

## Mitochondrial Disease Version 2.0 NINDS CDE Project Cognitive/Psychological Outcomes Subgroup Summary

The NINDS Mitochondrial Disease v2.0 Common Data Element (CDE) Cognitive/Psychological Outcomes Subgroup made recommendations based on validated tools that measure cognitive and psychological functions. More specifically, we describe cognition as neurological processes including attention, executive functioning, language, memory, visuospatial processing, motor/psychomotor skills, and intellectual abilities. Related functional and psychological areas to be measured include academic achievement, adaptive characteristics, and mood (e.g., depression and anxiety). Neuropsychological and psychological assessment tools were evaluated, and include question/answer, pencil/paper, and/or computerized methods.

Instruments that can be used across the life span were considered, as mitochondrial disease impacts both pediatric and adult populations. There are a broad range of clinical phenotypes as well as phenotypic heterogeneity with variable cognitive, motor, and psychiatric presentations.

### Summary of Recommendations

Subdomain	Instrument Name	Classification
Academic Achievement	American National Adult Reading Test (AMNART)	Supplemental
	Differential Ability Scales-II (DAS-II)	Supplemental
	Wechsler Individual Achievement Test-4 (WIAT-4)	Supplemental
Attention	Conners Continuous Performance Test 3rd Edition (Conners CPT 3)	Supplemental – Highly Recommended
	Test of Variables of Attention (TOVA)	Supplemental – Highly Recommended
Cognitive	Automated Neuropsychological Assessment Metrics (ANAM)	Supplemental
	Brief Test of Adult Cognition by Telephone (BTRACT)	Supplemental
	Cambridge Cognitive Examination - Revised (CAMCOG-R)	Supplemental
	Mini-Mental State Examination (MMSE)	Supplemental
	NEPSY-II	Supplemental
	Repeatable Battery for Assessment of Neuropsychological Status (RBANS)	Supplemental
	Symbol Digit Modalities Test	Supplemental
Dementia	Informant Questionnaire on Cognitive Decline in the Elderly (IQCODE)	Supplemental
	Montreal Cognitive Assessment (MoCA)	Supplemental
Emotional/Behavioral	ALS Depression Inventory (ADI-12)	Supplemental
	Apathy Evaluation Scale	Supplemental – Highly Recommended
	Apathy Scale (AS)	Supplemental – Highly Recommended
	Ask Suicide-Screening Questions (ASQ) Toolkit	Supplemental
	Autism Diagnostic Interview - Revised (ADI-R)	Supplemental
	Autism Diagnostic Observation Schedule, Second Edition (ADOS-II)	Supplemental

Subdomain	Instrument Name	Classification
Emotional/Behavioral	Beck Anxiety Inventory (BAI)	Supplemental – Highly Recommended
	Beck Youth Inventories - 2 <sup>nd</sup> Edition (BYI-II)	Supplemental – Highly Recommended
	Behavior Assessment System for Children, Third Edition (BASC-3)	Supplemental – Highly Recommended
	Brief Infant Toddler Social Emotional Assessment (BITSEA)	Supplemental
	Brief Psychiatric Rating Scale (BPRS)	Supplemental
	Cambridge Behavioral Inventory - Revised	Supplemental
	Center for Epidemiologic Studies - Depression Scale (CES-D)	Supplemental – Highly Recommended
	Child Behavior Checklist (CBCL)	Supplemental – Highly Recommended
	Children's Depression Inventory - 2 (CDI-2)	Supplemental
	Columbia - Suicide Severity Rating Scale (C-SSRS)	Supplemental
	Generalized Anxiety Disorder (GAD-7)	Supplemental – Highly Recommended
	Hamilton Anxiety Rating Scale (HAM-A)	Supplemental
	Hamilton Depression Rating Scale (HDRS)	Supplemental
	Hospital Anxiety and Depression Scale (HADS)	Supplemental
	Irritability Scale (IS)	Supplemental
	Modified Overt Aggression Scale (MOAS)	Supplemental
	Montgomery-Asberg Depression Scale (MADRS)	Supplemental
	Mood Disorder Questionnaire (MDQ)	Supplemental
Problem Behaviours Assessment HD - Short Version (PBA-s)	Supplemental	
Executive Functioning	Behavior Rating Inventory of Executive Function - Adult Version (BRIEF-A)	Supplemental – Highly Recommended
	Behavior Rating Inventory of Executive Function - Preschool Version (BRIEF-P)	Supplemental – Highly Recommended
	Behavior Rating Inventory of Executive Function - Second Edition (BRIEF-2)	Supplemental – Highly Recommended
	Delis-Kaplan Executive Function System (D-KEFS)	Supplemental – Highly Recommended
	Tower of London-Drexel University (TOLDX) - 2 <sup>nd</sup> Edition	Supplemental
	Trail Making Test (A, B)	Supplemental
Fatigue	Fatigue Severity Scale (FSS)	Supplemental – Highly Recommended
Intellectual Functioning	Kaufman Assessment Battery for Children-II	Supplemental
	Leiter International Performance Scale-3 (Leiter-3)	Supplemental
	Reynolds Intellectual Assessment Scale, Second Edition (RIAS-2)	Supplemental
	Wechsler Abbreviated Scale of Intelligence - Second Edition (WASI-II)	Supplemental
	Wechsler Adult Intelligence Scale - Fourth Edition (WAIS-IV)	Supplemental

Subdomain	Instrument Name	Classification
Intellectual Functioning	Wechsler Intelligence Scale for Children-V (WISC-V)	Supplemental
	Wechsler Preschool and Primary Scale of Intelligence (WPPSI-IV)	Supplemental – Highly Recommended
Language	Boston Diagnostic Aphasia Exam (BDAE-III)	Supplemental
	Boston Naming Test (BNT) - 30-Item Version	Supplemental
	Clinical Evaluation of Language Fundamentals - Fifth Edition (CELF-5)	Supplemental
	Comprehensive Assessment of Spoken Language, Second Edition (CASL-2)	Supplemental
	Controlled Oral Word Association Test (COWAT)	Supplemental
	Expressive One-Word Picture Vocabulary Test	Supplemental
	New Reynell Developmental Language Scale (NRDLS)	Supplemental
	Peabody Picture Vocabulary Test 5 <sup>th</sup> Edition (PPVT-5)	Supplemental
	Preschool Language Scales - Fifth Edition (PLS-5)	Supplemental
Memory	Brief Visuospatial Memory Test - Revised (BVMT-R)	Supplemental
	Child and Adolescent Memory Profile (ChAMP)	Supplemental
	Rey Auditory Verbal Learning Test (RAVLT)	Supplemental
	Rey Complex Figure Test and Recognition Trial (RCFT)	Supplemental
	Wechsler Memory Scale IV (WMS-IV)	Supplemental
	Wide Range Assessment of Memory and Learning, Third Edition (WRAML-3)	Supplemental
Motor Function	Beery-Buktenica Developmental Test of Visual-Motor Integration (Beery VMI)	Supplemental
	Grooved Pegboard Test	Supplemental
	Newcastle Mitochondrial Disease Adult Scale (NMDAS)	Supplemental – Highly Recommended
	Physical and Neurological Examination for Subtle Signs (PANESS)	Exploratory
	Purdue Pegboard	Supplemental
NIH Resources	National Institutes of Health (NIH) Toolbox	Supplemental
	Patient-Reported Outcomes Measurement Information System (PROMIS)	Supplemental
	Quality of Life in Neurological Disorders (Neuro-QOL)	Supplemental – Highly Recommended
Quality of Life/Patient Reported Outcomes	Child and Adolescent Scale of Environment (CASE)	Supplemental
	Child and Adolescent Scale of Participation (CASP)	Supplemental
	Newcastle Mitochondrial Quality of Life Measure (NMQ)	Supplemental – Highly Recommended
	Pediatric Quality of Life Inventory (PedsQL)	Supplemental – Highly Recommended
Sleep	Epworth Sleepiness Scale (ESS) - Adult Version	Supplemental
Visual-Spatial Processing	Judgment of Line Orientation (Benton JLO)	Supplemental
	Test of Visual Perceptual Skills - 4 <sup>th</sup> Edition (TVPS-4)	Supplemental