

# **Quality of Life in Primary Progressive Aphasia: Characteristics, Influences and Implications**

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## LIST OF ABBREVIATIONS

AC	Administering Clinician
ACE-III	Addenbrooke's Cognitive Examination - Third Edition
ACE-R	Addenbrooke's Cognitive Examination - Revised
AIQ	Aphasia Impact Questionnaire-21
CAT	Comprehensive Aphasia Test
CCRSA	Communication Confidence Rating Scale for Aphasia
GDS	Geriatric Depression Scale
GP	General Practitioner
HC	Healthy Controls
lv-PPA	Logopenic Variant of Primary Progressive Aphasia
MAE	Multilingual Aphasia Examination
nv-PPA	Non-fluent / Agrammatic Variant of Primary Progressive Aphasia
PPA	Primary Progressive Aphasia
QOL-AD	Quality of Life in Alzheimer's Disease Scale
RCSLT	Royal College of Speech Language Therapists
SydBat	Sydney Language Battery
sv-PPA	Semantic Variant of Primary Progressive Aphasia
WAIS-IV	Wechsler Adult Intelligence Scale - Fourth Edition
WHO	World Health Organization
WHO-BREF	World Health Organization Quality of Life Instrument - Abbreviated

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### ABSTRACT

Primary Progressive Aphasia (PPA) is an emerging area of speech pathology practice. Despite its cross over with stroke-aphasia and dementia presentations, both fields in which quality of life has been investigated, limited research has investigated quality of life in PPA. This study conducted four health-related quality of life assessments with six individuals with PPA. Health-related quality of life was found to be individual and heterogeneous: some individuals maintained good health-related quality of life despite linguistic impairment, whereas three out of six participants screened positively for depression. Factors noted to influence health-related quality of life, or reported as particularly important to participants, included: maintaining enjoyable activities, financial situation, family and speech pathology support, and time since and reaction to PPA diagnosis. Linguistic and cognitive impairment were not related to health-related quality of life in this study, although the small sample size is noted as a limitation. Those individuals without severe semantic impairment were able to more reliably report on their health-related quality of life. Implications for assessment of quality of life in PPA and multidisciplinary care of people with PPA are discussed.

### ORIGINALITY STATEMENT

To the best of my knowledge, this thesis contains no material previously published by another person except where acknowledged. I declare that this work has not been submitted for a higher degree to any other university or institution.

Leanne Ruggero, 24<sup>th</sup> April 2017



## DEDICATION & ACKNOWLEDGEMENTS

This thesis is dedicated to Alys: speech-language therapist and stroke-counsellor extraordinaire, manager, mentor and friend. Alys was diagnosed with a rare and aggressive terminal cancer while I worked in her department in 2014-15 and I subsequently took over her caseload. Her patients and colleagues constantly asked after her. Her gentle approachability and thoughtful, holistic approach to patient care had clearly left its mark on all of them. Alys wrote in my engagement card ‘Be happy. Do the things you want in life’. In her own way, she helped remind me that life is too short and encouraged me to make the decision to move home to Australia and try a research career.

I must firstly convey my gratitude to the six participants and their families who took part in this study. They have been willing to share their time, laughter and fears with me and it has been a privilege to meet them.

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On a personal level, a huge thank you to my parents-in-law Virginia and Nick and to my parents Susan and Joe for helping to organise my and David’s wedding during these busy past few months. And last but definitely not least, my husband David and close friends for their ongoing support and encouragement in this venture, even across continents and time-zones.

The impact of language disorders on people's lives is wide ranging. There is anecdotal, as well as emerging objective, evidence that Primary Progressive Aphasia (PPA), a progressive syndrome involving language impairment, can affect mood, identity, roles and relationships (Khayum, Wieneke, Rogalski, Robinson, & O'Hara, 2012; Korte & Rogalski, 2013; Taylor et al., 2014; Volkmer, 2013). By understanding the consequences of PPA on quality of life, the speech pathology profession can develop interventions which address PPA's communication changes within a holistic context, while also assisting people and their families adjust to these changes (Byng, Cairns, & Duchan, 2002; Cartwright, 2015; Cartwright & Elliott, 2009; Rogers & Alarcon, 1998).

This study is an initial exploration of health-related quality of life in PPA; investigating what characterises and influences it, and how speech pathologists might best assess it. Ultimately, it is hoped that this work can provide rationale and therefore advocacy for speech pathologists and multi-disciplinary teams to work with people with PPA in order to maximise communication as well as health-related quality of life.

### **Primary Progressive Aphasia**

PPA is a clinical syndrome arising from neurodegenerative disease in which progressive language impairment is the primary presenting feature. Although behavioural and cognitive changes can begin soon after onset of language symptoms, cognitive and social skills have been reported to remain intact for a period of up to 14 years (Mesulam, 2001). There are three currently accepted clinical variants of PPA. The semantic variant (sv-PPA) is characterised by impaired confrontation naming, single word comprehension and object knowledge but spared repetition and speech production (Gorno-Tempini et al., 2011). The symptoms of non-fluent/agrammatic variant (nfv-PPA) include agrammatism, effortful halting speech and impaired comprehension of complex sentences but spared object knowledge (Gorno-Tempini et al., 2011). Finally, individuals with the logopenic variant

(lv-PPA) present with impaired single word retrieval and repetition of sentences due to phonological errors, but spared single word comprehension, motor speech and grammar (Gorno-Tempini et al., 2011). It is also accepted that around one in six cases prove difficult to differentially diagnose and are thus referred to as ‘mixed’ variant (mixed-PPA; Sajjadi, Patterson, Arnold, Watson, & Nestor, 2012). In terms of neuropathological correlates, sv-PPA and nfv-PPA are usually associated with one of the pathological variants on the frontotemporal lobar degeneration spectrum while lv-PPA is often associated with Alzheimer’s pathology (Gorno-Tempini et al., 2011; Grossman, 2014). It is difficult to determine the prevalence of PPA due to its likely under-diagnosis and the fact that many studies report on prevalence of frontotemporal dementia, rather than PPA *per se*. However, estimates of its prevalence range from 3.1 per 100 000 (Magnin et al., 2016) to 3.4 per 100 000 (Coyle-Gilchrist et al., 2016) to 1.1 to 6 per 100 000 (Grossman, 2014). As awareness and diagnostic measures improve, individuals with PPA are being referred to speech pathology services in increasing numbers (Rubin, Croot, Nickels, & Brennan, 2016; Taylor, Kingma, Croot, & Nickels, 2009).

PPA can be extremely debilitating, as described in personal accounts of individuals with PPA (Douglas, 2014; Gould, 2015; Twigg & LaFontaine, 2016). The effect of PPA on conversation, for example, can lead to frustration and role changes (Nickels & Croot, 2014; Taylor et al., 2014). Encouragingly, there is an increasing body of evidence that language intervention and support groups can improve communication and participation (Jokel et al., 2014; Mioshi, McKinnon, Savage, O’Connor, & Hodges, 2013; Morhardt, O’Hara, Zachrich, Wieneke, & Rogalski, 2017; Nickels & Croot, 2014). In contrast, there remains limited evidence on the effectiveness of pharmacological interventions (Boxer et al., 2013; Kerchner, Tartaglia, & Boxer, 2011). In the face of no curative treatments for PPA, an understanding of how best to maximise quality of life in PPA is greatly needed.

PPA also has devastating effects on social and financial circumstances. For example, frontotemporal lobar degeneration has been associated with significant reduction in household income and increased financial burden due to medical appointments, respite stays and associated travel. Moreover, approximately 37 percent of caregivers of people with frontotemporal lobar degeneration will stop working in order to provide care (Galvin, Howard, Tatton, & Denny, 2016). Caregivers of people with PPA are known to experience carer-strain and depression (Diehl-Schmid et al., 2013; Roche, 2014). Roche (2014) suggested that caregiver satisfaction may be mediated by the positive mood of the person with PPA, which again provides motivation for maximising quality of life in people with PPA.

### Quality of Life

The World Health Organization (WHO) defines health as “a state of complete physical, mental and social wellbeing, not merely the absence of disease” (WHO, 1948, p.1) and further defines quality of life as:

The individual’s perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns. It is a broad ranging concept affected in a complex way by the person’s *physical health, psychological state, personal beliefs, social relationships and their relationship to salient features of their environment* (WHO, 1997, p.1) [emphasis added].

In health research, quality of life has been conceptualised in numerous ways, for example as a homeostatic system (Cummins, 2003; Dodge, Daly, Huyton, & Sanders, 2012) or as a dynamic construct (Allison, Locker, & Feine, 1997). This study has chosen to investigate the construct of ‘health-related quality of life’ due to its emerging adoption in the speech pathology field. Health-related quality of life can be considered a sub-field of quality of life, and is concerned with how an individual’s health state, in this case PPA, impacts on



their ability to lead a fulfilling life. Consistent with the WHO definition above, it involves exploring the individual's perception of their physical, mental/emotional, family and social functioning (Bullinger, Anderson, Cella, & Aaronson, 1993; Hilari, Wiggins, Roy, Byng, & Smith, 2003).

In considering the mental and emotional functioning of an individual, a distinction should be made between clinical depression and the use of depression as a vernacular expression. Major depressive disorder, as most recently defined in fifth edition of the Diagnostic and Statistical Manual of Mental Disorders (American Psychiatric Association, 2013), requires the presence of depressed mood and/or loss of interest or pleasure for at least a two week period. Individuals must show a minimum of five symptoms, including one of the above two, and any of the following; fatigue, feelings of worthlessness, suicidality and changes in appetite, weight, sleep patterns, psychomotor behaviour or concentration. Symptoms must cause significant distress or impairment in every day functioning and represent a change from previous functioning (American Psychiatric Association, 2013). The 15-Item Geriatric Depression Scale (Yesavage et al., 1982) used in this study is a widely used screener for depression, although it was developed on the basis of earlier definitions of depression.

**The importance of considering quality of life.** It is now well established in the field of stroke-aphasia that it is important to consider the quality of life of people with aphasia. Firstly, improving quality of life is the ultimate aim of speech pathology intervention, whether targeted explicitly, or indirectly by improving communication (Worrall & Holland, 2003). This view was shared by 74% of clinicians in a global survey who considered maximising quality of life to be the main aim of aphasia intervention (Hilari et al., 2015). Additionally, individuals' views on quality of life and what is valuable to them can help inform therapy options (Worrall & Holland, 2003) and may predict treatment compliance. Finally, quality of

life can be used as an outcome measure to demonstrate the effectiveness of intervention (Worrall & Holland, 2003). This last point is particularly pertinent in PPA. Those gains in PPA which can be made via impairment-based therapies may not reliably carry across into conversational contexts and, due to the progressive nature of the disorder, will ultimately be lost over time (Carthery-Goulart et al., 2013; Croot, Nickels, Laurence, & Manning, 2009). Thus, it is especially important to be able to demonstrate positive change in other areas of an individual's life, such as reduction of frustration or improvement in conversational satisfaction, as a consequence of speech pathology intervention.

In accordance with the above, key professional bodies and researchers have stressed the need for speech pathologists to view their patients holistically, also considering their emotional health. The Royal College of Speech Language Therapists (RCSLT) in the UK states that clinicians should “address emotional health, and enable participation in an individual's social context” (RCSLT, 2005, p. 98). Similarly, Speech Pathology Australia has highlighted that speech pathologists can provide counselling, administer quality of life outcome measures (Speech Pathology Australia, 2015) and enhance the wellbeing and participation of people with mental health disorders including dementia (Speech Pathology Australia, 2010). In the context of PPA, Cartwright (2015) has stated that people with PPA “need a realisation that life with PPA is possible” (p. 162) and stressed that health professionals working with PPA need to be able to foster a sense of hope in their patients. Khayum and colleagues have further highlighted the need for speech pathologists working with PPA to ensure patients are regularly assessed for clinical depression, offered counselling, medication and involvement in meaningful activities (Khayum et al., 2012). Anecdotal accounts and case studies suggest that clinicians working with PPA do regularly address quality of life with people with PPA and their families, even if only informally (C. Taylor-Rubin, personal communication, August 9, 2016; Summers & Cartwright, 2016;

Twigg & LaFontaine, 2016). In recent studies in the UK, clinicians have reported adopting predominantly functional and quality of life approaches to intervention when working with people with PPA, rather than choosing an impairment-directed approach (Kindell, Sage, & Cruice, 2015; Volkmer, Warren, Spector, & Beeke, 2016). Thus, investigating quality of life in PPA may also provide evidence to support emerging clinical practice.

The current knowledge of quality of life in PPA will now be discussed, followed by an overview of research on quality of life in stroke-aphasia and dementia due to their similarities with PPA and the paucity of research in the PPA field.

**Quality of life in primary progressive aphasia.** People with PPA are not only dealing with their changing language function over time but also facing the reality of their progressive diagnosis and its effect on mood, everyday participation and relationships. However, to date, there has been little research into quality of life for people with PPA, or what factors might influence it (Spreadbury & Kipps, 2016).

A search of the literature revealed only one unpublished research study that investigated quality of life in PPA (Cartwright, 2015). Analysing interviews with five individuals with PPA, Cartwright (2015) found that PPA was associated with significant, self-reported socio-emotional consequences such as embarrassment, self-consciousness, worry, frustration and anxiety. These consequences were further exaggerated by a lengthy differential diagnosis period. Cartwright (2015) suggested that negative emotional reactions to PPA can result in maladaptive coping strategies such as withdrawal, but could also be buffered by a variety of internal and external factors such as personality, awareness and family support.

Another study has used the Communication Confidence Rating Scale for Aphasia (CCRSA; Babbitt & Cherney, 2010) to measure confidence in a PPA sample (Rogalski et al.,

2016) pre and post intervention. Although participant profiles or trends in responses were not discussed in detail, overall confidence was noted to be reduced (Rogalski et al., 2016).

While these were the only two research studies identified, four recent case descriptions authored by a person with PPA have described the lived experience of PPA (Douglas, 2014; Gould, 2015; Summers & Cartwright, 2016; Twigg & LaFontaine, 2016). The authors with PPA highlighted factors which helped them to live with PPA such as: speech pathology services, accepting the diagnosis, being organised, being treated as an individual and periods of restorative silence (Douglas, 2014; Gould, 2015; Summers & Cartwright, 2016; Twigg & LaFontaine, 2016). Similar to several of Cartwright's (2015) participants, Gould (2015) outlined the difficulties he experienced as a result of PPA such as social isolation, hating his aphasia and low confidence. He also described the benefits of speech pathology and daily practice; concluding that this had been integral to improvements in his quality of life: *"Through speech therapy, I have learned that I don't have to feel embarrassed about my speech or memory difficulties. ... I am convinced that without it I would now be in a darker place"* (Gould, 2015, p. 17). Three of the individuals with PPA particularly discussed the fact that maintaining activities contributed to their own sense of quality of life: Summers involved herself in research projects, joined a writing group, enjoyed cooking and being in her husband's workshop (Summers & Cartwright, 2016); Douglas (2014) reported exercising and keeping up with literature in her professional field despite being retired; and Twigg reported going to the shops at quieter times of the day and having members of the congregation at her local church support her to continue her lay-minister work (Twigg & LaFontaine, 2016). These accounts provide a rich and varied account of life with PPA, however, they do not include any objective measures, nor suggestions for clinicians on how to enable their patients to explore what may facilitate their own quality of life, in the way these authors have done.

In addition to the above studies, a conference presentation reported on results from a clinician-led survey of a PPA support group in the US, which investigated what people with PPA and their families wanted from speech pathology services (Silverman, 2011). Responses included more education, more therapy for communication and generally to “Make life better ... with PPA” (Silverman, 2011, slide 14). Participants’ responses highlighted that people with PPA and their families find speech pathology services valuable, in line with Gould’s (2015) sentiments.

***Depression in primary progressive aphasia.*** It has been suggested that individuals with PPA may be at greater risk of developing depression than individuals with Alzheimer’s disease due to their preserved insight (Medina and Weintraub, 2007). A number of studies have reported on the prevalence of depression in PPA and a summary of these studies can be seen in Table 1, together with confidence intervals calculated by the current author for each study (Banks & Weintraub, 2008b; Medina & Weintraub, 2007; Riedl, Last, Danek, & Diehl-Schmid, 2014; Rohrer & Warren, 2010; Singh et al., 2015; Thompson, Patterson, & Hodges, 2003). As Table 1 shows, the method for determining depression varied across these studies, with only one study using self-reports from the person with PPA (Medina & Weintraub, 2007). Many of the studies performed a retrospective analysis of clinical data to diagnose depression, which may have prevented the researchers from identifying all potentially relevant information, if it was not recorded at the time. Due to the wide range of methodologies, these studies may not yield directly comparable prevalence rates and, indeed, the range of prevalence is extremely large (15.6-77.4%). The confidence interval for the weighted mean across all these studies was calculated to improve reliability of interpretation and suggests that roughly one third to one half (34.2-45.7%) of individuals with PPA will have depression.

Table 1

Prevalence of Depression in PPA Studies

Study	N	Reported depression prevalence	95% CI	Variant of PPA	Method of screening depression	Retrospective data analysis?
Banks & Weintraub (2008)	42	42% <sup>a</sup>	27.7 – 59.0%	All	NeuroPsychiatric Inventory <sup>b</sup> (proxy-report)	Yes
Medina & Weintraub (2007)	61	34.4%	22.7 – 47.7%	All	Geriatric Depression Scale <sup>c</sup> (self-report)	Yes
Riedl, Last, Danek, & Diehl-Schmid (2014)	43	42%	27.0 – 57.9%	Semantic and Non-Fluent	Prescription of anti-depressants at any point post-diagnosis	Yes
Rohrer & Warren (2010)	33	56%	36.4 – 71.9%	All <sup>d</sup>	NeuroPsychiatric Inventory (proxy-report)	No
Singh et al. (2015)	65	43%	53.4 – 77.4%	All <sup>d</sup>	NeuroPsychiatric Inventory (proxy-report)	No
Thompson, Patterson, & Hodges (2003)	47	27.7%	15.6 – 42.6%	Semantic only	Unclear from paper	Yes
Total	291	39.9% <sup>e</sup>	34.2 – 45.7% <sup>e</sup>			

<sup>a</sup> Reported in a bar graph and exact percentage is difficult to determine  
<sup>b</sup> Cummings et al. (1994)  
<sup>c</sup> Yesavage et al. (1982)  
<sup>d</sup> Data for each PPA variant are also individually specified in the article  
<sup>e</sup> Calculated as weighted mean and confidence intervals across all study participants

Medina and Weintraub (2007) found that the participants who scored in the depressed range on the Geriatric Depression Scale (Yesavage et al., 1982) had presentations characterised by social withdrawal and reduced mental and physical energy levels. In addition, these participants had lower naming scores than participants without depression, suggesting that depression and language ability may be related. Further analysis suggested that psychological distress develops as a response to the linguistic symptoms of PPA, rather than being a part of the underlying neuropathological disease process (Medina, 2009). It is worth noting that the opposite was true in Cartwright's (2015) small sample and that upon comparison of individual scores, one participant with poor naming scores had minimal impact to social-emotional domains while another participant with minimally impaired naming ability had profound impact in social-emotional domains.

The fact that one-third to one-half of people with PPA may have depression has significant ramifications for holistic patient management. Critically, depression may reduce an individual's motivation to seek or engage with relevant health services (Gillen, Tennen, McKee, Gernert-Dott, & Affleck, 2001; Rubin, Nickels, & Croot, 2016). Riedl and colleagues have aptly warned that even when anti-depressants are prescribed, little is known yet about their effect on mood, behaviour, language or cognition in people with PPA (Riedl et al., 2014). This review of the literature on depression in PPA clearly demonstrates that further investigation into prevalence rates, especially using self-reported and reliable measures, as well as treatment options, is vital.

**Health-related quality of life in stroke-aphasia.** The linguistic presentation of PPA can be strikingly similar to that of stroke-aphasia, hence it is relevant to consider the research on health-related quality of life in the stroke-aphasia field.

Of 75 diseases and conditions, aphasia has been reported to contribute more negatively to carer-reported health-related quality of life, than even cancer or Alzheimer's

disease (Lam & Wodchis, 2010). The factors demonstrated to contribute to health-related quality of life have varied across studies, depending whether qualitative or quantitative methods were employed and how long post-stroke the study was conducted. A recent review of the literature (Hilari, Needle, & Harrison, 2012) reported on the key factors known to influence health related quality of life. These have been summarised by the current author under the domains of communication, activity, social, health and psychological (see Table 2). Additionally, recent research by Nicholas, Hunsaker and Guarino (2017) with individuals with stroke-aphasia has revealed that cognitive ability may account for up to three times the variance in quality of life than linguistic ability does.

The concept of living successfully with aphasia is closely related to health-related quality of life and is defined by Holland (2006, p. 44) as “fitting it in, moving beyond, choosing and living one’s options, and dynamically accepting aphasia at a level that is far from passive or backward looking ... participating in life as fully as one chooses”. Hinckley (2006) and Holland (2006) first reported on factors which appeared to contribute to the ability to live successfully with aphasia, including; adaption of self-perception, taking charge of communication progress, looking toward the future (Hinckley, 2006), good physical and psychological health, social support, financial security, and, personality factors such as resilience and optimism (Holland, 2006). A series of qualitative studies have since determined that the main factors that people with aphasia, their families and speech pathologists identify as facilitating their successful living are: participation, meaningful relationships, support, communication, positivity, independence and autonomy, and, the concept of seeing living successfully with aphasia as a journey over time (Brown, Worrall, Davidson, & Howe, 2012).



Table 2

*Influences on Health-Related Quality of Life in Stroke Aphasia*

Domain	Factor of Influence	Studies	Study Type
Communication Factors	Aphasia severity	Bose, McHugh, Schollenberger, & Buchanan, 2009;	Quantitative
		Cruice et al., 2003;	Quantitative
		Franzén-Dahlin, Karlsson, Mejhert, & Laska, 2010;	Quantitative
		Hilari & Byng, 2009	Quantitative
	Communication disability or function	Cruice et al., 2003;	Quantitative
Social Factors	People and social support	Hilari et al., 2003;	Quantitative
		Sarno, 1997	Quantitative
		Cruice et al., 2010	Qualitative
		Cruice et al., 2010;	Qualitative
	Verbal communication	Hinckley, 2006;	Qualitative
Health Factors	Activity level	Parr, Byng, Gilpin, & Ireland, 1997	Qualitative
	Body functioning	Hilari et al., 2003	Quantitative
	Other medical problems	Cruice et al., 2010	Qualitative
Psychological Factors	Positive outlook	Hilari et al., 2003	Quantitative
	Emotional distress	Cruice, Hill, Worrall, & Hickson, 2010;	Qualitative
	Emotional health	Hinckley, 2006	Qualitative
		Hilari et al., 2003	Quantitative
		Cruice, Worrall, Hickson, & Murison, 2003	Quantitative

The implication drawn from the stroke-aphasia research is that if maintaining or optimising quality of life is the ultimate goal of speech pathology then intervention should focus not only on language impairment but also seek to measure and target factors which will promote quality of life (Hilari, Cruice, Sorin-Peters, & Worrall, 2016). However, speech pathologists can face a number of barriers when addressing quality of life or using a counselling approach in clinical practice including lack of training or feeling under-skilled (Northcott, Simpson, Moss, Ahmed, & Hilari, 2016; Rose, Ferguson, Power, Togher, & Worrall, 2014; Sekhon, Douglas, & Rose, 2015). Indeed, in an international survey, Hilari et al. (2015) found that those speech pathologists who addressed quality of life in stroke-aphasia tended to use informal approaches rather than standardised assessments.

**Quality of life in dementia.** Research into quality of life in dementia is also relevant as PPA is an atypical dementia syndrome and, unlike stroke-aphasia, involves symptom progression.

A national study in the UK interviewed people with dementia and found that they were able to be informants on their own quality of life, especially when supported to do so, such as through the use of visual aids (Alzheimer's Society, 2010). The findings suggested that people with dementia can indeed maintain a good quality of life, and that quality of life continues to be influenced by factors outside the person's dementia. The factors that were determined to facilitate quality of life are presented in Table 3, with the most important being 'relationships or someone to talk to'. Interestingly, of the ten factors, four (1, 5, 6 and 8) including this most important factor, are areas where speech pathologists can potentially make a valuable contribution.

Table 3

*Factors Found to Facilitate Quality of Life in Dementia.*


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1	Relationships or someone to talk to
2	Environment; both physical and ambient
3	Physical health, which was viewed as a vehicle to independence and engaging in activities
4	Sense of humour
5	Independence
6	Ability to communicate
7	Sense of personal identity
8	Ability or opportunity to engage in activities
9	Ability to practice faith or religion
10	Experience of stigma

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Note: Factors were weighted by importance to the interview respondents and number of times mentioned in interview transcripts, then analysed statistically to determine degree of importance. Factors are in order of descending importance from top to bottom.

Additional studies have highlighted factors such as depression, impaired activities of daily living and neuropsychiatric behaviours as being detrimental to quality of life at various stages of dementia severity (Bakker et al., 2014; Bruvik, Ulstein, Ranhoff, & Engedal, 2012; Giebel, Sutcliffe, & Challis, 2015).

As in the stroke-aphasia field, barriers to addressing quality of life in clinical practice have been reported. Vince, Clarke and Wolverson (2016) found psychiatrists reported having little time to address quality of life and, although they acknowledged it was a part of their role, felt they were not well supported to do this. In addition, some psychiatrists viewed dementia diagnosis as a point at which to engage with their patients about the idea of living well, whereas others reported believing this was a discussion to have later in disease progression, an issue that may prove similarly relevant in PPA.

### **Study Aims**

Recent gains in knowledge of quality of life in stroke-aphasia and dementia highlight the current gap in knowledge of quality of life in PPA. Speech pathologists are often the

professionals who are in the most regular or face to face contact with individuals with PPA and therefore the speech pathology profession needs to develop a greater understanding of the nature of quality of life in PPA so that management and intervention can best aim to maximise it. There is also a need to understand how best to assess quality of life in terms of the available assessments and the influence of linguistic and cognitive capabilities of individuals with PPA. Hence, the first aim of this study was to explore how people with PPA view their health-related quality of life. The second aim was to establish whether any particular factors mediate the impact of PPA on health-related quality of life.

To address these two aims, a series of assessments were administered that encompassed the key areas of physical, mental/emotional, family and social functioning highlighted in the definition of health-related quality of life. The assessments chosen were originally designed for either Alzheimer's disease or stroke-aphasia and were selected due to the absence of assessments designed specifically for PPA and the crossover of symptoms of these two conditions with those of PPA. Assessment choice was also motivated by whether assessments were aphasia-friendly or lent themselves to aphasia-friendly adaptation (see further below). Thus, a third aim of the study was to critique the selection and adaptation of stroke-aphasia and dementia assessments for use with people with PPA, and to make preliminary recommendations for assessment of quality of life in PPA for speech pathology clinical practice.

The participants in the study are described below, then the two main aims are addressed in sequence, followed by a General Discussion and Conclusion. Findings of the study's third aim are found embedded throughout as their significance becomes relevant. To anticipate the results, this study demonstrates that health-related quality of life in PPA is highly variable, and suggests that the factors which influence health-related quality of life are numerous and may be inter-related. It also finds that people with PPA without significant

semantic impairment are able to self-report on a range of health-related quality of life assessments and that those designed for the dementia population may be less suited for use in PPA.

### **Participants**

Ethical approval for the study was granted by the South Eastern Sydney Local Health District Human Research Ethics Committee (reference no 16/057) and by Macquarie University Human Research Ethics Committee (reference no 5201600718).

### **Recruitment**

Six<sup>1</sup> participants were recruited via health professionals in Sydney, Australia. Inclusion criteria required participants to be:

- i) diagnosed with PPA by an appropriate health professional according to the international consensus criteria (Gorno-Tempini et al., 2011);
- ii) proficient in English as their dominant language;
- iii) between 45 and 85 years of age;
- iv) living in the community
- iv) able to give consent to participate with sufficient comprehension and insight to do so, as determined by the participant's speech pathologist (not the researcher) during the recruitment process.

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<sup>1</sup> The small n in this study is a consequence of two factors. From conception to write-up, the Masters for which this study is assessable is a nine-month long program. Additionally, it is a well established fact that recruitment in PPA can be slow for a number of reasons including its rarity (Etcheverry et al., 2012). However, this study is still believed to hold significant value given it is the first known study of this particular nature in a PPA sample.

## **Participant Demographics**

Participants and their families answered a short demographic questionnaire regarding the participant's age, relationship, living situation, onset of symptoms, education, occupation, and medical, language and psychiatric history. Participants' ages, symptom duration (from noticeable symptom onset to present day) and time since formal diagnosis of PPA were calculated based on information obtained in the first session of the study. Participants' individual demographic details are listed in Table 4.

Participants were on average 68 years, 3 months of age (range = 60-77) and symptom duration averaged 33 months (range = 14-48). Four participants had a diagnosis of lv-PPA, one sv-PPA, and one mixed-PPA (with features of both lv-PPA and nvf-PPA).

## **Assessment of Language and Cognition**

Multiple language assessments were administered to evaluate expressive and receptive language in both spoken and written modalities as detailed in Tables 5 and 6. Assessments were conducted and scored by the current author, a trained speech pathologist, or by the participants' managing health care team as a part of their regular medical care. The Sydney Language Battery (SydBat; Savage et al., 2013) was chosen because of its ability to differentially diagnose the variants of PPA. It is a measure of single-word processing that involves four subtests all of which use 30 imageable nouns: confrontation naming, repetition, word-picture matching and semantic association (Savage et al., 2013). The sentence repetition subtest from the Multilingual Aphasia Examination (MAE) requires participants to repeat verbatim 14 sentences of increasing length from 3 to 24 syllables (Benton & Hamsher, 1989). The SydBat and the sentence repetition task have the advantage of reported data for PPA variants (Leyton et al., 2014; Savage et al., 2013).

Table 4

*Participant Demographics*

	P1-M	P2-S	P3-R	P4-G	P5-D	P6-J
Age at first session (years; months)	66;10	64;0	76;6	77;11	60;7	62;4
Gender	M	F	M	M	M	M
PPA variant	Logopenic	Logopenic	Logopenic	Logopenic	Semantic	Mixed
Symptom duration (months)	44	14	33	30	30	48
Time since diagnosis (months)	16	12	27	7	23	29
Co-morbid medical conditions	High cholesterol, Gout	Hormone replacement therapy	Nil	Nil	Nil	Nil
Closest communication partner	Wife	Daughter	Wife	Wife	Wife	Wife
Assistance at home?	No	No	No	No	Cleaner	Cleaner
Highest level of completed education	Diploma	Diploma	Degree	Military training	High school (age 15)	Diploma
Previous occupation	Military then travel agent	Youth worker: Senior case manager	Company director	Military then computer sales	Craftsman	Psychiatric nurse

\*Assistance at home determined by family / participant report of any external services assisting in the home such as cleaners, occupational therapists, professional carers.

In addition, several subtests of the Comprehensive Aphasia Test (CAT; Swinburn, Porter, & Howard, 2004) were used to assess additional linguistic skills: action naming, non-word repetition, spoken picture description, written picture description, word reading, spoken sentence comprehension, written sentence comprehension and paragraph comprehension.

The Addenbrooke's Cognitive Examination (ACE-III; Hsieh, Schubert, Hoon, Mioshi, & Hodges, 2013) was used to measure cognitive skills. The ACE-III was developed to assess cognition in dementia syndromes including frontotemporal dementia and PPA, and includes tasks that assess attention and orientation, language, verbal fluency, memory and visuospatial abilities. The ACE-III is scored out of 100, where a higher score indicates better cognitive ability, and a score under 88 is indicative of dementia (Hsieh et al., 2013). Scores below 88 are expected in PPA since the ACE-III has been designed to be sensitive to detecting language-based dementias. In a validation study, Hsieh et al. (2013) reported that individuals with PPA scored significantly lower than healthy controls in the domains of attention, memory, fluency and language. Participant 3-R had been assessed on an earlier, comparable, edition of the test (ACE-R; Mioshi, Dawson, Mitchell, Arnold, & Hodges, 2006) as part of his regular medical care only a month before taking part in the current study, hence those results are instead reported.

Non-verbal cognitive skills were assessed using three subtests of the Wechsler Adult Intelligence Scale (WAIS-IV; Wechsler, 2008). These were: forward and backward digit span, which are measures of immediate memory/working memory; matrix reasoning, a measure of non-verbal reasoning requiring completion of sequences of patterns; and coding, a measure of processing speed based on transcribing a series of symbols according to a key.

**Results.** Participants demonstrated a wide range of scores on language (see Table 5) and cognitive assessments (see Table 6) in accordance with their variant of PPA and symptom duration.



All participants scored below the normal range on the single word naming task of the SydBat, as is expected in sv-PPA and lv-PPA (Gorno-Tempini et al., 2011; Savage et al., 2013). Two participants with lv-PPA of protracted duration (P3-R, P4-G) plus one with sv-PPA (P5-D) also scored below the normal range on the semantic association subtest of the SydBat. At sentence level, four participants had performance below the normal range on the spoken picture description (P1-M, P3-R, P4-G, P6-J) and spoken sentence comprehension (P1-M, P2-S, P3-R, P6-J) subtests.

The range of scores obtained on the ACE-III in this study (including one participant assessed with the ACE-R) (Mean = 63.5, SD = 20.8, range = 35-90, n=6) was not dissimilar to the scores for people with PPA reported in the ACE-III validation study (mean = 58.7, SD = 21.4, n=21). All but one participant (P4-G) scored under the cut-off point for dementia. P3-R's overall ACE-III score was quite low (35/100) and this was observed by the administering clinician to be consistent with his poor orientation during sessions. A caveat here is that on tasks requiring verbal responses such as the memory items, points were lost either because the participant truly did not know, for example, what city they were in; or because they were experiencing word-finding difficulties and could not retrieve an answer. As such, Mesulam (2001) has cautioned against relying too heavily on the interpretation of neuropsychological test scores in PPA because language impairment may also affect scores on other domains.

In fact, on non-verbal assessments, all participants scored within the normal range for non-verbal reasoning, as measured by the matrix reasoning subtest of the WAIS-IV. One participant (P1-M) fell below the normal range on forward digit span and two (P1-M, P3-R) fell below the normal range on the coding (processing speed) subtest.

Table 5  
*Language Scores*

		Participant		Duration of symptoms (months)											
				PPA Variant											
				Subtest	Max Score	Raw Scores									
Expressive Language	SydBat <sup>a</sup>	30	18*	19*	7*	19*	7*	19*	7*	19*	7*	19*	7*	5*	
	CAT <sup>b</sup>	10	7**	10	2**	8*	0**	8*	0**	8*	0**	8*	0**	0**	
	SydBat	30	26*	28*	25*	30	29*	30	29*	30	29*	30	29*	29*	
	CAT	10	9	8*	8*	10	10	10	10	10	10	10	10	10	
	MAE <sup>c</sup>	14	3*	8	5*	7*	9	7*	9	7*	9	7*	6*	6*	
	CAT	65+	30**	42	25**	33*	47	33*	47	33*	47	33*	13**	13**	
	CAT	35+	14**	33	0**	28	15**	28	15**	28	15**	28	1**	1**	
Receptive Language	SydBat	30	30	28	21*	28	14*	28	14*	28	14*	28	27*	27*	
	SydBat	30	28	29	10*	24*	11*	24*	11*	24*	11*	24*	28	28	
	CAT	66	NA <sup>d</sup>	61	56**	66	60	66	60	66	60	66	18**	18**	
	CAT	32	25**	25**	20**	30	32	30	32	30	32	30	14**	14**	
	CAT	32	16**	27	17**	30	32	30	32	30	32	30	15**	15**	
	CAT	4	4	2*	2*	3.5	3	3.5	3	3.5	3	3.5	3.5	3.5	
	CAT	4	4	2*	2*	3.5	3	3.5	3	3.5	3	3.5	3.5	3.5	

\* <2SD from the mean for healthy controls

\*\* <cut-off point for healthy controls on the CAT

<sup>a</sup> SydBat: Sydney Language Battery, normative data from Savage et al. (2013)

<sup>b</sup> CAT: Comprehensive Aphasia Test, normative data from Swinburn, Porter and Howard (2004)

<sup>c</sup> MAE: Multilingual Aphasia Examination; Normative data from Leyton et al. (2014)

<sup>d</sup> Data not collected as subtest added to assessment battery after first participant completed

Table 6  
*Cognitive Scores*

		Participant	P1-M	P2-S	P3-R	P4-G	P5-D	P6-J
Duration of symptoms (months)			44	14	33	30	30	48
PPA Variant			lv-PPA	lv-PPA	lv-PPA	lv-PPA	sv-PPA	Mixed
Cognitive Screen	Subtest	Max Score	Raw Scores					
	Memory	26	16	18	4	20	13	9
	Attention	18	13	17	7	18	13	12
	Fluency	14	7	9	2	12	10	0
	Language	26	18	23	13	24	10	12
	Visuospatial	16	15	14	9	16	14	13
Total Composite		100	69 <sup>#</sup>	81 <sup>#</sup>	35 <sup>#</sup>	90	60 <sup>#</sup>	46 <sup>#</sup>
Non Verbal Intelligence	WAIS -IV <sup>b</sup>	Standard Scores						
	Forward Digit Span	3*	8	6	10	8	9	
	Backward Digit Span	6	11	5	10	7	8	
	Coding	2*	10	1*	13	7	11	
	Matrix Reasoning	11	5	7	14	14	11	

\* <2SD from the mean for healthy controls  
# Scores below 88 are indicative of dementia as per Hsieh, Schubert, Hoon, Mioshi and Hodges (2013)  
<sup>a</sup> ACE-III: Addenbrooke's Cognitive Examination - 3<sup>rd</sup> Ed.  
<sup>b</sup> WAIS-IV: Wechsler Adult Intelligence Scales - 4<sup>th</sup> Ed. Normative data from Wechsler (2008). Standard scores have a mean of 10 and SD of 3.

## **What Do People with Primary Progressive Aphasia Report About Their Health-Related Quality of Life Using the Chosen Assessments?**

The four anecdotal accounts of living with PPA described above (Douglas, 2014; Gould, 2015; Summers & Cartwright, 2016; Twigg & LaFontaine, 2016) and Cartwright's (2015) study have demonstrated that quality of life may vary from one individual with PPA to the next. This study therefore aimed to further investigate this heterogeneity objectively and obtain a profile of health-related quality of life in particular in participants with PPA using standardised assessments drawn from other neurological populations.

### **General Methods**

Four published assessments were administered and scored by the current author: The Quality of Life in Alzheimer's Disease Scale (QOL-AD; Logsdon, Gibbons, McCurry, & Teri, 1999), the Aphasia Impact Questionnaire-21 (AIQ; Swinburn, 2013), the Geriatric Depression Scale – Short Version (GDS; Yesavage & Sheikh, 1986) and the Communication Confidence Rating Scale for Aphasia (CCRSA; Babbitt & Cherney, 2010). These were chosen on the basis of previous literature, following the completion of an educational workshop on the use of quality of life assessments in aphasia and using clinical judgement. However, the adoption of assessments standardised on other neurological populations rendered the choice largely exploratory. A number of adaptations, described below, were made to the format of the assessments, either so they were appropriate to the PPA population or to make them more aphasia-friendly in line with suggested guidelines (T. A. Rose, Worrall, Hickson, & Hoffmann, 2011).

Throughout administration of the assessments, many participants spontaneously elaborated on the rationale for their ratings. This was permitted because it added additional context and insight to the ratings. The administering clinician did not act as an interviewer, *per se*, however did respond in a conversationally appropriate manner and assisted with

word-finding difficulties as required or requested. It has been suggested that this elaborative approach can be appropriate when administering quality of life assessments in clinical practice (Cruice, 2016). Thematic analysis was beyond the scope of this study, however, relevant quotes are included as supportive anecdotal evidence. These qualitative anecdotes were thought to be valuable for developing a holistic understanding of quality of life, especially given the small sample size and paucity of knowledge in this field.

### **Assessment of Overall Health-Related Quality of Life Using the QOL-AD**

**Assessment description.** The QOL-AD is a measure of an individual's perception of the impact of dementia on a wide range of domains such as energy levels, family and chores. In this assessment, participants rate themselves as poor (1), fair (2), good (3) or excellent (4) on thirteen items (see Appendix A), providing a composite score out of 52. For this study, the QOL-AD was reformatted from having all items on one page to one item per page in large text. A series of green ticks and red crosses were added underneath the written descriptors, as can be seen in the example in Appendix B. Participants' scores on the QOL-AD were compared to scores for 14 healthy ageing controls (ages 69-87), obtained from Cartwright (2015), using Singlims analyses (Crawford, Garthwaite, & Porter, 2010).

**Results and Discussion.** The wide range of composite scores (mean = 39.8, SD = 8.93, range = 26-49) demonstrated heterogeneity in health-related quality of life in the sample (see Table 7). P4-G's score was significantly reduced compared to healthy controls, indicating that his perception of his health-related quality of life was significantly lower than that of a healthy person of similar age. The remaining participants' composite scores fell within the normal range although some scores on individual items were noted to fall outside of the normal range. Notably, four out of the six participants (P1-M, P2-S, P5-D, P6-J) rated themselves significantly higher than healthy controls in their ability to do chores. Another trend across the sample was that every participant rated the Item 5: 'How do you feel about

your memory?’ as their lowest item on the QOL-AD. This suggests that people with PPA feel they are performing less well with regard to memory compared to other domains such as energy levels or mood.

The QOL-AD was easy to administer and the aphasia friendly version designed for this study was understood by all participants. One advantage of this assessment was that it invited participants to rate and discuss whether their financial situation was a source of stress for them without approaching the topic in a confrontational manner, and without needing to ask participants about their household income.

Overall, however, the assessment was not particularly sensitive for individuals with PPA, as similarly noted by Cartwright (2015). As an example, P6-J was identified as having depression on the GDS, had profound linguistic impairments and reported being bored and socially isolated, yet his score on the QOL-AD did not fall outside of the range for healthy controls. It is possible that broad assessments of quality of life designed for Alzheimer’s are not well suited to the PPA population due to the vast differences in the way that communication versus cognitive symptoms will affect activities of daily living in the two conditions (Mioshi et al., 2007).

Table 7

*Individual Participant Scores on the Quality of Life in Alzheimer's Disease Scale and Comparison to Healthy Controls*

Domain of Quality of Life	Current Study (n=6)						HC (n=14) <sup>†</sup>	
	P1-M	P2-S	P3-R	P4-G	P5-D	P6-J	Mean	SD
Physical Health	4 <sup>#</sup>	3	3	1 <sup>*</sup>	3	3	3	0.39
Energy	3	3	3	2	4 <sup>#</sup>	3	2.79	0.43
Mood	3	4	2	3	4	3	3.14	0.66
Living Situation	4	4	3	2 <sup>*</sup>	4	3	3.57	0.51
Memory	2	3	2	1 <sup>*</sup>	3	2	2.71	0.61
Family	4	4	3	2 <sup>*</sup>	4	3	3.71	0.47
Marriage	3	4	4	2 <sup>*</sup>	4	3	3.5	0.52
Friends	3	4	3	2	4	3	3.36	0.63
Self as a Whole	2	3	2	3	4	2	3.14	0.53
Ability to Do Chores	4 <sup>#</sup>	4 <sup>#</sup>	3	2	4 <sup>#</sup>	4 <sup>#</sup>	2.93	0.47
Ability to Do Things for Fun	4	4	2	2	4	3	3.14	0.77
Money	4	4	3	2	3	3	3	0.55
Life as a Whole	3	4	3	2 <sup>*</sup>	4	3	3.36	0.5
Total	43	48	36	25 <sup>*</sup>	49	38	41.36	4.78

Abbreviation: HC: Healthy Controls

<sup>†</sup>Healthy control data from Cartwright (2015), n=14

<sup>\*</sup>Significantly higher than HC (p<0.05 see Appendix F for full statistical details)

<sup>#</sup>Significantly lower than HC (p<0.05 see Appendix F for full statistical details)

**Assessment of Communication Activities, Participation and Mood Using the AIQ**

**Assessment description.** The AIQ is an assessment of an individual's perception of the impact of their aphasia on communication activities, participation (including activities of daily living, friends and family) and, mood. It consists of 21 items presented one-to-a-page with a five point pictorial rating scale supplemented by written descriptors (Swinburn, 2015). In this assessment, a score of zero represents no impact of aphasia (good quality of life) and a score of four represents significant impact of aphasia (poor quality of life). The 21 items are detailed in Appendix C. The second page of the AIQ was edited for this study so that the phrase "Let's look at your stroke and aphasia" was replaced with "Let's look at your life and primary progressive aphasia".

**Results and Discussion.** Individuals' scores on the AIQ are presented in Figure 1, Panels A-F. As defined by the authors of the assessment, items one to six pertain to activity (black bars), items 7 to 10 pertain to participation (white bars) and items 11 to 21 pertain to mood (red bars). P2-S and P5-D reported that their PPA was not having a substantial impact on their activities, participation or mood, which can be seen by the lack of bars in their corresponding panels. Conversely participants 1-M, 4-G and 6-J rated themselves as impacted by PPA on nearly every item.

When all scores were compared, no clear pattern emerged as to which domains participants rated as most affected. Instead the participants exhibited varying patterns of impact, discussed below with relation to the three domains the AIQ probes.



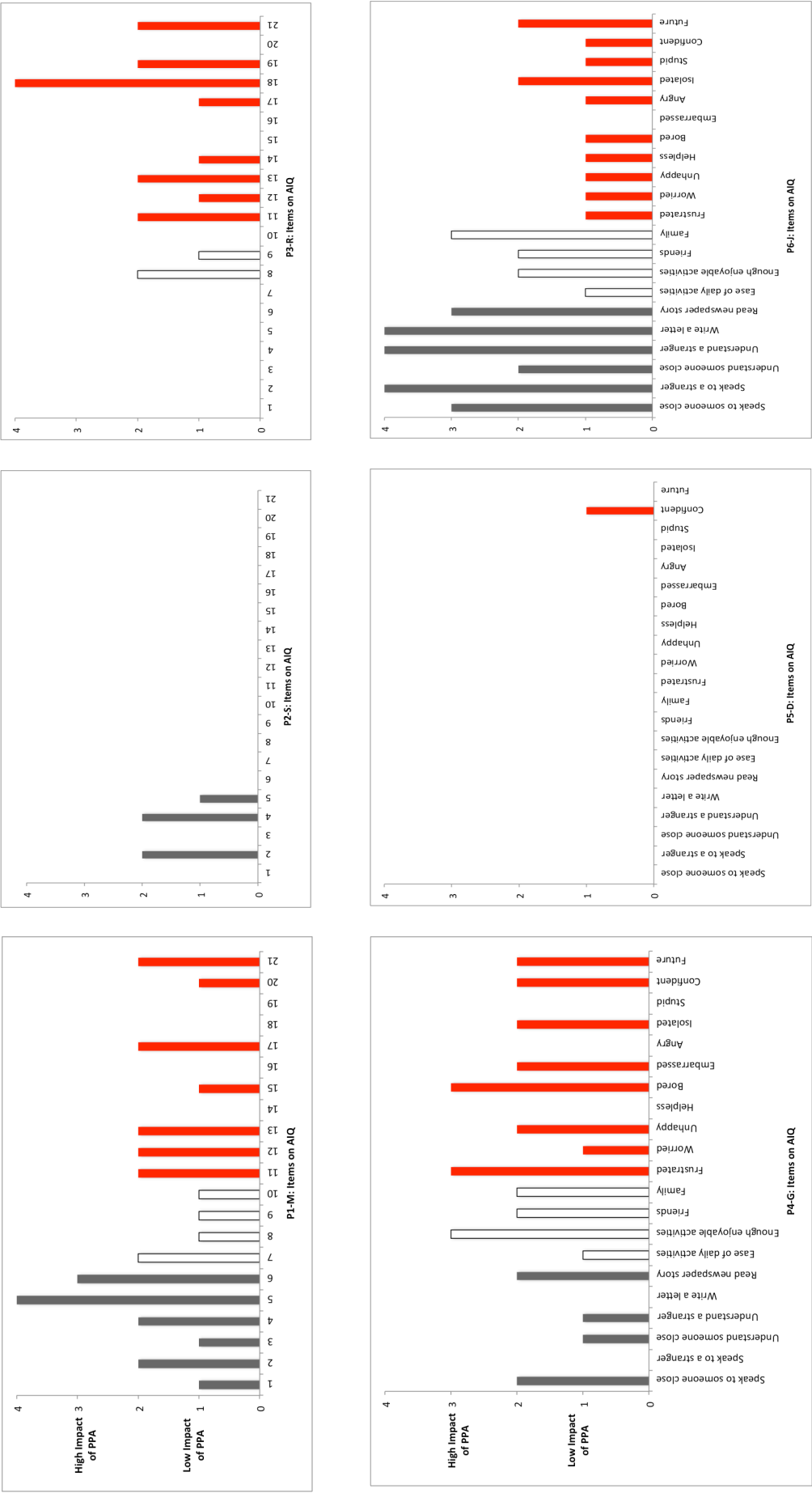


Figure 1 (A-F): Individual participant profiles for the AIQ.

AIQ: Aphasia Impact Questionnaire-21

**Communication activities.** In relation to the communication items, most participants commented that it was easier to talk to their families than to strangers. Four participants reported difficulty with reading, or that they were not reading at all any more (P1-M, P3-R, P4-G, P6-J) and all except P5-D rated themselves as having difficulty writing or no longer writing at all. In fact, for three out of six participants (P1-M, P2-S, P6-J), difficulty writing a letter received a rating that placed this item in the top three items on the AIQ impacted by PPA. These results are consistent with PPA (Brambati, Ogar, Neuhaus, Miller, & Gorno-Tempini, 2009; Graham, 2014) and the participants' respective language assessment scores.

P4-G additionally spoke about the difficulty of experiencing word finding difficulties in social situations:

*I was asked to talk about something at a ... at a military meeting I had with some ex-service people. And they asked me to get up and give a little talk about what we'd done in Vietnam. And I couldn't remember some of the words. Like I'd dealt with these people who had leprosy in leper colonies. And I couldn't remember that they were lepers and I couldn't remember the word leprosy. So it's very hard to try to [laughs as if amused] have this discussion with people when you can't bloody remember key things in it.*

P1-M demonstrated his resilient attitude when recounting an anecdote about cutting his finger but not being able to find the words to tell his wife what the problem was or explain that he needed a bandage. He was amused while retelling the story and surmised there was a need to laugh at these communication misunderstandings, which resonates with the fact that sense of humour was an influential factor for people with dementia (Alzheimer's Society, 2010).

***Participation.***

Some participants described having a busy schedule of enjoyable activities, while others did not. In relation to Item 8 ‘This week, did you have enough positive things to do, things you wanted to do?’ P4-G responded “*I don’t really do very much for fun at all*”. P2-S, conversely, rated herself as unaffected on participation items and likened being retired to “*being on a holiday*”. P1-M also rated himself as relatively unaffected and explained how he takes considerable pleasure walking, exploring the city, going to see movies and taking holidays.

In relation to Item 9 ‘This week how were things with friends?’, several participants spoke about the effect of PPA on maintaining friendships. For example, P4-G said “*I’ve got very few friends outside the family*” and P3-R noted:

*I don’t have much contact. I’ve gotta, I’m gon... I’m trying, I don’t know how but I’m ... I’m anxious to get off my butt and try and re-establish the the contact. See, at this stage in life there aren’t many **real** [his emphasis] friends ... very close friends and ... and so I don’t want this [PPA] for to stop me ... going to those people or even ringing them basically. It’s it’s already happened for some time that I’ve gone shy. But I hate that word. And fear, I hate the word fear.*

*... I’m not scared of what’s coming. What I’ve got to fight to do is to not let that stop me from friendships.*

This concurs with findings in the stroke-aphasia field that friendships can be particularly affected by aphasia (Hilari & Northcott, 2016). Several participants also noted that keeping up with friends was made more difficult by the need to interact with larger groups of people:

*P6-J: [laughs, bitterly] Er well .. every now and then I ... I can’t do anything with these huge people. I can do ... ah sometimes with boys, just one or two, that’s fine*

*Administering Clinician (AC): Just small groups, yeah*

*P6-J: Yeah that's it. That's about all*

*AC: Mmhmm*

*P6-J: But every now y'know I just yeah well I can't really speak very much really so ... anyway that's life*

Family was central to some participants' lives, while others implied that family interactions were strained. When asked about how things were with his family on Item 10, P6-J sighed audibly, shortly followed by “*not good*”, while P4-G simply said “*non-existent*”. P3-R explained that his happiness was affected because his son lived in another city “*I don't get the opportunity to spend enough time with him to for... get him ready for this [PPA]*”.

**Mood.** Mood was affected in five of the six participants, with a wide range of ratings on mood items from 0 (P2-S; no impact of PPA on mood) to 17 (P4-G; moderate impact of PPA on mood) out of 44. P2-S, for example, in response to Item 13; ‘This week, have you felt unhappy?’ described that PPA was not having a significant impact on her happiness: “*Nup. It's really weird, isn't it? Because you haven't got all of the ... the mind that I would have had a couple of years ago. Um, I can let things go.*” Conversely, in response to the same Item, P4-G expressed that his level of happiness was related to his communication: “*Yeah I've felt a ... well I feel a bit unhappy when I've got nothing ... I feel I've got nothing much to do and I'm not communicating with [wife's name] very well*”.

P3-R Highlighted on multiple occasions that his mood was heavily influenced by his aphasia and that he experience strong emotional reactions to word finding difficulties:

*AC: Yeah, it's harder with other people isn't it?*

*P3-R: Yeah. That's that's the part. That's the beginning. That's, that sort of thing has been the first stuff. Um. Making me nervous.*

*AC: Mmmmm, okay, yeah.*

*P3-R: But I ha, a f-f-few, Gone. ... See there? [Pointing out his Word Finding Difficulty and effortful speaking style]*

*AC: That's fine, yeah.*

*P3-R: When that [Word Finding Difficulty] starts, I'm thinking "Oh God"!*

It was positive that, across all participants on all items of the AIQ, four (P1-M, P2-S, P4-G, P5-D) rated helplessness and feeling stupid as not affecting them at all.

The results for Item 16 regarding embarrassment are worthy of mention.

Embarrassment was reported by some participants in their anecdotes, as it similarly was in interview transcripts from Cartwright's (2015) study, as in this example from P3-R's reaction to AIQ Item 14: 'Have you felt helpless?' where he is describing a recent visit to his son:

*Er I don't think ... but I know that that certainly to him and to a large ...[attempting to say extent] to [wife's name] that they don't understand me that ... this simple thing I've just stuffed up, is very embarrassing to me.*

However, on the AIQ Item 16: 'Have you felt embarrassed?' all participants except P4-G rated themselves as zero, or unaffected, by embarrassment. This is likely because some participants have experienced embarrassment in the past but had not experienced it 'in the past week' as the assessment asks because they have learnt to avoid potentially embarrassing situations. This avoidance strategy, similar to that described by Cartwright's (2015) participants, was indeed mentioned by several participants including P1-M:

*AC: It's hard in a group isn't it?*

*P1-M: It is very. Well I would find it very difficult. And and in that situation well I probably wouldn't be in there that situation because I guess when you want ... if you're not happy of doing that sort of stuff, the idea is then there's you just don't go with that way, if you know what I mean?*

*AC: Mmmm. You try and stick to smaller groups of people?*

*P1-M: Yeah. And I would probably just listen, if it was in a group.*

**Administration.** Although the AIQ produced some valuable results, its administration proved difficult with some of the participants with lower cognitive scores, who required further scaffolding or closed questions before they would rate themselves on the scale. Additionally, verbose conversation is often a feature of fluent variants of PPA and therefore some participants talked at length about the items. As an example, administering the AIQ was most challenging with P3-R, who did have a verbose conversational style and who scored 35 out of 100 on the ACE-III. He spoke for 12 minutes after being asked if he had felt angry this week despite three attempts to bring him back to the question and the rating scale. In total, five items could not be completed due to his verbose tangents. The AIQ administration guidelines do highlight that the person administering the assessment can “modify the questioning to make it more accessible to someone with aphasia” (Swinburn, 2013, p. 17), however, it is possible the AIQ involves too much abstraction for individuals who have concomitant cognitive deficits. Based on this study’s results, people with PPA with low cognitive scores may respond better to shorter questions or questions which require a yes/no response.

### **Assessment of Mood Using the GDS**

**Assessment description.** The GDS (Sheikh & Yesavage, 1986) is a screener consisting of 15 Yes/No questions, detailed in Table 8 alongside the participants’ results. The GDS has been demonstrated to be sensitive to detecting depression for individuals with and without aphasia (Schreiner, Morimoto, & Asano, 2001) and also with and without cognitive impairment, at least in individuals with a minimum Mini Mental State Examination score of 15 (Jongenelis et al., 2005; Smalbrugge, Jongenelis, Pot, Beekman, & Eefsting, 2008). The directionality of answers on the GDS changes throughout the assessment to avoid response bias. Responses are coded as either negative or positive for depression and totalled out of 15.

A score of five or more indicates possible depression and is cause for further investigation or onward referral to an appropriate health professional (Sheikh & Yesavage, 1986). For those participants who scored within the depressed range on the GDS, this was discussed with them in a subsequent session and the information also provided in writing with an aphasia-friendly format letter. The term ‘low mood’ but not ‘depression’ was used during these discussions, since the GDS is only a screener. In addition, participants were given a sealed letter to pass onto their GP if they wished, which described their study participation and GDS results.

The GDS is traditionally administered verbally, however, in the current study individuals who appeared to have sentence level auditory comprehension difficulties or exhibited confusion during this assessment also had a typed copy of the assessment in its usual format placed in front of them so they could read along. A yes/no visual chart was also placed in front of participants to supplement verbal responses, as required.

**Results and Discussion.** Two participants (P1-M, P2-S) did not provide any responses consistent with depression, however, fifty percent ( $n=3$ , P3-R, P4-G, P6-J) of the sample were found to be at risk of depression on this assessment (see Table 8). Although there was a wide confidence interval around this percentage (95% CI [11.8, 88.2]) because of the small sample, the figure is comparable to previous studies that have reported depression prevalence in PPA ranging from 27% to 56% (see Table 1, earlier). Upon discussion with these three participants, none appeared surprised that they scored at a low level on this mood screener and all indicated they were happy to take a summary letter to their GP.

The most commonly endorsed items on the GDS were ‘Do you think people are better off than you?’ and ‘Do you feel you have more memory problems than most?’ ( $n=4$ ; P3-R, P4-G, P5-D, P6-J). This latter finding corresponds to the same participants’ low self-ratings of memory reported on the QOL-AD. Positively, all participants, including those who overall

scored at risk for depression, endorsed feeling happy and in good spirits most of the time, and did not endorse helplessness or hopelessness.

Although Medina and Weintraub (2007) found that a significantly larger proportion of depressed participants compared to non-depressed participants had a previous history of depression prior to diagnosis with PPA, this was not the case in the present study. The fact that in this study only one of the three (P3-R) participants who were positive for depression on the screener had a prior history of depression may be due to the small sample size. However, it is also possible that there are multiple or interacting factors which contribute to one's risk of developing depression, especially following diagnosis with a progressive condition, such that past history alone is not predictive. These results suggest that at this point in time, attempting to predict who is at risk of depression is likely ineffective and individuals with PPA should instead have their mood monitored closely by their healthcare team.

In designing this study, a number of depression screeners were reviewed and considered inappropriate due to their complex use of language. Medina and Weintraub (2007) have previously reported that the yes/no response style in the GDS (Yesavage et al, 1982) was easy for their sample of participants with PPA to use. This study used a shorter version of the GDS (Sheikh & Yesavage, 1986) and all participants in this sample also appeared to grasp the questions and respond easily. One participant; P2-S, required a large-text 'Yes/No' visual to point to, as without it her verbal responses were inconsistent with the rationales she offered. Given that people with PPA do appear able to use the assessment, it seems preferable in many ways to allow people with PPA to self-report on their depression symptoms rather than relying on proxy-report measures as other studies have done (Banks & Weintraub, 2008b; Rohrer & Warren, 2010; Singh et al., 2015).



Table 8

*Participant Responses for the Geriatric Depression Scale*

Item on GDS	P1-M	P2-S	P3-R	P4-G	P5-D	P6-J	Total <sup>#</sup>
Are you basically satisfied with your life?	Y	Y	Y	N*	Y	Y	1
Have you dropped any of your activities and interests?	N	N	Y*	Y*	N	Y*	3
Do you feel that your life is empty?	N	N	N	Y*	N	N	1
Do you often get bored?	N	N	N	Y*	N	Y*	2
Are you in good spirits most of the time?	Y	Y	Y	Y	Y	Y	0
Are you afraid that something bad is going to happen?	N	N	Y*	N	Y*	Y*	3
Do you feel happy most of the time?	Y	Y	Y	Y	Y	Y	0
Do you often feel helpless?	N	N	N	N	N	N	0
Do you prefer to stay at home, rather than going out and doing things?	N	N	N	Y*	N	Y*	2
Do you feel you have more problems with memory than most?	N	N	Y*	Y*	Y*	Y*	4
Do you think it is wonderful to be alive now?	Y	Y	Y	N*	Y	Y	1
Do you feel pretty worthless the way you are now?	N	N	Y*	Y*	N	N	2
Do you feel full of energy?	Y	Y	Y	N*	Y	Y	1
Do you feel that your situation is hopeless?	N	N	N	N	N	N	0
Do you think that most people are better off than you are?	N	N	Y*	Y*	Y*	Y*	4
GDS Score /15	0	0	5	10	3	6	
Indicative of Depression?	No	No	Yes	Yes	No	Yes	

\* Item's endorsement indicative of depression.

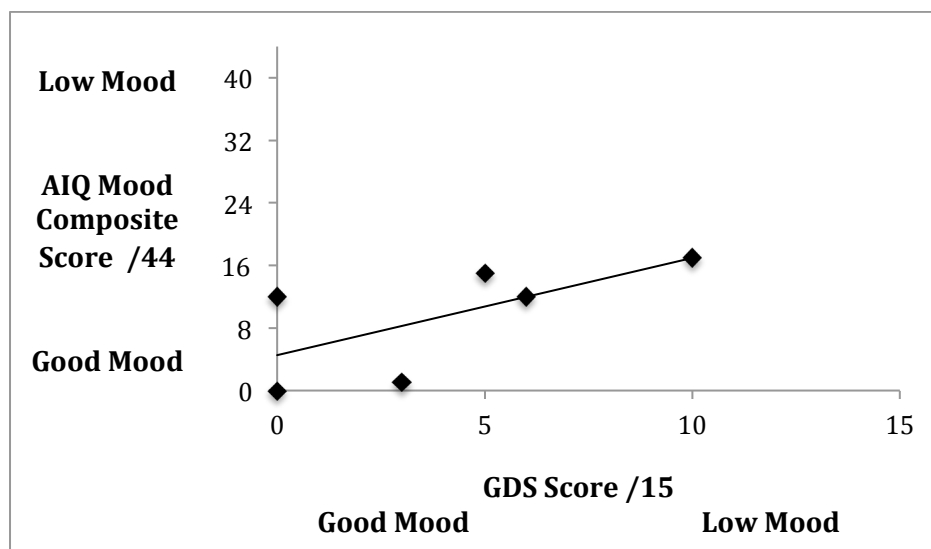
<sup>#</sup> Total number of participants who endorsed this Item for depression.

Abbreviation: GDS: Geriatric Depression Scale – 15 Item Version (Sheikh & Yesavage, 1986)

Item 15: ‘Do you think most people are better off than you are?’ was noted to be problematic for P1-M who misinterpreted the meaning as pertaining to financial situation, which was obvious from the rationale he gave for his rating. In a study administering the GDS with cognitively and non-cognitively impaired individuals in nursing homes, missing data was recorded more often for those with cognitive impairments and most often for this item (Koehler et al., 2005). Those authors did not explore this further, however, it is possible that the idiomatic nature of this expression makes it less appropriate for use in aphasic or cognitively-impaired populations.

A comparison of the AIQ and GDS assessments found that one participant (P1-M) was able to provide a more nuanced account of his mood with the five-point AIQ rating scale than with the yes/no ratings of the GDS. Although this was only observed in one participant in a sample of six, replication of this pattern would suggest that the AIQ was more sensitive to detecting slightly low mood in individuals with PPA than the GDS.

**Comparison of mood assessments.** Perception of mood was sampled across three of the assessments: a single item on the QOL-AD, 11 of the 21 items on the AIQ (as seen by the red coloured bars in Figure 1), and the entire GDS. The results from the mood items of the AIQ and the GDS are compared in Figure 2 to determine whether the assessment used would influence whether an individual was identified as having low mood. The correlation between these two assessments was moderate ( $r=0.66$ ,  $n=6$ ,  $p=0.153$ ), albeit non-significant given the small sample. This suggests that the two assessments may provide similar information in the measurement of mood in people with PPA. Of note, P1-M scored 0 out of 15 on the GDS (no depressive symptoms), however scored 12 out of 44 on the AIQ mood composite, which suggests that his mood was slightly low, but only detectable using one of the assessments.



*Figure 2.* Relationship between scores on the Geriatric Depression Scale and the mood Items of Aphasia Impact Questionnaire.

Note: Mood composite score is derived from the total score of Items 11 to 21 on the AIQ which measure various aspects of mood. Abbreviations: GDS: Geriatric Depression Scale-15 item version, AIQ: Aphasia Impact Questionnaire-21.

### **Assessment of Confidence in Communication Situations using the CCRSA**

**Assessment description.** The CCRSA is an assessment of the impact of aphasia on ten communicative situations (Babbitt & Cherney, 2010; items listed in Appendix D). Participants rank themselves on a scale from 0 to 100, which is accompanied by visual supports, such as a happy face and thumbs up sign. A score of zero represents “not confident”, a score of 50 represents “moderately confident”, and a score of 100 represents “very confident”. The CCRSA is formatted with five items per page, but was reformatted for this study to be administered with one item per page in large font (see Appendix E).

**Results and Discussion.** The average composite score on the CCRSA for the sample was 65.9 out of 100 (range = 48-97), which indicates a moderate to high level of confidence in the sample overall. This is not dissimilar to the baseline score reported for Rogalski et al.’s (2016) sample (mean = 68.2, SD 2.7) on this instrument, or Cartwright’s (2015) finding, via

interviews and a measure of self-efficacy in communication situations, that global confidence was reduced in her sample.

Individuals' scores on the CCRSA are presented in Figure 3, Panels A-F. For five out of six participants (except P5-D) confidence to follow movies was in their three highest rated areas of confidence. This is unsurprising, given that watching movies does not require any unexpected interactions or involve the use of expressive language, which is typically affected to a greater degree than comprehension in PPA (Mesulam, 2001).

Conversely, for four out of the six participants, their confidence to talk with others (P1-M, P2-S, P4-G, P6-J) and use the telephone (P1-M, P2-S, P3-R, P4-G) were in their three lowest rated areas of confidence, and three participants rated their confidence to stay in touch (P3-R, P4-G, P6-J), feel included in conversations (P1-M, P3-R, P4-G), and discuss their finances (P2-S, P3-R, P5-D) in their lowest three areas of confidence.

Interestingly, participants' confidence in their ability to make their own decisions varied considerably across the sample (P3-R=40, P6-J=40, P4-G=70, P1-M=80, P5-D=90, P2-S=100). P2-S actually described that she felt her diagnosis had made her more deliberate and had given her more desire to be an informed consumer. *"Yeah, I'm very much. Because I talk things through with people ... I think I'm a good decision maker. More so now than before this came. ... Because, I think it's that survivor thing? I have to really be on to everything."*

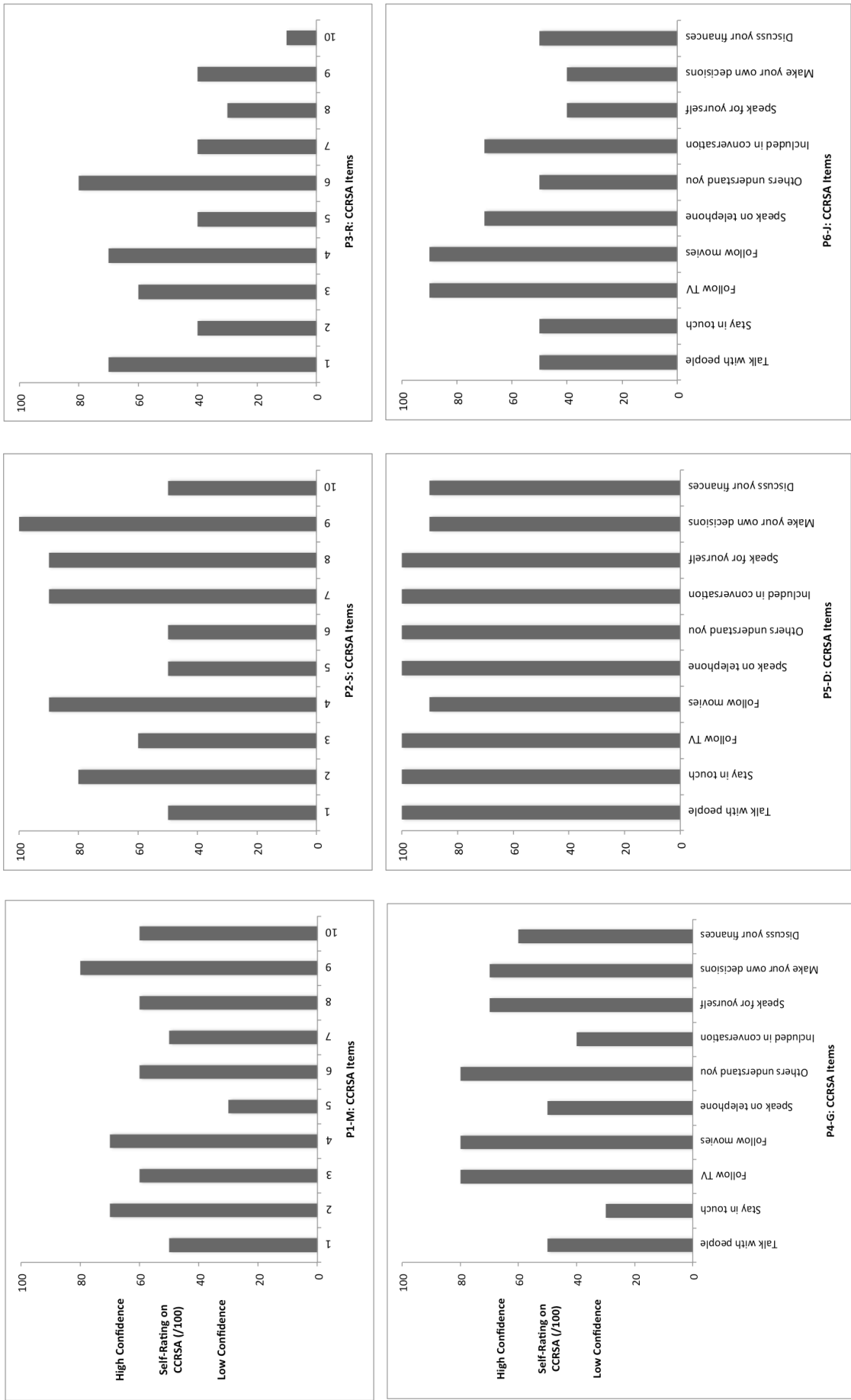


Figure 3 (A-F): Individual participant profiles for the CCRSA.

CCRSA: Communication Confidence Rating Scale for Aphasia

The adapted one-item-per-page CCRSA (see Appendix E) used for the current study was easy to administer and participants appeared to like it; “*Oh this looks very good. Really well put out. Isn’t it?*” (P2-S, upon seeing the large text version of the CCRSA). The assessment was able to reveal some interesting participant characteristics that did not arise from the other assessments administered in this study.

One disadvantage of the assessment was the fact that some items are fairly complex for individuals with receptive language or cognitive deficits, such as ‘How confident do you feel about your ability to follow movies on TV or in a theatre?’ All participants in this sample appeared to understand the object of the assessment however P2-S did use her own interpretation of the rating scale; by responding with a percentage with respect to her previous ability “*I can do about 60% what I used to be able to*”.

A second difficulty encountered was the general nature of certain items, which appeared to increase the cognitive demands of responding. For example, participants described struggling with the fact that their responses to Item 1: ‘How confident do you feel to talk with others?’ had to take into account that speaking to family members might be quite easy but speaking to strangers was near impossible.

### **Comparison of Assessment Scores**

To provide an overview of the participants’ health-related quality of life, a comparison of participants’ composite scores on all four assessments is shown in Figure 4.

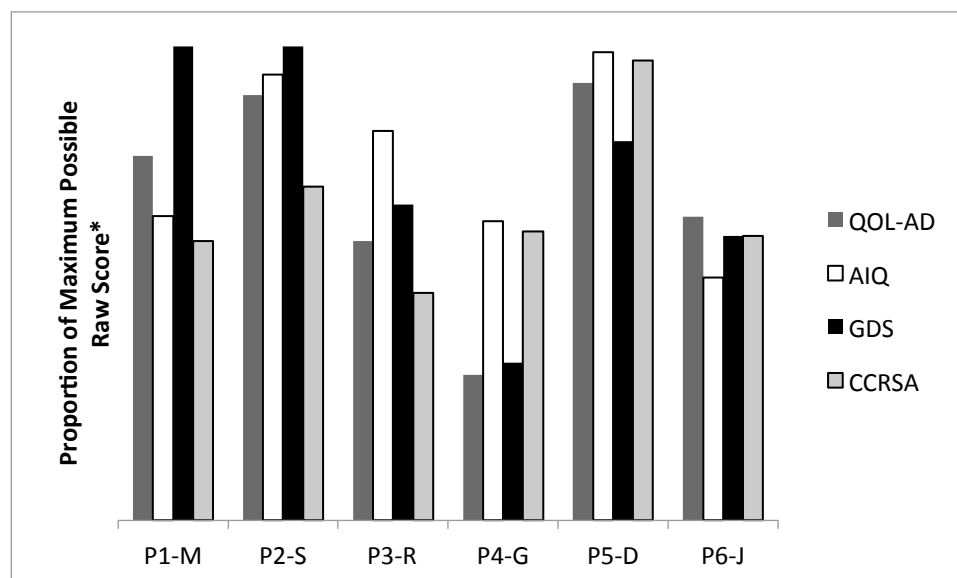


Figure 4. Comparison of composite scores on all quality of life assessments by individual.

\*Each raw score has been converted to a proportion of the maximum possible score for comparison. The minimum raw score on the QOL-AD is 13/52, thus 13 is illustrated as the minimum for the QOL-AD bars on this figure to allow for comparison across assessments.

QOL-AD: Quality of Life in Alzheimer's Disease Scale (Logsdon et al., 1999)

AIQ: Aphasia Impact Questionnaire-21 (Swinburn, 2015)

GDS: Geriatric Depression Scale – Short version (Sheikh & Yesavage, 1986)

CCRSA: Communication Confidence Rating Scale for Aphasia (Babbitt & Cherney, 2010)

### Assessing Health-Related Quality of Life in PPA

Administration of the four above assessments led to several conclusions about the practicalities of assessing health-related quality of life in PPA. A strength of this study is the attempt to include people with a range of language and cognitive skills. Hilari et al. (2012) highlighted in their systematic review that studies on health-related quality of life in stroke often do not report on how individuals with aphasia manage the assessments or exclude them entirely. Medina and Weintraub (2007) reported omitting some patients from their study who were unable to complete the GDS due to comprehension difficulties. They aptly described being “unable to avoid the pitfalls of excluding patients with significant comprehension deficits” (p. 156).

Overall, the six participants in this study engaged well with the administered assessments although two participants (P3-R and P5-D) demonstrated some difficulties with comprehension of abstract or emotional concepts and one (P3-R) with the use of the AIQ rating scale. Participants' scores on the semantic association subtest of the SydBat appeared to be an indicator of whether they would be able to comprehend the abstract concepts involved in the assessments. P3-R who scored poorly on this subtest (10 out of 30) at times discussed semantically related concepts not relevant to the question. For example, in response to Item 17 on the AIQ: 'This week have you felt angry?' he instead talked about being hurt and not wanting to "face the reality that I'm going stupid".

The provision of written materials appeared to facilitate comprehension in all four assessments. Cruice (2016) has suggested it can be appropriate to alter quality of life assessment administration such as using larger font or converting formats to one item per page insofar as it renders the content more accessible, without changing the content of the assessment itself. The AIQ, CCRSA and QOL-AD are designed for the participant to read along while the administrator reads the items aloud. In this study, the GDS was also given to participants to read along if the administering speech pathologist judged that they required written cues to better comprehend the questions. The written descriptors and visual scales developed for the QOL-AD specifically for this study were noted to also facilitate expression. An example of this is seen in the upcoming quote on p. 52-3, where P1-M pointed to key words on the page mid-sentence rather than saying them aloud.

**Insight.** Insight was not measured in this study, however, the concept of insight emerged as relevant following the assessment of P5-D. Individuals with semantic impairments have been found to be at risk of reduced insight (Banks & Weintraub, 2008a); often noticed early in relation to poor financial decisions (Hodges, Graham, & Patterson, 1995). P5-D consistently rated himself as having very high health-related quality of life



across all assessments (see Figure 4, p. 47). Tellingly, although he rated himself as 90% confident at managing his finances, his wife independently disclosed that she had had to restrict how much he did in his small business and that she was now managing his accounts.

Banks and Weintraub (2008a) have suggested that determining degree of insight may have clinical implications for treatment. For example, interventions which involve feedback contingent on responses may not prove appropriate for individuals with PPA who are unlikely to realise they have made an error. Indeed, it may also be that determining an individual's insight, via appropriate assessments or via comparison of scores from people with PPA and their partners' proxy-ratings, is similarly useful for determining which individuals with PPA are able to reliably report on their own health-related quality of life.

### **Which Factors Mediate the Impact of Primary Progressive Aphasia on Health-Related Quality of Life?**

The above results indicate that some people with PPA will report good health-related quality of life, while others will report poorer health-related quality of life or symptoms of depression. The second study aim addressed the factors that may be responsible for these differences in health-related quality of life. The literature on quality of life in stroke-aphasia and dementia has highlighted a number of factors which can influence quality of life, and three related sets of factors recurred in this literature: linguistic factors and cognitive factors, external factors such as maintaining enjoyable activities, financial situation, family and speech pathology support, and, response to diagnosis. The influence of these three sets of factors on quality of life in PPA is explored using the participants' case histories, assessment scores and qualitative comments. In most cases, the relationship between suspected influential factors and assessment scores was analysed visually and based on trends observed in the data, given the small sample size. Additional demographic factors were not examined in this study although a brief overview suggested there were no clear relationships.

### **Influence of Linguistic and Cognitive Factors on Mood**

The factors of aphasia severity, anomia severity and overall cognitive status were investigated in this study due to these factors proving influential to health-related quality of life for individuals with stroke-aphasia (Bose et al., 2009; Cruice et al., 2003; Franzén-Dahlin et al., 2010; Hilari & Byng, 2009; Worrall et al., 2017) or to depression in PPA (Medina & Weintraub, 2007).

**Linguistic function.** One participant, P6-J, described (see quote earlier, p. 36) that his increasing PPA severity had coincided with a loss of confidence and withdrawal from social situations, around two years after symptom onset. This aligned with his scores on the AIQ for friends (2 out of 4) and isolation (2 out of 4).

However, the more robust claim that anomia is related to mood in PPA (Medina & Weintraub, 2007) was not supported by the current study. As seen in Figure 5, there was no apparent relationship between naming accuracy on the SydBat and risk of depression on the GDS ( $r=-0.18$ ,  $p=0.728$ ). This relationship was also examined by correlating naming ability on the SydBat with mood composite score on the AIQ, overall language score on the ACE-III with GDS score and overall language score on the ACE-III with AIQ mood composite score. None of these analyses rendered a correlation larger than  $r=0.2$ . The lack of replication of Medina and Weintraub's (2007) finding may be an artefact of the small sample size in this study. However, these results, in addition to the lack of relationship between language impairment and social-emotional consequences in Cartwright's (2015) study, lend credence to the argument that the causes of depression are multifactorial and that knowing an individual's language ability cannot predict whether they will have depression.

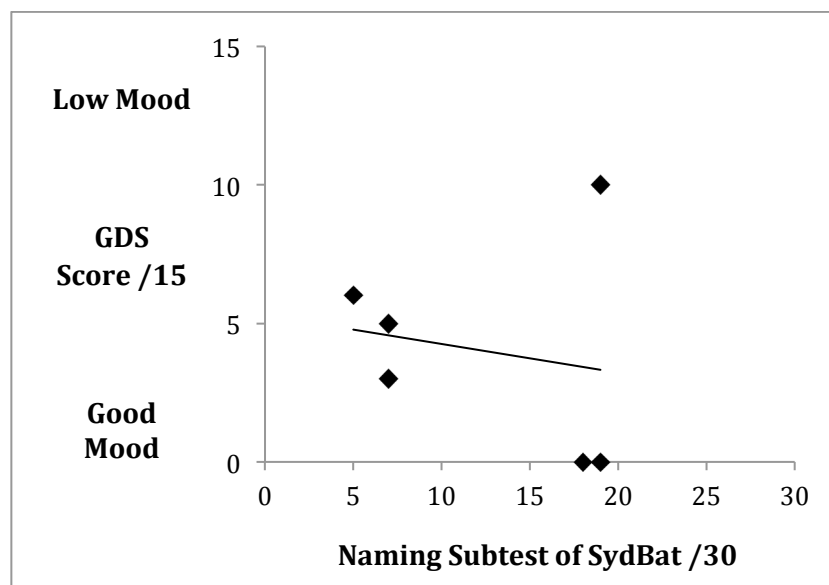


Figure 5. Comparison of Participants Mood to Naming Accuracy.

Abbreviations: SydBat: Sydney Language Battery, GDS: Geriatric Depression Scale – Short Version

**Cognitive status.** No clear relationships were found when overall ACE-III score was compared to mood on the GDS ( $r=0.02$ ,  $n=6$ ,  $p=0.974$ ), to the composite score on the QOL-AD ( $r=-0.13$ ,  $n=6$ ,  $p=0.802$ ) or to the AIQ composite score ( $r=-0.03$ ,  $n=6$ ,  $p=0.948$ ). On the basis of anecdotes, it would seem plausible that cognition has a role to play in health-related quality of life in PPA, as it has been found to in stroke-aphasia (Nicholas et al., 2017), however, this will need to be further investigated in a larger study.

### **Influence of External Factors on Health-Related Quality of Life**

The stroke aphasia and dementia literature has demonstrated that a wealth of external influences can mediate quality of life. In this study, a number of external factors were raised by participants as being influential: maintaining enjoyable activities, financial situation, and the support of family and speech pathology services.

**Maintaining enjoyable activities.** Some participants highlighted their satisfaction in pursuing non-verbal hobbies. P1-M and P6-J both reported that they considerably enjoyed

walking and that this gave them a reason to leave the house and interact with their environment. P1-M described that after being given his diagnosis, he stopped drinking, began walking everyday with a fitness tracker and lost weight. Similarly, P5-D enjoyed playing with his dog and working in his shed.

Some participants expressed enthusiasm in continuing to practice their language or cognitive skills with exercises from their therapist or on apps. P5-D practiced French on his tablet daily and P2-S reported doing at least two hours of daily naming practice. This was in contrast to P4-G who avoided crosswords or language-based activities and instead preferred television where he was not under pressure. These conclusions relate to findings from both the dementia (Alzheimer's Society, 2010) and stroke-aphasia fields (Cruise et al., 2010; Hilari et al., 2003) that have demonstrated that engaging in enjoyable activities can have a positive effect on health-related quality of life.

**Financial situation and stresses.** The QOL-AD entails participants rating their 'situation with money' (see Appendix A, Item 12), which elicited some important responses. P4-G described the fact that his financial situation was a source of stress:

*It causes me to sort of think about the future and how we'd exist if my wife wasn't working and I was just on the pension. We'd have to move out and the idea of going to live with my kids doesn't do much for me.*

During the QOL-AD, P1-M described the effect of finances on living with PPA:

*Ah, now this! [points to the word money on the page]. Once you've got that, erm ... it's er ... I think really [points to 'Excellent' on the Likert scale]  
...So we wouldn't go to ... first class or business class but we go [overseas] and ... and we have a really good time*

*...If you don't have ... I, I could think that if you didn't ... that sort of money erm ... it could be really, it it me ... it would ma- make what's happening to me much much ... not so much fun at all.*

These comments are similar to those expressed by the three participants in Holland's (2006) study who believed that financial security had helped their success post-stroke.

**Availability and quality of family support.** Family, support and meaningful relationships are known facilitators of quality of life in stroke-aphasia and dementia (Alzheimer's Society, 2010; Cruice et al., 2010; Hinckley, 2006; Parr, Byng, Gilpin, & Ireland, 1997). Thus, the family support available to participants was identified from their comments, case histories, and by observing the interaction between participants and their communicative partners during sessions.

For some participants, communication partners were more present in their day to day lives or involved in therapy than for others. Of the three depressed participants (P3-R, P4-G, P6-J), two of their wives continued to work and they were alone much of the time, such as P4-G who responded to Item 1 of the AIQ 'This week, how easy was it to talk to someone close to you [his wife]?' with: *"I always have problems talking to her because she's ... doesn't communicate with me very much"*. Both of these participants expressed that they were looking forward to their wives retiring so they could do more together or go on holidays.

P3-R expressed on multiple occasions that his mood was highly dependent on his wife's interactions with him. For example, he responded to Item 13 'Have you felt unhappy?' saying:

*P3-R: Yeah, sometimes*

*AC: Sometimes, okay. And how unhappy do you feel, would you say very unhappy, just a little bit unhappy? [pointing to rating scale]*

*P3-R: Oh you're going to be... [hesitates] have a giggle*

*AC: Oh, there's no right or wrong answers here*

*P3-R: You're going to think "Silly old bugger". ... I'm happy when my wife's happy.*

*AC: Yeah. That makes a lot of sense.*

*P3-R: And I'm really unhappy when she gets unhappy.*

*AC: Okay*

*P3-R: And some of the reasons that she might get ... is out um at the result of something I haven't been able to make it right to her, where she might turn around and sort of say mum-ma-makes of comment that is stinging or thinking like "don't be silly, you silly bugger" or [unintelligible]*

*AC: Hmmm*

*P3-R: And I still don't know how I stuffed it up, and I wouldn't go back then after that*

*AC: Yeah, yeah, okay*

*P3-R: So very sensitive... [significant word finding difficulties, attempting to say "with my wife"]*

*AC: Your relationship with your wife? Okay*

P3-R also acknowledged, in his response to Item 18 on the AIQ 'Have you felt isolated?', the tension between wanting his own needs filled and not wanting his wife to have to spend all her time with him:

*Little little bit. Down there [pointing to rating scale]. When [wife's name]'s gone. ...But I want her and hers life to not be inferred [interfered] with from anything by this. And she goes to the gym with the little jumping over thing with the girls and I don't want that to change. And I don't want her running home to look... so... so... I am isolas ... isolating myself so's that her activities are not taken away from her. ... It's really hard because I'm trying really hard to show that I'm not trying. ... I get jealous. I get jealous because I can't do it now. And I don't want her to stop.*

*... She's gotta be free to do that. And I'll turn on the television or whatever. And then I'll get lonely. If that were to happen regularly, then I'd be hurting. But I don't want to hurt her.*

With regard to the non-depressed participants (P1-M, P2-S, P5-D), P1-M's wife was a retired school teacher. She had commented during one session that she thought adapting activities for him was a lot like adapting activities for dyslexic children and she appeared to have a good sense of his strengths and weaknesses. P5-D's wife still had many activities she was independently involved in but was noted to have a good awareness of his comprehension deficits and the fact that he often needed things repeated or explained more clearly. P2-S was divorced but close to her children. She, like Douglas (2012), described deriving strength from her independence and the fact that she was not being treated differently by her family.

**Language intervention, speech pathology and support.** Two participants in particular discussed their strong feelings that speech pathology had contributed to their health-related quality of life, in a similar way to Gould (2015). P2-S described that she believed she would not be coping as positively with her PPA if she had not received the support of her speech pathologist, thorough and honest explanations what her variant of PPA entailed and the opportunity to engage in daily word retrieval exercises. Similarly, P5-D explained that he found daily language practice immensely helpful. He had even independently added several cognitive skill-training and French-language apps to his daily practice: *"I'm always trying to be working on expansion and new concepts ... Whether it's expansion or reviewing, and doing things again, I'm hoping that's what's going to be maintaining."*

### **Influence of Duration of and Reaction to PPA**

Participants' reactions to their diagnosis and the amount of time that had passed since their diagnosis appeared to make a difference to their outlook and mood. Although resilience

was not directly investigated, some individuals did appear to emerge from qualitative comments as more resilient than others.

In the case of P2-S, she described the few weeks post-diagnosis as an intensely difficult time. On some GDS items, she reported that she didn't rate herself as depressed on them "now" but that she would have immediately after diagnosis. She subsequently commented in responses to Item 13; 'Have you felt unhappy?', and 17; 'Have you felt angry?' of the AIQ that she felt PPA had caused her to adopt a 'life's too short' approach to negative emotions, such as in this example:

*Nup. ... I'm not an angry pers... I've never been a very angry person but I ... But I think even more so now ... You can't be bothered ... It's that survivor thing ... Yeah, there's ... There's some benefits in it [PPA].*

She further described her 'fight' against PPA during Item 14 of the GDS 'Do you think that your situation is hopeless?': *"No. We've been fighting this situation for more than a year. And we've achieved so much."*

P1-M also appeared to have moved on from the trauma of diagnosis and entered a period of acceptance. His philosophical outlook can be seen in the following responses from the GDS to Items 11; 'Do you think it is wonderful to be alive now?':

*P1-M: Yes. The option is [laughs - amused] ...*

*AC: Yep? Worse? [laughs with him]*

*P1-M: Erm really ... we grumble in the world, in Australia, y'know when you ... if you've been into India and Africa and places like that. And then you get here ... go back to ... you grumble about very famil- flimmy things, don't you?*

and Item 12; 'Do you feel pretty worthless the way you are?':

*I don't think pretty worthlessly ... but it's erm ... It's not as good as it could be obviously but ... But you've gotta take the cards. ... So I'd I'd ... There's a lot of well*



*... quite a lot of things that these ... I can't do well or, or at all that ... But I can walk, I can see. ... And all of that sort of stuff. ... So, I'd say no.*

In contrast, other participants appeared to have poor resilience post-diagnosis. For example, P6-J quite frequently discussed the fact that 'before' he had been okay but 'now' everything was much harder. "I could do it [reading] before but now it's getting worse and worse and worse. So I just have ... no". Similarly, P4-G expressed his sense of loss, in response to GDS Item 15; 'Do you think that others are better off than you are?': "*Boy that's loaded because ... yes, I have ... I've reached the point that I can't do some things and I'm really ... [unintelligible] ... facing the place that I can't do it, y'know?*"

### **A Multifactorial Account**

In addition to the influence of the individual factors described above, it is likely that many factors are inter-related and cannot easily be teased apart, as has been found previously in stroke-aphasia and dementia (Alzheimer's Society, 2010; Cruice et al., 2003; Hilari et al., 2012; Worrall et al., 2017). As an example, P4-G's financial situation was indirectly affecting his quality of life via his living situation, his wife's unavailability due to working full time and his difficulty in maintaining activities and social support. A multifactorial explanation would also account for P1-M's presentation. Although he missed reading for enjoyment and was aware of the progressive nature of his symptoms, he appeared to be bolstered by the benefits of financial stability, his self-described predisposition to chatty conversation, motivation to keep active, and, willingness to try new activities. The small sample size in this study precluded the ability to investigate the moderating and mediating relationships between factors which affect health-related quality of life but it would seem important for a larger study to investigate contributors to health-related quality of life in PPA using a multifactorial model.

### **General Discussion**

This study collected self-reports of health-related quality of life from six individuals with PPA using four assessments standardised for other neurological populations and by allowing additional qualitative comments. The first aim of the study was to establish what individuals with PPA would report about their health-related quality of life. They reported wide-ranging impacts of PPA on their mood, activities and participation including relationships. The second aim was to determine if any particular factors appeared to influence health-related quality of life in PPA. The results suggested that a number of factors were of influence and these included maintaining enjoyable activities, family support, speech pathology intervention and support, financial situation, time passed since diagnosis and reaction to diagnosis. Aphasia severity, anomia severity, cognitive status and prior history of mental illness were not found to be related to health-related quality of life in this study.

In addition to providing preliminary answers to the two main research aims, this study importantly demonstrated that people with PPA are largely able to self-report on their health-related quality of life, and that a range of assessments designed for other neurological populations can be utilised in PPA. Broader implications for the speech pathology profession and clinical practice will be discussed below.

### **Health-Related Quality of Life in PPA: Emerging Knowledge**

Although this study's results have only been drawn from six participants, these participants exhibited a range of quality of life presentations and each provided a valuable contribution to understanding of health-related quality of life in PPA and what fosters it.

**Self-reports on health-related quality of life in PPA.** A wide range of assessment scores and patterns emerged during study and these were grouped into either activities and participation, or mood, in line with the domains that the chosen assessments probed. Overall, three participants were identified as having depression and tended to have lower scores across

all assessments. Two other participants had very high scores across all assessments but for very different reasons; one because she was genuinely living well with PPA (P2-S) and the other because he appeared to lack insight or comprehension to answer all items appropriately (P5-D).

***Activities and Participation.*** Participants' results within activity and participation domains, as well as their confidence to engage in activities, revealed new insights into the impact of PPA. For example, despite significant linguistic or cognitive impairments, people with PPA may still consider themselves to be confident to make their own decisions.

In contrast, across a number of assessments, the people with PPA reported that they lack confidence to use the telephone, speak to strangers or with larger groups. Avoidance of using the telephone was also described by some of the participants in Cartwright's (2015) study and is predictable given the lack of nonverbal cues and the stress of unexpected questions when talking on the phone.

While Cartwright (2015) found that participants who had intact cognitive skills reported that they still managed the household finances, participants in this sample were more hesitant regarding their ability to manage finances. P1-M and P2-S reported being highly aware that they had difficulties with numbers. Both reported using 'tap and go' payment cards when out shopping, in order to avoid having to deal with change, similar to the way in which Twigg reported using strategies to make shopping easier (Twigg & LaFontaine, 2016).

On both the QOL-AD and GDS, some participants indicated they had reduced memory, which likely relates to their reduced memory for words. Others, meanwhile, made a deliberate choice not to rate themselves poorly on the 'memory' items, such as P2-S who responded to GDS Item 10 'Do you feel that you have more problems with memory than most?', by saying "*Oddly, I think no. Because that's not affected. I mightn't be able to express it because I can't spell it. But I know*". Her comment demonstrates her awareness that

memory and word finding difficulties are not the same and is likely a consequence of her education on the nature of PPA through speech pathology sessions.

Finally, participants reported experiencing very little difficulty completing chores. This is consistent with the fact that PPA initially affects linguistic domains in isolation, rather than activities of daily living (Gorno-Tempini et al., 2011). However, it is interesting that four of the six participants rated themselves significantly *higher* than healthy controls on their ability to do chores on the QOL-AD. It is possible that people with PPA inflate their judgements of non-communication domains, because they judge them in relation to communication-related domains which they are highly aware are deteriorating.

**Mood.** A number of important results emerged relating to mood. One unanticipated result in this study was that two participants (P1-M, P2-S) described a sense of purpose derived from PPA, as it had caused them to reflect and value what they had. P1-M compared his living situation positively relative to individuals living in third world countries and P2-S concluded that she didn't have the time or inclination to be unhappy.

The fact that the sample did not report feeling helpless or hopeless is positive, and suggests that people with various stages of PPA can continue to maintain a sense of hope or agency in their lives, an impression which is also gained from individual accounts of PPA (Douglas, 2014; Summers & Cartwright, 2016; Twigg & LaFontaine, 2016).

**Factors which influence health-related quality of life in PPA.** As outlined above, a range of factors were found to influence health-related quality of life. The inclusion of qualitative comments in this study proved valuable to fully understanding the experience of the individuals with PPA in this sample and has demonstrated the potential value of adopting such an approach in clinical practice to determine what factors may be important to a given patient.

***External factors.*** The value of engaging in enjoyable activities described by the participants in this study is comparable to the experiences recounted by Douglas (2014), Summers and Cartwright (2016), and Twigg and LaFontaine (2016) in their first person accounts, and one of the participants in Cartwright's (2015) study whose wellbeing reflected satisfaction with his activities.

The influence of financial situation on health-related quality of life indicated by participants' comments is not of surprise given that finances will have a cumulative effect on ability to engage in enjoyable activities or access helpful services. It is also related to similar findings in stroke-aphasia (Holland, 2006) and caregiver burden in frontotemporal dementia (Galvin & Schmidt, 2016).

Degree of support from communication partners and family emerged as highly influential, a notion echoed in Summers' (Summers & Cartwright, 2016) personal account. Cartwright's (2015) PPA sample similarly identified close relationships as being the most powerful external buffer for their difficulties. This is likely to have ramifications for how a person with PPA and their family are educated about the role of family and support networks in coping with PPA.

Regarding speech pathology support, some individuals in this sample demonstrated a real eagerness to understand their diagnosis, maintain linguistic skills and feel that they were doing something proactive. This concurs with comments made by Gould (2015) and by Silverman's (2011) survey participants about the value of speech pathology intervention, as well as the finding that individuals in PPA support groups benefit from education based sessions (Morhardt et al., 2017). This is significant and, certainly, establishing the value of speech pathology for health-related quality of life in PPA more objectively is a priority for the field.

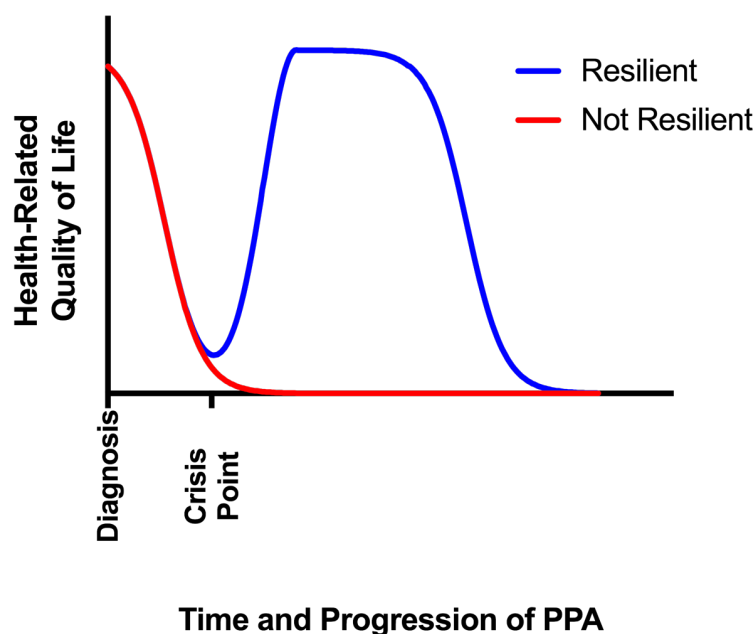
***Time passed since and reaction to diagnosis.*** There was qualitative evidence in this study that the effect of PPA on health-related quality of life was mediated by reaction to and time passed since diagnosis.

A wealth of literature has examined people's reactions to diagnoses of disease and other stressful events. Notably, Hewson (1997) proposed a model for coping with loss of ability. In this model, individuals must come to an awareness of their loss or diagnosis, then adjust to the loss, either by making modifications to the problem itself or lifestyle changes which will better accommodate the problem (Hewson, 1997). Hewson's (1997) model highlights that the threshold above which an event is deemed to be stressful changes based on an individual's personal circumstances and contextual factors.

A similar 'post-diagnosis reaction' model could be conceived, based on: the findings of this study, the comments made previously by other individuals with PPA about being proactive and facing their diagnosis (Douglas, 2014; Summers & Cartwright, 2016; Twigg & LaFontaine, 2016), as well as the intuitions of some clinical specialists (C. Taylor-Rubin, personal communication, August 9, 2016). In this model, there is a post-diagnostic dip in health-related quality of life, or 'crisis point', which then resolves in individuals who are able to gain back a sense of agency and reshape their lives around their diagnosis, such as P2-S, as seen in Figure 6. This concurs with Medina's (2009) report that 64% of people with PPA felt they had a new appreciation for life following diagnosis and also mirrors the findings that positive personal outlook (Cruice et al., 2010) and resilience (Holland, 2006) are influential on quality of life in stroke-aphasia. Ultimately, and despite this initial recovery, health-related quality of life can be expected to deteriorate as linguistic and cognitive skills decline over the longer term. However, a second potential trajectory for the seemingly less resilient individuals whose health-related quality of life does not recover following diagnosis, such as P6-J, is also shown in Figure 6. This second trajectory fits with Cartwright's (2015) finding

that people with PPA are intensely aware of their linguistic deficits and for some individuals this can cause negative emotional reactions and maladaptive coping strategies.

Further exploration of stress-coping strategies or the potential effect of resilience in PPA were unfortunately beyond the scope of this study, however, the results presented here make it clear that individuals with PPA are not all on the same emotional or coping trajectories and that this is an important area which warrants further consideration.



*Figure 6.* An illustration of changes in health-related quality of life over time for resilient and non-resilient individuals with PPA.

### **Applying Emerging Knowledge of Quality of Life in PPA to Clinical Practice**

The findings from this study have relevance for the management of individuals with PPA in clinical practice; both for speech pathologists and the wider multidisciplinary team. The heterogeneous presentation of health-related quality of life in this study's sample highlights that, although diagnostic criteria and knowledge of PPA are more clearly defined

than ever before, each individual with PPA has a unique presentation of quality of life and will require individualised clinical management and patient-centered goals.

Health professionals should be cognisant of the fact that individuals with PPA may still consider themselves to be quite confident at making their own decisions or managing finances despite linguistic and cognitive deficits. Speech pathologists are well qualified to support people with PPA to have a voice and make informed decisions together with their families and multidisciplinary team.

The fact that those individuals with PPA who had supportive and available partners and families were found to have better health-related quality of life, highlights the importance of all clinicians fostering an understanding in families of the value of their involvement in the life and therapies of their family member with PPA.

The finding that fifty percent of the sample had depression on a screener emphasises the need to monitor the mood of people with PPA and to be aware that low mood may have an impact on their ability to engage with interventions. This study has demonstrated that speech pathologists are able to effectively administer the GDS. However, it is important that in multidisciplinary teams it is clear whose role it is to formally assess depression and when to refer on to an appropriate health professional such as psychiatrist or clinical psychologist.

Lastly, the finding that those individuals who maintain enjoyable activities tended to have better health-related quality of life serves as a reminder that health professionals have a large role in facilitating activities and participation. Some individuals may find language-based activities motivating while others may prefer to avoid them. Involvement of occupational therapy may be worthwhile in order to assist people with PPA to adopt new non-verbal activities they can enjoy. This approach has proven successful in the dementia field (Laver et al., 2017; O'Sullivan & Hocking, 2013) and has emerging application for PPA (Morhardt et al., 2017; O'Connor, Ahmed, & Mioshi, 2014).



### **Study Limitations**

There are three clear limitations of this study. First, the small sample size (six participants) precluded the ability to robustly analyse relationships between variables. Larger scale replications of this study including all variants of PPA and more female participants are therefore advocated. Second, the researcher directly administered the health-related quality of life assessments with participants, which, despite the absence of clear hypotheses regarding the impact of PPA on health-related quality of life, has the potential to have unintentionally influenced the findings. These two considerations limit the ability to generalise this study's results to the wider PPA population. Third, the inclusion of individuals with lower cognitive and linguistic scores can be viewed both a strength, as previously indicated, and as a limitation given that some individuals may have answered questions while not fully comprehending their meaning. However, in the context of the paucity of research in PPA, it was felt important to include all individuals in this study as a starting point from which to determine who may or may not be able to self-report on quality of life in PPA. In order to determine the extent to which these participants can nonetheless self-report accurately, future studies may also consider inclusion of proxy-rated quality of life assessments for comparison.

### **Conclusion**

Although supporting quality of life is arguably the role of all multidisciplinary team members in dementia and PPA care, people with PPA are often in the most regular contact with their speech pathologists. Thus, speech pathologists are very likely to be the ones who first observe reduced quality of life in their patients and can build on the profession's growing understanding of quality of life in related communication disorders.

This study has demonstrated that people with PPA are a highly heterogeneous group who exhibit a wide range of health-related quality of life profiles. Individuals with PPA reported adapting to the realities of living with PPA and their futures differently, and each

with a unique set of influences and resources. This highlights the fact that assumptions about the experiences of living with PPA should not be made purely based on language and cognitive functions. The fact that some individuals with PPA are able to maintain good health-related quality of life is tremendously positive and necessitates further investigation into how best to facilitate this in as many people affected by PPA as possible.

This study has also demonstrated that people with PPA without significant semantic impairment are able to self-report on a range of health-related quality of life assessments and, with the assistance of a trained speech pathologist, expand on their experiences, just as has been previously found in the aphasia and dementia fields.

The results of this study are encouraging, yet given the small sample size, future studies would do well to recruit a larger sample of people with PPA that includes all the variants of PPA, and which investigates quantitative measures of quality of life through multifactorial analysis and qualitative findings in greater depth.

Encouragingly, however, there are already emerging and enlightening implications for clinical practice. When a spouse exclaims “Why are they happier now than six months ago?” or a newly diagnosed patient thinks “Why don’t I care anymore?” there are now some answers. The speech pathology profession should endeavour to assist people with PPA and their families to maintain positive mood, activities and participation with an emerging understanding the complexities of psychosocial reactions to PPA and those factors that influence health-related quality of life.

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## Appendices

### Appendix A.

Items for the Quality of Life in Alzheimer's Disease Scale (Logsdon et al., 1999).

1. First of all, how do you feel about your physical health? Would you say it's poor, fair, good, or excellent?
2. How do you feel about your energy level? Do you think it is poor, fair, good, or excellent?
3. How has your mood been lately? Have your spirits been good, or have you been feeling down? Would you rate your mood as poor, fair, good, or excellent?
4. How about your living situation? How do you feel about the place you live now? Would you say it's poor, fair, good, or excellent?
5. How about your memory? Would you say it is poor, fair, good, or excellent?
6. How about your family and your relationship with family members? Would you describe it as poor, fair, good, or excellent?
7. How do you feel about your marriage? How is your relationship with (spouse's name). Do you feel it's poor, fair, good, or excellent?
8. How would you describe your current relationship with your friends? Would you say it's poor, fair, good, or excellent?
9. How do you feel about yourself—when you think of your whole self, and all the different things about you, would you say it's poor, fair, good, or excellent?
10. How do you feel about your ability to do things like chores around the house or other things you need to do? Would you say it's poor, fair, good, or excellent?
11. How about your ability to do things for fun, that you enjoy? Would you say it's poor, fair, good, or excellent?
12. How do you feel about your current situation with money, your financial situation? Do you feel it's poor, fair, good, or excellent?
13. How would you describe your life as a whole? When you think about your life as a whole, everything together, how do you feel about your life? Would you say it's poor, fair, good or excellent?

Appendix B.

Example with the QOL-AD formatted for the current study: One item per page and visuals symbols.

1. Physical health	Poor	✖
	Fair	✓
	Good	✓✓
	Excellent	✓✓✓



## Appendix C.

Items for the Aphasia Impact Questionnaire-21 (Swinburn, 2015).

This week...

1. How easy was it to talk to someone close to you?
2. How easy was it for you to talk to a stranger?
3. How easy was it for you to understand someone close to you?
4. How easy was it for you to understand a stranger?
5. How easy was it for you to write a letter to a friend?
6. How easy was it for you to read a whole story in a newspaper?
7. How easy was it for you to do the things you have to do?
8. Did you have enough positive things to do – things you wanted to do?
9. How were things with friends?
10. How were things with your family?
11. Have you felt frustrated?
12. Have you felt worried?
13. Have you felt unhappy?
14. Have you felt helpless?
15. Have you felt bored?
16. Have you felt embarrassed?
17. Have you felt angry?
18. Have you felt isolated?
19. Some people tell us they feel stupid. I know you are not stupid. But this week.... Have you felt stupid?
20. Have you felt confident?
21. How do you feel about the future?

Appendix D.

Items for the Communication Confidence Rating Scale for Aphasia (Babbitt & Cherney, 2010).

1. How confident do you feel about your ability to talk with people?
2. How confident do you feel about your ability to stay in touch with family and friends?
3. How confident do you feel about your ability to follow news and sports on TV?
4. How confident do you feel about your ability to follow movies on TV or in a theatre\*?
5. How confident do you feel about your ability to speak on the telephone?
6. How confident do you feel that people understand you when you talk?
7. How confident do you feel that people include you in conversations?
8. How confident do you feel about your ability to speak for yourself?
9. How confident do you feel that you can make your own decisions?
10. How confident do you feel that you can participate in discussions about your finances?

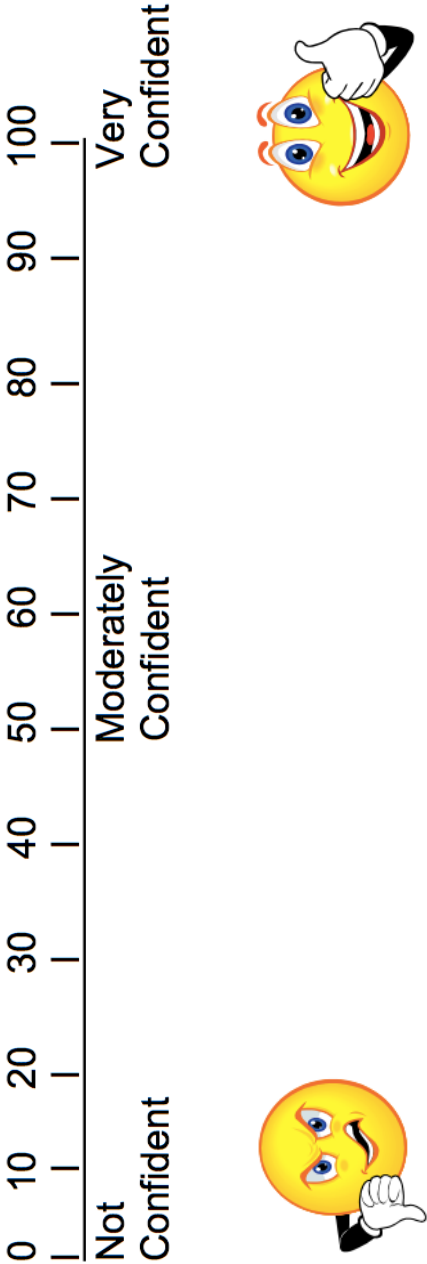
\*[NB Spelling of theater changed for Australian study]

Appendix E.

CCRSA as formatted for the current study with one item per A4 page.

Confidence

How confident do you feel about your ability to talk with people?



## Appendix F.

Singlims\* Statistical Results for Comparison of Participant Scores on QOL-AD scale to Healthy Controls from Cartwright (2015).

Domain of Quality of Life	Current Study (n=6)						HC <sup>#</sup> (n=14)	
	P1-M	P2-S	P3-R	P4-G	P5-D	P6-J	Mean	SD
Physical Health	4 <sup>a</sup>	3	3	1 <sup>b</sup>	3	3	3	0.39
Energy	3	3	3	2	4 <sup>c</sup>	3	2.79	0.43
Mood	3	4	2	3	4	3	3.14	0.66
Living Situation	4	4	3	2 <sup>d</sup>	4	3	3.57	0.51
Memory	2	3	2	1 <sup>e</sup>	3	2	2.71	0.61
Family	4	4	3	2 <sup>f</sup>	4	3	3.71	0.47
Marriage	3	4	4	2 <sup>g</sup>	4	3	3.5	0.52
Friends	3	4	3	2	4	3	3.36	0.63
Self as a Whole	2	3	2	3	4	2	3.14	0.53
Ability to Do Chores	4 <sup>h</sup>	4 <sup>h</sup>	3	2	4 <sup>h</sup>	4 <sup>h</sup>	2.93	0.47
Ability to Do Things for Fun	4	4	2	2	4	3	3.14	0.77
Money	4	4	3	2	3	3	3	0.55
Life as a Whole	3	4	3	2 <sup>i</sup>	4	3	3.36	0.5
Total	43	48	36	25 <sup>j</sup>	49	38	41.36	4.78

\*Crawford, Garthwaite and Porter (2010)

<sup>#</sup>Healthy Controls (HC) data from Cartwright (2015)

<sup>a</sup>Sig higher than HC (t=2.477, p=0.027, estimated effect size ( $z_{cc}$ )=2.564, 95%CI= 1.447 to 3.659)

<sup>b</sup>Sig lower than HC (t=-4.954, p<0.01, estimated effect size ( $z_{cc}$ )=-5.128, 95%CI= -7.137 to -3.107)

<sup>c</sup>Sig higher than HC (t=2.715, p=0.018, estimated effect size ( $z_{cc}$ )=2.814, 95%CI= 1.614 to 3.994)

<sup>d</sup>Sig lower than HC (t=-2.974, p=0.01, estimated effect size ( $z_{cc}$ )=-3.078, 95%CI= -4.350 to -1.788)

<sup>e</sup>Sig lower than HC (t=-2.708, p=0.02, estimated effect size ( $z_{cc}$ )=-2.803, 95%CI= -3.979 to -1.607)

<sup>f</sup>Sig lower than HC (t= -3.515, p<0.01, estimated effect size ( $z_{cc}$ )=-3.638, 95%CI= -5.107 to -2.153)

<sup>g</sup>Sig lower than HC (t=-2.787, p=0.015, estimated effect size ( $z_{cc}$ )=-2.885, 95%CI= -4.089 to -1.660)

<sup>h</sup>Sig higher than HC (t=2.199, p=0.047, estimated effect size ( $z_{cc}$ )=-2.277, 95%CI= 1.254 to 3.276)

<sup>i</sup>Sig lower than HC (t=-2.628, p=0.02, estimated effect size ( $z_{cc}$ )=-2.720, 95%CI= -3.868 to -1.551)

<sup>j</sup>Sig lower than HC (t=-3.307, p=0.03, estimated effect size ( $z_{cc}$ )=-3.423, 95%CI= -4.815 to -2.013)