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The use of a disability model in haemophilia research

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Summary. Most haemophilia research is medically orientated. However, assessment of the impact of disease on the individual is different when viewed from a rehabilitation perspective. Several models are available to explore functioning and health from this perspective. The disablement process (DP) is such a model, and the aim of this study was to introduce this process in haemophilia research to see whether this type of research could lead to meaningful data. Forty-three adult patients with severe haemophilia participated in this study in which the three 'main pathway' domains of the DP (impairments, functional limitations and disability) and two additional factors (intra-individual and risk factors) were addressed. Three questionnaires (HAL, Dutch-AIMS2 and IPA) were incorporated, and Pettersson scores for 21 patients were retrieved. Step-wise and hierarchical regression analysis was used to assess relationship between the various domains. Arthropathy explained 48% of the variance in functional limitations and nearly 25% of the variance in disability. Functional limitations explained 54% of the variance in disability. Patients identified pain as an important aspect of health which addressed 22% and 13% of the variance in functional limitations and disability respectively. Age was correlated with arthropathy ($r = 0.85$; $P < 0.001$), whereas psychological health correlated with pain ($r = 0.67$; $P < 0.001$). Both variables were also correlated with functional limitations and disability. Analyses adjusting for the effects of age and psychological health were subsequently performed resulting in more insight in the associations within the DP. The use of the DP in haemophilia research proved to be useful.

Keywords: disablement process, Dutch-AIMS2, haemophilia, HAL, questionnaire

Introduction

Patients with severe haemophilia (clotting factor activity <1% of normal) often experience haemorrhagic episodes, most commonly haemarthroses. Repeated joint bleeds (usually in the ankle, knee and elbow joints) ultimately result in crippling deformities, which cause chronic pain and result in functional limitations and disability and may require surgical interventions. The regular treatment for haemophilia is factor replacement therapy, either on-demand or prophylactic. Most research in haemophilia is therefore focused on the use and effects of clotting factors and is thus medically orientated. However, assessment of the impact of disease on functioning of the individual is different when viewed from a more broad rehabilitation perspective [1]. From this perspective, patients' functioning and health are associated with, and not merely a consequence of, a condition or disease as from a medical perspective. Additionally, functioning and health are not only associated with the underlying disease, but also with, and influenced by, personal and environmental factors [2].

Several conceptual approaches are available to describe and measure functioning and health from a rehabilitation perspective. One such model is the disablement process (DP) introduced by Nagi [3] and further elaborated by Verbrugge and Jette [4,5]. The

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DP is a conceptual scheme for disability that is used to describe how acute and chronic conditions may affect functioning in specific body systems, fundamental physical and mental actions, and activities of daily life [4]. The DP consists of a pathway with four distinct domains: 'pathology' (i.e. diagnosis or disease), 'impairments' (i.e. dysfunctions and structural abnormalities in specific body systems), 'functional limitations' (i.e. restrictions in basic physical and mental actions), and 'disability' (i.e. difficulty doing activities of daily life) [4]. These four domains make up the 'main pathway' of the process. Several factors may influence the outcome of this pathway; these factors can be classified into 'intra-individual factors' (e.g. coping, lifestyle), 'extra-individual factors' (e.g. medical care, rehabilitation) and 'risk factors' (environment, predisposition).

The DP has been successfully used in other patient populations, such as in those with rheumatoid arthritis [6–8], osteoarthritis [9], elderly [10], cervical spine disorders [11], pulmonary disease [12] and children with haemophilia [13]. These studies yielded interesting results that allow for a change of scope of therapy by describing the consequences of disease that are of great interest for rehabilitation; for example, children with moderate and severe haemophilia were comparable with their healthy peers regarding motor performance and activities of daily living. A majority of these children (79%) however perceived a significant impact of their disease, which was associated with pain and restriction in sports [13]. The DP has never been used to describe the health status of adult patients with haemophilia. However, it might prove its worth in identifying new targets for interventions (besides the 'usual' medical interventions such as replacement therapy) to prevent and/or reduce the long-term consequences of haemophilic arthropathy in adult patients with haemophilia. Therefore, the aim of this pilot study was to introduce the concept of the DP in adult patients with haemophilia and see whether this type of research could lead to meaningful data.

Methods

Forty-three consecutive patients (≥18 years old) with severe haemophilia were invited to participate in this pilot study during their annual check-up at the Van Creveldkliniek. No acute pathology (e.g. bleeding) was present in any patient at the time of assessment. The Medical Ethics Committee of the University Medical Centre Utrecht approved the study. Three questionnaires were administered: [1] the Haemophilia Activities List (HAL [14]), [2] the Dutch Arthritis Impact Measurement Scales-2 (Dutch-AIMS2 [15,16]) and [3] the Impact on Participation and Autonomy questionnaire (IPA [17–20]). The questionnaires were handed out and patients were asked to complete them at home and send them back using a return stamped envelope. No specific order of administration of the questionnaires was given. Besides the questionnaires, records of all patients were searched to retrieve the most recent Pettersson scores [21]. Only X-rays taken within 5 years of the current assessment were considered for evaluation (current assessment using the three questionnaires ±2.5 years).

Questionnaires

The HAL is a recently developed, haemophilia-specific questionnaire, evaluating the self-perceived abilities of patients with haemophilia with regard to performing activities of daily life [14]. Face and expert validity for the HAL were established, as well as the internal consistency and convergent validity [14]. This self-administered questionnaire encompasses 57 items, distributed over eight domains. For each domain a score was calculated, as well as an overall summarized score for the HAL (HAL_{sum}). Moreover, three component scores were calculated: Upper Extremity Activities (HAL_{upper}; e.g. reaching, carrying heavy objects), Lower Extremity Activities (HAL_{lower}; e.g. walking) and Key Activities (HAL_{key}; e.g. running, jumping). Additionally, an item assessing the use of walking aids was added to evaluate the needs for a walking aid in a non-bleeding situation.

The Dutch-AIMS2, originally designed for use in patients with rheumatoid arthritis [22], evaluates physical, psychological and social aspects of patients; its application in haemophilia was recently validated [15,16]. This self-administered questionnaire encompassed 81 items, distributed over 12 health status scales and five components: physical ($AIMS2_{physical}$), affect ($AIMS2_{affect}$), symptoms ($AIMS2_{symptom}$), social/interaction ($AIMS2_{social}$) and role ($AIMS2_{role}$). Moreover, a sum scores for the entire questionnaire was calculated ($AIMS2_{sum}$).

The IPA is a generic questionnaire, addressing the personal impact of illness on participation and autonomy and related experience of problems [17–20]. The self-administered questionnaire consisted of 31 items, distributed over five domains: autonomy indoors ($IPA_{auto\text{-}in}$), family role ($IPA_{famrole}$), autonomy outdoors ($IPA_{auto\text{-}out}$), social relations (IPA_{socrel}), and work and educational opportunities ($IPA_{workedu}$). A higher score represented increased

restrictions in participation or worse problems [20]. Because the IPA has not yet been validated haemophilia, the internal consistency of the questionnaire was assessed. Cardol *et al.* [17] obtained the following reliability coefficients (Cronbach's α coefficient) for each of the subscales: autonomy indoors (0.91), family role (0.90), autonomy outdoors (0.81), social relations (0.86), and work and educational opportunities (0.91). In our sample, we obtained similar coefficients (0.93, 0.93, 0.90, 0.90 and 0.92 respectively), which led us to consider the IPA as an instrument with satisfactory reliability indices in patients with haemophilia. The internal consistency for the summarized IPA score (IPA_{sum}) was 0.97.

Pettersson score

To assess haemophilic arthropathy, the Pettersson score was used [21]. Four separate scores were available: a summarized score, a score for the elbows, the knees and the ankles. Scores ranged from 0 (no abnormalities) to 13 (severe joint destruction) for each joint. This resulted in a range for the summarized Pettersson score from 0 to 78.

Domains of the disablement process

The first domain of the DP is pathology. In this study, only the medical diagnosis of severe haemophilia type A or type B was used in this domain. No differences were observed between the two types of haemophilia in this study ($P > 0.05$) and therefore this domain was omitted in further analyses. The results from the above mentioned questionnaires were used to address the other domains of the DP (i.e. impairments, functional limitations and disability) and the risk factors and intra-individual factors (Table 1). No extra-individual factors were recorded.

Data analysis

Data analysis was performed with SPSS 11.5, using a significance level of $\alpha = 0.05$. First, all data were assessed for normal distribution using the Kolmogorov–Smirnov test for normality. Descriptive statistics for all three questionnaires were calculated in order to give a detailed description of the population studied. To simplify interpretation of the scores of the different questionnaires (used only for the descriptive statistics), all scores were normalized to a score ranging from 0 (very good functional health status) to 100 (very bad functional health status). However, raw scores (not available for the five components of the Dutch-AIMS2) were used in all other statistical analyses. Subgroup analysis was performed using one-way analysis of variance (ANOVA) or a paired samples *t*-test, where appropriate.

Using the DP data were assessed in four consecutive steps. As a first step, correlation coefficients (Pearson's r or Spearman's ρ) were calculated to allow for linear regression analyses ($r = 0.4$–0.9). Secondly, (multiple) linear regression analyses (stepwise procedure) were used to investigate whether associations between the different domains of the main pathway existed. Thirdly, the effects of risk factors and intra-individual factors on the main pathway domains were calculated. As a last step, hierarchical linear regression analysis was applied (enter procedure) to adjust for possible effects of age and psychological health. For all regression analyses, the residuals (unstandardized) were assessed for normality using the Kolmogorov–Smirnov test.

Table 1. Variables used in the analysis of the disablement process (DP).

DP Domains	Variable	Instrument/outcome
Pathology*	Haemophilia type	n/a
	Haemophilia severity	n/a
Impairments	Haemophilic arthropathy	$Pettersson_{sum}$
		$Pettersson_{elbow}$
		$Pettersson_{knee}$
		$Pettersson_{ankle}$
	Pain due to haemophilia	$Dutch\text{-}AIMS2_{symptom}$
Functional limitations	Self-perceived ability to perform activities	HAL_{sum}
		HAL_{upper}
		HAL_{lower}
		HAL_{key}
Disability	Self-perceived autonomy and participation	IPA_{sum}
		$IPA_{auto\text{-}in}$
		$IPA_{famrole}$
		$IPA_{auto\text{-}out}$
		IPA_{socrel}
		$IPA_{workedu}$
Risk factors	Age	Age (years)
Intra-individual factors	Psychological health	$Dutch\text{-}AIMS2_{affect}$
Extra-individual factors	n/a	n/a

*Only patients with severe haemophilia type A or type B participated. No differences were observed between the two types of haemophilia in this study ($P > 0.05$) and therefore this domain was omitted in further analyses.
n/a, not applicable.
HAL_{sum}, IPA_{sum}: these variables were designated as dependent variables in the regression analyses, when appropriate.

Results

Forty-three patients [average age 45 years (SD 14; range 18–68)] participated in this study. All patients

were male and had severe haemophilia (<1% clotting activity), 37 had haemophilia A, six patients had haemophilia B.

Descriptives

Pettersson scores were available for 21 of the 43 patients (Table 2). The median Pettersson sum score was 40 (IQR 16–69). Eleven patients scored the maximum of 13 points for 17 knee joints and 15 ankle joints; arthroplasty was performed on six knees and arthrodesis on five ankles.

All 43 patients completed the three questionnaires. The results of the questionnaires are presented in Table 3. Eight variables differed significantly from a normal distribution: the HAL_{upper}, HAL_{key}, Dutch-$AIMS2_{affect}$, Dutch-$AIMS2_{role}$, $IPA_{workedu}$, $Pettersson_{elbow}$, $Pettersson_{knee}$ and $Pettersson_{ankle}$.

Haemophilic pain, as reported in the Dutch-AIMS2, was a major problem for most of the respondents. The highest scoring (i.e. worst) component of the Dutch-AIMS2 is the 'symptom'-component, which is directly related to pain due to haemophilia. Moreover, when asked to identify three areas of health in which they would most like to see improvement (item 60 of the Dutch-AIMS2), 63% of the patients ($n = 27$) indicated 'haemophilic pain' as the most important.

Patients reported problems with activities involving the lower extremities most frequently. For the HAL, the two domains relating directly to the legs (LEGS and LSKS) scored worst. In the Dutch-AIMS2, the 'Walking and Bending' scale was the highest scoring scale of this questionnaire, which corresponded to the findings in the HAL. Moreover, besides pain as being identified as an area for improvement, 'Walking and Bending' was mentioned by 27 patients (63%) as well, and 'mobility level' was the third area in which 21 patients (49%) most liked to see improvement.

Regarding the IPA, the patients identified the most problems in their autonomy outdoors ($IPA_{auton\text{-}out}$), and their family role ($IPA_{famrole}$), whereas few problems were reported regarding their social relations (IPA_{socrel}) and autonomy indoors ($IPA_{auton\text{-}in}$).

Table 2. Pettersson scores ($n = 21$).

	Median	IQR
$Pettersson_{sum}$	40	16–69
$Pettersson_{elbow}$*	10	5–25
$Pettersson_{knee}$*	13	0–26
$Pettersson_{ankle}$*	17	8–26

IQR, interquartile range.
*Data are not distributed normally.

Table 3. Results of the three questionnaires ($n = 43$).

	Normalized scores (0 = good, 100 = bad)	
	Median	IQR
HAL		
Separate domains		
Lying/sitting/kneeling/standing (LSKS)	40.0	28.2–58.2
Functions of the legs (LEGS)	66.6	42.2–80.0
Functions of the arms (ARMS)	16.0	8.0–44.0
Use of transportation (TRANS)	30.0	10.0–40.0
Self-care (SELFCA)	9.1	0.0–26.4
Household tasks (HOUSEHO)	26.2	11.4–42.9
Leisure activities and sport (LEISPO)	32.9	10.0–47.5
Other (OTHER)	0.0	0.0–15.0
SUM score (HAL_{sum})	31.3	20.7–44.7
Upper extremity component (HAL_{upper})*	14.7	4.2–32.6
Lower extremity component (HAL_{lower})	52.7	30.9–66.7
Key activity component (HAL_{key})*	86.7	53.3–100.0
Dutch-AIMS2		
SUM score ($AIMS_{sum}$)	29.3	18.9–35.0
Physical component ($AIMS_{phys}$)	16.3	10.8–28.5
Affect component ($AIMS_{affect}$)*	27.5	20.0–37.5
Symptom component ($AIMS_{symptom}$)	40.0	25.0–47.5
Social component ($AIMS_{social}$)	35.0	25.0–41.3
Role component ($AIMS_{role}$)*	18.8	0.0–37.5
IPA		
SUM score (IPA_{sum})	23.1	8.4–38.5
Autonomy indoors ($IPA_{auto\text{-}in}$)	25.0	5.4–30.4
Family role ($IPA_{famrole}$)	32.1	14.3–51.8
Autonomy outdoors ($IPA_{auto\text{-}out}$)	35.0	15.0–50.0
Social relations (IPA_{socrel})	20.8	8.3–31.3
Work & education ($IPA_{workedu}$)*	25.0	16.7–45.8

IQR, interquartile range.
*Data are not distributed normally.

Sixteen patients indicated using a walking aid (37.2%). Most frequently, one or two crutches were used ($n = 11$ and 8 respectively). A comparison between these patients and those not using a walking aid, showed that the former group had significantly more problems with performing activities than the latter (as represented by all HAL domains and components, the Dutch-AIMS2 component of 'mobility' and the 'autonomy indoors' and 'autonomy outdoors' domains of the IPA). Patients using a walking aid were also significantly older than those not using such an aid (on average 51 years vs. 41 years; $P = 0.017$).

Relationships within the DP

Analysis of the correlation coefficients revealed no multicollinearity ($r = 0.90$ or above; correlation coefficients are not published).

Pain was not dependent on haemophilic arthropathy, even when adjusting for age ($P = 0.899$) or

whether or not the patient received orthopaedic surgery ($P = 0.303$). Arthropathy in the knee explained nearly half of the total variance in 'functional limitations', and 24.2% of the total variance in the 'disability' domain (see Fig. 1). For pain, these associations were 22.5% and 13% respectively. When combining pain and arthropathy in the knee, over half of the total variance of 'functional limitations' could be explained. Of all variance in the 'disability' domain, over 38% could be explained by the combination of pain and haemophilic arthropathy. The sum score HAL was the primary variable to explain the variance in 'disability': over half of the total variance of IPA_{sum} was explained by HAL_{sum}.

Age was the primary 'predictor' of haemophilic arthropathy, but was also significantly associated (>20% variance explained) with the 'functional limitations' and 'disability' domains. Age did not associate with pain. Psychological health explained the variance in pain the most, but again significant associations existed with 'functional limitations' and 'disability' (>20% variance explained). There was no significant association between psychological health and haemophilic arthropathy or age.

When adjusting for age, haemophilic arthropathy explained nearly half of the total variance in 'functional limitations' and nearly a quarter in 'disability'. The contribution of arthropathy (i.e. $Pettersson_{knee}$) in these models was 15.1% and 4.1% respectively (see Fig. 2). Pain, when adjusted for psychological health, accounted for 5.1% of the total variance in 'functional limitations' and 0.1% of the total variance in 'disability'. Combining both haemophilic arthropathy and pain, and adjusting for age and psychological health, the 'impairment' domain accounted for 16.8% and 2.6% of the 'functional limitations' domain and 'disability' domain respectively. The association between 'functional limitations' and 'disability' remained stronger after adjusting for both age and psychological health: of the total amount of 61.5% explained variance in the IPA_{sum} ($P < 0.001$), the HAL_{sum} addressed 15.4%.

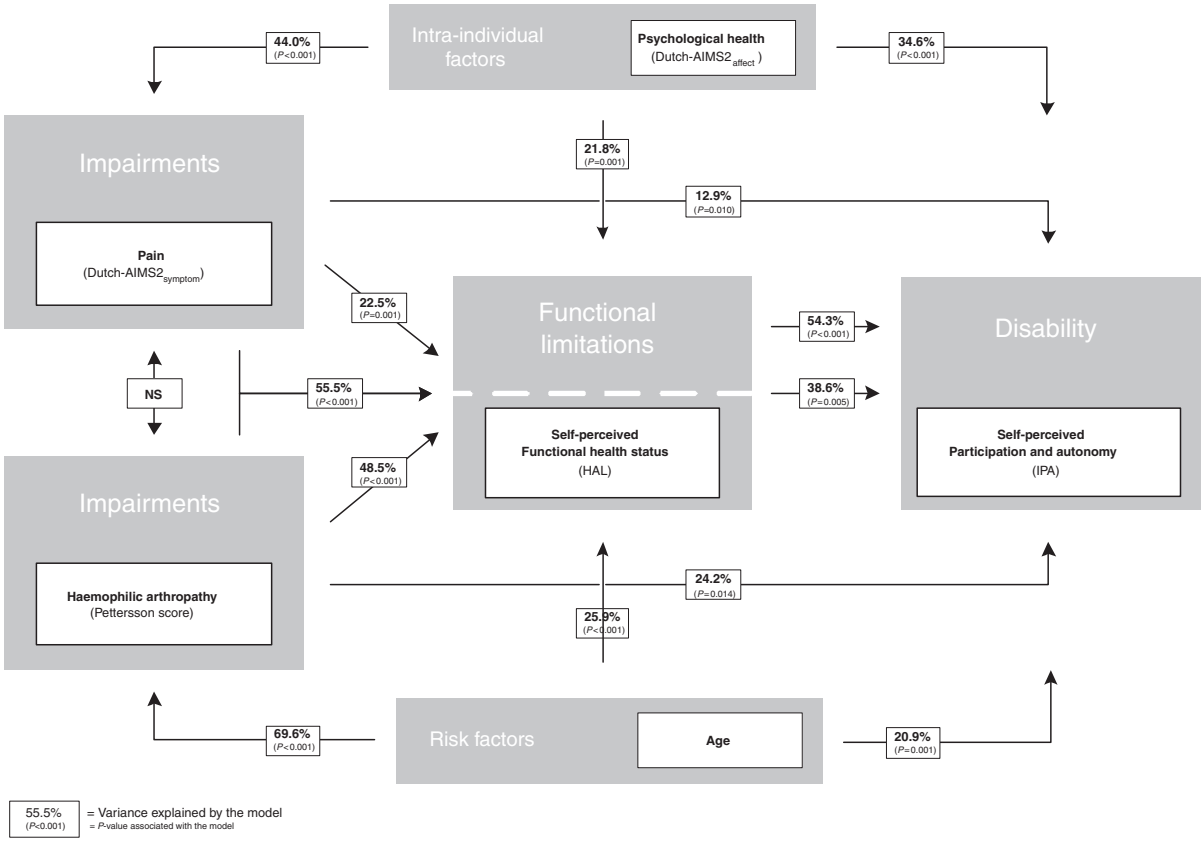

Fig. 1. Results of the linear regression analyses within the 'disablement process'. The domain of 'body structures' and 'functions' is divided into 'pain' and 'haemophilic arthropathy'. Interpretation (box starting with 55.5%): Analysis of the effect of the independent Impairments [represented by 'Pain' ($Dutch\text{-}AIMS2_{symptom}$) and 'Haemophilic Arthropathy' (Pettersson scores)] on the dependent 'Self-perceived Functional Health Status' (represented by HAL_{sum}) resulted in a model explaining a total of 55.5% (Adjusted $R^2 \times 100$) of the total variance in HAL_{sum} with a significance of $P < 0.001$.

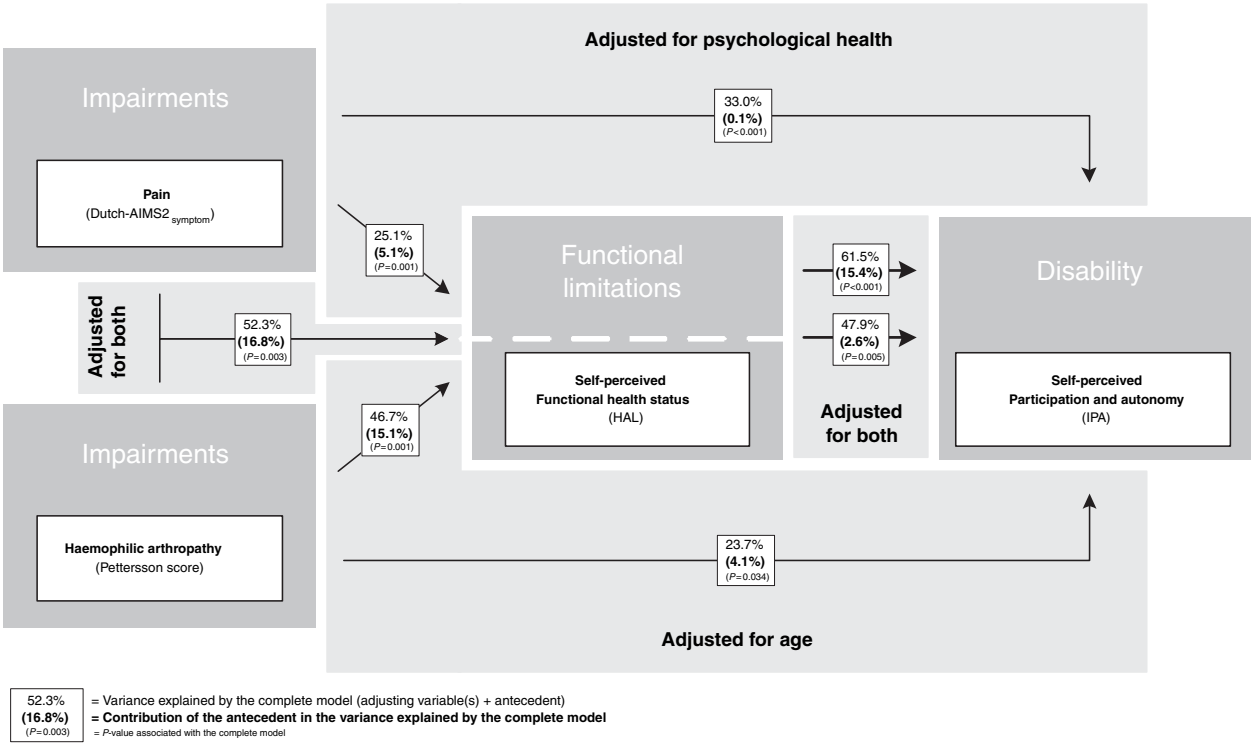

Fig. 2. Results of the regression analyses adjusting for age and/or psychological health. The domain of 'body structures' and 'functions' is divided into 'pain' and 'haemophilic arthropathy'. Interpretation (box starting with 52.3%): Hierarchical regression analysis of the independent Impairments [represented by 'Pain' (Dutch-AIMS2$_{symptom}$) and 'Haemophilic Arthropathy' (Pettersson scores)] on the dependent 'Self-perceived Functional Health Status' (represented by HAL$_{sum}$), adjusting for Psychological Health and Age, resulted in a model explaining a total of 52.3% (Adjusted $R^2 \times 100$) of the total variance in HAL$_{sum}$ with a significance of $P < 0.001$. The contribution of Pain and Haemophilic Arthropathy in the total amount of variance explained by the complete model was 16.8%.

Discussion

The aim of this study was to introduce the DP as a model for disability research in haemophilia. We found significant associations within the main pathway of the DP. Risk and intra-individual factors had an important influence on these associations, and therefore on the (final) outcome.

This pilot study was designed to see whether the introduction of the DP reveals meaningful data for haemophilia as a field of research. Nonetheless, the results from this study can be directive for both research and (clinical) care in future.

Over half of the total variance in the consecutive DP domains can be explained by the preceding domain (Fig. 1). The explanatory value of each domain becomes even more apparent when adjusting for both age and psychological health (Fig. 2). This phenomenon was also seen in immune-mediated polyneuropathies [23] and community-dwelling elderly (60–94 years [24]). Whether these relations are causal has to be investigated in a longitudinal study, for transversal data (as are used here) do not give insight into the causality of the relations found.

A dichotomy appears to exist within the 'impairments' domain: on the one hand, haemophilic arthropathy and on the other, pain. Both variables were found not to be associated, but age and psychological health were significantly associated with arthropathy and pain respectively. When using the DP as a research model in haemophilia, one has to take this dichotomy into account. It would be worthwhile to reassess this dichotomy in another set of data and, if it persists, to address both concepts separately, as is shown in Figs 1 and 2. Combining both concepts is acceptable when the contribution of each concept can be retrieved.

A notable finding is the fact that pain is an important issue for the patients. Not only was pain the highest scoring domain in the Dutch-AIMS2, but also a majority of the patients addressed pain resulting from haemophilia as an area of health in which they would most like to see improvement. Pain also plays a significant role when relating this

'impairment' variable with both the 'functional limitations' and the 'disability' domain. The finding that pain does not relate to age corresponds with previous research [25], but there is discord about the relationship between pain and arthropathy: Aznar *et al.* [26] found no relationship between the Bodily Pain dimension of the SF-36 questionnaire and the clinical GiIbert score, which corresponds to our data. Wallny *et al.* [27] and Triemstra *et al.* [28] however, found a significant relationship between pain in the knee and arthropathy in the knee or between pain and joint damage as reported by the patients themselves respectively. In the current study, patients who received orthopaedic surgery scored 13 points on the Pettersson score for the concerning joint, as was done in previous research [29,30]. In general, orthopaedic surgery is primarily performed to alleviate pain. Therefore, it can be hypothesized that patients who received orthopaedic surgery (and therefore a Pettersson score of 13 for that specific joint) might report less pain. Adjusting for surgery however had no effect on the significance of the relationship between arthropathy and pain. So, it seems that the use of the Pettersson score needs further investigation when using it as a measure within the DP.

The contribution of pain in the regression models where 'functional limitations' or 'disability' are the outcomes, is reduced to 5.1% and 0.1% respectively (Fig. 2) when adjusting for psychological health. Psychological health can therefore be regarded as a mediator, for it accounts for the relation between the antecedent and the outcome [31]. In the case of 'disability', psychological health might even be addressed as a full mediator, meaning that the effect of pain on disability seems to be fully mediated by psychological health. Thus, although the pain score is high in our study population, the effect it has on both 'functional limitations' and 'disability' seems of minor importance. Underlying psychological mechanisms, such as coping behaviours, may be more important here and future, in-depth research might shed some light on this paradox. In practice, it would mean that when one wants to control the effect pain has on functional limitations or disability, psychological health might be the starting point for the intervention. It has been suggested that cognitive behavioural therapy [32] may also be an efficient method to combat haemophilic arthropathy-induced pain [27,33]. This area definitely needs further exploration.

The use of a summarized score for the IPA questionnaire as the primary 'disability' outcome can be discussed. Such a score is not originally included in the questionnaire, but as a main outcome parameter for 'disability' was needed, we felt confident to summarize the different scales of the IPA to obtain such an outcome. The high Cronbach α statistic as a measure for internal consistency (0.97) supports the use of this score.

In this study, only self-rating instruments (i.e. questionnaires) were used. It is known from the literature that a discrepancy exists between self-rating instruments and performance-based instruments [34]. Therefore both types of instruments should be used in future studies [35].

The use of a disability model in haemophilia research is not entirely new. Triemstra *et al.* [28] introduced a model which was partially based on the International Classification of Impairments, Disability and Handicaps [36]. Furthermore, the successor of the ICIDH, the International Classification of Functioning, Disability and Health (ICF [37]) has been the basis for two literature studies to identify suitable measurements in haemophilia [35,38]. There are many similarities between the DP and the model proposed by the World Health Organization in the ICF. The view that overall disablement is represented by a series of related concepts that describe the consequences of a disease on different domains is similar [5] and the domains also share many communalities. The dichotomy (arthropathy and pain) that was found in the 'impairments' domain can easily be used in the corresponding domain of the ICF where such a split is already made: arthropathy resides under 'body structures', whereas pain is located in the 'body functions' domain. Using either of these disability models in haemophilia will help to further unravel the course of functional health status in this chronic disease and tailor individual care. Results from the studies by De Kleijn *et al.* [35,38] in which possible clinimetric instruments matching each of the three above-mentioned ICF domains are identified will be helpful to find suitable measurements.

Conclusion and recommendations

The results of this study suggest that the DP is a suitable model in haemophilia research to investigate associations between several aspects of a persons health status. When using the DP or a similar model in haemophilia research, one has to account for the dichotomy between pain and haemophilic arthropathy that seems to exist within the 'impairment' domain. Moreover, the role of pain and psychological health need further exploration. Future research has to incorporate measurement tools that fit all domains used within the research model, be it the DP or the ICF, including both the patient's as well as the

doctor's perspective on the patient's health status. Ultimately, a core set of instruments that cover all domains of the DP for patients with haemophilia should be composed and used in both research and care.

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References

1 Stucki G, Ewert T, Cieza A. Value and application of the ICF in rehabilitation medicine. *Disabil Rehabil* 2003; **25**: 628–34.
2 Stucki G, Sigl T. Assessment of the impact of disease on the individual. *Best Pract Res Clin Rheumatol* 2003; **17**: 451–73.
3 Pope, AM, Tarlov, AR. *Disability in America: Toward a National Agenda for Prevention*. Washington, D.C.: National Academy Press, 1991.
4 Verbrugge LM, Jette AM. The disablement process. *Soc Sci Med* 1994; **38**: 1–14.
5 Jette AM, Keysor JJ. Disability models: implications for arthritis exercise and physical activity interventions. *Arthritis Rheum* 2003; **49**: 114–20.
6 Escalante A, Del Rincon I. The disablement process in rheumatoid arthritis. *Arthritis Rheum* 2002; **47**: 333–42.
7 Escalante A, Del Rincon I. How much disability in rheumatoid arthritis is explained by rheumatoid arthritis? *Arthritis Rheum* 1999; **42**: 1712–21.
8 Hazes JM. Determinants of physical function in rheumatoid arthritis: association with the disease process. *Rheumatology (Oxford)* 2003; **42** (Suppl. 2): ii17–21.
9 Weigl M, Cieza A, Harder M *et al.* Linking osteoarthritis-specific health-status measures to the International Classification of Functioning, Disability, and Health (ICF). *Osteoarthritis Cartilage* 2003; **11**: 519–23.
10 Kempen GIJM, Van Heuvelen MJG, Van Sonderen E *et al.* The relationship of functional limitations to disability and the moderating effects of psychological attributes in community-dwelling older persons. *Soc Sci Med* 1999; **48**: 1161–72.
11 Hermann KM, Reese CS. Relationships among selected measures of impairments, functional limitations, and disability in patients with cervical spine disorders. *Phys Ther* 2001; **81**: 903–14.
12 Jette DU, Manago D, Medved E *et al.* The disablement process in patients with pulmonary disease. *Phys Ther* 1997; **77**: 385–94.
13 Schoenmakers MAGC, Gulmans VA, Helders PJ, Van den Berg HM. Motor performance and disability in Dutch children with haemophilia: a comparison with their healthy peers. *Haemophilia* 2001; **7**: 293–8.
14 Van Genderen FR, Van Meeteren NLU, Van der Bom JG *et al.* Functional consequences of haemophilia in adults: the development of the Haemophilia Activities List. *Haemophilia* 2004; **10**: 565–71.
15 De Joode EW, Van Meeteren NLU, Van den Berg HM, De Kleijn P, Helders PJM. Validity of health status measurement with the Dutch Arthritis Impact Measurement Scale 2 in individuals with severe haemophilia. *Haemophilia* 2001; **7**: 190–7.
16 Van Meeteren NLU, Strato IHM, Van Veldhoven NHMJ *et al.* The utility of the Dutch Arthritis Impact Measurement Scale 2 for assessing health status in individuals with haemophilia: a pilot study. *Haemophilia* 2000; **6**: 664–71.
17 Cardol M, De Haan RJ, De Jong BA, Van den Bos GA, De Groot IJM. Psychometric properties of the Impact on Participation and Autonomy Questionnaire. *Arch Phys Med Rehabil* 2001; **82**: 210–6.
18 Cardol M, Beelen A, Van den Bos GA *et al.* Responsiveness of the Impact on Participation and Autonomy questionnaire. *Arch Phys Med Rehabil* 2002; **83**: 1524–9.
19 Cardol M, De Haan RJ, Van den Bos GA, De Jong BA, De Groot IJM. The development of a handicap assessment questionnaire: the Impact on Participation and Autonomy (IPA). *Clin Rehabil* 1999; **13**: 411–9.
20 Cardol M, De Jong BA, Van den Bos GA *et al.* Beyond disability: perceived participation in people with a chronic disabling condition. *Clin Rehabil* 2002; **16**: 27–35.
21 Pettersson H, Nilsson IM, Hedner U, Norehn K, Ahlberg A. Radiologic evaluation of prophylaxis in severe haemophilia. *Acta Paediatr Scand* 1981; **70**: 565–70.
22 Riemsma RP, Taal E, Rasker JJ *et al.* Evaluation of a Dutch version of the AIMS2 for patients with rheumatoid arthritis. *Br J Rheumatol* 1996; **35**: 755–60.
23 Merkies ISJ, Schmitz PIM, Van der Meche FGA, Samijn JPA, Van Doorn PA. Connecting impairment, disability, and handicap in immune mediated polyneuropathies. *J Neurol Neurosurg Psychiatry* 2003; **74**: 99–104.
24 Jette AM, Assmann SF, Rooks D, Harris BA, Crawford S. Interrelationships among disablement concepts. *J Gerontol A Biol Sci Med Sci* 1998; **53**: M395–404.
25 Choiniere M, Melzack R. Acute and chronic pain in hemophilia. *Pain* 1987; **31**: 317–31.
26 Aznar JA, Magallón M, Querol F, Gorina E, Tusell JM. The orthopaedic status of severe haemophiliacs in Spain. *Haemophilia* 2000; **6**: 170–6.
27 Wallny T, Lahaye L, Brackmann HH *et al.* Clinical and radiographic scores in haemophilic arthropathies: how well do these correlate to subjective pain status and daily activities? *Haemophilia* 2002; **8**: 802–8.

28 Triemstra AHM, Van der Ploeg HM, Smit C *et al.* Well-being of haemophilia patients: a model for direct and indirect effects of medical parameters on the physical and psychosocial functioning. *Soc Sci Med* 1998; **47**: 581–93.
29 Fischer K, Van der Bom JG, Mauser-Bunschoten EP *et al.* Changes in treatment strategies for severe haemophilia over the last 3 decades: effects on clotting factor consumption and arthropathy. *Haemophilia* 2001; **7**: 446–52.
30 Fischer K, Van Hout BA, Van der Bom JG, Grobbee DE, Van den Berg HM. Association between joint bleeds and Pettersson scores in severe haemophilia. *Acta Radiol* 2002; **43**: 528–32.
31 Baron RM, Kenny DA. The moderator-mediator variable distinction in social psychological research: conceptual, strategic, and statistical considerations. *J Pers Soc Psychol* 1986; **51**: 1173–82.
32 Vlaeyen JW, Morley S. Cognitive-behavioral treatments for chronic pain: what works for whom? *Clin J Pain* 2005; **21**: 1–8.
33 Wallny T, Hess L, Seuser A *et al.* Pain status of patients with severe haemophilic arthropathy. *Haemophilia* 2001; **7**: 453–8.
34 Hidding A, Van Santen M, De Klerk E *et al.* Comparison between self-report measures and clinical observations of functional disability in ankylosing spondylitis, rheumatoid arthritis and fibromyalgia. *J Rheumatol* 1994; **21**: 818–23.
35 De Kleijn P, Van Genderen FR, Van Meeteren NLU. Assessing functional health status in adults with haemophilia: towards a preliminary core set of clinimetric instruments based on a literature search in rheumatoid arthritis and osteoarthritis. *Haemophilia* 2005; **11**: 308–18.
36 World Health Organization. *International Classification of Impairments, Disability, and Handicaps (ICIDH)*. Geneva: World Health Organization, 1980.
37 World Health Organization. *International Classification of Functioning, Disability and Health (ICF)*. Geneva: World Health Organization, 2001.
38 De Kleijn P, Heijnen L, Van Meeteren NLU. Clinimetric instruments to assess functional health status in patients with haemophilia: a literature review. *Haemophilia* 2002; **8**: 419–27.

