

ESCALA PARA MEDIR LA DEMENCIA FRONTOTEMPORAL

CDR® Dementia Staging Instrument
PLUS NACC FTLD Behavior & Language Domains

EUROPEAN REFERENCE NETWORKS
FOR RARE, LOW PREVALENCE AND COMPLEX DISEASES

Share. Care. Cure.



Disclaimer:

“The European Commission support for the production of this publication does not constitute endorsement of the contents which reflects the views only of the authors, and the Commission cannot be held responsible for any use which may be made of the information contained therein.”

More information on the European Union is available on the Internet (<http://europa.eu>).

Luxembourg: Publications Office of the European Union, 2019

© European Union, 2019

Reproduction is authorised provided the source is acknowledged.

RED EUROPEA DE REFERENCIA PARA ENFERMEDADES NEUROLÓGICAS RARAS (ERN-RND)

La ERN-RND es una red de referencia europea establecida y aprobada por la Unión Europea. La ERN-RND es una infraestructura de atención médica que se enfoca hacia las enfermedades neurológicas raras. Los tres pilares principales de la ERN-RND son (i) red de expertos y centros expertos, (ii) generación, agrupación y difusión de conocimiento sobre enfermedades neurológicas raras, y (iii) implementación de e-salud para permitir que viaje el conocimiento en lugar de los pacientes y sus familias.

La ERN-RND une a 32 de los principales centros de expertos de Europa en 13 Estados miembros e incluye organizaciones de pacientes muy activas. Los centros están ubicados en Bélgica, Bulgaria, República Checa, Francia, Alemania, Hungría, Italia, Lituania, Países Bajos, Polonia, Eslovenia, España y el Reino Unido.

Los siguientes grupos de enfermedades están cubiertos por la ERN-RND:

- Ataxias y paroplejías espásticas hereditarias
- Parkinsonismo atípico y enfermedad de Parkinson genética
- Distonía, trastornos paroxísticos y neurodegeneración con acúmulo cerebral de hierro
- Demencia frontotemporal
- Enfermedad de Huntington y otras Coreas
- Leucodistrofias

Puede encontrar información específica sobre la red, los centros expertos y las enfermedades cubiertas en el sitio web de la red: www.ern-rnd.eu.

Recomendación de uso clínico:

Red Europea de Referencia de Enfermedades Neurológicas Raras recomienda el uso de la siguiente escala como mejor práctica clínica para la evaluación y calificación de la Demencia Frontotemporal.

EXENCIÓN DE RESPONSABILIDAD

Las pautas clínicas, buenas prácticas, revisiones sistemáticas y demás indicaciones orientativas publicadas, avaladas o confirmadas por la ERN-RND son evaluaciones de la información científica y clínica actual que se ofrece como material educativo.

La información 1) no debe considerarse como inclusiva de todos los tratamientos y terapias apropiados ni como afirmación de un determinado estándar de cuidados; 2) no se actualiza constantemente y es posible que tampoco refleje el estado actual de conocimientos (desde el momento de la redacción de esta información y su publicación y lectura puede haber surgido nueva información); 3) se refiere solo a las cuestiones expresamente indicadas; 4) no prescriben ninguna medida sanitaria específica; 5) no sustituyen el criterio independiente y profesional del médico, ya que dichas informaciones no tienen en cuenta las diferencias individuales entre los pacientes. En todos los casos, el médico debe adaptar el modo de proceder elegido a cada paciente en su contexto individual. El uso de las informaciones es voluntario. La ERN-RND proporciona la información partiendo de la situación actual y no ofrece ninguna garantía expresa ni tácita respecto de ellas. La ERN-RND rechaza expresamente cualquier responsabilidad en relación con la comerciabilidad o adecuación para un uso o propósito específicos. La ERN-RND no se responsabiliza de ningún daño personal ni material resultante del empleo de esta información o en relación con ella ni de ningún error u omisión.

METODOS

El desarrollo de los Diagnostic Flowcharts para la Disonía fue realizado por el grupo de enfermedades para la Demencia Frontotemporal. Se trazaron escalas utilizadas en la práctica clínica de los miembros del grupo de enfermedades y la decisión sobre qué escala debía proponerse se tomó por mayoría.

Grupo de enfermedades para FTD:

Coordinadores de grupo de enfermedades:

Isabelle Leber¹; Markus Otto¹¹; Rik Vandenberghe³

Miembros del grupo de enfermedades:

Profesionales de la salud:

Alberto Albanese⁴; Adrian Danek⁵; Maria Teresa Dotti⁶; Barbara Garavaglia⁷; Zoltan Grosz⁸; Norbert Kovacs⁹; Milica Kramberger¹⁰; Bernhard Landwehrmeier¹¹; Johannes Levin⁵; Janne Papma¹²; Jonathan Rohrer²; Robert Rusina¹³; Harro Seelaar¹²; Matthis Synofzik¹⁴; Marc Teichmann¹; Pietro Tiraboschi⁷; John van Swieten¹²; Ione Wollacott²

Representantes de los pacientes:

Mary Kearney

¹ Assistance Publique-Hôpitaux de Paris, Hôpital Pitié-Salpêtrière, 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 INSTRUMENT¹

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				

¹Morris 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 #: _____

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 2: NACC FTLD BEHAVIOR & LANGUAGE DOMAINS

Please enter score below:

	IMPAIRMENT				
	None — 0	Questionable — 0.5	Mild — 1	Moderate — 2	Severe — 3
9. Behavior, compartment, and personality² <small>_____ - _____</small>	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. Language³ <small>_____ - _____</small>	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

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

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



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



European Reference Network
for rare or low prevalence complex diseases

Network
Neurological Diseases
(ERN-RND)

Coordinator
Universitätsklinikum
Tübingen – Deutschland

www.ern-rnd.eu

Co-funded by the European Union

