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Article

Exploring the reliability and validity of the Huntington's Disease Quality of Life Battery for Carers (HDQoL-C) within a Polish population

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Abstract: Huntington's disease (HD) is a rare genetic neurodegenerative disorder that causes motor disorders, neuropsychiatric symptoms and a progressing deterioration of cognitive functions. Complex issues resulting from the hereditary nature of HD, the complexity of symptoms and the concealed onset of the disease have a great impact on the quality of life of family carers. The caregivers are called as "forgotten people" in HD family, also in genetic counseling. This study aims to explore the reliability and validity of the Huntington's Disease Quality of Life Battery for carers (HDQoL-C) within a Polish population. 90 carers recruited from the Enroll-HD study in Polish research centres of the European Huntington`s Disease Network completed a polish translation of the HDQoL-C. Data was subjected to Principle Components Analysis and reliability measures. The Polish version of the shortened versions of the HDQoLC is similarly valid compared to the original English version and suitable for use within this population. The HDQoL-C has previously demonstrated a wide range of benefits for practitioners in capturing and understanding carer experience and these benefits can now be extended to Polish speaking populations.

Keywords: Huntington's disease; quality of life; family caregiving; reliability and validity; factor analysis; Poland

1. Introduction

Huntington's disease (HD) is a rare genetic neurodegenerative disorder caused by the mutation of the IT15 gene which codes the huntingtin protein located on the short arm of chromosome 4 [1]. The clinical presentation of the disease includes motor disorders, neuropsychiatric symptoms and a progressing deterioration of cognitive functions. The motor symptoms consist of involuntary choreatic movements and impaired saccadic eye movements. The symptoms that appear at the subsequent stages of the disease are dystonia, dysarthria, dysphagia, rigidity and bradykinesia, leading to death in 15-20 years [2,3]. The most frequent causes of death are aspiration pneumonia,

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injuries resulting from falls and suicides, which are recorded twice as often as in the total population [4]. In the juvenile form, usually presenting itself with mental degradation, motor disorders occur later, the course of the disease is more acute, followed by death after 8-10 years from the onset of the first symptoms. The range of neuropsychiatric symptoms in HD is within 33-76%, of which the most frequent are: depression, anxiety disorders, irritability, apathy, obsessive-compulsive disorders and psychotic symptoms [5,6]. Cognitive disorders include memory deterioration, slowed down thinking processes, disorders of executive and visual and spatial functions, problems with organisation, planning and multitasking, difficulties with decision-making and with dealing with new situations [7].

Despite the fact that diagnosing HD is traditionally based on motor symptoms, the highest impact on patients and carers is exerted by mental state and behaviour changes [8,9] as well as cognitive disorders, which may appear as early as 12-15 years before the diagnosis and delay the accurate diagnosis of HD [10,11].

Due to the autosomal dominant type of inheritance HD is passed over from generation to generation and has damaging effects both on the patients and their carers/families, who usually become responsible for caring for the family member with HD [12]. The disease's trajectory in the family may last up to 30 years or even more [13]. The disease being passed from generation to generation leads to situations when for each patient there are 20 people affected by the disease in various ways, such as the risk of HD, issues related to care or the effects of the disease on the family [14].

Complex issues resulting from the hereditary nature of HD, the complexity of symptoms and the concealed onset of the disease [15] have a great impact on the quality of life of family carers [16], who are aware that the disease might develop in other family members. Carers blame themselves for passing the disease on to the next generation [13,17–19].

For each child in a family with a parent having HD the risk of inheriting the gene is 50%, [12]while diagnostic and preclinical trials clearly confirm the genetic status of a person [1] . Carers fear that they might be forced to provide care for several generations, for a few people at the same time, and such a situation might last for years [20].

There are few studies which directly investigate the impact of HD on the quality of life (QoL) of family carers [21]. The existing studies demonstrate that the quality of life of carers is seriously compromised due to their burden [22] these capture a unique sense of long-term isolation and frustration [23] the performed role and conflicting roles, on the one hand requiring caring for the partner and, on the other, for the children, who they need to protect from unexpected behaviour, irritability or aggression of the HD patient [24], insufficient resources to perform the care duties is also of essence[25–27]. Carers experience substantial changes in communication with HD patients [28] and their ability to perform care and parental functions deteriorates as the disease progresses [29,30] and as their physical and mental health declines [18].

The quality of life of carers is considerably reduced due to the lack of access to specialised medical care, while medical personnel are often unaware of the immense impact of HD on the family and unprepared to solve specific problems resulting from the complex changes in HD families [31]. Feeling abandoned by the system, carers describe their experiences with medical professionals as "a lone journey"[27,32]. Family carers were described as "the forgotten" both in the families with HD [33], and in the genetic counselling system [34].

HD is currently incurable and there are no effective treatment methods, the concept of the quality of life is particularly important due to the unique burden carried by carers. Using specific questionnaires to evaluate the QoL of carers makes it possible for physicians and researchers to assess the emotional and physical functioning and lifestyle from the carer's perspective, and does not provide an objective interpretation [21]. It offers a possibility to assess changes in the QoL of carers over time, especially as despite many promising treatment-options, the disease is currently incurable, and one thing is certain: the path towards the solution is long [35].

The existing literature highlights the burden that family carers may face in supporting a loved one living with HD but as yet there is no validated quality of life measure to assess the impact of

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caring on the QoL of family members caring for a loved one with HD in Poland. In Poland there is a lack of studies on the impact of HD on the quality of life of family carers.

According to our knowledge, this is the first study with the participation of Polish family carers on such a numerous group with the use of a specific scale for evaluating the quality of life of carers in HD. Thus far only two reports referring to Polish carers of HD patients have been produced [26,36].

The aim of this study was therefore to validate a translated version of the Huntington's Disease Quality of Life battery for carers (HDQoL-C)[16] for use in Poland.

2. Materials and Methods

2.1. Instrumentation

The HDQoL-C is a multidimensional, disease-specific and subjective health related quality of life tool that incorporates the individual's physical health, psychological state, level of independence, social relationships and personal beliefs. The scale was developed as an outcome measure and in addition can be used to assess subjective QoL in family carers of people with HD. Cronbach's alpha scores for the three components of the original HDQoL-C scale demonstrate good internal consistency - 0.801 (Practical aspects of caregiving); 0.844 (Satisfaction with life) and 0.885 (Feelings about living with HD), with test re-test reliability for the same components being 0.86; 0.90; 0.92 respectively [16]. The scale demonstrates good congruent validity, good face validity and robust content validity. It has been translated and validated with success into French and Italian also[21]. The HDQoL-C is being used in ongoing Euro-Huntington's Disease Burden studies measuring the impact of Huntington's Disease in several European countries and in the USA. To develop the Polish version of the HDQoL-C, forward-backwards translation methods were applied to the original English version of the scale. The translation did not show any errors in translation.

2.2. Participant and Procedure

Participants were recruited among carers taking part in the Enroll-HD study in Polish research centres of the European Huntington`s Disease Network. Contact with carers was also established via the Polish Huntington's Disease Association during the annual conference in Warsaw, Poland devoted to HD, via carers involved in adding new members to the surveyed group, and through the "forum – HD zamki" website. The selection criteria of carers for the study was age ≥ 18 years, the presence of a patient living at home and written consent to participate in the study. All carers who took part in the study had a loved one under their care who had tested positive for HD.
From 100 carers contacted directly, through post or electronic mail, 90 responded to the invitation to take part in the study, which was carried out from June 2015 to December 2016. 18 of 20 carers answered a re-test questionnaire after a 2-week interval to gather data on test–re-test reliability. Because, HD is a rare disease and there is no detailed data indicating the prevalence of disease in Poland, the sample size is likely to be representative for the aim of this study. Ethical approval to conduct this study was granted by the Bioethics Committee of the Medical University of Lublin, Poland, (Protocol number KE-0254/134/2015). A written informed consent was obtained from each participant.

2.3. Data Analysis

Principal components analysis (PCA) was used to explore the inter-relationship between the variables on the HDQoL-C. Bartlett's test of sphericity and the Kaiser– Meyer–Oklin (KMO) measure of sampling adequacy were also used to assess the suitability of the data for PCA.
The Cronbach's α coefficient was used to assess the internal consistency of the scale. A reliability threshold level was considered acceptable when greater than 0,70. Reproducibility assesses if an instrument produces the same results on repeated administrations when respondents have not

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changed. The reliability coefficient was computed by correlating instrument scores for the two administrations.

3. Results

3.1. Respondent Characteristics

The sample comprised 90 carers. 57.8 % of carers were the main carers of a HD patient. The carer ages ranged between 20 and 80 years, with a mean of 48.78 years. Most carers were women (68%) and weren't carers before (81%). 57.8 % carers were married and 41.1% had a child with a risk of disease. Mean length of caring for an HD affected family member was 7.81 years. Table 1 presents the characteristics carers in family with HD.

Table 1. Characteristic of the researched group family carers (n= 90).

Characteristics of family carers	N	%
Women	61	68
Age (years)	48.78 (±15.21)	
Main carer	52	57.8
Marital status		
Married	52	57.8
Single	17	18.9
Widowed	13	14,4
Partnership	5	5.6
Divorced	3	3.3
Family situation		
Number of years since HD knowledge in family	11.76 (± 10.4)	
Have children at risk	37	41.1
Relation with HD patient		
Husband/wife	31	34.4
Parent	26	28.9
Child	14	15.6
Other	10	11
Sibling	8	8.9
Partner	1	1.2
Carer background		
Carer has previously cared any other HD affected person	17	19
Duration of caring (in years)	7.81 (±8.48)	

3.2. Principle Components Analysis and Reliability

In line with the English language version, separate Principle Components Analyses were conducted. Section One of the questionnaire is comprised of demographic information and so was not included in the analysis. For Section Two the Kaiser-Meyer Olkin measure of sampling adequacy showed that the sample was factorable (KMO=. 714). Bartlett's Test of Sphericity was highly significant (χ 2 = 210.4, df=36, p<.001), and low off-diagonal values in the anti-image correlation matrix demonstrated that the data were suitable for factor analysis [37].The analysis revealed two factors. The cross loadings displayed for item QoL2_2 in Factor 1 may indicate cause for concern as it

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also negatively loaded on to factor 2 at -.334. Table 2 outlines the Pattern Matrix of rotated factor loadings for section 2.

Table 2. Pattern Matrix of rotated factor loadings for section 2.

Item	Content	Factor	
		1	2
Qol2_3	How often do you have access to professionals that have specialized knowledge of HD and understand its implications?	.827	-.072
Qol2_4	How much support are you given by health care professionals?	.788	-.030
Qol2_6	How often do you have access to appropriate care facilities?	.766	.004
Qol2_7	How often do you receive any practical support you need?	.754	.036
Qol2_2	How often do you receive appropriate help from social services?	.610	-.334
Qol2_1	How often are you restricted by the need to maintain a regimented daily routine?	.039	.816
Qol2_8	How often do you experience a conflict of interest between what you want and what your HD affected relative wants?	.067	.741
Qol2_5	How often does the inherited nature of HD further complicate your caring role?	-.117	.667
Qol2_9	How often do you sleep well?	-.087	.337

Internal consistency was analysed using Cronbach's Alpha. The items in Factor 1 demonstrated high reliability (Cronbach's $\acute{\alpha}$= .81). Factor 2 demonstrated moderate reliability (Cronbach's $\acute{\alpha}$= .58), which increased to $\acute{\alpha}$= .64 if item QoL2_9 was deleted.

The PCA for Section 3 showed a Kaiser-Meyer Olkin measure of sampling adequacy indicating that the sample was factorable (KMO=. 858). Bartlett's Test of Sphericity was highly significant (χ 2 = 354.3, df=28, p<.001), and low off-diagonal values in the anti-image correlation matrix demonstrated that the data were suitable for factor analysis. The analysis produced a two-factor solution, however the second factor only featured a single item (Qol3_7). Table 3 outlines the Pattern Matrix of rotated factor loadings for section 3.

Table 3. Pattern Matrix of rotated factor loadings for section 3.

Item	Content	Factor	
		1	2
Qol3_2	How satisfied are you with what you achieve in life?	.860	-.128
Qol3_8	How satisfied are you with your overall quality of life?	.825	.246
Qol3_4	How satisfied are you with how safe you feel?	.811	.223
Qol3_1	How satisfied are you with your health?	.792	.000

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Qol3_6	How satisfied are you with your own happiness?	.763	.329
Qol3_5	How satisfied are you with feeling a part of your community?	.656	.373
Qol3_3	How satisfied are you with your close relationships with family or friends?	.566	.464
Qol3_7	How satisfied are you with the treatment that your HD affected relative receives?	.019	.926

The items in Factor 1 demonstrated high reliability (Cronbach's α= .90).

PCA for section 4 indicated a Kaiser-Meyer Olkin measure of sampling adequacy such that the sample was factorable (KMO=.795). Bartlett's Test of Sphericity was highly significant (χ 2 = 569.2, df=136, p<.001), and low off-diagonal values in the anti-image correlation matrix demonstrated that the data were suitable for factor analysis [38]. A four-factor solution was indicated, however, the items in the fourth factor were problematic with high cross loadings. Table 4 outlines the Pattern Matrix of rotated factor loadings for section 4.

Table 4. Pattern Matrix of rotated factor loadings for section 4

Item	Content	Factor			
		1	2	3	4
Qol4_7	I feel sad or depressed	.775	.052	.310	.097
Qol4_17	I feel like I don't know who I am anymore	.765	.087	-.017	.006
Qol4_8	I feel stressed	.749	.051	.143	.129
Qol4_5	I feel exhausted	.729	.126	.186	-.161
Qol4_16	I feel that i have had a "duty of care" forced on me	.711	-.166	-.269	.213
Qol4_10	I feel my own needs are not important to others	.645	-.063	.400	-.157
Qol4_3	I feel isolated	.592	.213	.282	-.154
Qol4_4	I feel there is hope for the future	.036	.797	.287	.004
Qol4_11	I feel comforted by the belief that one day there will be a cure for HD	.024	.773	-.155	.160

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Qol4_13	I feel comforted by my beliefs	.060	.674	.084	.253
Qol4_9	I feel worried about the genetic consequences of HD	.038	-.024	.793	.131
Qol4_1	I feel guilty	.220	.080	.590	-.006
Qol4_6	I feel supported	.176	.386	.474	.125
Qol4_12	I feel that HD brought something positive to my life	-.029	.334	.076	.701
Qol4_15	I feel that HD has made me a stronger person	.166	.404	.246	.673
Qol4_2	I feel financially disadvantaged	.424	.179	.170	-.583
Qol4_14	I feel that I can cope	.282	.322	.467	.546

Internal consistency was again analysed using Cronbach's Alpha. The items in Factor 1 demonstrated high reliability (Cronbach's $\acute{\alpha}$= .85). Factor 2 demonstrated -good reliability (Cronbach's $\acute{\alpha}$= .70), Factor 3 had moderate reliability (Cronbach's $\acute{\alpha}$= .51). Factor 4 was moderately reliable (Cronbach's $\acute{\alpha}$= .56), however, if item QoL4_2 was excluded the reliability of this sub-scale increased considerably (Cronbach's $\acute{\alpha}$= .78).

3.3. Test-Re-test

18 caregivers filled questionnaire again after 2 weeks. Coefficients are presented in the Table 5. All components present high statistically significant correlation ($p<0.001$) with satisfying coefficient of determination ($r2 > 0,6$).

Table 5. Test-retest correlations for Sub-scales

Sub-scales	II	III	IV
II	0.82	-	-
III	-	0.92	-
IV	-	-	0.83

4. Discussion

By measuring QoL in this Polish population we were able to build on our understanding of the issues surrounding caregiving in HD in order to establish ways of improving QoL for this carer group. The translation of the HDQoL-C into an additional language means the scale can be used even more widely, allowing for further comparisons across Europe. The need to translate and adapt QoL instruments for use in languages other than the source language (usually English) has increased

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with the internationalization of clinical trial programmes and cross-cultural research. For example, the ISPOR Task Force[37]note the importance of evidencing similarities in measurement properties between all versions of the same tool to pool analysis and facilitate comparability between countries. The HDQoL-C, has previously been translated into French and Italian [21] and the addition of this Polish translation has the potential to further our understanding of any cross-cultural differences in the resources, policies and practices that may influence the QoL HD family carers on a global level. The Polish version of the shortened versions of the HDQoL-C is similarly valid compared to the original English version. The scale instrument showed satisfactory face validity with little missing data (1,1%). The Cronbach's alpha coefficients demonstrate moderate to good reliability. There were two items, "How often do you sleep well?" (section 2, question 9) and "I feel financially disadvantaged" (section 4, question), that if removed increased the reliability of the subsection they sit within. The question "How often do you sleep well?", has consistently reduced factor reliability even in the original version and was kept in due to the emphasis that carers have placed on it despite not rotating well in the factor analysis. It may be that this question needs refinement in terms of wording to improve reliability or that the subjectivity of this item is difficult to articulate. With regards to the statement "I feel financially disadvantaged" it may be that this statement does not translate cross culturally or is not relevant for a Polish population. It may also be the case that the Polish carers were less focused on financial disadvantage than on the heritable or familiar elements of their experience of caring for someone with HD.

It should also be emphasised that the contact with carers during collecting data for the study was of a unique nature and had a therapeutic effect on them. They expressed that they were pleased that their roles were recognised, as most measures focus on HD patients. This is in line with evidence from [16] who also observed a cathartic benefit of engaging with family carers, many of whom described completing the questionnaire as an intervention itself.

A lack of items bias in English and Polish translations confirms the scale's multi-lingual, multi-cultural consistency and indicates that the scale is easily applicable in other languages. The Polish version of the HDQoL-C demonstrated good internal consistency and congruent validity. Further validation, such as test–retest validity and sensitivity to changes, would enhance this validation process.

5. Study limitations

Although this is the first study in Poland with the use of the HD-specific quality of life questionnaire for family carers, it has some limitations. The group of carers was quite small (90 family carers, only 10 did not respond to the invitation to study) due to the rare occurrence of HD. It should be emphasized that access to individual caregivers is difficult and it is only possible during a conference that is organized annually by the Polish Huntington's Disease Association (only those who benefit from such support) and an online forum during which there is no possibility of direct conversation. The most important thing, however, is that during conversations at the conference and in subsequent telephone conversations, the carers emphasized that just filling out the questionnaire and contacting researchers was a positive experience for them, thanks to which they had the opportunity to verbalize their feelings and difficulties related to the role of a caregiver. This is confirmed by the comments at the end of the questionnaire, in which the caregivers thanked for noticing them and understanding. At the same time, they emphasized that they do not have the closest people with whom they could talk, because the family avoids topics about HD. Of the entire study group, only 7 people did not leave contact, which also indicates their needs. The group of 90 respondents was also created thanks to carers who willingly gave contact to other caregivers.

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6. Practice Implications

Interest in the QoL of carers is crucial due to the practical, socio-economic aspects. In the situation when care-related costs are borne mainly by carers and not the State budget, incapacities of carers generate unplanned expenses for medical and social assistance [39]. Due to the fact that carers feel like a forgotten group in HD families, it is necessary to promote quality of life in primary healthcare. The time of caring for a patient with HD is much longer than in other neurodegenerative diseases and the need for environmental care is also longer. It is recommended to study the quality of life of HD caregivers in primary healthcare in order to implement appropriate support procedures at various stages of the disease, the more so that carers can also get ill and pass defective genes to their children.

7. Research Recommendations

This study is part of a wider project on family carers in HD in Poland. Further research, with quantitative and qualitative approach, can identify other areas of quality of life for HD families. Moreover, due to the multitude of symptoms (multifaceted disease) conducting interdisciplinary research would allow a better understanding of the needs of family carers and this would translate into providing them with practical support. Therefore, further research based on mixed methodology and done within multidisciplinary teams is recommended.

8. Conclusions

A lack of item bias in English and Polish translations confirms the scale's multi-lingual, multi-cultural consistency and indicates that the scale is easily applicable in other languages. The Polish version of the HDQoL-C demonstrated good internal consistency and congruent validity. Further validation, such as sensitivity to changes, would enhance this validation process. The HDQoL-C has demonstrated a wide range of benefits for practitioners in capturing and understanding carer experience and these benefits can now confidently be extended to Polish speaking populations.

Availability of data and materials: The HDQoL-C is the property of Dr Aimee Aubeeluck, CPsychol, FHEA. It has been developed for use by family members, researchers and clinicians and can be used and adapted freely for the benefit of improving the quality of life of families living with Huntington's Disease. To use the scale, contact is required: aimee.aubeeluck@nottingham.ac.uk and cite in any subsequent write up.

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References

1. MacDonald, M.E.; Ambrose, C.M.; Duyao, M.P.; Myers, R.H.; Lin, C.; Srinidhi, L.; Barnes, G.; Taylor, S.A.; James, M.; Groot, N.; et al. A novel gene containing a trinucleotide repeat that is expanded and unstable on Huntington's disease chromosomes. *Cell* **1993**, *72*, 971–983.
2. Ross, C.A.; Aylward, E.H.; Wild, E.J.; Langbehn, D.R.; Long, J.D.; Warner, J.H.; Scahill, R.I.; Leavitt,

Peer-reviewed version available at Int. J. Environ. Res. Public Health 2019, 16, 2323; doi:10.3390/ijerph16132323

Stout, J.C.; Paulsen, J.S.; et al. Huntington disease: Natural history, biomarkers and prospects for therapeutics. *Nat. Rev. Neurol.* **2014**, *10*, 204–216.

3. Novak, M.J.U.; Tabrizi, S.J. Huntington's disease. *BMJ* **2010**, *340*, 34–40.
4. Robins Wahlin, T.-B.; Larsson, M.U.; Luszcz, M.A.; Byrne, G.J. WAIS-R Features of Preclinical Huntington's Disease: Implications for Early Detection. *Dement. Geriatr. Cogn. Disord.* **2010**, *29*, 342–350.
5. Epping, E.A.; Paulsen, J.S. Depression in the early stages of Huntington disease Huntington disease: beyond motor symptoms. *Neurodegener. Dis. Manag.* **2011**, *1*, 407–414.
6. van Duijn, E.; Kingma, E.M.; van der Mast, R.C. Psychopathology in Verified Huntington's Disease Gene Carriers. *J. Neuropsychiatry Clin. Neurosci.* **2007**, *19*, 441–448.
7. Nopoulos, P.C. Huntington disease: A single-gene degenerative disorder of the striatum. *Dialogues Clin. Neurosci.* **2016**, *18*, 91–98.
8. Paulsen, J.S. Cognitive impairment in Huntington disease: Diagnosis and treatment. *Curr. Neurol. Neurosci. Rep.* **2011**, *11*, 474–483.
9. Eddy, C.M.; Parkinson, E.G.; Rickards, H.E. Changes in mental state and behaviour in Huntington's disease. *The Lancet Psychiatry* **2016**, *3*, 1079–1086.
10. Williams, J.K.; Kim, J.I.; Downing, N.; Farias, S.; Harrington, D.L.; Long, J.D.; Mills, J.A.; Paulsen, J.S. Everyday cognition in prodromal huntington disease. *Neuropsychology* **2015**, *29*, 255–267.
11. Stout, J.C.; Paulsen, J.S.; Queller, S.; Solomon, A.C.; Whitlock, K.B.; Campbell, J.C.; Carlozzi, N.; Duff, K.; Beglinger, L.J.; Langbehn, D.R.; et al. Neurocognitive Signs in Prodromal Huntington Disease. *Neuropsychology* **2011**, *25*, 1–14.
12. Williams, J.K.; Skirton, H.; Barnette, J.J.; Paulsen, J.S. Family carer personal concerns in Huntington disease. *J. Adv. Nurs.* **2012**, *68*, 137–146.
13. Williams, J.K.; Skirton, H.; Paulsen, J.S.; Tripp-Reimer, T.; Jarmon, L.; McGonigal Kenney, M.; Birrer, E.; Hennig, B.L.; Honeyford, J. The emotional experiences of family carers in Huntington disease. *J. Adv. Nurs.* **2009**, *65*, 789–798.
14. Hayden, M.R.; Ehrlich, R.; Parker, H.; Ferera, S.J. Social perspectives in Huntington's chorea. *South African Med. J.* **1980**, *58*, 201–3.
15. Hocaoglu, M.B.; Gaffan, E.A.; Ho, A.K. The Huntington's Disease health-related Quality of Life questionnaire (HDQoL): A disease-specific measure of health-related quality of life. *Clin. Genet.* **2012**, *81*, 117–122.
16. Aubeeluck, A.; Buchanan, H. The Huntington's Disease quality of life battery for carers: Reliability and validity. *Clin. Genet.* **2007**, *71*, 434–445.
17. Vamos, M.; Hambridge, J.; Edwards, M.; Conaghan, J. The Impact of Huntington's Disease on Family Life. *Psychosomatics* **2007**, *48*, 400–404.
18. Pickett, T.; Altmaier, E.; Paulsen, J.S. Caregiver Burden in Huntington's Disease. *Rehabil. Psychol.* **2007**, *52*, 311–318.
19. Soltysiak, B.; Gardiner, P.; Skirton, H. Exploring supportive care for individuals affected by Huntington disease and their family caregivers in a community setting. *J. Clin. Nurs.* **2008**, *17*, 226–234.
20. Aubeeluck, A.V.; Buchanan, H.; Stupple, E.J.N. 'All the burden on all the carers': exploring quality of life with family caregivers of Huntington's disease patients. *Qual. Life Res.* **2012**, *21*, 1425–1435.
21. Aubeeluck, A.; Dorey, J.; Squitieri, F.; Clay, E.; Stupple, E.J.N.; De Nicola, A.; Buchanan, H.; Martino, T.; Toumi, M. Further evidence of reliability and validity of the Huntington's disease quality of life battery for carers: Italian and French translations. *Qual. Life Res.* **2013**, *22*, 1093–1098.

Peer-reviewed version available at Int. J. Environ. Res. Public Health 2019, 16, 2323; doi:10.3390/ijerph16132323

22. Aubeeluck, A.; Buchanan, H. Capturing the Huntington's disease spousal carer experience: a preliminary investigation using the 'Photovoice' method. *Dementia* **2006**, *5*, 95–116.
23. Aubeeluck, A.; Moskowitz, C.B. Huntington's disease. Part 3: family aspects of HD. *Br. J. Nurs.* **2008**, *17*, 328–331.
24. Røthing, M.; Malterud, K.; Frich, J.C. Caregiver roles in families affected by Huntington's disease: a qualitative interview study. *Scand. J. Caring Sci.* **2014**, *28*, 700–705.
25. Roscoe, L.A.; Corsentino, E.; Watkins, S.; McCall, M.; Sanchez-Ramos, J. Well-Being of family caregivers of persons with late-stage Huntington's disease: Lessons in Stress and coping. *Health Commun.* **2009**, *24*, 239–248.
26. Domaradzki, J. Family caregivers' experiences with healthcare services – a case of Huntington's disease. *Psychiatr. Pol* **2016**, *50*, 375–391.
27. Skirton, H.; Williams, J.K.; Jackson Barnette, J.; Paulsen, J.S. Huntington disease: Families' experiences of healthcare services. *J. Adv. Nurs.* **2010**, *66*, 500–510.
28. Hartelius, L.; Jonsson, M.; Rickeberg, A.; Laakso, K. Communication and Huntington's disease: Qualitative interviews and focus groups with persons with Huntington's disease, family members, and carers. *Int. J. Lang. Commun. Disord.* **2010**, *45*, 381–393.
29. Keenan, K.F.; Miedzybrodzka, Z.; van Teijlingen, E.; McKee, L.; Simpson, S.A. Young people's experiences of growing up in a family affected by Huntington's disease. *Clin. Genet.* **2007**, *71*, 120–129.
30. Williams, J.K.; Ayres, L.; Specht, J.; Sparbel, K.; Klimek, M. Lou Caregiving by teens for family members with Huntington disease. *J. Fam. Nurs.* **2009**, *15*, 273–294.
31. Skirton, H.; Glendinning, N. Using research to develop care for patients with Huntington's disease. *Br. J. Nurs.* **1997**, *6*, 83–90.
32. Etchegary, H. Healthcare experiences of families affected by Huntington disease: Need for improved care. *Chronic Illn.* **2011**, *7*, 225–238.
33. Kessler, S. Forgotten person in the huntington disease family. *Am. J. Med. Genet.* **1993**, *48*, 145–150.
34. Decruyenaere, M.; Evers-Kiebooms, G.; Boogaerts, A.; Demyttenaere, K.; Dom, R.; Fryns, J.P. Partners of mutation-carriers for Huntington's disease: Forgotten persons? *Eur. J. Hum. Genet.* **2005**, *13*, 1077–1085.
35. Ross, R.A.C. Huntington's disease: A clinical review. *Orphanet J. Rare Dis.* **2010**, *5*, 40.
36. Banaszkiewicz, K.; Sitek, E.J.; Monika, R.; Sołtan, W.; Jarosław, S.; Szczudlik, A. Huntington's disease from the patient, caregiver and physician's perspectives: three sides of the same coin? *J Neural Transm* **2012**, *119*, 1361–1365.
37. Wild, D.; Grove, A.; Martin, M.; Eremenco, S.; McElroy, S.; Verjee-Lorenz, A.; Erikson, P. Principles of good practice for the translation and cultural adaptation process for patient-reported outcomes (PRO) measures: Report of the ISPOR Task Force for Translation and Cultural Adaptation. *Value Heal.* **2005**, *8*, 94–104.
38. Tabachnick, B., Fidell, L. *Using multivariate statistics.*; Pearson/Allyn & Bacon, 1996; ISBN 0205459382.
39. Ciałkowska-Kuźmińska, M.; Kasprzak, M. Obciążenie opiekunów jako czynnik delegowania opieki do instytucji psychogeriatrycznych. *Psychogeriatria Pol.* **2012**, *9*, 69–76.