Management of progressive speech and language disorders
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Overview
• Define primary progressive aphasia (PPA) and describe its variants
• Define primary progressive apraxia of speech (PPAOS)
• Describe the management principles for PPA and PPAOS
• Present case study – patient with PPAOS and aphasia

Primary Progressive Aphasia
• Aphasia of insidious onset
• Gradual progression of word finding, object-naming, or word-comprehension impairments
• Limitations in ADLs are attributed to language impairment at initial stages of disease and at time of testing
• Intact premorbid language function

Presentation
• Months, year, or years after onset
• First symptom is most commonly word finding deficit
• Individuals are aware of their deficits.
• Usually associate onset with a specific situation that required higher level linguistic demands

Disclosures
• No disclosures to report
PPA Subtypes

- PPA semantic variant
- PPA non-fluent/agrammatic variant
- PPA logopenic variant
Case BS

- Past Medical History: Per patient and wife, following urologic surgery in 4 years ago, the patient began to experience difficulty with self-expression.
- Patient reported this issue "was getting worse all the time."
- Spouse stated that the patient was beginning to isolate himself socially, although she was trying to prevent this.
- Patient stated: "I think right. I cannot articulate speech. Getting worse all the time."

Speech Sample

- Speech analysis was conducted
- Average word per minute = 43.1
- Average % CIUs = 83.7
- CIUs per Minute = 36
Case KR

- Noticed difficulty with word finding during conversations at home and work 3 years earlier.
- Word finding deficits have worsened. He understands most of what he hears in conversation, but sometimes has to ask for repetition.
- His wife reported that, recently, he has experienced some changes in memory function. He feels that it takes longer for him to get things done.

Nonfluent vs. Logopenic

- When agrammatism is not evident...
- Need to distinguish motor programming and planning errors from linguistic errors

Apraxia of speech vs. Phonemic paraphasia
• 21 patients with logopenic variant (lvPPA)
• 20 patients with nonfluent variant (nfvPPA)
• 17 age-matched controls

- Syllable segregation
  - Proportion of silence time
  - Median duration of silences
  - Variability of silence duration
- Lexical stress
  - Vowel duration
  - Peak intensity

Results

• Periods of silence during reading and relative vowel duration and intensity in polysyllable word repetition was associated with nfvPPA
• Relative vowel duration differentiated nfvPPA cases from both control and lvPPA cases with 88% agreement with expert judgment of presence of apraxia of speech in nfvPPA cases

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Primary Progressive AOS

• Apraxia of speech of insidious onset
• Gradual progression
• Absence of non-language cognitive impairments and aphasia for substantial period of time
• Resulting from a degenerative condition involving the left hemisphere

PPAOS With Aphasia

• Progressive AOS and aphasia may coexist
• If so, the most prominent feature is labeled first.

Primary Perceptual Characteristics

• Slow rate
• Distorted substitutions
• Syllable segmentation/excess & equal stress
• Poorly sequenced sequential motion rates
• Increased off-target errors with longer utterances
• Sound sequencing errors
• Articulatory groping
• Distorted additions
In ~ 20% of cases of nfv-PPA, AOS was the primary and sometimes only deficit.

Variability of vowel duration was useful in discriminating progressive AOS (with nfv-PPA) from control subjects and individuals with the lv-PPA.

Possibly associated with different neuro imaging loci and pathological diagnosis.

Duffy et al. (2015)

Factors Influencing Management

- Disease progression
  - Time from initial diagnosis
  - Severity of impairment
- Onset of additional disorders

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General PPA Treatment Guidelines

- Early speech-language-cognitive evaluation and follow-ups to determine pattern of decline
- Early treatment may focus on impaired language functions in addition to compensations.
- AAC should be introduced early for a more successful transition when need arises.
- Family members/caregivers must be involved.
- Treatment will not reverse progression, but may enhance communication.

Thompson (1997)

Application to PPAOS

- It is unknown if management principles apply to PPAOS. (Duffy, 2006)
- Alternative and augmentative communication may be more beneficial for individuals with PPAOS than PPA. (McNeil & Duffy, 2008)
Semantic Variant

Treatments
- Functional interventions
  - Narrative/conversational discourse
  - Participation in functional activities
- Impairment-based interventions
  - Picture naming skills
  - Lexical retrieval
  - Face-name associations
  - Object use

Outcomes
- Relearn target vocabulary
- Maintain gains above baseline levels—variable
- Generalization to untrained stimuli—limited
  - Personalized materials
  - Functional context

Carthey-Goulart et al. (2013)

Non-fluent Variant

Treatments
- Functional interventions
  - Augmentative and alternative communication devices
- Impairment-based interventions
  - Agrammatism
  - Phonological skills
  - Naming/lexical retrieval
  - Apraxia of speech

Outcomes
- Improved production of treatment targets
- Generalization to untreated items and/or different tasks
- Improved functional communication

Carthey-Goulart et al. (2013)

Logopenic Variant

Treatments
- No functional interventions have been reported
- Impairment-based interventions
  - Naming/lexical retrieval
  - Spelling

Outcome
- Improvement on trained items
- Generalization to untrained items and conversational skills

Carthey-Goulart et al. (2013)
Positive Effects of Language Treatment for the Logopenic Variant of Primary Progressive Aphasia

Nguyen M. Boren - Rachel M. King - Hanae Behbodipoor - Maya L. Henry - Hyunsoo Cho - Nevin Z. Ropack

- 77 year old man, 2.5 years post onset diagnosis of PPA-logopenic variant
- Treatment included generative naming tasks and semantic elaboration training

Training Approaches

In general...
- Errorless learning more effective than errorful learning
- Longer duration > shorter duration in maintenance of treatment gains
- Utilize stimuli that are familiar and high frequency

Across PPA Types

- Majority of studies provide Level III evidence
- Majority of studies targeted naming at level of impairment
- Few studies addressed functional communication
- All studies demonstrated positive outcomes for trained items

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Case Study

- Patient biographical and medical data
  - 64-year-old, right handed male
  - Employed as a Pharmacist
  - 4-year decline in communication function
  - Early, incorrect diagnosis of Alzheimer’s Disease
  - Later, diagnosed with PPAOS
  - MRI revealed minimal ischemic white matter changes in left periventricular regions

Case Study

- Patient and family report:
  - Initial difficulty “getting his words out”
  - Four year decline in communication function
  - Difficulty with initiating speech and word retrieval
  - Denied auditory comprehension and memory deficits
  - Primary concern was increased difficulty communicating with customers and pharmacy techs at work.
Speech and Language Testing

- Motor Speech Evaluation
- Revised Token Test (RTT)
- Story Retell—Arizona Battery for Communication Disorders of Dementia (ABCD)
- Picture description task—"cookie theft" picture
- Pyramid and Palm Trees Test (PPT)
- Boston Naming Test (BNT-Short form)
- Word fluency measure

Motor Speech Evaluation

- Perceptual signs
  - Slow rate
  - Inconsistent articulatory errors
  - Distorted substitutions
  - Sound repetitions
  - Voicing errors
  - Excess and equal stress
- Diagnosis—moderately severe AOS

Language Testing

- Revised Token Test
  - Overall score: 13.35 (80th %ile)
  - Primarily immediacy and self-correction errors
- Narrative discourse
  - 98 CIUs
  - 89 %CIUs
  - 29.4 CIUs/minute*
  - 53.4 %CIUs/minute*

*Consistent with aphasic performance—Nicholas & Brookshire (1993)

Language Testing

- Arizona Battery for Communication Disorders in Dementia—Story Retell
  - Immediate Recall 15/17
  - Delayed Recall 15/17

Story
While a lady was doing shopping, her wallet fell out of her purse, but she did not see it fall. When she got to the checkout counter, she had no way to pay for her groceries. So she put the groceries away and went home. Just as she opened the door to her house, the phone rang and a little girl told her that she had found her wallet. The lady was very relieved.

Patient read
The woman was doing ... shopping... uh... the wallet dropped... out of her purse... she went to the checkout counter... and oh she didn't have the wallet with her... and she put the groceries back... and... she left the store... and she went home... and the phone rang... and the little girl told her... the woman... she had found her purse... her wallet... and she was relieved.

Language Testing

- Pyramids & Palm Trees 52/52
- Boston Naming Test-SF 13/15
- Word Fluency—(f, a, s)
  - Oral naming 17 words
  - Written naming 19 words

Treatment Planning

- Educate patient and family
- Explore potential AAC options
- Implement speech/language treatment approach targeting patient’s impairments and functional needs.
- Consider patient’s situation
  - Over 1 hour drive to clinic
  - Only able to attend 1x every two weeks
Script Training

- Purpose—to develop an individual with aphasia’s ability to produce automatic, natural language
- Identify personally relevant and functional scripts
- Utilizes principles of motor learning
  - repetitive, cue-based, drill practice
  - train whole scripts in natural, conversational contexts

Script Training

- Therapeutic effect of script training reported for individuals with
  - Nonfluent aphasia without AOS (Youmans et al., 2005)
  - Fluent aphasia (Cherney et al., 2008)
  - Aphasia and apraxia of speech (Youmans et al., 2011)
  - Primary progressive aphasia (Holland et al., 2002)

Treatment

- Seven scripts were developed
- Modified training protocol (Youmans et al., 2005)
  - Reading
  - Repetition
  - Choral reading
  - Immediate spontaneous production
  - Delayed spontaneous production
- Daily home practice of all trained scripts
- Daily practice in functional settings

Example Script

P: Hi, have you been waited on?
C: No, I haven’t.
P: How can I help you?
C: I have a prescription to fill.
P: Do you have the ‘script number?
C: Yes, here it is.
P: This medication is out of refills. I’ll fax your doctor.
C: How long will that take?
P: It depends on when the doctor gets it.

Scoring

- Scripts were scored on-line
- Each response in a script was rated individually
- Accurate if:
  - Initiation of response was timely
  - Response included all words from script
  - Word production was accurate with exception of mild distortion or prosodic disturbance

Results

Session #

0 20 40 60 80 100

0 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15

% correct

Script 1 ‘customers’

Script 2 ‘customers’

Script 3 ‘customers’
Participant’s communication became more automatic,

Holland, A., Carthery. Computerized Eventually retired from his profession

Josephs, K. A., Duffy, J. R., Strand, E. A., Youmans. Data suggest script training was also successful for an

Beeson, N., Thera, N., Bier, N., Kertesz

Repetitive practice and articulatory modeling improved

Ballard, K. J., Savage, S., Leyton, C. E., Vogel, A. P., Hornberger, M. Therapeutic effect of script training for individuals with


Beeson, N., Bier, N., Kertesz


References

Results

Conclusions

Thanks for your attention.

Thank you

Long Term Outcome

- Patient’s performance continued to decline
- Eventually retired from his profession
- Evaluated for an AAC system
- Developed physical and behavioral deficits with symptoms consistent with a possible corticobasal degeneration and frontotemporal dementia-behavioral variant

Data suggest script training was also successful for an individual with PPAOS and aphasia.

Participant’s communication became more automatic, fluent, and communicative.

Repetitive practice and articulatory modeling improved production and fluency of trained stimuli.

Functional context of trained scripts facilitated generalization to functional interactions.

Conclusions

- Therapeutic effect of script training for individuals with aphasia has already been demonstrated
- Data suggest script training was also successful for an individual with PPAOS and aphasia.
- Participant’s communication became more automatic, fluent, and communicative.
- Repetitive practice and articulatory modeling improved production and fluency of trained stimuli.
- Functional context of trained scripts facilitated generalization to functional interactions.

Thank you