

GUIDED TOUR OF THE PRIMARY PROGRESSIVE APHASIA MODULE

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STRUCTURE OF THE PPA MODULE

ITEMS 1-3 ADDRESS THE ROOT DIAGNOSIS OF PPA.
ITEMS 4-13 ARE MEANINGFUL ONLY IF THE THE
ROOT DIAGNOSIS IS MADE
SO THE PATIENT
CAN BE CLASSIFIED INTO ONE OF THE THREE VARIANTS:
•NONFLUENT/AGRAMMATIC,
•SEMANTIC,
•LOGOPENIC.

(Gorno-Tempini et al., Neurology, 2011; 76:1006-1014)

**ROOT DIAGNOSIS OF PRIMARY PROGRESSIVE APHASIA
IS MADE IF ALL THREE CORE CRITERIA ARE MET**

(M. Mesulam, New England Journal of Medicine, 2003;348:1535-1542)

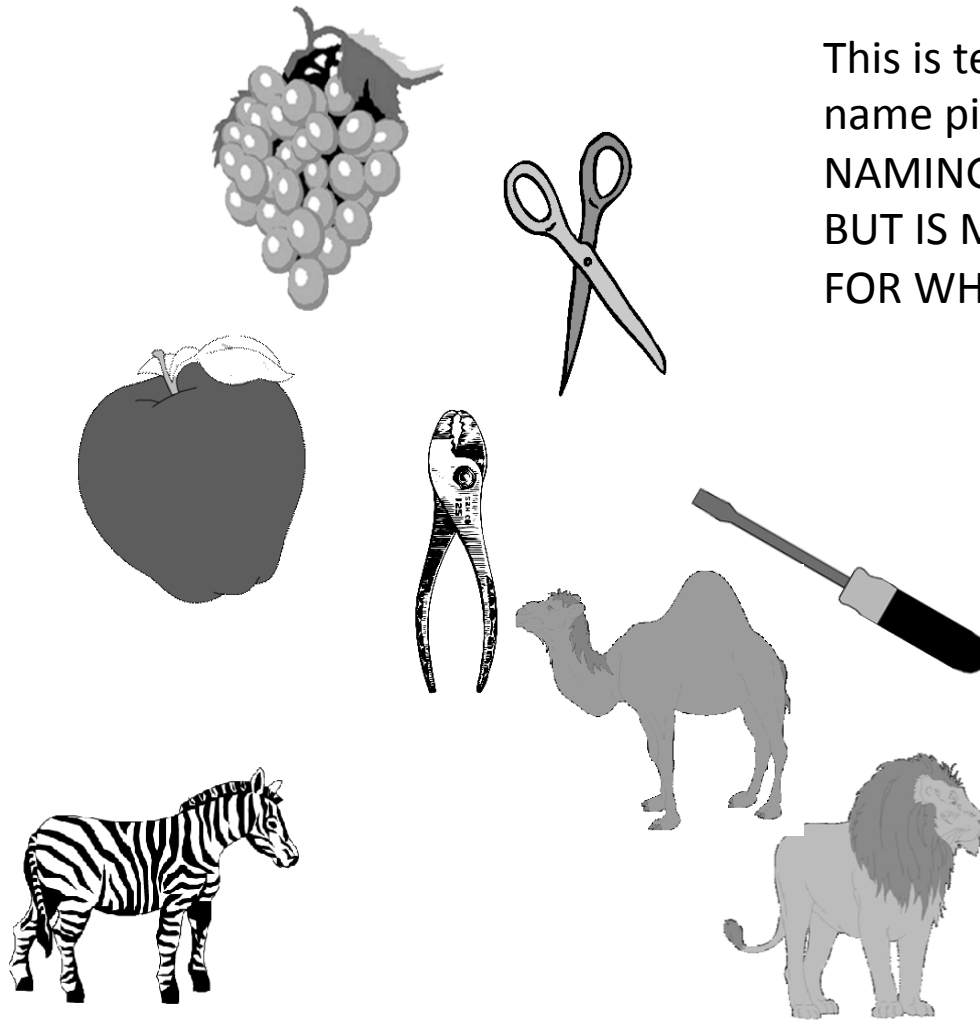
- A. There should be a language disorder of recent and insidious onset as manifested by gradually worsening impairments of word finding, usage or comprehension that cannot be attributed to more elementary motor or perceptual deficits.
(DETERMINED BY HISTORY, RECORDS, CLINICAL EXAMINATION)
- B. The underlying disease should be neurodegenerative, and all other causes should be excluded.
(DETERMINED BY IMAGING)
- C. The language impairment should constitute the most salient neurobehavioral deficit and the chief impediment to the pursuit of customary daily living activities during the initial stages of the illness. Consequential impairments of episodic memory, visuospatial processing, executive functions and comportment should be absent.
(DETERMINED BY HISTORY, CLINICAL EXAMINATION, NEUROPSYCHOLOGY)

QUESTION 1 ON FORM B9F IS ANSWERED 'YES' IF CRITERIA 'A' AND 'B' ARE BOTH MET.

QUESTIONS 2 AND 3 ARE BOTH 'YES' IF CRITERION 'C' IS MET.

IF ANY OF THE FIRST THREE QUESTIONS ON FORM B9F ARE EITHER NOT EVALUATED OR ANSWERED AS 'NO' THE PATIENTS DOES NOT HAVE PPA.

ITEM 4- POOR OBJECT NAMING



This is tested by asking the patient to name pictures of common objects.

NAMING CAN BE IMPAIRED IN ALL VARIANTS BUT IS MOST SEVERE IN THE SEMANTIC TYPE FOR WHICH IT IS A CORE CRITERION.

ITEM 5- IMPOVERISHED WORD SELECTION/RETRIEVAL

This aspect of PPA can be detected by giving the patient a picture of a scene that depicts an action (such as a picnic or sporting event) and asking for a narrative description. The presence of this feature is determined by word-finding pauses. Output is simplified and poor in content words such as verbs and nouns.

IMPOVERISHED WORD SELECTION/RETRIEVAL
OCCURS IN ALL PPA VARIANTS AND IS A CORE CRITERION FOR THE
LOGOPENIC TYPE.

ITEM 6- GRAMMATICAL SIMPLIFICATION OR ERRORS

Testing grammatical competence is probably the most challenging part of the assessment. When severe, this feature leads to abnormalities of verb tense agreement ('yesterday he go'), plural designations ('mices' rather than 'mice') and word order (syntax). Small grammatical words are omitted and output is telegraphic but informative.

THIS IS A CORE FEATURE OF THE NONFLUENT/AGRAMMATIC TYPE
AND TENDS TO BE MORE OBVIOUS IN WRITTEN THAN SPOKEN LANGUAGE.

ITEM 7- EFFORTFUL, HALTING SPEECH

This feature refers to the articulatory and executive aspects of speech, not to word-finding, which is addressed in item 5.

The speech abnormality designated by this item is distinct from upper or lower motoneuron dysarthria, Parkinsonian hypophonia, or cerebellar scanning speech. The type of speech disorder designated by item 7 is also referred to as apraxia of speech or phonological disintegration. Speech is effortful, occasionally distorted and very deliberate as if the patient has "marbles in the mouth."

THIS ITEM CONSTITUTES A CORE CRITERION FOR THE NONFLUENT/AGRAMMATIC TYPE AND ONE OF THE ANCILLARY EXCLUSIONARY CRITERIA FOR THE SEMANTIC TYPE.

ITEM 8- SPEECH SOUND/WORD ERRORS (PARAPHASIA)

This feature is detected either by listening to spontaneous speech or asking the patient to name objects. It should be differentiated from articulatory distortions that arise because of dysarthria or the type of speech disorder designated by item 7. A phonemic paraphasia is identified when there is a sound substitution in an otherwise correct word (for example, naming a pencil as 'plentil'). A semantic paraphasia is identified when an entirely different word that belongs to the same general category is substituted (for example, calling a radio a 'music').

PHONOLOGICAL PARAPHASIA IS AN ANCILLARY CRITERION FOR THE LOGOPENIC TYPE OF PPA AND CAN ALSO BE SEEN IN IN THE NONFLUENT/AGRAMMATIC TYPE.

SEMANTIC PARAPHASIAS ARE MOST COMMON IN SEMANTIC PPA.

ITEM 9- IMPAIRED WORD COMPREHENSION

This item is detected when the patient is unable to define the meaning of words. In addition to anomia, the patient also displays an inability to point to the appropriate object when the noun is provided by the examiner. THIS IS A CORE FEATURE OF THE SEMANTIC PPA TYPE AND AN ANCILLARY EXCLUSIONARY CRITERION FOR THE OTHER TWO TYPES.

ITEM 10- POOR OBJECT/PERSON KNOWLEDGE

This item designates impaired object (or face) recognition, not just anomia. In testing for this feature, the patient is usually given a picture and asked "what (or who) is this?" If naming fails, the examiner should produce the name and see if the patient points to the correct item, in which case a retrieval rather than recognition failure is reported. If pointing fails, the examiner should elicit a description of the item. If the patient provides plausible descriptions such as "an animal that has one or two humps and carries stuff in the desert" or "she is the head of England," than knowledge (recognition) is considered to be preserved. If the definition also fails, it is important to rule out a more basic perceptual impairment by asking the patient to identify the two faces or objects in the display that are identical.

If the patient cannot name the item, cannot point to it upon hearing its name, and has no perceptual impairment that would interfere with the sensory encoding of the object, the examiner can diagnose a failure of knowledge. POOR OBJECT/PERSON KNOWLEDGE IS AN ANCILLARY FEATURE OF THE SEMANTIC PPA TYPE AND AN ANCILLARY EXCLUSIONARY FEATURE OF THE OTHER TWO TYPES.



ITEM 11- CIRCUMLOCUTORY EMPTY SPEECH

This feature is almost always seen in conjunction with word-finding impairments. It reflects the patient's attempt to fill the void by words that approximate the intended message. The output is poor in verbs and nouns and contains many fillers such as "the thing," "stuff," "whachamacallit." The patient tries to convey the description of an object or scene with circumlocutions. This aspect of PPA is similar to the one designated by item 5 but without prominent word-finding pauses. CIRCUMLOCUTORY EMPTY SPEECH CAN BE PRESENT IN THE LOGOPENIC AND SEMANTIC TYPES OF PPA.

ITEM 12- SURFACE DYSLEXIA & DYSGRAPHIA

Words can be read and spelled through two interactive routes:

- A. By applying the rules of grapheme to phoneme conversion,
- B. By retrieving the entire orthographic pattern through stored expert knowledge of the word.

Spelling and reading familiar words that obey the phonological rules of English can be achieved through either route.

Damage to route A impairs the ability to spell and read plausible non-words (such as 'frimkin') whereas damage to route B impairs the ability to spell and read familiar but irregular words such as 'shoe' or 'soldier'. Damage to route A causes phonological dyslexia/dysgraphia whereas damage to route B causes surface dyslexia/dysgraphia.

The patient with surface dyslexia/dysgraphia spells and reads irregular words phonetically, that is *regularizes* them. For example, when the word 'shoe' is heard, it may be spelled as 'shew' or 'soldier' is spelled as 'solgier.' This type of impairment (inability to spell irregular but not regular words or nonwords) is known as surface dyslexia.

SURFACE DYSLEXIA/DYSGRAPHIA IS AN ANCILLARY FEATURE OF THE SEMANTIC PPA TYPE.

ITEM 13- POOR REPETITION

This item is tested by asking the patient to repeat phrases and sentences. Repetition impairments due to dysarthria do not count. Asking the patient to repeat a string of words that have no experiential associations (e.g., no, and, under) is usually more sensitive for eliciting this feature than meaningful phrases or tongue twisters.

POOR REPETITION IS A CORE CRITERION FOR LOGOPENIC PPA.

IT CAN ALSO EXIST IN THE NONFLUENT/AGRAMMATIC TYPE BUT SHOULD NOT BE PRESENT IN THE SEMANTIC TYPE.

ITEM 14- DOES THE PATIENT HAVE PPA?

ROOT DIAGNOSIS OF PRIMARY PROGRESSIVE APHASIA IS MADE IF ALL THREE CORE CRITERIA ARE MET

- A. There should be a language disorder of recent and insidious onset as manifested by gradually worsening impairments of word finding, usage or comprehension that cannot be attributed to more elementary motor or perceptual deficits.
- B. The underlying disease should be neurodegenerative, and all other causes should be excluded.
- C. The language impairment should constitute the most salient neurobehavioral deficit and the chief impediment to the pursuit of customary daily living activities during the initial stages of the illness.

ITEM 15- SUBTYPING

VIEWS & REVIEWS

Classification of primary progressive aphasia and its variants

Gorno-Tempini et al., Neurology, 2011

(CORE CRITERIA ARE IN RED,
ANCILLARY CRITERIA IN BLUE)

NONFLUENT/AGRAMMATIC

AGRAMMATISM IN LANGUAGE PRODUCTION

OR

EFFORTFUL APRAXIC SPEECH

+

TWO OF THREE ANCILLARY FEATURES

1. IMPAIRED COMPREHENSION OF COMPLEX SYNTAX
2. SPARED WORD COMPREHENSION
3. SPARED OBJECT KNOWLEDGE

SEMANTIC

IMPAIRED OBJECT NAMING
AND

IMPAIRED WORD COMPREHENSION

+

THREE OF FOUR ANCILLARY FEATURES

1. IMPAIRED OBJECT KNOWLEDGE
2. SURFACE DYSLEXIA/DYSGRAPHIA
3. SPARED REPETITION
4. SPARED GRAMMAR AND SPEECH

LOGOPENIC

IMPAIRED WORD RETRIEVAL
AND

IMPAIRED REPETITION

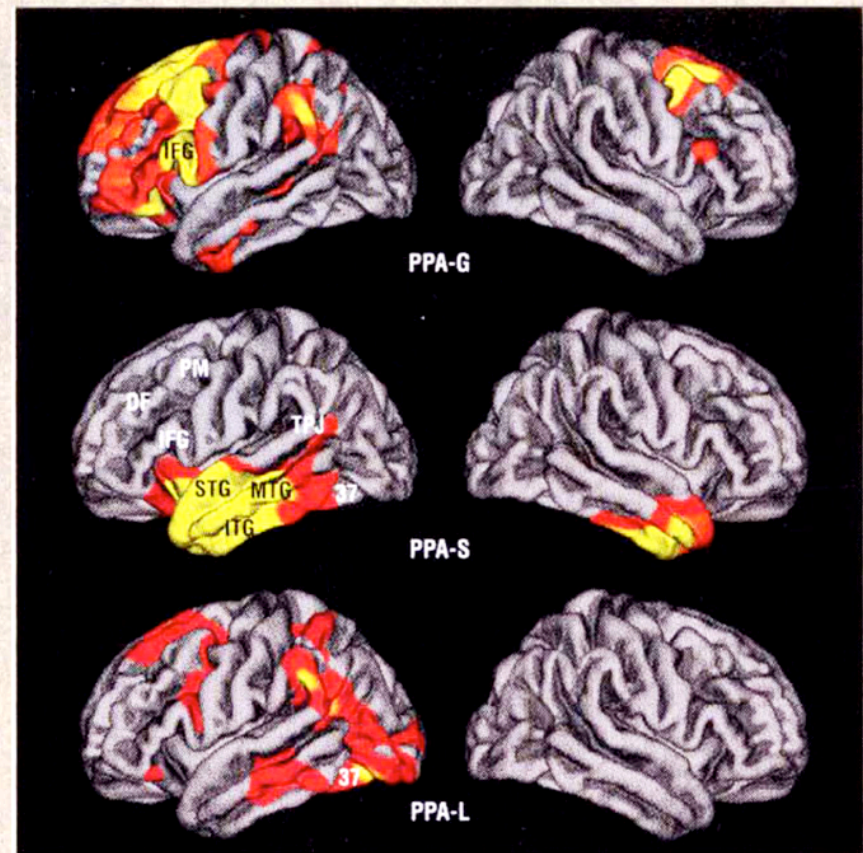
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THREE OF FOUR ANCILLARY FEATURES

1. PHONEMIC SPEECH ERRORS
2. SPARED WORD COMPREHENSION & OBJECT KNOWLEDGE
3. SPARED SPEECH
4. NO MAJOR AGRAMMATISM

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