Neurolinguistic and cognitive assessment for subtyping of primary progressive aphasia: Current practice

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Introduction to the European Reference Network for Rare Neurological Diseases (ERN-RND):

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ERN-RND unites 32 of Europe's leading expert centres in 13 Member States and includes highly active patient organizations. Centres are located in Belgium, Bulgaria, Czech Republic, France, Germany, Hungary, Italy, Lithuania, Netherlands, Poland, Slovenia, Spain and the UK.

The following disease groups are covered by ERN-RND:

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

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

Recommendation for clinical use:

The European Reference Network for Rare Neurological Diseases developed a current practice document for PPA to help guide the diagnosis of PPA subtypes in FTD patients.

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METHODOLOGY

Current practice document for PPA subtyping was done by the Disease group for FTD of ERN-RND.

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Development process:

- Listing of tests applied in ERN-RND FTD centres – November 2017 – April 2020
- Consent on diagnostic flowchart during ERN-RND annual meeting 2018 – 07/06/2018
- Discussion/Revision in ERN-RND disease group – June – September 2018
- Consent on document by whole disease group – June 2020

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Current practice document for neurolinguistic and cognitive assessment for PPA subtyping in FTD

This document lists a number of tests used at ERN-RND centres for subtyping of primary progressive aphasia in different EU languages: Dutch, English, French, German, Italian, Czech, Slovakian. The purpose is to serve as a practical guide to neurologists and other specialties for selecting tests that are currently used within ERN-RND for PPA subtyping. This may lead to more comprehensive assessment of different language and speech function and more accurate subtyping and communication of diagnosis and prognosis. The listing of tests is as they are currently used in the ERN-RND FTD centres and does not to pretend to be exhaustive or normative in any manner.

We distinguish between tests that are typically used by the clinician during the clinical assessment and tests to be administered by a neuropsychologist or speech therapist during a more formal and extensive neurolinguistic or neuropsychological assessment.

Two accessible clinical reviews explaining the basics of PPA subtyping are Vandenberghe, 2016, and Marshall et al, 2018.

This document remains within the framework of the Gorno-Tempini et al 2011 consensus guidelines (Gorno-Tempini et al, 2011). The purpose of this document is to provide a list of tests that can be used in clinical practice to operationalize the criteria, based on the current practice in the ERN-RND FTD centres. The authors are aware of the debates that are remaining about terminology and classification, the goal of the document is not to take a position in these debates. For instance, some authors draw a distinction between the nonfluent variant of primary progressive aphasia with predominant (or exclusively) speech apraxia, on the one hand, and primary progressive speech apraxia (Josephs et al, 2012), on the other hand. While this distinction may be theoretically and clinically of interest, the current document deals with both conditions within the framework of the consensus guidelines of Gorno-Tempini et al 2011, where, for clinical purposes, progressive speech apraxia would fall under the subtype nonfluent variant. This document is not meant to endorse one or the other view about progressive speech apraxia but to provide practical clinical tests for evaluating these language and speech disturbances. Likewise, outside of the three subtypes of the consensus criteria, other variants have been proposed, known under various names such as atypical PPA, PPA unclassifiable, mixed PPA (Mesulam et al 2012), or LV+. Again, the document does not take a position with regards to these additional subtypes and only provides a list of the tests that are applied in current practice to test different clinically relevant dimensions of PPA. Neither is the document meant to provide a systematic review of test for PPA subtyping and their validity metrics.

Schematically we distinguish three types of tests:

1. Widely used assessment instruments for use in aphasia in general and validated across many languages
2. Relatively concise language test batteries developed specifically for subtyping of PPA, which have been validated in a limited number of languages
    a. Sydney Language Battery (SYDBAT, Savage et al 2013) (also validated in Dutch)
    b. Screening for Aphasia in NeuroDegeneration (SAND) (also validated in Italian, Catricalà et al 2017)
3. Tests specific for a given language used for assessment of aphasia in general.

CLINICAL TESTS TO BE USED IN BY THE CLINICIAN DURING THE CLINICAL EVALUATION FOR PPA SUBTYPING

Root criterion of primary progressive aphasia

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There must be progressive aphasia. This can be documented by evaluating spontaneous speech, picture naming, word or sentence comprehension and/or sentence repetition. Preservation of other cognitive domains can be clinically evaluated by history (e.g. evidence for topographical memory or amnestic problems) or clinically (e.g. evidence for constructional problems in copying the overlapping pentagons).

The timecourse and preponderance of aphasia also is important. In PPA the language and speech problems should be leading and isolated for at least the first 12-24 months.

Is speech effortful and/or misarticulated?

Within the Gorno-Tempini et al recommendations for classification, speech apraxia is a characteristic feature of nonfluent variant (nfv) PPA. Clinical features characteristic of speech apraxia are the abnormal timing of speech, with lengthening of the duration of vowels or intersyllabic segments. Another useful feature is the effortfulness. Speech apraxia is most prominent for multisyllabic words and for words containing consonant clusters.

This can be assessed during confrontation naming, spontaneous speech or semispontaneous speech. Semispontaneous speech is typically elicited by the description of a scene, such as the Cookie Theft picture from the Boston Diagnostic Aphasia Examination, the PICNIC scene of the Western Aphasia Battery or the Summer Time picture of the SAND.

It can also be assessed during word repetition. To detect speech apraxia, repetition of multisyllabic nouns or nouns containing consonant clusters are useful. Examples are ‘constitutional’, ‘electricity’, ‘catastrophy’. The patient can also be asked to repeat a same multisyllabic word, such as ‘artillery’ or ‘cavalery’, three times.

It is critical to distinguish between repetition of words versus repetition of complex or long sentences/function words. Repetition of single words is useful as a test for speech apraxia, repetition of long sentences is useful as a test for the logopenic variant (lv) PPA.

Apraxia of speech is difficult to identify in clinical practice. It often mixes with dysarthria and the distinction between apraxia of speech and dysarthria may be difficult. As mentioned above, isolated apraxia of speech has also been referred to as ‘primary progressive apraxia of speech’ (Josephs et al, 2012). The aphasia features of nfvPPA are impairment of phonological encoding and of syntax. Apraxia of speech frequently adds to these language features. Presence of agrammatism may be clinically helpful for distinguishing aphasia from effortful speech in dysarthria.

Are there frequent grammatical errors?

Grammatical errors can clinically be assessed based on spontaneous speech or semispontaneous speech. Evaluate the length and complexity of the sentences and the presence of morphological or syntactic errors.

Sentence comprehension can be tested for reversible sentences (where word meaning does not disambiguate different possible thematic roles) or sentences with a non-canonical word order (i.e. “Peter is served by John. Who is the waiter?”).

Is repetition of phrases affected?

lvPPA typically fail on longer and more complex sentences, presumably due to phonological short-term memory deficits. Phrases that can be used are the phrases from the language subitem of the Montreal Cognitive Assessment (MoCA) test or the final few sentences of the AAT repetition test.

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The most sensitive are longer and semantically unpredictable sentences (i.e."He had no money, so he bought the whole pack of cigarettes")

Repetition of function words as in the MMSE is sensitive but not specific for the logopenic subtype.

Is understanding of word meaning affected?

This can clinically be evaluated by history or by asking the patient whether they recognize the word for words that they cannot retrieve during the Boston Naming test.

Single-word comprehension which is affected (mainly) in sv-PPA can be tested by pointing to pictures upon verbal entry.

STANDARDIZED TESTS WITH NORMATIVE DATA can be administered by the neuropsychologist or speech therapist to assess the different domains. This is non-restrictive list of tests that can help in differentiating between PPA subtypes. We distinguish three 'classes' of tests or test batteries

1. Normative tests such as Aachener Aphasie Test (AAT) (Huber et al, 1983) , Psycholinguistic Assessments of Language Processing in Aphasia (PALPA) (Kay et al 1992), the Comprehensive Aphasia Test (CAT) (Swinburn et al, 2005), or the Boston Diagnostic Aphasia Examination (Kaplan et al 1983, validated in French (Mazaux and Orgogozo 1982) and other European languages) are commonly used for language testing and available in nearly all EU languages. For these we will not mention the specific languages for which they have been validated (e.g. Mazeau and Orgogozo, 1986). An advantage of these tests is that they can be easily standardized across language groups. A disadvantage is that they have most often been developed mainly for testing types of aphasia other than PPA, most often stroke-induced aphasia. These tests will be referred to as 'Common' tests, referring both to their broad validation across languages and their use for aphasia in general.
2. A second class of tests has been specifically developed for PPA and is usually available in a limited number of languages: Sydney Language Battery (SYDBAT, Savage et al 2013) (also validated in Dutch), Screening for Aphasia in NeuroDegeneration (SAND) (also validated in Italian, Catricalà et al 2017). These sets of tests tailored to PPA often have to be validated per language. These tests will be referred to as PPA-specific tests.
3. Other tests have been developed for aphasia in general and are language-specific and for these tests, the language will be specifically mentioned. These will be referred to as language-specific tests.

CONFRONTATION NAMING

Common tests

1. Boston Naming test
2. Confrontation naming from the AAT, PALPA, or CAT

PPA specific tests

1. Naming part of the Semantic Association Test (SAT)
2. Naming subtask of the Sydney Language Battery (SYDBAT)
3. English and Italian: Picture Naming SAND (Catricalà et al 2017)

Language-specific tests

4. English: Graded Naming Test (Warrington & McKenna, 1980)

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5. Czech: Naming test of the Vyšetření fatických funkcí (VFF) - Czech language battery
6. Slovak: Test pomenovania obrázkov (Šteňová, Cséfalvay, 2011)
7. French: Picture Naming D080 (Deloche et al, 1997)
8. Italian: Test di denominazione visive (Sartori, 1988)

IS SPEECH EFFORTFUL AND/OR MISARTICULATED?

This can be assessed during testing of confrontation naming, repetition and semispontaneous speech.

Common tests

1. Repetition tests of AAT
2. Speech articulation subtasks of BDAE

PPA specific tests

3. Repetition test of SAND or SYDBAT

Language specific tests

1. Czech, Slovak: Repetition subtest of the comprehensive language batteries VFF (Czech); DgAAA (Slovak)
2. Dutch: Diagnostisch Instrument voor Apraxie van de Spraak (DIAS)
3. German: Repeat and Point Test, Hierarchische Wortlisten, a repetition test of hierarchical word lists

ARE THERE FREQUENT GRAMMATICAL ERRORS?

Common tests

1. Picture description (AAT, CAT)
2. Order execution subtask (BDAE)
3. Token test (working memory relevant to sentence comprehension)

PPA specific tests

4. Auditory sentence comprehension SAND

Language specific tests

5. Dutch:
    - Werkwoorden en Zinnen Test (WEZT)
    - Syntaxis uit de ScreeLing
6. English: Test of Reception of Grammar (TROG)
7. German: Komplexe Sätze, production of complex sentences

IS REPETITION OF PHRASES AFFECTED?

Common tests

1. Aachener Aphasie Test (AAT) repetition test
2. Repetition tests 12-14 of the CAT
3. Repetition subtask of BDAE

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PPA specific tests

4. Repetition subtask of the SYDBAT
5. Repetition of SAND

Language-specific tests

6. Czech, Slovak: Repetition subtest of the comprehensine language battery VFF (Czech); DgAAA (Slovak)
7. German: Repetition of the Aphasia Check List (ACL)
8. Dutch: Nazeggen uit subtest Fonologie van de ScreeLing

IS UNDERSTANDING OF WORD MEANING AFFECTED?

Common tests

1. Auditory and written single-word comprehension test (AAT, PALPA)
2. Associative Semantic test of the PALPA (subtest 45)
3. CAT auditory word comprehension (test 7) and written word comprehension (test 8)
4. BDAE subtest subtest II.02

PPA specific tests

5. Pyramids and Palm Trees test
6. Word comprehension and semantic association subtask of the SYDBAT or SAND

Language-specific tests

7. Dutch:
    - Test Relaties Abstracte concepten (TRACE)
    - ScreeLing substests Semantiek

8. English
    - Verbal and visual association subtask of the Semantic Association Task (SAT)
    - Abstract and concrete synonyms (Warrington et al., 1998)
    - British Picture Vocabulary Scale (Dunn & Whetton, 1982) (e.g. items 100-150 if there are time constraints)

9. German:
    - Auditory speech comprehension of the ACL

OBJECT IDENTIFICATION:

Common tests

1. BORB Object Decision test

PPA specific tests

2. Pyramids and Palm Trees test
3. Camel and Cactus Test
4. Semantic association subscale of the SAND

Language-specific tests

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5. Dutch: Semantische Associatie Test Benoemen
6. German: Kaffee und Kuchen Test

READING AND WRITING:

Common tests

1. Reading and writing subtests of the AAT.
2. Writing to dictation of the PALPA (subtest 42 and 43)
3. Written description of the CAT
4. Reading words of the CAT (test 20)

PPA specific tests

5. Reading and writing subtests of the SAND

Language-specific tests

6. Czech, Slovak: Reading and writing subtests from comprehensive language batteries VFF (Czech); DgAAA (Slovak)
7. Dutch: Hardop lezen screener DIAS

Abbreviations: AAT: Aachen Aphasie Test (translated in a wide range of languages); PALPA: Psycholinguistic Assessment of Language Processing in Aphasia (translated in a wide range of languages); SAND: Screening for Aphasia in Neurodegeneration (available in English and Italian); BDAE (translated in a wide range of languages); BORB: Birmingham Object Recognition Battery; PPT Pyramids and Palm Trees test (also available in a variety of languages eg French version (Merck et al. 2011); ACL Aphasie-Check-Liste

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