European Reference Networks

European Reference Network for rare or low prevalence complex diseases

Network: Neurological Diseases (ERN-RND)

Coordinator: Universitätsklinikum Tübingen — Deutschland

SCALA DI MISURAZIONE DELLA DEMENZA FRONTOTEMPORALE

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

EUROPEAN REFERENCE NETWORKS
FOR RARE, LOW PREVALENCE AND COMPLEX DISEASES

Share. Care. Cure.

Approvato da ERN-RND: 16 giugno 2020

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Luxembourg: Publications Office of the European Union, 2019

LA RETE EUROPEA DI RIFERIMENTO PER LE MALATTIE NEUROLOGICHE RARE (ERN-RND)

ERN-RND è una rete di riferimento europea istituita e approvata dall'Unione Europea. L'ERN-RND è un'infrastruttura sanitaria incentrata sulle malattie neurologiche rare (RND). I tre pilastri principali dell'ERN-RND sono (i) la rete di esperti e centri di competenza, (ii) la generazione, la condivisione e la diffusione delle conoscenze sulle RND e (iii) l'implementazione dell'ehealth per consentire alle competenze di viaggiare al posto dei pazienti e delle famiglie.

L'ERN-RND riunisce 32 dei principali centri europei di esperti in 13 Stati membri e comprende organizzazioni di pazienti molto attive. I centri si trovano in Belgio, Bulgaria, Repubblica Ceca, Francia, Germania, Ungheria, Italia, Lituania, Paesi Bassi, Polonia, Slovenia, Spagna e Regno Unito.

I seguenti gruppi di malattie sono coperti da ERN-RND:

- Atassie e paraplegie spastiche ereditarie
- Parkinsonismo atipico e malattia di Parkinson genetica
- Distonia, disturbo parossistico e neurodegenerazione con accumulo di ioni cerebrali
- Demenza frontotemporale
- Malattia di Huntingtons e altre cose da fare
- Leucodistrofie

Informazioni specifiche sulla rete, sui centri esperti e sulle malattie trattate sono disponibili sul sito web della rete ***www.ern-rnd.eu.***

Raccomandazione per l'uso clinico:
La Rete Europea di Riferimento per le Malattie Neurologiche Rare è fortemente impegnata
Raccomanda l'uso della seguente scala come migliore pratica clinica per il
valutazione e classificazione della demenza frontotemporale.

DICHIARAZIONE DI NON RESPONSABILITÀ

Le linee guida di pratica clinica, i consigli pratici, le revisioni sistematiche e le altre linee guida pubblicate, approvate o affermate dall'ERN-RND sono valutazioni di informazioni scientifiche e cliniche attuali fornite come servizio educativo. Le informazioni (1) non devono essere considerate come comprensive di tutti i trattamenti adeguati, dei metodi di cura o come una dichiarazione dello standard di cura; (2) non sono continuamente aggiornate e potrebbero non riflettere le evidenze più recenti (possono emergere nuove informazioni tra il momento in cui le informazioni vengono sviluppate e quello in cui vengono pubblicate o lette); (3) affrontano solo la domanda o le domande specificamente identificate; (4) non impongono un particolare corso di cure mediche; e (5) non sono intese a sostituire il giudizio professionale indipendente del curante, poiché le informazioni tengono conto delle variazioni individuali tra i pazienti. In tutti i casi, la linea d'azione prescelta deve essere considerata dal medico curante nel contesto della cura del singolo paziente. L'uso delle informazioni è volontario. L'ERN-RND ha fornito queste informazioni "così come sono" e non fornisce alcuna garanzia, espressa o implicita, in merito alle informazioni. L'ERN-RND declina specificamente qualsiasi garanzia di commerciabilità o di idoneità per un uso o uno scopo particolare. ERN-RND non si assume alcuna responsabilità per eventuali lesioni o danni a persone o cose derivanti o correlati all'uso di queste informazioni o per eventuali errori od omissioni.

METODOLOGIA

Lo sviluppo dei diagrammi di flusso diagnostici per la distonia è stato realizzato dal gruppo di malattia per la demenza frontotemporale. Le scale utilizzate nella pratica clinica dei membri del gruppo di malattia sono state mappate e la decisione su quale scala proporre è stata presa a maggioranza.

Gruppo di malattie per la FTD:

Coordinatori del gruppo malattia:

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

Membri del gruppo malattia:

Operatori sanitari:

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]

Rappresentanti dei pazienti:

Mary Kearney

1 Assistance Publique-Hôpitaux de Paris, Hôpital Pitié-Salepétrière, Francia: Centro di riferimento per le demenze rare; 2 University College London Hospitals NHS Foundation Trust, Regno Unito; 3 University Hospitals Leuven, Belgio; 4 IRCCS Istituto Clinico Humanitas - Rozzano, Italia; 5 Klinikum der Universität München, Germania; 6 AOU Siena, Italia; 7 Fondazione IRCCS Istituto Neurologico Carlo Besta - Milano, Italia; 8 Semmelweis University, Ungheria; 9 University of Pécs, Ungheria; 10 University Medical Centre Ljubljana, Slovenia; 11 Universitätsklinikum Ulm, Germania; 12 Erasmus MC: University Medical Center Rotterdam, Paesi Bassi; 13 Università Carlo, Praga; 14 Universitätsklinikum Tübingen, Germania.

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 #: ⎵⎵⎵

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, 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 Dementia 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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Co-funded by the European Union