

# Research on Primary Progressive Aphasia: What It Is Teaching Us About Brain Function and Progression

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Northwestern University Feinberg School of Medicine



**Cognitive Neurology and  
Alzheimer's Disease Center**



**Northwestern  
Medicine**

We study diseases to understand more about them in order to cure, treat or prevent them.

*The study of brain diseases also teaches us how the brain functions normally, if there are risk factors for disease, and the relationships between specific symptoms and the diseases that cause them.*

# Primary Progressive Aphasia: “A Language Dementia\*”

According to currently accepted criteria, the PPA diagnosis is made in a person with a dementia, in which language impairment (*aphasia*), caused by a neurodegenerative disease (*progressive*), is the earliest and most limiting symptom (*primary*).

\* There are also memory, visuospatial and behavioral dementias.

# What is Language?

**Brain processes that create words that express thoughts**

**Words: names of objects, colors, people,  
actions, adverbs, adjectives, connector words**

**Stringing words together expresses thoughts for  
the purpose of communication**

**Language: Speaking; understanding what others are  
saying; understanding words you read ; writing**

# What is Aphasia?

**Loss of the ability to use language (words) for communication in one or more forms:  
spoken, written, sign language**

**Caused by damage to the brain,  
in areas that control language function**

**Interventions help compensate  
for reduced ability to communicate with words**

**INTERVENTIONS AIMED AT MEMORY LOSS  
DEMENTIA DO NOT WORK FOR APHASIA!**



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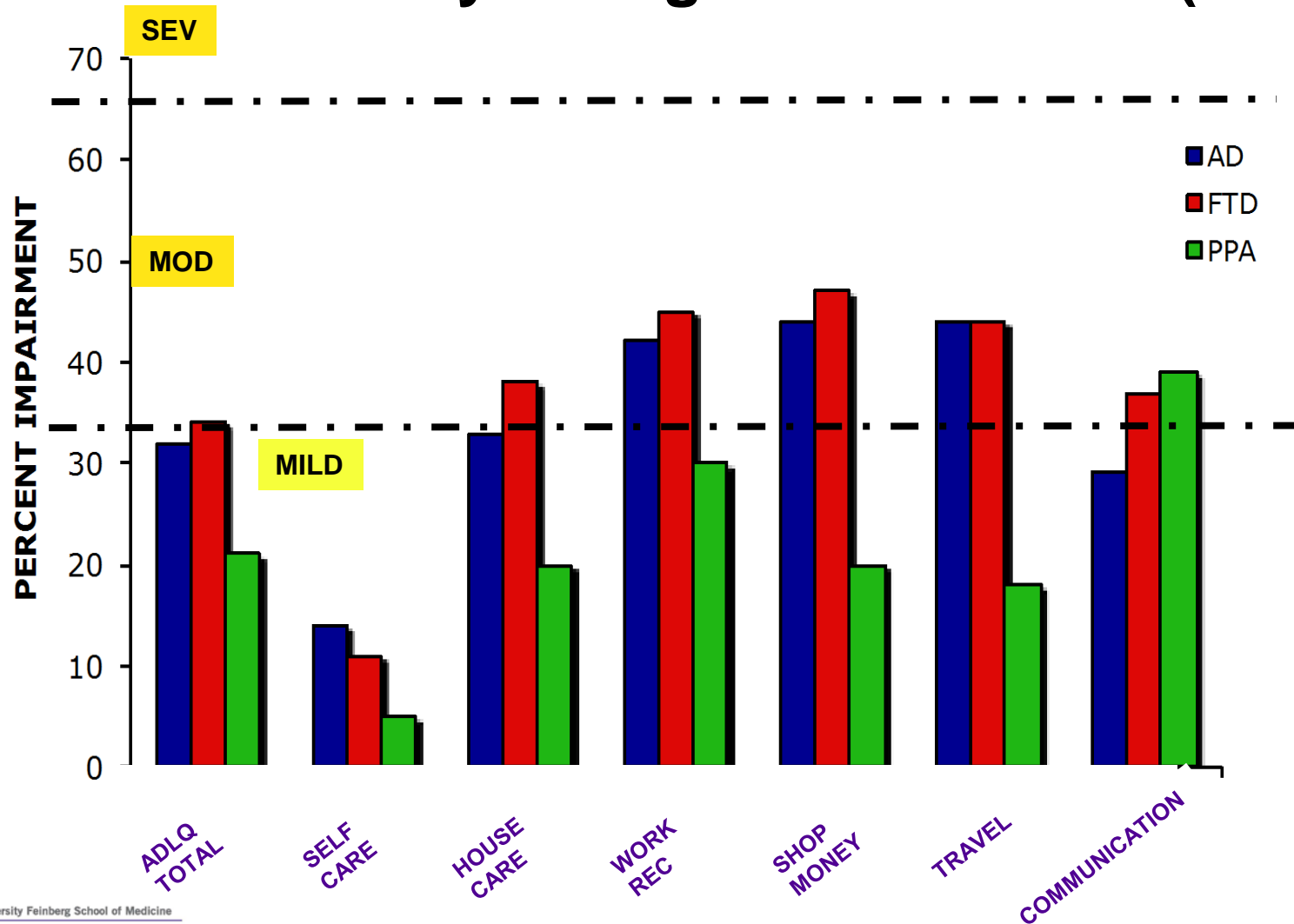
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# In early stages, PPA causes less functional Impairment than other forms of dementia

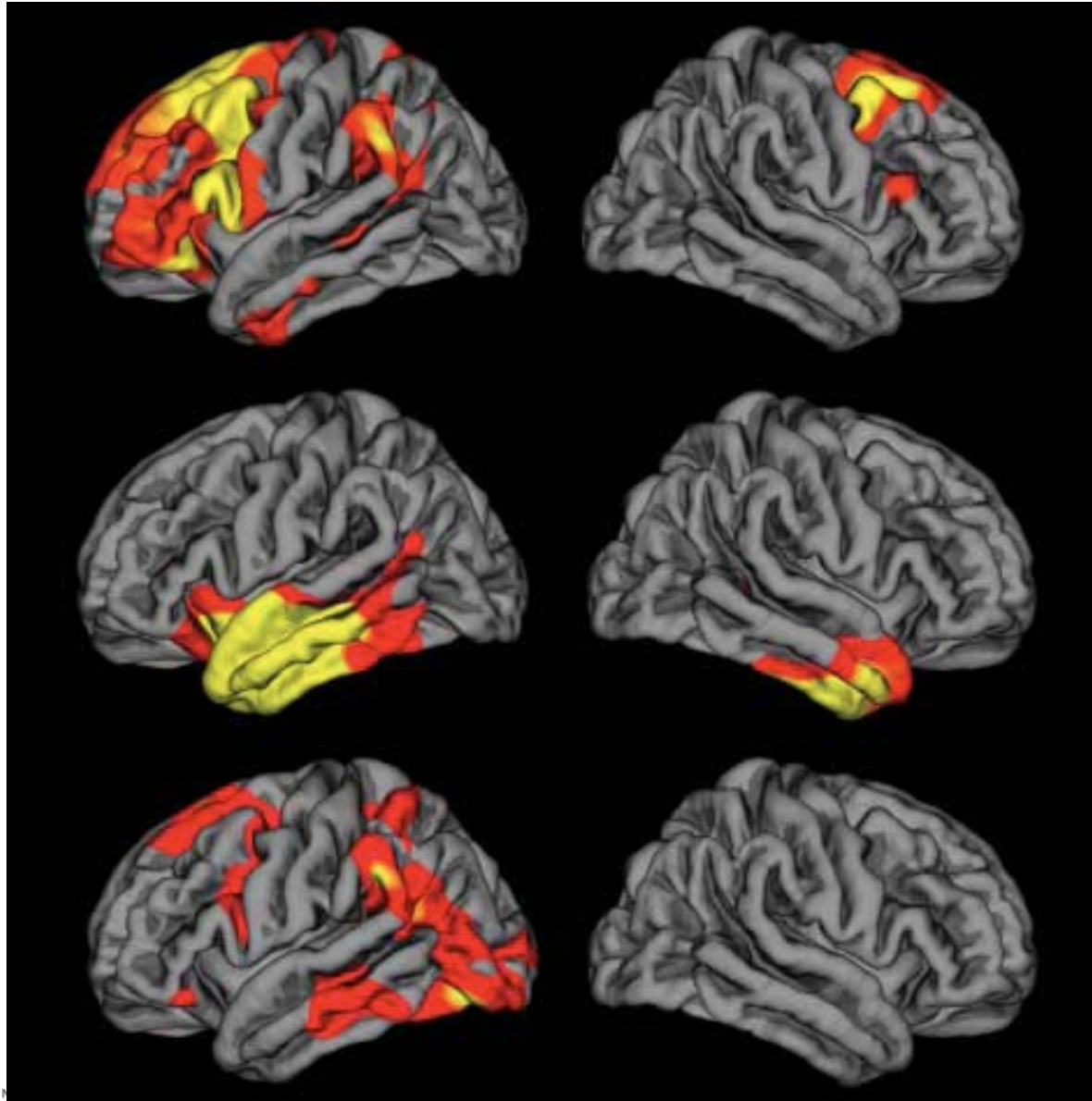
## Activities of Daily Living Questionnaire (ADLQ)



**LEFT**

**RIGHT**

**The disease  
affects left  
brain more than  
right**



**PPA-Agrammatic**

**PPA-Semantic**

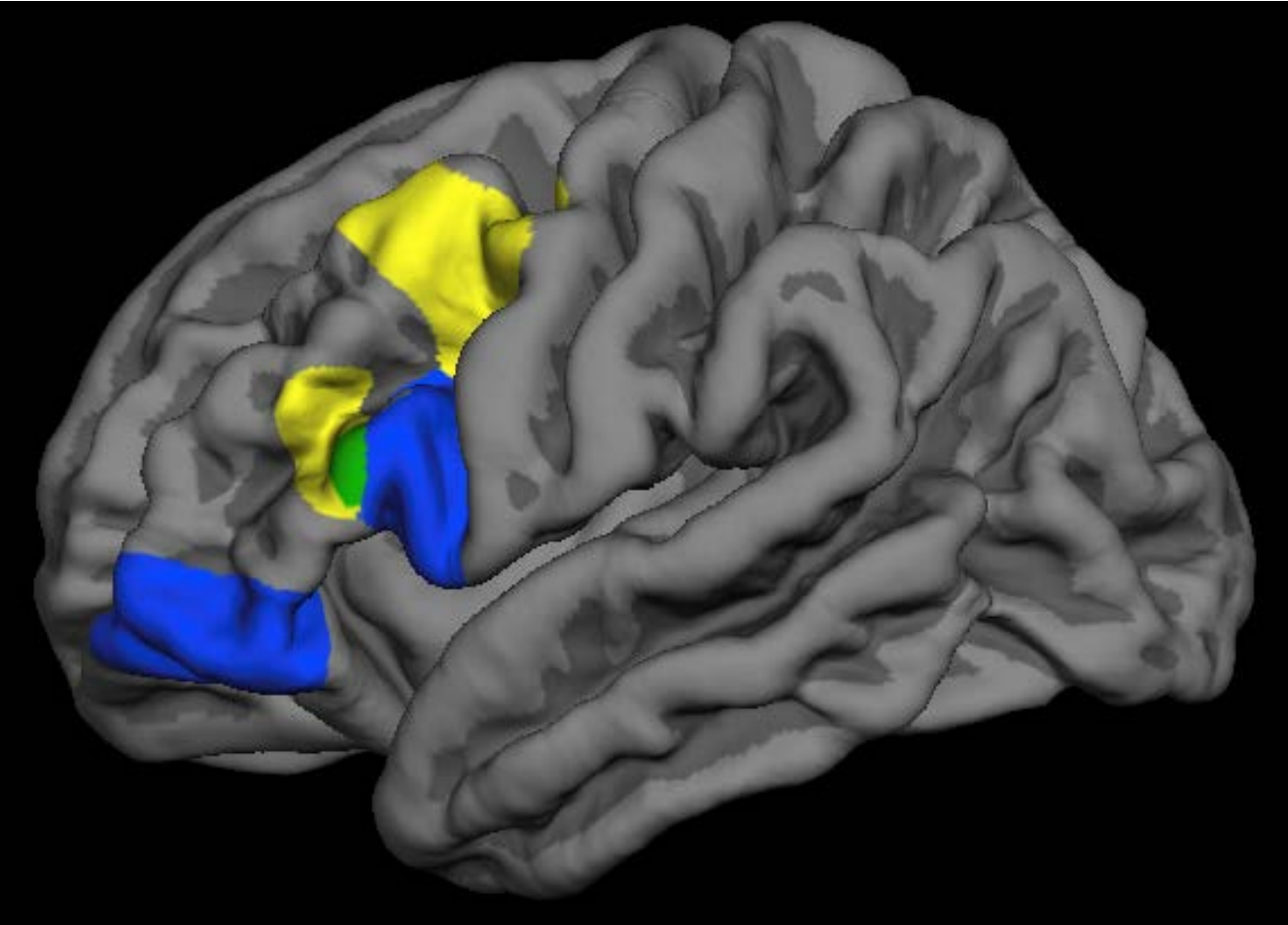
**PPA-Logopenic**



# DISSOCIATION OF FLUENCY FROM GRAMMAR IN THE FRONTAL LOBE

Northwestern Anagram Test ■

Mean Length of Utterance ■



Rogalski et al, J. Neurosci., 2011



# Brain Atrophy is Asymmetrical in PPA Regardless of Type of Neuropathology More Left Brain Atrophy

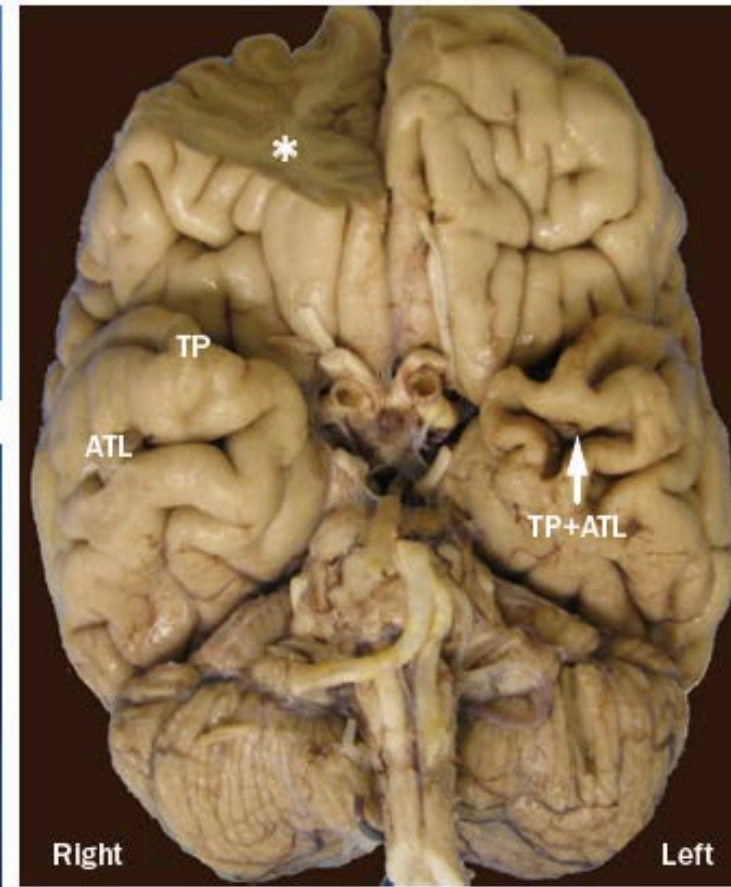
a PPA with Alzheimer disease pathology



b PPA with FTLD-tau pathology



c PPA with FTLD-TDP type B pathology



**We do not have blood or  
cerebrospinal fluid tests for FTLD**

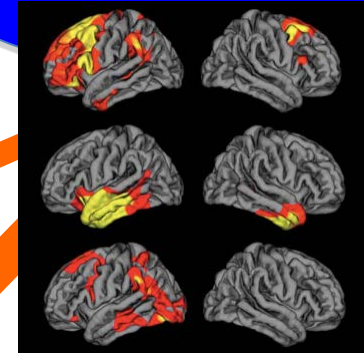
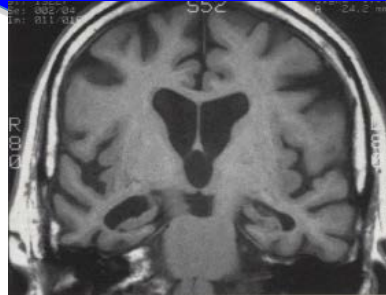
**Diagnosing PPA can help us predict  
what type of FTLD is in the brain or  
if it is more likely to be AD**

CLINICAL  
DEMENTIA  
SYNDROME

MEMORY LOSS  
DEMENTIA  
"Dementia of the  
AD type"

LANGUAGE LOSS  
DEMENTIA  
Primary Progressive  
Aphasia

BRAIN REGIONS  
AFFECTED

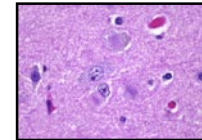
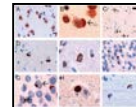
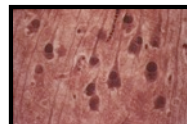
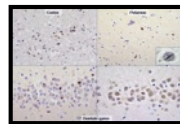
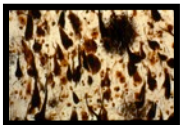


DISEASE

AD  
Neuropathology

FTLD  
Neuropathology

OTHER  
CJD, Cortical Lewy  
Body



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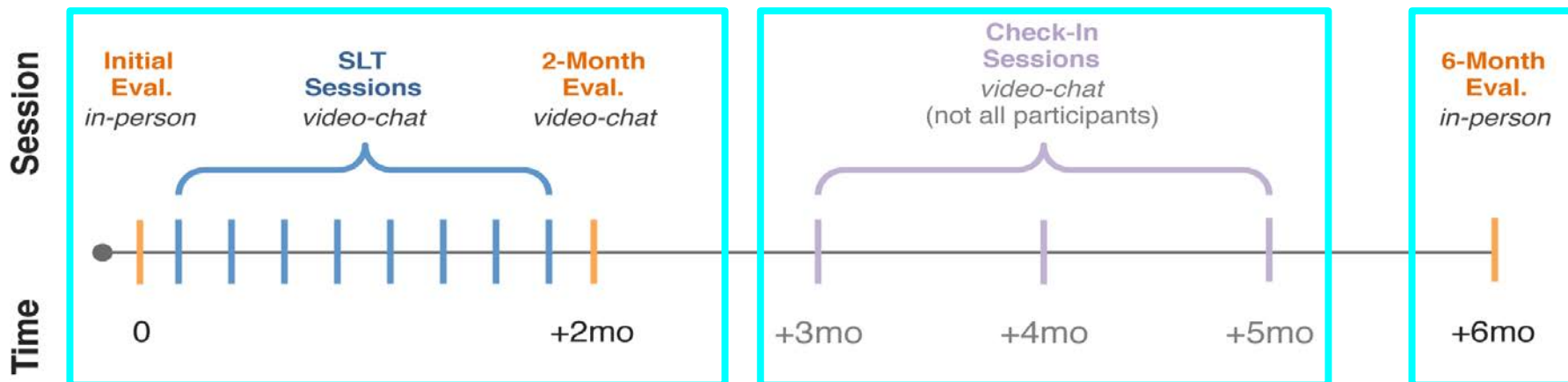
# Relationship of Neuropathology to PPA Subtype

<p>GOOD</p> <p>↑</p> <p>COMPREHENSION</p> <p>↓</p> <p>BAD</p>	<p><b>AGRAMMATIC</b></p> <p><i>n</i> = 17</p> <p>65% FTLD-TAU 18% FTLD-TDP 12% AD 6% DLBD &amp; AD</p>	<p><b>LOGOPENIC</b></p> <p><i>n</i> = 32</p> <p>56% AD 25% FTLD-TDP 16% FTLD-TAU 3% FTLD-TDP &amp; AD</p>
	<p><b>MIXED</b></p> <p><i>n</i> = 6</p> <p>67% AD 17% FTLD-TAU 17% FTLD-TDP</p>	<p><b>SEMANTIC</b></p> <p><i>n</i> = 3</p> <p>67% FTLD-TDP 33% AD</p>
	<p>BAD ←</p> <p><b>GRAMMATICALITY</b></p> <p>→ GOOD</p>	

# communication BRIDGE

E. Rogalski, Northwestern, PI

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





# communication BRIDGE

Improving access to speech-language therapy



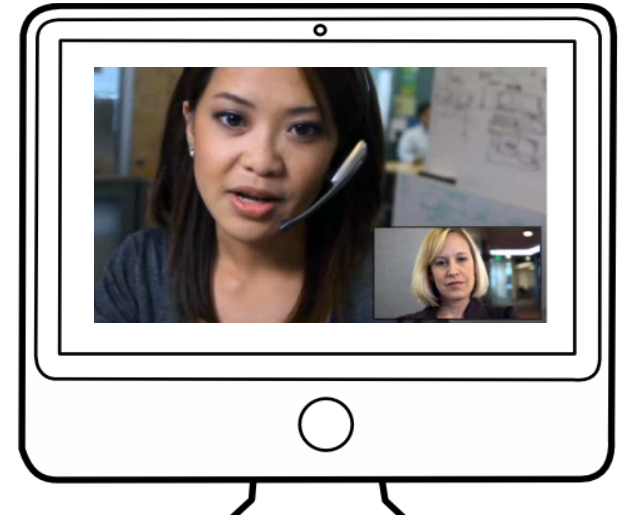
Geographic



Therapy approach



Financial



**E. Rogalski, Northwestern, PI**

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# CONCLUSION

- We understand that selective brain regions are vulnerable to neurodegenerative disease.
- We can predict the type of disease causing the PPA dementia with some accuracy.
- We can tailor our interventions to support communication and change strategies as symptoms progress.
- We can educate patients and caregivers about the diagnosis and coping strategies

