PRIMARY PROGRESSIVE APHASIA & APRAXIA OF SPEECH

AN INTRODUCTION TO ASSESSMENT AND MANAGEMENT

Michael de Riesthal PhD CCC-SLP
Kiiya Shibata MS CCC-SLP

DISCLOSURES

Michael de Riesthal PhD CCC-SLP receives a salary from Vanderbilt University Medical Center

Kiiya Shibata receives a stipend from Vanderbilt University

PRIMARY PROGRESSIVE APHASIA

- Aphasia of insidious onset
- Limitations in activities of daily living are attributed to language impairment at onset and throughout the initial stages of disease
- Intact premorbid language function
- Absence of prominent initial behavioral, memory, and visuospatial deficits
- Typical onset is between 55-65 years of age

(Gorno-Tempini et al., 2011)

PRESENTATION TO SPEECH

- Time course of initial presentation is highly variable
  - Some self-refer, with or without an official diagnosis
  - Others present at the recommendation of their neurologist
- During the initial interview, patients may say something like:
  - "I'm forgetting my words."
  - "I know what I want to say, but I can't say it."
  - "I can't keep up in conversations anymore."
  - "I need people to repeat things at work a lot."

(Gorno-Tempini et al., 2011)

PPA SUBTYPES

- Semantic Variant
- Logopenic Variant
- Non-fluent/ Agrammatic Variant

(Gorno-Tempini et al., 2011)

SEMANTIC VARIANT

CLINICAL CRITERIA

Core features (both must be present)

- Impaired confrontation naming
- Impaired single word comprehension

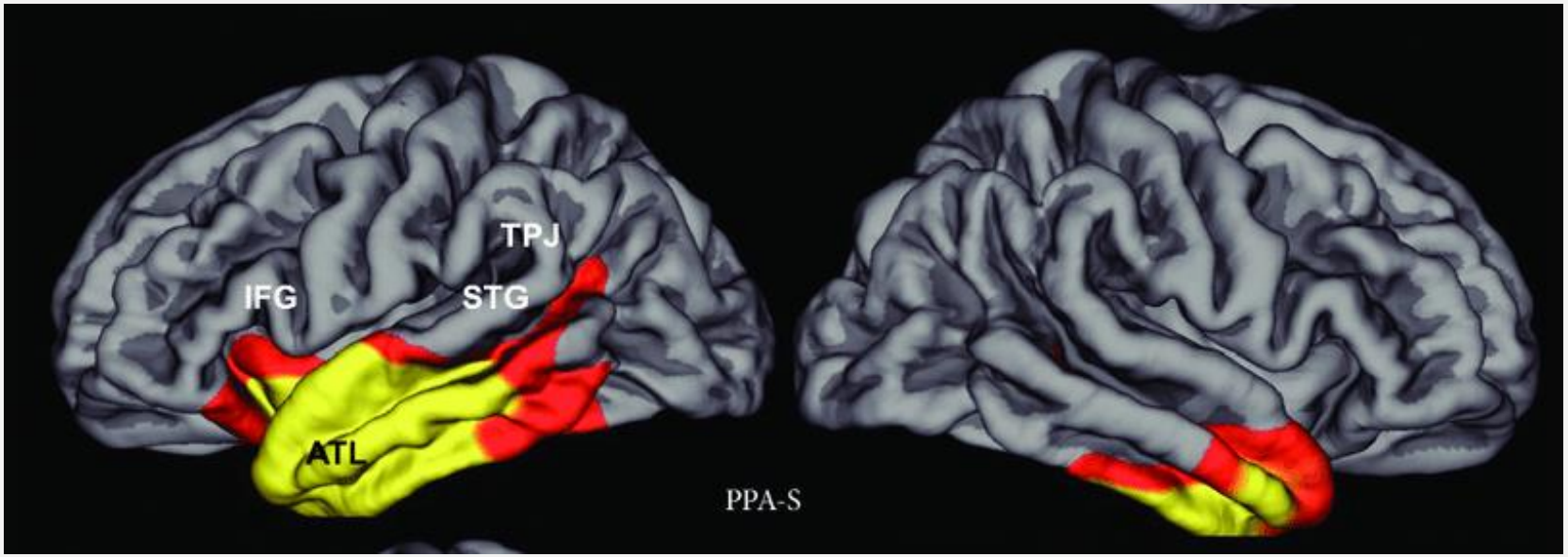
Additional features (at least three must be present)

- Impaired object knowledge
- Surface dyslexia or dysgraphia
- Spared motor-speech
- Spared repetition
- Spared speech production (grammar and motor-speech)

(Gorno-Tempini et al., 2011)

SEMANTIC IMAGING PATTERN

ANTERIOR TEMPORAL LOBE



(Gorno-Tempini et al., 2011; Mesulam, 2016)

LOGOPENIC VARIANT

CLINICAL CRITERIA

Core features (both must be present)

- Impaired single word retrieval in spontaneous speech and naming
- Impaired repetition of sentences and phrases

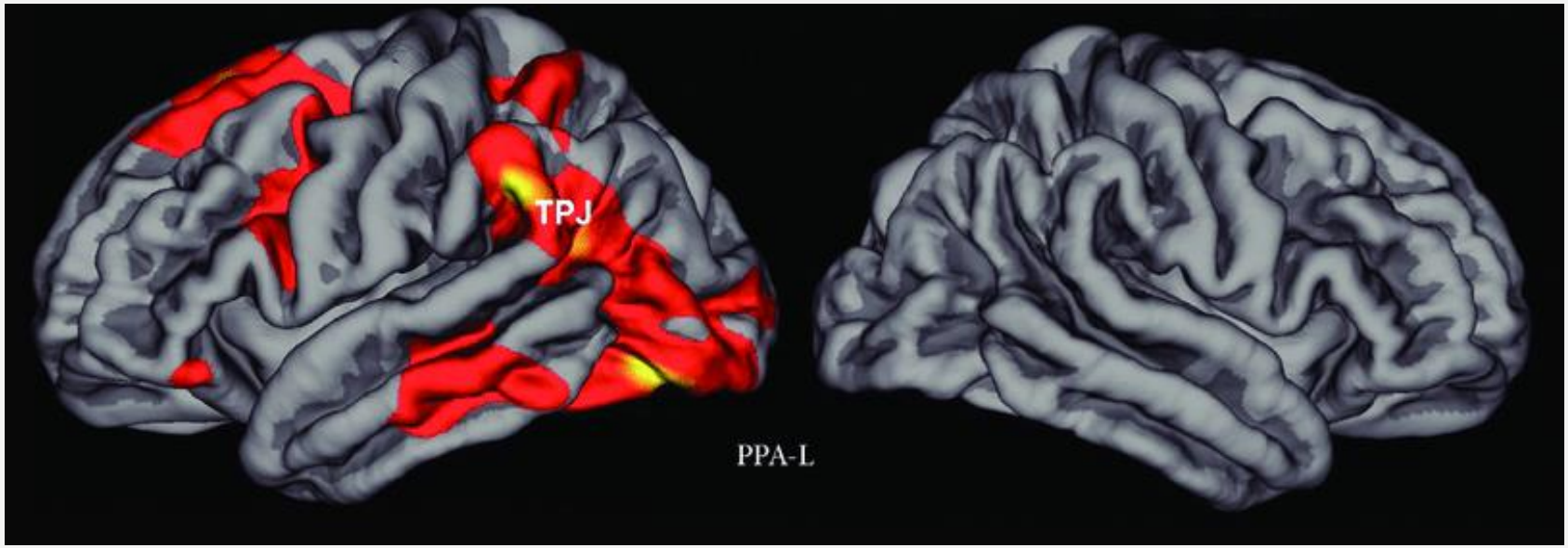
Additional features (at least two must be present)

- Phonologic errors in spontaneous speech and naming
- Spared single-word comprehension and object knowledge
- Spared motor-speech
- Absence of frank agrammatism

(Gorno-Tempini et al., 2011)

LOGOPENIC IMAGING PATTERN

LEFT POSTERIOR PERISYLVIAN OR PARIETAL



(Gorno-Tempini et al., 2011; Mesulam, 2016)

NON-FLUENT/ AGRAMMATIC VARIANT

CLINICAL CRITERIA

Core features (at least one must be present)

- Agrammatism in language production
- Apraxia of speech

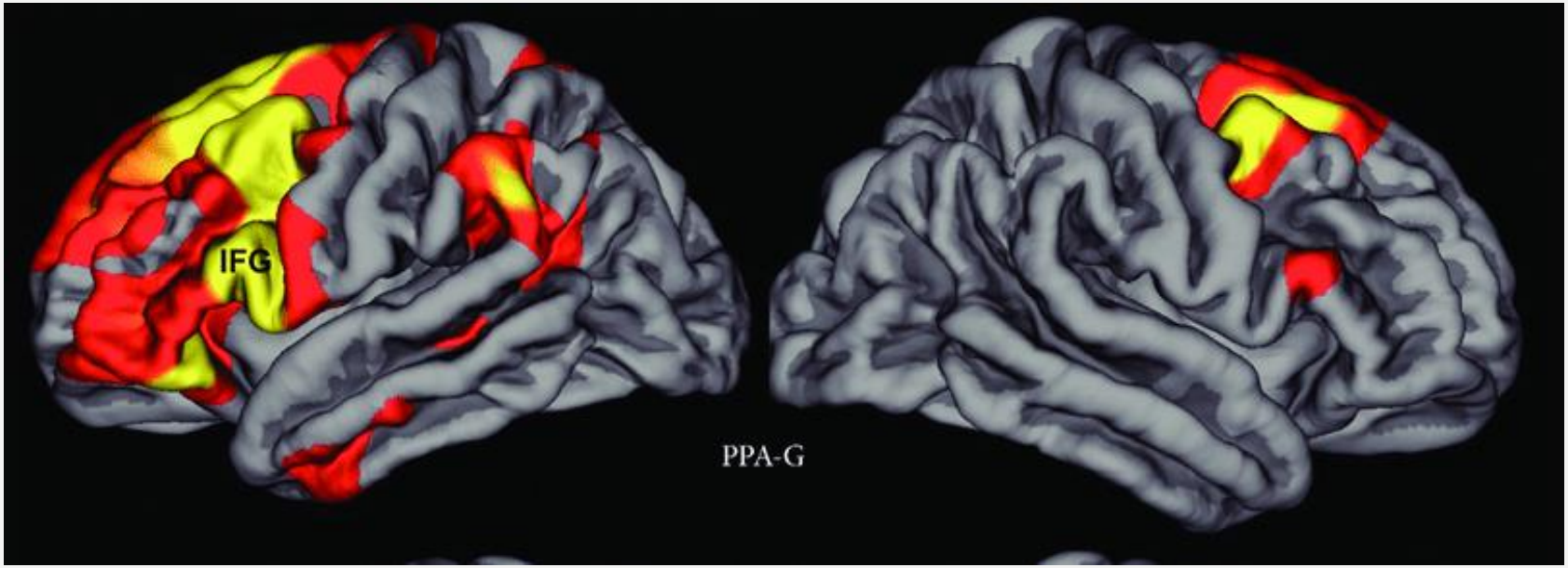
Additional features (at least two must be present)

- Impaired comprehension of syntactically complex sentences
- Spared single-word comprehension
- Spared object knowledge

(Gorno-Tempini et al., 2011)

NON-FLUENT/ AGRAMMATIC IMAGING PATTERN

LEFT POSTERIOR FRONTO-INSULAR



(Gorno-Tempini et al., 2011; Mesulam, 2016)

PRIMARY PROGRESSIVE AOS (PPAOS)

- Apraxia of speech of insidious onset, initially identified by Duffy (2006)
- Gradual progression
- Absence of non-language cognitive impairments and aphasia for substantial period of time
- Resulting from a degenerative condition involving the left hemisphere

(Duffy, 2006)

AOS SUBTYPES

AOS Type 1 describes a speech profile dominated by distorted sound substitutions or additions

AOS Type 2 describes a speech profile dominated by syllable segmentation or lengthened intersegment durations (sometimes referred to as scanning speech)

(Josephs et al., 2013)

AOS SUBTYPES

AOS Type 1 → AOS Type 2

Predominant agrammatism (nfvPPA) → Predominant AOS (nfvPPA) → AOS without language deficits (PPAOS)

(Josephs et al., 2013)

ASSESSMENT

REASON FOR EVALUATION

Diagnosis of PPA	Confirm language is primary impact on ADLs/IADLs
	Confirm variant
	Establish variant
No clear diagnosis	Symptom onset and timeline
	Impact on ADLs/IADLs
	Gather information for neurologist

SPECIAL CONSIDERATIONS

- Rare, unfamiliar diagnosis
- Young age of onset
- Slow progression that patients are acutely aware of
- Involve loved ones in all sessions
- Meet the patient and family where they are
- Setting expectations
- Medical identification card

TEST CORE FEATURES

svPPA	lvPPA	nfvPPA
• Impaired confrontation naming **AND** • Impaired single word comprehension	• Impaired single-word retrieval in spontaneous speech and naming **AND** • Impaired repetition of sentences and phrases	• Agrammatism **OR** • Apraxia of speech

TEST REMAINING ADDITIONAL FEATURES

svPPA	lvPPA	nfvPPA
• **Impaired object knowledge** • **Surface dyslexia or dysgraphia** • Spared repetition • Spared grammar and motor-speech	• Phonological paraphasias in spontaneous speech and naming • Spared motor speech • Spared object knowledge • No agrammatism	• **Impaired comprehension of syntactically complex sentences** • Spared object knowledge • Spared single word comprehension

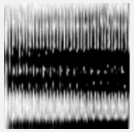
ASSESSMENT CHECKLIST

Discourse sample (word retrieval & agrammatism)	lvPPA/nfvPPA
Single word comprehension	svPPA
Confrontation naming	svPPA/lvPPA
Sentence repetition	lvPPA
Comprehension of syntactically complex sentences	nfvPPA
Motor speech	nfvPPA
Object knowledge	svPPA
Surface dyslexia or dysgraphia	svPPA

NONFLUENT VS. LOGOPENIC

- Particularly if agrammatism is not evident, the presence or absence of apraxia of speech (AOS) is critical to the differential diagnosis
- Regardless, AOS +/- will inform your treatment decisions, and must be assessed
- AOS vs phonemic paraphasias can be difficult to differentiate
  - How do yall assess for AOS? What tasks help you the most?

ACOUSTIC MEASURES



Relative vowel duration in repetition of multisyllabic words differentiates between lvPPA and nfvPPA (Ballard et al., 2014)

Frequency of distorted sound errors differentiated between lvPPA and nfvPPA in word repetition and connected speech tasks (Haley et al., 2021)

APRAXIA OF SPEECH RATING SCALE (ASRS)

- Strand et al. (2014)
- Perceptual tool for description and diagnosis of AOS
- Based on and developed for individuals with neurodegenerative disease
- Scores features on a 0-4 scale
  - 0= not present
  - 1= detectible but infrequent
  - 2= frequent but not pervasive
  - 3= nearly always evident but not marked in severity
  - 4= nearly always evident and marked in severity

(Strand et al., 2014)

1	**AOS - primary distinguishing features[a] (no overlap with dysarthria or aphasia). One or more must be present for diagnosis of AOS.**	**Score (0-4)**
1.1	Distorted sound substitutions	
1.2	Distorted sound additions (not including intrusive schwa)	
1.3	Increased sound distortions or distorted sound substitutions with increased utterance length or increased syllable/word articulatory complexity	
1.4	Increased sound distortions or distorted sound substitutions with increased speech rate	
1.5	Inaccurate (off-target in place or manner) speech AMR's (alternating motion rates, as in rapid repetition of "puh puh puh")	
1.6	Reduced words per breath group relative to maximum vowel duration	
2	**Distinguishing features unless dysarthria present[b]**	**Score (0-4)**
2.1	Syllable segmentation within words > 1 syllable	
2.2	Syllable segmentation across words in phrases/sentences	
2.3	Sound distortions	
2.4	Slow overall speech rate	
2.5	Lengthened vowel &/or consonant segments	
2.6	Lengthened intersegment durations (between sounds, syllables, words, or phrases; possibly filled, including intrusive schwa)	
3	**Distinguishing features unless aphasia present[c]**	**Score (0-4)**
3.1	Deliberate, slowly sequenced, segmented, &/or distorted (including distorted substitutions) speech SMRs in comparison to speech AMRs	
3.2	Audible or visible articulatory groping; speech initiation difficulty; false starts/restarts	
4	**Distinguishing features unless dysarthria &/or aphasia present[d]**	**Score (0-4)**
4.1	Sound or syllable repetitions	
4.2	Sound prolongations (beyond lengthened segments)	

SCORE OF 8 OR HIGHER INDICATIVE OF AOS

(Strand et al., 2014)

DISTINGUISHING FEATURES

AOS - primary distinguishing features[a] (no overlap with dysarthria or aphasia). One or more must be present for diagnosis of AOS.
Distorted sound substitutions
Distorted sound additions (not including intrusive schwa)
Increased sound distortions or distorted sound substitutions with increased utterance length or increased syllable/word articulatory complexity
Increased sound distortions or distorted sound substitutions with increased speech rate
Inaccurate (off-target in place or manner) speech AMR's (alternating motion rates, as in rapid repetition of "puh puh puh")
Reduced words per breath group relative to maximum vowel duration

(Strand et al., 2014)

TREATMENT & MANAGEMENT

BASIC PRINCIPLES

- Early intervention when possible
- Formal therapy and home practice facilitate maintenance, especially for trained items
- Errorless learning modes are generally favored
- Generalization is more likely with lvPPA and nfvPPA
- Spared semantic knowledge helps facilitate gains (critical to svPPA)
- Learning in typically highly context-bound for svPPA
- Gains are generally congruent to severity

(Cadório et al., 2017; Henry et al., 2018; Jokel, 2019)

BASIC COMPONENTS

- Heavy education and counseling
  - Voice or video recordings
  - Written materials
  - Teach/demonstrate back
- Speech binder
  - Introduce a communication book early, for later use as an AAC device as needed
  - Include scripts, target word stimuli, educational materials, and pictures of patient's everyday objects, activities, and people
- Identifying most important targets
  - Family, fun, function
  - Setting up compensatory strategies (AAC and self-cues)

LEXICAL RETRIEVAL TREATMENT (LRT)

- Originally developed for stroke aphasia
- Capitalizes on residual cognitive-linguistic systems
- Employs semantic, phonemic, and orthographic techniques
- Caregivers can be trained to administer LRT
- Comparable outcomes when administered via telehealth vs in-person

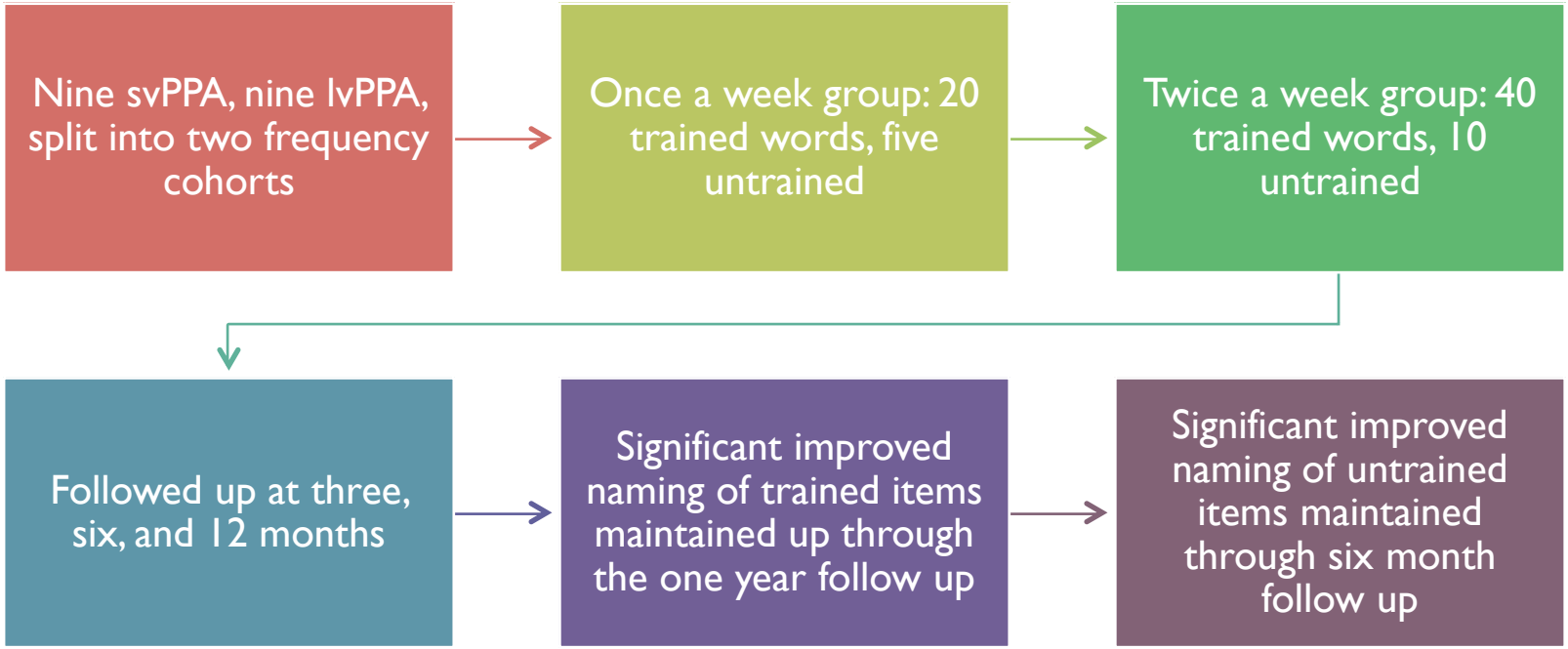
(Dial et al., 2019; Grasso et al., 2017; Henry et al., 2013; Henry et al., 2019)

LEXICAL RETRIEVAL TREATMENT

(Henry et al., 2013; Henry et al., 2019)

Cueing type	Prompt
Semantic self-cue	Prompt description: "Tell me about it."
Orthographic self-cue	Request written production: "Can you write the word?" Encourage any attempt
Phonemic self-cue	Point to the first letter (written either by the patient or clinician) "Think of the sound this letter makes. Now try to say the word"
Oral reading	If the patient does not produce the word (spoken or written), provide the written word and say, "What does this say?" Have the patient copy the written word three times
Repetition	Request repetition of the target word, after spoken model as needed
Yes/no questions	Ask five yes/no questions regarding the semantic features of the target word
Recall	Ask the patient to recall two semantic features and the spoken or written name of the target word

HENRY ET AL. (2019)



SCRIPT TRAINING

- Develop scripts
  - Tailor length and complexity to your patient
  - Encourage topics that come up most frequently in a patient's life
- Develop home practice videos for speech entrainment (i.e. video implemented script training for aphasia [VISTA])
  - Record a health speaker's mouth saying each script
  - Record directly on patient's phone (if they have one), or upload a video to a private youtube channel (if they have access)
- Practice in session via the hierarchy described by Henry et al. (2018)
- Note difficult utterances for targeted practice at home and in session
- Comparable outcomes when administered via telehealth vs in-person

(Dial et al. 2019; Henry et al., 2018)

Task Hierarchy	Implementation options
Recognize from foils	Ask patient to identify their written script from a field of related scripts.
Order sentences	Next, patients order the sentences of their script. If they make an error, ask them to check their work and then provide the correct order as needed.
Read script aloud	As they read the script, note words and phrases that are challenging, for home practice in isolation.
Produce script in response to questions	Ask patients questions that they can answer with their full script. You can also have them do this with family members, and unfamiliar partners.
Produce script from memory	Request repetition of the target word, after spoken model as needed
Answer questions with scripted sentences outside of the context of the script	Ask the patient questions that they can answer with one sentence from a trained script.

SCRIPT TRAINING

(Henry et al., 2018)

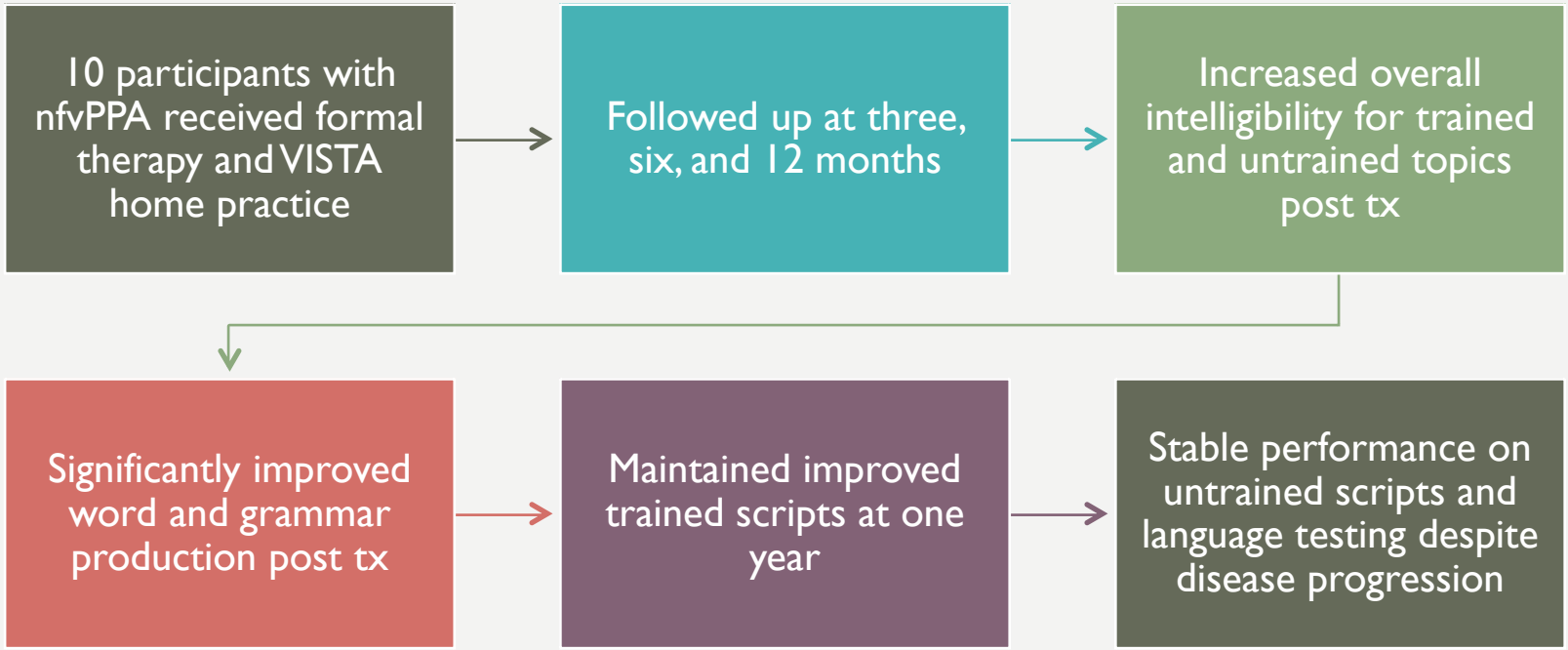
ST EXAMPLE

Examples from Henry et al. (2018)

"Football is a great sport. My favorite NFL team is the Green Bay Packers. My college team is the UW Badgers. I love to watch football all the time."

"Fly-fishing is a passion of mine for numerous reasons, but mostly for the wonderful places it takes me to. The waters and the ecosystems are inevitably beautiful and interesting. I also enjoy the fact that fly-fishing is so demanding, challenging and totally absorbing. It serves a therapeutic role that releases me from the stresses of everyday life. I often find myself planning a strip to one of the places I love to fish most, including Connecticut, Montana, Alaska, Canada, or Texas."

HENRY ET AL. (2018)



AAC

Readiness varies patient to patient

Ask family to provide pictures of the patient's everyday objects, activities, and people, if possible

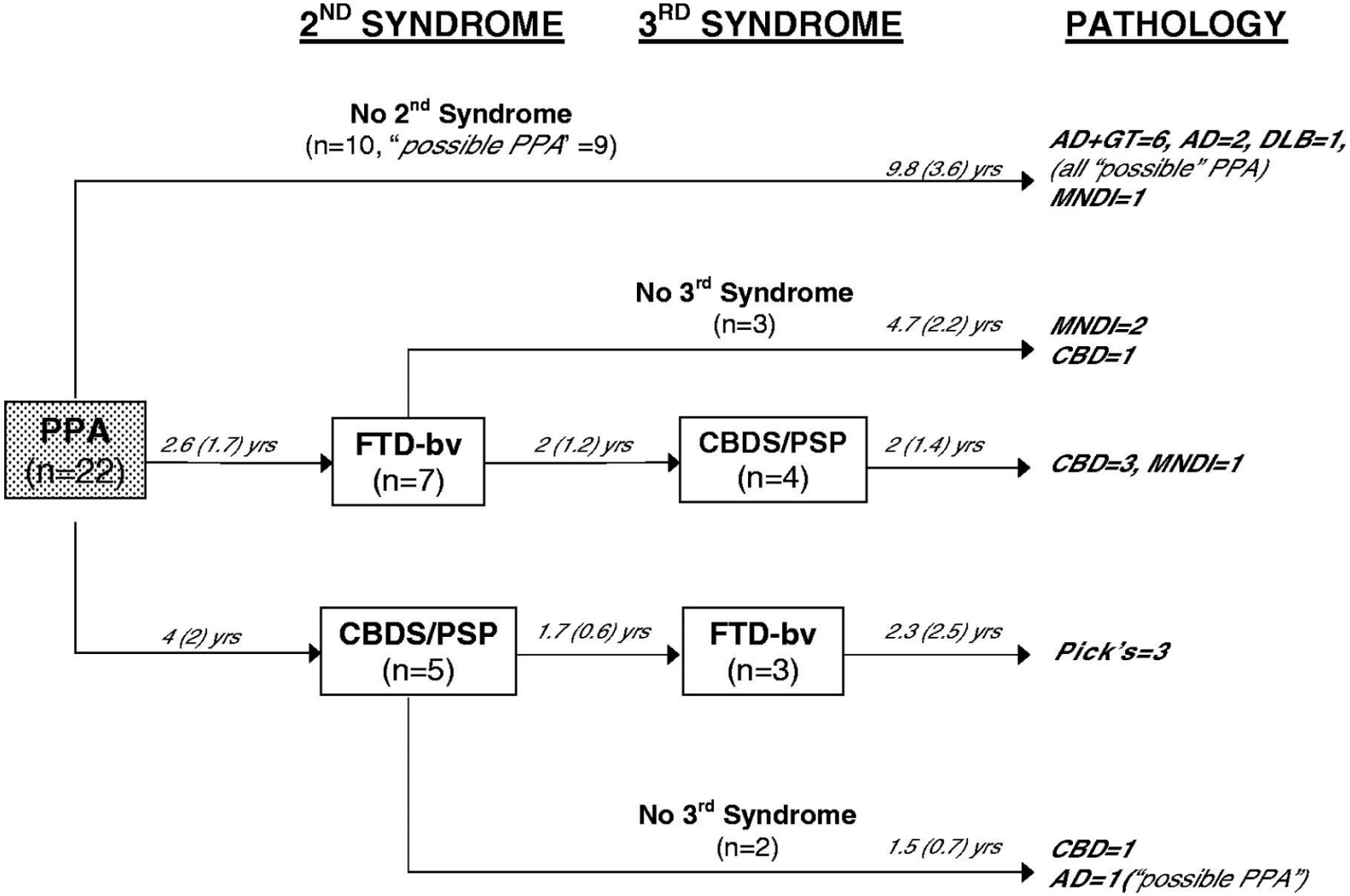
Use these as LRT targets, and compile them in a binder for the patient for later use as an AAC device if needed

Include scripts with visual aids/pictures in the binder, for later use as a communication book as needed

If completing over telehealth, you can use share drives and email to send materials to loved ones to print

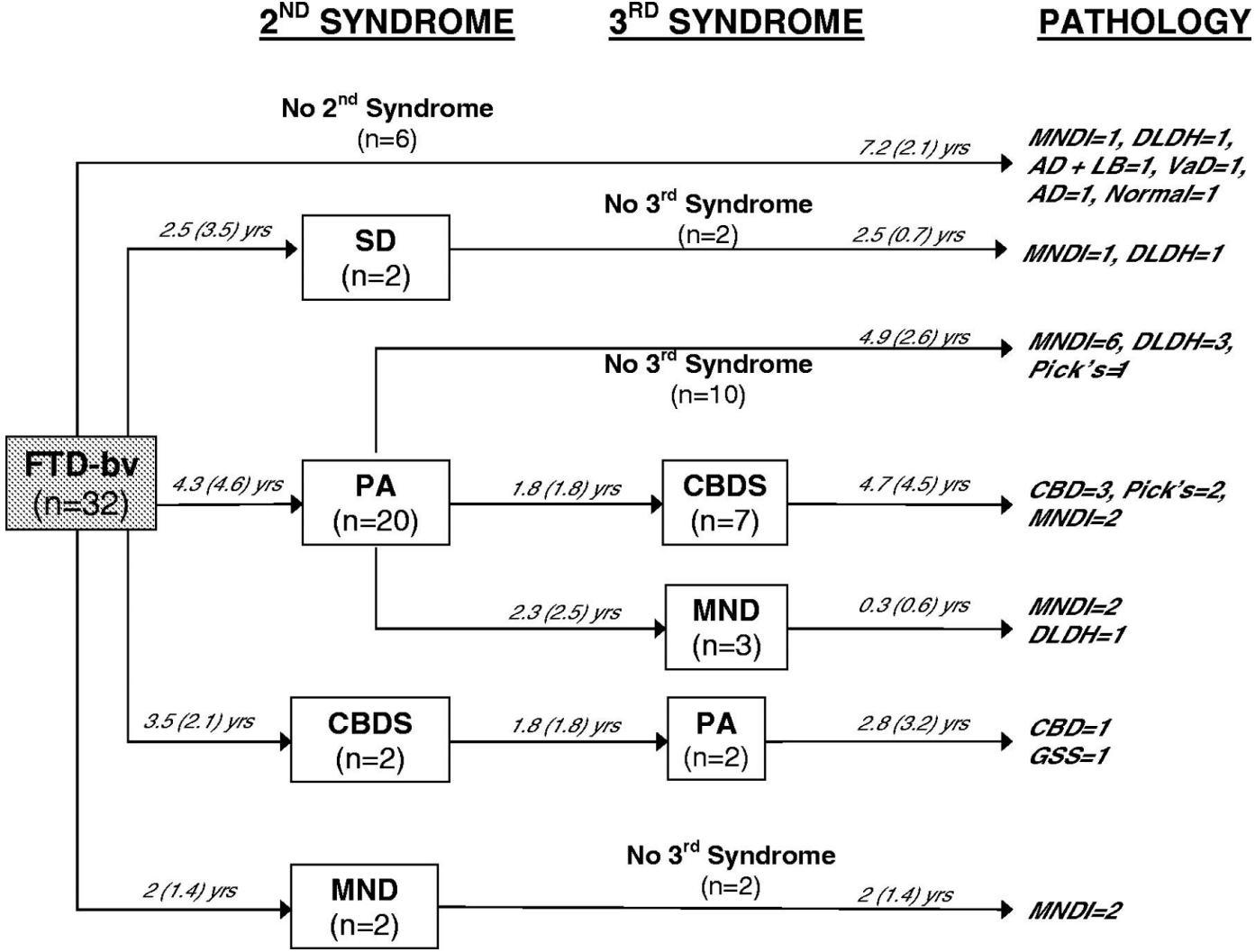
Teach loved ones to generate scripts and target words to add to patient's binder

PROGNOSIS & PROGRESSION



EVOLUTION OF CLINICAL SYNDROMES BEGINNING WITH PPA

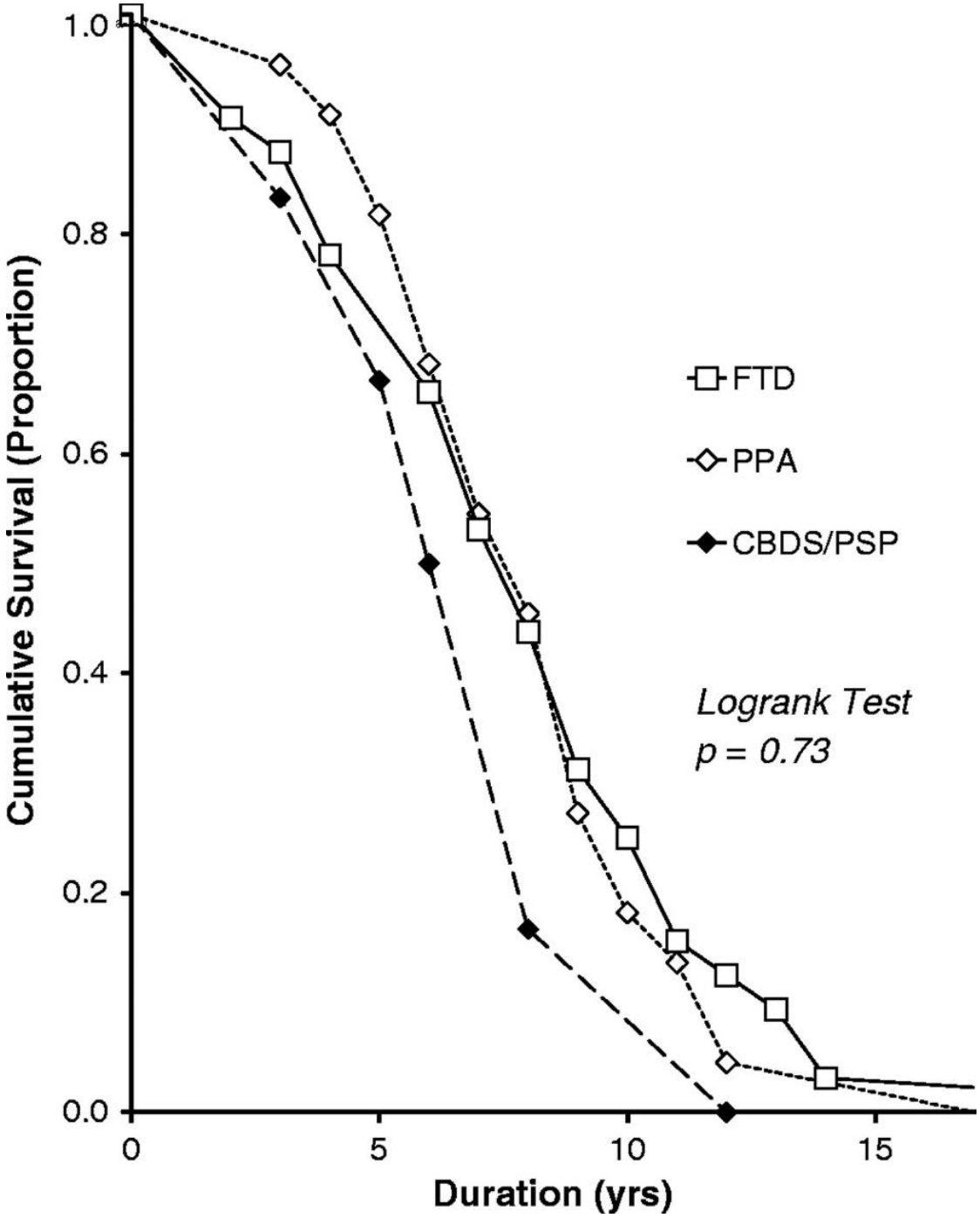
(Kertesz et al., 2005)



EVOLUTION OF CLINICAL SYNDROMES BEGINNING WITH FTD-BV

(Kertesz et al., 2005)

SURVIVAL CURVES FROM SYMPTOM ONSET ACCORDING TO FIRST CLINICAL SYNDROME



(Kertesz et al., 2005)

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