The Framingham Heart Study
Neuropsychology Group

Dementia Review Diagnostic Manual of Procedures

Version 6/23/2020

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Overview of Dementia Review

**** Please note that this protocol was written for the Brain Aging Program (BAP). The group led by Sudha Seshadri may have differences in procedures (see that group's documentation). However, the generation of the endpoints, we believe, remains consistent across the two groups.**

For a more comprehensive explanation of the different stages of the Dementia Review (DR) process, please see the *Dementia Review Manual of Procedures_[id]_[date]* ("id" is the Tech ID of the staff member who last edited the Manual of Procedures (MOP), and "date" is the date of that edit). This can be found on the N drive in this location: N:\Lab Activity\Dementia Review\~Protocols and Manuals~

The current document is more directly focused on the decision-making process involved in Dementia Review and guidelines for recording the data on the DR REDCap project.

Dementia Review is the process used at FHS for determine dementia-related endpoints. The stages of the process are

1. Participants for review are identified: The data management team generates of a list of participants who are flagged for possibly having cognitive decline. Flags include determination by a neurologist that there is impairment, the impression of a neuropsychological exam tester, a drop in MMSE by 3 points between any two consecutive Core cycle exams or 5 points between any two Core cycle exams, or referral (e.g., by Core staff, family, etc.). The list is generated based on the Principal Investigator's priorities.
2. A Research Assistant writes a summary of what is known about a participant (e.g., medical history, education, exam results) called a Dementia Review Case Summary (DRCS): The DRCS is compiled using several potential sources of information, although not every participant has every source available. These sources include the FHS Core exam(s), FHS Neuropsychological testing, FHS Neurology exam(s), FHS brain imaging, external medical records, and/or an interview with a family member (although the family interview is only done for participants who have donated their brain to the study).
3. A Dementia Review meeting is held: The DRCS is brought to a Dementia Review meeting, which includes an adjudication panel that must have at least one neuropsychologist and at least one neurologist. The panel evaluates each DRCS to identify whether there is evidence of cognitive decline, and, if so, the dates for last normal, impairment onset, mild dementia, moderate dementia, and severe dementia, as pertinent. If details in the DRCS are not clear, original source information is reviewed. A diagnostic impression is identified, as are additional possible contributors to observed cognitive impairment (e.g., toxic-metabolic, depression, brain tumor, etc.). In addition, the MCI stage is characterized (e.g., amnestic, non-amnestic, specific cognitive domains affected). Also noted at the time of DR are history of stroke or Parkinson's disease and results of brain scans. Although the brain scan findings have not contributed to clinical diagnoses, we have formalized the protocol to insure this by having all diagnostic decision-making done prior to reviewing the scans.
4. Dementia Review data is entered into a database: During the Dementia Review meeting, the Dementia Review form and CDR form (if applicable) are 1st and 2nd keyed immediately into a REDCap project. The REDCap project has extensive quality control built into it, via branching logic and data quality rules. The two keyed records are immediately compared, and any discrepancies are resolved at the time of the meeting.
5. For brain donors going to neuropath conference: Participants who are deceased and have donated their brain to our study are treated a little differently at DR because the case will be reviewed, with neuropathological findings, at our neuropath conferences (these occur a few times a year).

a. Brain imaging: Two weeks prior to the established time for the DR where a brain donor is to be reviewed, the RA in charge of DR informs the RA in charge of neuropath that the case is coming up. The latter then obtains any MRI/CT scans (the actual scans) to provide to the neurologist after DR decisions and prior to the neuropath conference. These scans will be found either at the Wellness Center (where we have been doing brain MRI scans since 1999) or the FHS Medical Records.
b. Family Interview: For participants who donate their brains, we do an extensive interview with a family – or another individual who knew the participant well – to gain additional information about the participant's cognition and adaptive behavior. This retrospective interview is aligned with the Clinical Dementia Rating (CDR)[1,2] interview, which provides information about a participants functioning which contributes to diagnosis and staging of dementia. If the participant had some cognitive impairment, the interviewer tries to ascertain the timeline of the impairment and any possible progression. A comprehensive view of the participant's functioning, starting at the beginning of impairment (if there is some) up until death, is obtained.
c. CDR: The Family Interview provides the information about functioning that is required for the panel to determine a CDR score, which classifies participants in terms of dementia severity (no dementia, mild cognitive impairment, mild dementia, moderate dementia, severe dementia) (see Appendix M).
d. Hachinski: The questions required to compute a Hachinski score are embedded in the Family Interview, and so are calculated from that source.
e. Blessed: The questions required to compute a Blessed score are embedded in the Family Interview, and so are calculated from that source.

Dementia Review Form

The Dementia Review Form is the data collection form on which all of the decisions made during the DR Meeting are recorded. See Appendix A for a copy of the DR form that was in use prior to December 2019 (recorded on paper), and Appendix B for a printout of the current REDCap form, launched in January 2019, which is electronic. The REDCap project is titled, ***Dementia Review**.* Also included in the REDCap ***Dementia Review*** project is an instrument for inputting CDR scores (see Appendix C).

What follows are notes on how to complete the Dementia Review Form. Embedded in these notes are guidelines that should be followed by the adjudication consensus panel in making determinations. It should be carefully reviewed by any neuropsychologist or neurologist on the panel, regardless of whether they will actually ever be required to enter the data on the form. However, it is primarily directed toward whoever is keying the data.

- There must be at least one neuropsychologist and at least one neurologist represented during the DR meeting, or the meeting has to be rescheduled. The FHS ID for these individuals is entered onto the form.
- Some participants have been reviewed previously, with some having multiple prior reviews. The Dementia Review Case Summary (DRCS) will be updated with any information that has become available since the last review. This new information will be presented in *italics* on the DRCS. It is not uncommon that newly obtained information requires a modification in data points (e.g., date of impairment onset,

dementia subtype). It should not be assumed that what was decided at a prior review meeting is still accurate.

- Participant ID – in REDCap, the ID must be entered without the dash, and all 0 place-holders should be included. For example, for ID 3-123, the entry in REDCap would be 30123.
- If the participant is deceased, answer "yes," and a new variable will be revealed asking for the date of death
- Beginning Information
    - The neurologist, neuropsychologist, RA who prepared the DRCS, and RA running the DR meeting should be coded here
    - If a new neurologist, neuropsychologist, or RA joins the FHS team, they should be added as options in the REDCap project. The people who currently have the ability to add new staff include: the project manager, the neuropsychologist, the RA(s) who manage or help to manage DR and data team members
- Short form or Long form
    - The short form includes only the following:
        - Identification information re participant and panel
        - Sources available for review
        - Date of last normal (with certainty ratings and supportive sources)
        - CT and MRI scan information
        - Cognitive status at time of death (with certainty ratings and supportive sources)
        - Whether the participant might have a developmental disorder or low education
    - The short form can be completed in these two situations:
        - It is determined that the participant did not have cognitive impairment
        - It is determined that the participant's impairment is non-progressive (e.g., developmental disorder)
    - If these two situations are not present, then answering "no" to the question, "Is this a Short Form?" will reveal the additional questions that need to be answered (i.e., the Long form).

Sources available for review

- **Neuropsychological Testing:** this refers only to NP testing done as part of a FHS brain research ancillary study (e.g., Kaplan-Albert, NP battery, MoCA); this does NOT include cognitive testing done by the Core (e.g., MMSE, CERAD, Stroop).
- **Neurological examination**: this refers only to neurology exams done as part of FHS research activities; exams done by outside neurologists go in the Medical Records category.
- **Family Interview Form**: Currently, the Family Interview (FI)is only done consistently for brain donors. The FI summary should be attached to the DRCS. Please note that this category does not include the Death Interview done by the Core—that is a different interview and it should be noted in the FHS Cycle Exam Records category.
- **FHS Cycle Exam Records:** this includes the MMSE, CERAD/Stroop, Medical History Updates (MHU), comments by Core study staff, chart summaries, and any other information obtained as part of the Core study research activities
- **Medical Records:** this includes all records generated from non-FHS related activities, such as hospital/ED notes, doctor notes, nursing home notes, neuropsych/neurology consults, etc.

Sources supporting…

- Many questions include an opportunity to indicate which sources were relied upon to make each decision (e.g., dates of onset, presence of dementia)

- REDCap will only expose the sources that were identified in the "Sources available for review" section (e.g., if you indicate there was no Family Interview (FI), then FI will not be an option throughout the form.
- For specific dates, in addition to the primary source of the date, sources can be considered supportive of the identified date when:
    - Supportive evidence falls within approximately 1 year of the identified date
    - The time course suggests support (e.g., mild dementia onset is 2010 based on medical records, and, based on NP testing, impairment onset is in 2009 and moderate dementia is in 2011; in this case, although the mild dementia date is based on medical records, that date is supported by NP testing)

Dates – general comments

- If the participant's cognitive impairment was caused by a discrete event, such as a stroke, with no evidence of earlier cognitive decline, the "cognitively intact" date and the "impairment onset" date will be the same.
- In general, the date from stronger sources of support and/or sources of support closest in time to transitions, should be used. There is often a trade-off here, which needs to be resolved with clinical judgment; however, typically the stronger source of support would be the better choice
- Although we really try to avoid coding dates as "unknown," sometimes it has to be done because the evidence is insufficient. For each date needed, you are first asked if the date has been identified. If you answer 1 (yes), then fields will open for you to enter the date and the degree of certainty; otherwise, these remain hidden
- All dates must have at least one source of support
- Occasionally it is not possible to identify the specific date or year, but evidence exists that can give you the month and/or the year. In these cases, a "dummy date" that is half-way through the month or year is given.
    - Unknown date (known month and year): MM/15/YYYY
    - Unknown month (known year): 06/30/YYYY
- After you enter a date into REDCap, you will be asked whether the date was an estimate. This is because it is possible that the 15th, or June 30, are real dates, and we want to keep these dates distinct.

Degree of certainty

- This is determined by the level of detail and reliability of the sources from which the diagnosis of dementia is made.
- It may be that sources of support clearly demonstrate particular stages of cognitive decline, but there is a large span of time for which no information is available. This would lower our degree of certainty regarding the date, because it may be that the onset of that stage of decline happened quite a bit earlier than the identified date. For example, last normal was in 1999 based on NP testing, and the first sign of cognitive impairment was on NP testing in 2009. We would choose the 2009 date, but with less certainty because the true date of onset may have been anytime during the 10 years between NP testings.

Last date documented to be cognitively intact

- This is the last date that the participant was known to be at baseline functioning.
- If onset was sudden, such as with stroke, the "cognitively intact" date may be the same as a date of impairment
- If cognitive impairment is thought to be exclusively due to a developmental disorder, then last cognitively intact will be the last date for which any data from any of the 5 sources provides documented evidence there was no decline (i.e., cognition determined to be baseline).

Cognitive impairment

- Whether a participant has cognitive impairment is determined by evaluating multiple factors (e.g., level of education, native language, age, baseline cognitive functioning) that contribute to cognitive functioning; if evidence exists that the participant had reduced cognition from baseline, this is marked "yes"
- See Appendix F. Staging Dementia
- If the cognitive impairment is thought to be exclusively due to a developmental disorder, then cognitive impairment should be marked "yes" and the question at the end of the form that asks about developmental disorder should also be "yes."
- In the past, this type of impairment would have resulted in an answer of "no" to the question of impairment. If needed, an appropriate adjustment can be made in the data set by setting the answer to "no" if the developmental disorder questions is "yes" and the date of onset is the participant's date of birth

Date of cognitive impairment onset

- If cognitive functioning is determined to be below baseline, the date of the source that reflects this performance earliest in the person's life should be used
- If the cognitive impairment is thought to be exclusively due to a developmental disorder, then the date of cognitive impairment should be the participant's date of birth

Cognitive decline

- Care should be taken with this variable because, unlike most other variables that are coded as "present/yes" or "not present/no," the cognitive decline variable has three options:
    - No
    - Yes, duration less than 6 months
    - Yes, duration greater than 6 months
- In addition, it is important to note that the cognitive decline referenced here is *additional decline after impairment onset date (i.e., it represents a decline from MCI to either a more pronounced MCI or to dementia). It is possible to have decline but never progress beyond MCI (e.g., participant has single domain MCI at start of impairment then progresses to multi-domain MCI).*
- If the decline only happens at or close to death, this is not considered decline; 6 months is the general guideline (i.e., if decline occurs within 6 months of death, record "no")
- The distinction between less than and greater than 6 months is meant to reflect the stability of the impairment; that is, if less than 6 months it may be a temporary condition, whereas that is less likely if it persists for greater than 6 months
- Code "0" if there are records that follow the date of impairment onset, and none of them provide evidence of decline
- Code "9" if there are absolutely no records after the date of impairment onset

Probable dementia present

- See Appendix D (DSM-IV Criteria for Dementia) and Appendix E (Alzheimer's Disease Diagnostic and Treatment Centers (ADDTC) Definition of Dementia)
- Whether you say "yes" or "no," the sources of support should be identified. That is, for example, if a neurology consultation suggests probable dementia, but all the remaining evidence suggests no dementia, then mark "no" for "probable dementia present" and indicate those other sources by coding as "1" (neurology would be coded as "0" because it does not support the lack of dementia)

Dates of diagnosis for Mild, Moderate, and Severe dementia

- Identifying the stages of dementia is a challenging task, with no clear-cut rules. See Appendix F for some examples of things you might see during the different stages of dementia. These should be used as a general guideline only
- Avoid using a date from the last months of life/terminal stage
- In the absence of an "event," do not date stages less than 6 months apart unless there is strong evidence

Definite stroke or TIA from Stroke Review

- This, quite literally, is the outcome of a FHS Stroke Review. Even if medical records seem to indicate there was a clinical stroke, if the stroke team decided it was not a stroke, then it is coded here as no stroke.
- If medical records suggest there was a stroke at some point in time *after* the Stroke Review was done, then the case should be referred back to the Stroke team for a re-review prior to completing the DR.
- Note that this is asking about stroke ***or*** TIA, not just stroke. If the stroke team indicates that there was a TIA, you will code "yes," and a new variable will show up where you can indicate whether it was determined to actually be a stroke or whether it was only TIA.

Parkinson's disease

- Code yes (1) if a diagnosis of PD was identified in any of the sources
- If the available information suggests the possibility of PD, the case should be referred for PD Review. However, the DR can be completed and this variable should be marked "no"

CT/MRI scan information

- First you are asked whether there are any CTs, and if you reply 1 (yes), then additional fields open. The same is true for MRI scans.
- The date and results from the *most recent* scan are coded
- Options for results are:
    - 1 = Normal
    - 2 = Atrophy only
    - 3 = Single Stroke
    - 4 = Two or more Strokes
    - 5 = Other,_____ (If you code "Other," a field will open for you to type in the findings)
    - 6 = Small Vessel Ischemic disease
    - 9= Unknown
- Changes in the brain (e.g., atrophy, white matter hyperintensities (WMH)) are expected as people age, and the majority of scans we see reflect this. Typically, the radiologist's impression is written to reflect this (e.g., "age-related atrophy").
- If the imaging report suggests "age-related" atrophy or WMH, the answer to the question, "CT [or MRI] scan results" should be 0 (**Normal**).
- If age-related changes are noted in the report, the question, "Are there other CT [MRI] results to note?" should be coded as 1 (yes), and then the specific findings can be coded. Depending on which options you choose, new questions will be revealed. For example, if you mark "atrophy" here, then you will be asked whether the atrophy was noted to be (or clearly implied to be) related to age and you will also be asked the severity (e.g., mild/age-related through extensive).
- Sometimes, the scan results reflect multiple findings of interest (e.g., atrophy, extensive white matter changes, brain tumor), but only one can be coded in the initial question asking about findings. Code the one that is most likely to be contributing to cognitive decline. Then, code the additional findings in the "other results to note" question.

- Do not include post-mortem MRI
- Radiology reports are dependent on terminology at the current time. (For ex: terms to describe white matter changes may be different in old CT or MRI scan reports)
- Synonyms for small vessel ischemic disease (#6) include microvascular ischemic disease, small vessel degeneration, and periventricular ischemic disease

Brain bank subject?

- This should be marked "yes" if the person is registered for the FHS brain donation program or is deceased and already donated their brain. If registered for a different brain bank, mark "no"
- If you mark "yes," a new field will open that asks whether the DR is taking place *after* the neuropath conference. Although DR after the neuropath conference is not currently part of the approved protocol, there has been internal discussions about re-reviewing cases after the neuropath conference, so this question was added to allow this to be clearly documented in the event that appropriate approval is obtained.

Brain autopsy performed?

- If the person is alive, this is clearly marked "no"
- If the person donated the brain to FHS, and we have received it, mark "yes" (whether or not the neuropath team has completed the autopsy)
- The brain autopsy report for our neuropath cases should never be available for the DR meeting; the panel should be blind to these results
- If the person had a brain autopsy performed by a different agency, and so the participant is not a FHS neuropath case, then the autopsy report can be made available to the panel for review

Hachinski[3] and Blessed[4] Scores

- See Appendix G (Hachinski) and Appendix H (Blessed)
- These scores can only be computed for cases for whom we have a Family Interview (i.e., neuropath cases)
- The person doing the Family Interview will calculate the scores and provide them to the panel

Cognitive status at the time of death

0 = No Dementia
0.5 = Cognitive Impairment – No Dementia
1 = Mild Dementia
1.5 = Greater than or equal to mild dementia
2 = Moderate Dementia
2.5 = Greater than or equal to moderate dementia
3 = Severe Dementia
4 = Alive ***Note that this must be coded as "4" if the person is still alive at the time of DR**
99 = Unknown

- Once a person becomes demented, you have the option of coding them *at* the highest level of dementia determined prior to death (i.e., 1, 2, 3) or coding them as *at least at* the highest level of dementia (i.e., 1.5, 2.5). The latter is used by considering the amount of time between the last documentation supporting the dementia stage and death, because you don't know whether they progressed further to the next stage.

Dementia subtype

NOTE:

Probable AD + Probable VaD = Mixed dementia (4)
Possible AD + Probable VaD = Mixed dementia (4)
Probable AD + Possible VaD = AD w/out stroke (1) OR AD w/stroke (2)

0 = None
1 = Alzheimer's Disease[5,6] Without Stroke
If AD is coded here (codes 1, 2, or 4), then the question "Alzheimer's disease by NINCDS-ADRDA criteria" must be coded 1 (yes)
See Appendix I
2 = Alzheimer's Disease With Stroke
This can be coded if Stroke Review determined there was no stroke, but there is clear evidence of stroke (e.g., a terminal stroke)
3 = Vascular Dementia[7,8] Without Alzheimer's Disease (see Appendix J)
If Vascular Dementia is coded here (codes 3 and 4), then the question "Vascular dementia present" must be coded 1 (yes)
4 = Mixed Dementia Type (Alzheimer's Disease + Vascular Dementia)
5 = Frontotemporal Dementia[9,10] (see Appendix K)
6 = Dementia with Lewy Bodies[11-13] (see Appendix L)
7 = Dementia that does not fit any other Category (progressive)
8 = Dementia that does not fit any other Category (non-progressive)
9 = Cognitive Impairment – No Dementia
This is coded when the participant is characterized as having mild cognitive impairment, but it has not progressed to dementia
10 = Dementia – Uncertain
The "uncertain" here refers to etiology – the person is deemed to have dementia but the etiology is uncertain
99 = Unknown

Severity of dementia subtype

- This should be coded as the highest level of dementia (0, 1, 2, 3)
- So if, for example, you coded "2.5" for the question "Cognitive status at the time of death," then the code here would be "2" (they have not achieved the level of severe dementia).

Cognitive impairment (i.e., MCI stage) subtype[14-16]

NOTE: If the Cognitive Impairment Onset Date is not known, these questions regarding the MCI stage will not be revealed, because you cannot characterize MCI if you don't know when the person had MCI

- First decide whether the MCI stage is best described as **amnestic** or **non-amnestic**. Whichever you choose will reveal the relevant questions
- Next decide whether there is only one affected domain ("**amnestic only**" or "**non-amnestic single domain**") or more than one ("**amnestic plus**" or "**non-amnestic multiple domain**")
- For "amnestic only," you are now done with this section
- For "non-amnestic single domain," you will need to identify the domain
- For "amnestic plus" or "non-amnestic multiple domain," you will need to identify all affected domains
- Domains include: **executive functioning, abstract reasoning, visuospatial functioning, language, and attention**

Criteria for DSM-IV

NOTE: At this point, you are no longer answering questions based *only* on the MCI stage (as with the previous question). Now you are indicating whether these problems were evident at any stage

- Memory impairment
  - You choose here whether memory impairments affected verbal memory, nonverbal memory, or both

    - Often, there is no direct reference to nonverbal memory – it is not part of routine mental status evaluations, so, unless the participant had NP testing, you may not have any indication of nonverbal memory. In this case, code 9 (unknown), which will reveal two variables that allow you to indicate yes/no/unknown for verbal and nonverbal memory separately. To illustrate, if you have NP testing that clearly shows verbal memory impairment but no nonverbal memory impairment, you would code 1 (Verbal only) for this variable. However, if you don't have NP testing, and are just going by medical records, for example, then you don't want to code "Verbal only" because you really don't know about nonverbal. Thus, you code "unknown" and are then able to specify "yes" to verbal memory and "unknown" to nonverbal memory. This is a relatively new choice (documenting them separately); in data post-processing, the "unknown" is assigned to whichever type of memory impairment is selected as "yes," so that it is equivalent to how the variable would previously have been coded.
- Aphasia – Partial or total loss of the ability to articulate ideas or comprehend spoken or written language.
- Apraxia – partial or total loss of the ability to perform coordinated movements or manipulate objects in the absence of motor or sensory impairment
- Agnosia – Loss of the ability to interpret sensory stimuli such as sounds or images
- Apraxia and Agnosia
    - Apraxia and agnosia are rarely mentioned in records
    - Code 9 (unknown) unless explicitly stated in records to be absent (0) or present (1)
- Executive functioning – Difficulties in planning and carrying out a task and manipulating with information, including trouble following commands, drawing, or working with numbers
- Abstraction – Deficits in similarities, difficulty interpreting a proverb or saying, concreteness in thinking.
- Impairment in function (social/occupational)
    - This is also rarely mentioned in records. We often have to surmise, based on level of impairment, whether it impacts function
    - Although this is a requirement for diagnosis of dementia, we would rarely be able to diagnose dementia if we needed hard, concrete evidence of a decline in functional abilities.
    - If a decline in functioning is explicitly documented, or if it is *highly likely* that functioning is impacted, code 1 (yes); if there is explicit documentation noting the lack of a decline in functioning, code 0 (no); if there is no indication of whether functioning is impaired or not, code 9 (unknown)
    - If "no" is chosen for decline in functioning, a logic check will not permit the diagnosis of dementia; thus, it is important to use the code of 9 (unknown) when functional abilities is truly unknown but the data otherwise supports a dementia diagnosis
    - Note: It is not uncommon to see a neurology report or a medical record in which the doctor says that cognitive problems do not affect functioning, but this may be based on self-report by the participant, who may be a poor informant. Always consider the source(s) of information

Dementia by DSM-IV criteria

- See Appendix D
- Memory impairment
- Impairment in at least one other cognitive domain
- Functional decline
- Not due to delirium, depression, or schizophrenia

Dementia by ADDTC criteria

- See Appendix E

- Impairment in two or more cognitive domains
- Functional decline secondary to cognitive impairment

Symptoms above present for at least six months

- The symptoms being referred to here are the ones that fall under the "Criteria for DSM-IV" section; that is, memory impairment, aphasia, apraxia, agnosia, executive functioning, abstraction, and impairment in function
- This question does NOT refer to the immediately preceding questions about whether DSM-IV and/or ADDTC criteria have been met

Cognitive deficits not related to DSM-IV criteria

- Language
- Visuospatial abilities
- Attention

Alzheimer's disease by NINCDS-ADRDA criteria

- See Appendix I
- Based on the NINCDS-ADRDA criteria, indicate whether AD is probable, possible, or definite
- NOTE: It is possible for a participant to have probable AD *and* probable vascular dementia
- If "possible" is chosen, you have the opportunity to define whether it is likely mixed with vascular disease, parkinsonism (including drug-induced), or "other"
- If you determine that there is more than one co-occurring etiology, code the one that seems to be most pertinent for the particular participant's presentation

Vascular dementia questions

- Dementia present – this must match the "Probable dementia present" variable
- Clinical stroke documented – this is based on the outcome of the Stroke Review; if you mark 1 (Yes, one stroke) or 2 (Yes, more than one stroke) or 3 (Terminal stroke only) here, then the question, "Definite stroke or TIA from Stroke Review" must be coded as 1 (yes)
- Suggestive Temporal Profile
    - Mark 1 "yes" if any of the following are indicated
        - Onset of dementia less than 3 months after stroke
        - Abrupt onset
        - Fluctuating stepwise decline
- Imaging (CT or MRI)
    - NOTE: This variable should be consistent with the CT and/or MRI results noted earlier. It does NOT need to be consistent with the conclusions of Stroke Review.
    - The following scan findings are documented here
        - Normal
        - One stroke
        - More than one stroke
        - Other _____
        - Extensive white matter changes
        - Unknown
    - "Atrophy only" is coded as 1 (Normal)
    - White matter changes need to be characterized as "extensive" (or phrasing suggesting extensive); otherwise, code 1 (Normal)
- Suggestive Temporal Profile
    - Onset of dementia is less than 3 months after stroke, or
    - Abrupt onset, or

    - Fluctuating stepwise decline
- Focal neurological signs suggestive of stroke
    - Weakness of an extremity
    - Exaggerated DTRs
    - Pseudobulbar palsy
    - Extensor plantar responses
    - Gait abnormalities
    - Hemianopsia
    - Facial weakness
    - Dysarthria
    - Sensory deficit if thought to be of vascular etiology
- Vascular dementia present
    - On REDCap, your answers to the Vascular Dementia questions (#29-32 in this outline) will be compiled to determine whether criteria is met for Probable or Possible Vascular Dementia.
    - See Vascular Dementia Example
    - If any of these fields are marked "1", then "Vascular dementia present" should be marked as 1 (yes); otherwise, mark 0 (no)
- If Vascular dementia, indicate Probable or Possible
    - This variable, too, will be computed for you.
    - If multiple possibilities are marked as 1 (criteria was met), code the lowest number (see Vascular Dementia Example)
- Coma/Persistent Vegetative State Post Stroke, Until Death
    - This is pretty straightforward

Other causes of dementia or impairment

- It is important to consider whether any of the things listed here have *contributed to cognitive impairment,* and only then should it be coded here. For example, there may be depression described in medical records, but there is no indication that the depression was significant enough to cause or contribute to cognitive impairment; in this case, you should not code depression here
- You can code up to three (3) of these "other causes"
- 1=PD prior to dementia onset
    i. The motor symptoms of PD are evident at some time prior to onset of cognitive decline; this is more likely to be PD with dementia
- 2=PD after dementia onset
    i. Cognitive decline comes first, then the motor symptoms of PD; this is more likely to be DLB
- 3=Dementia with Lewy Bodies
    i. If a different etiology (e.g., AD) is thought to be the primary cause of cognitive decline, and so you had to code one of the AD codes for Dementia Subtype, then you can capture it here if you believe there is also evidence for DLB
- 4=PSP
- 5=Shy-Dager syndrome
- 6=Striato-nigral degeneration
- 7=FTD with Parkinsonism
- 8=Wilson's disease
- 9=FTD (w/ and w/out atrophy on imaging)
- 10=Cortiocobasal ganglionic degeneration
- 11=Huntington's disease

- 12=Spino-cerebellar degeneration
- 13=Leukodystrophies
- 14=Post cardiac arrest
- 15=TBI
- 16=Post infectious sequelae (after meningitis, encephalitis, ADEM)
- 17=Malignancy (primary, secondary, para-neoplastic)
- 18=Subdural hematoma
- 19=NPH
- 20=CJD
- 21=Multiple sclerosis
- 22=AIDS associated dementias
- 23=Other infections (fungal meningitis, syphilis)
- 24=Alcoholic dementia
- 25=Toxic-Metabolic Encephalopathy
- 26=Dementia – Uncertain Etiology
- 27=Other Etiologies (specify _____)
- 28=History of Depression
- 29=History of Alcohol/Drug abuse
- 30=Unknown or N/A

2. Earliest Documented Date of Dementia (EDDD)

- This section is *only* completed if a Mild Dementia date could not be determined
- Indicate the earliest date for which you determined the participant to be demented
- Severity – indicate the severity of dementia at the time of this earliest date

Note: Not sure why there are options for "Mild" and "greater than or equal to Mild" on the original form because if you have a date for Mild then you wouldn't fill in this section

Vascular Dementia Example: REDCap equations will identify whether reported symptoms qualify for a diagnosis of Vascular Dementia, and provide the subtype(s) for which criteria have been met. In this case, criteria for both subtype = 1 and subtype = 5 have been met. Choose the lowest number to input for "If yes to Vascular Dementia Present, code…" (which is 1 in this example).

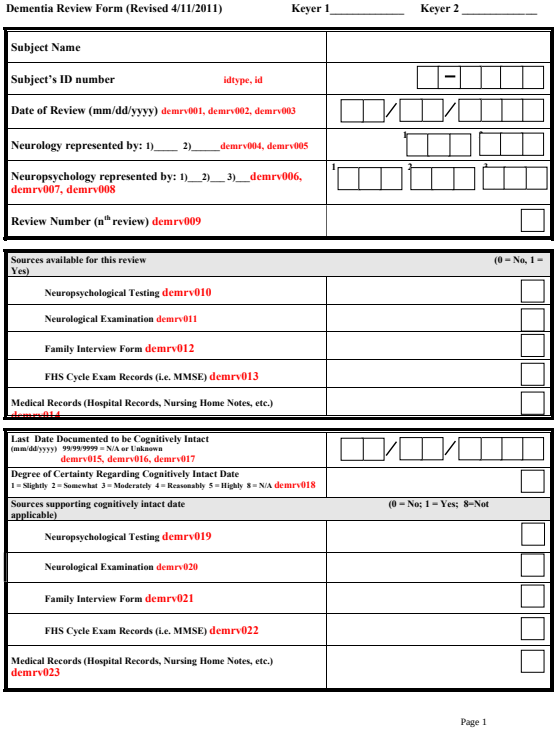
Vascular Dementia Questions (Ignore if completing the Short Form)	
Dementia Present	1 View equation
Clinical Stroke Documented ('definite' stroke at stroke review)	1 = Yes, One Stroke
Suggestive Temporal Profile (onset of dementia less than 3 months after stroke, abrupt onset or fluctuating stepwise decline)	1 = Yes
Imaging (CT or MRI)	2 = Yes, More Than One Stroke
Focal Neurological Signs Suggestive of Stroke	1 = Yes
Focal neurological signs include: **• Weakness of an extremity** **• Exaggerated DTRs** **• Pseudobulbar palsy** **• Extensor plantar responses** **• Gait abnormalities** **• Hemianopsia** **• Facial weakness** **• Dysarthria** **• Sensory deficit if thought to be of vascular etiology**	
Vascular Dementia subtype = 1?	1 View equation 1 = YES, 0 = NO
Vascular Dementia subtype = 2?	0 View equation 1 = YES, 0 = NO
Vascular Dementia subtype = 3?	0 View equation 1 = YES, 0 = NO
Vascular Dementia subtype = 4? **(**Note: in addition to a value of "1," there must be a diagnosis of Binswanger's disease)**	0 View equation 1 = YES, 0 = NO
Vascular Dementia subtype = 5?	1 View equation 1 = YES, 0 = NO
Vascular Dementia Present	1 = Yes
If yes to Vascular Dementia Present, code	1 = Probable (dementia present + clinical str
Coma/Persistent Vegetative State Post Stroke, Until Death	0 = No

Frequently Asked Questions

1. **If a participant is deemed cognitively impaired exclusively due to a developmental disorder, how should this be captured on the DR form?**
    a. Complete the "Short Form"
    b. "Last cognitively intact" date should be the date of death; or, if alive, the last known date (if info is from a long time ago) or date of review (if we have recent data)
    c. "Cognitive impairment" should be coded as "yes"
    d. "Cognitive status at death" should be coded as "0.5" if dead (if alive, code "4")
    e. "Is a Developmental Disorder (e.g., intellectual disorder, learning disorder) suspected, or does limited education possibly explain impairment?" should be yes
    f. Write in comments box that observed impairment is likely due to developmental disorder
    g. This will break Rule #75 ("If cognitive impairment is yes, subtype cannot be 0 (normal)"). When the warning screen pops up, choose "exclude" to bypass the logic check.
2. **If low scores on NP testing are thought to reflect low educational attainment, how should this be captured on DR form?**
    a. Complete the "Short Form"
    b. "Last cognitively intact" date should be the date of death; or, if alive, the last known date (if info is from a long time ago) or date of review (if we have recent data)
    c. "Cognitive impairment" should be coded as "yes"
    d. "Cognitive status at death" should be coded as "0" if dead (if alive, code "4")
    e. "Is a Developmental Disorder (e.g., intellectual disorder, learning disorder) suspected, or does limited education possibly explain impairment?" should be yes
    f. Write in comments box that observed low scores are likely related to low education

Appendix A. Dementia Review Paper form used through December 2018

See document entitled “Dementia Review Form 20110411, located on the N drive in this location: location



Dementia Review Form (Revised 4/11/2011) Keyer 1_____ Keyer 2 _____

Subject Name	
Subject's ID number idtype, id	
Date of Review (mm/dd/yyyy) demrv001, demrv002, demrv003	
Neurology represented by: 1)____ 2)_____demrv004, demrv005	
Neuropsychology represented by: 1)__2)___ 3)___demrv006, demrv007, demrv008	
Review Number (n[th] review) demrv009	

Sources available for this review	(0 = No, 1 = Yes)
Neuropsychological Testing demrv010	
Neurological Examination demrv011	
Family Interview Form demrv012	
FHS Cycle Exam Records (i.e. MMSE) demrv013	
Medical Records (Hospital Records, Nursing Home Notes, etc.) demrv014	

Last Date Documented to be Cognitively Intact (mm/dd/yyyy) 99/99/9999 = N/A or Unknown demrv015, demrv016, demrv017	
Degree of Certainty Regarding Cognitively Intact Date 1 = Slightly 2 = Somewhat 3 = Moderately 4 = Reasonably 5 = Highly 8 = N/A demrv018	
Sources supporting cognitively intact date applicable)	**(0 = No; 1 = Yes; 8=Not**
Neuropsychological Testing demrv019	
Neurological Examination demrv020	
Family Interview Form demrv021	
FHS Cycle Exam Records (i.e. MMSE) demrv022	
Medical Records (Hospital Records, Nursing Home Notes, etc.) demrv023	

Page 1

Cognitive Impairment 0 = No 1 = Yes 9 = Unknown demrv024	
Sources supporting presence/absence of cognitive impairment	**(0 = No; 1 = Yes; 8=Not applicable)**
Neuropsychological Testing demrv025	
Neurological Examination demrv026	
Family Interview Form demrv027	
FHS Cycle Exam Records (i.e. MMSE) demrv028	
Medical Records (Hospital Records, Nursing Home Notes, etc.) demrv029	
Date of Cognitive Impairment Onset (mm/dd/yyyy) 99/99/9999 = N/A or Unknown demrv031, demrv032, demrv033	
Degree of Certainty Regarding Impairment Onset Date 1 = Slightly 2 = Somewhat 3 = Moderately 4 = Reasonably 5 = Highly 8 = N/A demrv034	
Sources supporting cognitive impairment onset date	**(0 = No; 1 = Yes; 8=Not applicable)**
Neuropsychological Testing demrv035	
Neurological Examination demrv036	
Family Interview Form demrv037	
FHS Cycle Exam Records (i.e. MMSE) demrv038	
Medical Records (Hospital Records, Nursing Home Notes, etc.) demrv039	

Cognitive Decline demrv040 0 = No 1 = Yes, Duration Less Than 6 Months 2 = Yes, Duration Greater Than 6 Months 9 = Unknown	
Sources supporting the presence or absence of cognitive decline	**(0 = No; 1 = Yes; 8=Not applicable)**
Neuropsychological Testing demrv041	
Neurological Examination demrv042	
Family Interview Form demrv043	
FHS Cycle Exam Records (i.e. MMSE) demrv044	
Medical Records (Hospital Records, Nursing Home Notes, etc.) demrv045	

Page 2

Probable Dementia Present 0 = No 1 = Yes 9 = Unknown demrv046	
Sources supporting the presence or absence of dementia	**(0 = No; 1 = Yes; 8=Not applicable)**
Neuropsychological Testing demrv047	
Neurological Examination demrv048	
Family Interview Form demrv049	
FHS Cycle Exam Records (i.e. MMSE) demrv050	
Medical Records (Hospital Records, Nursing Home Notes, etc.) demrv051	

Date of Diagnosis of Mild Dementia (mm/dd/yyyy) 99/99/9999 = N/A or Unknown demrv052,demrv053,demrv054	
Degree of Certainty regarding Mild Dementia Date demrv055 1 = Slightly 2 = Somewhat 3 = Moderately 4 = Reasonably 5 = Highly 8 = N/A	
Sources supporting the date of mild dementia	**(0 = No; 1 = Yes; 8=Not applicable)**
Neuropsychological Testing demrv056	
Neurological Examination demrv057	
Family Interview Form demrv058	
FHS Cycle Exam Records (i.e. MMSE) demrv059	
Medical Records (Hospital Records, Nursing Home Notes, etc.) demrv060	

Date of Diagnosis of Moderate Dementia (mm/dd/yyyy) 99/99/9999 = N/A or Unknown (Dementia Diagnosis Date) demrv061, demrv062, demrv063	
Degree of Certainty regarding Moderate Dementia Date demrv064 1 = Slightly 2 = Somewhat 3 = Moderately 4 = Reasonably 5 = Highly 8 = N/A	
Sources supporting the date of moderate dementia	**(0 = No; 1 = Yes; 8=Not applicable)**
Neuropsychological Testing demrv065	
Neurological Examination demrv066	
Family Interview Form demrv067	
FHS Cycle Exam Records (i.e. MMSE) demrv068	
Medical Records (Hospital Records, Nursing Home Notes, etc.) demrv069	

Page 3

Date of Diagnosis of Severe Dementia (mm/dd/yyyy) 99/99/9999 = N/A or Unknown demrv070, demrv071, demrv072	
Degree of Certainty regarding Severe Dementia Date demrv073 1 = Slightly 2 = Somewhat 3 = Moderately 4 = Reasonably 5 = Highly 8 = N/A	
Sources supporting the date of severe dementia	**(0 = No; 1 = Yes; 8=Not applicable)**
Neuropsychological Testing demrv074	
Neurological Examination demrv075	
Family Interview Form demrv076	
FHS Cycle Exam Records (i.e. MMSE) demrv077	
Medical Records (Hospital Records, Nursing Home Notes, etc.) demrv078	

Definite Stroke or TIA from Stroke Review 0 = No 1 = Yes 9 = Unknown demrv079	
Parkinson's Disease demrv080 0 = No 1 = Yes 9 = Unknown	

CT Scan Information	
CT Scan Performed: demrv081 0 = No 1 = Yes 9 = Unknown	
Date of the Most Recent CT Scan demrv082, demrv083, demrv084 (mm/dd/yyyy) 99/99/9999 = N/A or Unknown	
CT Scan Results: demrv085 1 = Normal 2 = Atrophy only 3 = Single Stroke 4 = Two or more Strokes 5 = Other,____ 6 = Small Vessel Ischemic disease 8=Not Done 9= Unknown	

MRI Scan Information	
MRI Scan Performed: demrv086 0 = No 1 = Yes 9 = Unknown	
Date of the Most Recent MRI Scan demrv087, demrv088, demrv089 (mm/dd/yyyy) 99/99/9999 = N/A or Unknown	
MRI Scan Results: demrv090 1 = Normal 2 = Atrophy only 3 = Single Stroke 4 = Two or more Strokes 5 = Other,____ 6 = Small Vessel Ischemic disease 8=Not Done 9= Unknown	

Brain Autopsy Information	
Brain Bank Subject? 0 = No 1 = Yes 9 = Unknown demrv091	
Brain Autopsy Performed? 0 = No 1 = Yes 8 = N/A 9 = Unknown demrv092	
Brain Autopsy Report Available? 0 = No 1 = Yes 8 = N/A 9 = Unknown demrv092b	

Page 4

Hachinski Ischemia Score (Range 0 - 18) 88 = N/A demrv094	
Blessed Score (Range 0 - 25) 88 = N/A demrv095	

Cognitive Status at Time of Death demrv096 0 = No Dementia 0.5 = Cognitive Impairment – No Dementia 1 = Mild Dementia 1.5 = Greater than or equal to mild dementia 2 = Moderate Dementia 2.5 = Greater than or equal to moderate dementia 3 = Severe Dementia 4 = Alive 7=died, unknown at final review 9 or 9.9 = Unknown	
Certainty of Cognitive Status at Death demrv097 1 = Slightly 2 = Somewhat 3 = Moderately 4 = Reasonably 5 = Highly 8 = N/A	
Sources supporting the cognitive status at death applicable)	**(0 = No; 1 = Yes; 8=Not**
Neuropsychological Testing demrv098	
Neurological Examination demrv099	
Family Interview Form demrv100	
FHS Cycle Exam Records (i.e. MMSE) demrv101	
Medical Records (Hospital Records, Nursing Home Notes, etc.) demrv102	

Dementia Subtype (fill out at review only) demrv103 **0 = None** **1 = Alzheimer's Disease Without Stroke** **2 = Alzheimer's Disease With Stroke** **3 = Vascular Dementia Without Alzheimer's Disease** **4 = Mixed Dementia Type (Alzheimer's Disease + Vascular Dementia)** **5 = Frontotemporal Dementia** **6 = Dementia with Lewy Bodies** **7 = Dementia that does not fit any other Category (progressive)** **8 = Dementia that does not fit any other Category (non-progressive)** **9 = Cognitive Impairment – No Dementia** **10 = Dementia – Uncertain** **99 = Unknown**	If answer is dementia due to inability to fit other categories, specify:
Severity Of Dementia Subtype demrv104 **0 = None 1 = Mild 2 = Moderate 3 = Severe 9 = Unknown**	
For Cognitive Impairment (this only refers to impairment *during the MCI stage*), Code Subtype: **1= amnestic 2=non-amnestic 8=N/A 9=unknown** add01	
If Amnestic, Code Subtype: **1=amnestic only 2=amnestic plus 8=N/A 9=unknown** add02	
If 2 ("amnestic plus") code: 0= no 1= yes 9= unknown. If N/A, leave blank. Add03-add07 **executive function abstract reasoning visuospatial function language attention**	
If Non-Amnestic, Code Subtype: add08 **1=single domain 2=multiple domain 8=N/A 9=unknown**	
If 1 ("single domain") or 2 ("multiple domain") code: 0=no 1= yes 9= unknown. If N/A, leave blank. Add09-add13 **executive function abstract reasoning visuospatial function language attention**	

Dementia Review Form Supplement

Criteria for DSM-IV	
Memory Impairment **0 = No 1 = Yes 9 = Unknown** Demrv105	**If Memory Impairment, Code Subtype:** add15 **1= Verbal and Non-verbal memory 2=Verbal only 3= Non-verbal only** **8= N/A 9= Unknown**
Aphasia **0 = No 1 = Yes 9 = Unknown** demrv106	
Apraxia **0 = No 1 = Yes 9 = Unknown** demrv107	
Agnosia **0 = No 1 = Yes 9 = Unknown** demrv108	
Executive Dysfunction (planning, organizing, sequencing, abstracting) **0 = No 1 = Yes 9 = Unknown** demrv109	
Impaired Abstraction demrv110 **0 = No 1 = Yes 9 = Unknown**	
Significant Impairment in Function (Social/Occupational) **0 = No 1 = Yes 9 = Unknown** demrv111	
Dementia by DSM-IV Criteria Memory Impairment, Impairment in one other Cognitive Domain, Functional Decline, Not Due to Delirium, Depression, or Schizophrenia **0 = No 1 = Yes 9 = Unknown** demrv112	
Dementia by ADDTC criteria Impairment in two or more Cognitive Domains, Functional Decline secondary to Cognitive Impairment **0 = No 1 = Yes 9 = unknown** demrv113	
Symptoms Above Present for at least Six Months (refers to memory imp, aphasia, etc., *not* dementia) **0 = No 1 = Yes 8 = N/A 9 = unknown** demrv114	

Cognitive Deficits Not Related to DSM-IV Critera	
Language **0 = No 1 = Yes 9 = Unknown** add16	
Visuospatial Abilities **0 = No 1 = Yes 9 = Unknown** add17	
Attention **0 = No 1 = Yes 9 = Unknown** add18	

Alzheimer's Disease by NINCDS-ADRDA criteria (fill out at review only) **0 = No 1 = Yes 9 = Unknown** demrv115	
Classification of Alzheimer's Disease (fill out at review only; note that, in certain cases, a pt. *can* have both probable AD and probable vascular dementia) **1 = Probable AD (dementia, progression, and no other etiology)** **2 = Possible AD (dementia, progression, unusual clinical features or other contributory etiologies)** **3= Definite AD** **8 = N/A 9 = Unknown** demrv116	
If Possible AD, Code Subtype below: (fill out at review only) **1 = Mixed AD + Vascular** **2 = Mixed AD + Parkinsonism (includingdrug-induced)** **3 = Mixed AD + Other, Specify** _____ **8 = N/A 9 = Unknown** demrv117	

Notes for coding Vascular dementia: Focal neurological signs suggestive of stroke include: weakness of an extremity, exaggerated DTRs, pseudobulbar palsy, extensor plantar responses, gait abnormalities, hemianopsia, facial weakness, dysarthria and sensory deficit if thought to be of vascular etiology.	
1. Dementia Present (from page 3) **0 = No 1 = Yes 9 = Unknown** demrv118	
2. Clinical Stroke Documented ('definite' stroke at stroke review) **0 = No Stroke 1 = Yes, One Stroke 2 = Yes, More Than One Stroke** **3= Terminal Stroke Only 8 = N/A** demrv119	
3. Suggestive Temporal Profile (onset of dementia less than 3 months after stroke, abrupt onset or fluctuating stepwise decline) **0 = No 1 = Yes 8 = N/A 9 = Unknown** demrv120	
4. Imaging (CT or MRI) demrv121 (code 'atrophy only' or mild white matter changes' as 0) **0 = No Stroke 1 = Yes, One Stroke 2 = Yes, More Than One Stroke** **3 = Yes, Extensive White Matter Changes 8 = N/A 9 = Unknown**	
5. Focal Neurological Signs Suggestive of Stroke demrv122 **0 = No 1 = Yes 8 = N/A 9 = Unknown**	
Vascular Dementia Present (fill out at review only) demrv123 **0 = No 1 = Yes 9 = Unknown**	
If Vascular Dementia Present Code: (fill out at review only) demrv124 **1 = Probable Vascular Dementia:** **Criteria: #1 and #2 and #3 = 1 ('Yes')** **#4 = 1 or 2 ('One or More Strokes')** **Possible Vascular Dementia:** **2 = #1 and #2 = 1 ('Yes')** **#3 = 0 or 9 ('No' or 'Unknown')** **#4 or #5 = 1 ('Yes')** **3 = #1 and #2 and #3 = 1 ('Yes')** **#4 = 0 or 9 ('No Stroke' or 'Unknown')** **4 = #1= 1 ('Yes') and #4= 3 ('Extensive White Matter Changes') + diagnosis of Binswanger's disease** **5= #1= 1 ('Yes') and #4= 1, 2, or 3 and/or #5= 1 ('Yes') Or #1= 1 ('Yes') and #3= 1 ('Yes') and #4= 1, 2, or 3 and/or #5= 1 ('Yes')** **8 = N/A 9 = Unknown**	
Coma/Persistent Vegetative State Post Stroke, Until Death **0 = No 1 = Yes 8 = N/A 9 = Unknown** demrv125	

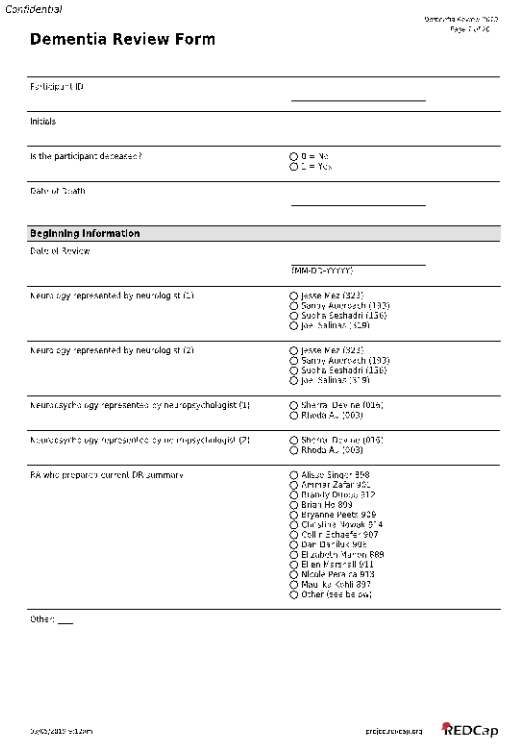
***Other Causes of Dementia or Impairment** demrv126, demrv127, demrv128 **0 = None 1 = PD prior to dementia onset 2 = PD after dementia onset 3 = Dementia with Lewy Bodies** **4 = PSP 5 = Shy-Dager syndrome 6 = Striato-nigral degeneration 7 = FTD with Parkinsonism** **8 = Wilson's disease 9 = FTD (w/ and w/out atrophy on imaging) 10 = Corticobasal ganglionic degeneration** **11 = Huntington's disease 12 = Spino-cerebellar degeneration 13 = Leukodystrophies** **14 = Post cardiac arrest 15 = TBI 16 = Post infectious sequelae (after meningitis, encephalitis, ADEM)** **17 = Malignancy (primary, secondary, para-neoplastic) 18 = Subdural hematoma 19 = NPH 20 = CJD** **21 = Multiple sclerosis 22 = AIDS associated dementias 23 = Other infections (fungal meningitis, syphilis)** **24 = Alcoholic dementia 25 = Toxic-Metabolic Encephalopathy 26 = Dementia – Uncertain Etiology** **27 = Other Etiologies (specify** _____**)**	1. 2. 3.

28 = History of Depression 29 = History of Alcohol/Drug abuse 99 = Unknown or N/A	

Earliest Documented Date of Dementia ("demented by" date) EDDD **(mm/dd/yyyy) 99/99/9999 = N/A or Unknown** Note: if the pt. has a "date of diagnosis of mild dementia," put 9s in the first line and 8s in the remaining lines. If the pt. is missing a "date of diagnosis of mild dementia," put the earliest documented date of dementia in the first line and then complete the rest of the boxes.	drfa_06,drfa_07,drfa_08
Severity: 1.0 = Mild Dementia 1.5 = Greater than or equal to mild dem. 2.0 = Moderate Dementia 2.5 = Greater than or equal to moderate dementia 3.0 = Severe Dementia 8.8= NA 9.9=Unknown (Dementia is present, and the pt. is at least mild, but severity is uncertain)	Drfa_09
Degree of Certainty Regarding Dementia Date **1 = Slightly 2 = Somewhat 3 = Moderately 4 = Reasonably 5 = Highly 8 = N/A**	Drfa_10
Sources supporting earliest documented date of dementia applicable)	**(0 = No; 1 = Yes; 8=Not**
Neuropsychological Testing	Drfa_11
Neurological Examination	Drfa_12
Family Interview Form	Drfa_13
FHS Cycle Exam Records (i.e. MMSE)	Drfa_14
Medical Records (Hospital Records, Nursing Home Notes, etc.)	Drfa_15

Appendix B. REDCap form launched on January 7, 2019

See document entitled "Dementia Review Form - REDCap," located on the N drive in this location: location



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Dementia Review Form

Participant ID

Initials

Is the participant deceased?
○ 0 = No
○ 1 = Yes

Date of Death

Beginning Information

Date of Review
(MM-DD-YYYY)

Neurology represented by neurologist (1)
○ Jesse Mez (823)
○ Sanjay Asthana (193)
○ Sophia Seshadri (136)
○ Joe Salinas (159)

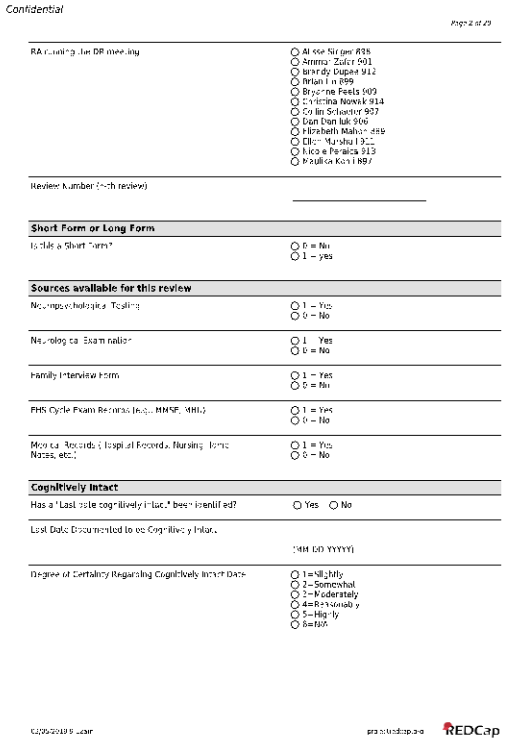
Neurology represented by neurologist (2)
○ Jesse Mez (823)
○ Sanjay Asthana (193)
○ Sophia Seshadri (136)
○ Joe Salinas (159)

Neuropsychology represented by neuropsychologist (1)
○ Sherita Devine (016)
○ Rhoda Au (003)

Neuropsychology represented by neuropsychologist (2)
○ Sherita Devine (016)
○ Rhoda Au (003)

RA who prepared current DR summary
○ Alisse Singer 858
○ Ammar Zafar 901
○ Mandy Dhugal 912
○ Brian Ho 899
○ Bryanne Peets 909
○ Christina Nowak 914
○ Collin Schaefer 907
○ Dan Dan Iuk 906
○ Elizabeth Mahon 888
○ Ellen Mansfield 911
○ Nicole Peracica 913
○ Paulika Kohli 897
○ Other (see below)

Other: ___



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RA completing the DR meeting
○ Alisse Singer 858
○ Ammar Zafar 901
○ Brandy Dupee 912
○ Brian Ho 899
○ Bryanne Peets 909
○ Christina Nowak 914
○ Collin Schaefer 907
○ Dan Dan Iuk 906
○ Elizabeth Mahon 888
○ Ellen Mansfield 911
○ Nicole Peracica 913
○ Paulika Kohli 897

Review Number (each review)

Short Form or Long Form

Is this a Short Form?
○ 0 = No
○ 1 = yes

Sources available for this review

Neuropsychological Testing
○ 1 = Yes
○ 0 = No

Neurological Examination
○ 1 = Yes
○ 0 = No

Family Interview Form
○ 1 = Yes
○ 0 = No

FHS Cycle Exam Records (e.g., MMSE, MHU)
○ 1 = Yes
○ 0 = No

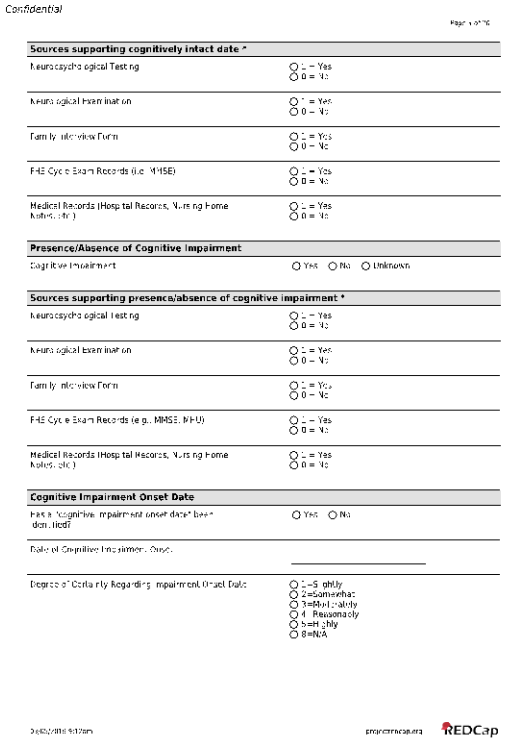
Medical Records (Hospital Records, Nursing Home Notes, etc.)
○ 1 = Yes
○ 0 = No

Cognitively Intact

Has a "Last date cognitively intact" been identified?
○ Yes ○ No

Last Date Documented to be Cognitively Intact
(MM-DD-YYYY)

Degree of Certainty Regarding Cognitively Intact Date
○ 1=Slightly
○ 2=Somewhat
○ 3=Moderately
○ 4=Reasonably
○ 5=Highly
○ 8=N/A



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Sources supporting cognitively intact date *

Neuropsychological Testing
○ 1 = Yes
○ 0 = No

Neurological Examination
○ 1 = Yes
○ 0 = No

Family Interview Form
○ 1 = Yes
○ 0 = No

FHS Cycle Exam Records (i.e. MMSE)
○ 1 = Yes
○ 0 = No

Medical Records (Hospital Records, Nursing Home Notes, etc.)
○ 1 = Yes
○ 0 = No

Presence/Absence of Cognitive Impairment

Cognitive Impairment
○ Yes ○ No ○ Unknown

Sources supporting presence/absence of cognitive impairment *

Neuropsychological Testing
○ 1 = Yes
○ 0 = No

Neurological Examination
○ 1 = Yes
○ 0 = No

Family Interview Form
○ 1 = Yes
○ 0 = No

FHS Cycle Exam Records (e.g., MMSE, MHU)
○ 1 = Yes
○ 0 = No

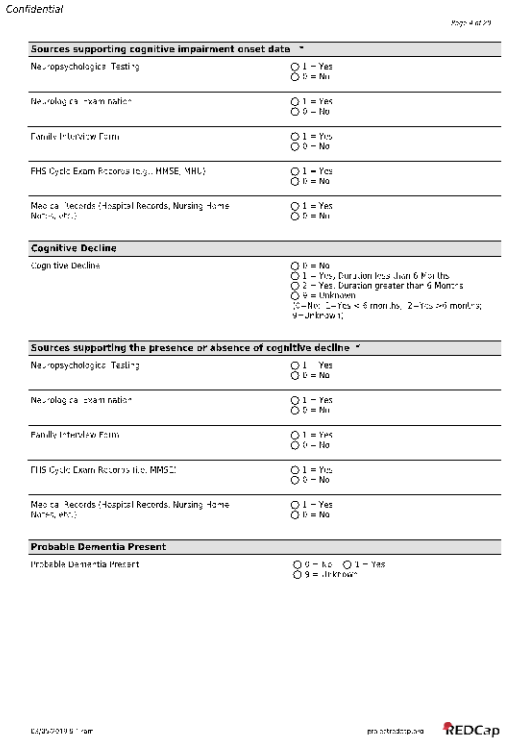
Medical Records (Hospital Records, Nursing Home Notes, etc.)
○ 1 = Yes
○ 0 = No

Cognitive Impairment Onset Date

Has a "cognitive impairment onset date" been identified?
○ Yes ○ No

Date of Cognitive Impairment Onset

Degree of Certainty Regarding Impairment Onset Date
○ 1=Slightly
○ 2=Somewhat
○ 3=Moderately
○ 4=Reasonably
○ 5=Highly
○ 8=N/A



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Sources supporting cognitive impairment onset date *

Neuropsychological Testing
○ 1 = Yes
○ 0 = No

Neurological Examination
○ 1 = Yes
○ 0 = No

Family Interview Form
○ 1 = Yes
○ 0 = No

FHS Cycle Exam Records (e.g., MMSE, MHU)
○ 1 = Yes
○ 0 = No

Medical Records (Hospital Records, Nursing Home Notes, etc.)
○ 1 = Yes
○ 0 = No

Cognitive Decline

Cognitive Decline
○ 0 = No
○ 1 = Yes, Duration less than 6 Months
○ 2 = Yes, Duration greater than 6 Months
○ 9 = Unknown
(0=No, 1=Yes < 6 months, 2=Yes >6 months, 9=Unknown)

Sources supporting the presence or absence of cognitive decline *

Neuropsychological Testing
○ 1 = Yes
○ 0 = No

Neurological Examination
○ 1 = Yes
○ 0 = No

Family Interview Form
○ 1 = Yes
○ 0 = No

FHS Cycle Exam Records (i.e. MMSE)
○ 1 = Yes
○ 0 = No

Medical Records (Hospital Records, Nursing Home Notes, etc.)
○ 1 = Yes
○ 0 = No

Probable Dementia Present

Probable Dementia Present
○ 0 = No ○ 1 = Yes
○ 9 = Unknown

Sources supporting the presence or absence of dementia *	
Neuropsychological Testing	○ 1 = Yes ○ 0 = No
Neurological Examination	○ 1 = Yes ○ 0 = No
Family Interview Form	○ 1 = Yes ○ 0 = No
FHS Cycle Exam Records (i.e. MYSEI	○ 1 = Yes ○ 0 = No
Medical Records (Hospital Records, Nursing Home Notes, etc.)	○ 1 = Yes ○ 0 = No

Mild Dementia	
Has a "Mild Dementia" date been identified?	○ Yes ○ No
Date of Diagnosis of Mild Dementia	(MM-DD-YYYY)
Degree of Certainty Regarding Mild Dementia Date	○ 1=Slightly ○ 2=Somewhat ○ 3=Moderately ○ 4=Reasonably ○ 5=Highly ○ 8=N/A

Sources supporting the date of mild dementia	
Neuropsychological Testing	○ 1 = Yes ○ 0 = No
Neurological Examination	○ 1 = Yes ○ 0 = No
Family Interview Form	○ 1 = Yes ○ 0 = No
FHS Cycle Exam Records (i.e. MYSEI	○ 1 = Yes ○ 0 = No
Medical Records (Hospital Records, Nursing Home Notes, etc.)	○ 1 = Yes ○ 0 = No

Moderate Dementia	
Has a "Moderate Dementia" date been identified?	○ Yes ○ No
Date of Diagnosis of Moderate Dementia	(MM-DD-YYYY)
Degree of Certainty Regarding Moderate Dementia Date	○ 1=Slightly ○ 2=Somewhat ○ 3=Moderately ○ 4=Reasonably ○ 5=Highly ○ 8=N/A

Sources supporting the date of moderate dementia	
Neuropsychological Testing	○ 1 = Yes ○ 0 = No
Neurological Examination	○ 1 = Yes ○ 0 = No
Family Interview Form	○ 1 = Yes ○ 0 = No
FHS Cycle Exam Records (i.e. MYSEI)	○ 1 = Yes ○ 0 = No
Medical Records (Hospital Records, Nursing Home Notes, etc.)	○ 1 = Yes ○ 0 = No

Severe Dementia	
Has a "Severe Dementia" date been identified?	○ Yes ○ No
Date of Diagnosis of Severe Dementia	
Degree of Certainty Regarding Severe Dementia Date	○ 1=Slightly ○ 2=Somewhat ○ 3=Moderately ○ 4=Reasonably ○ 5=Highly ○ 8=N/A

Sources supporting the date of severe dementia	
Neuropsychological Testing	○ 1 = Yes ○ 0 = No
Neurological Examination	○ 1 = Yes ○ 0 = No
Family Interview Form	○ 1 = Yes ○ 0 = No

FHS Cycle Exam Records (i.e. MYSEI	○ 1 = Yes ○ 0 = No
Medical Records (Hospital Records, Nursing Home Notes, etc.)	○ 1 = Yes ○ 0 = No

Stroke/PD	
Definite Stroke or TIA from Stroke Review	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Was it TIA only (i.e., no stroke)?	○ 0 = not TIA only, a stroke WAS identified ○ 1 = yes, TIA only
Parkinson's Disease	○ 1 = Yes ○ 0 = No ○ 9 = Unknown

CT Scan Information	
CT Scan Performed	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Date of the Most Recent CT Scan	(99-99-9999 if unknown)
CT Scan Results (Primary)	○ 1 = Normal ○ 2 = Atrophy only ○ 3 = Single Stroke ○ 4 = Two or more Strokes ○ 5 = Other, ____ (if yes, fill box below) ○ 6 = Small Vessel Ischemic disease ○ 9 = Unknown
CT Scan Results (Primary) = "Other, ____"	(Describe findings)
Are there other CT results to note?	○ 0 = no ○ 1 = yes
Other Notable CT Scan results	☐ Age-related (mild) atrophy ☐ Age-related (mild) white matter changes ☐ Atrophy (not noted to be age related) ☐ Atrophy (moderate) ☐ Atrophy (extensive) ☐ White matter changes (not noted to be age-related) ☐ Patchy white matter changes ☐ Moderate white matter changes ☐ Extensive white matter changes ☐ Old stroke(s) ☐ Silent stroke(s) ☐ Other notable findings

Other notable CT findings - Type:	☐ 0 = Atrophy ☐ 1 = White matter changes ☐ 2 = Old stroke ☐ 3 = Silent Stroke ☐ 4 = Other
Other notable CT findings - Age-related: Atrophy	○ 0 = No, not described as age-related ○ 1 = Yes, described as age-related (or inferred to be age-related, such as described as of abnormalities but noted to be normal)
Other notable CT findings - Severity: Atrophy	○ 0 = Mild (for age-related) ○ 1 = Mild to Moderate ○ 2 = Moderate ○ 3 = Moderate to Severe ○ 4 = Severe ○ 5 = Extensive ○ 6 = Not described
Other notable CT findings - Age-related: White Matter Changes	○ 0 = No, not described as age-related ○ 1 = Yes, described as age-related (or inferred to be age-related, such as described as of abnormalities but noted to be normal)
Other notable CT findings - Severity: White Matter Changes	○ 0 = Mild (for age-related) ○ 1 = Mild to Moderate ○ 2 = Moderate ○ 3 = Moderate to Severe ○ 4 = Severe ○ 5 = Extensive ○ 6 = Not described
Other notable CT findings - Quantity of strokes	○ 0 = One stroke ○ 1 = Multiple strokes
Additional Other Notable CT results "Other ___"	

MRI Scan Information	
MRI Scan Performed	○ 1 = Yes ○ 0 = No ○ 9 = unknown
Date of the Most Recent MRI Scan	(99-99-9999 if unknown)
MRI Scan Results (Primary)	○ 1 = Normal ○ 2 = Atrophy only ○ 3 = Single Stroke ○ 4 = Two or more Strokes ○ 5 = Other, (if yes, fill box below) ○ 6 = Small Vessel Ischemic disease ○ 9 = Unknown
MRI Scan Results (Primary) = "Other, ____"	(Describe findings)

Are there other MRI results to note?	○ 0 = no ○ 1 = yes
Other Notable MRI Scan results	☐ Age related atrophy ☐ Age-related white matter changes ☐ Atrophy (not noted to be age related) ☐ Atrophy (moderate) ☐ Atrophy (extensive) ☐ White matter changes (not noted to be age-related) ☐ Patchy white matter changes ☐ Moderate white matter changes ☐ Extensive white matter changes ☐ Old stroke(s) ☐ Silent stroke(s) ☐ Other notable findings
Other notable MRI findings - Type	☐ 0 = Atrophy ☐ 1 = White matter changes ☐ 2 = Old stroke ☐ 3 = Silent Stroke ☐ 4 = Other
Other notable MRI findings - Age-related: Atrophy	○ 0 = No, not described as age-related ○ 1 = Yes, described as age-related (or inferred to be age-related, such as description of abnormalities out-of-proportion to normal)
Other notable MRI findings - Severity: Atrophy	○ 0 = Mild (or age-related) ○ 1 = Mild to Moderate ○ 2 = Moderate ○ 3 = Moderate to Severe ○ 4 = Severe ○ 5 = Extensive ○ 6 = Not described
Other notable MRI findings - Age-related: White Matter Changes	○ 0 = No, not described as age-related ○ 1 = Yes, described as age-related (or inferred to be age-related, such as description of abnormalities out-of-proportion to normal)
Other notable MRI findings - Severity: White Matter Changes	○ 0 = Mild (or age-related) ○ 1 = Mild to Moderate ○ 2 = Moderate ○ 3 = Moderate to Severe ○ 4 = Severe ○ 5 = Extensive ○ 6 = Not described
Other notable MRI findings - Quantity of strokes	○ 0 = One stroke ○ 1 = Multiple strokes
Additional Other Notable MRI results = "Other"	_____

Brain Autopsy Information	
Brain Bank Subject?	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Brain Autopsy Performed?	○ 1 = Yes ○ 0 = No ○ 9 = Unknown (Leave blank if no this autopsy and no sign of outside autopsy)
Brain Autopsy Report Available?	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Is this review being done after the neuropath conference?	○ 0 = No ○ 1 = Yes
Hachinski/Blessed	
Was the Hachinski score computed?	○ Yes ○ No
Hachinski Ischemia Score	_____
Was the Blessed score computed?	○ Yes ○ No
Blessed Score	_____
Cognition at Death **	
Cognitive Status at Time of Death	○ 0 = No Dementia (0) ○ .5 = Cognitive Impairment-No Dementia (0.5) ○ 1 = Mild Dementia (1) ○ 1.5 = Greater than or equal to mild dementia (1.5) ○ 2 = Moderate Dementia (2) ○ 2.5 = Greater than or equal to moderate dementia (2.5) ○ 3 = Severe Dementia (3) ○ 4 = Alive (4) ○ 9 = Dead, unknown status at death (**A so code "7" as 9)
Certainty of Cognitive Status at Death	○ 1=Slightly ○ 2 = Somewhat ○ 3=Moderately ○ 4=Reasonably ○ 5=Highly ○ 8=N/A

Sources supporting the cognitive status at death	
Neuropsychological Testing	○ 1 = Yes ○ 0 = No
Neurological Examination	○ 1 = Yes ○ 0 = No
Family Interview Form	○ 1 = Yes ○ 0 = No
CTFS Cycle Exam Records (i.e. MMSE)	○ 1 = Yes ○ 0 = No
Medical Records (Hospital Records, Nursing Home Notes, etc.)	○ 1 = Yes ○ 0 = No
Subtypes (from here onward, fill out at review only)	
NOTE: Probable AD + Probable VaD = Mixed dementia (4) Possible AD + Probable VaD = Mixed dementia (4) Probable AD + Possible VaD = AD w/out stroke (1) OR AD w/Stroke (2)	
Dementia Subtype	○ 0 = None ○ 1 = Alzheimer's Disease Without Stroke ○ 2 = Alzheimer's Disease With Stroke ○ 3 = Vascular Dementia Without Alzheimer's Disease ○ 4 = Mixed Dementia Type (Alzheimer's Disease + Vascular Dementia) ○ 5 = Frontotemporal Dementia ○ 6 = Dementia with Lewy Bodies ○ 7 = Dementia that does not fit any other Category (progressive) (If yes, fill box below) ○ 8 = Dementia that does not fit any other Category (non-progressive) (If yes, fill box below) ○ 9 = Cognitive Impairment, No Dementia (MCI) ○ 10 = Dementia present - Uncertain Etiology(10) ○ 99 = Unknown (99)
If yes to "Dementia that does not fit any other Category" progressive or non-progressive, specify:	
Severity of Dementia Subtype	○ 0=None ○ 1=Mild ○ 2=Moderate ○ 3=Severe ○ 9=Unknown

Features of MCI Stage	
******************** Impairment onset date is not known, so MCI boxes cannot be completed ********************	
Subtype during the MCI stage	○ 1 = Amnestic ○ 2 = Non-Amnestic ○ 8 = N/A ○ 9 = Unknown (1=Amnestic, 2=Non Amnestic, 8=N/A, 9=Unknown)
If Amnestic, Code Subtype	○ 1 = Amnestic Only ○ 2 = Amnestic Plus ○ 8 = N/A ○ 9 = Unknown (1=Amnestic Only 2=Amnestic Plus 8=N/A 9=Unknown)
Executive Function	○ 1 = Yes ○ 0 = No ○ 9 = unknown
Abstract reasoning	○ 1 = Yes ○ 0 = No ○ 9 = unknown
Visuospatial function	○ 1 = Yes ○ 0 = No ○ 9 = unknown
Language	○ 1 = Yes ○ 0 = No ○ 9 = unknown
Attention	○ 1 = Yes ○ 0 = No ○ 9 = unknown
If Non-Amnestic, Code Subtype	○ 1 = single domain ○ 2 = multiple domain ○ 8 = N/A ○ 9 = unknown (1=single domain, 2=multiple, 8=N/A, 9=unknown)
Executive Function	○ 1 = Yes ○ 0 = No ○ 9 = unknown
Abstract Reasoning	○ 1 = Yes ○ 0 = No ○ 9 = unknown
Visuospatial Function	○ 1 = Yes ○ 0 = No ○ 9 = unknown
Language	○ 1 = Yes ○ 0 = No ○ 9 = unknown
Attention	○ 1 = Yes ○ 0 = No ○ 9 = unknown

Criteria for DSM-IV

Memory Impairment	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
If Memory Impairment, Code Subtype:	○ 1=Verbal and Non-verbal memory ○ 2=Verbal only ○ 3=Non-verbal only ○ 8=N/A ○ 9=Unknown
Verbal Memory Impairment?	○ 1 = Yes ○ 0 = No ○ 9 = Unknown (yes/no/unknown)
Nonverbal Memory Impairment?	○ 1 = Yes ○ 0 = No ○ 9 = Unknown (yes/no/unknown)
Aphasia	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Apraxia	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Agnosia	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Executive Dysfunction (planning, organizing, sequencing, abstracting)	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Impaired Abstraction	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Significant Impairment in Function (Social/Occupational)	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Dementia by DSM-IV Criteria: --Memory Impairment Impairment in one other Cognitive Domain --Functional Decline Not Due to Delirium, Depression, or Schizophrenia	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Dementia by ADDTC Criteria: --Impairment in two or more Cognitive Domains --Functional Decline secondary to Cognitive Impairment	○ 1 = Yes ○ 0 = No ○ 9 = Unknown

Symptoms above present for at least 6 months (refers to memory impairment, aphasia, etc; NOT dementia)	○ 1 = Yes ○ 0 = No ○ 9 = Unknown (yes/no/unknown)

Cognitive Deficits Not Related to DSM-IV Criteria

Language	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Visuospatial Abilities	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Attention	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Alzheimer's Disease by NINCDS-ADRADA Criteria	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Classification of Alzheimer's Disease (note that, in certain cases, a pt. can have both probable AD and probable vascular dementia)	○ 1 = Probable AD (dementia, progression, no other etiology) ○ 2 = Possible AD (dementia, progression, unusual clinical features or other contributory etiologies) ○ 3 = Definite AD ○ 8 = N/A ○ 9 = Unknown
If Possible AD, Code Subtype	○ 1 = Mixed AD + Vascular ○ 2 = Mixed AD + Parkinsonism (including drug-induced) ○ 3 = Mixed AD + Other (if yes, fill box below) ○ 8 = N/A ○ 9 = Unknown
If yes to for Mixed AD + Other, Specify	_____

Vascular Dementia Questions (Ignore if completing the Short Form)

Dementia Present:

Clinical Stroke Documented (definite stroke at stroke review)	○ 0 = No Stroke ○ 1 = Yes, One Stroke ○ 2 = Yes, More Than One Stroke ○ 3 = Terminal Stroke Only ○ 8 = N/A
Suggestive Temporal Profile (onset of dementia less than 3 months after stroke, abrupt onset or fluctuating stepwise decline)	○ 1 = Yes ○ 0 = No ○ 9 = Unknown

Imaging (CT or MRI)	○ 0 = No stroke OR atrophy only OR mild white matter changes ○ 1 = Yes, One Stroke ○ 2 = Yes, More Than One Stroke ○ 3 = Yes, Extensive White Matter Changes ○ 8 = N/A ○ 9 = Unknown
Focal Neurological Signs Suggestive of Stroke	○ 1 = Yes ○ 0 = No ○ 9 = Unknown
Focal neurological signs include: • Weakness of an extremity • Exaggerated DTRs • Pseudobulbar palsy • Extensor plantar responses • Gait abnormalities • Hemiparesis • Facial weakness • Dysarthria • Sensory deficit, although, to be of vascular etiology	
Vascular Dementia subtype = 1?	_____ (1 = YES, 0 = NO)
Vascular Dementia subtype = 2?	_____ (1 = YES, 0 = NO)
Vascular Dementia subtype = 3?	_____ (1 = YES, 0 = NO)
Vascular Dementia subtype = 4? (**Note: in addition to a value of '1,' there must be a diagnosis of Binswanger's disease)	_____ (1 = YES, 0 = NO)
Vascular Dementia subtype = 5?	_____ (1 = YES, 0 = NO)
Vascular Dementia Present	○ 1 = Yes ○ 0 = No ○ 9 = Unknown

If yes to Vascular Dementia Present, code	○ 1 = Probable (dementia present + clinical stroke documented + temporal profile + imaging 1 or more strokes) ○ 2 = Possible (dementia present + clinical stroke documented + no or Unknown temporal profile + imaging one stroke OR focal neurological signs suggestive of stroke) ○ 3 = Possible (dementia present + clinical stroke documented + temporal profile + imaging no stroke or unknown) ○ 4 = Possible (dementia present + imaging extensive white matter changes – diagnosis of Binswanger's disease) ○ 5 = Possible (dementia present + [imaging 1 or more strokes OR extensive WM changes] AND/OR focal neurological signs), with or without temporal profile ○ 9 = Unknown
Coma/Persistent Vegetative State Post Stroke, Until Death	○ 1 = Yes ○ 0 = No ○ 9 = Unknown

Other Causes of Dementia or Impairment

This part clean: Has PD prior to cognitive impairment, please code as 'PD prior to Dementia' in 'Other Causes of Dementia or Impairment' field

Other Causes of Dementia or Impairment

Other Causes of Dementia or Impairment	○ 0 = None ○ 1 = PD prior to dementia onset ○ 2 =2PD after dementia onset ○ 3 = Dementia with Lewy Bodies ○ 4 = PSP ○ 5 = Shy-Drager syndrome ○ 6 = Strato-nigral degeneration ○ 7 = FTD with Parkinsonism ○ 8 = Wilson's Disease ○ 9 = FTD (w/ and w/out atrophy on Imaging) ○ 10 = Corticobasal ganglionic degeneration ○ 11 = Huntington's disease ○ 12 = Spino-cerebellar degeneration ○ 13 = Leukodystrophies ○ 14 = Post cardiac arrest ○ 15 = TBI ○ 16 = Post infectious sequelae (after meningitis, encephalitis, ADEM) ○ 17 = Malignancy (primary, secondary, para-neoplastic) ○ 18 = Subdural hematoma ○ 19 = NPH ○ 20 = CJD ○ 21 = Multiple sclerosis ○ 22 = AIDS associated dementias ○ 23 = Other infections (fungal meningitis, syphilis) ○ 24 = Alcoholic dementia ○ 25 = Toxic-Metabolic Encephalopathy ○ 26 = Dementia-Uncertain Etiology ○ 27 = Other Etiologies (if yes, fill box below) ○ 28 = History of Depression ○ 29 = History of Alcohol, Drug abuse ○ 99 = Unknown or N/A

Other Causes of Dementia or Impairment	○ 0 = None ○ 1 = PD prior to dementia onset ○ 2 =2PD after dementia onset ○ 3 = Dementia with Lewy Bodies ○ 4 = PSP ○ 5 = Shy-Drager syndrome ○ 6 = Strato-nigral degeneration ○ 7 = FTD with Parkinsonism ○ 8 = Wilson's Disease ○ 9 = FTD (w/ and w/out atrophy on Imaging) ○ 10 = Corticobasal ganglionic degeneration ○ 11 = Huntington's disease ○ 12 = Spino-cerebellar degeneration ○ 13 = Leukodystrophies ○ 14 = Post cardiac arrest ○ 15 = TBI ○ 16 = Post infectious sequelae (after meningitis, encephalitis, ADEM) ○ 17 = Malignancy (primary, secondary, para-neoplastic) ○ 18 = Subdural hematoma ○ 19 = NPH ○ 20 = CJD ○ 21 = Multiple sclerosis ○ 22 = AIDS associated dementias ○ 23 = Other infections (fungal meningitis, syphilis) ○ 24 = Alcoholic dementia ○ 25 = Toxic-Metabolic Encephalopathy ○ 26 = Dementia-Uncertain Etiology ○ 27 = Other Etiologies (if yes, fill box below) ○ 28 = History of Depression ○ 29 = History of Alcohol, Drug abuse ○ 99 = Unknown or N/A

Other Causes of Dementia or Impairment	○ 0 = None ○ 1 = PD prior to dementia onset ○ 2 =2PD after dementia onset ○ 3 = Dementia with Lewy Bodies ○ 4 = PSP ○ 5 = Shy-Drager syndrome ○ 6 = Strato-nigral degeneration ○ 7 = FTD with Parkinsonism ○ 8 = Wilson's Disease ○ 9 = FTD (w/ and w/out atrophy on Imaging) ○ 10 = Corticobasal ganglionic degeneration ○ 11 = Huntington's disease ○ 12 = Spino-cerebellar degeneration ○ 13 = Leukodystrophies ○ 14 = Post cardiac arrest ○ 15 = TBI ○ 16 = Post infectious sequelae (after meningitis, encephalitis, ADEM) ○ 17 = Malignancy (primary, secondary, para-neoplastic) ○ 18 = Subdural hematoma ○ 19 = NPH ○ 20 = CJD ○ 21 = Multiple sclerosis ○ 22 = AIDS associated dementias ○ 23 = Other infections (fungal meningitis, syphilis) ○ 24 = Alcoholic dementia ○ 25 = Toxic-Metabolic Encephalopathy ○ 26 = Dementia-Uncertain Etiology ○ 27 = Other Etiologies (if yes, fill box below) ○ 28 = History of Depression ○ 29 = History of Alcohol, Drug abuse ○ 99 = Unknown or N/A
If Yes for Other Etiologies, Specify:	_____

Earliest Documented Date of Dementia

Earliest Documented Date of Dementia	_____
Severity	○ 1.0 = Mild Dementia ○ 1.5 = Greater than or equal to mild dementia ○ 2.0 = Moderate Dementia ○ 2.5 = Greater than or equal to moderate dementia ○ 3.0 = Severe Dementia ○ 8.8 = N/A ○ 9.9 = Unknown (Dementia is present, and the pt. is at least mild, but severity is uncertain)
Degree of Certainty Regarding Dement a Date	○ 1=Slightly ○ 2 = Somewhat ○ 3 = Moderately ○ 4 = Reasonably ○ 5 = Highly ○ 8 = N/A

Sources supporting earliest documented date of dementia

Neuropsychological Testing	○ 1 = Yes ○ 0 = No
Neurological Examination	○ 1 = Yes ○ 0 = No
Family Interview Form	○ 1 = Yes ○ 0 = No
FHS Cycle Exam Records (i.e. MMSE)	○ 1 = Yes ○ 0 = No
Medical Records (Hospital Records, Nursing Home Notes, etc.)	○ 1 = Yes ○ 0 = No

Developmental Disorder Suspected?

Is a Developmental Disorder (e.g., Intellectual disorder, learning disorder) suspected, or does limited education possibly explain impairment?	○ Yes ○ No

Comments

Comments	
Date Form Completed	_____
Person Completing DR Form	_____

Appendix C. Clinical Dementia Rating (CDR) form on REDCap

Confidential

Clinical Dementia Rating

Participant ID	_____
Participant ID	_____ (e.g., "3-98" entered as "30098")
Initials	_____
Date of Dementia Review	_____
Memory Domain CDR Score	◯ 0 ◯ .5 ◯ 1 ◯ 2 ◯ 3
Orientation Domain CDR Score	◯ 0 ◯ .5 ◯ 1 ◯ 2 ◯ 3
Judgment & Problem Solving Domain CDR Score	◯ 0 ◯ .5 ◯ 1 ◯ 2 ◯ 3
Community Affairs Domain CDR Score	◯ 0 ◯ .5 ◯ 1 ◯ 2 ◯ 3
Home & Hobbies Domain CDR Score	◯ 0 ◯ .5 ◯ 1 ◯ 2 ◯ 3
Personal Care Domain CDR Score	◯ 0 ◯ 1 ◯ 2 ◯ 3
Computed Global CDR Score	◯ 0 ◯ .5 ◯ 1 ◯ 2 ◯ 3
Date form completed	_____
Person completing form	_____

Appendix D. DSM-IV Criteria for Dementia

A. Development of multiple cognitive deficits that include memory impairment and at least one of the following cognitive disturbances
    a. Aphasia
    b. Apraxia
    c. Agnosia
    d. Disturbance in executive functioning
B. The cognitive deficits are sufficiently severe to cause impairment in occupational or social functioning.
C. The cognitive deficits must represent a decline from a previously higher level of functioning.

DSM-IV Criteria for the Diagnosis of Alzheimer's Disease

A. The development of multiple cognitive deficits manifested by both:
    1. Memory impairment (impaired ability to learn new information or to recall previously learned information).
    2. One (or more) of the following cognitive disturbances:
    a. Aphasia (language disturbance)
    b. Apraxia (impaired ability to carry out motor activities despite intact motor function
    c. Agnosia (failure to recognize or identify objects despite intact sensory function)
    d. Disturbance in executive functioning (i.e., planning, organizing, sequencing, abstracting)
B. The cognitive deficits in Criteria A1 and A2 each cause significant impairment in social or occupational functioning and represent a significant decline from a previous level of functioning.
C. The course is characterized by gradual onset and continuing cognitive decline.
D. The cognitive deficits in Criteria A1 and A2 are not due to any of the following:
    1. Other central nervous systems, conditions that cause progressive deficits in memory and cognition (e.g., cerebrovascular disease, Parkinson's disease, Huntington's disease, subdural hematoma, normal-pressure hydrocephalus, brain tumor)
    2. 2. Systemic conditions that are known to cause dementia (e.g., hypothyroidism, vitamin B12 or folic acid deficiency, neurosyphilis, HIV infection)
    3. Substance-induced conditions
E. The deficits do not occur exclusively during the course of a delirium.
F. The disturbance is not better accounted for by another disorder (e.g., major depressive disorder, schizophrenia).

DSM-IV criteria for the diagnosis of vascular dementia

A. The development of multiple cognitive deficits manifested by both:
1. Memory impairment (impaired ability to learn new information or to recall previously learned information)
2. One or more of the following cognitive disturbances:
(a) aphasia (language disturbance)
(b) apraxia (impaired ability to carry out motor activities despite intact motor function)
c) agnosia (failure to recognize or identify objects despite intact sensory function)
(d) disturbance in executive functioning (i.e., planning, organizing, sequencing, abstracting)

B. The cognitive deficits in criteria A1 and A2 each cause significant impairment in social or occupational functioning and represent a significant decline from a previous level of functioning.

C. Focal neurological signs and symptoms (e.g., exaggeration of deep tendon reflexes, extensor plantar response, pseudobulbar palsy, gait abnormalities, weakness of an extremity) or laboratory evidence indicative of cerebrovascular disease (e.g., multiple infarctions involving cortex and underlying white matter) that are judged to be etiologically related to the disturbance.

D. The deficits do not occur exclusively during the course of a delirium.

Appendix E. Alzheimer's Disease Diagnostic and Treatment Centers (ADDTC) definition of dementia

Dementia is a **deterioration** from a known or estimated prior level of **intellectual function** sufficient to interfere broadly with the conduct of the patient's **customary affairs of life,** which is **not isolated to a single narrow category of intellectual performance**, and which is **independent of level of consciousness.** This deterioration should be supported by **historical evidence** and documented by either bedside mental status testing or, ideally, by more detailed neuropsychological examination, using tests that are quantifiable, reproducible, and for which normative data are available.

Deterioration: Ideally, cognitive decline should be assessed using tests that are quantifiable, reproducible, and for which normative data are available**.** However, statistically derived cutoff scores should not be included in the definition of dementia because: (1) age-, education-, and gender-specific norms are not available for many cognitive functions in older populations; (2) standard deviation cutoffs may be inappropriate because many test scores used to assess dementia do not have a normal distribution; and (3) statistical cutoffs are inconsistent with the core meaning of dementia as a decline in an *individual's* mentation, because they establish arbitrary population standards to be applied to individuals with widely varying baseline function. Intellectual loss should remain a clinical decision.

Patient's customary affairs of life: Change is in functional impairment in *intellectual* activities of daily living rather than in social or occupational functioning. The degree of intellectual deterioration must be sufficient to interfere with the conduct of the patient's customary affairs of life.

Not isolated by a single narrow category: There is a significant gap between mental status testing and both the biologic status of the CNS and the intellectual activities of daily living[1]; thus,
the *number* or *type* of cognitive deficits is not specified. However, most patients with dementia will exhibit deficits on more than one type of intellectual task, and a distinction must be retained between the patient who has an isolated impairment such as aphasia as opposed to the broader intellectual loss connoted by dementia.

Independent of level of consciousness: In general, a diagnosis of dementia should not be made when there is clouding of consciousness (e.g., recent stroke). On the other hand, the presence of clouding of consciousness does not necessarily preclude a diagnosis of dementia. The important issue for a diagnosis of dementia is the ability to establish that the mental deterioration is not due to impairment of consciousness.

Historical evidence: Given the limits of mental status testing,[1] both historical evidence and clinical judgment should be considered in making a diagnosis of dementia.

[1]Some definitions of dementia require deficits in more than one "area of cognition." Problems with this include: (1) these so-called areas of cognition (such as attention, concentration, memory, language, and visual-spatial functions) are theoretical constructs that help us to conceptualize brain function but have imperfect biologic validity; (2) while assessment of these areas of cognition is conventionally based upon neuropsychological testing, there is no a priori basis and no universal system by which performance on given tasks can be specifically attributed to such discrete cognitive domains; and (3) in mental status testing, artificial challenges are substituted for actual intellectual activities of daily living; yet again, there is no one-to-one correlation between performance on such tasks and real-life intellectual function. Thus, criteria that attempt to operationalize the definition of dementia by specifying the number and types of cognitive or neuropsychological

deficits may actually sacrifice the essential meaning of a decline in mental status in favor of an arbitrary number of deficits identified in artificially delineated areas of cognition by insufficiently specific tests.

Appendix F. Staging Dementia

Dementia is a continuum, and there are no clear demarcations between different stages. In addition, some people may experience some symptoms earlier or later than is "typical." There is no universally accepted system for staging dementia, although the CDR is in widespread use, and can help to serve as a guideline. In addition to the CDR descriptions (Appendix M), the following features common for each stage of dementia may be useful in making determinations. It is important to note that none of these symptoms inevitably show up in the stage under which they are listed.

MCI

- Consistent slight forgetfulness, partial recollection of events
- Fully oriented except for slight difficulty with time relationships
- Slight impairments in solving problems and finding similarities
- Slight or no impairment in community affairs, home, or hobbies – some iADLs are more difficult – they take longer or more mistakes are made
- No impairments in activities of daily living (ADLs) or personal care

Mild Dementia

May…

- have moderate memory loss, more marked for recent events; deficit has begun to interfere with everyday activities
- have moderate difficulty with time relationships; oriented for place at examination but may have geographic disorientation elsewhere.
- have moderate difficulty in handling problems and similarities, but social judgment is usually maintained.
- be unable to function independently in community affairs such as shopping, finances or social groups, although they may still be involved and seem normal to casual inspection.
- there is mild but definite impairment of function at home, more difficult chores are abandoned, as well as difficult hobbies and interests.
- require prompting for personal care
- still be working and driving, but starting to have difficulty (making mistakes, getting confused)
- have decline in any cognitive domain (typical presentation varies with etiology), e.g.,
    - memory impairment (AD)
    - trouble thinking, organizing, planning (vascular)
    - fluctuations in attention (DLB)
    - behavior problems, such as being impulsive, rude, compulsive (bvFTD)
    - difficulty with speech and language (PPA)
        - non-fluent/agrammatic variant (nfaPPA)
        - semantic variant (svPPA)
        - logopenic variant (lvPPA)
- have decreased ability to perform complex tasks (e.g., paying bills, following a recipe)
- have subtle personality changes
- forget something they've just read or just been told
- misplace or lose important items
- repeat something they have just said (without awareness)
- have more pronounced difficulty coming up with words/names

- display problems with organization and planning
- try to hide symptoms

Moderate Dementia

May…

- have severe memory loss, only highly learned material are retained, new material is rapidly lost
- have severe difficulty with time relationships, usually disoriented to time and often place
- have severe impairment in handling problems and similarities, social judgment is usually impaired
- be unable to function independently outside the home, although they may be well enough to be taken to functions outside the family home
- only be able to do simple chores, there are very restricted interests that are poorly sustained
- require assistance in dressing, hygiene, and keeping of personal effects
- need some help with self-care or ADLs (e.g., dressing, bathing, eating)
- have personality and/or mood changes (e.g., more irritable, uncharacteristically refusing to do something, withdrawn)
- experience paranoia, confusion, or fear
- engage in repetitive/compulsive behaviors
- forget their address or other personal information (e.g., phone number)
- forget events or personal history
- have difficulty expressing self (e.g., confusing words, not making logical sense)
- have changes in sleep patterns (e.g., sleeping during the day, agitation at night)
- wander and have trouble getting home
- have delusions (e.g., someone is stealing from them, their home is not really their home)
- disorientation to place and/or time
- have trouble with bladder/bowel control

Severe Dementia

May…

- have severe memory loss, only fragments remain
- not be oriented at all or only to person
- be unable to make judgments or solve problems
- be unable to function independently outside the home, and they are too ill to be taken to functions outside a family home
- have no significant functioning inside the home, no activities or hobbies
- be unable to perform ADLs without significant assistance
- have difficulty chewing and swallowing
- only be able to perform very simple tasks
- not recognize family members, or confuse who is who
- be unable to have a conversation
- have limited or no speech
- be unable to communicate needs
- not understand what others are saying to them
- perseverate in expressing words or sounds
- have severe memory loss; be unable to recall recent events (e.g., what they had for last meal); be able to recall only fleeting (if any) personal history

- believe they are living in the past (e.g., going to school, in the service, living with parents)
- display restlessness (e.g., pacing, fidgeting)
- wring their hands, pick at something, pull at their clothes, touch themselves inappropriately in public
- display repetitive utterances, activities, gestures
- hallucinate
- have bladder and bowel incontinence
- be unable to control body movements (e.g., walk, sit up, move in bed)
- have significant behavior changes (e.g., agitation, aggressiveness, sundowning)
- need full-time supervision
- be unresponsive to environment
- have moments of lucidity

Appendix G. Hachinski Ischemia Scale

Table 1. Hachinski Ischemia Score

Abrupt onset*	2
Stepwise progression*†	1
Fluctuating course†‡	2
Nocturnal confusion‡	1
Relative preservation of personality†	1
Depression	1
Somatic complaints*	1
Emotional incontinence*†	1
History of hypertension*	1
History of strokes*†	2
Evidence of associated atherosclerosis	1
Focal neurologic symptoms*†‡	2
Focal neurologic signs*	2

* Items significantly more common in MID than AD.[41]
† Items significantly more common in MID than AD.[42]
‡ Items that explained a significance portion of the variance in logistic regression.[42]

Appendix H. Blessed Dementia Scale

Changes in performance of everyday activities (assign 1 point if true)

- **Inability to perform household tasks**
- **Inability to cope with small sums of money**
- **Inability to remember shortlist of items; for example, in shopping list**
- **Inability to find way about indoors**
- **Inability to find way about familiar streets**
- **Inability to interpret surroundings; for example, to recognize whether in hospital or at home; to discriminate between patients, doctors, nurse, relatives, other hospital staff, etc.**
- **Inability to recall recent events; for example, recent outings, visits of relatives or friends to hospital, etc.**
- **Tendency to dwell in the past**

Changes in habits (assign appropriate points)

- **Eating**
  (0) = cleanly, with proper utensils
  (1) = messily, with spoon only
  (2) = simple solids (for example, biscuits)
  (3) = has to be fed
- **Dressing**
  (0) = unaided
  (1) = occasionally misplaced buttons, etc.
  (2) = wrong sequence, commonly forgetting items
  (3) = unable to dress
- **Sphincter control**
  (0) = complete control
  (1) = occasional wet bed
  (2) = frequent wet bed
  (3) = doubly incontinent

Changes in personality, interests, and drive (assign 1 point if true)

- **Increased rigidity**
- **Increased egocentricity**
- **Impairment of regard of feeling for others**
- **Coarsening of affect**
- **Impairment of emotional control (for example, increased petulance and irritability)**
- **Hilarity in inappropriate situations**
- **Diminished emotional responsiveness**
- **Sexual misdemeanour (arising de novo in old age)**
- **Hobbies relenquised**
- **Diminished initiative or growing apathy**
- **Purposeless hyperactivity**

Appendix I. National Institute of Neurological and Communicative Disorders and Stroke (NINCDS) and the Alzheimer's Disease and Related Disorders Association (ADRDA) NINCDS-ADRDA Update (2011)

All-Cause Dementia

1. Interferes with the functioning
2. Decline from previous levels of functioning
3. Not explained by delirium or major psychiatric disorder
4. The cognitive or behavioral impairment involves a minimum of two of the following domains:
    a. Memory (Ability to acquire and remember new information)
    b. Reasoning and handling of complex tasks, poor judgment
    c. Visuospatial abilities—symptoms include
    d. Language functions (speaking, reading, writing)
    e. Personality, behavior, or comportment

Probable AD dementia

1. Insidious onset (months to years, not hours to days)
2. The initial and most prominent cognitive deficits are evident on history and examination in one of the following categories.
    - Amnestic presentation
        - Impairment in learning and recall of recently learned information
    - Nonamnestic presentations (Most prominent deficits are…):
        - **Language presentation**: Word-finding
        - **Visuospatial presentation**: Spatial cognition -
        - **Executive dysfunction**: Reasoning, judgment, and problem solving.
3. The diagnosis of **probable AD dementia should NOT** be applied when is evidence of
    - Substantial CVD (stroke temporally related, multiple/extensive infarcts or severe WMH
    - Core features of DLB other than dementia itself
    - Prominent features of behavioral variant FTD
    - Prominent features of semantic variant PPA or non- fluent/agrammatic variant PPA
    - Evidence for another concurrent, active neurological disease, or a non-neurological medical comorbidity or use of meds that could have a substantial effect on cognition

Possible AD dementia: Core clinical criteria

1. Atypical course
    a. Sudden onset of cognitive impairment or
    b. Insufficient historical detail or objective cog. documentation of progressive decline
2. Etiologically mixed presentation
    a. Concomitant CVD
    b. Features of DLB
    c. Other neuro/medical comorbidity or meds

Dementia unlikely to be due to AD

1. Does not meet clinical criteria for AD dementia.

2. Sufficient evidence for an alternative diagnosis such as HIV dementia, dementia of Huntington's disease, or others that rarely, if ever, overlap with AD

Appendix J. Criteria for the Diagnosis of Ischemic Vascular Dementia

Table 2. Criteria for the diagnosis of ischemic vascular dementia (IVD)

I. Dementia

Dementia is a deterioration from a known or estimated prior level of intellectual function sufficient to interfere broadly with the conduct of the patient's customary affairs of life, which is not isolated to a single narrow category of intellectual performance, and which is independent of level of consciousness.

This deterioration should be supported by historical evidence and documented by either bedside mental status testing or ideally by more detailed neuropsychological examination, using tests that are quantifiable, reproducible, and for which normative data are available.

II. Probable IVD

A. The criteria for the clinical diagnosis of PROBABLE IVD include ALL of the following:

1. Dementia;
2. Evidence of two or more ischemic strokes by history, neurologic signs, and/or neuroimaging studies (CT or T_1-weighted MRI);

   or

   Occurrence of a single stroke with a clearly documented temporal relationship to the onset of dementia;
3. Evidence of at least one infarct outside the cerebellum by CT or T_1-weighted MRI.

B. The diagnosis of PROBABLE IVD is supported by

1. Evidence of multiple infarcts in brain regions known to affect cognition;
2. A history of multiple transient ischemic attacks;
3. History of vascular risk factors (eg, hypertension, heart disease, diabetes mellitus);
4. Elevated Hachinski Ischemia Scale (original or modified version).

C. Clinical features that are thought to be associated with IVD, but await further research, include

1. Relatively early appearance of gait disturbance and urinary incontinence;
2. Periventricular and deep white matter changes on T_2-weighted MRI that are excessive for age;
3. Focal changes in electrophysiologic studies (eg, EEG, evoked potentials) or physiologic neuroimaging studies (eg, SPECT, PET, NMR spectroscopy).

D. Other clinical features that do not constitute strong evidence either for or against a diagnosis of PROBABLE IVD include

1. Periods of slowly progressive symptoms;
2. Illusions, psychosis, hallucinations, delusions;
3. Seizures.

E. Clinical features that cast doubt on a diagnosis of PROBABLE IVD include

1. Transcortical sensory aphasia in the absence of corresponding focal lesions on neuroimaging studies;
2. Absence of central neurologic symptoms/signs, other than cognitive disturbance.

III. Possible IVD

A clinical diagnosis of POSSIBLE IVD may be made when there is

1. Dementia;

   and one or more of the following:

2a. A history or evidence of a single stroke (but not multiple strokes) without a clearly documented temporal relationship to the onset of dementia;

   or

2b. Binswanger's syndrome (without multiple strokes) that includes all of the following:

   i. Early-onset urinary incontinence not explained by urologic disease, or gait disturbance (eg, parkinsonian, magnetic, apraxic, or "senile" gait) not explained by peripheral cause,

   ii. Vascular risk factors, and

   iii. Extensive white matter changes on neuroimaging.

IV. Definite IVD

A diagnosis of DEFINITE IVD requires histopathologic examination of the brain, as well as

A. Clinical evidence of dementia;

B. Pathologic confirmation of multiple infarcts, some outside of the cerebellum.

Note: If there is evidence of Alzheimer's disease or some other pathologic disorder that is thought to have contributed to the dementia, a diagnosis of MIXED dementia should be made.

V. Mixed dementia

A diagnosis of MIXED dementia should be made in the presence of one or more other systemic or brain disorders that are thought to be *causally* related to the dementia.

The degree of confidence in the diagnosis of IVD should be specified as possible, probable, or definite, and the other disorder(s) contributing to the dementia should be listed. For example: mixed dementia due to probable IVD and possible Alzheimer's disease or mixed dementia due to definite IVD and hypothyroidism.

VI. Research classification

Classification of IVD for RESEARCH purposes should specify features of the infarcts that may differentiate subtypes of the disorder, such as

Location:	cortical, white matter, periventricular, basal ganglia, thalamus
Size:	volume
Distribution:	large, small, or microvessel
Severity:	chronic ischemia versus infarction
Etiology:	embolism, atherosclerosis, arteriosclerosis, cerebral amyloid angiopathy, hypoperfusion.

Appendix K. Diagnosis of Frontotemporal Dementia

The Lund-Manchester Research Criteria (LMRC)[1]
Clinical diagnostic features of frontotemporal dementia

- CORE DIAGNOSTIC FEATURES
  - Behavioral disorder
    - Insidious onset and slow progression
    - Early loss of personal awareness (neglect of personal hygiene and grooming)
    - Early loss of social awareness (lack of social tact, misdemeanors such as shoplifting)
    - Early signs of disinhibition (such as unrestrained sexuality, violent behavior, inappropriate jocularity, restless pacing)
    - Mental rigidity and inflexibility
    - Hyperorality (oral/dietary changes, overeating, food fads, excessive smoking and alcohol consumption, oral exploration of objects)
    - Stereotyped and perservative behavior (wandering, mannerisms such as clapping, singing, dancing, ritualistic preoccupation such as hoarding, toileting, and dressing)
    - Utilization behavior (unrestrained exploration of objects in the environment)
    - Distractibility, impulsivity, and impersistence
    - Early loss of insight into the fact that the altered condition is due to a pathological change of own mental state.

  - Affective symptoms
    - Depression, anxiety, excessive sentimentality, suicidal and fixed ideation, delusion (early and evanescent)
    - Hypochondriasis, bizarre somatic preoccupation (early and evanescent)
    - Emotional unconcern (emotional indifference and remoteness, lack of empathy and sympathy, apathy)
    - Amimia (inertia, aspontaneity)

  - Speech disorder
    - Progressive reduction of speech (aspontaneity and economy of utterance)
    - Stereotypy of speech (repetition of limited repertoire of words, pharases, or themes)
    - Echolalia and perseveration
    - Late mutism.

  - Spatial orientation and praxis preserved (intact abilities to negotiate the environment)

  - Physical signs
    - Early primitive reflexes
    - Early incontinence
    - Late akinesia, rigidity, tremor
    - Low and labile blood pressure.

- Investigations
  - Normal EEG despite clinically evident dementia
  - Brain imaging (structural or functional, or both): predominant frontal or anterior temporal abnormality, or both
  - Neuropsychology (profound failure on "frontal lobe" tests in the absence of severe amnesia, aphasia, or perceptual spatial disorder)

- SUPPORTIVE DIAGNOSTIC FEATURES
  - Onset before 65
  - Positive family history of similar disorder in a first degree relative
  - Bulbar palsy, muscular weakness and wasting, fasciculations (motor neuron disease)

- DIAGNOSTIC EXCLUSION FEATURES
  - Abrupt onset with ictal events
  - Head trauma related to onset
  - Early severe amnesia
  - Early spatial disorientation, lost in surroundings, defective localisation of objects
  - Early severe apraxia
  - Logoclonic speech with rapid loss of train of thought
  - Myoclonus
  - Cortical bulbar and spinal deficits
  - Cerebellar ataxia
  - Choreo-athetosis
  - Early, severe, pathological EEG
  - Brain imaging (predominant post-central structural or functional deficit. Multifocal cerebral lesions on CT or MRI)
  - Laboratory tests indicating brain involvement or inflammatory disorder (such as multiple sclerosis, syphilis, AIDS and herpes simplex encephalitis).

- RELATIVE DIAGNOSTIC EXCLUSION FEATURES
  - Typical history of chronic alcoholism
  - Sustained hypertension
  - History of vascular disease (such as angina, claudication).

[1] **The Lund and Manchester Groups (1994). Clinical and neuropathological criteria for frontotemporal dementia.** ***Journal of Neurology, Neurosurgery, and Psychiatry, 57:*** **416-418.**

Appendix L. Diagnosis of Dementia with Lewy Bodies

1. **The central feature required for a diagnosis of DLB is progressive cognitive decline of sufficient magnitude to interfere with normal social or occupational function. Prominent or persistent memory impairment may not necessarily occur in the early stages but is usually evident with progression. Deficits on tests of attention and of frontal-subcortical skills and visuospatial ability may be especially prominent.**
2. **Two of the following core features are essential for a diagnosis of probable DLB, and one is essential for possible DLB:**
    1. **Fluctuating cognition with pronounced variations in attention and alertness**
    2. **Recurrent visual hallucinations that are typically well formed and detailed**
    3. **Spontaneous motor features of parkinsonism**
3. **Features supportive of the diagnosis are**
    1. **Repeated falls**
    2. **Syncope**
    3. **Transient loss of consciousness**
    4. **Neuroleptic sensitivity**
    5. **Systematized delusions**
    6. **Hallucinations in other modalities**
4. **A diagnosis of DLB is less likely in the presence of**
    1. **Stroke disease, evident as focal neurologic signs or on brain imaging**
    2. **Evidence on physical examination and investigation of any physical illness or other brain disorder sufficient to account for the clinical picture**

McKeith IG, Galasko D, Kosaka MD, Perry EK, Dickson MD, et al. (1996). Consensus guidelines for the clinical and pathologic diagnosis of dementia with Lewy bodies (DLB): Report of the consortium on DLB international workshop, *Neurology, 47,* 1113-1124.

(See 2005 revision on following page)

2005 Revision of criteria for diagnosing dementia with Lewy bodies[10]

Table 1 *Revised criteria for the clinical diagnosis of dementia with Lewy bodies (DLB)*

1. *Central feature* (essential for a diagnosis of possible or probable DLB)

   Dementia defined as progressive cognitive decline of sufficient magnitude to interfere with normal social or occupational function. Prominent or persistent memory impairment may not necessarily occur in the early stages but is usually evident with progression. Deficits on tests of attention, executive function, and visuospatial ability may be especially prominent.

2. *Core* features (two core features are sufficient for a diagnosis of probable DLB, one for possible DLB)

   Fluctuating cognition with pronounced variations in attention and alertness

   Recurrent visual hallucinations that are typically well formed and detailed

   Spontaneous features of parkinsonism

3. *Suggestive features* (If one or more of these is present in the presence of one or more core features, a diagnosis of probable DLB can be made. In the absence of any core features, one or more suggestive features is sufficient for possible DLB. Probable DLB should not be diagnosed on the basis of suggestive features alone.)

   REM sleep behavior disorder

   Severe neuroleptic sensitivity

   Low dopamine transporter uptake in basal ganglia demonstrated by SPECT or PET imaging

4. *Supportive features* (commonly present but not proven to have diagnostic specificity)

   Repeated falls and syncope

   Transient, unexplained loss of consciousness

   Severe autonomic dysfunction, e.g., orthostatic hypotension, urinary incontinence

   Hallucinations in other modalities

   Systematized delusions

   Depression

   Relative preservation of medial temporal lobe structures on CT/MRI scan

   Generalized low uptake on SPECT/PET perfusion scan with reduced occipital activity

   Abnormal (low uptake) MIBG myocardial scintigraphy

   Prominent slow wave activity on EEG with temporal lobe transient sharp waves

5. A diagnosis of DLB is *less likely*

   In the presence of cerebrovascular disease evident as focal neurologic signs or on brain imaging

   In the presence of any other physical illness or brain disorder sufficient to account in part or in total for the clinical picture

   If parkinsonism only appears for the first time at a stage of severe dementia

6. *Temporal sequence* of symptoms

   DLB should be diagnosed when dementia occurs before or concurrently with parkinsonism (if it is present). The term Parkinson disease dementia (PDD) should be used to describe dementia that occurs in the context of well-established Parkinson disease. In a practice setting the term that is most appropriate to the clinical situation should be used and generic terms such as LB disease are often helpful. In research studies in which distinction needs to be made between DLB and PDD, the existing 1-year rule between the onset of dementia and parkinsonism DLB continues to be recommended. Adoption of other time periods will simply confound data pooling or comparison between studies. In other research settings that may include clinicopathologic studies and clinical trials, both clinical phenotypes may be considered collectively under categories such as LB disease or alpha-synucleinopathy.

Appendix M. Clinical Dementia Rating (CDR)

TABLE I

Clinical dementia rating (CDR)

	Healthy CDR 0	Questionable dementia CDR 0.5	Mild dementia CDR 1	Moderate dementia CDR 2	Severe dementia CDR 3
Memory	No memory loss or slight inconstant forgetfulness	Mild consistent 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
Orientation	Fully oriented		Some difficulty with time relationships; oriented for place and person at examination but may have geographic disorientation	Usually disoriented in time, often to place	Orientation to person only
Judgment + problem solving	Solves every day problems well; judgment good in relation to past performance	Only doubtful impairment in solving problems, similarities, differences	Moderate difficulty in handling complex problems; social judgment usually maintained	Severely impaired in handling problems, similarities, differences; social judgment usually impaired	Unable to make judgments or solve problems
Community affairs	Independent function at usual level in job, shopping, business and financial affairs, volunteer and social groups	Only doubtful or mild impairment, if any, in these activities	Unable to function independently at these activities though may still be engaged in some; may still appear normal to casual inspection	No pretense of independent function outside home	
Home + hobbies	Life at home, hobbies, intellectual interests well maintained	Life at home, hobbies, intellectual interests well maintained or only 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 sustained	No significant function in home outside of own room
Personal care	Fully capable of self care		Needs occasional prompting	Requires assistance in dressing, hygiene, keeping of personal effects	Requires much help with personal care; often incontinent

Score as 0.5, 1, 2, 3 only if impairment is due to cognitive loss.

References

[1]Hughes CP, Berg L, Danziger WL, Coben LA, & Martin RL (1982). A New Clinical Scale for the Staging of Dementia. *British Journal of Psychiatry, 140:* 566-572.

[2]Morris JC. The Clinical Dementia Rating (CDR): current version and scoring rules. Neurology. 1993;43(11):2412–4.

[3]Hachinski VC, Iliff LD, Zilhka E, et al. (1975). Cerebral blood flow in dementia. *Archives of Neurology, 32,* 632-637.

[4]Blessed G, Tomlinson BE, Roth M. The association between quantitative measures of dementia and of senile change in the cerebral grey matter of elderly subjects. Br J Psychiatry. 1968;114(512):797–811.

[5]McKhann G, Drachman D, Folstein M, Katzman R, Price D, Stad-lan EM. Clinical diagnosis of Alzheimer's disease: report of the NINCDS-ADRDA Work Group under the auspices of Department of Health and Human Services Task Force on Alzheimer's Disease. Neurology. 1984;34(7):939–44.

[6]McKhann GM, Knopman DS, Chertkow H, Hyman BT, et al. (2011). The diagnosis of dementia due to Alzheimer's disease: Recommendations from the National Institute on Aging-Alzheimer's Association workgroups on diagnostic guidelines for Alzheimer's disease. *Alzheimers and Dementia, 7*, 263-269.

[7]Chui HC, Victoroff JI, Margolin D, Jagust W, Shankle R, and Katzman R (1992). Criteria for the diagnosis of ischemic vascular dementia proposed by the State of California Alzheimer's Disease Diagnostic and Treatment Centers, *Neurology*, *42,* 473-480.

[8]Roman GC, Tatemichi TK, Erkinjuntti T, Cummings JL, Masdeu JC, Garcia JH, et al. Vascular dementia: diagnostic criteria for research studies. Report of the NINDS-AIREN International Workshop. Neurology. 1993;43(2):250–60.

[9]The Lund and Manchester Groups (1994). Clinical and neuropathological criteria for frontotemporal dementia. *Journal of Neurology*, *Neurosurgery*, *and Psychiatry*, *57:* 416-418.

[10]Miller BL, Ikonte C, Ponton M, Levy M, Boone K, Darby A, Berman N, Mena I, Cummings JL (1997) A study of the Lund-Manchester research criteria for frontotemporal dementia: Clinical and single-photon emission CT correlations. *Neurology, 48(4):* 937-942.

[11]McKeith IG, Galasko D, Kosaka MD, Perry EK, Dickson MD, et al. (1996). Consensus guidelines for the clinical and pathologic diagnosis of dementia with Lewy bodies (DLB): Report of the consortium on DLB international workshop, *Neurology, 47,* 1113-1124.

[12]McKeith IG. Consensus guidelines for the clinical and pathologic diagnosis of dementia with Lewy bodies (DLB): report of the Consortium on DLB International Workshop. J Alzheimers Dis. 2006;9(3):417–23.

[13]McKeith IG, Perry EK, Perry RH. (1999) Report of the second dementia with Lewy Body international workshop: Diagnosis and treatment. Consortium on Dementia with Lewy Bodies. *Neurology, 53(5):* 902-905.

[14]Albert MS, DeKosky ST, Dickson D, Dubois B, Feldman HH, Fox NC, Gamst A, Holtzman DM, Jagust WJ, Petersen RC, Snyder PJ, Carrillo MC, Thies B, & Phelps CH. (2011) The diagnosis of mild cognitive impairment due to Alzheimer's disease: Recommendations from the National Institute on Aging-Alzheimer's Association workgroups on diagnostic guidelines for Alzheimer's disease. *Alzheimer's & Dementia, 7:* 270-279.

[15]Petersen RC, Smith GE, Waring SC, Ivnik RJ, Tangalos EG, Kokmen E. Mild cognitive impairment: clinical characterization and outcome. Arch Neurol. 1999;56:303–308.

[16]Winblad B, Palmer K, Kivipelto M, Jelic V, Fratiglioni L, Wahlund LO, et al. Mild cognitive impairment--beyond controversies, towards a consensus: report of the International Working Group on Mild Cognitive Impairment. J Intern Med. 2004;256:240–246.

Document Revisions

May 24, 2021: Clarified the use of images during the diagnostic process, formalized the procedure to wait until after the diagnoses are determined before looking at the scans.