

# **Supplementary Material**

## **Disease Burden of Huntington's Disease (HD) on People Living with HD and Care Partners in Canada**

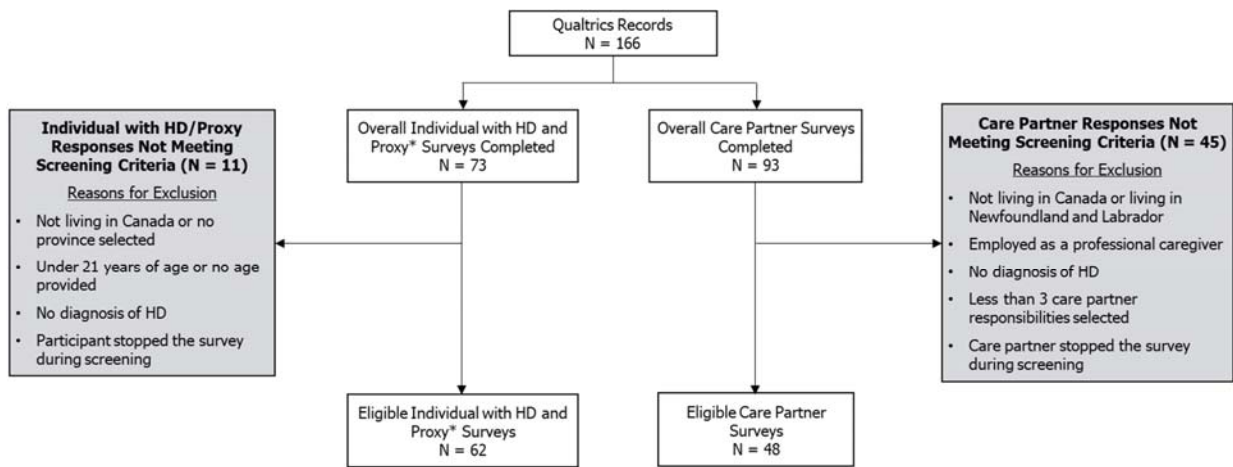
### **Survey Questionnaires**

HD Patient-Caregiver Survey\_CAN\_EN.pdf

HD Patient-Caregiver Survey\_CAN\_FR.pdf

HD Patient-Caregiver Survey\_AB\_EN.pdf

**Supplementary Figure 1. Patient and Care Partner Survey Respondent Inclusion Flow Chart**



\* Proxy surveys could be completed by a designated proxy or by care partners of persons with HD

HD, Huntington's disease

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**Supplementary Table 1. Self-Reported HD Stage Scoring Approach**

<b>Scale</b>	<b>Ability</b>	<b>Score</b>
<b>Occupation</b>	Unable	0
	Marginal work only	1
	Reduced capacity for usual job	2
	Normal	3
	Do not know / not sure	<b>End Scoring</b>
<b>Finances</b>	Unable	0
	Major assistance	1
	Slight assistance	2
	Normal	3
	Do not know / not sure	<b>End Scoring</b>
<b>Domestic chores</b>	Unable	0
	Impaired	1
	Normal	2
	Do not know / not sure	<b>End Scoring</b>
<b>Activities of daily living (i.e., self-care activities, such as self-feeding, bathing, dressing, grooming, etc.)</b>	Total care	0
	Major impairment	1
	Minimal impairment	2
	Normal	3
	Do not know / not sure	<b>End Scoring</b>
<b>Care level</b>	Full-time nursing care	0
	Home with chronic care	1
	Home independently	2
	Do not know / not sure	<b>End Scoring</b>
<b>IF 11-13 POINTS, CATEGORIZE STAGE 1</b> <b>IF 7-10 POINTS, CATEGORIZE STAGE 2</b> <b>IF 3-6 POINTS, CATEGORIZE STAGE 3</b> <b>IF 0-2 POINTS, CATEGORIZE STAGE 4</b>		

HD, Huntington's disease

Although this survey rating scale is similar to the clinical Shoulson-Fahn functional capacity rating scale, it is not directly comparable.

**Supplementary Table 2. SF-36v1 Scale and Summary Scores<sup>a</sup> for Individuals with HD – Stratified by Motor Transition Status<sup>b</sup>**

Scale	Have Not Transitioned			Transitioned			p <sup>c</sup>
	n	Mean (SD)	Median (IQR)	n	Mean (SD)	Median (IQR)	
Bodily Pain	29	76.0 (21.9)	74.0 (62.0 - 100.0)	19	71.9 (35.0)	100.0 (41.0 - 100.0)	0.836
Mental Health	29	69.8 (15.2)	72.0 (56.0 - 80.0)	18	58.2 (21.4)	60.0 (52.0 - 72.0)	0.068
Physical Functioning	29	81.9 (30.0)	95.0 (80.0 - 100.0)	19	33.2 (31.1)	25.0 (10.0 - 55.0)	<0.001
Social Functioning	29	72.4 (25.1)	75.0 (50.0 - 100.0)	19	42.1 (26.7)	37.5 (25.0 - 62.5)	<0.001
General Health	29	60.9 (22.0)	67.0 (45.0 - 75.0)	18	45.6 (22.0)	46.0 (30.0 - 55.0)	0.018
Vitality	29	59.8 (19.7)	60.0 (45.0 - 75.0)	18	43.9 (23.7)	47.5 (35.0 - 60.0)	0.040
Role – Emotional Scale	29	59.8 (43.1)	66.7 (0.0 - 100.0)	19	24.6 (36.6)	0.0 (0.0 - 66.7)	0.006
Role – Physical Scale	29	67.8 (39.6)	100.0 (33.3 - 100.0)	19	17.1 (26.4)	0.0 (0.0 - 25.0)	<0.001
<b>Physical Component Summary Score</b>	29	48.7 (11.3)	51.9 (45.9 - 54.8)	18	35.0 (11.5)	30.4 (27.2 - 38.9)	0.002
<b>Mental Health Component Summary Score</b>	29	45.1 (11.7)	47.4 (40.0 - 54.0)	18	38.5 (11.3)	41.5 (28.5 - 45.2)	0.080
<b>Mapped EQ-5D Utility Score<sup>d</sup></b>	29	0.82 (0.15)	0.88 (0.72 - 0.94)	18	0.57 (0.27)	0.62 (0.45 - 0.75)	<0.001

HD, Huntington's disease; IQR, interquartile range; SD, standard deviation

Results are not shown for those who did not answer the question on motor transition status.

<sup>a</sup> Individual SF-36v1 scales were scored following the SF-36 Interpretation and Scoring Guide [1]. Physical Component Summary and Mental Component Summary scores were calculated using the approach outlined by Ware et al. (1994) in the SF-36 Physical and Mental Health Summary Scales: A User's Manual utilizing Canadian normative values [2, 3]. SF-36v1 scales have been transformed to a 0-100 scale, with a lower score representing a higher burden.

<sup>b</sup> Motor transition status was self-reported and determined based on the question: "Have you/the person living with HD transitioned to early-stage motor onset (i.e., stage 1 or 2)?"

<sup>c</sup> P-values for continuous variables are based on the Kruskal-Wallis test comparing mean ranks of respondents based on self-reported motor transition status.

<sup>d</sup> The SF-36 was mapped to the EQ-5D using methodology outlined by Rowen et al. (2009) [4] to generate a utility score from 0-1, with a score of 1 representing full health.

**Supplementary Table 3. SF-36v1 Scale and Summary Scores<sup>a</sup> Stratified by Individuals with HD vs. Proxy<sup>b</sup> Survey Respondents**

Scale	Respondents with HD			Proxy Respondent			p <sup>c</sup>
	n	Mean (SD)	Median (IQR)	n	Mean (SD)	Median (IQR)	
Bodily Pain	36	78.3 (22.5)	84.0 (62.0 - 100.0)	13	62.5 (36.1)	62.0 (41.0 - 100.0)	0.211
Mental Health	35	67.0 (17.2)	72.0 (52.0 - 80.0)	13	62.2 (21.7)	64.0 (56.0 - 76.0)	0.552
Physical Functioning	36	78.5 (29.3)	92.2 (67.5 - 100.0)	13	14.2 (17.4)	10.0 (0.0 - 15.0)	<0.001
Social functioning	36	68.8 (25.1)	75.0 (50.0 - 87.5)	13	34.6 (27.6)	37.5 (12.5 - 50.0)	<0.001
General Health	35	61.1 (20.2)	62.0 (45.0 - 75.0)	13	38.5 (21.8)	42.0 (25.0 - 50.0)	0.002
Vitality	35	58.1 (20.1)	55.0 (40.0 - 75.0)	13	39.6 (24.0)	45.0 (25.0 - 50.0)	0.026
Role – Emotional Scale	36	56.5 (42.0)	66.7 (0.0 - 100.0)	13	20.5 (39.8)	0.0 (0.0 - 0.0)	0.010
Role – Physical Scale	36	62.3 (39.5)	75.0 (29.2 - 100.0)	13	3.8 (9.4)	0.0 (0.0 - 0.0)	<0.001
<b>Physical Component Summary Score</b>	35	48.5 (10.4)	51.9 (45.1 - 54.9)	13	28.0 (7.9)	28.9 (22.6 - 30.7)	<0.001
<b>Mental Health Component Summary Score</b>	35	43.5 (11.9)	43.8 (37.1 - 53.7)	13	41.0 (12.0)	42.3 (34.8 - 48.8)	0.570
<b>Mapped EQ-5D Utility Score<sup>d</sup></b>	35	0.81 (0.15)	0.86 (0.72 - 0.94)	13	0.48 (0.27)	0.48 (0.45 - 0.63)	<0.001

HD, Huntington's disease; IQR, interquartile range; SD, standard deviation

<sup>a</sup> Individual SF-36v1 scales were scored following the SF-36 Interpretation and Scoring Guide [1]. Physical Component Summary and Mental Component Summary scores were calculated using the approach outlined by Ware et al. (1994) in the SF-36 Physical and Mental Health Summary Scales: A User's Manual utilizing Canadian normative values [2, 3]. SF-36v1 scales have been transformed to a 0-100 scale, with a lower score representing a higher burden.

<sup>b</sup> A proxy respondent was defined as a helper/representative that completed the survey on behalf of the individual with HD if they were unable to complete the survey independently.

<sup>c</sup> P-values for continuous variables are based on the Kruskal-Wallis test comparing mean ranks of respondents based on responses from individuals with HD versus proxy respondents.

<sup>d</sup> The SF-36 was mapped to the EQ-5D using methodology outlined by Rowen et al. (2009) [4] to generate a utility score from 0-1, with a score of 1 representing full health.

**Supplementary Table 4. Caregiver Strain Index<sup>a</sup> Results for Care Partners**

<b>Caregiver Strain Index, n (%)</b>	<b>All Respondents (n=40)</b>
It is upsetting to find the person I care for has changed so much from his/her former self	30 (75.0)
I feel completely overwhelmed	27 (67.5)
Some behavior is upsetting	26 (65.0)
There have been other demands on my time	24 (60.0)
There have been emotional adjustments	23 (57.5)
My sleep is disturbed	20 (50.0)
Caregiving is confining	20 (50.0)
There have been family adjustments	20 (50.0)
There have been changes in personal plans	20 (50.0)
Caregiving is inconvenient	17 (42.5)
Caregiving is a financial strain	15 (37.5)
There have been work adjustments	12 (30.0)
Caregiving is a physical strain	11 (27.5)
<b>Total Caregiver Strain Index Score (n)</b>	38
Mean (SD)	6.9 (3.8)
Median (IQR)	8.0 (3.0 - 10.0)
<b>Respondents with high stress/burden (score <math>\geq 7</math>), n (%)</b>	21 (55.3)

HD, Huntington's disease; IQR, interquartile range; SD, standard deviation

<sup>a</sup> The Caregiver Strain Index was scored from 0-13 by summing the number of "Yes" responses. A score of  $\geq 7$  indicates a high level of stress and burden.

**Supplementary Table 5. HDQoL-C<sup>a</sup> Section 2, 3, and 4 Results for Care Partners**

<b>Characteristic</b>	<b>Respondents (n)</b>	<b>Mean (SD)</b>	<b>Median (IQR)</b>
<b>Feelings about role as a caregiver<sup>b</sup></b>			
How often are you restricted by need to maintain regimented daily routine	38	4.9 (3.0)	5.0 (2.0 - 7.0)
How often do you receive appropriate help from social services	38	3.3 (3.0)	3.0 (0.0 - 6.0)
How often do you have access to professionals that have specialized HD knowledge	38	5.3 (2.8)	6.0 (3.0 - 8.0)
How much support are you given by health care professionals	38	4.8 (2.8)	5.0 (2.0 - 7.0)
How often do the genetic consequences of HD impact your caring role	37	5.3 (2.6)	5.0 (4.0 - 8.0)
How often do you have access to appropriate care facilities	37	4.2 (3.0)	5.0 (2.0 - 6.0)
How often do you receive practical support you need	37	4.6 (3.0)	5.0 (2.0 - 7.0)
How often do you experience a conflict of interest between what you want and what your HD affected relative wants	38	5.3 (3.0)	5.0 (4.0 - 8.0)
How often do you sleep well	38	4.4 (2.6)	4.0 (2.0 - 7.0)
<b>Summary Score (%) – Section 2</b>	36	46.5 (16.3)	48.9 (35.0 - 56.1)
<b>Satisfaction with different areas of life</b>			
How satisfied are you with your health	38	5.8 (2.8)	6.0 (4.0 - 8.0)
How satisfied are you with what you achieve in life	38	5.9 (2.8)	6.0 (5.0 - 8.0)
How satisfied are you with your close relationships with family or friends	38	6.2 (2.7)	7.0 (4.0 - 8.0)
How satisfied are you with how safe you feel	38	7.4 (2.5)	8.0 (6.0 - 9.0)
How satisfied are you with feeling a part of your community	38	6.2 (2.7)	7.0 (5.0 - 8.0)
How satisfied are you with your own happiness	38	5.7 (2.8)	6.0 (4.0 - 8.0)
How satisfied are you with the treatment that your HD affected relative receives	38	6.2 (3.1)	7.0 (4.0 - 9.0)
How satisfied are you with your overall quality of life	38	6.1 (2.8)	7.0 (4.0 - 8.0)
<b>Summary Score (%) – Section 3</b>	38	62.1 (21.3)	60.0 (46.3 - 78.8)
<b>Feelings about different areas of life</b>			
I feel guilty	37	5.0 (3.0)	5.0 (3.0 - 7.0)
I feel financially disadvantaged	37	5.0 (3.5)	6.0 (2.0 - 8.0)
I feel isolated	37	6.0 (3.3)	7.0 (4.0 - 9.0)
I feel there is hope for the future	37	5.5 (3.3)	5.0 (2.0 - 8.0)

I feel exhausted	37	6.7 (2.6)	7.0 (5.0 - 9.0)
I feel supported	37	5.4 (2.7)	5.0 (4.0 - 7.0)
I feel sad or depressed	37	4.9 (3.0)	5.0 (3.0 - 6.0)
I feel stressed	37	6.7 (2.6)	7.0 (5.0 - 9.0)
I feel worried about the genetic consequences of HD	36	6.9 (3.5)	8.0 (5.0 - 10.0)
I feel my own needs are not important to others	37	6.5 (3.2)	7.0 (5.0 - 9.0)
I feel comforted by the belief that one day there will be a cure for HD	37	5.3 (3.1)	6.0 (3.0 - 7.0)
I feel that HD brought something positive to my life	37	2.3 (2.8)	1.0 (0.0 - 4.0)
I feel comforted by my beliefs	36	4.8 (3.5)	4.0 (2.0 - 8.0)
I feel that I can cope	37	5.9 (3.0)	6.0 (4.0 - 8.0)
I feel that HD has made me a stronger person	36	5.6 (3.7)	6.0 (2.0 - 9.0)
I feel that I have had a duty of care forced on me	37	5.9 (3.3)	6.0 (3.0 - 9.0)
I feel like I don't know who I am anymore	37	3.8 (3.3)	3.0 (1.0 - 7.0)
<b>Summary Score (%) – Section 4</b>	34	45.1 (17.9)	48.8 (32.4 - 55.9)

HD, Huntington's disease; HDQoL-C, Huntington's Disease Quality of Life Battery for Carers; IQR, interquartile range; SD, standard deviation

<sup>a</sup> The HDQoL-C was scored according to the HDQoL-C User Guide. HDQoL-C summary scores were transformed to a percent score for Sections 2-4, where a 100% score represents optimum quality of life.

<sup>b</sup> Responses may be skewed as the data analyzed from the survey excluded one level of response.



## **REFERENCES**

- [1] Ware JE, Jr., Snow KK, Kosinski M, Gandek B. SF-36 Health Survey Manual and Interpretation Guide. Boston, MA: Health Institute, New England Medical Center; 1993.
- [2] Ware JE, Jr., Kosinski M, Keller SD. SF-36 Physical and Mental Health Summary Scales: A User's Manual. Boston, MA: Health Assessment Lab; 1994.
- [3] Hopman WM, Towheed T, Anastassiades T, Tenenhouse A, Poliquin S, Berger C, et al. Canadian normative data for the SF-36 health survey. *CMAJ*. 2000;163(3):265-71.
- [4] Rowen D, Brazier J, Roberts J. Mapping SF-36 onto the EQ-5D index: how reliable is the relationship? *Health Qual Life Outcomes*. 2009;7:27.