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Management of progressive speech and language disorders: An update

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Pi Beta Phi Rehabilitation Institute
Vanderbilt Bill Wilkerson Center

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Learning Objectives

After this course, participants will be able to:

- List the three subtypes of PPA.
- Describe the distinguishing characteristics of each subtype.
- Describe the management principles for PPA.
- Describe three treatment approaches for PPA.

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Overview

- Define primary progressive aphasia (PPA) and describe its variants
- Describe the management principles for PPA
- Describe specific treatment approaches for the variants of PPA

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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

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Gorno-Tempini et al. (2011)

Primary Progressive Aphasia

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

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Gorno-Tempini et al. (2011)

Typical 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
- Type of aphasia will vary

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PPA Subtypes

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

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PPA-Nonfluent/Agrammatic

I. Clinical diagnosis of nonfluent/agrammatic variant PPA

At least one of the following core features must be present:

1. Agrammatism in language production
2. Effortful, halting speech with inconsistent speech sound errors and distortions (apraxia of speech)

At least 2 of 3 of the following other features must be present:

1. Impaired comprehension of syntactically complex sentences
2. Spared single-word comprehension
3. Spared object knowledge

II. Imaging-supported nonfluent/agrammatic variant diagnosis

Both of the following criteria must be present:

1. Clinical diagnosis of nonfluent/agrammatic variant PPA
2. Imaging must show one or more of the following results:
   a. Predominant left posterior fronto-insular atrophy on MRI or
   b. Predominant left posterior fronto-insular hypoperfusion or hypometabolism on SPECT or PET

III. Nonfluent/agrammatic variant PPA with definite pathology

Clinical diagnosis (criterion 1 below) and either criterion 2 or 3 must be present:

1. Clinical diagnosis of nonfluent/agrammatic variant PPA
2. Histopathologic evidence of a specific neurodegenerative pathology (e.g., FTLD-tau, FTLD-TDP, AD, other)
3. Presence of a known pathogenic mutation

Abbreviations: AD = Alzheimer disease; FTLD = frontotemporal lobar degeneration; PPA = primary progressive aphasia.

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Gorno-Tempini et al. (2011)

PPA Semantic Variant

I. Clinical diagnosis of semantic variant PPA

Both of the following core features must be present:

1. Impaired confrontation naming
2. Impaired single-word comprehension

At least 3 of the following other diagnostic features must be present:

1. Impaired object knowledge, particularly for low-frequency or low-familiarity items
2. Surface dyslexia or dysgraphia
3. Spared repetition
4. Spared speech production (grammar and motor speech)

II. Imaging-supported semantic variant PPA diagnosis

Both of the following criteria must be present:

1. Clinical diagnosis of semantic variant PPA
2. Imaging must show one or more of the following results:
   a. Predominant anterior temporal lobe atrophy
   b. Predominant anterior temporal hypoperfusion or hypometabolism on SPECT or PET

III. Semantic variant PPA with definite pathology

Clinical diagnosis (criterion 1 below) and either criterion 2 or 3 must be present:

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Gorno-Tempini et al. (2011)

PPA Logopenic Variant

I. Clinical diagnosis of logopenic variant PPA

Both of the following core features must be present:

1. Impaired single-word retrieval in spontaneous speech and naming
2. Impaired repetition of sentences and phrases

At least 3 of the following other features must be present:

1. Speech (phonologic) errors in spontaneous speech and naming
2. Spared single-word comprehension and object knowledge
3. Spared motor speech
4. Absence of frank agrammatism

II. Imaging-supported logopenic variant diagnosis

Both criteria must be present:

1. Clinical diagnosis of logopenic variant PPA
2. Imaging must show at least one of the following results:
   a. Predominant left posterior perisylvian or parietal atrophy on MRI
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III. Logopenic variant PPA with definite pathology

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Gorno-Tempini et al. (2011)

Overview

- Define primary progressive aphasia (PPA) and describe its variants
- **Describe the management principles for PPA**
- Describe specific treatment approaches for the variants of PPA

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CONTINUED™

Factors Influencing Management

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

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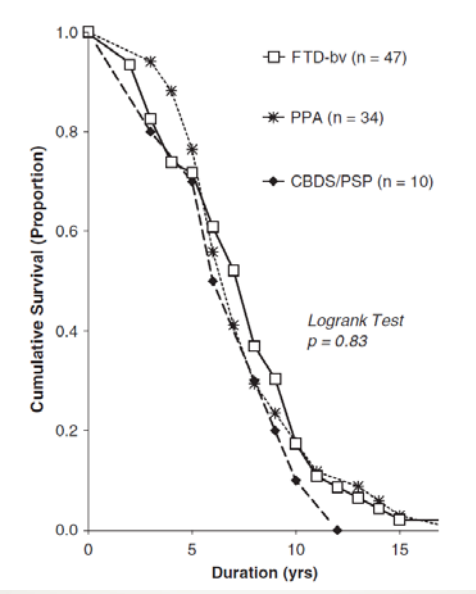
Survival curves from symptom onset according to first clinical syndrome



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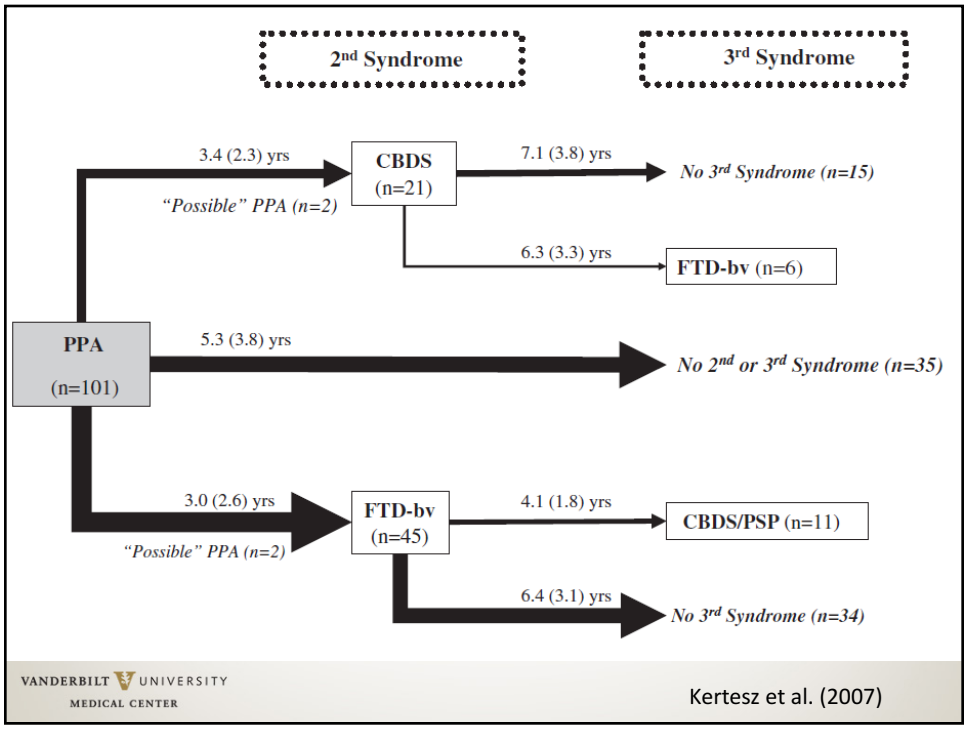
Kertesz et al. (2007)



Kertesz et al. (2007)

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.

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Thompson (1997)

Overview

- Define primary progressive aphasia (PPA) and describe its variants
- Describe the management principles for PPA
- **Describe specific treatment approaches for the variants of PPA**

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Specific Treatment Techniques?

- Growing number of published reports of specific treatment approaches applied to PPA
- One published report of treatment outcomes for an individual with PPA and AOS

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Dement Neuropsychol 2013 March;7(1):122-131

Original Article

Nonpharmacological interventions for cognitive impairments following primary progressive aphasia

A systematic review of the literature

Maria Teresa Carthery-Goulart[1], Amanda da Costa da Silveira[2], Thais Helena Machado[3], Leticia Lessa Mansur[4], Maria Alice de Mattos Pimenta Parente[2], Mirna Lie Hosogi Senaha[5], Sonia Maria Dozzi Brucki[6], Ricardo Nitrini[6]

39 articles met criteria (67 patients treated)

- Semantic variant (20 studies)
- Nonfluent/Agrammatic (8 studies)
- Logopenic variant (3 studies)
- General PPA diagnosis (8 studies)

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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

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Carthery-Goulart et al. (2013)

APHASIOLOGY, 2009, 23 (2), 210–235

Psychology Press
Taylor & Francis Group

Known, lost, and recovered: Efficacy of formal-semantic therapy and spaced retrieval method in a case of semantic dementia

Nathalie Bier
Université de Sherbrooke, Canada
Joël Macoir
Université Laval, Canada
Lise Gagnon
Université de Sherbrooke, Canada
Martial Van der Linden
University of Geneva, Switzerland
Stéphanie Louveaux
Research Center on Aging, CSSS-Sherbrooke Geriatric University Institute, Canada
Johanne Desrosiers
Université de Sherbrooke, Canada

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Purpose

1. Explore the effect of a formal-semantic therapy in relearning concepts
2. Compare the addition of the spaced retrieval to the use of a simple repetition
3. Explore the long-term maintenance treatment effects in both conditions
4. Explore possible effects of generalization within trained categories and between trained and non-trained categories

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Bier et al. (2009)

Participant

- 70 year-old woman
- Native French speaker
- 5-year history of progressive decline in word-finding and memory
- Mild to moderate cortical atrophy around the left sylvian fissure
- Left frontal hypoperfusion

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Bier et al. (2009)

Test Results and Diagnosis

- Impairment on all measures requiring semantic processing
- Normal performance on tests of visuo-perceptual abilities and episodic memory
- Testing results consistent with Semantic Dementia

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Bier et al. (2009)

Treatment

- Formal-semantic therapy and spaced-retrieval method:
  - Picture was presented along with its spoken name, specific attribute and written name of the category twice;
  - Pictures presented again with **increasing time-recall intervals** and patient was asked to name them and generate semantic attributes
  - Semantic feedback and cueing technique was used when patient was unable to name items.

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Bier et al. (2009)

Treatment

- Formal-semantic therapy with simple repetition:
  - Picture was presented along with its spoken name, specific attribute and written name of the category twice;
  - Pictures presented at **regular intervals** and patient was asked to name them and generate semantic attributes
  - Semantic feedback and cueing technique was used when patient was unable to name items.

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Bier et al. (2009)

Results

- Formal-semantic therapy led to better naming and generation of specific verbal attributes compared to baseline
- Spaced retrieval was not statistically superior to the simple repetition condition
- Gains maintained up to 5 weeks after the end of the intervention for both spaced retrieval and simple repetition

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Bier et al. (2009)

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

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Carthery-Goulart et al. (2013)

Behavioural Neurology 26 (2013) 77–88
DOI 10.3233/BEN-2012-120260
IOS Press

Treatment for apraxia of speech in nonfluent variant primary progressive aphasia

M.L. Henry[a,*], M.V. Meese[b], S. Truong[c], M.C. Babiak[a], B.L. Miller[a] and M.L. Gorno-Tempini[a]
[a]*Memory and Aging Center, Department of Neurology, University of California, San Francisco, CA, USA*
[b]*Alta Bates Medical Center, El Cerrito, CA, USA*
[c]*San Francisco State University, San Francisco, CA, USA*

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Purpose

1. To determine whether structured oral reading practice can improve production of multisyllabic words read aloud
2. To examine generalization to untrained speech behaviors and contexts
3. To assess long-term effects of training on oral reading and speech production

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Henry et al. (2013)

Participant

- 73 year-old right-handed woman
- English as her primary language
- Five-year history of progressive impairment of speech production
- Frontal/insular and temporoparietal atrophy in the left hemisphere.

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Henry et al. (2013)

Test Results and Diagnosis

- Language evaluation
  - Relatively fluent, grammatical spoken language
  - Semantic processing relatively spared
- Motor Speech Evaluation
  - slow rate; vowel and consonant distortions
  - sound insertions, deletions, and transpositions
  - pauses between words and syllables
  - increasing errors with increasing word length
  - reduced prosodic variation
- Testing consistent with nonfluent variant-PPA

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Henry et al. (2013)

Treatment

- Structured oral reading of text focusing on rehearsal of multisyllabic word production in sentence context
- Trained self-detection and correction of speech errors while reading text aloud
- Weekly treatment sessions and home practice

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Henry et al. (2013)

Treatment

- Text was read aloud until patient produced a word incorrectly
- For multisyllabic words, lines were drawn dividing the word into constituent syllables
- Word produced syllable-by-syllable
- Once produced correctly, whole word was repeated in a non-syllabified manner

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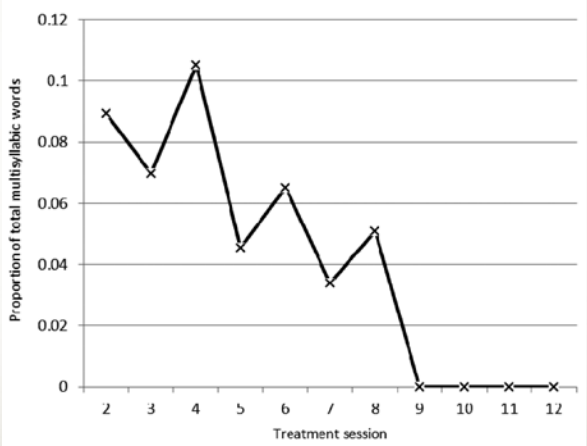
Henry et al. (2013)

Results

- Multisyllabic word production in **trained text** improved during treatment



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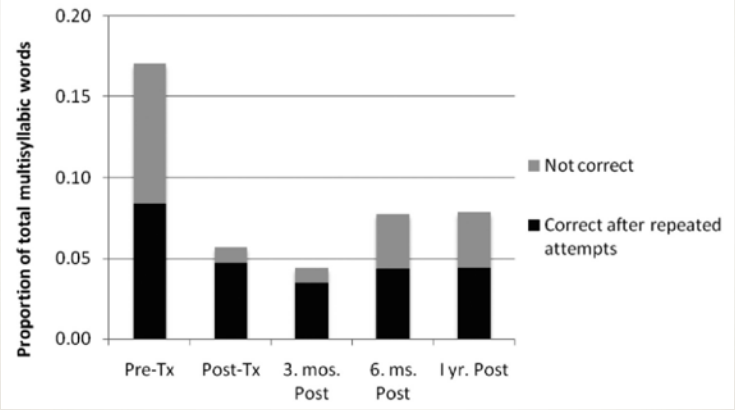
Henry et al. (2013)

Results

- Significant reduction in speech errors on single and multisyllabic words in **untrained text** at post treatment, 3 months, 6 months, and one year



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Henry et al. (2013)

Results

- One year post-treatment—significant increase in the proportion of multisyllabic words produced with at least one minor distortion
- Reading of novel text showed a gradual slowing over time

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Henry et al. (2013)

Results

- Patient perceived improved performance and comfort level in fluency, multisyllabic word production, sentence production, and stress assignment
- Reported improved confidence and reduced frustration in communication with both familiar and unfamiliar people

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Henry 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

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Carthery-Goulart et al. (2013)

J Mol Neurosci (2011) 45:724–736
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Positive Effects of Language Treatment for the Logopenic Variant of Primary Progressive Aphasia

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Participant

- 77 year old man
- Retired accountant
- 2.5 years post onset diagnosis of PPA-logopenic variant

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Beeson et al. (2011)

Test Results and Diagnosis

- Fluent spontaneous speech with normal syntactic structure
- Auditory comprehension—relative strength
- Repetition of sentences—mildly impaired
- Word finding—significant impairment
- Motor speech production—intact
- Primary Progressive Aphasia-Logopenic Variant

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Beeson et al. (2011)

Treatment

- Generative naming tasks were used to probe and to train lexical retrieval.
- 6 categories were trained (three living and three nonliving), and 6 semantically matched categories were probed but not trained
- Treatment included 2-hour treatment sessions, 6 days per week for 2 weeks, plus 1 h of daily homework.

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Beeson et al. (2011)

Treatment

- Each category trained over 2 days
- Day 1
  - Presentation of 30 colored photographs of example items with written labels
  - Practice naming pictures without labels
  - Elaboration of semantic features
- Day 2
  - Semantic feature analysis – attributes, functional use, context, similar items, superordinate category

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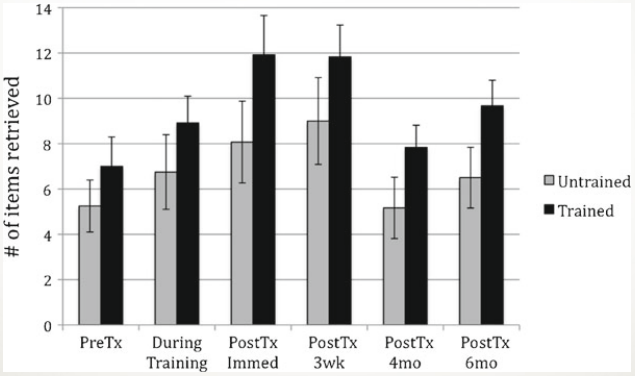
Results

- Improved ability to retrieve exemplars for the trained categories and untrained categories



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Beeson et al. (2011)

Results

- Patient perception
  - Overall confidence level - “a lot better”
  - Ability to name things – “better”
  - Overall speaking ability – “better”
  - Stress level during conversation – “better”
  - Ability to think of people’s names and come up with words in conversation – “somewhat better”

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Beeson et al. (2011)

Results

- Post-treatment fMRI activation suggested improvements were supported by increased reliance on left prefrontal cortex during word retrieval

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Beeson et al. (2011)

Final Thoughts

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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

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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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Carthery-Goulart et al. (2013)