Characterizing Primary Progressive Apraxia of Speech: A Case Study of a 76-Year-Old Female

Larissa M. Jordan, M.S., M.A., CCC-SLP; Karen Bryant, Ph.D., CCC-SLP; Mary Wille, & Abigail Simon

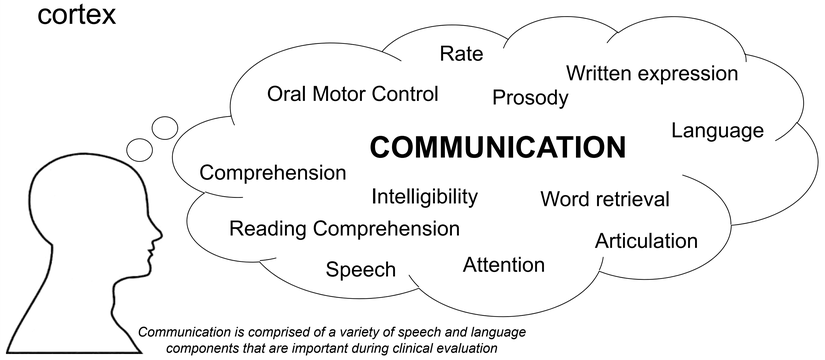
Department of Communication Sciences and Disorders, University of Iowa

BACKGROUND

- Primary Progressive Apraxia of Speech (PPAOS) is a progressive neurodegenerative speech disorder in which apraxia of speech (AOS) presents as the sole or dominant symptom (Jung, Duffy, & Josephs, 2013)
- Little is known about how PPAOS progresses over time, making it difficult to provide prognostic information
- Neuroimaging studies of progression demonstrate that PPAOS either remains the dominant symptom, with development of mild parkinsonism, or progresses fairly rapidly into symptoms aligned with atypical parkinsonian disorder (Josephs et al., 2014)
- No studies have formally evaluated prevalence, but it is estimated to occur in 4.4 per 100,000 people (Botha & Josephs, 2019)
- Expected prevalence of PPAOS is assumed to be higher than current diagnoses, due to limited research, clinical understanding, and recognition

INTRODUCTION

- Speech and language are composed of cohesive neural regions and networks
  - Damage to one component does not break down the entire system, but may demand new approaches to communication
- Dynamic networks create challenges for dissecting between symptoms and likely diagnoses
  - PPAOS is frequently misdiagnosed as primary progressive aphasia or dysarthria, yet in fact these diseases are distinct
- Presenting PPAOS symptoms include: isolated apraxia of speech and focal degeneration of the superior premotor cortex



Communication is comprised of a variety of speech and language components that are important during clinical evaluation

CASE STUDY

- We conducted a case study regarding a 76-year-old woman, "Anne"
- **Patient history:**
  - Anne's stated complaints of self-perceived difficulty to understand, slow speech, and trouble producing some words began three years before Wendell Johnson Speech and Hearing Clinic (WJSHC) visit
  - Referral diagnoses of 'verbal fluency disorder', moderate-severe dysarthria with reduced intelligibility in spontaneous conversation, and primary progressive aphasia

Anne's evaluation at WJSHC:

- **Oral motor exam:**
  - Tongue range of motion and labial function within normal limits
  - Unable to blow or click tongue
  - DDK rates all below average: 1.8 sec /p/, 1.6 sec /t/, 1.4 sec /k/, 0.8 sec /ptk/
  - Sustained /a/ below average, approximately 7 seconds
  - Not able to produce /z/ on command
- **Speech/Voice:**
  - Effortful speech with visible and audible trial-and-error groping behaviors
  - Spoken word intelligibility: 58% for words and 0% for sentences
  - Speech characterized by slow rate, monotone prosody and syllable segmentation
  - Slight huskiness in voice quality
- **Language:**
  - Written confrontation naming BDAE-Short: 100%
  - Able to write complex sentences without difficulty

- **The above symptoms led to diagnosis of probable PPAOS**
  - Score of 100% on written BDAE-Short ruled out primary progressive aphasia diagnosis
- **Clinical recommendations based on evaluation results and Anne's goals:**
  - Improve speech intelligibility using the "Integral Stimulation Approach" and graphic representation of the word segmented into syllables
  - Evaluate for Augmentative Alternative Communication (AAC) candidacy and devices
- **Outcome**
  - Anne preferred augmenting speech with pen and paper
  - Counseled on progressive nature of disease, urged to revisit AAC options

CLINICAL RELEVANCE

- Clinicians may work with patients to provide compensatory strategies that improve speech intelligibility
- Clinicians are responsible for counseling patient regarding progressive nature of disease and introducing options for new methods of communication, such as AAC
- Incorporate collaborative treatment team: neurologist, psychologist, social worker, speech-language pathologist, family members, etc.

KEY FINDINGS

- PPAOS is a progressive disease, therefore therapy goals must include management, counseling, and optimizing any new models of communication
- Appropriate AAC devices prove beneficial
- Future research could focus on clinical applications for management of PPAOS and progressing symptoms
- Greater awareness of PPAOS may contribute to accurate and timely diagnosis

SELECTED LITERATURE CITATIONS

Botha, H.A., & Josephs, K.A. (2019). Primary progressive aphasias and apraxia of speech. *CONTINUUM: Lifelong Learning in Neurology*, *25*(1), 101-127.

Josephs, K. A., Duffy, J. R., Strand, E. A., Machulda, M. M., Senjem, M. L., Master A. V., Gunter, J. L., Schwarz, C. G., Reid, R. I., Spychalla, A. J., Lowe, V. J., Jack, C. R. Jr, Whitwell, J. L. (2014). The evolution of primary progressive apraxia of speech. *Brain*, *137*(10), 2783–2795.

Jung, Y., Duffy J., Josephs, K. (2013). Primary progressive aphasia and apraxia of speech. Seminars in Neurology, 33(4): 342-347.

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