SUMMARY

What are progressive apraxia of speech (PAOS) & primary progressive apraxia of speech (PPAOS)?

- **Progressive Apraxia of Speech (PAOS)** - AOS of insidious onset & gradual progression due to a neurodegenerative condition. May be associated with aphasia &/or dysarthria.
- **Primary Progressive AOS (PPAOS)** - PAOS in which AOS is the first, only, or most salient feature of neurodegenerative disease & in which criteria are not met for diagnosis of another neurodegenerative disease (e.g., progressive supranuclear palsy syndrome, corticobasal syndrome)

Basic Demographics – PPAOS

- Age at onset: late 60s - early 70s (range = late 40s - early 80s)
- uncommon before age 65, but can occur earlier
- somewhat older than onset of agPPA (as much as a decade in some studies)
- Gender: probably more frequent in women (~50-70%)
- Education: ~ 15 years (10-20 years)
- Handedness: c/w population as whole (~ 90% R)
- Initial presentation for evaluation: ~ 2-3 years post onset

Speech features associated with PPAOS (most common, ordered; Josephs et al., 2012)

- Slow overall speech rate
- Lengthened intersegment durations (between sounds, syllables, words or phrases; possibly filled, including intrusive schwa)
- Increased sound distortions or distorted sound substitutions with increased utterance length or increased syllable/word articulatory complexity
- Syllable segmentation within multisyllabic words
- Sound distortions
- Syllable segmentation across words in phrases & sentences
- Audible or visible articulatory groping; speech initiation difficulty; false starts/restarts

Subtypes of PAOS or PPAOS? - Preliminary evidence (Josephs et al., 2013)

- **Type 1** – Predominated by articulatory abnormalities (e.g., distortions & distorted substitutions, repeated sounds, attempts at self-correction)
  - More evident when aphasia present & > AOS
  - Tends to be associated with widespread involvement in premotor, prefrontal, temporal-parietal lobes, caudate & insula
- **Type 2** – Predominated by prosodic abnormalities (e.g., segmentation of words & syllables)
  - More evident in PPAOS without aphasia or when AOS > aphasia
  - Tends to be associated with involvement in premotor cortex and midbrain atrophy
- **Type 3** – No clear difference in prominence of articulatory versus prosodic abnormalities
Accompanying Deficits @ initial evaluation - PPAOS

- Nonverbal oral apraxia: ~ 50% of cases at initial evaluation
  - % increases with disease progression
  - Probably more frequent when aphasia is also present (80-90%)
- Dysarthria: Present in ~ one-third
  - increased frequency with disease progression
  - most often spastic > hypokinetic, or mixed spastic-hypokinetic (Duffy, Strand & Josephs, 2015)
- Dysphagia: Not usually evident unless dysarthria also present

Acoustic Temporal Correlates

- Temporal measures of word and sentence duration, and maximum rate speech-like tasks distinguish PPAOS from normal speech and agrammatic PPA

Disease Course: Emerging Motor Deficits with Disease Progression

Josephs et al. (2014) – Followed evolution in 13 people with PPAOS without other neurologic signs at initial evaluation
- Initial evaluation at ~ 4 years post onset
- f/u at ~ 7 years post onset
- All developed extrapyramidal (parkinsonian) symptoms
- In 8/13 PPAOS remained predominant problem
- 5/13 evolved to a severe PSP-like syndrome
  - severe parkinsonism, near mutism, dysphagia, vertical supranuclear gaze palsy, urinary incontinence, balance difficulty with falls, & limb apraxia
- Conclusion: Some will fairly rapidly evolve by ~ 5 years to a devastating PSP-like syndrome, while others will retain PPAOS diagnosis, although some with mild parkinsonism, at ~7 years. Some evolve to a corticobasal (CBD)-like syndrome (CBS) with asymmetric rigidity, limb apraxia, & other extrapyramidal features

Neurologic underpinnings

- Primary neuroanatomic correlates
  - Grey matter - Superior lateral premotor cortex & supplementary motor area
  - White matter - Same as grey matter + inferior premotor cortex & body of corpus callosum, superior longitudinal fasciculus, esp. premotor components
  - Primary composite – Superior lateral premotor cortex & supplementary motor area.
- Pathology - PPAOS very consistently but not invariably associated with
  - tau biochemistry (tauopathy)
  - progressive supranuclear palsy or corticobasal degeneration
- In PAOS, when nonfluent/agrammatic aphasia present & AOS = or > nonfluent aphasia
  - Predicts PSP or CBS pathology in ~ 90%
  - PSP or CBD (tauopathy) possible in PPA (with no or less severe AOS), but not common (<20%)

Summary – Main take home points

- PPAOS exists - may not be as rare as literature implies.
- When primary, it should not be subsumed under classifications of primary progressive aphasia (PPA)
• It reflects L or L>R hemisphere abnormalities - frontal lobe (superior & mid premotor cortex, SMA)
• Tends eventually to be associated with conditions with prominent motor rather than cognitive deficits (e.g., PSPS, CBS)
• Tends to predict pathology consistent with tauopathy (e.g., PSP, CBD)

Selected References