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Diagnosis & Treatment of Complex Cases in Speech-Language Pathology

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Guest Editor: **Richard Peach, PhD, CCC-SLP, BC-ANCDS**

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Primary Progressive Apraxia of Speech and Aphasia in a Complex Case of Neurodegenerative Disease

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Primary Progressive Apraxia of Speech and Aphasia in a Complex Case of Neurodegenerative Disease

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Rush University Medical Center
Chicago, Illinois

Learning Objectives

- To describe the clinical presentation of progressive apraxia of speech and aphasia in a complex case of neurodegenerative disease
- To identify the speech and language characteristics of progressive apraxia of speech and aphasia
- To provide recommendations for the management of the disorder

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Case Study: Pt. M

- 67 y.o. female
- Right-handed
- January, 2012: Acute onset of speech disturbance
  - Slowed speech but without articulatory difficulty
  - Reading/writing preserved
- PMHx:
  - Depression
  - Chronic hearing loss
  - Smoking (1 pack/day x 40 years)
- Social Hx:
  - 2 yrs of college
  - Retired office manager
  - Lives with daughter

Evaluations: 2012 – 2013 (OSH)

- November, 2012; May, 2013: Neurology
  - Stroke, myasthenia gravis workups negative
  - EEG normal
  - MRIs/MRAs essentially normal with exception of very mild chronic white matter vascular changes
  - ?'s re: possible L > R temporal atrophy
- December, 2013: Speech-language
  - Mod-severe dysarthria
  - Poor intelligibility
  - ↓ labial, lingual strength, ROM, and coordination
  - Preserved auditory comprehension
  - Mild deficits in reading comprehension
  - Mild expressive aphasia (word-finding difficulty)
  - Mild cognitive deficit
  - No dysphagia

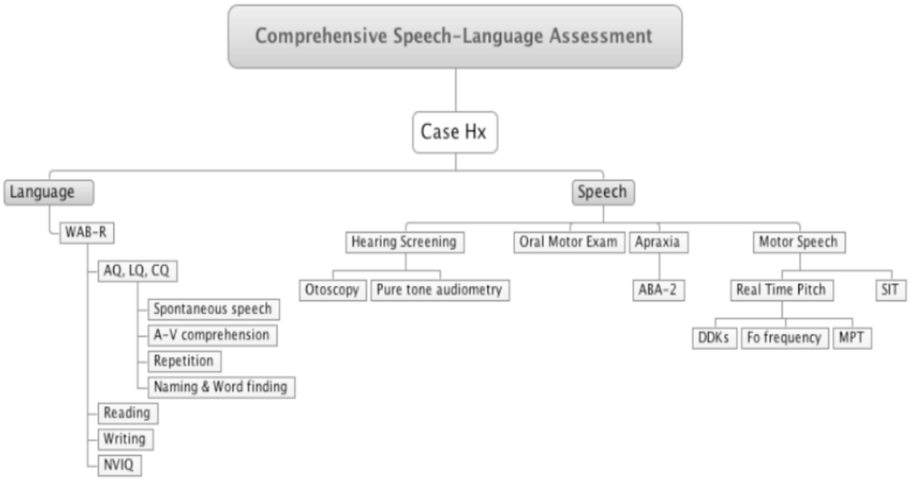
Evaluation: June, 2014 (RUMC)

- Neurology consultation
  - MMSE = 30/30
  - Significant speech fluctuation (variable slowing)
  - Normal prosody
  - Poor intelligibility
  - Paraphasic errors, esp. on confrontation naming
  - Difficulty with repetition
  - Speech for reading worse than spontaneous speech
  - Writes spontaneous sentence, unable to perform to dictation
  - No significant CN, motor, or sensory abnormalities other than dysarthria
  - Vibration minimally diminished in R toe
- MRI Brain
  - Minimal chronic small vessel ischemic changes within PVWM
  - No acute intracranial pathology
- DDX: PPA versus AOS

Progressive apraxia of speech as a sign of motor neuron disease (Duffy, Peach, & Strand, 2007)

- Level of Evidence: Case series
- Method:
  - Retrospective study of 7 pts with dx of MND and AOS
    - Identified from among 80 patients with AOS due to various neurodegenerative diseases (Duffy, 2006)
    - Age: mean= 67 years; range = 48-84 years
  - History, complaints, neurological and speech-language findings documented
- Findings
  - Speech first symptom in 5 pts (prominent complaint in all 7)
    - 1 with personality changes
    - 1 with toe paresthesias
  - Non-verbal oral apraxia in 5 pts; later emerged in remaining 2 pts
  - Dysarthria present in all (slow rate, reduced intelligibility, monopitch & monoloudness)
    - 3 with spastic
    - 2 with mixed flaccid-spastic
    - 2 with undetermined type (later mixed)
  - Variable presence of aphasia
    - No aphasia in 3 pts; 2 with mild-mod “nonfluent” aphasia; 2 w/ abnormal performance unable to be directly attributed to aphasia
  - Short breath groups during speech, despite adequate respiratory support (4 of 7)

Evaluation: July, 2014



Case history

- Primary complaints:
  - Poor speech
  - Overall fatigue
- Both have worsened since May 2014
- Difficulty in “getting words out”
  - Per daughter: “Knows what she wants to say, but can’t say it.”
- No changes in mental status reported

Clinical Observations

- No difficulty observed swallowing small amount of liquid
  - Eats several small meals/day
  - Recent weight loss
  - GERD reported
- Mildly unsteady gait
- Fatigue (increasing throughout session)
- Uses marker board to aid in communication

Assessment - WAB-R

Subtest	Score	Impressions
Spontaneous Speech:		Non-fluent production, reduced phrase length, incomplete sentences
Information Content	9/10	
Fluency	4/10	
Auditory Verbal Comprehension	7.95/10	Y/N responses preserved, moderately impaired sequential commands
Repetition	3.2/10	Hesitations, syllable segmentation, consonant and vowel substitutions, verbal substitutions
Naming & Word finding	8.3/10	Mildly impaired object naming and word fluency Moderately reduced sentence completion

Assessment - WAB-R (cont.)

Domain	Score	Interpretation
Reading	84/100	Mild-moderate impairment
Writing	78/100	Moderate impairment
Praxis	9.2/10	Mildly reduced
Raven's Coloured Progressive Matrices	29/36	51st percentile
Aphasia Quotient (AQ)	64.9	Cutoff = 93.8
Language Quotient (LQ)	72.7	
Cortical Quotient (CQ)	74.4	Cutoff = 90

Assessment (cont.)

- Hearing screening
  - Otoscopic exam
  - Pt report:
    - Difficulty in R ear
    - Uses phone on L only
  - Pure tone audiometry:
    - Did not pass 40 dB HL in either ear

- Oral Motor Exam
  - Gross facial symmetry at rest
  - **Mild flattening of R nasolabial fold**
  - Adequate labial, mandibular strength
  - Symmetrical tongue (at rest and upon protrusion)
  - **Mild R lingual weakness**
  - Adequate palatal symmetry and ROM
  - Gag present
  - Suck & snout absent
  - No lingual, chin fasciculations

Assessment - ABA-2

Subtest	Cutoff	Score	Impressions
Increasing word length	1	8	Severe impairment
Repeated trials of complex words	28	7	Moderate impairment
Utterance time (sec) polysyllabic words	15	22	Mildly lengthened
Limb apraxia	44	46	None
Oral apraxia	44	49	
Apraxic Speech Behaviors	4	14/15	Phonemic transposition errors Highly inconsistent errors Numerous and varied off-topic attempts Abnormal prosodic features

Assessment – Real Time Pitch

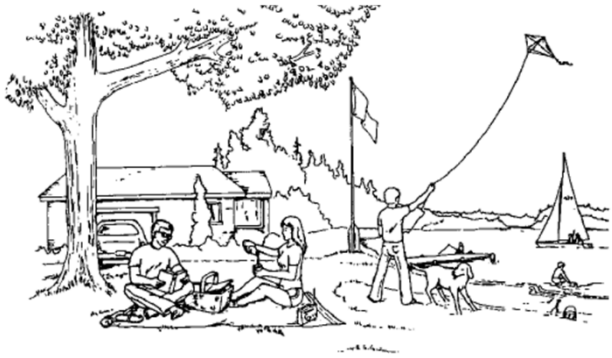
Measure	Results
AMRs	/p^/=2.5/s, /t^/=2.3/s, /k^/=2.3/s (Normal: /p^/=6.3/s, /t^/=6.2/s, /k^/=5.8)
SMRs	/p^t^k^/=1/sec (Normal: /p^t^k^/=5.0/sec (SD 0.7))
Mean Fundamental Frequency	Automatic speech: 131.1 Hz (SD 27.1) Reading: 122 Hz (SD 17.0)
Mean Loudness	Reading: 53.9 dB (SD 2.64)
Maximum Phonation Time	Average: 3.2 sec (Normal median= 14.4, SD 5.7)

- AMRs and SMRs
  - Slowed rate with irregular rhythm
  - Decreased coordination
- Maximum Phonation Time - Substantially reduced respiratory-phonatory support for speech production

Assessment – Speech

- Conversational Speech
  - Breath groups = 2-3 words
  - Short, visible inhalations
  - Marked vocal fatigue
  - Effortful, strained, hypernasal, imprecise, monotone/monopitch
- Sentence intelligibility: 53% (poor)
- Speaking rate = 35 WPM (normal = 190)
  - Rate of intelligible speech = 18.9 WPM
  - Rate of unintelligible speech = 16.9 WPM

Picture Description



Okay…father flying kite…kite…now daughters were (unintelligible)…fish her …her …and then a lot of stuff…The girl's uh…has her /s/ sting (sand) sting…and the man reading book and the woman's um…putting cup…pouring…drink…and the…and the…sailboat

Oral Reading

Impressions: Cognition

- Nonfluent aphasia:
  - Moderately reduced speech fluency, grammar, and phrase length
  - Mild impairments to auditory and reading comprehension
  - Mild naming deficits
  - Moderate writing impairment
- Nonverbal cognition: Low normal

Impressions: Speech

- Characteristics:
  - Slow rate
  - Frequent insertion of filled pauses w/in words
  - Syllable segmentation
  - Perceived sound substitutions
  - Reduced speech prosody (monotone & monopitch)
- Additional observations:
  - Hypernasality
  - Imprecise articulation
  - Strained vocal quality
  - Reduced respiratory-phonatory support
  - Poor speech intelligibility in both known and unknown contexts

Diagnosis

- Results are consistent with:
  - Primary progressive apraxia of speech & aphasia
- Co-occurring:
  - Spastic dysarthria
- Progressive AOS and aphasia can be first and only symptom of neurodegenerative disease for extended period of time
- Prognosis: guarded to poor
  - Based on suspected degenerative nature

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Apraxia of speech in degenerative neurologic disease (Duffy, 2006)

- Level of Evidence: IV
- **Method:**
  - Retrospective review of 80 pts seen between 1985 and 2004 who had AOS not less severe than any aphasia present in which cause was degenerative
    - Average age= 69 years (range=36-86 years)
    - Speech-language difficulty was first symptom in 80%
      - Was *only* initial pt complaint in 56%
- **Findings:**
    - Dx of AOS, majority displayed: slow rate, distorted substitutions, segmentation of syllables or excess and equal stress, poorly sequenced SMRs, and ↑off-target artic. errors with ↑ utterance length
    - Aphasia present in 49% (mild-mod severity, mostly non-fluent)
    - Dysarthria present in 50% (2/3 had spastic, hypokinetic, or mixed spastic-hypokinetic)
    - 77% had nonverbal oral apraxia
    - None had nonaphasic cognitive deficits worse than their AOS
    - Neurologic:
      - Grossly normal EEGs & CT scans; MRIs indicated atrophy in only L, or L> R; SPECT indicated L>R abnormalities in 48%

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Apraxia of speech in degenerative neurologic disease (Duffy, 2006)

- **Conclusions:**
  - 44% of sample received neurological dx from the neurologist based exclusively on speech-language findings or were strongly influenced by them
    - 90% of patients with neurological dx primarily based on:
      - presence of AOS
      - diseases lateralised to L hemisphere OR
      - Conditions assoc. with prominent motor manifestations (CBD, IPD, ALS/MND)
  - AOS can be the predominant and sometimes ONLY symptom in pts for whom S/L symptoms are first manifestation of degenerative disease
    - Distinguish between **PPA** and **primary progressive AOS and aphasia**
      - Implications for management (is there a language component?)

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Motor speech disorders associated with primary progressive aphasia (Duffy et al., 2014)

- AOS more strongly associated with PPA than dysarthria
- Among dysarthrias occurring with PPA, spastic and hypokinetic types occur most frequently
- AOS and dysarthria are uncommon in semantic and logopenic variants of PPA although features of AOS may be present in a minority of logopenic cases
- AOS is very common in nonfluent variant PPA; approximately one-third of patients with agrammatism and AOS have dysarthria

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Recommendations

- Audiologic examination
- Neurologic examination to further describe nature of neurodegenerative disorder
  - PET
  - Electromyography
- Continued speech-language treatment
- Follow-up assessment in 6 months

Treatment Recommendations

- Train *compensatory strategies* for enhanced verbal communication and QOL
  - Provide semantic cues and topic areas for semantic context
  - Use gestures and orthographic cues
- Continue use of writing tablet/other AAC
- Consider use of TTY
- Increase vocal loudness to improve respiratory support and speech intelligibility
- Develop plan for weekly homework to maintain current best levels of function
- Discontinue isometric/isotonic oral exercises

Wambaugh et al., 2006b; Abbs & De Paul, 1989

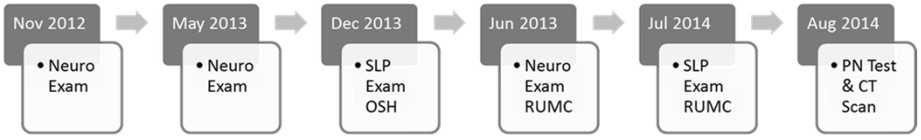
Patient and family education

- Describe components of speech-language assessment
- Review findings
  - Progressive AOS and aphasia with co-occurring spastic dysarthria
  - Not the result of a single degenerative condition; tends to be associated with diagnoses that have prominent motor rather than cognitive deficits (CBD, PSP, MND)
- Discuss recommendations
  - Compensatory strategies vs. oral exercises
  - Importance of tracking the progression

Follow-up: January, 2015

- Contact with Neurology to prepare for speech-language pathology follow-up testing
  - August, 2014: Blood tests for paraneoplastic antibodies (negative), chest CT for complaints of cough, weight loss
  - Neurology follow-up scheduled for November, 2014; patient refused to attend
  - No attempts to schedule speech-language pathology follow-up
- No further medical care provided at RUMC

Summary



Summary

Progressive AOS	Aphasia	Spastic dysarthria
-Slow rate -Insertion of pauses w/in words -Syllable segmentation -Perceived sound substitutions -Reduced speech prosody	-Reduced fluency -Poor sentence grammar -Reduced phrase length -Mildly impaired auditory and reading comprehen. -Mild naming deficits -Writing impairment	-Imprecise articulation -Strained vocal quality -Reduced respiratory-phonatory support -Hypernasality -Poor speech intelligibility

Additional findings:

-toe paresthesias

-possible L or L > R atrophy

-overall weakness/fatigue

-NVIQ: low range of normal

Conclusion

- AOS can be the first and most prominent manifestation of neurodegenerative disease (Duffy, 2006)
- Speech-language assessment essential to identifying and describing the communication deficits found in neurodegenerative disorders as well as to track their progression over time
- SLP must be cognizant of the nature and progression of a neurodegenerative disorder when planning intervention

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