I. Definition
   A. Form of cognitive impairment that involves degeneration of speech and language functions over time. Nerve cell death that occurs in speech and language neural area that is not caused by stroke, head trauma, infection or cancer (Cognitive Neurology and Alzheimer’s Disease Center, n.d.)
   B. Deterioration of language for at two years before decline in other cognitive functions (Mesulam, 1982)
   C. Inclusion Criteria (Gorno-Tempini et al., 2011)
      1. Most prominent clinical feature is difficulty with language
      2. Language difficulties are the principle cause of impaired daily living activities
      3. Aphasia is most prominent deficit at symptom onset and beginning of the disease
   D. Exclusion Criteria (Gorno-Tempini et al., 2011)
      1. Pattern of deficits cannot be accounted by other nondegenerative nervous or medical disorder
      2. Cognitive disturbance cannot be accounted by a psychiatric diagnosis
      3. No prominent initial episodic memory, visual memory, and visuoperceptual impairments
      4. No prominent, initial behavioral disturbance

II. PPA Variants (Gorno-Tempini et al., 2011)
   A. Patterns of language impairment and patterns of atrophy on imaging
   B. Prevalence of 2-15/100,000 individuals, survival of 7 years depending on when diagnosis is made and type of variant (Sapolsky et al., 2011)
   C. Average age onset of late 50s (Grossman, 2014)
   D. Reliably diagnosed in around 75-80% of individuals (Nickels & Croot, 2014)
   E. Etiology
      1. Alzheimer’s Disease: 30%-40% of individuals with PPA (Northwestern University)
      2. Frontotemporal Lobar Degeneration-tau (FTLD-tau): 60%-70% of individuals with PPA (Northwestern University)
      3. FTLD-TDP: 60%-70% of individuals with PPA (Northwestern University)
   F. Potential Factors
      1. Genetics: mutations in MAPT, GRN, or C9orf72 resulting in FTLD, ApoE, Prion Protein, Foxp2 (Rogalski, Weintraub, & Mesulam 2013)
      2. Focal structural lesions: late manifestation of developmental or genetic weakness of language network (Rogalski, Weintraub, & Mesulam 2013)
      3. Learning disability: developmental dyslexia (Rogalski, Weintraub, & Mesulam 2013)
   G. Subdivided into three groups
      1. Agrammatic/Nonfluent (nfv PPA) (Gorno-Tempini et al., 2011)
         Core features: One of these criteria must be present
- Agrammatic language (short, simple phrases, omits function words and inflections)
- Effortful and halting speech (apraxia of speech, slow labored speech production, distortions, deletions, substitutions, insertions, and disrupted prosody)

Secondary features: two of three criteria must be present
- Impaired comprehension of complex sentences
- Spared single word comprehension
- Spared object knowledge

Atrophy: Left posterior fronto-insular atrophy

Common Pathology: Frontolobar Temporal Deeneration (FLTD)-Tau, FTLD-TDP

Cognitive factors: (Bettcher & Sturm, 2014)
- Mild to moderate difficulty in executive function tasks
- Difficulty with tests of set-shifting
- Strengths: visuospatial processing and verbal episodic memory

Clinical Observations: (Bettcher & Sturm, 2014)
- Intact social skills
- Aware of performance on tasks and speech production
- Mild disinhibition and apathy as disease progresses

2. Logopenic (lv PPA) (Gorno-Tempini et al., 2011)

Core features: both criteria must be present
- Impaired word retrieval (phonological paraphasias, slow rate, frequent pauses)
- Impaired repetition of sentences and phrases

Secondary features: three of the four criteria must be present
- Speech errors in spontaneous and naming (phonologic paraphasias, no distortions)
- Spared single word comprehension and object knowledge
- Spared motor speech (good articulation and prosody)
- Absence of frank agrammatism (halting but due to word finding difficulties)

Atrophy: Left posterior perisylvian or parietal

Common Pathology: Alzheimer’s disease, FLTD-Tau, FTLD-TDP

Cognitive factors: (Bettcher & Sturm, 2014)
- Mild visuospatial deficits like localizing and constructing
- Difficulties in phonological working memory
- Impairments in numbers and complex calculations

Clinical Observations: (Bettcher & Sturm, 2014)
- Aware of difficulties
- May display apathy, anxiety, and mild irritability

3. Semantic (svPPA) (Gorno-Tempini et al., 2011)

Core features: both criteria must be present
• Impaired naming of objects (circumlocution, simplification, semantic paraphasia (Mesulam et al., 2014)
• Impaired single word comprehension (impairment in low frequency items, problems in object and person recognition)

Secondary features: three of the four criteria must be present
• Impaired object knowledge (especially for low frequency or low familiarity items)
• Surface dyslexia or dysgraphia (regularization errors)
• Spared repetition
• Spared speech production

Atrophy: Anterior temporal lobe
Common Pathology: FLTD-Tau, FTLD-TDP (Hillis, 2015)

Cognitive factors: (Bettcher & Sturm, 2014)
• Difficulties in verbal area in episodic memory

Clinical Observations: (Bettcher & Sturm, 2014)
• Changes in social and emotion

III. Assessment

A. Identify language impairment and the impact on communication activities/participation/quality of life to determine (Nickels, Taylor, & Croot, 2011)
B. Establish baseline for the nature, extent, and rate of change over time (Nickels, Taylor, & Croot, 2011)
C. Choose appropriately difficulty levels of tests, cutoff scores, normative values, and modes of assessment (Mesulam et al., 2014)
D. Sensitive measurement at early versus late stage (Harciarek, Sitek, Kertesz, 2014)
E. Initial assessment
   1. Detailed history
   2. Neurological examination
   3. Neuropsychiatric and Neuropsychological assessment
   4. Brain Imaging
F. Speech and Language Evaluation
   1. Speech Production (Gorno-Tempini et al., 2011), (Sapolsky et al., 2011)
      • Syntax and Grammar: Picture description, story retell, constrained-syntax production task, language sample analysis, BDAE seven point rating scale, Grammatical Competence and Paraphasias, Northwestern Anagram Test
      • Motor aspects of speech: Apraxia Battery for Adults, Fluency: WAB, BDAE seven point rating, scale for length, speech and picture description
   2. Word comprehension (Sapolsky et al., 2011)
      • Single word: BDAE Word Comprehension, WAB Auditory Word Recognition, Psycholinguistic Assessments of Language processing in Aphasia, Spoken Word-Picture Matching, Peabody Picture Vocabulary Test, Cambridge Semantic Category Comprehension
• **Sentence Comprehension**: WAB and BDAE tasks following commands tasks, BDAE Complex Ideational Material, PAL Sentence Comprehension, CYCLE Sentence Picture Matching

• **Semantics**: Pyramids and Palm Tree test, PALPA spoken-word picture matching (subtest 47), PALPA synonym judgments (subtest 49), Peabody Picture Vocabulary Test (Henry, Mooney, & Morhardt, 2015; Nickels, Taylor, & Croot, 2011)

3. **Repetition**: Repeat words, phrases and sentences from WAB and BDAE (Sapolsky et al., 2011)

4. **Object naming**: Boston Naming Test, Phonemic/category fluency tasks (Sapolsky et al., 2011; Nickels, Taylor, & Croot, 2011)

5. **Reading/Writing**: WAB and BDAE written language tasks (Sapolsky et al., 2011)

G. Functional Communication (Harciarek, Sitek, & Kertesz, 2014)

1. Social Networks Package (available online)
2. Aphasia Needs Assessment (available online)
3. Communicative Effectiveness Index-CETI (available online)
4. Frontotemporal Dementia Rating Scale (available online)

H. Cognitive Evaluation: Mini Mental State Exam, Digit Span Forward, Digit Span Backward, Cognitive Linguistic Quick Test, Trail Making Test, Addenbrooke’s Cognitive Examination-Revised

I. Longitudinal Monitoring: **Progressive Aphasia Severity Scale**: (Sapolsky et al., 2014)

   - Informant questionnaire
   - Interview the patient and care partner
   - Comprehensive language assessment
   - Rate domains of language (articulation, fluency, syntax and grammar, word retrieval and expression, repetition, auditory comprehension, single word comprehension, reading, writing, functional communication

IV. **Speech and Language Treatment**

A. Promising treatment effect, especially for participants with mild PPA. May not see generalization, but rather maintain language abilities (Rising, 2014).

B. Participation-directed and patient centered treatment: Identify strengths and weaknesses in communication to create individualized therapy goals/tasks/items. Focuses on functional outcome, partner involvement, adjust cueing level and use of strategies as disease progresses.

C. Language Retraining (Harciarek et al., 2014; Rogalski & Khayum, 2015)

   1. **Self-cueing strategies**: orthographic, semantic, phonological systems that capitalizes on the individual’s residual skills
   2. **Script Training**: formulate and rehearse scripts to increase ability to speak in different situations
   3. **Motor sequencing strategies**: hierarchy of fading cues to increase pronunciation of multisyllabic words

D. Compensatory Strategies (Harciarek et al., 2014)
1. Start early and support multi-modality communication through aided and unaided AAC, and communication partner support
2. Ipad Apps (Rogalski & Khayum, 2015)

E. Partner Training (Harciarek et al., 2014)
1. Improves quality of life by encouraging communication and participation in activities (Nickels & Croot, 2014; Simmons-Mackie et al., 2010)
2. Disease education: understand PPA and the progressive nature, care options and planning for the future (Khayum, et al., 2012)
3. Teach how to be an effective communicator: speak slow, simplify sentence structure and simple vocabulary, provide choices, eliminate distractions, increase use of nonverbal cues (Khayum & Rogalski, 2015)

F. Combine Approaches: use language retraining and compensatory strategies to improve participation (Harciarek et al., 2014)

G. Treatment Domains

1. Naming: Anomia is a common feature (Henry et al., 2013) and may result from damage to semantic or phonologic representations, or impaired transmissions of info between these components (Henry et al., 2008)
   - Evidence-based suggestions (Croot et al., 2009)
     - Generalization may not be a realistic goal
     - Patient-centered tasks/items
     - Errorless learning
     - Use phonological based (for nfvPPA), semantically based (svPPA) or both approaches
     - Semantic features matter
     - Lots of practice
     - Treatment effect appears to be most beneficial at early stages of the disease
     - Magnetic brain stimulation needs further research
   - Remediate impaired linguistic domain (Rising, 2014)
     - svPPA: retain specific concepts and engage residual semantic representations
     - lvPPA: engage residual phonological abilities
   - Capitalize on preserved ability
     - svPPA: phonological and orthographic cues (Rising, 2014), episodic memory/learning (Rising 2014), nonverbal problem solving and visual perceptual skills (Jokel, 2010), self-cueing strategies (Henry et al., 2013)
     - lvPPA: spared semantic and orthographic knowledge (Beeson et al., 2011) and spared cognitive linguistic processes (Henry et al., 2013)

2. Speech Production:
   - Combine pictures with gestures to facilitate sentence production (Schneider et al., 1996)
   - Script training (Khayum, et al., 2012)
• Personalize recording
• Oral reading treatment (Henry et al., 2013)

3. Writing:
• Two routes for spelling: Lexical & sublexical (Tsapkini et al., 2014)
• Teach phoneme to grapheme correspondences (Tsapkini & Hillis, 2013)

4. Transcranial Direct Current Stimulation (Tsapkini et al., 2014)

References


