

The House of Representative Standing Committee on Health and Ageing
An Inquiry into Dementia: Early Diagnosis and Intervention

By Margaret Pozzebon
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Background:

I am a Speech Pathologist who has worked in the field of adult neurological rehabilitation and aged care for over twenty-five years. In the past ten years I have specialised in the dementia, particularly in the early diagnosis and intervention of language dominant dementia. I am one of the very few speech pathologists who specialised in this area and work within the early diagnostic dementia clinics in Victoria (i.e. CDAMS clinic – Cognitive, Dementia And Memory service). I am also a PhD candidate with La Trobe University, and my research is focusing on spousal caregivers lived experience of living with a partner diagnosed with a language dominant dementia.

I welcome the opportunity to share my views on some aspects of the Terms of Reference of this inquiry, particularly as they relate to the valuable role of speech pathology from early diagnosis through to the end-stage of the disease process. I am representing myself as a clinician in the area of ageing and dementia and will seek to address the following Terms of Reference, specially:

- Early diagnosis and intervention are essential means by which to improve quality of life and maintain independence for people with dementia;
- Increase opportunity for continued social engagement and community participation for people with dementia

Introduction:

Despite the acknowledgement that dementia is devastating for the individual, their family, network of friends and wider community, very little attention or consideration is given to *how* the decline in speech and language skills affects interpersonal relationships and everyday lifestyles. For the most part, research literature and health service provision often focuses on dementia diagnosis and the impact of the degenerative process at the impairment level of functioning. There is minimal focus and follow-up supportive intervention services addressing broader quality of life, interpersonal and social participation aspects of dementia and causes attributed to this - apart from the wider known symptom of memory impairment in Alzheimer's Disease. In order to provide the best possible care and interventions for these individuals with dementia and their families, I believe it is essential for health clinicians to more fully appreciate the activity and participation implications of the linguistically based impairments often accompanying most dementia syndromes throughout the trajectory course of the disease.

Speech pathology has a critical role to play early diagnostic phase, subsequent intervention and supportive counselling in most forms of dementia. I seek to explain these contributions to you. I will also discuss the unique specialist expertise offered by speech pathology in the differential diagnostic process of dementia syndromes, and how the professional skill sets are ideally suited for delineating between the various speech and the language dominant dementia syndromes. Speech pathology service provision should also be seen as essential services for the provision of therapeutic intervention for the presenting linguistic–cognitive–behavioural issues and supportive services to

patients and their families during the often-long trajectory phases of the dementia condition.

In summary, my recommendations are:

Recommendation 1 – Promote the role of speech pathologists in the assessment of dementia, in providing 1:1 direct therapeutic intervention and support to the patient, their family and caregivers

Recommendation 2 – Ensure that all patients presenting with deteriorating speech and language-related skills are routinely assessed and clinically managed by a speech pathologist

Recommendation 3 – Promote a greater emphasis on a psychosocial approach to assessment and management of dementia throughout the long trajectory of the condition, and this is especially relevant to language dominant dementias.

Each of these recommendations will be discussed.

Recommendation 1 – Promote the role of speech pathologists in the assessment of dementia, in providing 1:1 direct therapeutic intervention and support to the patient, their family and caregivers

Effective communication skills are essential for people to establish and maintain interpersonal relationships with other people in everyday life. Communication skills however can be compromised by disease due to a myriad of medical conditions, such as dementia. The wide spectrum of different forms of dementia affect communication skills in a variety of ways, and these profiles are reflective on the underlying neuropathology of the specific dementia syndrome as well as the brain location of the infiltrating pathology. Aspects of individual's communication potentially compromised by dementia may include:

- *Interpreting interpersonal nonverbal language* (i.e. body language, vocal tone & intonation, facial expression of the speaker, context of the situation, etc.)
- *Processing and understanding the spoken-verbal message*. This includes accurately and timely 'linking' of conceptual meaning of individual words, phrases, sentences/utterances and the overall intent of the conversational discourse. Processing and understanding the various parts of an utterance (i.e. grammar, syntax & sentence structure) as well as the overall episode of interpersonal information exchange /interaction is also important.
- *Cognitive-behavioural skills essential for encoding and acting upon the incoming verbal and non-verbal information*. These skills include sustained and divided attention, auditory-verbal trace recall/retention skills, adequate hearing acuity, working memory, verbal reasoning, planning/organisation skills, and problems solving skills.
- *Formulation of thoughts into spoken language*. This involves 'transferring' ones thoughts and communicative intent into well select words and full-length utterances to effectively and efficiently meet the needs of the listener. The speaker needs to adjust their verbal output to suit the context and person/people involved in the communicative interaction. This may also involved speaking in different language codes as previously familiar and known by the individual.
- *Literacy skill*. This includes reading comprehension, reading aloud, spelling, written expression, and skilful use of various technology previously used by the individual.

- *Speech and voice production.* This involves clear and well-articulated speech sound production, voice quality including singing.
- *Swallowing skills* of various different food and fluid consistencies. Sharing a meal and socialising with other people is very important in our culture for social connectivity and one's general well being.

With respect to behaviour, speech and language characteristics are interestingly understood to be among the most reliable behavioural markers for distinguishing among dementia variants. Different patterns of impaired and preserved cognitive-communication related skills are associated to specific dementia syndromes. For this reason, speech-language pathology may be poised as a discipline to play an increasingly visible role in dementia diagnosis and management" (Reilly et al. 2010 p 439). One particular type of dementia prone to misdiagnosis by health clinicians is Primary Progressive Aphasia (PPA). PPA is an atypical dementia in which a decline in speech and language skills presents as the initial symptom of disease (Pijnenburg, 2004). PPA is a broad diagnostic category within the Fronto Temporal Dementia (FTD) spectrum of dementia disorders. There are three distinct clinical presentations: Semantic Dementia (SD), Non Fluent progressive Aphasia (NFPA) and Logopenic Phonological Aphasia (LPA) (Gorno-Tempini et al., 2011; Grossman et al. 2010; Croot et al. 2009). These three different variants of PPA have distinctive speech and language symptoms and distinctive features, and thorough evaluation of all aspects of communication needs to be conducted to differentially diagnose the particular dementia subtype.

Unlike most other forms of dementia with onset during later life, the onset of FTD / PPA usually occurs between 45 – 65 years of age. These newly diagnosed individuals with PPA are often still working, raising their family, and meeting financial and personal commitments. Symptoms and behaviours are often mistakenly attributed to depression, midlife concerns, marital conflict and other psychiatric conditions (Van Vliet et al. 2011, Merrilees et al. 2010). In the early years of the PPA condition cognitive skills are preserved, and thus most individuals with PPA are painfully aware and insightful of their declining speech and language skills. This intact insight of their communication-related difficulties places them at risk of increasing social withdrawal, anxiety and clinical depression if misdiagnosed and/or managed inadequately. Adding to the high risk of under diagnosis or indeed misdiagnosis in these atypical forms of dementias is the astute level of expertise by health clinicians in detecting and evaluating very specific and sometimes subtle communication-related characteristics associated with PPA. This level of assessment of neurolinguistics features and oral motor related disorders are often beyond the skills sets of the medical or Neuropsychology clinicians; areas language processing (i.e. aspects such as conceptual semantics, syntax, grammar, social pragmatics), various forms of verbal expression (such as underlying linguistic basis for word retrieval difficulties, cause for speech production problems, spelling and written expression deficits), and oral motor disorders (such as dysarthria subtypes, afferent / efferent dyspraxia, articulation disorders, problems with voice and swallowing skills). This was acknowledged by a large group of international experts in PPA and related disorders who in 2011 published the criteria for the three distinct variants of PPA (Gorno-Tempini et al. 2011). These authors acknowledged the highly valued expertise in actively contributing in the comprehensive evaluation by Speech-Language Pathologists: *"A 20-minute bedside language evaluation could be sufficient, although detail evaluation by a speech and language pathologist is likely to be more reliable"* p 1008.

Different aspects used to assess speech and language functions in PPA are outlined in a table below - from Gorni-Tempini et al. (2011).

Speech/language function	Task	Behavioral measures	Variant in which impaired
Speech production			
Grammar	Picture description task; story retelling (e.g., picture aided); constrained-syntax sentence production task	Grammatical structure; mean length of utterance; speech rate; accuracy of content; melody; prosody; specific error types in word selection; articulation	Nonfluent/agrammatic variant
Motor speech	Motor speech evaluation, including multiple repetitions of multisyllabic words; diadochokinesis of speech articulators; spontaneous speech	Effortfulness; hesitations; presence of apraxia of speech or dysarthria; specific types of speech sound errors; factors that affect articulation (e.g., word length in syllables)	Nonfluent/agrammatic variant
Confrontation naming	Single-word retrieval in response to pictures, sounds, foods, and odors	Error rate; delay in naming; factors that affect naming accuracy (e.g., familiar vs unfamiliar items, nouns vs verbs, semantic category); error types (e.g., semantic errors, phonemic errors)	Severe deficit in semantic variant with semantic errors; moderate impairment in logopenic variant with phonemic errors
Repetition	Oral repetition of words, pseudowords, phrases, and sentences	Factors that affect repetition accuracy (e.g., predictability of the phrase, sentence length, grammatical complexity); error types	Logopenic variant with phonological errors
Sentence comprehension	Matching orally presented sentences to pictures; answering yes/no questions; following directions	Factors that affect comprehension (e.g., grammatical complexity; reversibility of the sentence, e.g., The boy was kicked by the girl vs The ball was kicked by the girl)	Nonfluent/agrammatic variant, effect of grammatical complexity; logopenic variant, length and frequency effect
Single-word comprehension	Word-to-picture matching; Word-to-definition matching; Synonym matching	Factors that affect comprehension (e.g., familiarity; frequency; grammatical word class)	Semantic variant
Object/people knowledge	Picture-picture matching; odd-one-out; semantic associations; gesture-object matching; sound-picture matching	Factors that affect object knowledge (e.g., familiarity, semantic category)	Semantic variant
Reading/spelling	Lists including regular and irregular word lists, from various word classes, matched for other factors; pseudowords matched to words in length	Factors that affect reading/spelling accuracy (e.g., regularity, frequency, word class); error types (e.g., regularization, phonologically plausible errors; articulatory distortions)	Semantic variant with "regularization" errors; logopenic variant phonologic errors

Abbreviation: PPA = primary progressive aphasia.

Recommendation 2 – Ensure that all patients presenting with presenting with deteriorating speech and language-related are routinely assessed and clinically managed by a speech pathologist

There is great potential for speech-language pathology to emerge as a frontline discipline in dementia management (Rielly et al 2010) for a range of other therapeutic reasons. Recent developments in the field of language dominant dementias have also indicated there are additional reasons WHY it is important for health clinicians to identify and discriminate between the different forms of language dominant dementias, such as:

1. Clinical features and syndromes of the various language dominant dementias have different underlying neuropathologies. Differential diagnosis of the dementia is important for recommendations of various medications (such as anticholinergic medication 'Aricept' for Alzheimer's Disease variant condition such as PPA Logopenic Progressive Aphasia). Medications for the fronto temporal variant disorders (that includes PPA Tau pathologies) are not available at present however likely within the next decade.
2. Thorough, detailed and targeted appraisal of these communication-language abilities and oral motor skills are also important because it is now known that some patient who may initially present with cognitive-communication-behaviour features usually consistent with PPA may after a period emerge with a clinical profile consistent with different forms of dementia; such as Bulbar Onset Lower Motor Dementia, Cortico Basal Degeneration or Progressive Supranuclear Palsy. Speech production (i.e. dysarthria) and swallowing difficulties are most common in the latter mentioned dementia conditions.
3. Different language dominant dementias have different presenting symptoms and different rate and skills most vulnerable for decline. Accuracy in differentiating between

the various forms of dementia is essential in tailoring individual intervention and communication strategies to preserve skills as well as develop the individual's capacity to adjust and compensate for their evolving deficits with 'islands' of preserved cognitive-communication related skills. There is a growing body of research indicating that individual diagnosed with language dominant dementia indeed benefit from speech pathology therapeutic intervention. (Green-Heredia et al. 2009, Rapp and Glucroft 2009, Jokel et al. 2009, Wong et al. 2009).

Recommendation 3 – Promote a greater emphasis on a psychosocial approach to assessment and management of dementia throughout the long trajectory of the condition, especially in all forms of language dominant dementias.

Late disease identification and misdiagnosis often lead to personal and emotional trauma for the individual diagnosed with dementia and their immediate relatives, especially for atypical forms such as PPA (Van Vliet et al. 2010, 2011). Although the clinical features of language dominant dementias have gained increasing attention in the research literature since the early 1990s, little attention has been paid to understanding the everyday experiences of those who live with the disorder. In particular, there has been very little focus in the literature or clinical support/intervention services in regards to HOW the decline in speech and language skills affects interpersonal relationships. For the most part, all efforts have mostly focused on diagnosis and the impact of the degenerative process at the impairment level of functioning (Gorno-Tempini et al. 2011). In language dominant dementias the communication deficits are not only most prominent but also progressive in nature. Everyday communicative interactions become increasingly challenging for all. Patients diagnosed with these conditions and their families generally have little support or avenues for targeted interventions within the current health services (in Victoria) during the trajectory of the condition. In Victoria the CDAMS clinic are mainly set up for initial diagnosis only and very few CDAMS clinic actually employ speech pathologists as core members of the multidisciplinary team (i.e. only about four of the ten clinic in Victoria), and indeed most states in Australia do not have an equivalent dementia diagnostic services in place. There is no clear designated referral or intervention services to provide the critical and ongoing therapeutic and affective support for individuals with PPA, and this is especially important because these individuals are younger (i.e. 45 – 65 years), may still be employed, raising families with school-aged children and are usually very insightful of their evolving and slowly deteriorating speech and language skills. Individuals with PPA often are forced to compete for outpatient services with more acute neurological conditions such as stroke and traumatic brain injury, and thus are generally not regarded as a high priority by most outpatient health services.

Spouses typically provide the necessary care for their spouse/partner with dementia in the early until middle to later phases of the condition - though may have minimal, if any, speech pathology input to support them during the phases of insidious decline. Savundranayagam et al. (2005) investigated the impact of communication problems in Alzheimer's Disease on spouses. Their findings indicated that when the person with dementia was unable to communicate effectively, there was an inevitable change in the quality of that relationship. It was felt that it was not the lack of communication per se, but the 'problem behaviours' that stem from diminished communication skills that were most stressful for the spousal caregiver. This situation is likely to be exacerbated in language dominant dementia because the communication breakdown is an early presenting feature. (Kaiser, 2007) also drew attention to the fragile emotional health of

some individuals with PPA in the early to middle phases of the condition, in part due to preserved self-awareness during the early onset phase. It is therefore reasonable to contend that emotional reactions, feeling of loss and indeed depression may play a significant role in the communicative behavioural changes of the person with language dominant dementia. In order to develop appropriate services, a better understanding of the everyday consequences of the declining communicative – cognitive skills of those with language dominant dementia and the practical coping realities of spousal caregivers attempting to support and care for their communicatively impaired relative is required. Nevertheless, it is clear that this contribution demands an improved multidisciplinary-wide knowledge base and a commitment for the provision of necessary support and management services.

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