



Annotated Case Report Form (CRF) for Ad Hoc Periodic Dataset

Version 2016-10-R1

Enroll-HD

A worldwide observational study for Huntington's disease families

A CHDI Foundation Project

Table of Contents

1	Purpose of this Document	3
2	Forms and Data Files	4
3	Form “Enrollment”	5
4	Form “Variable Items”	6
5	Form “UHDRS Motor/Diagnostic Confidence (Motor)”	7
6	Form “UHDRS Total Funtional Capacity (TFC)”	12
7	Form “UHDRS Functional Assessment/Independence Scale (Function)”	13
8	Form “Mini Mental State Examination (MMSE)”	14
9	Form “Core and Extended Cognitive Assessment (Cognitive)”	15
	Revision History	18

1 Purpose of this Document

The purpose of this document is to provide an annotated view of the eCRF (electronic Case Report Form) of Enroll-HD's Ad Hoc Visits. It shows the source of the variable names provided in the Enroll-HD plus periodic dataset, which is available to the HD research community at large. The periodic dataset represents a data extract of the Enroll-HD Electronic Data Capture (EDC) database at a defined point in time. Information that could potentially identify a participant has been removed or de-identified within this dataset. When available, CISC SDMT naming conventions were used.

The annotated view shows all forms, variables, and variable names used within the periodic dataset. Forms and variables that are not provided in the dataset are not shown in the annotated forms. Variables that have been transformed for de-identification reasons are shown in the transformed manner in the annotated view.

The variables and data files of the dataset are described in detail within the [data dictionary](#) for the Enroll-HD plus periodic dataset.

2 Forms and Data Files

Forms are CRF entry screens that are displayed to data entry personnel and show how data is entered in the eCRF. Data files are sets of variables that define how the data are represented. The following table defines how forms and data files are related.

Form / Data File	Profile	Participation	Ad Hoc
<i>Enrollment</i>	✓	✓	
<i>Variable</i>			✓
<i>Motor</i>			✓
<i>TFC</i>			✓
<i>Function</i>			✓
<i>Cognitive</i>			✓
<i>MMSE</i>			✓

The data files “Profile” and “Participation” are not displayed on an entry form but consist of variables of various forms. It contains key information like subject, study and region identifiers and some visit-independent subject information, e.g. the subject’s latest HD classification. Please refer to the [data dictionary](#) for detailed information.

3 Form “Enrollment”

Enrollment [enrlmnt1] 5452_1	
Day of informed consent:	<input type="text"/> rfidy
Optional Components: [enrlmnt2] 359_1	Family History: <input type="radio"/> yes 1 <input type="radio"/> no 0 fhx

4 Form “Variable Items”

[varitems1]

General Variable Items [general] 652_1

[varitems2]

Weight (kg)	Weight (lbs)
<input type="text"/> weight	<input type="text"/> weight_2
Height (cm)	Height (inches)
<input type="text"/> height	<input type="text"/> height_2

BMI: bmi

5 Form “UHDRS Motor/Diagnostic Confidence (Motor)”

[motor2]

General [motor] 132_1

Motor score (TMS):

motscore

Motor score (TMS) incomplete:

miscore

Motor Assessment [135] 135_1

Ocular pursuit: [136] 136_1

Horizontal Vertical

- | | | |
|---------------------------------|---------------------------------|-------------------------------------|
| <input type="radio"/> 0 | <input type="radio"/> 0 | 0 = complete (normal) |
| <input type="radio"/> 1 | <input type="radio"/> 1 | 1 = jerky movement |
| <input type="radio"/> 2 | <input type="radio"/> 2 | 2 = interrupted pursuits/full range |
| <input type="radio"/> 3 | <input type="radio"/> 3 | 3 = incomplete range |
| <input type="radio"/> 4 ocularh | <input type="radio"/> 4 ocularv | 4 = cannot pursue |

Saccade initiation: [140] 140_1

Horizontal Vertical

- | | | |
|----------------------------------|----------------------------------|---|
| <input type="radio"/> 0 | <input type="radio"/> 0 | 0 = normal |
| <input type="radio"/> 1 | <input type="radio"/> 1 | 1 = increased latency only |
| <input type="radio"/> 2 | <input type="radio"/> 2 | 2 = suppressible blinks or head movements to initiate |
| <input type="radio"/> 3 | <input type="radio"/> 3 | 3 = unsuppressible head movements |
| <input type="radio"/> 4 sacinith | <input type="radio"/> 4 sacinitv | 4 = cannot initiate saccades |

<u>Saccade velocity:</u> [144] 144_1		<u>Horizontal</u>	<u>Vertical</u>	
<input type="radio"/> 0	<input type="radio"/> 0	0 = normal		
<input type="radio"/> 1	<input type="radio"/> 1	1 = mild slowing		
<input type="radio"/> 2	<input type="radio"/> 2	2 = moderate slowing		
<input type="radio"/> 3	<input type="radio"/> 3	3 = severely slow, full range		
<input type="radio"/> 4 <i>sacvelh</i>	<input type="radio"/> 4 <i>sacvelv</i>	4 = incomplete range		
<u>Dysarthria:</u>	<input type="radio"/> 0 = normal 0			dysarth
	<input type="radio"/> 1 = unclear, no need to repeat 1			
	<input type="radio"/> 2 = must repeat to be understood 2			
	<input type="radio"/> 3 = mostly incomprehensible 3			
	<input type="radio"/> 4 = anarthria 4			
<u>Tongue protrusion:</u>	<input type="radio"/> 0 = can hold tongue fully protruded for 10 sec 0			tongue
	<input type="radio"/> 1 = cannot keep fully protruded for 10 sec 1			
	<input type="radio"/> 2 = cannot keep fully protruded for 5 sec 2			
	<input type="radio"/> 3 = cannot fully protrude tongue 3			
	<input type="radio"/> 4 = cannot protrude tongue beyond lips 4			
<u>Finger taps:</u> [150] 150_1	<u>Right</u>	<u>Left</u>		
<input type="radio"/> 0	<input type="radio"/> 0	0 = normal ($\geq 15/5$ sec.)		
<input type="radio"/> 1	<input type="radio"/> 1	1 = mild slowing, reduction in amplitude (11-14/5 sec.)		
<input type="radio"/> 2	<input type="radio"/> 2	2 = moderately impaired (7-10/5 sec.)		
<input type="radio"/> 3	<input type="radio"/> 3	3 = severely impaired (3-6/5 sec.)		
<input type="radio"/> 4 <i>fingtapr</i>	<input type="radio"/> 4 <i>fingtapl</i>	4 = can barely perform task (0-2/5 sec.)		

<u>Pronate/supinate-hands:</u>	<u>Right</u>	<u>Left</u>		
[154] 154_1	<input type="radio"/> 0	<input type="radio"/> 0	0 = normal	
	<input type="radio"/> 1	<input type="radio"/> 1	1 = mild slowing and/or irregular	
	<input type="radio"/> 2	<input type="radio"/> 2	2 = moderate slowing and irregular	
	<input type="radio"/> 3	<input type="radio"/> 3	3 = severe slowing and irregular	
	<input type="radio"/> 4 prosupr	<input type="radio"/> 4 prosupl	4 = cannot perform	
<u>Luria:</u>	<input type="radio"/> 0 = ≥ 4 in 10 sec, no cue			luria
	<input type="radio"/> 1 = < 4 in 10 sec, no cue			
	<input type="radio"/> 2 = ≥ 4 in 10 sec with cues			
	<input type="radio"/> 3 = < 4 in 10 sec with cues			
	<input type="radio"/> 4 = cannot perform			
<u>Rigidity-arms:</u> [159] 159_1	<u>Right</u>	<u>Left</u>		
	<input type="radio"/> 0	<input type="radio"/> 0	0 = absent	
	<input type="radio"/> 1	<input type="radio"/> 1	1 = slight or present only with activation	
	<input type="radio"/> 2	<input type="radio"/> 2	2 = mild to moderate	
	<input type="radio"/> 3	<input type="radio"/> 3	3 = severe, full range of motion	
	<input type="radio"/> 4 rigarmr	<input type="radio"/> 4 rigarml	4 = severe with limited range	
<u>Bradykinesia-body:</u>	<input type="radio"/> 0 = normal			brady
	<input type="radio"/> 1 = minimally slow (?normal)			
	<input type="radio"/> 2 = mildly but clearly slow			
	<input type="radio"/> 3 = moderately slow, some hesitation			
	<input type="radio"/> 4 = markedly slow, long delays in initiation			

Maximal dystonia: [164] 164_1

Trunk	RUE	LUE	RLE	LLE	
<input type="radio"/> 0	<input type="radio"/> 0	<input type="radio"/> 0	<input type="radio"/> 0	<input type="radio"/> 0	0 = absent
<input type="radio"/> 1	<input type="radio"/> 1	<input type="radio"/> 1	<input type="radio"/> 1	<input type="radio"/> 1	1 = slight/intermittent
<input type="radio"/> 2	<input type="radio"/> 2	<input type="radio"/> 2	<input type="radio"/> 2	<input type="radio"/> 2	2 = mild/common or moderate/intermittent
<input type="radio"/> 3	<input type="radio"/> 3	<input type="radio"/> 3	<input type="radio"/> 3	<input type="radio"/> 3	3 = moderate/common
<input type="radio"/> 4 dysttrnk	<input type="radio"/> 4 dystrue	<input type="radio"/> 4 dystlue	<input type="radio"/> 4 dystrle	<input type="radio"/> 4 dystlle	4 = marked/prolonged

Maximal chorea: [171] 171_1

Face	BOL	Trunk	RUE	LUE	RLE	LLE	
<input type="radio"/> 0	<input type="radio"/> 0	<input type="radio"/> 0	<input type="radio"/> 0	<input type="radio"/> 0	<input type="radio"/> 0	<input type="radio"/> 0	0 = absent
<input type="radio"/> 1	<input type="radio"/> 1	<input type="radio"/> 1	<input type="radio"/> 1	<input type="radio"/> 1	<input type="radio"/> 1	<input type="radio"/> 1	1 = slight/intermittent
<input type="radio"/> 2	<input type="radio"/> 2	<input type="radio"/> 2	<input type="radio"/> 2	<input type="radio"/> 2	<input type="radio"/> 2	<input type="radio"/> 2	2 = mild/common or moderate/intermittent
<input type="radio"/> 3	<input type="radio"/> 3	<input type="radio"/> 3	<input type="radio"/> 3	<input type="radio"/> 3	<input type="radio"/> 3	<input type="radio"/> 3	3 = moderate/common
<input type="radio"/> 4 chorface	<input type="radio"/> 4 chorbol	<input type="radio"/> 4 chortrnk	<input type="radio"/> 4 chorruue	<input type="radio"/> 4 chorlue	<input type="radio"/> 4 chorrlle	<input type="radio"/> 4 chorlle	4 = marked/prolonged

Gait:

- 0 = normal gait, narrow base 0
- 1 = wide base and/or slow 1
- 2 = wide base and walks with difficulty 2
- 3 = walks only with assistance 3
- 4 = cannot attempt 4

gait

<p><u>Tandem walking:</u></p>	<p><input type="radio"/> 0 = normal for 10 steps 0</p> <p><input type="radio"/> 1 = 1 to 3 deviations from straight line 1</p> <p><input type="radio"/> 2 = >3 deviations 2</p> <p><input type="radio"/> 3 = cannot complete 3</p> <p><input type="radio"/> 4 = cannot attempt 4</p>	<p>tandem</p>
<p><u>Retropulsion pull test:</u></p>	<p><input type="radio"/> 0 = normal 0</p> <p><input type="radio"/> 1 = recovers spontaneously 1</p> <p><input type="radio"/> 2 = would fall if not caught 2</p> <p><input type="radio"/> 3 = tends to fall spontaneously 3</p> <p><input type="radio"/> 4 = cannot stand 4</p>	<p>retropls</p>

Diagnostic Confidence [motor3] .DCL 185_1

<p>Diagnostic confidence level (DCL):</p>	<p><input type="radio"/> 0 = normal (no abnormalities) 0</p> <p><input type="radio"/> 1 = non-specific motor abnormalities (less than 50 % confidence) 1</p> <p><input type="radio"/> 2 = motor abnormalities that may be signs of HD (50 - 89 % confidence) 2</p> <p><input type="radio"/> 3 = motor abnormalities that are likely signs of HD (90 - 98 % confidence) 3</p> <p><input type="radio"/> 4 = motor abnormalities that are unequivocal signs of HD (\geq 99 % confidence) 4</p>	<p>diagconf</p>
---	--	-----------------

6 Form “UHDRS Total Funtional Capacity (TFC)”

General [tfc] 893_1		
Functional score:	<input type="text"/>	tfcscore
Functional Capacity [functcap] 895_1		
Occupation:	<input type="radio"/> 0 = unable 0 <input type="radio"/> 1 = marginal work only 1 <input type="radio"/> 2 = reduced capacity for usual job 2 <input type="radio"/> 3 = normal 3	occupatn
Finances:	<input type="radio"/> 0 = unable 0 <input type="radio"/> 1 = major assistance 1 <input type="radio"/> 2 = slight assistance 2 <input type="radio"/> 3 = normal 3	finances
Domestic chores:	<input type="radio"/> 0 = unable 0 <input type="radio"/> 1 = impaired 1 <input type="radio"/> 2 = normal 2	chores
ADL:	<input type="radio"/> 0 = total care 0 <input type="radio"/> 1 = gross tasks only 1 <input type="radio"/> 2 = minimal impairment 2 <input type="radio"/> 3 = normal 3	adl
Care level:	<input type="radio"/> 0 = full time skilled nursing 0 <input type="radio"/> 1 = home or chronic care 1 <input type="radio"/> 2 = home 2	carelevl

7 Form “UHDRS Functional Assessment/Independence Scale (Function)”

General [function] 849_1	
Functional assessment score:	<input type="text"/> fascore
Functional score incomplete:	<input type="text"/> fscore

Independence Scale [indep1] .Independence 983_1	
Subject's independence in %:	indep1
	<input type="radio"/> 100 = no special care needed 100 <input type="radio"/> 95 95 <input type="radio"/> 90 = no physical care needed if difficult tasks are avoided 90 <input type="radio"/> 85 85 <input type="radio"/> 80 = pre-disease level of employment changes or ends; cannot perform household chores to pre-disease level, may need help with finances 80 <input type="radio"/> 75 75 <input type="radio"/> 70 = self-care maintained for bathing, limited household duties, e.g. cooking and use of knives, driving terminates; unable to manage finances 70 <input type="radio"/> 65 65 <input type="radio"/> 60 = needs minor assistance in dressing, toileting, bathing; food must be cut for subject 60 <input type="radio"/> 55 55 <input type="radio"/> 50 = 24-hour supervision appropriate; assistance required for bathing, eating, toileting 50 <input type="radio"/> 45 45 <input type="radio"/> 40 = chronic care facility needed; limited self feeding, liquified diet 40 <input type="radio"/> 35 35 <input type="radio"/> 30 = subject provides minimal assistance in own feeding, bathing, toileting 30 <input type="radio"/> 25 25 <input type="radio"/> 20 = no speech, must be fed 20 <input type="radio"/> 15 15 <input type="radio"/> 10 = tube fed, total bed care 10 <input type="radio"/> 5 5

8 Form “Mini Mental State Examination (MMSE)”




General [mmse] 3070_1	
MMSE score:	<input type="text"/>
	mmsetotal



9 Form “Core and Extended Cognitive Assessment (Cognitive)”

[cogs1]

Specifics [cogn3] .Cognitive Assessment 2288_1		
Did the participant complete the assessment in their native language and with normal or corrected-to-normal vision and hearing?	<input type="radio"/> yes 1 <input type="radio"/> no 0	gen1
Did the participant complete the assessment in their native language?	<input type="radio"/> yes 1 <input type="radio"/> no 0	gen2
At what age did the participant learn the language used?	<input type="text"/> years	gen3
Did the participant have normal/corrected-to-normal hearing and vision?	<input type="radio"/> yes 1 <input type="radio"/> no 0	gen4
Was vision uncorrected (e.g. no glasses during visit)?	<input type="radio"/> yes 1 <input type="radio"/> no 0	gen5
Was hearing uncorrected (e.g. no hearing aid worn)?	<input type="radio"/> yes 1 <input type="radio"/> no 0	gen6

Core Cognitive Assessment [cognitive] 4881_1		
Symbol Digit Modality Test completed: 	<input type="radio"/> yes 1 <input type="radio"/> no 0	sdmt
Total correct:	<input type="text"/>	sdmt1
Total errors:	<input type="text"/>	sdmt2

[verfct1]			
Verbal Fluency Test (Category) completed: 	<input type="radio"/> yes 1 <input type="radio"/> no 0		verfct
Category:	<input type="text" value="----"/> animals 1 other 2		verfctd
Total correct (1 min):	<input type="text"/>		verfct5
Total intrusion errors:	<input type="text"/>		verfct6
Total perseverative errors:	<input type="text"/>		verfct7
[scnt1]			
Stroop Color Naming Test completed: 	<input type="radio"/> yes 1 <input type="radio"/> no 0		scnt
Total correct:	<input type="text"/>		scnt1
Total errors:	<input type="text"/>		scnt2
Total self-corrected errors:	<input type="text"/>		scnt3
[swrt1]			
Stroop Word Reading Test completed: 	<input type="radio"/> yes 1 <input type="radio"/> no 0		swrt
Total correct:	<input type="text"/>		swrt1
Total errors:	<input type="text"/>		swrt2
Total self-corrected errors:	<input type="text"/>		swrt3

[sit1]			
Stroop Interference Test completed: 	<input type="radio"/> yes 1 <input type="radio"/> no 0		sit
Total correct:		<input type="text"/>	sit1
Total errors:		<input type="text"/>	sit2
Total self-corrected errors:		<input type="text"/>	sit3
[verfit1]			
Verbal Fluency Test (Letters) completed: 	<input type="radio"/> yes 1 <input type="radio"/> no 0		verfit
Total correct (3 min):		<input type="text"/>	verfit05
Total intrusion errors:		<input type="text"/>	verfit06
Total perseverative errors:		<input type="text"/>	verfit07

Revision History

Version	Summary of Changes
2016-10-R1	Initial version of annotated CRF for the Ad Hoc dataset.