European Reference Networks

European Reference Network for rare or low prevalence complex diseases

Network: Neurological Diseases (ERN-RND)

Coordinator: Universitätsklinikum Tübingen — Deutschland

SCALE TO MEASURE FRONTOTEMPORAL DEMENTIA

CDR® Dementia Staging Instrument

PLUS NACC FTLD Behavior & Language Domains

Endorsed by ERN-RND: 16th June 2020

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INTRODUCTION TO THE EUROPEAN REFERENCE NETWORK FOR RARE NEUROLOGICAL DISEASES (ERN-RND)

ERN-RND is a European Reference Network established and approved by the European Union. ERN-RND is a healthcare infrastructure which focuses on rare neurological diseases (RND). The three main pillars of ERN-RND are (i) network of experts and expertise centres, (ii) generation, pooling and dissemination of RND knowledge, and (iii) implementation of e-health to allow the expertise to travel instead of patients and families.
ERN-RND unites 32 of Europe's leading expert centres in 13 Member States and includes highly active patient organizations. Centres are located in Belgium, Bulgaria, Czech Republic, France, Germany, Hungary, Italy, Lithuania, Netherlands, Poland, Slovenia, Spain and the UK.

The following disease groups are covered by ERN-RND:

- Ataxias and Hereditary Spastic Paraplegias
- Atypical Parkinsonism and genetic Parkinsons' Disease
- Dystonia, Paroxysmal Disorder and Neurodegeneration with Brain Ion Accumulation
- Frontotemporal Dementia
- Huntingtons' Disease and other Choreas
- Leukodystrophies

Specific information about the network, the expert centres and the diseases covered can be found at the networks web site www.ern-rnd.eu.

Recommendation for clinical use:
The European Reference Network for Rare Neurological Diseases strongly recommends the use the following scale as best clinical practice for the assessment and rating of Frontotemporal Dementia.

DISCLAIMER

METHODOLOGY

The development of the Diagnostic Flowcharts for Dystonia was done by the Disease group for Frontotemporal Dementia. Scales used in the clinical practice of the disease group members were mapped, and the decision on which scale should be proposed was taken by majority voting.

Diesease group for Frontoemporal Dementia

Disease group coordinators:

Isabelle Leber[1]; Markus Otto[11];Rik Vandenberghe[3]

Disease group members:

Healthcare professionals:

Alberto Albanese[4]; Adrian Danek[5]; Maria Teresa Dotti[6]; Barbara Garavaglia[7]; Zoltan Grosz[8]; Norbert Kovacs[9]; Milica Kramberger[10]; Bernhard Landwehrmeier[11]; Johannes Levin[5]; Janne Papma[12]; Jonathan Rohrer[2]; Robert Rusina[13]; Harro Seelaar[12]; Matthis Synofzik[14]; Marc Teichmann[1], Pietro Tiraboschi[7]; John van Swieten[12]; Ione Wollacott[2]

Patient representatives:

Mary Kearney

[1] Assistance Publique-Hôpitaux de Paris, Hôpital Pitié-Salepétrière, France: Reference centre for rare dementias; [2] University College London Hospitals NHS Foundation Trust, United Kingdom; [3] University Hospitals Leuven, Belgium; [4] IRCCS Clinical Institute Humanitas – Rozzano, Italy; [5] Klinikum der Universität München, Germany; [6] AOU Siena, Italy; [7] Foundation IRCCS neurological institute Carlo Besta – Milan, Italy; [8] Semmelweis University, Hungary; [9] University of Pécs, Hungary; [10] University Medical Centre Ljubljana, Slovenia; [11] Universitätsklinikum Ulm, Germany; [12] Erasmus MC: University Medical Center Rotterdam, Netherlands; [13] Charles University, Prague, [14] Universitätsklinikum Tübingen, Germany

SCALE

INITIAL VISIT PACKET NACC UNIFORM DATA SET (UDS)

Form B4: CDR® Dementia Staging Instrument
PLUS NACC FTLD Behavior & Language Domains (CDR® Plus NACC FTLD)

ADC name: _____ Subject ID: _____ Form date: __ __ / __ __ / __ __ __ __ Visit #: __ __ __ Examiner's initials: __ __ __

INSTRUCTIONS: For information on the required online CDR training, see UDS Coding Guidebook for Initial Visit Packet, Form B4. This form is to be completed by the clinician or other trained health professional, based on co-participant report and behavioral and neurological exam of the subject. In the extremely rare instances when no co-participant is available, the clinician or other trained health professional must complete this form using all other available information and his/her best clinical judgment. Score only as decline from previous level due to cognitive loss, not impairment due to other factors, such as physical disability. For further information, see UDS Coding Guidebook for Initial Visit Packet, Form B4.

SECTION 1: CDR® DEMENTIA STAGING INSTRUMENT[1]

Please enter score below:	IMPAIRMENT				
	None — 0	**Questionable — 0.5**	**Mild — 1**	**Moderate — 2**	**Severe — 3**
1. Memory __.__	No memory loss, or slight inconsistent forgetfulness	Consistent slight forgetfulness; partial recollection of events; "benign" forgetfulness	Moderate memory loss, more marked for recent events; defect interferes with everyday activities	Severe memory loss; only highly learned material retained; new material rapidly lost	Severe memory loss; only fragments remain
2. Orientation __.__	Fully oriented	Fully oriented except for slight difficulty with time relationships	Moderate difficulty with time relationships; oriented for place at examination; may have geographic disorientation elsewhere	Severe difficulty with time relationships; usually disoriented to time, often to place	Oriented to person only
3. Judgment and problem solving __.__	Solves everyday problems, handles business and financial affairs well; judgment good in relation to past performance	Slight impairment in solving problems, similarities, and differences	Moderate difficulty in handling problems, similarities, and differences; social judgment usually maintained	Severely impaired in handling problems, similarities, and differences; social judgment usually impaired	Unable to make judgments or solve problems
4. Community affairs __.__	Independent function at usual level in job, shopping, volunteer and social groups	Slight impairment in these activities	Unable to function independently at these activities, although may still be engaged in some; appears normal to casual inspection	No pretense of independent function outside the home; appears well enough to be taken to functions outside the family home	No pretense of independent function outside the home; appears too ill to be taken to functions outside the family home
5. Home and hobbies __.__	Life at home, hobbies, and intellectual interests well maintained	Life at home, hobbies, and intellectual interests slightly impaired	Mild but definite impairment of function at home; more difficult chores abandoned; more complicated hobbies and interests abandoned	Only simple chores preserved; very restricted interests, poorly maintained	No significant function in the home
6. Personal care __.0	Fully capable of self-care (=0).		Needs prompting	Requires assistance in dressing, hygiene, keeping of personal effects	Requires much help with personal care; frequent incontinence
7. __ __.__	**CDR SUM OF BOXES**				
8. __.__	**GLOBAL CDR**				

[1]Morris JC. The Clinical Dementia Rating (CDR): Current version and scoring rules. Neurology 43(11):2412-4, 1993. Copyright© Lippincott, Williams & Wilkins. Reproduced by permission.

Subject ID: ⎵⎵⎵⎵⎵⎵⎵⎵⎵⎵ Form date: ⎵⎵ / ⎵⎵ / ⎵⎵⎵⎵ Visit #: ⎵⎵⎵

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SECTION 2: NACC FTLD BEHAVIOR & LANGUAGE DOMAINS

Please enter score below:	IMPAIRMENT				
	None — 0	**Questionable — 0.5**	**Mild — 1**	**Moderate — 2**	**Severe — 3**
9. Behavior, comportment, and personality[2] ⎵.⎵	Socially appropriate behavior	Questionable changes in comportment, empathy, appropriateness of actions	Mild but definite changes in behavior	Moderate behavioral changes, affecting interpersonal relationships and interactions in a significant manner	Severe behavioral changes, making interpersonal interactions all unidirectional
10. Language[3] ⎵.⎵	No language difficulty, or occasional mild tip-of-the-tongue	Consistent mild word-finding difficulties; simplification of word choice; circumlocution; decreased phrase length; and/or mild comprehension difficulties	Moderate word-finding difficulty in speech; cannot name objects in environment; reduced phrase length and/or agrammatical speech and/or reduced comprehension in conversation and reading	Moderate to severe impairments in either speech or comprehension; has difficulty communicating thoughts; writing may be slightly more effective	Severe comprehension deficits; no intelligible speech

[2]Excerpted from the Frontotemporal Demential Multicenter Instrument & MR Study (Mayo Clinic, UCSF, UCLA, UW).

[3]Excerpted from the PPA-CDR: A modification of the CDR for assessing dementia severity in patients with primary progressive aphasia (Johnson N, Weintraub S, Mesulam MM), 2002.

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https://ec.europa.eu/health/ern_e

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