National Alzheimer's Coordinating Center
Department of Epidemiology, School of Public Health, University of Washington

4311 11th Avenue NE #300
Seattle, WA 98105
phone: (206) 543-8637; fax: (206) 616-5927
e-mail: naccmail@u.washington.edu
website: https://www.alz.washington.edu

NACC Uniform Data Set (UDS)

CODING GUIDEBOOK for Telephone Follow-up Packet

Detailed, annotated explanations of each form on an item-level basis, with instructions, operational definitions, and references

(Version 2.0, February 2008)

NOTE: Version 2 is NOT the most current version of the UDS forms and is no longer used for data submission. For the most current version, please visit http://www.alz.washington.edu.

This guidebook was last modified January 14, 2014.

This publication was funded by the National Institutes of Health through the National Institute on Aging (Cooperative Agreement U01 AG016976)

The National Alzheimer's Coordinating Center (NACC)
Uniform Data Set (UDS) Telephone Follow-up Packet (TFP)

Introduction

This guidebook contains procedures to be followed when completing the data forms prepared for the Telephone Follow-up Packet of the NACC Uniform Data Set (UDS). The forms and guidebook are published by the National Alzheimer's Coordinating Center (NACC) with the cooperation and approval of the Alzheimer's Disease Centers (ADC) Clinical Task Force.[1]

The Clinical Task Force first convened in October 2002 to begin the process of improving the Minimum Data Set (MDS), which initially consisted of a small set of uniform and standardized data on ADC participants that was contributed by the Alzheimer's Disease Centers (ADCs). Over an 18-month period, the Task Force developed and revised the individual clinical and cognitive variables, and these changes were unanimously approved by the ADC Directors in April 2004. The Task Force then worked closely with NACC to develop the UDS data forms and guidebook to ensure standardization of the criteria and administered items for the database. Throughout this process, the Task Force received helpful input not only from the ADC Directors but also the Clinical Core Leaders, data managers, and many other interested individuals.

The development of the UDS required the adoption of the following principles and assumptions:

1. The UDS must contain sufficient data to be useful as a research database, but cannot represent an unacceptable burden to participants or the ADCs. Whenever possible, the UDS capitalizes on criteria, measures, and scales already administered by the majority of ADCs.
2. Assessments of all ADC participants, including nondemented controls, will include informant interviews.
3. Assessments will be obtained annually whenever possible, so that the UDS is a longitudinal database.

The UDS was the result of that development process, and the system was implemented in September 2005. The new data was intended to expand the MDS; standardize clinical and cognitive data on all ADC participants with uniform clinical assessments and diagnoses; provide data to support current research initiatives (e.g., the NIA's Genetics Initiative); and stimulate and facilitate future collaborative research. It is not hypothesis-driven; rather, it is designed to foster hypothesis-generating studies and, where appropriate, to test specific research questions. The Clinical Task Force subsequently authorized the development of Spanish translations of specific forms to allow the administration of the UDS to non-English speaking participants. A telephone follow-up packet has recently been created to allow data collection for subjects unable to continue in-person assessments, either temporarily or permanently. A milestones form is also required for these subjects.

The Task Force requires that the UDS be administered as a standard protocol, separate from protocols that have been developed for administration at individual ADCs. The ADCs may continue to separately administer their site specific protocols to maintain fidelity with data collected prior to the implementation of the UDS and to address research questions that are not addressed by the UDS.

More recently, the Task Force has developed additional standard assessments and criteria for more advanced stages of AD, as well as for non-AD disorders such as vascular dementia, dementia with Lewy bodies, and frontotemporal lobar degenerations. This current revision of the guidebook (version 2.0) incorporates those changes. Version 2.0 also includes some clarifications and modifications of previous form content and/or data elements.

[1] John C. Morris, MD, Washington University (*Task Force Chair*); Helena Chui, MD, University of Southern California; Jeffrey L. Cummings, MD, University of California Los Angeles; Charles DeCarli, MD, University of California Davis; Steven H. Ferris, PhD, New York University; Norman Foster, MD, University of Michigan (inactive); Douglas Galasko, MD, University of California San Diego; Neill Graff-Radford, MD, Mayo Clinic Jacksonville; Elaine Peskind, MD, University of Washington; Sandra Weintraub, PhD, Northwestern University.

Telephone Follow-up Form Z1: FORM CHECKLIST

The purpose of this form is to report the submission status of all forms in the UDS follow-up visit packet for each subject.

The Telephone Follow-up Packet (TFP) aims to obtain information from the informant when the subject is unable to attend an in-person UDS evaluation. NACC requires that Forms Z1, T1, A1, A2, A5, B4, B9, D1, and E1 be submitted with a telephone packet. This data must be obtained from an informant.

For forms not designated as required, if it is not feasible to collect all or almost all of the data elements for a subject and the ADC therefore decides not to attempt collection of those data, an explanation must be provided.

KEY: If the specified form was not completed, please enter one of the following codes:

95 = Physical problem
96 = Cognitive/behavior problem
97 = Other problem
98 = Verbal refusal

Form	Description	Submitted: Yes	Submitted: No	If not submitted, specify reason (see Key above)	Comments (provide if needed)
T1	Inclusion Form	**REQUIRED**		n/a	n/a
A1	Subject Demographics	**REQUIRED**		n/a	n/a
A2	Informant Demographics	**REQUIRED**		n/a	n/a
A3	Subject Family History	☐ 1	☐ 0	__ __	
A4	Subject Medications	☐ 1	☐ 0	__ __	
A5	Subject Health History	**REQUIRED**		n/a	n/a
B4	Global Staging – CDR: Standard and Supplemental	**REQUIRED**		n/a	n/a
B5 *or* B5S	Behavioral Assessment – NPI-Q	☐ 1	☐ 0	__ __	
B7 *or* B7S	Functional Assessment – FAQ	☐ 1	☐ 0	__ __	
B9	Clinician Judgment of Symptoms	**REQUIRED**		n/a	n/a
D1	Clinician Diagnosis – Cognitive Status and Dementia	**REQUIRED**		n/a	n/a
E1	Imaging/Labs	**REQUIRED**		n/a	n/a

Check "yes" if the specified form was completed for the subject during this visit. If a form is not designated as required and is not submitted, enter the appropriate Key code for the reason and provide a written explanation in the "Comments" section.

Telephone Follow-up Form T1: INCLUSION FORM

The purpose of this form is to obtain information from the informant when the subject is unable to attend an in-person UDS evaluation. The form should be completed by the clinician or clinical interviewer who will participation in the telephone follow-up.

Please complete the following before continuing with the Telephone Follow-up Packet.

1. Why is the UDS telephone follow-up protocol being used to obtain data about the subject?	Yes	No
a. Too cognitively impaired for in-person UDS visit.	☐ 1	☐ 0
b. Too physically impaired (medical illness or injury) to attend in-person UDS visit.	☐ 1	☐ 0
c. Homebound or in nursing home and cannot travel.	☐ 1	☐ 0
d. Subject or informant refused in-person UDS visit.	☐ 1	☐ 0
e. Other (*specify*): _____ (ADC staff convenience is <u>not</u> an acceptable reason.)	☐ 1	☐ 0
Self-explanatory.		

2. Has a UDS Milestones Form documenting the change to telephone follow-up been completed? *(If no, complete a Milestones Form now.)*	☐ 1	☐ 0
Self-explanatory.		

3. Is the subject likely to resume in-person UDS follow-up evaluations?	Yes ☐ 1	No ☐ 0	Unknown ☐ 9
Self-explanatory.			

Telephone Follow-up Form A1: SUBJECT DEMOGRAPHICS

The purpose of this form is to update basic descriptive information concerning subjects previously enrolled in the UDS. This form is to be completed by ADC clinician or interviewer with the informant plus ADC records and medical records.

1.	Subject's month/year of birth:	__ __/__ __ __ __
	Based on the best available information from the subject (or informant, if necessary), enter the subject's month and year of birth in the specified numerical format (e.g., March 1920 would be entered as "03/1920").	

2.	Subject's sex:	☐ 1	Male	☐ 2	Female
	Self-explanatory. (This information, in combination with month and year of birth, will allow subject verification.)				

3.	What is the subject's living situation?	☐ 1	Lives alone	☐ 4	Lives with group
		☐ 2	Lives with spouse or partner	☐ 5	Other (*specify*): _____
		☐ 3	Lives with relative or friend	☐ 9	Unknown
	Check the box for whichever category most accurately describes the subject's current living situation. Check number 4 if the subject lives with a group of related or non-related persons in a private residence. Check number 5 if the subject's living situation is other than those listed, and briefly describe in the space provided (e.g., assisted living, nursing home, adult family home, or other institutional settings). Check number 9 only if the subject or informant is unable or unwilling to identify the subject's living situation.				

4.	What is the subject's level of independence?	☐ 1	Able to live independently	☐ 3	Requires some assistance with basic activities
		☐ 2	Requires some assistance with complex activities	☐ 4	Completely dependent
				☐ 9	Unknown
	Check the box for whichever category most accurately describes the level of activity the subject is "able" to do. If the subject or informant indicates that the subject is able to perform complex activities but is not doing the activities because of her/his living situation, the subject is still considered to be "able" to live independently. Check number 2 if subject has deterioration in accustomed complex abilities (e.g., paying bills, shopping, remembering appointments, driving, cooking). Check number 3 if subject has deterioration in accustomed basic abilities (e.g., eating, dressing, personal hygiene). Check number 4 if subject is unable to perform basic activities of daily living. Check number 9 only if the subject or informant is unable or unwilling to identify the subject's living situation.				

5.	What is the subject's primary type of residence?	☐ 1	Single family residence	☐ 4	Skilled nursing facility/ nursing home
		☐ 2	Retirement community	☐ 5	Other (*specify*): _____
		☐ 3	Assisted living/ boarding home/adult family home	☐ 9	Unknown

This type of residence refers to the subject's living situation as reported above.

Check number 1 if the subject lives in an apartment, condominium, or house.

If the subject's current type of residence is other than those listed, check number 5 and briefly describe in the space provided.

Check number 9 only if the subject or informant is unable or unwilling to identify the subject's current type of residence.

6.	Subject's primary residence zip code (first 3 digits):	__ __ __ (*leave blank if unknown*)

Enter numerically the first three digits of the subject's primary residence (e.g., 981). If the subject or informant is unable or unwilling to provide the information, leave this space blank.

7.	Subject's current marital status:	☐ 1	Married	☐ 5	Never married
		☐ 2	Widowed	☐ 6	Living as married
		☐ 3	Divorced	☐ 8	Other (*specify*): _____
		☐ 4	Separated	☐ 9	Unknown

Check the box for whichever category most accurately describes the subject's current marital status. "Living as married" (number 6) may be applied to either heterosexual or same-sex relationships.

Check number 8 if the subject's current marital status is other than those listed, and briefly describe in the space provided.

Check number 9 only if the subject or informant is unable or unwilling to identify the subject's marital status.

Telephone Follow-up Form A2: INFORMANT DEMOGRAPHICS

The purpose of this form is to update descriptive information concerning the subject's informant. The form must be completed with the informant by the clinician/interviewer for a telephone follow-up.

1.	Informant's month/year of birth:	__ __/__ __ __ __ (*99/9999 = Unknown*)	
	Enter the informant's month and year of birth in the specified numerical format (e.g., March 1920 would be entered as "03/1920"). If the informant is unable or unwilling to answer, enter "99/9999".		
2.	Informant's sex:	☐ 1 Male	☐ 2 Female
	Self-explanatory. (This information, in combination with month and year of birth, will allow informant verification.)		
3.	Is this a new informant? (*If no, skip to item #9*)	☐ 1 Yes	☐ 0 No
	If this is the same informant who provided information at a previous UDS visit, check "no" and then answer only questions 9 thru 11. If this is a new informant, answer all questions below.		
4.	Does the informant report being of Hispanic/Latino ethnicity (i.e., having origins from a mainly Spanish-speaking Latin American country), regardless of race?	☐ 1 Yes	☐ 0 No ☐ 9 Unknown
	Ask the informant whether s/he considers her/his ethnicity to be Hispanic/Latino.		
	4a. If yes, what are the informant's reported origins?	☐ 1 Mexican/Chicano/ Mexican-American ☐ 2 Puerto Rican ☐ 3 Cuban ☐ 4 Dominican	☐ 5 Central American ☐ 6 South American ☐ 50 Other (*specify*): _____ ☐ 99 Unknown
	Ask the informant what s/he considers the her/his origins to be. Read or show the choices, if required, and allow only one category choice. Check number 1 if the informant reports having origins in Mexico. Check number 2 if the informant reports having origins in Puerto Rico. Check number 3 if the informant reports having origins in Cuba. Check number 4 if the informant reports having origins in the Dominican Republic. Check number 5 if the informant reports having origins in Belize, Costa Rica, El Salvador, Guatemala, Honduras, Nicaragua, or Panama. Check number 6 if the informant reports having origins in Argentina, Bolivia, Chile, Colombia, Ecuador, Paraguay, Peru, Uruguay, or Venezuela. Check number 50 if the informant reports origins other than those listed, and enter the origin in the space provided. Check number 99 only if the informant is unable or unwilling to identify her/his origins.		

5.	What does informant report as her/his race?	☐ 1	White	☐ 4	Native Hawaiian or Other Pacific Islander
		☐ 2	Black or African American	☐ 5	Asian
		☐ 3	American Indian or Alaska Native	☐ 50	Other (*specify*): _____
				☐ 99	Unknown

Ask the informant what s/he considers her/his race to be. Read or show the choices, and allow only one category choice. There will be an opportunity to record other applicable race categories in items 5 and 6.

Number 4: This includes Native Hawaiian, Guamanian or Chamorro, Samoan, or Other Pacific Islander.

Number 5: This includes Asian Indian, Chinese, Filipino, Japanese, Korean, Vietnamese, or other Asian.

Check number 50 if the informant reports a race other than those listed, and enter the race in the space provided. If the informant prefers to report her/his race as multiracial, check number 50 and specify "multiracial".

Check number 99 only if the informant is unable or unwilling to identify her/his race.

6.	What additional race does informant report?	☐ 1	White	☐ 5	Asian
		☐ 2	Black or African American	☐ 50	Other (*specify*): _____
		☐ 3	American Indian or Alaska Native	☐ 88	None reported
		☐ 4	Native Hawaiian or Other Pacific Islander	☐ 99	Unknown

If the informant reports an additional race, check the box that corresponds to this additional race.

Numbers 4 and 5: See previous inclusion list.

Check number 50 if the informant reports an additional race other than those listed, and enter the race in the space provided.

Check number 88 if the informant reports no additional race.

Check number 99 only if the informant reports an additional race but is unable or unwilling to identify it.

7.	What additional race, beyond what was indicated above in questions 5 and 6, does informant report?	☐ 1	White	☐ 5	Asian
		☐ 2	Black or African American	☐ 50	Other (*specify*): _____
		☐ 3	American Indian or Alaska Native	☐ 88	None reported
		☐ 4	Native Hawaiian or Other Pacific Islander	☐ 99	Unknown

If the informant reports another race, in addition to those already indicated in questions 5 and 6, check the box that corresponds to this additional race.

Numbers 4 and 5: See previous inclusion list.

Check number 50 if the informant reports an additional race other than those listed, and enter the race in the space provided.

Check number 88 if the informant reports no additional race.

Check number 99 only if the informant reports an additional race but is unable or unwilling to identify it.

8. Informant's years of education (report achieved level using the codes below; if an attempted level is not completed, enter the number of years completed). High school/GED = 12; Bachelors degree = 16; Master's degree = 18; Doctorate = 20 years: __ __ (*99 = Unknown*)

This question refers to achieved educational levels, rather than the number of years it took to complete that level. Use the following to describe achieved educational levels: High school or GED = 12 years, Bachelors degree = 16 years, Master's degree = 18 years, Doctorate = 20 years.

If the informant hasn't completed a level, enter the total number of years of education completed toward that level.

Examples: If the informant attended school for 8 years and did not earn a diploma or GED, enter "08". If the informant completed 17.5 years of school and earned a bachelor's degree but did not complete an attempted master's degree, enter "17". (However, if the informant attended school for 17.5 years to earn a bachelor's degree and that was the intended level of achievement, then enter "16".) If the informant attended school for 25 years to earn a PhD, enter "20" to indicate the achieved educational level.

If the informant is unable or unwilling to answer the question, enter "99".

9. What is informant's relationship to subject?

☐ 1 Spouse/partner
☐ 2 Child
☐ 3 Sibling
☐ 4 Other relative
☐ 5 Friend/neighbor
☐ 6 Paid caregiver/provider
☐ 7 Other (*specify*): _____

Self-explanatory. If the informant's relationship to the subject is other than those listed, check number 7 and briefly describe in the space provided.

10. Does the informant live with the subject?

☐ 1 Yes (*if yes, skip to #11*)
☐ 0 No

Self-explanatory.

10a. If no, approximate frequency of in-person visits:

☐ 1 Daily
☐ 2 At least 3x/week
☐ 3 Weekly
☐ 4 At least 3x/month
☐ 5 Monthly
☐ 6 Less than once a month

Self-explanatory.

10b. If no, approximate frequency of telephone contact:

☐ 1 Daily
☐ 2 At least 3x/week
☐ 3 Weekly
☐ 4 At least 3x/month
☐ 5 Monthly
☐ 6 Less than once a month

Self-explanatory.

11. Is there a question about the informant's reliability?

☐ 1 Yes
☐ 0 No

The informant's reliability should be based on a consensus opinion from the staff that interacted with the informant. If there is any reason to doubt the reliability of the informant, check "yes".

Telephone Follow-up Form A3: SUBJECT FAMILY HISTORY

The purpose of this form is to update descriptive information concerning the subject's family history. The form is to be completed by clinician/interviewer with the informant.

For the following questions:
Dementia refers to progressive loss of memory and cognition, and may be described as senility, dementia, Alzheimer's Disease, hardening of the arteries, or other causes that compromised the subject's social or occupational functioning and from which they did not recover.

Age at onset refers to the age at which dementia symptoms began, not the age at which the diagnosis was made.

Age should be identified through the clinical history, preferably given by a knowledgeable caregiver or family member. Age of mild memory difficulties of ambiguous significance, consistent with mild cognitive impairment, may not signal age at onset. Memory decline accompanied by symptoms that reflect significant functional change in the individual's abilities, e.g., in judgment, personal finances, home activities, orientation, such that the observed change(s) arouse caregiver concern over safety, determine age at onset of dementia symptoms.

Questions that probe for functional change may include the following:

- When did the individual manifest constant forgetfulness, resulting in an inability to manage her/his daily schedule?
- When did the individual display a significant failure in judgment in responding to solicitations or subscriptions?
- When did the individual manifest a significant change in cooking abilities or other home activities?
- When did the individual display a significant change in temporal or physical orientation (confusion regarding dates or locations)?

If you do not know or cannot elicit an exact age at onset, but have a general idea, please approximate to the nearest five-year period.

☐ Review with the subject/informant the data collected for this form at the previous UDS visit. If a version 2.0 Form A3 has been submitted previously and if there have been no changes, check this box and end form here.

This box may be checked in lieu of all other items below if none of the subject family history information has changed since the last visit.

If a version 2.0 form was not previously completed or if there are changes for any of the data, proceed to the next section.

Please consider blood relatives only.

PARENTS:

☐ Provide all information below if it has not been previously submitted. If there has been any change, enter all data in the row for the appropriate parent. Otherwise, check this box and proceed to the next section.

This box may be checked if a version 2.0 form was previously completed and none of the parent information has changed since the last visit.

	a. Year of birth (9999=unknown)	b. Is the parent still living? Yes	No	Unknown	c. If deceased, indicate year of death (9999=unknown)	d. Does/did this parent have dementia (defined above), as indicated by symptoms, history or diagnosis? Yes	No	Unknown	e. If yes, indicate age at onset (999=unknown)
1. Mother	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
2. Father	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __

If a version 2.0 form has not been previously submitted or if there are any changes, enter data for columns a, b and d (and, if applicable, c and e) for the appropriate parent.

(continued)

In column d: If the subject or informant provides no specific evidence (i.e., symptoms, history, or diagnosis) that the parent had dementia, check "0". If, after probing, evidence of the parent's dementia status is ambiguous, check "9".

In column e: If the parent had dementia, enter the age s/he first displayed symptoms of dementia (as described above); do not enter the parent's age at the time dementia was diagnosed. If the subject or informant is unable or unwilling to answer, enter "999".

SIBLINGS:

☐ Provide all information below if it has not been previously submitted. If there has been any change, enter all data in the row for the appropriate sibling. Otherwise, check this box and proceed to the next section.

This box may be checked if a version 2.0 form was previously completed and none of the sibling information has changed since the last visit.

3. How many full siblings did the subject have? (*99 = Unknown*) __ __

Self-explanatory.

4. For full siblings, indicate the following:

	4a. **Year of birth** *(9999=unknown)*	4b. **Is the sibling still living?** Yes	No	Unknown	4c. **If deceased, indicate year of death** *(9999=unknown)*	4d. **Does/did this sibling have dementia (defined above), as indicated by symptoms, history or diagnosis?** Yes	No	Unknown	4e. **If yes, indicate age at onset** *(999=unknown)*
Sibling 1	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 2	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 3	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 4	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 5	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 6	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 7	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 8	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 10	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 11	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 12	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 13	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 14	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 15	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 16	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 17	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 18	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 19	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Sibling 20	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __

If a version 2.0 form has not been previously submitted or if there are any changes, enter data for columns 4a, 4b and 4d (and, if applicable, 4c and 4e) for the appropriate sibling.

For column 4d, if the subject or informant provides no specific evidence (i.e., symptoms, history, or diagnosis) that the sibling had dementia, check "0". If, after probing, evidence of the sibling's dementia status is ambiguous, check "9".

For column 4e, if the sibling had dementia, enter the age the sibling first displayed symptoms of dementia (as described earlier); do not enter the sibling's age at the time dementia was diagnosed. If the subject or informant is unable or unwilling to answer, enter "999".

CHILDREN:

☐ Provide all information below if it has not been previously submitted. If there has been any change, enter all data in the row for the appropriate child. Otherwise, check this box and proceed to the next section.

This box may be checked if a version 2.0 form was previously completed and none of the information for children has changed since the last visit.

5. How many biological children did the subject have? (*99 = Unknown*) __ __

Self-explanatory.

6. For biological children, indicate the following:

	6a. **Year of birth** *(9999=unknown)*	6b. **Is the child still living?** Yes	No	Unknown	6c. **If deceased, indicate year of death** *(9999=unknown)*	6d. **Does/did this child have dementia (defined above), as indicated by symptoms, history or diagnosis?** Yes	No	Unknown	6e. **If yes, indicate age at onset** *(999=unknown)*
Child 1	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Child 2	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Child 3	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Child 4	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Child 5	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Child 6	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Child 7	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Child 8	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Child 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Child 10	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Child 11	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Child 12	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Child 13	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Child 14	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __
Child 15	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __

If a version 2.0 form has not been previously submitted or if there are any changes, enter data for columns 6a, 6b and 6d (and, if applicable, 6c and 6e) for the appropriate child.

For column 6d, if the subject or informant provides no specific evidence (i.e., symptoms, history, or diagnosis) that the child had dementia, check "0". If, after probing, evidence of the child's dementia status is ambiguous, check "9".

For column 6e, if the child had dementia, enter the age the child first displayed symptoms of dementia (as described earlier); do not enter the child's age at the time dementia was diagnosed. If the subject or informant is unable or unwilling to answer, enter "999".

OTHER DEMENTED RELATIVES:

☐ Provide all information below if it has not been previously submitted. If there has been any change, enter all data in the row for the appropriate relative. Otherwise, check this box and end form here.

This box may be checked if a version 2.0 form was previously completed and none of the information for other demented blood relatives has changed since the last visit.

7. Number of "other demented relatives" (cousins, aunts, uncles, grandparents, half siblings), as indicated by symptoms, history or diagnosis. (*99 = Unknown*) __ __

If the subject or informant provides no specific evidence (i.e., symptoms, history, or diagnosis) that the subject's other relatives had dementia, then enter "0".If, after probing, evidence of the dementia status for other relatives is ambiguous, enter "99".

8. For "other demented relatives" (cousins, aunts, uncles, grandparents, half siblings), indicate the following:

	8a. **Year of birth** *(9999=unknown)*	8b. **Is the relative still living?** **Yes**	**No**	**Unknown**	8c. **If deceased, indicate year of death** *(9999=unknown)*	8d. **Indicate age at onset** *(999=unknown)*
Relative 1	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __
Relative 2	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __
Relative 3	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __
Relative 4	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __
Relative 5	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __
Relative 6	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __
Relative 7	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __
Relative 8	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __
Relative 9	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __
Relative 10	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __
Relative 11	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __
Relative 12	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __
Relative 13	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __
Relative 14	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __
Relative 15	__ __ __ __	☐ 1	☐ 0	☐ 9	__ __ __ __	__ __ __

If a version 2.0 form has not been previously submitted or if there are any changes, enter data for columns 8a, 8b and 8d (and, if applicable, 8c) for the appropriate relative.

For column 8d, enter the age the relative first displayed symptoms of dementia (as described earlier); do not enter the relative's age at the time dementia was diagnosed. If the subject or informant is unable or unwilling to answer, enter "999".

Telephone Follow-up Form A4: SUBJECT MEDICATIONS

The purpose of this form is to record all prescription medications taken by the subject within the two weeks prior to the current visit. OTC/non-prescription medications and vitamins/supplements need not be recorded. *[Note: The preceding text came out of a 2009 CTF decision, but due to an oversight it was not immediately added to the form.]* This form lists the 100 drugs most commonly reported by subjects at many of the ADCs. The drugs are ordered alphabetically by their generic names and, if applicable, one or more commercial (brand) names are listed in parentheses.

Is the subject currently taking any medications? ☐ Yes ☐ No

Medication Name	drugID
☐ acetaminophen (Anacin, Tempra, Tylenol)	d00049
☐ acetaminophen-hydrocodone (Vicodin)	d03428
☐ albuterol (Proventil, Ventolin, Volmax)	d00749
☐ alendronate (Fosamax)	d03849
☐ allopurinol (Aloprim, Lopurin, Zyloprim)	d00023
☐ alprazolam (Niravam, Xanax)	d00168
☐ amitriptyline (Elavil, Endep, Vanatrip)	d00146
☐ amlodipine (Norvasc)	d00689
☐ ascorbic acid (C Complex, Vitamin C)	d00426
☐ aspirin	d00170
☐ atenolol (Senormin, Tenormin)	d00004
☐ atorvastatin (Lipitor)	d04105
☐ benazepril (Lotensin)	d00730
☐ bupropion (Budeprion, Wellbutrin, Zyban)	d00181
☐ calcium acetate (Calphron, PhosLo)	d03689
☐ calcium carbonate (Rolaids, Tums)	d00425
☐ calcium-vitamin D (Dical-D, O-Cal-D)	d03137
☐ carbidopa-levodopa (Atamet, Sinemet)	d03473
☐ celecoxib (Celebrex)	d04380
☐ citalopram (Celexa)	d04332
☐ clonazepam (Klonopin)	d00197
☐ clopidogrel (Plavix)	d04258
☐ conjugated estrogens (Cenestin, Premarin)	d00541
☐ conj. estrog.-medroxyprogesterone (Prempro)	d03819
☐ cyanocobalamin (Neuroforte-R, Vitamin B12)	d00413
☐ digoxin (Digitek, Lanoxin)	d00210
☐ diltiazem (Cardizem, Tiazac)	d00045
☐ divalproex sodium (Depakote)	d03833
☐ docusate (Calcium Stool Softener, Dioctyl SS)	d01021
☐ donepezil (Aricept)	d04099
☐ enalapril (Vasotec)	d00013
☐ ergocalciferol (Calciferol, Drisdol, Vitamin D)	d03128
☐ escitalopram (Lexapro)	d04812
☐ estradiol (Estrace, Estrogel, Fempatch)	d00537
☐ famotidine (Mylanta AR, Pepcid)	d00141
☐ ferrous sulfate (FeroSul, Iron Supplement)	d03824
☐ fexofenadine (Allegra)	d04040
☐ finasteride (Propecia, Proscar)	d00563
☐ fluoxetine (Prozac)	d00236
☐ folic acid (Folic Acid)	d00241
☐ furosemide (Lasix)	d00070
☐ gabapentin (Neurontin)	d03182
☐ galantamine (Razadyne, Reminyl)	d04750
☐ glipizide (Glucotrol)	d00246
☐ glucosamine (Hydrochloride)	d04418
☐ glyburide (DiaBeta, Glycron, Micronase)	d00248
☐ hydrochlorothiazide (Esidrix, Hydrodiuril)	d00253
☐ hydrochlorothiazide-triamterene (Dyazide)	d03052
☐ ibuprofen (Advil, Motrin, Nuprin)	d00015
☐ lansoprazole (Prevacid)	d03828
☐ latanoprost ophthalmic (Xalatan)	d04017
☐ levothyroxine (Levothroid, Levoxyl, Synthroid)	d00278
☐ lisinopril (Prinivil, Zestril)	d00732
☐ loratadine (Alavert, Claritin, Dimetapp, Tavist)	d03050
☐ lorazepam (Ativan)	d00149

Medication Name	drugID
☐ losartan (Cozaar)	d03821
☐ lovastatin (Altocor, Mevacor)	d00280
☐ medroxyprogesterone (Depo-Provera)	d00284
☐ memantine (Namenda)	d04899
☐ metformin (Glucophage, Riomet)	d03807
☐ metoprolol (Lopressor, Toprol-XL)	d00134
☐ mirtazapine (Remeron)	d04025
☐ multivitamin	d03140
☐ multivitamin with minerals	d03145
☐ naproxen (Aleve, Anaprox, Naprosyn)	d00019
☐ niacin (Niacor, Nico-400, Nicotinic Acid)	d00314
☐ nifedipine (Adalat, Procardia)	d00051
☐ nitroglycerin (Nitro-Bid, Nitro-Dur, Nitrostat)	d00321
☐ olanzapine (Zyprexa)	d04050
☐ omega-3 polyunsaturated fatty acids (Omacor)	d00497
☐ omeprazole (Prilosec)	d00325
☐ oxybutynin (Ditropan, Urotrol)	d00328
☐ pantoprazole (Protonix)	d04514
☐ paroxetine (Paxil, Paxil CR, Pexeva)	d03157
☐ phenytoin (Dilantin)	d00143
☐ potassium chloride (K-Dur 10, K-Lor, Slow-K)	d00345
☐ pravastatin (Pravachol)	d00348
☐ prednisone (Deltasone, Orasone)	d00350
☐ psyllium (Fiberall, Metamucil)	d01018
☐ pyridoxine (Vitamin B6)	d00412
☐ quetiapine (Seroquel)	d04220
☐ rabeprazole (Aciphex)	d04448
☐ raloxifene (Evista)	d04261
☐ ranitidine (Zantac)	d00021
☐ risperidone (Risperdal)	d03180
☐ rivastigmine (Exelon)	d04537
☐ sertraline (Zoloft)	d00880
☐ simvastatin (Zocor)	d00746
☐ tamsulosin (Flomax)	d04121
☐ temazepam (Restoril)	d00384
☐ terazosin (Hytrin)	d00386
☐ tolterodine (Detrol)	d04294
☐ trazodone (Desyrel)	d00395
☐ trolamine salicylate topical (Analgesia Creme)	d03884
☐ valsartan (Diovan)	d04113
☐ venlafaxine (Effexor)	d03181
☐ verapamil (Calan, Isoptin, Verelan)	d00048
☐ vitamin E (Aquavite-E, Centrum Singles)	d00405
☐ warfarin (Coumadin, Jantoven)	d00022
☐ zolpidem (Ambien)	d00910
☐ *Specify:*	d _ _ _ _ _
☐ *Specify:*	d _ _ _ _ _
☐ *Specify:*	d _ _ _ _ _
☐ *Specify:*	d _ _ _ _ _
☐ *Specify:*	d _ _ _ _ _
☐ *Specify:*	d _ _ _ _ _
☐ *Specify:*	d _ _ _ _ _
☐ *Specify:*	d _ _ _ _ _
☐ *Specify:*	d _ _ _ _ _

For each medication, find and mark the appropriate check box. If a reported drug is not on the list, enter the medication name in one of the open-text boxes listed as "Specify" at the end of the form. For all medications listed in the open-text boxes, associated drugIDs must also be recorded. The drugIDs may be determined by using the drugID Lookup Tool located on the NACC website at https://www.alz.washington.edu/MEMBER/DrugCodeLookUp.html. A tutorial for the Lookup Tool is also posted on this page.

Telephone Follow-up Form A5: SUBJECT HEALTH HISTORY

The purpose of this form is to record a history of conditions at the current visit as determined by the clinician's best judgment. The form should be completed by the clinician, based on subject/informant report, medical records, and/or observation.

- A condition should be considered "Absent" if it is not indicated by information obtained from informant report, medical records and/or observation.
- A condition should be considered "Active" if it happened within the last year or still requires active management, and is consistent with information obtained from informant report, medical records and/or observation.
- A condition should be considered "Inactive" if it existed or occurred in the past (greater than one year ago) but was resolved or there is no current treatment underway.
- A condition should be considered "Unknown" if there is insufficient information available from informant report, medical records and/or observation.

1. Cardiovascular disease	Absent	Active	Inactive	Unknown
a. Heart attack/cardiac arrest	☐ 0	☐ 1	☐ 2	☐ 9
b. Atrial fibrillation	☐ 0	☐ 1	☐ 2	☐ 9
c. Angioplasty/endarterectomy/stent	☐ 0	☐ 1	☐ 2	☐ 9
d. Cardiac bypass procedure	☐ 0	☐ 1	☐ 2	☐ 9
e. Pacemaker	☐ 0	☐ 1	☐ 2	☐ 9
f. Congestive heart failure	☐ 0	☐ 1	☐ 2	☐ 9
g. Other (*specify*): _____	☐ 0	☐ 1	☐ 2	☐ 9

Items 1a–1f are self-explanatory.

For item 1g, ask if the subject has any cardiovascular disease other than those listed. If no, check "0". If yes, record the condition in the space provided and check the appropriate box to specify whether active or inactive. (NOTE: "recent" and "remote" are not applicable to follow-up visits.)

2. Cerebrovascular disease	Absent	Active	Inactive	Unknown
a. Stroke	☐ 0	☐ 1	☐ 2	☐ 9
If active, indicate year(s) in which this occurred: (*9999 = Year unknown*)		1) __ __ __ __ 2) __ __ __ __ 3) __ __ __ __ 4) __ __ __ __ 5) __ __ __ __ 6) __ __ __ __		
b. Transient ischemic attack	☐ 0	☐ 1	☐ 2	☐ 9
If active, indicate year(s) in which this occurred: (*9999 = Year unknown*)		1) __ __ __ __ 2) __ __ __ __ 3) __ __ __ __ 4) __ __ __ __ 5) __ __ __ __ 6) __ __ __ __		
c. Other (*specify*): _____	☐ 0	☐ 1	☐ 2	☐ 9

Use the following criteria,[1] for stroke and recode either of two categories::

Clinical Stroke
Rapidly developing clinical symptoms and/or signs of focal and at times global loss of cerebral function (applied to patients in deep coma and to those with subarachnoid hemorrhage) with symptoms lasting more than 24 hours or leading to death (therefore excluding transient ischemic attacks), and with no apparent cause other than vascular.

Patients are also classified as having an ischemic stroke if they had computed tomographic or magnetic resonance imaging of the brain done within 7 days of onset of symptoms that either identified the symptomatic cerebral infarct or failed to identify an alternative cause of the symptoms.

Silent Stroke
Strokes determined by neuroimaging alone for which there is no history nor clinical sign (aka "silent stroke") should be captured by the UDS. This capture will be less than 100% as not all ADC participants have the necessary imaging study available at the time the clinician is completing the UDS form. For this question, a silent stroke, as defined here, would be coded as" remote/inactive," but the year may be unknown unless it can be documented to have occurred between visits, for example as a new MRI finding.

For subquestions *a* and *b*, enter each year of occurrence as a 4-digit number. If the event occurred more than once in a given year, make an entry for each occurrence (e.g., if the subject had three strokes in 2006, enter "2006" three times in the spaces provided).

TIA
Transient ischemic attack is rapidly developing clinical symptoms and/or signs indicating loss of cerebral function lasting less than 24 hours with no apparent cause other than vascular.

Other
If subject has a history of cerebrovascular disease other than those listed, briefly describe the condition in "Other" and check the appropriate box.

3. Parkinsonian features		Absent	Active	Unknown
a. Parkinson's disease		☐ 0	☐ 1	☐ 9
If active, indicate year of diagnosis:	(*9999 = Year unknown*)	__ __ __ __		
b. Other Parkinsonism disorder		☐ 0	☐ 1	☐ 9
If active, indicate year of diagnosis:	(*9999 = Year unknown*)	__ __ __ __		

Self-explanatory. Enter the year of diagnosis as a 4-digit number.

4. Other neurologic conditions	Absent	Active	Inactive	Unknown
a. Seizures	☐ 0	☐ 1	☐ 2	☐ 9
b. Traumatic brain injury				
1) with brief loss of consciousness (< 5 minutes)	☐ 0	☐ 1	☐ 2	☐ 9
2) with extended loss of consciousness (≥ 5 minutes)	☐ 0	☐ 1	☐ 2	☐ 9
3) with chronic deficit or dysfunction	☐ 0	☐ 1	☐ 2	☐ 9
c. Other (*specify*): _____	☐ 0	☐ 1	☐ 2	☐ 9

Self-explanatory. For item 4b3, check number 1 or 2 if sustained neurological impairment resulted from

[1] Cerebrovascular disease in the community: Results of a WHO Collaborative Study. (S Aho et al., 1980, Bull World Health Org., 58:113-130).

the head injury.

If subject has a history of neurologic condition other than those listed, briefly describe the condition in "Other" and check the appropriate box.

5. Medical/metabolic conditions	Absent	Active	Inactive	Unknown
a. Hypertension	☐ 0	☐ 1	☐ 2	☐ 9
b. Hypercholesterolemia	☐ 0	☐ 1	☐ 2	☐ 9
c. Diabetes	☐ 0	☐ 1	☐ 2	☐ 9
d. B12 deficiency	☐ 0	☐ 1	☐ 2	☐ 9
e. Thyroid disease	☐ 0	☐ 1	☐ 2	☐ 9
f. Incontinence – urinary	☐ 0	☐ 1	☐ 2	☐ 9
g. Incontinence – bowel	☐ 0	☐ 1	☐ 2	☐ 9

Self-explanatory.

6. Depression	No	Yes	Unknown

Include depressive disorders for which a clinician was consulted, whether or not treatment (behavioral or drug) was received. Depression includes major depressive disorder, situational depression, bipolar disorders, dysthymic disorders, and other mood disorders. Assessment can include DSM diagnoses, chart reviews, clinicians' opinion, or whether the subject is taking an SSRI for a depressive/mood disorder.

a. Active within past 2 years	☐ 0	☐ 1	☐ 9

Check "yes" if the subject has had a depression requiring medical attention within the last two years. If there have been no episodes of depression within the past two years, check "no". If, based on subject/informant report and/or medical records, it cannot be determined whether depression has occurred within the past two years, check "unknown".

b. Other episodes (prior to 2 years)	☐ 0	☐ 1	☐ 9

Check "yes" if episodes of depression occurred more than two years ago. If there were no episodes of depression prior to two years ago, check "no". If, based on subject/informant report and/or medical records, it cannot be determined whether depression occurred prior to the past two years, check "unknown".

7. Substance abuse and psychiatric disorders				
a. Substance abuse – alcohol	Absent	Active	Inactive	Unknown
1) Clinically significant impairment occurring over a 12-month period manifested in one of the following: work, driving, legal or social.	☐ 0	☐ 1	☐ 2	☐ 9

Self-explanatory.

b. Cigarette smoking history	No	Yes	Unknown

This section refers to cigarette smoking only. If your Center is interested in capturing information regarding chewing tobacco, snuff, etc., please use a separate non-UDS form.

1) Has subject smoked within last 30 days?	☐ 0	☐ 1	☐ 9

Self-explanatory.

2) Has subject smoked more than 100 cigarettes in her/his life?	☐ 0	☐ 1	☐ 9

If the subject has not smoked more than 100 cigarettes in her/his life, check "no" and then indicate

"N/A" for each of the remaining three questions below.

3) Total years smoked: (*88 = N/A; 99 = Unknown*) __ __

Self-explanatory.

4) Average number of packs/day smoked:

☐ 1 1 cigarette – < ½ pack
☐ 2 ½ – < 1 pack
☐ 3 1 – < 1½ pack
☐ 4 1½ – < 2 packs
☐ 5 ≥ 2 packs
☐ 8 N/A
☐ 9 Unknown

Check the appropriate box to indicate the average number of packs per day smoked by the subject while s/he was a smoker. Check number 9 only if the smoking history is unknown, based on available information or observation.

5) If subject quit smoking, specify age when last smoked (i.e., quit): (*888 = N/A; 999 = Unknown*) __ __ __

Self-explanatory.

c. Other abused substances

	Absent	Active	Inactive	Unknown
1) Clinically significant impairment occurring over a 12-month period manifested in one of the following: work, driving, legal or social.	☐ 0	☐ 1	☐ 2	☐ 9

If active or inactive, specify abused substance(s): _____

If number 1 or 2 is checked, briefly describe the other abused substance(s) in the space provided.

d. Psychiatric disorders ☐ 0 ☐ 1 ☐ 2 ☐ 9

If active or inactive, specify disorder(s): _____

If number 1 or 2 is checked, briefly describe the psychiatric disorder(s), other than depression (reported in item 6 above), in the space provided.

Telephone Follow-up Form B4: GLOBAL STAGING – CLINICAL DEMENTIA RATING (CDR): STANDARD AND SUPPLEMENTAL

The form should be completed by the clinician or other trained health professional, based on informant report and previous records of neurological exam of the subject. In the extremely rare instances when no informant is available, the clinician or other trained health professional must complete both the standard and supplemental versions of this form utilizing all other available information and her/his best clinical judgment. In support of the Uniform Data Set (UDS), NACC asked the Washington University ADC to create a CDR (standard version) training site for ADC personnel based on the training currently offered for staff working on the Alzheimer's Disease Cooperative Study (ADCS) trials. The UDS CDR Training Application (standard) may be accessed online at http://alzheimer.wustl.edu/cdr/Application/Step1.htm.

SECTION 1: STANDARD CDR

Use all information and make the best judgment. Score each category as independently as possible. Mark in only one box for each category, rating impairment as decline from the person's usual level due to cognitive loss alone, not impairment due to other factors such as physical handicap or depression. Occasionally, the evidence is ambiguous and the clinician's best judgment is that a category could be rated in either one of two adjacent boxes, such as mild (1) or moderate (2) impairment. In that situation, the standard procedure is to check the box of greater impairment.

Aphasia is taken into account by assessing both language and non-language function in each cognitive category. If aphasia is present to a greater degree than the general dementia, the subject is rated according to the general dementia. Supply evidence of non-language cognitive function.

Standard CDR Sum of Boxes

Calculate the sum of values for all answers and enter the total score in the space provided.

Standard Global CDR

The standard global CDR is derived from the scores in each of the six categories ("box score") as follows:

- Memory (M) is the primary category and all others are secondary.
- CDR = M if at least three secondary categories are given the same score as memory. Whenever three or more secondary categories are given a score greater or less than the memory score, CDR = score of majority of secondary categories on whichever side of M has the greater number of secondary categories. However, when three secondary categories are scored on one side of M and two secondary categories are scored on the other side of M, then CDR = M.
- When M = 0.5, CDR = 1 if at least three of the other categories are scored 1 or greater. If M = 0.5, then CDR cannot be 0; it can only be 0.5 or 1.
- If M = 0, then CDR = 0 unless there is impairment (0.5 or greater) in two more secondary categories, in which case CDR = 0.5.

Although applicable to most AD situations, these rules do not cover all possible scoring combinations. Unusual circumstances that occasionally occur in AD, and may be expected in non-Alzheimer's dementia as well, are scored as follows:

1) With ties in the secondary categories on one side of M, choose the tied scores closest to M for CDR (e.g., if M and another secondary category = 3, two secondary categories = 2, and two secondary categories = 1, then CDR = 2).
2) When only one or two secondary categories are given the same score as M, CDR = M as long as no more than two secondary categories are on either side of M.
3) When M = 1 or greater, CDR cannot be 0; in this circumstance, CDR = 0.5 when the majority of secondary categories are 0.

The CDR scoring algorithm can be accessed online at http://www.biostat.wustl.edu/~adrc/cdrpgm/index.html.

SECTION 2: SUPPLEMENTAL CDR

In addition to the factors investigated within the standard CDR, two additional constructs, "Behavior, Comportment and Personality" and "Language", have been appended as the UDS Supplemental CD, which will aid in the identification subjects with Frontotemporal Dementia and/or Primary Progressive Aphasia, respectively. Due to the specialized nature of these, instructions for the scoring of each item are outlined below.

Behavior, Comportment and Personality:

This item should be completed by a clinician or other trained health professional who is skilled in the assessment of FTLD, based on informant report and a review of the subject's cognitive, functional and behavioral status. This domain is intended to assess changes in personality, aberrant behaviors and changes in interpersonal relationships. The kinds of specific issues that might fall under these rubrics include: loss of insight, disinhibition, apathy, social withdrawal and disengagement, emotional lability, easy distractibility, reduced empathy for the feelings of others, impulsivity, and changes in eating habits and table manners. A key element of each of these is the degree to which these behaviors impact interpersonal relationships.

Language:

This item should be completed by a clinician or other trained health professional who is well versed in aphasia and aphasic disorders, based on information derived from the informant and also from clinical assessment of the patient's language functions in the course of the clinical interview and the mental status examination. This domain is intended to assess changes in language. The components of language that are primarily of interest include: spontaneous speech, auditory comprehension, object naming, reading and writing. Using these in combination, the goal is to assess the subject's ability to create and understand various forms of information. The rate at which these are generated or comprehended is also of interest.

CLINICAL DEMENTIA RATING (CDR): STANDARD AND SUPPLEMENTAL

SECTION 1: STANDARD CDR [1]

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

SECTION 2: SUPPLEMENTAL CDR

Please enter scores below	IMPAIRMENT				
	None 0	**Questionable 0.5**	**Mild 1**	**Moderate 2**	**Severe 3**
9. BEHAVIOR, COMPORTMENT AND PERSONALITY [2] __ . __	Socially appropriate behavior	Questionable changes in comportment, empathy, appropriateness of actions.	Mild but definite changes in behavior.	Moderate behavioral changes, affecting interpersonal relationships and interactions in a significant manner.	Severe behavioral changes, making interpersonal interactions all unidirectional.
10. LANGUAGE [3] __ . __	Normal speech, normal comprehension.	Minimal but noticeable word finding, minimal non-fluency. Comprehension normal in ordinary conversation.	Mild word finding problems event frequently, but does not significantly degrade broken speech. Or mild comprehension difficulties.	Moderate word-finding problems, interferes significantly with communication or moderate nonfluency or moderate comprehension difficulty in ordinary conversation.	Severe deficits in word finding, expressive speech, comprehension making communication virtually nil.

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

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

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

Telephone Follow-up Form B5: BEHAVIORAL ASSESSMENT – NEUROPSYCHIATRIC INVENTORY QUESTIONNAIRE (NPI-Q[1])

The form is to be completed by the clinician or other trained health professional per informant interview. ADC personnel must be certified as an NPI-Q interviewer through the online training system developed by the University of California Los Angeles and NACC. The NPI-Q Interviewer Certification system may be accessed through the NACC website at https://www.alz.washington.edu/npiq/signin.html. The procedures established in the training system must be followed to complete this form.

Please ask the following questions based upon changes. Indicate "yes" only if the symptom has been present within the past month; otherwise, indicate "no".

For each item marked "yes", rate the SEVERITY of the symptom (how it affects the patient): 1 = Mild (noticeable, but not a significant change)
2 = Moderate (significant, but not a dramatic change)
3 = Severe (very marked or prominent; a dramatic change)

1. NPI informant: ☐ 1 Spouse ☐ 2 Child ☐ 3 Other (specify): _____

		Yes	No		Severity		
2. DELUSIONS: Does the patient believe that others are stealing from him or her, or planning to harm him or her in some way?	2a.	☐ 1	☐ 0	2b.	☐ 1	☐ 2	☐ 3
3. HALLUCINATIONS: Does the patient act as if he or she hears voices? Does he or she talk to people who are not there?	3a.	☐ 1	☐ 0	3b.	☐ 1	☐ 2	☐ 3
4. AGITATION OR AGGRESSION: Is the patient stubborn and resistive to help from others?	4a.	☐ 1	☐ 0	4b.	☐ 1	☐ 2	☐ 3
5. DEPRESSION OR DYSPHORIA: Does the patient act as if he or she is sad or in low spirits? Does he or she cry?	5a.	☐ 1	☐ 0	5b.	☐ 1	☐ 2	☐ 3
6. ANXIETY: Does the patient become upset when separated from you? Does he or she have any other signs of nervousness, such as shortness of breath, sighing, being unable to relax, or feeling excessively tense?	6a.	☐ 1	☐ 0	6b.	☐ 1	☐ 2	☐ 3
7. ELATION OR EUPHORIA: Does the patient appear to feel too good or act excessively happy?	7a.	☐ 1	☐ 0	7b.	☐ 1	☐ 2	☐ 3
8. APATHY OR INDIFFERENCE: Does the patient seem less interested in his or her usual activities and in the activities and plans of others?	8a.	☐ 1	☐ 0	8b.	☐ 1	☐ 2	☐ 3
9. DISINHIBITION: Does the patient seem to act impulsively? For example, does the patient talk to strangers as if he or she knows them, or does the patient say things that may hurt people's feelings?	9a.	☐ 1	☐ 0	9b.	☐ 1	☐ 2	☐ 3
10. IRRITABILITY OR LABILITY: Is the patient impatient or cranky? Does he or she have difficulty coping with delays or waiting for planned activities?	10a.	☐ 1	☐ 0	10b.	☐ 1	☐ 2	☐ 3
11. MOTOR DISTURBANCE: Does the patient engage in repetitive activities, such as pacing around the house, handling buttons, wrapping string, or doing other things repeatedly?	11a.	☐ 1	☐ 0	11b.	☐ 1	☐ 2	☐ 3
12. NIGHTTIME BEHAVIORS: Does the patient awaken you during the night, rise too early in the morning, or take excessive naps during the day?	12a.	☐ 1	☐ 0	12b.	☐ 1	☐ 2	☐ 3
13. APPETITE AND EATING: Has the patient lost or gained weight, or had a change in the food he or she likes?	13a.	☐ 1	☐ 0	13b.	☐ 1	☐ 2	☐ 3

[1] Copyright© Jeffrey L. Cummings, MD. Reproduced by permission.

Telephone Follow-up Form B7: FUNCTIONAL ASSESSMENT – FUNCTIONAL ASSESSMENT QUESTIONNAIRE (FAQ[1])

NOTE: This form is to be completed by the clinician or other trained health professional, based on information provided by informant. For additional clarification and examples, see UDS Coding Guidebook for Telephone Follow-up Packet, Form B7. Indicate the level of performance for each activity by circling the one appropriate response.

The form is intended for completion by clinician or other trained health professional per informant interview. The intent of the FAQ is to assess change in an individual's functional activities, relative to previously attained abilities, that are caused by cognitive dysfunction. Circle the most accurate response, based on the informant's assessment.

In the past four weeks, did the subject have any difficulty or need help with:	**Not applicable (e.g., never did)**	**Normal**	**Has difficulty, but does by self**	**Requires assistance**	**Dependent**
1. Writing checks, paying bills, or balancing a checkbook.	8	0	1	2	3
2. Assembling tax records, business affairs, or other papers.	8	0	1	2	3
3. Shopping alone for clothes, household necessities, or groceries.	8	0	1	2	3
4. Playing a game of skill such as bridge or chess, working on a hobby.	8	0	1	2	3
5. Heating water, making a cup of coffee, turning off the stove.	8	0	1	2	3
6. Preparing a balanced meal.	8	0	1	2	3
7. Keeping track of current events.	8	0	1	2	3
8. Paying attention to and understanding a TV program, book, or magazine.	8	0	1	2	3
9. Remembering appointments, family occasions, holidays, medications.	8	0	1	2	3
10. Traveling out of the neighborhood, driving, or arranging to take public transportation.	8	0	1	2	3

Self-explanatory. If the informant indicates that the subject no longer performs a particular task, it is reasonable to probe further and ask if they think the subject could still do the task. This will help tease out the relevant cognitive impairment.

[1] Pfeffer RI, Kurosaki TT, Harrah CH, et al. Measurement of functional activities of older adults in the community. *J Gerontol* 37:323-9, 1982. Copyright© The Gerontological Society of America. Reproduced by permission of the publisher.

Telephone Follow-up Form B9: CLINICIAN JUDGMENT OF SYMPTOMS

The purpose of this form is to provide clinical determination of the onset of symptoms. The form should be completed by the clinician, and conclusions should be based on information obtained through subject, informant, medical records and/or observation. Neuropsychological test battery (except for the MMSE) and imaging results should not be used to determine answers for this form, but should be used to make the official clinical diagnosis on Form D1.

MEMORY COMPLAINT/AGE OF ONSET:	**Yes**	**No**
Relative to previously attained abilities:		
1. Does the subject report a decline in memory?	☐ 1	☐ 0

Decline refers to cognitive changes in the subject's usual or customary memory function. Check "yes" if the subject reports a current (i.e., recent) decline in memory function. This question refers to memory only and not behavior, motor, or other non-memory symptoms.

2. Does the informant report a decline in subject's memory?	☐ 1	☐ 0

Decline refers to cognitive changes in the subject's usual or customary memory function. Check "yes" if the informant reports a current (i.e., recent) decline in the subject's memory function. This question refers to memory only and not behavior, motor, or other non-memory symptoms. If there is no informant, leave this question blank.

3a. Does the clinician believe there has been a current meaningful decline in the subject's memory, non-memory cognitive abilities, behavior, or ability to manage her/his affairs, or have there been motor/movement changes?	☐ 1	☐ 0 (*If no, end form here*)

Cognitive decline refers to changes in the subject's usual or customary memory or non-memory cognitive abilities currently reported.

Decline or changes in behavior and ability to manage her/his affairs refers to meaningful change/decline from the subject's usual or customary behavior or functional ability levels currently reported.

Decline or changes in motor/movement refers to meaningful decline from the subject's usual or customary level (for example, as evidenced by gait disorder, falls, tremor and slowness) currently reported.

If the clinician is certain that there has been no meaningful decline (i.e., clinically significant) in the subject's memory, non-memory cognitive abilities, behavior, or ability to manage affairs, check "no" and do not complete the remainder of this form.

If the clinician is certain that there has been a meaningful decline, check "yes" and continue to question 3b.

If the clinician is uncertain whether there has been a meaningful decline, s/he should first complete questions 4 through 14 and then answer questions 3a and 3b.

3b. At what age did the cognitive decline begin (based upon the clinician's assessment)?	___ ___ ___	(*999=Unknown*) (*888= N/A*)

Cognitive decline refers to changes in the subject's usual or customary memory or non-memory cognitive abilities reported or observed at the current visit.

If cognitive decline is present, enter the age of onset. If the age of onset is unknown, enter "999". Any demented subject, as defined by Form D1, item 3, should have an age of onset entered.

If the subject does not exhibit cognitive decline, enter "888".

COGNITIVE SYMPTOMS:	Yes	No	Unknown
4. Indicate whether the subject currently is impaired meaningfully, relative to previously attained abilities, in the following cognitive domains, or has fluctuating cognition:			
a. **Memory** (For example, does s/he forget conversations and/or dates; repeat questions and/or statements; misplace more than usual; forget names of people s/he knows well?)	☐ 1	☐ 0	☐ 9
b. **Judgment and problem-solving** (For example, does s/he have trouble handling money (tips); paying bills; shopping; preparing meals; handling appliances; handling medications; driving?)	☐ 1	☐ 0	☐ 9
c. **Language** (For example, does s/he have hesitant speech; have trouble finding words; use inappropriate words without self-correction?)	☐ 1	☐ 0	☐ 9
d. **Visuospatial function** (For example, does s/he have difficulty interpreting visual stimuli; finding her/his way around.)	☐ 1	☐ 0	☐ 9
e. **Attention/concentration** (For example, does the subject have a short attention span or ability to concentrate? Is s/he easily distracted?)	☐ 1	☐ 0	☐ 9
f. **Fluctuating cognition** (Does s/he have pronounced variation in attention and alertness, noticeably over hours or days? For example, long periods of staring into space or lapses, or times when her/his ideas have a disorganized flow.)	☐ 1	☐ 0	☐ 9
g. **Other** (*If yes, then specify*): _____	☐ 1	☐ 0	☐ 9

Self-explanatory. Check number 9 only if the answer cannot be determined based upon information gathered from the subject, informant, medical records, and/or observation.

If the subject exhibits a meaningful decline in any ability (or abilities) other than those listed, briefly describe under "Other" and check number 1 (yes).

5. Indicate the predominant symptom which was first recognized as a decline in the subject's cognition:

☐ 1	Memory	☐ 6	Other (*specify*): _____
☐ 2	Judgment and problem solving	☐ 7	Fluctuating cognition
☐ 3	Language	☐ 88	N/A
☐ 4	Visuospatial function	☐ 99	Unknown
☐ 5	Attention/concentration		

This question refers to the onset of the cognitive change (i.e., when change in cognition was first noticed). If the informant or available information indicates that several symptoms occurred simultaneously, the clinician must ask the informant and/or use her/his best clinical judgment to commit to one of the symptoms as the predominant symptom.

If the predominant cognitive symptom first recognized as a decline was other than those listed, check number 6 and briefly describe in the space provided.

Check number 88 if there was no decline in the subject's cognition.

Check number 99 only if clinician is unable to ascertain the cognitive symptom predominant at onset, based on available information or observation.

6.	Mode of onset of cognitive symptoms:	☐ 1	Gradual (*> 6 months*)	☐ 4	Other (*specify*): _____
		☐ 2	Subacute (*≤ 6 months*)		
		☐ 3	Abrupt (*within days*)	☐ 88	N/A
				☐ 99	Unknown

This question refers to the onset of the cognitive change (i.e., when change in cognition was first noticed). The clinician should choose the option that most closely resembles the mode of onset of cognitive symptoms for the subject.

If the mode of onset was other than those listed, check number 4 and briefly describe in the space provided.

Check number 88 if there was no decline in the subject's cognition.

Check number 99 only if no information is available to allow the clinician to ascertain the mode of onset.

BEHAVIOR SYMPTOMS:	**Yes**	**No**	**Unknown**
7. Indicate whether the subject currently manifests the following behavioral symptoms:			
a. **Apathy/withdrawal** (Has the subject lost interest in or displayed a reduced ability to initiate usual activities and social interaction, such as conversing with family and/or friends?)	☐ 1	☐ 0	☐ 9
b. **Depression** (Has the subject seemed depressed for more than two weeks at a time; e.g., loss of interest or pleasure in nearly all activities; sadness, hopelessness, loss of appetite, fatigue?)	☐ 1	☐ 0	☐ 9
c. **Psychosis**			
1) Visual hallucinations	☐ 1	☐ 0	☐ 9
a) If yes, are the hallucinations well-formed and detailed?	☐ 1	☐ 0	☐ 9

Check 'yes' if the hallucinations are formed and detailed (e.g., people, animals or objects, not just vague visual images, blurs, lines or colors.

2) Auditory hallucinations	☐ 1	☐ 0	☐ 9
3) Abnormal/false/delusional beliefs	☐ 1	☐ 0	☐ 9
d. **Disinhibition** (Does the subject use inappropriate coarse language or exhibit inappropriate speech or behaviors in public or in the home? Does s/he talk personally to strangers or have disregard for personal hygiene?)	☐ 1	☐ 0	☐ 9
e. **Irritability** (Does the subject overreact, such as shouting at family members or others?)	☐ 1	☐ 0	☐ 9
f. **Agitation** (Does the subject have trouble sitting still; does s/he shout, hit, and/or kick?)	☐ 1	☐ 0	☐ 9
g. **Personality change** (Does the subject exhibit bizarre behavior or behavior uncharacteristic of the subject, such as unusual collecting, suspiciousness [without delusions], unusual dress, or dietary changes? Does the subject fail to take other's feelings into account?)	☐ 1	☐ 0	☐ 9
h. **REM sleep behavior disorder** (Does the subject appear to act out her/his dreams while sleeping (e.g., punch or flail their arms, shout or scream?)	☐ 1	☐ 0	☐ 9
i. **Other** (*If yes, then specify*): _____	☐ 1	☐ 0	☐ 9

If these symptoms are reported or observed to reflect the subject's condition at this clinical evaluation based upon information gathered from the informant, medical records, and/or observation, then answer "yes" (1); otherwise, answer "no" (0). Check "unknown" (9) only if the answer cannot be determined based upon information gathered from the informant, medical records, and/or observation.

If the subject exhibits a meaningful decline in any behavior other than those listed, briefly describe under "Other" and check number 1 (yes).

8.	Indicate the predominant symptom which was first recognized as a decline in the subject's behavioral symptoms:	☐ 1	Apathy/withdrawal	☐ 7	Personality change
		☐ 2	Depression	☐ 8	Other (*specify*): _____
		☐ 3	Psychosis	☐ 9	REM sleep behavior disorder
		☐ 4	Disinhibition	☐ 88	N/A
		☐ 5	Irritability	☐ 99	Unknown
		☐ 6	Agitation		

This question refers to the subject's symptoms at onset of behavior change. If the informant or available information indicates that several symptoms occurred simultaneously, the clinician must ask the informant and/or use her/his best clinical judgment to commit to one of the symptoms as the predominant symptom.

If the predominant behavioral symptom first recognized as a decline was other than those listed, check number 8 and briefly describe in the space provided.

Check number 88 if there was no decline in the subject's behavior.

Check number 99 only if clinician is unable to ascertain the behavioral symptom predominant at onset, based on available information or observation.

9.	Mode of onset of behavioral symptoms:	☐ 1	Gradual (> *6 months*)	☐ 4	Other (*specify*): _____
		☐ 2	Subacute (≤ *6 months*)	☐ 88	N/A
		☐ 3	Abrupt (*within days*)	☐ 99	Unknown

The clinician should choose the option that most closely resembles the mode of onset of behavioral symptoms for the subject.

If the mode of onset was other than those listed, check number 4 and briefly describe in the space provided.

Check number 88 if there was no decline in the subject's behavior.

Check number 99 only if no information is available to allow the clinician to ascertain the mode of onset.

MOTOR SYMPTOMS:	**Yes**	**No**	**Unknown**
10. Indicate whether the subject currently has the following motor symptoms:			
a. **Gait disorder** (Has the subject's walking changed, not specifically due to arthritis or an injury? Is s/he unsteady, or does s/he shuffle when walking, have little or no arm-swing, or drag a foot?)	☐ 1	☐ 0	☐ 9
b. **Falls** (Does the subject fall more than usual?)	☐ 1	☐ 0	☐ 9
c. **Tremor** (Has the subject had rhythmic shaking, especially in the hands, arms, legs, head, mouth, or tongue?)	☐ 1	☐ 0	☐ 9
d. **Slowness** (Has the subject noticeably slowed down in walking or moving or handwriting, other than due to an injury or illness? Has her/his facial expression changed, or become more "wooden" or masked and unexpressive?)	☐ 1	☐ 0	☐ 9

If these symptoms are reported or observed to reflect the subject's condition at this clinical evaluation based upon information gathered from the informant, medical records, and/or observation, then answer "yes" (1); otherwise, answer "no" (0). Check "unknown" (9) only if the answer cannot be determined based upon information gathered from the informant, medical records, and/or observation.

11. Indicate the predominant symptom which was first recognized as a decline in the subject's motor symptoms:

☐ 1 Gait disorder
☐ 2 Falls
☐ 3 Tremor
☐ 4 Slowness
☐ 88 N/A
☐ 99 Unknown

This question refers to the subject's symptoms at onset of decline in motor function. If the informant or available information indicates that several symptoms occurred simultaneously, the clinician must ask the informant and/or use her/his best clinical judgment to commit to one of the symptoms as the predominant symptom.

Check number 99 only if clinician is unable to ascertain the motor symptom predominant at onset, based on available information or observation.

12. Mode of onset of motor symptoms:

☐ 1 Gradual (> *6 months*)
☐ 2 Subacute (≤ *6 months*)
☐ 3 Abrupt (*within days*)
☐ 4 Other (*specify*): _____
☐ 88 N/A
☐ 99 Unknown

Check the option that most closely resembles the mode of onset of motor symptoms for the subject.

If the mode of onset was other than those listed, check number 4 and briefly describe in the space provided.

Check number 88 if there was no decline in the subject's behavior.

Check number 99 only if no information is available to allow the clinician to ascertain the mode of onset.

a. If there were changes in motor function, were these suggestive of Parkinsonism?

☐ 1 Yes
☐ 0 No
☐ 88 N/A

Self-explanatory.

13. Course of overall cognitive/behavioral/ motor syndrome:

☐ 1 Gradually progressive
☐ 2 Stepwise
☐ 3 Static
☐ 4 Fluctuating
☐ 5 Improved
☐ 9 Unknown

Check the appropriate number to indicate the overall decline in cognitive/behavioral/motor functions during the course of the illness. Fluctuating course does not refer to the short-term fluctuations that are part of DLB.

Check number 9 only if no information is available to allow the clinician to describe the overall course of the syndrome.

14. Indicate the predominant domain which was first recognized as changed in the subject:

☐ 1 Cognition
☐ 2 Behavior
☐ 3 Motor function
☐ 9 Unknown

Check the appropriate number to indicate which domain appears to be the first to have changed in the subject. Choose only one domain as predominantly changing first, based on the clinician's best judgment.

Check number 9 only if no information is available to allow the clinician to describe the predominantly changed domain.

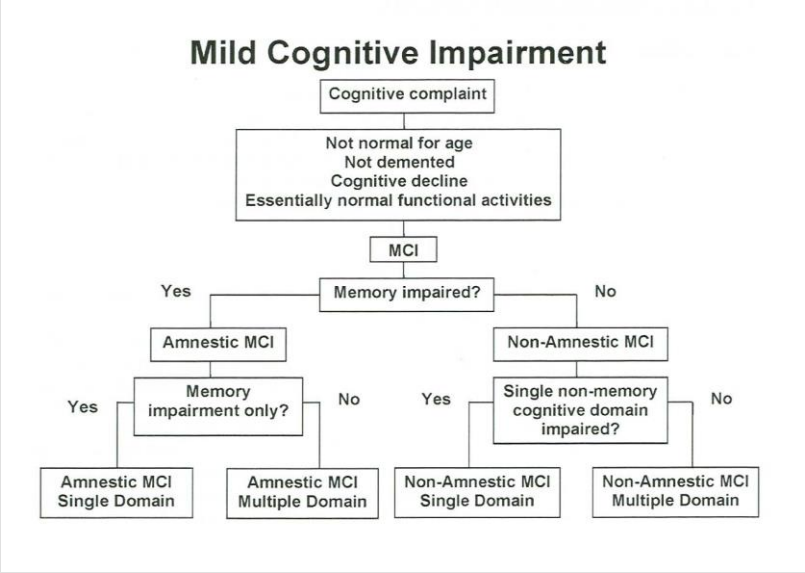
Telephone Follow-up Form D1: CLINICIAN DIAGNOSIS – COGNITIVE STATUS AND DEMENTIA

The purpose of this form is to record a diagnosis of the subject's current status relative to cognition and dementia. The form should be completed by the clinician, based on a review of all available information.

1.	Responses are based on:	☐ 1 Diagnosis from single clinician	☐ 2 Consensus diagnosis

2.	Does the subject have normal cognition (no MCI, dementia, or other neurological condition resulting in cognitive impairment)?	☐ 1 Yes (*If yes, skip to #14)*	☐ 0 No (*If no, continue to #3*)
3.	Does the subject meet criteria for dementia (in accordance with standard criteria for dementia of the Alzheimer's type or for other non-Alzheimer's dementing disorders)?	☐ 1 Yes (*If yes, skip to #5*)	☐ 0 No (*If no, continue to #4*)

After having determined that the subject does <u>not</u> have normal cognition (item #2 above) and does <u>not</u> have dementia (item #3 above), please use the following chart[1], excerpted from the Alzheimer's Disease Neuroimaging Initiative (ADNI) manual, page 18, to guide your answers to items 4a–4d:



First determine if memory impairment is present, based on your clinical judgment and/or neuropsychological memory test(s) (for example, logical memory sub-test of the WMS-R and/or other memory tests used at your ADC). Then apply the same clinical and/or psychometric approach to determine whether other cognitive domains are also impaired.

[1] *Arch Neurol*, 2005. 62;1160-63, Figure 1. Reproduced by permission of the publisher. Copyright© 2005. American Medical Association. All rights reserved.

4. If the subject does not have normal cognition and is not clinically demented, indicate the type of cognitive impairment (*choose only <u>one</u> impairment from items 4a thru 4e as being "present"; mark <u>all others</u> "absent"*) **and then designate the suspected underlying cause(s) of the impairment by completing items 5–30**:

(NOTE: Although items 5–30 were developed for individuals with clinical dementia rather than MCI, for the type of cognitive impairment indicated below (4a–4e), please designate the suspected cause(s) of the impairment in all cases by completing items 5–30.)

	Present	Absent
4a. Amnestic MCI – memory impairment only	☐ 1	☐ 0

If memory is impaired and memory is the <u>only</u> cognitive domain impaired, mark 4a as "present". (Note: Only <u>one</u> of items 4a–4e may be marked "present"; <u>all others</u> must be marked "absent".)

	Present	Absent	Domains	Yes	No
4b. Amnestic MCI – memory impairment plus one or more other domains (*if present, check one or more domain boxes "yes" and check all other domain boxes "no"*)	☐ 1	☐ 0	1) Language	☐ 1	☐ 0
			2) Attention	☐ 1	☐ 0
			3) Executive function	☐ 1	☐ 0
			4) Visuospatial	☐ 1	☐ 0

If memory is impaired, but is <u>not</u> the only cognitive domain impaired, mark 4b as "present", then mark on the list at right the other cognitive domain(s) which you judge to be impaired, based on your examination and/or neuropsychological tests.)

4c. Non-amnestic MCI – single domain (*if present, check only <u>one</u> domain box "yes"; check <u>all other</u> domain boxes "no"*)	☐ 1	☐ 0	1) Language	☐ 1	☐ 0
			2) Attention	☐ 1	☐ 0
			3) Executive function	☐ 1	☐ 0
			4) Visuospatial	☐ 1	☐ 0

If memory is <u>not</u> impaired, and <u>only one</u> other domain is impaired, mark 4c as "present" and mark on the list at right the <u>single</u> cognitive domain which you judge to be impaired, based on your examination and/or neuropsychological tests.)

4d. Non-amnestic MCI – multiple domains (*if present, check <u>two</u> or more domain boxes "yes" and check all other domain boxes "no"*)	☐ 1	☐ 0	1) Language	☐ 1	☐ 0
			2) Attention	☐ 1	☐ 0
			3) Executive function	☐ 1	☐ 0
			4) Visuospatial	☐ 1	☐ 0

(continued on next page)

If memory is <u>not</u> impaired, but <u>more than one</u> other cognitive domain is impaired, mark 4d as "present" and mark on the list at right each of those domains which you judge to be impaired, based on your examination and/or neuropsychological tests.)

4e. Impaired, not MCI	☐ 1	☐ 0

If you judge the subject to by cognitively impaired, but the subject's presentation, tests, symptoms, and clinical evaluation are <u>not consistent with MCI</u> and do not allow you to mark any of the above items (4a–4d) as "present", then mark 4e as "present".

Please indicate if the following conditions are present or absent. If present, also indicate if the condition is primary or contributing to the observed cognitive impairment (reported in items 3 or 4), based on the clinician's best judgment.

	Present	Absent		If Present: Primary	If Present: Contributing
5. Probable AD (NINCDS/ADRDA) (*if present, skip to item #7*)	☐ 1	☐ 0	5a.	☐ 1	☐ 2
6. Possible AD (NINCDS/ADRDA) (*if #5 is present, leave this blank*)	☐ 1	☐ 0	6a.	☐ 1	☐ 2

I. The criteria[1] for the clinical diagnosis of PROBABLE Alzheimer's disease include:
- dementia established by clinical examination and documented by the Mini-Mental Test, Blessed Dementia Scale, or some similar examination, and confirmed by neuropsychological tests;
- deficits in two or more areas of cognition;
- progressive worsening of memory and other cognitive functions;
- no disturbance of consciousness;
- onset between ages 40 and 90, most often after age 65; and
- absence of systemic disorders or other brain diseases that in and of themselves could account for the progressive deficit in memory and cognition.

II. The diagnosis of PROBABLE Alzheimer's disease is supported by:
- progressive deterioration of specific cognitive functions such as language (aphasia), motor skills (apraxia), and perception (agnosia);
- impaired activities of daily living and altered patterns of behavior;
- family history of similar disorders, particularly if confirmed neuropathologically; and
- laboratory results of:

  normal lumbar puncture as evaluated by standard techniques;

  normal pattern or nonspecific changes in EEG, such as increased slow-wave activity; and

  evidence of cerebral atrophy on CT with progression documented by serial observation.

III. Other clinical features consistent with the diagnosis of PROBABLE Alzheimer's disease, after exclusion of causes of dementia other than Alzheimer's disease, include:
- plateaus in the course of progression of the illness;
- associated symptoms of depression, insomnia, incontinence, delusions, illusions, hallucinations, catastrophic verbal, emotional, or physical outbursts, sexual disorders, and weight loss;
- other neurologic abnormalities in some patients, especially with more advance disease and including motor signs such as increased muscle tone, myoclonus, or gait disorder.
- seizures in advanced disease; and
- CT normal for age.

IV. Features that make the diagnosis of PROBABLE Alzheimer's disease uncertain or unlikely include:
- sudden, apoplectic onset;
- focal neurologic findings such as hemiparesis, sensory loss, visual field deficits, and incoordination early in the course of the illness; and
- seizures or gait disturbances at the onset or very early in the course of the illness.

(cont'd. on next page)

[1] McKhann G, Drachman D, Folstin M, Katzman R, Price D, and Stadlan 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 July, (34) 939-944.

V. Clinical diagnosis of POSSIBLE Alzheimer's disease:
   - may be made on the basis of the dementia syndrome, in the absence of other neurologic, psychiatric, or systemic disorders sufficient to cause dementia, and in the presence of variations in the onset, in the presentation, or in the clinical course;
   - may be made in the presence of a second systemic or brain disorder sufficient to produce dementia, which is not considered to be *the* cause of the dementia; and
   - should be used in research studies when a single gradually progressive severe cognitive deficit is identified in the absence of other identifiable cause.

VI. Criteria for diagnosis of DEFINITE Alzheimer's disease are:
   - the clinical criteria for probable Alzheimer's disease and
   - histopathologic evidence obtained from a biopsy or autopsy.

VII. Classification of Alzheimer's disease for research purposes should specify features that may differentiate subtypes of the disorder, such as:
   - familial occurrence;
   - onset before age of 65;
   - presence of trisomy-21; and
   - coexistence of other relevant conditions such as Parkinson's disease.

	Present	Absent		If Present: Primary	If Present: Contributing
7. Dementia with Lewy bodies	☐ 1	☐ 0	7a.	☐ 1	☐ 2

Revised (2005) criteria for the clinical diagnosis of dementia with Lewy bodies (DLB)[1]:

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 (it if 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 simple 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.

(For more information on the criteria above, visit the website of the Lewy Body Dementia Association at http://www.lewybodydementia.org/lbdsymptoms.shtml.)

[1] McKeith IG, Dickson DW, Lowe J, et al. Diagnosis and Management of Dementia with Lewy Bodies: Third report of the DLB Consortium, Neurology 2005; 65:1863-72.

	Present	Absent		If Present: Primary	If Present: Contributing
8. Vascular dementia (NINDS/AIREN Probable) (*if present, skip to item #10*)	☐ 1	☐ 0	8a.	☐ 1	☐ 2
9. Vascular dementia (NINDS/AIREN Possible) (*if #8 is present, leave this blank*)	☐ 1	☐ 0	9a.	☐ 1	☐ 2

This category is for dementia subjects that meet Probable NINDS-AIREN criteria for vascular dementia, which should therefore be designated as the primary etiology of the dementia. For mixed dementias or subjects meeting only Possible NINDS-AIREN criteria for vascular dementia, check number 1 ("present") for Question 23 below and indicate that stroke is contributory to the cognitive impairment.

NINDS-AIREN criteria for the diagnosis of vascular dementia[1]:

I. The criteria for the clinical diagnosis of *probable* vascular dementia include *all* of the following:

Dementia defined by cognitive decline from a previously higher level of functioning and manifested by impairment of memory and of two or more cognitive domains (orientation, attention, language, visuospatial functions, executive functions, motor control, and praxis), preferable established by clinical examination and documented by neuropsychological testing; deficits should be severe enough to interfere with activities of daily living not due to physical effects of stroke alone.

Exclusion criteria: cases with disturbance of consciousness, delirium, psychosis, severe aphasia, or major sensorimotor impairment precluding neuropsychological testing. Also excluded are systemic disorders or other brain diseases (such as AD) that in and of themselves could account for deficits in memory and cognition.

Cerebrovascular disease, defined by the presence of focal signs on neurologic examination, such as hemiparesis, lower facial weakness, Babinski sign, sensory deficit, hemianopia, and dysarthria consistent with stroke (with or without history of stroke), and evidence of nof relevant CVD by brain imaging (CT or MRI) including *multiple large vessel infarcts* or a *single strategically placed infarct* (angular gyrus, thalamus, basal forebrain, or PCA or ACA territories), as well as *multiple basal ganglia* and *white matter lacunes*, or *extensive periventricular white matter lesions*, or combinations thereof.

A relationship between the above two disorders, manifested or inferred by the presence of one or more of the following: (a) onset of dementia within 3 months following a recognized stroke; (b) abrupt deterioration in cognitive functions; or fluctuating, stepwise progression of cognitive deficits.

II. Clinical features consistent with the diagnosis of *probable* vascular dementia include the following:

(a) Early presence of gait disturbance (small-step gait or marche a petits pas, or magnetic, apraxic-ataxic or parkinsonian gait); (b) history of unsteadiness and frequent, unprovoked falls; (c) early urinary frequency, urgency, and other urinary symptoms not explained by urologic disease; (d) pseudobulbar palsy; and (e) personality and mood changes, abulia, depression, emotional incontinence, or other subcortical deficits including psychomotor retardation and abnormal executive function.

III. Features that make the diagnosis of vascular dementia uncertain or unlikely include (a) early onset of memory deficit and progressive worsening of memory deficit and progressive worsening of memory and other cognitive functions such as language (transcortical sensory aphasia), motor skills (apraxia), and perception (agnosia), in the absence of corresponding focal lesions on brain imaging; (b) absence of focal neurological signs, other than cognitive disturbance; and (c) absence of cerebrovascular lesions on brain CT or MRI.

(continued on next page)

[1] Roman GC, Tatemichi TK, Erkinjuntti T, Cummings JL, Masdeu JC, Garcia JH, Amaducci L, Orgogozo JM, Brun A, Hofman A, et al. Vascular dementia: diagnostic criteria for research studies. Report of the NINDS-AIREN International Workshop. *Neurology* 1993 Feb,43(2):250-60.

IV. Clinical diagnosis of *possible* vascular dementia may be made in the presence of dementia (section I-1) with focal neurologic signs in patients in whom brain imaging studies to confirm definite CVD are missing; or in the absence of clear temporal relationship between dementia and stroke; or in patients with subtle onset and variable course (plateau or improvement) of cognitive deficits and evidence of relevant CVD.

V. Criteria for diagnosis of *definite* vascular dementia are (a) clinical criteria for *probable* vascular dementia; (b) histopathologic evidence of CVD obtained from biopsy or autopsy; (c) absence of neurofibrillary tangles and neuritic plaques exceeding those expected for age; and (d) absence of other clinical or pathological disorder capable of producing dementia.

VI. Classification of vascular dementia for research purposes may be made on the basis of clinical, radiologic, and neuropathologic features, for subcategories or defined conditions such as cortical vascular dementia, subcortical vascular dementia, BD, and thalamic dementia.

The term "AD *with* CVD" should be reserved to classify patients fulfilling the clinical criteria for possible AD and who also present clinical or brain imaging evidence of relevant CVD. Traditionally, these patients have been included with VaD in epidemiologic studies. The term "mixed dementia," used hitherto, should be avoided.

		Present	Absent		If Present: Primary	If Present: Contributing
10.	Alcohol-related dementia	☐ 1	☐ 0	10a.	☐ 1	☐ 2
	Refer to the DSM-IV manual.[1]					
11.	Dementia of undetermined etiology	☐ 1	☐ 0	11a.	☐ 1	☐ 2
	Refer to the DSM-IV manual.					

[1] Diagnostic and statistical manual of mental disorders (DSM-IV). 4th ed. 1994, Washington, DC: American Psychiatric Association.

Use the following criteria to guide your answers to item 12:

Frontotemporal Lobar Degeneration:
A Consensus on Clinical Diagnostic Criteria (Neary et al., 1998)[1]

Criteria: The clinical criteria are set out in lists 1 through 4. The criteria for each of the three major clinical syndromes are divided into sections. The clinical profile statement together with the core clinical inclusion and exclusion features provide the necessary foundation for diagnosis. Additional clinical features, neuropsychological investigation, and brain imaging support the clinical diagnosis. Operational definitions of specific features are outlined later.

Clinical profile: This statement (seen in lists 1 through 3) summarizes the neurobehavioral profile necessary to fulfill criteria for diagnosis.

I. Core diagnostic features: These are features (see lists 1 through 3) integral to the clinical syndrome. All features must be present to fulfill the criteria for diagnosis.

II. Supportive diagnostic features:

Clinical: These are features (see lists 1 through 3) that are not present in all patients, or they may be noted only during one phase of the disease. They are therefore not necessary conditions for diagnosis. Supportive features are characteristic, often with high diagnostic specificity, and their presence adds substantial weight to the clinical diagnosis. The diagnosis becomes more likely when more supportive features are present.

Physical: In each of the clinical syndromes physical signs are few, in contrast to the prominent mental changes. Parkinsonian signs typically emerge only during late disease. The physical features outlined should be regarded as "supportive" rather than as necessary conditions for diagnosis.

Investigations: Formal neuropsychological assessment, EEG, and brain imagine each can provide support for and strengthen the clinical diagnosis. Such investigatory techniques are not available universally, and ought not to be considered a prerequisite for diagnosis. When neuropsychological assessment is performed, the profile of deficits must demonstrate disproportionate executive dysfunction in FTD or disproportionate language/semantic breakdown in PA and SD. With regard to brain imaging, the patterns of abnormality are characteristic, but not seen invariably. For example, prominent atrophy of the temporal lobes is well visualized by high-resolution MRI, but may be undetected by CT. Failure to demonstrate the prototypic appearances on imaging need not result in diagnostic exclusion.

III. Supportive features common to each of the clinical syndromes: These features (see list 4) support but are not a necessary condition for FTLD.

IV. Exclusion features common to each clinical syndrome:

Clinical: All features (see list 4) must be absent. Early severe amnesia, early spatial disorientation, logoclonic speech with loss of train of thought, and myoclonus are features designed to exclude AD.

Investigations: All features should be absent (when the relevant information is available).

V. Relative diagnostic exclusion features: These are features (see list 4) that caution against but do not firmly exclude a diagnosis of FTLD. A history of alcohol abuse raises the possibility of an alcohol-related basis for a frontal lobe syndrome. However, excessive alcohol intake may also occur in FTD patients as a secondary manifestation of social disinhibition or hyperoral tendencies. The presence of vascular risk factors such as hypertension ought to alert investigators to a possible vascular etiology. Nevertheless, such risk factors are common in the general population and may be present coincidentally in some patients.

[1] Neary D, Snowden JS, Gustafson L et al. Frontotemporal lobar degeneration: a consensus on clinical diagnostic criteria. *Neurology* 51:1546-54(1998).

		Present	Absent		If Present: Primary	If Present: Contributing
12.	Frontotemporal dementia (behavioral/executive dementia)	☐ 1	☐ 0	12a.	☐ 1	☐ 2

LIST 1. The Clinical Diagnostic Features of FTD

Clinical Profile: Character change and disordered social conduct are the dominant features initially and throughout the disease course. Instrumental functions of perception, spatial skills, praxis, and memory are intact or relatively well preserved.

I. Core diagnostic features

A. Insidious onset and gradual progression
B. Early decline in social interpersonal conduct
C. Early impairment in regulation of personal conduct
D. Early emotional blunting
E. Early loss of insight

II. Supportive diagnostic features

A. Behavioral disorder
   1. Decline in personal hygiene and grooming.
   2. Mental rigidity and inflexibility.
   3. Distractibility and impersistence.
   4. Hyperorality and dietary changes.
   5. Perseverative and stereotyped behavior.
   6. Utilization behavior.

B. Speech and language
   1 Altered speech output:
      a. Aspontaneity and economy of speech
      b. Press of speech
   2. Stereotypy of speech.
   3 Echolalia.
   4. Perseveration.
   5. Mutism.

C. Physical signs
   1. Primitive reflexes.
   2. Incontinence.
   3. Akinesia, rigidity, and tremor.
   4. Low and labile blood pressure.

D. Investigations
   1. Neuropsychology: significant impairment on frontal lobe tests in the absence of severe amnesia, aphasia, or perceptuospatial disorder.
   2. Electroencephalography: normal on conventional EEG despite clinically evident dementia.
   3. Brain imaging (structural and/or functional): predominant frontal and/or anterior temporal abnormality.

(*Review List 4 on page 39 for diagnostic exclusion criteria.*)

Use the following criteria to guide your answer to item 13:

Criteria for Primary Progressive Aphasia (PPA)[1,2]

Descriptive clinical profile: An aphasic dementia where the language impairment (aphasia) emerges in relative isolation and is the major determinant in the limitation of daily living activities. Perception, memory, personality are relatively preserved initially.

I. Core diagnostic features: These features are integral to the clinical syndrome.

A. Insidious onset and gradual progression.

B. Early onset of aphasic disturbance (including any combination of the following)

1. Word-finding pauses.
2. Word comprehension deficits.
3. Syntactic comprehension deficits.
4. Naming impairments.
5. Circumlocutious speech lacking nouns and verbs.
6. Agrammatic speech (abnormal syntax).
7. Pure word deafness.
8. Dysgraphia.

II. Supportive diagnostic features: These features are not present in all patients, but their presence serves further to support the diagnosis.

A. Clinical

1. Onset before the age of 65.
2. Dysarthria.
3. Ideomotor apraxia of the limbs.
4. Ideomotor apraxia of buccofacial musculature.
5. Dyscalculia.
6. Mild facial flattening on the side opposite the language dominant hemisphere (usually right face).
7. Asymmetrical upper extremity posturing upon stressed gait on the side opposite the language-dominant hemisphere (usually right arm).
8. Mild rigidity on the side opposite the language-dominant hemisphere (usually right side of body).

B. Investigations

1. Neuropsychology: Findings of aphasia and/or anomia in the absence of amnesia, prosopagnosia, associative visual agnosia, apathy, disinhibition. Scores on verbally mediated tests of memory and fluency may be abnormal because of the aphasia.
2. MRI or CT: Perisylvian atrophy that can extend to parietal cortex and/or inferior temporal cortex on the side of language dominance (usually left).
3. PET or SPECT: Asymmetrical hypometabolism in language-dominant hemisphere (usually left).
4. EEG: Asymmetrical slowing in the temporal leads of the language-dominant hemisphere (usually left).

(continued on next page)

[1] Mesulam M-M. Primary Progressive Aphasia. *Ann. Neurol.* 2001;49:425-432.

[2] Mesulam M-M. Primary progressive aphasia: A language-based dementia. *New Eng J Med.* 2003;348:1535-1542.

III. Exclusionary features

A. Historical or clinical

1. Abrupt onset.
2. Early amnesia.
3. Early prosopagnosia, visual agnosia.
4. Early spatial disorientation.
5. Early apathy or disinhibition.
6. Early motor neuron disease (if present, assign to relevant primary diagnosis).
7. Early major extrapyramidal signs/CBGD (if present, assign to relevant primary diagnosis).
8. Cerebellar signs.
9. Early eye movement abnormalities.
10. Head trauma related to onset.

B. Investigations

1. Brain imaging consistent with major stroke in the language dominant hemisphere (usually left).
2. Brain imaging showing asymmetrical moderate to severe lacunar stroke in the language-dominant hemisphere (usually left).
3. Brain imaging showing neoplasm or other space occupying lesion in the language-dominant hemisphere (usually left).
4. Brain imaging showing major trauma to language-dominant hemisphere (usually left).

		Present	Absent		If Present: Primary	If Present: Contributing
13.	Primary progressive aphasia (aphasic dementia)	☐ 1	☐ 0	13a.	☐ 1	☐ 2
	(*If PPA is present, specify type by checking <u>one</u> box below "present" and <u>all others</u> "absent"*):					
	1) Progressive nonfluent aphasia	☐ 1	☐ 0			

LIST 2. The clinical diagnostic features of progressive nonfluent aphasia

Clinical profile: Disorder of expressive language is the dominant feature initially and throughout the disease course. Other aspects of cognition are intact or relatively well preserved.

I. Core diagnostic features

A. Insidious onset and gradual progression

B. Nonfluent spontaneous speech with at least one of the following: agrammatism, phonemic paraphasias, anomia

II. Supportive diagnostic features

A. Speech and language
   1. Stuttering or oral apraxia.
   2. Impaired repetition.
   3. Alexia, agraphia.
   4. Early preservation of word meaning.
   5. Late mutism.

B. Behavior
   1. Early preservation of social skills.
   2. Late behavioral changes similar to FTD.

C. Physical signs: late contralateral primitive reflexes, akinesia, rigidity, and tremor

D. Investigations
   1. Neuropsychology: nonfluent aphasia in the absence of severe amnesia or perceptuospatial disorder.
   2. Electroencephalography: normal or minor asymmetric slowing.
   3. Brain imaging (structural and/or functional): asymmetric abnormality predominantly affecting dominant (usually left) hemisphere.

(*Review List 4 on page 39 for diagnostic exclusion criteria.*)

	Present	Absent
2) Semantic dementia – anomia plus word comprehension	☐ 1	☐ 0
3) Semantic dementia – agnosic variant	☐ 1	☐ 0

LIST 3. The clinical diagnostic features of semantic aphasia and associative agnosia (SD)

Clinical profile: Semantic disorder (impaired understanding of word meaning and/or object identity) is the dominant feature initially and throughout the disease course. Other aspects of cognition, including autobiographic memory, are intact or relatively well preserved.

I. Core diagnostic features

A. Insidious onset and gradual progression
B. Language disorder characterized by:
   1. Progressive, fluent, empty spontaneous speech;
   2. Loss of word meaning, manifest by impaired naming *and* comprehension;
   3. Semantic paraphasias; *and/or*
C. Perceptual disorder characterized by:
   1. Prosopagnosia: impaired recognition of identity of familiar faces; *and/or*
   2. Associative agnosia: impaired recognition of object identity.
D. Preserved perceptual matching and drawing reproduction
E. Preserved single-word repetition
F. Preserved ability to read aloud and write to dictation orthographically regular words

II. Supportive diagnostic features

A. Speech and language
   1. Press of speech.
   2. Idiosyncratic word usage.
   3. Absence of phonemic paraphasias.
   4. Surface dyslexia and dysgraphia.
   5. Preserved calculation.
B. Behavior
   1. Loss of sympathy and empathy.
   2. Narrowed preoccupations.
   3. Parsimony.
C. Physical signs
   1. Absent or late primitive reflexes.
   2. Akinesia, rigidity, and tremor.
D. Investigations
E. Neuropsychology
   1. Profound semantic loss, manifest in failure of word comprehension and naming and/or face and object recognition.
   2. Preserved phonology and syntax, and elementary perceptual processing, spatial skills, and day-to-day memorizing.
F. Electroencephalography: normal
G. Brain imaging (structural and/or functional): predominant anterior temporal abnormality (symmetric or asymmetric)

(*Review List 4 on page 39 for diagnostic exclusion criteria.*)

	Present	Absent
4) Other (e.g., logopenic, anomic, transcortical, word deafness, syntactic comprehension, motor speech disorder)	☐ 1	☐ 0

LIST 4. Features common to clinical syndromes of FTLD (extension of Lists 1 through 3)

III. Supportive features

A. Onset before 65 years: positive family history of similar disorder in first-degree relative

B. Bulbar palsy, muscular weakness and wasting, fasciculations (associated motor neuron disease present in a minority of patients)

IV. Diagnostic exclusion features

A. Historical and clinical

1. Abrupt onset with ictal events.
2. Head trauma related to onset.
3. Early, severe amnesia.
4. Spatial disorientation.
5. Logoclonic, festinant speech with loss of train of thought.
6. Myoclonus.
7. Corticospinal weakness.
8. Cerebellar ataxia.
9. Choreoathetosis.

B. Investigations

1. Brain imaging: predominant postcentral structural or functional deficit; multifocal lesions on CT or MRI.
2. Laboratory tests indicating brain involvement of metabolic or inflammatory disorder such as MS, syphilis, AIDS, and herpes simplex encephalitis.

V. Relative diagnostic exclusion features

A. Typical history of chronic alcoholism

B. Sustained hypertension

C. History of vascular disease (e.g., angina, claudication)

For subjects with normal cognition, indicate whether the following conditions are present or absent. If the subject is cognitively impaired, indicate also whether the condition is primary, contributing or non-contributing to the observed cognitive impairment, based on your best judgment. As an example, if subject is cognitively impaired and the impairment is due to Parkinson's disease dementia, then mark item #23 as "Present" and item #23a as ""Primary". If subject has other co-morbid conditions, these should be indicated as well. Mark only one condition as primary.

	Present	Absent		If Present: Primary	Contributing	Non-contrib.
14. Progressive supranuclear palsy	☐ 1	☐ 0	14a.	☐ 1	☐ 2	☐ 3

Use the following criteria, excerpted from *SIC Task Force Appraisal of Clinical Diagnostic Criteria for Parkinsonian Disorders* (Litvan et al., 2003):

NINDS-SPSP clinical criteria for the diagnosis of PSP

Diagnostic categories	Inclusion criteria	Exclusion criteria	Supportive criteria
	For possible and probable: Gradually progressive disorder with age at onset at 40 or later;	*For possible and probable:* Recent history of encephalitis; alien limb syndrome; cortical sensory deficits; focal frontal or temporoparietal atrophy; hallucinations or delusions unrelated to dopaminergic therapy; cortical dementia of Alzheimer type; prominent, early cerebellar symptoms or unexplained dysautonomia; or evidence of other diseases that could explain the clinical features.	Symmetric akinesia or rigidity, proximal more than distal; abnormal neck posture, especially retrocollis; poor or absent response of parkinsonism to levodopa; early dysphagia & dysarthria; early onset of cognitive impairment including > 2 of: apathy, impairment in abstract thought, decreased verbal fluency, utilization or imitation behavior, or frontal release signs.
Possible	Either vertical supranuclear palsy or both slowing of vertical saccades & postural instability with falls < 1 yr disease onset.		
Probable	Vertical supranuclear palsy and prominent postural instability with falls within first year of disease onset.[a]		
Definite	All criteria for possible or probable PSP are met and histopathologic confirmation at autopsy.		

Adapted from Litvan et al., 1996[1]

[a] Later defined as falls or the tendency to fall (patients are able to stabilize themselves).

NINDS-SPSP = National Institute of Neurological Disorders and Stroke, and Society for Progressive Supranuclear Pals, Inc.

PSP = progressive supranuclear palsy.

[1] Litvan I, Agid Y, Calne D, et al. Clinical research criteria for the diagnosis of progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome): report on the NINDS-SPSP international workshop. Neurology 1996;47:1-9.

		Present	Absent		Primary	If Present: Contributing	Non-contrib.
15.	Corticobasal degeneration	☐ 1	☐ 0	15a.	☐ 1	☐ 2	☐ 3

Use the following criteria, excerpted from *SIC Task Force Appraisal of Clinical Diagnostic Criteria for Parkinsonian Disorders* (Litvan et al., 2003)[1]:

Proposed research criteria for CBD

Diagnostic categories	Inclusion criteria	Exclusion criteria
Lang et al[2]	Rigidity plus one cortical sign (apraxia, cortical sensory loss, or alien limb) *Or* Asymmetric rigidity, dystonia and focal reflex myoclonus.	Early dementia; early vertical gaze palsy; rest tremor; severe autonomic disturbances; sustained responsiveness to levodopa; lesions on imaging studies indicating another pathologic condition.
Kumar et al[3]	Chronic progressive course; asymmetric onset; presence of: "higher" cortical dysfunction (apraxia, cortical sensory loss, or alien limb); *And* Movement disorders – akinetic rigid syndrome-levodopa resistant, and limb dystonia and reflex; focal myoclonus.	

CBD = corticobasal degeneration.

Qualification of clinical features: rigidity, easily detectable without reinforcement; apraxia, more than simple use of limb as an object, clear absence of cognitive or motor deficit; cortical sensory loss, asymmetric, with preserved primary sensation; alien limb phenomena, more than simple levitation; dystonia, focal in limb, present at rest at onset; myoclonus, reflex myoclonus spreading beyond stimulated digits.

16.	Huntington's disease	☐ 1	☐ 0	16a.	☐ 1	☐ 2	☐ 3

Refer to the DSM-IV manual.

17.	Prion disease	☐ 1	☐ 0	17a.	☐ 1	☐ 2	☐ 3

Refer to the DSM-IV manual.

18.	Cognitive dysfunction from medications	☐ 1	☐ 0	18a.	☐ 1	☐ 2	☐ 3

Refer to the DSM-IV manual.

19.	Cognitive dysfunction from medical illnesses	☐ 1	☐ 0	19a.	☐ 1	☐ 2	☐ 3

Refer to the DSM-IV manual.

[1] Litvan I, Bhatia KP, Burn DJ, et al. Movement Disorders Society Scientific Issues Committee report: SIC Task force appraisal of clinical diagnostic criteria for Parkinsonian disorders. Mov Disord. 2003 May; 18(5):467-86.

[2] Lang AE, Riley DE, Bergeron C. Cortico-basal ganglionic degeneration. In: Calne DB, editor. Neurodegenerative diseases. Philadelphia: WB Saunders; 1994. p 877-894.

[3] Kumar R, Bergeron C, Pollanen MS, Lang AE. Cortical basal ganglionic degeneration. In: Jankovic J, Tolosa E, editors. Parkinson's disease and movement disorders. Baltimore: Williams and Wilkins; 1998. p 297-316.

		Present	Absent		If Present: Primary	Contributing	Non-contrib.
20.	Depression	☐ 1	☐ 0	20a.	☐ 1	☐ 2	☐ 3

DSM-IV[1] criteria as summarized in The Multiplex Family Study Procedures Manual (created by Columbia University for the Alzheimer's Disease Genetics Initiative):

1. At least one of the following three abnormal moods that significantly interfered with the person's life:
   a. Abnormal depressed mood most of the day, nearly every day, for at least 2 weeks.
   b. Abnormal loss of all interest and pleasure most of the day, nearly every day, for at least 2 weeks.
   c. If 18 or younger, abnormal irritable mood most of the day, nearly every day, for at least 2 weeks.
2. At least five of the following symptoms have been present during the same 2-week depressed period:
   a. Abnormal depressed mood (or irritable mood if a child or adolescent), as defined in 1a.
   b. Abnormal loss of all interest and pleasure, as defined in 1b.
   c. Appetite or weight disturbance, either:
      1) Abnormal weight loss (when not dieting) or decrease in appetite; or
      2) Abnormal weight gain or increase in appetite.
   d. Sleep disturbance, either abnormal insomnia or abnormal hypersomnia.
   e. Activity disturbance, either abnormal agitation or abnormal slowing (observable by others).
   f. Abnormal fatigue or loss of energy.
   g. Abnormal self-reproach or inappropriate guilt.
   h. Abnormal poor concentration or indecisiveness.
   i. Abnormal morbid thoughts of death (not just fear of dying) or suicide.
3. The symptoms are not due to a mood-congruent psychosis.
4. There has never been a Manic Episode, a Mixed Episode, or a Hypomanic Episode.
5. The symptoms are not due to physical illness, alcohol, medication, or street drugs.
6. The symptoms are not due to normal bereavement.

[1] Diagnostic and statistical manual of mental disorders (DSM-IVE). 4th ed. 1994, Washington, DC: American Psychiatric Assoc.

		Present	Absent		Primary	If Present: Contributing	Non-contrib.
21.	Other major psychiatric illness	☐ 1	☐ 0	21a.	☐ 1	☐ 2	☐ 3
	Refer to the DSM-IV manual.						
22.	Down's syndrome	☐ 1	☐ 0	22a.	☐ 1	☐ 2	☐ 3
	Refer to the DSM-IV manual.						
23.	Parkinson's disease	☐ 1	☐ 0	23a.	☐ 1	☐ 2	☐ 3

Use the following criteria, excerpted from *SIC Task Force Appraisal of Clinical Diagnostic Criteria for Parkinsonian Disorders* (Litvan et al., 2003). [These are the suggested criteria included in the most recent AAN Practice Parameter; the UDS Instructions for Neuropsychological Tests (Form C1) will include the Practice Parameter as soon as it is published]:

UK Parkinson's Disease Society Brain Bank clinical diagnostic criteria

Inclusion criteria	Exclusion criteria	Supportive criteria
Bradykinesia (slowness of initiation of voluntary movement with progressive reduction in speed and amplitude of repetitive actions); And at least one of the following: Muscular rigidity. 4–6 Hz rest tremor. Postural instability not caused by primary visual, vertibular, cerebellar, or proprioceptive dysfunction.	History of repeated strokes with stepwise progression of parkinsonian features. History of repeated head injury. History of definite encephalitis. Oculogyric crises. Neuroleptic treatment at onset of symptoms. More than one affected relative. Sustained remission. Strictly unilateral features after 3 yr. Supranuclear gaze palsy. Cerebellar signs. Early severe autonomic involvement. Early severe dementia with disturbances of memory, language, and praxis. Babinski sign. Presence of cerebral tumour or communicating hydrocephalus on CT scan. Negative response to large doses of levodopa (if malabsorption excluded). MPTP exposure.	(Three or more required for diagnosis of definite PD): Unilateral onset. Rest tremor present. Progressive disorder. Persistent asymmetry affecting side of onset most. Excellent response (70–100%) to levodopa. Severe levodopa-induced chorea. Levodopa response for 5 yr or more. Clinical course of 10 yr or more.

UK = United Kingdom; PD = Parkinson's disease; CT = computed tomography.

The following information, published as Tables 10 and 11 in the *SIC Task Force Appraisal of Clinical Diagnostic Criteria for Parkinsonian Disorders* (Litvan et al., 2003)TP[1]PT, may be useful for differential diagnosis of non-Alzheimer's dementia.

Table 1. Consensus Criteria for the Diagnosis of MSA

Clinical domain	Features	Criteria
Autonomic and urinary dysfunction	Orthostatic hypotension (by 20 mm Hg systolic or 10 mm Hg diastolic); urinary incontinence or incomplete bladder emptying.[a]	Orthostatic fall in blood pressure (by 30 mm Hg systolic or 15 mm Hg diastolic) and/or urinary incontinence (persistent, involuntary partial or total bladder emptying, accompanied by erectile dysfunction in men).[a]
Parkinsonism	B, R, I, and T.	1 of 3 (R, I, and T) and B.
Cerebellar dysfunction	Gait ataxia; ataxic dysarthria; limb ataxia; sustained gaze-evoked nystagmus.	Gait ataxia plus at least one other feature.
Corticospinal tract dysfunction	Extensor plantar responses with hyperreflexia.	No corticospinal tract features are used in defining the diagnosis of MSA.[b]

MSA = multiple system atrophy; B = bradykinesia; R = rigidity; I = postural instability; T = tremor.

[a] Note the different figures for orthostatic hypotension, depending on whether it us used as a feature or a criterion.

[b] In retrospect, this criterion is ambiguously worded. One possible interpretation is that, while corticospinal tract dysfunction can be used as a *feature* (characteristic of the disease), it cannot be used as a *criterion* (defining feature or composite of features required for diagnosis) in defining the diagnosis of MSA. The other interpretation is that corticospinal tract dysfunction cannot be used at all in consensus diagnostic criteria, in which case there is no point mentioning it.

Table 2. Consensus Diagnostic Categories and Exclusion Criteria for MSA

Diagnostic categories	Inclusion criteria	Exclusion criteria
Possible	One criterion plus two features from separate other domains. When the criterion is parkinsonism, a poor levodopa response qualifies as one feature (hence, only one additional feature is required).	*For possible and probable:* Symptomatic onset <30 yrs of age; Family history of a similar disorder; Systemic diseases or other identifiable causes for features listed in Table 1; Hallucinations unrelated to medication; DSM criteria for dementia; Prominent slowing of vertical saccades or vertical supranuclear gaze palsy; Evidence of focal cortical dysfunction such as aphasia, alien limb syndrome, and parietal dysfunction; Metabolic, molecular genetic, and imaging evidence of an alternative cause of features listed in Table 1.
Probable	One criterion for autonomic failure/urinary dysfunction plus poorly levodopa responsive parkinsonism or cerebellar dysfunction.	
Definite	Pathologically confirmed by the presence of a high density of glial cytoplasmic inclusions in association with a combination of degenerative changes in the nigrostriatal and olivopontocerebellar pathways.	

MSA = multiple system atrophy; DSM = Diagnostic and Statistical Manual for Mental Disorders.

[1] Litvan I, Bhatia KP, Burn DJ, et al. Movement Disorders Society Scientific Issues Committee report: SIC Task force appraisal of clinical diagnostic criteria for Parkinsonian disorders. Mov Disord. 2003 May; 18(5):467-86.

		Present	Absent		If Present: Primary	Contributing	Non-contrib.
24.	Stroke	☐ 1	☐ 0	24a.	☐ 1	☐ 2	☐ 3

Use the following criteria,[1] for stroke and recode either of two categories as "present":

Clinical Stroke
Rapidly developing clinical symptoms and/or signs of focal and at times global loss of cerebral function (applied to patients in deep coma and to those with subarachnoid hemorrhage) with symptoms lasting more than 24 hours or leading to death (therefore excluding transient ischemic attacks), and with no apparent cause other than vascular.

Patients are also classified as having an ischemic stroke if they had computed tomographic or magnetic resonance imaging of the brain done within 7 days of onset of symptoms that either identified the symptomatic cerebral infarct or failed to identify an alternative cause of the symptoms.

Silent Stroke
Strokes determined by neuroimaging alone for which there is no history nor clinical sign (aka "silent stroke") should be captured by the UDS. This capture will be less than 100% as not all ADC participants have the necessary imaging study available at the time the clinician is completing the UDS form. For this question, a silent stroke, as defined here, would be coded as "present."

25.	Hydrocephalus	☐ 1	☐ 0	25a.	☐ 1	☐ 2	☐ 3

Self-explanatory.

26.	Traumatic brain injury	☐ 1	☐ 0	26a.	☐ 1	☐ 2	☐ 3

Self-explanatory.

27.	CNS neoplasm	☐ 1	☐ 0	27a.	☐ 1	☐ 2	☐ 3

Self-explanatory.

28.	Other (*specify*): _____	☐ 1	☐ 0	28a.	☐ 1	☐ 2	☐ 3
29.	Other (*specify*): _____	☐ 1	☐ 0	29a.	☐ 1	☐ 2	☐ 3
30.	Other (*specify*): _____	☐ 1	☐ 0	30a.	☐ 1	☐ 2	☐ 3

If there is an observed cognitive impairment that is not due to any of the above-listed conditions, mark this category as "present", enter the type of condition, and indicate whether it is primary, contributing, or non-contributing.

[1] Cerebrovascular disease in the community: Results of a WHO Collaborative Study. (S Aho et al., 1980, Bull World Health Org., 58:113-130).

Telephone Follow-up Form E1: IMAGING/LABS

The purpose of this form is to record any imaging or tests performed since the subject's previous visit or previous visit, from which images or samples are available. The form should be completed by ADC or clinic staff.

Since the last visit, has neuroimaging been completed and available at your ADC?		**Film** Yes	No		**Digital image** Yes	No
1. Computed tomography	1a.	☐ 1	☐ 0	1b.	☐ 1	☐ 0
2. Magnetic resonance imaging – Clinical study	2a.	☐ 1	☐ 0	2b.	☐ 1	☐ 0
3. Magnetic resonance imaging – Research study/structural	3a.	☐ 1	☐ 0	3b.	☐ 1	☐ 0
4. Magnetic resonance imaging – Research study/functional	4a.	☐ 1	☐ 0	4b.	☐ 1	☐ 0
5. Magnetic resonance spectroscopy	5a.	☐ 1	☐ 0	5b.	☐ 1	☐ 0
6. SPECT	6a.	☐ 1	☐ 0	6b.	☐ 1	☐ 0
7. PET	7a.	☐ 1	☐ 0	7b.	☐ 1	☐ 0
Self-explanatory. If neuroimaging has been performed since the last visit and the files/data are available at your ADC, check "yes".						

Are specimens of the following available at your ADC?	**Yes**	**No**
8. DNA	☐ 1	☐ 0
9. Cerebrospinal fluid – ante-mortem	☐ 1	☐ 0
10. Serum/plasma	☐ 1	☐ 0
If specimens were collected and available at your ADC, check "yes".		

Is genotype data available at your ADC?	**Yes**	**No**
11. APOE	☐ 1	☐ 0
If APOE genotyping has been performed and the slides/files/data are available at your ADC, check "yes".		