

Caring for patients with Huntington disease – A survey of caregivers' experiences and views

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Abstract

Introduction. Despite the growing interest in the consequences of caring for patients with Huntington disease (pHD), little is known about the family caregivers of such patients in Poland. Identification of their needs can improve caregivers' well-being, the quality of care and condition of pHD. The aim of this study was to understand the social functioning of family caregivers of pHD and their perception of the caregiving role.

Materials and methods. Data was collected from 55 family caregivers of pHD. A structured questionnaire was used consisting of 86 questions subsumed into five domains: 'Problems' and 'Feelings related to caregiving', 'Attitude toward caregiving', 'Satisfaction with life' and 'Perception of healthcare services'. Correlations between the different scales and other characteristics were measured as potential predictors of the burden. Non-parametric statistical methods were used in the analysis.

Results. Most respondents experienced a high (50.9%) or moderate (30.95%) feeling of burden. Although 70.9% of caregivers perceived caregiving positively, for many it was a source of negative feelings. Only 10.9% of respondents declared that caregiving decreased their QoL. Carers' perception of caregiving was mostly influenced by their negative experiences with the healthcare system. Respondents' domicile, religious practices, age, income, marital status, time of diagnosis and of caregiving, patient's age and stage of disease also influenced their experiences.

Conclusions. Health professionals and policy planners should focus on monitoring caregivers' health, identifying their needs, sources of distress, and supporting caregivers' coping strategies. They should also be better educated about the clinical and practical aspects of HD.

Key words

caregiving burden, carers, family caregiving, Huntington's disease.

INTRODUCTION

Huntington's disease (HD) is a rare chronic neurodegenerative disease of the brain that causes movement abnormalities, cognitive decline and behavioural disturbances. While its first symptoms usually begin at the age of 35–45, there is a wide variation in its onset. HD is characterised by progressive involuntary movements of the body and abnormal gait which result in problems with walking and clumsiness [1]. However, cognitive and behavioural changes may be present before the onset of movement abnormalities. The most characteristic initial symptom of HD are uncontrollable movements of the body followed by subtle changes in personality, cognition, and physical skills. As the disease progresses, the earlier symptoms escalate and other problems occur, including problems with concentration and memory loss, problems with planning and problem solving, mood and personality changes, depression episodes, impulsiveness, aggressive and anti-social behaviours, and suicidal ideation. Additionally, mood swings and emotional lability become more frequent. Patients also experience problems with swallowing and severe weight loss [1, 2]. For all these reasons, in advanced stages, HD patients require full nursing care. However, as there is no cure for HD, its treatment is purely palliative; consequently, the patient's death usually occurs after 15–20

years. Most frequently, it is caused by a secondary illness, mainly aspirational pneumonia [1, 2]. Other causes of death include suicides which are four to six times more common than in the general population.

It is equally important that the gradual loss of psychomotor and cognitive functions in a patient seriously affects the entire family as it provokes role changes and a loss of intimacy with the affected relative. Moreover, as HD progresses caregiving becomes more and more demanding and carers are often on 24/7 duty, forced to abandon their leisure activities and professional careers. As a result, caregivers have problems with maintaining a normal life, have little time for themselves and other family members. Thus, HD becomes a source of emotional burden, especially in that due to the complexity and severity of the symptoms of HD, the carers often experience social isolation and are overwhelmed by their caregiving responsibilities [3, 4, 5, 6, 7]. Frequently, they feel tired, anxious, depressed, lonely, fearful, frustrated, angry and helpless [2, 3, 8, 9, 10, 11, 12]. All in all, the available research suggests that, like with other dementias [2, 7, 12, 13], the quality of life of HD carers is seriously impaired in terms of burden [14], lack of social and emotional support, ability to cope with stress [15], social isolation [2, 7], financial costs [2, 6, 7], access to healthcare services [3, 5, 16], lack of information from health professionals [3, 5, 9, 16, 17, 18] and decline in own health [6, 10, 18].

At the same time, health professionals and social workers should be aware that HD is distinguished by some factors that are not present in other neurodegenerative diseases [2, 6, 10, 11, 13]. Firstly, HD usually begins much earlier,

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when a person is in middle-age. Consequently, HD carers are also younger and still have their own child-rearing and professional responsibilities [6, 15]. Moreover, HD has slower progression of symptoms which results in a prolonged disease trajectory which may last up to 30 years [6]. Additionally, due to its longer duration, it involves activities that are unique to HD [7]. Most importantly, in contrast to other dementias, HD is a hereditary disease and may be present in multiple family members who 'live in the shadow of HD' [2, 15, 19]. Thus, while still caring for the affected relative, HD carers may worry that they too will develop the disease and/or that they may have passed it to their children [2, 4, 6, 7, 10, 15]. Finally, in contrast to Alzheimer's or Parkinson's disease, health professionals lack basic knowledge about the clinical and practical aspects of HD and are unaware of caregivers' problems and needs [2, 3, 4, 5, 9, 16, 20]. Thus, it seems that the social implications of HD are much more serious and far-reaching [13, 21]. Nevertheless, most studies focus on the clinical dimension of HD and it is HD patients who become the subject of professional attention.

Meanwhile, the problems and needs of the caregivers are often overlooked. For that reason Kessler describes such carers as 'forgotten persons in the HD family' [22]. At the same time, while studies describe experiences of HD carers from many European countries, including the United Kingdom [3, 9, 18, 20, 23, 24], the Netherlands [25], Norway [26], Spain, [27] the United States and Canada [2, 4, 5, 6, 10, 11, 16, 22], less attention has been paid to HD carers in Poland [12, 28]. This is especially interesting because there are many studies on the needs, quality of life and caregiving burden of family carers of patients with Alzheimer's disease [29, 30], stroke [31, 32] or mental disorders [33, 34]. Therefore, the aim of this study is to understand the social functioning of family caregivers of patients with HD (pHD) and their perception of the caregiving role. The specific aims were: 1) to recognize the problems and needs of HD carers, 2) to evaluate their satisfaction with life, 3) to identify the factors that predict the burden experienced by caregivers.

MATERIALS AND METHOD

This research was part of a larger study on family caregivers of pHD. It was based on a pilot study which aimed to tackle the basic problems and needs of HD carers. The study was conducted between August – December 2014 among 55 family caregivers of pHD. The participants were recruited via the Polish Huntington's Disease Association and a discussion forum for pHD and their caregivers (<http://forum-hd.zamki.pl>). The potential participants received an invitation via the HD website and those who responded were interviewed via an e-mailed questionnaire. The inclusion criteria for the study were: 1) age over 18, 2) being a family member or significant other of HD subject, 3) being directly involved in caregiving, 4) willingness to participate in the study. Participants were excluded if they were professional or paid caregivers or under 18 years old.

A structured questionnaire was constructed from themes based on a review of the literature [35] and the study aims. Although tools exist to assess the quality of life of HD carers which measure caregivers' personal feelings about the caregiving role, life satisfaction and quality of life [8, 23], a more detailed questionnaire was constructed which included

items that are absent from the available tools, including caregivers' experiences with healthcare services. The questionnaire comprised of 86 questions: 20 items referred to caregivers' and patients' demographic data. The remaining 66 items were subsumed in 5 domains centred on the following topics: 'Problems related to caregiving', 'Attitude towards caregiving', 'Feelings related to caregiving', 'Satisfaction with life' and 'Caregivers perception of healthcare services'. The questions were open-ended. The respondents were asked to complete the questionnaire independently and to indicate whether they agreed or disagreed that the question was relevant to their caregiving experience.

The first scale, 'Problems related to caregiving' related to 13 items concerning the problems resulting from HD (loss of motor skills, personality changes, emotional lability, behaviour, loss of communication skills) and the factors affecting caregiving (costs of medications, home adaptation, lack of equipment, time spent on caregiving, number of caregiving tasks and available support). Each item was scored from 0 (never) to 4 points (always). The maximum score was 52. The following scale was assumed: 1–10 no feeling of burden, 11–20 low feeling of burden, 21–30 moderate feeling of burden, 31–40 high feeling of burden, and 41–52 very high feeling of burden.

The second scale: 'Attitude toward caregiving' related to 6 items asking whether the respondents considered caregiving as a duty, whether it was important for them, and if it was a source of satisfaction and personal growth, and whether they believed in the development of an effective cure for HD. For each answer, the respondents scored 0 (yes) or 1 (no), scoring a maximum of 6 points, where 0–2 meant a positive attitude, 3–4 – neutral and 5–6 – negative attitude.

The third subscale referred to the respondents' feelings toward caregiving. It focused on 25 items which asked whether caregiving made them feel physically and emotionally tired, uncomfortable, stressed, overloaded, angry, lonely, sad or depressed, guilty, insecure, hopeless and senseless. It also examined how caring for pHD influenced their lives and relationships with the affected relative and, finally, whether caregivers were concerned over developing HD or passing it to their children. The questions were scored from 0 (never) to 4 (always), and the maximum score was 100, where 0–20 meant very positive feeling, 21–40 – positive feelings, 41–60 – neutral, 61–80 – negative and 81–100 – very negative feelings towards caregiving.

The fourth subscale measured caregivers' satisfaction with life. The respondents were asked 10 questions about their life situation and satisfaction, health, well-being and sleeping problems, feeling of security and economic situation, and the influence caregiving had on their quality of life. The other questions referred to their feelings of happiness and whether they felt fulfilled in life. The questions were scored from 0 (very good) to 3 (very bad), with a maximum score of 30 points: from 0–6 signified very high satisfaction, 7–12 – high satisfaction, 13–18 – neutral, 19–24 – low satisfaction, and 25–30 – very low satisfaction with life.

The final subscale focused on carers' perception of healthcare services for pHD. It centred on the support system for pHD and their carers from the government, social institutions and physicians, the quality of medical care for pHD, including access to specialists, medications and information on HD, physicians' knowledge of clinical and practical aspects of HD, their communication skills and



empathy, and contacts with the genetic clinic. The answers ranged from 0 (very good) do 3 (very bad). The maximum score was 36 points. Scoring 0–9 meant very positive, 10–18 – positive, 19–27 – negative and 28–36 -very negative.

The StatSoft's Statistica 10. PL (StatSoft Inc., 2011) was used for a statistical analysis. Non-parametric statistical methods were used in the analysis: Spearman's rank correlation coefficient, Kruskal-Wallis one-way analysis of variance (ANOVA) and U Mann-Whitney test. Statistical significance was assumed to be $\alpha=0.05$. The results $p<0.05$ were recognized as statistically significant.

Participants. The sample consisted of 55 family caregivers of pHD, 39 (71%) were females and 16 (29%) males. The majority were aged between 50–70 ($n=24$; 61.8%). No significant difference in age between male and female caregivers was found. Most participants lived in a relationship with the pHD, either as spouses ($n=35$; 63.6%) or partners ($n=6$; 10.9%) and 10 (18.2%) were widowed. 46 respondents (83.7%) had children: 15 (27.3%) had one child, 20 (36.4%) had two, and 11 (20%) three or more children. While only 8 (13.8%) of those children were HD negative, 24 were affected in some way: 8 (13.8%) suffered from juvenile HD, 8 (13.8%) were HD carriers and one of the 8 (13.8%) suffered from HD ($n=8$; 13.8%). Another 26 (44.8%) were at risk of HD, but had not undergone testing. Most respondents lived either in small towns with less than 50,000 inhabitants ($n=24$; 43.6%) or in big agglomerations with a population of over 500,000 ($n=18$; 32.7%). 25 (45.5%) of them had university-level education and 15 (27.3%) had graduated from high school. 7 (12.7%) had received medical training. Over one-half of the participants ($n=30$; 54.5%) had a full-time job, while 13 (23.6%) were retired and 6 (10.9%) were unemployed. 36 carers (65.3%) reported an income of less than 2,500 PLN (€590) per month while 16 (29.1%) earned 3,000 PLN or more (€700). Of those reporting religious affiliation, 47 (85.5%) were members of the Roman Catholic Church and 6 (10.9%) were Greek Orthodox. While the majority of respondents declared themselves to be 'believers', 31 (56.4%) defined themselves as 'practicing believers' and 20 (36.4%) as 'believers who do not practice their religion'. 31 respondents declared that religion was important for their life decisions and choices (38.2%); for another 26 (47.3%), it was of little significance and for 8 (14.5%) it was 'irrelevant'.

44 respondents (80%) were primary caregivers of pHD, 24 them were patients' spouses (37.4%), 12 were their parents (18.8%) and 11 children (17.2%). The mean time of caregiving was 8.4 years ($SD = 4.9$; $-1-20$). For the majority, caregiving took 6–12 hours per day ($n=25$; 45.5%), while for 15 (27.3%) it was more than 12 hours per day. Only one-half of respondents ($n=28$; 50.9%) declared membership in a local support group. Detailed characteristics are shown in Table 1.

As for care recipients, this group was almost equally split according to their gender, as there were 26 females (47.3%) and 29 males (52.7%). The majority were aged over 50 ($n=30$, 54.6%). While all stages of the disease were present in the sample, most patients were in the advanced stage ($n=40$; 72.7%). The mean time of diagnosis was 10.5 ($SD=7.1$; $-1-30$) (Tab. 2.)

Table 1. Socio-demographic characteristics of the caregivers

Characteristics	No.	Percentage
Gender		
female	39	70.9
male	16	29.1
Age		
Under 20	0	0
20-29	6	10.9
30-39	2	3.6
40-49	13	23.6
50-59	18	32.7
60-69	10	18.2
Above 70	6	10.9
Marital status		
Single	2	3.6
Cohabiting	6	10.9
Married	35	63.6
Widowed	10	18.2
Divorced	2	3.6
Domicile		
Up to 10,000 inhabitants	12	21.8
10-50,000 inhabitants	12	21.8
51-100,000 inhabitants	4	7.32
101-500,000 inhabitants	9	16.4
Above 500,000 inhabitants	18	32.7
Education		
Elementary	2	3.6
Vocational	6	10.9
High School	15	27.3
University	25	45.5
Medical University	7	12.7
Employment status		
Full time	30	54.5
Part time	6	10.9
Retired/pension	13	23.6
Unemployed	6	10.9
Income		
1,000 PLN or less	2	3.6
1,001-1500 PLN	8	14.5
1,501-2000 PLN	19	34.5
2,001-2500 PLN	7	12.7
2,501-3000 PLN	3	5.5
More than 3,000 PLN	16	29.1
Religion		
Roman Catholic	47	85.5
Protestant	0	0.0
Greek Orthodox	6	10.9
Other	2	3.6
Religious practice		
Believing/practicing	31	56.4
Believing /not practicing	20	36.4
Non-believer/practicing	0	0.0
Non-believer/not practicing	4	7.3
What role does religion play in your life?		
Significant, it influences my life decisions and choices	11	20.0
Rather big, I try to follow religious principle in my life	20	18.2
Little, I separate religion from public issues	26	47.3
None, it is irrelevant to me	8	14.5
Children		
1	15	27.3
2	20	36.4
3 or more	11	20.0
No children	9	16.4
Child's health status (n=58)		
Juvenile HD	8	13.8
HD carrier (HD positive pre-symptomatic)	8	13.8
Suffers from HD	8	13.8
Negative test result	8	13.8
At risk, but did not undergo testing	26	44.8
Caregiver status		
Primary caregiver	44	80.0
Secondary caregiver	11	20.0
Relationship with HD patient (n=64)		
Spouse/partner	24	37.5
Parent	11	17.2
Child	12	18.8
Sibling	6	9.4
Son/Daughter-in-law	0	0.0
Mother/Father in law	0	0.0
Other relative	11	17.2
Time of caregiving, mean (SD) [years]	8.4 (4.9)	
Daily time dedicated to caregiving		
More than 12 hours per day	15	27.3
6-12 hours per day	25	45.4
Less than 6 hours per day	15	27.3
Membership of local support group		
Yes	28	50.9
No	27	49.1



Table 2. Socio-demographic characteristics of the HD patients

Characteristics	N	%
Gender		
Female	26	47.3
Male	29	52.7
Age		
Under 20	0	0.0
21-29	8	14.5
30-39	3	10.9
40-49	11	20.0
50-59	15	27.3
Over 60	15	27.3
Stage of disease		
Early	2	3.6
Middle	13	23.6
Late	40	72.7
Diagnosis, mean (SD) [years]	10.5(7.1)	

RESULTS

Most respondents experienced a high (50.9%) or moderate (30.95%) feeling of burden. Additionally, 6 caregivers (10.9%) experienced a very high feeling of burden. Nevertheless, the caregivers themselves were not negative as 70.9% of them described their attitudes toward caregiving as positive, while for 20.1% it was neutral, and none had a negative attitude. At the same time, for many, caregiving was a source of ambivalent feelings: while they described it as important, it was also a source of negative (29.1%) and neutral (54.5%) feelings, and only 9 caregivers (16.3%) described their feelings as positive. Just over one-half of respondents (52.8%) were satisfied with their lives, while 36.4% felt neutral about it. Only 10.9% declared that caring for pHD decreased their QoL. It was significant that apart from experiencing many problems and negative feelings resulting from caregiving, the participants' perception of problems was mostly influenced by their negative experiences with the healthcare system; while only 9.1% respondents described them as positive, for 41.8% it was negative and for the remaining 49.1% it was neutral (Tab. 3, 4).

Table 3. Mean, median, standard deviation and criterion variables for the scales

	Mean [range]	SD
Problems related to caregiving (range: 1-52/no feeling of burden-very high feeling of burden)	32.2 [1-43]	8.4
Attitudes toward caregiving (range: 0-6/positive-negative)	1.6 [0-4]	1.4
Feelings related to caregiving (range: 0-100/very positive-very negative)	53.8 [18-84]	16.1
Caregiving and perceived quality of life (range: 0-30/very high satisfaction-very low satisfaction)	12.7 [3-21]	4.3
Experiences with healthcare system (range: 0-36/very positive-very negative)	20.6 [8-31]	6.2

As shown in Table 5, analysis of the correlations between the different scales did not show any statistical significance, except in the case of 2 pairs of scales: 'Problems related to caregiving'/'Attitudes toward caregiving' ($r=0.12$; $p=0.3736$) and 'Problems related to caregiving'/'Caregiving and perceived satisfaction with life' ($r=0.04$; $p=0.7799$). All the other scores were statistically significant and the correlations were positive. The highest rank for Spearman's

Table 4. Scores on scales reported for HD caregivers

	N	%
Problems related to caregiving		
No feeling of burden	2	3.6
Low feeling of burden	2	3.6
Moderate feeling of burden	17	30.9
High feeling of burden	28	50.9
Very high feeling of burden	6	10.9
Attitudes toward caregiving		
Positive	39	70.9
Neutral	16	29.1
Negative	0	0.0
Feelings related to caregiving		
Very positive	2	3.6
Positive	7	12.7
Neutral	30	54.5
Negative	14	25.5
Very negative	2	3.6
Caregiving and perceived quality of life		
Very high QoL	4	7.3
High QoL	25	45.5
Neutral	20	36.4
Low QoL	6	10.9
Very low QoL	0	0.0
Experiences with healthcare system		
Very positive	0	0.0
Positive	5	9.1
Neutral	27	49.1
Negative	15	27.3
Very negative	8	14.5

Table 5. Correlations between scales

	r	p
Problems:		
related to caregiving & attitudes toward caregiving	0.12	0.3736
related to caregiving & feelings related to caregiving	0.30	0.0264
related to caregiving & caregiving and perceived satisfaction with life	0.04	0.7799
related to caregiving & experiences with healthcare system	0.36	0.0067
Attitudes:		
towards caregiving & feelings related to caregiving	0.46	0.0005
towards caregiving & caregiving and perceived satisfaction with life	0.60	0.0000
towards caregiving & experiences with healthcare system	0.44	0.0007
Feelings:		
related to caregiving & caregiving and perceived satisfaction with life	0.63	0.0000
related to caregiving & experiences with healthcare system	0.59	0.0000
Caregiving and perceived satisfaction with life & experiences with healthcare system	0.32	0.0189

correlation coefficient was observed between 'Feelings related to caregiving' and 'Caregiving and perceived satisfaction with life' ($r=0.63$; $p=0.0000$) as respondents' negative feelings resulting from caregiving strongly correlated with their negative perception of QoL and *vice versa*. Caregivers' feelings were also strongly correlated with their experiences with healthcare services ($r=0.59$; $p=0.0000$): the more negative experiences they had, the more negatively they perceived the caregiving role. Furthermore, such negative experiences were also correlated with the perceived number of problems related to caregiving ($r=0.36$; $p=0.0067$) and subjective QoL ($r=0.32$; $p=0.0189$). The worst experiences of caregivers were with the healthcare system and the scant support they received from physicians. They perceived caregiving as more burdensome and had a lower perception of QoL.

Respondents' attitudes toward caregiving were strongly correlated with their perceived QoL ($r=0.60$; $p=0.0000$).



On the other hand, moderate correlations were observed between respondents' attitudes and their feelings related to caregiving ($r=0.46$; $p=0.0005$) and the experiences with the healthcare system ($r=0.44$; $p=0.0007$). Thus, the caregivers' level of dissatisfaction with the caregiving role was related to the amount of negative feelings resulting from providing care, dissatisfaction with healthcare services, and the perceived decrease of QoL.

Finally, the problems related to caregiving were moderately correlated with the respondents' experiences with healthcare services ($r=0.36$; $p=0.0067$) and their feelings resulting from caregiving ($r=0.30$; $p=0.0264$), as the more negative these feelings were, the more problems the caregivers reported.

Statistical analysis showed that there were statistically significant differences between some caregiver/patient characteristics and the particular subscale scores (Tab. 6). There were 6 caregiver's and 5 patient's characteristics that were associated with at least one subscale. The most important factor that was strongly related and most consistently associated with all the scales was caregivers' domicile. Respondents who lived in small towns reported more problems related to caregiving, perceived the caregiving task more negatively, and felt more pessimistic about it. They also complained more about the influence caregiving had on their QoL, and had more negative experiences with the healthcare system.

Caregivers' perception of the problems related to caregiving also differed depending on the respondent's marital status, their commitment to religious practices and the caregiver's status. Thus, the respondents who were

married ($U=111$; $p=0.0007$), non-believers ($U=187$, $p=0.0017$) and were primary caregivers ($U=133$, $p=0.0224$) had the highest scores and felt more strain. It was also influenced by the patient's stage of the disease ($U=178$; $p=0.0217$).

On the other hand, respondents' attitudes toward caregiving differed according to their income ($U=209$; $p=0.0047$), religious practices ($U=2019$; $p=0.0097$) and age of the care recipients ($U=236.5$; $p=0.0197$). Caregivers who earned more and declared themselves as believers had a more positive attitude toward caregiving. At the same time, their feelings related to caregiving were statistically different depending on caregivers' age ($U=197$; $p=0.0056$), religious practices ($U=192$; $p=0.0023$), importance of religion in their lives ($U=192$; $p=0.0041$) and age of care recipient ($U=258$, $p=0.0489$). Younger caregivers and those who declared themselves as religiously indifferent or non-believers felt more pessimistic about their role as caregivers.

Apart from the domicile, the caregivers' perception of satisfaction with life differed depending on their income ($U=260$; $p=0.0495$) and religious practices ($U=219$; $p=0.0097$). Thus, the respondents who earned less than 2,000 PLN and declared themselves as non-believers had higher scores and declared that caregiving had a negative influence on their QoL. Finally, caregivers' experiences with the healthcare system differed depending on the caregivers' status ($U=98$; $p=0.0025$) and primary caregivers had more negative experiences with the healthcare system. On the other hand, no differences were observed in relationships between both caregivers' and patients' gender, caregivers' education and working status, and daily time spent on caregiving.

Table 6. Association between patient/caregiver characteristics and subscale scores

	Mean	Median	SD	Mean	Median	SD	U	P
AGE	Below 50 year old			Above 50 year old				
Feelings related to caregiving	61.2	60	15.1	49.3	48	3.9	197	0.0056
MARITAL STATUS	Married			Single				
Problems related to caregiving	34.5	37	6.1	25.4	28	10.7	111	0.0007
DOMICILE	Below 500,000 inhabitants			Above 500,000 inhabitants				
Problems related to caregiving	35.4	36	5.4	28.8	31	9.7	208	0.0043
Attitudes towards caregiving	2.3	2	1.2	0.9	0	1.2	149	0.0001
Feelings related to caregiving	58.6	57,5	12.8	48.9	48	17.8	250	0.0318
Caregiving and perceived satisfaction with life	14.1	14	3.7	11.2	10	4.5	235	0.0164
Experiences with healthcare system	22.4	23	5.5	18.7	17	6.5	247	0.0280
INCOME	Below 2000 PLN			Above 2000 PLN				
Attitudes towards caregiving	2.1	2	1.3	1.1	1	1.3	209	0.0047
Caregiving and perceived satisfaction with life	13.8	15	4.3	11.3	10	4.0	260	0.0495
RELIGIOUS PRACTICES	Practicing			Non-practicing				
Problems related to caregiving	35.2	34	4.8	28.3	29	10.4	187	0.0017
Attitudes towards caregiving	1.1	1	1.1	2.3	3	1.5	219	0.0097
Feelings related to caregiving	49.1	48	12.7	59.9	64.5	18.1	192	0.0023
Caregiving and perceived satisfaction with life	11.5	10	3.6	14.1	14	4.8	219	0.0097
IMPORTANCE OF RELIGION	Important or very important			Little or no importance				
Feelings related to caregiving	45.9	48	12.2	58.8	60	16.4	192	0.0041
AGE OF CARE RECIPIENT	Below 50 year old			Above 50 year old				
Attitudes toward caregiving	2.2	1	1.5	1.2	1	1.1	236,5	0.0197
Feelings related to caregiving	58.2	60	17.6	50.2	48	13.9	258	0.0489
STAGE OF DISEASE	Early or middle stage			Advanced stage				
Problems related to caregiving	29.5	28	6.4	33.2	35	8.9	178	0.0217
CAREGIVER'S STATUS	Primary caregiver			Secondary caregiver				
Problems related to caregiving	31.0	31.5	8.5	37.1	38	6.3	133	0.0224
Experiences with healthcare system	19.4	17	6.1	25.5	28	4.2	98	0.0025



Additionally, caregivers' perception of the problems related to caregiving was correlated with the time that had elapsed from the diagnosis ($r=0.30$; $p=0.0260$) and the number of years spent in caregiving ($r=0.47$; $p=0.0003$). The longer the time since the patient had been diagnosed with HD, and the longer the respondents had had the caregiving role, the more problems they experienced. At the same time, the number of years spent in caregiving was correlated with caregivers' experiences with the healthcare system: the more time the respondents spent struggling with and for healthcare services for their affected relatives, the more negative their perception of the healthcare system ($r=0.42$; $p=0.0013$).

DISCUSSION

The results of this pilot study provide evidence that the caregiver's burden is a multi-dimensional phenomenon, influenced both by the factors on the part of the patient as well as on the part of the caregiver. They show that while there are many objective burdens that affect one's ability to care, such as: time and energy devoted to the physical aspects of caregiving that may be unfamiliar or unpleasant, i.e. feeding, bathing, clothing and assisting with toileting, at the same time, its subjective dimension is equally important. In other words, the caregiver's experience of burden and the emotional reactions to caregiving may be more important than the actual problems experienced by the the pHD. This is of key importance, because caregivers' adaptation to stress, to a large part, is dependent on their subjective perception of one's caregiving situation [4]. This indicates the need for professional intervention aimed at both improving the pHD's condition and the caregiver's well-being.

The first specific aim of this study was to pinpoint the problems and needs of HD carers. The results show that caregivers were burdened by the time they had to spend in caregiving and the number of caregiving tasks. Additionally, they were preoccupied by the pHD's loss of motor skills, behaviour, emotional liability and personality changes, and complained about the access to modern medication for pHD. They also had problems with home adaptation to the patient's needs. Nevertheless, the caregivers in the study were mostly concerned with the accessibility and quality of medical care for their affected relatives [36]. The respondents reported problems with access to specialists, including neurologists, geneticists and psychologists, and complained about lack of support from both the government and social institutions, as well as from physicians and society. They were also in need of information on the clinical and practical dimension of HD as they were dissatisfied with physicians' knowledge about the disease. Thus, it was caregivers' negative experiences with healthcare services that strongly influenced the respondents' perception of the problems related to caregiving, and the influence it had on their QoL. It also had an impact on their feelings related to caregiving. Such results confirm the findings from other studies [3, 4, 5, 6, 7, 16, 19], as a comparative study between French, Italian, German, Polish and the USHD caregivers has shown that in Poland carers have the lowest level of use of resources, and spend the greatest amount of time looking after their HD relatives [37]. These results therefore reveal the caregivers' complex needs for healthcare assistance and emotional support.

Another aim of this work was to evaluate the caregivers' satisfaction with life. The study shows that most HD carers perceived their life situation as good and were satisfied with their lives (52.8%). However, while 10.9% of the respondents declared that caring for pHD decreased their QoL, the great majority defined their health as good or very good, and they had a general sense of security and wellbeing; they also felt happy and fulfilled in their lives. Nevertheless, despite these positive outcomes, the majority of respondents experienced a high or moderate feeling of burden and described their experiences as caregivers as negative (41.8%) or neutral (49.1%). Thus, although the participants stressed that caring for pHD did not influence their QoL, only a very few described their feelings related to caring as positive (16.3%), while the majority stressed having problems with maintaining a normal life. These findings are supported by other studies that prove that HD carers often feel overwhelmed by their caregiving responsibilities [3, 5, 6, 7, 18, 19], feel socially isolated and lonely, physically and emotionally tired, anxious, depressed, frustrated and helpless [2, 3, 8, 9, 10, 11, 12, 23]. Despite these negative feelings the majority of respondents had positive attitudes toward caregiving (70.9%).

Finally, the study aimed at identifying the factors that predict the caregiving burden. It has been shown that the most important factor affecting the respondents' experiences was their domicile which influenced the scoring on all the scales examined. This was related to the availability and quality of healthcare services, including neurologist, genetic clinic and psychiatric care. Many caregivers who lived in small towns or villages complained about barriers in access to professional care, lack of resources required for providing care, and lack of a support system for caregivers. At the same time, the study supports the findings from other studies that describe spirituality and religion as an important coping resource that helps many carers in finding meaning in times of hardship, and to accept things as they are [4, 6, 9]. Some relationships were also observed between caregivers' marital status, income, age and status as primary caregivers. Additionally, caregivers' experiences were also influenced by the age of the care recipient, time elapsed since the diagnosis, and the number of years spent in caregiving.

CONCLUSIONS

It should be emphasized that HD patients and their caregivers have a reciprocal impact on each other, as the care-recipient's state affects the caregiver's burden in both objective and subjective terms, and this, in turn, may lead to a worsening of care and undermine the quality of life of both parties. Most pHD are cared for by family caregivers in their own homes and need special attention and help. For that reason, health professionals and policy planners must develop effective strategies that will allow better monitoring of caregivers' mental health, identify their problems, (unmet) needs and the sources of their distress, as well as support effective strategies of coping with the stress. Moreover, they should be better educated about the clinical and practical aspects of HD.

Although this study brings a new insight into the situation of Polish family caregivers of pHD, it also has some limitations. First, as only 55 caregivers completed the questionnaire, the results prevent any generalisation about the whole population.



However, as HD is a rare disease the sample size is likely to be representative of HD carers in Poland. Secondly, the results from this pilot study describe the experiences only of those caregivers who volunteered and agreed to participate in the study and/or those who are members of the Polish Huntington Disease Association, and/or use social networks and chat forums for pHD and their carers. Thirdly, as this study was focused on caregivers' (unmet) needs, problems and concerns, this may have led to an over-representation of negative experiences, and an under-representation of positive ones. Nor does it present the other aspects of caregiving, such as the impact of HD on childbearing or those related to genetic testing. Consequently, a more in-depth study to clarify the meaning of the responses would be required to help clarify the issues of greatest concern. At the same time, this research has some advantages that should also be acknowledged. Most importantly, as HD carers in Poland seem to be neglected by the healthcare system, the study gives a new insight into their problems and needs. Consequently, it may stimulate further research on the situation of family caregivers of pHD. Finally, it seems that allowing caregivers' to share their experiences had a therapeutic value.

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