Progressive Speech and Language Disorders

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Outline
- Part One: Background
  - Primary Progressive Aphasias (PPA)
    - Semantic Variant of PPA
    - Logopenic Variant
    - Agrammatic/ non-fluent variant of PPA
  - Primary Progressive Apraxia of Speech (PPAOS)
- Part Two: Differential Diagnosis/ Case Studies
- Part Three: Treatment

Three General Components of Speaking

1. Cognitive linguistic processing
2. Sensorimotor planning/programming
3. Neuromuscular execution
What if something goes wrong?

- Brain lesion
  - Cerebrovascular Accident or Stroke (CVA)
  - Occlusive (ischemic) due to thrombosis or embolism
  - Hemorrhagic due to aneurysm or AVM
  - Trauma (TBI)
  - Tumor
  - Infection

What if something goes wrong?

- Brain lesion
  - Cerebrovascular Accident or Stroke (CVA)
    - Occlusive (ischemic) due to thrombosis or embolism
    - Hemorrhagic due to aneurysm or AVM
  - Trauma (TBI)
  - Neoplasm (tumor)
  - Infection/Toxin
  - Disease Process

Three General Components of Speaking:

1. Cognitive linguistic processing
   - Aphasia
2. Sensorimotor planning/programming
   - Apraxia of speech
3. Neuromuscular execution
   - Dysarthria

Three General Components of Speaking:

1. Cognitive linguistic processing
   - Dementia
   - Progressive aphasia
2. Sensorimotor planning/programming
   - Progressive apraxia of speech
3. Neuromuscular execution
   - Dysarthria
Primary Progressive Aphasia (PPA)

- A rare degenerative, neurological syndrome.
- Insidious onset.
- Progressive.
- Primary and initial complaint of language difficulties.

Gorno-Tempini et al. Diagnosis Criteria (2011)

- Pattern of deficits are not accounted for by other nondegenerative nervous system, medical, or disorder at onset.
- Absence of episodic memory, visual memory, and visuo-perceptual impairments at onset.
- Absence of behavioral disturbance at onset.

Variants of PPA

- Semantic variant
- Logopenic
- Agrammatic/ Non-fluent variant
Semantic variant of PPA

- Anomia with loss of single word meaning.
- Difficulty generating and **recognizing** familiar words.
- For rare words first and common nouns for later stages.
  - Verbs and abstract words are often spared.
- During confrontation naming, will demonstrate low confidence for identifying target word.

Semantic variant of PPA

- Fluent spontaneous speech is mostly retained.
- Some patients have problems recognizing familiar objects and faces.
  - The presence of this sign can help confirm the diagnosis.
- Report difficulty with writing and spelling, particularly with irregularly spelled words.
- Surface dyslexia

Logopenic variant of PPA

- A slow rate of speech, secondary to word retrieval difficulties.
- Phonologic errors may be present.
- Sentence and phrase repetition is impaired.
  - Repetition of single words is spared.
- Reading and writing abilities may be preserved longer than speech.
- Trouble understanding long or complex verbal information.

Agrammatic/ non-fluent variant of PPA

- **Speech is effortful and reduced in quantity.**
- Sentences become shorter and **word-finding hesitations** become more frequent.
- Pronouns, conjunctions and articles are lost.
- Word order may be abnormal, especially in writing or e-mails (agrammatic).
- Reversals (e.g., “he” for “she”; “yes” for “no”).
- Reduced comprehension for long and grammatically complex sentences.
PPA implies a disorder of language impairment. Aphasia may not even be present!

Primary Progressive Apraxia of Speech (PPAOS)

- Insidious
- Progressive
- Motor planning difficulty
PPAOS Diagnostic Criteria

• Initial complaint of speech difficulty.
• Unequivocal AOS.
• No evidence of aphasia.
• Normal neurological examination.
• Normal neuropsychological testing.

Patient Perceptions and Complaints

• “Pure” AOS:
  • “my speech won’t come out right”
  • “know what I want to say but doesn’t come out right”
  • not as fluent as before
  • mispronounce words
  • with mild or moderate AOS, patients report being surprised by errors that “sneak into” narratives
  • complaints usually center around articulation problems
  • some patients report having to speak slowly or more carefully to prevent errors
  • predict errors on multi-syllabic or difficult to pronounce words
  • recognize errors and attempt to correct them
  • problems may be more obvious when stressed or fatigued

Motor processes of speech

Prosody
Articulation
Resonance
Phonation
Respiration

PPAOS Types

PPAOS
Prosodic
Phonetic
Classification is **not** dichotomous, but a relative predominance

- Phonetic abnormalities
  - Sound additions
  - Sound substitutions
  - Sound distortions

- Prosodic abnormalities
  - Slow rate
  - Inter-word segmentation
  - Intra-word segmentation

Clinical Information

- Demographics
- Severity
- Language
- Cognition
- Neurologic

How does the initial presentation influence disease progression?

Some ideas...
Part Two
Differential Diagnosis

Assessment and differential diagnosis

Step 1: Is aphasia present?
Step 2: Is apraxia of speech present? If so, what dominates?
Step 3: Is dysarthria present?
Largely completed simultaneously!

First step
• Is aphasia present?
Variants of PPA: assessment tips

• Agrammatic variant
  • Loss of function words/ morphological markers
  • Assess spoken language AND writing

• Semantic variant
  • Surface dyslexia
  • Loss of word meaning

• Logopenic variant
  • Anomia without loss of single word meaning
  • Poor sentence repetition
  • Comprehension deficits
  • Phonological errors are present

Semantic dementia

Agrammatic
PPA
• Omission of articles

Apraxia of Speech

Logopenic
PPA
• Comprehension difficulties

Sound errors

Tasks for Assessing Motor Speech Programming Capabilities

• General conversational ability
• Imitation (sounds, words and sentences)
• Narrative picture description
• Automatic tasks (counting, days of the week, sentence completion)
• Singing a familiar song
• Reading aloud
• SMRs
• Writing sample

Second Step

• Is apraxia of speech present?
Tests to Assess and Characterize AOS

- Apraxia of Speech Rating Scale (ASRS)
- Articulatory Error Score (AES)

### Apraxia of Speech Rating Scale (ASRS)

<table>
<thead>
<tr>
<th>Score</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Not observed in any task</td>
</tr>
<tr>
<td>1</td>
<td>Infrequent</td>
</tr>
<tr>
<td>2</td>
<td>Frequent but not pervasive</td>
</tr>
<tr>
<td>3</td>
<td>Nearly always evident but not marked in severity</td>
</tr>
<tr>
<td>4</td>
<td>Nearly always evident and marked in severity</td>
</tr>
</tbody>
</table>

**Phonetic Features**

1. Sound distortions (excluding distorted substitutions or distorted additions)
2. Distorted sound substitutions
3. Distorted sound additions (including intrusive schwa)
4. Increased sound distortions or distorted sound substitutions with increased utterance length or increased syllable/word articulatory complexity

**Prosodic Features**

5. Syllable segmentation within words > 1 syllable (Brief silent interval between syllables and/or inappropriate equalized stress across syllables)
6. Syllable segmentation across words in phrases/sentences (Increased inter-word intervals and/or inappropriate equalized stress across words)
7. Slow overall speech rate (apart from pauses for word retrieval and/or verbal formulation)
8. Lengthened vowel &/or consonant segments independent of overall slow speaking rate

### Articulatory Error Score (AES)

- “Repeat each of the following words three times each.”
  - Cat
  - Catnip
  - Catapult
  - Catastrophe
  - Harmonica
  - Specific

- “Repeat these sentences one time.”
  - We saw several wild animals.
  - My physician wrote out a prescription.
  - The municipal judge sentenced the criminal.
Third step
• Is dysarthria present?

Differential diagnosis: Dysarthria
• Oral mechanism examination
• AMRs
• SMRs
• Groping?
• Distortions v. substitutions v. distorted substitutions

Other tips for diagnosis
• History is critical!
• Insidious onset
• Progression over time
• Asking the right questions is crucial
Case History Question Suggestions
• Any difficulties with speech or language as a child?
• How would you describe the difficulties you have been having?
• How long have the difficulties been occurring?
• Was the onset sudden? Or gradual?
• Any worse over time?

Case History: Speech
• Speech better at any time of the day?
• Things that make it worse?
• Does alcohol have any effect?
• Any point at which it completely returns to normal?
• Any situation in which people have a difficult time understanding you?
• Difficulty producing words, despite knowing what you want to say?
• Difficulty with longer/ harder words?

Case History: Language
• Difficulty understanding?
• Difficulty thinking of words?
• Mean to say one word and another word comes out? i.e. dog for cat or cone for comb?
• Hear a word and not know what it means?
• Difficulty with yes/ no?
• Difficulty with left/ right?
• Ever leave words out in speaking or writing?

Case History: Other
• Any changes in handwriting?
• Any changes in behavior or personality?
• Laughing or crying more easily? At inappropriate times?
• Any difficulty chewing or swallowing liquids or solids?
Summary

- Is the disorder progressive?
  - Change over time

- Is aphasia present?
  - Agrammatism
  - Loss of word meaning
  - Phonologic errors
  - Reduced comprehension

- Is apraxia of speech present?
  - Articulatory errors
  - Segmentation

Questions?

Case Study One

- 81 year old
- 2 year history of difficulty expressing himself ("executing") and reading
- Silent reading is normal in rate, accuracy, and comprehension
- Occasional word finding difficulties
- He denies difficulty with spoken or written language comprehension

Case Study Two

- 63 year old
- 2.5 years symptom duration
- Reports difficulty remembering names of people/ recalling their faces
- Reports word-finding difficulty and increasing difficulty with reading and spelling
- Denies difficulty with comprehension
Case Study Three

- 62 year old
- 2.5 year history of speech difficulty (noticeable to others only in the last 6 months)
- Longstanding lisp
- Denies difficulty with word retrieval
- Reports changes in handwriting (more laborious; larger print)

Case Study Four

- 69 year old
- 1.5 year history of “missing words,” confusing tenses, word order, and pronouns
- Denies difficulty with comprehension

Part Three

Treatment
Using a Theoretical Framework to Guide the Clinical Management of Intelligibility Disorders

What is an intelligibility disorder?
What is an intelligibility disorder?
A breakdown in the reception of a spoken message.

- Acoustic signal (sender)
- Transmission (environment)
- Reception (listener)

Possible sources of intelligibility breakdowns:
- Acoustic signal
  - Dysarthria
  - Stuttering
  - Cleft palate
  - Non-native speech
  - Cochlear implant signal
  - Synthesized speech (AAC)
- Transmission
  - Noisy environments
  - Cell phone
  - Acoustic reverberation
  - Cocktail party
- Reception
  - Hearing loss
  - Auditory comprehension deficits
  - Lack of experience or familiarity with distorted signal

Let's take a look at Dysarthria
- Motor speech disorder
- Results from neurological injury due to damage in the Central or Peripheral Nervous Systems
- Affects one, many, or all speech subsystems (respiration, resonance, phonation, articulation)

Differential diagnosis
- Examination of oral structure and function
- AMR, SMR
- Sustained /a/
- Measures of intelligibility
  - ...but largely, via the associated medical diagnosis
    - Hyperkinetic dysarthria- Huntington's Disease
    - Hypokinetic dysarthria- Parkinson's Disease
    - Mixed spastic-flaccid dysarthria- ALS
    - Ataxic dysarthria- cerebellar degeneration
(Mayo Classification System- Darley, Aronson, and Brown)
Mayo clinic classification approach

Hyperkinetic
Spastic
Hypokinetiс
Flaccid
Ataxic
Mixed

But does it matter?

• Does the “dysarthria subtype” dictate your clinical decision making?

• Probably not.

Mayo Clinic approach

1) there is considerable overlap in speech symptoms among the classification categories, and
2) speech symptoms within a given classification may differ along the severity dimension.

• This classification approach does not map well to the resulting communication disorders or to intervention targets.
  • It does help diagnose neurologic disease!
• Bottom line: we can’t treat the diagnosis, we need to treat the resulting intelligibility disorder!

What do I mean by that?

• We identify what is wrong and we try and fix it!

• Theory will tell us what will have the largest impact on the listener.
Speech Perception Theory

A theory is a set of interrelated principles and definitions that present a systematic view of phenomena by specifying relationships among variables with the purpose of explaining natural phenomena.

“Intelligibility is as much in the ear of the listener as it is in the mouth of the speaker.”
(Martin & Weismer, 1992)

Theoretical perspective

- Dysarthria interferes with perceptual processes:
  - Activation of an optimal *lexical candidate* pool
  - Competition among lexical candidates
  - Application of strategies to lexically segment the connected speech

How does it do that?

- To understand this interference, let’s take a look at “normal” speech perception

Namely:

- How do listeners understand single words? and connected speech?
- How does dysarthria interfere with these processes?
Models of Spoken Word Recognition
(e.g. Logogen, Cohort, Trace, Shortlist, Neighborhood Activation, PARSYN)

Commonly assume that at least two fundamental processes underlie spoken word recognition:

**lexical activation:** When a listener encounters a spoken word, certain aspects of the acoustic signal serve to activate a set of possible lexical candidates.

**lexical competition:** This is followed by competition among these candidates for the best fit with the input, and the winner of this competition is the recognized word.

Time Course

- Time course
  - Left-to-right processing of acoustic information
  - Delay decision until whole word is spoken

Dysarthric speech and word recognition models

- Distortions
- Omissions
- Substitutions
- Vowel reductions

"dip"
"_ip"

There is very little predictive value in single word intelligibility
Perceiving connected speech is a very different beast!
Deciphering Connected Speech

- Lexical activation and lexical competition are strongly influenced by the message
  - Semantics, syntactics, knowledge of topic and speaker: prime the lexical candidates and facilitate efficient competition
- The quality of the acoustic signal is not critical as listeners listen for words, not phonemes
  - Phonemic restoration
- Listeners perform “lexical segmentation”

Lexical Segmentation

- A fundamental process in deciphering degraded speech that determines “word size frames” for lexical access.

Metrical Segmentation Strategy

(Cutler & Norris, 1987)

When listeners encounter degraded speech
They listen for STRONG (stressed) syllables
Treat them as WORD ONSETS
And they’ll most often be right in English

Lexical Segmentation

- Semantically anomalous, syntactically correct
If you don’t “chop up” the acoustic stream into words correctly…

You have no chance at using all available acoustic and top-down information to make a best match for the words.

Regardless of “dysarthria type,” different types of degradation patterns (e.g. slow rate, reduced pitch and loudness variation, consonant imprecision), yield different perceptual errors.

THIS is the intelligibility disorder!

Listeners are flexible

• Use available acoustic cues, even when they’re degraded;
• Can switch the amount of reliance on various cues, depending on which are most robust and systematic;
• They use their “higher level” knowledge to facilitate and guide speech understanding
  • Semantics, syntax, phonotactics, knowledge of topic/ speaker

Context

Acoustic
“Signal-Complementary Information”
Lindblom, 1990

- Speech Signal transmitted
- Listener applies information
- Improved performance

Augmented perceptual processing

Listener manipulation

Increased intelligibility

Activating optimal lexical candidates

- Alphabet cueing
  - e.g. Hustad, Jones, & Dailey, 2003; Hustad, 2005

- Providing topic cues
  - e.g. Hustad, Auker, Natale, & Carlson, 2003; Jones, Mathy, Azuma, & Liss, 2004; Utianski, Azuma, and Liss, 2010

- Providing semantic context
  - e.g. Dongilli, 1994; Hammen, Yorkston, & Dowden, 1991

Lexical competition

- Providing a syntactic template or structure
- Creating semantically and syntactically predictable utterances
- Using supplementary cues such as first-letter, word-class cues, or gestures

Perceptual training

- Training material
  - Talker-specific
  - Disorder-specific

- Feedback type and frequency
  - Passive exposure vs. training
  - Generalization of minimal exposure/training

- Training regimen and intensity
Different patterns yield different errors

- Equivalent intelligibility
  - Phonetic errors
  - Segmentation errors

Intelligibility results

<table>
<thead>
<tr>
<th></th>
<th>Control</th>
<th>Familiarization</th>
<th>Topic Knowledge</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypokinetic</td>
<td>42%</td>
<td>45%</td>
<td>44%</td>
</tr>
<tr>
<td>Ataxic</td>
<td>43%</td>
<td>51%</td>
<td>50%</td>
</tr>
</tbody>
</table>

Metrical Segmentation Strategy

- Rhythmic structures of language to identify word boundaries
- Strong syllable as potential word onsets (Cutler & Carter, 1987)
- Based on the statistical probabilities of English
  - More likely to see insertions before strong syllables and deletions before weak syllables

Quality of lexical segmentation
Identifying the potential source of benefit

- Do listeners more correctly transcribe topic words, regardless of a lack of overall intelligibility for hypokinetic speech?

- Does the dysarthria type difference disappear when we look at the listener’s ability to use topic knowledge?

Take away points

- Topic knowledge and familiarization offer equal perceptual benefits for ataxic speech

- Sources of benefits for each speaker group
  - Listeners of hypokinetic speech rely on syllabic stress to segment words
  - Listeners of ataxic speech utilize lexical access and acoustic-phonetic remapping to segment words

Research-practice divide

Current pitfalls in the remediation of intelligibility disorders
**Theoretical and Clinical Perspectives**

**Theoretical Goal:** Advance knowledge about basic mechanisms and processes

**Clinical Goal:** Determines how characteristics of a particular clinical population relate to hypotheses generated from a theory or model.

*Bernstein & Weismer, 2000*

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**Treating Intelligibility Disorders**

- **Speaker:**
  - Respiration
  - Phonation
  - Resonance
  - Articulation
- **Listener:**
- **Intelligibility**

**Speaker-based approaches to remediation**

- Subsystem Approach
  - Respiration
  - Phonation
  - Resonance
  - Articulation
- Global Approaches
Respiration
- Modify posture, tone, strength
- Improve control of exhalation
- Use biofeedback or visual feedback
- Modify abnormal breathing pattern
- Compensation (breath groups)

Phonation
- Hyperadduction: relax
- Hypoadduction: pushing/pulling, maximum performance tasks
- Traditional voice therapy

Resonance
- Prosthetic device/surgery
- Tactile/kinesthetic feedback
- Visual feedback
- Contrast drills
- CPAP

Articulation
- Articulation drills
- Contrast drills
- Oral strengthening
- Range-of-motion exercises
Modification of speech signal

- Reducing speaking rate
- Speaking louder
  - Is reduced loudness the problem?
- Modifying prosody
  - Reduced pitch loudness and variation? Monopitch and monoloud?

Typical Approaches to Characterizing Intelligibility

- Single-word intelligibility tests
- Sentence intelligibility tests
- Estimates from reading passages or spontaneous connected speech

The estimates of single-word intelligibility are generally poor predictors of connected speech intelligibility, except at very mild and very severe ends of the continuum.

Why is this the case?

- Listeners don’t listen for phonemes in connected speech. They listen for words or even phrases.
- Listeners apply their knowledge of syntax, semantics, and phonotactics, and knowledge of the speaker to problem-solve degraded stretches of speech.

Treating Motor speech disorders
How can we use a theoretical framework to guide clinical practice?

Face-to-face communication
- Face the listener
- Get the listener's attention before speaking
- Audio-visual enhancement for comprehensibility

Transmission:
Minimize environmental interference
- Optimize the environment
- Reduce background noise (the dishwasher, too; not just the tv!)
- Avoid communication over a distance

Activate optimal lexical pool:
Alphabet Supplementation
- Use an alphabet supplementation board, pointing to the first letter of each word as it is spoken.
• What if hypernasality is interfering with activation of the target lexical item?

Syllable segmentation
• Slow speaking rate
• Pausing between words but without separating syllables within words

Influence of familiarity
• Measures of speech intelligibility are subject to bias.
• Are speakers improving, or are you becoming a better listener?

Is PPAOS a special case?
Ways to quantify change to overcome bias

• Rate
• Segmentation
• Loudness

• Digital advances

Cases? Questions? Comments?