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<RRH>*Perceived signs of PPA pre-diagnosis*

Research Report

Spousal recollections of early signs of primary progressive aphasia

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Abstract

Background: Although primary progressive aphasia (PPA) is characterized by progressive loss of language and communication skills, knowledge about the earliest emerging signs announcing the onset of this condition is limited.

Aims: To explore spousal recollections regarding the earliest signs of PPA and to compare the nature of the earliest perceived symptoms across the three PPA variants.

Methods & Procedures: In-depth interviews focusing on the earliest signs of illness onset were conducted with 13 spouses whose partners were diagnosed with PPA. The earliest recollections and observations described by the spouses were analyzed and coded according to the DSM-5 criteria for a mild neurocognitive disorder. These data were then compared across and within each of the three PPA variants.

Outcomes & Results: Spousal retrospective accounts indicated the three PPA variants (semantic, logopenic and non-fluent) had a signature profile announcing illness onset. Changes in social cognition presented in all three variants of PPA, but at different points in the illness trajectory. In particular, the findings suggest the possibility that PPA initially presents as subtle changes in social cognition for semantic variant PPA (svPPA) and logopenic variant PPA (IvPPA) rather than overt language impairments as defined in the current diagnostic criteria.

Conclusions & Implications: Understanding the nature of symptoms perceived in the earliest stages of PPA has potential to inform earlier and accurate diagnosis and interventions to assist those living with the illness.

Keywords: earliest signs, dementia, primary progressive aphasia (PPA), qualitative research, spouse.

<A>What this paper adds

<B>*What is already known on the subject*

The diagnostic criteria specify that PPA should only be considered in the context of relative preservation of cognitive–behavioural abilities for the first 2 years of the condition, though little research substantiates this 'two-year rule', or provides corroborative narrative accounts from those who have experienced and/or witnessed first hand the emergence of PPA.

<B>*What this paper adds to existing knowledge*

This paper provides clinicians with crucial insights into the earliest signs of emerging PPA. In particular, the retrospective accounts of the spouses in this study indicated that changes in social cognition presented as one of the earliest signs of PPA onset for svPPA and lvPPA, and that each of the three PPA variants may have a signature profile announcing illness onset.

<B>*What are the potential or actual clinical implications of this work?*

The delineation of early signs of PPA is important to facilitate timely diagnosis with support and intervention during the early illness phase, as well as for inclusion in clinical trials for potential disease-modifying treatments.

<A>Introduction

Primary progressive aphasia (PPA) is a neurodegenerative disorder characterized by an insidious and progressive loss of language abilities, though little is known about the earliest presenting symptoms. It falls under the collective banner of frontotemporal dementia (FTD), which is divided into two subtype forms: behavioural variant FTD (bvFTD) and PPA. PPA has three variant forms: semantic (svPPA), logopenic (lvPPA) and non-fluent (nfvPPA) (Gorno-Tempini *et al.* 2011, Mesulam *et al.* 2012), each with a specific profile of speech and language impairments. The consortium classification of PPA (Gorno-Tempini *et al.* 2011) was devised by a panel of experts, predominantly neurologists, for use by clinicians (such as geriatricians, psychiatrists, neurologist, neuropsychologists and speech–

language pathologists). The classification specifies the diagnostic criteria for PPA by identifying the salient speech, language and cognitive characteristics commensurate with each variant form of PPA. The criteria included in the classification are based on cross-sectional rather than longitudinal observation data (Harciarek *et al.* 2014). Because PPA is classified as a major neurocognitive disorder (NCD), by default the diagnosis should only be considered when the language-related deficits impinge on the individual's capacity for independence in their everyday life (*Diagnostic and Statistical Manual of Mental Disorders—5* (DSM-5); American Psychiatric Association (APA) 2013). The diagnostic criteria also stipulate that a diagnosis of PPA can only be considered in the context of relative preservation of cognitive–behavioural abilities for the first 2 years of the condition (Mesulam 2001, Mesulam *et al.* 2012). However, little research substantiates this arbitrary 'two-year rule' or provides corroborative narrative accounts from those who have experienced and/or witnessed first hand the emergence of PPA. FTD disorders are often mistaken for psychiatric and other neurological conditions (Hallam *et al.* 2008, Rosness *et al.* 2016). In addition, a 3–4-year lag from initial symptoms to confirmed diagnosis is typical (Mendez *et al.* 2007, Morgan *et al.* 2014, Speechly *et al.* 2008), often contributing to significant stress and burden during this time for those living with the condition. Thus, delineation of early signs of PPA is important to facilitate timely diagnosis with support and intervention during the early illness phase, as well as for inclusion in clinical trials for potential disease-modifying treatments.

In recent years there has been an increasing focus on identifying early clinical symptoms of decline in various neurodegenerative conditions, such as Alzheimer's disease (AD) (Dubois *et al.* 2014), Parkinson's disease (PD) and dementia with Lewy bodies (DLB) (Gaenslen *et al.* 2011, Fujishiro *et al.* 2013, McKeith *et al.* 2016). The latest edition of the DSM-5 (APA 2013) introduced the concept of mild NCD to promote early identification and treatment of cognitive decline before difficulties become too severe and progress to the status of a major NCD (i.e., 'dementia'). DSM-5 specifies six key neurocognitive domains with working definitions and examples of impairments in daily activities to assist clinicians in differentiating between mild versus major NCD. These NCD domains include complex attention, executive function, learning and memory, language, perceptual–motor, and social cognition.

Mild NCD shares many features of mild cognitive impairment (MCI) (Petersen *et al.* 2014), and is thought to be a transitional state between normal ageing and dementia, though not for all. DSM-5 specifically acknowledges the importance of obtaining 'evidence' from the individual, their close informants and clinician observations as vital components of the diagnostic work-up. Indeed, the clinical features characterized as indicative of emerging PD and DLB were derived by asking patients at the time of diagnosis about their experiences of symptoms in the years prior to presentation (Gaenslen *et al.* 2011, Fujishiro *et al.* 2013). In a 5-year follow up study of 358 subjects, Taragano *et al.* (2009) found that individuals who displayed emerging behavioural/personality changes (defined as mild behavioural impairment—MBI) without prominent cognitive decline were more likely to develop bvFTD than AD. Ismail *et al.* (2016) acknowledged this strong association of MBI with emerging bvFTD symptomology and proposed including MBI into the MCI framework because they can co-occur and together may signify a higher risk of dementia onset. In summary, the current literature reflects growing acknowledgement and motivation to detect early incipient neurodegenerative illness to target potential pharmacological interventions as well as to provide non-pharmacological support for those living with degenerative NCD.

Language deficits are considered to be the earliest signs of PPA above all other cognitive domains (Mesulam *et al.* 2012), yet little research has investigated the specific pattern of early symptom emergence in PPA as a dementia disease entity. Given that linguistic impairments develop as a consequence of neuropathology located in the frontotemporal cortical structures, it is conceivable that affected individuals may also exhibit subtle changes in social cognition during the early-onset phase prior to meeting the criteria for PPA diagnosis. Social cognition refers to the complex processes by which individuals detect, perceive and process how people are feeling or thinking, and this then influences their interpersonal responses within a social context (Forbes and Grafman 2010, Natelson-Love *et al.* 2015).

Symptomatic overlap between svPPA and bvFTD is recognized in the literature (Ahmed *et al.* 2015, Kamminga *et al.* 2015, Kertesz *et al.* 2007, Rosen *et al.* 2006). Social cognitive difficulties characteristically seen in bvFTD also emerge early in svPPA, but are believed to be exhibited *after* the onset of language deficits (Lui *et al.* 2004, Modirrousta *et al.* 2013, Rosen *et al.* 2006, Shinagawa *et al.* 2006). In nfvPPA the earliest symptoms are reportedly difficulties with spontaneous speech fluency, though pronounced behavioural signs are reported in the advanced phase of illness progression (Marczinski *et al.* 2004, Rosen *et al.* 2006). No information about the earliest signs of IvPPA was located in the literature. Solberger *et al.* (2011) compared interpersonal communication changes in bvFTD, svPPA and AD. Their results revealed specific changes in 'personality' at the very mild stage of the disease onset and these traits changed with disease progression. These authors reported that like bvFTD, individuals with svPPA may present as less extraverted and less warm in interpersonal interactions, however the nature and severity of aberrant behaviours remain more pronounced in the bvFTD throughout the illness trajectory. Singh *et al.* (2015) compared the prevalence and nature of neuro-behaviour symptoms across the three variants of PPA and progressive apraxia of speech (AOS). Their findings indicated that during the initial stages of confirmed PPA, mood changes such as anxiety were present, whereas behavioural symptoms such as apathy were associated with early onset AOS. Individuals with svPPA were also reported to display features of disinhibition, whereas individuals with nfvPPA were more likely to display appetite changes. Although these studies had methodological limitations (in terms of data collection methods and/or small sample sizes), the findings lend some support to the notion of changes in social cognition as possible early signs of PPA onset.

Given that communication 'is first and foremost about emotional connections and relationships' (Kindell *et al.* 2014: 409), it is possible that presenting signs of PPA may be perceived as a change in 'personality' by others who have close relationships with the individual. Krueger *et al.* (2000) defined 'personality' as the way an individual relates to the world. It incorporates predictable behavioural response patterns to a range of different environmental stimuli that close others have learnt to expect from an individual (Cipriani *et al.* 2015). Spouses typically share a close emotional bond established over time. As a consequence, their intimate relationship affords them person-specific knowledge, sensitivity and opportunity to detect any subtle changes in their partner's personality, behaviour and communication (Pozzebon *et al.* 2016). In our instrumental case study of a spouse whose husband was diagnosed with svPPA, the spouse reported explicit examples of changes in social–cognitive behaviour about 10 years *before* he exhibited prominent linguistic difficulties (Pozzebon *et al.* 2017). By the time, this couple presented for diagnostic work-up, social–relational issues and carer stress were well established. This case study highlighted the possibility that MBI or perceived 'personality changes' may present prior to the onset of PPA linguistic deficits. In summary, efforts to characterize the earliest symptoms of PPA could facilitate more timely and accurate diagnosis of illness onset, as well shed light on the resultant psychosocial adjustments imposed on individuals living through the emergence of PPA symptomatology.

<B>*Aims*

The aims of this qualitative study were twofold: (1) to explore the spousal recollections that for them signalled the earliest signs of PPA; and (2) to compare the nature of these earliest perceived symptoms across the three variants of PPA.

<A>**Method**

This project was part of a constructivist grounded theory inquiry (Charmaz 2006) exploring the lived experiences of spouses with partners diagnosed with PPA. During the analysis of the large data set, it emerged that all 13 spouses gave detailed accounts about their initial impressions and evolving concerns regarding behaviour changes displayed by their partner, often several years before a confirmed diagnosis. These data coded as spousal participant recollections of earliest signs of PPA are the focus of this study.

<B>*Recruitment process*

All spouses whose partner was diagnosed with PPA at the outpatient memory clinic of a large metropolitan health service in Melbourne were invited to participate. The diagnosis of PPA was established in accordance with the PPA consortium criteria (as outlined by Gorno-Tempini *et al.* 2011) via collaborative agreement of the multidisciplinary team. The diagnostic team consists of an aged care medical specialist (either a geriatrician or a psycho-geriatrician), speech–language pathologist and/or neuropsychologist. Recruitment of participants was in accordance with hospital and university ethics committees' approval stipulations.

<B>*Research design and data collection*

Because little is known about the spousal experience of living with a partner with PPA, a qualitative research methodology was selected. Given that the intent of the study was to understand the participants' world rather than predict, control or prove it, a constructivist grounded theory approach was considered most appropriate (Birks and Mills 2011, Charmaz 2006, Finlay 2006, Strauss and Corbin 1990). Participant narrative data were obtained via one-to-one, in-depth interview with each participant. The participants were asked open-ended and non-leading questions to encourage unstructured description of their personal experiences, such as 'I really want to know how it has been for you', 'Tell me about …'. See appendix A for examples of questions posed to the study participants.

Throughout the interview, the researcher only commented to seek clarification on issues raised, or made short remarks to maintain the flow of discussion (Charmaz 2014). This approach enabled issues of importance to be told by the research participants, giving them ample opportunity to explain their experiences including observational details of their partner's earliest signs of illness onset. Initial open coding to the category 'Initial/early signs' referred to the first time the spouse noticed a substantive change from the partner's previous state, rather than longstanding character–personality traits (Shinagawa *et al.* 2006). The interviews lasted 60–90 min and were audio recorded and transcribed verbatim.

<B>*Data analysis*

All spousal quotes relating to the partner's earliest signs that alerted them to a substantive change in status were collated for each of the PPA variants. These earliest recollections and observations described by the spouses were then further sub-coded according to the criteria of the six cognitive domains of DSM-5 (APA 2013) for mild NCD, to explore differentiation between the three variant forms of PPA. The six DSM-5 cognitive domains are: complex attention, executive function, learning and memory, language, perceptual motor, and social cognition (APA 2013: 593–595). Table 1 outlines the DSM-5 criteria of mild NCD for each of the NCD domains.

<tab 1>

The earliest signs as described by the spouses were then compared within each PPA variant group, with particular attention to the nature and sequence of language disturbances relative to the other cognitive domains. Each of the participant's quotes was independently analyzed and assigned to one of the six most suitable DSM-5 neurocognitive domains by the first and second authors. Agreement was reached on the majority of quotes, with further discussion required on one-fifth of the quotes. Where differences occurred the authors jointly reviewed the participant's quotes, the researcher's field and memos notes, and the DSM-5 cognitive domain definitions until 100% agreement was reached. Finally, to show the reader that the analysis was grounded in the spousal experience, illustrative quotes from the interviews are presented.

<B>*Demographic details of the spousal participants*

A total of 13 spouses participated in the study approximately 1–7 years after their partner's confirmed PPA diagnosis. All but one were female (12/13) and ages ranged between 54 and 82 years (mean ages of 70.9 years). Only one of the participants was employed at the time of the study; all others had retired from the workforce. The spousal participants were previously employed in a wide range of occupations from home duties to sales and managerial and professional roles (Australian and New Zealand Standard Classification of Occupations (ANZSCO) 2013). All participants were in a long-term relationship with the partner, ranging from 22 to 57 years (mean of 45.9 years) (table 2).

<tab 2>

<B>*Demographic details of their partners diagnosed with PPA*

All partners had a confirmed diagnosis of PPA (six svPPA, five lvPPA, two nfvPPA), and were male with one exception. They ranged in age from 61 to 87 years (mean age of 74.8 years). Only one was employed at the time of the study, while all others had retired. Compared with the spousal participants, the partners were generally employed in higher-level occupations according to the ANZSCO (2013) classification (four in management, two in professional roles, five in technical/trade, one in a clerical position and one in a labourer occupation). The time from earliest symptom as reported by their spouse to confirmed diagnosis of PPA ranged from 1 to 10 years, but also varied significantly between the three variant forms of PPA (svPPA: 2–10 years, lvPPA: 2–5 years, nfvPPA < 1 year) (table 2).

<A>**Results**

Analysis of the in-depth interview data revealed discernible patterns of earliest perceived signs for each PPA variant. These key findings are presented with participants' excerpts, including information about:

- the sequence of NCD presentation for each variant form of PPA;
- the most to least common earliest signs of PPA;
- the nature of the initial language difficulties;
- the time from earliest signs (as recognized by the spouses) to confirmed diagnosis; and
- the trigger reason for seeking professional advice.

To ensure confidentiality, pseudonyms are used throughout the reporting of participants' responses, including names of people and places.

<B>*Semantic variant PPA (svPPA)*

Six participants had a partner diagnosed with svPPA. All the spousal participants reported changes in social cognition presenting concurrently with language difficulties that for them signalled the start of illness onset. Initial recognition of decline was experienced by the spouses as ‘puzzling’ and distressing, particularly due to the ‘unusual’ nature of the semantic processing and word-retrieval difficulties in combination with unexpected ‘personality change’ considered to be to be out of character for their partner. This bewildering realization of changing circumstances was poignantly captured by a spouse describing a situation when their daughter drove her husband’s car on a family outing:

> And then we were going to go down to Wanga [a town] to see the movies because it was a real rainy day. ‘We’ll take your car dad.’ ‘Yeah.’ Well then coming back we got in a deluge of rain. It was just terrible. And she was asking him where the wipers were. He didn’t know the windscreen wipers. He couldn’t tell her. She had to pull over to the side of the road, and he he really lost it. So, from Christmas in 2014—all of this behaviour (i.e., sensitivity to noise, decreased empathy towards others, apathy). We couldn’t, we couldn’t make any sense of what he was saying. He was just yelling at both of us. ‘You, you’re driving too fast. That’s why the windscreen wipers won’t work.’ She said, ‘I don’t know where they are. Could you explain them to me?’ Then she asked me. Well, I don’t know anything about a car. Because I’ve NEVER driven. So, ah … so we sat there for about a half an hour before she got the manual out.

These spouses described an increasing awareness of their partner’s receptive and expressive language difficulties, specifically their partner’s inconsistent ability to understand common everyday words such as ‘coffee’ or ‘umbrella’. Making sense of these language processing difficulties at the single-word level was extremely perplexing and distressing for them.

> Oh, I can always remember this one instant when I thought, ‘Oh, there is something really not right here.’ Umm, he was going to go down the street and he came in and said, ‘Oh where’s

> the ….' And he's using all the hand movements to tell me what it was. And I thought, oh, he always wears a cap. I said, 'Oh you've got your cap on. What do you mean?' And in the end, it worked out to be … the umbrella. And I thought oh dear! This isn't right. And it just progressed from there.

Emotional withdrawal was described by all svPPA spousal participants as occurring parallel with the decline in language abilities. These spouses explained how their partners had unexpected 'mood swings' and completely 'switched-off' emotional connection with them and close others. This emotional 'disconnect' was experienced in various aspects of their relationship, including reciprocal conversations, sexual intimacy and sharing joint interests.

> But no, there was no physical side at all. Our chatting together also just stopped. I'd come home and say, 'Oh, Lori did this or the kids did that.' He'd just didn't seem as focused or as interested. He seemed to be just not as interested in the family. He just seemed to be in his little world. … He never took my hand when we walked down the street any more. Before … he was always happy to do things with me.

Four svPPA spousal participants reported perceptual motor processing difficulty as among their earliest initial observations of illness onset. This difficulty was always described as episodes of their partner becoming inexplicably 'lost' in a very familiar environment. For a moment in time their partner appeared to be unable to recognize previously familiar landmarks, such as the street where they lived, their local shopping centre and/or their workplace. For example, one participant explained her husband had worked at a food market for over 20 years—and yet he 'got lost'. This incident occurred about 3 years prior to the svPPA diagnosis and was very disconcerting for both of them:

> I said to him go buy some bread rolls and I'll go to the chemist. He went to buy rolls and then he doesn't come back to the car. It was half an hour … 35 minutes waiting. I saw him going this way up the street … and coming back to the same spot. I was screaming, 'What's wrong with you? Don't you know where the car is? What happened?' Of course, I start crying. Until now I didn't know … about … the dementia!

Two svPPA spousal participants described early difficulties with maintaining dual attention on tasks, such as driving and talking:

> He had a single mind just for that thing. He'd not talk about anything else except where you are going. And if you made any other comments, he got confused. 'We are now going to the concert.' He would say, 'That's where we are going, and that is all we're doing.'

One spouse described sensitivity to loud noise (hyperacusis) as one of the earliest signs that signalled 'the turning point' of indisputable change in her husband's behaviour. His negative reaction to his grandchild's shrill cries of excitement when unwrapping presents was completely out of character for him. He had previously enjoyed and relished the 'loud play' of interacting with their six young grandchildren:

> So on Christmas day we went to Nancy's. The kids are that excited. And Danny [grandson] got the first present. And he wanted these particular shoes—the Star Wars things. And when he opened them. He was SO excited. 'Oh Nan! It's the best thing out.' Well, he was really loud. The next minute I realised that Joe [husband] wasn't there. Next minute I saw our car drive off. And he was gone for three hours. I was frantic. I was ringing and he never answered his phone. For me that was the turning point. It was also when the anger started. He can't stand noises like that.

All six participants reported that the greatest issue for them was dealing with the partner's behavioural issues rather than the language-related difficulties. These difficulties with behavioural issues were experienced from the earliest onset of illness and became worse over time with progression of PPA:

> I think it's the behaviour is more of a … for me it's a battle … particularly social behaviour. The speech … oh we can laugh about it but we can cope with that. And I have the patience of

> waiting and … stop and just wait until he finds the words and he is communicating what it was.

Despite the perplexing and emotionally disturbing behaviour exhibited by the partner over a number of years, these couples mainly sought professional advice when the language difficulties became pronounced and were exhibited during every-day interpersonal interactions with other people. The time from initial symptom onset to a confirmed diagnosis varied widely in the svPPA group, ranging from 2 to 10 years. The point in time at which advice was sought did not appear to be shaped by the occupational background of the spouse. The findings relating to the svPPA group are presented in table 3.

<tab 3>

<B>*Logopenic variant PPA (lvPPA)*

Five spouses of partners diagnosed with lvPPA participated in the study. They all reported changes in social cognition as the earliest presenting symptoms preceding the language difficulties. These behavioural changes were described differently in comparison with those observed by spouses in the svPPA group. Reduced talking and cessation of emotional intimacy was the most prominent earliest signs of lvPPA, especially obvious in a social group setting. These spouses described their partners as ‘withdrawn’, ‘going into himself’ and ‘shutting off’. Their partners displayed increased introversion and reduced interpersonal interaction with them and close others in their shared life. Unlike the svPPA group, however, the appearance of mood swings was not a feature. Social awkwardness, ‘apathy’ and lack of interest in people was described by all the spouses:

> I noticed, in August 2014 before [2.5 years before the diagnosis] when we were on a cruise to the Baltic and it was a 30-day cruise. And he wasn’t talking much with his friends on that August Cruise. And one of my friends said to me, ‘Is Rob okay?’ And I said, ‘I don’t know. It’s weird. He doesn’t seem to be himself.’ Look, he didn’t talk! I guess that’s the hardest thing when you sit at a table with people or people come up and he just sits there and doesn’t talk. And you’re got to talk. Yeah, I can sit there and talk—like to help your husband to integrate a bit. He didn’t join in the conversation and, if anything, you sort of feel

> embarrassed. Embarrassed, but you know the man is trying to talk to him and he just doesn't answer.

These spouses all described how they noticed their partner 'just switched off' for some unknown reason, leaving them feeling 'lonely' and 'hurt'. Their partners were perceived as insensitive to the reaction and consequences of their actions:

> Oh no, no chatting. No chatting. We had a double bed and he put the blankets down [laughed] along the way. I reckon it could be twelve months [before the onset of language symptoms]. It was just strange. We never had sex from there on. Never. I was pretty crushed that he did that to me. And Carl [partner's best friend], he switched off him too. Then Bert [another close friend] … he seemed to find a reason to turn off him too. All his closest allies, … He switched off the people close to him.

As indicated in the spousal quote above, all five spouses reported very early cessation of emotional and sexual intimacy with them. This was perceived as a significant change in their previous spousal relationship:

> At first it probably didn't seem like that he would hug me or kiss me, but probably not as … he didn't and doesn't integrate as much terms of intimacy.

Subtle difficulty with complex attention was another less frequently mentioned earliest sign. This difficulty was described in terms of previously routine activities taking longer to execute, and errors being made—which the spouses thought was highly unusual:

> He had always done ALL the family's finances. All the probates, everything. All the extended family. Then suddenly he wasn't. I had to check things with him because he was less sure. And that's when I really noticed.

‘Forgetfulness’ and compromised working memory were also observed early:

> You could see it in his eyes. I’d be talking to him and then you can see that he’d just drifted off. And I’ll say, ‘Oh John! Are you listening?’ And it would just snap him back to where he was. His concentration span … very short. And his short-term memory was really—almost gold fish like. It’s there and it’s gone.

All five spouses in the lvPPA group reported that social–cognitive changes occurred about 1–3 years prior to their recognition of expressive language difficulties. The nature of the language and word-retrieval issues were different to that described by the svPPA spouses, and were commonly described as difficulties with ‘the thinking and the talking’ as well as ‘that level of difficulty getting the word out’. For one spouse, the word-finding difficulties became a serious safety issue:

> As you know he used to fly small planes, and I used to do the navigating. And Australia is not sign posted from the air. It looks exactly the same. A main road, a railway line and little towns [laughs]. And he used to have difficulty … that’s when I first realised the significance of it, of exactly remembering words when he was on air to the ground where we had been … and he couldn’t say something like, say it was Albany. But that’s when I first noticed that there was an issue for him. … So, it was a memory for a name.

The majority of lvPPA spousal participants had approached their local doctor to discuss the ‘peculiar’ earliest behaviour symptoms. Unfortunately, all five spouses expressed concerns about their partner were dismissed as insignificant and inconsequential, and the spouse usually felt foolish for raising the topic:

> With Ron after that one episode, you know when we’re in the air flying around in the cloud. I did speak to our local doctor who then did arrange for him to have a CT [computed

> tomography] brain scan, and there it stopped. There was never a question there after that you see. And yet I was right [laughed].

No additional advice or assistance was offered to these spouses during this early pre-diagnosis phase. It took 4–5 years for their partner's social behaviour and language issues to become much more apparent, and it was then that further medical opinion was sought for a diagnostic work-up. The lvPPA group findings are presented in table 3.

<B>*Non fluent variant PPA (nfvPPA)*

The nfvPPA group comprised two spouses, and both individuals reported speech–language difficulties as the earliest presenting sign. They experienced difficulty deciphering their partner's verbal output:

> My brother would say, 'Why are you looking at him like that?' And I would go up close to figure out what John [her partner] was saying. So, in a group of people I would be able to, not interpret but know what he was saying. He wasn't … he wasn't articulating properly but it didn't dawn on me. I was looking at him to try to work out the words. It was only 1 or 2 words then. … It wasn't like it is now.

Increasing difficulties with sentence construction was also acknowledged as one of the earliest signs of nfvPPA.

> The words were all over the place. Even to go down the street, it would be, 'Go street.' Things weren't linked up properly. That's when I started to pick things up then. 'Oh, what's going on here?'

Both spouses reported an interval of less than 1 year from their earliest observation of speech–language difficulties to obtaining a confirmed diagnosis of nfvPPA. The reason given for seeking medical opinion was the obvious speech impediments exhibited by their partner. Both participants reported that subtle social cognition issues emerged about 1–2 years post-diagnosis. One spouse expressed dismay that medical personnel dismissed their expressed concerns related to social behaviour changes and challenges exhibited by their partner during subsequent medical review appointments. The nfvPPA group findings are presented in table 3.

<A>**Discussion**

The findings of this qualitative study indicated that each variant form of PPA may have a signature profile announcing early illness onset. Also, speech–language difficulties were not perceived by the spousal participants as the primary signs announcing illness onset across all forms of PPA as reported in the current literature (Gorno-Tempini *et al.* 2011, Mesulam *et al.* 2012). The spousal retrospective accounts revealed that changes in social cognition presented as one of the earliest signs of decline specifically for svPPA and lvPPA. Although these social–cognitive changes were similar in nature, these difficulties became apparent at different points in the trajectory of each variant forms of PPA. For svPPA alterations in social cognition and language difficulties presented concurrently, for IvPPA social cognition changes preceded the language difficulties, whereas for nfvPPA the speech and language difficulties preceded social cognition changes that emerged 1–2 years after the diagnosis. These findings highlight the necessity for clinicians actively to pursue open and non-leading discussions with the individual and their significant–close others to determine if they have experienced or have observed any changes in social–cognitive–communication abilities during the diagnostic work-up (as promoted in the DSM-5), because emerging MBI may indeed signify a higher risk of PPA (Ismail *et al.* 2016, Taragano *et al.* 2009).

Given that PPA is a neurodegenerative condition that causes the gradual decline of language and communication skills, by default these changes impact on the individual's interpersonal interactions with others. As indicated by the results of this study, spouses are ideally positioned to provide astute observational information about any changes exhibited by their partner. All 13 spousal participants gave detailed accounts of various interpersonal situations in which their longstanding partner either behaved awkwardly or seemingly out of character for several years prior to a confirmed diagnosis. Spouses and close others have person-specific knowledge that enables them to detect any subtle changes in their partner's personality, behaviour and communication (Pozzebon *et al.* 2016). The spousal descriptions and context-specific examples of what their partner is 'doing' are likely to provide clinicians with vital insights to triangulate this information with findings obtained from formal assessments, as well greatly assist in earlier identification of an emerging NCD, or indeed a variant form of PPA. Supporting affective and interpersonal communication within spousal relationships should also be a primary consideration for intervention, particularly because research indicates that individuals diagnosed with PPA usually remain living at home and are cared for by their spouses well into the advanced stage of illness (Harciarek *et al.* 2014).

The DSM-5 classification and the PPA diagnostic criteria (Gorno-Tempini *et al.* 2011) do not provide explicit details to inform clinicians about how social–cognitive–communication changes are perceived and interpreted from the perspective of close others (e.g., the spouse) within the context of their pre-existing relationship. Indeed, the revealing content of the spousal quotes also act as reminders that changes in an individual's social–cognition and speech–language skills have significant ramifications for sustaining mutually satisfying interpersonal relationships. Such interpersonal consequences can clearly be expected as humans are innately social beings and communication is primarily about connections and relationships (Kindell *et al.* 2014). These spouses shared very personal accounts about how their partner gradually displayed increasing episodes of out-of-character 'behaviours' that had emotional consequences for them triggering feelings of anxiety, resentment, bewilderment and a growing sense of emotional detachment from their partner. Appreciating this emotional turmoil from the perspective of close others such as spouses is essential for clinicians seeking to provide efficacious therapeutic advice and intervention for those living with the reality of PPA. We need to know what they think in order to understand why they behave in the way that they do (Minichiello *et al.* 2008). In essence, these research findings not only challenge the long-held view of PPA as an illness entity that mainly impacts on receptive/expressive language abilities (at least in the initial years). There is a genuine need to empower individuals living with PPA to navigate the complexities of daily interpersonal interactions and social situations (Natelson-Love *et al.* 2015) starting from the earliest signs of illness onset.

From an early identification perspective, health professionals need to exercise diligence and actively listen for relevant issues and concerns raised by the individual, their spouse and/or relative. As demonstrated by the spousal participants' verbatim quotes, everyday-colloquial language was used to reflect a range of cognitive symptoms. Examples included use of the word 'forgetful' to describe both word-retrieval difficulties and deficits in working memory, 'personality change' to refer to emotional withdrawal and poor interpersonal engagement, and 'getting lost' to cover object-recognition issues and spatial disorientation. The onus is clearly on health professionals to interpret informant's lived experiences and descriptive narratives in order to differentiate and pick up on 'clues' to expose early signs of possible decline. Singh *et al.* (2015) explained that very early symptoms of NCD may be easily missed by the administration of questionnaires and scales, because these tools usually seek to measure symptoms by asking one question per symptom domain. Thus, quantitative measurement instruments 'may not necessarily capture the nuances of some particularly complex symptoms, particularly during the earlier stages of the illness' (236). It may be particularly difficult to determine with limited contextual information if the observed behaviour is due to mild depression/anxiety as a consequence of the language impairment, or indeed a manifestation of the NCD (Modirrousta *et al.* 2013, Singh *et al.* 2015). It is also acknowledged that behavioural–neuropsychiatric symptoms in FTD disorders are not as readily and reliably identified and captured by available neuropsychological measures and generally available neuroimaging techniques, such as CT and standard magnetic resonance imaging (MRI) (Hallam *et al.* 2008, Mesulam *et al.* 2012, Welsh-Bohmer 2008). Findings of this study further highlight the importance of obtaining qualitative information to identify emerging symptoms of PPA. Pursuing alternative yet practical methods of quantifying changes in 'personality' and 'behaviour' (i.e., social cognition) may assist clinicians in identifying the earliest signs of PPA illness, thus reducing misdiagnosis and allowing for earlier therapeutic interventions. The development of a structured interview and/or questionnaire for use with close others may be beneficial to guide clinician discussions about early-onset cognitive–communication changes as well as highlight useful strategies to deal with them. To this end, we note that a study focused on close others (predominantly spouses) using the La Trobe Communication Questionnaire (Douglas *et al.* 2000) has recently been found to be sensitive to social communication changes in partners with amyotrophic lateral sclerosis (Fisher *et al.* 2017).

<B>*Limitations*

Several limitations apply to this study. First, findings were based on spousal participants' recall of their initial impressions of illness onset that occurred several years earlier. The retrospective nature of the descriptions means they may have been influenced by experiences over time as well as by the partner's current cognitive–behavioural symptoms. Second, the sample size was small and varied across the PPA variants, potentially compromising the generalizability of the results to the wider PPA population. Third, spouses in the study were mainly women (12:1). It is well recognized that men are more difficult to recruit to health studies (Oliffe and Thorne 2007) and especially to qualitative research that explores emotionally sensitive topics via interview (Affleck *et al.* 2013, MacDonald *et al.* 2010). Future research may need to consider alternative methods of data collection to ensure equal gender representation. Notwithstanding these limitations, the study has highlighted the possibility that language issues may not be the primary presenting deficit across all variant forms of PPA. The results also emphasize the importance of obtaining qualitative information about cognitive–behaviour changes from those closest to individuals experiencing subtle to mild forms of neurocognitive decline. It is anticipated that this spousal perspective will influence further therapeutic endeavours and future research inquiries.

<A>**Conclusions**

This study sought to explore the earliest signs of PPA from those who witnessed its emergence. The findings suggest the possibility that PPA initially presents as subtle changes in interpersonal–relational contexts for svPPA and IvPPA rather than overt receptive and expressive language impairments. The nuances revealed through personal narratives illustrate the challenges associated with early identification, particularly as very early signs of PPA are unlikely to be easily captured in standardized clinical assessments, scales and questionnaires. Understanding the nature of symptoms perceived in the earliest stages of PPA has potential to inform earlier and accurate diagnosis and interventions to assist those living with the reality of the illness.

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<<t/s Set names in caps and scaps as per usual style>>

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<A>**Appendix A**

Table A1. Interview guide for in-depth spousal interviews

Broad topics	Prompt questions to spousal participants
Opening statement	'I really want to know what it has been like for you.'
First acknowledgement of change behaviour	'Could you tell me about the first moment of awareness of a likely problem with your partner.'
Progression of symptoms	'Tell me how X's communication has changed or is changing?'
Impact of PPA	'Tell me more about the things you do and share together.'
Supports	'Could I ask you, what helps you?'
Reflections about PPA	'What advice would you offer to a partner of newly diagnosed with this condition.' 'Any parting thoughts?

Table 1. DSM-5 neurocognitive domains for a 'mild' neurocognitive disorder (NCD)

Cognitive domain	Cognitive sub-skills	'Mild' NCD: early symptoms and observations
Complex attention	Sustained attention, divided attention, selective attention, processing speed	Normal tasks take longer than previously. Begins to find errors in routine tasks; finds work need more double-checking than previously. Thinking is easier when not competing with other things (radio, television, other conversations, mobile phone, driving)
Executive function	Planning, decision-making, working	Increased effort to complete multi-staged projects. Has increased difficulty multitasking or difficulty assuming a task interrupted by a visitor or phone call. May complain of increased fatigue from the

	memory, feedback/error utilization, overriding habits/inhibition, mental cognitive flexibility	extra effort required to organize, plan and make decisions. May report large social gatherings are more taxing or less enjoyable because of increased effort required to follow shifting conversations
Learning and memory	Immediate memory, recent memory (including free recall, cued recall and recognition memory). Very long-term memory (semantic, autobiographical, implicit learning).	Has difficulty recalling recent events, and relies increasingly on list-making or calendar. Need occasional reminders or re-reading to keep track of characters in a film or novel. Occasionally may repeat self over a few weeks to the same person. Loses track of whether bills have already been paid
Language	Receptive language. Expressive Language: naming, word finding, fluency, grammar and syntax	Has noticeable word-finding difficulty. May substitute general for specific terms. May avoid use of specific names of acquaintances. Grammatical errors involve subtle omissions or incorrect use of articles, prepositions, auxiliary verbs etc.
Perceptual motor	Visual perceptual, visuo-	May need to rely more on maps or others for direction. May find self lost or turned around when not concentrating on a task. Is less precise in parking. Needs to expend greater effort for spatial tasks such as

	constructional, perceptual-motor praxis and gnosis	carpentry, assembly, sewing or knitting.
Social cognition	Recognition of emotions, theory of mind	Has subtle changes in behaviour or attitude, often described as a change in personality, such as less able to recognize social cues or read facial expressions, decreased empathy, increase extraversion of introversion, decreased inhibition, or subtle or episodic apathy or restlessness

Source: Adapted from American Psychiatric Association (APA) (2013), pp. 593–595.

Table 2. Demographic data of study participants and their partner diagnosed with PPA

Participant number	Spouse's details			Partner's details						Relationship (years)
	Sex	Age (years)	Classification of occupation	Sex	Age (years)	Classification of occupation[a]	PPA variant diagnosis	Period of diagnosis	Years from earliest signs (recognized by spouse) to confirmed diagnosis	
P1	F	79	5	M	79	3	IvPPA	November 2013	4–5	52
P2	F	72	5	M	75	3	IvPPA	June 2013	5	45
P3	F	65	4	M	83	1	svPPA	Octobe	3–4	48

								r 2013		
P4	F	73	2	M	87	2	nfvPPA	October 2013	1	32
P5	M	71	6	F	67	5	nfvPPA	July 2013	1	48
P6	F	74	Home duties	M	75	3	svPPA	April 2013	3	53
P7	F	75	Home duties	M	76	1	svPPA	March 2008	10	52
P8	F	60	1	M	61	3	lvPPA	December 2014	4	40
P9	F	82	2	M	83	1	lvPPA	July 2013	5	57
P10	F	69	4	M	69	2	svPPA	August 2015	2	48
P11	F	81	1	M	81	1	svPPA	September 2015	3	54
P12	F	54	Home duties	M	69	8	svPPA	August 2015	3	22
P13	F	67	2	M	68	3	lvPPA	October 2015	2	46

Notes: P, participants; F, female; M, male.

[a] Australian and New Zealand Standard Classification of Occupations (ANZSCO) (2013): 1, Managers; 2, Professionals; 3, Technicians and trades workers; 4, Community and personal service

workers; 5, Clerical and administrative workers; 6, Sales workers; 7, Machinery operators and drivers; 8, Labourers.

Table 3. Summary of PPA early signs as reported by spousal participants

PPA variant subtype	Order and nature of mild neurocognitive disorder (NCD) symptoms (using DSM-5 cognitive domains descriptive definitions)	Key reason for seeking a diagnosis
svPPA, *N* = 6	Social cognition: subtle change in behaviour and attitudes, less able to recognize social cues and/or read facial expression, decreased empathy, apathy, mood swings, increased introversion, cessation of emotional and sexual intimacy, reduced interpersonal–social communication Language: receptive language (at single-word level), expressive language (noticeable word-finding and naming difficulty) Perception motor: getting lost in familiar places Complex attention: divided attention, speed of processing, noise sensitive (i.e., hyperacusis) *Social cognition // language difficulties*	Deterioration in language skills impact on lifestyle
lvPPA,	Social cognition: subtle change in behaviour and attitudes, increased introversion (especially withdrawal in social settings), apathy,	Escalation of behavioural,

N = 5	reduced empathy, mood swings, increased introversion, cessation of emotional and sexual intimacy, reduced interpersonal–social communication Complex attention: sustained attention Perception difficulties: getting lost in familiar places Executive function: working memory difficulties (especially following conversations in one-to-one and social settings), reduced cognitive flexibility Language: receptive language, expressive language difficulties (e.g., slowness in word-finding and naming difficulty, circumlocutory verbal output, talking less) *Social cognition → language difficulties*	memory and language issues
nfvPPA, *N* = 2	Language: speech fluency difficulties, grammar and syntax *Speech and language difficulties ----- social cognition*	Speech production and fluency issues