and the state of their joints, muscles and posture [9]. Today we have specific scales for haemophilia that allow us to measure the findings in both adults and children such as *World Federation of Haemophilia Physical Examination Score* (WFH-PES) [10], also called the Gilbert Score and *Haemophilia Joint Health Score 2.1* (HJHS 2.1) [11,12]. For a complete examination, imaging tests should also be run when the clinical situation of the PWH so requires (X-rays, ultrasound, MRI, CT). Functionality is assessed through the PWH's ability to carry out basic everyday activities in his or her own surroundings. Many generic tools for this have been

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described i.e. the *Health Assessment Questionnaire Disability Index* (HAQ-DI), the *SF-36* (Short-Form-36), the *Functional Independence Measure* (FIM) and the Barthel Index among others [13]. In terms of specific haemophilia questionnaires, the FISH (*Functional Independence Score in Haemophilia*) and the HAL (*Haemophilia Activities List*) questionnaires have been published and are extensively used, although the degree to which they have been transculturally validated is still low [14,15]. With respect to quality of life scales, there are many generic and specific questionnaires (*Haemo-QoL-A*, *A36 Hemofilia-QoL*, *HemoLatin-QoL*, *Hemo-A-QoL*) [15].

Both WFH-PES and HJHS 2.1 scales may be used to determine the intervention and treatment regimes to be used (haematological, rehabilitation or orthopaedic). It has also been stated that, with increased experience using the tool, less time may be required to administer and score the assessment.

The HAQ-DI is one of the most widely used comprehensive, validated and patient-oriented outcome assessment instruments [30]. The HAQ-DI is a tool that, in its complete form, assesses mortality, disability, symptoms, secondary effects of treatment and financial impact, but in clinical practice it is normally only the disability scale that is used (HAQ-DI) [16].

To date, culturally adapted HAQ-DI instruments have proved as equally reliable and valid as their parent. The HAQ-DI is one of the first instruments deliberately designed to capture prospectively and by protocol the long term influence of multiple chronic illnesses and to allow supplementation by additional measures for particular studies [16]. It has also been applied to a variety of diseases and conditions [17]. It has been used in other studies to assess the functional disability associated with haemophilia A and B [18]. It has been and continues to be administered across diverse disciplines and in different cultures, with properly designed adaptations that do not impact its reliability and validity. It should be considered a *generic* rather than a *disease-specific* instrument [16].

The HAQ-DI was initially validated in the English-speaking population. Later, since 2002, it has been translated and culturally adapted for over 60 languages and dialects, usually with minimal changes. The translation has been fully validated and some items have had to be modified to adapt it culturally. The HAQ-DI has been translated and adapted independently or by the Health Outcomes Group into the following languages: Arabic, Australian, Austrian, Dutch (Flemish), Belgian French, Canadian (French), Chinese, Chinese (Cantonese [Hong Kong]), Croatian, Danish, English (Canadian, United Kingdom), Finnish, French (France), German (Germany, Switzerland), Greek, Hebrew, Italian, Israel (English), Korean, Lithuanian, Netherlands, Norwegian, Portuguese (Brazil, Portugal), Romanian, Scandinavian (multiple languages), Scottish, Singapore (English, Malay, Mandarin), Slovenian, So. Africa (Afrikaans, English), Spanish (Argentine, Chilean, Costa Rican, Guatemalan, Mexican, Spain, Venezuelan), Swedish, Swiss, Thai, Turkish [16]. It has also been stated that the HAQ-DI can be completed by the patients themselves in <5 min [30].

This article proposes to present the HAQ-DI as a generic and self-administrable tool that evaluates how haemophilia impacts the ability of PWH to carry out everyday activities, which may be useful in environments for which the specific instruments cannot be used.

2. Methods

2.1. Participants

Between November 2013 and December 2014, data was collected on the physical and functional status of adult patients with haemophilia. These included people with haemophilia A and B, and having haemophilia with different degrees of severity depending on their clotting factor levels: severe (<1%), moderate (<5%) and mild (>5%). Both PWH undergoing prophylactic treatment and PWH being treated on demand were included. A musculoskeletal assessment in terms of

their haemophilia was made of all the patients who came to appointments at our centre's Haemophilia Unit. The impairment was quantified using the WFH-PES and the HJHS 2.1, two specific haemophilia scales. The same group of PWH was asked to fill in the Spanish version of the generic self-administrable functionality questionnaire (HAQ-DI), and they all did so. All patients having appointment in our institution in the aforementioned period were selected for the study. Those PWH who had an acute articular bleed at the time of the examination were excluded because the study was not designed to investigate acute haemarthroses.

2.2. Measurements

In all patients physical examination was performed and scored by the same examiner (HC-R) and always before the self-administered HAQ-DI disability scale. When filling in the disability scale, the patients did not know the score of the physical scales (WFH-PES and HJHS2.1). The HJHS 2.1 and the HAQ-DI were scored following the instructions recommended by the authors that developed such scales.

The first author of this article (Medical Specialist in Physical Medicine and Rehabilitation) carried out the physical examination in a consulting room with the following resources: stretcher, goniometer, measuring tape, and space to assess the gait. In addition, patients' footprints were analysed using a podoscope. The physical examination included data on pain, swelling, joint mobility, muscular strength and trophism, articular stability, deformity, footprint, balance and a visual analysis of the gait.

WFH-PES and HJHS 2.1 scales were chosen because they are tools that are haemophilia-specific and are commonly used in clinical practice [10–12]. The WFH-PES measures joint health by measuring, independently, the joints most frequently affected by haemophilia: ankles, knees and elbows. Until recently, it was the most frequently used scale in all contexts. Lately, the WFH recommends its use for adolescents and adults, but not for small children, since it seems to be less sensitive to early changes than the HJHS 2.1. The WFH-PES scores the following 7 items: *Swelling* (0–2, S if *synovitis*), *Muscle atrophy* (0–1), *Axial deformity* (0–2, measured only in knee and ankle), *Crepitus on motion* (0–1), *Range of motion* (0–2), *Flexion contracture* (0–2), and *Instability* (0–2). It is a cumulative scale, in which the higher the score, the worse the health of the joint; the minimum score is 0 and the maximum is 10 for an elbow and 12 for a knee or ankle. It was developed in English and is translated to Swedish and German [10–15,19,20].

The HJHS 2.1 is a physical assessment scale that measures the overall joint health of the joints most commonly affected by haemophilia, including the following 6 joints: left and right ankle, left and right knee, and left and right elbow. It was initially published to be used for children aged 4–18 with mild joint impairment, since it can pick up the early signs of joint disease. However, today it is also used in adult patients. The scale scores the following 9 parameters: *Swelling* (0–3), *Duration of swelling* (0–1), *Muscle atrophy* (0–2), *Crepitus on motion* (0–2), *Flexion loss* (0–3), *Extension loss* (0–3), *Pain* (0–2), *Strength* (0–4) and *Global gait* (0–4). It has not yet been adequately studied for use in adults, although some preliminary studies indicate that it is useful for that age group [21]. The manual is available in English, French, German and Spanish. It is a cumulative scale, in which the higher the score, the worse the musculoskeletal health. The minimum score is 0 and the maximum is 124: 20 per joint multiplied by 6 joints (two elbows, two knees, two ankles) equals 120 plus 4 for gait (124 overall).

Furthermore, information as to the patients' functional capacity was also collected; for this the HAQ-DI generic questionnaire was used. There are currently specific haemophilia tools, such as the FISH [14] and the HAL [15], both of which have been accepted by the WFH, with the recommendation to apply them in a complementary manner. They could not be used in our clinical context because there has been no transcultural validation for Spanish and we do not have the necessary material and time to complete the assessment.