Summary of Core and Supplemental – Highly Recommended Recommendations: Huntington's Disease CDEs

Start-up Resource –NINDS Huntington's Disease CDE Recommendations

The National Institute of Neurological Disorders and Stroke (NINDS) and other Federal agencies and international organizations have the common mission of developing data standards for clinical research. Through the efforts of subject-specific working groups, topic-driven data elements have been created. The first set of Common Data Elements (CDEs) for Huntington's Disease was developed in 2011. The Core data elements to be used by an investigator when beginning a research study in this disease/disorder are listed in this resource document. All other recommendations are listed on the website and should be considered based on study type.

Each CDE or instrument could be classified according to the definitions below:

General Core: A data element that is required for all NINDS funded studies.

Disease Core: A data element that collects essential information applicable to any disease-specific study, including all therapeutic areas. The NINDS and its appointed working groups assign the disease "Core" classification based on the current clinical research best practices. In each case, the disease Core CDEs are a small subset of the available CDEs, where it is anticipated that investigators will need to collect the disease Core CDEs on any type of study. These are required for all disease-specific studies.

Disease Supplemental - Highly Recommended: A data element which is essential based on certain conditions or study types in clinical research studies. In most cases, these have been used and validated in the disease area. These data elements are strongly recommended for the specified disease condition, study type or design.

Disease Supplemental: A data element which is commonly collected in clinical research studies. Use depends upon the study design, protocol or type of research involved. These are recommended, but not required, for studies.

Disease Exploratory: A data element that requires further validation but may fill current gaps in the CDEs and/or substitute for an existing CDE once validation is complete. Such data elements show great promise but require further validation before they are ready for prime-time use in clinical research studies. They are reasonable to use with the understanding that it has limited validation in the target group.

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National Institute of Health (NIH) Resources: *The NINDS also strongly encourages researchers to use these NIH developed materials for NINDS-sponsored research, when appropriate. Utilization of these resources will enable greater consistency for NINDS-sponsored research studies. These tools are free of charge.*	• **NIH Toolbox** • **Quality of Life in Neurological Disorders (Neuro-QOL)** • **Patient-Reported Outcomes Measurement Information System (PROMIS)**

Core CDEs for all NINDS Studies:

Domain/Sub-Domain	CDE Name	CDE ID	Study Type
Participant Characteristics; Demographics	Birth date	C00007	All studies
Participant Characteristics; Demographics	Ethnicity USA category	C00020	All studies
Participant Characteristics; Demographics	Race USA category	C00030	All studies
Participant Characteristics; Demographics	Birth sex assigned type	C58676	All studies
Participant Characteristics; Demographics	Gender identity type	C58677	All studies
Participant Characteristics; Demographics	Medical history condition text	C00322	All studies
Participant Characteristics; Demographics	Medical history condition SNOMED CT code	C00313	All studies

Supplemental- Highly Recommended CDEs for HD Studies:

CDE Domain; Sub-domain	**CDE Name**	**CDE ID**
Participant Characteristics; Demographics	Education year count	C00015[1]
Participant Characteristics; Demographics	Race/Ethnicity expanded category	C00031

[1] Note: Education year count C00015 is no longer a general Core CDE

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CDE Domain; Sub-domain	CDE Name	CDE ID
Assessments and Examinations; Laboratory Tests and Biospecimens/Biomarkers	Cytosine adenine guanine repeats larger allele number	C14936
Assessments and Examinations; Laboratory Tests and Biospecimens/Biomarkers	Cytosine adenine guanine repeats smaller allele number	C14937
Assessments and Examinations; Laboratory Tests and Biospecimens/Biomarkers	Cytosine adenine guanine repeat test indicator	C14938
Assessments and Examinations; Laboratory Tests and Biospecimens/Biomarkers	Cytosine adenine guanine repeats information source type	C17745
Assessments and Examinations; Laboratory Tests and Biospecimens/Biomarkers	Molecular study lab name	C17746
Assessments and Examinations; Laboratory Tests and Biospecimens/Biomarkers	Cytosine adenine guanine repeats information source other text	C19061
Assessments and Examinations; Laboratory Tests and Biospecimens/Biomarkers	Huntington's disease risk grade	C14939
Assessments and Examinations; Laboratory Tests and Biospecimens/Biomarkers	Cytosine adenine guanine repeat known indicator	C14940
Assessments and Examinations; Laboratory Tests and Biospecimens/Biomarkers	Cytosine adenine guanine repeat results provider type	C17747
Assessments and Examinations; Laboratory Tests and Biospecimens/Biomarkers	Cytosine adenine guanine repeat results provider other text	C19062
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - sit stand scale	C14693
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - unsupport stand scale	C14694
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - unsupport sit scale	C14695
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - stand sit scale	C14696

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CDE Domain; Sub-domain	CDE Name	CDE ID
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - transfer scale	C14697
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - unsupport stand closed eyes scale	C14698
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - unsupport feet together scale	C14699
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - reach forward scale	C14700
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - retrieve floor object scale	C14701
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - turn head scale	C14702
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - turn around scale	C14703
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - alternate foot step up scale	C14704
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - unsupport stand front foot scale	C14705
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - stand one leg scale	C17394
Outcomes and End Points; Motor Function	Berg Balance Scale (BBS) - scale total score	C14706

General Core for all Studies:

Investigators should review the FDA's "Guidance for Industry: Suicidal Ideation and Behavior: Prospective Assessment of Occurrence in Clinical Trials" for the most up-to-date information about suicidal ideation and behavior. One scale that FDA suggests is the Columbia Suicide Severity Rating Scale (C-SSRS) (available at Columbia Suicide Severity Rating Scale Website).

Core HD Instruments: These instruments and elements are recommended for use in all HD studies:

1. UHDRS (Unified Huntington's Disease Rating Scale) Motor Function, Total Functional Capacity, Functional Assessment Checklist, and Independence Scale

Supplemental – Highly Recommended Cognitive Instruments:

1. Montreal Cognitive Assessment (MoCA)
2. Symbol Digit Modality Test
3. Trail Making Test

Supplemental – Highly Recommended Quality of Life Patient Reported Outcome Instruments:

1. HDQLIFE
2. Quality of Life in Neurological Disorders (Neuro-QOL)

Supplemental – Highly Recommended Emotional/Behavioral Instruments:

1. Hospital Anxiety and Depression Scale (HADS)

Supplemental – Highly Recommended Motor Function Instruments:

1. Berg Balance Scale (BBS)

For the complete list of NINDS CDE recommendations for HD, please see the NINDS CDE website.