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The Patient Education Program for Huntington's Disease (PEP-HD)

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Abstract. The goal of the Patient Education Program for Huntington's disease is to improve quality of life for patients and caregivers, to educate and train them in order to develop coping strategies to deal with psychosocial stressors. The program was derived from a standardized evidence-based program for Parkinson's disease. This pilot study assessed the feasibility of the program in Huntington's disease. Forty manifest patients with 28 caregivers and 19 premanifest carriers with 14 partners participated. Assessments for depression and anxiety, psychosocial burden, need for help, quality of life, coping, behavioral, motor and cognitive status were performed. After program completion, significant improvement of behavioral symptoms and anxiety was found for manifest HD patients, and they used a less passive coping style and more social support. Their caregivers reported less psychosocial burden. Premanifest carriers and their partners improved their coping by seeking social support more often. This pilot study demonstrated the feasibility of the program in Huntington's disease, especially in the manifest stage of the disease. Further research to assess the effectiveness of the program seems warranted.

Keywords: Huntington's disease, psychosocial aspects, training support, caregivers

INTRODUCTION

Huntington's disease (HD) is an autosomal dominant inherited neurodegenerative disorder with mean age of onset in middle age. The disease is characterized by progressive motor, psychiatric and cognitive symptoms, causing functional decline [1]. Among the psychiatric symptoms, prevalence rates of depression, anxiety, apathy and irritability between 33% and 69% have been reported [2]. Possible psychosocial stressors are feelings like sadness and anxiety about the cognitive and physical decline, changes in social roles, and children at risk. Loss of social support is a risk factor for depression [3]. The most important sources of social support and daily care are informal caregivers, like spouses. Informal caregivers are at risk for caregiver burden due to the complexity of the HD symptoms and the psychosocial consequences [4–6]. Psychosocial challenges for patients and caregivers not only exist in the period of manifest symptoms and signs. With the discovery of the HD gene and the possibility of genetic testing, a stage before onset of apparent symptoms was created in which people have the knowledge to become ill. This knowledge may lead to anticipatory stress, anxiety, preoccupation with impending symptoms, suicidal ideation and feelings of hopelessness [7, 8]. It also may influence important future planning issues, like reproductive decisions. Despite many recommendations for psychological intervention studies in HD [4, 5, 9, 10], no such study was performed thus far. Therefore, we adapted an available standardized program from another neurodegenerative disease: the Patient Education Program for Parkinson's disease (PEPP). In a recent randomized controlled trial, benefits for this program were found regarding PD patients' Qol and caregivers' psychosocial problems and need for help [11]. The goal of the program is to improve quality of life for patients and caregivers, to educate and train them in order to develop coping strategies to deal with psychosocial stressors. Techniques from the cognitive behavioral therapy (CBT) [12] were implemented like

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cognitive restructuring, systematic relaxation training, situational behavioral analysis and training in social skills. The program was adjusted for use in HD and named: the Patient Education Program for Huntington's disease (PEP-HD). The aim of this pilot study is to evaluate the feasibility of the PEP-HD in premanifest and manifest Huntington's disease.

MATERIALS AND METHODS

Participants

Participants were HD mutation carriers without manifest symptoms (further labeled as PM carriers) and HD patients with known HD symptoms (further labeled as HD patients) and their primary caregivers. A database of patients attending the outpatient neurological department of the Leiden University Medical Center (LUMC) or the outpatient department for Huntington's disease Nij Friesma Hiem (NFH) in Grou was used to select eligible participants. Inclusion criteria were the following: 1) DNA confirmed diagnosis by expanded trinucleotide (CAG) repeat in the HD (*HTT*) gene; 2) a total functional score (TFC) $\geq$5); 2) a Mini Mental State Examination score (MMSE) $\geq$23); and 3) no current psychotic symptoms or severe behavioral problems. Inclusion criteria were carried out by means of the documentation in the medical file from the last visit at the hospital. If no recent data (last year) were available, then data were obtained at the initial patient screening.

Eventually, an invitation letter was sent to 106 HD patients and 54 PM carriers to participate in the study with their partner (Fig. 1). Participation without partner was also possible, but participation of both was encouraged. Patients, who were not able or willing to participate, were considered as non-participants, and participants who stopped during the study or missed more than two sessions were considered as drop-out. The study was approved by the Medical Ethical Committee and all participants gave informed consent.

Procedure

A two-period single group pre-post study, in which participants served as their own control, was used because of statistical efficiency considering the relatively small Dutch HD population. Groups of four to seven PM carriers or HD patients and groups of their partners subsequently entered the study. They first received baseline assessment at the hospital two months prior to the program, then served as a control during two months, and then received second assessment one week before participation in the program. After eight weeks of PEP-HD intervention, they received post-assessment within two weeks afterwards.

Intervention

Carriers/patients and partners participated in separate, but parallel groups of 4–7 members. The program consisted of eight two-weekly sessions of 90 minutes duration. The PEP-HD groups were trained by healthcare professionals who followed two days of training for this intervention. The program's content was standardized across groups (Table 1); it is adapted from the detailed manual for Parkinson's disease (PD) [13, 14].

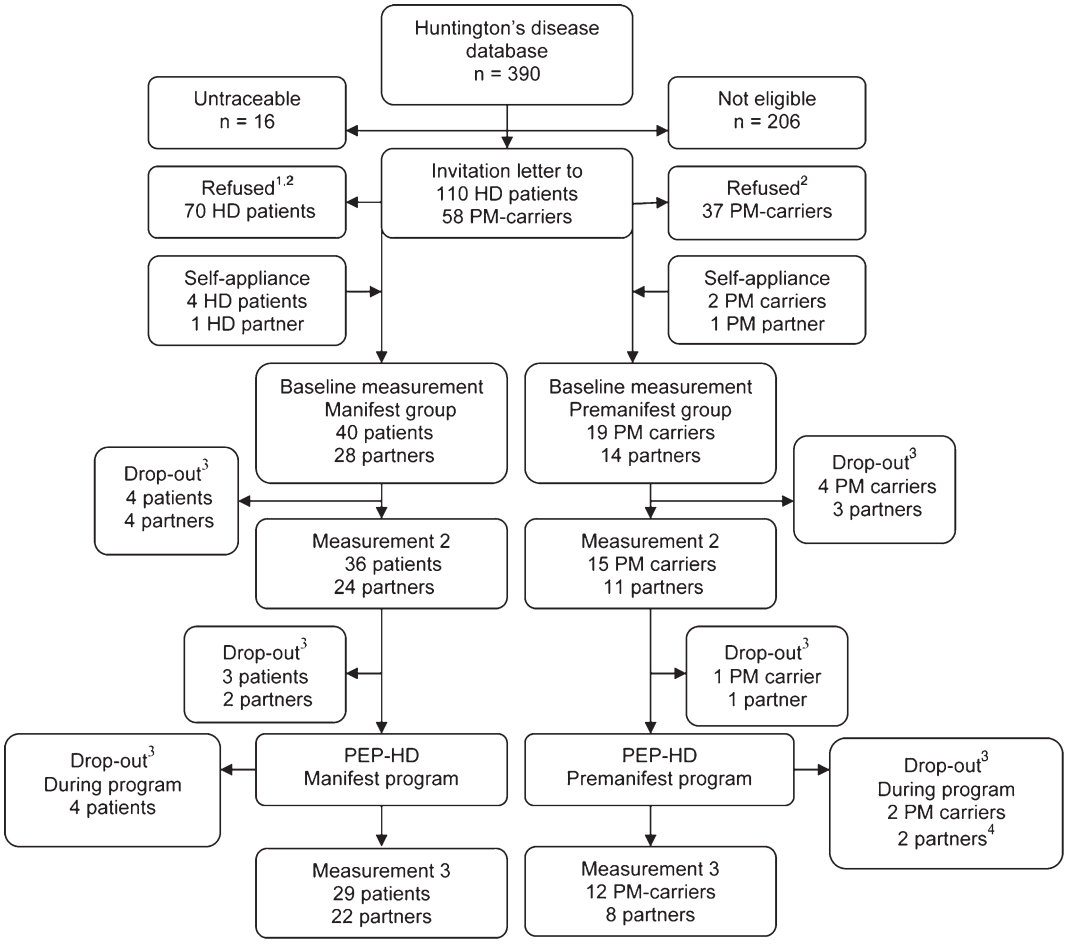
The core of the original program is based on generic coping strategies: session 1) taking a (pro) active role in treatment, seeking information about the disease; session 2) self-monitoring of body, behavior, cognitions, and mood; session 3) performing pleasant activities and relaxation; session 4) stress management by replacing unhelpful and unrealistic thoughts into helpful and realistic thoughts; session 5) dealing with or preventing depression and anxiety; session 6) social competence like communication and standing up for yourself; session 7) asking for social support. Session 8 is an overall rehearsal and program evaluation session. Because of these generic coping strategies, we hypothesized that the core of the program would be feasible for use in HD.

HD-specific adaptations to the program

Within the patient materials, examples specifically directed at PD were transformed into examples specifically directed at HD. For example, when discussing social stress due to visible symptoms, the tremor caused by PD was changed into involuntary movements caused by HD (chorea).

In session 1, we included HD specific information about where to find information about HD, such as HD websites, and names of HD professionals and HD specialized institutions.

Furthermore, the video materials used in session 5 and session 6 were made HD-specific. We developed video interviews about coping with HD for use in session 5. To respond to different needs of each subgroup, four interviews were recorded from the perspective of: 1) a HD patient, 2) a PM carrier, 3) a HD caregiver, or 4) a PM partner.



1 Two partners of manifest patients who refused to participate did participate themselves.

2 Arguments for non-participation: too much travel time (n = 30); too time-consuming (n = 27); too burdensome (n = 40); participation in group uncomfortable (n = 3); no need for/no interest (n = 19); not without partner (n = 1); unknown (n = 8).

3 Reasons for drop-out during study (n =8): too burdensome (3 M couples, 3 HD patients); participation in group not comfortable (1 HD patient and 1 PM carrier); personal circumstances (3 M and 3 PM couples, 1 PM carrier, 1 PM caregiver); death (1 HD patient); unknown (1 PM couple).

4 The two caregivers missed too much sessions due to personal circumstances.

Fig. 1. Flowchart of inclusion of subjects. [1]Two partners of manifest patients who refused to participate did participate themselves. [2]Arguments for non-participation: too much travel time (n = 30); too time-consuming (n = 27); too burdensome (n = 40); participation in group uncomfortable (n = 3); no need for/no interest (n = 19); not without partner (n = 1); unknown (n = 8). [3]Reasons lor drop-out during study (n = 8): loo burdensome (3 M couples, 3 HD patients); participation in group not comfortable (I HD patient and 1 PM carrier); personal circumstances (3 M and 3 PM couples, 1 PM carrier, 1 PM caregivcr); death (1 HD patient); unknown (1 PM couple). [4]The two caregivers missed too much sessions due to personal circumstances.

For session 6 (social competence) of the HD patients/PM carriers groups, video education material of the Dutch Huntington's Disease Association was selected to initiate a discussion about disclosure challenges of having HD, for example at work. For the PM carriers group, an interview was recorded with a HD caregiver discussing how to communicate with the patient about (beginning) HD symptoms. For the PM partner/HD caregiver group, video education material of the Dutch Huntington's Disease Association was selected about disclosure challenges about behavioral problems of the patient and challenges of asking for social support.

Assessment

Demographics were administered. The Unified Huntington's Disease Rating Scale (UHDRS) [15] was

Table 1
Thematic structure of PEP-HD

PEP-HD sessions	Structure	Main focus
1 Information	Introduction	The acquaintance of the participants and an overview of the program
	Active information	The importance of taking an active and central role in the health care system Advantages of information about HD. Where to find information
	Exercise	How to ask questions to health care professionals
	Homework	To draft questions for a visit to professionals
	Appetizer	Past experiences with keeping a diary/journal
2 Self-monitoring	Homework discussion	Homework discussion of session 1
	Active information	To learn about self-monitoring techniques, like a diary
	Exercise	An exercise 'body awareness' focused on breathing and muscular tensions
	Homework	Option 1: Using a diary to record i.e. fluctuations in mood or HD symptoms Option 2: Performing the exercise 'body awareness'
	Appetizer	Bringing something pleasant to the next session (i.e. an object or experience)
3 Health Promotion	Homework discussion	Homework discussion of session 2
	Active information	To improve wellbeing through pleasant activities
	Exercise	Exploring pleasant activities
	Homework	Performing a new pleasant activity every day
	Appetizer	Observing your own behavior in a stressful situation
4 Stress Management	Homework discussion	Homework discussion of session 3
	Active information	The role of unrealistic and unhelpful thoughts in stressful situations
	Exercise	Option 1: Learning to use alternative ways of thinking Option 2: Performing relaxation exercises to deal with stress
	Homework	Option 1: Trying out alternative ways of thinking Option 2: Relaxation training
	Appetizer	Observing changes of mood and causes of worry
5 Management of anxiety and depression (patients)/Caregiver's challenge	Homework discussion	Homework discussion of session 4
	Active information	To teach about the difference between normal feelings of anxiety and sadness and when they turn into anxiety disorders or depression/caregiver overload Second, learning about the role of unrealistic, unhelpful cognitions
	Exercise	Option 1: Positive thoughts Option 2: Maintaining healthy activities Discussion of a video clip of a HD patient/PM carrier/HD caregiver or PM partner telling about coping with the disease
	Homework	Option 1: Thinking of a positive event Option 2: Maintaining healthy activities
	Appetizer	Noticing situations in which you want to express your thoughts and feelings but not being able or having the confidence to do so
6 Social Competence	Homework discussion	Homework discussion of session 5
	Active information	Social skills like ways to communicate are discussed. Option 1: Unhelpful and helpful thoughts in communication Option 2: Ways of communication
	Exercise	Discussion of a video clip addressing communication problems
	Homework	(Patient/carrier group video: communication about having HD/being a HD gene carrier; caregiver/partner group video: communication about behavioral problems like aggression/communication about first symptoms) Option 1: Noting situations in which unhelpful thoughts contribute to a lack of socially competent behavior Option 2: Telling someone that you have HD
	Appetizer	To focus on the informal or formal support they would like to receive
7 Social Support	Homework discussion	Homework discussion of session 6
	Active information	To discuss the importance of and how to obtain social support
	Exercise	Role play/discussion
	Homework	Finding sources of support and asking for support
	Appetizer	Reflecting about the entire education program
8 Evaluation	Homework discussion	Homework discussion of session 7
	Active information	The group goes through the previous sessions and the program is evaluated. Expectations described in the first session and achievements are compared
	Exercise	Writing a postcard for each other and filling in a final evaluation questionnaire

Abbreviations: PEP-HD, Patient Education Program for Huntington's disease; HD, Huntington's disease; PM, premanifest.

used to assess disease signs. The UHDRS provides a motor, functional and cognitive score. A neurologist performed motor (a higher score indicating more motor symptoms) and functional assessment (Total Functional Capacity (TFC), a higher score indicating better functioning in daily life). Cognitive functioning was measured by the sum of raw scores (a higher score indicating better cognitive functioning) of three neuropsychological tests: Symbol Digit Modalities Test (SDMT) [16]; Stroop color-word test [17]; and Controlled Oral Word Association Test (FAS) [18]. Additionally, general cognitive functioning was assessed with the Mini Mental Status Examination (MMSE) [19] (a higher score indicating better general cognitive functioning). At last, the UHDRS provides a behavioral score by the sum of the product of severity and frequency per behavioral problem (a higher score indicating more behavioral problems). Cognitive and behavioral assessments were performed by a neuropsychologist. Medication and changes in medication were recorded by means of participants' self-report.

The following self-report questionnaires were administered. The Hospital Anxiety and Depression Scale (HADS) provides an anxiety and depression score (a higher score indicating more depression/anxiety) [20, 21]. Quality of life (divided into mental and physical) was measured with the generic 36-item Short Form health survey questionnaire (SF-36) (a higher score indicating better quality of life) [22, 23]. Psychosocial burden and need for help were assessed by an adapted version of the 'Belastungsfragebogen Parkinson kurzversion' (BELA-P-k) [24]. This questionnaire includes a partner version, the 'Belastungsfragebogen Parkinson Angehörigen kurzversion' (BELA-A-k) [25] (a higher score indicating more psychosocial burden or need for help). Coping strategies were measured with the Utrecht Coping List (UCL) [26, 27]. Before and after each session of the PEP-HD, participants were asked to rate their present mood on a 100-point Visual Analogue Scale (Mood-VAS) (a higher score indicating better mood) [28].

After completion of the program, participants were asked to fill out a program evaluation questionnaire.

Statistical analysis

The data were analyzed with the Statistical Package for the Social Sciences (SPSS 16.0). The significance level used was $p \leq 0.05$. Estimated age of symptom onset was calculated according to the equation of Langbehn [29]. Comparisons between participants versus non-participants were made (independent *t*-tests or Pearson Chi-Square). Participants were also compared with drop-outs (Mann-Whitney U Tests or Pearson Chi-Square). Changes (Δ) in the control period (measurement 1–2) were assessed to explore if scores within the same group changed without any intervention. If no important changes would occur, then the means of scores of measurement 1 and 2 would be used as baseline scores to assess the changes from pre- to post-intervention (dependent *t*-tests or Wilcoxon Rank tests). To compare pre/post-session Mood-VAS ratings, a linear mixed model with random participant effect, fixed time, fixed before-after session effect and fixed manifest-premanifest and fixed carrier/patient-partner effect was performed.

RESULTS

Of the 106 HD patients and 54 PM carriers who were invited to participate with their partner, eventually, 40 HD patients and 19 PM carriers were willing to participate in the study (Fig. 1). Demographics and clinical characteristics of all participants are presented in Table 2. Participating HD patients were significantly more often female ($p = 0.03$) and higher educated ($p = 0.04$) as compared to non-participating patients.

The drop-out rate during various moments in the study was 25% in the HD group (patients $n = 11$; partners $n = 6$) and 39% in the PM HD group (carriers $n = 7$; partners $n = 6$), of which most dropped out before the start of the program. HD patients who dropped out had significantly worse physical quality of life (SF-36, $p < 0.01$)) as compared to completers. PM-carriers who dropped out had significantly less motor symptoms (UHDRS-motor, $p = 0.03$) and better cognitive functioning (UHDRS-cognitive, $p = 0.03$) than completers. PM-partners who dropped out had significantly more psychosocial need for help (BELA-A-k, $p = 0.04$) as compared to completers.

No changes were found in HD patients during the control period (from measurement 1 to 2), PM carriers used more comforting cognitions (UCL, $p = 0.02$); HD caregivers experienced a worse physical Qol (SF-36, $p = 0.05$); and PM partners used less passive coping styles (UCL, $p < 0.01$) at measurement 2. Mean scores of measurement 1 and 2 were used as baseline scores (Table 3).

Pre- and post-intervention analyses are reported in Table 4. After participation, HD patients reported less behavioral problems (UHDRS, $p = 0.05$), less anxiety

Table 2
Demographics and clinical characteristics of all participants

	HD patients $n=40$	PM carriers $n=19$	HD caregivers $n=28$	PM partners $n=14$
Women, *n*	14 (35%)	13 (68%)*	16 (57%)	4 (29%)
Age, years	53.4 (9.0)	41.3 (10.4)*	55.6 (9.1)	44.9 (14.1)*
Having a partner, *n*	30 (75%)	16 (84%)	28 (100%)	14 (100%)
Participation in couple	26 (65%)[1]	15 (79%)[1]	26 (93%)	14 (100%)
Higher education level, *n*	16 (40%)	3 (16%)	11 (39%)	7 (50%)
Employed, *n*	9 (23%)	15 (79%)*	14 (50%)	12 (86%)*
Normal/increased CAG, range	15–31/40–53[2]	15–25/38–51	–	–
Years since genetic test	7.0 (6.1)	5.7 (5.5)	–	–
Estimated age of onset	48.6 (8.3)	49.5 (13.1)	–	–
UHDRS				
-Motor	32.8 (17.0)	4.7 (3.5)*[4]	–	–
-Independence scale	85.0 (13.0)	99.4 (1.6)*[4]	–	–
-Total functioning capacity	9.2 (2.5)	12.6 (0.8)*[4]	–	–
-Cognitive	210.7 (61.4)[3]	267.4 (64.1)*[4]	–	–
-Behavioral	10.8 (9.1)	9.6 (9.5)	–	–
MMSE, global cognitive functioning	27.8 (2.0)[5]	27.9 (1.3)	28.6 (1.2)	28.9 (1.4)
Medication use[6]				
-Antidepressants, *n*	18 (45%)	0 (0%)	3 (11%)	0 (0%)
-Neuroleptics, *n*[7]	9 (23%)	0 (0%)	0 (0%)	0 (0%)
-Benzodiazepines, *n*	7 (18%)	0 (0%)	1 (4%)	0 (0%)
- Anti-epileptics, *n*[8]	2 (5%)	0 (0%)	0 (0%)	0 (0%)
-Other, *n*[9]	23 (58%)	7 (37%)	15 (54%)	5 (36%)
- No medication, *n*	6 (15%)	12 (63%)	11 (39%)	9 (64%)

Values are mean (SD) unless otherwise indicated. Abbreviations: CAG, Cytosine-Adenine-Guanine repeat lengths; UHDRS, Unified Huntington's Disease Rating Scale; MMSE, Mini Mental State Examination; HD, Huntington's disease; PM, pre-manifest. *Significantly different from HD patients/HD caregivers (Mann Whitney U Test or Chi-square). [1]A primary informal care gives other than the spouse participated in the program; [2]In two HD patients, repeat lengths could not be verified, however DNA tests were performed; [3]$n=37$, three patients did not complete cognitive assessment because of color blindness ($n=2$) and too much burden ($n=1$); [4]$n=18$, in one PM carrier, no cognitive, motor and functional assessment was performed because of drop-out; [5]One missing value, because of drop-out; [6]Psychotropic medication use during the study changed in 4 HD patients: new antidepressant use ($n=1$); change of antidepressant ($n=1$); decrease of antidepressant dose ($n=1$); new benzodiazepine use ($n=1$).[7]Including Tiapride, primarily given as treatment for motor symptoms; [8]Primarily provided as mood stabilizers; [9]Other medication included all other medication than psychotropic, like medication for coronary, lung or stomach diseases.

Table 3
Baseline scores on questionnaires

	HD patients $n=40$	PM carriers $n=19$	HD caregivers $n=28$	PM partners $n=14$
HADS				
-Anxiety	6.0 (3.5)	5.9 (3.1)	5.3 (3.9)	4.3 (3.7)
-Depression	4.4 (3.3)	2.9 (3.2)*	2.5 (3.1)	1.5 (2.5)
BELA-P/A-k				
-Psychosocial burden	19.1 (14.4)[1]	7.4 (8.0)*	10.1 (7.7)	1.9 (2.2)*
-Psychosocial need for help	25.5 (18.1)[2]	11.5 (13.7)*	14.6 (12.2)[1]	6.4 (7.6)*
SF-36				
-Mental quality of life	40.2 (11.5)	43.6 (8.8)	47.7 (8.7)	50.2 (3.7)
-Physical quality of life	46.4 (9.6)	52.0 (8.9)*	51.4 (9.3)	53.9 (7.8)
UCL- coping strategies				
-Active coping	16.1 (4.2)	18.4 (4.1)	19.7 (3.6)	21.0 (3.9)
-Palliative reaction	17.5 (4.4)	17.6 (4.1)	17.7 (3.8)	15.7 (2.9)
-Avoidance	16.6 (3.6)	15.5 (3.2)	15.8 (3.5)	14.4 (2.9)
-Seeking social support	13.6 (3.6)	13.1 (2.7)	12.3 (2.7)	13.5 (4.4)
-Passive reaction	12.1 (3.4)	10.2 (2.3)*	10.3 (3.0)	10.2 (2.6)
-Negative emotion expression	5.4 (1.7)	5.7 (1.2)	5.7 (1.0)	5.4 (1.3)
-Comforting cognitions	11.8 (2.2)	11.3 (4.1)	12.2 (2.3)	11.8 (3.7)

Values are mean (SD) unless otherwise indicated. Abbreviations: HADS, Hospital Anxiety and Depression Scale; BELA-P/A-k, Belastungsfragebogen Parkinson/Angehörigen kurzversion; SF-36, 36-item Short Form health survey questionnaire; UCL, Utrecht Coping List. HD; Huntington's disease; PM; pre-manifest. *Significantly different from HD patients/HD caregivers (Mann Whitney U Test or Chi-square). [1]One missing value; [2]Two missing values.

Table 4
Change scores from pre- to post intervention for manifest and premanifest participants

	HD patients $n=29$	PM carriers[2] $n=12$	HD caregivers $n=22$	PM partners[2] $n=8$
	Mean change Δ	Mean change Δ	Mean change Δ	Mean change Δ
UHDRS-behavioral	–3.4 (8.8)*	2.1 (6.7)	–	–
HADS				
-Anxiety	–0.8 (2.2)*	–0.6 (1.4)	–0.4 (3.4)	–1.1 (2.8)
-Depression	–0.6 (2.1)	–0.3 (1.2)	–0.4 (1.9)	–0.6 (1.8)
BELA-P/A-k				
-Psychosocial burden	–1.8 (6.3)	1.11 (4.9)	–1.9 (3.4)*	–0.1 (0.8)
-Psychosocial need for help	0.2 (11.5)	1.8 (5.5)	–2.1 (5.7)	–0.1 (0.7)
SF-36				
-Mental quality of life	2.2 (8.0)	0.4 (3.3)	–1.2 (5.8)	1.8 (5.8)
-Physical quality of life	–0.4 (4.7)	–0.4 (3.7)	0.7 (6.2)	1.0 (2.7)
UCL-coping strategies				
-Active coping	0.5 (2.3)[1]	0.6 (2.3)	0.3 (2.3)	1.6 (2.2)
-Palliative reaction	0.1 (2.8)	0.2 (2.7)	0.3 (2.6)	1.5 (2.8)
-Avoidance	–0.6 (2.5)	0.3 (2.4)	0.2 (1.9)	–0.3 (1.6)
-Seeking social support	0.6 (1.6)*	0.9 (1.5)*	0.4 (2.1)	1.9 (1.8)*
-Passive reaction	–0.7 (1.6)*	0.2 (1.7)	–0.2 (0.8)	–0.8 (1.4)
-Negative emotion expression	0.1 (1.2)[1]	–0.1 (1.1)	0.3 (1.8)[1]	–0.1 (0.5)
-Comforting cognitions	–0.1 (1.5)	0.4 (2.1)	–0.2 (2.2)	0.9 (2.5)

Negative change scores reflect improvement on behavioral problems, anxiety, depression, psychosocial burden and need for help and worsening on quality of life; and less use of the particular coping strategy. UHDRS, Unified Huntington's Disease Rating Scale; HADS, Hospital Anxiety and Depression Scale; BELA-P/A-k, Belastungsfragebogen Parkinson/Angehörigen kurzversion; SF-36, 36-item Short Form health survey questionnaire; UCL, Utrecht Coping List. [1]A Wilcoxon Rank Test was used. [2]Wilcoxon Rank Tests were used for all variables. *$p \leq 0.05$.

Table 5
Summary of program evaluation questionnaire

	HD patients $n=29$	PM carriers $n=12$	HD caregivers $n=22$	PM partners $n=8$
Program rating (mean)	7.9	8.4	7.8	8.4
Most valued session,'Stress management', *n*	15 (52%)	6 (50%)	9 (41%)[1]	4[1] (50%)
Benefit from participation, *n*	23 (79%)	12 (100%)	17 (77%)	7 (88%)
Useful in daily life agree/agree somewhat, *n*	19 (66%)/8 (28%)[1]	11 (92%)/0 (0%)[1]	16 (73%)/5 (23%)	7 (88%)/0 (0%)
Difficulty to follow program agree/agree somewhat, *n*	1 (3%)/7 (24%)	1 (8%)/1 (8%)	1 (5%)/3 (14%)	0 (0%)/0 (0%)
Participation was tiresome, agree/agree somewhat, *n*	8 (28%)/7 (24%)[1]	2/0 (0%)[1]	3 (14%)/5 (23%)	2 (25%)/1 (13%)
Timing of intervention[2], *n*				
-Too early	–	1 (8%)[1]	–	2 (25%)
-Too late	–	1 (8%)	–	2 (25%)

[1]One missing value. [2]Item was not included in questionnaire for manifest HD.

(HADS, $p=0.05$), more use of seeking social support (UCL, $p=0.05$), and less use of passive reaction (UCL, $p=0.03$) as coping strategies after the program. HD caregivers reported less psychosocial burden (BELA-A-k, $p=0.02$). More use of seeking social support as a coping strategy was found in both PM carriers (UCL, $p=0.05$) and PM partners (UCL, $p=0.03$).

Participants' mood ($n=62$) significantly improved from pre- (M = 74.9) to post-sessions ($\Delta=5.7$, $p<0.01$) on the 100-point VAS. Mood also improved from session 1 (M = 76.6) to session 8 ($\Delta=7.1$, $p=0.01$), because of significant improvement between session 1 through 7 and 8 ($p<0.01$). Mood did not improve from session 1 to 7 (all $p>0.05$). There was no difference in effects between groups (manifest versus premanifest, patient versus caregiver) ($p>0.05$).

The overall program rating was good, premanifest participants rated the program somewhat higher than manifest participants (Table 5). Session 4 about stress management was most often reported as the most valuable session. Most participants experienced the program as useful in daily life. Contents of the program were not difficult to understand for most of the participants. Less than one third of the HD patients found it difficult to follow the program. Half of the HD patients and more than one third of the PM partners and

caregivers experienced participation in the program as tiresome. Most premanifest participants found the timing of the intervention right; 25% of them preferred participation in the program in an earlier stage.

DISCUSSION

Present pilot study is the first to assess the feasibility of a standardized psychosocial Patient Education Program for Huntington's disease (PEP-HD). The program was feasible in premanifest carriers as well as in HD patients and their partners. The assumption was that the program could diminish psychological distress and negative social impact and that it could stimulate the use of helpful coping strategies like active problem solving and seeking social support in order to improve quality of life. Psychosocial wellbeing did improve in HD patients and caregivers: they reported less anxiety, and less behavioral symptoms. Caregivers reported less psychosocial burden as was also found in the PD study [11]. The use of self-management intervention with cognitive behavioral strategies seems to be helpful to improve psychological well-being as hypothesized [12]. For example cognitive restructuring may have helped patients to use more helpful and realistic thoughts, which may have reduced anxiety. An additional value of the PEP-HD, besides the provision of knowledge and skills, is that participants experience attention from the trainers and interactions with the fellow-sufferers. A meta-analysis on behavioral therapy indicated that the specific therapy effects are larger than those achieved by placebo control conditions [30]. However, this has not yet been studied with regard to this particular education program and we recommend this for future research.

We also found improvements in coping strategies. The HD patients used more seeking social support and less use of passive coping. In the program, they learned to actively seek information, to stand up for themselves, use helpful communication skills and to seek social support actively. In pre-manifest HD, both carriers and partners used more seeking of social support after the program. This may be beneficial for coping with premanifest HD and psychological well-being in the future [3, 8]. However, this improvement in coping in the pre-manifest group was not accompanied by improvement on psychological outcome measures directly after the program. Premanifest carriers did have comparable psychological baseline scores regarding behavioral problems, anxiety and mental quality of life. This may indicate that the program is less effective in the premanifest stage of HD. However, we may not have been able to assess the specific premanifest psychosocial problems and possible improvements adequately. The BELA-P/A-k was developed originally for PD patients and caregivers with symptoms. Because some of the items are focused on consequences of disabilities, the questionnaire seems less relevant to the PM group, resulting in floor effects in scores. An outcome measure capturing the specific psychosocial problems in PM HD should be developed. Besides the effects of the program, the drop-out rate was also relatively high in premanifest (39%) compared to the manifest group (25%). Possibly, those premanifest carriers may have feared to be faced with the discussions about HD and its consequences as denial and avoidance are common in carriers [8]. Participating PM carriers and partners did evaluate their participation in the program as positive and most found timing of the intervention right.

The relatively low baseline scores on psychological self-report questionnaires may be the result of a selection bias of highly motivated and adjusted patients or impaired awareness [31]. Impaired awareness may lead to denial of (psychological) problems and overestimation of competencies, including behavioral and emotional control [31, 32]. Both neurological dysfunction and avoidant psychological coping may be causes of impaired awareness. It has been related to deficits in global cognition, memory and executive functioning [32]. Lack of self-awareness may also have contributed to non-participation or drop-out during the study. However, no differences on psychological outcome measures, and coping, were found between participants and drop-outs. Also, completers did not have better scores on cognitive tests as compared to drop-outs or non-participants.

Participants with higher education and female gender were more willing to participate. They may feel more attracted to an education program and to discuss their feelings than lower educated and male patients. HD patients who dropped-out had more physical problems, so the program and/or study may be too burdensome for some patients. Premanifest patients who dropped out had better motor and cognitive scores, but were not further away from disease onset according to our results. The higher psychosocial need for help in premanifest caregivers who dropped out could not be explained.

This study has its limitations as statistical power was reduced because of the small study sample. Because of this small study sample, a single-group design was used. Therefore we are not able to draw firm

conclusions about the effectiveness. In follow-up research, an international multicentre randomized controlled trial should be the next step to provide a larger sample and to enable drawing conclusions about the effectiveness. Follow-up research with follow-up measurements, for example after six months is recommended. Another limitation is that many outcome measures were used, and no statistical corrections for multiple testing were applied. Psychotropic medication changes were not likely to be of influence on the study results; they were reported only in four out of 29 patients.

In conclusion, this pilot study demonstrated the feasibility of the program in Huntington's disease, especially in manifest stages. Further research to assess the effectiveness seems warranted.

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