Haemophilia
The Official Journal of the World Federation of Hemophilia,
European Association for Haemophilia and Allied Disorders and
the Hemostasis & Thrombosis Research Society

Haemophilia (2014), **20**, e267–e274 DOI: 10.1111/hae.12435

ORIGINAL ARTICLE *Clinical haemophilia*

Reliability and validity of a novel Haemophilia-specific Self-Efficacy Scale

J. LOCK,* H. RAAT,† M. PETERS,‡ R. Y. J. TAMMINGA,§ F. W. G. LEEBEEK,¶ H. A. MOLL** and M. H. CNOSSEN*

**Department of Paediatric Haematology, Erasmus MC - Sophia Children's Hospital; †Department of Public Health, Erasmus MC, University Medical Centre Rotterdam, Rotterdam; ‡Department of Paediatric Haematology, Academic Medical Centre Amsterdam, Amsterdam; §Department of Paediatric Haematology, University Medical Centre Groningen, Groningen; ¶Department of Haematology, Erasmus MC, University Medical Centre Rotterdam; and **Department of General Paediatrics, Erasmus MC – Sophia Children's Hospital, Rotterdam, The Netherlands*

Summary. Higher self-efficacy in chronic disease patients is associated with higher development of self-management skills and increased quality-of-life. Quantification and monitoring of self-efficacy is therefore of importance. Self-efficacy in haemophilia patients has received little attention due to lack of standardized scales. To validate the novel Haemophilia-specific Self-Efficacy Scale (HSES) in haemophilia patients on prophylactic home treatment, haemophilia patients aged 1–18 years on prophylactic treatment ≥1 year were included from three Dutch Haemophilia Treatment Centres. The HSES consists of 12 items, relating to perceptions of the ability to function on a day-to-day basis with regard to patient's disease. Retest was performed in a subsample. Validity was proven by the General Self-Efficacy Scale and by the health-related quality-of-life assessment tool Haemo-QoL. Data were analysed from 53 children (response 75%), with a mean age of 9.8 years (SD 4.0). Mean total scale score of HSES was 55.5 (SD 4.7; range 38–60), with a ceiling effect of 17%. The HSES showed adequate *internal consistency* (Cronbach's alpha 0.72) and good *test–retest reliability* (Intra-Class-Correlation coefficient 0.75; $P < 0.01$; $n = 37$). The *convergent validity* was adequate as haemophilia-specific self-efficacy correlated significantly with general self-efficacy ($r = 0.38$; $P < 0.01$). High HSES scores correlated significantly with quality-of-life as measured by the Haemo-QoL ($r = -0.42$; $P \leq 0.01$). The novel HSES is a reliable and valid tool to assess self-efficacy in paediatric haemophilia patients on prophylactic home treatment. High self-efficacy correlated with higher quality-of-life, further underlining the importance to standardly assess, monitor and improve self-efficacy.

Keywords: children, haemophilia, health status, instrument development, self-efficacy, validity

Introduction

In haemophilia, as in other chronic diseases, self-management skills of patients and caretakers are of relevance for treatment adherence, prognosis of disease and quality-of-life [1]. Prophylactic replacement therapy with clotting factor concentrate in the home setting requires a high ability of self-management as organization of care is complex (Lock J, Raat H, Duncan N, Shapiro A, Beijlevelt M, Peters M, Tamminga RYJ, Leebeek FWG, Moll HA, Cnossen MH, Submitted; [2]). It includes insight on the necessity and dosing of clotting factor concentrate, taking prior prophylactic doses into account. Also practical and logistic capacities are of significance with regard to clotting factor concentrate infusion, stock and timely communication with the Haemophilia Treatment Centre (HTC).

Bandura developed the concept of 'self-efficacy'. This term describes the actual confidence an individual possesses with regard to specific actions necessary to achieve certain results [3]. It summarizes the integration of a motivated attitude towards a disease and its treatment, a capacity towards adequate judgment with regard to therapeutic interventions and demonstration of adherence to prescribed therapy [4]. Patients with low self-efficacy are less likely to persevere in a specific

Correspondence: Marjon. H. Cnossen, MD, PhD, Department of Paediatric Haematology, Erasmus University Medical Centre – Sophia Children's Hospital, P.O. Box 2040, 3000 CA Rotterdam, The Netherlands.
Tel.: +00 31 10 7036691; fax: +00 31 10 7036801;
e-mail: m.cnossen@erasmusmc.nl

Accepted after revision 9 March 2014

task when impediments arise, obliterating usual proceedings. Those with high self-efficacy will deploy all abilities to master obstacles. In clinical practice, self-efficacy is considered an antecedent for modification of behaviour [5,6]. Furthermore, development of disease-specific self-efficacy questionnaires is required to take disease-specific aspects into account not dealt with by the current validated general questionnaire.

In various chronic diseases high levels of self-efficacy are associated with higher quality-of-life and less clinical and psychological symptoms [5,7–9]. In addition, Richardson *et al.* reported that patients with a wide range of chronic diseases value self-efficacy highly, and are willing to trade reductions in health-related quality-of-life for improvements in their self-efficacy [10]. In haemophilia, a number of studies have evaluated general self-efficacy and possible training modules, but few have looked at disease-specific self-efficacy.

Kang *et al.* proved that a self-help program for mothers of children with haemophilia significantly improved knowledge, self-efficacy and quality-of-life [11,12]. Mulders *et al.* reported that an educational e-learning program in patients on prophylactic home treatment significantly improved general knowledge of treatment [13]. However, in this cohort, self-efficacy scores were relatively high at initiation and did not increase after intervention. Conflicting results were found by Barlow *et al.* and Buxbaum *et al.*, as the first documented high levels of self-efficacy in haemophilia patients, indicating a well-developed confidence with regard to disease management, whereas the latter found lower self-efficacy scores in haemophilia patients than in healthy controls [14,15]. All studies were performed using a general self-efficacy scale or a non-validated Haemophilia-specific Self-Efficacy Scale (HSES) as there was no validated HSES available. To adequately quantify and monitor self-efficacy and to identify subgroups at risk of higher morbidity and decreased quality-of-life, the HSES was recently developed and validated. This study aims to describe the psychometric properties of this novel instrument and the association between HSES and quality-of-life.

Patients and methods

Patients

Data for this cross-sectional, multicentre study were collected as part of a larger prospective study on the efficacy of home-treatment intervention by a trained haemophilia nurse (Netherlands Trial Register: 2543). Between June 2010 and December 2011, we enrolled children aged 1–18 years with haemophilia A or B on prophylaxis and home treatment for at least 1 year, from three HTC's in the Netherlands (Erasmus Medical Centre – Sophia Children's Hospital, Rotterdam; Academic Medical Centre – Emma Children's Hospital, Amsterdam; University Medical Centre Groningen). Patients and parents were required to speak and understand Dutch sufficiently. Patients with inhibitors against FVIII or FIX were excluded. One caregiver, primarily involved in the child's daily haemophilia treatment, and adolescents aged 10–18 years were asked to complete the questionnaire. To evaluate test–retest reliability of HSES, the questionnaire was sent two weeks after administration of the first questionnaire to consenting participants. Participants not returning the questionnaires within 2 weeks received reminders and were considered lost to follow-up after two unreturned messages. The Medical Ethical Committee granted permission to perform the study and written informed consent was obtained [MEC2010097].

Data collection

Socio-demographic data, including parental level of education, employment status and family structure were provided. For level of education the International Standard Classification of Education (ISCED) division into low, medium and high educational levels was applied [16]. Low is equivalent to ISCED 0–2, i.e. 'less than upper secondary level'; medium to ISCED 3–4, i.e. 'upper secondary level' and high to ISCED 5–6, meaning tertiary level, or minimally 2 years of education after upper secondary level. Haemophilia diagnosis, treatment and clotting factor consumption were extracted from medical files.

Self-efficacy

Haemophilia-specific Self-Efficacy Scale

To specify disease-specific self-efficacy qualities, a novel scale was developed and validated, specifically for haemophilia patients. The HSES was composed by a team of haemophilia professionals and psychologists with items from the validated Sickle Cell Self-Efficacy Scale [7,17], from the Pain Self-Efficacy Questionnaire [18] and from the validated General Self-Efficacy Scale [19,20]. The first two questionnaires were used as they specifically encompass disease aspects such as periodic immobilization and pain. The novel HSES consists of 12 items focusing on an individual's perceptions of haemophilia disease symptoms and the patient's abilities to cope with or reduce these symptoms. In our view, all aspects of treatment are incorporated: treatment efficacy, quality-of-life, infusion technique, state of mind in case of a bleed, pain modification, confidence, modification of prophylactic regimen, continuation of daily activities, other therapeutic interventions besides clotting factor treatment, belief in leading of a normal life, communication and attainment of personal goals. Items are scored ranging from 'I totally

disagree' to 'I totally agree' (see figure). On a five-point Likert-scale, the lowest score was given one point and the highest score five points. An unweighted sum score was calculated by adding the 12 item scores, with higher scores indicating greater self-efficacy (range: 12–60).

General Self-Efficacy Scale (GSES)

To assess the convergent validity of the HSES, we used the validated General Self-Efficacy Scale (GSES) [19,20]. The GSES consists of 10 items on a four-point Likert-scale, ranging from 'I totally disagree' to 'I totally agree', with sum scores ranging from 10 to 40. Higher scores also indicate greater general self-efficacy. Although self-efficacy is considered to be task-specific, we assumed the concepts of general self-efficacy and haemophilia-specific self-efficacy to be related, which is supported by literature in other diseases, when assessing self-efficacy [21].

Haemo-QoL

The disease-specific quality-of-life instrument Haemo-QoL was used to assess the divergent construct validity. This is a self-report measure for children with haemophilia and their parents, consisting of 21–77 items which cover 9–11 domains depending on the age group of the patient. Higher scores indicate lower disease-specific quality-of-life [22].

VERITAS-Pro

To quantify treatment adherence in children on prophylaxis, we used the Validated Haemophilia Regimen Treatment Adherence Scale–Prophylaxis (VERITAS-Pro) (Lock J, Raat H, Duncan N, Shapiro A, Beijlevelt M, Peters M, Tamminga RYJ, Leebeek FWG, Moll HA, Cnossen MH, Submitted; [2]). This instrument contains six subscales ('Time', 'Dose', 'Plan', 'Remember', 'Skip', 'Communicate'), each represented by four questions concerning a specific domain of haemophilia patient care. Cumulative score of all subscales ranges from 24 to 120 and cumulative scores per subscale range from 4 to 20. Lower scores reflect *higher* adherence.

Data analysis

Psychometric properties of HSES

The following psychometric properties of the HSES were evaluated as follows: feasibility, reliability and validity (convergent and divergent validity). Feasibility was expressed as response rate. Scale scores were described in terms of scale mean, SD, range, floor and ceiling effects and percentiles.

The total scale internal consistency reliability was assessed using Cronbach's alpha. Amidst varying standards in the literature, we considered 0.70 to be an acceptable alpha coefficient [23].

The test–retest reliability was assessed by the Intra-Class-Correlation Coefficients (ICC). The agreement between the perceived haemophilia-specific self-efficacy of parents and adolescents was also assessed by the ICC.

Validity was assessed by comparing HSES outcomes with the validated GSES and the Haemo-QoL. It was hypothesized that a low HSES outcome should correlate with low self-efficacy outcomes on the GSES and a low quality-of-life (i.e. higher score) by Haemo-QoL. As data were not normally distributed correlations in overall median sum scores were calculated and tested with Spearman's correlation coefficient. Low haemophilia-specific self-efficacy was defined as the lowest quartile of HSES scores, while high haemophilia-specific self-efficacy was defined as the highest quartile of HSES scores as data were not distributed normally.

Subgroup analyses were assessed by comparing HSES outcomes with age, duration of prophylactic home treatment, number of siblings, level of education, marital status and family composition. We compared the patient group with the lowest quartile of HSES scores with the patient group with the highest quartile of HSES scores. Due to non-parametrical data, the continuous outcomes were assessed using the Mann–Whitney *U*-test and categorical data were analysed by Chi-square test or the Fisher's exact test in case of low patient counts per subgroup.

Data were analysed separately for parent-reported and adolescent-reported scales, except for the inter-rater agreement analysis which compared adolescent-reported with parent-reported scales.

We considered P-values <0.05 as statistically significant; all tests were two-sided. All analyses were performed using SPSS 20.0 for Windows.

Results

Participants

A total of 71 patients of which 40 adolescents (10–18 years) were invited for study participation. Eighteen parents of children, including parents of 12 adolescents declined or did not fill out the questionnaire. Reasons for non-participation included time burden and logistical reasons. Fifty-three parents of both young children and adolescents (parent-reported questionnaires; response 75%) and 28 adolescents (adolescent-reported questionnaires; response 70%) were participated. Table 1 describes the baseline characteristics of all participants.

All 53 children were male with a mean age of 9.8 years (SD 4.0), 81% were diagnosed with

Table 1. Characteristics of the 53 participants at the time of study enrolment.

Characteristic	N (%)
Patient characteristics	
Age patients (years), mean (SD)	9.8 (4.0)
Sex patients, male	53 (100)
Diagnosis	
Haemophilia A	43 (81)
Haemophilia B	10 (19)
Severity of haemophilia	
Severe (<1%)	47 (89)
Moderate (1–5%)	5 (9)
Mild (6–40%)*	1 (2)
Duration of prophylactic treatment (years), mean (SD)	7.1 (3.6)
Parent characteristics	
Age parents (years), mean (SD)	39.8 (7.0)
Level of education†,‡	
Low level of education	4 (8)
Medium level of education	33 (66)
High level of education	13 (26)
Marital status†	
Married/registered partnership	11 (22)
Unmarried	30 (59)
Widow/widower	2 (4)
Divorced	8 (16)
Family composition†	
Living with partner and child(ren)	40 (78)
Single with child(ren)	10 (20)
Other	1 (2)
Individual completing scale†	
Mother/female guardian	45 (88)
Father/male guardian	6 (12)
Adolescent	28 (53)

*On prophylactic treatment due to bleeding tendency due to concomitant von Willebrand disease.
†Of two participants no information is available on marital status, family composition and whom filled out the questionnaire. Of three participants no information is available on level of education.
‡The usual ISCED division into Low, Medium and High is adopted here, as in the Eurostat Labour Force Survey. Low is equivalent to ISCED 0–2, i.e. 'less than upper secondary level of education'. Medium is given by ISCED 3–4, i.e. upper secondary level. High is ISCED 5–6, meaning tertiary level, or two more years of education after upper secondary level.

haemophilia A, 89% had severe haemophilia. Of the 53 children, 28 were adolescents. The mean age of this subgroup was 13.6 (SD 2.5). Mean duration of prophylactic treatment was 7.1 years (SD 3.6), with a median time span between prophylaxis initiation and start of the home treatment of 0.5 years. Of the 53 parents, the majority was female (88%), 8% were educated at a low level, 20% were single parents and 30% had two children or more.

Psychometric properties of the HSES

Table 2 displays the total scale scores. Mean total scale scores were relatively high (55.5 for the parent-report and 55.7 for the adolescent-report) as were the median scores (57.0 for both the parent- and the adolescent-report). Floor effects were absent. Ceiling effects were observed in 17% of the parents and in 29% of the adolescents.

The Cronbach's alpha of the total scale was $\alpha = 0.72$ for the parent-report and $\alpha = 0.86$ for the adolescent-report, indicating an adequate internal consistency (Table 2).

The test–retest reliability showed promising results, with a ICC of 0.75 (95%CI 0.56:0.86; $P < 0.01$; $n = 37$) for the parent-report and 0.67 (95%CI 0.29:0.87; $P < 0.01$; $n = 17$) for the adolescent-report. For the parent-report there was no significant difference between the test (mean 55.70; SD 4.16) and the retest (mean 55.88; SD 4.87; $P = 0.34$); also the adolescent-report showed identical results (test mean 55.88; SD 4.87; retest mean 55.18; SD 3.89; $P = 0.37$). The agreement between the perceived haemophilia-specific self-efficacy of parents and adolescents was however not significant (ICC −0.05; 95%CI −0.43:−0.35; $P = 0.59$).

Significant Spearman's correlations were observed with the General Self-Efficacy Scale (parent-report: $r = 0.43$; $P < 0.05$; adolescent-report: $r = 0.81$; $P < 0.01$). For the quality-of-life determined by the Haemo-QoL, the correlation of the total score was only significant for the parent-report of the HSES ($r = -0.45$; $P < 0.01$) and not significant for the adolescent-report HSES ($r = 0.02$; $P = 0.92$). Parents with a higher perceived self-efficacy (HSES) reported significantly less adherence with regard to subscales 'Plan', 'Remember' and 'Communication' on the VERITAS-Pro scale when compared with parents with a lower perceived self-efficacy (respectively $r = -0.28$; $r = -0.29$; $r = 0.37$; $P < 0.05$). No other correlations were seen between the HSES (parent-report), the HSES (adolescent-report) and other VERITAS-Pro (sub)scales (Table 3).

Parents with HSES scores in the lowest quartile reported significantly lower median scores on the: GSES ($P < 0.01$); the Haemo-QoL (sub)scales 'Total score' ($P < 0.01$), 'Feeling' ($P = 0.02$), 'View' ($P = 0.01$), 'Others' ($P = 0.03$), and 'Sport' ($P < 0.01$); and on the VERITAS-Pro subscales 'Remember' ($P = 0.05$), 'Skip' ($P < 0.01$) and 'Communicate' ($P = 0.01$), compared to parents with HSES scores in the highest quartile (Table 4). Adolescents with HSES scores in the lowest quartile reported significantly lower median scores on

Table 2. Score distribution and internal consistency reliability of the Haemophilia-specific Self-Efficacy Scale (HSES).

	Scale scores						Internal consistency reliability
	Mean (SD)		Range	Median [IQR]	Ceiling effect (%)*	Floor effect (%)†	Cronbach's alpha
Parent-report ($n = 53$)	55.45	(4.27)	45–60	57 [54–59]	17	0	0.72
Adolescent-report ($n = 28$)	55.68	(5.41)	38–60	57 [54–59]	29	0	0.86

*Ceiling effect; percentage of respondents with best possible score.
†Floor effect; percentage of respondents with worst possible score.

Table 3. Convergent and divergent validity of the Haemophilia-specific Self-efficacy Scale (HSES) with validation measures.

	HSES total scale	
	Parent-report	Adolescent-report
GSES*	0.43§	0.81¶
Haemo-Qol†		
Total score	−0.45¶	0.02
Physical	−0.11	−0.06
Feeling	−0.32§	−0.36
View	−0.38¶	−0.13
Family	−0.27	−0.25
Friends	−0.09	0.34
Support	−0.04	0.14
Others	−0.30§	−0.55¶
Sport	−0.30§	0.02
Dealing	0.05	0.31
Treatment	−0.13	−0.48¶
Future	−0.36	−0.23
Relation	−0.11	−0.14
VERITAS-Pro‡		
Total score	−0.12	0.08
Time	−0.10	−0.13
Dose	−0.11	0.16
Plan	−0.28§	−0.03
Remember	−0.29§	0.22
Skip	−0.26	−0.12
Communicate	0.37¶	0.14

*General self-efficacy scale.
†Haemophilia-specific health-related quality-of-life questionnaire.
‡Validated haemophilia regimen treatment adherence scale – prophylaxis.
§Spearman's correlation coefficient is significant at the 0.05 level (2-tailed).
¶Spearman's correlation coefficient is significant at the 0.01 level (2-tailed).

the: GSES ($P < 0.01$); the Haemo-QoL subscales 'Others' ($P = 0.04$) and 'Treatment' ($P = 0.02$), compared to adolescents with HSES scores in the highest quartile (Table 4).

Perceived disease-specific self-efficacy was not associated with age, duration of prophylactic treatment, level of education, number of siblings, marital status or with family composition. Neither in parents nor in adolescents (data not shown).

Discussion

The novel HSES is a feasible and reliable instrument to evaluate self-efficacy in Dutch paediatric patients with haemophilia on prophylactic home treatment. As timely communication and intervention is obligatory to modify prognosis in a disease with periodic episodes of pain and immobilization, we believe regular evaluation of self-efficacy is essential. In our study, HSES showed satisfactory psychometric properties and was able to discriminate between high and low self-efficacy. High HSES scores correlated significantly with quality-of-life measured by the Haemo-QoL. Further evaluation in other populations with regard to age and cultural background is necessary to broaden application possibilities of this valuable tool. Differentiation of subgroups within the haemophilia patient population with regard to self-efficacy is of paramount importance to identify potential high-risk patients with an increased risk of morbidity and decreased quality-of-life [24,25]. Subsequently, patients may undergo interventions aiming to increase self-efficacy, ultimately leading to cost-reduction of treatment in this era of rising health care costs.

Strengths of the HSES are diverse. Firstly, the 12 items chosen cover all aspects of haemophilia care in which self-efficacy plays a role and follow the definition of self-efficacy as described by Bandura in 1977 [3]. Secondly, general self-efficacy and disease-specific self-efficacy correlated significantly as did a higher self-efficacy with a higher quality-of-life as evaluated by HaemoQoL, a validated and widely used tool to analyse quality-of-life in children and adolescents with haemophilia. Furthermore, a high response rate was reached among the study population, leading to reliability of conclusions. Fourthly, as the HSES is an easily applied tool, it will allow monitoring of interventions aimed to improve haemophilia-specific self-efficacy. Finally, HSES is another example of a combination of qualitative research and quantitative survey techniques, such as seen in the development of the VERITAS-Pro by Duncan *et al.* [2]. In our opinion, this approach leads to richer, more valid and more reliable findings, with clear clinical implications, than when adopting qualitative or quantitative methods alone [26].

The limitations of our study are discussed. Firstly, some may deliberate the capturing of self-efficacy by a limited number of questions with fixed answering categories. However, we have chosen to make HSES a feasible tool in daily clinical practice: quick, reliable and valid. Secondly, the lack of patient report in constructing of the questionnaire is an omission as solely expert opinion of haematologists, haemophilia nurses and clinical psychologists was employed. Therefore, patient interpretation of questions may differ. Thirdly, due to practical reasons we were forced to exclude patients with language difficulties due to the questionnaire-based nature of the study. We are thoroughly aware, that specifically this group is characterized by low self-efficacy and decreased adherence to medical treatment [27]. Just as patients with inhibiting antibodies against FVIII/FIX, may also be characterized by low self-efficacy. We excluded this group, due to the fact that their intensive treatment has such a severe impact on daily life that it is not comparable to standard prophylactic treatment. Exclusion of these groups may have biased results towards underreporting of low self-efficacy. However, despite exclusion of these groups, HSES still differentiates between high and low self-efficacy [28], proving the sensitivity of the tool and its applicability in daily clinical practice. Fourthly, we administered a parent-report asking how

Table 4. Discrimination between validation measures between participants with low and high Haemophilia-specific Self-efficacy Scale(HSES) scores.

	HSES total scale					
	Parent-report			Adolescent-report		
Validity measures	Low HSES score* (n = 14); median [IQR]	High HSES score† (n = 9); median [IQR]	p-value‡	Low HSES score* (n = 8); median [IQR]	High HSES score† (n = 8); median [IQR]	p-value‡
GSES, mean§	32.50 [30.00–36.00]	38.00 [35.50–39.00]	<0.01	30.00 [27.00–31.75]	39.50 [38.00–40.00]	<0.01
Haemo-Qol¶						
Total score	33.77 [25.97–42.59]	20.13 [6.25–24.91]	<0.01	21.59 [16.75–33.33]	23.21 [20.45–24.59]	1.00
Physical	3.57 [0.00–25.89]	0.00 [0.00–8.93]	0.31	3.57 [0.00–25.89]	0.00 [0.00–26.79]	0.80
Feeling	10.94 [0.00–33.59]	0.00 [0.00–1.56]	0.02	4.69 [0.00–39.06]	0.00 [0.00–2.34]	0.13
View	20.14 [0.00–45.00]	0.00 [0.00–1.39]	0.01	8.75 [0.00–28.75]	3.75 [0.63–11.88]	0.72
Family	17.19 [6.25–33.59]	0.00 [0.00–25.00]	0.12	15.63 [0.00–35.16]	1.56 [0.00–14.06]	0.23
Friends	50.00 [25.00–70.31]	0.00 [0.00–84.38]	0.52	37.50 [0.00–65.63]	62.50 [34.38–95.31]	0.20
Support	53.13 [48.44–81.25]	100.00 [37.50–100.00]	0.25	75.00 [37.50–90.63]	81.25 [32.81–92.19]	0.73
Others	6.25 [0.00–30.21]	0.00 [0.00–2.08]	0.03	10.42 [0.00–78.13]	0.00 [0.00–0.00]	0.04
Sport	36.98 [6.25–62.50]	0.00 [0.00–5.56]	<0.01	2.78 [0.00–20.83]	5.56 [0.00–11.11]	1.00
Dealing	41.07 [20.54–50.89]	42.86 [32.14–58.93]	0.37	42.86 [30.36–57.14]	51.79 [44.64–59.82]	0.28
Treatment	20.31 [9.38–32.81]	9.38 [0.00–34.38]	0.37	43.75 [22.66–75.00]	15.63 [7.03–33.59]	0.02
Future	34.38 [25.00–37.50]	21.88 [18.75–25.00]	0.07	37.50 [28.13–46.88]	21.88 [4.69–35.94]	0.13
Relation	0.00 [0.00–37.50]	0.00 [0.00–0.00]	0.57	0.00 [0.00–25.00]	0.00 [0.00–0.00]	0.83
VERITAS-Pro**						
Total score	39.00 [29.75–43.00]	35.00 [29.00–38.50]	0.28	44.00 [40.50–54.75]	48.50 [39.25–55.25]	0.88
Time	5.50 [4.00–7.00]	5.00 [4.00–6.50]	0.48	6.50 [4.50–8.50]	6.00 [4.25–7.00]	0.72
Dose	4.50 [4.00–6.25]	4.00 [4.00–5.50]	0.48	4.50 [4.00–6.00]	5.00 [4.00–7.50]	0.72
Plan	8.00 [6.00–10.50]	4.00 [4.00–9.50]	0.10	10.50 [6.50–15.00]	9.00 [5.00–13.75]	0.80
Remember	8.00 [4.00–8.25]	4.00 [4.00–4.50]	0.05	5.00 [4.25–8.75]	7.00 [4.50–8.00]	0.72
Skip	5.00 [4.00–6.00]	4.00 [4.00–4.00]	<0.01	5.00 [4.25–5.75]	4.50 [4.00–5.75]	0.50
Communicate	5.00 [4.00–6.25]	10.00 [6.00–11.50]	0.01	11.00 [7.50–16.00]	14.50 [8.50–16.00]	0.44

*Lowest 25% of HSES scores.
†Highest 25% of HSES scores.
‡Mann–Whitney *U*-test.
§General self-efficacy scale.
¶Haemophilia-specific quality-of-life questionnaire.
**Validated haemophilia regimen treatment adherence scale – prophylaxis.

parents perceive their own self-efficacy, but unfortunately omitted how they perceive the self-efficacy of their children, which would have been a valuable addition. Furthermore, statistical analysis of subgroups was of course limited by small sample size. We therefore recommend future studies to assess reliability and validity in other subgroups of patients, and in other settings and to utilize other qualitative research methods such as cognitive debriefing.

Haemophilia-specific Self-Efficacy Scale properties were satisfactory. A floor effect was absent as is frequently the case in positively skewed assessments of reported self-efficacy [21,29,30]. Skewing was observed towards the most positive category ('ceiling effect') as often reported in other surveys on self-efficacy in chronic diseases [21,29,30]. This limitation effects the discrimination between participants with a high self-efficacy and restricts participants with a high self-efficacy to acquire better scores in a follow-up assessments. The ceiling effect can be explained by several factors such as the extensive education of patients and parents with regard to disease. In addition, current treatment focuses intensively on self-management skills, patients and parents have been dealing with the disease for a longer period of time, and multiple family members may be affected, leading to more disease experience.

The HSES scale questionnaire's internal consistency was good. Cronbach's alpha coefficients in similar questionnaires were comparable [7,17,31]. The test–retest reliability was adequate both in parents and adolescents. The agreement between the perceived haemophilia-specific self-efficacy of parents and adolescents showed no correlation, as is often seen when comparing parent-reported and adolescent-reported outcomes on self-efficacy and quality-of-life questionnaires [32,33]. This is most likely explained by the differences in treatment experience between parents and adolescents as well as diverging management responsibilities between parents and adolescents.

The convergent and divergent validity analyses of HSES showed promising test results, which is of paramount importance to discriminate between optimal and less optimal self-efficacy and to promote its future use. Both parents and adolescents clearly expressed similar opinions on their HSES report and their general self-efficacy report. In addition, parents also showed similar opinions on their disease-specific self-efficacy as expressed by the HSES and quality-of-life measurements by Haemo-QoL. However, the latter was only observed in some subscales of the Haemo-QoL in adolescents. Most probably, outcome is influenced by growing and not yet complete responsibility of adolescents for their disease, which directly

correlates with self-efficacy outcome and not with quality-of-life outcome. Adequate transition towards disease responsibility is of course expected of the adult haemophilia patient. This development could be measured and monitored by HSES, making it an important tool in the challenging transitional period [7,34,35].

In line with our hypothesis, we found that adherence to administered clotting factor concentrate doses in relationship to prior prophylactic doses (subscales 'Remember' and 'Skip') was higher in parents with a higher perceived haemophilia-specific self-efficacy. In contrast, we found evidence that adherence to communication with the HTC determined by the VERITAS-Pro was significantly lower in parents with a high perceived disease-specific self-efficacy than in parents with a low perceived self-efficacy. Our hypothesis is that the latter may be the consequence of the well-developed self-management strategies of these parents, decreasing communication moments. Further research is necessary to objectify patient outcome in these patients.

Conclusion

The HSES shows satisfactory psychometric properties to describe the self-efficacy in paediatric haemophilia patients on prophylactic home treatment. HSES parent-report correlated with quality-of-life measures, further underlining the importance to standardly assess, monitor and improve self-efficacy. Validation in other cohorts is impending to augment the value of HSES.

Acknowledgements

J. Lock, H. Raat, F.W.G. Leebeek, H.A. Moll and M.H. Cnossen designed the research study; J. Lock, M. Beijlevelt, M. Peters and M.H. Cnossen performed the research; J. Lock, H. Raat and M.H. Cnossen analysed the data; J. Lock and M.H. Cnossen wrote the manuscript. All authors critically reviewed and approved the final version of the manuscript. We thank A. de Goede-Bolder and C.T Hijmans for their valuable advice at the initiation of the project. The Haemophilia-specific Self-Efficacy Scale is added as an online supplement.

Disclosures

This study was made possible by an investigator-initiated unrestricted research grant from Bayer Schering Pharma (J. Lock) and an unrestricted research grant from Pfizer (M.H. Cnossen). F.W.G. Leebeek received an unrestricted research grant from CSL Behring and has served on advisory boards of CSL Behring and Baxter. MHC has received unrestricted research/educational funding for various projects from the following companies: Bayer Schering Pharma, Baxter, Novo Nordisk, Pfizer and Novartis. M. Peters received unrestricted research grants from Baxter, Novo Nordisk and Pfizer. H.R, R.Y.J.T, and H.A.M. stated that they had no interests which might be perceived as posing a conflict or bias.

References

1 Teitel JM, Barnard D, Israels S, Lillicrap D, Poon MC, Sek J. Home management of haemophilia. *Haemophilia* 2004; **10**: 118–33.
2 Duncan N, Kronenberger W, Roberson C, Shapiro A. VERITAS-Pro: a new measure of adherence to prophylactic regimens in haemophilia. *Haemophilia* 2010; **16**: 247–55.
3 Bandura A. Self-efficacy: toward a unifying theory of behavioral change. *Psychol Rev* 1977; **84**: 191–215.
4 Marks R, Allegrante JP, Lorig K. A review and synthesis of research evidence for self-efficacy-enhancing interventions for reducing chronic disability: implications for health education practice (part I). *Health Promot Pract* 2005; **6**: 37–43.
5 Smarr KL, Parker JC, Wright GE *et al*. The importance of enhancing self-efficacy in rheumatoid arthritis. *Arthritis Care Res* 1997; **10**: 18–26.
6 Jones F. Strategies to enhance chronic disease self-management: how can we apply this to stroke? *Disabil Rehabil* 2006; **28**: 841–7.
7 Clay OJ, Telfair J. Evaluation of a disease-specific self-efficacy instrument in adolescents with sickle cell disease and its relationship to adjustment. *Child Neuropsychol* 2007; **13**: 188–203.
8 Syrjala AM, Ylostalo P, Niskanen MC, Knuuttila ML. Relation of different measures of psychological characteristics to oral health habits, diabetes adherence and related clinical variables among diabetic patients. *Eur J Oral Sci* 2004; **112**: 109–14.
9 Tedesco LA, Keffer MA, Davis EL, Christersson LA. Effect of a social cognitive intervention on oral health status, behavior reports, and cognitions. *J Periodontol* 1992; **63**: 567–75.
10 Richardson G, Bojke C, Kennedy A *et al*. What outcomes are important to patients with long term conditions? A discrete choice experiment *Value Health* 2009; **12**: 331–9.
11 Kang HS, Kim WO, Cho KJ, Jeong Y. Development, implementation and evaluation of a new self-help programme for mothers of haemophilic children in Korea: a pilot study. *Haemophilia* 2010; **16**: 130–5.
12 Kang HS, Kim WO, Jeong Y, Kim SY, Yoo KY. Effect of a self-help program for mothers of hemophilic children in Korea. *Haemophilia* 2012; **18**: 892–7.
13 Mulders G, de Wee EM, Vahedi Nikbakht-Van de Sande MC, Kruip MJ, Elfrink EJ, Leebeek FW. E-learning improves knowledge and practical skills in haemophilia patients on home treatment: a randomized controlled trial. *Haemophilia* 2012; **18**: 693–8.
14 Barlow JH, Stapley J, Ellard DR, Gilchrist M. Information and self-management needs of people living with bleeding disorders: a survey. *Haemophilia* 2007; **13**: 264–70.
15 Buxbaum NP, Ponce M, Saidi P, Michaels LA. Psychosocial correlates of physical activity in adolescents with haemophilia. *Haemophilia* 2010; **16**: 656–61.
16 National Science Foundation. User Friendly Handbook for Mixed Methods Evaluations, 1997Available at http://www.nsf.gov/pubs/1997/nsf97153/. Accessed November 07, 2013.
17 Edwards R, Telfair J, Cecil H, Lenoci J. Reliability and validity of a self-efficacy instrument specific to sickle cell disease. *Behav Res Ther* 2000; **38**: 951–63.
18 Nicholas MK. The pain self-efficacy questionnaire: taking pain into account. *Eur J Pain* 2007; **11**: 153–63.
19 Luszczynska A, Scholz U, Schwarzer R. The general self-efficacy scale: multicultural validation studies. *J Psychol* 2005; **139**: 439–57.
20 Schwarzer R, Jerusalem M. Generalized Self-Efficacy Scale. In: Weinman J, Wright S, Johnston M eds. Measures in Health Psychology: a User's Portfolio Causal and Control Beliefs. Windsor, England: NFER-NELSON, 1995: 35–7.
21 Freund T, Gensichen J, Goetz K, Szecsenyi J, Mahler C. Evaluating self-efficacy for managing chronic disease: psychometric properties of the six-item Self-Efficacy Scale in Germany. *J Eval Clin Pract* 2013; **19**: 39–43.
22 von Mackensen S, Bullinger M, Haemo-Qo LG. Development and testing of an instrument to assess the quality of life of children with haemophilia in Europe (Haemo-QoL). *Haemophilia* 2004; **10**(Suppl 1): 17–25.
23 Nunnally JC, Bernstein IH. Psychometric Theory, 3rd ed. New York: McGraw-Hill 1994.

24 Marks R, Allegrante JP, Lorig K. A review and synthesis of research evidence for self-efficacy-enhancing interventions for reducing chronic disability: implications for health education practice (part II). *Health Promot Pract* 2005; **6**: 148–56.
25 Schrijvers LH, Uitslager N, Schuurmans MJ, Fischer K. Barriers and motivators of adherence to prophylactic treatment in haemophilia: a systematic review. *Haemophilia* 2013; **19**: 355–61.
26 National Science Foundation. User Friendly Handbook for Mixed Methods Evaluations, 1997. Available at http://www.nsf.gov/pubs/1997/nsf97153/. Accessed November 07, 2013.
27 Frayne SM, Burns RB, Hardt EJ, Rosen AK, Moskowitz MA. The exclusion of non-English-speaking persons from research. *J Gen Intern Med* 1996; **11**: 39–43.
28 Keane MC, Morgan BS. Perceived self-efficacy and language differences. *Psychol Rep* 1991; **69**: 291–8.
29 Brady TJ. Measures of self-efficacy: Arthritis Self-Efficacy Scale (ASES), Arthritis Self-Efficacy Scale-8 Item (ASES-8), Children's Arthritis Self-Efficacy Scale (CASE), Chronic Disease Self-Efficacy Scale (CDSES), Parent's Arthritis Self-Efficacy Scale (PASE), and Rheumatoid Arthritis Self-Efficacy Scale (RASE). *Arthritis Care Res (Hoboken)* 2011; **63**(Suppl 11): S473–85.
30 Miles CL, Pincus T, Carnes D, Taylor SJ, Underwood M. Measuring pain self-efficacy. *Clin J Pain* 2011; **27**: 461–70.
31 Bland JM, Altman DG. Cronbach's alpha. *BMJ* 1997; **314**: 572.
32 Upton P, Lawford J, Eiser C. Parent-child agreement across child health-related quality of life instruments: a review of the literature. *Qual Life Res* 2008; **17**: 895–913.
33 van der Ende J, Verhulst FC. Informant, gender and age differences in ratings of adolescent problem behaviour. *Eur Child Adolesc Psychiatry* 2005; **14**: 117–26.
34 Breakey VR, Blanchette VS, Bolton-Maggs PH. Towards comprehensive care in transition for young people with haemophilia. *Haemophilia* 2010; **16**: 848–57.
35 Hauser ES, Dorn L. Transitioning adolescents with sickle cell disease to adult-centered care. *Pediatr Nurs* 1999; **25**: 479–88.

