

Amyotrophic Lateral Sclerosis (ALS) CDE Highlight Summary

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.

- National Institutes of Health (NIH) Toolbox
- Quality of Life in Neurological Disorders (Neuro-QOL)
- Patient-Reported Outcomes Measurement Information System (PROMIS)

Suicidal Ideation

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](#)).

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Disease; Domain	Recommendations and List of Forms
ALS	<p>These instruments and elements are recommended for use in all Amyotrophic Lateral Sclerosis Studies:</p> <p>Core:</p> <ol style="list-style-type: none"> 1. Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised (ALSRFS-R) / ALS Functional Rating Scale-Revised-Self-Explanatory (ALSRFS-R-SE) 2. Manual Muscle Testing or Quantitative Dynamometry (Hand Held Dynamometry and Fixed Dynamometry are available on the website) <p>Supplemental – Highly Recommended instruments:</p> <ol style="list-style-type: none"> 3. One of the following three Supplemental - Highly Recommended cognitive/behavioral screening tools should be used based on study requirements: <ul style="list-style-type: none"> o Abrahams Written Verbal Fluency o ALS Cognitive Behavioral Screen (ALS-CBS) o Edinburgh Cognitive and Behavioral Scale (ECAS) 4. Rasch-Built Overall Amyotrophic Lateral Sclerosis Disability Scale (ROADS) <p>CRF Modules Containing Core CDEs:</p> <p>Demographics; Social Status; Family History; Medical History; Genetics; Symptom, Sign, Diagnosis Criteria; Vital Signs; Physical Examination; Pulmonary Function; Myocardial Oxygen Consumption, Gas Exchange and Nocturnal Oximetry</p> <p>Core Elements: See Start-Up Resource Listing document (ALS Start-Up Resource Listing)</p>
Participant/Subject Characteristics; Demographics:	<p>Core: Demographics*</p> <p>Supplemental: Demographics*</p>
Participant/Subject Characteristics; Social Status:	<p>Core: Social Status*</p> <p>Supplemental – Highly Recommended: N/A</p> <p>Supplemental: Social Status*</p> <p>Exploratory: N/A</p>

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Disease; Domain	Recommendations and List of Forms
Participant/Subject History and Family History; General Health History:	Core: Family History*, Medical History* Supplemental – Highly Recommended: N/A Supplemental: Family History*, Medical History* Exploratory: N/A
Disease/Injury Related Events; Classification	Core: Genetics*, Symptom, Sign, Diagnosis Criteria* Supplemental – Highly Recommended: N/A Supplemental: Genetics*, Symptom, Sign, Diagnosis Criteria* Exploratory: N/A
Assessments and Examinations; Physical/Neurological Examination:	Core: Physical Examination* Supplemental – Highly Recommended: N/A Supplemental: Neuromuscular Examination, Physical Examination* Exploratory: N/A
Assessments and Examinations; Vital Signs and Other Body Measures:	Core: Vital Signs* Supplemental – Highly Recommended: N/A Supplemental: Vital Signs* Exploratory: N/A
Assessments and Examinations; Laboratory Tests and Biospecimens/ Biomarkers:	Core: N/A Supplemental – Highly Recommended: N/A Supplemental: Biomarkers Other Collection Information Guidelines, Collection of Fluid: Blood Guidelines, Collection of Fluid: CSF Guidelines, Collection of Fluid: Plasma Guidelines, Collection of Fluid: Urine Guidelines, Fasting Information, Laboratory Tests, Exploratory: Fasting Information, Resting Energy Expenditure, Tissue Based Marker Collection, Tissue Based Marker Collection

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Assessments and Examinations; Imaging Diagnostics:	<p>Core: N/A</p> <p>Supplemental – Highly Recommended: N/A</p> <p>Supplemental: Brain Perfusion Magnetic Resonance Imaging, Diffusion Tensor Imaging (DTI), Functional Magnetic Resonance Imaging (fMRI), Magnetic Resonance Imaging (MRI), Magnetic Resonance Spectroscopy</p> <p>Exploratory: Brain Perfusion Magnetic Resonance Imaging, Diffusion Tensor Imaging (DTI), Functional Magnetic Resonance Imaging (fMRI), Magnetic Resonance Imaging (MRI), Magnetic Resonance Spectroscopy</p>
Assessments and Examinations; Non-Imaging Diagnostics:	<p>Core: N/A</p> <p>Supplemental – Highly Recommended: N/A</p> <p>Supplemental: Electrophysiology</p> <p>Exploratory: N/A</p>
Treatment/Intervention Data; Drugs:	<p>Core: N/A</p> <p>Supplemental – Highly Recommended: N/A</p> <p>Supplemental: Prior and Concomitant Medications</p> <p>Exploratory: N/A</p>

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<p>Outcomes and End Points; Cognitive:</p>	<p>Core: N/A</p> <p>Supplemental – Highly Recommended: Abrahams Written Verbal Fluency*, Edinburgh Cognitive and Behavioral Scale (ECAS)*, ALS Cognitive Behavioral Screen (ALS-CBS)*.</p> <p>*One of the following three Supplemental – Highly Recommended cognitive/behavioral screening tools should be used based on study requirements.</p> <p>Supplemental: ALS Depression Inventory (ADI-12), ALS-Frontotemporal Dementia Questionnaire (ALS-FTD-Q), Beck Depression Inventory II, Cambridge Behavioral Inventory- Revised, Center for Neurologic Study-Lability Scale for pseudobulbar affect (PBA), Emotional Lability Questionnaire: Persons with MND (ELQ-MND), Emotional Lability Questionnaire: Relatives and Carers (ELQ-R and C), Frontal Behavior Inventory (FBI), Frontal Behavioral Inventory-ALS Version (FBI-ALS), Frontal Behavioral Inventory-Modified by Heidler Gray, Frontal Systems Behavior Scale, Hospital Anxiety and Depression Scale, Motor Neuron Disease Behavioral Instrument (MiND-B), Neuropsychiatric Inventory – Questionnaire (NPI-Q), Penn State Screen of Frontal and Temporal Dysfunction Syndromes, and UCSF Screening Exam.</p> <p>Exploratory: Geriatric Depression Scale (GDS), Hamilton Depression Rating Scale (HDRS), and Neuropsychiatric Inventory – Clinician Version</p>
<p>Outcomes and End Points; Functional Outcomes</p>	<p>Core: ALS Functional Rating Scale – Revised (ALSFRRS-R)/ ALS Functional Rating Scale – Revised – Self-Explanatory (ALSFRRS-R-SE)</p> <p>Supplemental – Highly Recommended: Rasch-Built Overall ALS Disability Scale (ROADS)</p> <p>Supplemental: 6 Minute Walk Test, ALS Swallowing Severity Score – (ALSSS), Barthel Index, Center for Neurologic Study-Bulbar Function Scale (CNS-BFS), Clinical Milestones/Events, Death Report, Dynamic Gait Index, Edmonton Symptom Assessment System (ESAS), EuroQoL-5 Dimension Questionnaire, Modified Fatigue Impact Scale (MFIS), Performance-Oriented Mobility Assessment (POMA), Penn Upper Motor Neuron Score (PUMNS), Schwab and England Activities of Daily Living Scale (ADL-Scale), Sensory Organization Test (SOT), Timed 25-Foot Walk (T25FW), Timed Up and Go</p> <p>Exploratory: Checklist for Individual Strength – Fatigue (CIS), Clinical Milestones/Events , Fatigue Severity Scale (FSS), Madrid Quantitative Neuromuscular Assessment (MAQUINA), and Modified Fatigue Impact Scale (MFIS)</p>

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Disease; Domain	Recommendations and List of Forms
Outcomes and End Points; Summary of Outcome Recommendations	<p>Core: N/A</p> <p>Supplemental – Highly Recommended: N/A</p> <p>Supplemental: ALS CDE Instrument Recommendations by Outcome Domain and Classification, ALS Outcomes Recommendations Guidelines for Use</p> <p>Exploratory: N/A</p>
Outcomes and End Points; Muscle Strength Testing:	<p>Core: Manual Muscle Testing – Using the Medical Research Council Muscle Grading Scale</p> <p>Supplemental – Highly Recommended: N/A</p> <p>Supplemental: Pinch Strength, Fixed Dynamometry, Grip Strength Fatigue, Hand Held Dynamometry</p> <p>Exploratory: Grip Strength</p>
Outcomes and End Points; Pulmonary Function Testing / Respiratory Status:	<p>Core: Myocardial Oxygen Consumption, Gas Exchange and Nocturnal Oximetry*, Pulmonary Function*</p> <p>Supplemental – Highly Recommended: N/A</p> <p>Supplemental: Borg Dyspnea Scale, Myocardial Oxygen Consumption, Gas Exchange and Nocturnal Oximetry*, Pulmonary Function*</p> <p>Exploratory: Baseline Dyspnea, Myocardial Oxygen Consumption, Gas Exchange and Nocturnal Oximetry*, and Transition Dyspnea Index</p>
Outcomes and End Points; Quality of Life:	<p>Core: N/A</p> <p>Supplemental – Highly Recommended: N/A</p> <p>Supplemental, Amyotrophic Lateral Sclerosis Assessment Questionnaire (ALS-AQ40), EuroQoL-VAS, McGill Quality of Life Questionnaire (MQOL), Quality of Life in Neurological Disorders (Neuro-QOL, Quality of Life in Swallowing Disorders (SWAL-QOL), Short Form 36-Item Health Survey (SF-36)</p> <p>Exploratory: Schedule for the Evaluation of Individual Quality of Life – Direct Weighting (SEIQoL-DW) and Sickness Impact Profile)</p>
Outcomes and End Points; Subjective Assessments / Patient and Caregiver Reported Outcomes	<p>Core: N/A</p> <p>Supplemental – Highly Recommended: N/A</p> <p>Supplemental: Zarit Caregiver Burden Scale</p> <p>Exploratory: Caregiver Burden Scale (CGBS), Clinical Global Impression Scale,</p>

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Outcomes and End Points; Upper Motor Neuron Signs / Neuromuscular Excitability	<p>Core: N/A</p> <p>Supplemental – Highly Recommended: N/A</p> <p>Supplemental: Modified Ashworth Scale for Grading Spasticity, NINDS Myotatic Reflex Scale, Tardieu Scale</p> <p>Exploratory: N/A</p>

*Include one or more data elements that are Core or Supplemental – Highly Recommended