Primary Progressive Aphasia:

A Language Dementia

Sandra Weintraub PhD, ABCN/ABPP
Clinical Core Leader
Professor of Psychiatry, Neurology and Psychology
Northwestern Alzheimer’s Disease Center
Clinical Core Leader
I have no financial relationships to disclose:

Employee of: Northwestern University
Consultant for: None
Stockholder in: None
Research support from: NIA, NIDCD, NINDS
Honoraria from: None
OBJECTIVES

Discuss the diagnostic criteria and expected neuro-imaging and neuro-pathology associations for primary progressive aphasia and its subtypes.
OUTLINE

Same Page Context: What is Dementia and where does PPA fit?
History: PPA: A Language Dementia
Neuropsychology: Language and Non Language
Clinical Diagnosis:
  PPA and Subtypes
  Neuroimaging Patterns
  Neuropathology
Interventions
DEMENTIA: A CLINICAL SYNDROME

- Progressive *DECLINE from a prior level in*: any one or more cognitive and/or behavioral functions (e.g., memory, language, judgment)

- interferes with customary activities and social relationships, causes dependence

- Caused by intrinsic brain disease
CAUSES OF DEMENTIA

Neurodegenerative: NERVE CELL DEATH selective for cognitive neuroanatomical networks

- Non Alzheimer’s Disease
  - Diffuse Lewy Body
  - Prion Diseases
  - Others
- Alzheimer’s Disease
  - Amyloid Plaques, Tau Tangles
- Frontotemporal Lobar Degeneration
  - Tauopathies (e.g. Pick disease, CBD, PSP, others)
  - TDP-43 Proteinopathies

Other
- Vascular
- Metabolic
- Tumor
- Reversible
- Others
  - FUS, other
3 LEVELS OF DEMENTIA DESCRIPTION RECOGNIZED IN LATEST DIAGNOSTIC CRITERIA

EARLY CLINICAL/NEUROPSYCHOLOGICAL SYNDROME (E.G., APHASIA, AMNESIA, BEHAVIOR)

NEUROANATOMICL CHANGES ATROPHY, PHYSIOLOGICAL DYSFUNCTION IN RELEVANT NETWORK (E.G., LEFT HEMISPHERE=LANGUAGE)

POST MORTEM NEUROPATHOLOGIC DISEASE (ALZHEIMER VS TAUOPATHY VS TDP-43 PROTEINOPATHY)

Weintraub in Dickerson and Atri, 2014
EARLY PROFILE: AMNESIA
AKA “Dementia of the AD type”

EARLY PROFILE: APHASIA
Primary Progressive Aphasia

AD
Neuropathology

FTLD
Neuropathology

OTHER
CJD, Cortical Lewy Body

WEINTRAUB IN DICKERSON AND ATRI, 2014

Do not copy or distribute without permission
HISTORY OF PPA

Pick, 1892
Sérieux, 1893
Pick, 1904
Franceschi, 1908
Rosenfeld, 1909

Mesulam, 1982: 6 cases of “Slowly Progressive Aphasia”


Gorno-Tempini et al, 2011: PPA Subtypes Diagnostic Criteria
Primary Progressive Aphasia: A Language Dementia

According to currently accepted criteria, the PPA diagnosis is made in any patient in whom a language impairment (aphasia), caused by a neurodegenerative disease (progressive), constitutes the most salient aspect of the initial clinical presentation (primary).

LANGUAGE IS EARLIEST AND MOST PROMINENT SYMPTOM

Percent Change in Test Scores Over 2 Years

**Language and Related Tests**
- Auditory Comprehension
- Repetition-Words
- Repetition-Sentences
- Oral Reading- Words
- Oral Reading-Sentences
- Confrontation Naming
- Word Fluency
- Reading Comprehension
- Praxis-Buccofacial
- Praxis Limb
- Calculation

**Non Language Tests**
- Memory-Orientation
- Memory 3W3S
- Line Orientation
- Facial Recognition
- Hooper VOT
- Reasoning-Raven’s Matrices
- Reasoning- Shipley
- Reasoning-Visual-Verbal
Neuropsychological Assessment
COMPONENTS OF LANGUAGE

Aphasia Severity (Western Aphasia Battery AQ)
Spontaneous Speech (fluency, content, grammar)
Language Semantics (naming, word comprehension)
Grammar (production, comprehension; morphology)
Reading (single word, regular vs irregular)
Writing (form, semantic content, grammar)
Single Word Comprehension
Semantic Word-Picture Matching
FTLD Module of the Uniform Data Set
https://www.alz.washington.edu/WEB/forms_ftld.html
Noun and Verb Naming
FTLD Module of the Uniform Data Set
https://www.alz.washington.edu/WEB/forms_ftld.html
Challenges For Testing Grammar Production

Motor speech disorders and word finding difficulty interfere with output.

Therefore, need a test of grammatical processing that:
- eliminates speech production
- reduces working memory load
- reduces impact of word finding deficits
NORTHWESTERN ANAGRAM TEST (NAT)
Weintraub, Mesulam, Thompson
Table 1. Samples of Canonical and Noncanonical Sentence Types on the Northwestern Assessment of Verbs and Sentences (NAVS)

<table>
<thead>
<tr>
<th>Type of Sentence</th>
<th>Structure</th>
<th>Example</th>
</tr>
</thead>
<tbody>
<tr>
<td>Canonical</td>
<td>Actives</td>
<td>The groom is carrying the bride</td>
</tr>
<tr>
<td></td>
<td>Subject-extracted</td>
<td>Who is carrying the bride?</td>
</tr>
<tr>
<td></td>
<td>Wh-questions</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Subject clefts</td>
<td>It was the groom who carried the bride</td>
</tr>
<tr>
<td>Noncanonical</td>
<td>Passives</td>
<td>The bride was carried by the groom</td>
</tr>
<tr>
<td></td>
<td>Object-extracted</td>
<td>Who is the groom carrying?</td>
</tr>
<tr>
<td></td>
<td>Wh-questions</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Object clefts</td>
<td>It was the bride who the groom carried</td>
</tr>
</tbody>
</table>
Consensus guidelines for unifying classification of PPA and its three variants, Agrammatic (aka PPA-G), Semantic (aka PPA-S) and Logopenic (aka PPA-L)

1. **Clearly delimits clinical diagnosis from sources of supporting evidence and etiology (i.e., structural/functional imaging, neuropathology)**

2. Detailed clinical descriptors

3. Supportive Neuroimaging (must fulfill clinical criteria AND have imaging evidence)

4. Supportive Neuropathology (must fulfill clinical criteria AND have post mortem autopsy verification or known genetic mutation)

5. Systematic data collection

Caveat: clinical instruments not specified; needs validation
Root Clinical Diagnosis of PPA (Mesulam 2003):

1. Clinical Examination
   - Language deficits (e.g., anomia, word comprehension) most prominent
   - Language deficits cause difficulties in activities of daily living, otherwise normal
   - Language deficits earliest and most prominent thereafter

2. Exclusionary Criteria
   - Non-neurodegenerative explanation (e.g., stroke, neoplasm, psychiatric)
   - Prominent episodic memory and visuospatial/perceptual deficits early
   - Prominent behavioral disturbance early

Non Fluent Agrammatic PPA (PPA-G):

1. Agrammatism in language production  OR
2. Effortful, halting speech (“apraxia of speech”)

PLUS 2 OF 3:
1. Poor syntactic comprehension
2. Spared single word comprehension
3. Spared object knowledge

IMAGING-SUPPORTED (clinical features must be present)
1. Fronto-insular atrophy on MRI
2. Posterior left fronto-insular hypoperfusion or hypometabolism (PET, SPECT)

WITH DEFINITE PATHOLOGY (clinical features must be present)
1. Post mortem histopathologic diagnosis OR
2. Presence of a known pathogenic mutation
PPA-Agrammatic

PPA-Semantic

PPA-Logopenic
Semantic Variant PPA (PPA-S):

1. Poor confrontation naming of objects AND
2. Impaired single word comprehension

PLUS at least 3:
1. Poor object knowledge
2. Surface dyslexia and/or dysgraphia
3. Spared repetition
4. Spared motor speech and grammar

IMAGING-SUPPORTED
1. Anterior temporal lobe atrophy and/or
2. Anterior temporal hypoperfusion or hypometabolism (PET, SPECT)

WITH DEFINITE PATHOLOGY
1. Post mortem histopathologic evidence OR
2. Presence of a known pathogenic mutation

Logopenic Variant PPA (PPA-L):

1. Impaired single word retrieval in spontaneous speech and naming AND
2. Impaired repetition of sentences and phrases

PLUS at least 3:
1. Phonological errors in speech and naming
2. Spared single word comprehension and object knowledge
3. Spared motor speech
4. Absence of frank agrammatism

IMAGING-SUPPORTED
1. Left posterior perisylvian or parietal atrophy on MRI AND OR
2. Left posterior perisylvian or parietal hypoperfusion or hypometabolism (PET, SPECT)

WITH DEFINITE PATHOLOGY
1. Post mortem histopathologic evidence
2. Presence of a known pathogenic mutation

Dissociations Between Fluency And Agrammatism In Primary Progressive Aphasia
Aphasiology, 2012

□=PPA-G; □=PPA-L; △=PPA-S

Do not copy or distribute without permission
DISSOCIATION OF FLUENCY FROM GRAMMAR IN THE FRONTAL LOBES: NAT VS MLU

Rogalski et al, J. Neurosci., 2011
NEUROPSYCHOLOGY OF PPA: HOW DO YOU DEMONSTRATE THE RELATIVE PRESERVATION OF NON LANGUAGE FUNCTIONS?

CHALLENGES FOR TESTING NON LANGUAGE DOMAINS

Tests Contain Complex Verbal Instructions: Block Designs, WCST, Picture Completion

Tests Contain Verbal Stimuli: Word list memory, story memory, MMSE, Trail Making B

Tests Require Spoken Output: Similarities, word list/story memory, MMSE

CHOOSE TESTS THAT MINIMIZE VERBAL INTERFERENCE
Neuropsychological Assessment:

NON LANGUAGE FUNCTIONS

Retentive episodic memory
Visuospatial perception
Object perception
Reasoning, cognitive flexibility
Executive functions (attention, working memory, shifting)
Activities of Daily Living
DEMENTIA SEVERITY: MMSE

Annualized percentage change on the MMSE and the ADLQ

Change Scores

<table>
<thead>
<tr>
<th>% Change</th>
<th>MMSE</th>
<th>ADLQ</th>
</tr>
</thead>
<tbody>
<tr>
<td>-25%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>-20%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>-15%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>-10%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>-5%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>0%</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* p < .05

Osher, Wicklund, Rademaker, Johnson, Weintraub, 2007
LEVEL OF IMPAIRMENT MEASURED BY THE ADLQ
IN THREE NEUROPSYCHOLOGICAL
PROFILES OF DEMENTIA

PERCENT IMPAIRMENT

SEV

MOD

MILD

AD
FTD
PPA

ADLQ TOTAL
SELF CARE
HOUSE CARE
WORK REC
SHOP MONEY
TRAVEL
COMMUNICATION

Wicklund et al, 2007
Do not copy or distribute without permission
Visual-Verbal Test (Feldman & Drasgow, 1959)

SORT 1

SORT 2

Do not copy or distribute without permission
Wicklund, Johnson, Weintraub, 2004

**SHIFTS**

- NC
- ONC
- PPA

**SORTS**

- NC
- ONC
- PPA

---

Do not copy or distribute without permission
THREE WORDS THREE SHAPES MEMORY TEST (3W3S)
Verbal and Nonverbal Material
Both in Visual Modality

<table>
<thead>
<tr>
<th>CONDITION</th>
<th>COMPONENT TESTED</th>
</tr>
</thead>
<tbody>
<tr>
<td>COPY</td>
<td>Assures attention to stimuli, verifies impact of writing and drawing ability on performance</td>
</tr>
<tr>
<td>EFFORTLESS ENCODING</td>
<td>How much is remembered without forewarning?</td>
</tr>
<tr>
<td>ACQUISITION TRIALS</td>
<td>Two additional learning trials (30 secs each) followed by immediate reproduction to assure encoding</td>
</tr>
<tr>
<td>DELAYED RECALL</td>
<td>Spontaneous retrieval after a delay</td>
</tr>
<tr>
<td>MULTIPLE CHOICE</td>
<td>Recognition (retention) after a delay</td>
</tr>
</tbody>
</table>

Weintraub et al, 2003
THREE WORDS THREE SHAPES TEST

PPA (Aphasic Dementia)

Cognitively Normal Control

DAT (Amnestic Dementia)

RECOGNITION

PPA Similar to Normal Control For Both Words and Shapes

DAT worse than PPA and Control For Both Words and Shapes

Weintraub et al, Behavioural Neurolog 2012

Do not copy or distribute without permission
THREE WORDS THREE SHAPES TEST

PPA (Aphasic Dementia)

DAT (Amnestic Dementia)

Cognitively Normal Control

RECOGNITION

PPA Similar to Normal Control For Both Words and Shapes

DAT worse than PPA and Control For Both Words and Shapes

Weintraub et al, Behavioural Neurology 2012

Do not copy or distribute without permission
THREE WORDS THREE SHAPES TEST

PPA (Aphasic Dementia)

Cognitively Normal Control

DAT (Amnestic Dementia)

**RECOGNITION**

PPA Similar to Normal Control For Both Words and Shapes

DAT worse than PPA and Control For Both Words and Shapes

Weintraub et al, Behavioural Neurolog 2012

Do not copy or distribute without permission
THREE WORDS THREE SHAPES TEST

PPA (Aphasic Dementia)

Cognitively Normal Control

DAT (Amnestic Dementia)

RECOGNITION

PPA Similar to Normal Control For Both Words and Shapes

DAT worse than PPA and Control For Both Words and Shapes

Weintraub et al, Behavioural Neurolog 2012

Do not copy or distribute without permission
3W 3S
PPA

Effortless Encoding

15 minute Delay

30 min Delay

Do not copy or distribute without permission
Northwestern Naming Battery- Semantic Associates Subtest

- Sweater
- Blanket
- Pillow
Northwestern Naming Battery - Semantic Associates Subtest
Northwestern Naming Battery- Semantic Associates Subtest

http://northwestern.flintbox.com/public/project/22014/ Thompson and Weintraub
Do not copy or distribute without permission
PPA Subtyping Algorithm

1. Single Word Comprehension- PPVT IV
2. Grammar- NAT
3. Naming- BNT
4. WAB Repetition
5. BDAE Fluency Rating (scale of 7)

Mesulam, Wienecke, Rogalski, Cobia, Thompson, Weintraub, Arch Neurol 2009
PPA Subtyping Algorithm

(1) PPA-S if the PPVT-4 score is less than 60% and the NAT score is 60% or greater

(1) PPA-G if the NAT score is less than 60% and the PPVT-4 score is 60% or greater

(1) PPA-L if the PPVT-4 and NAT scores are both 60% or greater

(1) Mixed PPA if the PPVT-4 and NAT scores are both less than 60%.
FGD PET HYPOMETABOLISM IN LEFT CEREBRAL HEMISPHERE IN PPA

Alzheimer and frontotemporal pathology in subsets of primary progressive aphasia

<table>
<thead>
<tr>
<th>Subject</th>
<th>Early Problems of Memory/Behavior</th>
<th>Neuropathology</th>
<th>Aphasia Subtype</th>
<th>Asymmetry</th>
<th>ATAC</th>
<th>TDP-43</th>
<th>Braak Stage</th>
<th>Onset Age/Sex</th>
<th>Death Age</th>
<th>ApoE</th>
<th>Imaging</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>No/No</td>
<td>AD</td>
<td>Mixed</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>6</td>
<td>58 / M</td>
<td>73</td>
<td>3.4</td>
<td>MRI=atrophy, most prominent L temporal lobe</td>
</tr>
<tr>
<td>2</td>
<td>No/No</td>
<td>AD</td>
<td>Logopenic</td>
<td>sm (F, T, P) L&gt;R</td>
<td>Multiple</td>
<td>No</td>
<td>6</td>
<td>48 / F</td>
<td>59</td>
<td>3.3</td>
<td>SPECT=decreased perfusion R hemisphere</td>
</tr>
<tr>
<td>3*</td>
<td>No/No</td>
<td>AD</td>
<td>Mixed</td>
<td>a (T) L&gt;R</td>
<td>No</td>
<td>No</td>
<td>6</td>
<td>51 / M</td>
<td>72</td>
<td>3.3</td>
<td>CT=L perisylvian atrophy</td>
</tr>
<tr>
<td>4*</td>
<td>No/No</td>
<td>AD</td>
<td>Semantic</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>6</td>
<td>70 / F</td>
<td>76</td>
<td>4.4</td>
<td>MRI=small cerebellar infarct, otherwise unremarkable</td>
</tr>
<tr>
<td>5*</td>
<td>No/No</td>
<td>AD</td>
<td>Logopenic</td>
<td>a (T) L&gt;R</td>
<td>No</td>
<td>No</td>
<td>6</td>
<td>67 / M</td>
<td>73</td>
<td>3.3</td>
<td>MRI=Non-specific subcortical changes, SPECT=normal</td>
</tr>
<tr>
<td>6</td>
<td>No/No</td>
<td>AD</td>
<td>Logopenic</td>
<td>sm (F) L&gt;R</td>
<td>No</td>
<td>No</td>
<td>6</td>
<td>71 / M</td>
<td>78</td>
<td>3.4</td>
<td>SPECT=L temporoparietal hypoperfusion</td>
</tr>
<tr>
<td>7</td>
<td>No/No</td>
<td>AD</td>
<td>Mixed</td>
<td>a (H, T, P) L&gt;R</td>
<td>Multiple</td>
<td>No</td>
<td>6</td>
<td>59 / M</td>
<td>71</td>
<td>3.4</td>
<td>MRI=unremarkable</td>
</tr>
<tr>
<td>8</td>
<td>No/No</td>
<td>AD</td>
<td>Logopenic</td>
<td>sm (F, T, P) L&gt;R</td>
<td>Single Cluster</td>
<td>No</td>
<td>6</td>
<td>71 / F</td>
<td>78</td>
<td>3.3</td>
<td>MRI=L, perisylvian atrophy</td>
</tr>
<tr>
<td>9*</td>
<td>No/No</td>
<td>AD</td>
<td>Logopenic</td>
<td>a, nlg (T) L&gt;R</td>
<td>Single Cluster</td>
<td>ND</td>
<td>5</td>
<td>47 / M</td>
<td>70</td>
<td>3.3</td>
<td>PET=L, perisylvian and parietofrontal hypometabolism</td>
</tr>
<tr>
<td>10</td>
<td>No/No</td>
<td>AD</td>
<td>Logopenic</td>
<td>a, sm (F, T, P) L&gt;R</td>
<td>Single Cluster</td>
<td>No</td>
<td>6</td>
<td>58 / F</td>
<td>66</td>
<td>3.3</td>
<td>PET=L, temporoparietal hypometabolism</td>
</tr>
<tr>
<td>11</td>
<td>No/No</td>
<td>AD</td>
<td>Logopenic</td>
<td>?</td>
<td>ND</td>
<td>ND</td>
<td>6</td>
<td>80 / M</td>
<td>87</td>
<td>ND</td>
<td>MRI=atrophy L temporal pole and bilateral frontal</td>
</tr>
<tr>
<td>12</td>
<td>No/No</td>
<td>FTLD-U</td>
<td>Logopenic</td>
<td>?</td>
<td>Yes</td>
<td>0</td>
<td>53 / M</td>
<td>62</td>
<td>3.3</td>
<td>MRI=L, perisylvian atrophy; PET=L temp hypoperfusion</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>No/No</td>
<td>FTLD-U</td>
<td>Agrammatic</td>
<td>a (F, T, P) L&gt;R</td>
<td>Yes</td>
<td>2</td>
<td>56 / F</td>
<td>62</td>
<td>3.3</td>
<td>SPECT=decreased L frontotemporal hypoperfusion</td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>No/No</td>
<td>FTLD-U</td>
<td>Logopenic</td>
<td>No</td>
<td>Yes</td>
<td>2</td>
<td>63 / F</td>
<td>69</td>
<td>3.4</td>
<td>MRI=unremarkable</td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>No/No</td>
<td>FTLD-U</td>
<td>Mixed</td>
<td>No</td>
<td>Yes</td>
<td>2</td>
<td>65 / F</td>
<td>69</td>
<td>3.4</td>
<td>MRI=unremarkable; SPECT=L, temporal hypoperfusion</td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>No/No</td>
<td>FTLD-U</td>
<td>Logopenic</td>
<td>a, nlg (T) L&gt;R</td>
<td>Yes</td>
<td>2</td>
<td>58 / M</td>
<td>65</td>
<td>3.3</td>
<td>MRI=atrophy L frontotemporal</td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>No/No</td>
<td>FTLD-T</td>
<td>Agrammatic</td>
<td>a, nlg (F, T, P) L&gt;R</td>
<td>No</td>
<td>0</td>
<td>45 / F</td>
<td>60</td>
<td>2.3</td>
<td>EEG=left slowing; SPECT=L frontotemporal hypoperfusion</td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>No/No</td>
<td>FTLD-T</td>
<td>Logopenic</td>
<td>No</td>
<td>ND</td>
<td>0</td>
<td>56 / M</td>
<td>70</td>
<td>3.4</td>
<td>MRI=L, perisylvian atrophy</td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>No/No</td>
<td>FTLD-T</td>
<td>Agrammatic</td>
<td>?</td>
<td>No</td>
<td>0</td>
<td>56 / M</td>
<td>66</td>
<td>3.3</td>
<td>MRI=L, perisylvian and inferotemporal atrophy; SPECT=patchy hypoperfusion mostly L</td>
<td></td>
</tr>
<tr>
<td>20</td>
<td>No/No</td>
<td>FTLD-T</td>
<td>Agrammatic</td>
<td>No</td>
<td>No</td>
<td>1</td>
<td>59 / F</td>
<td>65</td>
<td>3.3</td>
<td>MRI=L, perisylvian and temporal/parietal atrophy</td>
<td></td>
</tr>
<tr>
<td>21</td>
<td>No/No</td>
<td>FTLD-T</td>
<td>Mixed</td>
<td>sm, nlg (F) L&gt;R</td>
<td>No</td>
<td>0</td>
<td>67 / M</td>
<td>78</td>
<td>3.3</td>
<td>MRI=unremarkable; EEG=L frontotemporal slowing</td>
<td></td>
</tr>
<tr>
<td>22</td>
<td>No/No</td>
<td>FTLD-T</td>
<td>Agrammatic</td>
<td>No</td>
<td>ND</td>
<td>0</td>
<td>71 / M</td>
<td>77</td>
<td>2.3</td>
<td>MRI=L, temporal atrophy</td>
<td></td>
</tr>
<tr>
<td>23</td>
<td>No/No</td>
<td>FTLD-T</td>
<td>Agrammatic</td>
<td>?</td>
<td>ND</td>
<td>0</td>
<td>57 / F</td>
<td>67</td>
<td>2.4</td>
<td>SPECT=L hypometabolism</td>
<td></td>
</tr>
</tbody>
</table>

5/6 Agrammatic PPA= FTLD-T
7/11 Logopenic PPA= AD; 4 FTLD
M. K.- PPA. Female, onset at age 44. Died 16 years after disease onset.
Risk Factors

? Male gender (Mesulam & Weintraub, 1992)


Genetic:
• ApoE ξ4 allele is NOT a risk factor for PPA, even in those with AD pathology at death (Rogalski et al, 2011)
• Heterozygosity of Met/Val at codon 129 of prion protein (Li et al, 2005)
• Over-representation of H1H1 haplotype (also in PSP, CBD) (Geschwind et al, 2004)
• Two PPA families with PGRN mutation (Mesulam et al, 2006)
Primary Progressive Aphasia (PPA)

Later Symptoms

1. Mutism
2. Severe speech comprehension deficits
3. Personality changes
4. Memory loss
5. Daily living activities severely limited
1. Determine the feasibility of Internet-based video speech-language therapy in PPA

2. Identify the most effective speech-language therapy strategies

3. Evaluate longitudinal impact on functional communication, quality of life and interpersonal communication
Improving access to speech-language therapy

Geographic

Therapy approach

Financial

E. Rogalski, PI