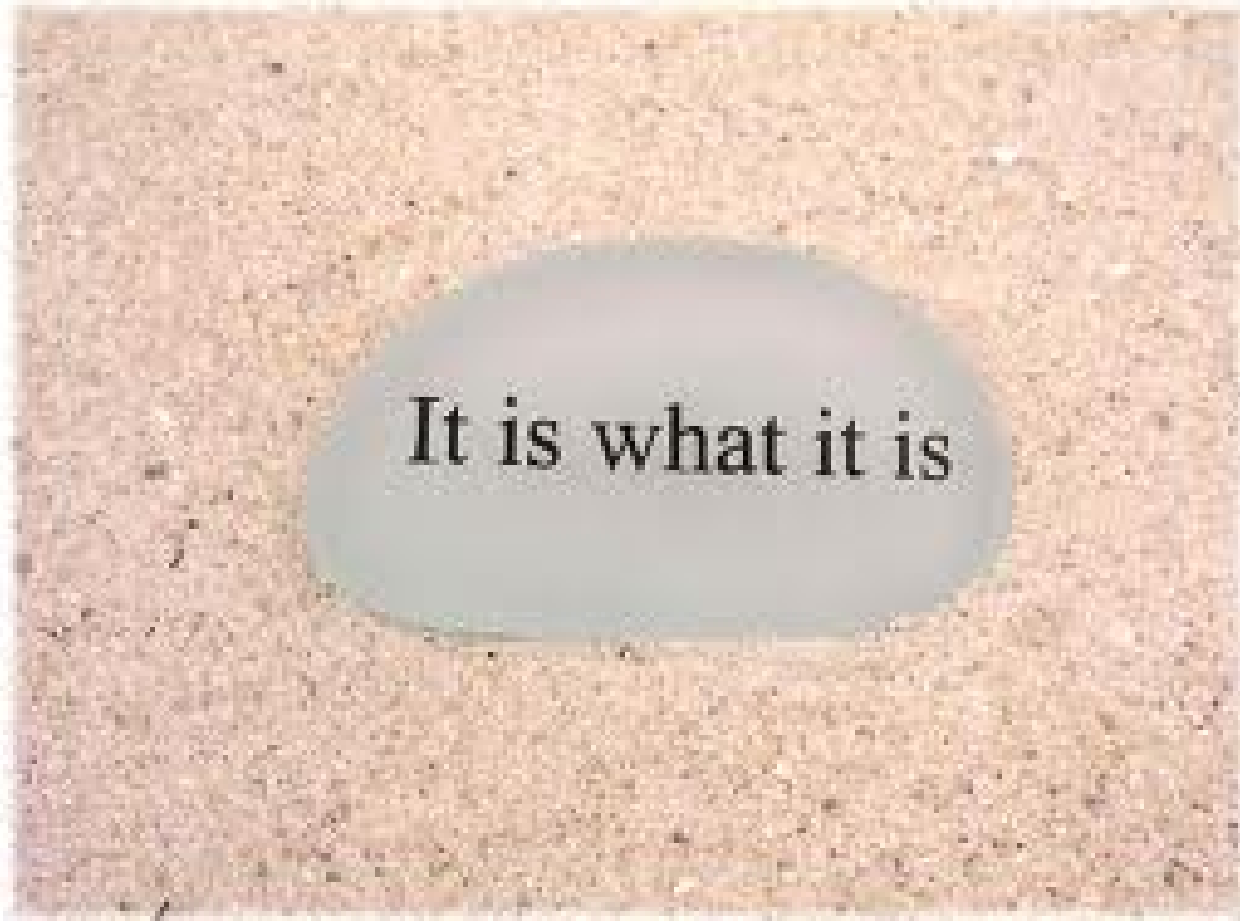


# Taking first steps: towards a speech pathology management pathway for primary progressive aphasia.

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# Primary Progressive Aphasia PPA



Progressive  
non-fluent  
variant PPA



Logopenic  
variant PPA

Semantic  
Variant PPA



# Treatment

- Is Tx indicated in neurodegenerative conditions?
- What is a Tx gain in a progressive condition?
- How do Tx goals and approaches change with disease progression?

*“The presenting speech and language complaints can be strikingly similar to those of people with stroke- induced aphasia.”*

( McNeil & Duffy, 2001, p.245)

- ▶ Retention of reasoning and cognitive flexibility in early stage (*Shany-Ur et al 2011*)
- ▶ No significant socio emotional deficits in PPA and LPA (*Wicklund et al, 2004*)
- ▶ PPA patients have insight into their symptoms, especially in the earlier stages of the disease, (*Banks & Weintraub 2008*).
- ▶ Ability to learn in early stages (*Cartwright & Elliott 2009*)

What is the speech pathology  
management pathway for PPA in NSW?

# Speech pathology services in NSW, Australia

## **Taylor & Miles-Kingma survey (Taylor et al., 2009)**

- (To our knowledge) the only survey of service provision for PPA
- Prompted by an increase in referrals to their service in Sydney
- 34 speech pathology services with an adult neurological caseload in rural and metropolitan regions of NSW asked about referrals between June 2004 and July 2005
- 77% reply rate (26 surveys returned)



# Speech pathology services in NSW, Australia

## Taylor & Miles-Kingma results

- 50% respondent services received **NO** referrals
- Range of services provided.
- Emerging area of practice
- Need more information about PPA for clinicians and health care consumers
- Need to promote referrals and role of speech pathologist for this client group & families
- **Need to develop management pathway**



International Frontotemporal Dementia Conference  
 Northwestern University Cognitive Neurology and Alzheimers  
 Disease Center (CNADC) Chicago  
 Massachusetts General Hospital, Boston  
 University of California, Memory & Ageing Center, San  
 Francisco



# Clinical assessment of progressive aphasia

Lyndsey Nickels, Cathleen Taylor, and Karen Croot

There is an increasing awareness that language impairments can be the most prominent initial symptom of people with a number of neurodegenerative disorders. Consequently, speech pathologists are increasingly required to apply their skills to the communication needs of this group. While the literature addressing the nature and treatment of the language impairments of individuals with progressive aphasia is growing, little guidance is available regarding assessment. In this paper we review the assessment requirements of this population, arguing that assessment needs to a) identify the current status of the person's language impairment and the impact it has on their communication activities, participation, and quality of life, to enable goal-planning for treatment, and b) establish the nature, extent, and rate of change in language skills over time. We argue that, while many factors influencing choice of assessment are similar to those for people with non-progressive aphasia, important factors that are particular to people with progressive aphasia need to be considered.

In the past, the primary role for speech pathologists in the area of acquired neurogenic language impairments was restricted to the assessment and treatment of language impairments resulting from stroke, traumatic brain injury and, less often, from tumour, infection, and surgery. Occasionally the speech pathologist might have been called on to perform a differential diagnosis between aphasia and dementia, or to facilitate communication in people with dementia using techniques such as reality orientation or reminiscence therapy (e.g., Baines, Saxby & Ehrlert, 1987; Spector, Davies, Woods & Orrel, 2000). However, more recently there has been a realisation that deterioration in language processing can be the most prominent initial symptom in a number of neurodegenerative diseases. The resulting syndrome is primary progressive aphasia,

which typically occurs in one of three behaviourally defined variants (Gorno-Tempini et al., 2011): semantic dementia (Snowden, Goulding, & Neary, 1989), nonfluent progressive aphasia (Gorno-Tempini et al., 2004), and logopenic progressive aphasia (Gorno-Tempini et al., 2004, 2006; Mesulam et al., 2008). Over the last thirty years or so, a wide range of other syndrome labels have also been applied to individuals with progressive language impairments, including pure progressive anomia, primary progressive conduction aphasia, primary progressive apraxia of speech, language- or temporal-variant frontotemporal dementia (see Croot, 2009, for more details). Many people with these syndromes are found on post-mortem investigation to have frontotemporal lobar degeneration neuropathology, but others have Alzheimer disease pathology (Gorno-Tempini et al., 2011).

There is currently a growing recognition of, and evidence for, the role of the speech pathologist in the treatment of individuals with communication disorders associated with dementia (e.g., Royal College of Speech and Language Therapists [RCSLT], 2005a, 2005b). In the Australian context, Taylor, Mies-Kingma, Croot, and Nickels (2009) surveyed speech pathology service provision for people with primary progressive aphasia in New South Wales. The survey gave a clear picture that speech pathologists viewed progressive aphasia as an emerging field of practice and revealed that when clients were referred, all centres provided assessment services. However, the survey responses also indicated that speech pathologists lacked confidence in the appropriate service provision for this population. While literature is emerging on treatment for progressive language impairments (e.g., Croot, Taylor & Nickels, 2011; Nickels & Croot, 2009), little guidance is available for the speech pathologist regarding assessment of progressive language impairments. This article aims to address this issue.

As McNell and Duffy (2001, p. 475) note, the speech and language symptoms in people with progressive language impairments "can be strikingly similar to those of people with stroke-induced aphasia". They argue that decisions about treatment in progressive language impairments can thus be based on the same philosophical, clinical, theoretical and practical considerations that apply in stroke-related aphasia. Thus, the approach to assessment of progressive language impairments should also be similar to the assessment of non-progressive language impairment. However, because of the different long-term prognosis

## KEYWORDS

ASSESSMENT

PRIMARY  
PROGRESSIVE  
APHASIA

PROGRESSIVE  
APHASIA

PROGRESSIVE  
LANGUAGE  
IMPAIRMENT

SEMANTIC  
DEMENTIA

## THIS ARTICLE

HAS BEEN  
PEER-  
REVIEWED



Lyndsey Nickels (top), Cathleen Taylor (centre), and Karen Croot

# Professional development

## Assessment of PPA

1. Identify current status of language, speech & communication & impact on communication activities, participation & quality of life to allow goal-planning for treatment
2. Nature & extent of change over time



# What's the evidence?

## Evidence for speech, language, and communication interventions in progressive aphasia

Karen Croot, Cathleen Taylor, and Lyndsey Nickels

### Clinical scenario

You are the manager of the speech pathology department in a large metropolitan hospital with an outpatient rehabilitation service. A local geriatrician refers a 52-year-old man with a diagnosis of progressive aphasia. You see him for initial interview and he reports a gradually worsening problem with talking that he first began to notice about 2 years ago. He has recently decided to take early retirement from his professional life as an architect because of the impact of these speech and language changes. His word finding difficulties and articulatory errors are readily apparent in conversation. He reports anxiety and frustration in speaking situations, and withdraws from communicative situations for fear of making errors. He speaks of wanting to be able to communicate, and is seeking treatment due to the distress and frustration caused by his communication impairment. Apart from his difficulties in speaking, he is in excellent physical health with no reported changes to his behaviour and personality or in his activities of daily living. He keeps up his hobby of kayaking, and has plans to travel with his wife. At the conclusion of the initial interview, you wonder what services you can offer to this client.

### Response to this scenario

Your speech pathology team has previously assessed clients with progressive aphasia, provided education to clients and their families about the speech, language and communication changes that can be seen with the disease, and suggested ways to reduce associated activity limitations and participation restrictions. However, you have seen a small but steady increase in the number of referrals for various progressive language impairments over the last few years, and have been thinking for a while that you would like to develop a management pathway that includes a more systematic approach to intervention. You are not sure what evidence is available to guide your decision-making.

Some health care providers question whether there is a place for interventions with this population, since there is currently no cure for the underlying neuropathological changes that cause progressive aphasia, and because the person's communication and cognitive abilities can only be expected to decline with disease progression. However, you disagree with these views. You agree instead with McNell and Duffy (2001), who advise that since a person with progressive aphasia has impairments similar to those seen in other adult neurogenic populations (including some with neurodegenerative disease), intervention is appropriate, guided by the same general philosophical, clinical, theoretical and practical considerations about treatment that you would apply in other neurogenic populations.

You also reflect that because there is an expectation of decline without treatment, the question about what intervention outcomes to expect in this client group is not straightforward. Although improvement above the level seen at initial assessment is one possible outcome if therapy is effective, it is not the only one. There might also be an outcome of no change (i.e., the client maintains his or her current level of ability), or a slowing of deterioration that allows the individual to continue in desired activities for a longer period of time than would have been possible without the intervention (Rapp & Glucroft, 2009). Education of the significant communication partner may also result in more successful communicative interactions both immediately and in the future.

### Developing an answerable clinical question

To respond to this scenario you first develop an answerable clinical question using guidelines provided on a website promoting evidence-based practice in speech pathology ([http://www.clap.health.nsw.gov.au/specialties/tbbp\\_so\\_patn/resources.htm](http://www.clap.health.nsw.gov.au/specialties/tbbp_so_patn/resources.htm)) and within a previous "What's the evidence?" column (O'Halloran & Rose, 2010). These guidelines suggest you should first define the patient or problem, the intervention, the comparison intervention, and the outcome.

#### Patient or problem

"Primary progressive aphasia" is a broad diagnostic category in the sense that people with this diagnosis can have a diverse range of progressive language difficulties, sometimes accompanied by speech motor impairments (apraxia of speech, various types of dysarthria) and/or by other more or less severe cognitive impairments (e.g., memory problems). You realise that other diagnostic labels are used for people with progressive language impairments, such as semantic dementia, nonfluent progressive aphasia and frontotemporal dementia (and others, see Croot, 2009). To ensure that you pick up all of the relevant studies about intervention in this population you will have to search the scientific literature on a range of syndrome names and combine the results.

#### Intervention

Here you are willing to look broadly at what evidence there is for speech, language and communication interventions across this population, so you do not specify a particular type of intervention.

#### Comparison intervention

This is not relevant at this stage, because you're not yet trying to weigh up the effectiveness of one intervention against another,



Karen Croot (top), Cathleen Taylor (centre) and Lyndsey Nickels

# Professional development

## Evidence-based interventions

- How to do a literature search
- Level 4 evidence for word retrieval interventions (high SCED ratings)
- Limited evidence for other interventions
- Collaborative decision-making with person with PPA and partner: experimental, need for rehearsal
- Regular review



Alzheimer's Australia Research Hazel  
Hawke Research Grant in Dementia Care:  
word relearning study



## Research questions

1. Does treatment that improves word retrieval in aphasia caused by stroke improve word retrieval in progressive aphasia?
2. Does treatment improve word retrieval in connected speech as well as picture naming?

# Hypotheses

- **Treatment effect:** significantly better picture naming for treated items compared to untreated items at post-test than baseline
- **Generalisation to untreated words:** not expected
- **Transfer to connected speech:**
  - treated words occur more often in connected speech at post-test compared with baseline
  - more words produced in the treated than the untreated topic at post-test compared to pre-test

# Treatment

- Half the items in one topic treated (matched on frequency, accuracy at Baselines 1 and 2)
- Untreated words from treated topic and all words from untreated topic serve as controls.
- Repetition and reading in the presence of picture (RRIPP): pictures presented in powerpoint with auditory file of word & word printed below picture. Different pictures treated than in pre- and post-tests
- Approx 15 mins/day, 10 days over 2 weeks

# Outcome measures

## Three baselines and two post-test measures of...

- **Naming:** Photographs of stimulus items sourced for naming, presented with definitions to assist recognition as required.
- **Word retrieval in connected speech:** elicited using a structured interview focusing on each topic, e.g.

Tell me something about your life today. What do you do?

Prompts: What type of things do you enjoy doing with other people?

What do you enjoy doing at home?

Can you tell me more about things you do away from home?



# Participants

- 80 yr old man **PNFA**
- 4 year h/o speech & language deterioration
- 2010 MRI: Frontal and L perisylvian atrophy
- 54 yr old woman **LPA**
- 5 yr h/o speech & language deterioration
- 2010 MRI: dilatation of L lateral ventricle, mild parietal atrophy (particularly L)

## Language symptoms

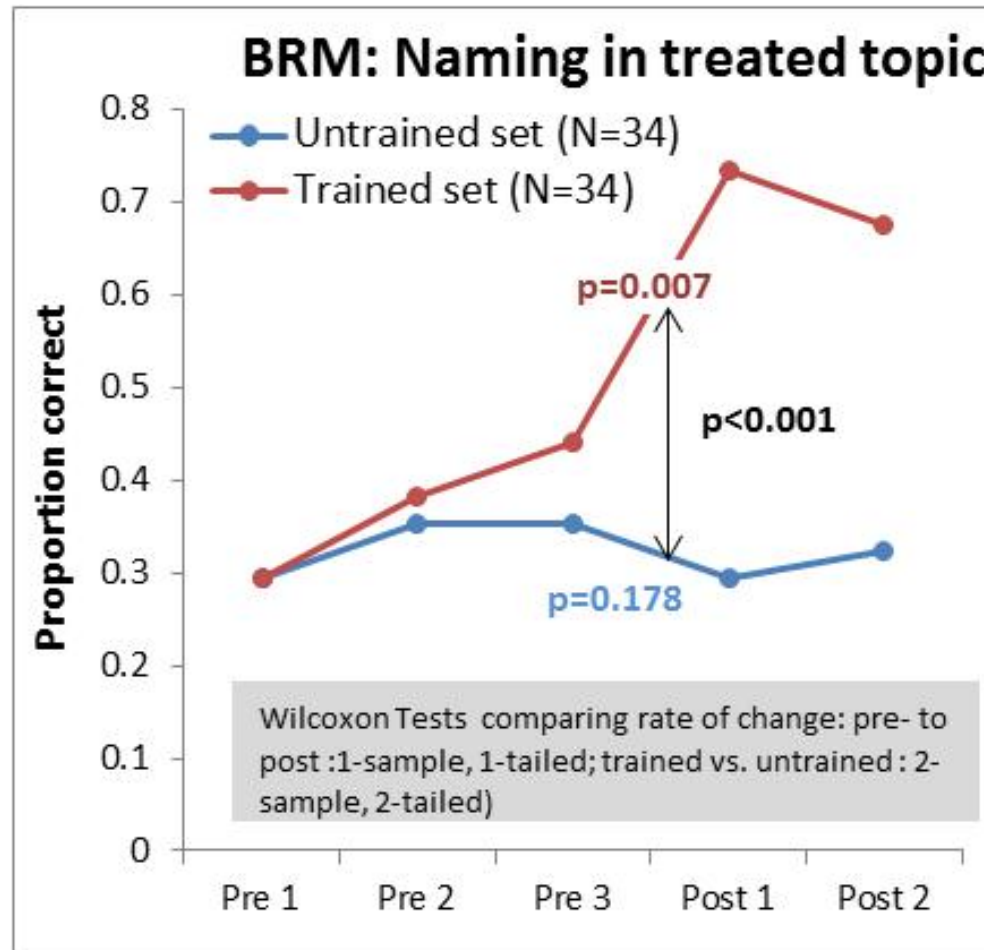
- effortful, halting speech and impaired word retrieval
- Good comprehension; good repetition; poor spontaneous speech.
- Graded Naming Test 18/30
- TROG-2 9 blocks passed

## Language Symptoms

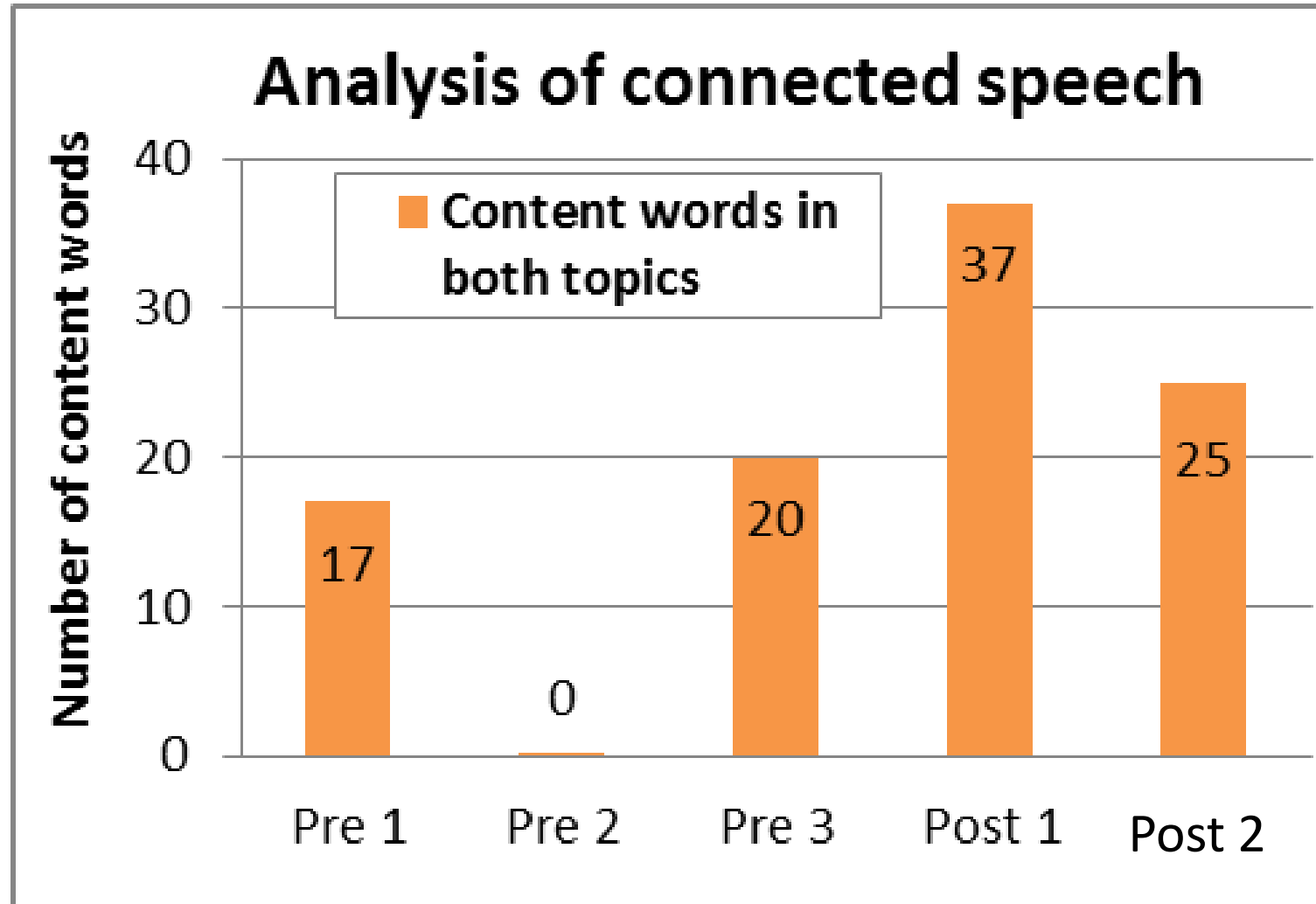
- fluent, grammatical well articulated expressive language with impaired word retrieval
- good comprehension; word repetition; poor spontaneous speech.
- Graded Naming Test 5/30, TROG-2 9 blocks passed



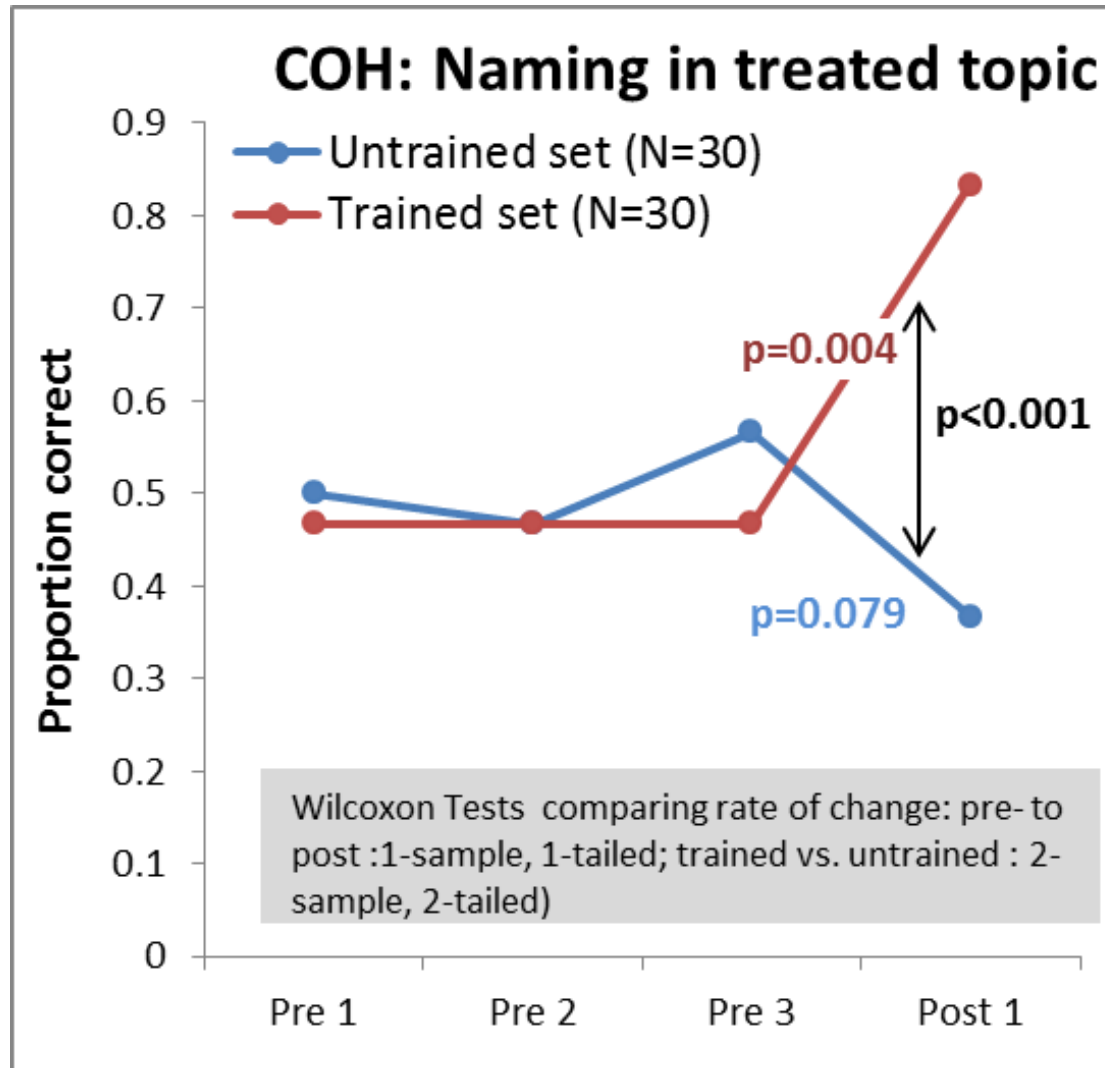
# Results BRM



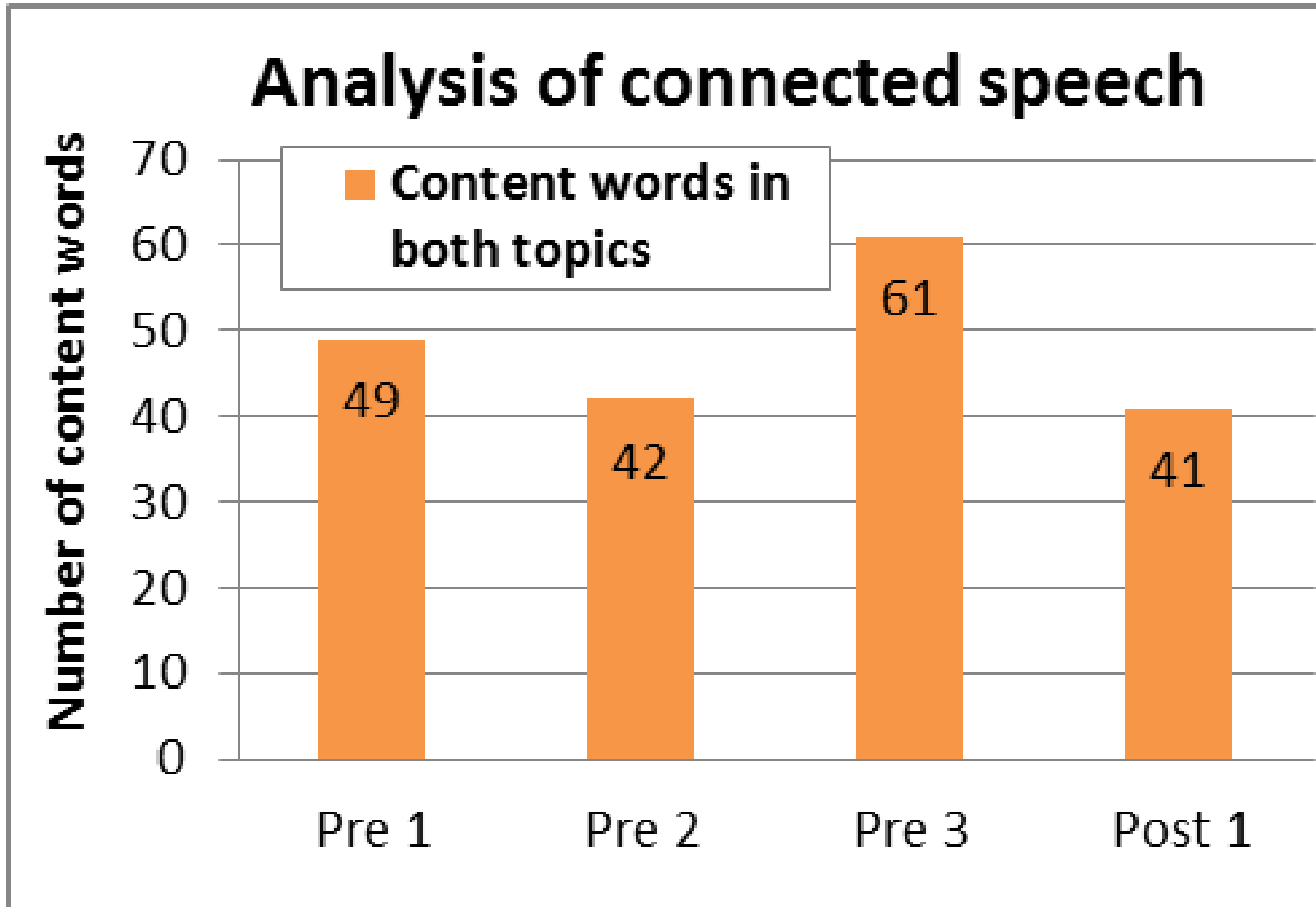
# Results BRM



# Results COH



# Results COH



# Summary of results so far

- BRM & COH significantly improved in naming treated items (as predicted)
- No generalisation to untreated items or topics (as predicted)
- No generalisation into (our measures of) connected speech (not as predicted)
- More to come.

# Other initiatives include

- Training at undergraduate and GEM levels
- Continuing professional development to Speech Pathologists in LHD, greater Sydney and NSW Regions



# Outreach and support



## **Communication and Support group**

- Monthly session held at War Memorial Hospital for individuals with PPA and their families/carers

## **PPAmpulet**

- 2-3x year newsletter for individuals with PPA and their families/carers

# Primary Progressive Aphasia Pathway

- assessment
- intervention
- ongoing review
- education
- support

