

Work Productivity and Activity Impairment of Huntington’s Disease patients by disease stage in the US and EU5: Evidence from the Huntington’s Disease Burden of Illness Study (HDBOI)

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Introduction

- Huntington’s Disease (HD) is a rare, inherited, and highly complex neuro-degenerative disorder, affecting all aspects of an individual’s life, including physical, cognitive, and behavioural processes.¹
- The symptoms associated with HD have a strong impact on functional capacity and affect a person’s ability to perform work functions and maintain employment and may lead to reduced work hours or cessation of work.^{2,3}
- The literature suggests that people with HD generally overestimate their abilities and under-report the severity of their deficits.⁴

Objective

The aim of this research is to descriptively explore differences in Work Productivity and Activity Impairment Specific Health Problem (WPAI-SHP) of HD participants by disease stage.

Methods

- The HDBOI study is a retrospective, cross-sectional dataset that captures demographic, health resource utilization, and health-related quality of life of a cohort of HD participants and their caregivers.
- Countries included in the study were Germany, France, Italy, Spain, the UK and the USA.
- Data was collected via a **Patient and Public Involvement and Engagement (PPIE-P) questionnaire**; an optional questionnaire completed by participants capturing information on WPAI-SHP.
 - Caregivers of HD participants who were unable to complete the PPIE-P due to physical or cognitive impairments, were asked to complete it as proxy-respondents.
- Participants were categorized as early, mid, or advanced stage by the treating physician. Physicians were provided with the Unified Huntington’s Disease Rating Scale Total Functional Capacity score⁵ and corresponding disease stages to guide their assessment of disease stage.

Outcomes

- The **WPAI-SHP** questionnaire was administered to participants and is a validated and widely-used instrument for measuring the impact of a condition on an individual’s work and activities during the previous seven days.⁶
 - It is a 6-item instrument which measures the following, attributed to HD related health problems: (i) Work time missed (Absenteeism), (ii) Impaired productivity at work (Presenteeism), (iii) Impairment in daily activities, and (iv) Overall work productivity loss (overall work impairment/ absenteeism plus presenteeism)
- WPAI-SHP outcomes were explored by disease stage. Each outcome is expressed as impairment percentages, with higher numbers indicating greater impairment and less productivity.

Results

Sample characteristics

- The analytic sample with information on WPAI-SHP was comprised of 482 HD participants, of which 42% were early-stage (ES), 34% mid stage (MS), and 24% advanced stage (AS).
- Approximately 9% (N=42) of responses were proxy-reported, with 29% in ES, 21% in MS, and 50% in AS.
- Within the PPIE-P sample 42% (N=185) of participants reported being employed, compared with just 7% (N=3) in employment in the proxy-reported group (See **Table 1**).
- Approximately 27% of the PPIE-P sample reported stopping work early (i.e., retiring) due to HD.

Table 1. Employment status of self and proxy reported PPIE-P by disease stage

	All Sample		Early		Mid		Advanced	
	N	Percent	N	Percent	N	Percent	N	Percent
Self-reported	440	91	192	44	155	35	93	21
Proxy-reported	42	9	12	29	9	21	21	50
All sample	482	100	204	42	164	34	114	24
Self-reported	N	Percent	N	Percent	N	Percent	N	Percent
Employed	185	42	110	57	53	34	22	24
Not employed	255	58	82	43	102	65	71	76
Proxy-reported	N	Percent	N	Percent	N	Percent	N	Percent
Employed	3	7	2	17	0	.	1	5
Not employed	39	93	10	83	9	100	20	95

Note: Early/Mid/Advanced stage is based on disease progression as defined by the treating physician

WPAI-SHP outcomes

- Overall **impairment in daily activities** increased with disease severity for both self-reported and proxy-reported groups (See **Table 2**).
 - Generally, reported impairment in daily activities was larger in the proxy-reported group compared with the self-reported group; 79% versus 58% respectively.
- Of those who reported being employed for pay overall **work productivity loss** increased with disease severity: 43% in ES, 55% in MS, and 57% in AS.
- It is unusual for MS or AS participants to maintain employment given the progressive worsening of symptoms at later stages of the disease. The percentages employed for pay in the MS and AS groups in the HDBOI study were higher than expected and this may be attributable to an overestimation of abilities by respondents.⁴

References

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Results continued

Table 2. Work Productivity and Activity Impairment Specific Health Problems outcomes by disease stage

Impairment in daily activities	All Sample			Early Stage			Mid Stage			Advanced Stage		
	N	Mean	Std. Dev.	N	Mean	Std. Dev.	N	Mean	Std. Dev.	N	Mean	Std. Dev.
Self-reported	440	58.1%	0.25	192	46.4%	0.26	155	63.6%	0.21	93	72.9%	0.20
Proxy-reported	42	78.8%	0.19	12	68.3%	0.23	9	80.0%	0.12	21	84.3%	0.16
Work time missed	Std.			Std.			Std.			Std.		
	N	Mean	Dev.	N	Mean	Dev.	N	Mean	Dev.	N	Mean	Dev.
Self-reported	134	11.5%	0.19	83	9.1%	0.13	35	14%	0.26	16	18.3%	0.20
Proxy-reported	-	-	-	-	-	-	-	-	-	-	-	-
Impaired productivity at work	Std.			Std.			Std.			Std.		
	N	Mean	Dev.	N	Mean	Dev.	N	Mean	Dev.	N	Mean	Dev.
Self-reported	134	42.2%	0.24	83	38.3%	0.27	35	49.1%	0.19	16	47.5%	0.17
Proxy-reported	-	-	-	-	-	-	-	-	-	-	-	-
Percent overall work productivity loss	Std.			Std.			Std.			Std.		
	N	Mean	Dev.	N	Mean	Dev.	N	Mean	Dev.	N	Mean	Dev.
Self-reported	134	48%	0.27	83	43%	0.29	35	55%	0.23	16	57%	0.20
Proxy-reported	-	-	-	-	-	-	-	-	-	-	-	-

Note: Those who reported being employed for pay but had not attended work in the previous 7 days were excluded from WPAI-SHP outcomes, hence numbers in employment in Table 1 and Table 2 do not match.

Proxy-reported WPAI-SHP

- The results of WPAI-SHP self-reported contrast with proxy-reported, where only three participants (of 42) were employed for pay, with the majority either retired (N=15), unemployed (N=11), or on sick leave (N=10).
 - Of those who reported being employed in the proxy-reported group, all had not attended work in the previous 7 days and were not included in WPAI-SHP outcomes for this reason.

Conclusion

- Results from the HDBOI study show that people with HD bear a significant economic and humanistic burden, affecting both daily activity and work-related productivity, which is greater at later stages of disease.
- Observed discrepancies between self-reported and proxy-reported WPAI-SHP and higher than anticipated employment levels among MS and AS groups may be related to the well-documented unawareness of deficits exhibited by people with HD, and this topic warrants further research.
- WPAI-SHP may not be the most appropriate tool for assessing productivity outcomes, due to the self-reported nature of this tool and the difficulties with insight in the HD population, hence it may not reflect the actual work productivity loss at later stages of disease.

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